EUPSA 2018
European Paediatric Surgeons Association

ABSTRACT BOOK

19th EUPSA Congress
Paris, June 20th – 23rd, 2018
Cité des Sciences et de l’Industrie
ASSISTED REPRODUCTIVE TECHNOLOGY AND ESOPHAGEAL ATRESIA: DANGEROUS LIAISONS?

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Aim of the study: Assisted reproductive technologies (ART) are becoming widespread, accounting for approximately 2% of all births in the western countries. Concerns exist on the potential association of ART with congenital anomalies. Few studies have addressed if a relationship exist between ART and the development of esophageal atresia (EA). Our aim was to analyze the relationship between EA and ART.

Methods: Retrospective case control study of all patients treated for EA between 2011 and 2016. For each patient with EA (case), 2 controls with bronchiolitis (and no congenital anomaly) treated during the same period were analyzed. Variables analyzed: prevalence of ART, gestational age, birth weight, and maternal age. Patients with EA born after ART were also compared to those born after non-assisted conception for disease complexity. Fisher’ and Mann-Whitney test were used as appropriate.

Results: 339 patients were analyzed (113 cases and 226 controls). Tables show main findings. Exposure to ART was significantly higher in EA patients than in controls [12.7% vs 3.0%; odds ratio: 4.0 (95%CI 1.7-9.6); p=0.0015] (table 1). Patients with EA born after ART do not appear to have a more complex anomaly than those born after non-assisted conception (table 2).

Conclusions: Patients with EA were more likely to be conceived following ART as compared to controls without congenital anomalies. Patients with EA born after ART seem comparable to those born after non-assisted conception in terms of anomaly complexity.

<table>
<thead>
<tr>
<th></th>
<th>EA</th>
<th>Controls</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of pts</td>
<td>113</td>
<td>226</td>
<td></td>
</tr>
<tr>
<td>ART</td>
<td>14 (12.7%)</td>
<td>7 (3.0%)</td>
<td>0.0015</td>
</tr>
<tr>
<td>Gestational age (wks)</td>
<td>37 (27-42)</td>
<td>39 (37-42)</td>
<td>0.0001</td>
</tr>
<tr>
<td>Birth weight (g)</td>
<td>2522 (1020-3700)</td>
<td>3210 (1890-4500)</td>
<td>0.0001</td>
</tr>
<tr>
<td>Maternal age (yy)</td>
<td>34 (18-51)</td>
<td>33 (19-49)</td>
<td>0.373</td>
</tr>
</tbody>
</table>

Table 1. EA group versus control

<table>
<thead>
<tr>
<th></th>
<th>ART EA</th>
<th>non-ART EA</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of pts</td>
<td>14</td>
<td>95</td>
<td></td>
</tr>
<tr>
<td>Gestational age (wks)</td>
<td>38 (27-41)</td>
<td>37 (30-42)</td>
<td>0.682</td>
</tr>
<tr>
<td>Birth weight (g)</td>
<td>2510 (1020-3250)</td>
<td>2525 (1050-3700)</td>
<td>0.555</td>
</tr>
<tr>
<td>Mother age (yy)</td>
<td>40 (28-51)</td>
<td>33 (18-44)</td>
<td>0.552</td>
</tr>
<tr>
<td>Long gap</td>
<td>4 (28.6%)</td>
<td>38 (38.3%)</td>
<td>0.565</td>
</tr>
<tr>
<td>CHD</td>
<td>10 (73%)</td>
<td>52 (52.5%)</td>
<td>0.253</td>
</tr>
<tr>
<td>VACTERL</td>
<td>5 (35.7%)</td>
<td>25 (25.2%)</td>
<td>0.516</td>
</tr>
<tr>
<td>Death</td>
<td>1 (7%)</td>
<td>6 (6%)</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 2. ART EA group versus non-ART EA group.
SCI-UG02
HEALTH STATUS AND QUALITY OF LIFE OF SCHOOL-AGED CHILDREN WITH ESOPHAGEAL ATRESIA

Chantal A. ten Kate, Gyan Ramsingh, Yannick van de Wijngaert, Dick Tibboel, René M.H. Wijnen, Claudia M.G. Keyzer-Dekker, Hanneke IJsselstijn
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Aim of the study To compare Health Status (HS) and Quality of Life (QoL) of school-aged esophageal atresia (EA) patients with reference norms and to investigate the longitudinal development of HS and QoL.

Methods During the neuropsychological assessment within the infrastructure of a routinely offered standardized follow-up program EA patients born between 1999-2008 filled in two questionnaires. PedsQL measures the perception of children on their HS (objective), DUX25 measures the perception on their QoL and feelings about their daily functioning (subjective). Comparison with validated Dutch reference norms (t-test; Engelen 2009, van Doorn 2008) and change over time (paired t-test; assessment at 8 and 12 years). Ethical approval was obtained.

Main results In total we included 81 children (61.7% boys, 33.3% born <37 weeks of gestational age, 84.0% Gross type C, 13.6% thoracoscopic repair). For comparison with the reference norms see table 1. EA patients reported significantly lower scores on the PedsQL at 8 years. Scores on the PedsQL improved significantly from 8 to 12 years on all domains except for school functioning (n=23; figure 1). The DUX25 scores did not change over time.

Conclusions Children with EA reported impaired HS at 8 years but not at 12 years, whereas scores on a subjective QoL questionnaire were normal. HS improved significantly from 8 to 12 years..

<table>
<thead>
<tr>
<th></th>
<th>PedsQL</th>
<th>DUX25</th>
</tr>
</thead>
<tbody>
<tr>
<td>8 years</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Norm</td>
<td>82.11 ± 8.87 (219)</td>
<td>80.00 ± 12.00 (991)</td>
</tr>
<tr>
<td>Questionnaire</td>
<td>75.18 ± 11.80 (56)</td>
<td>81.69 ± 9.61 (55)</td>
</tr>
<tr>
<td>Mean difference</td>
<td>6.9 (p&lt;0.01)</td>
<td>-1.7 (NS)</td>
</tr>
<tr>
<td>12 years</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Norm</td>
<td>82.11 ± 8.87 (219)</td>
<td>80.00 ± 12.00 (991)</td>
</tr>
<tr>
<td>Questionnaire</td>
<td>82.73 ± 11.08 (32)</td>
<td>79.25 ± 9.71 (40)</td>
</tr>
<tr>
<td>Mean difference</td>
<td>-0.6 (NS)</td>
<td>0.75 (NS)</td>
</tr>
</tbody>
</table>

Figure 1: Longitudinal development from 8 to 12 years for the PedsQL self-reports (n=23). * = p<0.01
RESULTS OF MODIFIED STEP WITH OR WITHOUT DUODENAL LENGTHENING IN CHILDREN WITH SHORT BOWEL SYNDROME

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AIM OF THE STUDY. The STEP intestinal lengthening procedure has a 47% success rate (International STEP-Registry) to achieve a free-state of parenteral nutrition (PN) in pediatric short bowel syndrome (SBS). We have describe a modified STEP (MSTEP) which can be applied to the duodenum. The aim of this study is to describe the outcome of children with SBS who underwent MSTEP.

METHODS. Between 2005-2017, 14 (7M,7F) children (median age:5 years (range:6 months-18 years) with neonatal SBS underwent MSTEP. Median weight was 13.4 kg (range:3.4-32 kg). Of those, 5 patients simultaneously underwent duodenal lengthening. Causes of SBS were: gastroschisis (n=4), Hirschsprung (n=2), NEC (n=6), and volvulus (n=2). Median intestinal length at the time of neonatal surgery was 32 cm (range:0-75 cm). 1/14 retained the ileocecal valve, and 8/14(61%) preserved <50% of colon. STEP indication were: nutritional autonomy achievement (n=10) (and metabolic disturbances/failure to thrive (n=4). Five patients only preserved one central vein access.

MAIN RESULTS. The mean intestinal length pre-STEP and post-STEP are shown in table. Two patients underwent re-STEP procedures. Complications included 1 leak, 1 intestinal obstruction, 1 stapler line bleeding. With a mean follow-up of 4.6 years (range:9m-13 yrs) there was only one exitus due to central line sepsis 9 months after surgery (child already weaned off PN). 7/10 patients achieved nutritional autonomy (one after teduglutide administration) including 4/5 patients who underwent duodenal lengthening. 4/4 children improved their metabolic/nutritional status.

CONCLUSIONS. The MSTEP success rate is similar or even superior to the standard technique, maybe duodenal lengthening related.

<table>
<thead>
<tr>
<th>STEP Indication</th>
<th>#</th>
<th>Median Intest. length pre-STEP (cm)</th>
<th>Int. length post-STEP (cm)</th>
<th>Duodenal lengthening</th>
<th>Mean stapler firings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nutritional autonomy achievement</td>
<td>10</td>
<td>50 (R:0-120*) *1/10 stoma</td>
<td>134 (R:40-230)</td>
<td>5/10</td>
<td>17 (R:6-28)</td>
</tr>
<tr>
<td>Metabolic disturbances</td>
<td>4</td>
<td>89 (R:50-140)</td>
<td>150(R:105-180)</td>
<td>0/4</td>
<td>14 (R:9-22)</td>
</tr>
</tbody>
</table>
Aim: Chewing dysfunction (CD) may cause restrictions in solid food intake and can be seen in 37% of children with esophageal atresia-tracheoesophageal fistula (EA-TEF). The Functional Chewing Training (FuCT) is a holistic approach to improve chewing function (CF) in children. A retrospective study was performed to evaluate the effect of FuCT on CF in children with EA-TEF.

Methods: Eleven children with CD were included. Patients who received 12 weeks FuCT including impairment-based (positioning, sensory stimulation, chewing exercise) and adaptive (adjustment of food consistency) components were investigated for age, sex and type of atresia. Chewing performance level was scored with Karaduman Chewing Performance Scale (KCPS), and tolerated food texture was determined by International Dysphagia Diet Standardization Initiative (IDDSI). The baseline and final KCPS and IDDSI levels were compared to evaluate the effect of FuCT on CF.

Results: 63.6% of cases were isolated-EA, and 36.4% were EA-distal TEF with a median age of 25 (25-84 months) months, of which 63.6% (n=6) were male. Baseline evaluation showed that 6 cases were in level-I, 4 in level-III and 1 case in level-IV according to KCPS. Five children with CD (45.5%) had IDDSI level-3 and six (54.5%) had level-7. There was a significant improvement in KCPS scores and IDDSI scores after 12 weeks training (p=0.003, p=0.025; respectively). KCPS scores showed level-0 in 9 cases, and level-1 in 2 cases. All children had IDSS1 level-7.

Conclusions: The FuCT is an effective method to improve chewing function in children EA-TEF who had CD.
SCIENTIFIC SESSION I: UPPER GASTROINTESTINAL TRACT

Thursday June 21st

SCI-UG05
DEVELOPMENTAL STATUS OF CHILDREN OPERATED FOR EA & TEF ALONG WITH CAREGIVERS’ STRESS, THEIR QUALITY OF LIFE AND COPING ABILITIES

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All India Institute of Medical Sciences, Delhi, India

Aim of the study: To assess the developmental status of children operated for esophageal atresia with or without trachea-esophageal fistula (EA & TEF) along with caregivers’ stress, their quality of life and coping abilities.

Methods:
Descriptive cross sectional survey with institutional ethical permission on children up to 5 years of age after EA & TEF surgical repair and their caregivers in a single tertiary care centre. The tools used were: Demographic datasheet of child and caregivers, anthropometry assessment, child behaviour checklist (1.5-5 years) caregiver report form, Developmental Assessment Scale (DASII) and modified Vineland Social Maturity Scale (VSMS) for developmental assessment, parental stress scale (PSS), WHO QOL Bref and coping strategies checklist

Main results: Out of 51 children, 47.1% of children had low & very low weight (VLW) for age, and 31.4% had height less than normal, for the reference age. Other anthropometric parameters like mid-upper arm circumference (MAC), skinfold thickness and head circumference were less than the reference age in 68.9%, 51.1% and 41.1% respectively. The developmental delay as measured by DASII and VSMS was observed in 40.63% and 37.5% of children respectively. Among the caregivers, 47% had moderate stress, poorest quality of life in environmental (60.1± 18.9) and psychological (60.8±18.8) domain. Most commonly used coping strategy by the caregivers was emotional outlet (29.41%).

Conclusions: Developmental delay was present in children operated for EA & TEF. Significant number of caregivers had stress affecting their quality of life and were using emotional outlet as their coping strategy.
**SCU-UG07**

**CLINICAL FACTORS AFFECTING CONDITION-SPECIFIC QUALITY-OF-LIFE DOMAINS IN PEDIATRIC PATIENTS WITH ESOPHAGEAL ATRESIA: THE SWEDISH-GERMAN EA-QOL©-STUDY**

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¹Department of Pediatric Surgery, Queen Silvia Children’s Hospital, Gothenburg, Sweden. ²Center of Pediatric Surgery, Hannover Medical School, Hannover, Germany. ³Department of Medical Psychology, University Medical Center Hamburg-Eppendorf, Hamburg, Germany. ⁴Department of Pediatrics, Institute of Clinical Sciences, University of Gothenburg, Gothenburg, Sweden

**Aim of the study** To determine clinical factors which influence condition-specific quality-of-life (EA-QOL©) in esophageal atresia (EA) children.

**Methods** The EA-QOL©-questionnaires were developed/validated using a child-and-parent-derived approach (international standards). Following ethical approval, 124 families of EA-children (Gross A-E) from Sweden and Germany participated (response rate 68%): 53 parents of EA children 2-7 years old completed the 17-item-version; 62 children/71 parents completed the 24-item-version for children aged 8–17 years (Table 1). Items were linearly transformed to a 0–100 scale. 70% item-responses were required for domain-scores-calculations. Clinical data were collected through medical records, and standardized questionnaires. Data were analyzed using Mann-Whitney U-test and linear regression analysis (p< 0.05).

**Main Results** Primary repair was related to better Eating-QOL and Health & well-being in 8-17-year-olds. Prior gastrostomy insertion was associated with worse Eating-QOL in both age-groups; worse Social isolation & stress in 2-7-year-olds; worse Health & well-being and Body perception in 8-17-year-olds (p<0.05). Dysphagia negatively influenced EA-QOL©-scores on all domains (p<0.05), except one (Physical health & treatment in 2-7- year-olds). An increased number of digestive symptoms decreased EA-QOL©-domain-scores (p<0.001) with one exception. Wheezing and dyspnea impaired Physical health dimensions across child age-groups. An increased number of respiratory symptoms decreased scores on two EA-QOL©-domains in 2-7-year-olds, and one domain in 8-17-year-olds (Table1, p<0.05).

**Conclusions** The first EA-QOL©-questionnaires revealed surgical and symptom variables to influence EA-QOL©-areas in children with EA, with prominent influence of digestive symptoms.
Table 1. The impact of digestive symptoms and respiratory symptoms on the last four weeks on scores of the EA-QOL-D domains in children 2-7 years old (parent-report) and children 8-17 years old (child and parent-report) according to linear regression analysis

<table>
<thead>
<tr>
<th>Domain</th>
<th>Digestive symptoms</th>
<th>Respiratory symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>b_0</td>
<td>b_1</td>
</tr>
<tr>
<td><strong>The EA-QOL-D questionnaire for children 2-7</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eating (7 items)</td>
<td>46</td>
<td>90.3</td>
</tr>
<tr>
<td>Physical health &amp; treatment (6 items)</td>
<td>46</td>
<td>77.6</td>
</tr>
<tr>
<td>Social isolation &amp; stress (4 items)</td>
<td>42</td>
<td>91.1</td>
</tr>
<tr>
<td><strong>The EA-QOL-D questionnaire for children 8-17</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eating (8 items)</td>
<td>58</td>
<td>86.6</td>
</tr>
<tr>
<td>Social relationships (7 items)</td>
<td>59</td>
<td>84.2</td>
</tr>
<tr>
<td>Body perception (5 items)</td>
<td>60</td>
<td>92.1</td>
</tr>
<tr>
<td>Health &amp; well-being (4 items)</td>
<td>59</td>
<td>91.4</td>
</tr>
<tr>
<td><strong>The EA-QOL-D questionnaire for children 8-17</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eating (8 items)</td>
<td>65</td>
<td>124.3</td>
</tr>
<tr>
<td>Social relationships (7 items)</td>
<td>63</td>
<td>86.6</td>
</tr>
<tr>
<td>Body perception (5 items)</td>
<td>63</td>
<td>89.9</td>
</tr>
<tr>
<td>Health &amp; well-being (4 items)</td>
<td>63</td>
<td>93.4</td>
</tr>
</tbody>
</table>

*Digestive symptoms (n=4): dysphagia, heartburn, vomiting
*Respiratory symptoms (n=5): cough, wheezing at physical activity/rest, airway infections, dyspnea on physical exertion/rest, chest tightness
**GASTROSTOMY COMPLICATIONS REQUIRING SURGERY – DEFINING THE RISK**

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Great Ormond Street Hospital for Children NHS Foundation Trust, London, United Kingdom

**Aim of the study:**  
Gastrostomy insertion is regarded as safe. The reported incidence of complications ranges from 5-17%. We sought to quantify the incidence of surgical complications in this large series.

**Methods:**  
Retrospective review with institutional approval was performed of gastrostomy insertions in a single centre from January 2011 to December 2015. Demographic and operative data were obtained from medical records. Complications including buried bumper, gastrocolic fistula, persistent gastrectaneous fistula and need for resiting were identified. Data are presented as median [range] and analysed using Fisher’s exact test.

**Main Results:**  
1169 patients had a gastrostomy inserted aged 2.3 years [1 day–19 years]. Follow up was a minimum of 2 years, median 3.5 years. 140 (12%) patients died, only 1 death was related to the gastrostomy. Table shows techniques and complication rates. Techniques included percutaneous endoscopic gastrostomy (PEG), laparoscopic assisted gastrostomy (LAG), radiologically inserted gastrostomy (RIG) or open insertion. In 1 patient the insertion technique was not identified. 153 (13%) patients had a jejunal extension inserted. The incidence of buried bumper was 19/1010 (1.8%) with gastrostomy alone and 14/153 (9.2%) with jejunal extension (p<0.0001, Relative risk 3.45, 95% CI 2.25-5.3). The overall incidence of surgical complications was 91/1169 (7.8%).

**Conclusions:**  
The complication rate of gastrostomy insertion is low. There are no significant differences in complication rates between visualised and non-visualised techniques, although laparoscopic/open techniques prevent gastrocolic fistula. Patients who require a gastro-jejunostomy are at higher risk of buried bumper. These data provide information for informed consent for gastrostomy insertion.
<table>
<thead>
<tr>
<th>Table of Results</th>
<th>PEG (n=456)</th>
<th>RIG (n=356)</th>
<th>LAG (n=301)</th>
<th>Open (n=55)</th>
<th>TOTAL (n=1169)</th>
<th>p value*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gastrocolic fistula</td>
<td>7 (1.5%)</td>
<td>2 (0.6%)</td>
<td>0</td>
<td>0</td>
<td>9 (1%)</td>
<td>p=0.06</td>
</tr>
<tr>
<td>Buried Bumper</td>
<td>17 (3.7%)</td>
<td>7 (2%)</td>
<td>7 (2.3%)</td>
<td>2 (3.6%)</td>
<td>33 (3%)</td>
<td>p=0.8</td>
</tr>
<tr>
<td>Resiting</td>
<td>10 (2.2%)</td>
<td>13 (3.7%)</td>
<td>9 (3%)</td>
<td>3 (5.4%)</td>
<td>35 (3%)</td>
<td>p=0.7</td>
</tr>
<tr>
<td>Gastrocutaneous fistula</td>
<td>4 (1%)</td>
<td>3 (0.8%)</td>
<td>5 (1.6%)</td>
<td>2 (3.6%)</td>
<td>14 (1.1%)</td>
<td>p=0.1</td>
</tr>
<tr>
<td>Total</td>
<td>38 (8.3%)</td>
<td>25 (7%)</td>
<td>21 (7%)</td>
<td>7 (11.1%)</td>
<td></td>
<td>p=1</td>
</tr>
</tbody>
</table>

*p value of comparison between PEG/RIG (blind techniques) and LAG/Open (visualised techniques)
SCI-UG09
CISAPRIDE® USE IN PATIENTS WITH INTESTINAL FAILURE AND ITS IMPACT ON PROGRESSION OF ENTERAL NUTRITION

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AIM: Gastrointestinal dysmotility is common in pediatric intestinal failure (PIF) patients, leading to delays in advancement of enteral nutrition (EN). There is limited literature regarding the safety and efficacy of Cisapride® for improvement of enteral tolerance and ability to wean PN. Our objective was to describe a single center experience on the use of Cisapride® in patients with PIF.

METHODS: Retrospective chart review of patients was performed. Demographic, intestinal anatomy and outcome data was collected. Percentage of EN prior to Cisapride®, progression of EN at 3 months and 6 months and ability to wean PN was calculated.

RESULTS: Use of prokinetics was identified in 60/106 patients (56.6%), 29/60 patients (48.3%) failed to progress EN on other prokinetics and started on Cisapride®. Prior to Cisapride® the progress of EN had plateaued for a mean of 42.3 days (SD 60.2). The rate of feed progression pre-Cisapride® was 0.14%/day (SD 0.19) and rate of progression after Cisapride® initiation improved significantly (0.69%/day [SD 0.31]) (p<0.001). Percentage of EN improved significantly from baseline to 3 months post-initiation (23.9% vs 79.4 % respectively; p<0.001). ECG was performed on initiation of Cisapride® and after every dose change. Medication was discontinued in 2/29 (6.8%).

CONCLUSION: Cisapride® can be beneficial in PIF patients who have failed to progress EN on first line prokinetics. The most significant period of improvement occurred within 3 months of Cisapride® initiation. Cardiac side effects in our cohort were lower than previously reported.
SCI-UG10
LONG-TERM EFFECTIVENESS OF ANTI-REFLUX SURGERY IN ESOPHAGEAL ATRESIA PATIENTS

Anders Sandin, Lars-Göran Friberg, Oskar Enoksson, Vladimir Gatzinsky, Michaela Dellenmark-Blom, Linus Jönsson, Kate Abrahamsson

Abstract


Methods: A retrospective study on 99 patients with EA at a single pediatric surgical center in Scandinavia 1997-2010 were included. 26 patients had anti-reflux surgery (ARS) during the study period due to severe gastroesophageal reflux (GER) disease. The patients were followed with a 24-hour pH study at 1, 7 and 15 yrs of age from where the reflux index (RI) was calculated. Fisher’s Exact test (2-sided), Chi Square test and Fisher’s non-parametric test was used for statistical analysis. P<0.05 was considered significant. Study approval by the ethical committee.

Main results: Antireflux surgery was significantly more common in patients with EA Gross A+B, and when a primary anastomosis wasn’t possible. At age 1 yr, RI was worse in ARS patients prior to surgery compared to non-ARS patients (32% and 10% respectively). After surgery, the groups were similar (14% and 10% respectively). At age 7 yrs, RI in the ARS-group was 16% and 5% in the non ARS-group. At age 15 yrs RI was 26% and 6% respectively.

Conclusions: In short term, ARS is effective to decrease gastrointestinal reflux in EA patients, but in the long-term gastroesophageal reflux seems to return and is worse in the operated group. Therefore, ARS should be used with great caution in EA patients.
Aim:
Mothers of children with esophageal atresia (EA) may experience increased psychological distress. Aim of the study was to report psychological distress in mothers of adolescents with EA, and to seek clinical factors affecting maternal distress.

Methods:
Mothers and patients with EA born 1996-2002 were enrolled. 125 EA patients were identified, and 102 invited. Maternal psychological distress was assessed by General Health Questionnaire 30-item version (GHQ-30), reflecting sub-scores on different parts of psychological distress. Clinically important psychological distress was defined as GHQ-30 case-score ≥ 6, which may raise suspicion of significant stress-related problems. Mothers’ sociodemographic characteristics were recorded, and adolescent clinical data as number of days on ventilator, dysphagia, gastro-esophageal reflux and gastrostomy in history were retrieved from medical records. Correlation is reported by Spearman’s R.

Main results:
65/102 (63.7%) mothers (median age 44) and 68/102 (66.7%) adolescents took part in the follow-up study. GHQ-Likert score was 24 (11-59). GHQ sub-score for depression was significantly correlated to gastrostomy (r=0.278, p=0.026) and number of days on ventilator (r=0.385, p=0.003). GHQ case-score was mean 3.23 (0-23). We have earlier examined mothers of healthy- and renal transplanted children, with case-scores mean 1.2 and 2.1, respectively. In the present study 15 (22%) mothers reported clinically important psychological distress, and gastrostomy was significantly correlated to case-score (r=0.304, p=0.014).

Conclusion:
Clinically important psychological distress was diagnosed in 15 (22%) EA mothers. Surprisingly, mothers of EA adolescents reported significantly more stress problems compared to mothers of healthy controls and renal transplanted patients.
SCIENTIFIC SESSION I: UPPER GASTROINTESTINAL TRACT

Thursday June 21st

SCI-UG12

OESOPHAGEAL DIAMETERS IN CHILDREN CORRELATED TO BODY WEIGHT. A RETROSPECTIVE ANALYSIS

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Aim of the study

After surgery of oesophageal atresia or ingestion of alkali or acid corrosive substances oesophageal strictures are handled by balloon dilatation. There is currently no common agreement on the dilatation expanse. In this retrospective study we wanted to find out the normal size of oesophageal diameters in relation to the body weight of children.

Methods

Our study includes 53 children from 2011 – 2016. All these children had no oesophageal strictures and underwent radiology contrast investigations for other reasons such as reflux disease. Evaluations were blinded to different examiners who measured the maximum contrast filling between the second and third rib as our cranial diameter (Fig.1B) and between 7th and 8th rib as our caudal diameter (Fig.1A&C). It was not possible to include healthy children because of ethical aspects and radiation doses.

Main Results

We analyzed all children (n=29) within the 3rd-97th weight percentile and found 8-mm mean cranial diameter for a 10 kg child (Fig.2). We find linear correlation between weight and oesophageal diameters (Correlation coefficient cranial: 0.70; caudal 0.67). Mean weight of these children (Standard deviation) is 26.5 (18.1) kg and median age is 7.0 years.

Conclusions

Beside some limitations, the weight of children is associated with oesophageal diameters. We propose this as a first recommendation for subsequent oesophageal dilatation expanse in children with esophageal stenosis.
Fig. 1

Mean cranial (upper) diameter

- Linear equation: \( y = 0.088x + 7.2515 \)
- \( R^2 = 0.48678 \)

Mean caudal (lower) diameter

- Linear equation: \( y = 0.1259x + 8.065 \)
- \( R^2 = 0.4515 \)

Fig. 2
SCII-LG01
ARE ANORECTAL MALFORMATIONS ASSOCIATED WITH AN INCREASED RISK OF ADHD?

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Aim of the study
The aim of the study was to investigate if patients with anorectal malformations (ARM) have an increased risk to develop attention deficit hyperactivity disorder (ADHD).

Methods
This was a nationwide population-based cohort study using register data from Swedish national health care registers. The study includes all patients born in Sweden 1973-2009 with ARM diagnosis in the National Patient Register. For each patient, five randomly selected age and sex-matched children without ARM were added. All individuals were evaluated for occurrence of ICD-code suggesting ADHD in the National Patient Register during the period 2001-2014. As a proxy for ADHD, prescription of drugs to treat ADHD was analyzed in the Swedish Prescribed Drug Register during the period July 2005-2014. Logistic regression was used to calculate odds ratios (OR) and 95% confidence interval (95% CI). The study was approved by the Regional Ethics Review Board.

Main results
The cohort comprised 1024 ARM patients and 5073 non-ARM individuals. Thirty (2.9%) of the ARM patients and 79 (1.6%) of the non-ARM individuals had an ICD-code suggesting ADHD (OR 1.91 (95% CI 1.25-2.92)) registered. Forty (3.9%) of the ARM patients and 151 (3.0%) non-ARM subjects were prescribed ADHD drugs (OR 1.32 (95% CI 0.93-1.89)).

Conclusions
We did not find evidence of a more widespread use of ADHD medication among patients with ARM, which we find the most robust way to study ADHD. However, the higher occurrence of ADHD diagnosis among ARM patients warrants further clinical studies.
SCII-LG02
SONOGRAPHIC SACRAL RATIO

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Abstract

The purpose of this study was to evaluate in a cohort of healthy newborns, whether sonography is a reliable tool to examine the sacrococcygeal region and allows to reproducibly determine a sonographic sacral ratio (sSR) comparable to Peña's.

High-resolution sonography of the sacrum and coccyx was performed on 298 healthy newborns (age: 0-7 days; m=f=149), with the parents’ approval. On longitudinal images of the sacrum and coccyx two lines were drawn, measuring the distance between the upper edge of the first sacral vertebra to the lower edge of the fourth sacral vertebra (‘S1-S4’), and measuring the distance between the lower edge of the fourth sacral vertebra to the bottom of the coccyx (‘S4-Bottom’). The sSR was calculated by dividing 'S4-Bottom' by 'S1-S4'. Correlations between sSR and bodyweight or height were tested.

For 'S1-S4' 520 measurements were evaluated, averaging at 29,2mm (SD 1,7mm; range: 21-34mm). For 'S4-Bottom' 518 measurements were suited, averaging at 17,6mm (SD 3,2mm; range: 7,6-27mm). The average sSR of 518 measurements was 0,6 (SD 0,11; range: 0,26-1). There was very weak correlation between sSR and both weight and height, with a Pearson correlation coefficient of -0,081 and -0,059 respectively.

In conclusion, ultrasound is a reproducible and easy-to-learn method to examine and measure the sacrococcygeal region. It allows to calculate what we called “sSR” in newborns, analogous to Peña’s sacral ratio. Its use as a prognostic tool for continence in children with anorectal malformations has to be evaluated in further multicentric prospective studies.
SCII-LG03
TESTICULAR FUNCTION IN ADULT MALE PATIENTS WITH ANORECTAL MALFORMATIONS: A BI-CENTRIC STUDY

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Aim of the study

Patients with anorectal malformation (ARM) are expected to have a normal life span, therefore issues about fertility are important. Little is known about these aspects in adult patients. Aim of the study was to assess the potential for fertility in adult males with history of ARM.

Methods

Twenty-eight patients from two referral centers, aged ≥ 18 male, were enrolled. Doppler ultrasound of testis, semen analysis, testosterone, and administration of IIEF-5 questionnaire for erectile dysfunction were performed. Data were compared with 15 controls.

Main results

Cryptorchidism and recurrent episodes of epididimo-orchitis were reported 28.6% and 39.3% of cases, respectively. Erectile dysfunction was reported by a single patient and ejaculatory anomalies by 35.7%. Testicular hypotrophy (<10 ml) was detected in 53.5% patients. ARM patients showed an average testicular volume lower than controls (11.3 vs 14.3 ml, \( p=0.0033 \)) and in 96.4\% of cases the rete testis was absent or very scarce. Half patients were azoospermic/cryptozoospermic, 6 of them presented a reduced peripheral sensitivity to androgens (ASI >139). Coital debut resulted delayed at 18 years old (vs 15 years of age in the control group). Overall 57.1\% reported their condition didn’t affect the sexual sphere.
Conclusions

A careful study of reproductive tract and testicular function is recommended in ARM patients to detect and treat low fertility/infertility conditions. It's also important to recognize those clinical conditions that could affect the spermatogenesis during childhood and to guarantee an appropriate psychological support.
ANALYSIS OF SACRAL PLEXUS ANATOMY USING MRI BASED NEUROTRACTOGRAPHY, ACCORDING TO SPINAL DYSRAPHISM, SACRAL ANOMALY AND TYPE OF ANORECTAL MALFORMATION

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Aim of the study: Evaluation of functional outcome in anorectal malformations (ARM) is currently based on type of malformation, sacral anomaly (SA), and associated spinal dysraphism (SD). These parameters could be enriched by the knowledge of patient’s sacral plexus anatomy, using diffusion tensor MRI based neurotractography.

Methods: From 2016 to 2017, 26 ARM were prospectively included in this study, recording clinical and radiological data. Each patient underwent a pelvic MRI with a coronal volumic sequence for 3D modeling of pelvic organs and an axial diffusion sequence for diffusion tensor imaging (DTI) and tractography of the sacral plexus. 3D Slicer ® software enriched by house-made plugins was used for segmentation and qualitative evaluation of tractograms.

Main results: Repartition of ARM types (11 female - 15 male, median age at MRI of 1,2 years - min-max: 0,3-13) was similar to Krickenbeck’s classification: perineal fistula n= 8 (30%), recto-urethral fistula n=6 (23%), recto-vesical fistula n=1 (4%), recto-vestibular fistula n=4 (15%), cloaca n=2 (7%), no fistula n=1 (4%), rare forms n= 4 (15%). 7 patients (27%) presented either SA or SD, 4 with both, 1 with isolated SA, 2 with isolated SD. Patients with either SA or SD presented a very much altered sacral plexus anatomy (disappearance of sacral roots) on tractography.

Conclusions: This original preliminary study of sacral plexus anatomy using diffusion tensor MRI based neurotractography opens a new field of research in ARM phenotype characterization. Collection of more data and controls is currently on-going to improve both qualitative and quantitative analysis.
Figure: Examples of ARM patients with and without SA/SD.
Left = 7 months-old boy with recto-bulbar fistula, sacral agenesis and short spinal cord.
Right = 6 months-old girl with recto-vaginal fistula, normal sacrum and spine.
MECHANICAL BOWEL PREPARATION BEFORE PEDIATRIC COLORECTAL SURGERY

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Aim of the Study
The usage of mechanical bowel preparation (MBP) before colorectal surgery (CRS) in children remains the standard of care for many pediatric surgeons. However, the value of MBP in children undergoing CRS is unclear. The aim of this study was to systematically review and analyze the effect of MBP on the incidence of major complications, specifically anastomotic leak, intra-abdominal infection, and wound infection following CRS in infants and children.

Methods
Embase, MEDLINE, Web of Science and CINAHL databases were searched. Papers discussing MBP in patients aged 0-16 years undergoing various types of elective CRS (e.g. colonic resection, pull-through for Hirschsprung Disease, anorectal malformation repair) were included if they reported quantitative data regarding the effects of MBP on postoperative complications. Studies reporting adult patients, emergency procedures, or non-colorectal surgery were excluded. The odds ratios for the postoperative complications were calculated using a random-effects model.

Main Results
Out of 1731 papers retrieved, 6 studies (2132 patients) met the inclusion criteria (2 randomized controlled trials, 3 retrospective studies and 1 case-control study). MBP before CRS did not significantly decrease the occurrence of anastomotic leak, intra-abdominal infection, and wound infection compared to no MBP.

Conclusions
The usage of MBP before CRS in infants and children does not decrease the incidence of postoperative complications. Therefore, MBP before CRS in infants and children can be safely omitted. To overcome confounding factors such as the usage of oral antibiotics and variation of MBP mixture, a large prospective study is needed to validate these results.
### Randomized Controlled Trials

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<th>N of patients</th>
<th>Odds Ratio [95% CI]</th>
<th>P-value</th>
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<td>76</td>
<td>1.08 [0.11, 10.74]</td>
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<tr>
<td>Wound infection</td>
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<td>1.10 [0.22, 5.48]</td>
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### Non-Randomized Studies

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<th>N of patients</th>
<th>Odds Ratio [95% CI]</th>
<th>P-value</th>
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<td>Intra-abdominal infection</td>
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<td>Wound infection</td>
<td>4</td>
<td>2056</td>
<td>1.22 [0.60, 2.47]</td>
<td>0.59</td>
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Aim of the study: to introduce a new simplified MR score for Crohn’s disease (CD) and to compare it with clinical, laboratory and endoscopic indexes, in order to ascertain whether it may be used to evaluate the CD activity in children.

Methods: We retrospectively included children affected by CD, with endoscopic confirmation, and studied by contrast-enhanced MR enterography (MRE). Our Paediatric CD MR Index (PCDMRI) was based on mural/perimural parameters of the most affected intestinal tract and on extramural features. Correlation analysis was performed between the PCDMRI and: Paediatric CD Activity Index (PCDAI), Simple Endoscopic Score for CD (SES-CD), serum C-Reactive Protein (CRP) and fecal Calprotectin (fC). Agreement on disease location and inter-reader reproducibility of PCDMRI were also estimated.

Main results: Forty-two children, aged 13.5±2.6 years, were included in the study, for a total of 80 examinations. Median calculated scores were: 23(8-38) for PCDAI, 22(10-29) for SES-CD, 15 (9-19) for PCDMRI. At bivariate analysis, PCDMRI positively correlated with PCDAI (r=0.635, P<0.001), SES-CD (r=0.598, P<0.001), fC (r=0.402, P=0.005), and CRP (r=0.500, P<0.001). A positive association was found between PCDAI and all segmental parameters defining PCDMRI (P<0.001). At multivariate analysis, only the contrast enhancement pattern and mural signal intensity on T2-weighted images were independent predictors of PCDAI (P≤0.046). The agreement on disease localization between MRE and endoscopy was substantial (κ=0.754; P<0.001). The inter-reader reproducibility was 91%.

Conclusions: PCDMRI significantly correlates with PCDAI, SES-CD, CRP and fC, proving to be a reliable parameter to assess the severity of the disease.
**SCII-LG07**

**PATIENTS WITH HIRSCHSPRUNG DISEASE DO NOT HAVE AN INCREASED RISK OF ATTENTION DEFICIT HYPERACTIVITY DISORDERS**

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**Aim of the Study:** Hirschsprung disease (HSCR) has previously been associated with increased need for remedial teaching at school, despite normal intelligence, and problems with sustained attention. The hypothesis was that HSCR is associated with an increased risk for attention deficit hyperactivity disorders (ADHD).

The aim of this study was to assess the risk of ADHD in individuals with HSCR in a population-based cohort.

**Methods:** This was a nationwide, population-based cohort study. The study exposure was HSCR and the study outcome was ADHD. The cohort included all individuals with HSCR registered in the Swedish National Patient Register between 1964-2013 and ten age- and sex-matched controls per patient, randomly selected from the Swedish Population Register. Prescription of drugs for ADHD, registered in the Swedish Prescribed Drug Registry, was used as a proxy to identify individuals with ADHD.

**Main results:** The cohort comprised 739 individuals with HSCR (565 male) and 7390 controls (5650 male). Twenty-six individuals with HSCR and 202 controls had ADHD (Odds ratio (OR) 1.30, Confidence interval (CI) 95% 0.84-1.93), indicating a similar risk for ADHD in the groups. The mean age at diagnosis of ADHD was not different between the groups; 18.1 years (SD +/- 8.4) versus 16.7 years (SD +/- 7.8), p=0.39. Down syndrome did not affect the risk for ADHD (OR 2.26, CI 95% 0.68-5.53)). Female gender decreased the risk for ADHD (OR 0.58, CI 95% 0.40-0.83).

**Conclusions:** There was no increased risk of ADHD in patients with Hirschsprung disease.
MATERNAL USE OF SELECTIVE SEROTONIN REUPTAKE INHIBITORS DURING PREGNANCY DOES NOT INCREASE THE RISK OF HIRSCHSPRUNG'S DISEASE IN NEWBORNS

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**Aim of the Study:** Hirschsprung’s disease (HSCR) is a multifactorial disease caused by both genetic and environmental factors. Maternal intake of selective serotonin reuptake inhibitors (SSRI) during early pregnancy has been associated with increased risk for HSCR in the offspring in a Danish nation-based study.

The aim of this study was to assess the risk for HSCR in newborns after maternal intake of SSRI in a population-based Swedish cohort.

**Methods:** This was a Swedish nationwide, population-based, case-control cohort study containing all children born in Sweden between 1/1 2006 till 31/12 2012. The cases were identified in the Swedish National Patient Register and the controls (five age- and sex-matched controls per case) were randomly selected among children without HSCR in the cohort. Data on maternal use of SSRI use during pregnancy were collected from the Swedish Prescription Drug Register.

**Main results:** The study included 150 cases of HSCR (120 males) and 665 controls (560 males). Five (3.4%) mothers to newborns with HSCR had used SSRI during pregnancy compared to 16 (2.2%) mothers among the controls (p=0.372). There was no difference in maternal mean age in mothers who used SSRI compared to mothers who had not used SSRI (30.9 (SD +/- 5.1) versus 30.6 (SD +/- 5.0), p=0.81).

**Conclusions:** There was no increased risk of HSCR due to maternal intake of SSRI in this cohort. Limitations include a risk for type 2 errors.
SCIENTIFIC SESSION II: LOWER GASTROINTESTINAL TRACT

SCII-LG09

ARE WE DOING TOO MANY RECTAL BIOPSIES TO RULE OUT HIRSCHSPRUNG'S DISEASE?

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Aim: NICE (National Institute for Health and Care Excellence) outlines five indications for performing rectal biopsies to rule out Hirschsprung's Disease (HSD). The aim of this study was to investigate the compliance of a tertiary referral centre with these guidelines.

Methods: Retrospective analysis of case notes was performed for all patients undergoing open or suction rectal biopsies to rule out HSD from 2009-2014.

Results: 188 patients underwent 214 biopsies (suction n=154, open n=60). 128 patients (68.1%) had NICE indicated biopsies (group A, figure 1), 84.4% of these were ganglionic. 60 patients underwent unindicated biopsies (group B, figure 1): presentations included constipation, distension or vomiting, not meeting NICE criteria. 25 patients were diagnosed with HSD (mean age 13.2 weeks, range 3 days-3.2 years). 80% of HSD patients had NICE indicated biopsies (delayed meconium n=2, neonatal constipation n=2, chronic distension and vomiting n=11, multiple indications n=5). Of the 5 HSD patients with unindicated biopsies, 3 patients presented neonatally with only distension or vomiting. The additional 2 patients presented atypically: one at 14 months with new obstructive symptoms having had neonatal constipation settling with dietary change, and the other at 40 months, with 22 months of constipation and new distension with vomiting.

Conclusion: A significant number of unindicated biopsies were performed, the majority of which were ganglionic. HSD patients in group B had atypical presentations that could be related to inaccurate documentation or history, however deferring these biopsies would lead to missed HSD diagnoses.

Figure 1. Indicated vs Unindicated Biopsies.
**SCII-LG10**

**Nutritional needs in patients with total colonic aganglionosis - a patient reported outcome in the Nordic countries**

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**Aim:** The aim of this study was to evaluate the growth and nutritional support in patients with total colonic aganglionosis (TCA).

**Methods:** A patient-reported multicenter study from five centres in three Nordic countries. Patients born between 1987-2016 answered a questionnaire including questions about growth, medications and nutritional needs.

**Results:** Of a total of 72 patients, four had died. The response rate was 84%. Respondents were a median of 10 (range, 1-29) years of age. The lengths of resected bowel included colon (n=11), small bowel 0-50 cm (n=29), 50-150 cm (n=7) and >150 cm (n=5; one transplanted) and unknown (n=2). Seventy per cent (n=38/54) had undergone a successful intestinal reconstruction operation (mucosectomy with short muscular cuff with J-pouch (n=17) or straight ileo/jejunal-anal anastomosis (n=8), Duhamel (n=8) and Rehbein (n=5)) and 30% (n=16/54) had an enterostomy. Any previous or present growth problem was reported by 35% (n=19) and the use of additional nutrition by 52% (n=28). Nineteen per cent were underweight by BMI. Supportive parenteral nutrition was in use by 30% (n=16) at survey. Another 35% (n=19) had previously used to have parenteral nutrition up to a median of 5 (range, 2-8 years) of age. Supplemental iron and B-vitamins were required by 41% (n=22) and 37% (n=20) respectively at survey, and additional salt and/or oral rehydration was taken by 61% (n=33) and 40% (n=22) respectively.

**Conclusions:** Patients with TCA report a considerable prevalence of reduced BMI and need of nutritional support, which need consideration during clinical follow-up.
SCII-LG11
LONG-TERM OUTCOME AND QUALITY OF LIFE IN PATIENTS WITH TOTAL COLONIC AGANGLIONOSIS IN THE NETHERLANDS
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Aim of the study
Total colonic aganglionosis (TCA) is a severe form of Hirschsprung’s Disease (HD) associated with a high morbidity. This study assesses long-term functional outcome and Quality of Life (QoL) of TCA patients in a national consecutive cohort.

Methods
Medical records of all patients (n=53) diagnosed between 1995 and 2015 were reviewed on surgical and demographic characteristics. Functional outcome of all non-syndromal patients, aged ≥ four years (n=35) was assessed in medical records and with a questionnaire. Generic and disease-specific QoL was assessed using standardized validated questionnaires.

Main Results
Of 35 patients eligible for follow-up, 18 responded to the questionnaires (51%). They were aged 4-19 years. In 67% of these patients a Duhamel procedure was performed, in 33% a Rehbein. In the questionnaire 65% of the patients reported complaints of constipation, 47% of faecal incontinence, 53% of soiling. 18% of patients used bowel management (flushing or laxatives) and 29% an adapted diet only. Children and adolescents with TCA had worse perception of their general health and were more limited by bodily pain and discomfort compared to healthy peers. Their quality of life is influenced most by frequent complaints of diarrhea and other physical symptoms.

Conclusions
Children and adolescents with TCA report lower health-related quality of life compared to normative data, especially in the physical domain. We suggest standardized follow-up and prospective longitudinal future research on functionality and quality of life of these patients.
SCII-LG12
MATERNAL METHAMPHETAMINE ABUSE AND NEC IN TERM NEONATES

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Aim of the Study:

NEC in premature neonates remains a potentially fatal disease. NEC can also occur in term baby and the risk factors are limited to major cardiac anomalies. Methamphetamine (MA) is an addictive recreational drug, which causes euphoria due to reuptake inhibition of dopamine in the brain. Maternal MA abuse is known to cross the placental that can affect on the fetus but there is no literature to its relation to NEC.

Methods:
A retrospective study had done over an 8-Year Period (January 2009 – March 2017) for NEC term neonates (G.A≥37 weeks).
We included a comparable group of 30 term Non-NEC neonates over the same period to assess the maternal factors, in comparison to findings obtained those in NEC neonates.
Ethics approved: IRB0005239

Main results:
The total number of NEC in the study period was 194 patients, 14 of them were term neonates.
4 patients with NEC term neonates had maternal MA abuse in comparison to Non-NEC term neonates (*P value <0.05)
Tabulated drug/medical usage in this cohort:

Conclusions:
Maternal MA can induce NEC in term neonates, 2 hypotheses might explain this:

- MA causes increased dopamine, norepinephrine, and serotonin from presynaptic neurons. This may cause excessive sympathetic activity and vasoconstriction (either in placenta or in fetal gut vessels).
- MA inhibits the immune system by reducing the number and function of macrophages, NK, monocytes and granulocytes, contributing to the increased susceptibility to infections
PW1-LG01
CARDIOGENIC NECROTIZING ENTEROCOLITIS: A SYSTEMATIC REVIEW OF THE LITERATURE AND META-ANALYSIS

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Aim of the study: To investigate if necrotizing enterocolitis (NEC) has a different presentation and outcome in patients with or without congenital heart disease (CHD).

Methods: A systematic review of the literature on the characteristics of CHD infants with NEC was performed by two independent investigators using a defined strategy (PubMed, Cochrane, Embase and Web of Science). A meta-analysis was conducted on studies comparing NEC in CHD and non-CHD infants using RevMan 5.3.

Results: Systematic review: Of 7,108 abstracts screened, 159 full-text articles were analyzed and 65 studies were included. NEC had an incidence of 5.1% in CHD infants (7,651/148,984, range 0-24%) and 0.8% in non-CHD infants (26,430/3,256,891, range 0.1-8.9%). In very low birth weight infants, NEC occurred in 6.3% of CHD patients (6,361/100,454pts) and in 8.9% of non-CHD (23,201/257,794pts). In CHD cases, NEC occurred before cardiac surgery in 43% cases and surgery for NEC was required in 31% infants (1,967/6,273). Meta-analysis: In 21 comparative studies, the incidence of NEC was higher in CHD infants (6%, 768/13,145) than in infants with no CHD (0.9%, 32,625/3,354,323pts; p<0.00001, OR 1.84, 95% CI 1.7 to 1.9). The mortality was higher in infants with CHD and NEC (38%, 243/640) than in those without CHD (27%, 6651/24810; p<0.00001, OR 3.4, 95% CI 2.8 to 4.1, Figure).

Conclusions: The risk of developing NEC is higher for infants with CHD disease than in those without. However, NEC in CHD infants has different characteristics than in infants without cardiac anomalies. Mortality remains very high for both populations.

Figure 1: Mortality in infants with NEC with or without CHD
Introduction: Hirschsprung’s-associated enterocolitis (HAEC) incidence varies widely due to lack of an agreed HAEC definition or cause. In 2009, a panel of experts proposed a clinical score to help classify the episodes and homogenize diagnosis and treatment. Our aim was to characterize HAEC episodes and compare them to the established score.

Methods: Retrospective study of patients with HAEC admitted between 2000-2016. Clinical data, time of pull-through or intestinal transit reconstruction were collected. Episodes of HAEC were identified and Pastor HAEC-Risk-Score was calculated, with ≥10 considered indicative of HAEC. A Pearson correlation coefficient was performed.

Results: 21/93 patients (22.6%) with HD presented 50 episodes of HAEC with a median of 2 (1-5) episodes during an 8.3-yr (2-15.6) follow-up. Most common symptoms were foul-smelling 43/50 and explosive 30/50 diarrhea. Physical findings showed a distended abdomen 38/50 and low-grade fever 30/50. Upon abdominal x-rays, a dilated bowel 41/50 and rectosigmoid cut-off 40/50 were identified most frequently. Only 17/50 showed abnormal laboratory findings. Forty-nine out of 50 were admitted to a surgical ward with IV-antibiotics 47/50, NPO 46/50 and nursing 37/50 for a mean length-of-stay of 7±1days. Pastor HAEC-Risk-Score was 9.4±0.9, only 25/50 met the criteria (≥10). Pearson correlation coefficient for the length-of-stay and IV-antibiotic requirement was r=0.3 and r=0.4 respectively.

Conclusion: There is not a standardized definition or diagnostic criteria for HAEC, the Pastor HAEC-Risk-Score correlates with the accepted management of this condition. However its restrictive criteria(≥10) failed to identify milder HAEC episodes.
LONGITUDINAL BOWEL FUNCTION AFTER TRANSANAL ENDORECTAL PULL-THROUGH FOR HIRSCHSPRUNG DISEASE

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Department of Pediatric Surgery, Oslo University Hospital, Oslo, Norway

Background:

It is assumed that bowel function improves over time in patients operated for Hirschsprung disease (HD). The aim of this study was to compare bowel function at two time points in a cohort of HD patients to assess whether bowel function actually improves with increasing age.

Methods:

Functional outcome, classified according to the Krickenbeck scoring system, was assessed in HD patients operated with transanal endorectal pull-through 1998-2007 at two time-points: 2008-2011 (FU1) and 2017-2018 (FU2). Fecal incontinence was defined as involuntary leaking of stool requiring change of underwear or diapers. The study was approved by the hospital Data Protection Official.

Results:

Of the 64 patients examined at FU1, 34 (53%) participated at FU2. 3/34 patients had Downs syndrome. Median age at FU1 and FU2 was 8.1 years (3.4-16.6) and 16.1 years (10.3-25) respectively. Among these 34 patients, fecal incontinence was reported by 41% at FU1 and 53% at FU2. The number of patients with daily incontinence was constant; 35%. 15% was constipated at first follow-up and 24% at FU2. The need for bowel management and stoma was higher at FU2 than FU1 (see table).

<table>
<thead>
<tr>
<th></th>
<th>Appendicostomy</th>
<th>Stoma</th>
<th>Rectal enemas</th>
</tr>
</thead>
<tbody>
<tr>
<td>FU1</td>
<td>1 pt</td>
<td>3 pts</td>
<td>5 pts</td>
</tr>
<tr>
<td>FU2</td>
<td>7 pts</td>
<td>1 pt</td>
<td>4 pts</td>
</tr>
</tbody>
</table>

Conclusion:

Impaired bowel function is common in HD patients several years after surgery, and no significant improvement was found in this patient cohort. Long-term follow up is recommended, and transitional care should be implemented.
PW1-LG04
MANAGEMENT AND OUTCOME OF PATIENTS WITH TOTAL COLONIC AGANGLIONOSIS – A NORDIC MULTICENTER STUDY

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1Skåne University Hospital Lund Lund University, Lund, Sweden. 2Helsinki University Children’s Hospital, Helsinki, Finland. 3Oslo University Hospital, Oslo, Norway. 4Karolinska University Hospital, Stockholm, Sweden. 5Skåne University Hospital Lund, Lund University, Lund, Sweden. 6University Children’s hospital, Uppsala, Sweden

Aim: To report the outcomes of Nordic patients with total colonic aganglionosis (TCA).

Methods: An international cross-sectional multicenter study. Patients born with TCA 1987-2016 in five Nordic centers were invited to answer a survey on bowel function using the Rintala bowel function score (BFS; 1-20). Data are reported as medians.

Results: Of 72 patients, four (6%) had died. The 68 survivors had been followed up for a for 11 (range, 0-29) years. Intestinal reconstructions had been performed in 76% at a 11 (0.5-156) months of age comprising mucosectomy with a short muscular cuff and J-pouch (JIAA) (20), straight ileo-anal anastomosis (IAA) (12), Duhamel (13) or Rehbein operations (6). One quarter had a jejunostomy (8), ileostomy (7) or had been transplanted (1). The thirty-day complication rate was 36% (15% Clavien-Dindo grade 3-4). Further surgery during follow-up was performed in 18%, including adhesiolysis, additional resections, myotomy and STEP-procedures. 26% received botox injections and 46% were treated for enterocolitis. Eventually, 9 (18%) underwent permanent stoma formation at 26 months (1-66) after reconstruction for diarrhea, stricture or more extended aganglionosis than expected. Patients >4 years old with intact reconstructions (n=36; age 10 years) had a BFS of 15 (range, 5-19; JIAA 16, IAA 13, Duhamel 15 and Rehbein 15; p=0.282). Full bowel control was reported by 52% and 83% had >two bowel movements/day.

Conclusion: In Nordic countries, four different reconstructions for TCA were used during the last 30 years. 18% needed stomas after reconstructions, whilst 52% with intestinal continuity were continent.
Background: The relative efficacy of surgical versus non-operative treatment (NOT) for acute uncomplicated appendicitis in children is uncertain and a definitive trial challenging to conduct. We performed a feasibility randomised controlled (RCT) trial to establish whether a definitive RCT is possible.

Methods: Three-centre feasibility RCT of children (4-15yrs) with acute uncomplicated appendicitis using pragmatic inclusion criteria (no specific clinical, laboratory or radiological criteria). Participants were randomised to NOT pathway (10 days broad-spectrum antibiotics, analgesia, close observation) or laparoscopic appendicectomy. Main end points were feasibility of recruitment, adherence to treatment allocation, adverse events and completeness of follow up data. Ethical approval: 16/SC/0596.

Results: Randomisation was acceptable with 46/100 eligible patients being enrolled (27 male, age 10.5yrs [5.5-15.3]). In appendicectomy arm (n=23) all received surgery, hospital stay was 2.6 days (0.9-6), with 3 readmissions for abdominal pain (2 with intra-abdominal collection). In NOT arm (n=23), 14 were successfully treated and discharged and 9 underwent appendicectomy during initial admission (Table). Hospital stay in NOT arm was 3.4 days (1.4-8.1). There were 2 readmissions (fever and abdominal pain) and 5/14 developed recurrent appendicitis (confirmed at appendicectomy). Overall, all but one participant complied with follow up.

Conclusions: An RCT of appendicectomy versus NOT for acute uncomplicated appendicitis is possible, adverse events are as expected and patients completed follow up. A definitive RCT is needed to understand the clinical effectiveness of these two approaches.

<table>
<thead>
<tr>
<th>Appendixectomy during initial admission in NOT arm (n=9)</th>
</tr>
</thead>
<tbody>
<tr>
<td>withdrew trial consent</td>
</tr>
<tr>
<td>deterioration &lt;24hrs</td>
</tr>
<tr>
<td>no improvement by 48hrs</td>
</tr>
<tr>
<td>parental request (clinically improving)</td>
</tr>
</tbody>
</table>
NECROTIZING ENTEROCOLITIS IN INFANTS WITH CONGENITAL HEART DISEASE: THE IMPACT OF DUCTAL DEPENDENT ANOMALIES

Elisa Siano¹, Vincenzo D Catania¹, Elke Zani-Ruttenstock¹, Alejandro Floh², Agostino Pierro¹, Augusto Zani³

¹Division of General and Thoracic Surgery, The Hospital for Sick Children, Toronto, Canada. ²Department of Critical Care Medicine and Labatt Family Heart Centre, The Hospital for Sick Children, Toronto, Canada

Aim: To analyze the impact of ductal dependent anomalies in infants with congenital heart disease (CHD), who develop necrotizing enterocolitis (NEC).

Methods: Following ethical approval, the charts of infants with CHD and NEC managed at our Institution (2014-2017) were reviewed. Infants with ductal dependent (DD) anomalies were compared with those with non-ductal dependent (nDD) anomalies. CHD severity was measured via the Risk Adjustment for Congenital Heart Surgery (RACHS-1) in infants undergoing cardiac surgery. Data are reported as mean±SD or median (range).

Results: Of the 81 infants treated for CHD and NEC, 56 (69%) had DD and 25 (31%) had nDD anomalies. Demographics of DD and nDD infants are reported in Table 1. Cardiac surgery was performed in 55 (98%) DD and 21 (84%) nDD infants. Only 11 (14%) required surgery for NEC [DD= 7 (13%); nDD= 4 (16%); p=n.s.]: 10 had bowel resection and stoma formation and 1 had primary anastomosis. At surgery, NEC was multifocal in 9% and unifocal in 91% (colon: 60%, small intestine: 30%; ileocecal region: 10%). Postoperative complications occurred in 6 (60%) and included anastomosis leak (n=1) and adhesive obstruction (n=1). NEC recurred in 38% infants (DD=22; nDD=9; p=n.s.). The were no differences in length of stay [DD=60 (11-329); nDD=60 (5-323); p=n.s.] and mortality [DD=18%; nDD=20%; p=n.s.].

Conclusions: Infants with DD anomalies who develop NEC have different demographics, severity of CHD and NEC presentation characteristics to those with nDD anomalies. However, the severity of bowel disease and mortality rate are similar in both groups.

Table 1.

<table>
<thead>
<tr>
<th></th>
<th>DD (n= 56)</th>
<th>nDD (n= 25)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth weight (grams)</td>
<td>2787±105</td>
<td>2107±200</td>
<td>0.001</td>
</tr>
<tr>
<td>Gestational age (weeks)</td>
<td>38 (25-39)</td>
<td>36 (25-38)</td>
<td>0.03</td>
</tr>
<tr>
<td>Age at NEC diagnosis (days)</td>
<td>15 (1-278)</td>
<td>32 (1-289)</td>
<td>0.01</td>
</tr>
<tr>
<td>RACHS-1 – n (%)</td>
<td>DD (n= 55)</td>
<td>nDD (n= 21)</td>
<td></td>
</tr>
<tr>
<td>category 1</td>
<td>4 (7)</td>
<td>4 (19)</td>
<td></td>
</tr>
<tr>
<td>category 2</td>
<td>2 (4)</td>
<td>7 (33)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>category 3</td>
<td>11 (20)</td>
<td>2 (10)</td>
<td></td>
</tr>
<tr>
<td>category 4</td>
<td>11 (20)</td>
<td>8 (38)</td>
<td></td>
</tr>
<tr>
<td>category 5</td>
<td>-</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>category 6</td>
<td>27 (51)</td>
<td>-</td>
<td></td>
</tr>
</tbody>
</table>
THROMBOSPONDIN 4: A NEW MARKER OF INTERSTITIAL CELLS OF CAJAL IN THE HUMAN COLON

Anne-Marie O’Donnell, Prem Puri
National Children’s Research Centre, Our Lady’s Children’s Hospital Crumlin, Dublin, Ireland

Abstract

Aim of the Study: Thrombospondins are a family of extracellular matrix proteins, made up of five members (TSP-1-5). Thrombospondin-4 (TSP-4) has reported roles in the cardiovascular and nervous systems, inflammation and cancer. Lee et al. have investigated the transcriptomic profile of Interstitial cells of Cajal (ICCs) in the murine colon and jejunum, and revealed TSP-4 as a highly specific marker for these cells. We designed this study to investigate the expression of TSP-4 in the normal human colon and in Hirschsprung’s disease (HSCR).

Methods: HSCR tissue specimens (n=5) were collected at the time of pull-through surgery, while colonic control samples were obtained at the time of colostomy closure in patients with imperforate anus (n=5). Western blot analysis was undertaken to quantify TSP-4 protein expression, and immunolabelling of TSP-4 was visualized using confocal microscopy to assess protein distribution.

Main results: Western blot analysis revealed high levels of TSP-4 protein expression in normal controls and ganglionic HSCR, while there was a marked decrease in TSP-4 protein expression in aganglionic HSCR. Confocal microscopy revealed colocalization of TSP-4 to ICCs within normal colon specimens and ganglionic HSCR, with a reduction in TSP-4-labelled ICCs in aganglionic HSCR colon.

Conclusions: To our knowledge, this is the first report describing TSP-4 as a new marker of ICCs in the human colon. These findings suggest a role for TSP-4 in pacemaker activity of the gut, with many potential applications in further HSCR research.
CORRELATION OF THE SACRAL RATIO WITH THE SEVERITY OF THE ANORECTAL MALFORMATION AND THE PRESENCE OF TETHERED CORD

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1Columbus Childrens Hospital, Columbus, USA. 2Medical University Of Vienna, Vienna, Austria

Introduction

The sacral ratio (SR) has been used as a tool to predict fecal continence in patients with anorectal malformations (ARM). We sought to determine whether the sacral ratio additionally would correlate with the complexity of the ARM, and with the presence of tethered cord (TC).

Methods

All patients seen in our Center with an ARM were divided by diagnosis following the Krickenbeck classification. We measured their SR using their AP and lateral films, assessing for ARM diagnosis and tethered cord on MRI/US of the spine. Analysis of variance (ANOVA) and t-tests, were used to define differences in SR, type of ARM, and the presence of tethered cord.

Results

283 patients with 10 ARM diagnoses were reviewed. Higher sacral ratios (a more normal sacrum) were observed in those patients having less severe malformations in a linear distribution. The lower SR's (hypodeveloped sacrum) were found in the most malformations (table 1). The SR was lower in those patients with tethered cord compared with those with no TC (0.54 [0.54-0.63] vs 0.72 [0.70-0.75] (p > 0.001).

Conclusions

The more normal the sacrum (the higher the SR) the less complex the malformation and vice versa. Patients with tethered cord have a more hypodeveloped sacrum (lower SR). Therefore the SR (a measure of sacral hypodevelopment and caudal regression) correlates with the complexity of the malformation and the presence of tethered cord.
T-CELL RESPONSES IN THE APPENDIX DIFFER BETWEEN CHILDREN WITH SIMPLE AND COMPLEX APPENDICITIS

SML The¹, RRCE Schreurs²,³, R Bakx¹, AA Drewniak³, JH Van der Lee⁴, HA Heij¹, LWE Van Heurn¹, RR Gorter¹, MJ Bunders²,³,⁵

¹Paediatric Surgical Centre Amsterdam. VU University Medical Centre & Emma Children’s Hospital AMC, Amsterdam, Netherlands. ²Department of Pediatrics, Emma Children’s Hospital, Academic Medical Center, University of Amsterdam, Amsterdam, Netherlands. ³Department of Experimental Immunology, AMC, UvA, Amsterdam, Netherlands. ⁴Pediatric Clinical Research Office, Academic Medical Centre, Amsterdam, Netherlands. ⁵Heinrich-Pette-Institute, Hamburg, Germany

Aim: There is increasing evidence that simple and complex appendicitis are different entities, which may require different treatment strategies. Therefore, we assessed if there are differences in the immunological response and determined whether numbers and function of tissue-resident T-cells in the appendix in children with simple or complex appendicitis differ.

Methods: Children (0-17 years old) undergoing an appendectomy for appendicitis were included. Classification of entities was based upon intraoperative and histopathological criteria. The mononuclear cell fractions from the epithelial layer (IEL), lamina propria (LPL) and muscular layer were isolated from the appendix. T-cells were analysed using seventeen-parameter flow cytometry. Cytokine production was assessed after stimulation.

Main results: Twenty children were included (simple n=8, complex n=12). Complex appendicitis was associated with an absolute decrease of mononuclear cells in all layers (figure 1). Moreover, CD103+CD69+CD4+ effector (CCR7−CD45RA−) T-cells were decreased in patients with complex appendicitis (IEL median simple 48% versus complex 16%, p=0.002; LPL 59% versus 23%, p=0.014; Muscle 41% versus 6%, p=0.028) suggesting a decrease of tissue-resident memory T-cells or an influx of peripheral T-cells in complex appendicitis. Furthermore, mucosal CD4+ T-cells of patients with complex appendicitis produced relatively more interleukin (IL)-17 (n=10) (figure 2).

Conclusion: The immunological response in simple and complex appendicitis differs. Complex appendicitis is characterized by a loss of mononuclear cells, specifically of tissue-resident CD4+ T-cells. In addition, mucosal CD4+ T-cells produced relatively more IL-17, which can attract neutrophils. Further studies are required to understand underlying mechanisms.
Figure 1. Mononuclear cell counts per cm$^2$ in simple and complex appendicitis

Figure 2. IL-17 production of mucosal CD4 T cells in simple and complex appendicitis
**PW1-LG11**

**URINARY TRACT DYSFUNCTION AND ANOMALIES IN CHILDREN WITH HIRSCHSPRUNG DISEASE**

Christina Granéli1, Hedda Marschall Sima2, Einar Arnbjörnsson1, Pernilla Stenström1

1Department of Pediatric Surgery, Skane University Hospital Lund, Lund University, Lund, Sweden.
2Department of Pediatric Surgery, Skane University Hospital Lund, Lund University, L, Sweden

**Aim**

To evaluate the prevalence of concomitant urinary tract anomalies and urinary tract dysfunction in children with Hirschsprung disease (HD).

**Method**

This was an observational case-control study. Children with HD operated on with transanal endorectal pull-through technique 2005-2017 were invited to participate in the study. Ultrasound examinations of the urinary tract were performed. All children older than four years old were asked to answer a questionnaire regarding urinary tract function. Age-matched healthy children were used as controls. Ethical approval was obtained.

**Results**

71 children were identified of whom one had deceased and one had migrated. Ultrasound examinations were performed in 54 children (78%). Ten anomalies were found in 6 children (11%). Hydronephrosis was the most common anomaly (4%, n=2). One had a urological intervention (Wilm’s tumour). All 37 children older than four years old (27 boys and 10 girls), median age eight years (range 4-12), answered the questionnaire and so did 284 healthy controls (144 boys and 140 girls). Boys with HD reported a higher frequency of enuresis 65% (n=13) versus 9% (n=14) (p<0.05), urinary tract infections 18% (n=4) versus 3% (n=4) (p<0.05) and pyelonephritis 10% (n=2) versus 0 (p<0.05). Girls with HD reported enuresis more frequently 60% (n=16) than healthy girls 7% (n=10) (p<0.05).

**Conclusion**

Urinary tract symptoms and infections were more common among boys with HD than healthy boys. Anomalies of the urinary tract were more common than in the normal population. The results suggests that the urinary tract deserves attention in the follow-up of children with HD.
Aim of the study

Total colonic Hirschsprung Disease (TCHD) presents a postoperative challenge due to hypermotility and perineal rash, and is thought to be worse in younger children, leading some to delay the pull-through until the age of potty training. Because treatments have improved in recent years, we hypothesized that surgery could be done earlier in life (before age 2) without affecting postoperative outcomes.

Methods

TCHD patients (2014-2017) were treated preoperatively for hypermotility with thickening of ileostomy stools, PPIs, constipating diet, water soluble fiber, loperamide, and application of a cyanoacrylate-based barrier for prevention/treatment of the perineal rash. Pull-through timing, number of bowel movements, perineal skin status, and quality of life were assessed (pedsQL).

Main Results

19/26 patients (mean age 17.5 months) and were not toilet trained, and 7/26 were toilet trained at the time of their pull-through. There was no difference in the number of bowel movements [(5 vs 4(p=0.5)], in quality of life, 81 (SD 14.53) vs 78.61 (SD 12.59)(p=0.5), or incidence of severe perineal rash [12/18 vs 4/7(p=0.99)] in these two groups. In all patients the rash resolved within the first 3 months.

Conclusion

TCHD patients who undergo definitive pull through before the age of toilet training have similar postoperative outcomes related to hypermotility, stool frequency, quality of life, and perineal rash compared with those who were operated on later in life. With the implementation of bowel management care and a perineal skin protocol, there appears to be no reason to delay the pull-through.
PW2-UG03  
EFFECTS OF ESOPHAGEAL ATRESIA ON INTESTINAL MUCOSA

Francesco Macchini¹, Anna Morandi¹, Stefano Mazzoleni², Giorgio Fava¹, Ernesto Leva¹  
¹Fondazione IRCCS Ca’ Granda, Ospedale Maggiore Policlinico, Milan, Italy. ²Fondazione IRCCS Ca’ Granda - Ospedale Maggiore Policlinico, Milan, Italy

Abstract
Introduction: Esophageal atresia (EA) prevents the fetus to swallow amniotic fluid (AF). AF promote the maturity of the intestinal epithelial absorptive system. The alteration of the amniotic circle may determine mucosal alterations and interfere with growth. Aim of our study is to evaluate alterations of intestinal mucosa in children with EA.

Materials and Methods: Since November 2016 we performed esophagogastroduodenoscopy with multiple biopsies to all patients operated on for EA in our Center. Biopsies were taken at three esophageal levels and distal duodenum. Mucosal alterations were evaluated, focusing on Peptic and Eosinophilic Esophagitis (EoE) and Celiac Disease (CD). We included children on free diet and without therapy. Clinical, anthropometric and surgical data were collected.

Results: Thirty-one patients were studied (mean age 5±3 years). 69% of patients had polyhydramnion. Gestational age was 36±2 weeks, birth weight 2.387±595 g, and 41% neonates were SGA. 78% were type C, 15% type A and 7% type D EA. At the time of endoscopy 50% had a weight < 3° while 43% a height <3°. Twenty six children (26/31, 84%) presented a peptic esophagitis. Five children (16%) had EoE and 4 (13%) CD.

Conclusions: Our data confirm the high prevalence of peptic and EoE in EA patients. The prevalence of CD in EA resulted to be superior to the general population (13% vs 1%). These findings could be related to the alteration of the AF circle during intrauterine life. Further studies are needed to deepen the subject.
STATE OF PLAY: 8 DECADES OF SURGERY FOR ESOPHAGEAL ATRESIA

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1Center of Pediatric Surgery, Hannover Medical School, Hannover, Germany. 2Surgery Unit, UCL Institute of Child Health and Great Ormond Street Hospital, London, United Kingdom

Aim of the Study: Surgical expertise and advances in technical equipment and perioperative management has let to enormous progress in survival and morbidity of patients with esophageal atresia (EA) in the last decades. We aimed to analyse the available literature on surgical outcome of EA for the last 80 years.

Methods: A Pubmed literature search was conducted for the years 1944-2017 using the key words “esophageal/oesophageal atresia”, “outcome”, “experience”, “management” and “follow-up/follow up”. Reports on long-gap EA only, non-English articles, case reports and reviews without original patient data were excluded. We focused on mortality and rates of recurrent fistula, leakage and stricture.

Main results: Literature search identified 747 articles, 123 manuscripts met the inclusion criteria. The first open end-to-end anastomosis and fistula ligation was reported in 1941. Thoracoscopic fistula ligation and primary anastomosis was performed first in 2000. Reported mortality rate decreased from 100% before 1941, to 54% in the 1950-1959, 28% in 1970-1970, 16% in the 1990-1999 to nowadays 9%. Rates of recurrent fistula varied over time between 4% and 9% with a current rate of 4.4%. Leakage rate remained stable between 11% and 16%. However, stricture rate increased from 25% to 38% (Figure).

Conclusions: Including a full range of articles reflecting the heterogeneity of EA, mortality rate significantly decreased during the course of 80 years. Along with the decrease in mortality, there is a shift to importance of major postoperative complications and long-term morbidity regardless of surgical technique.
Aim of the Study
Patients with esophageal atresia (EA) often experience physical limitations. With increasing life expectancy over the past decades, focus in care shifted to improving the long-term quality of life. We performed validated testing of fitness and gross motor skills in children born with EA.

Methods
Patients with EA were evaluated using the standardized Kinderturntest Plus after caregiver’s written consent. Test scores range from 1 to 5 (quintile of age and gender matched standard population). Caregivers completed an online-questionnaire on patient history. The local ethics board was consulted, who granted an exemption.

Main results
Thirteen patients (mean age 7.2[4-12] years) were included. Comorbidities were prematurity (54%), birth weight <1500g (23%), congenital heart disease (46%), developmental delay (38%), skeletal deformity (28%). The mean test score was significantly lower in children with EA (2,05) compared to the general population (3), independent of prematurity, birthweight, or diagnosis of congenital heart disease (p<0,01). Patient scores were below average in 54%-63% (Figure).

Caregivers identified notable deficits of strength and endurance. All but one patient complained about discomfort during physical exercise, most commonly dyspnea (46%) and reflux symptoms (31%). Notably, 93% of subjects participated in regular school physical education classes, and 86% participated regularly in additional organized sporting activities.

Conclusions
Children after EA repair have decreased physical fitness and impaired locomotor function compared to the general population on a standardized test. Physical discomfort is frequent during exertion. To avoid demotivation, locomotor skill should be promoted at each individual’s comfortable level of exertion.
Figure: Locomotor test scores of children with EA compared to healthy controls.
PW2-UG06
PREVALENCE, RISK FACTORS AND MANAGEMENT OF ANASTOMOTIC STRUCTION FORMATION AFTER ESOPHAGEAL ATRESIA REPAIR: A MULTICENTER STUDY

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¹Department of Pediatric Surgery and Intensive Care, Erasmus MC University Medical Center - Sophia Children’s Hospital, Rotterdam, Netherlands. ²Department of Gastroenterology and Hepatology, Erasmus MC University Medical Center, Rotterdam, Netherlands. ³Department of Biostatistics, Erasmus MC University Medical Center, Rotterdam, Netherlands. ⁴Department of Pediatric Surgery, Pediatric Surgical Center of Amsterdam (Academic Medical Center and VU Medical Center), Amsterdam, Netherlands. ⁵Department of Pediatric Surgery, University Medical Center Groningen – Beatrix Children’s Hospital, Groningen, Netherlands. ⁶Department of Pediatric Surgery, Maastricht University Medical Center, Maastricht, Netherlands. ⁷Department of Pediatric Surgery, Radboud University Medical Center – Amalia Children’s Hospital, Nijmegen, Netherlands

Aim of the Study To determine the prevalence of anastomotic strictures (AS) after esophageal atresia (EA) repair, to identify associated risk factors and to evaluate its management.

Methods Retrospective study in EA patients born between 1999-2013 in five centers in the Netherlands. According to ESPGHAN guidelines refractory strictures were defined as AS requiring ≥5 dilations at maximally four-week intervals, clinically relevant strictures as AS requiring ≥3 dilations. Risk factors were identified with multivariable logistic regression analysis. Ethical approval was obtained.

Main results We included 454 children (7% Gross type A) with end-to-end anastomosis performed in 436 (96%). Anastomotic leakage occurred in 13%. Fifty-eight percent of children with an end-to-end anastomosis developed an AS. Refractory strictures were found in 32/436 (7%) children and required a median of 10 (5-34) dilations (28.6% balloon dilatation, 68.1% bougienage, 3.3% unknown). Clinically relevant strictures were found in 61/436 (14%). Gross type A, anastomotic leakage and need for dilation within 28 days after repair were risk factors for development of refractory strictures (table 1). Adjuvant medical management included esophageal stents in 7 (1.5%) children and topical mitomycin in 2 (0.5%) patients. Intralesional steroid injections were not performed. All children still required additional dilations (2-30).

Conclusions The incidence of refractory strictures in end-to-end anastomosis after EA repair was 7%. Risk factors were Gross type A, anastomotic leakage and the need for dilation within 28 days after repair. Further research to additional treatment options could be useful in prevention of refractory stricture formation.

Table 1: Multivariable logistic regression

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>OR (95% CI)</th>
<th>p-value</th>
<th>OR (95% CI)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gestational age (weeks)</td>
<td>0.98 (0.85-1.12)</td>
<td>0.711</td>
<td>0.96 (0.86-1.06)</td>
<td>0.403</td>
</tr>
<tr>
<td>Gross type A</td>
<td>5.71 (1.48-22.13)</td>
<td>0.012</td>
<td>5.79 (1.80-18.57)</td>
<td>0.003</td>
</tr>
<tr>
<td>Thoracoscopic repair</td>
<td>0.45 (0.14-1.50)</td>
<td>0.191</td>
<td>0.72 (0.30-1.72)</td>
<td>0.460</td>
</tr>
<tr>
<td>Anastomotic leakage</td>
<td>5.03 (1.88-13.43)</td>
<td>0.001</td>
<td>2.93 (1.33-6.43)</td>
<td>0.007</td>
</tr>
<tr>
<td>Dilation ≤28 days after repair</td>
<td>15.90 (5.89-42.92)</td>
<td>&lt;0.001</td>
<td>10.59 (4.50-24.90)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>
EOSINOPHILIC ESOPHAGITIS AND ESOPHAGEAL ATRESIA. IS THERE AN ASSOCIATION?

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AIM OF THE STUDY

Eosinophilic esophagitis (EoE) is a recently diagnosed entity and seems to be more prevalent in patients with esophageal atresia (EA). It can mimic dysphagia and gastroesophageal reflux (GER) symptoms frequently observed in EA, but treatment is different.

METHODS

Retrospective review (2002-2017) of patients with EA who underwent an esophagogastroduodenoscopy (EGD) and esophageal biopsy. EGD was performed in symptomatic patients and in all patients at 12-15 years. Diagnostic criteria for EoE included ≥15 eosinophils for high-power field (hpf) in the esophageal mucosa.

MAIN RESULTS

From a total of 100 patients with EA, 27 lost follow-up. In the remaining 73, 48 patients (65.7%) underwent EGD because of dysphagia (16) or GER studies (32). Esophageal biopsies were performed in 27 patients and diagnostic criteria for EoE was achieved in 6 (22.2%). Mean age at diagnosis was 10 ± 2.1 years and indication for EGD was GER symptoms (4) or dysphagia (2). In 2 cases an anti-reflux surgery was previously performed but symptoms slightly persisted, while in 4 cases contrast studies or phmetry showed no or minimal GER. EGD demonstrated light distal esophagitis in 2 cases, white exudates in 2, and was completely normal in the last 2.

CONCLUSIONS

Patients with EA seems to have a higher risk to develop EoE at early puberty. Follow up should not only be focused on studying GER but also on actively search for EoE with an esophageal biopsy, even when macroscopic appearance is normal.
PW2-UG08
GENETIC ANALYSIS IN PATIENTS WITH TRACHEOESOPHAGEAL ANOMALIES AND INFANTILE HYPERTROPHIC PYLORIC STENOSIS
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1Department of Pediatric Surgery and Intensive Care, Erasmus MC University Medical Center - Sophia Children’s Hospital, Rotterdam, Netherlands. 2Department of Clinical Genetics, Erasmus MC University Medical Center, Rotterdam, Netherlands. 3Department of Biomics, Erasmus MC University Medical Center, Rotterdam, Netherlands.

Aim of the study Patients born with esophageal atresia (EA) appear to have a 30 times higher prevalence of infantile hypertrophic pyloric stenosis (IHPS). This and the fact that EA and IHPS feature together in genetic syndromes, suggests the involvement of a genetic component for both. We explored whether genetic defects disturbing developmental pathways important for foregut morphogenesis are responsible for this specific combination of congenital anomalies.

Methods EA/IHPS patients born between 1988-2012 and where possible their parents were included after written informed consent. We performed whole exome sequencing and copy number profiling. We searched for mutations in mutation intolerant genes. Segregation analysis of possible candidate variants was performed.

Main results 15 out of 26 potential cases were included, in which 150 ultra-rare putative deleterious variants in relevant genes were found. However, all proved to be inherited from one of the unaffected parents. Burden analysis revealed enrichment in ten genes intolerant to genetic variation: 9 in heterozygous models (ALMS1, SLC28A3, SP2, EPB41, AMBRA1, VWA8, CLGN, SDK2, PDLIM7) and one in a homozygous model (GUCY2F).

Conclusions Neither deleterious de novo variations nor recessive variants were identified. As no single gene seems to explain this rare combination of phenotypes, we suspect a complex inheritance pattern. To evaluate this hypothesis, we will compare the individual patient genetic burden for overlap with specific biological pathways enriched in mouse transcriptome profiles of the developing foregut.
PW2-UG09
MUCOSAL PROLIFERATION IN PEDIATRIC SHORT BOWEL SYNDROME

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Aim of the Study: We investigated mucosal proliferation in duodenum biopsies of pediatric short bowel syndrome (SBS) patients in relation to healthy controls.

Methods: Duodenum biopsies were obtained from 69 SBS patients at the median age of 5.2 years (IQR 2.5-11.75), of whom 20 (29%) were on parenteral nutrition (PN) and 49 had weaned off PN (71%), and 27 age-matched controls without intestinal disease. Proliferative enterocytes were identified using MIB-1 immunohistochemistry. Extension of MIB-1 staining towards villus tips was graded (1-3) and percentage of positively stained enterocytes (MIB proliferative index) was counted.

Main results: Remaining bowel anatomy and post-resection time were comparable (P>0.05) between patients on PN and weaned off patients, except for slightly shorter remaining small intestine length in patients on PN [21% (8-25) vs 29 (21-43), P=0.001]. Patients and controls had comparable MIB grades [1.33 (1.00-1.83) vs 1.33 (1.00-1.67)] and MIB indexes for villi [3.22 (1.48-7.23) vs 3.00 (0.86-6.23)] and crypts [46.20 (34.42-55.69) vs 47.08 (37.50-58.13)] (P>0.05 for all). A positive correlation between MIB index of villi and crypts (r= 0.465-0.606, P<0.05) was seen only in patients weaned off PN and controls. The remaining colon length correlated with duodenum MIB grade (r=0.302, P=0.035) only in patients weaned off PN.

Conclusions: After weaning off PN an increase in duodenal proliferation associated to remaining colon length. In contrast, in patients on PN no association between villi or crypt enterocyte proliferation was seen suggesting possible PN related decrease in mucosal proliferation.
PW2-UG10: SURGICAL REPAIR OF LONG-GAP ESOPHAGEAL ATRESIA - A RETROSPECTIVE STUDY COMPARING THE MANAGEMENT OF LONG-GAP ESOPHAGEAL ATRESIA IN THE NORDIC COUNTRIES

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Introduction
Several surgical procedures have been described in the reconstruction of long-gap esophageal atresia (LGEA). We reviewed the surgical methods used in children with LGEA in the Nordic countries (Norway, Sweden, Finland, Iceland and Denmark) over a 15-year period and the postoperative complications within the first postoperative year.

Materials and Methods
Retrospective multicenter medical record review of all children born with Gross type A or B esophageal atresia between 01/01/2000 and 12/31/2014.

Results
We included 71 children; 56 had Gross type A and 15 had Gross type B LGEA. Delayed primary anastomosis (DPA) was performed in 52.1% of the children and an esophageal replacement procedure in 47.9%. Gastric pull-up (GPU) was the most frequent procedure (25.4%). Patient characteristics did not differ significantly apart from the frequency of chromosomal abnormalities, congenital heart defects and other anomalies, which was significantly higher in patients who had a replacement procedure. The frequency of leakage and stricture didn’t differ significantly between the two groups, whereas the frequency of GER was significantly higher after DPA compared to esophageal replacement (67.6% and 38.2% respectively, p = 0.013). At 1-year follow-up the mean body weight was higher after DPA (9046 g) than after organ interposition (8278 g) (p = 0.043).

Conclusion
DPA and esophageal replacement procedures were equally applied. Postoperative complications and
follow-up were similar except for the development of GER and the body weight at 1-year follow-up. Long-term results should be investigated.
PW2-UG11
DUODENAL ATRESIA: DO NOT OVERLOOK ASSOCIATED INTESTINAL ATRESIAS!

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AIM OF THE STUDY: Duodenal atresia (DA) is a congenital malformation that requires surgery to re-establish bowel continuity. The classical surgical repair mandates to rule out the presence of possible associated intestinal atresias. Laparoscopic repair of duodenal atresia is safe and effective, but does not easily allow to rule out associated intestinal atresias. Herein, we aimed to investigate the incidence of DA associated intestinal atresia.

METHODS: After obtaining ethical approval, we reviewed the medical charts of all consecutive neonates who underwent DA repair at our Institution between 2001 and 2016. All infants with associated esophageal atresia were excluded. Associated intestinal atresias were classified according to Bland-Sutton types.

MAIN RESULTS: All 140 neonates with DA underwent open repair and were intraoperatively investigated for associated intestinal atresia. Of these, 10 (7%) babies (birth weight: 2560±752; gestational age: 35±2) had an associated intestinal atresia (Table). In 4 cases, the associated atresia was within the duodenum. None of the 10 infants had trisomy 21 or other genetic anomalies. One patient underwent re-laparotomy for a missed duodenal web proximal to the first duodenal atresia.

CONCLUSIONS: The association between duodenal atresia and other intestinal atresias is not as rare as reported in the literature. Surgeons should be aware of the risk of missing another intestinal atresia when performing open or laparoscopic DA repair.
TREATMENT OUTCOME OF CONGENITAL MICROGASTRIA

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Radboudumc, Nijmegen, Netherlands

Aim of the study
Congenital microgastria is an extremely rare birth defect. Treatment options can be divided into two groups: conservative/less invasive treatment (C/LT, i.e. diet/gastrostomy/jejunostomy) and more extensive gastric surgery (EGS, i.e. Hunt-Lawrence pouch/Total Esophageal Gastric Dissociation). Aim of this study was to review existing literature on the outcome of the treatment options.

Methods
Using Prisma Guidelines a Pubmed-Medline search was conducted. Clinical outcome (way of feeding and growth) and mortality of the different treatment modalities were pooled and compared.

Main results
Of the 71 manuscripts found, 38 describing 51 cases were included, published in 1973-2017. In 46 patients, type of treatment and outcome were described of whom 19 were treated by C/LT (Table 1). Mortality was 47% in the C/LT group versus 15% in the EGS group. In the C/LT group mortality was related to co-morbidity, not to the microgastria. Median follow-up was 42 months (range 1-240). Enteral or oral feeding was poorly described but in at least 7 of the 33 surviving patients oral feeding was described as normal, of whom 6 were in the EGS group. Growth was described as present but final body length and weight remained behind compared to their peers, without difference between the two treatment groups.

Conclusion
Although evidence is scarce, the two treatment groups in the treatment of microgastria show only minimal difference in terms of feeding and growth. Mortality was higher in the C/LT group, possibly because of worse comorbidity.

<table>
<thead>
<tr>
<th>Table 1: Summary of treatment options, mortality rate and outcome</th>
</tr>
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<tbody>
<tr>
<td><strong>Conservative or less invasive treatment (C/LT)</strong></td>
</tr>
<tr>
<td>Non-operative</td>
</tr>
<tr>
<td>N of Patients</td>
</tr>
<tr>
<td>N of Mortality</td>
</tr>
<tr>
<td>Percentage</td>
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<tr>
<td>Total mortality per treatment group</td>
</tr>
<tr>
<td>Feeding</td>
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PW3-G01
FETAL CARDIAC ADAPTATION IN DIAPHRAGMATIC EXPERIMENTAL HERNIA: THE EFFECT OF THE SIMVASTATIN ANTENATAL TREATMENT

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3IRCCS Centro Neurolesi “Bonino-Pulejo, Messina, Italy.
4Department of Radiology, Studio Diagnostico Eco, Vimercate, Milano, Italy.
5School of Medicine and Surgery, University of Milano-Bicocca, Milano, Italy.
6School of Medicine and Surgery, University of Milano-Bicocca, Milano, Italy.
7Hopitaux Pédiatriques de Nice CHU-Lenval, Nice, France.
8Pediatric Unit, University of Pavia and Fond. IRCCS Policlinico S. Matteo, Pavia, Italy.

Aim of the study Statins and sildenafil have been shown to exert beneficial effects in cardiac injury. We hypothesized that antenatal maternal administration of simvastatin and/or sildenafil might also promote benefit in cardiac remodeling of congenital diaphragmatic hernia (CDH). Therefore, we performed micro-CT image analysis and histology of the heart after antenatal treatment in the experimental nitrofen-induced CDH rat model. Fetal growth and organ development were also evaluated.

Methods At 9.5 days postconception (dpc), pregnant rats were exposed to nitrofen. At +16 and +20 dpc, fetuses were treated with simvastatin or sildenafil or simvastatin+sildenafil. At +21 dpc a post-mortem micro-CT and autopsy were performed.

Main results All nitrofen-treated fetuses had a lower birth weight compared to controls. In the simvastatin-treated group, a significant improvement was noted in CDH fetuses. Impairment of the kidney, lung and liver was also noted in CDH. Compared to controls, CDH fetuses showed a lower ventricular mass, with a greater left ventricular thickness. Simvastatin treatment decreased the ventricular mass and improved wall thickness in CDH. CDH animals exhibited myocardial hypotrophy, severe vascular depression in the left ventricle and intense intestinal edema in comparison with controls and nitrofen exposed animals without CDH (NDH). In CDH animals, the cardiac morphology appeared severely deformed with left ventricular wall verticalization. Simvastatin treatment improved cardiac myocyte appearance and heart morphology.

Conclusions cardiac impairment and remodelling were confirmed in CDH. The potential to treat CDH with antenatal simvastatin may improve the management of this malformation.
PROPOFOL USE IN NEONATES AND YOUNG INFANTS UNDERGOING SURGERY ALLOWS A QUICKER AND EASY INTUBATION

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**Aim of the study:** To evaluate the safety, effectiveness and speed of propofol induction during the intubation phase of anaesthesia in full-term neonates and young infants. **Methods:** All consecutive infants below 6 months of age, undergoing general anaesthesia between 2011 and 2013, were enrolled. Propofol induction (4 mg/kg iv) was compared with sevoflurane induction (4%). Recorded parameters were: time required for oro tracheal intubation (OTI), quality of OTI, heart rate, blood pressure and oxygen saturation before OTI (T0), at OTI (T1), and after recovery of spontaneous breathing (T2), time required for recovery of spontaneous breathing, prevalence of complications (seizures, skin rash). Mann-Whitney and Fisher’s exact test were used as appropriate. Results are median (IQ range) or prevalence; p<0.05 was considered significant. **Results:** 160 infants were enrolled in the study, 80 in each group. Major surgery (involving organs in the thoracic, abdominal or pelvic cavities) was performed in 64% and 54% of patients in the propofol and sevoflurane group, respectively (p=0.07). Patients in the propofol group showed shorter time for OTI [11.5 (4.0-65) versus 360.0 (228.0-720.0) seconds, (p<0.0001)] and recovery of spontaneous breathing [59.0 (4.0-140.0) versus 480.0 (420.0-960.0) seconds (p<0.001)]. No significant complications were recorded in both groups. **Conclusions:** In neonates and infants up to 6 months of age, a single bolus of 4 mg/kg of propofol is a safe and effective alternative to sevoflurane. In addition, propofol use allows a quicker intubation, without affecting the hemodynamic parameters.
OUTCOMES OF LAPAROSCOPIC REPAIRS OF INCARCERATED INGUINAL HERNIA IN CHILDREN

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Aim: Laparoscopic hernia repair (LHR) role in management of incarcerated cases is unclear. This review analyzes the outcomes after laparoscopic repair of incarcerated inguinal hernia in children.

Methods: Literature was searched on PubMed® using terms “laparoscopic”, “incarcerated”, “inguinal”, “hernia” and “children”. Age, sex, side, sac content, operative technique, follow-up period, complication and recurrence rate were analyzed.

Results: Fifteen articles with n=689 incarcerated inguinal hernias were found. Median age was 22.4 month (2 weeks-16 years). In n=355 (51.5%) manual reduction and delayed surgery (MRDS), n=34 (4.9%) manual reduction in general anesthesia (MRGA) followed by emergency LHR and n=300 (43.5%) intraoperative reduction (IOR) was necessary. Incarcerated contents were documented in n=68: intestine n=36 (52.9%), ovary n=14 (20.6%), omentum n=11 (16.2%), appendix n=5 (7.4%) and Meckel’s diverticulum n=2 (2.9%). Among the n=18 girls in IOR group n=14 (77.8%) had ovarian incarcerated.

For LHR, hook method was used in n=376 (54.6%) and purse string suture in n=313 (45.4%); with 2 conversions in IOR group. Mean follow-up was 15 months (3-80 months); with testicular atrophy n=1 (0.15%), recurrence n=4 (0.58%) in MRDS and n=1 (0.15%) in IOR, all 5 cases were closed with purse string technique. Total recurrence rate was 0.73%; significantly higher (p=0.014) with purse string (n=5, 1.6%) than with hook (n=0).

Conclusion: Hook and purse string methods are equally popular in LHR for pediatric incarcerated hernias, with 50% hernia reductions possible intraoperatively. Recurrence rate is low and comparable with non-incarcerated hernias, however significantly higher with purse string than hook technique.
PW3-G04
3D PATIENT SPECIFIC MODELS FROM MRI SEGMENTATION AND TRACTOGRAPHY TO ENHANCE SURGERY PLANNING OF PELVIC TUMORS AND MALFORMATIONS
Cecile Olivia Muller1,2, Alessio Virzi1,2, Jean-Baptiste Marret1,2, Eva Mille1,2, Laureline Berteloot4,2, Pietro Gori3, Thomas Blanc1, David Grevent4, Frederic Bary1, Nathalie Boddart4,2, Isabelle Bloch1,3, Sabine Sarnacki1,2
1Pediatric surgery department, Necker Enfants Malades Hospital, Paris, France. 2IMAG2 laboratory, Imagine Institute, Paris, France. 3LTCI Telecom Paris Tech, Paris, France. 4Pediatric radiology department, Necker Enfants Malades Hospital, Paris, France

Aim of study: Image guided surgery is the next revolution in surgery that should be part of daily practice in management of pediatric patients with rare pelvic tumors and malformations. The first step requires 3D patient specific models derived from medical imaging data, including nervous anatomy to improve outcome of pelvic surgery.

Methods: Since 2016, clinical and radiological data of 65 patients between 3 months to 18 years of age were prospectively collected: 28 tumors, 34 malformations and 3 controls. Radiological examination was performed on a 3Tesla MRI with a standard protocol and two additional sequences: coronal 3D T2 for 3D modeling and axial diffusion tensor (25 directions, b=600) for neurotractography. Segmentation was performed using semi-automatic and manual methods.

Main results: 3D Slicer® was chosen as a relevant software tool for segmentation and development of in-house semi-automatic plugins to build patient’s specific 3D models of the pelvis. These models were computed for 35 pediatric patients using semi-automatic methods (pelvic bones and bladder), and manually for the other organs. After some learning, segmentation time dropped from 40 to 7 hours on average, including pelvic neurotractography.

Conclusions: These preliminary results are the first steps towards creating patient specific 3D models from MRI, including neurotractography, for pediatric malformations and tumors of the pelvis. Development of additional segmentation methods for other pelvic structures is mandatory to routinely use 3D patient specific models and evaluate their added value for surgical management.
PW3-G05:
LAPAROSCOPIC INGUINAL HERNIA REPAIR UNDER CAUDAL ANESTHESIA IS FEASIBLE IN CHILDREN

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¹Center of Pediatric Surgery, Hanover Medical School, Hannover, Germany. ²Clinic for Anesthesiology and Intensive Care Medicine, Hanover Medical School, Hannover, Germany

Aim of the Study:
To avoid potential side effects of general anesthesia, open inguinal herniotomy can be performed under caudal anesthesia in children. To date, for laparoscopic inguinal hernia repair, general anesthesia was mandatory. To combine the advantages of the minimal invasive surgery with the advantages of caudal anesthesia we examined the feasibility of performing laparoscopic hernia repair under caudal anesthesia in children.

Methods:
Six children (age 23 - 74 months) with diagnosis of inguinal hernia underwent laparoscopic herniorrhaphy under caudal anesthesia. Analgosedation was administered before placing the caudal blockade. The sedation was subsequently reduced to evaluate the feasibility of spontaneous breathing during laparoscopy. Routine monitoring of anesthesiological parameters was performed.

Main results:
During laparoscopy all children breathed spontaneously without the necessity for further supportive airway measures.
The caudal block was sufficient to provide analgesia during the laparoscopy. Intraoperatively we did not experience any episode of awakening or uncontrolled movements of the patients. Patients left the operating theater with an average time of 4.6 min after the end of the surgery. Enteral feeding could be started immediately after the operation.

Conclusion:
Laparoscopic inguinal hernia repair is feasible under caudal blockade with mild sedation. Promoting initial results we have introduced this approach to our routine concepts and will collect more data before a general recommendation can be given.
**PW3-G06**

**EFFECTS OF CARBON DIOXIDE INSUFFLATION AND TRENDELENBURG POSITION ON BRAIN OXYGENATION DURING LAPAROSCOPY IN CHILDREN**

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**Background/aim:** Laparoscopic appendectomy has become more popular compared to the open appendectomy in children nowadays. There are limited data on the effects of pneumoperitoneum and Trendelenburg position on cerebral oxygenation. This study was designated to evaluate the changes in cerebral saturation using near-infrared spectroscopy during laparoscopic surgery in children.

**Methods:** The children underwent laparoscopic (LAP Group, n=22) or open appendectomy (OPEN Group, n=22). Right and left cerebral oxygenation (RScO₂-LScO₂), heart rate (HR), mean arterial pressure (MAP), end-tidal CO₂ pressure (PETCO₂) and peripheral oxygen saturations (SpO₂) were recorded before anesthesia induction (T₀,baseline), after induction (T₁), after intubation (T₂), 5 minutes after intubation (T₃), 5 minutes after pneumoperitoneum-15th minute at OPEN (T₄), 5 minutes after Trendelenburg-20th minute at OPEN (T₅), 30 minutes after pneumoperitoneum-45th minute at OPEN (T₆), 5 minutes after supine position-skin suturing at OPEN (T₇), 5 minutes post-extubation (T₈).

**Results:** Groups were similar with respect to their demographic data. In LAP group, a significant increase in HR was recorded at T₅. No significant difference was observed in the MAP, PETCO₂, SpO₂, RScO₂ and LScO₂ values between the groups. There was a significant increase in the perioperative T₁-T₈ values compared to the T₀ values in LScO₂ of the LAP group.

**Conclusion:** Our results suggest that pneumoperitoneum and Trendelenburg position does not alter the hemodynamic values and can be safely performed in children without altering regional brain oxygenation levels.
PW3-G07
PEDIATRIC ENDOSCOPIC PILONAL SINUS TREATMENT (PEPSiT) FOR SUCCESSFUL MANAGEMENT OF PILONAL SINUS DISEASE RECURRENCE AFTER OPEN REPAIR
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Federico II University of Naples, Naples, Italy

Aim of the Study: This study aimed to report application of pediatric endoscopic pilonidal sinus treatment (PEPSiT) for an effective treatment of pilonidal sinus disease (PSD) recurrence after open repair.

Methods: Ten patients (6 boys and 4 girls with an average age 16.8 years [14-18]) with a recurrent PSD after open excision were operated in our unit using PEPSiT over the last 2 years. In regard to operative technique, we introduced the fistuloscope through a fistula hole. All the hairs were removed with endoscopic forceps. The cavity was abraded with endoscopic brush and finally cauterized with a monopolar electrode. External openings were not closed.

Main results: Average operative time was 27.7 minutes [24-43]. We did not report intraoperative neither postoperative complications. As for postoperative course, the average analgesic requirement was 20 hours [16-26] and the average hospital stay length was 22.4 hours [18-36]. The average time to return to full daily activities was 2.3 days [1-4]. At 1 month postoperatively, the external openings were closed in all patients, who were highly satisfied with cosmetic results. No cases of recurrence was recorded at a mean follow-up of 18 months.

Conclusions: Our preliminary results demonstrate that PEPSiT is an excellent technique for surgical treatment of PSD in children. In fact, it is technically easy, with a short and painless hospital stay and it allows to the operated patients an early return to full daily activities. In addition, it is also effective as second-line treatment option of PSD recurrence after open repair.
WHAT IS BEHIND THE ACUTE OVARIAN PAIN? A 6-YEAR MONOINSTITUTIONAL EXPERIENCE

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Hospital Infantil Universitario Niño Jesus, Madrid, Spain

Objectives: Evaluation of the treatment applied in patients with suspicious symptoms of ovarian torsion in our center and its subsequent evolution, for the proposal of a consensus therapeutic approach in these surgical emergency.


Results: The median age at diagnosis was 13 years (r: 4-17). In 11/27 we performed a laparotomy (all of them before 2015, except for one performed in 2017) and in 16/27 we opted for a laparoscopic approach. We performed an oophorectomy in 14/27 patients (10/19 between 2012-2014 and 4/8 between 2015-2017) and in 13/27 the ovary was preserved (4/12 between 2015-2017) after detorsion and/or extirpation of the ovarian tumor. Ovarian torsion was confirmed in 14/27 cases and ovarian mass in 17/27. The pathological anatomy confirmed 8 hemorrhagic cysts, 7 mature cystic teratomas, 2 serous cystadenomas, one granulosa cell tumor. No recurrent episodes of ovarian torsion were recorded. In the follow-up period (median of 10 months, r: 0-55), 1 case of ovarian atrophy was observed and in 12/13 cases in which the ovary was preserved, the viability of normal ovarian parenchyma was confirmed.

Conclusions: Our results confirm the high prevalence of benign lesions that cause ovarian torsion in the pediatric population; as well as the high survival of the ovarian parenchyma after detorsion, which justifies surgical treatments with ovarian preservation in these patients.
PW3-G09: DIAGNOSTIC ACCURACY OF PREOPERATIVE ULTRASOUND IN PREDICTING CONTRALATERAL INGUINAL HERNIA IN CHILDREN: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Aim: The incidence of children developing metachronous contralateral inguinal hernia (MCIH) is 7-15%. Exploration of the contralateral groin during unilateral hernia repair can prevent development of MCIH and subsequent surgery and anesthesia. Preoperative ultrasound is a less invasive strategy and potentially able to detect contralateral patent processus vaginalis (CPPV) and prevent MCIH.

Methods: We queried MEDLINE, Embase and Cochrane Library to identify studies regarding children aged <18 years diagnosed with unilateral inguinal hernia, who underwent preoperative ultrasound of the contralateral groin without clinical signs of contralateral hernia. Studies included in the meta-analysis compared ultrasonographic test results to contralateral exploration or clinical follow-up. Random-effects model was used to obtain pooled estimates of sensitivity, specificity, positive and negative likelihood ratios (LR+ and LR-), and area under the receiver operating characteristic curve (AUC).

Results: Thirteen studies (2091 patients) were included, seven (1013 patients) in the meta-analysis. Pooled sensitivity and specificity was 91% and 94% respectively. LR+ and LR- was 11.0 and 0.1 respectively. The AUC (0.973) shows high diagnostic accuracy of preoperative ultrasonography for detecting CPPV, although diagnostic ultrasonographic criteria largely differ. Three studies reported the diameter of the inguinal canal: 6.8±1.3 mm in children with CPPV, 2.70±1.17 mm and 9.0±1.9 mm in children with contralateral hernia.

Conclusion: Diagnostic accuracy of preoperative ultrasound to detect CPPV seems promising, although it might result in an overestimation of MCIH prevalence, since CPPV not invariably leads to MCIH. Unequivocal ultrasonographic criteria are inevitable for proper diagnosis of CPPV and subsequent prediction of MCIH.
MONITORING CEREBRAL AND RENAL OXYGENATION STATUS DURING NEONATAL DIGESTIVE SURGERIES USING NEAR INFRARED SPECTROSCOPY (NIRS)

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Introduction:
Neonates’ tolerance can be a crucial parameter in the choice of surgical management. NIRS, a non-invasive regional monitoring of tissue perfusion, can detect adverse events associated with surgery such as abdominal compartment syndrome.

Objective:
Evaluating changes in renal and cerebral regional oxygen saturation (rSO2) during neonatal digestive surgeries to detect and prevent tissue’s hypoperfusion.

Methods:
Prospective monocentric study. NIRS was evaluate during and after neonatal digestive surgeries in parallel to commonly used monitoring parameters.

Results:
Nineteen patients included operated mainly for congenital diaphragmatic hernia, gastroschisis, omphalocle, necrotizing enterocolitis, occlusion.
During surgery, renal rSO2 values tended to decrease with time (72% of decline after the first 30 min). Events related to pathological values were bowel manipulation, viscera reintegration, abdominal wall closure. There was a significant correlation between renal rSO2 and SpO2 (p < 0.01). Cerebral rSO2 values were stable. Interestingly, hemodynamics and respiratory parameters were stable during surgery. After surgery, the lowest mean values were in the first 6 h (60% of rSO2 anomalies). It gradually reverses to normal values over time, probably due to decrease of inflammation and edema, progressive disappearance of ileus, evacuation of intra-intestinal content, and adaptation of the abdominal wall.

Conclusion:
NIRS is a non-invasive bedside tool to monitor tissue perfusion. It guides surgery by evaluating tolerance of herniated viscera reintegration methods (one or two surgical times) in neonatal digestive surgeries. Monitoring during surgery and in the first 6 h postsurgery is essential, this period bearing the most risk of systemic organ failure.
PW3-G11
ASSESSMENT OF OVARIAN RESERVE BY SERUM ANTI-MÜLLERIAN HORMONE LEVELS AFTER PEDIATRIC OVARIAN TORSION SURGERY
Meltem Çağlar Oskaylı, Neslihan Gülçin, Erdem Öztatman, Gonca Gerçel, Murat Mutuş, Burhan Aksu, Çiğdem Ulukaya Durakbaş

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Aim of the Study: Determination of ovarian reserve by serum anti-Müllerian hormone (AMH) levels in children who underwent either ovarian preserving surgery or oophorectomy because of ovarian torsion.

Methods: Patients operated for unilateral ovarian torsion over ten years were contacted for the study with ethical committee approval. Seventeen patients agreed to be included.

Main Results: There were 10 patients who underwent ovarian detorsion and seven who underwent oopherectomy. The mean age at operation was 11.6±2.23 (8-15) years and 13.2±2.17 (10-16) years, respectively (p>0.05). The ovarian torsion was isolated in four in the first group and in three in the second. At the time of this study, the mean age was 18±2.11 (14-21) years with a mean postoperative follow-up of 5.9±2.8 (2-10.5) years. Echogenicity of all preserved ovaries was normal by ultrasonography with presence of antral follicles in 6. Three ovaries were smaller than expected for age, yet two of these had antral follicles. The mean AMH level was 5.54±2.25 ng/ml in the detorsion group and 2.70±2.11 ng/ml in the oopherectomy group with a statistically significant difference (p<0.05).

Conclusions: Presence of follicles in preserved ovaries after detorsion was reported previously. AMH is expressed in granulosa cells of growing follicles and its serum level is valuable in assessing the quantitative aspects of the ovarian reserve. This study is unique by indicating preservation of the ovary in children with torsion is well justified in terms of future ovarian reserve.
PW3-G12
FOLLICULAR ADENOMA IN CHILDREN - CLINICAL AND PATHOLOGICAL SPECTRUM
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Aim: Assessment of clinical aspects of thyroid follicular adenoma in children and an attempt to elaborate management algorithm useful in clinical practice.

Material and methods. Clinical database of all children operated due to thyroid disease in years 1993-2017 at was reviewed. The children in whom postoperative histology report showed follicular adenoma were selected and their medical files were analysed in detail. Clinical course of the disease, medical and family history, results of imaging studies, details of surgical management and final outcome were assessed in each case.

Results: Among 183 children operated on for thyroid pathology there were 56 patients with follicular adenoma (30,6%). Their age ranged from 7 to 18 years. Associated defects or diseases of other systems were noted in 13 of them (23,3%). Ultrasound scan showed a solitary nodular nodule in 37 children (66,1%). Multiple nodules in one lobe were detected in 5 patients, while further 14 children showed bilateral thyroid pathology. The result of preoperative cytologic examination was suspicious in 12 children (21,4%). Intraoperative assessment showed congenital anatomical anomalies of the thyroid gland in 24 children (42,8%). Forty-one children (73,2%) underwent at least unilateral total lobectomy. In six patients with multifocal disease, both lobes were affected by adenoma. One patient had two adenomas in one lobe.

Conclusions: Follicular adenoma in children present a wide clinical spectrum. High incidence of associated diseases of other systems and congenital anomalies of the thyroid gland may indicate on complex etiology of follicular adenoma.
PW4-TH01
ASSESSMENT OF THE CONCERNS OF CAREGIVERS OF CHILDREN WITH ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA RELATED TO FEEDING-SWALLOWING DIFFICULTIES
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**Aim:** Children with esophageal atresia-tracheoesophageal fistula (EA-TEF) may experience feeding and swallowing difficulties (FSD), which result in stressful interactions between children and caregivers. The aims of this study were to assess concerns of caregivers of children with EA-TEF related to FSD and its relationship with type of atresia, repair time and time to start oral feeding (TSOF).

**Methods:** Caregivers accompanying 16 children with EA-TEF were evaluated for age, sex, type of atresia, repair time and TSOF. Parents completed the Feeding/Swallowing Impact Survey (FS-IS) to assess the concerns of caregivers related to FSD. It has 3 subscales including daily activities, worry and feeding difficulties. Average scores range between 1 to 5, of which increasing scores reflect more caregiver concern.

**Results:** The mean age was 3.15±2.19 years, of which 68.8% were male. 56.3% of cases were isolated-EA, and 43.8% were EA-distal TEF. 62.5% of cases received early repair, and 37.5% had delayed repair. The median TSOF was 4 weeks (2-72 weeks). The mean scores of daily activities, worry, and feeding difficulties were 2.98±1.07, 2.67±1.06, and 2.44±0.92, respectively. No correlation was found between the subscale “daily activities” of FS-IS and atresia type, repair time and TSOF (p>0.05). Moderate to strong correlations were found between the subscales called “worry” and “feeding difficulties” of FS-IS and atresia type, repair time and TSOF (p<0.05, r=0.61-0.84).

**Conclusions:** The concerns of caregivers of children with EA-TEF about FSD are related with the type of anomaly and surgical outcome.
SUCCESSFUL TREATMENT OF RECURRENT POST-SURGICAL ANASTOMOTIC STENOSIS OF THE LEFT MAIN BRONCHUS WITH ENDOBRONCHIAL METALLIC STENT

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2Istanbul University, Cerrahpasa Medical Faculty, Department of Radiology, Istanbul, Turkey. 3Istanbul University, Cerrahpasa Medical Faculty, Department of Anesthesiology, Istanbul, Turkey

Aim: To report successful treatment of recurrent post-surgical stenosis of a case with traumatic left main stem bronchial rupture with endobronchial stenting.

Case: A 2.5-year-old male patient with blunt chest trauma due to a traffic accident was admitted to an outside center 7 months ago. Chest tube drainage was performed due to bilateral pneumothorax. CT detected rupture in the left main bronchus. Bronchoscopy confirmed that the rupture was in the left main bronchus. The patient was initially followed with intubation for twelve days. There was atelectasis at one month after trauma. Bronchoscopy revealed obliteration 1 cm below the carina. Balloon dilatation was performed three times due to stenosis at the anastomosis. Pneumonectomy was planned due to persistence of symptoms. Following admission, thoracic CT scan of the left main bronchus showed complete obliteration and total atelectasis of the lung. Bronchoscopy confirmed the radiological findings. Seven months after trauma, left thoracotomy was performed. One cm gap was found between the proximal and distal bronchial ends and an anastomosis was done. Postoperatively, the left lung progressively collapsed. Bronchoscopy at the 3rd postoperative week showed an intact but collapsing anastomosis. An endobronchial transanastomotic metallic stent was placed with successful expansion of the left lung. The stent was removed after 3 months. The lungs are normal two years after the operation and the patient is free of symptoms.

Conclusion: Endobronchial metallic stenting may successfully alleviate symptoms following recurrent post-surgical anastomotic stenosis of traumatic bronchial rupture.
PW4-TH03
CONGENITAL DIAPHRAGMATIC HERNIA: A SCIENTOMETRIC ANALYSIS OF THE GLOBAL RESEARCH ARCHITECTURE AND SCIENTIFIC ADVANCES

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Aim of the Study: To evaluate the global research architecture and scientific advances relating to congenital diaphragmatic hernia (CDH) using a combination of scientometric methods and visualization techniques.

Methods: A search strategy for the Web of Science™ database was designed for CDH publications from 1900 to 2016. Research output of countries, institutions, individual authors and collaborative networks was analyzed. Semi-qualitative research measures including citation rate and h-index were assessed. Choropleth mapping and network diagrams were employed to visualize results.

Main Results: A total of 3,669 publications were identified, originating from 76 countries (Figure 1). The largest number was published by the USA (n=1,250), UK (n=279) and Canada (n=215). The USA combined the highest number of cooperation articles (n=152), followed by Belgium (n=115) and the Netherlands (n=93). The most productive collaborative networks were established between UK/Belgium (n=53), Belgium/Spain (n=47) and UK/Spain (n=34) (Figure 2). Canadian publications received the highest average citation rate (22.8), whereas the USA had the highest country-specific h-index (72). Eighty-five (2.3%) articles were published by international multicenter consortiums and national research networks. The most productive institutions and authors were based in North America and Europe.

Conclusions: CDH-related research has constantly been progressing, involving today many disciplines with main research endeavors concentrating in a few high-income countries. Recent advances in prenatal interventions and regenerative medicine therapy hold now the promise of improving CDH outcome in the 21st century. International collaborations and translational research should be strengthened to allow further evolution in this field.
PW4-TH04
FROM A REPORT OF 114 BRONCHOGENIC CYSTS (BCs) ON ADULTS AND CHILDREN: EARLY EXCISION MANAGEMENT APPEARS NOW CLEARLY MANDATORY

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¹Department of Pediatric Surgery, CH Henri Duffaut, Avignon, France. ²IMM, Paris, France. ³CHU, Strasbourg, France. ⁴Department of Thoracic Surgery, La Timone Hospital, Marseille, France. ⁵Department of Pediatric Surgery, La Timone Hospital, Marseille, France

Aim of the study: We established previously on a small pediatric cohort that an early surgical resection could bring a real benefit to children in terms of evolutive complications. Herein, we go further, by presenting the largest combined series of children and adults BCs ever described in the literature to date, we finally hope to demonstrate whether or not, BCs must be operated preventively or when they become symptomatic.

Methods: In this multicentric retrospective study (from 2000 to 2015), 114 patients aged from 3 months to 77 years were included. Epidemiological, clinical and histopathological data were collected and analyzed separately by age and by symptomatology.

Main results: Of the 32 children, 19 (41%) were operated preventively without any complications, the other 13 symptomatic children (59%) required surgery and developed the same order of complications than the asymptomatic adults; the most complications being seen for the symptomatic adults. Only 4 thoracotomies, 49 thoracoscopies and 4 minimal pulmonary resections were performed for asymptomatics (children and adults combined), while 22 thoracotomies, 32 thoracoscopies, and 7 conversions were required for symptomatics, associated with 11 pulmonary resections (8 lobectomies), 7 bronchial or esophageal closing fistulas, and 4 pneumolyses. The post-operative duration was 6 d for symptomatics Vs 4 d for asymptomatics (p<0.01). Inflammatory elements were observed in 5 children (15%) and 61 adults (74%) (p<0.01).

Conclusions: Here, we demonstrated that BCs must be operated preventively. Early surgery appears safe, and permit to reduce the duration of hospitalization as well as complications associated with evolutive BCs.
PW4-TH05
ULTRASONOGRAPHIC ASSESSMENT OF MEDIASTINAL SHIFT ANGLE (MSA) IN ISOLATED LEFT CONGENITAL DIAPHRAGMATIC HERNIA FOR THE PREDICTION OF POSTNATAL SURVIVAL
Anita Romiti, Milena Viggiano, Laura Valfré, Andrea Conforti, Lucia Aite, Lucilla Ravà, Marta Ciofi Degli Atti, Piero Bagolan, Leonardo Caforio
Bambino Gesù Children’s Hospital Clinical and Research Institute, Rome, Italy

Objectives: To quantify mediastinal shift in isolated CDH, by the introduction of a new ultrasonographic (US) marker, defined as mediastinal shift angle (MSA) and to evaluate its ability in predicting postnatal survival.

Methods: Twentyfour fetuses from singleton pregnancies with isolated left-sided CDH were included in the study group and subdivided in group A (16 survivors) and group B (8 non survivors). Study group was matched with a control group of 95 fetuses from singleton pregnancies free from chromosomal anomalies. On the same US stored images commonly used for LHR measurement, a landmark line was drawn from posterior face of the vertebral body, splitting it into two equal parts, to the mid surface of the sternum. Another landmark line was traced from the vertebral body to the lateral wall of the right atrium. The angle between these two lines defined mediastinal shift and called MSA.

Results: Median MSA was significantly different between group A (34.3°) and group B (42.7°) (p<0.001) and between study group and the control group (19°) (p<0.001).

Statistical analysis confirmed an inverse correlation between MSA values and survival (P=0.004). The best cut-off value for MSA was 43.7° which demonstrated the highest discriminatory power (sensitivity 63%; specificity 93.75%). In fetuses with isolated CDH, mediastinal shift may be quantified using MSA and this US marker seems to reliably predict survival.

Conclusions: In conclusion, our results show that, similarly to LHR and O/E LHR, the MSA has a good correlation with survival in isolated left sided CDH.
**PW4-TH06**  
**CONGENITAL DIAPHRAGMATIC EVENTRATION AND HERNIA SAC: CLINICALLY ONE ENTITY WITH UNEXPECTED OUTCOMES**

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Radboudumc - Amalia Children's Hospital, Nijmegen, Netherlands

**Aim of the study:** Congenital diaphragmatic eventeration (CDE) is considered a specific type of congenital diaphragmatic hernia (CDH). As it is mostly described as part of a CDH cohort, little is known on outcomes and prognosis. This study provides insights in the prognosis of CDE compared to classical CDH and evaluates whether CDH with hernia sac is comparable to CDE.

**Methods:** All surgically treated patients with CDE or Bochdalek type CDH (CDH<sub>Bochdalek</sub>) from 2000-2016 were included in this retrospective analysis. Demographics, CDH-characteristics, treatment, survival, complications and pulmonary status were retrospectively evaluated. CDE and CDH<sub>Bochdalek</sub> patients were compared.

**Main results:** In total 189 patients were included. There were no significant differences in demographics or outcome parameters in patients with a CDE (n=16) or CDH with hernia sac (n=19). Therefore, these were combined for the comparison (CDE<sub>total</sub>, n=35). The 1-year survival rate of CDE<sub>total</sub> compared to CDH<sub>Bochdalek</sub> was significantly higher. However, there was no significant difference in recurrence rate after one year (Table 1). They required significantly more oxygen suppletion at 30 days and at discharge than primary repaired CDH<sub>Bochdalek</sub> patients with higher recurrence rates (Table 2).

**Conclusion:** A congenital diaphragmatic eventration and Bochdalek CDH with hernia sac seem to form one entity. They have significantly better survival than Bochdalek type CDH. Though they have better short term outcome than patch repaired CDH patients, the recurrence rate and requirement of pulmonary support is higher than primary repaired patients.

**Table 1. Treatment and surgical outcomes of CDE<sub>total</sub> versus CDH<sub>Bochdalek</sub>**

<table>
<thead>
<tr>
<th></th>
<th>CDE&lt;sub&gt;total&lt;/sub&gt; (n=35)</th>
<th>CDH&lt;sub&gt;Bochdalek&lt;/sub&gt; (n=154)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Survival, n(%)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30-days</td>
<td>35 (100)</td>
<td>127 (83)</td>
<td>0.007</td>
</tr>
<tr>
<td>1-year</td>
<td>34 (97)</td>
<td>117 (76)</td>
<td>0.005</td>
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<tr>
<td><strong>Recurrence</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30-days</td>
<td>3 (9)</td>
<td>0 (0)</td>
<td>0.006</td>
</tr>
<tr>
<td>1 year&lt;sup&gt;1&lt;/sup&gt;</td>
<td>5 (15)</td>
<td>7 (5)</td>
<td>0.13</td>
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<tr>
<th></th>
<th>CDE&lt;sub&gt;total&lt;/sub&gt; (n=35)</th>
<th>CDH&lt;sub&gt;Bprim&lt;/sub&gt; (n=84)</th>
<th>P-value</th>
<th>CDH&lt;sub&gt;Bpatch&lt;/sub&gt; (n=70)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Survival, n(%)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 year</td>
<td>34 (97)</td>
<td>80 (95)</td>
<td>1.00</td>
<td>37 (53)</td>
<td>&lt;0.001</td>
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<td>Recurrence</td>
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<tr>
<td>30 days</td>
<td>3 (9)</td>
<td>0 (0)</td>
<td>0.02</td>
<td>0 (0)</td>
<td>0.04</td>
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<tr>
<td>1 year&lt;sup&gt;1&lt;/sup&gt;</td>
<td>5 (15)</td>
<td>1 (1)</td>
<td>0.008</td>
<td>6 (9)</td>
<td>0.50</td>
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<tr>
<td><strong>Pulmonary state 30 days&lt;sup&gt;2&lt;/sup&gt;</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Oxygen supplication</td>
<td>14 (44)</td>
<td>7 (9)</td>
<td>&lt;0.001</td>
<td>24 (60)</td>
<td>0.17</td>
</tr>
<tr>
<td>Mechanical ventilation</td>
<td>13 (41)</td>
<td>6 (8)</td>
<td>&lt;0.001</td>
<td>18 (45)</td>
<td>0.71</td>
</tr>
</tbody>
</table>

Missing values: <sup>1</sup>6, <sup>2</sup>41
PW4-TH07
CONGENITAL LUNG ABNORMALITY QUANTIFICATION BY COMPUTED TOMOGRAPHY: THE CLAQ SCORE

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Aim of the study: Objective chest CT scoring is needed in order to execute clinical trials for the management of congenital lung abnormalities (CLA). We aimed to develop a reliable quantitative scoring method for CLA’s to aid in clinical decision-making.

Methods: In congenital lung abnormality quantification (CLAQ) scoring, axial slices every 5 mm are overlaid with a grid and annotated according to the abnormality within. Grid size is relative to maximal lung width. Blinded for diagnosis, random CT-scans of children with CLA were reviewed for all visible parenchymatic abnormalities and assigned a certain hierarchy according to clinical relevance by a pediatric pulmonologist, radiologist and surgeon (Figure 1). Using this method, two observers scored 19 out of 120 randomly selected anonymized CT-scans in children with CLA twice. Inter- and intra-observer agreement was calculated using a two-way mixed-effects model in which kappa-values (intraclass correlation coefficient) greater than 0.8 were considered excellent.

Main results: Clinical diagnosis of these 19 patients were congenital pulmonary airway malformation (n=4), bronchopulmonary sequestration (n=4), congenital lobar emphysema (n=3) and a bronchogenic cyst (n=1). Bronchial atresia was observed in 7 patients. Inter-observer correlation coefficients for the total disease percentage and most frequently scored abnormalities (CRSW/CRHypoD/PRHypoD) were all excellent. Intra-observer scores for the two individual observers were excellent for the aforementioned abnormalities and total disease percentage.

Conclusions: The CLAQ score is a quantitative method for objective characterization of CLA with excellent inter – and intraobserver reliability scores. The remaining scans will be scored for further validation.
PW4-TH08
IS EARLIER BETTER? LONGTERM PULMONARY FUNCTION AFTER LOBECTOMY

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Introduction

Recommendation of early pulmonary resection in asymptomatic congenital pulmonary airway malformations (CPAMs) is based on the presumed compensatory lung growth during the first years of life, along with avoidance of future infectious complications. The aim is to analyse the long-term pulmonary function after lobectomy before and after one year of age.

Methods

Retrospective review of children who underwent lobectomy for CPAM (2001-2016) was performed. Patients who were old enough (>5 years) for spirometry were included in the study and were divided in two groups (surgery before or after 12 months of age). Pulmonary function testing values were considered normal if they were >80% of predicted.

Main Results

Forty-seven patients underwent pulmonary lobectomy for CPAM, 23 of them met the inclusion criteria and prospectively performed a spirometry. Among them, 7 had surgery before and 16 after one year of age (0.1 vs 2), being both groups comparable in terms of sex, type of CPAM and surgical approach. Time from surgery until pulmonary function testing was longer in patients who had surgery before one year of age (9.1 vs 4.6, p=0.003). After correcting results by time from surgery until spirometry, a better FEV1/FVC was found in patients who had surgery after one year of age (90 vs 77%, p=0.043).

Conclusion

Although spirometry may be influenced by many other variables, this preliminary study could not prove better pulmonary function in patients with early lobectomy. Further studies are required in order to resolve the best age to perform lobectomy in CPAMs.
PW4-TH09
RADIATION-FREE IMAGING OF PECTUS EXCAVATUM
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Aim of the Study
The severity of Pectus Excavatum PE is usually evaluated by computed tomography imaging (CT). This evaluation should not be repeated to avoid excessive irradiation. The OrtenBodyOne scanner is a noninvasive 3-dimensional imaging system recently developed for evaluation of PE severity. It uses depth sensors to scan the entire 3D external body surface of a patient. This study aims to evaluate the utility of this new imaging system for the evaluation of PE severity.

Methods
Patients treated for a PE from April 2015 to January 2017 with available CT and OrtenBodyOne data were included. PE depth, thoracic width and length, Haller Index, Anthropometric index and Asymmetric index were calculated from CT and OrtenBodyOne images. Correlations between PE indexes calculated from CT and from OrtenBodyOne were calculated applying the non-parametric Spearman correlation procedure with Bonferroni correction adjusting for multiple comparisons. This study was approved by the local ethics committee.

Main results
Forty males (90.9%) and 4 females (9.1%), 18 with symmetric (40.9%) and 25 with asymmetric PE (56.8%) were included. The mean age was 16.2 years (4.3–63.5). The following measures and indexes acquired by OrtenBodyOne and by CT were significantly correlated: PE depth (r= 0.84, p=0.002), Anthropometric index (r= 0.81, p=0.002) and Asymmetric index (r= 0.67, p=0.002). The correlation between CT Haller Index and External Haller index was not significant (r= 0.44, p=0.05).

Conclusion
OrtenBodyOne imaging system can be used to evaluate the severity of PE. Measures can be repeated throughout treatment while avoiding unnecessary irradiation.
NEW INSIGHT ON MORTALITY RISK FACTORS IN ESOPHAGEAL ATRESIA PATIENTS: RESULTS FROM THE EUPSA OESOPHAGEAL ATRESIA REGISTRY

Andrea Conforti1, Francesco Morini1, Laura Valfrè1, Michael Hollwarth2, Ernest van Heurn3, Prem Puri4, Pietro Bagolan1, on behalf of the EUPSA Oesophageal Atresia Registry

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Aim: improving survival rate in infants with oesophageal atresia (OA) with/without trachea-oesophageal fistula (TOF) has been extensively reported. Low birth weight and cardiac anomalies are commonly considered as the most influencing risk factors. Nonetheless, the impact of others risk factors in predicting mortality has not been extensively investigated. Aim of this study was to describe the role of several risk factors in predicting mortality in OA/TOF patients enrolled in an International multicentre registry.

Methods: All patients consecutively registered from July 2014 to December 2017 in the EUPSA Oesophageal Atresia Registry (EUPSA-OAR) were included in the study. Patients were categorised based on survival, and possible risk factors were analysed. Fisher's exact and Mann-Whitney test were used as appropriate. Results: During the study period, 374 patients were consecutively recorded by 24 active Centres. Table shows main findings. Conclusions: Present data from a large cohort of patients from the EUPSA-OAR suggest that, besides classical risk factors, perinatal indicators of health, associated malformations, and post-operative complications (anastomotic leak, pneumothorax and sepsis) are associated to mortality in OA/TOF infants.

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Survivors (n=94)</th>
<th>Non-survivors (n=280)</th>
<th>p-Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth weight (&lt;2500 g)</td>
<td>25 (26.6)</td>
<td>131 (46.8)</td>
<td>0.097</td>
</tr>
<tr>
<td>GA, median (w)</td>
<td>38 (31-39)</td>
<td>38 (30-39)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>GA, median (d)</td>
<td>270 (223-330)</td>
<td>270 (222-330)</td>
<td>0.601</td>
</tr>
<tr>
<td>Prematurity, %</td>
<td>5</td>
<td>85</td>
<td>0.406</td>
</tr>
<tr>
<td>Type of presentation (newborn)</td>
<td></td>
<td></td>
<td>0.763</td>
</tr>
<tr>
<td>Neonatal age, median (d)</td>
<td>11 (7-19)</td>
<td>11 (7-19)</td>
<td>1.000</td>
</tr>
<tr>
<td>Low birth weight, %</td>
<td>1</td>
<td>27</td>
<td>0.706</td>
</tr>
<tr>
<td>Leukocytes, mm³</td>
<td>270 (223-330)</td>
<td>270 (222-330)</td>
<td>0.601</td>
</tr>
<tr>
<td>Oxygen saturation (mmHg)</td>
<td>100 (95-100)</td>
<td>98 (90-110)</td>
<td>0.008</td>
</tr>
<tr>
<td>Cardiac anomalies, %</td>
<td>10</td>
<td>14</td>
<td>0.005</td>
</tr>
<tr>
<td>Cardiac anomalies, %</td>
<td>1</td>
<td>0</td>
<td>1.000</td>
</tr>
<tr>
<td>Cardiovascular malformations, %</td>
<td>5</td>
<td>14</td>
<td>0.371</td>
</tr>
<tr>
<td>Chromosomal malformations, %</td>
<td>5</td>
<td>14</td>
<td>0.971</td>
</tr>
<tr>
<td>CLSI, %</td>
<td>5</td>
<td>0</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Severe, %</td>
<td>2</td>
<td>37</td>
<td>0.022</td>
</tr>
<tr>
<td>Venous, %</td>
<td>9</td>
<td>26</td>
<td>0.886</td>
</tr>
<tr>
<td>Respiratory infections, %</td>
<td>6</td>
<td>14</td>
<td>0.826</td>
</tr>
<tr>
<td>Primary respiratory, %</td>
<td>1</td>
<td>2</td>
<td>0.796</td>
</tr>
<tr>
<td>Infant infection 1 to 3 weeks, %</td>
<td>6</td>
<td>38</td>
<td>0.044</td>
</tr>
<tr>
<td>Anastomosis complications, %</td>
<td>4</td>
<td>6</td>
<td>1.000</td>
</tr>
<tr>
<td>Primary respiratory, %</td>
<td>11</td>
<td>366</td>
<td>0.978</td>
</tr>
<tr>
<td>Electrolyte balance, %</td>
<td>12</td>
<td>117</td>
<td>0.418</td>
</tr>
<tr>
<td>Surgical complications, %</td>
<td>15</td>
<td>52</td>
<td>0.034</td>
</tr>
<tr>
<td>Anastomotic leak, %</td>
<td>0</td>
<td>30</td>
<td>0.04</td>
</tr>
<tr>
<td>Urine culture positive, %</td>
<td>2</td>
<td>19</td>
<td>0.851</td>
</tr>
<tr>
<td>Renal failure, %</td>
<td>2</td>
<td>19</td>
<td>0.851</td>
</tr>
<tr>
<td>Pneumothorax, %</td>
<td>5</td>
<td>14</td>
<td>0.515</td>
</tr>
<tr>
<td>Cardiac arrest, %</td>
<td>1</td>
<td>7</td>
<td>0.468</td>
</tr>
<tr>
<td>Death, %</td>
<td>1</td>
<td>8</td>
<td>1.000</td>
</tr>
</tbody>
</table>
**Aim:** Since the introduction of the Nuss’ procedure, there has been a certain preference for its excellent outcomes. Nonetheless, it requires mediastinal and pleural invasion with plausible fatal complications. Procedures such as Taulinoplasty avoids entering the thoracic cavity with a completely extra thoracic approach. The aim of this study is to compare the results of Taulinoplasty and Nuss procedure.

**Methods:** In the last 5 years, 49 patients with pectus excavatum (PE) underwent corrective surgery. Patients were evaluated for the surgical approach, surgical outcome and quality of life surveys (QoL) comparing both procedures.

**Results:** Taulinoplasty (TP) was performed in 23 patients while Nuss procedures (NP) in 26. Both groups had similar Haller index before surgery (TP:4.34 NP:4.63) with an immediate correction of the sternal defect in all cases. TP had shorter surgical time (TP:55±5min NP:120±60min), reduced the hospital stay (TP:5±1days NP:7±2.5days), and required less analgesics. Complications were similar in both groups. 4 cases (17.3%) of TP recurred due to plate dislodging while 3 cases (11.5%) had mobilization of the bar. QoL had a positive improvement in most cases for both groups.

**Conclusions:** Taulinoplasty is an effective treatment with similar corrective results as in Nuss procedure, with shorter operative time, requires less analgesia and shortens the hospital stay. But its main advantage is the extra-thoracic approach that avoids the risk of injury of vital organs providing a safe alternative for the repair of PE.
PW4-TH12
MID-TERM FUNCTIONAL OUTCOME AND NUTRITIONAL STATUS FOLLOWING GASTRIC TRANSPOSITION: COMPARISON OF ESOPHAGEAL ATRESIA AND CORROSIVE ESOPHAGEAL STRicture CASES
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Aim of the study: We aimed to evaluate mid-term functional and nutritional status of the patients who had undergone gastric transposition (GT) for esophageal replacement.

Methods: Hospital records of consecutive esophageal atresia (EA,n=23) and corrosive esophageal stricture (CES,n=13) patients who had undergone GT between November 2012-June 2017 were reviewed. Demographics, previous surgical history, clinical characteristics, biochemical and radiologic workup were evaluated. Patients were grouped according to primary diagnosis and were compared in terms of mid-term functional and nutritional states.

Main results: Mean age at surgery was 43.8±39.6 months. Laparoscopic GT was performed in 6 (16.7%). Pyloroplasty and Witzel-jejunostomy were not performed in 13 (36.1%). One major (opening of tracheoesophageal fistula) and two minor (bleeding and pneumothorax) complications developed intraoperatively. Means for age at operation and weight were significantly lower in EA group that had a higher rate of comorbidities. Pyloroplasty rate and intraoperative complications were similar in both groups. Time for extubation, oral feeding, hospital stay, need for dilatation, postoperative hoarseness and dumping syndrome findings were higher in EA group. Mean duration for scintigraphic gastric emptying was longer and need for metoclopramide use was higher in CES group. Both groups had similar hemoglobin, iron, iron binding capacity, B12, folic acid, glucose levels.

Conclusions: In the early postoperative period, anastomotic problems is more likely to occur in EA group. Dumping syndrome is more frequent in EA patients whereas gastric emptying is more problematic in CES patients following GT. Nutritional status is similar in both groups.
PW5-HB02
TRANS-UMBILICAL LAPARO-ENDOSCOPIC SINGLE SITE SURGERY WITH HEPATIC DUCTOPLASTY IN MANAGEMENT OF CHILDHOOD CHOLEDOCHAL CYST

Tran Son, Duong Mai
Saint Paul Hospital, Hanoi, Vietnam

**Aim:** To present our techniques and results of trans-umbilical laparo-endoscopic single site surgery (TULESS) with hepatic ductoplasty in management of childhood choledochal cyst (ChC).

**Methods:** All ChC cases undergoing TULESS excision of ChC, ductoplasty and hepaticojejunostomy by the same surgeon from October 2012 to October 2017 were reviewed. For TULESS, 3 trocars were placed at a single umbilical skin incision and conventional instruments were used.

**Results:** 46 patients (35 girls, 11 boys) with hepatic ductoplasty were identified from total 237 patients with ChC undergoing TULESS by the same surgeon for that period. The median age was 17 months. Ductoplasty for a small common hepatic duct (less than 5mm) was carried out in 38 patients and for presence of an aberrant duct in 8 patients. The median operative time was 195 minutes. There was no intraoperative complication, no conversion to open surgery. Additional trocars (conversion to conventional laparoscopic surgery) were required in just the first case of aberrant duct. Postoperative bile leak was noted in one patient (2.2%), which was resolved with non-operative treatment. The median postoperative hospital stay was 5 days. At a median follow up of 36 months (range: 3 months to 60 months), one patient needed a redo surgery for anastomotic stenosis; all other patients were in good health, with excellent postoperative cosmesis.

**Conclusions:** TULESS with conventional instruments is feasible and safe for hepatic ductoplasty in most cases of childhood ChC with small common hepatic duct or aberrant duct.
PW5-HB03: WHAT HAPPENS AFTER KASAI FOR BILIARY ATRESIA? A EUROPEAN MULTICENTRE SURVEY

Joel Wong, Mark Davenport
Kings College Hospital, London, United Kingdom

**Aim of study**: Biliary atresia (BA) is rare and while Kasai portoenterostomy (KPE) is the standard there is a lack of evidence for optimum post-operative management. We sought to determine the range of options across Europe.

**Methods**: Survey of current pre- and post-operative practice, Data are quoted as median (range). Centres known to have an interest in BA were approached preferentially.

**Results**: 13 respondent centres (from 9 countries) with wide range new cases seen (1 - >15) per year. 5 centres see ≤5 patients/year, and 3 centres >15 patients/year.

- 82(60–100) % of infants were described as isolated BA, 10(3-20)% as syndromic BA, 5(1-10)% as cystic BA and 4(0-15)% as CMV-positive BA.
- Ultrasound was universal, liver biopsy (n=5); radio-isotope (n=3); MRCP(n=2); ERCP (n=1) and laparoscopic cholangiogram(n=1). Viral serology was tested in 11 centres.
- Modal age at KPE was 61–70 days old. All centres perform an open KPE. A steroid-based regimen was used in 8, although there was marked variation in regimens. Modal perioperative antibiotics were a combination of beta-lactamase inhibitor and gentamicin (n=8). 10 centres prescribe oral antibiotics, with varying duration (4–52 weeks). If CMV IgM serology was positive, 5 would treat with specific anti-viral therapy. Ursodeoxycholic acid (n=13), phenobarbitone (n=4) and cholestyramine (n=2) were used. Declared clearance of jaundice varied from 30% (1/13) to >60% (5/13).

**Conclusion**:

- All perform an open Kasai portoenterostomy.
- No consensus on a standard post-KPE drug regimen.
- Wide variability of outcome.
FIGURE: countries with centralisation of BA.
DEMOGRAPHIC ANALYSIS OF PRAGMATIC CLASSIFICATION OF BILIARY ATRESIA IN ENGLAND AND WALES (1999-2015)

Maria Asimakidou¹, Kat Ford¹, Jess Burns¹, Michael Dawrant², Naved Alizai³, Khalid Sharif³, Evelyn Ong³, Erica Makin¹, Mark Davenport¹

¹Kings College Hospital, London, United Kingdom. ²Leeds General Infirmary, Leeds, United Kingdom. ³Birmingham Childrens Hospital, Birmingham, United Kingdom

AIM OF STUDY: Biliary atresia (BA) is characterised by aetiological heterogeneity with a number of possible variants. We sought to define their demographic features.

METHODS: Prospective national dataset entry since 1999. All were histologically confirmed cases of BA born in England or Wales; demographic details were added later from medical records. BA was divided into four pragmatic groups based on presumed identity [isolated biliary atresia (IBA); Biliary Atresia Splenic Malformation (BASM); Other anomalies+BA (ANOM) and cystic biliary atresia (CBA).  ANOVA and Chi² were used and a P value of 0.05 was considered significant.

RESULTS: There were 703 (54% female) infants with BA that could be divided into IBA (n = 516, 73.3%); ANOM(n=33, 4.6%); BASM(n=96, 13.5%) and CBA (n =61, 8.7%) (N.B. possible variant overlap included). Mean gestational age was lower in BASM and ANOM compared to CBA (37.8±0.53, 38.1±0.29 vs. 39.1±0.26 weeks; P =0.04). Mean birth weight varied significantly in the order CBA(3.3kg) >IBA(3.1kg)> BASM (3.1kg)>ANOM (2.7kg)(ANOVA, P = 0.004). Gender varied with significant female predominance in BASM (65%) and CBA (67%) but not IBA (50%) and ANOM (54%)(P = 0.001). Parental racial background varied significantly with decreasing Caucasian proportion: CBA(84%);BASM(77%);IBA(73.4%) and ANOM(64%)(P=0.02, for trend).

CONCLUSIONS: There is a remarkable demographic variation between pragmatic variants in gender, racial background and birth attributes.
PW5-HB05
RESULTS AMD ANALYSIS OF THE CURRENT SITUATION OF URINARY SULFATED BILE ACID (USBA) EXAMINATION FOR DIAGNOSIS OF BILIARY ATRESIA
Masayuki Obatake¹, Yasuaki Taura², Yusuke Yamane², Takuya Yoshida², Koichi Sakamoto¹, Yuki Fujieda¹, Kazuhiro Hanazaki¹

¹Kochi Medical Scholl, Kochi, Japan. ²Nagasaki University, Nagasaki, Japan

Aim of the study: Urinary sulphated bile acid (USBA) has been used as an index for early diagnosis of biliary atresia (BA) since 2007, and the feasibility of UABA was introduced at the 2010 EUPSA in Bern. We reported experiences of operating USBA and evaluated the current situation of USBA for diagnosis of BA.

Methods:
USBA samples were collected from infants 14 - 28 days old in Nagasaki prefecture. The infants whose USBA showed more than 55 µmol/g creatinine underwent a detailed examination, including a blood test and abdominal ultrasonography.

Results:
From August 2007 to November 2017, the total number of USBA examinations were 52972. The examination rate has increased to more than 60% of prefectural annual births since 2013. USBA value of 1122 (2.1%) of 52972 infants exceeded the cut-off value. During the period of using USBA, 12 infants were diagnosed as BA. Seven of 12 received USBA examination. They received examinations at the age of 9 - 32 days and Kasai procedures at the age of 32 - 111 (60.1 ±26.1) days. The patients of extrahepatic BA type were 6 in type III and one in aplasia of hepatic radicles.

Conclusions:
USBA could note the possibility of BA in neonatal period, however necessity of further examination for BA diagnosis delayed the Kasai procedure. Comparing the time of Kasai procedure 87.6±54.6 days of age without USBA (from 1991 to 2017), USBA can provide to get an earlier Kasai procedure done.
MULTIMODAL MANAGEMENT OF MAIN BILE DUCT STONES IN INFANTS AND OLDER CHILDREN

Silvia Scarvaglieri Mazzeo1, Marianna Iaquinto1, Maria Grazia Scuderi1,2, Emmanuel Gonzalez1, Stéphanie Franchi1, Virginie Fouquet1, Hélène Martelli1, Sophie Branchereau1, Florent Guérin1

1Bicêtre Hospital, Le Kremlin Bicêtre, France. 2Policlinico Rodilico Vittorion Emanuelle, Catane, Italy

Introduction:
Our goal is to understand the most effective treatment, ranging from observation, interventional radiology, endoscopy, or surgical intervention for main bile duct stones (MBDS) in children according to age groups.

Methods: We conducted a monocentric retrospective study of patients admitted in our centre for MBDS between 2005 and 2016. We looked at age, symptoms, management and outcome, we distinguished patients over (Group A) and under one year of age (Group B).

Results:

<table>
<thead>
<tr>
<th></th>
<th>Group A: &lt;1Years old</th>
<th>GroupB:&gt;1 Years old</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>n</td>
<td>48</td>
<td>42</td>
<td></td>
</tr>
<tr>
<td>age at diagnosis</td>
<td>2.5[1-6]Mo</td>
<td>11[2-16]</td>
<td></td>
</tr>
<tr>
<td>hemolytic disease</td>
<td>4</td>
<td>22</td>
<td></td>
</tr>
<tr>
<td>neonatal cause</td>
<td>15 (31%)</td>
<td>2(5%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Symptoms</td>
<td>24(50%)</td>
<td>28(66%)</td>
<td>0.058</td>
</tr>
<tr>
<td>pancreatitis/angiocholitis</td>
<td>15(31%)</td>
<td>7(16%)</td>
<td>0.011</td>
</tr>
<tr>
<td>Treatment(success/n)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Observation</td>
<td>20/44 (45%)</td>
<td>15/24 (62%)</td>
<td>0.096</td>
</tr>
<tr>
<td>percutaneous BD lavage</td>
<td>15/23 (65%)</td>
<td>not performed</td>
<td></td>
</tr>
<tr>
<td>Cholecystectomy+ BD lavage</td>
<td>1/3</td>
<td>3/4</td>
<td></td>
</tr>
<tr>
<td>ERCP+sphincterotomy</td>
<td>not performed</td>
<td>4/5</td>
<td></td>
</tr>
<tr>
<td>ending with cholecystectomies</td>
<td>12 (25%)</td>
<td>32(76%)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Complications occured in 3 urgent cholecystectomies in infants and 1 cholecystectomy+lavage and 1 sphincterotomy in older children resulting in hepaticojejunostomy

Conclusion: MBDS in infants are more often complicated. Spontaneous resolution occurs in 50% of cases with a median of 3 weeks (max 3 Months). Delayed cholecystectomies may be avoided in infants.
Aims. No unified treatment practices for choledochal malformations (CMs) and pancreaticobiliary maljunction (PBM) exist. We evaluated how CMs and PBMs in children are currently managed in the Nordic countries.

Methods. A questionnaire was sent to pediatric hepatobiliary centers in Sweden, Norway, Denmark, and Finland.

Main results. Eight centers treating median 3.5 cases/year replied. In all units, diagnostic magnetic resonance cholangiopancreatography (MRCP) was routinely performed and CM surgery was confined to 1-2 surgeons. Preferred operation age for antenatally detected CMs was <6 months in four, <12 months in two, and <24 months in one center. While all reported open hepaticojejunostomy as the preferred surgical approach for CMs, management of PBMs without associated proximal dilatation was variable. Definition for a pathological common channel length varied between 4-10 mm. Four centers (50%) favored hepaticojejunostomy and three (38%) would consider ERCP and stenting for symptomatic PBMs without biliary tree dilatation. The preferable approach for asymptomatic PBMs without biliary tree dilatation was either cholecystectomy (n=2), hepaticojejunostomy (n=1), or merely follow-up (n=3) while two centers had no opinion on their management. Follow-up after CM or PBM surgery included abdominal ultrasound in 8 (100%), liver biochemistry in 7 (88%), and MRCP in 4 (50%) centers, respectively. Transition and continued follow-up during adulthood was reported by 5 (63%).

Conclusions. While open hepaticojejunostomy was a uniformly established approach for CMs, definition and management of PBMs without proximal biliary tract dilatation varied widely. Further studies are needed to provide evidence-based guidelines for the optimal management of PBMs in children.
PW5-HB08
OUTCOMES OF KASAI FOR BILIARY ATRESIA BEYOND THE AGE OF THREE MONTHS
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Aim: To assess the outcome of patients who underwent a late Kasai (i.e. after 90 days old) in order to know whether there is a benefit rather than referring the patient straight to liver transplantation (LT).

Methods: We conducted a monocentric retrospective study from 1995 to 2017, looking at post-operative complications, overall and transplant free survival of patients referred for a Kasai after 90 days old and we compared them to patients who did not undergo a Kasai because of were referred very lately or with severe portal hypertension or ascites which contraindicated the Kasai.

Results: We identified 70 out of 424 (16%) Late Kasai performed at a maximum of 198 days, 13 of them were born premature, they were compared to 24 No Kasai. Complications after Kasai included 10 ascites and 3 cholangitis and 3 cardiac issues. Median Follow-up was 109 Months [1-262]. The success rate of Kasai was 17/70 (16%). Overall 5 years survival was 85% in the Kasai group and 83% in the No Kasai group (P=0.7). Transplant free survival was 53% and 28% at 2 and 5 years in the Late Kasai group vs. 12% and 4% in the No Kasai group (P<0.001). Median TFS was 24 months in the Kasai group vs. 12 months in the no Kasai Group.

Conclusion: Despite a low success rate, Kasai performed beyond three months of age has advantages in delaying and sometimes avoiding the need for LT, therefore it helps to overcome technical issues in small recipients.
NEAR-TOTAL PANCREATECTOMY RESULTS FOR PERSISTENT HYPERINSULINEMIC HYPOGLYCEMIA OF NEONATE

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**Aim:** Congenital hyperinsulinism (CHI) is a rare condition in neonates which is mostly resistant to medical therapies. Here we present Near-total pancreatectomy (95% to 98% resection) in a series of CHI as treatment of choice to prevent long-term neurologic sequelae secondary to hypoglycemia-induced brain injury, without causing diabetes mellitus.

**Method:** Neonates suffering from severe hyperinsulinism for whom pediatric surgery consultation was requested during the study period (2006-2017) were included in the study. Patients whose condition was successfully controlled with medical treatment and those suffering from other congenital abnormalities were excluded. Patients were followed-up for 5 years.

**Results:** During the study period 20 neonates (12 girls and 8 boys) with an average weight of 4045 ± 895 grams, were evaluated. Four patients with the appropriate response to medical therapy were excluded from the study. None of the patients suffered from comorbidities. Sixteen patients underwent Near-total pancreatectomy (95% to 98%). Follow-up ranged from 5 months to 8 years. On follow up two patients died from surgical complications, and one required glycemic medication. Diabetes mellitus was not seen in any of 13 patients. None of the patients needed re-operation. Common postoperative complications such as malabsorption in 11 patients treated with Fat-soluble vitamins supplementation.

**Conclusion:** Near-total pancreatectomy is feasible alternative to other treatments of medical refractory hyperinsulinism with less frequent need for re-operation due to inadequate pancreatic tissue removal and minimal diabetes mellitus complication due to total removal tissue. This surgical technique can be considered as the treatment of choice for these patients.
PW5-HB10
MANAGEMENT OF FOCAL NODULAR HYPERPLASIA - SINGLE INSTITUTION EXPERIENCE

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Aim of the Study: Focal nodular hyperplasia (FNH) is a benign lesion that accounts for 2% all pediatric liver tumors. Management strategies for children vary between institutions. Our aim was to describe our experience and assess optimal surgical treatment.

Methods: We reviewed records of all children with FNH managed from 1992 to 2016 at our institution.

Main results: A total 15 patients with FNH were identified, including 9 male and 6 female. The diagnosis of FNH was histologically confirmed in eight (53%) patients, either by open biopsy sample (n=1) or hepatic resection (n=7). Imaging methods were not able to rule out malignancy in six of these patients and two children underwent surgery for mass effect. The median time interval between the diagnosis and surgery was 16.5 days (range 11-165). Remaining seven (47%) patients were diagnosed only by imaging and were observed with regularly imaging study. During follow up period (median 4.5 years, range 1.3-13) lesion regressed in 2 cases and increased in 4 cases in whom two patients underwent resection after 4 and 5.2 years, respectively. Tumor size was slightly larger for patients requiring surgery (8.0±1.5 cm versus 6.2±2.6 cm, p=0.14) without significant difference in the age at diagnosis (p=0.44).

Conclusion: FNH is rare in children. Malignant transformation has not been reported. Surgical treatment is indicated in symptomatic and progressive FNH or in children with diagnostic difficulties.
PW5-HB11
VACUUM DEVICE IN LAPAROSCOPIC TREATMENT OF THE HIDATID CYST OF LIVER IN CHILDREN

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**Aim.** Outcome of laparoscopic treatment of hidatid cyst of liver in children with a vacuum device.

**Methods.** 21 patients with hidatid cyst of the liver, in the period 2013 - 2017. Dominant cyst of the right lobe of the liver (73.3%) in the 6-8 segment. The volume of cysts is from 425 to 2600 ml. All patients underwent laparoscopic liver echinococcectomy. Opening of the fibrous membrane with the help of the ultrasound scalpel "Harmonics" and bipolar coagulation Enseal. After 12 mm of trocar we insert an end-bag to evacuate the chitinous membrane. Under the control of the vacuum device, the vacuum extractor is fed to the immediate site of the cyst removal. With the help of tools, we immerse the chitin sheath in the end-bag. Since the tube is transparent, the control of the material being removed is also provided. After extracting chitin cyst wall was treated with argon plasma coagulation, allowing not only possible to carry out coagulation residual cavity, but also achieve elimination of residual hydatid scoleces.

**Results.** There were no intraoperative complications in our observations. In the postoperative period, bile fistula was noted in the 1st (10%) patient. There were no recurrences of the device disease.

**Conclusions.** Laparoscopic echinococcectomy with device vacuum, and handling an argon-plasma coagulation were effective in children hidatid cyst of liver.
PW5-HB12
THE ROLE OF ANTIBIOTIC PROPHYLAXIS FOR THE PREVENTION OF CHOLANGITIS POST-KASAI PORTOENTEROSTOMY FOR BILIARY ATRESIA

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Aim
To investigate whether routine antibiotic prophylaxis following Kasai portoenterostomy (KP) in infants with biliary atresia (BA) influences the incidence of cholangitis.

Methods
Children were diagnosed with true cholangitis only if presenting all of the following: jaundice, fever, positive blood culture.

Single-center review: Following ethical approval, all consecutive patients who underwent KP for BA at our Institution (2000-2017) were included. All our patients routinely received long-term prophylactic Trimethoprim-sulfamethoxazole.

Systematic review: Using a defined strategy (PubMed, Cochrane, Embase), two independent investigators reviewed all articles reporting the incidence of post-KP cholangitis.

Results
Single-center review: The incidence of true cholangitis was 19% (18/92 patients), with the first episode occurring within the first year after KP in 74% cases. Suspected cholangitis (negative blood cultures) occurred in 23 (25%). Patients who developed cholangitis had similar age and biochemistry (conjugated bilirubin, AST, APRI index, γGT) at KP to patients who never had cholangitis (P=0.2; P=0.1; P=0.6; P=0.9; P=0.3). Jaundice clearance at 3 and 6 months post-KP was more common in patients who developed cholangitis than those who did not (P<0.0001 and P=0.01).

Systematic review: Of 571 abstracts screened, 121 articles were included (7,898 patients). The incidence of post-KP cholangitis was 42% among children who received routine antibiotic prophylaxis and 60% in children who did not (P<0.0001).

Conclusions
Cholangitis is a common complication following Kasai portoenterostomy. Routine antibiotic prophylaxis is an effective strategy to minimize the risk of developing cholangitis. A multicentre randomized controlled study, with stringent criteria to diagnose cholangitis, is necessary to confirm these findings.
PW6-UR01
ANO-SCROTAL DISTANCE (ASD) IS IT A MARKER FOR THE SEVERITY OF CHORDEE?
Ahmed Hadidi, Emir Haxhija
Emma and Sana Offenbach Hospital, Offenbach, Germany

Aim: to evaluate the relationship between the severity of chordee and Ano-Scrotal Distance (ASD)

Methods: boys younger than 2 years undergoing hypospadias repair between January 2015 and December 2017 were included. ASD was measured from the Anal verge to the scrotum. 321 boys with hypospadias and 25 age-matched controls undergoing circumcision (median age 1.17 years, range 0.8-1.9) had ASD measured under anaesthetic. The patients were grouped according to the Hypospadias Grade; Grade I (glanular, n= 86), Grade II (distal, n=135), Grade III (proximal, n= 69) and Grade IV (perineal, n= 31). Chordee was classified into 3 groups; no chordee, superficial chordee corrected by degloving and severe chordee requiring additional procedures for correction.

Results: The median ASD for controls was 4.2 cm (range 3-5.5). The median ASD for Glanular hypospadias was 3.8 cm (range 2-5), for distal was 3.6 (range 2-5) for proximal was 3 cm (range 2-5) and for perineal 2.5 cm (range 1.5-4).

Considering chordee, 10 boys with glanular hypospadias (11%) had severe chordee and ASD was < 3cm, 7 with distal hypospadias (5%) had severe chordee and ASD< 3cm, 24 with proximal hypospadias (35%) had severe chordee and ASD distance < 3 cm and 17 patients with perineal hypospadias (55%) had ASD < 3 cm.

Conclusion: Severe chordee was commonly associated with shorter ASD. Boys with ASD < 3 cm are likely to require additional procedures to correct chordee. This suggests that arrested distal migration of the urethra plays a role in the aetiology of hypospadias and chordee.
PW6-UR02
MENTAL HEALTH STATE OF PERSONS WITH SEXUAL DEVELOPMENT DISORDERS: A REVIEW OF ACTUAL LITERATURE
Marine Bohet¹, Rémi Besson², Renaud Jardri¹, François Medjkane¹

¹Child and psychiatry department, Hôpital M.Fontan, CHRU Lille, University of Lille, Lille, France.
²department of pediatric surgery, hôpital Jeanne de Flandres, CHRU Lille, University of Lille, Lille, France

Aim of the study: Sexual Development Disorders (DSD) are congenital conditions characterised by an atypical development of chromosomal, gonadal or anatomical sex. Since the Chicago consensus, mental health is an active preoccupation of the multidisciplinary teams in charge, which includes psychology and psychiatry members. Therefore, it is crucial to identify the difficulties and needs of this population. This work aims to review actual knowledge about mental health and psychological state of persons with DSD.

Methods: PUBMED and SCIENDIRECT Databases have been interrogated with the algorithms « DSD and psychology » and « DSD and psychiatry ». Studies included were formally about morbid psychological aspects and excluded case reports and qualitative studies. Main results: 18 studies from 2006 to 2016 were included. Ten were about DSD without aetiologic discrimination, four were about Congenital Adrenal Hyperplasia and four were about malformative origins. Heterogeneous methodologies require a qualitative analysis. 16 studies show more psychological distress in the DSD group, characterised by anxiety, depression and interpersonal sensibility. Three out of five show an elevated risk of suicide. No determinant factor could be identified because of inconsistent data. The main limits of the studies were their cross-section method, small sample size and recruitment bias. Conclusions: DSD persons are more likely to suffer from psychological distress. Being aware of this is crucial for clinician in terms of prevention and precocious concerns. In the future, longitudinal and multicentric studies, with standardised measures compared with the control group will give more consistent and precise knowledge.
PW6-UR03
FREQUENCY OF ANTIBIOTIC-ASSOCIATED DIARRHEA AND RELATED COMPLICATIONS IN PEDIATRIC PATIENTS UNDERWENT HYPOSPADIAS REPAIR: A COMPARATIVE STUDY USING PROBIOTICS VS PLACEBO
Ciro Esposito, Agnese Roberti, Francesco Turrà, Mariapina Cerulo, Giovanni Severino, Alessandro Settimi, Fulvia Del Conte, Luca Vaccaro, Serena Izzo, Maria Escolino
Federico II University of Naples, Naples, Italy

Aim of the Study: This study aimed to evaluate the effectiveness of probiotics (Lactobacillus rhamnosus GG), as a preventive measure of antibiotic-associated diarrhea (AAD) in children underwent hypospadias repair and its effects on postoperative outcome, comparing the group treated with probiotics +antibiotics with two control groups (only antibiotics and antibiotics + placebo).

Methods: We performed a prospective, randomized, placebo controlled study with three groups of patients (30 boys for each group) underwent hypospadias repair in our unit from March 2016 to December 2016. G1 received antibiotics +probiotics (Lactobacillus rhamnosus GG), while G2 and G3 respectively received only antibiotics or antibiotics + placebo (glucose solution at 5%) for the same period. We evaluated number of evacuations/day, stool consistency, and number of dressings/day.

Main results: Overall incidence of postoperative AAD was 33.3% (30/90) and it was statistically lower in G1 patients compared to G2 and G3 ones (p=0.002). Duration of AAD was significantly longer in G2 and G3 compared to G1 (p=0.001). In G1, frequency of dressing change was significantly lower compared to G2 and G3 (p=0.001).The incidence of post-operative complications (fistula and dehiscence) was significantly higher in G2 and G3 compared to G1 (p=0.001).

Conclusions: Our study confirmed that the use of probiotic Lactobacillus rhamnosus GG associated with antibiotics significantly reduced the incidence and duration of postoperative AAD. In addition, the use of probiotics LGG reduced the frequency of dressing changes and the incidence of postoperative complications, such as urethral fistula and foreskin dehiscence.
PW6-UR04
LONG-TERM RESULTS IN CONGENITAL ADRENAL HYPERPLASIA
Javier Serradilla, Susana Rivas, María José Martínez-Urrutia, Roberto Lobato, Alba Bueno-Jiménez, Manuel Gómez-Cervantes, Virginia Amesty, Pedro López-Pereira
Hospital Universitario La Paz, Madrid, Spain

Aim
The sex of a newborn with congenital adrenal hyperplasia (CAH) is often initially unclear because of genital ambiguity. Nowadays, its surgical treatment has gone through a phase of controversy because of the physical and psychological consequences for the patient. Our aim is to evaluate the current situation of our patients according to the treatment received.

Methods
Our series of patients treated of CAH between 1990-2010 was analyzed. We assessed their baseline situation (physical examination, genetics, family history), type of surgery performed (section or plication of corpora cavernosa (CC), vaginoplasty, genitoplasty) and the follow-up events (secondary sexual development, medication side effects). The long-term results were analyzed through a questionnaire about their current sexual situation, self-esteem and satisfaction, excluding patients under-age.

Results
Thirty-one patients (21 years, [6-42]) were reviewed. Some had several surgeries (2, [1-7]). Partial section of CC was performed in 80% while their plication was carried out in 19%. Glans reduction was practiced in 36% and vaginoplasty in 83%. Twenty-one (26 years, [18-42]) met the questionnaire inclusion criteria. The response rate was 71% (15). Section or plication of CC did not show differences in satisfaction and sexual function (p>0.05) while glans reduction showed a significant decrease in clitoral sensivity (p<0.05). Although 30% referred some discomfort with their female assignment, 60% were satisfied with their genitalia appearance.

Conclusions
Although most patients were satisfied, there was a considerable percentage who showed consequences in their sexual and social development. This should lead us to a multidisciplinary, more conservative management in this disease.

PW6-UR05: 3MM LAPAROSCOPIC PYELOPLASTY AND NEPHROSTENTS IN CHILDREN UNDER 2 – TOWARDS A TRUE MINIMALLY INVASIVE APPROACH
Mohammad Bader, Abraham Cherian
PW6-UR06
INTRA-OPERATIVE MEASUREMENTS TO PREDICT SUCCESS OF FORESKIN RECONSTRUCTION IN DISTAL HYPOSPADIAS REPAIR: A SINGLE CENTRE PROSPECTIVE SERIES

Alexander Macdonald, Kalpana Patil, Arash Taghizadeh, Massimo Garriboli

Evelina London Children's Hospital, London, United Kingdom

Aims of the study:

Foreskin reconstruction (FR) at distal hypospadias repair is a recognised option but it is not always technically achievable. We sought to determine the relationship between intra-operative anatomical measurements and success of FR.

Methods:

Intra-operative foreskin and penile measurements were prospectively collected in infants undergoing distal hypospadias repair between 2016-2017. Parameters measured included: foreskin length (FL); stretched foreskin length (sFL) and glans width (GW).

Univariate non-parametric analysis and data quoted as median [range].

Main results:

We identified 51 infants, in 41 parents requested FR. Reconstruction was undertaken at 16 [9-119] months (n=4 infants <12 months old at repair; n=18 aged 12-17 months; n=9 aged 18-24 months; n=9 >24 months at repair). Parameters measured: FL= 30mm [16-40]; sFL= 40mm [21-53]; GW= 14mm [12.5-17]. FR was performed in n=39 (95%) and not technically possible in n=2. At a follow-up of 5.5 months (3-15) a fully retractable foreskin was observed in 34/39 (87%). In 3 foreskin was intact but narrow; dehiscence occurred in 1 and foreskin fistula in 1. Median FL:GW and sFL:GW ratio was 2.1 and 2.9 in successful FR and 2 and 2.7 in unsuccessful FR (p=NS). Measured parameters, foreskin to glans ratios and incidence of complications did not significantly change with age.

Conclusions:

Age at reconstruction does not influence anatomy or success of reconstruction. Although statistical significance has not been achieved in this initial small cohort of children, intra-operative measurements as described may represent an objective means to quantify feasibility and outcome of FR.
Aim of the study: Kidney functional capacity correlates with anthropometric data but there are conflicting reports concerning transplant outcomes, in the presence of a high donor/recipient weight mismatch. Furthermore, there are sparse studies in the pediatric population. The aim of this study was to access the relation between the donor/recipient body weight ratio (D/RBWR) and kidney graft outcomes. We hypothesized that lower ratios would associate with poorer graft function.

Methods: We retrospectively analyzed all renal transplanted children in a transplant center, since 2004, all with a minimum follow-up of 6 months. Liver donor transplants were excluded. We calculated the D/RBWR and divided patients into 3 groups: low (<0.9), medium (0.91–1.2) and high (>1.2). Data was collected on graft function, acute and chronic rejection, post-transplant complications and graft survival. We calculated estimated glomerular filtration rate (Schwartz formula) 48h and 6 months after transplantation.

Main Results: Of 50 patients, the median age was 9 years and 52% were males. Median donor weight was 50kg [10-90] and recipient was 19,5kg [13-75], with a median D/RBWR of 1,82. Lower D/RBWR associated with increased incidence in hypertension and oncologic complications. D/RBWR and creatinine at 48h or 6 months had a negative significant correlation. Graft function at 48h and 6 months was significantly lower in the low D/RBWR group. Linear regression showed that the receptor weight was the only independent factor.

Conclusions: We conclude that a low D/RBWR may contribute to inferior renal allograft function, in the pediatric population.
Aim of the study: Post traumatic High flow priapism (PTHFP) is a rare condition in children, usually due to an arterio-cavernous fistula. There is no risk of ischemic lesion because of high blood flow in the cavernosa tissue, unlike low-flow priapism. There is actually no consensus between a conservative treatment and a selective arterial embolization of the fistula. We report 3 cases of PTHFP with initial conservative management.

Method: Since 2012, 3 boys aged of 7, 11 and 16 years old were admitted for PTHFP with painless erection. Two of them had a history of perineal trauma and one presented a HFP after a penile nerve block for a circumcision. Arterio-cavernous fistula was assessed by doppler ultrasound in all cases. Conservative management was decided.

Results: Conservative treatment was successful for the two younger boys. Complete penile detumescence was achieved in 7 and 25 days, respectively. Spontaneous fistula occlusion was confirmed by doppler ultrasound. For the third boy, selective embolization of the fistula was performed 6 months after the initial trauma. Penile detumescence occurred 72 hours later. Normal erections were subsequently noticed in all cases.

Conclusion: Conservative management of PTHFP with a protocolised follow-up is a valid and safe option in case of painless erection. Selective embolization is the alternative of choice in persistent PTHFP, although quite challenging in small children. In our experience delayed embolization does not compromise erectile function.
**PW6-UR09**

**SURGICAL MANAGEMENT OF DISORDERS OF SEX DEVELOPMENT WITH SEVERE HYPOSPADIAS. A SINGLE SURGEON’S EXPERIENCE OF 58 CASES**

Takanori Ochi¹, Asuka Ishiyama¹, Yuta Yazaki¹, Hiroshi Murakami¹, Masahiro Takeda¹, Shogo Seo¹, Ryo Sueyoshi², Geoffrey J. Lane¹, Hidenori Haruna¹, Toshiaki Shimizu¹, Atsuyuki Yamataka¹

¹Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan. ²Department of Pediatrics and Adolescent Medicine, Tokyo, Japan

**Purpose:** To review the surgical treatment of disorders of sex development (DSD) with severe hypospadias (SH).

**Methods:** DSD was diagnosed by gross examination of external genitalia (micropenis, hypospadias, bifid scrotum) and assessment of undescended testis, hormones, and karyotype. DSD+SH (n=58) treated between 1999 and 2017 by primary (n=6) or staged (n=52) urethroplasty (UP) were reviewed for DSD classification, type of hypospadias, sex assignment, hormonal abnormality, surgical strategy, and post-UP complications (post-UPC).

**Results:** DSD were associated with sex chromosome abnormalities (n=4), 46,XY (n=51), 46,XX (n=1), and 47,XY +21 (n=2). (Table) Of these, 2 cases were recognized in older children by karyotyping/gonadal histopathology with subsequent sex reassignment from female to male. Hypospadias was perineal: (n=26), scrotal: (n=16), penoscrotal: (n=15), and midshaft: (n=1). Mean age at final UP was 4.12±0.21 years; all cases had soft tissue interposition at UP. At mean follow-up 5.16±0.56 years after final UP, observed post-UPC (n=8; 13.8%) were urethral stenosis (n=3), urethral diverticulum (n=2), urethrococutaneous fistula (n=2), and curvature (n=1). Mean onset of post-UPC was 1.24±0.77 years (range: 0.1-6.3). All 17/52 staged-UP cases who had ventral penile shaft reinforcement with pedicled fat tissues (PFT) as soft tissue interposition at initial surgery (+PFT), had significantly less post-UPC than (-PFT) cases; (0/17; 0%) versus (8/35; 22.9%) (p=.032) at mean follow-up 4.58±0.49 years after final UP.

**Conclusions:** Although UP for DSD+SH was formidably challenging, PFT was effective for preventing post-UP with satisfactory outcome.

**Table: Case details**

<table>
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<tr>
<th>Number</th>
<th>Sex chromosome DSD (n=4)</th>
<th>Karyotype</th>
<th>Type of UP at last UP</th>
<th>Type of UP at last UP</th>
<th>Period since last UP (years)</th>
<th>Post-UPC</th>
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</thead>
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<td>1</td>
<td>45,XY/46,XY</td>
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<td>8 (0.2)</td>
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<td>4.1</td>
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</tr>
<tr>
<td>2</td>
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<td>-</td>
<td>4.5</td>
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</tr>
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<td>Scrotal</td>
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<td>-</td>
<td>5.0</td>
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<tr>
<td>4</td>
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<td>Perineal</td>
<td>8 (4.0)</td>
<td>-</td>
<td>5.0</td>
<td>None</td>
</tr>
<tr>
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<td>Fatula (0.1)</td>
</tr>
<tr>
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<td>10.3</td>
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</tr>
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<td>-</td>
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<td>-</td>
<td>4.2</td>
<td>Fatula (0.1)</td>
</tr>
</tbody>
</table>

Figure Case 8: 46,XY DS: Testosterone Insufficiency, sex reassignment female to male
A: Initial inspection, B, C: Initial surgery, D, E, F: First urethroplasty after bilateral orchiectomy
PW6-UR10
ENDOUROLOGICAL TREATMENT OF SECONDARY OBSTRUCTIVE MEGAURETER
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Aim of the study: We present our experience and long-term results of endoscopic balloon dilatation of secondary stenosis after vesicoureteral reimplantation.

Methods: Retrospective study of the patients with ureterovesical strictures after reimplantation surgery treated by high pressure balloon dilatation. Clinical data, renal function, surgical records, dilatation technique, postsurgery complications and ultrasonographic and isotopic data (pre and post dilatation) were evaluated.

Main results: Five patients were treated in our department: 4 of them had a Cohen procedure and 1 had a Politano-Leadbetter surgery. In all the patients, ultrasound studies presented progressive ureterohydronephrosis. Diuretic renogram was obstructive with T ½ > 20 minutes that was unilateral in 4 patients and bilateral in the patient with the Politano-Leadbetter procedure. Three patients presented lumbar pain and febrile urinary tract infection. Endoscopic treatment was started performing a retrograde pyelography. High pressure balloon dilatation (2.7 Fr) with a nominal profile of 6-9 mm was used in all cases.

Median operative time was 30 (19-75) minutes and hospital stay was 24 hours in all patients except in the one with bilateral stenosis and febrile urinary tract infection (72 hrs). No intra or postoperative complications occurred. Postoperative renogram and ultrasound showed a significant improvement in elimination curves and dilatation (p<0.05) after a median follow-up of 5 years (2-8).

Conclusions: Endoscopic dilatation of strictures secondary to ureterovesical reimplantation is technically feasible and allows good long-term results. Given its low complication rate, it should be considered as the initial treatment of these patients.
**PW6-UR11**

MATERNAL FIRST TRIMESTER SERUM LEVELS OF FREE BETA HUMAN CHORIONIC GONADOTROPHIN AND HYPOSPADIAS

Matthieu Peycelon$^{1,2,3,4}$, Lea Carlier$^{3,5,6}$, Alienor De Chalus$^{5,6}$, Adeline Bonnard$^{5,6}$, Myriam Rachid$^{5,6}$, Liza Ali$^{1,2,4}$, Muriel Houang$^7$, Annabel Paye-Jaouen$^{1,4}$, Georges Audry$^{5,8}$, Serge Amselem$^{3,5,6}$, Francoise Muller$^{2,9}$, Alaa El Ghoneimi$^{1,2,4}$, Alexandra Benchi$^{10,11}$, Jean-Pierre Siffroi$^{3,5,6}$, Capucine Hyon$^{3,5,6}$

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**Aim.** Although the causes of hypospadias remain often unknown, endocrine, vascular, genetic and environmental factors have been implicated. Human chorionic gonadotrophin (hCG) peaks in the first trimester of pregnancy stimulate foetal testosterone production and normal male genital development. One potential etiological pathway could be an altered release of hCG leading to androgen deficiency. The aim was to identify an association between maternal first trimester levels of serum free-beta hCG (fbhCG) and occurrence of hypospadias.

**Methods.** A cohort of 301 pregnant women was established in 2015 of whom 149 boys had surgery for hypospadias. Serum levels of fbhCG were ascertained from laboratory databases and fbhCG multiple of the median (MoM) were compared between affected and unaffected boys. Statistical analysis: Fisher test and logistic regression.

**Results.** Median fbhCG values and MoM were 25.92 ng/mL (2.7-224) and 1.47 (0.3-7.6) respectively amongst women with an infant with hypospadias and 1.0 in case of unaffected boy. No correlation was found (p>0.05). Stratified by suspected placenta dysfunction (prematurity, intrauterine growth retardation or low birth weight), median fbhCG MoM was 1.36 (0.7-7.6) (p>0.05). However, there was interestingly a significant trend towards high levels of fbhCG for severe types (1.87 (0.5-4.9) in proximal hypospadias (N=47) vs. 1.14 (0.3-7.6) in distal hypospadias (N=102), p<0.05).

**Conclusions.** Our findings do not support the hypothesis that alteration in maternal hCG levels is associated with the development of hypospadias. However, higher fbhCG values were found in children with proximal hypospadias. Because of the small number of patients, further studies are needed.
HISTONE METHYLTRANSFERASE PRDM9 mRNA LEVEL INCREASE IN RESPONSE TO CURATIVE HORMONE TREATMENT OF CRYPTORCHIDISM-DEPENDENT MALE INFERTILITY

Faruk Hadziselimovic1, Katharina Gegenschatz-Schmid1, Gilvydas Verkauskas2, Philippe Demougin3, Vytautas Bilius2, Darius Dasevicius4, Michael B. Stadler5,6

1Cryptorchidism Research Institute, Kindermedizinisches Zentrum Liestal, Liestal, Switzerland. 2Children’s Surgery Centre, Faculty of Medicine, Vilnius University, Vilnius, Lithuania. 3Biozentrum, Life Sciences Training Facility, University of Basel, Basel, Switzerland. 4Institute for Pathology, National Centre of Pathology, Affiliate of Vilnius University Hospital Santaros Klinikos, Vilnius, Lithuania. 5Friedrich Miescher Institute for Biomedical Research, Basel, Switzerland. 6Swiss Institute of Bioinformatics, Basel, Switzerland

Aim of the study: To investigate the role of positive regulatory domain containing (PRDM) family genes in the cryptorchidism induced infertility. Results of abrogated mini-puberty are impaired translation of germ cells into Ad (dark) spermatogonia, increased germ cells loss, transposon deregulation and infertility development. Here, the dataset is interpreted with a focus on members of the positive regulatory domain containing (PRDM) family, some of which encode histone methyltransferases that are important for reproduction and transposons silencing.

Methods: During orchidopexy for bilateral cryptorchidism as a part of randomized prospective study, biopsies are performed to confirm the presence of type Ad spermatogonia using semi-thin section method. Part of the biopsy was utilized for RNAseq analysis in 7 low infertility risk (LIR) and in 8 high infertility risk (HIR) boys, median age of 18.5 (8–59) months. An absence of Ad spermatogonia identifies HIR patients, who are treated with a gonadotropin-releasing hormone agonist having second testis biopsied after 6 month of GnRH treatment (4 boys). This study was approved by the Vilnius Regional Biomedical Research Ethics Committee.

Main results: We found that three genes that are essential for male germ cell development, namely PRDM1, PRDM9, and PRDM14, were down-regulated in testes from HIR patients and that PRDM9 (central regulator of crossover distribution) was significantly induced after GnRHa treatment.

Conclusions: Conserved chromatin modification enzymes are involved in cryptorchidism and in its treatment with curative GnRH therapy. We provide additional observation consistent with the idea that infertility in cryptorchidism is a consequence of transposon deregulation.
COMBINING SIGMOID RESECTION, ANTEGRADE ENEMA AND AUGMENTATION CYSTOPLASTY: A COLLABORATIVE APPROACH FOR BLADDER AND BOWEL MANAGEMENT

Aim of the study

Patients with neurogenic bladder in need of urologic reconstruction often have associated bowel dysfunction. The augmentation cystoplasty plan should take the patient’s bowel status into account and those patients with fecal incontinence in need of an augmentation cystoplasty may benefit from sigmoid resection with or without antegrade enema at the time of the urologic reconstruction. The resected sigmoid then becomes a readily available option for augmentation cystoplasty obviating the need for a second bowel anastomosis.

Methods

We reviewed charts (2014-2017) of patients who underwent combined sigmoid resection and augmentation cystoplasty utilizing the sigmoid segment. Enema volume, enema duration, and stool continence before and after surgery was recorded.

Main Results

13 patients underwent sigmoid resection with sigmoid-cystoplasty, Mitrofanoff, and Malone (6 neomalone, 2 split Malone). Median follow up was 18 months [2-36]. Nine patients had anorectal malformation and 4 myelomeningocele. Preoperatively 10 patients were on rectal enemas, 3 were on laxatives, and 6 of 13 (43%) were continent of stool. After surgery 12/13 (92%) were continent of stool (p<0.05). There was no difference in the enema volume (p=0.6) or in the duration of the enema before and after resection (p=0.053).

Conclusion

Patients with fecal incontinence and constipation who are in need of a bladder augment can benefit from a combined sigmoid resection, antegrade enema (if needed) and cystoplasty using sigmoid colon. This collaborative approach can both increase stool continence and obviate the need for a second bowel anastomosis when obtaining the segment for augmentation cystoplasty.
UNDESCENDED TESTIS TORSION: A MULTICENTER REVIEW

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**Aim of the Study** Torsion of an undescended testis (UT) is a surgical emergency, difficult to diagnose, whose prognosis depends on a quick management. Only case reports or monocentric reviews have been reported in the literature so far. We led a multicenter retrospective study to evaluate the management and outcome of these patients.

**Methods** We reviewed and analyzed all cases of UT torsion operated on in eight French hospitals between 1997 and 2017. We divided patients in two groups: patients referred less than 6 hours after the onset of symptoms (group A) or more than six hours (group B).

**Main results** We collected 57 cases (17 in group A and 40 in group B). Median age was 4.6 years [1 month-18 years] (4y in group A and 4.8y in group B). Symptoms were pain (n=55, 96%), inguinal mass (n=53, 93%) and vomiting (n=15, 26%). 8/9 patients with neurological disorders are from group B. An ultrasound scan performed in 24 patients led to the diagnosis in 13 patients (54%). At surgery, an orchidectomy was performed in 4 patients (24%) of group A and 22 patients (55%) of group B. At a mean follow-up of 2 years, 11 patients in group A (65%) and 6 patients in group B (15%) had a normal testis.

**Conclusions** UT torsion can happen at any age. Early diagnosis in front of a painful inguinal mass with an empty scrotum is essential to improve the salvage rate of UT torsion.
IDIOPATHIC DETRUSOR UNDERACTIVITY IN CHILDREN: A CHALLENGING DIAGNOSIS

Mohammad Bader, Massimo Garriboli, Anne Wright, Jo Clothier

Guy's and St Thomas NHS Foundation Trust, Evelina London Children's Hospital, London, United Kingdom

Aim:

Underactive bladder is poorly reported in the paediatric population. Urodynamically, underactive detrusor (DU) is defined as contraction of reduced strength and/or duration resulting in prolonged or incomplete emptying.

Clinically, it is characterised by abdominal straining and interrupted uroflowmetry. We postulate that these characteristics are unreliable in predicting detrusor underactivity.

Methods:

Children were identified from our prospectively collected database of 1575 video urodynamic studies (VUDS) (January 2010 - September 2017). Children with anatomical outflow obstruction, neurological lesions or inhibited voiding during urodynamics were excluded. We review urinary tract symptoms, status, flow patterns and urodynamics in children with diagnosed detrusor underactivity. Values displayed as percentage and range.

Main Results:

Out of 58 children, 45 (78%) were females. Median age at presentation was 10 years (5.2-15.4). 78% of children had day and/or nighttime incontinence, 53% voided with abdominal straining and 65% had recurrent UTIs.

Uroflows were staccato in 31%, fractionated in 14% and tower shaped in 19%. Incomplete emptying was seen in 22%. Renal scarring present in 21% on DMSA.

VUD studies showed average bladder capacity of 125% of EBC (55-255%). Post-void residual 49% of total bladder capacity (0-100%). 26% showed detrusor overactivity and 33% had reduced bladder sensation. 95% had unsustained bladder contractions (one patient had insufficient contraction p det 29). Vesicoureteric reflux present in 19%. Dysfunctional voiding was seen in 29%.

Conclusions:

Abdominal straining and uroflow patterns do not reliably characterise the underactive bladder in childhood.

Significant co-existent renal and bladder abnormalities are frequent.

Invasive urodynamics is recommended.
ROBOTIC VERSUS LAPAROSCOPIC ASSISTED SURGERY IN PEDIATRIC GYNECOLOGY: IS OPERATIVE TIME A PROBLEM?

Fabrizio Vatta¹, Piero Romano¹, Noemi Pasqua¹, Claudia Filisetti¹, Ilaria Brambilla², Gloria Pelizzo³, Luigi Avolio³

¹Fondazione IRCCS Policlinico San Matteo, Pediatric Surgery Dpt, Pavia, Italy. ²Fondazione IRCCS Policlinico San Matteo, Pediatric Dpt, Pavia, Italy. ³Ospedale Di Cristina, Pediatric Surgery Dpt, Palermo, Italy

Aim of the Study: Since its introduction, robotic surgery (RS) in Pediatrics struggled to thrive due to high costs, difficulties on small children and allegedly longer operative time. Aim is to compare RS and laparoscopic surgery (LS) in pediatric gynecology, particularly in terms of surgical time.

Methods: 30 patients (15 LS, 15 RS) operated upon from 2010 to 2017 for ovarian and/or tubal lesions were reviewed. To randomly compare the two populations, adnexal torsions, urgent LS procedures and neonates were excluded. Patients’ and pathologies’ characteristics, operative time, conversion and complication rate, length of hospitalization were statistically analyzed.

Main results: RS and LS group were comparable in terms of age [11.8±4.2 vs 12.0 (8.4-13.7), p value 0.36] and weight (Kg 43.8±15.3 vs 43.3±24.8, p value 0.53). Operative time was 136.1±55.7’ for RS and 110±29.5’ for LS, length of hospitalization 2 days (2-2) for RS and 2 (1.5-2.5) for LS. Neither complications nor conversions were recorded. No statistically significant differences were found by comparing the two populations (Table 1). Data on involved pathologies and type of procedure are summarized in Table 2.

Conclusions: Our study shows how RS is a valid alternative to LS in pediatric gynecology, providing acknowledged technical advantages with no differences in terms of mean operative time as well as patients’ outcome, conversion rate and length of hospitalization. Nevertheless, known disadvantages are high costs, lack of availability in emergency and difficult use in small children.

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Significant p-value <0.05
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Aim: To evaluate the children with late-presented posterior urethral valves (PUVs).

Methods: Between January 2003 and December 2017 files of patients who were diagnosed with late-presented PUVs were analyzed retrospectively.

Results: Seventeen boys were diagnosed with late-presented PUVs (mean age: 7.35 years). The most common symptoms at presentation was lower urinary tract symptoms (LUTS) including: urinary frequency (n:10), day and night time incontinence (n:8), hesitancy (n:2) and straining (n:2). Urinary ultrasound examination revealed upper urinary tract impairment (n:9), bladder wall thickening (n:4), bladder diverticula (n:2), postvoid residual urine (n:4). VCUG showed PUV in 10 patients. Six patients had normally appearing urethra on VCUG. Urethra couldn’t be evaluated in one patient who didn’t able to pee. Reflux was detected in 14 of 34 renal units. Urodynamic studies were available in 10 patients and revealed detrusor overactivity (n:5), significant post-void volume (n:3), decreased capacity (n:4). All patients had PUVs and Type 1 PUV was the most common valve form (n:13), followed by type 3 PUV (n:4). The mean follow-up period was 52.8 months. LUTS have been resolved in 11 patients. Reflux spontaneously improved in 9 renal units.

Conclusion: Late-presented PUVs may be missed on VCUG. Cystoscopic examination should be preferred if a boy has repetitive daytime and urge incontinence, recurrent UTI and persistent VUR regardless of VCUG results. Prognosis of these patients is variable, and reports on the long-term follow-up after valve ablation of late-presented PUVs are insufficient.
EVALUATION OF PEDIATRIC VCUG AT A CHILDREN’S HOSPITAL: INDICATIONS, COMPLICATIONS, FINDINGS

Doğuş Güney¹, Can İhsan Öztorun², Müjdem Nur Azılı¹, Bilge Karabulut¹, Hüseyin Tuğrul Tiryaki³

¹University of Health Sciences Ankara Children’s Hematology Oncology Training and Research Hospital, Ankara, Turkey. ²Yıldırım Beyazıt University, Ankara, Turkey

Aim of study

We aim to determine any data which supports 'normal or pathologic' voiding cystourethography (VCUG) examination, to reduce the number of unnecessary VCUG and VCUG related complications.

Methods:

We reviewed 670 VCUG examinations. Age, gender, urinary ultrasound finding, indication for VCUG, VUR grade, complication after VCUG were recorded. Patients were compared in terms of complications and findings of imaging studies.

Main results:

VCUG (65.2% female; mean age, 6.24±4.24 y) examinations were requested by pediatricians (54.6%) and surgeons (45.4%). The number of normal VCUG was 315. The rate of severe reflux in the right kidney was 19.5% and in the left kidney was 17.2%. Post-imaging complication rate was 8.5%. Complication rate in female patients was 10.5% while 4.7% in males (p <0.05). The risk of complication in patients with hydronephrosis, and thinned renal parenchyma was high (19.3% - 6.9%, 13.9% - 7.1%, p<0.05). Complication rate was high in patients with ureter dilatation (right 17.6% - 7.4%, p<0.01, left 14.8% - 7.7% p<0.05). The risk of VUR was 47.7% in patients with normal urinary system, 57% in patients with hydronephrosis examined by ultrasonography (p<0.001). The reflux grade was higher in patients with hydronephrosis in the right kidney (17.5% - 6.3%), those with renal parenchyma-thinned (13.4% - 8.7%) and those with ureter dilatation (22% - 8.6%), (p <0.001, p <0.01, p <0.05).

Conclusion:

Risk of complication was higher in girls, patients with hydronephrosis, parenchymal thinning, and ureter dilatation. Although VUR rate was not low in the patients with normal US, VCUG and US findings were correlated.
PW7-UR07
MEATOTOMY AND MUCOSAL GRAFT FOR THE TREATMENT OF MEATAL STENOSIS AFTER HIPOSPADIAS SURGERY

Javier Rojas-Ticona¹, Gerardo Zambudio Carmona², Maria José Guirao Piñera², Vanesa Villamil², Angela Sanchez-Sanchez³, Maria Cristina Gimenez-Aleixandre², Kevin Carlos Mansilla-Eguia³, Juan Pedro Hernandez Bermejo²

¹University Hospital Complex of Albacete, Albacete, Spain. ²“Virgen de la Arrixaca” Clinical University Hospital, Murcia, Spain. ³National University of Saint Augustine, Arequipa, Peru

Aim of the Study: To present our experience and results of surgical treatment in patients with meatal stenosis after hypospadias surgery using meatotomy and mucosal graft.

Methods: Descriptive and observational study. We selected from our registry of 895 patients operated by hypospadias those who presented meatal stenosis and were treated surgically by dorsal meatotomies and grafts of the lingual or preputial mucosa. We performed descriptive statistical analysis and comparisons between previous and post-intervention uroflowmetry values (SPSSv22).

Main results: 26 patients with meatal stenosis were operated using meatotomy with mucosal graft. The predominant clinic was the presence of a fine-caliber stream in nineteen patients (73.1%). The stenosis was objectified calibrating a meatus inferior to 8Fr in 15 patients (57.7%) and by altered uroflowmetry. The meatotomy was performed at a mean age of 7 years (3-13 years). In 14 patients, we used lingual mucosa and in 12 preputial mucosae. The mean pre-intervention peak flow was 4.3ml/s with a significant improvement at 15.2ml/s after the intervention (p = 0.0002). Three patients with lingual graft had a history of failed preputial graft. There was no complication of the donor area in cases of the lingual mucosa.

Conclusions: Dorsal meatotomy with mucosal graft is a suitable surgical option in the treatment of meatal stenosis in patients operated of hypospadias. In our experience the use of lingual mucosa grafts is a useful technique with a low risk of complications and well tolerated.
POSTER WALK 7 UROLOGY Thursday June 21st

PW7-UR08
POSTOPERATIVE ASSESSMENT OF HYPOSPADIAS REPAIR IN CHILDREN WITH UROFLOWMETRY AND OBJECTIVE SCORING: OUR CENTER EXPERIENCE

Leily Mohajerzadeh, Ahmad Khaleghnejad Tabari, Mohsen Rouzrokh, Hasan Tabiee, Khashyar Atqiaee
Pediatric Surgery Research Center (PSRC), Research Institute for Children Health (RICH), Mofid Children’s Hospital (MCH), Shahid Beheshti University of Medical Sciences (SBMU), Tehran, Iran, Islamic Republic of

Purpose:
Functional outcome of hypospadias repair is more significant than cosmetic outcome. Aim of this study was to evaluate the functional outcome of hypospadias after distal shaft hypospadias surgery over a long-term follow-up.

Patients and method: Study designed Files from patients who underwent primary hypospadias repair at our institution between 2006 and 2011 were reviewed. Over a period of 6 years, 572 primary hypospadias patients were treated in our center. Then we called and requested them for visit and uroflowmetry in children more than 5 years old. However only 78 children and their parents agreed to participate in this study. HOSE (Hypospadias Objective Scoring Evaluation) questionnaire and uroflowmetry were obtained to evaluate long-term outcome of hypospadias repair.

Results: The age at time of assessment ranged from 5 to 11 year-old, with mean follow up of 60 months. In MAGPI group 100% had acceptable HOSE. In TIP group 82% had acceptable score and in Onlay group 90% had acceptable score. Uroflow rates of MAGPI group: 5 (12%) subjects were below the fifth centile, in 11 (31%) patients equivocal (between 5th and 25th centile) and 23 (58%) patients above 25th centile. Uroflow rates of TIP group: 11 (32%) subjects were below fifth centile, 17 (50%) patients were equivocal (between 5th and 25th centile) and 6 (18%) patients were above 25th centile. Uroflow rates of Onlay flap group: one patient (20%), equivocal (between 5th and 25th centile) and 4 (80%) patients above 25th centile.

Conclusion: HOSE and uroflowmetry are non-invasive and simple tools to assess long-term outcomes of children after hypospadias repair. Because studies reported remarkable improvement at puberty so watchful waiting with following the objective parameter by examination and uroflowmetry is proposed to avoid unnecessary intervention.

Key words: Hypospadias, objective assessment of hypospadias repair, uroflowmetry
THE PROBLEMS OF MANAGEMENT OF DISORDERS OF SEX DEVELOPMENT IN DEVELOPING COUNTRY

Manuela EHUA, Martial Olivier Moulot, Kouame Agbara, Roumanatou BANKOLE
Treichville teaching hospital, Abidjan, Côte d'Ivoire

Aim of the Study: Identify the limits of the management of abnormalities of sexual development in our context.

Methods: The study was retrospective from January 2007 to December 2017 and patient record was done in the pediatric surgery department. Epidemiological, clinical, paraclinical, therapeutic and evolutionary settings have been studied.

Main results: We collected 12 patients whose average age was 5.33 years. The distribution of rearing sex was as follows: 6 females (50%), 4 males (33.3%) and 3 indeterminates (25%). Sexual dimorphic were the reason for consultation of all patients. Hormonal analysis were incomplete in all patients. Testosterone (50%), progesterone (25%) have been dosed. Karyotype was performed in 75% of cases. 50% of patients had a masculine pseudo-hermaphroditism. Female pseudo-hermaphroditism, congenital adrenal hyperplasia and gonadic dysgenesis were the diagnosis in 8.33% each. Diagnostic and therapeutic management started in all patients. They consisted of feminizing genitoplasty in six patients (50%), two masculinizing genital surgeries (16.66%), topical administration of androgen in 33.33%. 9 out of 12 patients are now lost to follow-up.

Conclusion: The socio-economic difficulties of the populations, the impossibility of access to the basic diagnostic and paraclinical methods coupled with the pejorative cultural representations of the pathology constitute the limits for the management of abnormalities of the sexual development in our practice.

Key words: Disorders of sex development - Pediatric
Aim of the study: Post-operative routine control after endoscopic PUV ablation is currently done either by Micturating Cysto-Urethro-Gram (MCUG) or cystoscopy or both. We investigated the impact of routine postoperative MCUG on the management of PUV.

Methods: Neonates, under one month, managed for endoscopic PUV ablation from January 2012 to January 2017 were included. Patients managed by diversion or managed at later age were excluded. All children had MCUG, ultrasound and metabolic assessment before PUV ablation and at 4 weeks postoperative. All had PUV ablation by endoscopic cold knife incision at 5, 7, and 12 O’clock. We have reviewed the outcome of each child, MCUG, upper tract, and kidney function. Confirmed febrile urinary tract infections (UTI) were analyzed. Reoperation type and indications were reviewed.

Main results: Thirty-one patients met the inclusion criteria. Median weight was 3380gr (2320-4440). Median follow-up was 49 months (12-56). Nine children (29%) had indications for second surgery mainly febrile UTI: 4 at one month (2 circumcisions, 1 missed associated anterior urethral valves, 1 vesicostomy) and 5 children at 6 months (4 circumcisions, 1 ureterostomy). Routine post-operative MCUG showed residual posterior urethral dilatation in 23/31 (74%) There was no correlation between residual dilatation and indication for second look or another surgery (p = 0.005).

Conclusions: None of our patients had second look cystoscopy or change of management based on MCUG results. Post-operative routine MCUG after neonatal endoscopic PUV ablation may be reconsidered. MCUG may be indicated in selected group of children based on strict follow-up criteria.
HOW TO DECREASE OPERATIVE TIME WITHOUT ALTERING THE RESULTS OF LAPAROSCOPIC PYELOPLASTY

Eric Dobremez¹, Carmen Trabanino Garcia¹, Francois Semjen², Luke Harper¹

¹Pediatric Surgery Unit, Bordeaux, France. ²Anaesthesiology, Bordeaux, France

Introduction

The results of transperitoneal laparoscopic pyeloplasty for pelvi-ureteral junction obstruction (PUJO) are good. Operative time can be long and is due to the complexity of the anastomosis and performing knots laparoscopically. We aimed to evaluate if knotless sutures could decrease operative time without affecting results.

Patients and Methods

Within the past 12 months we operated four children using the Stratafix Spiral suture (Stratafix Spiral PDO, Absorbable, 7cm x 7cm, 4-0, Ethicon). The patients were 6, 9 and 14 (2) years-old. They all presented PUJO without polar vessels. We performed a transperitoneal laparoscopic pyeloplasty with pelvi-ureteric anastomosis using a double running suture. A transanastomotic double J stent was inserted and left in place for 6 weeks; a postoperative ultrasound was performed at 6 months.

Results

In this video presentation we show the technique. The average operative time was 96.4 min. We compared it to the average operative time of the 5 previous PUJO operated using classic sutures, including knots which was 117.6 min, meaning the knotless sutures allowed for a 20% decrease in operative time. Both groups had identical postoperative courses and results.

Conclusion:

These preliminary results suggest using knotless sutures reduces operative times without altering results.
PW7-UR12
BILATERAL TESTICULAR VOLUMEN AFFECTATION IN ADOLESCENTS WITH LEFT VARICOCELE

Sara Hernandez-Martin¹, Lidia Ayuso¹, Pedro Lopez-Pereira², Ada Y. Molina¹, Alberto Perez-Martinez¹, Carlos Bardaji¹

¹Complejo Hospitalario De Navarra, Pamplona, Spain. ²Hospital Universitario La Paz, Madrid, Spain

Aim: To determine if bilateral testicular volume is diminished in adolescent patients with left varicocele.

Methods: Retrospective case-control study. Charts of patients with left varicocele followed in Pediatric Surgery consultation in the last 15 years were reviewed. Right or bilateral varicocele cases were excluded. Patients in which bilateral testicular volume was measured by US prior to treatment were selected as cases. Controls included patients in the same range of age, examined clinically and sonographically during that period for anomalies without theoretical involvement of the testicular volume.

The size of both testicles was compared between the groups. Differences among left (LTV) and right testicular volume (RTV) were investigated in each group. Finally, the analysis was performed according to varicocele grade.

Results: 206 cases and 94 controls were found. Mean age and range were similar in both groups (p=0.06), 13±1.4 years (7.6-16.1) in cases and 12.7±1.9 years (7.8-15.8) in controls. Cases showed lower bilateral testicular volume than aged-matched controls, with an absolute difference of 1.57 mL (p=0.007) for LTV and 1.18 mL (p=0.046) for RTV. LTV was lower than RTV in both cases (-0.67±1.6 mL) and controls (-0.25±1.7 mL), although no statistically significant differences were found in controls. Results were consistent in every age group. Patients with grade III varicocele showed the smallest bilateral volume.

Conclusions: Both testis can be affected in patients with left varicocele regarding its volume. Comparing LTV with RTV might result insufficient to determine if there are disease induced changes.
MEDIAN SACRAL ARTERY EMBRYOLOGY AND ANATOMY: MICRO-CT FETAL VASCULARIZATION STUDY AND LITERATURE REVIEW

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¹Hôpital Clocheville, Tours, France. ²EA2415, Aide à la décision médicale personnalisée, Université de Montpellier, Montpellier, France. ³Laboratoire d’anatomie de Montpellier, UFR médecine, Université Montpellier, Montpellier, France

Aim of the Study
The median sacral artery (MSA) is the termination of the dorsal aorta which undergoes a sequence of regression and remodeling during embryo and fetal development. The MSA takes part in the pelvic vascularization and may be injured during pelvic surgery. The embryological steps of its development, anastomosis and the anatomical variations are linked but not fully understood.

Methods
The pelvic vascularization and more precisely the MSA of a 22 gestational weeks (GW) fetus were studied using micro-CT imaging. Arterial segmentations, 3D visualization and a literature review concerning MSA anatomy and embryology was performed.

Main results
At 6GW the MSA distributes segmental arteries in front of the sacrum and coccyx, forming a plexus with ganglia and nerves. Then it bends with the acute anterior flexure of the vertebrae and the regression of the second neural tube. Between 10 and 18GW it becomes very tortuous at the lower sacrum and coccygeal levels, accompanying veins, nerves and sympathetic ganglions to form a complex with no evidence of arterio-venous anastomosis.

In our specimen the MSA is a well-developed strait artery in front of the sacrum longer than the abdominal aorta. Anastomosis between the MSA and internal pudendal arteries and between the MSA and the superior rectal artery were detected.

Conclusions
The analysis of embryo and fetal microscopic anatomy, dissections and 3D visualization help us understand how the MSA has a central role in pelvic vascularization through ilioaortic anastomotic system.
Fig 1. Arterial vasculature of our specimen.

Fig 2. Anastomotic network.
OPTIMIZATION OF 3D PHENOTYPING OF PULMONARY VASCULATURE IN RAT FETUS

Emrah Aydin, Brittany Levy, Sam Nachabe, Marc Oria Alonso, Foong-Yen Lim, Jose Luis Peiro
Cincinnati Children’s Hospital Medical Center, Cincinnati, USA

Aim of the study

Comparative, functional, and developmental studies of animal morphology require accurate visualization of three-dimensional structures, but few widely applicable methods exist for non-destructive whole-volume imaging of animal tissues. The purpose of this study was to develop a method for quantitative analysis of the total fetal pulmonary vasculature using a technique that could be translated into the human studies.

Methods

Rats were harvested at E21. Embryos were fixed overnight in 4%. They were treated with 25% Lugol solution for 72 hours for perfusion. The µCT scan was performed, and pulmonary vasculature was segmented. Vessels were analyzed per diameters, length and branch generation.

Main results

Partially peeled skin technique was the only method that proved the microarchitecture, the side branches of the high generation vessels and 3D interconnectivity of the whole lung prenatally. The 3D images gathered by µCT and processed by the software showed pulmonary vasculature distributed through the lung on E21 of a rat fetus which was representative of the shape and structure of the lungs. The mean number of vessels segmented in the pulmonary tree was 900 with a mean diameter of 134.13µ (range 40.72-265.69µ). While up to 30th generation of the vessels were able to be segmented both in arteries and veins, the majority of the branches were between 11th and 20th generations.

Conclusion

Passive diffusion of the contrast material enables quantitative analysis of the fetal pulmonary vasculature. This technique is a useful tool to analyze the effects of congenital lung diseases on pulmonary vasculature.
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<td><strong>Diameter of the vessel</strong></td>
<td>122.97 ± 46.07</td>
<td>145.78 ± 60.18</td>
<td>131.14 ± 45.46</td>
<td>137.68 ± 57.72</td>
</tr>
<tr>
<td><strong>Circumference of the vessel</strong></td>
<td>386.32 ± 144.75</td>
<td>457.98 ± 189.05</td>
<td>411.97 ± 142.82</td>
<td>432.53 ± 181.34</td>
</tr>
<tr>
<td><strong>L/D ratio</strong></td>
<td>0.20 ± 0.15</td>
<td>0.18 ± 0.14</td>
<td>0.18 ± 0.14</td>
<td>0.21 ± 0.16</td>
</tr>
</tbody>
</table>

Table 1: The morphometric results per vessel type. (RPA: Right pulmonary artery, RPV: Right pulmonary vein, LPA: Left pulmonary artery, LPV: Left pulmonary vein, L/D ratio: Vessel length to diameter ratio.)

Figure 1: Three-dimensional images of the entire lung by μCT scan after in-vivo lugol application (Left), Skeletonization of the entire lung (Right above), Skeleton of one group of vessels with mapping (Right below).
FETAL TRACHEAL OCCLUSION IN MICE: A NOVEL TRANSUTERINE METHOD

Emrah Aydin, Rashika Joshi, Marc Oria Alonso, Brian Varisco, Foong-Yen Lim, Jose Luis Peiro

Cincinnati Children’s Hospital Medical Center, Cincinnati, USA

Aim of the study

Fetal tracheal occlusion (TO) is an emerging surgical therapy in congenital diaphragmatic hernia (CDH) that improves the fetal lung growth. Different animal models of CDH and TO present advantages and disadvantages regarding ethical issues, cost, surgical difficulty, size, survival rates and available genetic tools. We developed a minimally invasive murine transuterine TO model, which will be useful in defining how TO impacts lung molecular biology, cellular processes, and overall lung physiology.

Methods

Time-mated C57BL/6 mice underwent laparotomy at E16.5 with transuterine TO performed on 2 fetuses in each uterine horn. At E18.5, fetuses harvested. The lungs of the TO fetuses were compared with the non-manipulated counterparts by morphometric and histologic analysis.

Main results

Sixteen of twenty TO fetuses survived to E18.5 of which successful TO was confirmed in 12 (75%). Fetal weights were comparable, but lung weights were significantly greater in TO (28.41±5.87 vs. 23.38±3.09, p=0.043). Normalized lung-to-body weight ratio was also greater (1.22±0.10 vs. 1.02±0.13, p=0.006). E18.5 TO lungs demonstrated dilated central and distal airspaces with cellularity. Protein/DNA ratio was decreased 5.4-fold (p=0.005) in TO vs. control while total lung DNA/fetus weight ratio was increased 3.3-fold (p=0.048).

Conclusions

Mice fetal transuterine TO is feasible with comparable outcomes to other current animal models. The increase in lung weight, lung-to-body weight ratio, and DNA/protein ratio indicate organized lung growth rather than edema or cell hypertrophy, and therefore an effective fetal TO.

Figure 1: Schematic representation of the transuterine suture through the neck, respecting one side vascular (carotid and jugular) elements and surrounding trachea. (C: Carotid artery, J: Jugular vein, T: Trachea, E: Esophagus, V: Vertebra)
PW8-BS04
REDUCTION OF ADHESION FORMATION AND DERMAL SCAR FORMATION IN MICE
Michael Boettcher, Carolin Götz, Stefan Mietzsch, Michaela Klinke, Tobias Fuchs, Konrad Reinshagen
UKE Medical School, Hamburg, Germany

Introduction: Peritoneal adhesions are a common complication of surgical interventions of the abdomen, affecting between 60-100% of patients with the pathomechanism evolving from peritoneal healing, a complex process involving i.e. hemostasis and inflammation. As extracellular traps, including neutrophil extracellular traps (NETS), contribute to the pathogenesis of various diseases and affect both hemostasis and inflammation, the aim of the study was to evaluate the effects of DNase1 on peritoneal adhesion and dermal scaring formation.

Methods: In a total of 117 mice adhesions were induced by coagulation and 2 6x0 Vicryl sutures. The following treatment groups were assessed: sham, control, DNase1 single, DNase1 multi. Additionally, a PAD4 knockout model (resulting in the inability to form NETS) was used. Defined endpoints were: (1) adhesion formation (Leach, Near score), (2) scar formation, (3) inflammation (NE, MPO, TLR4, H3cit), and (4) wound healing (collagen, SMA, fibrin).

Results: DNase1 significantly reduces adhesion formation and scarring in various clinical settings, i.e. anastomosis, deserosation and heat exposure. DNase1 did not cause adverse effects and did not negatively affect collagen or SMA formation. Moreover, DNase1 improved dermal wound healing.

Discussion: We were able to show that peritoneal formation is associated with fibrin production and NETs formation. Extracellular DNA induces inflammation, neutrophil activation, and NETosis, ultimately leading to adhesion formation. As such, DNase 1 treatment reduces fibrin production, inflammation, and neutrophil recruitment leading to a significant reduction and even prevention of peritoneal adhesions in mice.
Aim of the study
We have recently developed a three-dimensional bioreactor-based culture system to produce tissue engineered oesophagi (TEO). Previous attempts at orthotopic implantations in rats failed due to intrinsic weakness of the model. To evaluate our construct in vivo we have established two different xenogenic implantation murine models.

Methods
Oesophageal constructs based on a bio-reactor culture of human mesoangioblasts and murine fibroblasts in decellularized rat oesophagi were prepared for transplantation. Model 1: TEO were wrapped in the omentum of 4 NOD-SCID mice; at 2 weeks they were harvested and seeded with rat oesophageal epithelia and cultured in vitro for 4 days. Model 2: TEO underwent a further seeding with epithelial progenitors from rat oesophagi prior to implantation as a patch repair to fill a defect in SCID mice for 4 weeks. For both the models, histology and immunofluorescence analyses were performed to define the microstructure.

Main Result
Model 1: the engineered oesophageal muscle components retained a smooth muscle differentiation of human cells and preserved their native orientation after 2 weeks. Epithelial cells seeded over the implanted oesophagus survived and differentiated after seeding ex vivo. Model 2: all the animals survived without complication. TEO integrated in the transplanted gut.

Conclusion
Using model 1 we underlined the feasibility of a 2-stage approach where pre-vascularization is achieved with an omental wrap. Model 2 demonstrated the capability of this scaffold to regenerate the forestomach wall in mice, the latter being histologically comparable to the oesophagus.
PW8-BS07

INTESTINAL INJURY IS PREVENTED BY AMNIOTIC FLUID STEM CELL ADMINISTRATION BEFORE THE ONSET OF NECROTIZING ENTEROCOLITIS

Marissa Cadete, Bo Li, Carol Lee, Hiromu Miyake, Joshua O’Connell, Agostino Pierro
The Hospital for Sick Children, Toronto, Canada

Aim
Amniotic fluid stem (AFS) cells can attenuate necrotizing enterocolitis (NEC) when administered at the onset of NEC, but it is not clear if this effect is preventative or therapeutic. The aim of this study is to examine the effect of AFS cells on the prevention of intestinal injury during experimental NEC.

Methods
Following ethical approval (AUP32238), NEC was induced in 5-day old C57BL/6 mice by gavage feeding of hyperosmolar formula, hypoxia, and oral lipopolysaccharide (4mg/kg) for 4 days. On postnatal days 4 and 5 prior to NEC induction, mice received intraperitoneal injections of phosphate buffered saline (PBS; n=6) or 2x10^6 AFS cells (n=6). Breastfed pups served as control (n=6). At P9, pups were sacrificed and distal ileum was harvested. NEC severity was scored by three blinded investigators (H&E staining), and confirmed by RT-qPCR for inflammation (Il-6). Intestinal epithelial maintenance was evaluated by immunofluorescence staining and quantitative analysis of proliferation marker Ki67, and by RT-qPCR for intestinal stem cell marker Lgr5.

Main Results
AFS cell administration prior to NEC induction led to a reduction in NEC severity score (Fig1A) and inflammatory cytokine Il-6 (Fig1B). Intestinal epithelial proliferation (Ki67, Fig1C) and intestinal stem cell activation (Lgr5, Fig1D) were elevated with AFS cell administration compared to NEC, indicating maintenance of epithelial homeostasis.

Conclusions
These data demonstrate that when administered before NEC induction, AFS cells prevent intestinal injury and prompt intestinal growth. AFS cells can be used as clinical prevention strategy for infants at risk of NEC.
ENHANCED PROPAGATION OF PAEDIATRIC UROTHELIAL CELLS IN CULTURE FOR BLADDER TISSUE ENGINEERING

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1Stem Cell and Regenerative Medicine Section, University College London Great Ormond Street Institute of Child Health and Great Ormond Street Hospital, London, United Kingdom. 2Department of Paediatric Urology, Evelina London Children’s Hospital, London, United Kingdom. 3Department of Paediatric Urology, Great Ormond Street Hospital NHS Foundation Trust, London, United Kingdom. 4The Francis Crick Institute, London, United Kingdom. 5Jack Birch Unit of Molecular Carcinogenesis, Department of Biology, University of York, York, United Kingdom

Abstract

Aim of the Study: Bladder tissue engineering represents a promising alternative to circumvent adverse events of conventional bladder augmentation. Current culture systems are unable to generate sufficient cells for therapeutic use from diseased bladder urothelium, due to both limited expansion and differentiation capacities. The aim of the study was to investigate if co-culture with irradiated 3T3-J2 feeder cells would enhance the in vitro expansion of human paediatric urothelial cells derived from small biopsies of healthy and diseased urothelia.

Methods: Resected ureter or bladder specimens from 10 children (3 healthy bladders, 4 healthy ureters, 2 exstrophy bladders and 1 neuropathic bladder) were collected. Urothelial cells were cultured on plates pre-seeded or not with 3T3-J2 feeder cells. The population growth for each condition was calculated. The capacity for differentiation and barrier function of expanded cells was analysed by immunofluorescence and transepithelial electrical resistance (TEER).

Main results: Co-culture with 3T3-J2 feeder cells significantly improved the in vitro expansion of urothelial cells from both healthy and diseased urothelia. The mean cell yields at 30 days after isolation were 6.84 x 10^10 with feeder cells and 0.83 x 10^10 without, respectively (p<0.05). Both groups were capable of differentiation in vitro, generating comparable tight junction structures and barrier function (TEER: 4082 with feeder cells versus 3495 without, Ω·cm^2).

Conclusions: Irradiated 3T3-J2 feeder cells supported the generation of greater numbers of urothelial cells that retained differentiation and functional capacity. This evidence supports the future use of this method for bladder tissue engineering.
**PW8-BS09**

**COMPARISON BETWEEN THE SIZES OF PULMONARY ARTERIOLES AFTER VENTILATION IN CDH RABBIT MODEL**

Rebeca Lopes Figueira, Erika Matsushita, Karina Miura da Costa, Alexandre Todorovic Fabro, Thamires Melchiades, Marcos Borges, Walusa Assad Goncalves, Lourenco Sbragia

Ribeirao Preto Medical School - University of Sao Paulo, Ribeirao Preto, Brazil

Congenital Diaphragmatic Hernia (CDH) is frequently associated with pulmonary hypoplasia and hypertension. Pulmonary hypertension (PH) manifests itself early, especially in ventilated patients with high oxygen dependence. Furthermore, the PH is related to the response of pulmonary arterioles. **Aim:** To evaluate the effect of ventilation response on two different sizes of pulmonary arterioles in CDH rabbit model. **Methods:** It was approved by CAR# #100/2017. *New Zealand* rabbits were divided into 4 groups (n=10 per group): Control (C), Control Ventilated (CV), CDH and CDHV. The surgery was performed on E25 and harvested on E30 (term=30 days). The neonates were ventilated with FlexiVent (Scireq, Montreal, QC, Canada) with the following parameters: respiratory rate 150 breaths/min, PEEP 4 cmH₂O, inspiratory time 0.1s and expiratory time 0.3s. Dynamic compliance (CRS), dynamic elastance (ERS) and dynamic resistance (RRS) were measured every 4 min during 24 min. After that, the lungs were expanded with 20 cm H₂O and fixed in PFA. The slides were prepared with Masson Trichome. The arterioles were divided between two groups < 30μm and 30-60μm and the media wall thickness (MWT) were measured in 40 fields and compared. **Results:** The CRS was increased. ERS and RRS were increased in CDHV (*p*<0.05). Pulmonary arterioles <30μm showed changes in the MWT after ventilation in comparison with >30-60μm (*p*<0.05) (Figure). **Conclusion:** The ventilation response of CDH is changed concerning compliance, elastance and resistance. The bigger arterioles presented smaller response to ventilation and might justify the difficulties in ventilating this population.

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**Figure 1.** CRS Complacence, Elastance and Resistance graphics and photomicrography of pulmonary vessel. A) Control. B) Control ventilated. C) CDH. D) CDH Ventilated. Black dotted lines represent the thickness of pulmonary vessels.
PW8-BS10
ALTEPLASE AND DEOXYRIBONUCLEASE (DNASE) REDUCE MOLECULAR SIZE AND VISCOSITY OF PLEURAL FLUID IN A RAT MODEL OF EMPYEMA

Mario Gehlen, Jose Carlos Fraga, Sergio Amantea, Nadya Silveira, Jane Kulczynski, Eliane Roesch, Kalyana Portal, Paulo Sanches

Aim of the study: To investigate the effects of fibrinolytic therapy with alteplase and/or deoxyribonuclease (DNase) on physical and chemical properties of empyema fluid.

Methods: After ethical approval by the Institutional Review Board, Streptococcus pneumoniae was introduced into the pleural cavity of 67 Wistar rats using a pleural pressure monitoring device. The animals were euthanized after 24 hours. After volume determination, intrapleural fluid was stored at –80ºC. Selected samples were thawed at room temperature before exposure to one of the following: GI=alteplase (n=12), GII-DNase (n=12), GIII=alteplase+DNase (n=12), or GIV=saline (n=6). The mean size of empyema fluid molecules was determined using dynamic light scattering. Viscosity was measured using the drip method.

Main results: Macroscopic analysis confirmed the presence of loculated fluid containing pus and fibrin, for a mean volume of 4.16 mL (0.5 to 8 mL). All samples were culture-positive for Streptococcus pneumoniae. Use of alteplase, DNase, or combined treatment with alteplase+DNase entailed a reduction of more than 135nm in the size of fluid molecules; however, this reduction was statistically significant only for alteplase (p=0.021). Viscosity was reduced in all experimental groups. A decrease in median viscosity was detected following DNase treatment (–5mPa/s), with statistical difference in comparison to controls (8mPa/s; p = 0.048).

Conclusions: In a rat model of empyema, alteplase significantly reduced molecular size, whereas DNase reduced the viscosity of pleural fluid.
PW8-BS11
HUMAN FOETAL GUT MESOANGIOBLATS-LIKE CELLS FOR SMOOTH MUSCLE REGENERATION

Silvia Perin¹, Conor McCann¹, Dipa Natarajan¹, Giulio Cossu², Simon Eaton¹, Paolo De Coppi¹,³, Nikhil Thapar¹,⁴

¹Stem Cells and Regenerative Medicine, UCL Great Ormond Street Institute of Child Health, London, United Kingdom. ²Division of Cell Matrix Biology and Regenerative Medicine, University of Manchester, Manchester, United Kingdom. ³Specialist Neonatal and Paediatric Surgery (SNAPS) Department, Great Ormond Street Hospital NHS Foundation Trust, London, United Kingdom. ⁴Neurogastroenterology and Motility Unit, Department of Gastroenterology, Great Ormond Street Hospital NHS Foundation Trust, London, United Kingdom

Aim of the Study:

Very little is known about regeneration of damaged smooth muscle intestine. The aim of this study is to determine potential use of human foetal gut mesoangioblast-like cells in gut smooth muscle regeneration as a possible treatment for gastrointestinal disorders.

Methods:

Mesoangioblasts-like cells (MABS), isolated from human foetal midgut (UK-HDBR ethic), were expanded and TGFβ was used to induce smooth muscle differentiation in vitro. Immunohistochemistry, FACS analysis and calcium imaging were performed to characterise MABS and smooth muscle derived cells. After eGFP-lentiviral labelling, MABS were transplanted in vivo on injured terminal ileum of NOD-SCID mice (under UK licence, PPL 70/7622) and cell engraftment was assessed with immunohistochemistry after 4 weeks.

Main Results:

MABS showed high proliferative capacity, positivity for mesenchymal markers CD90 and CD146 and for pericyte markers NG2 and PDGFRβ. After TGFβ-treatment MABS expressed smooth muscle proteins SM22 and calponin, and showed intracellular calcium transit in response to carbachol. In vivo, transplanted MABS were able to engraft, distribute in the muscle layers of the injured ileum and differentiate in smooth muscle cells.

Conclusion:

Here we demonstrate that MABS can be isolated from human foetal gut and in vitro, after TGFβ treatment, MABS generate smooth muscle cells being able to respond to carbachol stimulus, suggesting their potential to contract in vivo. MABS ability to differentiate in smooth muscle cells and engraft in vivo candidate them as potential treatment for gut regeneration.
PW8-BS12
NECROTIZING ENTEROCOLITIS IS IMPROVED BY NESFATIN-1 VIA THE MODULATION OF INFLAMMATION-RELATED TRANSCRIPTION FACTORS AND TIGHT JUNCTION PROTEINS

Kivilcim Karadeniz Cerit¹, Turkan Koyuncuoglu¹, Damla Yagmur³, Damla Anıl⁴, İrem Peker Eyuboglu⁵, Serap Sirvanci⁶, Mustafa Akkiprik¹, Burak Aksu⁶, Tolga Dagli¹, Berrak Yegen²

¹Marmara University School of Medicine, Department of Pediatric Surgery, Istanbul, Turkey. ²Marmara University School of Medicine, Department of Physiology, Istanbul, Turkey. ³Marmara University School of Medicine, Istanbul, Turkey. ⁴Marmara University School of Medicine, Department of Histology and Embryology, Istanbul, Turkey. ⁵Marmara University School Of Medicine, Department of Medical Biology, Istanbul, Turkey. ⁶Marmara University School Of Medicine, Department of Medical Microbiology, Istanbul, Turkey.

**Aim:** It was aimed to investigate therapeutic impact of nesfatin-1 and involvement of capsaicin-sensitive afferent neurons in a neonatal rat model of necrotizing enterocolitis (NEC).

**Methods:** To induce NEC, newborn Sprague-Dawley rat pups were separated and fed with a hyperosmolar formula, while control pups (n=16) had no intervention. NEC-induced rats received intraperitoneally saline or nesfatin-1 for 3 days, while a subgroup of nesfatin-treated rats was injected with capsaicin once on the 3rd day. On the 3rd day, pups were exposed to hypoxic chamber for 45 sec. After clinical states of pups were assessed using a clinical sickness score on the 4th day, pups were sacrificed and intestinal tissues were macroscopically scored. Using RT-PCR, gene expressions of occludin, claudin-3, cycloxygenase-2, nuclear factor (NF)-κB-p65 (RELA), and vascular endothelial growth factor (VEGF) were determined in intestines, and relative abundance of bacteria in feces were quantified by qPCR. Immunohistochemical staining for NF-κB and VEGF was performed in terminal ileum. Study approved by the Ethics Committee (058.2016.mar).

**Main Results:** Along with the increased abundance of Proteobacteria and Actinobacteria, clinical and macroscopic scores of intestine, gene expressions of claudin-3 and RELA, and NF-κB immunoreactivity were higher in saline-treated NEC group as compared to control rats (p<0.001). Increased expressions of occludin, claudin-3, cycloxygenase-2 and RELA genes along with increased VEGF immunostaining were evident in nesfatin-1-treated NEC group (p <0.05).

**Conclusion:** Nesfatin-1 alleviated NEC-induced injury by modulating microbiota, expressions of inflammation-related transcription factors and tight junction proteins.
SCII-YI01
NONOPERATIVE TREATMENT VERSUS APPENDECTOMY FOR ACUTE
NONPERFORATED APPENDICITIS IN CHILDREN: FIVE-YEAR FOLLOW UP OF A
RANDOMISED CONTROLLED PILOT TRIAL

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Svensson1,3

1Department of Women's and Children's Health, Karolinska Institutet, Stockholm, Sweden. 2Department of
Surgery, Västmanland Hospital Västerås, Västerås, Sweden. 3Department of Pediatric Surgery, Astrid
Lindgren Children's Hospital, Karolinska University Hospital, Stockholm, Sweden. 4UCL Great Ormond Street
Institute of Child Health, London, United Kingdom

Aim of the study:
To evaluate the outcome of nonoperative treatment of acute appendicitis in children; five-year outcome.

Methods:
A randomised controlled pilot trial (the CONSAPP trial), including 50 children with non-perforated acute
appendicitis, was conducted in 2012. The patients were randomised to treatment with antibiotics or
appendectomy with one year follow-up reported. The children were followed up for at least five years (5,3 (5,0-5,6)) after inclusion. Data was extracted from the computerised notes and telephone interviews.
The primary outcome was treatment failure, defined as need for a secondary intervention under general
anaesthesia, related to the previous diagnosis of acute appendicitis. The study was approved by the Regional Ethics Review Board.

Main results:
There were no failures in the appendectomy group (0/26) and 11 failures in the nonoperative group
(11/24). Nine failures had occurred during the first year after inclusion, two of whom had histologically
confirmed appendicitis. There were two further patients with recurrent acute appendicitis 1 to 5 years after
inclusion. Both these patients had uncomplicated laparoscopic appendectomies for histologically confirmed
acute appendicitis. There were no losses to follow-up.

Conclusions:
At five years of follow-up 46% of children treated with antibiotics for acute appendicitis, had undergone
appendectomy, although acute appendicitis was only histologically confirmed in 17%. Treatment with
antibiotics appears to be safe in the long term; none of the children previously treated conservatively re-
apresented with complicated appendicitis. Long-term data from larger trials are needed to clarify the role of
treatment of nonperforated acute appendicitis in children with antibiotics.
SCIII-YI02
THE SPACED LEARNING CONCEPT SIGNIFICANTLY IMPROVES TRANSFERABILITY AND LONG-TERM ACQUISITION OF LAPAROSCOPIC SUTURING SKILLS: A PILOT RANDOMIZED CONTROLLED STUDY

Johannes Boettcher, Lea Klippgen, Stefan Mietzsch, Robert Bergholz, Konrad Reinshagen, Michael Boettcher
UKE Medical School, Hamburg, Germany

Background: Spaced learning has been shown to be effective in various areas like traditional knowledge or motor skill acquisition. By using a pre-post-follow-up-design the aim of the study was to evaluate the long-term impact of implementation of the spaced learning concept in laparoscopic training.

Methods: To evaluate the effectiveness of spaced learning, subjects were asked to perform four surgeon’s square knots on a bowel model within 30min - prior and post 3h of hands-on training. To examine the long-term skills the same students were asked to perform comparable but more complex task (four slip knots in a model of esophageal atresia) 1y later as follow-up measurement. All subjects were medical students and novice in laparoscopic suturing. Total time, knot stability (tensiometer), suture accuracy, knot quality (Muresan score) and laparoscopic performance (Munz checklist) were assessed. Moreover, motivation was accessed using QCM.

Results: Twenty students were included in the study; after simple randomization, ten were trained using “spaced learning” concept and ten conservatively. Two were lost to follow-up. Both groups had comparable baseline characteristics and improved after training significantly regarding all aspects assessed in this study. Subjects that trained via spaced learning were superior in terms of speed, knot quality and suture strength.

Conclusion: The spaced learning concept is very suitable for long-term complex motor skill acquisition like laparoscopic suturing and knot tying. It is superior to conventional training regarding speed and most importantly knot quality. Thus, we strongly recommend to incorporate the spaced learning concept into training courses and surgical programs.
SCIII-YI03
FIRST POPULATION-BASED REPORT OF INFANTS WITH CDH: 30-DAY-OUTCOMES FROM THE AMERICAN COLLEGE OF SURGEONS NATIONAL QUALITY IMPROVEMENT PROGRAM

Elke Zani-Ruttenstock, Augusto Zani, Annie Fecteau
Division of General and Thoracic Surgery, The Hospital for Sick Children, Toronto, Canada

Aim of the study:
The American College of Surgeons (ACS) has developed a registry, the National Quality Improvement Program Pediatric (NSQIP-P), that provides participating centers with high quality surgical outcome data for children. Herein, we aimed to analyze for the first time the short-term outcomes of live-born infants with congenital diaphragmatic hernia (CDH) registered on this large North American database.

Methods:
During 2015-2016, 101 participating centers uploaded 95 peri-operative data points on the NSQIP-P database for patients that underwent surgical repair of CDH. Following ethical approval, we reviewed the demographics, peri- and post-operative data (up to 30 days following surgical repair) of infants with CDH.

Main results:
There were 432 (61% male) infants, who underwent CDH surgical repair during the study period. The prematurity rate (gestational age <37 weeks) was 17%. The majority of infants (82%) had cardiac risk factors identified (72% were reported as major/severe). Extra-corporeal membrane oxygenation (ECMO) was employed in 13% of patients prior to surgery. The majority of infants (83%) were ventilated preoperatively, 34% received inotropes. Median age at surgery was 5 (0-74) days. CDH repair was attempted via thoracoscopy in 18% (n=79) infants, but with a high rate of conversion to open surgery (n=32, 41%). The post-operative 30-day mortality rate was 9%.

Conclusions:
This is the first report on CDH outcomes from the NSQIP-P database. Utilization of ECMO was low compared with single-center studies from North America. The post-operative mortality rate of babies with CDH considered suitable for surgery remains alarmingly high.
SCIENTIFIC SESSION III: YOUNG INVESTIGATOR AWARD

Thursday June 21st

SCIII-YI04
LONG-TERM FUNCTIONAL OUTCOMES AND QUALITY OF LIFE IN HIRSCHSPRUNG’S DISEASE: A NATIONWIDE SURVEY

Rob J. Meinds1, Alida F.W. van der Steeg2,3, Cornelius E.J. Sloots4, Marieke J. Witvliet5, Ivo de Blaauw6, Wim G. van Gemert7, Monika Trzpis1, Paul M.A. Broens1

1University Medical Center Groningen, Groningen, Netherlands. 2Emma Children's Hospital, AMC and VU University Medical Center, Amsterdam, Netherlands. 3Center of Research on Psychology in Somatic diseases (CoRPS), Tilburg University, Tilburg, Netherlands. 4Erasmus MC Sophia Children’s Hospital, Rotterdam, Netherlands. 5Wilhelmina Children’s Hospital, University Medical Center Utrecht, Utrecht, Netherlands. 6Radboudumc-Amalia Children’s Hospital, Nijmegen, Netherlands. 7University Medical Center Maastricht, University of Maastricht, Maastricht, Netherlands

Aim of the Study: It is unclear whether functional outcomes improve or deteriorate with age following surgery for Hirschsprung’s disease (HD). Our aim was to determine the long-term functional outcomes, to determine factors associated with poor outcomes, and to evaluate their influence on quality of life (QoL).

Methods: We performed a nationwide, cross-sectional survey in all known HD patients of 8 years and older (N = 619). Patients with a permanent stoma or intellectual disability were excluded. Functional outcomes were assessed by Rome IV criteria and validated defecation scores. QoL was assessed by the CHQ-CF87 questionnaire and WHOQOL-100 questionnaire in pediatric and adult patients, respectively.

Main results: A total of 55.9% (n = 346) patients responded (median age 18 years). The prevalence of constipation was comparable in pediatric and adult patients, and in patients and controls. The prevalence of fecal incontinence was lower in adult patients compared to pediatric patients (16.8% versus 37.6%, P<.001), but remained higher than in controls (16.8% versus 6.1%, P=.003). Patients that required a redo pull-through procedure had an increased likelihood of fecal incontinence (odds ratio 3.88, P=.002). There was a negative relationship, and moderate, (r>0.40) between several QoL domains and constipation.

Conclusions: Functional complaints, especially fecal incontinence, improve with aging but persist in a substantial group of adult patients. Patients who required a redo pull-through procedure suffered more often from fecal incontinence. Transitional care is recommended in a select group of patients, as poor functional outcomes were negatively related to QoL.
SCII-YI05

DOES MECHANICAL BOWEL PREPARATION REDUCE THE RISK OF DEVELOPING INFECTIOUS COMPLICATIONS IN PAEDIATRIC COLORECTAL SURGERY? A SYSTEMATIC REVIEW AND META-ANALYSIS

Koen Zwart¹, Dirk-Jan van Ginckel¹, Caroline Hulsker², Marieke Witvliet², Maud van Herwaarden²

¹University Medical Center Utrecht, Utrecht, Netherlands. ²Wilhelmina Children’s Hospital, Utrecht, Netherlands

AIM OF THE STUDY – Recent meta-analyses concerning mechanical bowel preparation (MBP) prior to colorectal surgery in the adult population have shown that MBP does not significantly reduce the overall infectious complication rate. The aim of this systematic review and meta-analysis is to determine whether MBP prior to colorectal surgery reduces the risk of developing infectious complications in the paediatric population.

METHODS – PubMed, Embase and the Cochrane library were systematically searched to identify articles comparing paediatric patients receiving MBP with paediatric patients not receiving MBP prior to colorectal surgery. Results are presented with weighted risk differences based on the number of events and sample size per study.

MAIN RESULTS – Six original studies were included comparing MBP (n=810) versus non-MBP (n=1167). The meta-analysis shows the overall risk of developing infectious complications in patients with MBP is 10.1% compared to 9.1% in patients without MBP, resulting in a non-significant risk difference of -0.03% (CI -0.09% – +0.03%). Concerning the number of wound infections and anastomotic leaks specifically, a non-significant risk difference of -0.03% (CI -0.08% – +0.02%) and of 0.01% (CI -0.01% – +0.02%) was found, respectively.

CONCLUSION – MBP prior to colorectal surgery does not reduce the risk of developing infectious complications in the paediatric population.
Figure 1. Forest plot of the overall risk of developing infectious complications in patients receiving MBP

<table>
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<th>Study or Subgroup</th>
<th>Favours No MBP Events</th>
<th>MBP Events</th>
<th>Total Events</th>
<th>Total Weight</th>
<th>Risk Difference M-H, Random, 95% CI</th>
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<td>20</td>
<td>24</td>
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<td>-0.17 [-0.44, 0.09]</td>
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</tr>
<tr>
<td>Shah et al. 2016</td>
<td>4</td>
<td>34</td>
<td>38</td>
<td>3.8%</td>
<td>0.12 [-0.17, 0.41]</td>
<td>2016</td>
</tr>
<tr>
<td>Subtotal (95% CI)</td>
<td>34</td>
<td>42</td>
<td>76</td>
<td>8.4%</td>
<td>-0.04 [-0.32, 0.25]</td>
<td></td>
</tr>
<tr>
<td>Total events</td>
<td>8</td>
<td>12</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heterogeneity: Tau² = 0.02, Chi² = 21.16, df = 1 (P = 0.14); I² = 54%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Test for overall effect: Z = -0.24 (P = 0.81)</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

1.1.2 Retrospective comparative studies

<table>
<thead>
<tr>
<th>Study or Subgroup</th>
<th>Favours No MBP Events</th>
<th>MBP Events</th>
<th>Total Events</th>
<th>Total Weight</th>
<th>Risk Difference M-H, Random, 95% CI</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leys et al. 2005</td>
<td>1</td>
<td>33</td>
<td>34</td>
<td>26.0%</td>
<td>0.00 [-0.06, 0.07]</td>
<td>2005</td>
</tr>
<tr>
<td>Brezler et al. 2009</td>
<td>0</td>
<td>8</td>
<td>8</td>
<td>8.0%</td>
<td>-0.17 [-0.35, 0.02]</td>
<td>2009</td>
</tr>
<tr>
<td>Serrurier et al. 2011</td>
<td>7</td>
<td>85</td>
<td>92</td>
<td>22.8%</td>
<td>-0.08 [-0.16, -0.00]</td>
<td>2011</td>
</tr>
<tr>
<td>Arès et al. 2017</td>
<td>90</td>
<td>1007</td>
<td>1097</td>
<td>29</td>
<td>34.7%</td>
<td>2017</td>
</tr>
<tr>
<td>Subtotal (95% CI)</td>
<td>1113</td>
<td>768</td>
<td>781</td>
<td>91.6%</td>
<td>-0.03 [-0.09, 0.04]</td>
<td></td>
</tr>
<tr>
<td>Total events</td>
<td>98</td>
<td>70</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heterogeneity: Tau² = 0.00, Chi² = 9.31, df = 3 (P = 0.03); I² = 68%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Test for overall effect: Z = 0.82 (P = 0.41)</td>
<td></td>
<td></td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

Total (95% CI)  

<table>
<thead>
<tr>
<th>Study or Subgroup</th>
<th>Favours No MBP Events</th>
<th>MBP Events</th>
<th>Total Events</th>
<th>Total Weight</th>
<th>Risk Difference M-H, Random, 95% CI</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total events</td>
<td>106</td>
<td>82</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heterogeneity: Tau² = 0.00, Chi² = 11.79, df = 5 (P = 0.04); I² = 58%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Test for overall effect: Z = 0.91 (P = 0.37)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Test for subgroup differences: Chi² = 0.00, df = 1 (P = 0.95); I² = 0%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Aim of the Study: Necrotizing enterocolitis (NEC) is the leading cause of death in preterm infants. However, the progression of intestinal injury during NEC is not understood. We sought to investigate the development of intestinal injury, regeneration impairment, and gut dysbiosis in a mouse model of NEC.

Methods: Experimental NEC was induced at postnatal days 5-9 using hypoxia, oral lipopolysaccharide, and hyperosmolar formula gavage (n=6-10 mice/group; ethics-approved protocol #32238). Breastfed (BF) pups served as non-stressed controls. Pups were sacrificed at P5, P7, and P9. Three blinded investigators scored NEC injury in the distal ileum. Ileal inflammation (IL-6) and intestinal stem cell levels (Lgr5) were measured using qPCR. Goblet cell differentiation (Muc2) in the ileum was determined with immunofluorescence staining (*p<0.05; ***p<0.001). Ileal microbiota was assessed using 16S rDNA sequencing and QIIME analysis with PICRUSt (11,618 reads/sample).

Main Results: NEC-induced mucosal injury (A) and inflammation (B) were exacerbated from P5-P9. Intestinal stem cells (C) and goblet cell differentiation (D) both decreased from P5-P9. At P9, the NEC microbiota was enriched in Proteobacteria (q=0.024; red bars) and deficient in Firmicutes (q=0.035; blue bars) (E), while the NEC functional profile for bacterial disease pathogenesis was enhanced (F).

Conclusions: Experimental NEC induces progressive worsening of intestinal injury and inflammation, impairment of regeneration, and gut dysbiosis in mouse pups. These findings highlight changes in the intestinal epithelium and gut microbiota during NEC development and offer new targets for preventing NEC in at-risk human preterm infants.
MIRIAM DUCI1, KANJI CEladin2, MARTA ERCULIANI3, SILVIA VISENTIN2, GIOVANNA VERLATO3, ANNA CHIARA FRIGO4, PIERGIORIO GAMBA1, FRancesco Fascetti-Leon1

1Division of Paediatric Surgery, Department of ‘Salute della Donna e del Bambino’, University of Padova, Padova, Italy. 2Division of Obstetrics and Gynecology, Department of ‘Salute della Donna e del Bambino’, University of Padova, Padova, Italy. 3Division of Neonatal Intensive Care Unit, Department of ‘Salute della Donna e del Bambino’, University of Padova, Padova, Italy. 4Department of Cardiac, Thoracic and Vascular Sciences, University of Padova, Padova, Italy

Antenatal factors may play an important role in NEC. This study aimed to identify antenatal risk factors associated with the development of NEC and the role of the placental alterations to this context. Data of NEC patients including antenatal findings [preeclampsia, diabetes, cholestasis, abnormal antenatal umbilical flow (AAUF), clinic chorioamniositis (CC) and histology of placentas] were compared to unaffected cases between 2002 and 2016 in a single centre. Unaffected infants were matched for gestational age. Newborns with cardiovascular diseases were excluded. Bivariate and multivariate analysis were performed. We identified 136 NEC cases and 134 controls in the study period. The group of mothers of NEC-neonates had an higher prevalence of preeclampsia [p=0.0024], CC [p<0.001] and AAUF [p<0.0001]. There weren’t differences in terms of diabetes and cholestasis (respectively p=0.9 and p=0.13). Histology of Placentas from 123/136 NEC cases and 126/133 unaffected newborns were available. Chorioamniositis was significantly more present in NEC cases vs controls (p=0.0001). There weren’t differences in vascular anomalies and necrotic alterations (table-1). Multivariate analysis identified AAUF (p<0.001), CC (p=0.021) and histological chorioamniosis (HC) (p=0.0002) as significant predictors of NEC (table-2). Bivariate tests show that preeclampsia and HC occurred more often in severe cases of NEC (respectively p=0.002 and p=0.0044).

This study suggests that AAUF, CC and HC can independently predict the risk of NEC. Preeclampsia and HC seems associated to more severe cases. As AAUF and CC are immediately available data, they allow to raise the alert level towards newborns at higher risk.
<table>
<thead>
<tr>
<th>Variables</th>
<th>Mothers of NEC-neonates</th>
<th>Mothers of unaffected neonates</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preeclampsia</td>
<td>54 (39.7%)</td>
<td>33 (24.63%)</td>
<td>0.0024</td>
</tr>
<tr>
<td>Abnormal Antenatal Umbilical Flow</td>
<td>91 (66.91%)</td>
<td>13 (9.7%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Diabetes</td>
<td>9 (6.62%)</td>
<td>10 (7.46%)</td>
<td>0.9</td>
</tr>
<tr>
<td>Cholestasis</td>
<td>16 (11.76%)</td>
<td>9 (6.71%)</td>
<td>0.13</td>
</tr>
<tr>
<td>Clinic Chorioamnitis</td>
<td>23 (16.91%)</td>
<td>4 (2.98%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Histological Chorioamnitis</td>
<td>69 (56.1%)</td>
<td>40 (31.75%)</td>
<td>0.0001</td>
</tr>
<tr>
<td>Necrotic Alterations</td>
<td>20 (16.2%)</td>
<td>16 (12.69%)</td>
<td>0.64</td>
</tr>
<tr>
<td>Vascular Anomalies</td>
<td>25 (20.32%)</td>
<td>34 (27.64%)</td>
<td>0.23</td>
</tr>
</tbody>
</table>

Table 1

<table>
<thead>
<tr>
<th>Variables</th>
<th>OR</th>
<th>P value</th>
<th>95%CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abnormal Antenatal Umbilical Flow</td>
<td>28.486</td>
<td>&lt;0.001</td>
<td>11.263-63.049</td>
</tr>
<tr>
<td>Clinic Chorioamnitis</td>
<td>5.078</td>
<td>0.021</td>
<td>1.260-19.716</td>
</tr>
<tr>
<td>Histological Chorioamnitis</td>
<td>38.283</td>
<td>0.0002</td>
<td>2.172-718.005</td>
</tr>
</tbody>
</table>

Table 2
TRACHEAL REPLACEMENT USING AN IN-BODY-TISSUE-ENGINEERED COLLAGENOUS TUBE “BIOTUBE” WITH A BIODEGRADABLE STENT IN A BEAGLE MODEL: A NOVEL TRACHEAL SCAFFOLD

Shohei Hiwatashi¹, Yasuhide Nakayama², Yuichi Takama¹, Satoshi Umeda¹, Takeshi Terazawa², Hiroomi Okuyama³

¹Osaka university, Suita, Japan. ²National Cerebral and Cardiovascular Center, Suita, Japan

Aim: To investigate the usefulness of BIOTUBE, an in-body-tissue-engineered collagenous tube with a biodegradable stent, as a novel tracheal scaffold in a beagle model.

Methods: We prepared BIOTUBEs by embedding specially designed molds, including biodegradable stents, into subcutaneous pouches in beagles. After 2 months, the molds filled with ingrown connective tissues and were harvested to obtain the BIOTUBEs. Tubular BIOTUBEs, cut to 2 cm length, were implanted to replace the same-length defects in the cervical trachea of 5 beagles (Figure 1). Endoscopic and X-ray fluoroscopic evaluations were performed every week until the state of the lumen became stable. The trachea, including the BIOTUBE, was harvested and subjected to histological evaluation, 3 to 7 months after implantation. The study protocol was approved by the National Cerebral and Cardiovascular Center Committee (No. 17013).

Results: One beagle died 28 days after implantation because of insufficient expansion and retentive force of the BIOTUBE. The remaining four beagles were transplanted with a BIOTUBE reinforced by strong stent; all four survived throughout the observation period. Endoscopy revealed narrowing of the BIOTUBEs in all 4 beagles due to inflammatory reaction, but patency was maintained by steroid application at the implantation site and balloon dilation against the stenosis. After 2 months, the lumen gradually became wider (Figure 2). Histological analyses demonstrated that the internal surface of the BIOTUBEs was covered with tracheal epithelial cells.

Conclusion: This study demonstrated the usefulness of the BIOTUBE with a biodegradable stent, as a novel scaffold for tracheal regeneration.
SCII-YI09
SONS WITH CRYPTORCHIDISM BORN TO MOTHERS WHO HAVE SMOKED DURING PREGNANCY HAVE IMPAIRED TESTICULAR FUNCTION COMPARED TO CRYPTORCHIDS OF NON-SMOCKERS

Simone Hildorf¹, Erik Clasen-Linde², Lihua Dong³, Dina Cortes⁴,⁵, Jorgen Thorup¹,⁴

¹Dept. Paediatric Surgery, Rigshospitalet, Copenhagen, Denmark. ²Dept. Pathology, Rigshospitalet, Copenhagen, Denmark. ³Lab. reproductive Biology, Rigshospitalet, Copenhagen, Denmark. ⁴Faculty of Health and Medical Sciences, University of Copenhagen, Copenhagen, Denmark. ⁵Dept. Paediatrics, Hvidovre Hospital, Copenhagen, Denmark

Aim of the Study: A meta-analysis including 11,900 cases showed that maternal gestational smoking was associated with increased risk of cryptorchidism. The aim of study was to investigate whether a hormone profile of cryptorchid boys supplementing histopathological evaluation of testicular biopsies could add detailed knowledge to the impact of maternal gestational smoking on pathogenesis of cryptorchidism.

Methods: 601 boys 4 months-14 years old were included. Because of the pronounced age dependency of normal hormone levels the evaluated parameters in boys whose mothers had smoked heavily (>10 cigarettes/day) during pregnancy were compared to age matched cryptorchid controls of non-smoking mothers (1:6). Birthweight, unilateral/bilateral cryptorchidism, germ-cell number/tubular cross section(G/T), gonadotropins and inhibin-B were evaluated.

Main results: 501 boys were sons of non-smokers, 72 boys of intermittent smokers and 28 boys of heavy smokers. (38.7%, 44.4% and 60.7% respectively had bilateral cryptorchidism (p=0.021)). Table shows statistical significant (*p<0.05) results of 28 sons of heavy smoking mothers compared to 168 age matched controls of non-smokers.

Conclusions: All findings could be associated with placental dysfunction with impaired hCG production well described in women smoking during pregnancy.

<table>
<thead>
<tr>
<th>Data matched by age (1:6)</th>
<th>Heavy smoking mothers</th>
<th>Non-smoking mothers</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of boys</td>
<td>28</td>
<td>168</td>
</tr>
<tr>
<td>Median age (years) at orchiopexy (range)</td>
<td>1.8 (0.8-6.4)</td>
<td>1.8 (0.8-6.5)</td>
</tr>
<tr>
<td>Median birth weight (range)</td>
<td>3096 (1200-5500)*</td>
<td>3500 (680-5800)*</td>
</tr>
<tr>
<td>Median pg/ml serum inhibin-B (range)</td>
<td>96 (20-281)*</td>
<td>115 (27-260)*</td>
</tr>
<tr>
<td>Median IU/L serum FSH (range)</td>
<td>0.9 (0.2-3.9)</td>
<td>0.7 (0.17-3.1)</td>
</tr>
<tr>
<td>Median G/T (range)</td>
<td>0.444 (0-3.132)*</td>
<td>0.539 (0-2.830)*</td>
</tr>
</tbody>
</table>
SCIII-YI10
ALTERATIONS OF TIGHT JUNCTIONS EXPRESSION IN INJURED MOUSE INTESTINAL ORGANIDS

Carol Lee¹, Bo Li¹, Yuhki Koike², Hiromu Miyake¹, Shogo Seo¹, Agostino Pierro¹,³

¹The Hospital for Sick Children, Toronto, Canada. ²Mie University, Tsu, Japan. ³University of Toronto, Toronto, Canada

Aim: Necrotizing enterocolitis (NEC) is associated with disrupted intestinal barrier and administration of amniotic fluid stem cells (AFSCs) decreased intestinal permeability in experimental NEC. Intestinal organoids are 3-D structures that contain all cell types of the intestinal epithelium. We hypothesize that injured intestinal organoids permeability is increased and that this increase is due to the change in the tight junctions which can be reversed by AFSCs.

Methods: Following ethical approval (#32238), mouse intestinal organoids were established using ileal crypts. To induce injury, organoids were treated with lipopolysaccharide (LPS) (200µg/ml) and hypoxia for 48 hours. AFSCs were used as treatment. Organoids permeability was investigated using FITC-dextran, 4kDa (FD4) and tight junction markers (claudin 2, 3 and 4) were assessed by immunofluorescence staining.

Results: Translocation of FD4 was not observed in control organoids, increased in injured organoids, and not observed in organoids treated with AFSCs. Immunofluorescence staining shows that tight junction ZO-1 was lower in injured organoids compared to control and administration of AFSCs restored ZO-1 expression (A-C). Pore-forming claudin 2 (D-F) was upregulated in injured organoids while claudin 3 (G-I) and claudin 4 (J-L) were deranged in injured organoids and administration of AFSCs prevented these changes.

Conclusions: Intestinal organoid permeability was impaired by hypoxia and LPS. This damage can be reversed by AFSCs administration as shown by permeability assay, decreased of pore-forming claudin2 and restoring of tight junction proteins claudin 2 and 3. These findings indicate that AFSCs play an important role in restoration of intestinal permeability.
WHY PEDIATRIC SURGEONS ARE NEEDED - A SYSTEMATIC REVIEW ON THE EVIDENCE

Oliver Muensterer
University Medicine Mainz, Mainz, Germany

Background: Pediatric surgeons believe to have an indispensable role in treating the most important part of society. However, models in which general and/or specialist adult surgeons take over pediatric care are practiced or increasingly proposed.

Purpose: This systematic review examines the available evidence on outcome when children are cared by pediatric versus non-pediatric general or specialist surgeons.

Methods: A search was performed on using the terms <adult> AND <pediatric> <surgery> <care> was performed. Titles and abstracts were screened for studies that compared outcome when treatment was provided by either pediatric or non-pediatric surgeons. Multiple studies on one diagnosis were pooled, when available.

Results: Twenty-one relevant full-text articles were extracted out 3587 titles and 328 abstracts. Benefits of pediatric specialist care were found for several indications. Specifically, recurrence after inguinal hernia repair (OR 0.42), mucosal perforation during pyloromyotomy (OR 0.19), risk of operative treatment for intussusception (OR 0.20) or solid organ injury (OR 0.19), complication rate after neonatal surgery (OR 0.37), as well as chance and amount of bowel resection during surgery for intussusception (-17% resected bowel) or volvulus (OR 0.63) were all significantly lower if care was provided by pediatric versus non-pediatric surgeons. There was no significant difference in outcome for the treatment of appendicitis.

Conclusions: Care for children is more effective, less invasive, more economic, and safer when provided by pediatric rather than adult surgeons for a broad spectrum of diagnoses. In contrast, outcome for appendicitis is equivalent. These findings may help define patterns for optimal resource utilization.
SCIENTIFIC SESSION IV: GENERAL

Thursday June 21st

SCIV-G02

GASTROSCHISIS: THE IMPACT OF PERINATAL FINDINGS ON OUTCOME

Linus Jönsson¹, Viktor Husmark¹, Lars-Goran Friberg², Michaela Dellenmark-Blom¹, Vladimir Gatzinsky², Kate Abrahamsson¹

¹Dept of Pediatric Surgery, Gothenburg, Sweden. ²Dept of Pediatric Surg, Gothenburg, Sweden

Aim of the Study: To investigate pre- and postnatal factors affecting outcomes among children with gastroschisis (GS).

Methods: A retrospective journal study on 169 consecutive patients treated at a single pediatric surgical center in Scandinavia 1975-2017. Data from maternity records and medical charts of GS patients up to 15 years of age was used. Appendectomy en passant was performed if easily achieved during primary surgery. Unavailable logistic regression was used to calculate odds ratio (OR, 95% confidence interval). The study was approved by the local ethical committee.

Main results: 8 patients (5%) died, all within 2 years of age. Mortality was related to antenatal factors and factors related to intestinal failure postnatally; polyhydramnios seemed particularly detrimental (Table 1). Staged closure increased the risk of reoperations within 1 year of age (OR 2.67, 1.10-6.46). Child growth at 10 years was associated to intestinal dilation in utero and weight, stature and BMI of the mother at five and 10 years of age (Table 2). Appendectomy en passant was associated with an increased use of laxantia at 1 year of age (OR 2.9, 1.2-6.8).

Conclusions: Antenatal factors implying fetal intestinal compromise increase the risk of death within two years of age. Nutritional status of the mother and antenatal intestinal dilation might affect the growth of the child from birth until 15 years of age. Appendectomy en passant at primary closure may affect early intestinal function and should be further investigated.
SCIV-G03
OUTCOME OF REFUSED NEONATAL SURGICAL CASES IN A LOW TO MIDDLE INCOME COUNTRY

Aly Shalaby, Moutaz Ragab, Khaled Bahaaeldin
Cairo University Specialized Pediatric Hospital, Cairo, Egypt

Aims of the study: The combination of a high birth rate and limited resources leads to a strain on health services with dire consequences to the patient. We aimed to track the outcome of refused cases at our tertiary center in the university teaching hospital.

Methods: Data was collected prospectively on all refused surgical neonates at our institution. The families were later contacted to document outcomes.

Main results: Over a 10 month period from January 2017 to October 2017 we received 495 surgical referrals. Of these 209 (42.2%) were refused due to lack of incubators or nursing staff. Eighty one (44%) were from our city. The majority were cases of esophageal atresia with trachea-esophageal fistula (32.5%), followed by bowel obstruction (19%). Thirty five cases were lost to follow up, of the remaining 174 cases 84 (48.2%) died, 36 (20%) were referred to public hospitals, 48 (27.5%) to private hospitals and 6 (3.4%) were managed conservatively.

Conclusions: In our country nearly half of refused neonatal surgical cases die while waiting for surgical intervention. Cases were shared equally between the public and private sector. This data is crucial in planning service provision for neonates requiring surgical treatment.
VALUE OF NEAR INFRARED SPECTROSCOPY (NIRS) AS ADDITIONAL INTRA-OPERATIVE MONITORING IN INFANTS UNDERGOING MAJOR ABDOMINAL SURGERY

Anna Morandi¹, Stefania Franzini², Maria Teresa Ambrosini², Edoardo Calderini², Ernesto Leva¹

¹Department of Pediatric Surgery, Fondazione IRCCS Ca' Granda - Ospedale Maggiore Policlinico, Milano, Italy. ²Anaesthesia and Paediatric Intensive Care Unit, Fondazione IRCCS Ca' Granda - Ospedale Maggiore Policlinico, Milano, Italy

Aim of the Study: NIRS allows non-invasive, continuous monitoring of regional cerebral and splanchnic O2 saturation (rSO2), mirroring the balance between oxygen delivery and consumption at tissue level. Falls in rSO2>20% from baseline or values <50% are pathological, as they may cause, if overlooked, organ damage. In infants undergoing major abdominal surgeries we added NIRS to ASA monitoring.

Methods: we report our experience between March 2017 and January 2018 with 6 infants (mean age 3.2 months, SD±5.9) undergoing general anaesthesia for major surgery. Two patients underwent a Kasai’s procedure, two had Congenital Diaphragmatic Hernia (CDH) repair, one gastroschisis repair and the last one received nephrectomy. All patients received standard monitoring (NBP, HR, ECG, SpO2, Temperature, UO) and NIRS (INVOS 5100, Somanetics®, Troy, Michigan). We elaborated NIRS traces by the Shortcut INVOS software and analysed them along with intra-operative vital signs recordings.

Main results: we observed a splanchnic rSO2 decrease > 20% in all patients during key surgical manoeuvres (i.e. reducing viscera into the abdominal cavity in CDH and gastrochisis, liver dislocation in Kasai’s procedure, contralateral packing during nephrectomy). Three patients showed splanchnic rSO2<50% during intra-abdominal reduction of viscera. Both Kasai’s patients showed a decrease > 20% in cerebral and splanchnic rSO2 much earlier than any monitoring change. The last patient showed significant fall in splanchnic rSO2 without any standard monitoring change.

Conclusions: in infants NIRS may prevent potential organ damage earlier than or in presence of an uneventful standard monitoring, allowing to timely amend ongoing physiological derangements.
SCIV-G05
PARENTAL PERCEPTIONS OF CLINICAL RESEARCH IN PAEDIATRIC SURGERY: A PILOT STUDY

Li Wen Lee¹, Yoong Wend Chen¹, Candy SC Choo², Yong Chen², Shireen A Nah²,¹

¹Duke-NUS Medical School, Singapore, Singapore. ²KK Women’s and Children's Hospital, Singapore, Singapore

Aim: Participant recruitment in paediatric research requires parental consent. We evaluate parental attitudes to clinical research in a general paediatric surgical setting.

Methods: An ethically approved prospective survey was administered to parents/legal guardians accompanying their children in the paediatric surgical outpatient clinic (OC) or day surgery (DS) using convenience sampling in September-November 2017. We modified a published survey (Abernathy et al) employing Likert scale responses. Questions included demographics, parental willingness to enrol children in specified types of research and beliefs regarding conduct of research. Responses were converted to scores for analysis. Chi-squared tests or Mann-Whitney U tests were used with p<0.05 significance.

Results: Eighty-four parents were surveyed (OC, n=67; DS, n=17) of 100 approached. Most were willing to enrol their child in research involving sample collection: urine (96%), saliva (86%), extant blood samples (81%); but this dropped significantly when additional blood draws were required (49%, p<0.001). Parents were significantly less amenable to studies involving investigational compared to common therapies (p<0.001), and significantly less likely to enrol their children in studies with elements of randomisation compared to common therapies (p<0.001). Barriers to enrolment included child’s young age (45%) and perceived compromise to privacy (32%). Majority were amenable to research relating to their child’s condition (82%) and felt that research should be conducted to help others (89%).

Conclusion: Most parents see the benefit and necessity of clinical research. However, studies involving novel investigational treatments or elements of randomisation create uncertainty, making parents less amenable to enrolling their child in research.
SCIV-G06
PERIOPERATIVE MUSIC INTERVENTIONS FOR INFANTS: THE MUSIC STUDY, A RANDOMIZED CONTROLLED TRIAL

Rosalie Kühlmann¹, Joost van Rosmalen², Lonneke Staals¹, Claudia Keyzer-Dekker¹, Jaap Dogger¹, Tom de Leeuw¹, Fred van der Toorn¹, Hans Jeekel², René Wijnen¹, Monique van Dijk¹,²

¹Erasmus University Medical Center – Sophia Children’s Hospital, Rotterdam, Netherlands. ²Erasmus University Medical Center, Rotterdam, Netherlands

Aim of the study: This study aimed to investigate the effects of music interventions on discomfort, anxiety and pain in infants having surgery.

Methods: In a parallel single-blinded randomized controlled trial, 0-3-year-old infants undergoing elective surgery with general anesthesia and caudal block were randomly allocated via a computer-generated list with use of opaque envelopes to a preoperative music intervention (PM), or pre- and intraoperative music intervention (PIM), or no music intervention (control). Outcomes were level of distress and pain following the COMFORT-B scale, preoperative anxiety, and physiologic measurements. Primary outcome assessment was blind. Bonferroni correction adjusted multiple comparisons. Trial registration: NTR5402.

Main results: 195 infants (median age 6.9 months (IQR 3.3-11.1)) were randomized. 178 infants were included in intention-to-treat primary analysis. There was no significant difference in COMFORT-B scores between the PIM-group and control-group at 4 hours after surgery (mean difference -1.22(95%CI - 2.60;0.17); p=0.085). No statistically significant differences in pain scores were found. Additional analysis showed a borderline significant interaction effect between music exposure and a greater reduction from high COMFORT-B score at baseline; p=0.027. General linear modelling showed a statistically significantly reduced heart rate after the preoperative music intervention in the holding area in the combined PM- and PIM group compared to the control-group; p=0.003. A tendency towards lower heart rate was found in the PIM-group compared to the control-group at four hours postoperative; p=0.069.

Conclusions: Music interventions do not reduce postoperative pain in infants, but preoperatively distressed infants seem to benefit with less discomfort postoperatively.
Aim of the study: To investigate if children with IgE-mediated allergy have lower risk of complicated appendicitis, regardless of age and sex.

Methods: The study was approved by the regional ethical board. All children undergoing appendectomy for acute appendicitis at a tertiary center of pediatric surgery between January 2007 and July 2017 were retrospectively reviewed. The degree of appendicitis was classified as uncomplicated (phlegmonous) or complicated (gangrenous, perforated) based on the intraoperative picture and histopathological analysis. The presence of IgE-mediated allergy was routinely asked for and collected from the surgical and anesthesia charts. Primary outcome was complicated or uncomplicated appendicitis. Independent variables were occurrence of any prior IgE-mediated allergy, as well as age, sex, and seasonal antigen exposure.

Main results: Of 605 included children, 349 (58%) had uncomplicated appendicitis and 256 (42%) had complicated disease. IgE-mediated allergy occurred in 24% of children with uncomplicated appendicitis, compared with 8% in children with complicated disease (p<0.0001). The presence of allergy was associated with shorter hospital stay (p=0.004). After adjusting for age and sex, children with allergies were three times less likely to have complicated appendicitis (adjusted OR 0.33 [95% CI 0.19-0.56], p<0.001). The protective association with allergy was particularly strong at times of seasonal antigen exposure during spring (adjusted OR 0.54 [95% CI 0.34-0.86], p=0.01) and summer (adjusted OR 0.56 [95% CI 0.34-0.93], p=0.03).

Conclusion: Children with allergy seem protected from complicated appendicitis. These findings introduce novel opportunities for prevention and prediction of one of childhood’s most feared surgical emergencies.
SCIV-G08
UTILIZATION OF MINIMALLY-INVASIVE PEDIATRIC SURGERY IN GERMANY, AUSTRIA AND SWITZERLAND

Caroline Fortmann¹, Jan-Hendrik Gosemann², Benno M. Ure¹, Martin Metzelder³, Joachim F. Kübler¹

¹Pediatric Surgery, Hannover Medical School, Hannover, Germany. ²Pediatric Surgery, University of Leipzig, Leipzig, Germany. ³Pediatric Surgery, Medical University of Vienna, Vienna, Germany

Aim of the study: Minimally-invasive techniques have become standard of care for an increasing number of surgical procedures in pediatric patients. We evaluated utilization of minimally-invasive techniques in pediatric surgical centers in german-speaking countries to illustrate development and identify improvement potential.

Methods: A survey was sent to all 102 pediatric surgery departments in Germany, Austria and german-speaking Switzerland to evaluate spectrum of minimally-invasive operations, quantity of procedures, conversions, major complications, number of performing surgeons and residents in the year 2016.

Main results: 68 institutions (66.7%) took part in our survey of which 41.2% were academic hospitals. Laparoscopy is performed in all clinics. For basic procedures (e.g. appendectomy and cholecystectomy) laparoscopy is established in most centers. But numerous institutions do not perform more demanding procedures, even if minimally-invasive techniques are considered gold standard (Figure 1). There are only few centers that offer full spectrum of minimally-invasive surgery. The education of the majority of pediatric surgical residents is restricted to simple procedures such as appendectomy. More demanding laparoscopic procedures such as inguinal herniotomy, pyloromyotomy and fundoplicatio are rarely taught during residency (Figure 2). Regarding thoracoscopy, majority of centers (94.1%) perform simple thoracoscopic procedures such as tumor biopsy or lung biopsy. Complex thoracoscopic procedures as well as training are restricted to only few centers.

Conclusions: Basic minimally-invasive procedures are broadly used, but complex operations are restricted to few centers. It is alarming that the majority of residents in pediatric surgery are not exposed to or trained in minimally-invasive surgery beyond the basic procedures.

Figure 1: Percentage of institutes who offer selected basic, moderate and advanced procedures.

Figure 2: Percentage of institutes who teach selected basic, moderate and advanced procedures.
EFFECT OF A CLINICAL PATHWAY ON ANTIBIOTIC PERIOPERATIVE PROPHYLAXIS IN PEDIATRIC SURGICAL PATIENTS

Daniele Donà¹², Dora Luise¹, Enrico La Pergola³, Genni Montemezzo³, Theoklis Zaoutis⁴, Piergiorgio Gamba³, Carlo Giaquinto¹

¹Division of Pediatric Infectious Diseases, Department of Women’s and Children’s Health, University of Padua, Padova, Italy. ²Division of Infectious Diseases and the Center for Pediatric Clinical Effectiveness, Children’s Hospital of Philadelphia, Philadelphia,, USA. ³Pediatric Surgery Unit, Department of Women’s and Children’s Health, University of Padua, Padova, Italy. ⁴Division of Infectious Diseases and the Center for Pediatric Clinical Effectiveness, Children’s Hospital of Philadelphia, Philadelphia, USA

Aim

Appropriate perioperative antibiotic prophylaxis (PAP) is essential to prevent surgical site infections (SSIs) and to avoid the misuse of antibiotics. We aimed to determine the effectiveness of a Clinical Pathway (CP) on PAP appropriateness and to evaluate the effect CP implementation on SSIs.

Methods

The study was set at the Paediatric Surgery Unit of a tertiary referral Centre where on 31/01/2017 the CP for PAP was implemented. This is a pre-post quasi-experimental study to assess the changes in PAP during a 6-month period pre (1/02/2016-31/07/2016) and post CP implementation (1/02/2017-31/07/2017). All children aged 1 month-15 years subjected to one or more surgical procedures were included in the study.

Results:

797 children were included (412 pre vs 385 post). 49.0% and 43.1% patients received a PAP during the two periods (p=0.09). The majority of patient receiving an antibiotic prophylaxis had an indication for PAP in both periods (84.6% vs 87.3%, p=0.38). After CP implementation there was an increasing of correct PAP (44.6 % vs 56.0 %; p=0.03). The selection of the appropriate antibiotic improved, both for monotherapy (81.0% vs 91.9%; p=0.02) and combination therapy (47.7 % vs 69.0%; p=0.03). Also the duration of prophylaxis decreased, with an increase of PAP discontinuation within 24 hours (41.0 % vs 61.4%; <0.001). There was no increase in treatment failures (3.9 % vs 2.6% post, p=0.3).

Conclusions:

CP with a proper educational intervention can be a useful tool to improve the choice of first-line antibiotic and the duration of PAP in surgical paediatric patients.
SCIENTIFIC SESSION V: THORACIC (PARALLEL SCIENTIFIC SESSION)

Friday June 22\textsuperscript{nd}

SCV-TH01

PATCH REPAIR FOR CONGENITAL DIAPHRAGMATIC HERNIA: A SYSTEMATIC REVIEW OF THE LITERATURE AND META-ANALYSIS

Vincenzo D Catania\textsuperscript{1,2}, Giuseppe Lauriti\textsuperscript{3}, Elke Zani-Ruttenstock\textsuperscript{1}, Mario Lima\textsuperscript{2}, Pierluigi Lelli Chiesa\textsuperscript{3}, Augusto Zani\textsuperscript{1}

\textsuperscript{1}Division of General and Thoracic Surgery, The Hospital for Sick Children, Toronto, Canada. \textsuperscript{2}Department of Pediatric Surgery, S.Orsola Malpighi University Hospital, Bologna, Italy. \textsuperscript{3}Department of Pediatric Surgery, "Spirito Santo" Hospital and "G. d'Annunzio" University, Chieti-Pescara, Pescara, Italy

Aim of the study: Different patches are used to close large congenital diaphragmatic hernia (CDH). Our aim was to compare the outcomes of commonly used absorbable (Surgisis [SIS], small intestinal submucosal patch) and non-absorbable (Gore-Tex [GTX], polytetrafluoroethylene) patches.

Methods: Using a defined strategy (PubMed, Cochrane, Embase and Web of Science MeSH headings), two investigators independently reviewed the literature from 2000 to 2017. Only studies comparing SIS versus GTX were included in the meta-analysis. Outcome measures included recurrence and small bowel obstruction (SBO). Meta-analysis was conducted using RevMan 5.3.

Results: Systematic review: Of 228 abstracts screened, 18 full-text articles were analyzed and 13 retrospective studies met our criteria. Out of 1,660 patients, 535 (32\%) underwent patch repair. The recurrence rate after primary closure was 5\% (32/644 pts, range 0-10\%), 39\% using SIS (50/127 pts, range 22-100\%), and 19\% using GTX (65/337 pts, range 0-45\%). The incidence of SBO following primary closure was 5\% (25/469 pts, range 0-9\%), 18\% using SIS (23/124 pts, range 7-35\%), and 7\% using GTX (10/149, range 4-22\%). Meta-analysis: In 5 comparative studies, SIS was associated with an increased risk of recurrence (55\%; 36/66) compared to GTX (24\%; p=0.01, OR 2.3, 95\%CI 1.2 to 4.5, I\textsuperscript{2}=60\%; Fig.1). No difference was observed for SBO following both SIS (17\%) and GTX (7\%; p=ns; Fig.2).

Conclusions: Patch repair of CDH is at increased risk of recurrence and SBO compared to primary closure. Comparing absorbable and non-absorbable patches, absorbable patches are at higher risk of recurrence in comparison to non-absorbable ones.
**Fig. 1 - Risk of hernia recurrence**

<table>
<thead>
<tr>
<th>Study or Subgroup</th>
<th>SIS</th>
<th>Events</th>
<th>Total</th>
<th>GTX</th>
<th>Events</th>
<th>Total</th>
<th>Weight</th>
<th>Odds Ratio M-H, Fixed, 95% CI</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grethel 2006</td>
<td>12</td>
<td>27</td>
<td>17</td>
<td>45</td>
<td>60.7%</td>
<td>1.32</td>
<td>[0.50, 3.47]</td>
<td>2006</td>
<td></td>
</tr>
<tr>
<td>Jancelewicz 2010</td>
<td>15</td>
<td>23</td>
<td>7</td>
<td>16</td>
<td>24.6%</td>
<td>2.41</td>
<td>[0.65, 8.92]</td>
<td>2010</td>
<td></td>
</tr>
<tr>
<td>Jawaid 2013</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>35</td>
<td>0.1%</td>
<td>355.00</td>
<td>[5.74, 21949.77]</td>
<td>2013</td>
<td></td>
</tr>
<tr>
<td>Jancelewicz 2013</td>
<td>7</td>
<td>14</td>
<td>4</td>
<td>19</td>
<td>14.5%</td>
<td>3.75</td>
<td>[0.82, 17.17]</td>
<td>2013</td>
<td></td>
</tr>
<tr>
<td><strong>Total (95% CI)</strong></td>
<td>66</td>
<td>115</td>
<td>100.0%</td>
<td></td>
<td></td>
<td>2.33</td>
<td>[1.22, 4.46]</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Total events 36 28
Heterogeneity: Chi² = 7.41, df = 3 (P = 0.06); I² = 60%
Test for overall effect: Z = 2.55 (P = 0.01)

**Fig. 2 – Risk of small bowel obstruction**

<table>
<thead>
<tr>
<th>Study or Subgroup</th>
<th>SIS</th>
<th>Events</th>
<th>Total</th>
<th>GTX</th>
<th>Events</th>
<th>Total</th>
<th>Weight</th>
<th>Odds Ratio M-H, Fixed, 95% CI</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grethel 2006</td>
<td>2</td>
<td>27</td>
<td>2</td>
<td>45</td>
<td>32.0%</td>
<td>1.72</td>
<td>[0.23, 12.98]</td>
<td>2006</td>
<td></td>
</tr>
<tr>
<td>Jancelewicz 2010</td>
<td>8</td>
<td>23</td>
<td>1</td>
<td>16</td>
<td>17.7%</td>
<td>8.00</td>
<td>[0.89, 72.10]</td>
<td>2010</td>
<td></td>
</tr>
<tr>
<td>Romao 2012</td>
<td>1</td>
<td>13</td>
<td>2</td>
<td>9</td>
<td>50.3%</td>
<td>0.29</td>
<td>[0.02, 3.83]</td>
<td>2012</td>
<td></td>
</tr>
<tr>
<td><strong>Total (95% CI)</strong></td>
<td>63</td>
<td>70</td>
<td>100.0%</td>
<td></td>
<td></td>
<td>2.12</td>
<td>[0.69, 6.53]</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Total events 11 5
Heterogeneity: Chi² = 3.72, df = 2 (P = 0.16); I² = 46%
Test for overall effect: Z = 1.30 (P = 0.19)
FUNCTIONAL EVALUATION OF THE HUMAN FETAL LUNG WITH BOLD MRI: PRELIMINARY REPORT

Naziha Khen-Dunlop\textsuperscript{1,2,3}, Gihad Chalouhi\textsuperscript{3}, Daniel Balvay\textsuperscript{3}, Augustin Lecler\textsuperscript{3}, Laurence Bussieres\textsuperscript{2}, Afef Bouchouicha\textsuperscript{3}, Olivier Clement\textsuperscript{3}, Bertrand Tavitian\textsuperscript{3}, Nathalie Siauve\textsuperscript{3}, Laurent Salomon\textsuperscript{2,3}

\textbf{Background:} Congenital diaphragmatic hernia prognosis is mainly based on prenatal lung volume. But this parameter fails to appropriately predict the outcome in many cases. Blood-oxygen-level dependent effect (BOLD) is developed in adults functional imaging and placenta dysfunctions. It uses the paramagnetic properties of deoxyhemoglobin as a natural contrast agent. The aim of our study was to evaluate if a BOLD response can be detected in fetal lung.

\textbf{Methods:} From January 2014 to December 2016, 38 healthy pregnant women, followed for fetal disease with an indication for prenatal MRI, were enrolled. After a conventional scan on a clinical 1.5T MRI (normoxic period), maternal hyperoxia was induced for 5 minutes and 5 min were added for the BOLD sequence (hyperoxic period). Results are given as mean ± SEM.

\textbf{Results:} Within the 38 mothers, 3 cases (8\%) were excluded because of major artefact movements. For the placenta, the mean BOLD response decrease was 19.1 ±5.8\% (p<0.0001). No significant BOLD effect was observed for the brain nor the liver, with a mean BOLD response decrease of 3.6 ±1.3\% (p=0.64) and 2.8 ±0.9\% (p=0.23) respectively. In the lungs, a positive response was observed, with a mean BOLD response decrease of 11.1 ±3.6\% (p=0.0012).

\textbf{Conclusion:} Our results demonstrated for the first time that normal fetal lung, despite its “non-functional status”, reacts to hyperoxia with a BOLD effect. The next step will be the evaluation of fetuses with congenital diaphragmatic hernia, looking for correlations between the level of BOLD response and post-natal evolution.
SCV-TH03
THE ROLE OF CENTRALIZATION IN CONGENITAL DIAPHRAGMATIC HERNIA (CDH) – COMPARISON OF TWO INTERNATIONAL TREATMENT CENTERS

Julia van Ling1, Kim Heiwegen1, Melanie Drewett2, Ivo de Blaauw1, Michael Stanton2, Sanne Botden1
1Radboudumc, Nijmegen, Netherlands. 2Southampton University Hospital NHS Foundation Trust, Southampton, United Kingdom

Aim of study: In the Netherlands, the care for Congenital Diaphragmatic Hernia (CDH) patients is subspecialized into two expertise centers, however, in the United Kingdom (UK), the care is spread over approximately 27 units. This study compares surgical outcome between two international treatment centers with different CDH-treatment protocols.

Method: One centralized ECMO-center and one none-centralized, non-ECMO center participated in this retrospective study. Demographics, treatment characteristics and multiple outcome measures were gathered from all CDH patients between 2000–2016. Study groups were created based on the severity of the pulmonary hypertension (PPHN). Differences between the patients in both centers were analyzed.

Results: A total of 337 patients were included. The baseline characteristics were similar for both centers, except a lower Apgar-score at 1-minute in the ECMO-center (6, range 0-10; versus 5, range 0-9; p=0.03). In the ECMO-center, more patients received ECMO-support (Table 1), while the number of diagnoses of severe PPHN was comparable (41.6% versus 43.8%, p=0.73). Also, significantly more patients got surgical repair of their diaphragm in the ECMO-center (Table 1). The 1-years survival between both centers was similar (72.9% versus 73.0%, p=1.00), however, when compared for patients with none or mild-moderate PPHN the survival was lower in the non ECMO-center (96.0% versus 83.9%, p=0.008).

Conclusion: Centralization can play an essential role in the care of CDH patients nowadays. This cross-national multicenter study seems to favor centralization of the treatment of CDH and seems to particularly favor the outcome of the less severe cases of CDH.

Table 1. Treatment characteristics; expertise center versus none expertise center

<table>
<thead>
<tr>
<th>Treatment characteristics</th>
<th>Expertise center (n = 225)</th>
<th>None expertise center (n = 112)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>ECMO treatment, n (%)</td>
<td>69 (30.8)</td>
<td>3 (2.7)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Repaired patients, n (%)</td>
<td>204 (90.7)</td>
<td>87 (77.7)</td>
<td>0.002</td>
</tr>
<tr>
<td>Patch repair, n (%)</td>
<td>82 (40.2)</td>
<td>201 (23.5)</td>
<td>0.007</td>
</tr>
<tr>
<td>Abdominal patch, n (%)</td>
<td>292 (14.6)</td>
<td>221 (2.4)</td>
<td>0.002</td>
</tr>
<tr>
<td>Chest drain peri-operative, n (%)</td>
<td>1074 (56.0)</td>
<td>35 (3.7)</td>
<td>&lt; 0.001</td>
</tr>
</tbody>
</table>

Missing values: 12, 25, 33, 413, 56
<table>
<thead>
<tr>
<th></th>
<th>Expertise center (n = 225)</th>
<th>None expertise center (n = 112)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Survival, n (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30 days</td>
<td>177 (78.7)</td>
<td>87 (77.7)</td>
<td>0.89</td>
</tr>
<tr>
<td>1 year</td>
<td>164 (72.9)</td>
<td>81 (73.0)</td>
<td>1.00</td>
</tr>
<tr>
<td>Pulmonary state at 30 days of life, n (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>On room air</td>
<td>114(^1) (69.9)</td>
<td>49(^1) (67.1)</td>
<td></td>
</tr>
<tr>
<td>On oxygen supplementation</td>
<td>12(^1) (7.4)</td>
<td>12(^1) (16.4)</td>
<td>0.004</td>
</tr>
<tr>
<td>Mechanical ventilation</td>
<td>36(^1) (22.1)</td>
<td>8(^1) (11.0)</td>
<td></td>
</tr>
<tr>
<td>Surgical complications, n (%)</td>
<td>61 (29.9)</td>
<td>15(^2) (17.9)</td>
<td>0.04</td>
</tr>
<tr>
<td>Recurrent hernia of repaired patients, n (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 year</td>
<td>12 (5.9)</td>
<td>2 (2.3)</td>
<td>0.24</td>
</tr>
<tr>
<td>Length of stay in the hospital, median (range)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>All patients</td>
<td>18(^3) (0 – 329)</td>
<td>11 (1 – 254)</td>
<td>0.03</td>
</tr>
<tr>
<td>Survivors</td>
<td>21(^3) (3 – 329)</td>
<td>14 (3 – 254)</td>
<td>0.03</td>
</tr>
<tr>
<td>Number of surgeries in the first year, median (range)</td>
<td>1</td>
<td>1 (0 – 3)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Missing values: \(^1\)14, \(^2\)3, \(^3\)16
SCV-TH04
IS PATCH IN CONGENITAL DIAPHRAGMATIC HERNIA A REAL PROBLEM?

Etienne Suply1, Clare Rees1, Kate Cross1, Hesham Elagami1, Stefano Giuliani1, Simon Blackburn1, Joe Curry1, Simon Eaton2, Paolo De Coppi1,2
1Great Ormond Street Hospital, London, United Kingdom. 2University College London, London, United Kingdom

Aim of the study
This study aimed to compare the recurrence rate of congenital diaphragmatic hernia (CDH) repaired with or without a patch using a tension-free technique.

Methods
After ethical approval, all patients with postero-lateral CDH born between 2000 and 2016 were studied retrospectively. This study provides updated data and excluded patients with Morgagni-Larrey hernias, death before surgery or initially operated on in other hospitals. Recurrence was analysed with univariate analysis and binary regression, adjusting for year, sex, defect side, liver up, patch, surgical approach, absence of postero-lateral rim and follow-up length.

Main results
Over 17 years, 96 CDH were repaired without patch, 107 with patch. Gestational age, birth weight, sex, side, age at surgery and minimally-invasive approach were not different between the groups. We observed a low recurrence rate (6.4%), with no difference between groups (patch 8.4% vs. no patch 4.2% p=0.21) despite higher incidence of liver herniation (p<0.01) and absence of postero-lateral rim (p<0.01) in patients with patch. Preoperative ECMO use was also higher (p<0.01) as was mortality (p<0.05) after patch repair. Multivariate analysis showed that recurrence was higher after thoracoscopic repair compared to open (OR 12.2 [2.2-68], p<0.01); neither patch use (OR 2.3, [0.5-10.4], p=0.28) nor any other factors were significant predictors of recurrence.

Conclusion
In this large single centre series where tension-free repair was advocated, we have shown that using a patch to repair a CDH was not associated with a higher recurrence rate.

<table>
<thead>
<tr>
<th>Demographics</th>
<th>Total (n=203)</th>
<th>Patch (n=107)</th>
<th>No patch (n=96)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Males, n (%)</td>
<td>123 (60.6)</td>
<td>64 (59.8)</td>
<td>59 (61.4)</td>
<td>ns</td>
</tr>
<tr>
<td>Left side, n (%)</td>
<td>161 (79.3)</td>
<td>80 (74.8)</td>
<td>81 (84.4)</td>
<td>p=0.09</td>
</tr>
<tr>
<td>Preoperative ECMO, n (%)</td>
<td>34 (16.7)</td>
<td>31 (29.0)</td>
<td>3 (3.1)</td>
<td>p&lt;0.01</td>
</tr>
<tr>
<td>Age at surgery (days), median [range]</td>
<td>7 [0-2170]</td>
<td>10 [1-2170]</td>
<td>4.4 [0-1662]</td>
<td>ns</td>
</tr>
<tr>
<td>Surgery before 30 d, n (%)</td>
<td>170 (83.7)</td>
<td>97 (90.7)</td>
<td>73 (76.1)</td>
<td>p&lt;0.01</td>
</tr>
<tr>
<td>Mortality, n (%)</td>
<td>22 (10.8)</td>
<td>16 (15.0)</td>
<td>6 (6.2)</td>
<td>p&lt;0.05</td>
</tr>
<tr>
<td>Follow-up (months), median [range]</td>
<td>26.8 [0-185]</td>
<td>19 [0-178]</td>
<td>29.3 [0-185]</td>
<td>ns</td>
</tr>
<tr>
<td>Recurrence, n (%)</td>
<td>13 (6.4)</td>
<td>9 (8.4)</td>
<td>4 (4.2)</td>
<td>p=0.21</td>
</tr>
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<table>
<thead>
<tr>
<th>Surgery</th>
<th>Total (n=203)</th>
<th>Patch (n=107)</th>
<th>No patch (n=96)</th>
<th>p value</th>
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</thead>
<tbody>
<tr>
<td>Open surgery, n (%)</td>
<td>151 (74.4)</td>
<td>82 (76.6)</td>
<td>69 (71.9)</td>
<td>ns</td>
</tr>
<tr>
<td>Liver herniated, n (%)</td>
<td>84 (41.4)</td>
<td>62 (58.0)</td>
<td>22 (22.9)</td>
<td>p&lt;0.01</td>
</tr>
<tr>
<td>Absence of postero-lateral rim, n (%)</td>
<td>72 [35.5]</td>
<td>64 [59.8]</td>
<td>8 [8.3]</td>
<td>p&lt;0.01</td>
</tr>
</tbody>
</table>
Aim of the study: Prosthetic patches are often used for repairing the large defect of congenital diaphragmatic hernia (CDH). The aim of this study was to identify whether material of the prosthetic patches influences the recurrence rate after CDH repair.

Methods: This is a multicenter retrospective review of 446 CDH patients born from January 2011 to December 2016. Inclusion criteria were left-side CDH survivors that the initial diaphragm repair was performed with prosthetic patch closure by laparotomy. Development of recurrence were analyzed.

Main results: Four hundred forty-six CDH patients collected from 15 institutions whom 394 (88.3%) underwent surgery. Ninety-eight (22%) met the inclusion criteria and survived to discharge or 90 days, were analyzed for recurrence in the follow-up. The patients number of defect size A, B, C, D (proposed by the CDH Study group) defined at surgery were 0, 19 (19.4%), 42 (42.9%), 35 (35.7%), respectively and 2 (2%) undescibed. The material of the prosthetic patch were dichotomized as following groups; Gore-Tex (polytetrafluoroethylene: PTFE) patch group and Mesh (polyester/polypropylene: Mesh plug and Polypropylene+PTFE: Composix Mesh) patch group. Of the 87 Gore-Tex patch group survivors, 14 (16%)
developed recurrence, while 3 out of 11 (27%) had recurrence in Mesh patch group (p value=0.38, not significant).
Conclusions: There were no significant difference in the incidence of recurrence regarding the material of patch in our cohort.
FOREIGN BODY ASPIRATION IN INFANTS: ROLE OF SELF FEEDING

Gül Özyüksel, Tutku Soyer, Sule Yalcin, Saniye Ekinci, İbrahim Karnak, Arbay Özden Cifci, Feridun Cahit Tanyel
Hacettepe University Faculty of Medicine Department of Pediatric Surgery, Ankara, Turkey

**Aim:** ‘Baby led weaning’ is a popular method in which the babies are encouraged to self feeding to gain oral-motor abilities. It’s role in foreign body aspiration (FBA) is controversial. A retrospective study was performed to evaluate the results of FBA in infants and it’s relation with feeding method.

**Methods:** Children who underwent bronchoscopy for FBA for the last 10 years were evaluated. Infants (<1 year of age) were evaluated for age, sex, clinical findings and results of bronchoscopy. Infants with FBA were investigated for feeding characteristics (self feeding, caregiver-assisted feeding).

**Results:** Totally 826 patients were underwent bronchoscopy with a presumptive diagnosis of FBA. FBA was noted in 50.2% (n:417). Only 9.07% (n: 75) of cases were under 1 year of age and 67% of (n:50) them had FBA. The mean age was 9 months (5-12 months) and male:female was 18:32. 80% of cases aspirate when self feeding and 14% of them aspirate during caregiver-assisted feeding. Three cases aspirated inorganic substance. The most common aspirated foods were nuts, raw vegetables and meat. Five of the cases underwent emergent bronchoscopy because of desaturation and respiratory distress. During bronchoscopy, FBA was detected at left main bronchus in 27 cases, right main bronchus in 20 cases and trachea in 3 cases.

**Conclusion:** Self feeding to promote oral motor functions may cause FBA in infants. Emergent bronchoscopy is more common in infants and reveals aspiration of foods that can not be consumed safely in this age group.
THE PROGNOSTIC VALUE OF PRENATALLY DETECTED SMALL OR ABSENT FOETAL STOMACH WITH PARTICULAR REFERENCE TO OESOPHAEGAL ATRESIA

Lucinda Tullie¹, David Howe², Nigel Hall¹,³, Melanie Drewett¹, David Burge¹, Diana Wellesley⁴
¹Department of Paediatric Surgery and Urology, Southampton Children’s Hospital, Southampton, United Kingdom. ²Department of Maternal and Fetal Medicine, University Hospital Southampton, Southampton, United Kingdom. ³Faculty of Medicine, University of Southampton, Southampton, United Kingdom. ⁴Wessex Clinical Genetics Service, University Hospital Southampton, Southampton, United Kingdom

Aim: Many prenatally detected anomalies are well understood but some findings have less certain implications. We report outcomes of pregnancies with a prenatally detected small or absent stomach.

Methods: Institutionally approved retrospective review of a regional foetal anomaly register over 22 years (1994-2015). Data were collected for pregnancies with a small or absent foetal stomach on second trimester scan and correlated with postnatal outcomes.

Results: A small or absent stomach was detected in 78 pregnancies. In 47 of these, this was the only prenatally detected anomaly. Eighteen of these had concurrent 3rd trimester polyhydramnios.

In 28 with an isolated absent stomach, 10 pregnancies had 3rd trimester polyhydramnios and all had significant upper gastrointestinal pathology (9 oesophageal atresia [OA] +/-trache-oesophageal fistula [TOF] and 1 microgastria). In the remaining 18 cases without polyhydramnios, 12 had a chromosomal or structural abnormality and 6 had no abnormality. Of 19 with an isolated small stomach, just one case (with polyhydramnios) had OA+/-TOF.

In 31 pregnancies, absent (22) or small (9) stomach was detected alongside other anomalies. Six had OA+/-TOF with other associated anomalies, 7 had chromosomal abnormality, 17 had a variety of other structural anomalies, and 1 no abnormality.

Conclusion: All cases of small or absent stomach should be offered prenatal chromosomal testing and 1 in 5 will have OA+/-TOF. An isolated absent stomach, particularly in conjunction with polyhydramnios, strongly predicts foetal pathology. Cases of absent stomach with polyhydramnios should be delivered in a paediatric surgical centre.
SCV-TH08
RIGHT AND LEFT DIAPHRAGMATIC HERNIA: MORTALITY AND MORBIDITY COMPARED

Pinton Anne¹, Sica Marina², Sananes Nicolas¹, Favre Romain¹, Benachi Alexandra¹, Boubnova Julia¹, Lehn Anne², Lachkar Amane², Alshammari Deidhan², Becmeur Francoise², Schneider Anne²
¹Gynecology et obstetrics, Strasbourg, France. ²Pediatric Surgery, Strasbourg, France

Aim of the study

The objective of this study was to evaluate the mortality of right congenital diaphragmatic hernia (CDH) in comparison to left CDH.

Methods

This is a retrospective cohort study of pregnancies with CDH from 2008 to 2016 recorded in the database of the French National Center for Rare Diseases. The primary outcome was survival to discharge. We made a comparative study between right and left CDH by univariate and multivariate analysis. Terminations and stillbirths were excluded from analyses of neonatal outcomes.

Main Results

A total of 506 CDH were included, 67 (13%) right and 439 left side (87%) CDH. Rate of survival was respectively 49% right and 74% left CDH (p<0.01). Multivariate analysis showed two significantly factors associated to mortality: thoracic herniation of liver (OR 2.27; IC 95% [1.07-4.76]; p=0.03) and lung-to-head-ratio (OR 2.99; IC 95% [1.41-6.36]; p<0.01). Side of CDH was not significantly associated to mortality (OR 1.87; IC 95% [0.61-5.51], p=0.26). No significant differences were found between the two groups as regards eating disorders, non-invasive ventilation, pulmonary arterial hypertension. The only difference is complementary surgery necessary in the right-side hernia group.

Conclusions

Rate of right CDH mortality is more important than left CDH. Nevertheless after adjusting by lung-to-head-ratio and thoracic herniation of liver, right CDH has not more risk of mortality than left CDH.
CORRECTION OF FUNNEL CHEST USING SELF DESIGNED SINGLE-COMPONENT T-SHAPED METAL BAR WITH EXTRA PLEURAL PLACEMENT UNDER CONTROL OF SUBXIFOIDAL WINDOW

Danylo Krivchenya¹, Vasyl Prytula¹, Anatolii Levytskyi¹, Oleksii Bebeshko², Eugene Rudenko¹, Michael Silchenko², Faizullah Hussaini¹, Valentyn Rogozinskiy¹

¹Bogomolets National Medical University, Kyiv, Ukraine. ²National pediatric specialized hospital Ohmatdyt, Kyiv, Ukraine

Aim: to optimize the minimally invasive treatment of Funnel chest (FC) using a self designed single-component T-shaped metal bar.

Methods: From 2001-2017 we treated various variants of FC in 168 children aged 6 months to 18 years with technique of Nuss. T-shaped bar with a leading wedge-shaped end was used in 121 and Lorentz-bar in 33, both bars at the same time in 10, and in stages in 4. A subxifoidal window was used for controlling the safety of mediastinum and mobilization of the sternum.

Since 2011, element of the Ravich was added to our modification the dissection of the anterior plate of the sternum over its distortion from the partial horizontal incision in 20 patients. FC was corrected simultaneously with other anomalies in 42 patients, from which cardiac defects in 16, lung in 7, esophagus in 1, and diaphragm in 18.

Main results: Positive functional and cosmetic result was achieved in 97% of patients. With no deaths and severe life-threatening complications. We had hemothorax in 2, reactive pericarditis in 1, suppuration of the bar bed in 1, intraoperative pneumothorax in 22, horizontal displacement of the bar in 1, dislodging of bar in 3, relapse of the FC in 2 and hypercorrection in 2.

Conclusion: The placement our T-shaped bar with our modification allows safe installation without pneumothorax, or guiding instruments. Operation was significantly simplified for prevention of intense pain, displacement or dislodging of bar and relapse of FC after bar removal. The T-shaped bar is removed from single incision.
Asymptomatic CPAM and Malignancy Risk: CPAM-Derived Mesenchymal Stem Cells Study to Support Surgical Treatment

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¹Pediatric Surgery Unit, Children’s Hospital, Istituto Mediterraneo di Eccellenza Pediatrica, Palermo, Italy.
²Immunology and Transplantation Laboratory/Cell Factory/Pediatric Hematology/Oncology Department, Pavia, Italy.
³Pediatric Anesthesiology and Intensive Care Unit, Children’s Hospital, Istituto Mediterraneo di Eccellenza Pediatrica, Palermo, Italy.
⁴Pediatric Unit, University of Pavia and Fond. IRCCS Policlinico S. Matteo, Pavia, Italy.

Aim of the study
The association between congenital pulmonary airway malformations (CPAM) and malignancy is reported in the literature. Interactions between the tumor, immune, and mesenchymal stromal/stem cells (MSCs) have been recognized as crucial for understanding tumorigenesis. We characterized MSCs isolated from CPAM lesions in order to define potential malignancy risks.

Methods
CPAM pulmonary tissue (type I, II and CLO) was used for MSC expansion. Morphology, immunophenotype, differentiation and immunological capacity, proliferative growth, gene signature telomerase activity and in vivo tumorigenicity in nude mice were evaluated.

Main results
MSCs were successfully isolated and propagated from CPAM tissue. CPAM-MSCs presented the typical MSC morphology and phenotype, while exhibiting high proliferative capacity, reaching confluence at a median time of 5 days as well as differentiation capabilities. CPAM-MSCs at early passages were not neoplastic and chromosomally normal, even though unbalanced chromosomal rearrangements were noted by molecular karyotype.

Conclusions
CPAM-MSCs exhibited specific features similar to tumor derived MSCs. Whilst there was no evidence of malignant transformation in the cystic tissue, our results provide evidence that this abnormal tissue has malignant potential. MSCs are considered important players in the tumor microenvironment and they have been closely linked to regulation of tumor survival, growth and progression. Thus, early lesion resection also in asymptomatic patients might be indicated to exclude that the microenvironment may be potentially permissive to cancer development.
SCV-TH11
PREDICTORS OF LONG-TERM PULMONARY MORBIDITY IN CHILDREN WITH CONGENITAL DIAPHRAGMATIC HERNIA (CDH)

Robin Wigen\(^1\), Wenming Duan\(^1\), Monping Chiang\(^1\), Theo Moraes\(^1\), Priscilla Chiu\(^1\)\(^2\)
\(^1\)The Hospital for Sick Children, Toronto, Canada. \(^2\)Department of Surgery, University of Toronto, Toronto, Canada

**Purpose:** The aim is to identify prognostic markers of long-term pulmonary morbidity among CDH survivors.

**Methods:** A single institution, retrospective review was performed on all CDH patients from 2000 and 2012 (REB#1000053383). Liver position, patch USE and pulmonary function tests (PFT) (forced expiratory volume at 1 second [FEV\(_1\)] and forced vital capacity [FVC] expressed as mean % predicted ±SD) were recorded. Data were analyzed using ANOVA.

**Results:** Patients with acceptable and reproducible PFT (n=72 for 202 total PFT) with patch repair and liver down (n=40) had significantly lower FEV\(_1\) (62.9±17.6) than those with no patch and liver down (n=98, FEV\(_1\)=85.2±15.9, \(p<0.0001\)) or liver up (n=36, FEV\(_1\)=79.3±14.4, \(p<0.0001\)). Patients with patch repair and liver up (n=28) also had significantly lower FEV\(_1\) (71.9±14.4) than those with liver down and no patch (\(p=0.0006\)). Patients with liver down and patch repair had PFT results consistent with moderate reduction of lung function while the remainder had mild to no decrease in lung function. All CDH patients older than 14 years of age had a reduction in FEV\(_1\)/FVC consistent with obstructive phenotype, with a mean FEV\(_1\)/FVC=62.3 for patch repair group and FEV\(_1\)/FVC=76.1 in the no patch group.

**Conclusion:** Decreased pulmonary function of CDH survivors correlated with use of patch repair and liver position. CDH lung disease should be monitored into adulthood.

**Fig. 1-** PFT showed decreasing FEV\(_1\) with increasing age for CDH patients

A. No patch repair (left panel) vs. patch repair.
B. Liver down (left) vs liver up
SCIENTIFIC SESSION V: THORACIC (PARALLEL SCIENTIFIC SESSION)  
Friday June 22nd

SCV-TH12
LONG-TERM PREVALENCE, CUMULATIVE HAZARD AND PREDICTIVE FACTORS OF INTESTINAL AND GASTRIC METAPLASIA AFTER REPAIR OF OESOPHAGEAL ATRESIA

Koivusalo Antti, Sistonen Saara, Rintala Risto, Pakarinen Mikko
Children’s Hospital, Helsinki, Finland

Aim of the study: We assessed the long-term hazard and predicting factors of gastric and intestinal metaplasia (GM and IM) in oesophageal atresia (OA).

Patients and methods: After ethical consent we reviewed 1289 surveillance endoscopies in 361 OA patients from 1949 to 2016 (type A n=30, B n=6, C n=290, D n=15, E n=17, F n=3). Age, long-gap (A or B) (n=36), primary anastomosis (n=322), fundoplication (n=86), oesophagitis and anastomotic complications (n=66) were tested as predictive factors.

Main Results: During a median follow-up of 28(IQR13–41) years, 3(1–5) endoscopies/patient, we found metaplasia in 58 (16%) patients, IM in 10(3%), type A (3;10%) and C (7;2.4%), and GM in 54(15%). Detection of IM occurred after a median of 24(16–44) years and of GM after 17(7.3–35) years,(p=0.13). At the latest endoscopy metaplasia was found in 49(14%) patients, IM in 7 (2%) type A(1; 3%), C(6; 2.1%) and GM in 44(12%), type A10%), B(17%), C(12%), D(13%), E(6%) and F(0%), 91(25%) had oesophagitis. Cumulative hazard for IM and GM increased two and three fold by the age of 50. Anastomotic complications and fundoplication predicted GM. (Table 1 and Figure1)

Conclusions: Cumulative hazard for IM and GM increased with age. After 28 years cumulative incidence of IM and GM reached 3% and 15% with 2% and 12% point prevalence. Fundoplication and anastomotic complications predicted GM.

Table 1 Independent risk factors for gastric metaplasia (GM) and intestinal metaplasia (IM)

<table>
<thead>
<tr>
<th></th>
<th>All Metaplasia</th>
<th>IM</th>
<th>GM</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>RR (95% CI)</td>
<td>p</td>
<td>RR (95% CI)</td>
</tr>
<tr>
<td>Age</td>
<td>1.03 (1.10–1.05)</td>
<td>0.008</td>
<td>1.1 (1.0–1.1)</td>
</tr>
<tr>
<td>Long Gap</td>
<td>1.6 (0.6–3.8)</td>
<td>0.32</td>
<td>1.3 (0.2–11)</td>
</tr>
<tr>
<td>Primary anastomosis</td>
<td>0.6 (0.3–2.0)</td>
<td>0.78</td>
<td>0.7 (0.1–6.1)</td>
</tr>
<tr>
<td>Fundoplication</td>
<td>1.7 (0.9–3.3)</td>
<td>0.12</td>
<td>2.4 (0.5–11)</td>
</tr>
<tr>
<td>Anastomotic complications</td>
<td>1.7 (0.8–3.5)</td>
<td>0.15</td>
<td>0.7 (0.1–5.8)</td>
</tr>
<tr>
<td>Oesophagitis</td>
<td>1.9 (01.0–3.7)</td>
<td>0.05</td>
<td>4.1 (0.9–19)</td>
</tr>
</tbody>
</table>
Figure 1 Cumulative hazard of gastric (GM) and intestinal metaplasia (IM)
Mélodie JURICIC¹, Thomas PRUDHOMME¹, Emmanuelle GODEAU², Isabelle CLAUDET¹, Ourdia BOUALI¹
¹HOPITAL DES ENFANTS, TOULOUSE, France. ²RECTORAT DE L’ACADEMIE DE TOULOUSE, TOULOUSE, France

Aim of the study

To evaluate urinary continence prevalence in schoolchildren population by examination of their perceptions of school toilets and their bladder habits.

Methods

We performed a transversal descriptive study, with a survey (20 questions), distributed to parents in 5 schools: 3 elementary schools (ESch) and 2 secondary schools (SSch) in the same region, with school directors and parents consents. We obtained University Rector autorization, CNIL agreement and positive statement of the Ethical Comity of University Hospital. Answers content was grouped and analyzed thematically. Statistical analysis was made using Chi-test.

Main results

With a mean response rate of 54.6% (68% in Esch, 34% in SSch), 943 children aged 6 to 15 years old were included (sex ratio 1/1). Half of them were in ESch. In Esch, 17% of children reported never using school toilets, whereas this rate was 34% in SSch, mostly due to the fear to be seen (76%) and uncleanliness (64.2%). Overall, 50% and 41.6% supported an unavailibility of toilets or paper respectively.

Urinary tract disorders as incontinence, pyelonephritis, cystisis, nocturnal enuresis and genital inflammation were present in 7.42% and constipation affected 10.2% of schoolchildren in this study. Urinary or fecal elimination problems were related to toilets access conditions for 40.7% parents.

Conclusions

Many children influenced by negative perceptions of school toilets have adopted unhealthy toilet habits, factor conductive to incontinence, UTI and constipation. This public health problem causes physical, mental, scholar and social consequences. Individual and collective preventive measures have to be made.
SCVI-UR02

DOES PRENATAL URINARY TRACT OBSTRUCTION INFLUENCE THE LEVEL OF BIOMARKERS IN NEWBORN SCREENING?

Live Lundar, Lars Mørkrid, Alexander Rowe, Trine Tangaas, Ingjerd Sæves, Rolf Pettersen, Gunnar Aksnes, Ragnhild Emblem
Oslo University Hospital, OSLO, Norway

Aim of the study
Biomarkers of renal disease in dry spot-blood samples is largely unexplored. We aimed to map the pattern of screening biomarkers in neonates with posterior urethral valves (PUV).

Methods
We included 21 PUV-patients born 2012-2016. The national newborn screening program (NBS) analyzes disease markers in dry spot blood collected 2 days after birth. Results from 37 biochemical analyzes (2 hormones, 2 enzymes, 7 amino acids, 29 acylcarnitines and 1 organic acid) were compared with data from the entire neonatal population = reference.

Main results
Z-score means of 12 of 37 (32.4%) markers showed P-values < 0.05.

<table>
<thead>
<tr>
<th>Marker</th>
<th>Mean</th>
<th>SD</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>alanine</td>
<td>1.403</td>
<td>1.544</td>
<td>0.000</td>
</tr>
<tr>
<td>C160 H-carn.</td>
<td>0.698</td>
<td>0.896</td>
<td>0.002</td>
</tr>
<tr>
<td>leucine</td>
<td>0.936</td>
<td>1.207</td>
<td>0.008</td>
</tr>
<tr>
<td>OH-pro</td>
<td>1.393</td>
<td>1.832</td>
<td>0.017</td>
</tr>
<tr>
<td>C180 H-carn.</td>
<td>0.083</td>
<td>1.199</td>
<td>1.30</td>
</tr>
<tr>
<td>tyrosine</td>
<td>0.747</td>
<td>1.129</td>
<td>4.02</td>
</tr>
<tr>
<td>C18:2 carn.</td>
<td>0.822</td>
<td>1.268</td>
<td>0.986</td>
</tr>
<tr>
<td>phenylalanine</td>
<td>0.837</td>
<td>1.478</td>
<td>0.925</td>
</tr>
<tr>
<td>C10-car.</td>
<td>0.936</td>
<td>1.30</td>
<td>1.333</td>
</tr>
<tr>
<td>C12-car.</td>
<td>-0.71</td>
<td>0.83</td>
<td>0.040</td>
</tr>
<tr>
<td>C3-car.</td>
<td>0.537</td>
<td>4.02</td>
<td></td>
</tr>
<tr>
<td>C5-car.</td>
<td>0.496</td>
<td>0.925</td>
<td></td>
</tr>
<tr>
<td>C12:1-car.</td>
<td>-0.639</td>
<td>1.333</td>
<td></td>
</tr>
</tbody>
</table>

By combining markers, either by ratios or principal component analysis more sensitive and specific variables for PUV may be obtained: e.g. the ratio phenylalanine/tyrosine has a z-score mean=1.385 (SD=1.547), P=0.0006.

Conclusions
PUV obviously affects disease markers in NBS. To which extent these new unexpected findings constitute biochemical fingerprints specific of urinary tract obstruction must be confirmed in a larger cohort of patients. Post-analytical tools that utilizes others combinations of markers may aid in the identification of cases with high sensitivity and acceptable specificity.
**SCVI-UR03**

**CHILDREN WITH CLOACAL ANOMALIES ARE BORN WITH SIGNIFICANTLY REDUCED TOTAL RENAL VOLUME**

Molly Fuchs¹, Yuri Sebastiao¹, Laura Weaver¹, Benjamin Thompson¹, Kristina Booth¹, Alejandra Vilanova-Sanchez², Daniel Dajusta¹, Christina Ching¹, Richard Wood³, Venkata Jayanthi³, Marc Levitt¹  
¹Nationwide Children’s Hospital, Columbus, USA. ²Hospital Universitario La Paz, Madrid, Spain

Aim of the study: To determine if children with cloacal anomalies have reduced renal volume at birth compared to controls.

Methods: An IRB approved database was reviewed. All children with cloacal anomaly with renal ultrasound (RUS) in the first 3 months of life were included. Horseshoe and cross-fused ectopic kidneys were excluded due to complexity of volume measurement. We identified a control group that underwent normal screening RUS for ear tags or 2-vessel cord. The earliest RUS was reviewed and renal volume measured.

Main results: 30 cloacas were compared to 52 controls, all age < 3 months. Cloacas had significantly reduced total renal volume compared to controls (table 1). Cloacas had reduced volume of each renal unit compared to controls but this only approached significance (table 2). Solitary kidneys were not significantly different than controls.

Conclusions: Children with cloacal anomalies have significantly reduced total renal volume from the time of birth compared to controls. Assuming renal volume represents glomerular mass, inherent difference in renal volume may predispose children with cloaca to renal insufficiency.

**Table 1: Total renal volume (cm³)**

<table>
<thead>
<tr>
<th></th>
<th>Cloaca</th>
<th>Controls</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>n (patients)*</td>
<td>Mean (SD)</td>
<td>Median (IQR)</td>
<td>n (patients)</td>
</tr>
<tr>
<td>23</td>
<td>19.42 (6.22)</td>
<td>19.50 (14.66-22.61)</td>
<td>52</td>
</tr>
</tbody>
</table>

* excluded patients with solitary kidney

**Table 2: individual renal volumes (cm³)**

<table>
<thead>
<tr>
<th></th>
<th>Solitary kidney</th>
<th>n (renal units)</th>
<th>Age-adjusted mean</th>
<th>Age-adjusted difference from controls (95% CI)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cloaca</td>
<td>Yes</td>
<td>7</td>
<td>8.85</td>
<td>-1.26 (-4.71, 4.38)</td>
<td>0.6</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>46</td>
<td>8.73</td>
<td>-1.38 (-2.64, 0.10)</td>
<td>0.07</td>
</tr>
<tr>
<td>Control</td>
<td></td>
<td>104</td>
<td>10.11</td>
<td>Reference</td>
<td></td>
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</table>
USEFULNESS OF ULTRASOUND MEASUREMENTS TO PREDICT SUCCESS IN THE POSTOPERATIVE EVOLUTION OF THE URETEROPELVIC JUNCTION OBSTRUCTION IN CHILDREN

Laura Pérez-Egido, Alberto Parente, Laura Burgos, Ruben Ortiz, Juan Carlos De Agustín, Jose María Angulo
Gregorio Marañón University Hospital, Madrid, Spain

During laparoscopic pyeloplasty removal of the ureteropelvic junction inevitably results in an anatomical modification of the pelvis, but endourological procedures do not allow modification of the pelvis. We evaluated postoperative ultrasound (US) parameters of the ureteropelvic junction obstruction (UPJO) treated endourologically as success markers.

Methods: This retrospective study included all patients <18 months old who underwent endourological intervention (high-pressure balloon dilatation) between 2007-2016. Preoperative and postoperative US (3, 6 and 12 months), anteroposterior diameter (APD), pelvis to cortex ratio (PCR) and percent improvement in APD (PI-APD) were evaluated.

Results: A total of 51 patients were included. The reduction in APD and PCR were significant at 3 months, 6 months and 1 year (P<0.05). Statistically significant AUC was found for 3 months postoperative APD and PCR. Good prognosis cutoff values were calculated and no patient with APD 19mm or less or/and postoperative PCR3.3 or less underwent re-intervention (S:100%E:70%). Statistically significant AUC was found for 6 months postoperative APD, PCR and PI-APD. Good prognosis cutoff values were calculated and patients with APD 18.5 or more, RPC 3.1 or more and PI-APD 35% or less constitute a group of high risk of re-intervention (S:85%E:90%).

Conclusion: Our study supports that combination of US measurements in the postoperative follow-up of UPJO can be used to identify an early low risk of recurrence group, avoiding the postoperative renogram. Also, after 6 months postoperative, US can detecte a high risk of recurrence group who needs further investigation as a renogram.
LONG-TERM UROLOGICAL OUTCOMES IN CHILDREN WITH HIRSCHSPRUNG DISEASE

Colin Brook¹, Aurore Bouty¹, Lucy Collins², Shreya Sood¹, Rija Khanal¹, John Hutson¹,²,³, Mike O'Brien¹, Warwick Teague¹,²,³, Yves Heloury¹, Sebastian King¹,²,³
¹Royal Children’s Hospital, Melbourne, Australia. ²Department Of Paediatrics, University Of Melbourne, Melbourne, Australia. ³F. Douglas Stephens Surgical Group, Murdoch Children’s Research Institute, Melbourne, Australia

Aim of the Study
The urological outcomes for children following pull-through surgery for Hirschsprung Disease (HD) are poorly understood. We aimed to study these outcomes using validated questionnaires in patients at a large tertiary referral centre.

Methods
All HD patients aged between 5 – 18 years, managed at our tertiary referral centre, were identified. We utilised the Vancouver Dysfunctional Elimination Syndrome Survey (VDESS) for patients aged 5 –10 years, and the urological parameters of the Fecal Incontinence and Constipation Quality of Life (FICQOL) questionnaire for patients aged 11-18 years.

Main Results
A total of 38 patients [M:F 31:7; median age 8.1 years (5.0-10.7)] completed the VDESS (43% participation), whilst 58 patients [M:F 49:9; median age 14.9 years (11.1-18.7)] completed the FICQOL questionnaire (73% participation). 64% of patients had short segment disease, and the Soave pull-through was the most common procedure (82%).

One in eight patients aged 5-10 years were wet during the day, ranging from almost dry (2.6%) to soaked (5.2%); 31% experiencing nocturnal enuresis more than twice a week. Almost half the patients void 5-6 times daily. 13% have urgency every day.

In patients aged 11 – 18 years, 6% were found to have issues with erections. In this older cohort, 12% had had previous urinary tract infections, and 5% experienced daily urinary incontinence. None required catheterisations.

Conclusions
Significant urological morbidity exists following pull-through for HD. Appreciation of these urological issues is important in the long-term follow-up of HD patients.

Ethics
HREC 36003A Human Research Ethics Committee
HYPOSPADIAS AND THE USE OF THE AGES AND STAGES QUESTIONNAIRE TO EVALUATE NEURODEVELOPMENTAL DEVELOPMENT

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Introduction: Placental insufficiency is one of the possible reasons in the etiology of hypospadias. The studies have shown that lower testosterone levels at gestation cause fetal growth retardation and genital abnormalities. Placenta-related intrauterine growth restriction may also result in neurodevelopmental impairment. We aimed to evaluate the impairment of neurodevelopmental status of the children with hypospadias.

Material and Methods: The children were divided into two groups as hypospadias (distal-proximal) and traditional circumcision group. ASQ and ASQ-SE(Ages and Stages Questionnaire; communication, gross motor, fine motor, problem solving, personal-social skills) tests which are validated and extensively used as a screening tool for detecting impaired neurodevelopment were applied to children.

Results: 137 children were enrolled to the study. Seventy-eight of them had hypospadias (27 proximal, 51 distal) and 59 of them were admitted for traditional circumcision. The mean age of the patients was 25.6 months (hypospadias group: 25.5, circumcision group: 25.8). The hypospadias group had statistically significant impaired points than circumcision group in communication, gross motor and personal-social skills (p<0.05). In fine motor skills, hypospadias group had also impaired points but this was not significant statistically (p=0.053).

Conclusion: In the present study it is demonstrated that children with hypospadias have impaired neurodevelopmental status at communication, gross motor and personal-social skills. It should be kept in mind that the follow up of children who were operated due to hypospadias should not only include surgical worries but also neurodevelopmental issues. Evaluation and early guidance to related departments when needed may result in better neurodevelopmental status for these children.
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**Aim of study:** Urethrocutaneous fistulas are the most common late complication of hypospadias repair. It seems that one of the most important factors in reducing the likelihood of fistula formation is the use of an intermediate layer of healthy tissue between the neo urethra and the skin. In this study, we tried to introduce a new formulation of glue made up from cryoprecipitate, calcium gluconate and Packed RBC.

**Methods:** From April 2014 to January 2016, patients with distal hypospadias that referred for operation were selected. Hypospadias repair performed by tabularized incised plate urethroplasty. Four hundred cases of distal penile hypospadias were selected for study (300 cases vs 100 controls). Three hundred cases underwent TIPS urethroplasty with application of cryocalcium glue. The control group underwent the same procedure without application of glue by the same surgeon. All patients were followed 2 weeks of surgery and then 3 monthly. The range of follow up time was 6 to 30 months.

**Main Results:** The mean age of patients was 13.03±8.34 in case group and 13.59±7.48 months for control group (P=NS). The mean length of reconstructed urethra was 18.12±4.18 mm in case group and 16.17±3.74 mm in control group (P=NS). Urethrocutaneous fistula was occurred in 5 patients of case group and 6 patients in control group (1.6% vs 6%, P=0.001).

**Conclusion:** Cryocalcium glue used to reinforce urethroplasty anastomosis appears to be a safe and inexpensive alternative for fibrin to reduce the risk of post-operative fistula formation.
18 YEARS OF NUTCRACKER SYNDROME: EXPERIENCE AT A TERTIARY REFERRAL HOSPITAL

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AIM OF THE STUDY
Nutcracker Syndrome (NS) defines an entity generated by the compression of the left renal vein that associates development of renal venous hypertension, which transmitted in a retrograde direction may cause hematuria, proteinuria and varicocele. We present our experience with NS in the last 18 years.

METHODS
Retrospective review of the patients followed in our center with diagnosis of NS based on clinical and imaging tests (ultrasound, CT/MRI, phlebography).

MAIN RESULTS
21 patients, mean follow-up 52.3 months (37.1-67.5). Mean age of presentation was 11.7 years (9.9-13.4). Most frequent presentation: hematuria 16 patients (76.2%), macroscopic in 75% and in 42.9% related to physical exercise. Other symptoms: left varicocele 7 patients (33%), proteinuria 6 patients (21.6%). Mild-moderate cases received conservative treatment (change of physical activity, postural hygiene), which was effective in 16(76.2%). In the remaining 5 cases (23.8%), 2(9.5%) required endovascular procedures (intravascular stent in the renal vein in 1 case, and embolization of the spermatic vein in 1 case), in 1(4.8%) of the patients, transposition of the renal vein and renal autotransplantation was performed, and 2(9.5%) of the patients with mild cases required surgical correction of the varicocele.

CONCLUSIONS
Hematuria, usually macroscopic and related to physical exercise, is the most frequent symptom in patients with NS. Diagnosis is based on non-invasive tests; phlebography should be reserved for severe cases that require an interventionist attitude. Endovascular or surgical treatment is indicated in severe cases with failure of conservative treatment.
SCVI-UR09

RISK FACTORS FOR DEVELOPING HYPOSPADIAS AND CRYPTORCHIDISM: A CASE-CONTROL STUDY

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Aim of the study: To investigate the association between endocrine disrupting chemicals (EDC) exposure and paternal factors in the etiology of hipospadias and cryptorchidism.

Methods: A case-control study. Cases were infants between 6 months and 14 years of age with hypospadias or cryptorchidism attended in our hospital over a period of 18 months. Controls were infants with the same range of age without any urological problem. Data on parental occupational exposures and other socio-demographic variables were collected by face-to-face interviews. Logistic regressions were used for the analysis.

Results: 420 patients were studied, 210 cases (107 hypospadias/103 cryptorchidism) and 210 controls (mean age=3.37±2.64 [range 0.5-12]). Birth weight was significantly lower in hypospadias group (median 2.92±0.67kg versus 3.36±0.59Kg in controls; p=0.015). Both maternal and paternal age were significantly higher in cases (median 33.28±5.14 versus 32.25±5.08 years; p=0.04 and 36.25±6.28 versus 34.74±5.77 years; p=0.01, respectively). Significant differences were observed between cases and controls regarding maternal and paternal occupational exposure to EDC, adjusted odds ratios (OR) were 3.19 [95% confidence interval (CI): 1.48-6.88] and 1.89 [95% CI: 1.08-3.28] respectively. An increased risk in fathers with urological problems was observed in cases (OR= 2.17 [95% CI: 1.01-4.69]). In hypospadias group, an increased risk in smoking fathers was also observed (OR=2.10 [95% CI: 1.25-3.53]). Antiabortive drugs (OR=2.17 [95% CI: 1.01-4.69]) and father’s high educational level (OR=2.17 [95% CI: 1.01-4.69]) appeared to be protecting factors.

Conclusions: Exposure to EDC could increase the risk of hypospadias and cryptorchidism.
SCVI-UR10
AGE AT ORCHIDOPEXY ...DOES IT REALLY MATTER? A SYSTEMATIC REVIEW

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Aim of the study

To assess the ideal timing for orchidopexy with respect to fertility using different measures.

Methods

We performed a critical review of the literature published between 2004 and 2017 using MEDLINE, EMBASE and Pre-MEDLINE databases. The search strategy used the terms ‘orchidopexy’ or ‘cryptorchidism’ in combination with ‘age’ or ‘timing’.

87 papers were found. Studies included were randomized and/or controlled trials, case-control studies, and cohort studies. 12 articles were found

Main results

Articles used different measures to estimate fertility due to the difficulty in assessing this outcome. It included testicular growth (7), histology (3) and semen analysis in adulthood (2). For the studies that estimated fertility potential based on testicular volume, the overall consensus was that orchidopexy performed less than one year resulted in greater testicular growth, although long term volumes were significantly less than the normative values at all ages.

Three studies reported on testicular histology at the time of orchidopexy with various outcomes examined to determine fertility potential (seminiferous tubule diameter, germ cells per tubule). It showed that histological parameters in patients aged less than one year were significantly higher than in patients aged more than one year.

In the two studies reporting semen analysis, sperm count were significantly increased when orchidopexy was performed at one year.

Conclusion

There is a paucity of higher-quality studies investigating the effect of age at orchidopexy on fertility but orchidopexy performed before one year can optimize fertility.
SCVI-UR11
INDICATORS FOR OUTCOMES IN THE LAST DECADE CONCERNING LAPAROSCOPIC FOWLER-STEPHENS

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Aim of the Study: This study analyzed indicators for outcomes in undescended testes (UDT) managed with laparoscopic 2-stage Fowler-Stephens orchidopexy (LFSO) in the past decade.

Methods: Literature was searched on Pubmed® (2007-2017) using terms “2 step orchidopexy”, “Fowler-Stephens”, “undescended testis” and “laparoscopic”.

Main Results: Twenty reports of LFSO with 706 testes were analyzed that included n=145 (20%) bilateral UDT. Testicular vessels were divided by clipping+/-division (13 reports; n=367 testes), diathermy (2 reports; n=56 testes), ligation (2 reports; n=25 testes) and not specified (5 reports; n=258 testes). Time interval for 2nd stage was 2-19 months. At 2nd-stage, the testis was channeled through a medial neohiatus (17 reports, n=631 patients), or internal ring preserving collateral vessels within gubernaculum (4 reports; n=75 patients). Outcomes were successful in n=591 (85%). In bilateral UDT, testicular atrophy (TA) was not observed after (6 reports; n=71 patients) synchronous approach, however in 3 (5 reports; n=43 patients) after metachronous approach (p=0.05). Orchidectomy for TA after 1st stage was n=9 (1.25%). Re-ascent after orchidopexy was in n=27 (3.8%). TA was observed in 16/367 after clipping and 6/56 after diathermy (N.S.) Successful outcomes were in 72/75 (96%) gubernaculum sparing LFSO, versus 535/631 (84.8%) non-gubernaculum sparing (p=0.05).

Conclusions: In the past decade, LFSO has 85% success. TA after 1st stage LFSO was 1.25% and 2nd stage was 15%. TA is independent of the type of testicular vessel ligation technique. However, TA is significant in non-gubernaculum saving procedures and when metachronous orchidopexy is performed for bilateral UDT.
SCIENTIFIC SESSION VI: UROLOGY (PARALLEL SCIENTIFIC SESSION)  Friday June 22nd

SCVI-UR12
PARENTAL HOME REMOVAL OF URETHRAL CATHETERS FOLLOWING UROLOGICAL SURGERY – A PROSPECTIVE ONE YEAR AUDIT

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Aim of the study

Many urological operations require placement of a urethral Foley catheter which needs to remain in situ for a period of time after discharge. Patients require a further hospital admission or community nurse review for catheter removal.

We identified that parents can remove the catheter at home by cutting the balloon port and introduced this practice to our institution. Cutting the balloon port deflates the retaining balloon, and facilitates spontaneous passage of the catheter. Aim of this audit was to assess safety and success of home catheter removal.

Methods

A prospective audit was performed over 12 months. Parents of eligible patients were instructed verbally and with an information leaflet, including date for removal. Telephone follow-up post-removal was undertaken to assess the outcome.

Main results

38 patients were included from 07/2016 to 06/2017; age 9m-12y (median 2.5y). 31 patients had required a catheter following hypospadias repair, 7 patients for other urological procedures.

Home catheter removal was successful in 35 (92%) cases. 3 children required professional support for catheter removal. Median time until catheter passage was 3 hours (range 0-24hrs). Considering that cost for day case admission for catheter removal averages at 130£ per patient, home catheter removal saved the NHS 4550£ in the time period.

Conclusions

Parental removal of a urethral catheter is a safe and successful alternative to catheter removal by a professional. In addition to significant cost savings, it minimizes parental anxiety and inconvenience related to the catheter removal appointment.
Background

This study analyses the prognosis of Biliary Atresia (BA) in France since 1986, when both Kasai operation (KOp) and Liver Transplantation (LT) became widely available.

Methods/Materials

The charts of all patients diagnosed with BA born between 1986 and 2015 and living in France were reviewed. Patients were classified into 4 cohorts according to their years of birth: 1986-1996 (477), 1997-2002 (278), 2003-2009 (364), 2010-2015 (237).

Results

1356 patients were included; 1276 (94%) underwent KOp. Age at KOp (median 59 days, range 6-199) was stable over time. Survival with Native Liver (SNL) after KOp was 42%, 36%, 27% and 24% at 5, 10, 20 and 30 years. Independent prognostic factors for SNL after KOp were: age at KOp, polysplenia syndrome, anatomical pattern of the extra-hepatic biliary remnant.

16%, 7%, 7%, 7% of patients died without LT in the 4 cohorts (p=0.0001).

753 patients (55%) underwent LT. Patient survival after LT was 79% at 30 years. 5-year patient survival was 76%, 91%, 88%, and 93% in cohorts 1 to 4, respectively (p<0.0001).

Actual BA patient survival (from diagnosis) was 80%. 5-year BA patient survival was 72%, 88%, 87%, 88% in cohorts 1 to 4, respectively (p<0.0001).

Conclusion

With the sequential treatment of Kasai operation and liver transplantation if needed, 24% of BA patients reach the age of 30 years with their native liver, and 88% of BA patients can live. Continuous improvement of BA prognosis was mainly due to reduced mortality without LT, and better outcomes after LT.
SCVII-HB02

OUTCOME OF LOCALIZED LIVER-BILE DUCTS RHABDOMYOSARCOMA ACCORDING TO LOCAL THERAPY. A REPORT FROM THE EUROPEAN SOFT TISSUE SARCOMA STUDY GROUP

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Objectives: To evaluate the impact of local treatment therapy on outcome in patients with liver-bile ducts rhabdomyosarcoma (BD-RMS)

Methods: We analyzed the data of 30 patients (1 liver, 29 bile ducts) included in the RMS 2005 protocol until December 2016. Delayed surgery and/or External Beam Radiation Therapy (EBRT) were performed after at least 4 courses of chemotherapy.

Results: All were embryonal RMS, 15 had a tumour > 5cm, 6 were N1. Median age at diagnosis was 3 years (11 months - 8 years). Eight patients (26%) had primary surgery (1 IRS I, 6 IRS II, 1 IRS III) consisting of bile duct excision (BDE). Six of them also received EBRT. All are in first complete remission (CR1) except one (IRS II, EBRT+, local relapse, death). Six patients (19%) received EBRT alone (one local relapse, death). Sixteen patients (%) underwent secondary surgery (7 with additional EBRT): 14 BDE +/- partial hepatectomy (PH). Two had PH alone. Four R1 patients underwent EBRT without recurrence. Among 3 R0 patients who had EBRT, 1 had a metastatic relapse and died. Nine R0 patients did not receive EBRT: 3 relapsed locally, 2 died and 1 is in CR2. Local relapse was only associated with size >5cm. Five-year OS and EFS were 85% and 76.47%.

Conclusion: Local relapse in BD-RMS is fatal. There is no difference in outcome according to local treatment type. However, possible long-term sequelae of EBRT on hepatic pedicle could be balanced with the safety of BDE + Roux en Y loop.
ADJUVANT ANTI-VIRAL THERAPY FOR CYTOMEGALOVIRUS IgM+ve ASSOCIATED BILIARY ATRESIA

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Aim of Study: CMV-IgM+ve associated biliary atresia (CMV-BA) is a distinct aetiological subgroup characterized by inflammatory infiltrate and greater degree of hepatic fibrosis, leading to worse outcome. We report our experience with adjuvant antiviral therapy after Kasai portoenterostomy (KPE).

Methods: Single-centre prospective database between 2003 and 2017. Since 2011, valganciclovir (10 mg/kg/day) was started early post-operatively, in some cases followed by valganciclovir (60 mg/kg/day) (Group 1). Clearance of jaundice (CJ) was defined as achieving a total bilirubin ≤ 20μmol/L in post-KPE period and tested with Fisher test; native liver (NLS) and overall survival (OS) were compared with untreated BA CMV IgM+ve patients (Group 2) using LogRank test. P value of <0.05 was regarded as significant. Data are quoted as median (IQR range).

Results: During the 14-year period, 376 infants with histologically confirmed BA were treated, of those 37 (10%) were CMV IgM+ve at presentation. Racial background was Caucasian (n=16, 43%) or non-Caucasian (n=21, 57%). 36 underwent KPE [Group 1 (n=8) and Group 2 (n=28)]. Overall age at surgery was 67 (53-77) days: total bilirubin: 169 (140-211) μmol/L; AST/platelet ratio (APRi): 1.0 (0.64-2.5). There was no difference in age at surgery (P=0.13); bilirubin (P=0.12); or APRi (P = 0.13) between Group 1 and Group 2.

CJ was higher in Group 1 (75% vs 21%, P=0.007). The 2 year NLS and OS were improved (75.6% vs 26% and 100% vs 82%, respectively).

Conclusions:

- Adjuvant antiviral therapy improved outcome in infants with CMV-BA.
AIM OF THE STUDY: Liver transplantation (LT) in small recipient is challenging. The use of whole liver graft (WLG) is usually appreciated because it could facilitate the procedure (less bleeding, shorter ischemia time and no size mismatch). In this study, we questioned the impact of such graft when they come from deceased donors (DD) weighing less than 10 Kg.

METHODS: We retrospectively reviewed all LT of pediatric recipients that received a WLG from donor weighing 10 Kg or less between 2000 and 2016. The analysis was focused on outcomes and early vascular complications.

MAIN RESULTS: Among the 590 children identified that underwent LT during the study period, 12 received a WLG from a DD weighing from 8 to 10 Kg (12/590 = 2%). The mean age of recipients was 14 months and indication for LT was biliary atresia in 9 patients. The main vascular complication was early hepatic artery thrombosis occurring in six patients (50%). Only four patients had no vascular complication in the post-operative course (33%). Graft survival in this series was 85% at 1 year and 59% at 5 years. Overall survival at 10 years was 80%.

CONCLUSION: Pediatric LT using a WLG from donor weighing less than 10 Kg are rare. In our experience, this graft type carries a very high rate of vascular complications and should be used when no other solution is available (in emergency cases).
SCVII-HB05
MONITORING PROGRESSION OF THE CHRONIC LIVER DISEASE AFTER SUCCESSFUL PORTOENTEROSTOMY FOR BILIARY ATRESIA

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Aim of the study. To identify noninvasive follow-up markers for the chronic liver disease affecting biliary atresia (BA) patients after successful portoenterostomy.

Methods. Among patients clearing their jaundice after portoenterostomy (n=39), evolution of native liver histology was analyzed in follow-up biopsies taken at median age of 1.6 (n=26), 4.3 (n=27), or 10.7 years (n=27). Liver biochemistry, APRI, and liver stiffness were correlated with histological findings and development of portal hypertension.

Main results. Increasing cytokeratin 7-positive hepatocyte-cholangiocyte-metaplasia (HCM) after portoenterostomy (p=0.005-0.016) was observed in patients who developed portal hypertension (n=15) or were transplanted (n=6), while others (n=18) showed unchanged HCM (p=ns) and decreasing fibrosis after age 1.6 years (p=0.036). HCM correlated with increasing fibrosis (r=0.525, p<0.001). Bile acids>57 µmol/L (sensitivity 0.83, specificity 0.88), GT>65 U/L (0.88, 0.87), and conjugated bilirubin>7 µmol/L (0.75, 0.95) predicted high-grade HCM (AUROC ≥0.90, p<0.001 for each), with their combination providing an AUROC of 0.94 (p<0.001). Bile acids<21 µmol/L and GT<52 U/L predicted decreasing HCM (p<0.001 for both). The combination of liver stiffness (cutoff >17 kPa) and APRI (cutoff >0.70) was highly accurate in predicting portal hypertension (AUROC 0.96, p<0.001). Liver stiffness cutoff 24 kPa (0.76, 0.75) was the best prognosticator of cirrhosis (AUROC 0.81, p<0.001).

Conclusions. After successful portoenterostomy, degree of histological HCM associates closely with fibrosis progression, portal hypertension, and the need for transplantation. Conjugated bilirubin, bile acids, and GT accurately reflect the extent of HCM, while liver stiffness and APRI are reliable markers of portal hypertension.
Figure 1. A) HCM and B) fibrosis scores in liver biopsies according to development of portal hypertension during follow-up. *p<0.05 when compared to patients with portal hypertension at corresponding age.

Figure 2. The best noninvasive predictors for A) score 3–4 HCM and B) portal hypertension.
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**AIM OF STUDY:** Type 4 (intra and extrahepatic dilatation) choledochal malformation (CM) represents the extreme of the spectrum. The aim was to identify mechanistic factors and post-operative course with liver conservation surgery.

**METHODS:** Prospective single-centre dataset review of operation series 1996-2017. Definition included all CM with ultrasound defined intrahepatic dilatation (either > 5 mm) in addition to either fusiform or cystic extrahepatic dilatation. All were treated by excision of the extrahepatic component and hepaticojejunostomy only. Non-parametric correlation and Chi² analysis were used and a P value of 0.05 was considered significant. Data are described as median (IQR).

**RESULTS:** There were 37 (27 female) of 192 (19.2%) CM in total of which 5 (13%) were detected antenatally. Age and weight at surgery was 3.8 (1.8 – 8.5)years and 15(12–26)kg respectively. Intraoperative bile amylase levels correlated significantly with age ($r_S= 0.53; P=0.0005$) and inversely with choledochal pressure ($r_S=-0.63;P<0.0001$) and CBD diameter($r_S=-0.35; P=0.01$). CBD diameter was 20(14-43)mm with a distinct bimodal distribution in relation to age (FIG 1) and no correlation with age($P=0.4$). Older children had high amylase bile. Right and left duct diameter was similar at 8(4.5-12) and 7(4.5-13)mm respectively but with predominantly left-side correlation with CBD size ($r_S=0.43;P=0.004$). FIG.2 illustrates post-operative left-sided intrahepatic duct dimensions.

**CONCLUSIONS:**

- There appears to be a bimodal pattern of Type 4 CM
- Conservative hepaticojejunostomy is associated with reversal of intrahepatic dilatation in all, presumably due to relief of intraductal pressure.
SCVII-HB07
PREDICTING BILIARY ATRESIA OUTCOMES AFTER SUCCESSFUL PORTOENTEROSTOMY

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Aim of the study. To identify early predictors of outcome after successful portoenterostomy for biliary atresia (BA).

Methods. Among patients who normalized their bilirubin after portoenterostomy (n=30), liver biochemistry was measured one, three, six, and 12 months postoperatively and related with native liver survival, follow-up liver histology, and portal hypertension; defined as esophageal varices or spleen size >2 SD associated with thrombocytopenia.

Main results. Median follow-up time with native liver was 6.8 (interquartile range 2.4-8.7) years, during which portal hypertension had developed in 12 (40%), 5 (25%) were transplanted, and none died. Liver biopsies taken at age of 4.3 (1.3-6.4) years showed cirrhosis in 15 patients (50%). Bile acids one, three, six, and 12 months (p=0.010-0.053, Figure 1), AST three and six months (p=0.050 for both), ALT three months (p=0.036), and APRI six months (p<0.001) after portoenterostomy predicted native liver survival. At three and six months postoperatively, bilirubin (optimal cutoff >9 µmol/L), GT (>130-180 U/L), and APRI (>0.47-0.55) predicted development of portal hypertension (area under receiving operating characteristic, AUROC 0.84-0.91, p<0.001-0.003 for each) while bilirubin >9-14 µmol/L (AUROC 0.85-0.88, p≤0.001) and APRI >0.55-0.61 (AUROC 0.84-0.85, p=0.002-0.006) predicted progression to histological cirrhosis. Conclusion. Liver biochemistry already three months postoperatively predicts the long-term outcomes after successful portoenterostomy. Bile acids most consistently reflect native liver survival after clearance of jaundice. After its normalization, bilirubin is unrelated with native liver survival but together with GT and APRI predicts development of cirrhosis and portal hypertension.
Figure 1. Bile acids six months after successful portoenterostomy A) in relation to native liver survival time and B) in the prediction of native liver survival.

Figure 2. A) Bilirubin six months after successful portoenterostomy in relation to survival without portal hypertension and B) bilirubin, GT, and APRI in the prediction of portal hypertension development.
LAPAROSCOPIC CHOLECYSTECTOMY USING NEAR-INFRARED FLUORESCENCE IMAGING WITH INDOCYANINE GREEN IN PEDIATRIC PATIENTS AFFECTED BY COMPLICATED CHOLELITHIASIS

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AIM OF THE STUDY
Although rare, common bile duct (CBD) and cystic duct (CD) injuries during laparoscopic cholecystectomy (LC) can be reduced and avoided using Indocyanine green (ICG) fluorescence, which leads to an easier intraoperative recognition of the extrahepatic biliary anatomy. Intravenous (IV) injection of this dye has become extensively used for confirming anatomy in adult laparoscopic cholecystectomy. We describe eight cases of pediatric patients who underwent LC with near-infrared fluorescence (NIRF) imaging after an IV injection of ICG.

METHODS
Eight pediatric patients (mean age 8.1 yrs; range 3 – 17 yrs.) with complicated cholelithiasis and scheduled for elective LC underwent IV injection of ICG (Verdy Diagnostic Green; dosage 0.25-0.5mg/kg) 12 hours before surgery. The CD and the CBD, highlighted by the ICG, were visualized and dissected.

MAIN RESULTS
In all cases the CD and the cystic artery were safely and effectively identified. They were both clipped with Hem-o-loks and dissected. The dissection of the gallbladder from the liver bed was challenging in 3 cases due to inflammatory adhesions. The mean surgical time was 136 minutes, while the mean length of hospital stay was 2.5 days. There were no postoperative complications.

CONCLUSIONS
In pediatric patients affected by complicated cholelithiasis, laparoscopic cholecystectomy using near-infrared fluorescence (NIRF) imaging with ICG provides an immediate and distinct visualization of extrahepatic biliary structures and allows a safe dissection of the cystic duct and artery.
Aim: To present a single surgeon’s experience with transumbilical laparoendoscopic single site surgery (TULESS) for childhood choledochal cyst (ChC).

Methods: Medical records of all children undergoing TULESS for ChC by the same surgeon from September 2012 to August 2017 were reviewed. For the TULESS, three 3-5mm ports were placed at separate points in the same umbilical incision site. Roux-en-Y loop was created extracorporally through the umbilical incision. Excision of ChC and hepatico-intestinal anastomosis were performed using conventional laparoscopic instruments.

Results: 237 patients were identified with median age of 26 months (range 1 month -12 years). The median diameter of ChC (measured by the largest size) was 3.1 cm (range: 1.0 – 12 cm). The ChC was successfully excised by TULESS in all cases. Hepatico-jejunostomy was performed in 235 cases and hepatico-duodenostomy in 2 cases. Additional trocars were needed in just one case (0.4%). There was no conversion to open surgery. The median operative time was 180 minutes. No drain was used in 94.5% of cases. Early postoperative complications were noted in 2.1%, including bile leak (0.8%), intestinal obstruction (0.4%) and mild umbilical infection (0.8%). The median postoperative hospital stay was 5 days. At follow up of 4 – 64 months, one patient needed a redo-surgery for anastomotic stenosis; all other patients were in good health. The postoperative cosmesis was excellent, as all patients were virtually scarless.

Conclusions: In experienced hands, TULESS with conventional laparoscopic instruments for childhood ChC is feasible, safe, with excellent postoperative cosmesis.
SCVII-HB11
OUTCOME OF MULTIMODAL MANAGEMENT OF CONGENITAL PORTOSYSTEMIC SHUNT AND THE IMPACT OF ASSOCIATED ANOMALIES

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Introduction: Congenital portosystemic shunt (CPS) is a rare condition which may lead to serious complications. We wanted to assess the multimodal management (observation, interventional radiology, surgery) and the impact of associated anomalies (AA) on the outcome.

Method: We conducted a review of patients with CPS referred to our centre between 1998 to 2017. We looked at presentation, AA, anatomic form, intervention to close the shunt and the outcome.

Results: 145 children were identified. AA included cardiovascular (38), liver (10), urologic (7) anomalies, with 54 children having more than one. 1 child died of severe pulmonary hypertension before treatment and one of liver malignancy after closure.

<table>
<thead>
<tr>
<th></th>
<th>AA(n=65)</th>
<th>Not AA (n=80)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prenatal diagnosis</td>
<td>19</td>
<td>42</td>
<td>0.002</td>
</tr>
<tr>
<td>age at diagnosis</td>
<td>78.6</td>
<td>64.7</td>
<td></td>
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<tr>
<td>anatomic form</td>
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<td></td>
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<tr>
<td>extrahepatic</td>
<td>13</td>
<td>5</td>
<td>0.007</td>
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<tr>
<td>porto hepatic vein</td>
<td>23</td>
<td>48</td>
<td>0.001</td>
</tr>
<tr>
<td>left sided (incl. Aranthius)</td>
<td>8</td>
<td>9</td>
<td>0.422</td>
</tr>
<tr>
<td>right terminolateral</td>
<td>7</td>
<td>7</td>
<td>0.345</td>
</tr>
<tr>
<td>right laterolateral</td>
<td>14</td>
<td>11</td>
<td>0.114</td>
</tr>
<tr>
<td>PCS Complications</td>
<td>45</td>
<td>46</td>
<td>0.075</td>
</tr>
<tr>
<td>liver nodules</td>
<td>40</td>
<td>40</td>
<td>0.085</td>
</tr>
<tr>
<td>neurologic</td>
<td>20</td>
<td>17</td>
<td>0.099</td>
</tr>
<tr>
<td>cardiac-pulmonary</td>
<td>4</td>
<td>7</td>
<td>0.290</td>
</tr>
<tr>
<td>shunt closure at last FUP</td>
<td>50</td>
<td>60</td>
<td>0.392</td>
</tr>
<tr>
<td>lost fup</td>
<td>2</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>delay at closure</td>
<td>11</td>
<td>14</td>
<td></td>
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<tr>
<td>spontaneous</td>
<td>9</td>
<td>26</td>
<td>0.004</td>
</tr>
<tr>
<td>delay of closure (Mo)</td>
<td>3</td>
<td>5.5</td>
<td></td>
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<tr>
<td>radiologic</td>
<td>19</td>
<td>15</td>
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<tr>
<td>surgical</td>
<td>19</td>
<td>17</td>
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<tr>
<td>ri+surgical</td>
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<td>2</td>
<td>0.263</td>
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<tr>
<td>post op complication</td>
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<td>8</td>
<td>0.243</td>
</tr>
<tr>
<td>Death</td>
<td>1</td>
<td>1</td>
<td>0.448</td>
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</tbody>
</table>
Conclusion: patients with PCS and associated anomalies have less prenatal detection, porto-hepatic shunts, and spontaneous closure rate. Intervention to close the shut does not involve more complications than patients without AA.
SCVII-HB12
SURGICAL ANATOMY AND RADIOLOGICAL CHARACTERISTIC OF CONGENITAL RETROHEPATIC PORTO-CAVA SHUNTS (ABERNETHY MALFORMATION)

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AIMS OF THE STUDY. Congenital portosystemic shunts (CPSS) can develop life–threatening complications including liver tumors. Retrohepatic porto-cava shunts (RPCS) are considered unsuitable for radiological treatment. The distinction between end-to-side (ES-RPCS) and side-to-side (SS-RPCS) can be difficult, but essential for treatment strategies (one or two step closure). We analyze our experience with RPCS.

METHODS: Between 2007-2017, 6/18 children with CPSS treated were RPCS (3 ES-RPCS, 3 SS-RPCS). Radiological studies included doppler-ultrasounds, CT/MRI and angiography with occlusion test (5/6). Four (3/3 ES-RPCS, 1/3 SS-RPCS) developed liver tumors.

MAIN RESULTS: ES-RPCS showed normal portal vein diameters without intrahepatic portal vein branches (IHPVB), one with a “puff of smoke” pattern and absence in the remaining. SS-RPCS showed 3/3 aneurismatic dilatation of the portal vein with patent IHPVB (1mild, 2moderate). Portal pressure (occlusion test) was higher in ES-RPCS (>40 mmHg) than SS-RCPs (<25 mmHg) (failed to occlude in one due to fistula diameter). ES-RPCS surgical anatomy findings were caudate lobe absence (one with liver accessory lobe) with the fistula entering on the left aspect of the cava (free of liver parenchyma) while in SS-RPCS entered upon the upper cava aspect through the hepatic parenchyma. 2/2 ES-RPCS and banding surgery showed PV cavernomatous transformation with IHPVB patency at 3 months. 1/2 patient developed portal hypertension favoured by descompensated Fallot anomaly. 2/2 SS-RPCS surgery occlusion /banding patients cured with complete IHPVB development.

CONCLUSION. Fistula shape, relationship with vena cava, portal pressure and IHPVB are different in both types of RPCS. Heart disease may contraindicate conservative surgery.

<table>
<thead>
<tr>
<th>Shunt type</th>
<th>Age/sex</th>
<th>Symptoms</th>
<th>PVP (mmHg)</th>
<th>Treatment</th>
<th>F/up (yrs)</th>
<th>Status</th>
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</thead>
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<tr>
<td>ES-RPCS</td>
<td>16/F</td>
<td>Liver tumors (HNF-adenoma)</td>
<td>ND</td>
<td>Left lateral segmentectomy</td>
<td>9</td>
<td>Awaiting transplant</td>
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<tr>
<td>ES-RPCS</td>
<td>11/F</td>
<td>Liver tumor (adenoma)</td>
<td>51</td>
<td>Banding</td>
<td>2.5</td>
<td>Portal HT (30% mass reduction)</td>
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<tr>
<td>ES-RPCS</td>
<td>4/F</td>
<td>Liver tumors (HNF)</td>
<td>40</td>
<td>Banding</td>
<td>1.6</td>
<td>Tumor resolution</td>
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<tr>
<td>SS-RPCS</td>
<td>5/M</td>
<td>Liver tumors (adenoma)</td>
<td>Failed (FistulaØ&gt;2 cm)</td>
<td>Liver transplant (HCC)</td>
<td>4</td>
<td>Exitus. HCC recurrence</td>
</tr>
<tr>
<td>SS-RPCS (H-Type)</td>
<td>2/M</td>
<td>Encephalopathy</td>
<td>24</td>
<td>Failed amplazter Surgical oclusion</td>
<td>3</td>
<td>Cured</td>
</tr>
<tr>
<td>SS-RPCS (short fistula)</td>
<td>2/M</td>
<td>Encephalopathy</td>
<td>22</td>
<td>Banding</td>
<td>1.7</td>
<td>Cured</td>
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</table>
**Aim of the Study**

Hepatoblastoma is the most frequent hepatic tumor in children and its initial presentation will affect treatment and prognosis. The aim of this study is to evaluate long-term results of liver transplantation in children with unresectable hepatoblastoma.

**Methods**

Retrospective review of patients with hepatoblastoma who underwent liver transplantation in the last 25 years was performed, analyzing risk factors, tumor presentation, treatment and long-term survival in order to identify prognostic factors.

**Main Results**

Thirty-one patients underwent liver transplantation in the context of unresectable hepatoblastoma, mainly male (67%) and presenting risk factors such as prematurity (12.9%), maternal smoking (6.5%) and familial adenomatous polyposis (3.2%). Most frequent presentation was multifocal PRETEXT III with vascular involvement (51.6%) and PRETEXT IV (45.2%). Four patients had metastasis at diagnosis (12.9%).

Twenty one patients received a living-donor (67.7%) and 10 a cadaveric graft (32.3%), at 31.7 months of age (5-125). Most transplants were primary and only two were performed as rescue therapy after an attempt of surgical resection of the tumor.

Overall survival one and five years after transplantation were 93.3% (± 4.6%) and 86.4% (± 6.3%) respectively. We could not find any statistically significant differences between risk factors, tumor presentation, type of graft or type of transplant.

**Conclusions**

Liver transplantation has increased hepatoblastoma survival in unresectable patients. Due to this good results, we have not been able to find significant prognostic factors in this cohort.
SCIENTIFIC SESSION VIII: UROLOGY

SCVIII-UR01
FERTILITY POTENTIAL IN PATIENTS TREATED FOR VARICOCELE IN PEDIATRIC AGE

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Aim of the study

to assess the fertility potential of patients treated for varicocele without post-operative complication with complete ipsilateral testicular catch-up growth and normal hormonal values.

methods

We considered data about adolescent with varicocele aged between 10 and 16 years treated at our institution between September 2010 and September 2015. all patients were followed clinically and with hormonal tests before and after surgery and re-evaluated after semen analysis. Semen quality was correlated with hormonal status, surgical complications and testicular volume.

Main Results

133 subjects without recurrence and with testicular catchup growth were enrolled; 52 patients had grade III varicocele, 80 had grade II varicocele and 1 patients had grade I varicocele. Surgical complications at 18 months were 6 hydroceles (2 in grade III and 4 in grade II varicocele). Among 41 patients with semen analysis, 75% had a good semen quality, 9,7% had a fairly good semen quality and 14,6% had a poor semen quality. Among grade III (12 patients), 7 (58%) had a good quality, 3 (25%) a fairly good quality and the other 2(16,6%)a poor quality. In grade II, 23(79,3%) had a good quality, 4(13,7%) a fairly good quality and the other 2(6,8%) patients had a poor quality.(p>0.05).there were no correlation between semen quality, hormonal status, grade of varicocele before surgery and testicular volume after surgery

Conclusions

Our results demonstrate that varicocelectomy alone in full well studied and treated patients in pediatric age doesn’t preserve the fertility potential
The radical soft-tissue mobilisation (RSTM, Kelly repair) is an anatomical reconstruction of bladder exstrophy generally been performed as part of a two-step strategy following successful neonatal closure.

**Aim:** To determine the feasibility of a combined procedure of delayed bladder closure and RSTM in one stage without pelvic osteotomy.

**Methods:** From 11/2015 to 12/2017, 18 bladder exstrophy patients (12M-6F) underwent combined closure with RSTM by a single surgeon at three institutions, including 13 intentionnally delayed repairs (median age 12 weeks [2-33]), and 5 cases presenting late (age 24m [10-37]). RSTM included mobilisation of the bladder plate, urogenital diaphragm and corpora cavernosa from the pelvic walls, followed by anatomical reconstruction with anti-reflux procedure, bladder closure, urethro-cervicoplasty, muscle sphincter approximation, and penile/clitoral reconstruction. Examined criteria were bladder dehiscence/prolapse, bladder-neck or urethral fistula, urethral stenosis, and parietal hernia. Continence and voiding have not been addressed at this stage.

**Main results:** All bladder exstrophy cases were successfully closed without osteotomy, with no case of bladder dehiscence after 11 months [3-24m] follow-up.

Urethral fistula or stenosis occurred in four patients: all 3 fistulae closed spontaneously in less than 3 months; one urethral stenosis was successfully treated with endoscopic high-pressure balloon dilatation. Although the follow up is short, it does allow examination of the main outcome criterion, namely bladder dehiscence, which is usually expected to happen very early after surgery.

**Conclusion:** The Kelly RSTM can be safely combined with delayed bladder closure without osteotomy in both primary and redo cases in classic bladder exstrophy.
SCIENTIFIC SESSION VIII: UROLOGY

SCVIII-UR03
RETROGRADE INTRARENAL SURGERY IN THE MANAGEMENT OF RENAL LOWER POLE STONES IN CHILDREN

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Introduction. Renal stones located in the lower pole of the kidney represent a serious challenge for surgical treatment in children. The options are: open surgery, extracorporeal shock-wave lithotripsy, percutaneous nephrolithotomy and retrograde intrarenal surgery. Reports about the endoscopic treatment in children are limited. The aim of the study was to evaluate the effectiveness of retrograde intrarenal surgery in pediatric patients with renal stones in lower pole of the kidney.

Patients and Methods. We retrospectively analyzed the results of the retrograde intrarenal surgery in 24 patients with renal stones in lower pole, between April 2012 and September 2016. Flexible ureterorenoscopy in combination with holmium laser lithotripsy were performed. We considered stone fragment size 3 mm or less as a measure for sufficient fragmentation of the stone.

Results. Mean duration of general anesthesia was 68 minutes (range, 40-90). Duration of hospitalization was 1-3 days (mean, 1.6 days). Complications were found after two (8.4%) surgical procedures: perirenal haematoma in one (4.2%) and urinoma in one (4.2%) patient. The stone was completely fragmented in 18 (75%) patients. In 3 (12.5%) patients the stone was incompletely fragmented and in 3 (12.5%) patients the stone was not fragmented. Mean follow up was 9 (range, 6-18) months.

Conclusion. Retrograde intrarenal surgery in children is the least invasive, effective and safe surgical procedure for stones in lower pole of the kidney, with minimal complication rate. Unsuccessful treatment in some patients was due to loss of ureterorenoscope deflection with laser probe in working channel.
Aims of the study: The efficacy of endoscopic management of primary obstructive megaureter (POM) has been proved, but the need for postoperative double J stenting is still a matter of debate. We aimed to compare the outcomes of dilatation with or without ureteral stenting.

Methods: Multicentre retrospective study on children operated on from May 2010 to November 2017 following the rules of the local ERB. Data were given in median (range) and analysed retrospectively using a Chi2 test.

Main results: 47 dilatations were performed in 43 patients (27 with prenatal diagnosis). Surgery was indicated for function decrease at renal scan (25), urinary tract infections (26), upper urinary tract dilation increase (22). Size of the renal pelvis, pelvic ureter and parenchyma thickness were 22mm, 16mm, and 4mm respectively. Age at surgery was 13 months (0.3-166). 22 procedures were performed with a double J stent insertion versus 25 without. 21 patients developed postoperative complications: 13 in the JJ group vs 8 in the non-stented group (p=0.08) including 10 vs 4 UTI (p=0.05), 1 vs 3 dilation increases (p=0.6), 0 vs 1 renal colic, 2 vs 0 JJ related pain, respectively. After a follow-up of 26 months (2-66), 37 ureters required only 1 dilatation (18 vs 19), 4 two dilatations (1 vs 3), 6 a Cohen procedure (2 vs 4).

Conclusions: Balloon dilatation without double J stenting for POM offers a comparable success rate with a lower risk of UTI and a tendency towards lower overall complication rate in comparison with the stenting group.
SCVIII-UR05
ROLE OF WINTER SHUNT IN POSTOPERATIVE VIABILITY IN POST CIRCUMCISION GLANS SECTION: ABOUT THREE CASES AND LITERATURE REVIEW

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Department Of Pediatric Surgery, Université De Tunis El Manar, Faculté De Médecine De Tunis, Tunis, Tunisia

Introduction

The aim of this study is to report three cases of glans amputation secondary to circumcision, discuss therapeutic means, and review the role of spongiocavernosal shunts.

Materials and methods

We performed a retrospective study based on 26 circumcision complications observed over a period of 17 years (2000-2017). Section of the glans was found in 13 cases, among which 5 were total amputations. Three of them have benefited from glans replantation using winter shunt.

Results

The age of our patients ranged between 3 and 5 years. In all cases, the circumcision was made by a paramedical practioner. They consulted a specialist hours following the circumcision. Surgical treatment was the same for all three patients: Glans replantation was performed using three anastomoses. The urethra was repaired with an end-to-end anastomosis of the urethral serosa with 6-0 PDS sutures over an 8 FR silicone catheter. A Corpus spongiosum was anastomosed with a single layer of interrupted 6.0 PDS sutures. The fascial layer and skin were reapproximated with interrupted 5-0 PDS sutures. Winter’s spongiocavernosal shunt was performed to maximize venous drainage. The length of hospital stay ranged from 10 to 21 days with good cosmetic and functional results.

Conclusion

In glans amputation, venous drainage following replantation is vital to the success of surgery. However, anastomosis of the dorsal veins is not always possible. In these instances, we propose spongiocavernosal shunt as an intervention which may aid in avoiding complications associated with venous congestion of the replanted glans.
ROBOT-ASSISTED EXTRAVESICAL URETERAL REIMPLANTATION (REVUR) FOR UNILATERAL VESICO-URETERAL REFLUX IN CHILDREN: RESULTS OF A MULTICENTRIC INTERNATIONAL SURVEY

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Aim of the Study: This multicentric international retrospective study aimed to report the outcome of robot-assisted extravesical ureteral reimplantation (REVUR) in patients with unilateral vesico-ureteral reflux (VUR).

Methods: The medical records of 55 patients (35 girls, 20 boys) underwent REVUR in 4 international centers of Pediatric Robotic Surgery for primary unilateral VUR were retrospectively reviewed. Patients’ average age was 4.9 years. The preoperative grade of reflux was III in 12.7%, IV in 47.3% and V in 40%. Twenty-six patients (47.3%) presented a loss of renal function preoperatively and 10 (18.1%) had a duplex system.

Main results: Average robot docking time was 16.2 minutes [5-30]. Average total operative time was 92.2 minutes [50-170]. No conversions or intra-operative complications were recorded. All patients had a bladder Foley catheter for 24 hours post-operatively. Average hospital stay was 2 days [1-3]. Average follow-up length was 28 months [9-60]. We recorded 3 (5.4%) postoperative complications: 1 small urinoma resolved spontaneously (II Clavien) and 2 persistent reflux, only one requiring redo-surgery using endoscopic injection (IIIb Clavien).

Conclusions: REVUR is a safe and effective technique for treatment of primary unilateral VUR. The procedure is easy and fast to perform thanks to the 6° of freedom of robotic arms. The learning curve is short and it is useful to begin the robotics experience with a surgeon expert in robotic surgery as proctor on the 2nd robot console. The high cost and the diameter of instruments remain the main challenges of robotics applications in pediatric urology.
SCVIII-UR07

SURGERY FOR URETEROPELVIC JUNCTION OBSTRUCTION (UPJO) – COMPARISON OF OUTCOMES UND COSTS BETWEEN PEDIATRIC SURGEONS AND UROLOGISTS IN GERMANY

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Aim of the Study:

Surgery for UPJO in Germany is performed by both Pediatric Surgeons and Urologists. The aim of this study was to analyze outcome and costs of surgery for UPJO in relation to the operating discipline.

Methods:

Data of patients aged 0-18 years were extracted from a major public health insurance (covering ~5.6 million clients) during 2009-2016 including socio-demographic variables, treating discipline and costs. Logistic regression analysis was performed for the risk of a complication within the first postoperative year.

Results:

229 children (31% female) were included (171 OPEN (75%; mean age 1.0 years) and 58 LAP procedures (25%; mean age 8.5 years). Pediatric Surgery operated on 162 patients (125 OPEN (77%); 37 LAP (23%)) whereas Urology performed surgery on 67 patients (46 OPEN (69%), 21 LAP (31%)). The median age of children operated by Pediatric Surgery was 0.6 years (OPEN) and 4.2 years (LAP) and therefore significantly younger compared to Urology (OPEN: 9.1 years; LAP: 15.8 years; p< 0.0001). After one year of follow-up, 5 children needed redo surgery after OPEN (N=4; 2.3%) and LAP (N=1; 1.7%) pyeloplasties. Case costs were equal for both techniques. However, the length of hospital stay was 4.3 days (p=0.0005) shorter in LAP compared to OPEN pyeloplasties.

Conclusions:

Pediatric Surgeons operate on younger patients compared to Urologists. LAP pyeloplasty is associated with a shorter length of stay, equal costs and similar complication level independent of the operating specialty. Therefore, laparoscopic pyeloplasty should be further promoted in specialized centers.
SCVIII-UR08

COMPLICATIONS AFTER VAGINOPLASTY IN ADULT WITH CONGENITAL ADRENAL HYPERPLASIA

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Aim of the Study: To evaluate the morbidity of vaginoplasty in adult women with congenital adrenal hyperplasia (CAH).

Methods: A gynaecological examination was performed in 44 CAH adult women regularly followed in endocrinology. Surgical reports were reviewed. Six of them had vaginoplasty during adulthood (17-29 years).

Main results: Feminising genitoplasty was performed in one single operation in 2 patients (22 and respectively 29 years of age) including clitoral reduction, vaginoplasty and perineoplasty. In the other 4 cases, clitoral surgery was performed during infancy (1 month – 1 year) with subsequent vaginoplasty in adulthood (17-28 years). Average age of examination was 39 years (22-55 years). Five out of this 6 patients had bleeding complications after vaginoplasty. One patient presented further complications (vaginal stenosis, urethrovaginal fistula). Overall in this series, each patient received 2.3 procedures (1-5).

Conclusions: The timing of genital reconstruction in Anomalies of Genital Development (AGD) is a controversial issue. This study confirms that the magnitude and morbidity of late vaginoplasty during adulthood is significantly higher than in childhood.
SCVIII-UR09

FEMALE GENITAL APPEARANCE IN 44 ADULTS WITH CONGENITAL ADRENAL HYPERPLASIA

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2Hôpital Louis Pradel, Hospices Civils de Lyon, Lyon, France.
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Aim of the Study: To describe variations in genital measurements in adult women with congenital adrenal hyperplasia (CAH).

Methods: Gynaecological examination was performed prospectively in 44 adult women with CAH (1 had no surgery, 43 had surgery). Clitoral size, labial aspect, vaginal length and width, anogenital distance were measured. Average age of examination was 30 years (17-55 years). Clitoral surgery was performed in 34/44 patients (6 had re-do surgery). Vaginal surgery was performed on 39/44 patients.

Main results: Seven patients had no measurable clitoris. The clitoral size for the 37 others was on average 18.3 mm long (3 – 55 mm). The mean vaginal length was 77 mm (65-100). Six patients had introital stenosis. The mean distance between the tip of the clitoris and the urethral meatus was 27.3 mm (1 – 60mm) and the mean distance from the posterior fourchette to the anterior anal margin was 24.3 mm (15 – 50mm).

Conclusions: Anogenital distance and clitoral length widely vary in this cohort. The vaginal length was normal compared with reference values in literature. A more systematic anatomical examination in Anomalies of the Genital Development (AGD) adults who underwent previous genital surgery is one important part of the overall evaluation of AGD.
USE OF SALINE FILLED EXPANDERS IN PELVIC OSTEOSARCOMA TREATED WITH RADIOTHERAPY: PRELIMINARY EXPERIENCE WITH 5 PEDIATRIC CASES

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Aim of the Study
Radiation enteritis is a side effect of pelvic radiotherapy. The placement of an expander in the abdomen can successfully isolate small bowel and rectum from the radiation field. The saline filled expanders are adjustable to the pediatric pelvic anatomy. We describe our experience using these expanders to prevent radiation enteritis.

Methods
A retrospective study of pediatric ilio-sacral osteosarcoma treated by radiotherapy was conducted in our unit. The expanders were placed two weeks before radiotherapy by Pfannenstiel incision to push the bowel or rectum out of the radiation field. The expanders have rectangular or “croissant” shape with a volume between 60 and 300 ml. They are filled with a mixture of saline and contrast solution using a subcutaneous chamber. A CT scan was performed to control the radiation dose near the bowel.

Main results
Five children, 7 to 17 years old, were treated with radiotherapy for pelvic osteosarcoma since 2007. Four expanders were placed near iliac bone to displace the small bowel. One near the sacrum to protect the rectum. The patients received the scheduled radiotherapy. Local discomfort related to the expander was resolved by partial emptying. No enteritis was reported. Three were removed by laparoscopy, one by laparotomy and one is scheduled for.

Conclusions
The use of saline filled expanders in the abdomen before radiation in children treated for pelvic osteosarcoma is feasible with good tolerance and without surgical complications. We can adapt their size, form and volume to the local anatomy and area of protection.
Aim of the Study
To analyze indications, surgical intervention methods and long-term treatment results in children with pancreatic tumors.

Methods
Retrospective analysis of 31 patients treated for pancreatic tumors between 1991-2017. We recorded data relating to the histological diagnosis, surgical technique and treatment outcome.

Main results
Benign tumors were diagnosed in 22 patients (the most common was solid pseudopapillary tumor - 13 patients), malignant tumors in 9 patients (pancreatoblastoma – 1, adenocarcinoma – 2, acinar cell carcinoma – 1, insulinoma – 1, gastrinoma – 1, Burkitt lymphoma – 3). Tumors were localized in the head of pancreas – 14 patients, body – 11 and tail - 6. Tumor resection was performed in 28 patients: distal pancreatic resection – 13, enucleation – 7, modified Whipple procedure – 6, subtotal pancreatic resection – 2. The patient with malignant insulinoma also underwent liver transplantation because of multiple unresectable liver metastases. Biopsy was performed in 3 patients with unresectable tumors. 7 patients with malignant diseases received chemotherapy. Early reoperation was required in 5 patients (pancreatic leakage - 3, bowel necrosis – 1, bowel anastomosis ulcer – 1). Follow-up ranged from 1 to 146 months (median 25 months). Tumor recurrence occurred in 5 patients between 6 to 24 months after surgery. 4 patients with local recurrence underwent reoperation. 1 patient died due to neoplastic disease progression and 1 from surgical complications. Two patients with tumor spread were transferred to regional palliative care units.

Conclusions
Surgical treatment depends on the anatomical localization and the tumor type. Histopathologic diagnosis is the most important factor for long term prognosis.
PW9-ON03
MANAGEMENT OF PNEUMOPERITONEUM IN PATIENTS WITH PNEUMATOSIS INTESTINALIS: DIFFICULT SURGICAL DECISION-MAKING PROCESS

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Aim of the study: Pneumatosis intestinalis (PI) is a rare complication among patients following allogeneic hematopoietic stem cell transplant (HSCT), frequently associated with the treatment of graft versus host disease (GVHD). Management of the pneumoperitoneum accompanying PI is one of the most serious surgical decision-making processes. The aim of the study is to evaluate the outcome of patients with pneumoperitoneum accompanying PI.

Methods: Between 2007 – 2017 we retrospectively reviewed medical data of 269 patients after allogeneic HSCT, 5 of them had pneumoperitoneum accompanying PI.

Main results: Over the 10-year period we treated 5 patients with PI, who received allogeneic HSCT, all suffered from chronic GVHD. Symptoms of PI were abdominal pain and distension. All of the patients were managed without surgical intervention, despite having pneumoperitoneum at the time of diagnosis of PI. 4 of them were treated by hyperbaric oxygen therapy with good results. None of the patients died as a direct consequence of PI.

Conclusions: It is a difficult surgical decision not to operate on the pneumoperitoneum. We recommend that PI as a benign self-limiting process should be treated with conservative expectant management. Surgical intervention is non-therapeutic and exceedingly unnecessary.
Aim of the study:

To show the interest and perspectives of virtual surgical planning in Nephron Sparing Surgery (NSS) for Wilms tumor cases.

Methods:

A retrospective review was performed including children treated at our institution for a Wilms Tumor with a NSS using a 3D modeling software. Patients had chemotherapy pre operatively as recommended by the SIOP. Images from CT scan were analyzed with Visible Patient Planning Suite 1.0.11 to make 3D reconstruction. This software allows us to analyze the tumor in all the dimensions of the space by rotating images or watching through organs. Vascularization is studied using the clip applying system which enables us to simulate vessels’ clamping. This function shows the resulting ischemia and estimates the volume of the vascularized territory.

Results

3 hemi nephrectomies were performed. The first case concerned a 21-months boy with an extensive left Wilms tumor in the inferior and middle pole measuring 25*18*17 mm. The second case concerned a 31-months boy with a bilateral Wilms tumor; both are extensive in the superior and middle renal pole measuring 55*56*47mm on the right and 74*61*57mm on the left. Mean operative time was 136 min and the length of stay was about 7 days. No complications occurred and renal function was preserved. A CT scan carried out post-operatively for each patients shows the proper functioning of the remaining hemi kidneys.

Conclusion:

Virtual surgical planning with 3D reconstruction is an innovating procedure. This personalized tumor mapping allows surgeons to perform a tailored surgery.
Aim: was to assess the incidence and outcome of surgical management of perforated pediatric intestinal lymphoma.

Patients and methods:

After the IRB approval, retrospective analysis of the hospital database between June 2007 and June 2017 of all cases of perforated intestinal lymphoma was done. All patients were ≤18 years old.

Results:

Perforation developed in 4.7% (16 of 340) of patients with intestinal lymphoma. Most of the patients were Burkitt’s lymphoma with a median age of 5.3 years. The ileum was the most common site of perforation n=10, followed by ileocecal n=4, and jejunum n=2. The timing of perforation was at presentation n=2, during chemotherapy n=12 and at relapse n=2. Primary resection anastomosis was done in 13 patients, 2 patients had resection with a stoma and 1 patient had stoma with no resection. The resected specimen showed a viable tumor in 12 patients, no viable tumor n=3 and chronic inflammation in 1 patient. Median delay to restart chemotherapy was (16 days). Wound infection n=4 (25%) and dehiscence n=2 (12.5%) were the most common complications. The 3-year OS and EFS of perforated patients were 83.4% and 78.9% respectively compared with 88.7% and 79.2% OS and EFS of non-perforated patients with no statistically significant different OS (P=0.425) and EFS (P=0.827).

Conclusion

Perforation is not an adverse prognostic factor for survival in pediatric intestinal lymphoma patients. Primary resection anastomosis is a safe option for most of the patients.
Aim of the Study

LSD1 (KDM1A) is expressed in neuroblastoma and other malignancies. Like a putative oncogene LSD1 epigenetically regulates gene expression patterns. In neuroblastoma LSD1 is a co-factor of MYCN. LSD1 knockdown resulted in transcriptional program alterations coinciding with neuronal differentiation and impaired tumor growth. For AML and other cancers LSD1 is a candidate therapeutic target. We investigated LSD1 inhibition in neuroblastoma cell lines and evaluated the therapeutic potential.

Methods

LSD1-specific inhibitor compounds NCL-1, GSK2879552 and RG6016 were tested on neuroblastoma cell lines IMR-5/75, SK-N-BE(2), SK-N-AS and SCLC cell line NCI-H526 as positive control. Cell viability was assessed using CellTiterGlo luminescent assay. Compound IC50 values were derived using GraphPad Prism.

Main Results

Our results confirm inhibitory effects of NCL-1 on neuroblastoma cell lines with IC50s in low micromolar ranges. Its effect, though, seems unspecific as results of the positive control show. RG6016 seems more selective as differences between dose-response curves indicate. GSK2879552 shows strongest efficacy after 3 days with an average IC50 of 25 µM. Therefore, inhibition of neuroblastoma cell viability using NCL-1 is more effective than GSK2879552 or RG6016. Yet, more selective and less toxic inhibitions seems viable using RG6016.

Conclusion

Targeting LSD1 is a therapeutic option for neuroblastoma treatment. As observed IC50 values exceed typical therapeutic concentrations, additional steps in LSD1 inhibitor optimization and dosage finding are required prior to potential transition into clinical trials. Future experiments should investigate inhibitory mechanisms of compounds and decipher specific downstream effects on neuroblastoma biology.
THE ELEVATED RISK OF RECURRENCE OF MATURE OVARIAN TERATOMA IN LONG-TERM FOLLOW-UP SUGGESTS OVARIAN-SPARING SURGERY AS PREFERABLE INITIAL SURGICAL APPROACH

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AIMS OF THE STUDY. Mature ovarian teratoma (MOT) are benign tumors with low risk of recurrence in short to mid-term follow up, potentially amenable to ovarian-sparing surgery (OSS). Despite the advantages of OSS for reproductive and hormonal health, ovariectomy (OVC) is still the most practiced approach to MOT among pediatric surgeons. We aimed to evaluate the risk of recurrence of MOT in the long term follow up, depending on the initial surgical choice.

METHODS. Retrospective analysis of patients operated for MOT from 2004 to 2016 in two pediatric surgical unit, invited to ultrasound follow-up (FU) by independent operators.

MAIN RESULTS. Twenty-eight patients agreed to participate (mean age 10.7 years): 14 treated with OSS (mean diameter 52.3 mm, in 2 cases after ovarian detorsion), 14 treated with OVC (mean diameter 97.7 mm, 8 out of 14 presenting with torsion). During follow-up (median 72 months, range 5-156 months), MOT recurrence was observed in 4 OSS-pts (28.5%, 1 ipsilateral, 3 contra-lateral after 7, 6, 3 and 3 years from surgery) and in 3 OVC-pts (21.4%, after 10, 6 and 6 years from surgery), without significant difference among surgical approach (Fisher’s exact test).

CONCLUSIONS. The elevated risk of MOT recurrence, irrespective of the surgical approach, imposes the need of a prolonged ultrasound follow up and suggests the application of OSS as preferable initial therapeutic choice, to preserve the follicular reserve.
EVALUATION OF THE INTRAPELVIC EXTENSION OF SACROCOCCYEAL TERATOMA IN PRENATAL LIFE: ROLE OF FETAL MRI

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Aim of the study
Prenatal assessment of sacroccygeal teratoma (SCT) is critical in order to plan the delivery and the immediate postnatal care. Our purpose was to study the intrapelvic extension of a series of fetal SCT and to compare prenatal sonography and magnetic resonance imaging (MRI) findings.

Methods
In the period between 2005 and 2017, seventeen pregnant women underwent fetal MRI (21st–38th week of gestation) after sonographic suspicion of SCT. Images were analysed to detect intrapelvic extension and classified according to Altman system. MRI grading was compared with sonographic one.

Main Results
At prenatal MRI 9/17 SCT were classified as type I (predominantly external mass), 6/17 as type II (predominantly external mass with internal extension), and 2/17 as type III (external and internal mass with extension into the abdominal cavity). In 10/17 cases there was complete agreement between MRI and sonography, in 5/7 sonography was not able to classify the mass and in 2/7 intrapelvic extension was underestimated by sonography. MRI findings were confirmed by postnatal MRI in 8/17 patients, in 7/17 a CT-angiography was indicated for a better definition of the vascularisation and 2 patients with huge masses were submitted to early surgery in the neonatal period.

Conclusions
Prenatal MRI allows good evaluation not only of the size and the composition (cystic or solid) of SCT but also of its intrapelvic extension. We consider it a necessary tool to better complete the diagnostic definition of fetal SCT.
PW9-ON09
ENDOGLIN (CD105) AS AN INDEPENDENT PROGNOSTIC FACTOR IN NEUROBLASTOMA

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Aim of the Study. To determine if the clinical outcome of children with neuroblastoma is correlated with intratumoral microvessel density (IMVD), as determined by endoglin (CD105) staining and other prognostic factors.

Methods. Tumor tissue specimens from 38 patients with peripheral neuroblastic tumors who underwent surgical resection or biopsy of their primary tumor without any preoperative therapy were retrospectively reviewed. IMVD was identified immunohistochemically using monoclonal antibodies against CD105. Prognostic factors, such as the MYCN oncogene, disease stage, histopathology and age, were correlated with outcome.

Main results. Among 38 examined specimens, the median IMVD value was 23.2 (15.1–28.4). The IMVD identified by CD105 was significantly higher in patients with unfavorable histology, metastatic disease, MYCN amplification and COG high risk group. ROC analysis was used to find significant IMVD level regarding EFS. The cut-off >18 was selected according to the greatest sensitivity (100%) and specificity (68.42%). The multivariate Cox proportional hazards analysis demonstrated that MYCN amplification and IMVD were significant prognostic factors in predicting EFS (hazard ratio for MYCN amplification: 3.61; 95% CI: 1.20–10.90; P ¼ 0.023 and for IMVD: 1.05; 95% CI: 1.00–1.09; P ¼ 0.037).

Conclusion. IMVD determined by CD105 appeared to be an independent prognostic factor for neuroblastoma.
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**Objective:** To evaluate the effect of neo-adjuvant intra-arterial chemotherapy (IAC) and systemic chemotherapy for treatment of invasive bladder/prostate rhabdomyosarcoma (BP RMS) in children.

**Methods:** Between 1999 to 2013, 8 patients (6 males and 2 female), aged 0.5-14.4 years (median 6 years) with invasive BP RMS received preoperative intra-arterial chemotherapy (IAC) and systemic chemotherapy. According to the IRSG staging system, 2 patients had stage II, 5 stage III and 1 stage IV(with lung metastasis).

IAC was performed using Seldinger technique. The femoral artery was catheterized and a catheter placed in the contralateral internal iliac artery for anticancer agents infusing. The total amount of agents infused in bilateral internal iliac arteries was cisplatin 90 mg/m², pirarubicin 40 mg/m², and vindesine 3 mg/m². Intravenous chemotherapy with vindesine, ifosfamide, and etoposide was administered 3 weeks after IAC. Surgery was carried out 3 weeks after Intravenous chemotherapy. Radiotherapy and multidrug chemotherapy were administered after surgery.

**Results:** No cardiotoxicity, renal insufficiency, or hepatic dysfunction were found. Grade III marrow suppression developed in 4 patients. Bladder-sparing surgeries were carried out after neo-adjuvant therapy: partial cystectomy in 6, prostatectomy plus partial cystectomy in 2 patients. The median length of follow-up was 9.5 years (range 5.6-18.1 years). Six patients were recurrence free survival with functioning bladder. Two patients had pelvic cavity recurrence 1 year after surgery.

**Conclusion:** Neo-adjuvant IAC combined with systematic chemotherapy for the treatment of invasive BP RMS in children is safe and effective.
CHEST WALL RECONSTRUCTION AFTER INTRAOPERATIVE ELECTRON RADIATION THERAPY (IOERT): EXPERIENCE IN OUR INSTITUTION

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Abstract

Aim of the study:

IOERT (Intraoperative Electron Radiation Therapy) is a valid option for the treatment in selected oncologic pediatric patients. This study analyzes the application of this therapy in patients who require a costal reconstruction after a thoracic wall tumor resection.

Methods:

An analytic retrospective study was designed including all patients undergoing surgery with IOERT and thoracic wall reconstruction in our institution between 2011 and 2017, including both primary tumors and recurrences. Local disease control, metastatic recurrence, overall surveillance, postsurgical complications and functional and aesthetic result were analyzed.

Main results:

5 patients were included (4 with Ewing’s sarcoma and an embryonic rhabdomyosarcoma recurrence). All cases were advanced stages of the disease and were treated following our institution’s latest oncological protocol. The radiation dose varies between 7.5 and 15 Gy and all had chest wall reconstruction with different materials (goretex, titanium, resorbable plates, muscle flaps). With a mean follow-up of 109 weeks, local disease control was achieved in 100% of the patients. 3 patients presented complete remission, and 2 presented a distant metastatic recurrence. Cerebrospinal fluid fistula to pleura and a resorbable plate rejection were present as postoperative complications. Two patients developed long term postoperative scoliosis, and in all cases a good aesthetical result was achieved. Total survival was 80% (one death due to pulmonary metastasis).

Conclusions:

Our study supports IOERT as an effective therapy in the local control of the disease in thoracic wall tumors without leading to a poor evolution of the surgical reconstruction of the area.
Aim of the Study: The aim of the study was to analyze PRETEXT and POSTTEXT extent of hepatoblastoma with respect to survival and surgical complications.

Methods: From 2006 to 2016, 22 patients with HB had imaging data available and underwent surgery at our institution, the median follow up 4.49 (0.7 – 8.45) years. Event free survival (EFS) and overall survival (OS) was analyzed using Stat View program.

Main results: At diagnosis of HB, the median age was 1.44 (0-6.46) year. Eighteen out of 22 patients (82%) were treated with pre-operative chemotherapy, the rest underwent up-front liver resection. Pre-operative PRETEXT or POSTTEXT analysis: 6 patients had stage I, 10 stage II, 4 stage III and 2 stage IV. VPEFR staging was positive in 11 patients. Three patients died (1 in the pre-surgery PRETEXT/POSTTEXT tumor stage I, 1 in stage III, 1 in stage IV); 1 had local relapse and is alive, 1 had secondary malignancy, EFS is 77.3%, OS 86.4%. Any statistically significant differences in EFS, in term of pre-surgical stage of tumor (p=0.15), VPEFR positivity (p=0.66), radically resection (p=0.8), histology (p=0.45) was found, except of metastatic disease (p=0.006). Surgical complications (assessed in 20 patients, bleeding in 1, biliary leak in 3) occurred in 0/5 stage I, 3/10 stage II, 1/4 stage III, 0/1 stage IV (p=0.09); no difference was found in VPEFR factors.

Conclusions: We found no significant correlation between PRETEX/POSTEX stages, VPEFR positivity and event free survival in hepatoblastoma.

Supported MH CZ–DRO, Motol University Hospital, CZ 00064203
PW10-LG01
LONG-TERM OUTCOMES IN PEDIATRIC APPENDEICEAL CARCINOIDS

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Aim of the study
The tendency of nonoperative management of appendicitis let us explore the natural history of appendiceal carcinoids, compare them with appendicitis patients, and determine the possibility of deciding the extent of the surgery and post-operative follow-up on behalf of the intraoperative findings.

Methods
A retrospective review was performed of patients with appendicitis and appendiceal carcinoids between 2009 and 2017.

Main results
Of 2781 patients, 10 (0.36%) were diagnosed with appendiceal carcinoids. 60% were female with an average age of 13.10±1.73. The mean tumor size was 0.97±0.34 cm with 70% located at the tip. Majority had an insular pattern (n=9), six had subserosal fat tissue invasion, one had an extension to mesoappendix, one had a vascular invasion, and two had a lymphatic invasion. The average mitotic index was 3.20±1.40/50HPF, and the average Ki67 activity was 3±1.7%. 50% of the patients were pathologic grade 2. The mean follow-up period was 66.40 ± 25.92 months. All patients were further evaluated with ultrasonography (n=10), CT (n=3) and MRI (n=10). Serum markers including chromogranin (n=9), NSE (n=6) and 5-HIAA (n=6) were normal. None required further treatment and had any sign and symptoms of carcinoid syndromes or recurrences post-operatively.

Conclusions
Other than appendectomy, no additional surgery or follow-up is required in appendiceal carcinoids less than 1.5 cm in size, regardless of the lymphoid or vascular invasion. The surveillance of the larger tumors should be based on clinical findings of the patients.
Background: Enterocolitis (EC) is the most common and serious complication of Hirschsprung's disease (HD). The prognosis and outcome of EC depend on early diagnosis and effective management. The present study was established to evaluate the role of (C-159t) gene polymorphism biomarkers in patients developed such complication.

Methods: We conducted a prospective study for all the patients with Hirschsprung's disease complicating EC admitted and managed in our pediatric surgery unit, from the period 1st of January 2014 to the end of December 2016. All patients developing EC were evaluated with sepsis screen tests, Blood culture, subjected to broad-range PCR and PCR–restriction fragment length polymorphism (RFLP). The ethics committees of the health authorities approved the study.

Results: The distribution of (C-159t codon) polymorphism was detected by PCR-RFLP technique. At this locus there are three genotypes; homozygote (TT) at 353bp, heterozygous (CT) at 497bp, 353bp and 144bp, and wild-type (CC) which still undigested. TT genotype was significantly increased by 13.1 times for those patients with EC associated sepsis. Having a CC genotype significantly decreases the risk of EC associated sepsis by 5.9 times. Presence of T allele significantly increased the risk of having sepsis by 5.9 times in the patients with EC.

Conclusion: There is a significantly higher prevalence of detection of (C-159t) gene polymorphism among the Hirschsprung's patients associated EC. This gene may play a major role in the pathogenesis and EC development. More studies incorporating this gene are warranted.
PW10-LG03: UPFRONT AND INTERVAL APPENDICECTOMY FOR APPENDICULAR MASS HAVE LITTLE DIFFERENCE IN MAJOR OUTCOMES

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Aim of the Study:

There is no consensus in the management of paediatric patients presenting with Appendicular mass or Abscess. In this study we aim to compare between two management approaches for this condition, specifically Emergency Appendicectomy (EA) and Interval Appendicectomy (IA).

Methods:

In this retrospective study, we included 64 patients with Appendicular mass or Abscess operated in our Paediatric Surgery Department over three and a half years. These patients were classified into group I (IA) and group II (EA). Clinical characteristics & outcomes were investigated. The main outcomes were compared based on three parameters: the duration of hospital stay, the duration of intravenous antibiotics and post-operative morbidities. Data were analysed using MS Excel 2013.

Main Results:

Group I, represents 31.25% (n=20) and group II represents 68.75% (n=44). The average age is 8.9±3.7 years. No statistically significant difference in the duration of symptoms and other clinical characteristics (heart rate, temperature and CRP) between the two groups. Average duration of hospitalization in group I was found 11±3.7 days, versus 5.6±2.6 days in group II; while the average duration of IV antibiotics in group I, was 10.5±3.3 days versus 5.3±2.6 days in group II; both of these results are statistically significant. There was no statistically significant difference in the overall complication rates between the two groups.

Conclusions:

We found that EA as a treatment for an appendicular mass or abscess, is as safe as IA and is more cost-effective with shorter hospital stay and reduced duration of IV antibiotics.
PW10-LG04
LONG-TERM OUTCOMES AFTER PROCTOCOLECTOMY WITH ILEOANAL ANASTOMOSIS FOR PEADIATRIC ULCERATIVE COLITIS

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Aim of the Study

To evaluate long-term outcomes of proctocolectomy with ileoanal anastomosis (IAA) for ulcerative colitis (UC).

Methods

Medical records of children undergoing proctocolectomy with IAA for UC or unclassified inflammatory bowel disease (IBD-U) during 1985-2016 were retrospectively assessed. Risk factors for IAA failure, defined as ileostomy at last follow-up, were analysed with Cox regression.

Main results

Of the 87 patients, 85 (98%) had UC and 2 (2%) IBD-U preoperatively. Altogether 57 underwent two-stage and 30 three-stage procedures, and a pouch reconstruction was performed in 76 (87%) while 11 (13%) underwent straight IAA. Median age at surgery was 14.2 (interquartile range 12.3-16.1) years. During 7.8 (4.1-14.5) years’ follow-up, 9 (10%) patients were diagnosed with Crohn’s disease (CD). Postoperative leakages (n=8, 9%) and strictures (n=10, 11%) were equally common while fistulas (78% vs. 9%, p<0.001) and abscesses (56% vs 11%, p<0.001) more frequent among patients with later CD diagnosis. At last follow-up, eight patients (9%) of whom 7 had CD diagnosis had been converted to ileostomy while others reported daytime stooling frequency of 5 (4-7) and 0.5 (0-1) at night (Figure). CD diagnosis (hazard ratio, HR = 23.3, p<0.001), postoperative abscesses (HR 16.3, p<0.001) and fistulas (HR=20.9, p<0.001) as well as 3-stage surgery (p=0.004) increased risk for an ileostomy.

Conclusions

In children with UC, long-term outcomes after proctocolectomy with IAA are excellent. Need for 3-stage surgery, postoperative fistulas and abscesses, but not leakages or strictures, associate with postoperative
CD diagnosis and increase the risk for ileostomy.
PW10-LG05
EUROPEAN PAEDIATRIC SURGEONS’ ASSOCIATION SURVEY ON THE MANAGEMENT OF APPENDICITIS

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Aim of the Study: To define patterns in the management of appendicitis.

Methods: 169 delegates from 42 (24 European) countries completed a validated survey administered at the EUPSA 2017 annual meeting.

Results: In the work-up of children with suspected acute appendicitis, most surgeons rely on full blood count (92%), C-reactive protein (82%), and ultrasonography (76%), but rarely on CT-scans or MRI. In suspected simple appendicitis, most surgeons (76%) do not perform appendectomy at night in clinically stable patients, start antibiotic preoperatively (64%), but only 15% offer antibiotic therapy alone (no appendectomy). In suspected perforated appendicitis, 96% start antibiotic preoperatively and 92% perform an appendectomy. Presence of phlegmon/abscess is the main contraindication to immediate surgery. In case of appendix mass, most responders (75%) favour a conservative approach, and perform interval appendectomy always (56%) or in selected cases (38%) between 2 and 6 months from the first episode (81%). Children with large intraperitoneal abscesses are managed by percutaneous drainage by 59% and operated on by 37%. Laparoscopy is the preferred surgical approach for both simple (89%) and perforated appendicitis (81%). Most surgeons send the appendix for histology (96%) and pus for microbiology if found (78%). At the end of the procedure, 58% irrigate the abdominal cavity only if contaminated using saline solution (93%). In selected cases, 52% leave a drain in situ.

Conclusions: Some aspects of appendicitis management lack consensus, particularly appendix mass and intraperitoneal abscess. Evidence based guidelines should be developed which may help standardise care and improve clinical outcomes.
Purpose: Pilonidal sinus disease (PSD) is associated with a complex disease process in children, and its management remains controversial. There are few published studies on PSD in the pediatric literature; therefore, we present our experience with conservatively treating PSD in children.

Methods: This study involved a retrospective review with telephone follow up. All children diagnosed PSD in 2012–2017 were identified at the outpatient clinic of department of Pediatric Surgery. All patients initially underwent conservative treatment (meticulous hair removal, improved perianal hygiene, warm sitz baths, and drainage for abscess). Data collection included demographics, type of management, recurrence, presence of infection, and total healing time.

Results: In the study period, 29 children were identified. Their mean age was 14.94 ± 1.09 (range: 12–16) years, and 51.7% were girls. The mean length of follow up was 8.34± 6.36 (range: 1-25) months. Complete healing occurred in 79.3% of patients. Recurrence was evident in 12% of patients. Four patients underwent surgery.

Conclusion: PSD is being seen in children more frequently in recently. Conservative treatment may be the recommended initial approach for PSD in children.
**UNPLUGGING THE DIAGNOSIS – SHOULD RECTAL SUCTION BIOPSY BE PERFORMED IN ALL NEONATES PRESENTING WITH MECONIUM PLUG OBSTRUCTION?**

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**Aim of the study**
Limited data exists on the association and clinical overlap between meconium plug obstruction (MPO) and Hirschsprung’s disease (HD). The aim of this study was to determine the incidence of HD in children presenting with MPO.

**Methods**
All patients with MPO were identified through retrospective review of medical and radiological records between 2008 and 2017. The underlying diagnosis, demographics, clinical course, radiological findings, and long-term outcomes were recorded and analysed.

**Main results**
A total of 29 patients with MPO were identified. Of these, 5 were diagnosed with HD (17.2%) and 24 as meconium plug syndrome (82.8%). Neonatal rectal suction biopsy was performed in 22 of the 29 patients (75.9%), 1 patient had an open rectal biopsy in the postneonatal period (3.4%), and in 6 patients (20.7%) no rectal biopsy has been performed to-date. No patients were found to have cystic fibrosis, intestinal atresia, or malrotation.

**Conclusions**
In our series, a 17.2% incidence of HD was found in patients presenting with MPO. Although all diagnoses of HD were made within the neonatal period, delayed diagnosis has been reported in the literature. The results of our study support the use of rectal suction biopsy within the neonatal period in all patients with MPO. This is to ensure that underlying HD is identified promptly to reduce complications associated with delayed diagnosis.
PW10-LG08
ANORECTAL MALFORMATIONS AND PERINEAL HEMANGIOMAS: THE ARM-NET CONSORTIUM EXPERIENCE

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Aim
Perineal hemangiomas (PH) in patients with anorectal malformations (ARM) are rare but can pose a significant challenge and warrant particular attention. Surgical incision of posterior sagittal anorectoplasty (PSARP) may involve the hemangioma site resulting in hemorrhage, damage to blood supply, leading to dehiscence. The aim of this study was to review the experience of the ARM-net consortium in the management of PH associated with ARM and evaluate treatment strategies.

Methods
Data were collected retrospectively by questionnaire from all members of the ARM-net consortium. The cohort included all patients with ARM and a PH located in planes of dissection of PSARP.

Results
Ten patients, managed at eight colorectal centers, met the study criteria. Three patients each had rectobulbar or rectovestibular fistula, two had rectoperineal fistula and one rectoprostatic fistula; in one the hemangioma was too disfiguring to determining malformation type. Mean follow-up time was 35 months. Colostomies were constructed before definitive repair in eight patients. Five patients received systemic beta-blockers before PSARP; three were operated uneventfully following partial/complete involution of the hemangioma and two are awaiting surgery. The two patients with rectoperineal fistula were managed expectantly. The remaining three patients underwent surgery with no preoperative medical treatment, and all had complications: mislocated neoanus in two and complete perineal dehiscence in one.

Conclusions
Attempting PSARP in the presence of a PH may lead to complications and adversely affect outcome. This study confirms the benefits of beta-blockers treatments prior to surgical reconstruction.
EARLY VERSUS LATE MEGARECTUM IN ANORECTAL MALFORMATION

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Aims:
Anorectal malformation (ARM) associated megarectum has increased risk of complications and poorer functional outcomes. This study aimed to prove the hypothesis that early (preoperative) megarectum (EM) has higher morbidity than late (postoperative) megarectum (LM).

Methods:
We retrospectively reviewed 125 non-cloacal ARM who had undergone reconstruction in 2000-2016, and subsequent stoma closure, to identify megarectum based on radiological and surgical findings. Type of malformation, sacral ratio, and spinal cord anomalies were assessed for risk stratification. Primary end point was the need for excision of megarectum, quantified by binary logistic regression; secondary end points were stricture, faecal incontinence, constipation, and other surgeries. Continuous data were median (range). Statistics: Mann-Whitney-U-test, Fisher’s exact test.

Results:
22 patients with megarectums were included, 12 (55%) were females, 7 (32%) were EM; 15 LM were related to stricture or constipation. EM were found in low-type ARM (two were Currarino), but had comparable sacral ratio and spinal cord anomalies to LM.

EM was associated with higher megarectum excision than LM (4/7 vs 3/15, P=0.145). Binary logistic regression corrected for ARM anomaly type, sacral ratio of 0.52, and spinal anomaly, showed EM was associated with near-significant increased risk of megarectum excision (OR11.1, 95% CI 0.70,176, P=0.088).

EM and LM had comparable stricture (57%vs73%), incontinence (57%vs33%), constipation (100%vs87%), and need for further surgeries (86%vs80%), P>0.10.

Conclusion:
ARM-associated megarectum carries significant morbidity and need for further surgery. EM detected before reconstruction has a trend towards significant increased need for megarectum excision compared to LM.
PW10-LG10

GAPS IN PARENTAL KNOWLEDGE AND UNDERSTANDING OF APPENDICITIS

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Aims: Knowledge and understanding is required for parents to be able to make appropriate decisions regarding their children’s treatment and involvement in research. We aimed to assess parental knowledge regarding appendicitis and its treatment.

Methods: Parents attending children’s outpatients’ department over a 2 month period in 2017 were invited to complete a questionnaire that assessed knowledge about appendicitis and its treatment. Short educational statements were included to rectify any knowledge gaps. Ethical approval 17/EM/0344.

Results: Three hundred and ninety-six questionnaires were completed. There were significant parental knowledge gaps. Just 2/3 of respondents identified the anatomic location of the appendix in the correct abdominal quadrant, and fewer (57%) identified appendicitis as an inflammatory or infectious condition. Just under half correctly identified the incidence of appendicitis from a range of options (Figure). Parents significantly overestimated the risk of complications and death from appendicitis. For simple appendicitis 30% believed there is a ≥10% chance of significant complications and 5% believed there was a ≥25% chance of death. For perforated appendicitis 73% believed there was a ≥25% chance of significant complications and 30% believed there was a ≥25% chance of death. However, the majority (90%) knew that surgery was the most frequently used treatment for appendicitis.

Conclusions: There are noticeable knowledge gaps in parents’ understanding of appendicitis and evidence that parents overestimate the severity of the consequences of appendicitis. This information may help clinicians and researchers when communicating with parents regarding treatment options and involvement in research.

![How common is appendicitis?](image_url)
Aim of the study: To assess the frequency of, and identify contributing factors for wound dehiscence after posterior sagittal anorectal plasty (PSARP).

Methods: Ethical approval was obtained (2017/191). Charts of all children with anorectal malformations (ARM) reconstructed with PSARP at a tertiary centre of paediatric surgery between 2001–2016 were reviewed retrospectively. Wound dehiscence within 30 days postoperatively was analysed regarding gender, prematurity, birth weight, type of ARM, other congenital malformations, single- or multi-staged reconstruction, age and weight at reconstruction, postoperative antibiotics and fasting.

Main results: Ninety patients were included, 53 (59 %) males, 15 (17 %) born prematurely and 23 (26 %) small for gestational age. There were 41 (46%) with a perineal fistula, 28 (31%) had heart malformations, 35 (39%) urinary tract malformations, and 14 (15%) spinal malformations. Single-stage reconstruction was performed in 40 (44%) patients and 50 (56%) had a multi-stage reconstruction with a divided colostomy. Prolonged antibiotic prophylaxis (2-14 days) was used in 33 (37%) patients. Postoperative fasting for 0-3 days was used by 84 (93%) and for 4-7 days by 6 (7%) patients. Wound dehiscence occurred in 28 (31%) patients and was significantly more common among patients without a colostomy; 17 (43%) vs 11 (22%) (p=0.042). No other analysed demographical or surgical variables were proved to influence the rate of wound dehiscence.

Conclusions: A divided colostomy protected against wound dehiscence after PSARP. No other risk factor for wound dehiscence was identified. Further studies are needed to establish contributing factors to uncomplicated wound healing after PSARP.
Purpose: A complication of the surgical management of anorectal malformations (ARMs) is a retained remnant of the original fistula (ROOF) formerly called a posterior urethral diverticulum (PUD). A ROOF may have multiple presentations or may be incidentally discovered. We sought to define the entity and the surgical indications for excision of a ROOF when found.

Methods: Retrospective cohort review of all male patients who presented to our Center following previous repair for ARM at another institution to evaluate for problems with urinary and/or fecal continence, (2014-2017). Charts were reviewed for symptoms, original malformation, pre-operative imaging, treatment, and postoperative follow-up.

Results: Of 105 referred male patients, 13 had a ROOF. 11 underwent reoperation, and 2 await repair. 9 patients had an additional reason for a redo such as anal mislocation or rectal prolapse. Indications for ROOF excision were for associated urinary symptoms, to make a smoother posterior urethra, to help with intermittent catheterization, or for prophylactic reasons. All patients were repaired at an average age of 4.2 years, using a PSARP approach with excision of the ROOF. Preoperative evaluation included pelvic MRI, VCUG, and cystoscopy. Urinary symptoms associated with a ROOF and ease of catheterization were improved in all repaired cases.

Conclusion: The high incidence of ROOF requires evaluation of male ARM patients with urinary symptoms and includes both an MRI and cystoscopy. The ROOF related symptoms can be improved with excision. The theoretical oncologic risk requires prophylactic excision of a ROOF.
PW11-UG01
ENDOSCOPIC TREATMENT OF PEDIATRIC PRIMARY MORBID OBESITY USING INTRAGASTRIC BALLOON: SHORT AND MID-TERM RESULTS

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Aim of the Study:
Obesity is the most prevalent nutritional disorder among children and adolescents leading to several comorbidities. Intragastric balloon (IGB) is a temporary endoscopic treatment totally reversible and repeatable. We retrospectively analyzed short/mid-term results of obese pediatric patients treated by IGB at our Institution.

Methods:
Eighty-nine patients, mean age 15.8 years (range 8-19) underwent to IGB endoscopic insertion for severe obesity (BMI>35). A liquid-filled nonadjustable IGB till a final volume of 600 ml of saline solution was used for a period of 6 months. Seventy-one patients (81%) had one or more associate comorbidities [hyperinsulinemia 24 patients (38%), steatosis 45 (51%), dyslipidemia 9 (10%), nocturnal apnoea 6 (7%), hypertension 13 (15%)].

Main results:
Mean follow-up was 17 months (range 6-32). There were no serious complication or death. Mean BMI, at the end of the treatment, decreased from 38.74±0.6 to 32.25±0.4 (p<0.05), mean weight from 101,8±18.5 Kg to 93,8±16.9 Kg (p<0.05), while, after the follow-up mean BMI was 33.7±3.6 (p<0.05) mean weight 93.7±0.9 Kg (p<0.05). Nine-teen patients (20%), solved their comorbidities.

Conclusions:
The IGB is associated with significant short/mid-term weight loss with no serious complications. Intragastric Balloon may have a potential role as a stand alone procedure or may be used in preparation for a subsequent bariatric surgery in extreme obese patients, reducing both anesthesiological and surgical risks. The IGB is safe and effective allowing also the resolution of the co-morbidities in the 20% of cases.
PW11-UG02

DIAGNOSTIC WORKUP OF NEONATES WITH OESOPHAGEAL ATRESIA: RESULTS FROM THE EUPSA OESOPHAGEAL ATRESIA REGISTRY

Francesco Morini, Andrea Conforti, Augusto Zani, Sanja Sindjic-Antunovic, Antti Koivusalo, Florian Friedmacher, Ernest van Heurn, Agostino Pierro, Michael Hollwarth, Pietro Bagolan, on behalf of the EUPSA Oesophageal Atresia Registry

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Aim of the study: Controversies exist on the optimal diagnostic workup for neonates with oesophageal atresia (OA) with/without trachea-oesophageal fistula (TOF). Aim of this study was to describe the current diagnostic policies in OA/TOF patients enrolled in an International multicentre registry. Methods: All patients consecutively registered from July 2014 to December 2017 in the EUPSA Oesophageal Atresia Registry (EUPSA-OAR) were included in the study. The difference in prevalence of diagnostic studies among Centres forming the EUPSA-OAR was analysed. Chi-square test or Fisher’s exact test were used as appropriate. Results: During the study period, 374 patients were consecutively recorded by 24 Centres. Table shows main findings. Chest X rays, cardiac ultrasound, abdominal ultrasound, and abdominal X rays were granted to the majority of patients. Preoperative bronchoscopy and oesophageal gap measurement were performed in about one third of the patients. Conclusions: Present data from a large cohort of patients from the EUPSA-OAR show both inter-Institutional and intra-Institutional variability in diagnostic workup of patients with OA/TOF. Efforts should be made to develop shared guidelines on diagnostic workup for OA/TOF patients.

<table>
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<th>Study</th>
<th>Patients (%)</th>
<th>Median</th>
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<tr>
<td>Chest X-ray</td>
<td>372 (99%)</td>
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<td>(100-100)</td>
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<td>Cardiac US</td>
<td>365 (98%)</td>
<td>100</td>
<td>(100-100)</td>
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<tr>
<td>Abdominal US</td>
<td>353 (94%)</td>
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<td>(90-100)</td>
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<td>Gap measurement</td>
<td>112 (30%)</td>
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<td>(0-27)</td>
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<tr>
<td>Gap in VB</td>
<td>80 (71%)</td>
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<td>(10-72)</td>
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<td>Chromosomal analysis</td>
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<td>10-46</td>
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<td>Esophagoscopy</td>
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PW11-UG03
SURGICAL MANAGEMENT OF GASTROESOPHAGEAL REFLUX IN CHILDREN WITH NEUROLOGICAL IMPAIRMENT

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Aim of the Study
To evaluate neurologically impaired children with gastroesophageal reflux (GER) who underwent antireflux surgery (ARS) between January 2011 and June 2016 in our clinic retrospectively.

Methods
Eighty-four patients were reviewed. Data including patient demographics, indications for surgery, preoperative diagnostic tests, additional surgical procedures, complications and reoperations were collected. The study was approved by the Ethics Committee (09.2016.496).

Main Results
There were 50 boys and 34 girls with a mean age of 44.4 months. The most frequent underlying pathology was cerebral palsy (67.4 %). Main indication was recurrent aspiration pneumonia (77.1 %). Fifty-three patients were unable to feed orally. Preoperative diagnostic tests were upper gastrointestinal series (78.6%), gastric emptying study (69%), endoscopy (8.3%) and pH monitoring impedance (21.4%). Type of surgical procedure was laparoscopic Nissen fundoplication (LNF) and gastrostomy in 61 patients, only LNF in 12 patients, open Nissen fundoplication (ONF) and gastrostomy in 5 patients. LNF and pyloroplasty (PP) in 3 patients and LNF, gastrostomy and PP in 3 patients. In the postoperative period 58.3 % of patients were symptom free. Only one patient died in the early postoperative period due to nonsurgical reason. Early postoperative complications occurred in 6 patients. Two patients underwent redo NF and gastrostomy was added in one patient.

Conclusions
Children with neurological impairment and GER are difficult to treat. The main indication for ARS is recurrent aspiration pneumonia. These patients may also have delayed gastric emptying and may need pyloroplasty. Postoperative success rate is relatively low. Complication and reoperation rate is low.
**PW11-UG04**

**PARTIAL GASTRECTOMY FOR SEVERE GASTROPARESIA IN CHRONIC INTESTINAL PSEUDO-OBSTRUCTION**

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**Aim of the study:** Gastroparesis is characterized by delayed gastric emptying in the absence of obstruction and may lead to feeding intolerance requiring total parenteral nutrition, particularly in chronic intestinal pseudo-obstruction (CIPO). Gastroparesis Cardinal Symptoms Index (GCSI) was developed to evaluate the severity. The aim was to assess the efficacy of partial gastrectomy in severe gastroparesis secondary to CIPO.

**Methods:** Patients with CIPO who underwent partial gastrectomy for gastroparesis between 2005 and 2015 were included. Efficacy of gastrectomy on preoperative symptoms was assessed using the GCSI score, evaluating 9 clinical symptoms (nausea, retching, vomiting, stomach fullness, not able to finish normal-sized meal, feeling excessively full after meals, loss of appetite, bloating, stomach visibly larger) from “none” to “very severe”.

**Main results:** Three patients underwent partial gastrectomy at 3, 15 and 17 years. Preoperative GCSI scores were 38/45, 35/45 and 35/45. All had total parenteral nutrition with failure of intensive medical management before surgery. Postoperative GCSI scores decreased to 13/45, 11/45 and 11/45 with a follow-up of 29 months, 10 years and 6 months, respectively. Parenteral nutrition was discontinued 12 months and 3 months for the 2 first patients. At last follow-up, part of oral intakes/parenteral nutrition (expressed as percentage of total daily caloric intakes) was for each patient respectively 85%/15%, 60%/40%, 80%/20%.

**Conclusions:** Partial gastrectomy should be considered in severe gastroparesis in CIPO failing medical therapy to improve quality of life with reduction of parenteral supplementation and restoration of oral intake.
AIM: Small bowel bacterial overgrowth (SBBO) is a challenge in the management of pediatric intestinal failure (IF). Our goal was to determine the proportion treated for SBBO by an intestinal rehabilitation program and factors related to its development.

METHODS: A retrospective analysis of IF patients referred between 2008-2014. Data was collected on factors related to IF and SBBO. The cohort was stratified on the diagnosis of SBBO and refractory SBBO. Statistical testing completed using T-test, Chi square and logistic regression.

RESULTS: 35/102 patients developed SBBO (34%) and 16 (16%) had refractory SBBO. Gastroschisis patients were more likely to develop SBBO (40.0 vs 19.4%, p=0.025), were more likely in those with shorter residual small bowel (SB) (45.4 vs 66.5%, p=0.004) and patients with SBBO were less likely to wean from parenteral nutrition (PN) (51.4 vs 85.1%, p<0.0001). Refractory SBBO patients were likely to have gastroschisis (50.0 vs 22.1%, p=0.020), have a shorter residual SB and LB remaining (23.2 vs 65.9%, p<0.0001 and 60.6 vs 79.4%, p=0.03) and less likely to wean from PN (37.5 vs 80.2%, p=0.001). Logistic regression demonstrated that longer SB residual was protective (p=0.001, OR 0.95; CI 0.93-0.99) and SBS as a cause of IF was a risk factor (p=0.001, OR 0.95; CI 0.93-0.99) and SBS as a cause of IF was a risk factor (p=0.001, OR 0.95; CI 0.93-0.99) and SBS as a cause of IF was a risk factor.

CONCLUSION: A longer SB remnant was protective against SBBO. Patients with SBBO were more likely to have IF caused by SBS rather than primary dysmotility or mucosal enteropathy. The presence of the ileocecal valve or method of delivery did not predict development of SBBO.
Our 12 Years’ Experience of Esophageal Replacement Surgeries

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Aims of the Study: To describe our experience of esophageal replacement in terms of clinical scenarios requiring replacement, its technical details, complications encountered and their management, outcome and follow-up.

Methods: The retrospective case series (IRB approval taken) was conducted between Jan 2005 and Dec 2017 by reviewing the medical record of all the patient undergoing esophageal replacement surgeries during the study tenure.

Results: A total of 78 patients (Males, 51(65.38%); Females, 27 (34.61%) M:F 1.8:1) underwent esophageal replacement surgeries. Esophageal replacement was done due to corrosive esophageal strictures, long gap esophageal atresia. Esophageal replacement was done by Gastric Transposition/pull up in 73 cases (93.59%), Colon interposition in 03 cases (3.85%), and Jejunal interposition in 02 cases (2.56%). The route of esophageal replacement was trans-hiatal in 65 (83.33%), retrosternal in 06 (7.69%), and trans-hiatal with thoracotomy in 07 (8.97%) cases. Majority of the patients tolerated the replacement well. Regarding complications there was no graft necrosis; wound infection was encountered in 7 (8.97%), wound dehiscence in 4 (5.13%), anastomotic leak in 8 (10.26%), anastomotic stenosis in 8 (10.26%), fistula formation in 1 (1.28%), dumping syndrome in 4(5.13%), reflux in 7(8.97%), and dysphagia in 9 (11.54%) patients. These complications were dealt accordingly. One patient needed revision of colonic interposition. Ten (12.82%) patients expired in our series.

Conclusion: We have described our experience from a resource constrained set-up. Gastric conduit is the best conduit for esophageal replacement as to technical aspects. Complications are not uncommon and need respective management.
PW11-UG07
RISK FACTORS OF DUMPING SYNDROME IN CASES AFTER TOUPET FUNDOPLICATION

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Shizuoka Children’s Hospital, Shizuoka, Japan

Aim of the Study
In postoperative cases of fundoplication, gastric emptying ability is promoted and sometimes exhibits dumping syndrome. It is reported that the necessary water amount and the energy requirement is large for physique, the retention capacity is decreased due to shrinkage of the fundus of the stomach due to fundoplication, and the retention time is shortened due to immaturity of the pyloric function in childhood. We examined the risk factors of dumping syndrome after toupet fundoplication.

Methods
We studied 190 patients who were performed toupet fundoplication during 2003 to 2017, of which 157 cases were laparoscopic surgical cases. Regarding the risk factors of dumping syndrome, gender, age at the time of surgery, body weight at the time of surgery, Cerebral palsy, severe scoliosis, microgastria, chromosomal abnormalities, and complex cardiac anomalies were retrospectively studied.

Main results
After surgery, 16 patients (8.5%) presented dumping syndrome. Significant risk factors of dumping syndrome for univariate analysis were those who underwent surgery within 8 months of age (crude OR 5.7), severe scoliosis (crude OR 6.8), microgastria (crude OR 26.5), and major cardiac anomaly (crude OR 3.6). Significant risk factors of dumping syndrome for multivariate analysis were those who underwent surgery within 8 months of age (adjusted OR 10.2, 95% CI: 2.6 - 45.2), severe scoliosis (adjusted OR 19.3, 95% CI: 4.4 - 91.1), and microgastria (adjusted OR 26.5, 95% CI: 1.4 - 896.4).

Conclusions
Cases with surgery within 8 months of age, patients with severe scoliosis, microgastria were high-risk group of dumping syndrome.
PW11-UG08
META-ANALYSIS OF GASTROSTOMY IN CHILDREN BY USING LAPAROSCOPY-ASSISTED OR PERCUTANEOUS ENDOSCOPIC TECHNIQUE

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Aim of the study: To conduct a metanalysis to compare the rate of major complications associated with the two most used techniques for gastrostomy placement in children i.e. Laparoscopy-Assisted Gastrostomy (LAG) and Percutaneous Endoscopic Gastrostomy (PEG).

Methods: Major electronic databases were queried for comparative studies of the two techniques in publications from 2002 through 2017. The main outcome measure was the frequency of serious postoperative complications defined as reoperation within 30 days or death. Screening of eligible studies, data extraction, and assessment of methodological quality were conducted. Forest and funnel plots were generated for outcomes using RevMan 5.1, with p<0.05 considered significant.

Main results: Ten reports with a total of 2020 patient met the inclusion criteria. Major complications were reported in 16 of 1012 (1.6%) compared with 80 of 1008 (7.9%) children undergoing LAG and PEG respectively. Studies comparing LAG and PEG revealed a significantly lower risk for major complications with LAG, n=10 studies, OR 0.19 (95% CI 0.11 – 0.32) p<0.001. During the study period all the publications reported lower frequencies of major complications when using LAG compared with PEG. The number needed to treat to reduce one major complication by abandoning PEG was 25-50. None of the studies analyzed was a randomized controlled trial which constituted a bias.

Conclusions: In children LAG was associated with a decreased risk of major complications compared to PEG, which should be avoided in children.
PW11-UG09
DIFFERENCES OF GENERIC HEALTH-RELATED QUALITY OF LIFE AFTER REPAIR OF ESOPHAGEAL ATRESIA IN A GERMAN-SWEDISH COHORT

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Aim

Despite advances of outcomes of esophageal atresia (EA), knowledge on patients’ health-related-quality-of-life (HRQOL) is sparse. Due to heterogeneity of EA, large cohorts need to be investigated to ensure reliability of data. Endpoint was generic HRQOL after EA-repair.

Methods

After ethical approval, 192 patients (2-18 years;134 Swedish;58 German) were included. Clinical data were reviewed. EA was classified (“severe” and “mild-moderate”). PedsQL 4.0 was used in appropriate versions (2-7 years;8-18 years;self- (SR) and proxy report(PR)) to determine generic HRQOL.

Results

Swedish and German samples were clinically and demographically comparable. HRQOL was lower in “severe EA” vs. “mild/moderate” (total score;PR 85.61 vs. 73.57;p<0.001) and Gross A vs. Gross C-type EA (2-7 years;total score;PR 60.97 vs. 79.25;p 0.035). Total HRQOL was higher in Swedish vs. German sample (PR 82.27 vs. 72.67;p 0.002). HRQOL was impaired in German sample vs. healthy population (total score;PR 72.67 vs. 82.70;p 0.001). In German patients (8-18 years), HRQOL was higher in SR vs. PR (80.66 vs. 74.71;p 0.044). Patients’ age and presence of VACTERL/ARM didn’t affect HRQOL. Various differences were detected regarding different dimensions of PedsQL.

Conclusions

In this first international study we found several differences in perception of generic HRQOL. HRQOL appears to be determined by type of EA rather than patients’ age or the presence of typical associated malformations. Country-specific differences may be culturally dependent, but further investigations are suggested. A condition-specific instrument validated for EA may provide additional insights.
INTESTINAL INFLAMMATION IN CHILDREN WITH SHORT BOWEL SYNDROME

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Aim of the Study: Short bowel syndrome (SBS) has been associated with intestinal inflammation. We studied histological mucosal inflammation in children with SBS.

Methods: Thirty-six patients at median age 2.5 years (IQR 1.1-4.1) and thirteen age-matched controls underwent endoscopic duodenal biopsies. At 2.2 years (0.4-4.4) after last intestinal resection fourteen (29%) patients were on PN. Twenty-two had weaned off PN after 2.6 years (1.1-4.1). Mucosal inflammation was assessed by histological inflammation grade (1-4) of lamina propria and intraepithelial leukocytes count (IELC)/100 enterocytes.

Main results: The median small bowel length was significantly shorter [18% (9-22) vs 29 (23-45), P<0.001] and duration of PN longer [32 months (13-102) vs 6 (2-11)] in patients on PN. SBS patients had comparable inflammation grade [2.00 (1.67-2.67) vs 1.67 (1.33-2.17), P=0.130] and significantly lower IELC [0.07 (0.04-0.09) vs 0.09 (0.07-0.14), P<0.05] than controls. Both inflammation grade [2.00 (1.58-2.42) vs 2.33 (1.58-3.00), P=0.441] and IELC [0.07 (0.05-0.09) vs 0.06 (0.04-0.09), P=0.516] were similar in PN and weaned off patients. Age, post-resection time and time after weaning off PN associated positively with inflammation grade and IELC (r=0.435-0.519, P<0.05), and remaining ileum length inversely with IELC (r=0.495, P=0.022) after weaning off PN. Absence of the ileocecal valve (ICV) associated with increased IELC only in weaned off patients [0.08 (0.06-0.14) vs 0.05 (0.04-0.08), P=0.014].

Conclusions: No abnormal duodenal mucosal inflammation was observed in children with SBS either during or after weaning off PN. However, absence of ileocecal region associated with increased intraepithelial inflammation after weaning off PN.
PW11-UG11
GER IN EA/TEF: SOME COMPLICATIONS DURING SURGICAL CORRECTION MAY BE RELATED TO THE NEED OF FUNDOPPLICATION

Marta Martos-Rodríguez, Carles Giné Prades, Ana Laín Fernández, Laura García Martínez, Carlos Leganés Villanueva, Patricia Barila Lompe, Marta Pilar Martín Giménez, Manuel López Paredes
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INTRODUCTION

Fundoplication in patients with EA/TEF has a high failure rate mainly because of the short esophagus and the inherent motility dysfunctions. As complications during primary anastomosis may influence on those causes, we aim to correlate them with the need for fundoplication.

METHODS

Single-center retrospective analysis of patients with EA/TEF that benefited from a fundoplication during a 7-year period. The variables were: complications after primary anastomosis (early stenosis, leakage), need for fundoplication and complications (recurrence, valve migration and RE DO surgery). Contingency tables and Pearson’s Chi-square was used for the statistical analysis.

RESULTS

From a total of 73 EA, 62 (3 type 1, 51 type 3, 6 type 4 and 2 type 5) were evaluated excluding those with esophageal substitution and neonatal death. Complicated anastomosis was found in 29 (46.8%). Reflux was identified in 37 (59.7%) and fundoplication was indicated in 18 (29%). Mean age at intervention was 4.21 years. Nissen fundoplication was performed in all cases. We found 11 complications in 6 (33.3%) patients: valve migration 5 (27.7%), GER recurrence 6 (33.3%). 4 (22.2%) patients needed a RE DO procedure. Between those EA with or without fundoplication, the stenosis rate was 44.4% vs 20.5% (p=0.05) and leakage 16.7% vs 20.5% (NS). In failed fundoplication, the complication rate on primary anastomosis was no different from the successful fundoplication patients (NS).

CONCLUSION

Early stenosis of primary anastomosis seems to be related to the further need for fundoplication in patients with EA, but not to the potential failure of this fundoplication.
LONG-TERM FOLLOW UP AFTER SURGERY FOR ESOPHAGEAL ATRESIA: SOMATIC AND NEURO-BEHAVIORAL DEVELOPMENT IN PRE-SCHOOL AGED CHILDREN

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AIMS OF THE STUDY. Children treated for esophageal atresia (EA) are notoriously at risk for somatic growth delay. Less investigated are mid-long terms effects on neuro-psychomotor function. Increased incidence of anxiety disorders and need of scholastic support are reported in school-aged and adolescent patients. We aimed to evaluate neuro-behavioral development of pre-school EA patients.

METHODS. Parents of 11 EA patient (6 males, 5 females – 10 type C, 1 type A) out of 18 operated in our Unit from 2012 to 2015 agreed to participate to the study. Somatic growth (gestational age, weight, length at birth, 1-3-6-12-24 months) and neuro-behavior development (Child Behaviour Check-List – CBCL version 1½-5 years) were evaluated. Somatic growth of patients was compared to CDC charts (t-test, p significant<0.05). CBCL results of patients were compared to those of 12 age-matched healthy children (Mann-Whitney U-test, p significant<0.05).

MAIN RESULTS. Patients presented a significant constant difference on weight growth, mainly in males (FIG). CBCL results evidenced sub-clinical ranges for EA patients in all subcategories and no differences versus controls. DSM-oriented scales resulted within normal ranges in all patients, but a significant difference versus control was evident for Oppositional Defiant Problems (mean overall score: EA patients 3.0 vs controls 1.0, p=0.01).

CONCLUSIONS. Our small cohort of EA patients is not at significant risk for neuro-behavior problems in pre-school age, but presents a significant sub-clinical major incidence of a mental disorders considered precursor of anxiety disorder. If confirmed in longitudinal larger follow up studies, this evidence could require preventive strategies.
OPERATIVE TIME AND LENGTH OF HOSPITAL STAY FOLLOWING LAPAROTOMY OR LAPAROSCOPY IN INFANTS AND CHILDREN: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Aim of the Study: The aim of the study was to determine whether laparoscopy is associated with shorter operative time (OT) and length of hospital stay (LOS) compared with laparotomy for gastrointestinal procedures in infants and children.

Methods: Using a defined search strategy (PubMed, Medline, OVID, Embase, Cochrane databases), two investigators independently identified all comparative studies reporting OT and LOS following laparotomy or laparoscopy in infants and children. A meta-analysis was conducted using RevMan 5.3. Data expressed are mean[SD].

Main results: Of 13,189 titles or abstracts screened, 680 full-text articles were analyzed and 165 comparative studies (720,337 children, 10 randomized controlled trials, RCTs) were included. Overall, OT was shorter at laparotomy (99.2[77.1] minutes) compared to laparoscopy (116.5[91.4] minutes; p<0.00001), as confirmed by RCTs included (Figure). However, laparoscopy had similar OT to open pyloromyotomy for hypertrophic pyloric stenosis (HPS, 29.9[8.5] versus 30.4[6.8] minutes; p=ns), and shorter OT than open Ladd’s procedure for intestinal malrotation (58[7.1] versus 72[5.7] minutes; p<0.00001). Patients undergoing laparoscopy had shorter LOS (4.9[3.6] days) compared those who had open procedures (6.8[4.8] days; p<0.00001). This outcome was consistent in all procedures, except for pyloromyotomy to treat HPS (open 2.4[1.0] versus laparoscopy 2.3[0.9] days; p=ns).

Conclusions: This meta-analysis shows that laparoscopy for gastrointestinal procedures is advantageous in reducing the LOS in infants and children. Conversely, the OT of laparoscopy is longer than that of open procedures. These findings are supported by a high level of evidence in the literature.
PW12-G02
LONG TERM RESULTS ON USE OF OMEGA-3 FATTY ACID IN PEDIATRIC PATIENTS WITH SHORT BOWEL SYNDROME

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Introduction

Parenteral nutrition (PN) associated liver disease remains as a major comorbidities in patients with short bowel syndrome (SBS). Use of omega-3 fatty acid (Omegaven) has been proposed as a rescue therapy with promising results. However the long term outcomes is still not fully investigated. Herein, we try to summarize our experience on the use of Omegaven in SBS patients on a long term follow up basis.

Methods

13 patients with SBS who had been given Omegaven as part of the PN were followed up for 5 years. Patients’ demographics, operations performed, remaining bowel length, the PN regimen given, liver function test results and growth parameters were studied.

Results

There were 8 boys and 5 girls. The median gestational age is 30 weeks with birth weight of 1.53kg. 12 patients suffered from necrotizing enterocolitis and one patient from midgut volvulus. The median small bowel length remaining was 25cm (range 9.5 to 59cm). 10 patients tolerated full enteral feeding and weaned off from PN at median age of 11 months. All patients achieved jaundice clearance in 5 months after using Omegaven. Bilirubin and liver enzymes level remained normal in all patients since 1 year of age. Interval ultrasonography assessment did not show any cirrhotic change or splenomegaly. Patients have adequate weight gain till the end of study period.

Conclusion

Use of Omegaven in SBS patients is safe and showed good long term outcomes. It has the potential to reduce the incidence of PN associated liver disease in SBS patients.
PW12-G03
LAPARO-ENDOSCOPIC SINGLE-SITE APPROACH IN PAEDIATRIC SURGERY: SEVEN YEARS OF EXPERIENCE

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Aim of the study: to give an overview of the surgical interventions eligible for laparo-endoscopic single site (LESS) approach in paediatric surgery.

Methods: retrospective analysis of all surgical interventions performed through a LESS approach in a single centre, excluding appendectomies.

Main results: Eighty-one patients have had LESS procedures since 2011. Fifty-four procedures consisted in organ resections: cholecystectomies (29), colectomies (8), nephrectomies (7), ovariectomies or ovarian cyst resections (7) and splenectomies (3). Among them, all cholecystectomies and some nephrectomies required the use of an additional 2 mm percutaneous clamp for a better exposure. The 27 other procedures with suturing were Nissen fundoplications (14), gastric banding (5), vascular hitch (4), Heller myotomies (3) for mega-oesophagus, and gastropexy (1) for gastric volvulus. The JAIMY™ robotized needle-holder was used for 18/27 (67%) of those cases. An additional 5 mm trocar was required in 25/81 (31%) cases and 2 trocars in 6/81 (7.4%) cases. Five (6.2%) interventions were converted into traditional laparoscopy and no laparotomy was performed. We experienced 5 (6.2%) per-operative complications: 3 gallbladder perforations during cholecystectomy, one intestine perforation during umbilical opening, and one gastric plication ischemia during a Nissen procedure, requiring repositioning through conventional laparoscopy.

Conclusions: the miniaturization and robotization of laparoscopic instruments now allows for LESS approach in paediatric surgery procedures through a wide range of digestive, urologic and gynaecologic indications. Surgeons’ gain of experience will decrease the number of conversions and need for additional trocars, converging towards a "zero scars" paradigm.
HOSPITALISATION VERSUS EMERGENCY DEPARTMENT MANAGEMENT OF INTUSSUSCEPTION IN CHILDREN: A SYSTEMATIC REVIEW AND META-ANALYSIS OF OUTCOMES

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¹National University of Singapore, Singapore, Singapore. ²KK Women’s and Children's Hospital, Singapore, Singapore. ³Duke NUS Medical School, Singapore, Singapore

Aim: Recent literature advocates outpatient emergency department (ED) management of intussusception citing low recurrence rates and post-reduction events after uncomplicated ileo-colic reduction. However, few studies include both inpatient and outpatient cohorts. We performed a systematic review and meta-analysis to compare outcomes between the groups.

Methods: Studies published in English until Jan 2018 were searched from Medline, Embase, and Cochrane databases, using a combination of the terms ‘intussusception’, ‘reduction’ and ‘management’. A meta-analysis of studies comparing outcomes after successful intussusception reduction in children between inpatients and ED patients was performed.

Results: No randomized controlled trials were found. Nine observational studies (8 retrospective, 1 prospective) were included, comprising 546 inpatients and 776 ED cases. There was no statistical difference in overall recurrence rate between inpatients (8.8%) and ED (10.1%) [pooled odds ratio (OR)=1.09;95% confidence interval (CI) 0.74 to 1.62; P=0.66; I²=0]. Each of the following outcomes were reported by 5 studies (different studies per outcome) without significant differences: early recurrence (<48 hours) [pooled OR=1.27;95% CI 0.46 to 3.48;P=0.65;I²=0], post-discharge recurrence (pooled OR=1.57;95% CI 0.71 to 3.48;P=0.27;I²=34%); and recurrence requiring surgery (pooled OR=0.99;95% CI 0.32 to 3.06;P=0.99;I²=0). Methods of reduction were air, barium or other contrast enema. All studies described protocols with exclusion criteria for ED management of intussusception.

Conclusions: Emergency department management of intussusception after uncomplicated reduction appears acceptable. However, evidence levels are low and randomized controlled trials should be performed to evaluate its safety.
PW12-G05: PRELIMINARY EXPERIENCE WITH A THREE-DIMENSIONAL HIGH-DEFINITION ENDOSCOPE FOR MINIMALLY INVASIVE SURGERY IN CHILDREN

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¹IRCCS Burlo Garofolo, Center for Mother and Child, Triest, Italy. ²Regina Margherita - Children’s Hospital, Turin, Italy. ³Pediatric Surgery Department – Schneider Children’s Medical Center of Israel, University of Tel Aviv, Tel Aviv, Israel. ⁴Pediatric Surgery Department – Soroka University Medical Center, Ben Gurion University, Beer Sheva, Israel

Introduction
In paediatric surgery only a few experiences with a 3D system are presented. We could show in an earlier experimental study that 3D visual information helps inexperienced surgeons to learn faster complicated procedures. To create a 3D image, systems require a dedicated monitor and passive-polarised glasses. This pilote study is focusing on the application of a small size 3D 4mm endoscope (Visionsense, Neuromed Spa), initially designed for ENT and Neurosurgery in small cavities.

Material and Methods
In 2 centers. 26 patients (age 1month - 17yrs) underwent thoracoscopic, laparoscopic and retroperitoneoscopic procedures. All 14 surgeons answered a detailed questionary concerning the subjective feeling and experience, side effects, neuro-sensory feeling, the visional quality and the impact on procedures.

Results
All procedures were successfully finished (OR time 45min - 235min), 4mm scope , 0-40°; no complications ore technical problems occured. Results of questionaries (n=14): 15% fatigue for eyes; no nausea or headache. 80% better view and illumination; 90% positive effect on learning, tissue handling and intracorporal suturing. 80% believe in a future of this technique, 70% are favourable for its use in neonatal and small cavity surgery. 60% believe that this technique is expensive and needs refinement.

Conclusion
This 3D system can be applied safely and effectively in complex procedures. The image is accurate with a great depth perception, perfect lighting. Our study confirms a generally positive impression and a very positive judgement by most surgeons. Nevertheless the system is still expensive and needs a further adaption for our needs.
**PW12-G06**

**GYNECOMASTIA IN ADOLESCENTS: MAJOR IMPROVEMENT IN SELF-ESTEEM AND BODY-IMAGE AFTER SURGERY**

Javier Serradilla, Alba Bueno-Jiménez, Miriam Miguel, Esaú Fernández-Pascual, Mariela Dore, Paloma Triana-Junco, Javier Jiménez, Mercedes Díaz, Juan Carlos López-Gutiérrez

*Hospital Universitario La Paz, Madrid, Spain*

**Aim**

The prevalence of gynecomastia is up to 65% in adolescents. Although it regresses spontaneously in most cases, it sometimes persists causing body-image disorders. Surgery is a well-established treatment for these patients. We assessed the impact of surgery on the self-image of adolescents with gynecomastia and their overall postoperative satisfaction.

**Methods**

Sixty adolescents (12-18 years) who had surgery for gynecomastia between 2010-2017 were asked to complete an anonymous, online questionnaire (a shortened version of the Nuss paediatric questionnaire for thoracic malformations). It consists of 15 questions about self-perception, pre and postoperative self-esteem and general satisfaction with surgical results.

**Results**

The response rate was 63% (38). Thirty-two patients (84%) improved their overall happiness and their body-image after surgery. Twenty-eight (74%) reported changes in their social life, being able to face social situations that they previously avoided. There was a high level of satisfaction after surgery. When compared with the preoperative period, patients showed postoperatively an improvement of 4 points ([1-9], p<0.001) in a ten-point self-esteem scale. Only eight adolescents (21%) reported being unhappy with the scars. Nonetheless, up to thirty-six (95%) referred they would happily undergo surgery again.

**Conclusions**

Mastectomy is an effective treatment for gynecomastia with high levels of postoperative satisfaction. It significantly improved patients’ body-image and self-esteem. Our modified version of Nuss questionnaire proved to be useful to measure patient satisfaction. Therefore, surgical treatment should be offered to all adolescents with symptomatic gynecomastia. Concurrently, we cannot obviate the increasing prevalence of gynecomastia; addressing epidemiological issues is mandatory.
RADIOLOGICAL RISK FACTORS OF RELAPSE AFTER MEDICAL TREATMENT FOR UNCOMPLICATED APPENDICITIS: A RETROSPECTIVE STUDY

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Aim
Non operative managment (NOM) for uncomplicated appendicitis (UA) can be a potential good alternative for some selected patients with a 10 to 15% recurrence rate. The aim of our study was to determine risk factors of recurrence according to ultrasound (US) features at diagnosis and 7 days after the beginning of the treatment.

Methods
A monocenter retrospective study including all patients treated for UA from March 2014 to May 2017 has been performed. Patients treated were included according to the previous published study. Statistical analysis included X2 test for non parametric features and T test for parametric datas.

Main results
298 patients were included (120 girls, 178 boys) with a mean age at diagnosis of 9.9 years old. Among them 61 patients presented with a relapse after 5,2 months and a mean follow up of 23 months. No clinical or US features at diagnosis have significant odd ratios. Analysis of the US scan at day 7 indicate that increased parietal thickness (>2.5mm, p=0.0033, OR=2.3), local hyperhemia (p=0.0319, OR=1.7), apparition of a visible fecolith (p=0.0003, OR =3.9), a diameter over 7mm (p=0.052, OR =1.9) and a retrocaecal position (OR=2, p=0.0579) are statistically associated with relapses.

Conclusion
Risk factors of relapse have been highlighted at the 7 day control ultrasound scan such as a diameter over 7 mm, a fecolith apparition, an internal diameter of 2.5mm.. Further studies will help us to determine which subpopulation will benefit the most of NOM in UA.
**PW12-G08**

**3D PRINTING OF COMPLEX TUMORS ENHANCES COMPREHENSION OF SURGICAL ANATOMY**

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Aim of the Study

3D printing involves the creation of a physical model from a 3D image. In recent years there have been an increasing number of reports on the use of 3D models in medicine. Learning complex surgical oncological anatomy is challenging. We present our experience with 3D printing of complex tumors and its anatomical relations regarding teaching aspects.

Methods

Pediatric Surgery Trainees, last year medical school students and Pediatric Clinical Oncologists participated. A questionnaire including surgical anatomy aspects was designed by 3 surgeons with more than 5y of pediatric surgical oncology experience. In addition, participants rated their satisfaction levels on a 5-point Likert scale. Medical students received a class on the subject prior to the test. Medical students were divided in 2 groups and a case was presented. Group_A answer with the images. Group_B had the support of 3D printed models. Trainees and clinical specialists responded before and after consulting the 3D models. Scores and satisfaction levels were compared.

Main results

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Conclusions

3D printed models that can be manipulated in a 3D space can significantly benefit visuo-spatial understanding of complex surgical anatomy enhancing learning.
PW12-G09
COMPLICATIONS AFTER EMBOLIZATION OF PERIPHERAL ARTERIOVENOUS MALFORMATIONS

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Radboud University Medical Center, Nijmegen, Netherlands

**Aim of the study:** We aimed to evaluate the occurrence of complications after embolization of peripheral arteriovenous malformations (AVM) in an expertise center.

**Methods:** All patients with a peripheral AVM, treated with embolization between January 2010 and July 2016, were retrospectively reviewed. Patient characteristics included age, AVM location, AVM angioarchitecture (Yakes classification), and number of procedures. Complications were graded: minor complications were transient and without surgical reintervention, major complications were permanent or required surgical reintervention. Complications were compared between patients and procedures. Embolization materials and expected side effects were also reported.

**Main results:** During the study period 442 interventions were performed in 93 patients (median 2, range 1-82). In most cases, ethanol was used (n=428; 96.8%). The cohort included 21 children (age <18), in whom 38.5% (n=170) of the interventions were performed. A total of 53 complications were identified (12%) in 36 patients. Six complications were graded as major (1.4%) and 47 were minor complications (10.6%). Children seem to develop less complications per procedure compared to adults but this was not significant (Figure). Other factors, such as AVM location and type of angioarchitecture, did not significantly affect complication risk (Table).

**Conclusions:** This study shows few severe complications after use of ethanol in the treatment of AVM. Nevertheless, as a complete treatment course usually consists of multiple embolization procedures, overall complication risk in an individual patient may be substantial and must always be weighed against the treatment indication. Therefore, treatment should take place in centers with experience and a multidisciplinary approach.
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THE DEVELOPMENT OF A 3D PEDIATRIC MODEL TO TRAIN COMPONENT TASKS OF ECMO CANNULATION

Sanne Botden, Erik Leijte, Ivo de Blaauw
Radboumc - Amalia children’s hospital, Nijmegen, Netherlands

Aim of the Study
Extra-Corporeal Membrane Oxygenation (ECMO) therapy is used as rescue therapy for patients with (cardio)respiratory failure. In neonates or infants the carotid artery and/or jugular vein are used for ECMO cannulation. The procedure of locating the cannulas in the proper position and connecting them to the system is high risk, with dramatic consequences if not performed correctly. It would be beneficial to train the subsequent cannulation steps outside the clinical setting, to ensure more routine in this emergency high procedure.

Methods
To train the component steps of the cannulation, a 3-Dimensional model was developed, resembling the carotid artery and jugular vein, which could be cannulated to an ECMO system. Other important features of the model had to be low cost, reusable and the opportunity to simulate blood flow.

Main results
Several models were created using plaster. The model resembling the clinical setting most, was further developed to a 3-D drawing and printed. Figure 1 shows the 3D printed model, the vessels are simulated by small water balloons, fixated with small surgical clamps. Figure 2 shows the model when used for training. The following component tasks can be performed: 1: Securing the vessels. 2: opening of the vessels. 3: Inserting the cannulas (with proper haptics). 4: connecting the cannulas to ECMO system. 5: fixation of cannulas.

Conclusion
We have developed a easy to use 3D model for the training of ECMO cannulation, because training of this high risk procedure could reduce complications.
IMPLEMENTATION AND EVALUATION OF A MANDATORY NATIONAL PEDAGOGIC PROGRAM USING SIMULATION TRAINING IN FRANCE FOR RESIDENTS IN PEDIATRIC SURGERY

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Pediatric surgery resident training in France relies on theoretical teaching and bedside training. Due to the characteristics of the specialization itself (three-party relationship between parents-child-physician, wide variability and complexity of procedures, “specialization in rare conditions”), a resident must develop technical skills and capacities in communication and multidisciplinary work.

To implement the resident training, the national board of pediatric surgery has developed a national program in simulation for all residents. The process took several years from inception, to implementation, and addresses cognitive skills, technical skills, communication, collaboration, leadership, advocacy, and professionalism.

Methods:
The timeline and crucial elements involved in the creation of the inaugural simulation-based pediatric surgical training program in France and follow up after 2 years of existence.

Results:
Yearly, 40 residents (>90% of all the French residents in pediatric surgery) attend a mandatory, two day simulation-based course. Scenario-based simulation is used to cover cognitive skills, as well as issues surrounding communication, collaboration, leadership, advocacy, professionalism and team building. Traditional simulators are used for upskilling of psychomotor skills.

Evaluation focus the pedagogic value (rated 4.8/5), the benefit of simulation concerning communications and team training, the efficiency of a 2 day training, the necessity to increase the number of sessions.

Conclusion:
Important lessons have been learned in the creation and implementation of France’s, nation-wide, mandatory simulation-based program. The introduction a national simulation-based mandatory training program for residents help ensure a measurable level of competence, and allow targeting the highest possible level of expertise among all trainees.
PW12-G12

LAPAROSCOPY IN A MODEL OF PREMATURE NEONATES WITH NEC AND PERSISTENT DUCTUS ARTERIOSUS

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Aim of the study: Laparoscopy in premature neonates is still controversial. The physiological impact of pneumoperitoneum in premature babies with NEC has not previously been systematically evaluated. We studied the respiratory and cardiovasculary changes during pneumoperitoneum in an animal model of prematurity, necrotizing enterocolitis (NEC) and with presence or absence of persistent ductus arteriosus (PDA).

Methods: 33 preterm piglets were delivered by caesarean section, fed with parenteral nutrition and increasing amounts of infant formula. Presence or absence of PDA was diagnosed using echocardiography before surgery. Laparoscopy was obtained by pneumoperitoneum with inflation of CO2. We increased the intra-abdominal pressure (IAP) from baseline to 6 mmHg and hold it steady during laparoscopy. Study approved by animal ethics committee.

Main results: The incidence of PDA was 43 %. We found a significant association between severe NEC-lesions and PDA (p < 0.05). We found significant changes in heart rate, airway pressure and lactate in the PDA group when we elevated the IAP from baseline to 6 mmHg (p < 0.05). In the group without PDA we only found a significant change in heart rate (p < 0.05).

Conclusions: This animal model of premature neonates with NEC and or PDA indicates that laparoscopy is safe when an IAP of max six mmHg is used. The study also indicates, it might be recommended to perform an echocardiography before laparoscopy. We found a significant association between severe NEC-lesions and PDA.

<table>
<thead>
<tr>
<th>Changes</th>
<th>PDA (n = 14)</th>
<th>Without PDA (n = 15)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hb (g/dl)</td>
<td>16.3 ± 0.70</td>
<td>14.2 ± 0.35 ***</td>
</tr>
<tr>
<td>MAP (mm Hg)</td>
<td>62.43 ± 4.68</td>
<td>62.30 ± 2.40</td>
</tr>
<tr>
<td>SAP (mm Hg)</td>
<td>34.95 ± 3.38</td>
<td>31.34 ± 2.01</td>
</tr>
<tr>
<td>Res (KPa)</td>
<td>95.7 ± 13.57</td>
<td>94.2 ± 1.34</td>
</tr>
<tr>
<td>AP (mm Hg/L)</td>
<td>13.34 ± 0.32</td>
<td>14.91 ± 1.06 *</td>
</tr>
<tr>
<td>TV (ml)</td>
<td>37.5 ± 0.98</td>
<td>18.29 ± 0.94</td>
</tr>
<tr>
<td>CO2 (mm Hg)</td>
<td>80.47 ± 9.40</td>
<td>99.31 ± 6.64</td>
</tr>
<tr>
<td>O2 (mm Hg)</td>
<td>314 ± 36.5</td>
<td>306.3 ± 36.4</td>
</tr>
<tr>
<td>pH</td>
<td>7.32 ± 0.05</td>
<td>7.32 ± 0.04</td>
</tr>
<tr>
<td>Lactate (mmol/L)</td>
<td>2.33 ± 0.09</td>
<td>1.92 ± 0.48 *</td>
</tr>
</tbody>
</table>

Mean ± SE

* p < 0.05, ** p < 0.005, *** p < 0.001

Notes: Map = systolic arterial pressure; SAP = diastolic arterial pressure; AP = airway pressure; TV = tidal volume
Aim of the Study: We use intra-cavitary electrocardiography technique (IC-ECG) to avoid need for intraoperative radiological control in the assessment of the proper location of the central venous catheters (CVC): the correct CVC tip position at the caval-atrial junction (CAJ) is indicated by negative deflection of the P wave; in fact the catheter itself acts as an exploring electrode; when the tip approaches the sino-atrial node a small negative deflection of the P wave appears.

Methods: We retrospectively reviewed all CVC positioned in the last 3 years with IC-ECG (Group-1) or classical fluoroscopic control (Group-2). All patients were postoperatively examined by chest X-ray, according to hospital protocol.

Main Results: 389 CVC were positioned: Group-1 (323, 83%) IC-ECG, Group-2 (66, 17%) fluoroscopy, main age 7,42 years, 80,46% in the left internal jugular vein, 16,96% in the right (2,57% required two accesses). In Group-1 average duration for the procedure was 20 min, overall match between IC-ECG and x-ray was 98,67% while in 1,23% (4 patients) position was wrong (all due to caval thrombosis). In group-2 average duration was higher (45 min) and repositioning was necessary in 7 patients (10,6%). No differences for age or complications in both groups.

Conclusions: CVC placement with IC-ECG technique in children is a reliable method, with short procedure time and very low rate of complications. As shown in our series only in 1% of cases the position was wrong for caval thrombosis. This method could prevent children from unnecessary X-ray exposure.
AIM OF STUDY: The recently 2018 updated ISSVA classification of vascular anomalies doesn’t include Primary Intestinal Lymphangiectasia (PIL) as a recognized entity among lymphatic anomalies.

We evaluated our cohort of patients with such a diagnosis to eventually demonstrate that PIL results from a disruption of lymphatic circulation, thus corresponding to a secondary rather than a primary event in the context of generalized lymphatic anomaly (GLA).

METHODS: An analysis of intestinal lymphatic involvement was performed on patients diagnosed with PIL between 1965 and 2016. Of the 21 included, 10 had been diagnosed before 5 years of age (1 prenatal), 8 between 5 and 18 years and 3 at older than 18 years. Endoscopy, histological study, magnetic resonance imaging and lymphoscintigraphy were performed on all patients. Dynamic intranodal lymphangiography was performed on 8 patients.

RESULTS: Twenty-one patients diagnosed of PIL were evaluated. All patients had diagnosis of GLA afterwards, with 12 patients presenting a central lymphatic channel obstruction. Associated lymphatic malformation was present in 16, diarrhea in 10, chylothorax in 11, chylous ascites in 10, pericardial effusion in 6, coagulopathy in 3 and osteolysis in 7.

CONCLUSION: We consider intestinal lymphangiectasia not an entity in itself, but a consequence of lymphatic flow impairment in the thoracic duct, producing chylous reflux into the intestinal lymphatics. Therefore, we propose the term “primary intestinal lymphangiectasia” to be replaced by the more accurate “protein-losing enteropathy” in the context of a channel-type lymphatic malformation or generalized lymphatic anomaly.
COMPARISON OF EFFECTIVENES OF INGROWN TOENAIL SIMPLE WEDGE RESECTION VS. ABLATION WITH CARBON DIOXIDE LASER – A CLINICAL RANDOMISED STUDY

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Aim of the Study

Ingrown toenail is a common and painful nail disease. The most commonly used method of treatment is the wedge resection of the nail or an offending piece of nail. The aim of the study was to compare this technique with a new method of wedge excision utilizing laser ablation with carbon dioxide laser.

Methods

This was a prospective randomized single-blind study. One hundred and twelve patients were included into the study. Control visits were performed systematically. We analyzed the time needed for complete healing of the wound, number of recurrences, pain levels and complication rates. Also, the need for analgesics was assessed and the time of complete pain relief.

Main results

Eighty four percent of wounds after CO2 laser healed completely before week three visit, whereas 75% in the wedge excision group (p <0.05). Three recurrences were observed in CO2 laser group and 11 in wedge excision group (6% and 24% respectively, P < 0.05). Eighty eight % of patients in CO2 laser group did not need analgesics more than 24 hours after the procedure and in all cases no pain was noted 48 hours after the procedure. In the wedge excision group 80% of patients did not need analgesics more than 24 hours after procedure.

Conclusions

Carbon dioxide laser ablation should be acknowledged as a superior method of ingrown toenail treatment compared with surgical wedge excision as the wound heals more rapidly and less recurrences are observed.
PW13-G04
WHICH ARE THE OPTIMAL CUT-OFF VALUES FOR AIR-SCORE IN PEDIATRIC APPENDICITIS?

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Aim of the Study: Since the Appendicitis Inflammatory (AIR)-score seem to outperform other clinical prediction scores for pediatric appendicitis, the aim of this study was to evaluate the optimal cut-offs for AIR-score in a pediatric population.

Methods: The study was approved by the regional ethical committee. Data was prospectively collected at the pediatric ED on all children with suspected appendicitis during 22 months. The diagnosis was confirmed by histopathological examination. Sensitivity, specificity and positive/negative predictive values (PPV/NPV) were calculated for each cut-off. Primary outcome was missed appendicitis and negative appendectomy.

Main results: A total of 263 children (55% boys, median age 9 years) were included of which 120 (46%) had appendicitis. Regarding the low cut-off value to rule out appendicitis, no difference could be seen in sensitivity, NPV or rate of missed appendicitis between ≤3 or ≤4 points, while ≤5 points had a significantly higher rate of missed appendicitis (15% vs 7% and 5%, p = 0.02). Regarding the high cut-off value, no differences could be seen in specificity, PPV or rate of negative appendectomy between >8, >9 and >10 points.

Conclusions: The high cut-off value of the AIR score can be lowered to >8 points when used in children, resulting in more patients taken straight to surgery without affecting the negative appendectomy rate significantly. The low cut-off value should be kept at ≤4 points since a higher value results in significantly more patients with missed appendicitis.

<table>
<thead>
<tr>
<th>Low risk of appendicitis</th>
<th>Low AIR-score</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0-3 (n=85)</td>
<td>0-4* (n=122)</td>
</tr>
<tr>
<td>NPV</td>
<td>95 [88–98]</td>
<td>93 [87–96]</td>
</tr>
<tr>
<td>Missed appendicitis</td>
<td>4 [5%]</td>
<td>9 [7%]</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>High risk of appendicitis</th>
<th>High AIR-score</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>8-12 (n=53)</td>
<td>9-12* (n=37)</td>
</tr>
<tr>
<td>Specificity</td>
<td>99 [95–100]</td>
<td>99 [96–100]</td>
</tr>
<tr>
<td>PPV</td>
<td>96 [86–99]</td>
<td>97 [83–100]</td>
</tr>
<tr>
<td>Negative appendectomy</td>
<td>2 [4%]</td>
<td>1 [3%]</td>
</tr>
</tbody>
</table>

Values presented as % [95% CI], and as the number and absolute percentage of patients; n (%). PPV/NPV: Positive/negative predictive value; * Original cut-off values; ** significant when comparing 0-3 and 0-4 to 0-5
Aims of the study: A closed-member group was set up to improve communication between team members, discuss new neonatal surgical admissions and share regular updates. It rapidly took on a teaching dimension involving case-based discussions and image review. The aim of this study is to evaluate the impact of this teaching method on training.

Methods: An online questionnaire was distributed to all current and past surgical trainees who took part in the neonatal surgery rotation from April 2016 to December 2017. Questions covered their level of training, rotation duration, and perceived effect of the chat discussion on their training and their intended learning outcomes (ILOs). Answers were on a sliding scale. Free text sections described strengths and drawbacks of this communication method. We asked what they did with the chat history once they left the group.

Main results: Twelve trainees (100%) covered a period of 20 months. The majority were junior trainees, 60% spent 3 months and 40% spent 6 months on the unit. The majority felt that the chat adequately supplemented their training, and strongly agreed that the platform was useful. The majority strongly agreed that they were confident with most of the ILOs. A closed-group membership was favoured by 9 (75%). Strengths included interactivity and focused learning while the main drawback was the delayed response time. Five (41.7%) saved the chat history, and 4 (33.3%) sometimes returned to it for reference.

Conclusions: Pediatric surgery trainees felt that an instant messaging group strongly supplemented their learning.
LONG-TERM HEALTH RELATED QUALITY OF LIFE IN A COHORT OF PATIENTS TREATED WITH SCLEROTHERAPY FOR LYMPHATIC MALFORMATION

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\textsuperscript{1}Karolinska University Hospital, Stockholm, Sweden. \textsuperscript{2}Karolinska Institutet, Stockholm, Sweden

Aim of the study

The aim of this study was to assess the Health-related quality of life (HRQOL) in a long-time follow-up cohort of children and adolescents with LM.

Methods

All patients who underwent sclerotherapy with OK-432 for LM at our institution, between 1998 and 2013 were reviewed. A study specific questionnaire was sent to all patients with at least five years’ follow up after the first injection treatment asking for persisting symptoms and satisfaction with the treatment and care. KIDSCREEN-52 was used for assessing HRQOL.

Results

One hundred and thirty-eight patients were treated with OK-432 for LM from 1998-2013. There were statistically relevant negative correlations between the total number of injection treatments and HRQOL dimensions Autonomy (p=0.013), Parent Relation and Home Life (p=0.014), Financial Resources (p=0.025), and School Environment (p=0.037).

Localization in the head neck area was a negative predictor throughout all studied dimensions with the strongest association with Psychological Well-being (p=0.009), Parent Relation and Home Life (p=0.017) and School Environment (p=0.006).

Patients with persistent visible signs were less satisfied with treatment (p = <0.001). However, the degree of experienced difficulty of remaining problems including signs of illness was negatively correlated with overall satisfaction with treatment (r=-0.384, p = 0.011).

Conclusion

LM patients with malformations localized in the head and neck area requiring multiple treatments constitute a risk group for affected HRQOL. Multiple treatments per se imply a risk for impaired HRQOL.
THE COMBINATION OF SERUM WHITE BLOOD CELL COUNT, NEUTROPHIL PERCENTAGE AND C-REACTION PROTEIN IN DIAGNOSIS OF PEDIATRIC APPENDICITIS

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Aim: We aim to evaluate the diagnostic value of three inflammatory markers, white blood cell count (WBC), neutrophil percentage (NEU), C-reactive protein (CRP) and combined triple-marker in pediatric appendicitis.

Methods: Clinical data of 1392 inpatients admitted for suspicion of appendicitis were collected from 2012-2017. Inflammatory markers were tested upon admission with the cut-off points of WBC(10000 per ul), NEU(75%) and CRP(5 mg/L). Appendicitis was confirmed histologically post-appendectomy. The diagnostic value of inflammatory markers was trended in relation to duration of abdominal pain on admission.

Results: In 718 children with appendicitis, the percentage of results above cutoff value (sensitivity) of WBC and NEU were 94.6% and 80.5% at day 1 of pain but decreased to 64.9% and 51.1% at day 5. Conversely, CRP sensitivity was only 60.9% at day 1 but increased to 97.9% at day 4 of pain. When any marker exceeded their cut-off value (any triple-marker positive), sensitivity was 99.7% throughout the course of appendicitis with a specificity of 23.6%. When all 3 markers were below cut-off (all triple-marker negative), negative predictive value (PPV) was 98.8%. In our cohort, overall negative appendectomy rate was 7.2% (52 cases). However, among 19 patients with all negative triple-marker who underwent appendectomy, 17(89.5%) were histologically normal.

Conclusions: Individual inflammatory markers are unreliable in different stages of appendicitis. Combined triple-marker demonstrates excellent sensitivity throughout the progress of appendicitis with high negative predictive value. All triple-marker negative can confidently rule out appendicitis, therefore avoiding costly imaging studies and negative appendectomy.
Aim: The aim of this study is to present our technique and results of single trocar laparoscopy-assisted surgery (STLAS) for BOC in children.

Methods: Medical records of patients with diagnosis BOC undergoing STLAS at our centers from 2009 to 2017 were reviewed. For the STLAS, an 11 mm umbilical trocar was placed and a 10 mm camera with engrafted 5 mm working channel was used. The ovarian cystic wall was grasped and after fluid aspiration, the cyst was brought out of abdomen via the umbilical or small suprapubic incision and it was excised extracorporally, sparing ovarian tissue when possible.

Results: 45 patients were identified, with median age of 3 years. The median size of the cysts was 6.0 cm. In 30 cases the cyst was mature teratoma, in 11 – simple cyst and in 4 – dermoid cyst. In 39 patients (86.7%) the cyst was excised via the umbilical incision and in 6 - via the suprapubic incision. Ovary sparing was achieved in 42 cases (93.3 %). The mean operative time was 32.5 minutes. There was no intra- or postoperative complication. The mean postoperative hospital stay was 1.8±0.6 days. At a median follow up of 36 months, recurrence was detected in 2 patients and they underwent 3-trocar laparoscopic total oophorectomy. All other patients were in good health with excellent postoperative cosmesis.

Conclusions: STLAS is simple, safe, with excellent cosmesis. This technique also facilitates ovary sparing and can be a viable approach in minimally invasive management of BOC in children.
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¹Ankara Yıldırım Beyazıt University Medical Faculty Pediatric Surgery, Ankara, Turkey. ²Health Sciences University Ankara Child Health And Diseases Hematology Oncology Training And Research Hospital, Department Of Pediatric Surgery, Ankara, Turkey. ³Health Sciences University Ankara Child Health And Diseases Hematology Oncology Training And Research Hospital Department Of Interventional Radiology, Ankara, Turkey

Aims of the Study:

Hydatid cyst is endemic in our country and contaminated by the eggs of the parasites named echinococcus granulosus (EG). PAIR, laparoscopic and open surgery are preferable treatment methods. Aim of this study is to discuss the data and results of hydatid cyst cases who are treated in our clinic.

Method:

163 hydatid cyst cases who are treated in our clinic, have been analysed between the 1997-2016 years retrospectively.

Main Results:

90 girl and 73 boy cases were evaluated. The mean age of cases was 10.28 (5-17) years. Albendazole treatment were given for 3 weeks before operation, except in emergencies. Hydatid cyst was seen in the liver in 105 cases, liver and lung in 30 cases, the lung in 18 cases, liver and spleen in four cases, liver and the omentum in two cases, liver and kidney in one case, liver and adrenal gland in one case, spleen in one case and the rib in one case. There was single cyst in 115 cases and multiple cysts were in 48 cases. PAIR treatment were used in 41 liver hydatid cyst cases by interventional radiologist. The other cases were treated with open surgery. Infection, perioperative bile duct opening and recurrence were seen only cases with surgical treatment.

Conclusion:

The hydatid cyst should be in the preliminary diagnosis in cases of suspicious radiological and clinical findings in endemic areas. Percutaneous treatment should be considered in appropriate elected hepatic hydatid cases due to minimally invasive, low morbidity and mortality rates.
Aim of the study: To show our experience in the treatment of children with laryngeal stenosis.

Methods: Retrospective and descriptive study of patients treated in our unit with the diagnosis of laryngeal stenosis between 2000 and 2017. The following variables were studied: demographic data, type and location of the lesion, associated anomalies, initial tracheotomy, type of treatment and outcomes.

Results: 78 children were included in the study (39 boys) with a median age at diagnosis of 9 months; 33 (42.3%) showed an associated anomaly. 76.9% of the lesions were located in the subglottis, 12.8% were glotto-subglottic stenosis and isolated glottic involvement was detected in 8 cases (10.2%). 84.6% were acquired stenosis and the remaining were congenital. 30 patients (38.5%) had a severe stenosis (Myer-Cotton grades III and IV) and a tracheotomy was performed as an initial treatment in 37 patients (47.4%). The decannulation rate was 70%; 4 patients are under survey and another 6 with incomplete follow-up. 41 patients (52.6%) did not have a tracheotomy as an initial treatment and 33 (80.5%) of them showed clinical improvement. Overall, 26 patients were treated surgically, 19 with endoscopic procedures, and 14 required both types of treatment. There were 3 deaths in the series but none of them were related to their airway disease.

Conclusion: Laryngeal stenosis in children are usually located in the subglottic region and most of them are acquired. A tracheotomy is usually required as an initial treatment in severe lesions and the management may include surgical and endoscopic techniques.
PW13-G11
ANALYSIS OF 3D PRINTING MATERIALS AND ITS CORRELATION TO HUMAN ANATOMY

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Aim of the Study

3D printing involves the creation of a physical model from a 3D computer version. Different 3D printers and materials are available on the market. The hardness of materials is expressed on a Shore scale. The objective is to analyze the physical characteristics of available 3D printing materials and compare them to human structures.

Methods

Shore scale for 3D printing materials is given by the provider. Shore scale for human structures was calculated using Bulk modulus (K), Shear modulus (G) and Young modulus (Y) obtained from published ultrasound elastography.

Main results

<table>
<thead>
<tr>
<th>Tissue</th>
<th>Hardness (Shore)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lung</td>
<td>40 Shore_OOO 10 Shore_OO</td>
</tr>
<tr>
<td>Liver</td>
<td>52 Shore_OOO 25 Shore_OO</td>
</tr>
<tr>
<td>Fat</td>
<td>54 Shore_OOO 35 Shore_OO 3 Shore_A</td>
</tr>
<tr>
<td>Kidney</td>
<td>56 Shore_OOO 40 Shore_OO 4 Shore_A</td>
</tr>
<tr>
<td>Pancreas</td>
<td>58 Shore_OOO 45 Shore_OO 5 Shore_A</td>
</tr>
<tr>
<td>Mammary gland</td>
<td>56_Shore OO 10 Shore_A</td>
</tr>
<tr>
<td>Parotid Gland</td>
<td>60 Shore_OO 13 Shore_A</td>
</tr>
<tr>
<td>Brain</td>
<td>70_Shore OO</td>
</tr>
</tbody>
</table>
Conclusions
Bony structures have a similar hardness to 3D printing materials. Few soft resins hardness values are similar to some human structures like muscle, brain or breast tumors. 3D printing materials are still limited. Research should focus on new materials that mimic human anatomy characteristics.
PW13-G12: EVALUATION OF PATIENTS WITH PERIPHERAL ARTERIOVENOUS MALFORMATIONS IN AN EXPERTISE CENTER

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Aim of the study: Arteriovenous malformations (AVM) are the most complex lesions in the group of congenital vascular malformations. This study evaluates the population of patients treated for a peripheral AVM in one of the largest expertise centers in Europe.

Methods: All patients with a peripheral AVM, treated using embolization between January 2010 and July 2016, were retrospectively reviewed. Patient characteristics included age, AVM location, AVM angioarchitecture (Yakes classification) and number of procedures. In addition, we evaluated minor and major procedure-related complications.

Main results: A total of 442 interventions were performed in 93 patients. The cohort included 21 children (age <18), in whom 38% (n=170) of the interventions were performed. The median age was 31 (range 2-66) at the first procedure during the study period. Figure 1 and 2 show the distribution of AVM locations and angioarchitecture types respectively. Most patients had an AVM type 2 (n=57; 61%) and almost 50% of all procedures (n=219) were performed in these patients. Overall, the median number of procedures per patient was two (range 1-82). The average complication risk per procedure was 12%; 10.6% minor and 1.4% major.

Conclusions: In this expertise center a diverse group of patients has been treated, including a substantial number of patients with complex type 2 and type 4 arteriovenous malformations. Most AVMs were located in the head and neck region and the extremities. The required number of procedures per patient was for a large group very low but varied widely, with a low overall complication rate.

Figure 1:
Figure 2:

Angioarchitecture types of the arteriovenous malformations

- Percentage of patients (total n=93)
- Percentage of procedures (total n=442)
PW14-TH01
PNEUMONIA AFTER REPAIR OF OESOPHAGEAL ATRESIA – INCIDENCE AND MAIN RISK FACTORS

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Aim of the Study: Oesophageal atresia (OA) is associated with significant respiratory mortality. We assessed retrospectively the incidence and predicting factors of pneumonia five years after repair of OA.

Methods: Institutional ethical consent was obtained. Hospital records of patients with OA from 2002 to 2017 were reviewed including episodes of pneumonia that required hospital admittance in university or regional hospitals within five years OA repair. Type of OA and repair, anastomotic complications, fundoplication for GER, associated major cardiac disease and chromosomal anomalies or neurological impairment (CN) were tested as potential risk factors for pneumonia.

Main Results: A total of 107 patients (58 males), (type A 7, B 3, C 86, D 3, E 6, F 2), median age 8.1 (IQR 4.1-12) years, were included. Thirty-nine (36%) patients had 82 episodes of pneumonia corresponding a median of 2(IQR 1-4) episodes /patient. The majority of pneumonias occurred before the age of three years. (Figure1) The cause of pneumonia was RS-virus infection in eighteen (17%) and aspiration in eight (8%) episodes. Four (80%) of five patients with recurred TEF had at least one episode of pneumonia. Independent risk factors for pneumonia were CN, fundoplication and reoperations for anastomotic complications Type of OA, type of repair and cardiac disease did not predict pneumonia. (Table1) Five (5%) patients died of cardiac disease (n=4) and asphyxiation (n=1).

Conclusions: Episodes of pneumonia occurred in one third of patients with OA. Patients with chromosomal anomalies or neurological impairment, fundoplication and with major anastomotic complications were at greatest risk.
Figure 1 Occurrence of pneumonia episodes (n=82) by age

Table 1 Risk factors for pneumonia in 107 patients with oesophageal atresia

<table>
<thead>
<tr>
<th>Risk factor</th>
<th>RR (95%CI)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Long gap (type A or B ,n=10) vs others (n=97)</td>
<td>1.9 (0.5 − 7.2)</td>
<td>0.32</td>
</tr>
<tr>
<td>End to end (n=95) vs reconstruction (n=11)</td>
<td>2.4 (6.8 − 8.5)</td>
<td>0.17</td>
</tr>
<tr>
<td>Major cardiac disease (n= 22)</td>
<td>1.7 (0.7 − 4.4)</td>
<td>0.27</td>
</tr>
<tr>
<td>Chromosomal anomaly or neurologic impairment (n=6)</td>
<td>10.0 (1.2 − 91)</td>
<td>p=0.04</td>
</tr>
<tr>
<td>Fundoplication for GER (n=25)</td>
<td>2.8 (1.1 − 6.8)</td>
<td>p=0.03</td>
</tr>
<tr>
<td>Reoperations for anastomotic complications (n =14)</td>
<td>5.8 (1.7 − 20)</td>
<td>p= 0.006</td>
</tr>
</tbody>
</table>
FILAMIN A MUTATION ASSOCIATED WITH LUNG DISEASE: PITFALL IN PEDIATRIC SURGERY

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Mutations in the X-linked gene encoding filamin A (FLNA) have been reported to cause a wide range of human diseases, such as cerebral periventricular nodular heterotopia (PVNH), cardiac valvular disease and skeletal anomalies. Recently lung involvement has been also defined. We reported a male infant was referred to our surgical department, with presumptive diagnosis of congenital lobar emphysema. He developed a significant lung disease resulting in lobar emphysema of the left upper lobe and a subsegmental atelectasis and areas of air trapping into the lower lobe. Following lobar resection, chronic respiratory failure occurred leading the mechanical ventilation to assist the patient’s breathing. The histological features showed a combination of alveolar simplification, emphysematous changes and perivascular and interstitial fibrosis. Genetic testing showed a new pathogenic variant of the FLNA gene c.7391_7403del; p.Val2464AlafsTer5. Brain magnetic resonance imaging depicted PVNH. Mesenchymal stromal cells (MSCs) were successfully isolated and propagated from lung tissue and they presented an high proliferative and fibrotic capacity. This report confirms an association between FLNA gene mutation and lung disease. Because of the role of the action of FLNA in mesenchymal migration, the alteration in mesenchimal properties could be directly related to the defects in cell migration during the embryonic development and in pulmonary damage described in FLNA-defective patients. The lung disorder leded to respiratory failure during infancy and these pulmonary complications may be the presenting feature of this disorder. An early recognition is important to guide potential therapies, genetic testing and brain imaging.
PW14-TH03
PERIDIAPHRAGMATIC LOCATION: A CLUE TO THE ORIGIN OF RHABDOMYOMATOUS DYSPLASIA IN CONGENITAL PULMONARY ADENOMATOUS MALFORMATIONS (CPAM)

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Aim of the study: Rhabdomyomatous dysplasia (RD) is a pathologic finding in CPAMs that was (wrongly) attributed to their malignant potential. The increasing recognition of extrathoracic (intradiaphragmatic and intraabdominal) CPAMs offers a clue to the origin of RD. We hypothesize that the presence of RD is related to the CPAM’s anatomic location.

Methods: Retrospective review was performed of all children who underwent resection of a CPAM during a 10-year period. The age at the time of operation, location of the CPAM, and pathologic findings were collected. Peridiaphragmatic location was defined as within the inferior pulmonary ligament, deep to the diaphragmatic portion of the parietal pleura (“intradiaphragmatic”) or adjacent to the abdominal side of the diaphragm. Statistical analysis was performed using Fisher Exact test for 2x2 tables.

Main results: Twenty-six patients with CPAM were identified. Preoperative imaging was performed by CT scan (16/26), ultrasound (5/26), MRI (1/26) and chest radiograph (4/26). The mean age at resection was 15 months. Of these, 16 were pure cystic adenomatoid malformations, 4 were extralobar sequestrations, 4 were intralobar sequestrations and 2 were bronchogenic cysts. Nine lesions were peridiaphragmatic with 4 being intradiaphragmatic (44%). Eight of the 9 resected peridiaphragmatic lesions contained histologic evidence of rhabdomyomatous changes (89%, CI 52-99%). None of the other lesions contained RD (CI 0-19%, P<0.001).

Conclusions: Rhabdomyomatous dysplasia was seen exclusively (and in virtually all) peridiaphragmatic CPAMs. While the exact significance of RD remains unclear, it may represent incorporation of striated muscle tissue associated with the developing diaphragm.
Aim of the study: Determine the incidence of congenital diaphragmatic posterior-lateral hernia with late revelation and specify its' epidemiological, semiological and therapeutic characteristics.

Materials and methods: Descriptive retrospective study of children with a diaphragmatic posterior-lateral hernia after 30 days of life between 1994 and 2017 in three French centers.

Results: The incidence was 8.5% of all Bochdalek congenital diaphragmatic hernias (46 out of 538 cases). Forty-six patients were included aged from 34 days to 11 years old. The sex-ratio was 1/1.1. The average age of discovery was 9 months (6-22). The hernia was located on the left side in 96% of cases and in right side in 4% of cases. In 65% of cases, it was revealed in emergency. The revealing symptoms were respiratory (24%), digestive (26%) or mixed (36%). This was an asymptomatic accidental discovery in 13% of the cases. The initial diagnosis was wrong in 36% of the cases, mostly confused with a pneumonia or pleuropneumonia. Thirty-seven percent of patients had an associated malformation. The hernia was operated by thoracoscopy in 51% of cases and required a patch in 3 patients (7%). Pulmonary hypoplasia or pulmonary arterial hypertension were not found.

Conclusion: Congenital diaphragmatic hernia with late revelation is a poorly known pathology, but requiring fast intervention to avoid complications. The modes of discovery are clinically misleading, with the expression being sometimes pulmonary, sometimes abdominal, or mixed. Unlike neonatal hernias, prognosis and evolution seem favorable.
The aim:
Surgery is the mainstay of the treatment of pulmonary hydatid disease.
The aim of this study is to discuss Thoracoscopic approach of pulmonary hydatid disease

Patients and Methods:
Retrospective study: 60 cases of pulmonary hydatid cyst treated using the thoracoscopic approach among 252 subject to surgical treatment. 12 years [January 2004 - February 2016]. Pediatric Surgery Department

Main Results:
The mean age is 9 years [4 years-15 years]. The Sex-ratio is 0.8. Symptoms were dominated by coughing. The X-ray chest is done in all the cases with signs of rupture in 18%. CT has completed the assessment in 10 cases where the diagnosis were uncertain. Conversion to thoracotomy in 6 cases (uncontrollable air leakage). Mean duration of the intervention: 80 minutes. The mean duration of the drainage is 24h (cysts less than 5cm) and 4 days if more than 5 cm. The mean hospital stay is 3 days (2 – 8 days). In the post-operative course 7 patients presented lung infection. 2 required a prolonged drainage. In all the other cases, the follow-up was uneventful. At mean follow-up of 38 months, all patients were asymptomatic without recurrence.

Conclusion:
All stages of the surgical treatment of hydatid cyst of the lung can be easily accomplished by thoracoscopy, with less morbidity and early recovery in cysts less than 5 cm.
UNDETTREATING PEPTUS? COMPARISON BETWEEN HALLER INDEX WITH CORRECTION AND DEPRESSION INDEX

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AIM OF THE STUDY
In the last years new indexes are gaining ground in the evaluation of Pectus Excavatum (PE). The aim of our study is to compare surgical indication established by the classical Haller Index (HI) in relation to the Correction (CI) and Depression Index (DI).

METHODS
Retrospective study (2010 and 2017) of all CT scans and MRI of PE patients evaluated in our paediatric outpatient clinic. Each index (HI, CI, DI) was measure twice, by a facultative and a trainee. Since indication for corrective surgery is established and internationally accepted for a HI >3.25, we evaluated CI and DI in patients with HI inferior and superior to 3.25. Reviewing literature CI >20% and DI >0.2 were considered indicative for surgery.

MAIN RESULTS
81 studies in 77 patients (mean age of 12 years) were analysed. Correlation for measures done by a trainee versus facultative was significant: HI 0.942 (p<0.0001), CI 0.862 (p<0.0001), and DI 0.762 (p<0.0001). 25 had a HI ≤3.25: 21 (84%) had a CI >20%, 24 (96%) a DI >0.2 indicating surgery. Changing the set of CI to 10% (proposed by St.Peters 2011) all 25 should have undergone surgery. 56 had a HI >3.25: 1 (1.78 %) presented a CI < 20%, 3 (5.35%) a DI <0.2.

CONCLUSIONS
Following the criteria published for the new indexes, PE patients may be undertreated in 26% and overtreated in 3.7%. An international consensus is mandatory to establish the most reliable index and its limits for surgery indication.
PW14-TH08
INTRAOPERATIVE TRANSESOPHAGEAL ECHOCARDIOGRAPHY: CARDIOLOGIC CHANGES DURING PECTUS EXCAVATUM REPAIR

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AIM OF THE STUDY
Cardiac compression in children with pectus excavatum is debated. Transthoracic echocardiography (TTE) at rest is usually used, but it can be challenging to perform because of anterior wall deformation. Our aim is to assess right heart changes during pectus excavatum correction by intraoperative transesophageal echocardiography (TEE)

METHODS
A prospective study (2017) of patients who underwent pectus excavatum repair was conducted. Under general anesthesia, cardiologic changes were addressed by three-dimensional TEE before correction, during sternum elevation and after correction. Diastolic diameters of right ventricle (RVEDD), right atrium (RA) and tricuspid annulus (TA) were measured.

MAIN RESULTS
Eight patients (2 females/6 males) with mean age of 14,3 years (range 12,5-16,6) were included. Mean preoperative Haller Index was 5,3 (range 3,4-6,6) and mean Correction Index was 41,9 (range 31,7-59,5). Preoperative TTE at rest showed right heart chamber compression in 2 patients, and was normal in 6.

Before correction, intraoperative TEE showed in all cases compression of the right heart and deformation of the TA. Nuss procedure (with insertion of 2 bars) was performed in 7 cases, and the Pectus-Up technique in 1. During sternum elevation, RVEDD, RA and TA improved: mean augmentation was 6,50mm (range 0-10) for RVEDD, 11mm (range 0-13) for RA and 7 mm (range 3,9-11,6) for TA.

CONCLUSIONS
Preoperative transthoracic echocardiography at rest may underestimate right chamber compression in pediatric patients with pectus excavatum. This preliminary study suggests that immediate structural and functional cardiac improvement during surgical repair can be assessed with intraoperative transesophageal echocardiography.
SLIPPING RIB SYNDROME IN PAEDIATRICS: REPORT OF 4 CASES

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Aim of the study

The Slipping Rib Syndrome (SRS) is an unknown pathology for the paediatric surgeon due to its low incidence in children. The weakness of the costal ligaments allowing an area of rib hypermobility has been postulated recently as the main ethiology. It produces an intermittent pain in the lower thorax or upper abdomen that can affect to the daily activities and can be the origin of unspecific chronic pain.

Methods

A retrospective review of patients diagnosed of SRS between October 2012 and March 2017 was performed. Data of demographics, symptoms, imaging studies, surgical findings and long-term follow-up were collected.

Results

During this period, 4 patients (2M/2F) were diagnosed of SRS. Median age at diagnosis was 13y [12-15] with a mean duration of symptoms of 13m [12-36]. In 2 patients the SRS was associated with Costal Dysmorphia (CD). The initial diagnosis was clinical with posterior ultrasound confirmation. Resection of the affected cartilages was performed in 3 patients and after a follow-up of 6m [3-30], they all are painless and refer a good cosmetic result. One patient refused the intervention.

Conclusions

The SRS is an infrequent cause of thoracic pain with an ethiology not well understood. The awareness of this disease and its typical presentation can avoid unnecessary studies and excessive work up. The resection of the affected cartilages is a safe and effective treatment.
PW14-TH10
POSTNATAL MANAGEMENT PLANNING FOR PRENATALLY DIAGNOSED CONGENITAL CYSTIC LUNG DISEASE

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【Aim of the Study】We evaluated the appropriate management for prenatally diagnosed congenital cystic lung disease.

【Methods】Postnatal clinical course and accuracy of diagnostic imaging were investigated in 59 patients who were prenatally diagnosed with cystic lung lesions including congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), or bronchial atresia (BA) in our hospital between 2001 and 2017.

【Main results】Fetal ultrasound detected the lesions at median 24 weeks of gestational age, and fetal MRI was performed in 45 patients. Pulmonary resection was needed during the neonatal periods because of respiratory failure in 23 patients, eleven of whom underwent fetal treatment. Prenatal and postnatal diagnostic accuracy of imaging confirmed by pathological diagnosis was 94% and 100% in CPAM, 75% and 100% in BPS, and 100% and 100% in BA, respectively. Elective surgery was performed in 25 patients at median age of 14 months. Incidence of pneumonia before elective surgery was 14.2% in CPAM, 37.5% in BPS, and 16.7% in BA, and the median age at an initial episode of infection was 12.5 months. Prenatal, postnatal, and late infant diagnostic accuracy of imaging confirmed by pathological diagnosis was 64.7%, 73.3%, and 84.6% in CPAM, 50%, 75%, and 87.5% in BPS, and 80%, 80%, and 100% in BA. Seven patients with suspected imaging of BA were observed conservatively because of spontaneous reduction.

[Conclusions] Elective surgery should be performed within one year after birth to avoid infection. Spontaneous reduction may be expected in BA on the basis of high accuracy of imaging diagnosis.
PW14-TH11
STRUCTURED AND UNIFORM REPORTING OF CT-IMAGING FINDINGS IN CONGENITAL LUNG ABNORMALITIES

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Aim of the study: While common in pathology reports, clear radiological nomenclature and reporting of congenital lung abnormalities (CLA) is lacking. To develop a uniform and structured way of reporting, a review of the literature for CT-scan anomalies in patients with CLA was done.

Methods: In accordance with PRISMA guidelines, a systematic literature search was conducted. All articles in English published before August 2017 were reviewed and selected by two independent reviewers. Abnormalities included congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), congenital lobar emphysema (CLE) and a bronchogenic cyst (BC). Patient cohorts with a median age <18 years were included. Only reports describing CLA features and case-series exceeding 5 cases were included in this review.

Main results: 1581 papers were found, 158 remained after title-abstract screening. After assessing full-text, 29 papers remained. All included papers were retrospective cohort studies.

In CPAM, CT-scans show a single or multiple, air-containing cysts, sometimes fluid-filled or containing fluid-levels. Depending on the type, a soft-tissue mass can be distinguished. BPS are described as multiple aerated cysts or soft-tissue mass with anomalous arterial blood supply. Perilesional hypodensity, atelectasis and mediastinal shift are often seen. CLE is characterized by lobar hyperinflation and emphysematous changes accompanied by mediastinal shift with atelectasis of neighboring lobes. BC are described as solitary, round/ovoid cystic structures with a well-defined, smooth border and homogenous water/soft-tissue attenuation.

Conclusions: CT findings and nomenclature in CLA are very variable. Because of this variation, better nomenclature and structured reporting in imaging is needed.
PW14-TH12
PLEURAL EFFUSION IN CONGENITAL DIAPHRAGMATIC HERNIA

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AIM: Pleural effusion is a known complication of congenital diaphragmatic hernia (CDH), described in 5 to 30% of the cases. The aim of the present study was to examine the clinical features of CDH that may contribute to an association with postoperative pleural effusion.

METHODS: Records for 52 babies with CDH born between January 2013 and December 2017 in our institution were retrospectively analyzed. Pre-operative and post-operative variables were compared in presence/absence of post-operative pleural effusion.

RESULTS: Among the 52 patients, 20 (38%) developed a pleural effusion, and 13 of them (25%) were confirmed chylothorax. All were managed non-operatively. We observed that the LHR (38% vs 48%; p=0.03), the need of patch repair (50% vs 15%; p=0.006), the duration of mechanic ventilation (16 days vs 10 days; p=0.002) and the length of stay (46 days vs 26 days; p=0.002) were significantly associated with post-operative pleural effusion. The incidence of post-operative infections was also higher in case of pleural effusion (30% vs 7%; p=0.02). The 2 groups were however comparable for survival and there was no difference for secondary pulmonary arterial hypertension (25% vs 15%; p=0.37). The incidence of recurrence was also comparable (15% vs 3%; p=0.1).

CONCLUSIONS: These data confirmed that pleural effusion after CDH repair is related to increased disease severity and to significant morbidity. However, it seems associated to the lung dysfunction but not to the cardiopulmonary morbidity.
PW15-OH01
CENTRAL VENOUS CATHETER POSITIONING IN PEDIATRIC ONCOLOGY: SYSTEMATIC REVIEW AND META-ANALYSIS OF THE GROUP OF YOUNG PEDIATRIC SURGEONS OF EUROPE

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Aim of the study: The aim of this study is to compare the outcomes of open and ultrasound (US) guided approach for placement central venous catheter (CVC) in pediatric oncology.

Methods: Using defined search strategy systematic review and meta-analysis were performed following "Cochrane Handbook for systematic reviews of intervention" and "preferred reporting item for systematic reviews and meta-analysis" (PRISMA, fig1). Inclusion criteria were defined and data collection included factors related to patient, CVC placement technique and complications. Maneuvers were compared by appropriate statistical methods, and meta-analysis was conducted using RevMan 5.3.

Main Results: Systematic review: of 1831 abstracts screened, 128 full-text articles were analyzed. Of these, 8 papers (629 infants) met our inclusion criteria. There were no randomized studies and only one prospective. 4 articles (322 infants) reported CVC placement with ultrasound percutaneous guided approach and 4 articles (307 patients) reported open placement of CVC fitting inclusion criteria. Success and failure definition were not specified in most reports. Meta-analysis: 8 studies compared CVC placement US guided percutaneous approach (444 procedures) versus open approach (346 procedures) to the occurrence of complications (immediate and delayed). Forest plot (fig2) shows a prevalence of immediate complications in the US guided procedures compared, while in open procedures delayed complications are prevalent than immediate (p<0.001).

Conclusions: The meta-analysis shows a potential to reduce delayed complications when ultrasound is used in CVC placement. However, further randomized control trials are needed to identify risk factors to prevent immediate and delayed complications in both approaches.
USE OF INTRAOPERATIVE ULTRASOUND IN TESTICULAR SPARING SURGERY IN ASYMPTOMATIC, NON-PALPABLE, SMALL LESIONS

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Background: With modern easily accessible ultrasonography many asymptomatic, small volume testicular lesions are picked up nowadays. They should be approached with the same oncological principles however organ preservation must be considered since a large number of these masses are benign. The benefits of preserving testicular parenchyma are preserving function, psychological and cosmetic.

Method: Five pre-pubertal boys with small volume testicular lesions underwent sparing surgery in our institute in the last five years. All patients underwent ultrasound of the testicles, serum alpha-feto protein and human chorionic gonadotropin level measurements pre-operatively. The inguinal approach was used for all patients, the cord was gently slung, and the testicle was delivered from the external ring. Intra-operative USS helped locate the lesion in impalpable cases. The tunica vaginalis and albuginea were incised and the lesion enucleated and sent fresh for histological evaluation. No frozen section was utilised. The tunica albuginea and tunica vaginalis were closed separately.

Results: Tumour markers were normal in all cases. Four of the lesions were identified as epidermoid, with one also having a haemangioma. The fifth case was a Leydig tumour. All were less than 10mm in diameter. The Leydig tumour underwent a subsequent radical orchiectomy. There was no recurrence in any of the cases.

Conclusion: Testicular sparing surgery can be the treatment modality in selected cases in all age groups. Children are expected to benefit most from this approach. Intraoperative ultrasound can aid in localization of these small volume lesions.
Aim of the study: Primary dislocation of tunnelled central venous catheters (CVCs) is still major complication among pediatric patients, with previously reported rate up to 50% of all CVC. Role of subcutaneously anchored suturless device (SASD) in prevention of this event is still debated, especially regarding its safety, effectiveness and cost-effectiveness.

Methods: we reviewed use of SASD for tunnelled CVCs in pediatric patients inserted over a period of 10 months. All CVC were inserted according to institutional insertion bundle and managed according institutional policies. Complications during placement and removal, pain/exit site inflammation and rate of dislocation were evaluated.

Main Results: 105 central lines (37 Peripherally inserted central catheters and 68 centrally inserted central catheters/CICCs) were secured with SASD in 93 patients (age range 20 days – 12 years, median age 5.6 years). Median duration of the CVC was 3 weeks (range 5 days - 7 months). No complication occurred during SASD placement. One patient experienced accidental CVC removal for skin erosion of the exit site after 4 weeks. SASD removal – performed by splitting in two halves with scissors - did not require sedation or local anesthesia and was easy and uneventful in all cases.

Conclusions: SASD is effective in preventing dislocation in 99% of patients, with negligible complications both at insertion and during maintenance and removal. Considering that SASD saved approximately $6,00 per week (cost of traditional sutureless devices) and avoided CVC primary dislodgement in this high-risk patient population, it can be also considered cost-effective.
PW15-OH04
ROBOT-ASSISTED LAPAROSCOPIC MANAGEMENT OF RENAL TUMORS IN CHILDREN: PRELIMINARY RESULTS

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Aim
We present preliminary results of robot-assisted laparoscopic (RAL) total and partial nephrectomy for renal malignant tumours.

Methods
For total nephrectomy, a four-trocar transperitoneal approach was used. The tumour was extracted inside a plastic bag without morcellation through a Pfannenstiel incision. Partial nephrectomy was performed using a three-trocar retroperitoneal approach with renal artery clamping. Renorrhaphy was done with sliding-clip technique. Patients with Wilms were treated according to SIOP-2001.

Results
Seven children underwent RAL nephrectomy at a mean age of 5.7 years (3.2–14.1). Total nephrectomy for Wilms tumour was done in 4 cases: one was converted due to renal vein injury, one was converted after the nephrectomy for renal vein tumour thrombus misdiagnosed on preoperative imaging. Staging was I (n=2); II (n=1), III (lymph nodes involvement, n=1). Histology showed regressive type (n=3) and diffuse anaplasia (n=1).

Three patients had partial nephrectomy: increasing size of one nodule in a bilateral nephroblastomatosis after Vincristin-Actinomycin, onset of right tumour after left nephrectomy for Wilms and left tumour ten years after liver transplantation for hepatoblastoma. Warm ischemia time was 40-29-26 minutes. Pathology was Wilms in 2 cases (stage I with standard histology) and tubulopapillary renal cell carcinoma (n=1). Tumour was completely removed in all cases (R0). No ruptures occurred.

Postoperative course was uneventful. Patients were discharged from day 2 to day 7. No recurrence or long-term complications occurred (1–15 months follow-up).

Conclusions
RAL total and partial nephrectomy for renal tumours in children is feasible and safe in highly selected children.
Objectives: In this study, it was aimed to evaluate long term results of the cases who were operated due to ovarian tumor in childhood.

Methods: Hospital records of patients who underwent surgery in our clinic between 1995 and 2015 were reviewed retrospectively. Demographic data, clinical, operative, histopathologic findings and long term results of the cases were evaluated.

Results: 90 operations were performed on 85 patients. The median age of operation was $13.3 \pm 3.8$ years. 84.7% ($n = 72$) of the cases were benign and 15.3% ($n = 13$) were malignant. Over-conservative surgery (OCS) could be performed in 72.2% of benign masses. The most common benign mass was cystic teratoma (MCT) (54.2%) and malignant mass was mixed ovarian germ cell tumor (30.8%). In long-term follow-ups patients who underwent oophorectomy due to disgerminoma and mixed germ tumor were undergone contralateral oophorectomy due to asynchronous metachronous tumor in adulthood. Asynchronous contralateral mass was found in 5.1% and ipsilateral recurrence in 5.1%. There was no difference in rates of ipsilateral recurrence and asynchronous metachronous teratoma between OCS and oophorectomy in MCT. There was no difference between the age of interrogation of in groups. Metrorrhagia (54.1%) and dysmenorrhea (51.4%) in OCS group were higher than oophorectomy group. There was no difference between two groups in pregnancy success and blood hormone levels ($p = 1.00$).

Conclusion: There was no difference between oophorectomy and OCS in terms of fertility. However, in the OCS group, dysmenorrhea and metrorrhagia were more frequent than oophorectomy. In oophorectomy group, the contralateral similar mass in MCT isn’t higher than OCS group.
Background: The use of permanent central vein catheters (CVC) are essential component of pediatric oncological treatments. The two most commonly used CVC types are port-a-cath (PAC) and Hickmann line (HIC). Monitoring of the surgical complications and nursing helps improve the results and maximize the CVCs life.

Method: In a retrospective study, we analysed the CVCs inserted in a Central European Oncological Center between 2006 and 2014. We investigated the insertions, the CVC-related infections and the removal causes. Fischer's Exact tested the relationship between CVC types.

Results: In the examined period it was implanted 592 CVCs in the Clinic. 69 CVCs were excluded for various reasons. Among the residual 523 CVCs it was 423 (80.88%) PAC and 100 (19.12%) HIC type.

The complication rate of insertion for PAC was 8.51%, and for HIC was 3%. There was no significant difference between the CVC types (p = 0.05912).

We could follow the care of 491 CVCs, which means 182230 nursing days. During this period 183 CVCs had 316 catheter related sepsis. 46 (9.37%) catheter was removed due to sepsis. The investigated 409 PAC has 272 sepsis, and the 100 HIC has 44 sepsis during nursing period.

106 (21.59%) catheter removal occurred due to catheter-related complications. 85 (20.78%) PAC and 21 (25.61%) HIC were removed due complications. There was no significant difference between the CVC types (p=0.3773).

Conclusion: There is no significant difference between the complication rate of PAC and HIC catheter types, either of insertion or nursing.
LONG-TERM RESULTS AFTER DIVERSION SURGERY IN EXTRAHEPATIC PORTAL VEIN OBSTRUCTION

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Aim of the Study

Extrahepatic portal vein obstruction (EHPVO) is the most frequent cause of portal hypertension in children. The aim of this study is to analyze long-term results after diversion surgery.

Methods

Retrospective review of EHPVO patients who underwent diversion surgery analyzing number of platelets, leucocytes, prothrombin activity, splenomegaly and gastrointestinal bleeding 10 years after surgery.

Main Results

Thirty-three patients were evaluated, being mostly males (64%) and presenting with gastrointestinal bleeding (46%). Mesoportal shunt (Rex) was performed in 19 patients, mesocaval in 7, distal splenorenal in 7 and proximal splenorenal in 3.

When comparing mesoportal shunt to the other portosystemic shunts an increase in platelets was found with every technique, but clearly higher in mesoportal shunt. The highest increase was 6 months after surgery (p=0.0015) as well as prothrombin activity (p=0.0003). Leucocytes level also increased without statistically significance. Spleen size (cm) and spleen size Z score (SSAZ) decreased significantly 6 months after mesoportal shunt (p=0.0168).

Before surgery over 94% patients suffered gastrointestinal bleeding, reducing significantly afterwards with bleeding episodes in only 4 of them (12%).

Conclusions

Diversion surgery in EHPVO, especially mesoportal shunt of Rex, improves hepatic function (prothrombin activity), reduces hipersplenism (platelets, leucocytes and spleen size) and decreases gastrointestinal bleeding episodes.
ABERNATHY SYNDROME: VARIED PRESENTATIONS, MANAGEMENT AND OUTCOME

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Introduction: Abernathy syndrome or Congenital portosystemic shunt (CPSS) are very rare abnormalities which have varied presentation like cholestasis, hepatopulmonary syndrome, encephalopathy or liver tumours. We discuss our experience in managing such patients.

Aims: To evaluate the presentation, anatomy, management and outcome of children with congenital portosystemic shunt managed by us.

Materials: Patients with congenital portosystemic shunt treated between 2013-2017 were evaluated for their presentation, management and outcome.

Results: Five patients with median age of 6 years (3.5 years -13 years) were included in this study. Three patients, aged 3.5, 5 and 13 years presented with cyanosis, respiratory distress and clubbing, 1 (12 years) with jaundice, while 1 patient (6 years) had presented with a hepatic mass (Focal nodular hyperplasia). Occlusion angiography was performed in all patients and was diagnostic for type 2 CPSS in 4 patients and was suspect in 1 patient. Liver biopsy was performed latter. Operative ligation of the CPSS was performed in all patients with good outcome. The features of Hepatopulmonary syndrome resolved in the 3 patients while in the patient with FNH, the lesions have decreased in size and no new lesions have appeared.

Conclusions: CPSS can have varied presentation. ligation of the shunt resulted in good outcome in these patients with type 2 CPSS.
Aim of this study

Cholelithiasis is the rare condition in children. Congenital heart disease (CHD) is considered to be a risk factor for cholelithiasis. In this study, we investigate what is the optimal treatment of gallstones in CHD patients.

Methods

In the last 5 years, 11 patients with CHD and cholelithiasis were identified and reviewed.

Main result

The CHD of all 11 children with cholelithiasis was cyanotic. In eight patients, gallstones were found at the studying heart disease and 3 patients were noticed by the symptoms related to cholelithiasis. Ten children with cholelithiasis received ursodeoxycholic acid and no one was observed on dissolution of gallstones. Four patients treated with cholecystectomy after reparable surgery for CHD and 2 of them underwent urgent surgery. The indications for urgent surgery were acute cholangitis and choledocholithiasis. One patient required urgent surgery presented liver abscess. Two other children are ready for cholecystectomy because gallstones are developing.

Conclusion

Prophylactic administration of ursodeoxycholic acid is not effective in children with cyanotic CHD and gallstones. The children with cyanotic CHD and gallstones regardless of cardiac repair always have the risk of urgent cholecystectomy. We recommend the elective cholecystectomy to children with cyanotic CHD if gallstones develop, for avoiding urgent surgery.
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**Aim:** Biliary atresia (BA) is one of the most frequent causes of obstructive jaundice in neonates. In the porta-hepatis, typical histological features are of chronic inflammation with proliferation of bile micro-tubules. Cytomegalovirus (CMV) infection is thought to be a triggering mechanism for BA by activating innate immunity against the bile duct epithelium. We would like to present a cohort of GB histology in BA cases with particular attention to CMV infection.

**Material:** Over a 14-year period (2004-2017), 51 Kasai procedures were performed for BA. In 15 cases, no discernible GB was present. In a further 4 cases histological specimen was untraceable. In the remaining 32 cases, a GB was present and was resected and sent for histological examination. 10 patients were CMV IgM positive at surgery. Ethics was obtained.

**Results:** A wide variety of histological features was present. This variation was from; normal histology (9 cases) which was noted only in CMV IgM negative patients, to cuboidal epithelium with variable replacement of smooth muscle by fibrosis (10 cases) and finally, obliteration of epithelium with dense fibrosis (13 cases).

**Conclusion:** BA is considered to be a singular disease entity and histologically it has unique features in the porta-hepatis. However, the GB with its smooth muscle wall and columnar epithelium lining is affected to a varying degree. This is suggestive that BA is a myriad of different diseases and CMV is possibly a triggering mechanism which causes obliterating cholangitis in a minority of patients.
PW15-OH11
TREATMENT OF INTRACTABLE CHOLESTATIC PRURITUS WITH PARTIAL BILIARY DIVERSION

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Abstract

Aim of the study:

Partial biliary diversion (PBD) may relieve intractable cholestatic pruritus caused by various rare liver diseases. The aim of this study was to report results of PBD from three Nordic referral centers.

Methods:

Retrospective review of medical records of all patients operated with PBD. Approval from ethical review boards was obtained.

Results:

30 patients (17 female) with Progressive Familial Intrahepatic Cholestasis (PFIC)1 (4 pts), PFIC2 (17 pts), PFIC3 (1 pt), Aagenaes syndrome (1 pt), Alagille syndrome (2 pts), and unknown (5 pts) were included. The initial procedures were cholecystojejunostomy (73%), button in the gallbladder (23%) and button in the jejunal conduit (3%). 3/30 operations were laparoscopic. Median age at PBD was 1.6 yrs (0.3-13). Serum levels of bile acids preoperatively and at discharge were median 334 (65-687) and 82 (2-383) mmol/L respectively. Stoma complications were common; leakage, skin irritation and infection (23%), poor bile flow needing stoma revision (7%), and prolapse (7%). Three patients with button in the gallbladder had conversion to cholecystojejunostomy due to insufficient drainage, and five had external PBD converted to internal PBD. Median use of the PBD is 83 (1-298) months. At present, 40% has undergone a liver transplantation, and they had the PBD for median 70 (2-256) months. In those with the PBD still in use, 57% has no itching, 29% some itching and 13% troublesome itching.

Conclusion:

A significant number of patients may achieve satisfactory relief of pruritus after PBD and thereby postpone the need for liver transplantation.
In increased prevalence of pancreaticobiliary maljunction in biliary malignancies

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Aim: The incidence of pancreaticobiliary maljunction (PBM) approximates 1:100,000 within Western populations. We aimed to study the significance of PBM in biliary tract malignancies.

Methods: Medical records and magnetic resonance cholangiopancreatography (MRCP) findings of 252 consecutive patients treated for biliary malignancies during 2005-2016 were reviewed. Patients with other known risk factors for biliary cancers (n=27) were excluded. A common pancreaticobiliary channel located outside the duodenal wall and measuring >10mm was defined as PBM.

Main results: Of the 225 patients, a reliably interpretable preoperative MRCP was available for 73 patients (32%). Sex (47% vs. 57% females) and age at diagnosis (67 vs. 66 years) were similar among patients with or without an MRCP (p=ns for both). In MRCP a PBM with a median length of 20mm (range 10-23mm) was identified in four patients (5.5%, 95% confidence interval 1.6-14), while none had accompanying biliary tree dilatation. PBM patients were significantly more often females (100% vs. 43%, p=0.043), less likely to have intrahepatic bile duct cancer (0% vs. 65%, p=0.019) while more likely to have gallbladder cancer (75% vs. 22%, p=0.044) compared to the others (Figure). Age at diagnosis (66 vs 67 years, p=0.898), extrahepatic bile duct cancer incidence (25% vs. 13%, p=0.453) and survival status at last follow-up (50% vs. 42% alive, p=1.000) were comparable between subgroups.

Conclusion: The prevalence of PBM is substantially higher in adults with biliary malignancies than one would expect based on its incidence, reinforcing the etiologic role of PBM especially in females with gallbladder cancer.
**PW16-BS01**

**LASER SPECKLE CONTRAST IMAGING TO EVALUATE INTESTINAL MICROCIRCULATION IN NEC**

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**Aim of the study:** The role of ischemia in the pathogenesis of necrotizing enterocolitis (NEC) remains unclear. We have previously used laser speckle contrast imaging (LSCI) to identify reduced intestinal microcirculation in cases of mild NEC lesions in the small intestine in an animal model. In this study we examined piglets with both mild and severe NEC-lesions.

**Methods:** 13 preterm piglets were delivered by caesarean section and fed with parenteral nutrition and increasing volumes of infant formula. We used high volumes (15 ml/kg every 3hrs) of formula in order to induce severe NEC-lesions. During surgery at day 3-5 macroscopic NEC-lesions were detected using a macroscopic scoring system (1-6 for increasing NEC severity). A picture of the bowel was taken with the LSCI exposure.

**Main Results:** We found significant lower flux-values in the intestine with NEC-lesions score 3-4 compared to intestine with NEC-lesions score 1-2. The same applies to NEC-lesion score 5-6, where the intestinal microcirculation was reduced significant compared to NEC-lesion score 3-4.

**Conclusions:** This study confirms our earlier findings that a decreased microcirculation seems to be an important part of the pathogenesis of NEC. We found a significant association between the extent of NEC-lesions and the microcirculation in both colon and the small intestine. This animal model might be used to further explore microcirculation in the pathogenesis of NEC and to identify a cut of value to evaluate whether a NEC-lesion is potentially reversible.
PIK3CA RELATED OVERGROWTH SYNDROME (PROS): TOWARDS A PHARMACOGENETIC THERAPY WITH RAPAMYCIN FOR COMPLEX VASCULAR MALFORMATIONS

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Aim of the Study
Combined capillary-lymphatic-venous malformations (CVM) associated with segmental overgrowth (SO) in a syndromic pattern have been recently related to mosaic somatic mutations of PIK3CA gene, belonging to PI3K/mTOR pathway, and to both somatic and germinal mutations in complementary pathways (TIE2, RASA1, PTEN). This clinical condition have been recently defined with the acronym PROS. Aim of this study was to explore the molecular basis of a pharmacologic inhibition of mTOR by Rapamycin (Sirolimus) in CVM therapy.

Methods
From January 2015 to December 2017 sixty-nine patients affected by CVM underwent genetic-molecular studies through Next-Generating Sequencing (NGS) on pathologic tissue biopsies and peripheral blood. NGS was performed using a MiSeq Illumina instrument optimizing a combination of different approaches. Deep sequencing of multiplex amplicon library generated from custom oligo panel of pre-selected 25 genes involved in vascular anomalies, was performed using an Illumina TSCA kit.
Two patients affected by PROS were enrolled for Rapamycin therapy.

Main Results
29 mutations were identified: 19 in PIK3CA gene; 6 in GNAQ gene, 4 in TEK gene with a 2-30% mosacism rate. In 2 cases (mother and daughter) familial germinal mutation in TEK gene was detected.
The 2 patients receiving Rapamycin therapy had a significant favourable response without side effects (follow-up one year).

Conclusions
Genetic-molecular studies through NGS in patients affected by CVM have shown a significant correlation between SO and somatic mutations of PIK3CA gene. NGS could be employed as a pharmacogenetic screening tool to select patients possibly responsive to systemic Rapamycin.
PW16-BS03
WEIGHT DETERMINES THE RESILIENCE OF THE RODENT OESOPHAGUS SUBJECTED TO TRACTION

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Aim of the study: Anastomotic tension is a relevant problem in the repair of oesophageal atresia, especially in cases with a long-gap variant. As it is often advocated that the infant’s weight and size has to be taken into account and therefore a long-gap needs to be classified based on vertebral bodies, we hypothesized that weight may influence the resistance of the oesophagus to traction forces.

Methods: We subjected the explanted oesophagi of 18 Sprague-Dawley rats, aged 15 to 444 days (n=2/timepoint), to linear traction using a motorised horizontal test stand until oesophageal mucosa became visible. Experiments complied with the directive 2010/63/EU. We powered our analysis to 95% using preceding exploratory analyses (n=5).

Main results: Age and weight correlated highly with oesophageal resilience (R=0.731 and R=0.846). Both were significant univariate predictors of anastomotic durability (traction=1.103N+0.005*weight[grams], P<0.001; traction=1.861N+0.009*age[days], P<0.001). However, age and weight strongly correlated (R=0.911), and age was a predictor of weight (P<0.001; weight=130.4g+1.83*age[days]). The multivariate model traction=0.82N-0.002*age[days]+0.008*weight[grams] to predict sustained traction forces based on weight, age and their interaction reached statistical significance (P=0.009) with adequate goodness-of-fit (adjusted R²=0.707). Only weight was a significant predictor of traction resilience (P=0.003), whereas age (P=0.566) and the interaction term (P=0.187) were not.

Conclusions: Weight determined oesophageal anastomotic resilience in a multivariate rat model. This suggests that weight may play a more crucial role in the timing of surgery than age.
PW16-BS05
FEASIBILITY OF FLUORESCENCE ANGIOGRAPHY TO DETECT BOWEL ISCHEMIA IN A PIGLET MODEL OF PREMATURE NEONATES WITH NEC

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Aim of the study: Reduced intestinal microcirculation are thought to be a part of the pathogeneses in NEC. The aim of this study was to evaluate the feasibility and usefulness of intraoperative assessment of vascular perfusion in NEC-lesions using indocyanine green fluorescence angiography (ICG-FA) both during laparoscopic and open surgery.

Methods: 10 piglets were delivered by caesarean section and fed with parenteral nutrition and increasing volumes of infant formula. During laparoscopic or open surgery macroscopic NEC-lesions were detected using a macroscopic scoring system (1-6 for increasing NEC severity until necrosis). The intestinal perfusion was assessed by ICG-FA laparoscopy (n = 6) and laparotomy (n= 4). ICG was given as a bolus of 0.25 mg/kg injected in the umbilical artery, then the tissue of interest was exposed to light at an excitation wavelength of approximately 750-800 nm.

Results: We found a significant decrease in ICG-FA values in NEC-lesions score 3-4 compared to NEC-lesions score 1-2 and in NEC-lesions score 5-6 compared to score 3-4 in both colon and the small intestine.

Conclusions: Our result supports the hypothesis that ischemia is an important part of the pathogenesis in NEC. ICG-FA seems to be a feasible and useful technique both during open and laparoscopic surgery to evaluate microcirculation in NEC-lesions.
MATERNAL PUFA N-3 SUPPLEMENTATION PREVENTS HYPEROXIA-INDUCED PULMONARY HYPERTENSION IN OFFSPRING

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Background: Bronchopulmonary dysplasia (BPD) associated to pulmonary hypertension (PH) affect 18-25% of premature infants, with significant perinatal mortality. Polyunsaturated fatty acids ω-3 (PUFA ω-3) has been shown to improve vascular remodeling under pathophysiological conditions. However, no study examining such effects of PUFA ω-3 supplementation in vascular remodeling, or on angiogenic markers in BPD-associated PH was reported.

Objectives: To evaluate the effects of maternal PUFA ω-3 supplementation on pulmonary vascular injuries occurring in rat PH model.

Methods: From embryonic D15, pregnant Spague-Dawley rats were supplemented daily with PUFA ω-3, PUFA ω-6 or normal saline (0.2 ml/day). Within 24h after birth, pups were randomly assigned to either in room air or hyperoxia (O2) exposure (FiO2=85%) for 20 days and then sacrificed for pulmonary hemodynamic and morphometric analysis.

Results: Maternal PUFA ω-3 supplementation significantly improved survival rate, the increase in right ventricular systolic pressure caused by O2, and the alveolarization and arterial remodeling. Following PUFA ω-3 supplementation, fatty acids analysis revealed a consistent higher level in total ω-3 together with lower levels of total ω-6, as measured in lung and breast milk. Furthermore, reduced mRNA expressions of VEGFA, VEGFR-2, ANG-1, eNOS and PECAM-1 and NO concentration in lung, together with increased ANG-2 mRNA level in O2-exposed rats, were reversed by PUFA ω-3 supplementation.

Conclusion: These data indicate that maternal PUFA ω-3 supplementation strategies could be effective in the protection of BPD-associated PH in preterm newborns.
Aim of the study: In this study, we intend to establish an experimental pathway for biobanking human Amniotic Fluid Stem Cells (AFSCs) under GMP conditions.

Methods: We performed a comparative study between the standard media Chang C and the xeno-free media Chang D, in an effort to disentangle the expression profile of hAFSC, ensure minimum variations owing to the source and yield results with consistent quality. We compared the characteristics of n=48 amniotic fluid samples originating from the 2nd trimester (Gestational Age, GA:15-27 weeks (w)) and 3rd trimester (GA:28-40w). We assessed proliferation, viability and cell cycle phase in CD117+ hAFSC (passage 0-5; P0-P5) and characterized for embryonic (ESC) and mesenchymal stem cell (MSC) markers using FACS and qPCR.

Results: hAFSC cultured in Chang D media took longer to adhere on petri dishes (240±6.2 hours vs. 276±1.5 hours, p=0.05, n=50), but there was no difference from cell cultured in Chang C in both doubling time (36±5.8 hours vs. 36±6.1 hours) and cell viability thereafter (P0: 1.57±0.98, vs. 1.41±0.62, P3: 41.73±3.13 vs. 44.01±1.07). Cell cycle analysis showed that, for both culture conditions, the majority of cells were at G0/G1 (59±0.85% vs. 62±2.32%) with a reduced percentage of S phase cells (11±0.2%) at P3.

Conclusions: Our findings have significant translational value and may be utilized for the development of GMP protocols for hAFSC biobanking.
PW16-BS08: DYNAMIC 3D CULTURE IMPROVES IPSCS DIFFERENTIATION TOWARDS MATURE HEPATOCYTE-LIKE CELLS

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Aim of the study
Transplantation is the only treatment for liver failure however donor shortage and immunologic response represent an issue in this clinical field. Both these limitations could be overcome by whole organ tissue engineering. Decellularised livers preserve the complex anatomy and the biochemical niche of the organ. Induced Pluripotent Stem cells (iPSc) represent a valuable cell source for the regeneration, however they require a three-dimensional environment to reach a mature phenotype. The aim of this study is to investigate the differentiation and growth of iPSCs towards hepatocyte phenotype when cultured on decellularised livers inside a custom-made bioreactor.

Methods
Mice livers were decellularised using a detergent-enzymatic approach. Human iPSCs were differentiated in vitro into hepatic endoderm. Cells were harvested at the endoderm stage, injected into the decellularised scaffolds and cultured in a bioreactor for 14 days. Cells cultured in static conditions on decellularised liver lobes and Matrigel-coated plates were used as control.

Main Result
We observed a faster differentiation of iPSCs with higher expression of mature hepatocyte markers (albumin) and lower expression of fetal markers (AFP) in the decellularised scaffold compared to 2D culture. The bioreactor cultured scaffold showed an increased distribution and spreading of the cells throughout the scaffold in comparison to static culture.

Conclusion
Here we demonstrated how a 3D culture on extracellular matrix scaffold inside a bioreactor facilitates a more mature cell phenotype. These findings pave the way to the engineering of whole livers.
**PW16-BS09**

**INHIBITION OF HEDGEHOG SIGNALING CASCADE IS COORDINATED WITH ACCELERATED INTESTINAL EPITHELIAL CELL TURNOVER DURING INTESTINAL ISCHEMIA-REPERFUSION IN A RAT**

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**Aim of the study:** The purpose of this study was to evaluate the role of hedgehog (Hh) signaling 24 and 48 hours following intestinal ischemia-reperfusion (IR) in a rat.

**Methods:** Rats were divided into four experimental groups: Sham(24h) rats underwent laparotomy and were sacrificed after 24 hours, Sham(48h) rats underwent laparotomy and were sacrificed after 48 hours, IR(24h)-rats underwent occlusion of both superior mesenteric artery and portal vein for 20 minutes followed by 24 hours of reperfusion, IR(48h)-rats underwent IR followed by 48 hours of reperfusion. Intestinal structural changes, enterocyte proliferation and enterocyte apoptosis were determined by immunohistochemistry. Hh-related genes and protein expression were determined by using Real-Time PCR, Western blot and immunohistochemistry.

**Main results:** IR-24 rats demonstrated a significant decrease in GIL mRNA in jejunum and in Shh and PTCH2 mRNA in ileum compared to Sham-24 animals that was accompanied by a significant decrease (2.3-fold decrease in jejunum) in the number of Shh-positive cells (Immunohistochemistry). After 48 hours, IR rats demonstrated a significant increase in Ihh, PTCH2 mRNA in jejunum and in Dhh, Ihh, SMO, GIL, PTCH2 mRNA in the ileum compared to IR-24 animals that was coincided with increased number of Shh-positive cells in jejunum (2.6-fold increase) and ileum (1.4-fold increase).

**Conclusions:** 24 hours following intestinal IR, inhibited cell turnover was associated with inhibited Hh signaling pathway. Signs of intestinal recovery appeared 48 hours after IR and were correlated with increase in Hh signaling pathway activity.
PW16-BS10
QUANTIFICATION OF SERUM TRYPPTASE LEVEL IN PRETERM INFANTS TO EVALUATE MAST CELL ACTIVITY

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Introduction: Prematurity exposes newborns to infectious complications due to an immature immune system. Mast cells seem to have a central place in their defenses.

Objective: Evaluate mast cell activity in preterm infants using assay of serum tryptase level, a specific protease of mast cell activation.

Material and methods: A case-control study compared serum tryptase levels of preterm infants to full-term infants. We also evaluated serum tryptase level kinetic’s in preterm infants, with and without infectious complications.

Results: 51 patients were included per group. Serum tryptase levels were homogenous in full-term infants and variable in preterm infants. Mean serum tryptase level was lower (p = 0.02) in preterm infants (5.02 μg/L) than in full-term infants (6.17 μg/L) after excluding 4 preterm infants presenting confounding factor (prolonged premature rupture of membranes). This result reflects a quantitative deficit in immunity effector cells of preterm infants. Preterm infants at risk of enterocolitis had a higher basal tryptase level (7.02 μg/L) than the others (4.84 μg/L) (p = 0.04). Prolonged premature rupture of membranes is associated with a higher level of tryptase: 12.37 μg/L versus 5.06 μg/L (p <0.0001).

Conclusion: Prognosis of the most vulnerable preterm infants seems to be dependent on unregulated mast cell hyperactivity leading to a deleterious inflammatory response in different epithelia. Serum tryptase assay has an interest in screening these at-risk patients. Additional data are needed to establish threshold values.
BIOMECHANICAL ANALYSIS OF MUSCLE WOUND HEALING IN A MURINE MODEL

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INTRODUCTION

Skeletal muscle tissue engineering is a new strategy of tissue repair. We have already shown that human progenitor muscular cells are able to be integrated in a wounded mouse muscle without immune rejection. We want to show the functional recovery of the repaired tissue repair of the same wounded mouse muscle treated with progenitor skeletal muscle cells. The aim of this study is to analyze the biomechanic properties of this muscle wound healing in our mouse model.

METHODS

Human muscle progenitor cells (14 w.of gestation) were expanded for the experimentation. These cells were associated with a collagen scaffold. The right gastrocnemius muscle of C57BL/6 mice were injured. The injured thigh muscles were partially replaced (hole of 4 mm) either with a collagen scaffold disc with human marked fetal muscle cells (1x10⁵), with the scaffold disc alone or with nothing. An analysis of the force of the muscle contraction as well as the tetanization and the endurance of the muscle was performed at day one and at 8 weeks.

RESULTS

Our mouse model is well adapted to test the muscle contractility even after a severe injury. Our preliminary results show that the absolute peak twitch obtained from the injured muscle is lower than the one obtained from the non injured contra-lateral muscle at different time point after the intervention.

CONCLUSIONS

Fetal human cells are able to engraft on mouse muscle. These cells associated with a collagen scaffold are able to improve the functional recovery after an muscle injury.
LUNG TRANSPLANTATION FOR LATE ONSET PULMONARY HYPERTENSION IN CONGENITAL DIAPHRAGMATIC HERNIA: CASE REPORT AND LITERATURE REVIEW

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Aim of the study: Lung hypoplasia and pulmonary hypertension (PH) associated with congenital diaphragmatic hernia (CDH) may cause fatal respiratory failure. Lung transplant (LTx) may represent an option for CDH-related end-stage pulmonary failure. Aim of present study is to report a patient with CDH who underwent Ltx and to review the relevant literature on the topic.

Case report: GO was born at 33 weeks of gestation, with a prenatally diagnosed isolated left CDH. Twenty-four hours after birth, she underwent surgical repair of a type D defect. Post-operative course was unexpectedly uneventful and she was discharged home at 58 days of life. Echocardiography before discharge was unremarkable. Periodic follow-up revealed gastroesophageal reflux (GER) and initial scoliosis. At the age of 10 she was re-admitted for severe PH. Lung function progressively deteriorated and at the age of 14 she underwent heart-lung transplantation due to end-stage respiratory failure. After discharge, she developed recurrent respiratory tract infections, severe malnutrition, and drug-induced diabetes. Scoliosis and GER progressed, requiring posterior vertebral arthrodesis and antireflux surgery. Bronchiolitis obliterans further impaired her respiratory function, and despite she had a second Ltx she lastly died at the age of 18, 4 and 1.5 years after the first and the second Ltx, respectively.

Table 1 shows 7 further cases of LTx in CDH children collected from the Literature.

Conclusions: Late-onset PH is an ominous complication of CDH. LTx may be considered as a last-resource treatment in CDH patients with irreversible and fatal respiratory failure, although its prognosis seems unfair.

<table>
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<th>Author</th>
<th>Year</th>
<th>Pts</th>
<th>Prenat. Diag.</th>
<th>ECMO</th>
<th>CDH side</th>
<th>Type of Tx</th>
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<tr>
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<td>L</td>
<td>Heart-lung 12 years</td>
<td>Alive for 2 years</td>
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Aim of the study:

The appendix has been used as a catheterisable conduit since the 1980s. We describe a case of incidental carcinoid tumour in the tip of an appendix used for an Antegrade Continence Enema (ACE) procedure.

Methods:

We undertook an institutionally approved single patient case-note review and surveyed paediatric surgical and urological practice in the UK.

Results:

A 9-year old boy, with previous anorectal malformation, neuropathic bladder and bowel, underwent ileocystoplasty, Monti-Mitrofanoff and appendix ACE procedure. The macroscopically normal appendix tip was sent for routine histopathology, standard practice for the operating surgeon. Microscopy demonstrated a 5mm well-differentiated neuroendocrine tumour extending into muscularis propria, Ki-67 index <2%. Due to margin involvement, and following multidisciplinary recommendation, the appendix conduit and surrounding skin were re-excised and a tube caecostomy created through a separate incision. On full appendix microscopy, there was no residual tumour and no further treatment required.

Two-thirds of all GI tract carcinoid tumours are in the appendix tip. To our knowledge, this is only the second reported paediatric case of incidental carcinoid in an appendix conduit. Whilst a variety of techniques for Mitrofanoff and ACE formation are reported, none mention whether the appendix conduit tip should be sent for histopathology. However, the majority of UK surgeons surveyed do not request routine histology on the appendix tip.

Conclusions:

In an era of increased litigation and with the recognised, albeit small, risk of carcinoid in the appendix conduit, we recommend routine histological examination of the appendix tip.
SCIX-CR03

POSTPNEUMONECTOMY SYNDROME IN A NEW BORN AFTER RIGHT PNEUMONECTOMY DUE TO CONGENITAL BRONCHIAL ATRESIA

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Miguel Servet Universitary Hospital, Zaragoza, Spain

Introduction. Postpneumonectomy syndrome (PS) is a rare and severe complication of pneumonectomy, more frequent in infants and young children than in adults. It is caused by a severe mediastinal shift and rotation, causing bronchial stretching of the remaining lung with severe respiratory failure. Surgical treatment includes placing a prosthesis in the empty chest cavity.

Case report. We report a case of an eight day old patient admitted to our hospital with respiratory distress and prenatal diagnosis of congenital pulmonary airway malformation. After a CT and flexible bronchoscopy the patient was diagnosed with right congenital bronchial atresia. A right pulmonary hiperinsuflation with a severe mediastinal displacement, compressing left hemitorax was observed on the CT. A right pneumonectomy was performed. During the postoperative course, the patient experienced respiratory failure secondary to a mediastinal shift causing tracheal deviation and obstruction of the left main bronchus. Due to the PS an expandable prosthesis was implanted temporarily in the right hemitorax, re-establishing symmetry of the mediastinum and trachea and reopening the left main bronchus. However, the patient continued with respiratory distress despite an absence of airway obstruction. The patient died at 3 months of age.

Conclusions. PS is a life-threatening complication of pneumonectomy secondary to congenital bronchial atresia. The insertion of an expandable prosthesis can improve the respiratory symptoms; however, treatment is generally unsatisfactory. Preventing PS implanting the tissue expander at the time of pneumonectomy is probably the best viable option.
INTERCOSTAL GASTROSTOMY: A STEP TO SURVIVAL FOR CDH WITH A OA/TOF

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*Sheffield Childrens Hospital, Sheffield, United Kingdom*

**Aim**
The combination of oesophageal atresia and distal tracheo-oesophageal fistula (OA-TOF) with a congenital diaphragmatic hernia (CDH) conveys an 80% mortality. We present a neonate with an OA-TOF and left CDH who had a large stomach in the chest impeding ventilation. Our aim is to demonstrate the timely intervention of a pigtail chest drain to treat a tension gastro-thorax.

**Methods/Results**
35+5, 2.2kg, female, antenatally diagnosed left CDH with predicted lung volumes of 80%. She was intubated but had recurrent desaturations and profound bradycardias requiring chest compressions. Inability to pass a nasogastric tube demonstrated she also had OA-TOF. At 2 hours, she had worsening respiratory acidosis despite oscillation. CXR showed a very inflated stomach in the left hemithorax causing mediastinal shift and effectively a ‘tension gastro-thorax’. The stomach needed decompressing within the thoracic cavity. We inserted an 8Fr Fuhrman seldinger pigtail chest drain (Cook Medical Europe Limited®) through the left thorax into the stomach (Figure 1). Over the next 12 hours her respiratory acidosis improved and we ligated her fistula. We subsequently repaired her CDH and OA and she is thriving at home 18 months later.

![Figure 1](image)

**Conclusion**
Surgical dogma dictates that decompressing the stomach in a patient with OA-TOF can lead to preferential ventilation through the fistula and should never be done. We present the rare case of a patient with an OA-TOF and CDH who required just that. In cases of tension gastro-thorax, consider the use of a pigtail chest drain as an intrathoracic gastrostomy.
SCIX-CR05
CALCIFICATION IN ASSOCIATION WITH A BENIGN THYROGLOSSAL DUCT CYST IN A CHILD

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¹mugla Sitki Kocman University Research And Training Hospital Department Of Pediatric Surgery, Mugla, Turkey. ²mugla Sitki Kocman University Faculty Of Medicine Department Of Pediatric Surgery, Mugla, Turkey. ³mugla Sitki Kocman University Faculty Of Medicine Department Of Pediatrics, Mugla, Turkey

Aim

Thyroglossal duct cysts (TDCs) are the most common congenital anomalies of the midline cervical region. Carcinoma arising within a TDC has been reported to occur in approximately 0.7% of cases. Thyroglossal duct cyst calcification is a very rare condition. Calcification in TDC is thought to be a specific marker for malignancy, which is seen %1 of TDCs. We here present the first benign recurrent TDC case whose histopathological examination revealed calcification of the cyst wall.

Case presentation

A 5-year-old boy presented with troublesome dischaging, tenderness and redness in front of the neck. He had a history of neck abscess for three times since the operation for TDC at another clinic performed a year ago. Ultrasonography showed a supralaryngeal subcutaneous cyst in size of 24x12 mm with anechoic content. Antibiotics were prescribed before surgical resection. A standard Sistrunk procedure was performed for recurrent TDC. The histopathological findings confirmed calcification in association with a benign TDC. One-year clinical follow up is uneventful without any complication or recurrence.

Discussion

Calcification is an important ultrasonography or computed tomography feature of malignancy in TDC, especially in adults. To the best of our knowledge, there has been only one case reported a 3-year-old boy with calcification occurring in a benign TDC. Our patient is the first reported recurrent benign TDC in association with calcification. Since both cases of benign TDC and calcification comorbidity were reported in children, unlike adults, calcification can not be accepted as a specific marker for malignancy in childhood.
Asynchronous anisoperistaltic and isoperistaltic double longitudinal intestinal lengthening and tailoring

Zafer Dokumcu\textsuperscript{1,2}, Javid Naghiyev\textsuperscript{1}, Emre Divarci\textsuperscript{1,2}, Funda Özgenc\textsuperscript{3,2}, Ahmet Celik\textsuperscript{1,2}
\textsuperscript{1}Ege University Faculty of Medicine Department of Pediatric Surgery, Izmir, Turkey. \textsuperscript{2}Pediatric Intestinal Failure and Rehabilitation Center, Izmir, Turkey. \textsuperscript{3}Ege University Faculty of Medicine Department of Pediatrics, Division of Pediatric Gastroenterology, Izmir, Turkey

Aim of the study: Short bowel syndrome (SBS) results from the alteration of intestinal digestion and absorption. It requires an aggressive multidisciplinary approach that is often tailored to the individual needs of the patient. We aimed to present the first successful case of the asynchronous anisoperistaltic and isoperistaltic double longitudinal lengthening and tailoring (LILT) in English literature.

Methods: Medical records of an infant with SBS after an extensive bowel resection due to gastrochisis and complicated ileal atresia was reviewed.

Main results: A 6-months old boy (weight: 3740 gr, Z-score: -5.93) with SBS (Bowel length: 50 cm jejunum, 15 cm colon) was referred to our center. He was 100\% total parenteral nutrition (TPN) dependent due to malabsorptive diarrhea from the proximal jejunostomy. First, dilatation of the jejunum was achieved with the closure of the stoma and a narrow anastomosis. Four weeks later, an anisoperistaltic LILT (20 cm) was performed in the distal jejunum for retardation of the enteral flow. Seven months later, he was on cyclic TPN and hence, an isoperistaltic LILT (15 cm) was added to the proximal jejunum as a second stage for obtaining adequate bowel length. He was TPN-independent with full oral feeding and gained weight at 18 months of follow-up (weight: 8000 gr, Z-score: -3.63).

Conclusions: Asynchronous anisoperistaltic and isoperistaltic double LILT may offer a good option for the treatment of SBS in selected patients.
SUCCESSFUL PERITONEAL DIALYSIS USING A PERCUTANEOUS TUBE FOR PERITONEAL DRAINAGE IN AN EXTREMELY LOW BIRTH WEIGHT INFANT: A CASE REPORT

Satoshi Yokoyama, Tatsuo Nakaoka
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Background: Although Peritoneal dialysis (PD) has been used occasionally in extremely low birth weight (ELBW) infants, prognosis is poor. Several reports have described successful PD in these infants, but no guideline-based evidence concerning indications for renal replacement therapy in ELBW infants are currently available.

Case presentation: A male was born at 24W3D gestation weighing 264 g by emergency cesarean section due to complications of pregnancy in a patient with HELLP syndrome. On day of life (DOL) 15, the inability to ventilate, along with cardiovascular dysfunction, AKI, and ascites under tension led to the tentative diagnosis of abdominal compartment syndrome. On DOL 17, placement of a percutaneous drainage tube immediately released compression of the tense abdomen. Although intraabdominal pressure reduction with percutaneous drainage temporarily improved respiratory status, circulatory impairment persisted and infections were not well controlled. Finally, the patient developed anuria. On DOL 21, PD was started by initially inserting a drainage tube. Although the patient had catheter-associated peritonitis, urine output improved by DOL 44 and PD was discontinued on DOL 53. On DOL 75, extubation was conducted without circulatory dysfunction. The patient was discharged on DOL 224.

Conclusions: We emphasize that starting PD treatment before the onset of anuria is important in ELBW infants with AKI. Although the catheter used in our case was initially inserted for drainage of ascites, this type of catheter is sufficiently useful for PD in ELBW infants, and PD using a drainage tube may represent a safe, effective, and minimally invasive treatment for ELBW infants.
HERLYN-WERNER-WUNDERLICH SYNDROME: A NEONATALLY DIAGNOSED CASE

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1Centro Hospitalar São João, Porto, Portugal. 2Faculty of Medicine - Porto University, Porto, Portugal

Introduction: Herlyn-Werner-Wunderlich syndrome (HWWS), defined by the triad of didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis, is a very rare complex Mullerian malformation. It is usually diagnosed after menarche, when symptoms related to hematocolpos arise. Most evidence available on HWWS comes from case reports, the majority of them concerning postpubertal girls.

Case report: Herein, we report a case of a female newborn, delivered at 38 weeks and 5 days of gestation, with prenatal US and MR showing right renal agenesis and a cystic pelvic mass. Physical examination after delivery revealed a permeable vaginal introitus in addition to a vestibular bulging, suggesting the presence of a septated/duplicated obstructed vagina. At the 4th day of life, vaginoscopy revealed the presence of an obstructed right hemivagina that underwent septotomy, confirming HWWS. The postoperative course was uneventful.

Discussion: In the presence of a solitary kidney and a pelvic mass on prenatal US, HWWS should be considered, enabling neonatal treatment. Early and accurate diagnosis and treatment are of utmost importance to avoid complications and preserve the reproductive potential.
INTRODUCTION

Combination of posterior urethral valves (PUV) and urachal remnants (UR) are rare. We report a case of a baby with normal antenatal scans, in whom an urachal cyst (UC) and PUV were found.

CASE DESCRIPTION

A 1 day-old baby was transferred to our neonatal intensive care unit because of respiratory distress and acute kidney failure. Baby was stabilised and renal function improved. An abdominal USS demonstrated a midline, anechoic cyst cranially to the bladder. A micturating cystourethrogram showed PUV associated with left vesico-ureteric reflux (VUR). At 23 days of life, baby was well enough to undergo a cold-knife ablation of PUV while the excision of urachal cyst was performed at the age of 4 months. A subsequent videourodynamic assessment demonstrated a small capacity bladder with reduced compliance and bilateral grade 5 VUR.

DISCUSSION:

Association between PUV and UR is extremely rare and there is only one published case reporting PUV and UC.

A proposed theory suggests that a patent urachus can be secondary to the presence of increased pressure in the developing bladder. The presence of a patent urachus as well as VUR would act as a “pop-off” mechanism lowering the intravesical pressure and preserving the kidneys. Although attractive, this theory is not supported by clinical evidence. We could speculate that, despite the presence of a demonstrated high-pressure bladder, the vesical end of the urachus spontaneously closed and we would have expected to find a patent urachus rather than a UC if the pop-off mechanism would have occurred.
CONGENITAL PULMONARY AIRWAY MALFORMATION (CPAM) PRESENTING AS AN SPONTANEOUS PNEUMOTHORAX IN A NEWBORN

Maria Alvarez-Barrial, Borja Nava, Carlos De La Torre, Saturnino Barrena, Francisco Hernandez, Carlos Delgado, Antonio Munoz, Javier Serradilla, Alba Bueno, Leopoldo Martinez, Manuel Lopez-Santamaria
La Paz Hospital, Madrid, Spain

INTRODUCTION
Congenital pulmonary airway malformation (CPAM), is a rare developmental anomaly of the lower respiratory tract. Patients with CPAM are usually asymptomatic, but some may present with respiratory distress. We report a rare presentation as an image compatible with persistent and localized spontaneous pneumothorax.

PRESENTATION OF CASE
A 2-month-old male infant without prenatal diagnosis, postnatal distress or barotrauma, was admitted with acute respiratory symptoms and an image diagnosed as a right tension pneumothorax on chest X-ray. Despite placement of a chest drain, radiological image persisted. CT confirmed the presence of a CPAM consisting of an only large upper–middle right lobe bullae. An open surgical approach was decided and a huge bullae depending from the right upper apex lobe was found and resected. Pathological report disclosed type 4 CPAM.

DISCUSSION/CONCLUSION:
Despite the negative prenatal screening, the diagnosis of CPAM should be considered in a patient with sudden respiratory distress and spontaneous pneumothorax. Type 4 CPAM may appear like unique lung cyst mimicking a spontaneous bullae or a massive pneumothorax.
CONTROLLED ELASTIC LOOP FOR INTESTINAL KINKING (CELIK): A NOVEL SURGICAL TECHNIQUE TO IMPROVE INTESTINAL ADAPTATION IN SHORT BOWEL SYNDROME

Emre Divarci\textsuperscript{1,2}, Zafer Dokumcu\textsuperscript{1,2}, Miray Karakoyun\textsuperscript{1,2}, Funda Ozgenc\textsuperscript{1,2}, Ahmet Celik\textsuperscript{1,2}

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Lengthening procedures could not be performed in some of the patients with short bowel syndrome (SBS) due to narrow diameter of intestine. In this study, we aimed to report our initial experience as a novel surgical technique “Controlled Elastic Loop for Intestinal Kinking (CELIK)”. A 3 months old infant was referred to our center with ultrashort bowel syndrome. Only 8 cm jejunum with proximal jejunostomy and half of colon was present due to midgut volvulus. The diameter of proximal jejunum was not suitable for lengthening. Dilatation attempts such as controlled jejunostomy obstruction with insufflated foley catheter balloon in the stoma and narrow jejuno-colonic anastomosis were failed. An experimental surgical technique was planned to dilate jejunum in the 15\textsuperscript{th} month of life. A vascular elastic tape is passed from mesentery of jejunum, and pulled out from a mini hole in the abdominal wall (Figure.1). In the postoperative period, elastic tape on the skin is squeezed by inserting plastic tube fragments for intestinal obstruction (Figure.2). The diameter of proximal jejunal segment was controlled by x-rays. After a period of 3-4 weeks, patient underwent re-exploration. The diameter of jejunum was enlarged from 2.5 cm to 4 cm, and Bianchi procedure (LILT) could be performed successfully. Patient is in the second month of postoperative period. Intestinal adaptation with enteral feeding improved gradually.

Bowel diameter is crucial for lengthening procedures in SBS. This technique (CELIK) provides safe and controlled bowel obstruction to achieve effective intestinal adaptation before definitive surgery.
UMBILICAL CORD TERATOMA ASSOCIATED WITH OMPHALOCELE MINOR

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¹University of Manitoba, Winnipeg, Canada. ²Diagnostic Services Manitoba, Winnipeg, Canada. ³HSC Children’s Hospital, Winnipeg, Canada

AIM
Teratomas are tumors arising from totipotent embryonic germ cells. The umbilical cord is a rare site for these tumors with only 17 cases reported in the literature. We report a newborn with omphalocele minor associated with an umbilical cord teratoma.

METHODS
A 27-year-old had a normal 20 week ultrasound and delivered at 38 weeks gestation via spontaneous vaginal delivery. At birth, the infant was found to have an omphalocele minor and no associated anomalies. The infant was otherwise well and underwent primary repair of the omphalocele minor on day 3 of life.

MAIN RESULTS
At surgery, a small mass (1.5x1.5x1 cm) was resected and thought to be an umbilical remnant. Review of the pathology demonstrated a mass consisting of a reddish-brown nodular tissue discovered to be an umbilical cord teratoma with mature skin, adipose, and cartilaginous tissue present. There was no evidence of immature elements or somatic cell malignancies.

CONCLUSION
Our case is a rare incidental post-natal finding of umbilical cord teratoma, probably due to the small size of the lesion. Of the 17 reported cases, only 6 involved both an omphalocele and an umbilical cord teratoma. Differential diagnosis of a mass located in the umbilical cord may include small acardiac twins that can be differentiated from umbilical cord teratomas by rudimentary second umbilical cord and craniocaudal skeletal organization. Umbilical cord teratomas may be associated with other midline defects, such as omphalocele and bladder extrophy. Associated anomalies should always be ruled out in presence of umbilical cord teratoma.
WHOOSH SENSATION AND SALTY TASTE IN THE BACK OF THE THROAT AFTER FLUSHING A LIVER ABSCESS WITH NORMAL SALINE!

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University of Manitoba, Winnipeg, Canada

We present a 16-year-old male with a two-week history of nausea, vomiting, diarrhea, abdominal pain and fever. The past medical history was remarkable for well-controlled asthma. The patient had no recent travel history. Physical examination revealed right upper abdominal tenderness and hepatomegaly. Laboratory investigations demonstrated elevated WBC and CRP. CT revealed two liver abscesses (Figure 1A, B) that were subsequently drained using CT-guided insertion of two 10 French pigtail drains. The patient was concomitantly treated with intravenous ceftriaxone for a Streptococcus anginosus infection. The drain in the smallest abscess was removed after 10 days. The patient was discharged home with continuing intravenous ceftriaxone and flushing of the largest abscess with normal saline. One month after the admission, he complained of a sensation of a whoosh and salt water in the back of his throat after flushing of the abscess. A drainogram identified a hepatobronchial fistula (Figure 1C, D). We reduced the volume to flush the abscess from 10 to 2 ml twice daily and continued the intravenous antibiotics. One week later, the salt taste and coughing resolved, and the liver abscess further reduced in size on ultrasound investigation. The drain was removed two months after admission when a follow up drainogram confirmed that the hepatobronchial fistula had healed. All symptoms had resolved on an outpatient clinic appointment five months after the admission. A hepatobronchial fistula is rare, but should be considered when patients complain of a whoosh sensation and salt taste in the back of their throat.

Figure 1
Introduction: Peritoneal tuberculosis is rare in immune-competent infants. We report two cases of peritoneal tuberculosis in infants.

Observations

Case N°1: 6 month old female infant, referred for a day history of postprandial bile stained vomiting, associated with obstipation. On clinical examination, she had a temperature of 38°C, her abdomen was soft and non distended. An abdominal ultrasound revealed an intussuception. At laparotomy, a haemoserous fluid was identified, as well as adhesion, diffuse granulation, and multiple mesenteric adenopathies. We proceeded to sampling the peritoneal fluid, and a nodal biopsy. Postoperatively, a tuberculine skin test came back positive. The pathological analysis of the specimen confirmed the presence of a tuberculoid follicle. The infant was prescribed antituberculous medication. After 6 months, she has achieved full recovery.

Case N°2: 8 month-old male infant referred for vomiting and constipation that began 15 days prior to the presentation. On clinical examination, he had a temperature of 38°C, his abdomen was distended. Laparotomy revealed a peritonitis with a straw coloured fluid, granulations and adhesions. We performed an adhesiolysis, biopsies, as well as drainage of the peritoneal cavity. Postoperatively, the Gene Xpert and Tuberculin Skin Test were negative. The pathological analysis of the specimens revealed caseous necrosis, in keeping with a diagnosis of peritoneal tuberculosis. The medical management was identical to that of the previously mentionned patient, and resulted in a good recovery.

Conclusion: Abominal tuberculosis in infants simulate a surgical emergency. The diagnosis often requires a surgical exploration and biopsies.
PRENATAL DIAGNOSIS OF JUVENILE GRANULOSA CELL TUMOR OF THE TESTIS: CASE REPORT

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Fondazione IRCCS Policlinico San Matteo, Pediatric Surgery Dpt, Pavia, Italy

Prepubertal primary testicular tumors account for approximately 1% of all pediatric solid tumors. Gonadal stromal tumors, including Leydig cell, Sertoli cell and granulosa cell, represent about 8% of these neoplasms. Juvenile-type granulosa cell tumor (JGCT) of the testis is extremely rare and, even if it is the most common neoplasm of the testis in the first 6 months of life, only 2 cases have been described at prenatal ultrasound (PU). We report a new case of prenatal JGCT diagnosis: while performing PU in a male at 38th week of gestation, for a suspected podalic presentation, a left heterogeneous solid and multilocular septated cystic testicular mass was identified. Hypothesis of JGCT was formulated. At birth a left painless scrotal mass was detected. Ultrasound excluded torsion of the testis and confirmed the presence of a left lesion without normal-appearing parenchyma. Tumors markers were normal for age. Inguinal exploration revealed a cystic mass beneath tunica albuginea replacing almost all normal parenchyma: accordingly orchiectomy was performed (figure 1). On the basis of microscopic examination (presence of follicular structures, irregular in size and shape, with intraluminal basophilic fluid - figure 2) and immunohistochemical findings (lack of reactivity for AFP, CD30, vimentin and the positivity for a-inhibin), the histologic diagnosis of juvenile-type granulosa cell tumor (JGCT) was confirmed. At 7-year-follow-up the child presents an uneventful outcome.
GLANS ISCHEMIA AFTER CIRCUMCISION IN A 16-YEAR-OLD BOY: FULL RECOVERY AFTER ANGIOGRAPHY WITH LOCAL SPASMOLYSIS, SYSTEMIC VASODILATATION AND ANTICOAGULATION

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¹Department of Pediatric Surgery, University of Leipzig, Leipzig, Germany. ²Department of Radiology, University of Leipzig, Leipzig, Germany. ³Hospital for Children and Adolescents, University of Leipzig, Leipzig, Germany

Introduction:
Circumcision is one of the most frequent procedures performed by pediatric surgeons. A dorsal penile nerve block (DPNB) is commonly used for perioperative analgesia. The total complication rate of this technique has been reported to be as low as 0.5%. However, few case reports describe ischemia of the glans penis following DPNB. Therapeutic options include hyperbaric therapy, pentoxifylline, iloprost, enoxaparine, anticoagulation, and peridural anesthesia. We report the case of a boy with ischemia of the glans after DPNB.

Case report:
The 16-year-old boy presented on the first postoperative day after circumcision under DPNB (0.25% bupivacaine) at an outside institution. On clinical examination the distal glans was found to be ischemic (A). The patient underwent immediate angiography under sedation. An intra-arterial spasmolysis (alprostadil and nitroglycerine) was performed and a sufficient perfusion of the glans penis was confirmed (B). Subsequently systemic sildenafil, arginine and heparin were given. Following this three-day medical treatment ischemia resolved completely without adverse events (C).

Conclusion:
Ischemia of the glans is a rare complication after circumcision under DPNB. Our case emphasizes the role of angiography in the diagnostic workup and the therapeutic efficacy of local spasmolysis, systemic vasodilatation and anticoagulation.
SERIAL REDUCTION OF AN EXTREMELY LARGE GASTROCHISIS USING VACUUM-ASSISTED CLOSURE

Marilyn Butler¹, Julie Fuchs², Matias Bruzoni²
¹Oregon Health and Science University, Portland, OR, USA. ²Stanford University, Palo Alto, CA, USA

Introduction: Despite widespread use of silos for reduction of gastroschisis, closure remains challenging when the fascial defect is large, eviscerated contents are substantial, medical status is precarious, or abdominal domain is limited.

Case: A boy was born at 35 5/7 weeks with gastroschisis. A spring-loaded silicone silo was placed at birth. By day of life (DOL) 22, minimal visceral contents had been reduced, and the silo was difficult to maintain due to a large fascial defect. A bespoke Vacuum-Assisted Closure (V.A.C.) dressing was constructed as follows: Whitefoam™ and GranuFoam™ dressings were cut to half their thickness, fashioned in the shape of a cup by sewing strips together, and placed over the eviscerated bowel. Strips of adhesive drapes were used to secure the dressing, circumferentially wrapping the infant. After puncturing the drape, a SensaT.R.A.C.™ Pad was placed over the dressing, connected to a V.A.C.® Therapy canister, and placed to negative pressure. Endotracheal intubation was necessary for bi-weekly dressing changes until the viscera were consolidated. Then Mepitel® was placed over the viscera prior to Whiteform™ and GranuFoam™ dressings. When the viscera were completely reduced, Mepitel® was placed until the wound was closed.

Results: By DOL 50 the viscera were completely reduced, and VAC® Therapy was discontinued. Feeds were commenced on DOL 57 and increased to goal. He was discharged home on DOL 115.

Conclusions: V.A.C.® dressings can be used to aid gradual reduction of extremely large gastroschises, particularly in medically fragile infants.
Introduction

Fowler’s syndrome is a rare cause of chronic urinary retention in teenage girls. We report a typical case of a pubertal girl who lost her ability to void after uncomplicated laparoscopic appendectomy.

Case Report

A 14 year old girl presented two weeks after an uncomplicated appendicectomy with reduced urinary frequency and prolonged micturition time. Two months later she completely lost her ability to void after acute cystitis. She underwent a comprehensive set of investigations to elucidate the aetiology of her urinary retention, including MRI of brain and spine. The only abnormal findings were polycystic ovaries, asensitive and atonic bladder with capacity 1200 millilitres and extremely high maximum urethral closure pressure of 120 cm of water on videourodynamics. She did not tolerate clean intermittent catheterisation and suffered from recurrent urinary tract infections with a suprapubic catheter. Finally, after 2 years, based on the history, symptoms and urodynamic findings she was diagnosed Fowler’s syndrome. Implantation of S3 neurostimulator restored her voiding completely back to normal.

Discussion

Fowler’s syndrome is an extreme form of dysfunctional voiding with sphincter hyperactivity followed by atonic bladder. The sacral nerve stimulation is the only known effective treatment. Despite peak incidence between 20 and 30 years of age, it can happen in pubertal and adolescent girls, often after general anaesthesia or use of opiates. It does not occur in men and about 50% percent of Fowler’s syndrome patients have associated polycystic ovaries or endometriosis. It should be thought of in all cases of unexplained chronic urinary retention.
INTRADIAPHRAGMATIC SEQUESTRATION: INTRAOPERATIVE DIAGNOSIS USING MINIMAL INVASIVE APPROACH

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Extralobar pulmonary sequestrations (EPS) are most commonly found within the thorax and occasionally in the abdominal cavity, but intradiaphragmatic location is extraordinarily rare.

We report a case of a 5-years-old-girl with antenatal diagnosis of pulmonary sequestration on the left lower lobe (LLL). Postnatally the patient remained asymptomatic and further studies were performed. The ultrasound and a computer tomography (CT) revealed a focal soft tissue density in the posterior medial LLL that measured 33 × 19 × 38 mm with one vessel extending from the abdominal aorta to the mass. No venous drainage could be identified. With the preoperative diagnosis of intralobar pulmonary sequestration, a thoracoscopic left lower lobectomy was planned. However, LLL seemed normal and the systemic vessel was not found. The whole hemithorax was explored and a mass confined to the diaphragm was observed. After dissection of a superficial muscular layer the mass was completely identified, vascular supply arising from the aorta was controlled and the the EPS was thoracoscopically resected. Postoperative course was uneventful. The pathologic study confirmed diagnosis of a hybrid lesion: type II cystic adenomatoid malformation and EPS.

Preoperative diagnosis of an intradiaphragmatic EPS is challenging and a definite diagnosis is usually found at surgical exploration. Minimally invasive approaches can allow exploration of both the thoracic and abdominal cavities in search for these rare lesions, especially when they are not found in the expected location.
Aim of the study: Gastric transposition (GT) has been a good option for esophageal replacement in children. As one of its advantages, it may be performed with minimally invasive techniques but single incision laparoscopy-assisted GT (SILA-GT) has not yet been reported. We aim to present the first case of thoracoscopic esophagectomy and SILA-GT in English literature.

Methods: Medical records of a child with long-segment corrosive esophagus stricture (CES) due to caustic ingestion were reviewed. Parental informed consent has been obtained.

Main results: A 2.5 years old boy with persistent long-segment CES and chronic aspiration necessitated esophageal replacement at the end of eight months of dilatation programme including topical mitomycin-C application. The scarred esophagus was freed via right thoracoscopic esophagectomy in the first phase of the operation. Following re-positioning, stomach was prepared for transposition via SILS approach using Octoport™. In the third phase, the native esophagus was dissected, excised and gastric transposition and a single layer esophagogastric anastomosis were performed through the left neck incision. The operation was terminated after bilateral gastric fixation suturing at the hiatal level. Operation time was 6 hours and there was no perioperative complication. The child was extubated on second and fed on 7th postoperative days.

Conclusions: Thoracoscopic esophagectomy provides a perfect advantage for secure esophageal dissection and SILA-GT is feasible, safe and efficient in patients with severe CES.
SCIX-CR21
RETRO-RECTAL HERNIA AS A COMPLICATION OF HIRSCHSPRUNG DISEASE SURGERY

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We report the case of an eight-year-old male diagnosed with Hirschsprung Disease (HD) at infant time. He underwent endo-rectal pull-through at the age of six months with no incidences during surgery. Two months later he suffered an episode of Hirschsprung-associated enterocolitis, managed conservative with antibiotics with good response.

After that, the patient had a good outcome but he presented mild fecal incontinence, frequently after meals. Complementary test were all normal, including contrast enema. Treatment with oral laxatives was attempted but the patient presented constant leakage so it was dropped off, and the patient was managed with diet and hygienic measures with no optimal results.

A psychological evaluation was required without any pathological findings. Contrast enema was repeated during follow up, showing a significantly increased presacral space, previously not described. MRI was indicated in order to discard a presacral mass. On the MRI the space between the rectum and the sacrum was fulfilled with small bowels. An upper intestinal transit confirmed the diagnoses of Retro-rectal Hernia. Surgical treatment was decided and laparoscopic rectopexy was performed. The retro-rectal space was closed using absorbable braided 2,0 suture with no complications.

After the intervention, the fecal incontinence disappeared and two years later the patient remains asymptomatic with dairy bowel movements and no leakage.
Hirschsprung disease (HD) and, rarely, intestinal duplication (ID) are causes of neonatal intestinal obstruction. We present a case with the coexistence of these two congenital pathologies. A two-day-old neonate presented with bilious vomiting and abdominal distension referable to intestinal obstruction; abdominal ultrasound revealed a cystic mass in right hypochondrium (38x27mm). We performed an explorative laparotomy and found cystic cecal duplication. Isolated resection of ID was not possible so ileocecal region exeresis and ileocolic end-to-end anastomosis were performed. Post-surgical course was characterized by recurrent bilious vomiting, abdominal distension, constipation and weight loss. At one month of life, due to exacerbation of symptoms, explorative laparotomy was performed. Several adhesions were resected and anastomotic stenosis was excluded. Despite surgery, abdominal distension persisted associated with absence of spontaneous evacuation, thus HD was suspected. Therefore, another explorative laparotomy was planned. Multiple intestinal biopsies and ileostomy were performed. As the diagnosis of HD was confirmed, transanal endorectal pull-through was performed. Intestinal recanalization operation followed three months later. At one-year follow-up the child has normal bowel function and no more episodes of abdominal distension have occurred. Coexistence of ID and HD is extremely rare and only three cases (1 with intestinal neuronal dysplasia) have been previously described in literature. This association could be secondary to an ischemic damage occurring during the embryogenesis, interfering both with the intestinal canalization and the neural crest migration. As showed by our report, the presence of both HD and ID can mislead the diagnostic work up and delay an appropriate therapy.
SCIENTIFIC SESSION IX: CASE REPORTS

SCIX-CR23
UMBILICAL APPENDIX OR APPENDICOUMBILICAL FISTULA: CONGENITAL OR TRAUMATIC?

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BACKGROUND: Congenital umbilical anomalies are mostly represented by urachal or omphalomesenteric remnant. We present the case of a patient presenting umbilical granuloma. Persistent urachus was suspected. At surgical exploration, the granuloma turned out to be the tip of the vermiform appendix (Figure 1). Some very rare cases of congenital appendico-umbilical fistula (AUF) are described. We aim to discuss the origin of AUF.

METHODS: Report of a case of AUF from our institution. A review was performed to collect the cases of AUF from the literature. Informed consent from the reported patient was obtained.

RESULTS: Two origins are described: (1) AUF results from an injury of the tip of the appendix, located at the umbilicus (Figure 2). The umbilical location of the appendix is either caused by failure of the appendix to re-enter the abdomen during embryogenesis, or due to secondary herniation of the appendix, after re-entering of the midgut into the abdomen. (2) AUF is a patent omphalomesenteric duct developed from the tip of the appendix. In every reported case, the congenital aspect cannot be strictly declared.

CONCLUSIONS: AUF appears to be the consequence of an injury of the tip of the appendix, located into an unrecognized umbilical hernia. Surgical exploration allows diagnosis and treatment. Surgeon has to look for an associated malrotation disease.

Figure 1: Intra operative view.
Tip of the vermiform appendix drained to the umbilicus

Figure 2: Hypothesis of appendico-umbilical fistula caused by appendix injuries with the neonatal clamp.
“Gastrostomy” (Gr. *gaster* – stomach + *stomoun* – to provide with an opening, or mouth) is one of the oldest abdominal operations in continuous use. Initially suggested in 1837, the first recorded operation occurred in 1849. Survival was achieved nearly three decades later. Some of the early patients were children with caustic esophageal stricture. The basic gastrostomy types are: A- Formation of a gastric cone through the abdominal incision (1846) or a counter incision (1890); B-Formation of a channel for a catheter from the anterior gastric wall, parallel to the stomach (1891) or perpendicular to it (1894); C-Formation of a tube from the gastric wall (1899); D-Formation of a tube from an interposed bowel segment (1906); E- Gastrostomy catheter placement without laparotomy (percutaneous endoscopic or PEG) (1980), F- Gastrostomy catheter placement with minimally invasive techniques, radiologically (1987) or laparoscopically (1991).


Although the indications for gastrostomy have changed significantly over the last one-and-a half centuries, multiple factors have contributed to its increased use, including longer survival of adults and children with temporary or permanent inability to swallow, need for long term continuous feedings, and administration of frequent or non-palatable medications.
Aim: Since the birth of pediatric surgery as an independent medical specialty, there has been a distinct evolution in the German-speaking world. This expansion can be traced back to few founding pioneers. In this study, we construct a detailed pedigree of pediatric surgery based on a comprehensive survey and face-to-face interviews with German-speaking retired pediatric surgeons.

Methods: A detailed survey was sent to all heads of pediatric surgery departments in Germany, Austria and Switzerland. Participants were asked to name up to 3 most influential teachers in their careers. The results were used to construct a comprehensive pedigree to trace back pediatric surgery training and mentorship in time. Data was analyzed using descriptive statistics and graphics. For supplemental information and corroboration of findings, twelve retired pediatric surgeons were interviewed face-to-face. The recorded audio files were professionally transcribed and correlated with the survey data.

Results: A total of 94 departments were contacted, 69 of which provided usable data (recall 73%). Using the number of respondents identifying specific individuals as their teacher, a pedigree was constructed with 5 main truncs: 1) the German Democratic Republic trunc (Tischer, Meissner, Krause, Benneck, Mau), 2) the Austrian trunc (Hartl, Sauer, Menardi, Wurnig), 3) the Swiss trunc (Grob), 4) a German Federal Republic trunc (Drachter, Rehbein, Oberniedermayr, Hecker, Waag, Willital), as well as 5) an international trunc (Rickham, Georgeson, others) which seemed to have had significant influence on contemporary pediatric surgery in German-speaking countries, since 22% of surveyed German-speaking pediatric surgeons received training abroad. Strong interactions between the 5 main truncs of the pedigree of German-speaking pediatric surgery.

Conclusion: Pediatric surgery developed on the foundation of a number of prominent individuals who the current leadership in German-speaking countries identifies as their teachers and mentors. Our study underscores the affiliation with historically formed schools that characterize the pedigree of pediatric surgeons up to the modern age.
Ordinarius Professor Cemil Topuzlu (1866 – 1958), was a leading surgeon in the Ottoman Empire who was internationally recognized for his pioneering work in many areas of surgery.

Early in his career, he worked for three years as an assistant for the world-recknowned French surgeon Jules-Émile Péan, and then returned to Istanbul to establish the modern surgical principles and surgical training in his country. In a short duration of time, he also became the private surgeon of the Ottoman Imperial family. He later served as the Mayor of Istanbul at the beginning of the 20th century, and became the first Dean of Istanbul University Medical Faculty. He was one of the founding members of the French Surgical Association and one of the first members of the ‘Societe Internationale de Chirurgie’ founded in 1902.

Some of his internationally acknowledged surgical innovations and contributions to Pediatric Surgery can be listed as follows:

- He reported the removal of a pen cover from the right main bronchus of a 7-year-old girl through a tracheotomy in 1903.

- On 27 August 1903, one of his patients undergoing external urethrotomy under chloroform anesthesia developed cardiac arrest, and he performed open chest cardiac massage.

- He introduced novel vascular suture techniques, which he presented at the International Medical Congress in Moscow in August 1897 and at the annual Congress of the ‘Société de Chirurgie de Paris’ in July 1904, where he reported two cases of arterial tear during breast carcinoma resection and repair within the same session.

- He also defined the "do not resuscitate" code in cases involving serious heart disease and other diseases, where life expectancy was very short.

Beside his innovations in the field of surgery, he was the founder of the first Ottoman Pediatric Hospital named Hamidiye Etfal Sisli Etfal Hospital in 1899. Aware of the importance of specialization in surgery, he sent trainees to Europe to specialize in Pediatric and Orthopedic Surgery.

Ordinarius Professor Cemil Topuzlu, is recognised as the leading surgeon who brought modern concepts and new innovations in surgical practice and specialty training in the Ottoman Empire and modern Turkey.
HISTORY OF CONJOINED TWINNING

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**Introduction:** Conjoined twinning (CT) occurs in roughly 1:100,000 pregnancies and results from the incomplete division of the embryonal disk destined to yield monozygotic twins. It is a conspicuous malformation and therefore, when delivery is possible, diagnosis is immediate. For this reason, at issue with other malformations, CT is known from ancient times. The purpose of this presentation is to review documents about CT.

**Methods:** Bibliographic review and search of images from different sources.

Results: There are paintings and sculptures from the neolithic age to these days. Most documents come from central Turkey, Syria and Iraq but there are also some from Egypt, Mesoamerica, Africa and Europe. Among the more relevant, the first autopsy performed in America at the beginning of the 16th century on a set of twins and the history of the original Siamese twins, Chang and Eng Bunker will be shown.

**Conclusions:** The public attention given to CT exploded with the introduction of current social media and press and TV coverage. Many cases are publicized and the tough ethical and technical decisions involved are subjected to public scrutiny. These aspects will be discussed.
In 1990, the Lancet published an original article by Malone, Kiely and Ransley, ‘Preliminary report: the antegrade continence enema’. This reported the use of the appendix to produce a continent catheterisable colonic stoma to deliver an antegrade and complete enema. Five patients with faecal incontinence related to either a myelomeningocele or an ano-rectal anomaly were successfully managed. The technique was rapidly adopted by many surgeons and indications were intractable incontinence secondary to spina bifida, ARMs, Hirschsprung disease and idiopathic constipation. Several publications confirmed the benefits of this management for the majority of patients.

However, as early as 1890, Keetley had suggested using the appendix as a spout when emptying the caecum at open laparotomy. Weir in 1903 reported using the appendix as a conduit for irrigating the colon of a patient with ulcerative colitis, thus achieving a remission. For 40 years, an appendicostomy was recommended for this indication and was also briefly tried for patients with intractable constipation or abdominal distension (irritable bowel syndrome).

In the 1940’s, an ileostomy became the preferred diversion for colitic patients and appendicostomies completely fell out of clinical practice. Up till then, they had never been recommended for the management of faecal incontinence.

28 years after the Lancet paper, the use of the appendix stoma is being challenged by more effective and acceptable irrigations per rectum. Is ‘the wheel turning full circle’ again? The many editions of a standard English surgical textbook – Bailey and Love - chart the ‘ebb and flow’ of the appendicostomy in clinical practice.


Aim of study

The purpose of this study is to contribute to the standardization of treatment and to identify factors related to mortality of children with inhalation injury.

Methods:

We retrospectively evaluated patients with a diagnosis of inhalation injury in our pediatric burn intensive care unit between December 2009- February 2017. Clinical characteristics and / or bronchoscopic findings were used to recognize inhalation injury. Patients divided into two groups according to treatment protocol. Protocol A: nebulized adrenaline, albuterol. We defined protocol B as; Protocol A and nebulized Heparin - N-acetylcysteine (NAC) combination.

Main results:

Forty four children met the inclusion criteria. This study include 63,6% male with a mean age of 86,1 ± 56,4 months and a mean total burn surface area (% TBSA) of 38,4 ± 20,9%. Mechanical ventilation (MV) support was applied to 75% of patients. Of the 26 patients performed flexible bronchoscopy, 13 patients were classified as grade 1, 6 patients were grade 2, 6 patients were grade 3, and 1 patients were grade 4. Protocol B group needed MV support more than protocol A group (100%-%57,7 p=0,001). Extubated patients’ grades of inhalation injury were significantly lower than those of the intubated patients (grade 1: 100%- 61,5%, p=0,034). The average percentage of cutaneous burns in survivor patients is lower than non-survivors (34,5%- 51,6%, p=0,01)

Conclusion: Heparin-NAC therapy was preferred more frequently in entubated patients. High grade inhalation injury was associated with need of mechanical ventilation. Mortality was higher in patients with higher % TBSA burn.
Abstract

Introduction: Burns and scalds are among the most severe injuries in childhood. In spite of enormous efforts, their treatment too often leads to unsatisfactory results. Diagnostic uncertainty about the burn depth contributes significantly to the duration of treatment and to the amount of skin grafting. Thus, an improvement in diagnostic accuracy would be an important step towards better results.

Materials and Methods: Optical coherence tomography (OCT) is a new laser based technique for the examination of the skin with a depth of penetration of ca. 2 mm. In this pilot study, a new OCT technology is applied to children who sustained burns or scalds. It was the aim of the study to identify OCT changes associated with burns in order to improve diagnostic accuracy. Results: We observed epidermal loss, irregular skin surface, loss of dermal papillary pattern, loss of skin lines or disappearance or characteristic variations in the vascular pattern in burns and scalds of variable depth.

Conclusions Vascular changes were found in deeper burns requiring skin grafts. This is the first systematic study of OCT in the assessment of burn wounds in children. A number of new burn-associated skin changes were identified. Thus, OCT provided an “optical biopsy” of burn wounds that adds significant information.
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AIM OF THE STUDY

Nonoperative management (NOM) of blunt renal trauma (RT) in children is standard of care and is reliable and safe even in high grade injuries (grade IV-V). We aimed to evaluate the outcomes of NOM.

METHODS

Retrospective study (2002-2017) of children with RT treated at our institution. We analyzed epidemiology, diagnostic procedures, associated injuries, treatment and follow-up. All patients were initially managed nonoperatively (including angioembolization), regardless of injury grade. Failure of NOM was considered as the need for operative intervention within the first 72 hours.

MAIN RESULTS

We treated 60 patients with a mean age of 10.3 years (SD: 4.1). The most frequent injury mechanism was direct impact (26). Injury grades: I (20), II (12), III (11), IV (15) and V (2). Fifty-three (88.3%) had hematuria. Twenty-nine (48.3%) sustained associated injuries. In 32 (53.3%) physical exploration of the abdomen was normal. Six patients (10%) had initial hemodynamic instability.

All patients were managed with NOM; 5 (8.3%) required angioembolization and 1 (1.7%) stent insertion. NOM was successful in 59 (98.3%), one patient underwent urgent nephrectomy due to persistent bleeding after angioembolization. Complications of NOM appeared in 8 (13.3%): urinoma (2), pseudoaneurysm (1), lithiatis (2) and pleural effusion (3). Mean hospital stay was 9.8 days (SD:10.5). Mean follow-up was 91.38 months (SD:53.55).

CONCLUSIONS

NOM in children with RT is highly successful even in high-grade injuries and associated with a low complication rate. Angioembolization should be considered the treatment of choice in patients with RT and active bleeding.
Aim Numerous studies established a link between socioeconomic status (SES) and several dimensions of general health. This study examines the association between maternal SES and outcome in newborns requiring surgery for congenital anomalies. Results should provide the basis for future strategies to improve perinatal healthcare.

Methods Ambispective data analysis of newborns with isolated esophageal atresia (EA), intestinal atresia (IA), congenital diaphragmatic hernia (CDH), omphalocele (OC), gastroschisis (GS) undergoing surgery between 01/2008-11/2017 accessing the databases Neodat and Viewpoint. Maternal SES was determined according to the validated education classification CASMIN and graduated into “lower” or “higher”. Endpoints were number of prenatal screenings, incidence of postoperative complications (mortality, reoperation, sepsis, pooled minor complications), length of mechanical ventilation, and readmission to NICU. Ethical approval obtained.

Results Inclusion of 170 patients with EA (n=32), IA (n=25), CDH (n=47), OC (n=19), GS (n=47). Women of lower SES (n=71, 42%) attended fewer prenatal screenings (EA, 3.7 vs. 7.1, p=0.001; IA, 3.5 vs. 9.2, p=0.001; OC, 2.5 vs. 8.8, p=0.009; GS, 4.1 vs. 7.0, p=0.002). Lower SES was associated with higher incidence of postoperative complications (EA, 60% vs. 23%, p=0.041; IA, 67% vs. 20%, p=0.022; CDH, 83% vs. 46%, p=0.009; GS, 74% vs. 25%, p=0.002), and higher readmission-rate to NICU (IA, 0.33 vs. 0, p=0.042; GS, 0.37 vs. 0.04, p=0.008).

Conclusions Lower maternal SES is associated with a reduced uptake of prenatal screening and a higher incidence of postoperative complications in newborns with congenital anomalies. Specific support should be provided prenatally and perioperatively for families with low SES.
LONG-TERM RESULTS FOLLOWING INTRAARTICULAR FRACTURES OF THE MEDIAL MALLEOLUS IN CHILDREN AND ADOLESCENTS WITH SPECIAL EMPHASIS ON MRI

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Abstract

Aim of the Study: There is no publication combining clinical follow-up with MRI to determine possible posttraumatic alterations following pediatric intra-articular fractures of the medial malleolus. The aims of this study were to retrospectively analyze a cohort of patients with Salter-Harris III and IV fractures of the medial malleolus and to evaluate their long-term outcome. MRI was used to assess possible changes of the articular surface that cannot be diagnosed on radiographs.

Methods: Following ethical approval, 54 patients with Salter-Harris III (n=38) or IV fractures (n=17) of the medial malleolus treated between 2001 and 2011 were invited for a follow-up examination. Clinical outcome was assessed with the Weber score, osteoarthritis with the Kellgren and Lawrence classification. MRI were rated by the Outerbridge classification. Correlations between the clinical and radiological outcomes were calculated.

Main Results: 17 patients were recruited for MRI long-term follow-up at a mean of 112 (range, 65-184) months. The Weber score was very good for 5, good for 10 and poor for 2 patients; the Kellgren and Lawrence score revealed a favorable grade 0 in 15 and grade 1 in 2 patients. The MRI based Outerbridge classification yielded grade 0 for 12, grade 1 for 1, grade 2 for 2, and grade 3 for 1 patient. The Outerbridge score significantly (p<0.05) correlated with the Weber score.

Conclusions: This study shows excellent and good outcome of Salter-Harris III and IV fractures of the medial malleolus. Worse clinical outcome correlated with posttraumatic changes of the articular cartilage seen on MRI.
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Aim of the study

Preoperative information is of paramount importance. Unanswered questions or misunderstood information generate anxiety in parents. We decided to evaluate how various aspects of preoperative information were perceived by parents.

Method

We gave out 101 questionnaires containing the Amsterdam Preoperative Anxiety and Information Scale (APAIS), as well as various other questions pertaining to preoperative information. All procedures were performed as inpatient procedures, under general anaesthesia. All families agreed to answer the questionnaire.

Main results

Forty-four percent of parents demonstrate significant anxiety. All responders found that use of a drawing during consultation had made explanations clearer and 91% found it reassuring. Preoperative consultation with the anesthetist contributed to clarifying the information and reassured parents (85%). Twenty-eight percent of parents went to internet for further information but only half of these would recommend its uses to others. The degree of education of the parents did not impact the way the quality of information was perceived, nor did type of surgery. For 90% of patients, it is the relationship created during consultation which contributes to building trust in the surgeon, more than any other criteria. When asked which aspect of preoperative information could be improved, one in 5 parents answered that it was information concerning the practical aspects of the hospital stay such as clothing and eating.

Conclusion

Having the parents' input on what information they need is essential and contributes to the global shift towards patient or parent-centered outcome.
Abstract
Introduction: Injury is the leading cause of death in childhood being thoracic trauma (TT) the second death cause after traumatic brain injury. Thorax-CT is the most sensitive radiation-associated test in TT. We aim to analyse the diagnostic value of Chest-CT in TT.

Methods: Retrospective study was performed of all the patients with TT treated at our institution from 2005 to 2017. Demographic, clinical, radiological and surgical data were included for statistical analysis.

Results: A total of 131 patients were reviewed, being male 73%. Mean age and weight were 8±1y and 32±3Kg respectively, with a mean Injury Severity Score (ISS) of 9±2. Major traumatic mechanism was motor vehicle crash (46%). Chest x-ray was performed in 125 patients and CT-Scan in 24 patients whose mean ISS was 23±5. Predominant diagnoses were lung contusion and bone fractures. Isolated thoracic injuries accounted for 3.8%.

Chest-CT was performed in 12 patients with a previous abnormal x-ray and was decisive for management in 3. In the 6 patients with normal previous x-ray, 2 showed abnormal findings on CT and none of them required surgical treatment. Among the 6 patients without any previous x-ray, CT showed life-threatening injuries in 2 (traumatic diaphragmatic hernia and laceration of innominate vein).

Conclusion: Chest-CT improved accuracy of patient diagnosis but didn’t affect patient management with the exception of vascular lacerations, airway injury or diaphragmatic hernias. Spreading adoption of bronchoscopy and more tailored used of Chest-CT is needed in order to reduce radiation burden in TT patients.
Aim of the study

The use of 3D laparoscopic surgery in the pediatric population is still uncommon and few are the studies reported in vivo. This prospective study compares advantages and disadvantages of 3D and 2D endosurgery, in the treatment of congenital and acquired conditions in children. Both technologies were compared in terms of applicability and the effectiveness. Patients’ outcome and surgeons’ response were evaluated along with a hospital costs analysis.

Methods

Group A included all patients who underwent a 3D laparoscopic procedure from May 2016 to September 2017. Group B included patients undergoing standard laparoscopic 2D procedures during the same period. Demographics, operative report and postoperative parameters of each group were collected and compared. A laparoscopic optic evaluation questionnaire was filled out by the equipe at the end of each surgical procedure. Moreover, a cost assessment of the 3D and 2D procedures (operating room and surgical instrumentation costs) were evaluated.

Main results

3D laparoscopic procedure were 30 while the classical 2D surgeries were 22. Mean patients’ age was 82.8 months in Group A and 92.2 months in Group B. No differences were found between the two groups concerning preoperative and operative parameters as well as complication rates and length of hospital stay. No differences in operating room’s costs were found. Instead, cost of surgical instrumentation was significantly lower in Group A (p< 0.05).

Conclusions

In pediatric patients, 3D laparoscopic surgery while maintaining the benefits of MIS facilitates the surgeon’s performance and proves more cost effective than the 2D technology.
Aim of the Study: Angio-embolization (AE) is widely used in hemodynamically unstable adult patients with solid organ injury (SOI). It has limited use in pediatric patients. We present our experience of AE to control bleeding in patients with SOI.

Methods: AE applied trauma patients were evaluated in our clinic between 2012-2017, retrospectively. Information regarding patients’ demographics, type and mechanism of injury, injury severity, length of hospital stay, requirement for transfusion, complications and the results were presented.

Main Results: Eleven patients consist of 7 boys and 5 girls aging from 1 to 16 are taken in the study. AE is performed on two liver injury (grade IV), two splenic injury (grade IV) and seven renal injury (six, grade IV; one, grade V) patients. Three of these children had multiple SOI. However, AE was performed on the actively bleeding organ. The average time between patients’ admission and AE was 22 hours. Blood loss measured by the decrease in hematocrit values from admission to AE was 7.33± 5.3 %. Thrombocyte counts are found to be decreased by 129.8 ± 64.6x10³/µL between the admission and the AE.

Mean duration of intensive care unit stay and hospital stay were 4 and 12 days, respectively.

AE failed in one patient and was repeated successfully. There was no mortality.

Conclusions: AE is safe and effective treatment option for unstable patients with grade IV to grade V SOI. Bleeding control with AE saves patients from open surgery.
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Aim: Investigate the utility of urinary intestinal fatty acid binding protein (i-FABP) in infants (<6m) to predict intestinal ischaemia due to necrotizing enterocolitis (NEC) or other pathology.

Methods: Ethically approved (12/LO/1898), multicentre prospective study. Three groups were enrolled: (i) clinical diagnosis of NEC who underwent emergency laparotomy (NEC group); (ii) acute abdominal pathology of unknown pre-operative diagnosis necessitating emergency laparotomy (AA group); (C) healthy infants undergoing elective laparotomy or laparoscopy (Control). Pre-operative urine samples were collected. Urinary i-FABP/Creatinine was compared between groups and compared with presence or absence of intestinal ischaemia or necrosis. Data are median (IQR). Comparisons were made using Mann Whitney and Kruskal-Wallis tests. ROC analysis was used to determine the optimum test threshold.

RESULTS: Forty-two infants were enrolled: 16 NEC, 12 AA and 14 Control. i-FABP/Cr was highest in NEC group but there was considerable overlap (Figure). Infants with intestinal ischaemia or necrosis had higher i-FABP/Cr than those without ([53-10-80] vs 1.8[0.2-12]; p=0.0004). An i-FABP/Cr threshold of 46.4 pg/nmol detected intestinal ischaemia with a likelihood ratio of 8.5 and high specificity (94%) but low sensitivity (55%); AUC 0.85; p=0.0007). Using this threshold of i-FABP/Cr alone to guide surgical decision making would miss a significant proportion of infants with ischaemic bowel.

CONCLUSION: Urinary i-FABP appears an unreliable predictor of intestinal ischaemia in this setting since it may be raised in infants without ischaemia and values in infants with ischaemia vary widely.
Aim: Splenic pseudo-aneurisms are rare arteriovenous fistulas that may affect children after closed abdominal trauma. Splenic rupture or late hemorrhage are the most common complication leading to hemodynamic shock if unattended. In adults the management has been widely studied, but in children its incidence is unknown, and treatment is not well defined. The diagnostic tests include Doppler-ultrasound or AngioTC, however percutaneous arteriography is both diagnostic and therapeutic with selective embolization. Surgery is necessary if embolization fails or hemodynamic instability.

Methods: A five-year retrospective study of closed abdominal trauma with splenic involvement analyzing the development of splenic pseudo-aneurisms and its treatment.

Results: 32 patients with splenic trauma with a mean grade III (GI-IV) managed conservatively. In 9.3% (n=3) a splenic pseudo-aneurism was diagnosed with a mean diagnostic time of 6.5 days (5-8 days). The development of Splenic pseudoaneurism was not associated to other injured organs neither to the grade of spleen injury. Two cases underwent percutaneous embolization, one was successful and the other had post-embolization hemorrhage requiring urgent splenectomy. The third case was managed conservatively with spontaneous resolution after two months of strict imaging surveillance.

Conclusion: Splenic pseudo-aneurisms in pediatric population is probably underdiagnosed with a higher incidence than previously reported. We consider it is advisable to perform an image study at day 5-7 before discharging a patient with splenic trauma to discard pseudo-aneurisms. Treatment is still controversial, but we advocate for percutaneous embolization with USG-Doppler controls saving splenectomy if hemodynamically unstable.
SCX-G12
VASCULAR ANOMALIES: FROM PRENATAL EVALUATION TO PERINATAL MANAGEMENT

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Aim of the study

Vascular anomalies (VA) are grouped in vascular tumors and vascular malformations (VM). Although prenatal US identification is possible, VA remains rarely diagnosed during pregnancy. We reviewed a series of patients with VA identified during prenatal US discussing the perinatal management.

Methods

We collected data of patients with VA over a period of 8 years (January 2010–January 2018): prenatal evaluation (US and MRI), postnatal diagnosis and perinatal management (US, MRI at 2 months in complex forms, airways endoscopy in case of cervical and thoracic location).

Main results

We identified 15 cases of VA at a mean gestational age of 24 weeks [range 20-31].

- 3/15 complex lymphatic: 2/3 cases of PROS-A (one of them associated with multiple malformations that lead to voluntary termination of pregnancy at 22nd wog) and 1/3 case of Nonne-Milroy syndrome (voluntary termination of pregnancy at 20th wog).

- 11/15 common lymphatic: macrocystic (5), microcystic (3) and mixed (3), localized in the neck (6), thorax (2), thorax and abdomen (2) and axilla (1). 1/11 (cervical) recovered by the 23th wog and 2/11 (cervical) caused airways compression and required EXIT intubation; both babies breathed spontaneously after 2 days. One of them underwent sclerotherapy; the other one had recurrent infections, tracheostomy and surgical mass removal.

- 1/15 congenital hemangioma

Prenatal evaluation allowed the diagnosis in 83% of cases.

Conclusions

Lymphatic VM and congenital hemangioma are VA that can be identified prenatally. Complex forms with segmental hypertrophy require further evaluations, including the genetic one. The multidisciplinary approach is crucial for patient’s management, especially in the perinatal period.
USE OF THERAPEUTIC PLASMA EXCHANGE IN CHILDREN WITH THROMBOCYTOPENIA-ASSOCIATED MULTIPLE ORGAN FAILURE IN A PEDIATRIC BURN INTENSIVE CARE UNIT

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Aims of the Study:
Thrombocytopenia- associated multiple organ failure (TAMOF) has evolved to an accepted therapy for selected indications such as Therapeutic Plasma Exchange (TPE). The aim of this study is to describe effectiveness of TPE in pediatric burn cases.

Methods:
All TPE procedures performed at a children burn intensive care unit during a 2-year period (2016-2018) were retrospectively evaluated.

Main Results:
Eleven patients with a median age of 6.3 (1-16) years underwent a total of 33 TPE sessions. All patients were treated for burn. All procedures were performed using the filtration technique and heparin anticoagulation. Eight of the cases were male, three were female. Burn percentage was between 23% and 80% (mean 45.7%). Four of the patients were flammable and two were flame and inhalation burns. In other cases; two were hot water, one was milk and one was concentrated liquid (soup) burn. TPE was performed in an average of fifth day after TAMOF. Four (36.3%) patients showed full or partial recovery after TPE, seven had no response. Minor adverse events occurred in 4/33 (12.1%) procedures was reported.

Conclusions:
TAMOF is the most important cause of mortality in pediatric burn intensive care units. TPE is a safe apheresis method in children, even when performed as a long-term therapy. Efficacy is high under selected conditions. The TPE procedure is a promising treatment modality in addition to standard treatment in treatment of pediatric burn cases with TAMOF.
**SCX-G14**

**EXCISION OF PRENATALLY DIAGNOSED ENTERIC DUPLICATION CYSTS**

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*The Hospital for Sick Children, Toronto, Canada*

**Aim:** With improvements in prenatal ultrasound, more abdominal enteric duplication cysts (EDCs) are diagnosed prenatally. The ideal treatment approach remains unclear. The aim of this study was to review the evidence supporting the indication and timing of surgery for prenatally diagnosed EDCs. **Methods:** A systematic review was performed using defined search strategies identifying articles with inclusion criteria of prenatal diagnosis of histologically-confirmed EDCs. Outcomes included i) age at operation, ii) indications for surgery (symptomatic versus prophylactic). Data is reported as median (range) or mean±SD (where normally distributed). **Results:** Of 1444 articles screened, prenatal diagnosis of histologically-confirmed EDCs was made in 58 articles, detailing timing and indications for excision for 68 patients. Twenty nine patients (43%) became symptomatic early and were excised at a median age of 2 days (Table 1). Indications for intervention included bowel obstruction, volvulus, bleeding, recurrent pain and diagnostic dilemma (concern for cystic neuroblastoma). There were 39 patients (57%) who were resected prophylactically at a median age of 90 days without any interval symptoms. There was an increased proportion of ileal duplications in the symptomatic patients and an increased proportion of gastric duplications in the asymptomatic patients (Table 1). **Conclusions:** The optimal approach to management of EDCs diagnosed prenatally is not supported by strong evidence in the literature. Our systematic review indicates that almost half of the infants diagnosed prenatally become symptomatic early and are operated. In asymptomatic patients, close observation can be considered. However, it is debatable whether or when prophylactic excision is indicated.

| Table 1. Symptomatic and asymptomatic patients with prenatally diagnosed EDCs. |
|---------------------------------|-----------------------------|-----------------------------|-----------------------------|
| **Detected on prenatal US at week [median (range)]** |
| Symptomatic  | Asymptomatic |
| n=29  | n=39  | P=0.9  |
| 23w (16w–16w)  | 21w (16w–36w)  |
| **Gestational age in weeks [mean (±SD)]**  | 28.5w (±1.8w)  | 39w (±1.7w)  | P=0.3  |
| **M±F**  | M=14 : F = 8 (ND = 14)  | M=14 : F=11 (ND =11)  | P=0.5  |
| **Age at surgery in days [median (range)]**  | 2 (1–60)  | 90 (2–870)  | P=0.001 (Mann-Whitney)  |
| **Largest cyst dimension in cm [mean (±SD)]**  | 4.6cm (±2cm)  | 3.7cm (±1.5cm)  | P=0.1  |
| **Anatomic location**  |  |  |
| Gastric  = 2 (7%)  | Duodenal = 10 (26%)  |
| Jejunal  = 4 (14%)  | Duodenal = 7 (18%)  |
| Ileal  = 8 (29%)  | Jejunal = 0 (0%)  |
| Ileocecal  = 7 (25%)  | Ileal = 5 (13%)  |
| Colonic  = 2 (7%)  | Ileocecal = 10 (26%)  |
| Isolated retroperitoneal  = 2 (7%)  | Colonic = 2 (5%)  |
|  | Isolated retroperitoneal  = 5 (13%)  |
|  | P=0.03 (ANOVA)  |
Aim: Determine the incidence of non-accidental trauma (NAT) among children with major traumatic injuries attending the emergency department (ED) shockroom. Traumatic injury of children visiting the ED may have been caused by NAT i.e. physical abuse or traumatic injury due to neglect. Distinguishing NAT from accidental injuries is challenging.

Methods: Between 2010 – 2015 data from medical files were retrospectively collected of all children who attended the ED shockroom. Demographic data and information regarding; mechanism of injury, injury type, Injury Severity Score (ISS) and child abuse screening were collected. Children with suspected NAT were evaluated by the Suspected Child Abuse & Neglect Team (SCAN) to confirm or reject child abuse. An ISS of 16 or more was considered as major trauma.

Results: The cohort contains 421 children and 31 (7.4%) were evaluated by SCAN. In 10 (2.4%) NAT was confirmed. In total 81 (19.2%) had an ISS ≥ 16. The prevailing trauma mechanism in major trauma were traffic accidents (54.3%) and “fall from height” (24.7%). In major trauma NAT was confirmed in 2 (2.5%), 1 caused by physical abuse, 1 as a result of neglect and an accidental cause in 4. One case remained unclear. Nine children have died, but have not been evaluated by SCAN.

Conclusion: The incidence of non-accidental trauma in children attending the shockroom of the emergency department is 2.4% and the incidence among major trauma is 2.5%. However, the incidence of NAT in major trauma in our study is lower than previously published articles have reported (5 – 7%).
Differential Effects of Amniotic Fluid and Mesenchymal Stem Cells on Intestinal Epithelial Cells

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Aim: Stem cell therapy has been shown to be beneficial in intestinal injury such as necrotizing enterocolitis (NEC). Proteomic analysis indicated that amniotic fluid stem cells (AFSCs) play greater role in cell growth and development, while bone marrow derived mesenchymal stem cells (MSCs) are involved in immune regulation. The purpose of this study is to compare the effects of these stem cells on intestinal epithelial cells in normal and injury conditions.

Methods: Intestinal epithelial cells (IEC18) were exposed to lipopolysaccharide (LPS) to induce injury. They were co-cultured with AFSCs and MSCs using Transwell. Cell migration was measured by wound healing assay, cell proliferation was assessed by immuno-staining of Ki67 and gene expression of stem cell marker Lgr5 was analyzed by RT-PCR. Data was compared using one-way ANOVA with Bonferroni post-test.

Results: In normal condition, AFSCs but not MSCs had better effects on cell migration (Ai). Both AFSCs and MSCs showed similar improvement in wound healing in LPS-induced injury (Aii). AFSCs increased Ki67 in both normal and injury conditions (Bi-Bii) whereas MSCs upregulated proliferation only in the LPS-induced injury (Bii). Both AFSCs and MSCs increased stem cell Lgr5 after injury (Cii).

Conclusions: AFSCs improved growth in normal condition while both AFSCs and MSCs showed beneficial effects after injury. These findings indicate that AFSCs and MSCs have different mechanism of action on intestinal epithelial cells. These differential effects should be considered when planning stem cell therapy in NEC.
DECREASED LUNG-TO-LIVER-RATIO INDICATES A RELEVANT ROLE OF THE LIVER FOR THE PATHOGENESIS OF PULMONARY HYPOPOLASIA IN EXPERIMENTAL CONGENITAL DIAPHRAGMATIC

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Aim of the study: Prenatal liver position and lung-to-liver-signal-intensity-ratios (LLSIR) can be correlated with clinical outcome in congenital diaphragmatic hernia (CDH). Furthermore, it has been hypothesized that abnormal fetal liver growth might impair lung growth, leading to pulmonary hypoplasia (PH). However, the role of the liver during pathological lung development remains unclear. We aimed to study fetal lung and liver development and to evaluate pathomorphological changes in experimental CDH using micro-computed tomography (µCT).

Methods: Fetal rats (control, n=6) were harvested on embryonic-day 21 (ED21), processed, µ-CT-scanned, and analyzed to establish normal values for lung and liver volumes. Thereafter, fetuses (ED21, n=3) with nitrofen-induced CDH (administration ED11) were analyzed and pathomorphological changes were evaluated.

Main results: Normal values for lung and liver development were established by virtual dissection and reconstruction of control fetuses (Table). Nitrofen-exposure on ED11 resulted in right-sided CDH (Figure) with liver herniation into the chest. Both, lung and liver volumes were reduced in CDH fetuses compared to controls. The resulting lung-to-liver ratio was decreased by 33.3% in CDH fetuses demonstrating an impairment of lung growth in favor of liver growth in CDH, independent of the amount of intrathoracic liver.

Conclusions: µCT is an excellent tool to study prenatal features of nitrofen-induced CDH in fetal rats. Besides the high-resolution 3-dimensional visualization, µCT-imaging allows objective evaluation of prenatal anatomical changes in experimental CDH. A decreased lung-to-liver ratio indicates a relevant impairment of lung growth in favor of liver growth, suggesting a significant impact of the liver for the pathogenesis of PH in experimental CDH.
SCXI-BS03

ENDOGENOUS OMEGA-3 FATTY ACIDS PREVENT THE DEVELOPMENT OF EXPERIMENTAL NECROTIZING ENTEROCOLITIS INDEPENDENTLY OF PHYLUM LEVEL CHANGES IN GUT MICROBIOTA

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Aim: Changes in intestinal microbiome are associated with development of necrotizing enterocolitis (NEC). Previous reports indicate that dietary omega-3 PUFA induce changes in gut microbiota. The aim of this study was to investigate whether endogenously produced omega-3 PUFAs alter gut microbial composition and affect disease progression in experimental NEC.

Methods: Following ethical approval (#32238), an omega-6 PUFA enriched diet was fed to C57BL/6 mice with 2 genetic backgrounds: (i) Wild type (WT) and (ii) Heterozygous Fat-1. In Fat-1 mice, omega-6 PUFA are converted to omega-3 PUFA. From postnatal day 5 to 9, NEC was induced by hypoxia, gavage feeding of hyperosmolar formula, and lipopolysaccharide administration. The ileum was harvested on day 9 to evaluate the severity of mucosal injury (hematoxylin/eosin staining), inflammation (IL-6 by qPCR), and microbiota profile (16S rRNA sequencing).

Main Results: Mucosal injury and IL6 expression were significantly lower in Fat-1 NEC compared to WT NEC (Figure A,B). NEC induction was associated with a higher proportion of Firmicutes and lower proportion of Proteobacteria in both WT and Fat-1 controls (Figure C-E). However, there was no difference in bacterial composition between WT and Fat-1 NEC (Figure C-E).

Conclusions: Gut microbiome at the phylum level are not different between WT and Fat-1 NEC, even though the intestinal injury was less severe in the latter group. This finding suggests that endogenously produced omega-3 PUFA can prevent the development of NEC independently of the intestinal microbiome.
PREDICTIVE FACTORS OF POSTOPERATIVE COMPLICATIONS IN HIRSCHSPRUNG’S DISEASE: CHARACTERISATION OF THE INTESTINAL EPITHELIAL BARRIER AND THE ENTERIC NERVOUS SYSTEM

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Aim of the study: In Hirschsprung’s disease (HD), the postoperative course remains unpredictable, with enterocolitis occurring in one third and functional intestinal disorders, particularly obstructive symptoms (OS), in half. The aim was to identify predictive factors of postoperative complications by characterising the intestinal epithelial barrier (IEB) and the enteric nervous system (ENS) in the ‘healthy’ ganglionic zone in HD.

Methods: This multicenter translational study included full term HD neonates. The IEB assessment included following studies performed on resected colon during curative surgery: para/transcellular permeability, expression of tight junction (ZO-1, Occludin, Claudin1, JAMA, Cinguline) and Caveolin-1 proteins, mRNA expression of inflammatory markers (TNFa, IFNg, IL-8/10/1b, TLR-2/4/9). In addition, the ENS phenotype was characterized using an immunochemistry approach, with whole mounts of myenteric plexus stained with antibodies against calretinin, neuronal nitric oxide synthase (nNOS), and Hu (pan-neuronal marker). During the clinical prospective follow-up, postoperative complications were noted.

Main results: Eighteen HD were included (median age at surgery: 48 days). 8 of them presented postoperative enterocolitis (n=4), OS (n=4) and chronic liquid stools (n=4) (median follow-up: 21 months). Para and transcellular permeabilities were significantly higher in patients with postoperative enterocolitis (p=0.002 and p=0.001), nevertheless no statistical difference was found concerning tight junctions, caveolin-1 and inflammatory markers expression. Patients presenting OS were found to have a decreased number of nNOS neurons (p=0.1).

Conclusions: Studying IEB and ENS markers at time of curative surgery may help to predict postoperative complications, stratify HD patients and propose a personalised prophylactic treatment.
SCIENTIFIC SESSION XI: BASC SCIENCE  Saturday June 23rd

SCXI-BS05
EXPERIMENTAL AND HUMAN NECROTIZING ENTEROCOLITIS IS ASSOCIATED WITH IMPAIRMENT OF EPIGENETIC REGULATOR HMGA1

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Aim
HMGA1 is a nonhistone chromatin remodeling protein and architectural transcription factor. It is highly expressed in Lgr5+ intestinal stem cells (ISC) and is involved in upregulating genes for stem cell maintenance. Lgr5+ ISC impairment is associated with necrotizing enterocolitis (NEC). The aim of this study is to determine the role of HMGA1 in NEC.

Methods
Mice (AUP32238): NEC was induced in 5-day old C57BL/6 mice by gavage feeding hyperosmolar formula, hypoxia, and oral lipopolysaccharide (4mg/kg) for 4 days. P9 pups were sacrificed and ileum was harvested. Breastfed pups served as control.
Human neonates (REB1000056881): Intestinal ileal specimens were obtained from resections for active NEC and from control neonates undergoing resections for conditions other than active NEC in areas with no pathological abnormality. HMGA1, Ki67, and Lgr5 expression was assessed by immunohistochemistry staining and RT-qPCR.

Results
Compared to controls, both mice and humans with NEC exhibited a decrease in HMGA1 protein expression at the bottom of the crypts (Fig1). This was confirmed by gene expression analysis (Fig2). Ileum from mice and humans with NEC also had less proliferation (Ki67; control: 8.6±1.4; NEC: 1.955±1.075, p<0.001) and ISC activation (Lgr5; control: 1±0.32334; NEC: 0.15151±0.04903, p<0.05) than controls.

Conclusion
Our rodent and human studies indicate for the first time that HMGA1 is downregulated in NEC, which may account for the intestinal epithelial damage and impaired ISC proliferation. HMGA1 represents a novel target to enhance intestinal homeostasis in infants with NEC.
SCXI-BS06
TRANSPLANTED SOX10+VENUS MOUSE ENTERIC NEURAL CREST-DERIVED CELLS MIGRATED INTO AGANGLIONIC COLON IN THE HIRSCHSPRUNG’S MOUSE MODEL

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Aim: Failure of the enteric neural crest-derived cells (ENCC) of enteric nervous system (ENS) to colonize perfectly the embryonic gut results in Hirschsprung’s disease (HD). Cell based therapies have potential for the treatment of enteric neuropathies by generating enteric neurons in regions that entirely lack an ENS due to developmental defects such as HD. The Endothelin-B receptor (Ednrb) null mouse model has been widely used as a model for HD. Previously, we developed a Sox10 transgenic version of the Ednrb mouse to visualize ENCC with green fluorescent protein, known as the Venus mouse. The aim of the present study was to evaluate cellular migration into aganglionic gut using Sox10-Venus mice.

Methods: Fetal guts from Sox10-Venus+ mice were dissected on embryonic day 15.5 (E15.5) and cells were dissociated. These cells were cultured for 21 days under non-adherent conditions to generate neurospheres which were implanted into aganglionic region of E15.5 Ednrb−/− mouse hindgut explants on a membrane. After 4 days, the recipient gut was fixed and examined histochemically using confocal microscopy.

Results: By 5 days, cells had proliferated to form neurosphere-like aggregates that continued increase in size up to 14 days (Figure1). Four days after neurosphere transplantation, Sox10-Venus+ cells extended from the neurosphere within the aganglionic gut wall (Figure2).

Conclusion: These findings suggested that ENCC from the gut of Sox10-Venus+ mice can migrate into explants of recipient aganglionic bowel of Ednrb−/− mice. Thus, further study of cell therapy is needed to
determine whether they generate functioning neuronal networks in aganglionic gut.
SCXI-B507
TUMOR NECROSIS FACTOR-STIMULATED GENE 6 PROTEIN ATTENUATES THE INTESTINAL INJURY ASSOCIATED WITH ISCHEMIA-REPERFUSION

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Aim
Intravenous administration of amniotic fluid stem (AFS) cells protects the intestine from ischemia-reperfusion (IR) injury. However, it is difficult to administer stem cells during IR injury. Tumor necrosis factor-induced protein 6 (TSG6) is produced by stem cells and has anti-inflammatory effect. We hypothesized that intravenous administration of TSG-6 can reduce the intestinal injury related to IR.

Methods
With ethical approval, the superior mesenteric artery of 4-week old rats was occluded for 90 minutes (ischemia) and unclamped (reperfusion). After reperfusion phosphate buffered saline (PBS) \[n=9\] or green fluorescent protein (GFP) labeled AFS cells \[2\times10^6\] \[n=9\] were injected via tail vein. Sham rats underwent laparotomy with no IR or injection \[n=5\]. 48 hours after IR, rats were sacrificed and ileum harvested. TSG-6 mRNA expression and protein level were measured. Second experiment; TSG-6 was intravenously administrated to rats exposed to IR \[n=7\] \[Sham \(n=4\), and control PBS \(n=7\)\]. Inflammation (qPCR) and intestinal histology (hematoxylin/eosin) were measured. Data was compared using one-way ANOVA.

Results
Tsg-6 mRNA and protein were significantly overexpressed in animals receiving AFS cells compared to sham and PBS controls (Figure A). TSG-6 administration blunted intestinal inflammation (Figure B) and intestinal injury caused by IR (Figure C).

Conclusion
TSG-6 protein is produced in response to stem cells injection after IR injury. TSG-6 administration attenuates the intestinal injury associated with IR. TSG-6 is a potential novel therapeutic agent for conditions associated with IR such as midgut volvulus.
**SCXI-BS08**

**KNOWN AND NOVEL MICRORNAS CONTAINED IN AMNIOTIC FLUID STEM CELL EXOSOMES RESCUE PULMONARY HYPOPLASIA IN EXPERIMENTAL CONGENITAL DIAPHRAGMATIC HERNIA**

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**Aim of the Study:**

Exosomes secreted by amniotic fluid stem cells (AFSC) have a regenerative effect on hypoplastic lungs in experimental congenital diaphragmatic hernia (CDH). In this study, we aimed: 1) to investigate whether this effect could be replicated by exosomes from another source, mesenchymal stem cells (MSC); 2) to analyze whether AFSC and MSC contain microRNAs important for promoting lung growth.

**Methods:**

Exosomes were isolated by ultracentrifugation of AFSC and bone marrow-MSC conditioned medium and assessed for size using nanoparticle tracking.

**Ex vivo studies:** Rat fetal lungs were harvested at E14.5 from fetuses of dams that received nitrofen to induce pulmonary hypoplasia at E9.5. Explants were treated with medium alone, AFSC-exosomes, MSC-exosomes. Groups were compared for bud count and surface area. Fetal lungs from untreated dams served as control.

**small RNA-sequencing:** Exosome RNA was isolated using SeraMir, constructed into libraries (CleanTag Small RNA), and sequenced on NextSeq High Output single-end sequencing run.

**Main results:**

The improvement in lung growth parameters observed in hypoplastic lungs following treatment with AFSC-exosomes was not replicated by MSC-exosomes (**Fig. A**). We identified 127 microRNAs differentially expressed in AFSC-exosomes, some of which are known to have aberrant expression in CDH lungs (mir-33, mir-200c) and some that are novel (mir-743, mir-871; **Fig. B**).

**Conclusions:**

The microRNA cargo of AFSC-exosomes contains a differential profile that allows for improved regenerative potential for lung development and growth compared to MSC-exosomes. AFSC-exosomes may represent a better source of stem cells to promote lung growth in babies with CDH.
**A**

**Mean Surface Area**

**Lung Bud Count**

**B**

<table>
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<th>miRNA</th>
<th>AFSC-Exo alignments</th>
<th>MSC-Exo alignments</th>
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<td>rno-mir-465</td>
<td>2167</td>
<td>0</td>
<td>Infinity</td>
<td>?</td>
</tr>
<tr>
<td>rno-mir-881</td>
<td>2211</td>
<td>0</td>
<td>Infinity</td>
<td>?</td>
</tr>
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</table>
Aim of the study: The examination of volatile organic compounds (VOCs) has emerged as a promising area of cancer research. The aim of this study was to examine whether neuroblastoma causes alterations of fecal VOCs.

Methods: Following ethical approval, 10 Hsd:Fox1nu mice received 4 subperitoneal depots with 500,000 neuroblastoma cells each (MHH-NB11). 10 mice with inoculation of culture media served as shams. Following 9 weeks of tumor growth mice were euthanized and gas-chromatography-mass spectroscopy was used to measure VOCs in the supernatant space above isolated stool samples. VOCs were compared between the two groups. ROC analysis was performed and the area under the curve calculated.

Main Results: Eight animals developed a neuroblastoma with a mean weight of 3.9±2.3g. Neuroblastoma caused a significant reduction of total white adipose versus sham animals (0.02±0.01g vs. 0.04±0.02g, p=0.012). Out of 27 measured VOCs, three showed a different pattern between the groups. Benzaldehyde and 2-hexanone were significantly increased in the tumor animals (619±1105 vs. 78±62pbb, p=0.043 and 3.9±1.3 vs. 1.1±1.8pbb, p=0.005). In contrary, neuroblastoma caused a significant decrease of 3-methybutanal (201±139 vs. 434±350pbb, p=0.043). The area under the curve to predict presence of neuroblastoma was >0.8 for benzaldehyde, ethanol and 2-hexanone.

Conclusions: Neuroblastoma causes significant alterations of the fecal volatilome. The possibility of VOCs to serve as biomarkers for diagnosis, patient response and/or monitoring disease recurrence has to be examined in future studies.
SCXI-BS10
COMBINED GENETIC EFFECTS OF RET, NRG1 AND SEMA3 SUSCEPTIBILITY VARIANTS ON MULTIFACTORIAL HIRSCHSPRUNG DISEASE IN INDONESIA

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Abstract

Aim of the Study: Specific genetic variants at RET (rs2435357), NRG1 (rs7835688, rs16879552), and SEMA3C/D (rs1583147, rs12707682, rs11766001) are associated with Hirschsprung disease (HSCR) in Indonesia. This study aimed to investigate the additional effect of RET rs2506030 to determine its potential interactions with other known susceptibility polymorphisms of HSCR in Indonesian.

Methods: Sixty HSCR patients and 122 non-HSCR controls were ascertained for this study and genotyped using the TaqMan assay.

Main Results: RET rs2506030 was marginally associated with HSCR both by case-control analysis (OR=1.68; P=0.043) and the transmission disequilibrium test (TDT) (P=0.034). Furthermore, individuals with 5 or 6 risk alleles at RET rs2506030, rs2435357, and NRG1 rs7835688 showed ~23-fold higher HSCR risk than those with 0 or 1 risk allele.

Conclusion: Disease risk of HSCR is increased by the combination of specific RET and NRG1 susceptibility variants.
**Aim of the Study:** Pulmonary hypertension (PH) is an important contributor of morbidity and mortality in infants with congenital diaphragmatic hernia (CDH). It is well known that nitric oxide (NO), generated by endothelial NO synthase (eNOS), is a major endogenous regulator of vascular tone and plays a key role in pulmonary vasodilatation. Recently, a novel protein, nitric oxide synthase interacting protein (NOSIP) has been described, which reduces the production of NO by interacting with eNOS. We designed this study to investigate the pulmonary vascular expression of NOSIP in the nitrofen-induced CDH.

**Methods:** Following ethical approval (REC1103), time-mated Sprague-Dawley rats received nitrofen or vehicle on gestational day 9 (D9). Fetuses were sacrificed on D21 and lung specimens divided into CDH and control (n=6 for each group). Quantitative RT-PCR and western blotting were performed to analyze pulmonary gene and protein expression of NOSIP. Immunofluorescence-double-staining for NOSIP was combined with a specific SMC marker to evaluate protein expression in the pulmonary vasculature.

**Main Results:** Relative mRNA and protein expression of NOSIP was significantly decreased in CDH lungs compared to controls (Figure 1). Confocal-laser-scanning-microscopy revealed markedly diminished NOSIP immunofluorescence in CDH lungs compared to controls, mainly in the muscular and endothelial components of the pulmonary vasculature (Figure 2).

**Conclusion:** This study demonstrates for the first time decreased expression of NOSIP in the pulmonary vasculature of the nitrofen-induced CDH. We speculate that NOSIP underexpression interferes with NO production, impairing vascular remodeling resulting in PH.
SCXI-BS12

EPHRIN-B1, -B2 AND -B4 EXPRESSION IS DECREASED IN DEVELOPING DIAPHRAGMS AND LUNGS OF FETAL RATS WITH NITROFEN-INDUCED CONGENITAL DIAPHRAGMATIC HERNIA

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National Children’s Research Centre, Dublin, Ireland

Aim of the Study:
Congenital diaphragmatic hernia (CDH) is assumed to originate from a malformation of the amuscular mesenchymal component of the primordial diaphragm. Mutations in ephrin-B1, a membrane protein that is expressed by mesenchymal cells, have been found in humans with CDH and pulmonary hypoplasia (PH), highlighting its important role during diaphragmatic and airway development. Ephrin-B1, -B2 and -B4 are expressed in fetal rat lungs and have been identified as key players during lung branching morphogenesis. We hypothesized that diaphragmatic and pulmonary expression of ephrin-B1, -B2 and -B4 is decreased in the nitrofen-induced CDH model.

Methods:
Time-mated rats received nitrofen or vehicle on day 9 (D9) (ethics:REC668b). Fetal diaphragms (n=72) and lungs (n=72) were harvested on D13, D15 and D18, and divided into control and nitrofen-exposed specimens. Ephrin-B1, -B2 and -B4 gene expression was analyzed by qRT-PCR. Immunofluorescence-double-staining for ephrin-B1, -B2 and -B4 was combined with mesenchymal and epithelial markers (Gata4/Fgf10 and CGRP) to evaluate protein expression/localization.

Main Results:
Ephrin-B1, -B2 and -B4 gene expression was significantly reduced in pleuroperitoneal folds/primordial lungs (D13), developing diaphragms/lungs (D15), and fully muscularized diaphragms/differentiated lungs (D18) of nitrofen-exposed fetuses compared to controls (Tables). Confocal-laser-scanning-microscopy demonstrated markedly diminished ephrin-B1 immunofluorescence in diaphragmatic and pulmonary mesenchyme of nitrofen-exposed fetuses on D13, D15, and D18 compared to controls, whereas ephrin-B2 and -B4 expression was mainly decreased in airway epithelium (Figure1a-f).
Conclusions:
Decreased ephrin-B1, B2 and B4 expression may disrupt diaphragmatic development and lung branching morphogenesis by interfering with epithelial-mesenchymal interactions, causing diaphragmatic defects and PH.
SCXI-BS13
NECROTIZING ENTEROCOLITIS INDUCES DETRIMENTAL EFFECTS ON THE CEREBRAL DISTRIBUTION OF MATURE NEURONS AND OLIGODENDROCYTES

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1Developmental and Stem Cell Biology Program, PGCRL, The Hospital for Sick Children, Toronto, Canada.
2Division of General and Thoracic Surgery, The Hospital for Sick Children, Toronto, Canada.
3Translational Medicine Program, PGCRL, The Hospital for Sick Children, Toronto, Canada.

Aim of the Study: The pathogenesis of neurodevelopmental delay in neonates with necrotizing enterocolitis (NEC) remains poorly understood. We previously reported that experimental NEC induces neuroinflammation by activating microglia and astrocytes in the cortex and hippocampus. The aim of the present study was to investigate if neurons, the principal brain cell population, and oligodendrocytes, neuroglial cells responsible for white matter development, were also affected by experimental NEC.

Methods: NEC was induced in 5-day old neonatal mice using hypoxia, gavage feeding with hyperosmolar formula and oral lipopolysaccharide (4mg/kg) (protocol n.32238). The presence of NEC in the intestine was confirmed at histology by two blinded investigators. Breastfed pups served as control. NEC pups and control were blindly compared for the number of mature neurons (NeuN+) and oligodendrocytes (Olig2+) in the cerebral cortex, ganglia/thalamus, and hippocampus.

Main results: Compared to control, NEC pups had fewer mature neurons in the hippocampus (p=0.0001), but similar distribution of mature neurons in other regions. The brain of NEC pups also had decreased numbers of oligodendrocytes in the cortex compared to control (p=0.0001), but similar representation in the hippocampus (p=0.12) and ganglia/thalamus (p=0.79) (Fig A-B).

Conclusions: This study shows that the number of mature neurons and oligodendrocytes in the brain of pups with NEC is depleted in regions of the brain that control cognitive function and memory/learning. Further studies are needed to identify potential strategies to prevent the neurodevelopmental delay that occurs in infants with NEC.
RNA CARGO OF AMNIOTIC FLUID STEM CELL EXOSOMES IS ESSENTIAL FOR IMPROVING PULMONARY HYPOPLASIA IN EXPERIMENTAL CONGENITAL DIAPHRAGMATIC HERNIA

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2Division of General and Thoracic Surgery, The Hospital for Sick Children, Toronto, Canada.
3Translational Medicine Program, PGCRL, The Hospital for Sick Children, Toronto, Canada

Aim:
It has been reported that Amniotic Fluid Stem Cells (AFSC) promote lung growth in experimental congenital diaphragmatic hernia (CDH), through an undefined paracrine mechanism. Exosomes are extracellular vesicles, that mediate cellular paracrine effects. Herein, we investigated if the beneficial effect of AFSC was due to AFSC-exosome RNA transfer.

Methods:
AFSC-exosomes were isolated from conditioned medium using ultracentrifugation. To investigate the role of RNA, AFSC-exosomes were treated with RNase-A (100μg/mL) that degraded AFSC-exosome RNA. 

In vitro and ex vivo studies: E14.5 primary lung epithelial cells (EpCAM+) were isolated from fetuses of dams that received nitrofen to induce pulmonary hypoplasia at E9.5. Cells were treated with medium alone, AFSC-exosomes, or RNase-A pre-treated AFSC-exosomes (AFSC-ExoR). Groups were compared for proliferation and apoptosis rates. Fetal lung explants were harvested at the same time points, grown for 72h with same conditions, and assessed for bud count and surface area. Fetal lungs and epithelial cells from untreated dams served as control.

RNA sequencing: AFSC-exosome RNA was isolated, constructed into libraries and sequenced. Gene-set enrichment analysis was performed.
Statistics: one-way ANOVA (Tukey post-test).

Results:
AFSC-exosome treatment on nitrofen-exposed epithelial cells has pro-proliferative and anti-apoptotic effects, which are ablated with treatment by AFSC-ExoR (Fig.A). Measures of lung growth are not rescued in hypoplastic lungs treated with AFSC-ExoR (Fig.B). AFSC-exosomes show abundant expression of 296 mRNAs enriching for functions in Fig.C.

Conclusions:
RNA transfer of cargo contained inside AFSC-exosomes plays a crucial role in their beneficial effect on different models of pulmonary hypoplasia secondary to CDH.
PERI-OPERATIVE CARE IN RECONSTRUCTIVE SURGERY FOR RECTOVESTIBULAR FISTULA WITHIN THE ARM-NET CONSORTIUM: DOES IT INFLUENCE COMPLICATIONS AND SHORT-TERM COLORECTAL FUNCTION?

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¹Radboudumc Amalia Children’s Hospital, Nijmegen, Netherlands. ²Radboud University Medical Center, Nijmegen, Netherlands. ³Emma Children’s Hospital, AMC and VU University Medical Center, Amsterdam, Netherlands

Background/Purpose
Peri-operative care in patients with an anorectal malformation (ARM) type rectovestibular fistula is managed differently amongst centers of the ARM-Net consortium. We aimed to evaluate the impact of preoperative bowel preparation, perioperative antibiotics, and postoperative (par)enteral feeding on complications after reconstructive surgery and colorectal function at 1 year follow up.

Methods
A retrospective cohort study in ARM patients with a rectovestibular fistula, extracted from the ARM-Net consortium database, was performed. Data concerning preoperative bowel preparation, perioperative antibiotic use divided into prophylaxis (max. 24 hours postoperatively) and therapeutic use (>24 hours postoperatively), postoperative (par)enteral feeding, complications after reconstruction, and constipation 1 year postoperatively were retrieved from medical files. Univariate analyses were performed.

Results
In total, 169 patients were included in the study. Some kind of preoperative bowel preparation was given in 64% of the patients. Antibiotics were administered as prophylaxis in 33% whereas the majority (67%) received it on a therapeutic basis. Fifty patients (30%) were allowed to receive normal enteral feeds immediately after surgery, whereas 52% received total parenteral nutrition postoperatively. Overall complications were encountered in 29% of the patients, ranging from wound infection, to dehiscence and stenosis. No significant differences were found between bowel preparation, antibiotic use, and type of postoperative feeding and postoperative complications. Half of the patients experienced constipation 1 year after reconstruction.

Conclusion
Within the ARM-Net consortium postoperative complications and short term colorectal function were not significantly influenced by differences in peri-operative care in patients with an ARM-type rectovestibular fistula.
Aim of the study: To evaluate our short-term outcome of patients with short bowel syndrome-intestinal failure (SBS-IF).

Methods: Hospital records of patients with SBS-IF who had undergone surgical interventions between January 2013 and December 2017 in our center were retrospectively reviewed. Demographics, diagnosis, clinical features, nutritional status and need for hospitalization were evaluated.

Main results: A total 27 patients (16 boys, 11 girls) with a mean age of 29.7±47.1 months were included. The two major underlying conditions were necrotizing enterocolitis in 12 (46.2%) and volvulus in 6 (22.2%). Two patients had congenital SBS-IF. There were 17 (63%) patients with <50% small bowel length and 9 (33.3%) patients with <50% colon length. Half of the patients lacked ileocecal valves. Majority of the patients (77.8%) were referred from outer centers, all required parenteral fluid resuscitation and 85.2% were full total parenteral nutrition (TPN) dependent. The most common surgical interventions were; 10 stoma closures, 3 stoma creations, 1 elastic loop intestinal kinking, 1 stoma tightening, 4 tapered anastomosis, 8 LILT and 1 jejuno-colic patch procedures. Ten (37%) patients died whereas the most common reason was sepsis (n=6). There were 5 (18.6%) in-stay TPN-dependent patients and 2 (7.4%) intermittent TPN-receivers. TPN treatment was ceased in 10 (37%) patients.

Conclusions: Sepsis is the leading reason for death in SBS-IF patients. TPN dependence may be decreased with multi-disciplinary patient-specific management and bowel adaptation procedures.
Aim of the study:
Ostomy construction is considered a relatively safe alternative in neonates with acute abdomen. However, ostomy closure carries its own risks. We aimed to determine morbidity and mortality after ostomy closure and searched for risk factors for adverse outcome.

Methods:
Retrospective study including infants, in whom an ostomy was constructed in the first three months of life, who undergoing ostomy closure in a tertiary referral center in the Netherlands between 2006 to 2016. Primary outcome measurements were complications and mortality directly related to ostomy closure. Severity of complications was graded according to Clavien-Dindo. We compared patients with and without complications regarding patient and disease characteristics.

Main results:
85 patients were included, 57 (67.0%) boys. Indications for ostomy construction included anorectal malformation (40.0%) and necrotizing enterocolitis (30.6%). Mean (SD) age at closure was 206 (103) days. Postoperatively 18 patients (21.1%) developed 23 complications (table 1), four patients >1 complication. Patients with complications had lower gestational age (228 versus 250 days; p=0.04) and birth weight (2043 versus 2561 gram; p=0.04). One patient died after seven months because of comorbidity. Closure of small bowel ostomy and colostomy (12.9% versus 8.2%; p=0.37), and also weight (6397 versus 6520 gram; p=0.85) and gestational age (450 versus 458 days; p=0.80) at time of closure were not significant.

Conclusions:
20% of patients develop complications after ostomy closure, 9% need reoperation. Predictive factors are lower gestational age and birth weight at primary surgery, not so much the patient characteristics at the time of ostomy closure.

Table 1. Complications of ostomy closure sorted by Clavien-Dindo classification

<table>
<thead>
<tr>
<th>Grade</th>
<th>Complication</th>
<th>No. of complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Superficial wound infection</td>
<td>6 (7.1%)</td>
</tr>
<tr>
<td></td>
<td>Paralytic ileus</td>
<td>2 (2.4%)</td>
</tr>
<tr>
<td></td>
<td>Anaplastis</td>
<td>1 (1.2%)</td>
</tr>
<tr>
<td></td>
<td>Gastropares</td>
<td>1 (1.2%)</td>
</tr>
<tr>
<td></td>
<td>Central line infection</td>
<td>2 (2.4%)</td>
</tr>
<tr>
<td></td>
<td>Brontocardia</td>
<td>1 (1.2%)</td>
</tr>
<tr>
<td></td>
<td>Pneumonia</td>
<td>1 (1.2%)</td>
</tr>
<tr>
<td></td>
<td>Upper respiratory tract infection</td>
<td>1 (1.2%)</td>
</tr>
<tr>
<td>II</td>
<td>Anastomotic leakage</td>
<td>3 (3.5%)</td>
</tr>
<tr>
<td></td>
<td>Intraluminal abscess</td>
<td>2 (2.4%)</td>
</tr>
<tr>
<td></td>
<td>Anastomotic stenosis</td>
<td>1 (1.2%)</td>
</tr>
<tr>
<td></td>
<td>Intestinal perforation</td>
<td>1 (1.2%)</td>
</tr>
<tr>
<td></td>
<td>Intraluminal bleeding</td>
<td>1 (1.2%)</td>
</tr>
</tbody>
</table>

1 Grade I: Any deviation from the normal postoperative course without the need for pharmacological treatment or surgical, endoscopic, and radiological interventions.
2 Grade II: Requiring pharmacological treatment with drugs other than such allowed for grade I complications.
3 Grade III: Intervention under general anesthesia.
PW17-LG04
ANORECTAL MANOMETRY MAY REDUCE THE NUMBER OF RECTAL SUCTION BIOPSY PROCEDURES NEEDED TO DIAGNOSE HIRSCHSPRUNG’S DISEASE

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Aim of the Study: To evaluate whether anorectal manometry (ARM), used to test the rectoanal inhibitory reflex (RAIR), is a safe alternative for reducing the number of invasive rectal suction biopsy (RSB) procedures needed to diagnose Hirschsprung’s disease (HD).

Methods: Between 2010 and 2017, we prospectively collected the ARM results of 105 patients suspected of having HD. Following the outcome, the patients either underwent RSB to confirm HD or they were treated conservatively. Primary ARM-based diagnoses were compared with the definitive diagnoses based on the pathology reports and/or clinical follow-ups. Additionally, we analyzed whether modifications to our ARM protocol improved diagnostic accuracy.

Main results: The sensitivity of ARM and RSB was comparable (97% versus 97%). The specificity of ARM, performed according to our initial protocol, was significantly lower than that of RSB. After we modified the protocol the difference between the specificity of ARM and RSB was no longer statistically significant (74% versus 84%, respectively, P = .260). The negative predictive value of ARM was 100%, while their positive predictive value was significantly lower than that of RSB (56% versus 97%, P < .001).

Conclusions: ARM is a viable screening tool for HD and, provided it is performed properly, it can be used to exclude HD with absolute certainty once the RAIR is observed. By contrast, an absent RAIR on ARM should always be followed by a RSB to confirm the diagnosis of HD. Using ARM as the diagnostic of first choice could reduce the number of invasive biopsies.
Aim of the study
Anastomotic ulcer (AU) is an uncommon complication after bowel resection. It is mainly reported in small bowel to colon anastomosis, usually in short bowel syndrome. Etiology is unknown. Presentation is refractory anemia but it can also appear as an acute life-threatening rectal bleeding. Treatment is not well established, with high recurrence, even with surgical resection. Aim: to present our experience with AU.

Methods
Records of patients with AU at our institution since 2006 were retrospectively reviewed.

Main results
Four patients were identified. Extensive intestinal resection and small bowel to colon anastomosis was performed in three cases. Underlying diseases were gastroschisis, necrotizing enterocolitis and total colonic aganglionosis. Another patient had a sigmoid volvulus: sigmoidectomy with colorectal anastomosis. Clinical presentation was iron deficiency anemia in two patients and rectal bleeding in the other two (one needing resuscitation). Ileocolonoscopy confirmed AU in all cases (Figures). Medical treatment with iron, antacids and antibiotics was effective in one case. Two patients received blood transfusion. Endoscopic coagulation was useful in three cases. None of the patients underwent surgical resection. After a mean follow-up of 42.5 months (17-167), two patients are free of symptoms, one is under iron supplementation and another presents sporadically rectal bleeding.

Conclusions
Severe bowel resection, especially when ileocecal valve is involved, may lead to AU. It shoud be suspected when chronic anemia or rectal bleeding appears in these patients. Management is not defined, being medical treatment and endoscopic fulguration the first step. Surgery should be restricted to non-responding cases.
AIM OF THE STUDY
To present our variant of transumbilical laparoscopic-assisted appendectomy and to compare it with three-port conventional laparoscopic appendectomy (CLA) in terms of surgical time, postoperative complications and economic cost.

METHODS
This technique provides access to the abdominal cavity through an umbilical incision without the use of trocars (insertion of a probe for pneumoperitoneum and a laparoscopic camera and forceps through a single 8-mm incision), followed by an extracorporeal appendectomy.

We performed a retrospective review of patients with diagnosis of phlegmonous appendicitis who underwent surgery at our center (public tertiary referral hospital) over the last year, comparing the results of our trocar-free transumbilical appendectomy (TFTA) and CLA.

MAIN RESULTS
148 patients were analized. In 30(42.9%) of them TFTA was practiced and in the remaining 118(57.1%) CLA was performed. Both groups were comparable in their demographic and clinical features. In 4 of the TFTA cases (13.3%), introduction of 2 trocars for appendicular adhesiolysis was required. Operative time for TFTA (47 minutes, 31.6-63.6) was significantly lower than for CLA (70.4 minutes, 44.9-95.9) (p = 0.01). There were no differences in postoperative complications (abscess, wound infection, intestinal occlusion) between both groups (0 events in the ALST group and 3 in the ALC group, p = 0.82). Each TFTA represented an average saving of 270.73(213.31-328.15) euros in surgical material when compared to CLA.

CONCLUSIONS
For selected cases of uncomplicated appendicitis, TFTA represents a safe option when compared to conventional laparoscopic appendectomy, associating a shorter operative time and a lower economic cost.
PW17-LG07
IMMATURITY OF THE RECTOANAL INHIBITORY REFLEX AS A CAUSE FOR CONSTIPATION IN INFANTS

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Aim of the Study: The rectoanal inhibitory reflex (RAIR) plays an important role in the defecation process and is responsible for the relaxation of the internal anal sphincter upon rectal dilatation. Based on literature, we hypothesized that the RAIR may not always have reached full maturity at birth. In addition, we hypothesized that immaturity of the RAIR may play a role in symptoms associated with constipation in a subgroup of patients.

Methods: Between 2011 and 2017 we prospectively gathered data on newborns who presented themselves with severe constipation to our tertiary center, who had an absent or immature RAIR, as measured with anorectal manometry (ARM), and who underwent at least one follow-up ARM measurement. Maturity of the RAIR was based on the changes in sphincter pressure following rectal dilatation. Patients with an organic cause for constipation were excluded (e.g. Hirschsprung’s disease, congenital anorectal malformation).

Main results: A total of nine patients were included, all males. None of the patients had a fully matured RAIR on the initial measurement, all patients required high dose laxatives, and six out of nine patients required daily rectal washouts. At follow-up measurement, eight out of nine patients had developed a mature RAIR, eight patients’ parents reported decreased severity of symptoms associated with constipation, whereas only two patients still required rectal washouts.

Conclusion: Severe constipation in infants and newborn can be caused by a dysfunctional and immature RAIR, which may be able to further mature after birth. Correspondingly, constipation complaints may decrease as the RAIR matures.
PW17-LG08
RULING OUT APPENDICITIS IN CHILDREN: CAN WE USE CLINICAL PREDICTION RULES?

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1VU medical centre, Amsterdam, Netherlands. 2Academic Medical Centre, Amsterdam, Netherlands. 3Red Cross Hospital, Beverwijk, Netherlands

Aim of the study: To identify available Clinical Prediction Rules (CPRs) and investigate their ability to rule out appendicitis in children presenting with abdominal pain at the emergency department.

Methods: A literature search was conducted to identify potential CPRs. These CPRs were subsequently tested in a historical cohort from a general teaching hospital, consisting of all children (<18 years) that visited the emergency department between January 2012 and December 2014 with abdominal pain (identified through ICD-codes). Data were extracted from the electronic patient files and scores of the identified CPRs were calculated for each patient. Negative likelihood ratios with 95% confidence intervals were calculated for those CPRs that could be calculated for at least 50% of patients.

Main results: 12 CPRs were tested in a cohort of 291 patients, of whom 87 (29.9%) suffered from acute appendicitis. The Fenyö-, Modified Lindberg- and Raja Isteri Penigran Anak Saleha Appendicitis score could not be calculated for at least 50% of patients and thus these CPRs were excluded. Outcomes of the remaining 9 CPRs are displayed in Table 1. The Ohmann score, Alvarado score, Modified Alvarado Score, Pediatric Appendicitis Score, Low Risk Appendicitis Rule Refinement, Christian score and Low Risk Appendicitis Rule had a negative likelihood ratio point estimate <0.1.

Conclusion: This study identified seven CPRs that could reliably rule out appendicitis in the pediatric population. Prospective studies are needed to investigate if clinical monitoring and re-evaluation is a safe treatment strategy in case of low suspicion of appendicitis according to these CPRs.

<table>
<thead>
<tr>
<th>Table 1. Accuracy statistics</th>
<th>Negative likelihood ratio</th>
<th>Sensitivity</th>
<th>Negative predictive value</th>
</tr>
</thead>
<tbody>
<tr>
<td>LRAR (n=262)</td>
<td>0.09 (0.04-0.25)</td>
<td>95.3 (87.9-98.5)</td>
<td>95.6 (88.5-98.6)</td>
</tr>
<tr>
<td>LRARR (n=260)</td>
<td>0.07 (0.02-0.23)</td>
<td>96.5 (89.4-99.1)</td>
<td>96.3 (89.3-99.1)</td>
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<tr>
<td>Christian (n=242)</td>
<td>0.08 (0.00-0.22)</td>
<td>94.8 (86.5-98.3)</td>
<td>96.3 (90.2-98.1)</td>
</tr>
<tr>
<td>Lintula (n=219)</td>
<td>0.50 (0.38-0.65)</td>
<td>54.0 (41.0-66.4)</td>
<td>83.2 (76.8-88.3)</td>
</tr>
<tr>
<td>MASS (n=206)</td>
<td>0.30 (0.20-0.46)</td>
<td>73.0 (60.1-81.1)</td>
<td>88.2 (81.5-92.8)</td>
</tr>
<tr>
<td>Alvarado (n=203)</td>
<td>0.03 (0.00-0.20)</td>
<td>98.4 (90.0-99.9)</td>
<td>98.8 (92.5-99.9)</td>
</tr>
<tr>
<td>MAS (n=199)</td>
<td>0.03 (0.00-0.23)</td>
<td>98.4 (90.5-99.9)</td>
<td>96.3 (90.9-99.9)</td>
</tr>
<tr>
<td>PAS (n=199)</td>
<td>0.07 (0.00-0.22)</td>
<td>95.3 (86.0-98.8)</td>
<td>96.7 (90.0-99.1)</td>
</tr>
<tr>
<td>Ohmann (n=148)</td>
<td>0 (100)</td>
<td>100 (90.2-100.0)</td>
<td>100 (88.6-100.0)</td>
</tr>
</tbody>
</table>

Data is displayed as value (95% CI)
*Data is displayed as percentage (95% CI)
LRAR: Low Risk Appendicitis Rule
LRARR: Low Risk Appendicitis Rule Refinement
MASS: Modified Alvarado Scoring System
MAS: Modified Alvarado Score by Shen
PAS: Pediatric Appendicitis Score
PW17-LG09
PREDICTING THE ODDS OF HIRSCHSPRUNG DISEASE IN PATIENTS REFERRED TO PEDIATRIC SURGERY

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Aim of the study
The purpose of this study was to determine the incidence of Hirschsprung disease (HD) in all patients referred to pediatric surgery with symptoms suggestive for HD. This information will be useful when advising parents on the potential utility of a rectal biopsy to diagnose HD.

Methods
Referrals to a pediatric surgery service from 2006 to 2017 were screened for complaints of constipation, abdominal distension, failure to thrive, and failure to pass meconium within 48 hours of birth. Data collected included demographics, age of surgical assessment, age at onset of symptoms, and results of contrast study or biopsy. The incidence of a biopsy positive for HD was calculated for the entire cohort and for sub-groups based on symptoms, age at the time of assessment and age at onset of symptoms. Chi-squared tests were used to compare incidences of HD.

Main results
Of the 355 patients referred for surgical assessment, 13.5% had biopsy proven HD; the incidence was 49.2% for patients assessed in the neonatal period (n=63), 10.7% in infancy (n=75) and 4.3% in childhood (n=209). These rates were significantly different (p<0.05) from each other. For children in whom symptoms reportedly started after their first birthday (n=47) the likelihood of HD was 2.1%.

Conclusions
Although the incidence of HD in all children referred to pediatric surgery is 13.5%, a rectal biopsy for patients presenting after their first birthday is unlikely to prove positive for HD, particularly if symptoms reportedly started beyond infancy.
THE ALTERATION OF THIOL/ DISULPHIDE HEMOSTASIS AND BILIRUBIN LEVELS IN CHILDREN WITH APPENDICITIS

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Introduction: A prospective study was planned to find out if there is an alteration in thiol/disulfide homeostasis and bilirubin levels in children with both acute(AA) and complicated appendicitis(CA)

Patients and the Method: 82 children consisted of 25 children with AA, 27 children with CA and 30 healthy children were included in the study. WBC, CRP, total(TB) and direct bilirubin(DB), disulfide, serum natural thiol, total thiol values disulfide/serum natural thiol, disulfide /serum total thiol and serum natural thiol/total thiol ratios were determined and compared between groups

Results: The mean disulfide/serum total thiol ratio was significantly higher in both appendicitis groups. The difference between acute and complicated appendicitis groups regarding parameters of Thiol/disulfide hemostasis was not significant. The cut off value of disulphide/total thiol ratio was 6.5 with sensitivity of 51% and specificity of 96%. While the mean TB and DB levels were significantly higher in both acute and complicated appendicitis groups, there was no difference between AA and CA groups. The cut off value of TB and DB to predict AA or CA in all population was 0.74 and 0.24 with a sensitivity of 54% and a specificity of 94%,respectively.

Conclusions: Thiol/ disulfide homeostasis shifted towards disulfide direction in children with appendicitis. When the test is positive in patient with suspected appendicitis, the likelihood of appendicitis is high. We couldn’t find any evidence that hyperbilirubinemia distinguish CA in children. The elevation in TB and DB levels may have diagnostic value in children with appendicitis.
Caecal volvulus (CV) and appendiceal volvulus (AV) are manifestations of intestinal malrotation/malfixation and can represent life-threatening conditions. This study reviewed outcomes of CV/AV in the pediatric group.

MEDLINE/PubMed were reviewed from 1990-2017 using the keywords "caecal" or "appendiceal", "volvulus" and "children". Data was collected for symptoms, morbidities, previous surgery, management, complications and mortality. Articles were selected by 2 reviewers.

Twenty-two articles with n=27 (n=26 CV, n=1 AV) were included, median age 9 years (5 days-15 years) with n=20 (74%) males. Abdominal distention n=20 (74%) and abdominal pain (n=18, 66.7%) were the common symptoms. Cornelia de Lange syndrome (CdL) was present in n=6 (22.2%). Radiological investigations were performed in n=26 (96.3%) with plain radiographs (XR) being most common n=23 (85.2%). Correct interpretation was observed: XR 3/23 (13%), contrast enemas 4/5 (80%), ultrasound 1/2 (50%) and computed tomography (CT) 2/4 (50%). For the other cases, intervention was indicated for signs of bowel obstruction n=18 (66.7%) and appendiceal abscess (CT) n=1 (3.7%). Treatment was surgical n=26 (n=25 open, n=1 laparoscopy) and endoscopic n=1. Stoma was placed in n=9 (33.3%). There was no mortality. Complications in n=7 (25.9%): n=5 sepsis and n=2 enterocutaneous fistula. There was n=1 CV recurrence after 6 years.

CV/AV are rare pathologies in children. Since 1/4th CV/AV patients have CdL syndrome, it is paramount to consider CV/AV as differential diagnosis when abdominal symptoms occur in CdL. Investigations are not always conclusive, but with prompt surgical management complications are minor and mortality is rare.
**PW17-LG12**

**IMPACT ON PATIENT CARE OF A MULTIDISCIPLINARY CENTER SPECIALIZING IN COLORECTAL AND PELVIC RECONSTRUCTION**

Alejandra Vilanova-Sanchez, Carlos Reck-Burneo, Alessandra Gasior, Laura Weaver, Karen Diefenbach, Dennis Minzler, Robert Dyckes, Christina B Ching, Venkata R. Jayanthi, Katerine McCracken, Geri Hewitt, Richard J Wood, Marc A. Levitt

*Nationwide Children’s Hospital, Columbus, USA*

**Aim**

Many patients with an anorectal malformation (ARM) or pelvic anomaly have associated urologic or gynecologic problems. We hypothesized that our multidisciplinary center, which integrates pediatric colorectal, urologic, gynecologic and GI motility services, could impact a patient’s anesthetic exposures and hospital visits.

**Methods**

We tabulated during 2015 anesthetic/surgical events, endotracheal intubations, and clinic/hospital visits for all patients having a combined procedure.

**Main results**

82 patients underwent 132 combined procedures (table 1). The median age at intervention was 3 years [0.2-17], and length of follow up was 25 months [7-31]. The number of procedures in patients who underwent combined surgery was lower as compared to if they had been done independently [1(1-5) vs 3(2-7); p<0.001]. Intubations were also lower (1[1-3] vs 2[1-6]; p<0.001). Hospital length of stay was significantly lower for the combined procedures vs the theoretical individual procedures (8 days [3-20] vs 10 days [4-16]; p<0.05). Postoperative clinic visits were fewer when combined visits were coordinated as compared to the theoretical individual clinic visits (urology, gynecology and colorectal) (1[1-4] vs 2[1-6]; p<0.001).

**Conclusions**

Patients with anorectal and pelvic malformations are likely to have many medical or surgical interventions during their lifetime. A multidisciplinary approach can reduce surgical interventions, anesthetic procedures, endotracheal intubations, and hospital/outpatient visits.

**Malformation**

- ARM: 75
- Spinal Anomaly: 5
- Sacrococcygeal Teratoma: 1
- Hirschsprung’s: 1
- Colorectal+Urology + Gynecology: 87

**Specialty**

- Colorectal+Urology: 34
- Colorectal+Gynecology: 11
- Practical Advantage: 115

**Reason for Collaboration**

- Tissue sharing: 9
- Single Pelvic Exploration: 8
PW18-UG01
TOTAL OESOPHAGO-GASTRIC DISSOCIATION: INDICATIONS AND RESULTS IN TWO THIRD-LEVEL CENTERS

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Aim of the study:
Fundoplication is the first choice in gastro-oesophageal reflux (GOR) refractory to medical therapy. Literature describes significant morbidity and recurrences in children with cerebral palsy and seizures. Total oesophago-gastric dissociation (TOGD) as described by Bianchi is a valid alternative. The aim of the study is to evaluate the indications and results of TOGD in two third-level centers.

Methods:
We included all patients with severe GOR who underwent a TOGD between February 2016 and June 2017. Previous operations, presence of epilepsy, tetra paresis and ab ingestis pneumonia were considered. Postoperative complications, re-do surgery, contrast study after one month from the intervention and life quality after 6 months were recorded.

Main results:
Fifteen children underwent laparoscopic or robotic TOGD. Five patients had already undergone fundoplication. Seizures and tetra paresis were present in 10/15 patients, ab ingestis pneumonia in 8/15. No intraoperative complications, conversions to open surgery and vagal lesions were recorded. One oesophago-jejunal dehiscence was detected after 5 days in a patient with oesophageal atresia history. One patient had an hemorrhagic gastric complication and one needed a dilatation for a stenosis after 3 months. Most of the children gained weight after 6 months and no ab ingestis pneumonia were recorded after the TOGD.

Conclusion: TOGD could be considered a valid option for GOR, not only as "rescue" intervention, but also as primary one in neurologically impaired children. Short term major complications are present. Long term follow up shows a better quality of life.
PW18-UG02
COMPARISON OF DELAYED ANASTOMOTIC REPAIR AND PRIMARY ANASTOMOTIC REPAIR IN PATIENTS WITH INTESTINAL ATRESIA

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Aim of the Study

Intestinal atresia is a congenital defect which results in intestinal obstruction. Atresias are classified by location and appearance. There are various surgical approaches to atresias including primary anastomotic repair (PAR) versus ostomy with delayed anastomotic repair (DAR). The aim of this study was to compare these two approaches to repair in regard to patient outcomes.

Methods

A retrospective review was performed of patients admitted with the diagnosis of intestinal atresia from 2007-2017. Patients with gastoschisis, duodenal and colonic atresia were excluded from analysis.

Main Results

The review included 45 cases of intestinal atresia.

<table>
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<th>Type</th>
<th>PAR (%)</th>
<th>DAR (%)</th>
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<td>16.7 (1/6)</td>
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<tr>
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<td>11.1 (4/36)</td>
<td>0.0 (0/6)</td>
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<td>16.7 (1/6)</td>
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<td>19.4 (7/36)</td>
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<td>IV</td>
<td>16.7 (6/36)</td>
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<tr>
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<td>66.7 (4/6)</td>
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<td>33.3 (2/6)</td>
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<td>2.0 (range 2.0-3.0)</td>
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<tr>
<td>Number of Surgeries</td>
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<td>Return to OR after anastomotic repair (%)</td>
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<tr>
<td>I</td>
<td>7.7 (1/13)</td>
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</tr>
<tr>
<td>II</td>
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<tr>
<td>IV</td>
<td>23.1 (3/13)</td>
<td>100.0% (1/1)</td>
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<td>Length of Stay (days)</td>
<td>29.0 (20.0-66.0)</td>
<td>112.5 (76.5-139.5)</td>
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<td>Time to Full Feeds (days)</td>
<td>19.0 (13.5-42.0)</td>
<td>93.0 (76.0-114.5)</td>
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<tr>
<td>TPN (days)</td>
<td>16.0 (12.0-26.0)</td>
<td>142.0 (126.0-174.0)</td>
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Conclusion
Although limited by a small sample size for subgroup analysis, our data supports improved outcomes with primary anastomosis for most patients with intestinal atresia. The more complex patients with types III and IV atresia are susceptible to complications if not diverted and likely benefit from a staged approach.
CONGENITAL PYLORIC ATRESIA: ASSESSMENT OF MANAGEMENT AND OUTCOMES

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Aim: This study aimed to analyze congenital pyloric atresia (CPA) with regards to diagnosis and management in children.

Methods: The literature was reviewed for articles on Pubmed with search terms “congenital”, “pyloric” and “atresia”. The articles were reviewed by 2 surgeons to reduce bias in selection.

Results: The search revealed 50 articles involving n=113 with CPA from 1995-2017. The diagnosis was established in n=108 in the newborn period and in n=6 between 2-6 years. Prematurity was associated in n=37 (32.74%) and polyhydramnios in n=44 (38.93%). CPA had a family history in n=6 and consanguinity was accounted in n=11. Associated anomalies were found in n=91: epidermolysis bullosa (EB) 41.59% and other atresias 17.69%. Type-I (pyloric diaphragm) was diagnosed in n=41 (n=2 pyloric diaphragmatic duplication and n=6 complete pyloric diaphragm), Type-II (pyloric atresia without gap) in n=18 and Type-III (pyloric atresia with gap) in n=12. Therapeutic management was Heineke-Mickulicz pyloroplasty n=49, gastroduodenostomies n=40, gastrojejunostomies n=7, Finney pyloroplasties n=2 and gastrostomies n=15. Additional procedures for associated anomalies were performed in n=39. Only n=1 was approached with laparoscopic surgery. N=2 needed re-intervention for a leak (n=1) and pyloric lumen stenosis (n=1). Complications arose in n=58 (51.32%) leading to death in n=50 (44.24%), n=36/58 (62%) of which had EB.

Conclusions: Over 80% cases of CPA have associated anomalies. Open Heineke-Mickulicz pyloroplasty and gastroduodenostomy are the preferred surgical procedures, with low rates of re-interventions. Around half the patients developed complications after surgery and mortality is high especially with EB association.
PW18-UG04
LAPAROSCOPIC GASTROSTOMY VERSUS PERCUTANEOUS ENDOSCOPIC GASTROSTOMY: OUTCOMES AND COMPLICATIONS

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Miguel Servet University Hospital, Zaragoza, Spain

Objective. To compare the outcomes and complications between percutaneous endoscopic gastrostomy (PEG) and laparoscopic gastrostomy (LG).

Methods. Retrospective review of 78 patients who had their gastrostomies inserted in our hospital (2010-2017). Surgical techniques and minor and major complications were compared.

Main results. 24 LG (14 males) and 54 PEG (28 males) were performed. The mean age was 4'77 [0'15-15'56] years for LG and 5'37 [0'12-16'58] years for PEG. Mean operative time for the PEG group was significantly lower when compared with the LG technique (p<0'05). No significant differences were found in major and minor complications between both techniques, however, there is a clear tendency to minor complications in LG (n = 13 in LG versus n = 19 in PEG) and major complications in PEG (n = 1 in LG versus n = 11 in PEG). The median time of presentation of complications was 59 [2-975] days. The minor complications were, mainly, overgranulation and peristomal leakage. We found a statistically significant association between patients with oncological comorbidity and minor complications (p <0.05) The major complications were, a tube dislodgement in LG group and 8 Buried-Bumper syndrome, a colon caught without perforation, a colon perforation, an omentum caught, two dislodgement, a small bowel perforation in PEG group.

Conlusions. Despite not finding statistically significant differences in the major and minor complications between both groups, the higher frequency and severity of major complications in the PEG group is evident; considering LG a safer technique. However, studies with larger number of patients are necessary.
PW18-UG05
TOTAL OESOPHAGO-GASTRIC DISSOCIATION: A 20-YEAR RETROSPECTIVE

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Background: Total oesophago-gastric dissociation (TOGD) has been performed in our institution since 1994, pre-dating its published description by Bianchi in 1997. Originally developed as a “rescue” procedure for failed anti-reflux procedures in severely neuro-developmentally impaired children, it has become a viable primary option in this group, as well as in neurologically normal children with severe reflux secondary to oesophageal pathology.

Objective: We describe our institution’s experience of TOGD between 1994-2015.

Methods: Three papers were analysed detailing our total experience of TOGD between 1994-2015; outcome data was merged to form a complete picture of the history of this procedure in our institution.

Results: In total 77 patients underwent TOGD between 1994-2015 (34 males, 43 females). Eleven patients (14.3%) were neurologically normal children with oesophageal pathology, the remainder were children with gastro-oesophageal reflux disease (GORD) secondary to severe neurological impairment. Fifty-six procedures (72.7%) were primary procedures, the remainder performed as rescue procedures following the failure of fundoplication. No patients reported a recurrence of reflux symptoms; the mean hospital in-patient stay was 10.6 days. There were 15 reported complications in 10 patients representing 13% of the cohort; two deaths were attributable to the procedure (2.6%).

Conclusion: TOGD remains a good option for children suffering with reflux - both neurologically normal with complex oesophageal pathology and those with severe neurological impairment – in the primary setting as well as a rescue procedure following failure of fundoplication. Our experience suggests no recurrence of reflux symptoms post-TOGD, and a comparable morbidity and mortality to other anti-reflux procedures.
Aim: Percutaneous gastrojejunalostomy (PEGJ) feeding is a minimal access approach to provide nutritional support when gastric feeds are difficult. This study performs an analysis of PEGJ techniques and outcomes in children.

Methods: Literature was searched on Pubmed® using terms: “percutaneous gastrojejunostomy”, “gastrojejunal feeding” and “children”. Endpoints of the study were age, associated morbidities, type of procedure, complications, failure and reoperation rate, mortality.

Results: 18 articles with a total of n=555 PEGJ tube placements were analyzed which indicated a mean age 4.25 years (1.4 months-19 years). Comorbidities for PEGJ patients were divided into morphological groups: neurologic n=330 (59.4%), respiratory n=77 (13.9%), cardiac n=69 (12.4%) and gastroenterological n=67 (12%). Image-guided technique was used for tube placement in n=250 (45%) performed by Interventional Radiologist, and endoscopic technique (orally or through an existing gastrostomy) in n=305 (55%) performed by Gastroenterologists/Surgeons. Failure rate was 2.8% for all PEGJ inserted (3.1% for image-guided, 2.2% for endoscopic technique). Surgery was subsequently required in n=92 (16.6%) as fundoplication or surgical jejunostomy placement. Minor complications were tube related in n=364 (65.5%). Major complications included intussusception n=38 (6.9%), bowel perforation n=14 (3.5%), and infection n=35 (6.25%). Lethal outcome was reported in n=53 (9.5%), mostly related to comorbidities.

Conclusions: Neurological affected patients account for >50% of the clinical conditions that require PEGJ in pediatric age group. PEGJ placement is evenly distributed between Interventional Radiologists and Gastroenterologists/Surgeons. Image-guided technique presents a comparable complication rate to endoscopic technique. Intussusception and bowel perforation account for 10% of complications in PEGJ’s.
PW18-UG07
LAPAROSCOPIC VERSUS OPEN PYLOROMYOTOMY IN INFANTS: FOCUS ON COMPLICATIONS

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Aim of the study

To determine the impact of laparoscopic pyloromyotomy (LP) versus open circumumbilical pyloromyotomy (OP) on per and post-operative complications in infants with hypertrophic pyloric stenosis.

Methods

A retrospective study between 2002 and 2017 was performed, 247 infants with congenital pyloric stenosis were included. Patients aged more than 1 year old were excluded. The length of operating room time, surgical procedure, postoperative stay, time to refeeding, and complications were evaluated by review of the hospital record.

Main results

Out of 247 pyloromyotomy patients, 100 underwent LP (40.5%) and 147 underwent OP (59.5%). There was no difference in the incidence of per or post-operative complications in the 2 groups. The duration of surgery was significantly longer in the OP group (28.9+-9.1minutes vs. 21.9+-8.3minutes, p=0.009) and so was the postoperative length of stay (2.6+-1.4 days vs. 2.4+-2.8days, p=0.001). The overall incidence of unspecific complications (hematoma, wound infection, wound dehiscence) was higher in the OP group and there were more specific complications (incomplete pyloromyotomy and mucosal perforation) in the LP group, but it was not statistically significant.

Conclusions

Laparoscopic pyloromyotomy has a similar complication rate compared with the open umbilical approach. Because duration of surgery and postoperative hospital length of stay are at best a few minutes shorter after LP than after OP, we cannot acknowledge the superiority of one approach over the other one.
GASTRIC VOLVULUS IN CHILDREN: MANAGEMENT AND OUTCOMES

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Gastric volvulus (GV) is a rare entity that requires high suspicion for diagnosis and prompt management. This study reviewed outcomes of GV in children.

MEDLINE/PubMed databases were searched for articles (2008-2017) using key-words "gastric", "volvulus" and "children". Data was collected for symptoms, morbidities, previous surgery, diagnosis, management, complications, and follow-up. Articles were selected by 2 reviewers.

Fifty papers with n=76 patients were included, median age 3.3 years (newborn - 17 years), with n=35 (46.1%) females. Vomiting n=46 (60.5%) and abdominal pain n=28 (36.8%) were the most common symptoms, and acute presentation occurred in the majority of cases n=56 (73.7%). Associated conditions, such as congenital heart defects and cerebral palsy, were present n=18 (23.7%) and history of previous surgery or abdominal trauma in n=8 (10.5%). Radiological examinations were performed in all cases, and upper gastrointestinal series (UGI) was diagnostic for n=41/42 (97.6%). Management was surgical in n=53 (n=48 open, n=14 laparoscopic approach) with n=40 with gastropexy/gastrostomy, conservative management in n=9, endoscopic management n=2 and n=3 lethal outcomes before commencement of treatment. Complications occurred in n=15 (19.7%), with esophageal stenosis being the most common (n=4). There were n=9 (11.8%) deaths (n=8 after acute presentation), and only n=1 recurrence 6 months post endoscopic management, that was treated conservatively. The mean follow-up was 8.4 months.

GV occurs at a median age of 3.3 years and requires prompt management as mortality >10%. The preferred surgical approach for GV is gastropexy/gastrostomy. Esophageal stenosis is the most common morbidity post GV management.
OUTCOME OF PATIENTS AFTER NECROTIZING ENTEROCOLITIS - DOES THE TYPE OF SURGICAL PROCEDURE AFFECT MORTALITY RATE AND MORBIDITY?

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**Background.** Necrotizing enterocolitis is one of the most serious cause of acquired abdominal emergency in premature newborns. The thesis evaluates postoperative outcome of patients after surgical NEC.

**Methods.** The single center retrospective analysis of 10-years period was performed in tertiary hospital to assess risk factors (prematurity, birth weight, postnatal course, additional diseases, bacterial infection) of necrotizing enterocolitis. The authors evaluated outcome after different surgical procedures.

**Results.** Overall number of patients treated for NEC in the reviewed period was 157. Surgery was performed in 56.7% (n=89). Significant risk factors for NEC outbreak were caesarean section (p<0.05), prematurity (p<0.01), very/extremely low birth weight (p<0.001), which was significant also in prediction of mortality in operated patients (p<0.05). Four types of surgical procedure were compared: primary anastomosis, terminal enterostomy, duplex enterostomy and enterotomy with abdominal drainage. Total mortality in surgically treated patients reached 29% and there was no significant difference between the type of enterostomy in relation to mortality rate. Significantly lower morbidity was observed after duplex enterostomy.

**Conclusion.** Risk factors of NEC are well known and include prematurity, low birth weight, thrombocytopenia. Clinical symptoms lead to necessary therapeutic steps with surgery inevitable in indicated cases. The best surgical approach for NEC is not clear and depends on the extent of intestinal involvement. In presented study authors did not confirm the significance of surgical approach to mortality rate, however best postoperative outcome was achieved by duplex ileostomy.
ENDOSCOPIC MANAGEMENT OF FOREIGN BODY INGESTION AND FOOD IMPACTION IN CHILDREN. WHICH ENDOSCOPIC METHOD AND RETRIVAL DEVICE TO USE?

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Background: The aim of this study is to review the experience on foreign body ingestion (FBI) in children with special emphasis on the endoscopic techniques and retrieval devices used for FB extraction.

Material Method: The charts of 268 children who presented with FBI and food impaction and underwent removal of FB between 2002 and 2016, were reviewed retrospectively. Demographic data, ingested material, removal technique and tool, level of FB, complications and outcomes were recorded.

Results: 294 foreign bodies removed from 268 children with mean age of 34 months. 54% of the FB were entrapped in esophagus and 32% were in stomach. 8% in both stomach, duodenum and intestine, 6% only in intestine. The most frequently ingested item was coin (39%), followed by small disk batteries (20%) and safety pins (15%). The preferred endoscopic technique for esophageal FB was rigid endoscopy (74%) and the most common retrieval tool was optical forceps. Retrieval net (44%) was the most common tool for the FB in stomach followed, retrieval grasper (25%) and grasping forceps (19%) and polypectomy snare (12%).

Conclusions: Rigid endoscopy should be preferred for esophageal FB since optical forceps allow strong grasping of the coin. However retrieval of sharp objects like safety pins requires flexible endoscopic view for a safe FB extraction. Retrieval nets are the most effective tools for the removal of gastric foreign bodies as they provide safe grasp for coins, batteries and magnets which are the commonly ingested FB by children.
PW18-UG11
BOLUS FEEDING AFTER FUNDOPLICATION IS SAFE

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Aim of the Study: Failure of fundoplication has been linked to several causes. We evaluated the effect of early bolus feeding on rates of wrap disruption in a minimal dissection paradigm.

Methods: A retrospective review was performed of patients undergoing laparoscopic fundoplication from 2014 to 2016. Data were collected on demographics, hospital course, and one-month follow up. All values are reported as medians with interquartile ranges (IQR).

Main results: 58 patients were enrolled. Median age was 200 days (118, 538). The most common preoperative symptom was retching (n=33, 58%). 39 patients (67%) had a gastrostomy tube placed during fundoplication and 8 patients (14%) already had a gastrostomy tube.

Median time to initiation of feeds was 0 days (0, 1) with 28 patients (51%) receiving bolus feeds and 26 (48%) on continuous. Three patients (10%) on continuous were started at their full rate. Median time to bolus conversion was 2 days (1, 6).

Median follow up was 1.9 months (.83, 2.2) with 23 patients (39.6%) reporting retching. There was no difference in the rate of retching between patients on continuous or bolus feeds (34.4% vs 44.8%, p=0.59). 18 patients (37%) underwent workup for ongoing reflux symptoms. One patient from the bolus group was diagnosed with a wrap disruption – a non-significant difference between the groups (p=1). This patient’s symptoms were controlled and she was allowed to gain weight with a successful repair 18 months later.

Conclusions: Bolus feeding after laparoscopic fundoplication does not lead to increased rates of retching or wrap disruption.
PW18-UG12
LAPAROSCOPIC-ASSISTED GASTRIC TRANSPOSITION: A STANDARDISED AND RISK-ASSESSED APPROACH

Charlotte Holbrook, Paolo De Coppi, Kate Cross, Joseph Curry
Great Ormond Street Hospital, London, United Kingdom

Aim:
Our 14-year experience with laparoscopic gastric transposition has been modified over time, aiming to improve operative safety and reduce complications. Additionally, standardised pre-operative investigations have been instituted to identify anatomical anomalies. Our aim was to review our outcomes following introduction of these changes 4 years ago.

Methods:
Retrospective case note review was undertaken of patients who had laparoscopic-assisted gastric transposition between 2013 and 2017.

Results:
12 patients were identified. Mean age was 3.5 years (range 0.9 to 8.6). Mean follow-up was 1.5 years (range 0.2 to 3.9). Indications were long gap oesophageal atresia (8) and caustic injury (4). Preoperatively all patients had chest CT, identifying abnormal vascular anatomy in 2, and microlaryngobronchoscopy, identifying abnormalities in 5. Operative technique involved mini-laparotomy for preparation of the stomach and laparo-thoracoscopy for mediastinal dissection and transposition. One procedure was converted to laparotomy due to dense adhesions (subsequently excluded from analysis). Median operating time was 351 minutes (245 to 515). Postoperative complications are shown in Table 1. There were no anastomotic leaks and no early postoperative mortality. 4 require supplemental feeding via jejunostomy due to oral aversion.

<table>
<thead>
<tr>
<th>Early</th>
<th>Late</th>
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</thead>
<tbody>
<tr>
<td>Pneumonia</td>
<td>Stricture</td>
</tr>
<tr>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>Post extubation stridor</td>
<td>Poor weight gain</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
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<tr>
<td>Chylothorax</td>
<td>Recurrent vomiting</td>
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<td>1</td>
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</table>

Conclusion:
- The safety of posterior mediastinal dissection is increased by use of the laparoscope and augmented by standardised preoperative investigations which help define surgical strategy.
- Retaining a mini-open technique for part of the procedure improves the security and safety of gastric suturing (there were no anastomotic leaks).
PW19-G01
TANNING VERSUS SCHUSTER IN THE MANAGEMENT OF THE OMPhALOCELE: WHAT TO CHOOSE?

Aurélien BINET1, Sarah AMAR3, Khalid ALZAHRA1, Bertin KOUAME2, Ossenou OUATTARA3, Maxime KOFFI3, Soukere MOUFIDATH1, Dorian NASSER3, François BASTARD4, Francis LEFEBVRE5, Aurélien SCALABRE6, Hubert LARDY1
1Universitary Hospital Center - Pediatric Surgery Unit, TOURS, France. 2Universitary Hospital Center - Pediatric Surgery Unit, ABIDJAN, Afghanistan. 3Universitary Hospital Center - Pediatric Surgery Unit, ABIDJAN, Côte d'Ivoire. 4Universitary Hospital Center - Pediatric Surgery Unit, ANGERS, France. 5Universitary Hospital Center - Pediatric Surgery Unit, REIMS, France. 6Universitary Hospital Center - Pediatric Surgery Unit, SAINT ETIENNE, France

Aim of the study
Different treatments are currently proposed in front of a giant omphalocele, depending on the easy access or not to a technical platform resuscitation. The purpose of this study is to compare the conservative and surgical management of the giant omphalocele in terms of morbidity and mortality.

Methods
Multicentric retrospective study between January 1996 and December 2016 including all giant omphaloceles. Epidemiology was studied as well as medical and surgical managements.

Main results
147 patients included (98 patients in the "tanning" group and 49 in the "surgery" group). Hospital length of stay (resuscitation / surgery) is significantly shorter in the "tanning" group (11 versus 95 days ; p = 0.0001). No significant differences between the two groups in terms of mortality (p = 0.1529). The average duration for oral empowerment was acquired at 179 days in the "surgery" group, whereas from the date of birth in the tanning group. No significant differences in terms of closure time.

Conclusion
The increase in survival in the "surgery" group is at the cost of significant morbidity. Access to a technical intensive care unit could reduce the mortality rate of children cared for by tanning. Immediate feeding in the "tanning" group and early return home is a real benefit in the overall development of the child. This technique should be reconsidered in developed countries considering the global benefits and available techniques of remote/delayed abdominal wall reconstruction.
MANAGEMENT OF HYDATID DISEASE IN CHILDREN: A REPORT OF 135 PEDIATRIC HYDATIDOSIS

Gizem Beril Ozdemir, Zehra Gunyuz Temir, Ayse Basak Ucan, Belkis Deniz Ozbilek, Ozkan Okur, Incinur Genisol, Volkan Altnok, Gokce Sonmez, Hunkar Erdogan, Ayberk Cubukcu, Munevver Hosgor

Health Medical Sciences University, Izmir Dr. Behcet Uz Children’s Hospital, Izmir, Turkey

AIM OF THE STUDY: The aim of this study is to investigate demographic features and results of surgical and medical treatment of children with hydatid disease.

METHODS: The clinical records of 135 children treated for hydatid disease from January 2000 to December 2016 were retrospectively reviewed. Data were collected in terms of demographic characteristics, methods and results of treatment and complications.

MAIN RESULTS: There were 85 boys and 50 girls, age between 3-16 years. %41 of patients were living in the countryside and %89 had a contact with animals previously. Lung cysts were present in 77 cases, followed by liver cysts in 67, other organ involvement was detected in 7. 24 cases were treated with only albendazole chemotherapy, 13/24 had liver cysts and 9/24 had lung cysts. %83 of all cases were treated by combined albendazole chemotherapy and surgery. Percutaneous drainage was the first surgical procedure in appropriate patients with liver cysts. Thoracotomy was performed after 7 days albendazole treatment in lung cysts larger than 5 cm. Complication rate of only albendazole treatment (urticarial reaction, %5) was lower than surgery combined with albendazole (pneumothorax, pneumonia, septicemia, biliary fistula, %20). There were 6 recurrences after surgery (4 percutaneous drainage, 2 thoracotomy). 1 patient with recurrent liver cyst and 2 patients with recurrent pulmonary cysts were re-operated.

CONCLUSIONS: Hydatid disease still remains a challenging condition in our country and leads to higher necessity for pediatric surgeons to be more aware of clinical features and treatment strategies of pediatric hydatidosis.
PW19-G03
MANAGEMENT AND OUTCOMES OF CONGENITAL ANOMALIES IN LOW-, MIDDLE-, AND HIGH-INCOME COUNTRIES: PROTOCOL FOR A MULTI-CENTRE, INTERNATIONAL, PROSPECTIVE COHORT STUDY

Naomi Wright¹, Niyi Ade-Ajayi², Justine Davies¹, Dan Poenaru³, Nick Sevdalis⁴, Andy Leather¹, Global PaedSurg Research Collaboration¹

¹King’s Centre for Global Health and Health Partnerships, London, United Kingdom. ²King’s College Hospital, London, United Kingdom. ³McGill University, Montreal, Canada. ⁴King’s College London, London, United Kingdom

Aim: Congenital anomalies have risen to become the 5th leading cause of death in children below age 5-years globally, yet limited data exists from low- and middle-income countries where the majority of these deaths occur. This collaboration aims to undertake a multi-centre prospective cohort study of congenital anomalies globally to compare outcomes between low-, middle- and high-income countries (LM&HICs).

Methods: The Global PaedSurg Research Collaboration will be established consisting of children’s surgical care providers from around the world participating in the study; active collaborators will be co-authors of resulting presentations and publication(s). Data will be collected on patients presenting primarily with anorectal malformation, intestinal atresia, oesophageal atresia, gastroschisis, exomphalos, congenital diaphragmatic hernia, and Hirschsprung’s disease for a minimum of 30-consecutive days between Oct 2018 - April 2019. Data will be collected using REDCap and will include patient demographics, clinical status, interventions and outcome.

Results: The primary outcome will be all-cause in-hospital mortality, with post-operative complications being secondary outcomes. Chi-squared analysis will be used to compare mortality between LM&HICs. Multivariate logistic regression analysis will be used to identify factors affecting outcomes. Ethical approval will be sought from all participating centres. Funding has been granted by the Wellcome Trust.

Conclusion: The study aims to be the first large-scale, geographically comprehensive, multi-centre prospective cohort study of a selection of common congenital anomalies across the globe to define current management and outcomes, aid advocacy and global health prioritisation, and inform future interventional studies aimed at improving outcomes.
ADVERSE EVENTS OF DIFFERENT SCLEROSING AGENTS IN THE TREATMENT OF LYMPHATIC MALFORMATIONS IN CHILDREN

Silje Kooijman, Frédérique Bouwman, Bas Verhoeven, Leo Schultz Kool, Carine van der Vleuten, Sanne Botden, Ivo de Blauuw
Radboudumc, Nijmegen, Netherlands

Aim of study: Lymphatic malformations (LMs) often require treatment, because of lesion-related complications. Sclerotherapy is considered the primary treatment of choice. Bleomycin has proven to be effective, but a major concern is the risk of pulmonary toxicity. This study compares the differences in complications between sclerosing agents in the treatment of LMs in children.

Methods: All sclerotherapy procedures for children with LMs during 2011-2016 at a national referral center were retrospectively reviewed. Demographics and procedure-related complications were recorded and compared between different sclerosing agents. Patients treated with ethanol were excluded.

Main results: A total of 217 procedures in 109 patients were performed using bleomycin (63%), aethoxysclerol (22%), doxycycline (7%) or bleomycin and aethoxysclerol combined (9%). The median number of procedures per patient was two (range 1-13). Table 1 shows the procedure-related complications; there were no significant differences. The two cases of functional impairment required an intervention and were thus considered major. Less invasive treatment, including analgesics (7%) and antibiotics (3%), was required in 22 cases (10%). One patient developed pneumonia after bleomycin injection, which resolved after antibiotic treatment. No known cases of pulmonary fibrosis occurred.

Conclusions: Only one patient developed pneumonia during the study period. This institutional study shows no significant differences in complications between the sclerosing agents, although the design of the study allows patient-specific treatment regimes. The risk of bleomycin on pulmonary toxicity seems to be negligible when used correctly, so it appears to be a safe agent in the treatment of LMs in children.

<table>
<thead>
<tr>
<th>Procedure-related complications after sclerotherapy</th>
<th>Total Group N=217</th>
<th>Bleomycin n=136</th>
<th>Aethoxysclerol n=47</th>
<th>Doxycycline n=15</th>
<th>Combi bleo/aethoxy n=19</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Short term (&lt; 30 days)</strong></td>
<td></td>
<td></td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Minor</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Pain</td>
<td>12 (6%)</td>
<td>8 (6%)</td>
<td>1 (2%)</td>
<td>2 (13%)</td>
<td>1 (5%)</td>
<td>0.411</td>
</tr>
<tr>
<td>Swelling</td>
<td>43 (20%)</td>
<td>24 (18%)</td>
<td>12 (26%)</td>
<td>1 (7%)</td>
<td>6 (32%)</td>
<td>0.199</td>
</tr>
<tr>
<td>Fever</td>
<td>12 (6%)</td>
<td>9 (7%)</td>
<td>1 (2%)</td>
<td>1 (7%)</td>
<td>1 (5%)</td>
<td>0.708</td>
</tr>
<tr>
<td>Infection</td>
<td>8 (4%)</td>
<td>3 (2%)</td>
<td>2 (4%)</td>
<td>1 (7%)</td>
<td>3 (15%)</td>
<td>0.631</td>
</tr>
<tr>
<td>Hematoma</td>
<td>16 (7%)</td>
<td>10 (7%)</td>
<td>2 (4%)</td>
<td>1 (7%)</td>
<td>3 (15%)</td>
<td>0.449</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>1 (1%)</td>
<td>1 (1%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0.897</td>
</tr>
<tr>
<td>Other</td>
<td>4 (2%)</td>
<td>3 (2%)</td>
<td>1 (2%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0.659</td>
</tr>
<tr>
<td><strong>Major</strong></td>
<td></td>
<td></td>
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<td></td>
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<tr>
<td>Functional impairment</td>
<td>2 (1%)</td>
<td>1 (1%)</td>
<td>1 (2%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0.773</td>
</tr>
<tr>
<td><strong>Long term</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Minor</td>
<td>3 (1%)</td>
<td>3 (2%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0.612</td>
</tr>
<tr>
<td>Scarring</td>
<td>1 (1%)</td>
<td>1 (1%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0.897</td>
</tr>
<tr>
<td>Hyperpigmentation</td>
<td>1 (1%)</td>
<td>1 (1%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0.897</td>
</tr>
<tr>
<td><strong>Major</strong></td>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lymphedema</td>
<td>1 (1%)</td>
<td>1 (1%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0.897</td>
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</table>
PW19-G05:
STANDARD OF CARE IN RARE DISEASES: PREPARING FOR ERNICA’S UPCOMING GUIDELINES ON HIRSCHSPRUNG’S DISEASE

Kristiina Kyrklund¹, Francesco Fascetti-Léon², René Wijnen³, Mikko Pakarinen¹
¹Helsinki University Children’s Hospital, Helsinki, Finland. ²Pediatric Surgery, Department of Women’s and Children’s Health, University of Padova Italy, Padova, Italy. ³Erasmus Medical Centre, Rotterdam, Netherlands

Aims of the study –
To enable European Reference Networks (ERNs) including ERNICA, the ERN for inherited and congenital malformations, to develop clinical practice guidelines (CPGs) for improving standards of care in rare diseases (RDs).

Methods –
The RD action & DG Santé workshop (Rome, Italy, December 2017) was attended by 63 delegates, including representatives from 23/24 ERNs, European Project/ Domain/National Experts and Patient Advisory Groups. Strategies for enabling the generation and uptake of CPGs were identified.

Results –
Developing adequate mechanisms of care in RDs requires standardization of care pathways and terminology, and registries for outcome reporting to guide evidence-based practice. CPGs should aim for practicality, cost-effectiveness, definition of safe standards and incorporate strategies for disease prevention. An open call for experts and systematic pooling of knowledge through ERNs are needed to overcome geographical and case volume limitations. High-quality evidence is often not available in RDs. Transparent approaches, external review and patient involvement ensure quality control and demonstrate a view of healthcare targets that incorporates lived experience of the disease. Lay versions are a powerful means for enhancing uptake and dissemination of CPGs by placing pressure on national systems for care delivery according to established standards of good practice.

Conclusions –
ERNs are uniquely suited for developing high-quality CPGs to improve healthcare outcomes in RDs. Outcomes from this workshop provide a foundation for ERNICA’s upcoming CPGs on Hirschsprung’s disease, esophageal atresia and other malformations, with applicability across RDs within pediatric surgery. Because of cooperation of expertise centers within ERN’s, implementation is better ensured.
COAGULOPATHY IN NEONATAL SURGERY

Pamela Middleton, Odette Pheiffer, Behrouz Banieghbal
Tygerberg Hospital, Cape Town, South Africa

Aim:

Pro- and anti-coagulant components remain in homeostasis in healthy neonates however during illness this equilibrium is disturbed leading to coagulopathy. In neonates with severe necrotising enterocolitis (NEC) coagulopathy is common, particularly with thrombocytopenia. This study's aim was to compare the rate of abnormal INR as a marker for coagulopathy in neonates with NEC requiring surgery to that of other neonatal patients undergoing surgery.

Methods:

A review of all neonates undergoing surgical procedures over the period January 2016 – December 2017 was undertaken. Ethical approval was obtained. The patients were grouped according to underlying diagnosis and the procedure they underwent. The coagulation screen for each patient was recorded and patients with an INR greater than 1.5 were labelled as coagulopathic.

Main Results:

Of 209 patient requiring surgery 94 underwent laparotomies. Of these 30% (n=29) were for NEC related complications and 48% (n=14) of the patient with NEC had abnormal preoperative INR. Patients undergoing laparotomy for other causes had an abnormal INR in 12.3% (n=8) of cases. Patients undergoing a laparotomy for NEC were found to have a relative risk of 2.9 of having abnormal INR compared with neonates undergoing exploratory laparotomy for another reason.

Conclusion:

NEC is associated with a significantly higher rate of coagulopathy than other surgical conditions in the neonatal period. This is possibly due to increased activation of immunological factors which interact with the balance of procoagulatory and anticoagulatory factors in neonates. Risk of coagulopathy should not be overlooked in neonatal patients with or without NEC.
LOW COST TRAINING OF PEDIATRIC MINIMALLY INVASIVE PROCEDURES

Sanne Botden, Erik Leijte, Ivo de Blaauw
Radboudumc - Amalia children’s hospital, Nijmegen, Netherlands

Aim of the Study
Minimally invasive surgery is used more frequently for the correction of rare congenital malformations, such as congenital diaphragmatic hernia (CDH) and esophageal atresia (EA). Because these conditions are rare, these specific techniques are not used often. Moreover, they are difficult and require specific surgical skills. Because it important to master these skills, to avoid complications, the aim of this research is to develop readily available models to train these procedures outside the clinical setting.

Methods
The EoSim augmented reality laparoscopic simulator was used for this research, which is a relatively low cost and portable simulator, usable for training of pediatric procedures (including 3mm instruments). The aim was to develop three models to train the most important component tasks of the CDH and EA repairs. The most important feature was that it should be easy available and reuse.

Main Results
Model EA repair (Figure 1): two water balloons are fixated on the flat tray. The upper balloon is upright, resembling the pouch, and the lower one upside down (with the cuff cut off), resembling the ligated trachea-esophageal fistula.
Model CDH repair (Figure 2): The upper part of a size 8 sterile glove are cut of and pulled over a transparent cup, with the floor cut out. This is placed upright on the flat tray and fixed with an elastic band.

Conclusions
We have developed two low cost models for the training of difficult minimally invasive skills that can be made and used by everyone.

1 2
PW19-G08:
PEDIATRIC TRACHEOSTOMY: A COMPARISON OF OUTCOMES BETWEEN DIFFERENT INDICATIONS

Leonor Melero Guardia, Juan Luis Antón-Pacheco, Rocío Morante Valverde, Isabel Carrillo Arroyo, Belén Aneiros Castro, Jesús Vicente Redondo Sedano, Rubén Martín Alelu, María Isabel Benavent Gordo, Andrés Gómez
Hospital Universitario 12 de Octubre, Madrid, Spain

Aim of the Study
To define the mortality and long-term outcomes of children undergoing tracheostomy in our airway unit.

Methods
Retrospective and descriptive study of patients that required a tracheostomy between 2008 and 2017 (10 years). The following variables were studied: demographic data, age, indication, surgical technique, complications, decannulation and mortality rates. These rates were estimated according to the underlying tracheostomy indication by means of Chi Square test.

Main results
A total of 104 patients were included in the study. The median patient age was 2.8 years (range, 3 days-15 years). Cotton’s procedure was the most frequent tracheotomy technique (87.5%). Primary indications for tracheostomy were divided into five groups: (1) airway obstruction (n=64), (2) acquired respiratory disease (n=15), (3) congenital respiratory disease (n=5), (4) acquired neurologic disease (n=15), and (5) congenital neurologic disease (n=5). Overall decannulation rate was 49% with an average time until decannulation of 1.6 years. Decannulation rate for each of the 5 groups was: 68.6%, 63.6%, 0%; 50%; y 0%, respectively (p=0.0137). Most frequently detected complications were persistent tracheocutaneous fistula (14.4%), granuloma and suprastomal collapse (5.7%). Overall mortality rate was 15.3% and related to each indication group: 6.2%, 20%, 40%, 33.3% and 40%, respectively (p=0.0128). In only one patient mortality was due to the tracheostomy.

Conclusions
Tracheostomy is a relatively common procedure in our setting because of the specific features of our patients. Significant prognostic differences in mortality and decannulation rates exist based on the underlying indication of the tracheostomy.
Kathryn Ford¹, Hannah Thompson², Niyi Ade-Ajay³, Oliver Burdall⁴
¹John Radcliffe Hospital, Oxford, United Kingdom. ²Great Ormond Street Hospital, London, United Kingdom. ³Kings College Hospital, London, United Kingdom. ⁴Norfolk and Norwich University Hospital, Norwich, United Kingdom

Abstract

AIM: Simulation training is now a mainstay of surgical training but expense often limits the availability to trainees. We aimed to use household items to develop a home-made, low cost, low fidelity appendicectomy model and validate its efficacy across varied settings.

METHODS: An appendix model was designed, ‘kit list’, instructions (including video) produced and validated by a range of surgeons, from novice to senior registrar, in three cohorts (2016 – 2017): laparoscopic use in a (A) low- and (B) high- income setting and (C) open use in a high-income setting. Validation was based on delegate feedback using a Likert scale (1-10) regarding tactile feedback; complexity; realism and usefulness of the model. Data are presented as median (range).

MAIN RESULTS: Each model costs 0.62Euro to produce. 40 candidates in 3 cohorts (A: n=11; B: n=8; C:n=21) covering junior and senior trainees from high and low income settings attended the courses. All candidates would recommend the model and 90% (n=36) felt that the model was easily reproducible. All cohorts rated the tactile feedback and realism at a median of 6/10 and 5/10 respectively, for both laparoscopic and open techniques. The more junior trainee and low income cohorts rated the model as more useful (P=0.02; one-way ANOVA, F-value 4.1).

CONCLUSIONS: This easily reproducible model is unique and has a major role in training for junior trainees maximising learning with limited cases or for surgeons in low-resource settings.
LAPAROSCOPIC HERNIA REPAIR: EVALUATION OF THE IDES TECHNIQUE IN A PEDIATRIC POPULATION

Lucile Fievet¹, Salman Ghazwani¹, Claire Gallinet², Ahmad Fakhro³, Sophie Vermersch², Jean-Luc De Lucas¹, François Varlet², Aurelien Scalabre²
¹CH Henri Duffaut, Avignon, France. ²University hospital, Saint Etienne, France

**Aim of the study:** Recently several laparoscopic techniques have emerged for repair of inguinal hernia in children, the gold standard still being the open herniotomy. This study evaluates the inguinal dissect, excise and suture (IDES) technique which includes incision of the peritoneum at the internal ring, dissection of the vas and vessels, excision of the hernia sac and closure of the peritoneal defect.

**Methods:** This retrospective study includes all consecutive children operated on for inguinal hernia using the laparoscopic IDES technique in 2 centers from December 2015 to December 2017. Clinical data were collected from medical charts.

**Main results:** A total of 155 children were included, 76 females (49%) and 79 males (51%). The mean age at surgery was 55 months +/- 43. Asymptomatic contralateral hernias were found in 20% of cases during laparoscopy and were systematically treated. The mean operating time was 46 min [21;106] for unilateral cases, and 55 min [27;103] for bilateral cases. No intraoperative complications were reported. Clavien grade 1 complications occurred in 8 cases, mainly postoperative vomiting. 112 patients under 3 months old were discharged on the same day, and younger patients were usually discharged the next morning. All patients had a follow-up within a 6 weeks period after surgery. No recurrence was observed. In only one case recurrence was suspected but not confirmed during re-do procedure.

**Conclusions:** Laparoscopic inguinal hernia repair using the IDES technique appeared safe and effective in children, suited to ambulatory surgery.
ROLE OF ROUTINE ANALYTICAL STUDIES IN THE PREPARATORY OF PEDIATRIC SURGERY

Cristina Garces Visier\textsuperscript{1,2}, Cristina Bailen Ruiz\textsuperscript{1}, Jaime Rodriguez De Alarcon García\textsuperscript{1}, Eva Dominguez\textsuperscript{1}, Luis Felipe Ávila Ramirez\textsuperscript{1}, Carmen Soto Beauregard\textsuperscript{1}

\textsuperscript{1}hospital Universitario Clinico De San Carlos, Madrid, Spain. \textsuperscript{2}hospital Infantil Universitario Niño Jesus, Madrid, Spain

OBJECTIVES: To determine the value of preoperative tests for scheduled surgeries with general anesthesia.

MATERIAL AND METHODS: A multidisciplinary group was established to improve the surgical procedures that are carried out in pediatric surgery. The requirements of the analytical profiles were established in relation to age, surgical risk and type of surgery. Pre-operative blood tests were reviewed in pediatric patients who underwent surgery with general anesthesia from June 2015 to January 2017. The incidences detected were analyzed in relation to compliance with the agreed profiles (blood count, coagulation, glucose, creatinine and ions) as well as the values that were not normal and their possible influence on the perioperative management of the patient.

RESULTS: During the study period, 846 scheduled minor surgeries and 1185 blood tests were performed. 8.6% presented abnormal results. 19 children had hemoglobins <8.7mg/dl. 58 children presented platelet counts <50.000, which in 97% were false decreases due to platelet aggregation due to difficult extraction that were not confirmed in a second test. 92 presented hyponatremia that in 78% were due to inadequate technique. No result forced to suspend the surgery. No relationship was observed between the preoperative blood test alterations and postoperative complications nor could they have been prevented by said results.

CONCLUSIONS: The results suggest that in pediatric patients undergoing minor surgical procedures, the blood tests performed routinely in the preoperative period have a very limited value.
Aim of the study: In this study aimed to compare the results of subclavian (SCV) and internal jugular (IJV) veins for port catheterization.

Methods: We retrospectively analysed intravenous port catheterization records in our clinic. SCV punction was performed between 2009-2015, from 2015 to 2017 IJV punction was preferred. Surgical intervention complicatons and the revision reasons were addressed.

Main Results: 436 patients (249 M, 187 F) underwent port catheterization with a mean age of 6.4± 5.3 years (2 months - 20 years). The indications were malignancies (372 patients), chronic diseases (52 patients) and hematological diseases (12 patients). SCV was preferred for 308 patients, whereas for 128 patients IJV punction with ultrasound guidance was chosen. Seven patients had complications associated with surgical intervention in SCV group (2.2%). (Pneumothorax in three, two catheters and the reservoirs were disconnected and two had wound dehiscence). In IJV group, 2 patients had surgical intervention complications (1.5%). (One patient had a kinking of catheter, one patient’s catheter and the reservoir were disconnected. In total, 31 patients (7.1%) required port revision. Major causes of revision were resistant catheter infection (16 patients) and thrombosis obstruction (15 patients).

Conclusions: Permanent vascular access is mandatory, especially for malignancy, chronic and hematological diseases. SCV, due to its anatomical location may not always be suitable for ultrasound guidance. IJV seems as a safer option for intravenous catheterization under ultrasound guidance as it doesn’t carry the risk of pneumothorax. For intravenous port catheterization, internal jugular vein should be preferred in children requiring permanent vascular access.
Aim: To present the epidemiologic data of Turkish Esophageal Atresia Registry (TEAR)

Method: The data of TEAR between 2014-2017 were evaluated for gestational age, birth weight, height, associated anomalies, type of atresia, surgical treatment modalities and postoperative outcome. The data was obtained from electronic submissions of two forms including medical recordings of neonatal period and infancy from 23 pediatric surgery clinics in Turkey.

Results: Data of 305 cases with male-female ratio of 157:148 was evaluated. The mean gestational age was 36.07 weeks (26-42), birth weight 2372.7 grams (700-4500) and height 45.8 cm’s (30-56). Prenatal diagnosis was obtained in 29.7% of cases. The incidence of associated anomalies was 68.6% with cardiac anomalies (54.2%) being the most common. The Gross classification distribution were: C in 226 (73.9%), A in 51 (16.7%), B in 10 (3.3%), D in 9 (2.9%), and E in 8 (2.6%). Congenital stenosis was detected in 19 (6.2%). Primary anastomosis was achieved in 78.4% with thoracotomy (n=228) and thoracoscopy (n=12). Primary anastomosis could not be achieved in 20.3% with long-gap atresia. %61.4 were discharged from hospital on the day of discharge.
full oral feeds. At the end of one year, anastomotic stricture requiring dilatation was present in 34 cases (11.1%) and fistula recurrence in 1 case (0.3%). Antireflux surgery was performed in 5.2% of cases. The mortality rate was 19.3% in neonatal period and 1.5% at the end of first year.

**Conclusion:** TEAR allows us to evaluate the greatest epidemiological data on esophageal atresia in Turkey.
PERINATAL PREDICTIVE FACTORS OF SHORT-TERM MORBIDITY IN PATIENTS WITH OESOPHAGEAL ATRESIA IN A DEVELOPING COUNTRY

Sahla Sallemi¹, Saloua Ammar¹, Manar Hbaieb¹, Mohamed zouari¹, Hayet Zitouni², Mahdi Ben Dhaou¹, Riadh Mhiri¹, Amel Ben Hmed¹, Chiraz Rgaieg³, Afef Ben Thabet³, Abdellatif Gargouri¹
¹Department Of Pediatric Surgery, Hedi Chaker Hospital, Sfax, Tunisia. ²Department Of Pediatric Surgery, Hedi Chaker Hospital, Sfax, Tunisia. ³Department Of Neonatology, Hedi Chaker Hospital, Sfax, Tunisia

Aim of the study: To describe early postoperative outcomes in children with esophageal atresia (OA) and to identify early predictive factors (first month of life) of short-term complications (STC).

Methods:

Charts of children operated for OA in our department from 2007 to 2017 were reviewed. A short-term complicated evolution was defined by the occurrence during the first year of age of at least one of these complications: Esophageal stricture requiring dilatation, recurrent fistula, severe reflux or tracheomalacia, recurrent pneumopathy, prolonged feeding by gavage or parenteral nutrition or death. The statistical analysis was done by SPSS version 2.0.

Main results:

Forty-two cases were collected. Type III accounted for 76%. Thirty-nine patients underwent oesophageal anastomosis and 3 patients underwent gastrostomy with oesophagostomy. Twenty-three child (54%) developed STC. Ten patients (23%) died during hospitalization. Perinatal predictive variables of STC were: delayed diagnosis \(p = 0.03\), low birth weight \(p = 0.03\), and prolonged intubation \(> 4j\) \(p = 0.042\). The presence of VACTERL syndrome and long hospital stay weren’t significant factors of STC.

Conclusions:

The short-term prognosis of OA still reserved. The predictors of STC are delayed diagnosis, low birth weight, and prolonged intubation.

<table>
<thead>
<tr>
<th>Variables</th>
<th>STC Number (%)</th>
<th>No STC Number (%)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delayed diagnosis</td>
<td>12 (92%)</td>
<td>1 (7%)</td>
<td>0.03</td>
</tr>
<tr>
<td>Low birth weight</td>
<td>8 (100%)</td>
<td>0 (0%)</td>
<td>0.03</td>
</tr>
<tr>
<td>Prolonged intubation</td>
<td>14 (87%)</td>
<td>2 (12%)</td>
<td>0.041</td>
</tr>
<tr>
<td>VACTERL</td>
<td>3 (75%)</td>
<td>1 (25%)</td>
<td>1</td>
</tr>
<tr>
<td>Cardiopathie</td>
<td>8 (80%)</td>
<td>2 (20%)</td>
<td>0.45</td>
</tr>
<tr>
<td>Hospital stay&gt;30 days</td>
<td>5 (71%)</td>
<td>2 (28%)</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 1: Predictors of STC
THE USE OF AUTOPERICARDIUM IN SURGICAL TREATMENT OF TRACHEOMALACIA IN CHILDREN

Danylo Krivchenya¹, Alexander Dubrovin¹, Eugene Rudenko¹,², Vasyl Prytula¹,²
¹Bogomolets National Medical University, Kyiv, Ukraine. ²National Specialized Children Hospital "OKHMATDYT", Kyiv, Ukraine

Aim of the Study was to optimize surgical treatment of tracheomalacia (TM) with variable use of autopericardium in children.

Methods. Seventy-three patients aged from 2 weeks to 16 years (mean 9.6 months) operated on for idiopathic TM (n=17, 23.3%), TM associated with esophageal atresia (n=37, 50.7%) or innominate artery compression (n=19, 26.0%) were selected for the study. TM surgery included aortopexy, tracheoplasty or combination of both procedures. Autopericardium was used for TM surgery in 47 (64.4%) patients (group I) as pericardial flaps for aortopexy (n=31) or free native or modified with glutaraldehyde pericardial patch for tracheoplasty (n=16). In 11 of these cases, aortopexy and tracheoplasty were performed simultaneously.

In 26 (35.6%) cases (group II) different methods of aortopexy or tracheoplasty were used. Aorta was sutured to the sternum and trachea was reinforced (n=17) with siliconized rubber disks or patches of fascial-pleural tissue or preserved pericardium. Combination of both aortopexy and tracheoplasty in this group was performed in 4 cases.

Main results. Total of 69 (94.5%) patients survived, 63 (91.3%) remained asymptomatic at the late follow-up. Total mortality was 4 (5.5%), including 2 deaths in the I group (4.3%) and 2 deaths in the II group (7.7%), p=0.89. Complication rates in I and II groups were 8.5% (n=4) vs 23.1% (n=6), respectively, p=0.52. There were no pericardium-related complications.

Conclusions. Autopericardial flap aortopexy is expedient for tracheomalacia due to innominate artery compression. In idiopathic and "esophageal" tracheomalacia with an unstable trachea tracheoplasty with modified autopericardium and simultaneous aortopexy are indicated.
THE ROLE OF FLEXIBLE BRONCHOSCOPY ACCOMPLISHED THROUGH A LARYNGEAL MASK AIRWAY IN THE TREATMENT OF TRACHEABRONCHIAL FOREIGN BODIES

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1mugla Sitki Kocman University Research And Training Hospital Department Of Pediatric Surgery, Mugla, Turkey. 2mugla Sitki Kocman University Faculty Of Medicine Department Of Pediatric Surgery, Mugla, Turkey

Aim
Rigid bronchoscopy is considered to be the standard way for extraction of tracheobronchial foreign bodies (TBFBs). Although flexible bronchoscopy is considered to be a diagnostic tool, we here present our experience with flexible bronchoscopy as a therapeutic device for TBFBs in children.

Methods
A retrospective study included 19 patients underwent flexible bronchoscopy under general anesthesia using a laryngeal mask airway (LMA) between July 2016 and January 2018 for suspicion of TBFBs.

Results
Demographic characteristics, preoperative signs, symptoms and radiologic findings of patients are given in Table 1. Twelve (63.2%) patients were found to have TBFBs. All procedures were successfully accomplished through LMA by using basket forceps (75%), a Fogarty catheter (16.7%) and a suction (8.3%). 9 (75%) of TBFB locations were right bronchial, 2 (16.7%) were left bronchial and 1 (8.3%) were tracheal. The types of extracted TBFBs were organic in 11 (91.7%) and non-organic in 1 (8.3%). There were no complications except laryngeal edema noted in 2 (16.7%) patients and relieved within 48 hours. The mean time of postoperative hospitalisation was 2.84±0.76 (1-4) days.

Conclusion
Flexible bronchoscopy accomplished through a LMA is a safe and effective technique in retrieving TBFBs in children with favorable success and low complication rates.

Table 1

<table>
<thead>
<tr>
<th>Table 1</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mean age, years</strong></td>
</tr>
<tr>
<td>Wheezing</td>
</tr>
<tr>
<td>Cough</td>
</tr>
<tr>
<td>Dyspnea</td>
</tr>
<tr>
<td><strong>Mean duration of hospitalisation, hours</strong></td>
</tr>
<tr>
<td><strong>Physical examination</strong></td>
</tr>
<tr>
<td>Normal</td>
</tr>
<tr>
<td>Abnormal</td>
</tr>
<tr>
<td><strong>Chest X-ray</strong></td>
</tr>
<tr>
<td>Normal</td>
</tr>
<tr>
<td>Abnormal</td>
</tr>
</tbody>
</table>
EARLY VIDEO ASSISTED THORACOSCOPIC (VATS) PLEURODESIS AND BLEBECTOMY - THE OPTIMAL TREATMENT FOR SIGNIFICANT PRIMARY SPONTANEOUS PNEUMOTHORAX IN CHILDREN

Samuel Davies¹, Anna Wells¹, Alan James², Rajendra Kumar¹, Devesh Kaushal²
¹John Hunter Childrens Hospital, Newcastle, Australia. ²John Hunter Hospital, Newcastle, Australia

Background

The current management of primary spontaneous pneumothorax (PSP) in children is not standardized. This study aims to formulate optimal surgical treatment of PSP in children.

Methods

A retrospective review of consecutive children (< 19 years of age) presenting to a single tertiary hospital with PSP between 2002 and 2017. Moderate and large PSP at presentation were considered significant.

Outcomes of children undergoing initial non-operative treatment versus video-assisted thoracoscopic (VATS) pleurodesis with blebectomy were compared.

Results

61 children with PSP included 50 males (82%) and 11 females (18%), median age at presentation 16.35 (57%) had left sided PSP and 26 right sided (43%). The pneumothorax was small in 10 (16%) and significant in 51 children (84%).

44 children (72%) were initially managed non-operatively. This group included 10 with small and 34 with significant PSP. At follow up only one child with small pneumothorax had recurrence compared to 24 with significant pneumothorax (71%).

17 children (38%) with significant large pneumothorax at presentation required a VATS with blebectomy and pleurodesis. None of these patients required further surgery.

The mean operative time for VATS was 52.8 minutes (range 32 to 80 minutes). The mean duration for chest tube was 3.9 days (range 2-5 days).

At follow up (median 7 years) Post VATS, late recurrence was noted in 3 (7%) and contralateral pneumothorax 13 (21%).

Conclusions

Primary VATS with blebectomy is the optimal management for all significant PSP at first presentation. Non-operative treatment should be reserved for small PSP.
FIBRINOLYSIS WITH UROKINASE AS FIRST LINE TREATMENT FOR PEDIATRIC PLEURAL EMPyEMA: IS THERE ANY FACTOR ADVISING FOR AN INITIAL VATS?

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Hospital Universitary Vall d’Hebron, Barcelona, Spain

INTRODUCTION
Management of pleural empyema in children is still controversial. Marhuenda published in 2014 the results of a prospective, randomized multicentric trial (2008-2010) comparing VATS versus chest drain and fibrinolysis with urokinase (UK) concluding that there were no differences. The aim of the study was to evaluate effectiveness and efficiency of UK protocol and to identify any possible bad prognostic factor that would make initial VATS advisable.

METHODS
Single center retrospective review (2011-2018) of patients treated with protocol of thoracic drainage + 6 doses of UK as first line treatment. Data regarding symptoms, complications, requirement of second surgical procedures (VATS), forecast factors and outcome were collected.

RESULTS
A total of 85 patients were enrolled in this period. Mean age was 4.8 years (2m-17y). Twenty-one patients (24.7%) presented complicated pneumonia (7 necrotizing pneumonias, 5 bronchopulmonar fistula, 9 residual fluid collections) but only 8 (9.4%) required a second surgical procedure (VATS). None required a third intervention or a decortication by thoracotomy. Median hospital stay was 10 days (4-36). There were no statistical differences in prognostic factors such as comorbidities, sonographic characteristics or microbiological cultures (multiple logistic regression). Six months after the pneumonia all the cases had a normal x-ray. We didn’t observe complications related to the use of urokinase.

CONCLUSION
In our experience, fibrinolytic therapy is still recommended as first line treatment for pleural empyema. VATS should be reserved for patients with poor response or complications, as we did not find any prognostic factor that could advise for an initial surgery.
THORACOSCOPIC RESECTION OF CONGENITAL PULMONARY AIRWAY MALFORMATION: DOES AGE MATTER?

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Introduction
The rate of antenatal detection for congenital pulmonary airway malformation (CPAM) has increased in the last decade and many centres would recommend excision to prevent potential complications. In the same period thoracoscopic resection has gained greater attention. However, the optimal timing to perform resection is still controversial. In this study we reviewed our experience after 105 operations performed and attempted to see if the age of surgery played a difference in terms of intra and peri-operative outcomes.

Methods
All patients who underwent thoracoscopic resection for CPAM in a tertiary referral centre were recruited into the study. Patients’ demographics, peri-operative data and post-operative outcomes were analyzed.

Results
105 consecutive patients were identified in the study period, 52 males and 53 females. The mean age and mean body weight at operation was 14.9 ± 2.3 months and 9.75 ± 0.8 kg respectively. 96 patients (91.4%) were detected antenatally by ultrasound. 21 patients experienced chest infection before the operation (20%). The mean operative time was 141.0 ± 8.5 minutes. 15 patients required conversion to open thoracotomy (14.3%). The mean hospital stay was 4.82 ± 0.3 days. There was no mortality. Sub-group analysis showed that if operation was performed before 1 year old the operative time (130 vs 173 minutes, p=0.02), blood loss (29.3 vs 192.8 ml, p=0.04) and intensive care duration (0.9 vs 1.6 days, p=0.02) were all significantly less.

Conclusion
Thoracoscopic resection of CPAM is safe and effective. Late resection after 1 year old may increase operative difficulties.
PW20-TH08
VIDEO-ASSISTED THORACOSCOPY SUPERIOR TO CHEST TUBE DRAINAGE AS A PRIMARY TREATMENT FOR CHILDHOOD EMPYEMA

Anna Mäkinen1, Janne Suominen1,2, Jukka Salminen1,2
1University of Helsinki, Helsinki, Finland. 2Department of Pediatric Surgery, Helsinki Children’s Hospital, Helsinki, Finland

Aim of the study
The incidence of empyema in children is increasing, yet the preferable first-line treatment remains controversial. The aim of this study was to compare video-assisted thoracoscopy (VATS) and chest tube drainage as a primary treatment for childhood empyema.

Methods
All the children diagnosed with empyema as a complication to pneumonia in our tertiary care hospital during 2001-2016 were included, resulting in 50 patients receiving either VATS (25 patients) or chest tube drainage (25 patients) as a first-line treatment. The groups were similar in terms of age, gender and hospital admission from the onset of symptoms. The Mann-Whitney U test and Fischer’s t test were used for the analysis.

Main Results
Compared to the chest tube group, patients receiving VATS as a primary treatment had a shorter hospital stay (11 days vs. 18 days, p=0.006), a lower total cost (15747e vs. 25037e, p=0.0011), and a lower reoperation rate (20% vs. 88%, p=0.0001). There was no statistically significant difference in the duration of antibiotic treatment (19 d vs. 22.5 d, p=0.076). Patients in the chest tube group had a higher complication rate (28% vs. 56%, p=0.0845), yet this did not reach statistical significance.

Conclusions
Early VATS reduces the length of hospital stay and decreases total cost of care compared with chest tube drainage.

<table>
<thead>
<tr>
<th></th>
<th>VATS</th>
<th>Chest tube</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of subjects</td>
<td>25</td>
<td>25</td>
<td></td>
</tr>
<tr>
<td>Age (y)</td>
<td>5 [1-15.8]</td>
<td>3.5 [0.7-13.9]</td>
<td>0.3416</td>
</tr>
<tr>
<td>Gender [M/F]</td>
<td>14:11</td>
<td>12:13</td>
<td>0.7775</td>
</tr>
<tr>
<td>Complication * rate</td>
<td>7/25 (28%)</td>
<td>14/25 (56%)</td>
<td>0.0845</td>
</tr>
<tr>
<td>Reoperation rate</td>
<td>5/25 (20%)</td>
<td>22/25 (88%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Hospital admission from the onset of symptoms (d)</td>
<td>5 [0-15]</td>
<td>4.5 [0-15]</td>
<td>0.6349</td>
</tr>
<tr>
<td>Intervention from hospital admission (d)</td>
<td>2 [0-8]</td>
<td>3 [0-13]</td>
<td>0.4580</td>
</tr>
<tr>
<td>Length of hospital stay after intervention (d)</td>
<td>7 [4-12]</td>
<td>11 [4-40]</td>
<td>0.0002</td>
</tr>
<tr>
<td>Length of hospital stay (d)</td>
<td>11 [6-20]</td>
<td>18 [5-37]</td>
<td>0.0006</td>
</tr>
<tr>
<td>ICU length of stay (d)</td>
<td>0 [0-4]</td>
<td>0 [0-17]</td>
<td>0.0134</td>
</tr>
<tr>
<td>Duration of antibiotic treatment (d)</td>
<td>19 [10-29]</td>
<td>22.5 [14-49]</td>
<td>0.0760</td>
</tr>
<tr>
<td>Duration of chest tube treatment (d)</td>
<td>5 [3-10]</td>
<td>8.5 [3-23]</td>
<td>0.0011</td>
</tr>
<tr>
<td>Total cost (e)</td>
<td>15747 (8932-26889)</td>
<td>25037 (6474-108375)</td>
<td>0.0011</td>
</tr>
</tbody>
</table>

Values are expressed as median and range.

*Complications include admission to the ICU, pneumothorax requiring intervention, bronchopleural fistula, need for urgent bronchoscopy, pus leakage from thoracostomy wound and blood aspiration during chest tube thoracostomy followed by emergency CPR.
PW20-TH09
VATS APPROACH FOR THORACIC BRONCHOGENIC CYST IN CHILDREN: A GOLD STANDART

YAQOUB JAFAR1, FREDERIC HAMEURY2, aurélien scalabre3, PIERRE YVES MURE2, YOHAN ROBERT1, PIERRE Yves Rabattu1, Remi Dubois2, Francois Varlet3, Christian Piolat1
1centre Hospitalier Universitaire De Grenoble, Grenoble, France. 2hôpital Femme Mère Enfant , Hospices Civil De Lyon, Lyon, France. 3centre Hospitalier De Saint Etienne, Saint Etienne, France

Background:

Bronchogenic cyst (BC) represents a rare type of bronchial tree malformation sometimes detected by prenatal ultrasounds. There is no general consensus on the approach and timing of intervention for BC. However, it’s advocated by many pediatric surgeons to resect it early in life. Aim of this study is evaluating video assisted thoracic surgery (VATS) as first line approach for BC management in the largest pediatric published series, in our knowledge.

Material and methods:

We have performed a retrospective (2007-2017) multi-center study from three French referral centers. We included all pediatric patients (< 18 years) operated by VATS and thoracotomy from well documented (histopathology examination) thoracic BC.

Results:

Twenty four patients had been operated for BC. The median age for the patients that was operated was 45 months (2 months -16 years). The median size of the cyst that was resected was 27mm in its greatest diameter (range 15mm-55mm). Four conversions to open thoracotomy occurred because of bad exposure. One patient operated directly by thoracotomy because of hemodynamic instability. The median hospital stay was 3. Days (range 1-7 days). There was no mortality or major morbidity post operatively. During follow up, there was no long term recurrence or major complication.

Conclusion:

VATS is considered feasible and safe for BC in experienced hands in children. The size and location of the BC does not affect the decision to select this approach. For many advantages, VATS can be considered as an effective first line management for BC.
Aim of the study:
To understand the current practice of thoracoscopic approach for treating type C esophageal atresia and to report short and midterm outcomes of this technique.

Methods:
A detailed survey was sent to 6 Italian institutions, performing advanced minimally invasive surgery concerning about exclusion criteria applied, operative details, postoperative management and outcomes of this approach.

Results:
57 cases of Type C esophageal atresia were operated thoracoscopically in the period from 2009 to 2017. The mean weight was 2.69 kg (+/- 0.48), mean operative time was 197 minutes (+/- 71), mean length of hospitalization was 21.2 days (+/- 15.7). 8 patients (14%) developed early leak, which managed conservatively. 21 patients (37%) developed esophageal stricture, half of them required less than 4 sessions of endoscopic dilatation. 2 cases had recurrent tracheoesophageal fistula which operated through open thoracotomy approach within 3 months. The procedure was successfully completed in 45 babies, while 9 cases were converted to open approach. Staged operation by ligating the fistula only as a 1st staged was applied in 3 cases due to severe prematurity and the 2nd stage was performed through thoracoscopic approach in one case and open approach in the others. 6 cases (10.5%) required fundoplication for severe GERD and 5 patients (8.8%) operated for severe tracheomalacia by aortopexy procedure.

Conclusions:
Thoracoscopic repair of Type C esophageal atresia can be safely performed by experienced endoscopic pediatric surgeons. Outcomes are comparable to those of open procedure. Careful selection of cases and standardizing the technique could improve the outcome.
MINIMALLY INVASIVE TREATMENT OF HYDATID CYST OF THE LUNG IN CHILDREN

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1Military Hospital, Oran, Algeria. 2Military Hospital, Algiers, Algeria. 3Military Hospital, Algiers, Algeria

The videothoracoscopic approach of lung hydatid cyst in children obeys the same rules conventional surgery. This technique was long controversial due to the risk of hydatid dissemination during surgery and the difficulty of closure of bronchial pleural fistulas. Following A review of the literature shows that only a few publications have been reported in relation with children, concluding in the feasibility of the technique for small cysts only.

The purpose of our work is to define the place of videothoracoscopy in the treatment of pulmonary hydatid disease of children.

MATERIALS AND METHODS:
This is a prospective study over a period of 3 years, involving 25 patients, operated by surgical videothoracoscopy. We have studied the epidemiological characteristics of the patients, the clinical and para-clinical data, as well as the per operating data, and the follow-up after surgery.

RESULTS:
The average age of our patients was 7.15 ± 3.14 years, and the sex ratio was 1.55. The average operative time was 115.9 ± 50 minutes. We recorded no conversion. The postoperative course was, complicated with a quick resolutive subcutaneous emphysema in 4 interventions. The average duration of postoperative hospitalization of our patients was 3.13 ± 1 day. No recurrences in our series after follow-up of 18 to 48 months.

CONCLUSION:
The videothoracoscopy is a feasible safely technique that undoubtedly has a place in the surgical treatment of pulmonary hydatid cyst in children.

Keywords:
Videothoracoscopy, child, lung, hydatid cyst.
A HYBRID TECHNIQUE IN PATIENTS WITH EXTREME HALLER INDEX: STERNAL ELEVATION WITH PECTUS-UP BEFORE INSERTION OF THE PECTUS BAR

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¹Pediatric Surgery Department, Corporació Sanitaria Parc Taulí, Sabadell (Barcelona), Spain. ²Pediatrics Department, Corporació Sanitaria Parc Taulí, Sabadell (Barcelona), Spain. ³Institut d’Investigació i Innovació Parc Taulí (I3PT), Fundació Parc Taulí, Sabadell (Barcelona), Spain

Abstract

Aim: Cardiac puncture and aortic injury have been described in patients with severe chest wall defects undergoing minimally invasive repair of pectus excavatum (MIRPE). We present an easy technique using the screw of the Taulinoplasty (Pectus Up technique) to elevate the sternum before passing the pectus bar, improving visualization and facilitating the placement of substernal bars with minimal morbidity during MIRPE.

Methods: In patients with severe Haller index and Correction Index there is no space between sternum and heart. Although this cardiac compression could be assessed thoracoscopically, the dissection while inserting the introducer (sword) could be very dangerous. In a supine position and arms tucked to the sides, a single 1 cm incision is made over the sternal defect. A traction screw is placed into the sternum at the middle of the defect and with a retractor plate the sternum is elevated (Taulinoplasty technique). After the elevation of the defect and under thoracoscopic vision of the anterior mediastinum a safe dissection across to the left thorax can be performed.

Results: A total of 5 patients underwent this hybrid technique, achieving a complete correction of the sternal defect and significant reduction of the Haller and Correction index before MIRPE without life-threatening complications.

Conclusion: After acquiring some experience with the Pectus-up technique, we consider this a safe and reproducible procedure complementing the minimally invasive repair for extreme Pectus Excavatum reducing its potential fatal risks inherent to the technique.
pw21-ur01
Correlation of Anorectal Malformation Severity and Associated Urologic Abnormalities: A Review of 329 Patients

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1Nationwide Children's Hospital, Columbus, USA. 2Ohio State University, Columbus, USA. 3Hospital Universitario La Paz, Madrid, Spain

Aim of the study: To determine the incidence of urologic diagnoses in children with anorectal malformation (ARM).

Methods: An IRB approved database of children with ARM was reviewed. Urologic diagnoses were tabulated. ARM types were defined by distal rectal anatomy.

Main results: Of 329 patients, 50% were male and 50% were female. 46.9% of females with ARM had one or more urologic diagnosis which trended upwards with more severe ARM type. 84.6% of long common channel cloacas (>3 cm) had one or more urologic diagnosis. 60.5% of males had one or more urologic diagnosis, which trended upwards with height of malformation.

Conclusions: Urologic anomalies are highly prevalent in children with ARM and more severe ARM seem to have higher incidence of urologic diagnoses. This data stresses the importance of proper ARM screening and for having a pediatric urology specialist involved in the management of such children.

Table 1:

<table>
<thead>
<tr>
<th>Type of Female Fistula</th>
<th>Type of Male Fistula</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perineal n=38</td>
<td>Perineal n=37</td>
</tr>
<tr>
<td>Vestibular n=54</td>
<td>Bulbar n=56</td>
</tr>
<tr>
<td>Vaginal n=16</td>
<td>Prostatic n=48</td>
</tr>
<tr>
<td>Cloaca &lt; 3 cm n=28</td>
<td>Bladder Neck n=26</td>
</tr>
<tr>
<td>Cloaca &gt; 3 cm n=26</td>
<td>All Male ARM n=167</td>
</tr>
<tr>
<td>All Female ARM n=162</td>
<td></td>
</tr>
<tr>
<td>Hydro</td>
<td>Hydro</td>
</tr>
<tr>
<td>13.16%</td>
<td>20.37%</td>
</tr>
<tr>
<td>6.25%</td>
<td>35.71%</td>
</tr>
<tr>
<td>35.71%</td>
<td>69.23%</td>
</tr>
<tr>
<td>Cloaca &lt; 3 cm</td>
<td>VUR</td>
</tr>
<tr>
<td>27.78%</td>
<td>52.00%</td>
</tr>
<tr>
<td>25.00%</td>
<td>45.83%</td>
</tr>
<tr>
<td>46.15%</td>
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<tr>
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PW21-UR02
URINARY TRACT INFECTIONS AFTER VOIDING CYSTOURETHROGRAPHY: IS ANTIBIOTIC PROPHYLAXIS NECESSARY?

Pilar Guillén, Manuel Espinoza, Francesco Ecclesia, Marta De Lucio, Cristina Garcés, Henar Souto, Ana Luis Huertas, Cristina Riñón, Rafael Arteaga
Hospital Infantil Universitario Niño Jesús, Madrid, Spain

AIM OF STUDY: To assess the frequency of urinary tract infection (UTI) in children following voiding cystourethrography (VCUG) in our centre.

METHODS: Retrospective study of patients who underwent VCUG between December 2015 and November 2017. Demographic, clinical, microbiological and radiological data were recorded. Post-VCUG UTI was defined as a clinically compatible case confirmed by positive urine culture within the following 10 days.

MAIN RESULTS: We reviewed 256 patients, 145 females and 111 males, with a mean age of 5 years. The main test indications were febrile UTI in 118 (46%) and vesicoureteral reflux (VUR) in 61 (24%). 177 patients (69%) received antimicrobial prophylaxis; in the remaining 79 patients a urine culture was obtained 6 days before. Among the 99 abnormal VCUG, the most frequent diagnosis was VUR (26.5%), followed by neurogenic bladder (6%). Post-VCUG UTI was diagnosed in six patients (2.34%), 3 males and 3 females, with a median age of 27 months [3-120]. Four of them presented radiological abnormalities (3 VUR and 1 ureterohydronephrosis) and five had received antimicrobial prophylaxis. The isolated germs in urine cultures were Escherichia coli (2/6), Klebsiella (2/6), Enterobacter (1/6) and Pseudomonas (1/6). There was no statistically significant difference in UTI frequency between prophylaxis and prior culture (p =0.66).

CONCLUSIONS: The frequency of UTI following VCUG is low, with a higher rate of atypical etiological germs. A prior urine culture might be a useful and safe alternative to prophylaxis for the prevention of post-VCUG UTI.
PW21-UR03
RITUAL CIRCUMCISION: A HAPPY EVENT BUT NOT ALWAYS

Mohamed Amine MSEDDI, Anis MASMOUDI, Sahla SEILLAMI, Nouri REBAI, Walid SMAOUI, Mohamed FOURATI, Mohamed JALLOULI, Mahdi BEN DHAOU, Mourad HAJSLEMEN, Riadh MHIRI, Nabil MHIRI
1Urology department of Habib Bourguiba Hospital, Sfax, Tunisia. 2Pediatric surgery department of Hedi Chaker Hospital, Sfax, Tunisia

Aim of the study:

Circumcision is a ceremonial and ritual act practiced in some religions, but this procedure is not harmless. Serious complications can be observed such as glandular amputation, which is considered as a family tragedy. The Purpose of this study was to describe clinical features and treatment difficulties associated with glandular amputation during circumcision.

Methods:

We report a retrospective series of 15 patients who suffered of glandular amputation during circumcision performed within traditional methods out of hospital settings.

Main results:

The mean age of those patients was 5 years (1 to 8 years). The average delay for consultation was 2.4 days. Glandular amputations consisted on partial section (6 cases) and complete (3 cases). In the other cases, it was about complete glandular necrosis caused by inadequate use for an electric device. Surgical treatment consisted on neomeatoplasty in 73%, 3% of cases. Four cases were treated by glandular reimplantation which failed in 2 cases unfortunately.

Conclusion

Glandular amputation is a severe and dramatic complication that remains difficult to manage. Prevention is required, which goes through performing this procedure by urologists or pediatric surgeon in order to limit the risk of potential complications of this gesture apparently inoffensive.
Aida Daib, Dorra Tarchella, Malak Boughdir, Youssef Hellal, Rabiaa Ben Abdallah, Riadh Ben Malek, Youssef Gharbi, Najib Kaabar
Hbib Thameur Hospital, Tunis, Tunisia

Aim of the study: To evaluate the impact of drainage technique in pyeloplasty in children, comparing double J stent to uretero-pyelostomy.

Methods: We did a retrospective study including 80 children who underwent pyeloplasty according to AndersonHeynes as treatment of a ureteropelvic junction obstruction between 2011 and 2016 in our department. They were divided into two groups. In group 1 a double J stent was inserted after pyeloplasty and in group 2 an external ureteropelvic stent was inserted. The uretero-pyelostomy was removed 7 days after surgery, while double J stent was removed under general anesthesia 4 to 6 weeks postoperatively. We compared operative time, hospital stay, intra and postoperative complications.

Main results: The mean age at surgery was 23.42 months in double J stent group and 38.81 months in uretero-pyelostomy group. Mean operative time was similar in the two groups (63.4 minutes in group 1 versus 75 minutes in group 2). No intraoperative complications were found during the placement of stent. Mean hospital stay was 3.8 days in group 1 versus 10.16 days in group 2. In postoperative followup, we reported 9 complications in group 1 (urinary tract infection, calculus and migration of stent) and 5 complications in group 2 (urinary leakage and stenosis). The median followup period was 4.24 years.

Conclusions: In children, the outcome of uretero-pyelostomy in pyeloplasty compared to double J stent placement is similar. The double J stent is associated with a shorter hospital stay. However, a second general anesthesia is needed to remove it.

Figure 1: double J stent complicated by calculus

Table 1: outcome and complications depending on the drainage technique:
<table>
<thead>
<tr>
<th></th>
<th>Double J stent</th>
<th>Uretero-Pyelostomy</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>No patients</td>
<td>50</td>
<td>30</td>
<td>/</td>
</tr>
<tr>
<td>Mean operative time (min)</td>
<td>63.4</td>
<td>75</td>
<td>0.99</td>
</tr>
<tr>
<td>Mean hospital stay (days)</td>
<td>3.8</td>
<td>10.16</td>
<td>4.9 e10-9</td>
</tr>
<tr>
<td>Complications</td>
<td>9</td>
<td>5</td>
<td>0.12</td>
</tr>
<tr>
<td>Follow-up (years)</td>
<td>4.24</td>
<td>4.24</td>
<td>/</td>
</tr>
<tr>
<td>Success rate (%)</td>
<td>100%</td>
<td>93.33%</td>
<td>0.98</td>
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PW21-UR05
OUTCOMES OF TRANSPERITONEAL LAPAROSCOPIC HEMINEPHRECTOMY IN DUPLEX KIDNEYS IN INFANTS AND CHILDREN

Delphine Demède\textsuperscript{1}, Pierre-Yves Rabattu\textsuperscript{2}, Jacques Birraux\textsuperscript{3}, Pierre DE Mouriquand\textsuperscript{1}, Pierre-Yves Mure\textsuperscript{1}
\textsuperscript{1}HFME, Lyon, France. \textsuperscript{2}HCE, Grenoble, France. \textsuperscript{3}HUG, Genève, Switzerland

Aim of the Study: To report the feasibility, safety and results of transperitoneal laparoscopic heminephrectomy (TLHN) for no or poor functional moiety in duplex kidneys.

Methods: Sixty-twow TLHN (50 upper pole and 12 lower-pole nephrectomies) were retrospectively reviewed in a single-institution. Median age at surgery was 35 months (range 7-204). The mean follow up was 13 months (range 2-56) and was based on clinical review, renal ultrasound, and nuclear investigation in a subgroup of 27 patients. Per and postoperative morbidity was studied.

Results: TLHN was feasible in all patients. No conversion was needed. The median operating time was 120 min (range 70-240), and the median hospital stay was 2.5 days (range 1- 29). We had no significant bleeding or digestive injuries. Three complications were observed: one persistent secreting moiety, one ureteral injury requiring open surgical repair, and one abscess of the nephrectomy space. Postoperative nuclear renographies showed no significant loss of function of the remaining moiety with a median difference of 2% (range 0-8%).

Conclusion: TLHN in duplex kidneys appeared to be a safe and effective procedure even in small children. Advantages compared to open surgery are a shorter hospital stay and a better cosmetic result. When compared to retroperitoneal approach, TLHN seems to provide a smaller conversion rate maybe due to a better operating space especially in infants. No digestive injury was observed. Furthermore, results are similar with no significant loss of function and no major complication.
XANTHOGRANULOMATOUS PYELONEPHRITIS IN CHILDREN: A 24-CASE SERIES

Mohamed Amine MSEDID1, Anis MASMOUDI1, Sahla SELLAMI2, Nouri REBAI1, Walid SMAOUI1, Mohamed FOURATI1, Mohamed JALLOULI1, Mahdi BE? DHAOU2, Mourad HAJSLIMEN1, Riadh MHIRI2, Nabil MHIRI1

1Urology Department Of Habib Bourguiba Hospital, Sfax, Tunisia. 2Pediatric Surgery Department Of Hedi Chaker Hospital, Sfax, Tunisia

Aim of the study:

Xanthogranulomatous pyelonephritis (XGP) is a non-specific chronic inflammation of the kidney which is extremely rare in children. Chronic renal infection usually associated to urolithiasis is the main cause. Throughout our experience of XGP in children, were studied differential diagnosis, surgical treatment and outcome.

Methods:
We reviewed retrospectively analysis of clinical records of 24 children who underwent nephrectomy for XGP among 114 nephrectomises performed in children (21%), between 1985 and 2017.

Main results:
The mean age was 8 years (ranges between 4 to 16 years). The commonest clinical presentation was lumbar pain (66,6%), lumbar mass (54%), fever (41,6%), weight loss (50%) and hematuria (12,5%). Biological abnormalities noted were leukocytosis (66,6%), anemia (54%) and pyuria (87,5%). Urine culture identified Proteus mirabilis (in 10 cases), Esherichia Coli (5 cases), Klebsiella (2 cases), morganella (1 case). IVP and isotopic studies showed a non-functioning kidney in 23 cases (95,8%) and renal stones in 18 cases (75%). Diagnosis was made post-operatively in all cases by pathology examination. Treatment consisted of antibiotic therapy followed by nephrectomy. Only one case of a focal XGP underwent conservative treatment. One infant died post-operatively due to septicemia and acute renal failure complications.

Conclusion:
The clinical features of the XGP remain not specific. The definitive diagnosis is done by histological examination. Total nephrectomy is the treatment of choice for the diffuse form, whereas partial nephrectomy may be curative for focal disease if the remaining kidney is functional.
 WHICH PSYCHOMETRIC EVALUATION FOR NEUROGENIC BLADDER AND BOWEL DYSFUNCTION IN CHILDREN WITH SPINA BIFIDA?

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¹Hospices Civils de Lyon, Pediatric Surgery unit, Lyon, France. ²Hospices Civils de Lyon, Neurosurgery Unit, Lyon, France. ³Hospices Civils de Lyon, Pediatric Surgery, Lyon, France. ⁴CHU Rennes, pediatric readaptation, Rennes, France. ⁵Hospices Civils de Lyon, pediatric readaptation unit, Lyon, France

Introduction: In patients with Spina Bifida (SB), main goals of elimination disorders treatment are clear but achievement is complex, due to a wide range of impairments, social and environmental factors. We elaborated a global score (MAFUR) to situate the child on his therapeutic path, with motricity, independence, fecal, urinary and renal scales. After the content validity we already started, one of the next step will be the criterion validity against Gold Standards. The aim of this study is to definite the gold standards in elimination disorders classifications in SB.

Method: our multidisciplinary team performed a Pubmed research of scores, scales and classifications validated and used in SB elimination disorders evaluation.

Results: Based on the ICF-CY (International classification of functioning, disability and health: children & youth version) we elaborated a model of main interactive factors that interfere in SB elimination disorders. A classification of spinal disorders was based on two published reports. The ASIA score gave the functional level. The ISI-P (Incontinence Symptom Index-Paediatric) and the Hostility Score (urodynamic) are specific of SB bladder elimination disorders. The NBDS (Neurogenic Bowel Dysfunction Score) has a paediatric validation, and the A-FICS (Adolescent Fecal Incontinence and Constipation Symptom Index) was validated for adolescents with SB. The HOFFER score allow functional ambulation evaluation in SB patients. The WeeFIM is a functional independence score for children.

Conclusion:
Despite the need for comparative studies, these psychometric evaluations are barely used in every day care and publications. This research emphasise the need for a specific global score like MAFUR.
Aim of the study: There are various clinical approaches in non-palpable testis management. We aimed to present our results of 152 children.

Methods: Medical records of patients who were operated for unilateral non-palpable testis in four pediatric surgery units between January 2008 and June 2017 were retrospectively reviewed. Demographics, findings of physical examination, ultrasonography, and surgical exploration were evaluated. A criterion was not evaluated if missing data was higher than 50%.

Main results: There were 152 patients with a median age of 22.4 months (6-180 months). The left side was predominant (65.7%). Records lacked data for presence or absence of nubbin, scrotal hypoplasia and contralateral hypertrophy in 16.4%, 80.3% and 39.5% of patients, respectively. Physical examination revealed nubbin in 48.8% and hypertrophy on the contralateral side in 25% of the patients. Majority of the explorations were laparoscopic (81.1%) whereas inguinal (18.1%) and scrotal (0.8%) exploration were also performed. Palpation of nubbin had a positive predictive value (PPV) of 78.2% in the inguinoscrotal region. Testes were normal in 16.1% of patients with palpated nubbin. PPV of contralateral testicular hypertrophy was 82.6%. Ultrasonography could detect 70.6% of intraabdominal testes. Ductus ending (with or without testis attached) was inguinal in 57.4%, intraabdominal in 39.9% and scrotal 2.7%.

Conclusions: Non-palpable testes are mostly seen on the left side. Contralateral testis hypertrophy is the most reliable physical examination finding. Seventy percent of intraabdominal testes could be identified with ultrasound. Pre-structured evaluation forms and meticulous physical examination may prevent these patients from unnecessary interventions.
HANAN SAID1, SHAMIM HASHIM2, ABDUL AZIZ AL GHAMDI1, SYED SALAHUDDIN3, ALI ELSAYED2
1International Medical Center, Jeddah, Saudi Arabia. 2King Fahd Armed Forces Hospital, Jeddah, Saudi Arabia. 3Leeds Children Hospital, Leeds, United Kingdom

PURPOSE

TO EVALUATE THE EFFICACY AND FEASIBILITY OF ENDOPYELOTOMY IN RECURRENT PUJ OBSTRUCTION IN PAEDIATRIC AGE FROM 1 YEAR TO 12 YEARS

MATERIAL AND METHODS

Total of 6 patients were operated, all done by retrograde approach with laser ablation. Five of them had endopyelotomy after open pyeloplasty, 4 (age range from 9-11) out of the 5 were symptomatic. The six-case had duplex system with PUJ obstruction in lower moiety, for which the initial surgery was in ureteroureterostomy. He had persistent dilatation of lower moiety on follow-up ultrasound and decreased perfusion on CT angiography. Patient had division of lower calyceal shelf / septum.

RESULTS

The elder patients, between 9 year and 11 years old became completely asymptomatic and improvement of radiological study, that is with ultrasound and MAG3 scan. In the other two patients, 3 year and 1-1/2-year-old the improvement was confirmed radiologically.

CONCLUSIONS

Endopyelotomy in a recurrent PUJO is feasible and safe procedure and it could replace redo surgery. Further studies are needed to evaluate this procedure.
Aim of the study: To evaluate the effects of repeated Botulinum-A Toxin Injections (BTI) (≥5) on bladder compliance in children treated for refractory neurogenic detrusor overactivity (DOA).

Methods: We retrospectively identified 14 patients with refractory DOA who received at least 5 BTI in our hospital, between November 2006 and November 2017. Patients who had non-neurogenic DOA and those who underwent augmentation cystoplasty were excluded. To assess the effects of repeated BTI on bladder compliance, we compared urodynamic testings performed before the first BTI and after the last BTI (Student’s test for paired samples, statistical significance p<0.05). Of the 14 patients, 3 were not exploitable (missing urodynamic data), 2 were excluded (1 bladder dysfunction due to posterior urethral valves, 1 enterocystoplasty). The remaining 9 patients (1 microcephaly, 6 dysraphia, 1 spinal cord tumor, 1 traumatic spinal cord injury) received 9 BTI (n=1), 7 BTI (n=1), 6 BTI (n=4) and 5 BTI (n=4) full dose, spaced 13 months (6-29) apart, with a mean age of 6 years (1-13) at the first BTI.

Main Results: In all patients, bladder compliance improved after repeated BTI (mean bladder compliance raised from 5 to 19 mL/cmH(2)O; p=0.04), with a mean follow-up of 3 months since the last BTI.

Conclusions: Repeated BTI in children with refractory neurogenic DOA are safe regarding bladder compliance. Moreover, it might improve this parameter. A longer follow-up is needed to support this hypothesis.
ENDOSCOPIC HOLMIUM LASER TREATMENT OF URINARY STONE DISEASE: A SINGLE-CENTER EXPERIENCE

Federica Marinoni, Giorgio Giuseppe Orlando Selvaggio, Giovanni Di Iorio, Andrea Pansini, Giovanna Riccipetitoni
V. Buzzi Children’s Hospital, Milano, Italy

Aim of the Study
Stone disease is increasing in children. Standard treatment procedures are: extracorporeal shock wave lithotripsy (ESWL), ureteroscopy (URS/RIRS), percutaneous nephrolithotomy (PCNL/MicroPNL) and surgery. Aim of our study is to demonstrate the feasibility of endoscopic treatment.

Methods
We collected data about 31 patients diagnosed between 2008 and 2017, of which 22 were endoscopically treated with Holmium Laser. Demographics, etiology, stone location, number and type of procedures, technical details, stone-free rate at 6 months were recorded.

Main results
Age ranged between 2-15 years. The etiology was metabolic (7 patients) or secondary to: neurogenic bladder (4), urinary tract malformation (1), Kabuki Syndrome (1). Twenty-nine procedures were performed in 22 patients: 16 URS, 9 RIRS, 1 PCNL, 1 MicroPNL, 2 bladder lasers, without complications. Hospitalization time ranged between 36-48 hours. Seven patients required pre-stenting due to the ureteral diameter and 7 had a ureteral sheath placed during treatment. All patients were stented at the end of the procedure and found stone free at 6-months follow-up.

Conclusions.
Endoscopic treatment with Holmium Laser is a valid and safe procedure, favored by the miniaturization of the instruments. URS/RIRS minimize risks of bleeding and visceral injury compared to PCNL/MicroPNL. The use of rigid and flexible ureterorenoscope allows the treatment of renal and proximal ureteral calculi. The ureteral sheath makes multiple or recurrent accesses into the kidney easier and decreases the intrarenal pressure by drainage of the irrigation fluid. In younger children pre-stenting may be necessary to dilate the ureteral papilla.
Medical Expulsive Therapy Versus Operation in Pediatric Distal Ureteral Stones: Which Is Advantageous?

Asya Eylem Boztas, Ozge Ozturk, Kamer Polatdemir, Gokce Sonmez, Ozge Atacan, Aytac Karkiner, Arzu Sencan, Akgun Oral

Health Medical Sciences University, Izmir Dr. Behcet Uz Children’s Hospital, Izmir, Turkey

Aim of the Study: Incidence of ureterolithiasis in pediatric population is increasing in recent years. Recently medical treatment of distal ureteral stones with doxazosin has advantages to have patients stone free within a shorter period, less stay in hospitals and causing cost saving.

Methods: Between 2015-2017 total 46 Patients with ureteral stones analyzed. 38 were eligible for the study to the exclusion criterias. Complete data were available for all patients. Their age range was 1-17 years (mean 8,3 years). Patients were divided into 2 main groups: group 1 (n =17), who received doxazosin once daily, before bed; and group 2 (n =21), who had ureteroscopy, uretherolitotripsy. All patients had distal ureteral stone. Datas were abstracted using manual chart review. Passage was defined as radiographic clearance.

Main Results: Mean expulsion times for groups 1 and 2 were 13,1 and 29,8 days(p<0,005), avarage hospitalization days for group 1 was 2,7 days and fort group 2 was 8,04 days (p<0,005). And the mean cost for groups 1 and 2 were 863,9 – 2009,7 Turkish Liras (p<0,005). In group 1, 2 of the patients underwent surgery (11%) and 1 patient had urinary tract infection(5,8%). In group 2, 1 patient had urinoma after surgery and 3 of patients had urinary tract infection(9,5%).

Conclusions: As it is discussed in many studies alpha blockers are shown to be a safe and effective therapy option for ureteral stones in pediatric patients. Use of alpha blockers have advantages in stone free preiod, shortened hospitalizations and lower cost.
PW22-G01
LAPAROSCOPIC HIATAL HERNIA REPAIR IN CHILDREN

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¹Division of Pediatric Surgery, Department of Pediatrics, University of Szeged, Szeged, Hungary.
²Department of Pediatric Surgery, Chelsea Children’s Hospital, Chelsea and Westminster NHS Fdn Trust, Imperial College London, London, United Kingdom

Aim: This study analyzed the literature with regards to outcomes of laparoscopic hiatal hernia (HH) repair.

Methods: Pubmed® was searched (2000-2015) using terms ‘laparoscopic hiatal hernia repair’ and ‘children’. Data collected included age, comorbidities, symptoms, diagnostics, treatment and outcomes.

Results: 10 articles met the inclusion criteria with n=125 patients ranging from 0.4-8 years (mean 4.16 years). HH was primary (congenital) n=15 and secondary (post fundoplication) n=110. Symptoms included vomiting n=48, failure to thrive n=12, chest pain n=8, gastro-esophageal reflux n=29, dysphagia n=8, respiratory infection n=8 and unknown n=12. Comorbidities included neurologic impairment n=10, cerebral palsy n=2, esophageal atresia n=26, trisomy 47XX n=2, malrotation n=1, Marfan’s n=1 and gastrochisis n=1. Chest films (n=5), upper gastrointestinal contrast study (n=39), barium swallow (n=20), CT (n=3) or esophagogastroduodenoscopy (n=6) led to diagnosis. N=92 underwent Nissen, n=6 Collis-Nissen, n=3 Toupet, n=2 Dor and n=1 Thal (unknown n=21). Laparoscopic repair was done n=93 (3 conversions: adhesions, respiratory distress, and unknown), robotic-assisted n=7, open n=16, and unknown n=19. Patches were employed in n=46 (37%). Complication rate was 6.4% (n=8); gastric perforation n=1, mesh penetration into esophagus n=1, postoperative dysphagia n=1. N=5 (4%) recurrences were managed with redo fundoplication (1 Nissen, 4 unknown), among which n=1 had a previous mesh.

Conclusion: HH is congenital in 12% and post fundoplication in 88% children; with esophageal atresia being the predominant comorbidity in 1/5. Laparoscopic Nissen is the preferred technique for HH repair with patch requirement in 1/3. Complications and recurrences after laparoscopic HH repairs are 6.4% and 4% respectively.
Background. Owing to the rarity of pancreatic cancer in children, Pancreticoduodenectomy (PD) is rarely performed in this age group. Aim of this retrospective study was to focus on long-term functional outcome of PD, with particular interest on late surgical complications, nutritional status and glucose homeostasis.

Materials and results. From 2010 to 2017, 6 children underwent pylorus preserving PD for various indications: pancreatic acinar cell carcinoma in two cases; solid pseudo-papillary tumor in three cases and endocrine tumor in the remaining. Median follow up was 4.25 years (range: 6 months- 10 years). Two patients developed stenosis of the pancreatico-jejunal anastomosis (11 and 4 months post-surgery, respectively) needing surgical treatment and one ulcer of the duodenum-jejunum anastomosis, treated by high dose- PPI. All patients were treated by pancrealipase and multivitamins for subclinical pancreatic insufficiency. Other chronic medications include PPI and periodic iron supplements. None of the patients developed post-operative malnutrition as showed by the BMI Z-score higher than -2 in all patients at the final anthropometric assessment ( BMI Z-score was 0.8 ±0.7 at surgery and -0.2±1.4 at follow up). No patients suffered for glucose intolerance needing treatment by insulin or oral hypoglycemic agents.

Conclusions. In surviving children and adolescents affected by pancreatic malignancy treated by pylorus preserving PD, long-term outcome is satisfactory; this procedure seems to have no impact on growth, nutritional status and glucose homeostasis if early detected and treated. Surgical complications can occur but they can be treated and are not life-threatening.
AIM: Neonatal burns are a challenge for paediatric surgeons, and their management differs significantly from older children due to the special physiology of newborns. Our aim was to describe the epidemiology of neonatal burns, their specific physiopathology and the difficulties encountered during their treatment.

METHODS: Medical records of 60 neonates (30 preterm) who had received treatment for burns during the last 20 years were reviewed. We collected data on burn aetiology, fluid resuscitation, type of dressings employed and need for surgical treatment.

RESULTS: Most newborns (41/60; 68%) presented with chemical burns, mainly due to extravasations (38/41), affecting small TBSA (<1-2%); and, therefore, did not require specific fluid resuscitation. Thirty-four (97%) healed by primary intention. Babies with thermal injuries underwent a more complicated clinical course. Six required fluid resuscitation per neonatal hypovolemic shock protocols (which proved to be more effective than standard burn fluid resuscitation formulas); and 9% (2/22) needed skin grafting, which was technically difficult owing to the lack of neonatal-sized dermatomes. Toxicity from the topical agents employed (nitrofurazone, mupirocin, silver sulfadiazine) was not observed; but its use in low-weight preterm babies was carefully addressed.

CONCLUSIONS: Neonatal burns are unique and represent a different entity from burns in other stages of life. Many difficulties are encountered by the medical team, such as ineffectiveness of standard fluid resuscitation guidelines; the potential toxicity of topical agents and the lack of appropriate-sized surgical material. On the other hand, the outstanding healing potential of babies allows for primary intention healing in most cases.
PW22-G04
POSTERIOR CRUCIATE LIGAMENT (PCL) TEARS IN PAEDIATRIC AND ADOLESCENT PATIENTS: MID-TERM FOLLOW-UP AFTER SURGICAL MANAGEMENT

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Aim of the study: The present study describes epidemiology, management and outcome of pediatric and adolescent patients with PCL lesions.

Methods: Following ethical approval, all patients <18 years with PCL lesions were included. Follow-up examination consisted of clinical examination and Pedi-IKDC and Tegner Lysholm (TLS) scores. Posterior shift was measured in kneeling view radiographs. Patients were grouped into those who underwent screw fixation, reconstruction with semitendinosus/gracilis or quadriceps tendon (STG/QT), and those who received bone-patellar tendon-bone (BPTB) reconstruction.

Main Results: 16 patients (mean age 14.5 years) with 7 PCL avulsion fractures and 9 PCL tears were included. 13 patients suffered from multiple injuries to the knee. 6 patients had open physes at time of surgery. Treatment was epiphyseal screw fixation in 5 (Group 1), STG/QT graft in 8 (Group 2) and BPTB graft in 3 cases (Group 3). Group 2 showed a significantly higher posterior shift of the injured knee, which did not translate into significant difference in Pedi-IKDC and TLS scores. Patients with single PCL injuries showed a clear trend to better Pedi-IKDC and less posterior shift. Arthrosis grade 1 or 2 was seen in 8 patients, 11 patients were not able to perform deep squats. None of the patients with open physes showed growth disturbances.

Conclusions: Despite multiple injuries in 13 of 16 patients, Pedi-IKDC and TLS scores at follow-up were comparable to previous studies in young patients with single PCL injuries. In case of open growth plate, careful techniques can prevent growth disturbances.
CONGENITAL POUCH COLON (CPC): SYSTEMATIC REVIEW AND WESTERN COUNTRY EXPERIENCE

Miriam Duci¹, Enrico La Pergola¹, Paola Midrio², Francesco Fascetti - Leon¹, Piergiorgio Gamba³
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The management of CPC, a rare variant of anorectal malformation, is still controversial. We aimed to systematically review the existing evidences about treatments and outcomes of CPC and to report the experience with preserving pouch of two two European Centers.

A systematic review was performed in the “MEDLINE” database using “congenital pouch colon” words. Articles with comprehensive description of surgical management and defined follow-up were included. Results from our two-centers series were incorporated. Non parametric test were performed. P<0.05 was considered significant.

56 out of 96 studies identified met our research criteria including 617 patients (353 type 1-2CPC, 257 type 3-4CPC, 7 type 5CPC (Saxena classification)). We added 4 CPC cases from our Centres (3 type 1CPC, 1 type 3CPC). The pouch was preserved more frequently in type 1-2 (p<0.0001)(tab 1). Pouch re-dilatation occurred in 31. In 94 patients (66 spared pouch, 28 resected) the follow-up was reported. No significant differences in terms of outcomes were found (P=0.58)(Table2). Patients with spared pouch underwent tapering and pull-through (103/335) or tubularization (163/335). In this group the tubulised pouch was used to increase bladder capacity (4/163) and to do pull-trough (5/163). In 69/335 the pouch use was not specified.

The existence of an ideal management for CPC is not supported by robust evidence in literature. However, our limited “sparing-pouch” experience might encourage utilising the dilated colon.

<table>
<thead>
<tr>
<th>Pouch Type</th>
<th>N° of patients with Pouch Spared (%)</th>
<th>N° patients with Pouch Resected (%)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 and 2</td>
<td>250 (76.4%)</td>
<td>100 (35%)</td>
<td>F &lt; 0.0001</td>
</tr>
<tr>
<td>3 and 4</td>
<td>75 (22.4%)</td>
<td>183 (64%)</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>4 (12%)</td>
<td>3 (1%)</td>
<td></td>
</tr>
</tbody>
</table>

Table 1

<table>
<thead>
<tr>
<th>Outcomes</th>
<th>N° of patients with Pouch Spared (%)</th>
<th>N° patients with Pouch Resected (%)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Good QoL</td>
<td>33 (33.7%)</td>
<td>7 (25%)</td>
<td></td>
</tr>
<tr>
<td>Poor QoL (patients depending on intestinal irritation and soiling)</td>
<td>53 (80.3%)</td>
<td>21 (75%)</td>
<td>P &lt;0.05</td>
</tr>
</tbody>
</table>

Table 2
KIDS AND TEENS ON WHEELS: A SIX-YEAR POPULATION-BASED STUDY

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Aims of the study
Traffic injuries are a major cause of morbidity among children and adolescents. Our aim was to study the epidemiology of school aged children’s trauma focusing on vehicles the children use independently.

Methods
After ethical approval data from N=1945 children age 7-17 years treated for trauma at our institutions between 2008 and 2013 was included. Vehicles included bicycles, mopeds, skateboards and kick-scooters.

Main results
Mean patient age was 13.1 years (SD 2.5), with moped patients being the oldest (15.4 years, SD 0.88). Most patients were males in all vehicle groups (70-95%). The most common injury was a fracture of the upper limb (37%; 730/1945) with higher frequency among skateboard and scooter riders than other vehicles, p<0.001. Lower limb fractures, internal organ injuries, multiple injuries, hospital admissions and operative treatment were more common among moped drivers than other vehicles, p<0.001.
Head injuries were more frequent among bicycle riders (20.8%) and moped drivers (16.9%, p<0.001) than other vehicles. Only a helmet that stayed intact was associated with a reduced number of head injuries, p<0.001.
The median ISS-score was 4 in all vehicle groups. Mean ISS varied from 2.96(bicycles) to 3.84(mopeds). The only small statistic difference in ISS was between bicycles and skateboards.

Conclusions
Moped drivers had a higher burden of injuries than patients injured while using other vehicles although ISS score didn’t show significant difference. Helmet use doesn’t affect the number of head injuries, if it is
broken or loosely attached.

Table 1. Demographic, injury and treatment data of 1945 children and adolescents.

<table>
<thead>
<tr>
<th></th>
<th>Scooter (n=171)</th>
<th>Skateboard (n=341)</th>
<th>Bike (n=842)</th>
<th>Moped (n=591)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male sex, n (%)</td>
<td>78%</td>
<td>95%</td>
<td>72%</td>
<td>70%</td>
<td></td>
</tr>
<tr>
<td>Age, mean (SD)</td>
<td>10.8(2.12)</td>
<td>13.1(1.80)</td>
<td>11.9(2.38)</td>
<td>15.4(0.88)</td>
<td>2</td>
</tr>
<tr>
<td>ISS, mean (SD)</td>
<td>3.19(1.6)</td>
<td>3.32(1.6)</td>
<td>2.96(2.0)</td>
<td>3.84(3.9)</td>
<td>3</td>
</tr>
<tr>
<td>ISS, median (range)</td>
<td>4.00(1-10)</td>
<td>4.00(1-9)</td>
<td>4.00 (1-18)</td>
<td>4.00(1-29)</td>
<td></td>
</tr>
<tr>
<td>Head injury, n (%)</td>
<td>19 (11%)</td>
<td>21(6%)</td>
<td>175(21%)</td>
<td>100(17%)</td>
<td>4</td>
</tr>
<tr>
<td>Fracture, n (%)</td>
<td>114(67%)</td>
<td>245(72%)</td>
<td>426(51%)</td>
<td>292(49%)</td>
<td></td>
</tr>
<tr>
<td>Upper extremity, n (%)</td>
<td>90(53%)</td>
<td>187(55%)</td>
<td>341(41%)</td>
<td>110(19%)</td>
<td>6</td>
</tr>
<tr>
<td>Lower extremity, n (%)</td>
<td>23(14%)</td>
<td>56(16%)</td>
<td>67(8%)</td>
<td>162(27%)</td>
<td>7</td>
</tr>
<tr>
<td>Spine, n (%)</td>
<td>0(0%)</td>
<td>0(0%)</td>
<td>2(0.2%)</td>
<td>5(0.8%)</td>
<td></td>
</tr>
<tr>
<td>Internal organ, n (%)</td>
<td>3(2%)</td>
<td>1(0.3%)</td>
<td>18(2%)</td>
<td>33(6%)</td>
<td>7</td>
</tr>
<tr>
<td>Multiple injuries, n (%)</td>
<td>4(2%)</td>
<td>5(2%)</td>
<td>55(7%)</td>
<td>72(12%)</td>
<td>8</td>
</tr>
<tr>
<td>Operative treatment, n (%)</td>
<td>26(15%)</td>
<td>65(19%)</td>
<td>123(15%)</td>
<td>180(31%)</td>
<td>7</td>
</tr>
<tr>
<td>Days hospitalized, median (range)</td>
<td>0(0-9)</td>
<td>0(0-6)</td>
<td>0(0-55)</td>
<td>1(0-162)</td>
<td></td>
</tr>
<tr>
<td>Hospitalized &gt;1 day, n (%)</td>
<td>33(19%)</td>
<td>76(22%)</td>
<td>230(27%)</td>
<td>328(56%)</td>
<td>7</td>
</tr>
</tbody>
</table>

2 Moped patients oldest, p<0.001

3 Difference between skateboards and bicycles, p<0.002

4 Difference in frequency of Head injuries between moped/bicycle group and others, p<0.001, no difference between mopeds and scooters.

6 Difference in frequency of upper extremity fractures between skateboard/scooter riders and bike/moped riders p<0.001

7 Difference in frequency of lower extremity fractures/Internal organ injuries/operative treatment/admittance to hospital between moped rides and other groups p<0.001

8 Difference in frequency of multiple injuries between moped riders and other groups, p<0.001 and bicycle riders compared to other groups (mopeds excluded), p<0.001
ABDOMINAL TRAUMA: ARE THE APSA GUIDELINES APPLIED AND ARE THEY APPLICABLE?

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Aim of the study
The American paediatric surgical Association (APSA) published guidelines for management of isolated spleen or liver injury in Children in 2000, proposing Length of Hospital stay (LOS) according to CT injury Grade. We hypothesized that these guidelines are not widely applied in our country. We decided to evaluate management of isolated abdominal trauma and see whether application of the APSA guidelines would have modified outcome.

Methods
We retrospectively reviewed data of children admitted for spleen, liver or kidney injuries over a 5-year period, in 2 university paediatric hospitals in France. Patients with associated abdominal trauma, with other extra-abdominal injury or with initial operative management were excluded.

Results
122 patients with isolated trauma (62 spleen, 37 liver, 23 kidney) were included. Average LOS for Grades I, II, III, IV and V were respectively 8.2, 9.4, 12.8, 13.8 and 22.5 days. 11 patients presented post-trauma complications. Only 2 of these occurred after the required LOS proposed by APSA: one embolization on the 6th day post-trauma for a Grade III spleen injury and a peritoneal drainage on the 15th day post-trauma for a Grade IV liver injury.

Conclusion
LOS in these 2 centres is at least 3 times longer than the APSA recommendations. Only 2 patients (1,6%) required further treatment for a complication which was diagnosed after the required LOS guidelines. We think it’s safe to follow APSA recommendations for Liver, Spleen or Kidney isolated trauma. Applying these guidelines would decrease hospital stay and costs without impacting patient outcome.
PW22-G09
A LARGE SERIES OF PEDIATRIC SILS SPLENECTOMY: SINGLE CENTER EXPERIENCE

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King Fahad Armed Forces Hospital, Jeddah, Saudi Arabia

Aims: To assess the feasibility, safety, and expediency of SILS splenectomy in pediatric patients. To highlight the difficulties in this technique and discuss our strategies to address these issues and compare it with other published series.

Material & Methods: Retrospective study of all patients operated with SILS splenectomy from April 2011 to August 2017. Forty nine patients had been operated. Age ranged from 2.5 year to 14.8 years. One type of port was used in all cases about 95% through 1.5 cm umbilicus. Mainly straight regular instruments were used in the procedures. One extra port in the left flank was used for splenic-retraction.

Results: Forty nine patients underwent SILS splenectomy. Eighty % were done by trainees. Six cholecystectomies were done simultaneously, 2 conversion due to bleeding. Forty five patients with sickle cell disease, two with thalassemia one spherocytosis and one Fanconi’s anemia. There is no wound infection, one post operative bleeding. Mean operative time for splenectomy was 182.26 minutes (130-190) & 251min for both splenectomy and cholecystectomy (230-270) min depending on the operator’s skill, severity of adhesions and size of the spleen. It is comparable with conventional technique.

Conclusion: SILS splenectomy is safe, feasible and better cosmesis with almost invisible scar. More than one procedure could be done at the same time. The effects of instrument crowding, as well as the absence of triangulation, are among the common technical challenging issues. The confusion of crossing image will be adapted after the first 10-15 min. It can be done by trainees under supervision of experienced mentor.
PW22-G10
LONG TERM FOLLOW-UP EXAMINATION OF THE PERFUSION OF THE INTERNAL JUGULAR VEIN AFTER VESSEL-SPARING IMPLANTATION OF A HICKMAN-CATHETER OR PORT-A-CATH

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Aim of the study: Hickman-Catheter (HC) and Port-a-Cath (Port) are frequently used catheters in paediatrics for long-term venous access. Catheters are inserted in a vessel-sparing technique when placed in the internal jugular vein (IJV). Even if this is a common technique, only little data exists about the vessel’s state in the long-term follow-up. Aim of this study is to determine the perfusion of the IJV after vessel-sparing implantation of a HC or Port.

Methods: 100 patients (aged ≤ 18 years), who underwent a vessel-sparing implantation of a HC/Port in the IJV at a single institution, were included. Patency and shape of the IJV were determined by a Doppler-ultrasound examination. Operation time, catheters indwelling time, drugs administered via the catheter, catheter-associated thrombus or sepsis, patient’s BMI, coagulopathies, and surgeon’s level of training were collected from medical chart.

Main Results: All catheters were inserted in the correct vessel. 91% of our patients had a patent IJV. 47% had no signs of structural abnormalities. 44% showed venous scars, stenosis, or dilatation. In 9% no perfusion of the IJV was detectable. 26% of our patients had relevant findings of the blood flow-pattern, caused by relevant structural alterations. Statistical analysis did not reveal any specific parameter as a risk-factor for relevant structural abnormalities of the IJV.

Conclusion: Even if one quarter of our patients had relevant structural alterations in the follow-up examination, vessel-sparing venous cut-down is a save approach to place the catheter in the correct vessel.
OUTCOME OF PAEDIATRIC PANCREATIC TRAUMA: A TRI-CENTRIC EXPERIENCE

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AIM OF THE STUDY Pediatric pancreatic traumas are rare and usually associated with traumas of other abdominal organs. Management of intermediate grades of injuries is controversial. Aim of the study was to assess the outcome of pancreatic traumas in children.

METHODS Pancreatic traumas occurring between 2004 and 2017 were collected from 3 referral pediatric centers and classified according to AAST classification (I-V grades, being IV and V extremely rare).

RESULTS 22 patients (mean age 8.2 years, ranges: 5-12) were collected with 12 grade I, 5 grade II, and 5 grade III. The most common mechanism of injury was bicycle handlebar (76%). 16 patients were managed conservatively, including 4 with pseudocyst. Six other patients, instead, developed a pseudocyst which required treatment: 3 percutaneous drainages, 1 each endoscopic drainage, subtotal pancreasectomy, cyst-jejunumstomy. ERCP was used in 1 patient with complete pancreatic duct transaction to introduce a stent into the main pancreatic duct. No complications were registered. The mean length of stay was 13 days for grade I, 48 days for grade II-III. Patients with grade I were discharged with normal biochemical and ultrasound pattern. At 3 months from discharge persistence of disomogeneous parenchymal area was evident in 6/10 grade II-III patients that eventually solved at 6 months. The 4 pseudocysts, conservatively treated, required a median of 60 days to disappeared.

CONCLUSIONS The conservative management of grade II and III pancreatic lesions is safe and effective although associated with higher rate of pseudocyst formation and longer hospitalization. Low grade injuries have excellent outcome.
DISTAL RADIAL FRACTURES IN CHILDHOOD: IS THE PROXIMAL TITAN ELASTIC NAILING A SAFE SURGICAL METHOD?

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Heim Pal National Pediatric Institute, Dept. of Surgery and Traumatology, Budapest, Hungary

Aim of the study

The treatment of distal radial fracture with severe displacement in childhood can be done by Kirschner wiring (KW), by titan elastic nailing from distal direction (DTEN), and by “short TEN” technique. Every technique has shortages giving not the ideal solution always. An alternative method was introduced in our practice, the titan elastic nailing from proximal direction (PTEN), being suitable and effective. The aim was to study the effectiveness and possible complications of PTEN, especially the radial nerve injury.

Methods

The cases were studied between 1st of July 2012 and 1st of July 2017. All applied PTEN cases were included.

Results

28 pts were treated by PTEN. The technique was chosen as first in 20/28 cases. 4/28 pts were treated conservatively initially, but due to later fracture re-displacement PTEN procedure was applied as secondary method. 2/28 pts were treated by KW as first, but after later re-displacement the PTEN technique was chosen at reoperation. 2/28 pts were treated by DTEN, but due to intraoperative complication the method was converted immediately to PTEN. Radial nerve injury was not found in any case. 1/28 severe re-displacement was found after PTEN, that was corrected by osteotomy combined with KFI plate. Other complication was not found.

Conclusions

The PTEN technic is an easy, safe and suitable alternative method. The feared radial nerve injury should be avoided by inserting the nail to the adequate anatomic position (above radial neck) at forearm pronation. The procedure time is shorter to the other techniques.
Trans-anal rectal injuries is uncommon, and the diagnosis is usually delayed because of the patients unwillingness to give correct history. These injuries may be caused by the introduction of foreign body, sexual assault, iatrogenic lesions and rarely by barotrauma by compressed air jet through the anus which may lead to colonic distension and perforation. This report describes a case of anorectal barotrauma due to insufflation of high pressure air directly from the anus. While playing two children inserted a compressor in the anus of a 3 years old child. After the joke the patient presented severe rectal bleeding, abdominal distension and severe pain. At the emergency department x-rays showed massive colonic distension, pneumoscrotum, generalized subcutaneous emphysema that extended from the pelvis to the thorax and suspected signs of pneumoperitoneum. Examination under anaesthesia showed two anterior perineal lacerations, and during proctoscopy no macroscopic lesions of the rectum were found. Laparoscopy was performed to exclude colonic perforations: no blood or fecal soiling was identified; the only relevant finding was distension and parietal emphysema of the recto-sigmoid colon. Spontaneous resolution of emphysema and surveillance of further complications were obtained with periodic abdominal x-rays. Trans-anal barotrauma by compressed air causing injuries of the intestine is rare in the pediatric population, but can be fatal. Surgical exploration has the priority for the early treatment of potential bowel perforation thus leading to an improved survival and lower morbidity.
TORSION OF CECAL APPENDIX. REPORT OF THE FIRST ITALIAN CASE AND REVIEW OF THE LITERATURE

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¹Department of Pediatric Surgery and Pediatric Minimally Invasive Surgery and New Technologies, San Bortolo Hospital, Vicenza, Italy. ²Department of Pediatric Surgery and Pediatric Minimally Invasive Surgery and New Technologies, San Bortolo Hospital, Vicenza, Italy

Abstract

In pediatric patient appendicitis is the most common cause of abdominal pain and surgery. Torsion of the vermiform appendix is a rare cause that is clinically indistinguishable from appendicitis and usually the diagnosis is intraoperative. The first description of torsion of the vermiform appendix was made by Payne in 1918. The clinical presentation is the same of the acute appendicitis. The preoperative investigations play a minimal role. The etiology of this condition is unclear, but it is possible to distinguish a primary and a secondary torsion. We report a case of 5-year old boy who presented with right lower quadrant abdominal pain. Its clinical signs and symptoms (vomiting, increasing tenderness and later muscular rigidity in the right lower abdomen) and investigations (ultrasonography proved appendicular involvement) were similar to that of acute appendicitis. Intraoperatively we found a torsion of the vermiform appendix of 720° on its base with its mesentery rotated in counter-clockwise direction. The appendix was gangrenous in appearance. A video-assisted trans umbilical appendectomy was performed. We reviewed the clinical presentation and management of this rare condition performing a review of the literature.
Alba Bueno, Javier Serradilla, Jose Luis Encinas, Pilar Durán, Francisco Hernández, María Álvarez, Francisco de Borja Nava, Carlos Delgado, Antonio Jesús Muñoz, Manuel López-Santamaría, Carlos Andrés De la Torre

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**INTRODUCTION:**

Pectus excavatum (PE) is the most common congenital chest wall malformation. It is sometimes associated to entities such as congenital diaphragmatic hernia (CDH). It is viable to repair by Nuss technique in patients with CDH, however some technical aspects must be taken into account.

**CASE REPORT:**

We report a 13-year-old patient with a severe PE who had a history of a corrected left CDH. During growth she developed severe PE becoming deeper every year. Functional tests were normal but the girl and her family complaint about cosmetic appearance and the impact on quality of life.

An asymmetric PE with Haller's Index of 6.8 and a correction index of 59% was found by cardiac MRI. The most interesting finding was a complete cardiac dextroposition, with the left ventricle wall in contact to the sternum.

We discussed about which approach would be more appropriate and safer for the patient.

Finally, we performed a Nuss procedure from right side. We bewared of the proximity of the heart at the time of trocar placement. Sternal elevation made easier the tunneling from right to left. Some pleural adhesions were found and removed by blunt dissection. Two bars were placed with no complications.

The girl presents optimal aesthetic and functional results in the follow-up time.

**CONCLUSIONS:**

Nuss technique is feasible for these patients. Special attention on cardiac position and possible pulmonary pleural adhesions is mandatory. The use of sternal elevation during the procedure is highly recomened.
**PW23-CR05**

**ARTERIAL PRIAPISM IN CHILDREN: TREATMENT BY AN ORAL ALPHA-ADRENERGIC STIMULANT AGENT (ETILEFRINE)**

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**Background:**

In children, selective arterial embolization could be challenging for arterial priapism. We have tried Etilefrine, an alpha-adrenergic agonist, since its efficiency has been reported as preventive treatment of priapism in sickle-cell disease.

**Material and method:**

**Case 1:** 13 year old boy admitted with a 24 hours of post-traumatic priapism. Blood cavernous gas showed a well oxygenated blood which confirmed the diagnosis. We have tried a decompressive puncture, then an intracavernous injection of Ephedrine but failed after 48 hours. Therefore, Etilefrine (20 mg/d) orally was given for one month. **Case 2:** 9-year old boy admitted with a 10-day history of painless priapism arising after a penile injury. A small painless induration was noted at the base of the penis. Doppler ultrasonography (US) confirmed the diagnosis showing an arterio-venous fistula. Etilefrine (15 mg/d) was given orally.

**Results:**

**Case 1:** After one month follow up, there was no recurrence. The penis was flaccid with normal spontaneous erections. **Case 2:** after 30 days, the penis was flaccid, with normal spontaneous erections. Etilefrine was stopped because of headache afterword. Four months follow up showed the penile induration has disappeared. In addition, The Doppler US showed decreased vascular flow of the arterio-venous fistula and size of the hypoechochogenic node. Four years after, the patient was still having spontaneous erections and normal penile development.

**Conclusions:**

Etilefrine appears as an effective new modality to treat the arterial priapism in children. It has low morbidity and it seems to have no erectile consequence.
Persistent anastamotic leak post oesophageal atresia tracheo-oesophageal fistula (OA-TOF) repair is difficult to manage, sometimes requiring oesophageal replacement. Recurrent TOF have previously been successfully treated with fibrin sealant (Tisseel®). We present the first case of the use of endoscopic Tisseel® to treat a persistent anastamotic leak post OA-TOF.

32/40, 1.3kg female with post-natally diagnosed OA-TOF and segmental 7th left rib. She underwent ligation of the fistula and gastrostomy on day 1 due to a long gap. Gap assessment and OA-TOF anastomosis was performed at 12 weeks old with a transanastamotic tube (TAT) and chest drain left in situ due to a tight anastomosis. Unfortunately she developed a persistent pneumothorax and a contrast swallow confirmed an anastamotic leak. This was managed conservatively with a chest drain and gastrostomy feeds. Repeat contrast swallow showed a significant, persistent leak (Figure 1).

She returned to theatre 7 weeks post anastomosis. Under endoscopic and fluoroscopic guidance, a guidewire was passed down the oesophageal lumen and Tisseel® applied to the anastamotic defect (Figure 2). A TAT was passed over the guidewire and a chest drain left in situ. Surprisingly, not only did the anastamotic leak resolve completely, but she did not require an oesophageal stretch until more than 10 weeks post Tisseel®.
We present the first case of the use of Tisseel® to treat a significant anastamotic leak, without complication and preventing the requirement for oesophageal replacement.
Partial Duplication of the Genitourinary System with Total Colonic Duplication in a Boy After In Vitro Fertilization: A Case Report

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¹Riga Stradins University, Riga, Latvia. ²University Children's Hospital, Riga, Latvia

Abstract

Children with combinations of large colorectal and genitourinary duplications are rare, however, there is an elevated prevalence suggested with the rise of fertility treatments.

Some malformations are so rare that the occurrence worldwide is less than 100 reported cases. Since these cases are so rare, it is believed that this is the very first case of such magnitude to be fully documented. This report follows the patient from delivery through all surgical treatments to the current health status.

This case report depicts a boy born after in vitro fertilization (IVF) presenting with two anuses, true diphallia, hypogonadism, spina bifida and myelocele. Upon further diagnostic investigations, the patient had total duplication of the colon, rectum, anus and urinary bladder each with its own urethra.

The excretory malformations include one imperforate anus, one anal stenosis, and a rectourethral fistula. During his treatment, the medical team consisted of neonatologists, pediatricians, gastroenterologists, and endocrinologists, while the surgical team included pediatric surgeons and pediatric urologists.

Postoperatively, the patient regularly followed up with the medical team as he frequently developed constipation. The current health status shows improvement in the quality of life with continuous interdisciplinary treatment.
DOUBLE AORTIC ARCH: ABOUT UNUSUAL CLINICAL PRESENTATION IN 2 CHILDREN

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Introduction
The double aortic arch is a rare congenital malformation resulting from the persistence of the fourth arches and dorsal aortas forming a complete vascular ring encircling and compressing the esophagus and trachea. It is usually diagnosed during neonatal period with respiratory symptoms like stridor, wheezing or pneumonias. We present two unusually late diagnosis evoked on digestive symptoms.

Case reports
Two boys aged 13 and 14 presented in the emergency with a history of recent dysphagia. The first one had lost 4kgs in one month. The second patient was a known asthmatic who recently described food hanging up in the mid esophagus. Both patients benefited from a computed tomography angiography (fig 1) which confirmed the diagnosis and showed an associated persistent arterial duct. The treatment consisted in a division of the atretic segment and a ligation of the arterial duct through a left thoracotomy. They are asymptomatic one year later.

Discussion
Our two described cases are unusual because true tracheoesophageal compression was found per operatively and we could not explain why both boys became dysphagic so late and why they did not suffer from more severe respiratory symptoms before. The treatment is always surgical and the approach depends on the possible associated cardiac malformations. The functional prognosis is good after surgery but it can be altered by tracheomalacia if associated.

Conclusion
Double aortic arch can be diagnosed in older patients with unusual digestive symptoms.

Fig 1. Axial CT picture showing the double aortic arch.
Here we present two cases of uterine duplication, with a non-communicating rudimentary horn and a unicornuate uterus.

The first case was a 13-year-old woman who came to emergency room for acute abdominal pain and severe dysmenorrhea. She had been normally menstruated for 1 year. In her medical history, we noticed that she was the second born of a twin pregnancy provided by in vitro fertilization. On examination, her external genitalia were normal. MRI showed uterine duplication with a unicornuate left uterus and a right non-communicating hemi-uterus that contained important hematometra. There was only one cervix, one vagina and no renal anomaly. The patient underwent a complete laparoscopic excision of her right hemi-uterus and the fallopian tube. Laparoscopy revealed no endometriosis lesions. Post operative recovery was uneventful.

The second case is a 14-year-old woman who was hospitalized for abdominal pain, vomiting and nausea. Transabdominal US-scan revealed a large lateral uterine mass and MRI showed a right unicornuate uterus communicating with the vagina and a non-communicating left horn. She currently undergoes a progesterone treatment waiting for surgical resection of her rudimentary left hemi-uterus. To conclude, uterine duplication with non-communicating hemi-uterus is a very rare malformation that is to be known because of its complications. This type of uterine malformation is associated with infertility, endometriosis and a risk of uterine rupture during pregnancy. It must be suspected in case of dysmenorrhea, and management relies on laparoscopic surgical resection.
We report the case of a 7-month-old boy presenting with upper airway obstructive symptoms since the age of 15 days. During the initial diagnostic workout, a fiberoptic bronchoscopy showed a bulging in the subglottic region, below the left vocalis cordis. A cervical MRI showed the same intensity of another mass located in the left paratracheal cervical region that was diagnosed as an ectopic thymus. The normal thymus gland was visualized in the usual cervical position. At the age of 7 months a clinical worsening required a surgical management. Through a standard transverse neck incision, the anterior thyroid and cricoid cartilages and the first three tracheal rings were exposed. We performed a modified laringofissure with a vertical midline incision through the cricoid cartilage up to the lower border of the thyroid cartilage just below the level of the vocal cords. Gained access to the endotracheal lumen, the lesion was covered by normal-appearing mucosa. A mucosal flap was raised at the level of the bulging, and the mass presented a soft consistence being densely adherent to the surrounding submucosal tissues. While dissecting submucosally the lesion, it appeared to have continuity with the left paratracheal mass identified by the MRI as ectopic thymus. The lesion was isolated and the final histology showed thymic tissue.

In conclusion, ectopic thymus should be considered in differential diagnosis of subglottic masses in order to guide correctly the surgical choice of resection.
PW23-CR11
CHALLENGING DIAGNOSIS FOR A BIG MEDIASTINAL TUMOR IN A 11-YEAR OLD GIRL

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Aim:
We would like to demonstrate diagnostic difficulties in a 11 year old girl sent to our center because of a big tumor of the ventral mediastinum.

Methods:
Because of dyspnea and fatigue a 11 year old girl presents to the clinic. Exams show a big tumor of the ventral mediastinum causing atelectasis because of compression of the left bronchial system and pleural effusion. Patient gets drainage of effusion with unspecific cytological findings and patient is referred to your center. After CT- and MRI-exams and two biopsies of the tumor without clear diagnosis, interdisciplinary tumor board suggests primary resection because of recurrent severe dyspnea and suspicion for tumor of the thymus.

Main Results:
Histological analysis of specimen shows a nodular Hodgkin-lymphoma IIBE and patients is included in EURONET-PH2-C2-trial. Because of nodal recurrence in early response assessment in PET-CT after 2 cycles of OEPA-schema, patient receives chemotherapy with cyclophosphamide, vincristine, prednisone, dacarbazine, doxorubicine and etoposide. During operation left laryngeal recurrent nerve and pericard must be resected because of involvement in tumor. Patient tolerates operation and following chemotherapy well, but experience recurrent episodes of dyspnea.

Conclusions:
Tumors of thymus are the most common tumors in ventral mediastinum in children. In our case two biopsies only showed normal thymustissue so interdisciplinary tumor board suggest resection. Because of lack of right diagnosis patient received excessive surgery with high morbidity. 6 Months after initial operation patient is in complete remission.

PW23-CR12: CAVERNOUS HEMANGIOMA OF THE BLADDER IN CHILDREN: ABOUT TWO CASES

Nizar Sassi, Sondes Sahli, Wiem Hannachi, Nesrine Chebil, Amina Karray, Mourad Hamzaoui
Abstract

Oesophageal lung (OL) is a very rare anomaly wherein the bronchus of a lung connects directly with the oesophagus. Repeated lower respiratory tract infection (LRTI) is the hallmark of this condition. We present the clinical images of a five-month-old boy who presented with repeated LRTI since birth.

A contrast-enhanced CT scan revealed a small collapsed right lung being supplied by the pulmonary artery and drained by the pulmonary vein. The right-sided main bronchus was not identifiable.

A contrast oesophagogram showed prompt delineation of the right bronchial subdivisions. A communication of the lung with oesophagus was established at the junction of middle and lower one-third of the intrathoracic oesophagus.

The child underwent an exploratory thoracotomy. The preoperative findings were confirmed. The bilobed lung was non-aerated and had a liver like consistency. There was a direct communication between the oesophagus and the lung across a very short 0.5 cm wide stump Excision of the lung with primary repair of the oesophagus was curative.

This report is expected to help understand the abnormal anatomy and surgical repair in OL. Management is excision of the involved lobes and primary repair of the oesophageal defect.
ABDOMINAL PAIN REVEALING A DUPLICATION OF VERMIFORM APPENDIX IN A 6-YEAR-OLD CHILD

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Aim of study:
Duplication of the vermiform appendix is an extremely rare congenital anomaly, with a reported incidence of 0.004%. Even though the abnormality is rare, the complications that might arise from an unidentified duplicate appendix may have serious consequences for the patient. We present a case of appendicular duplication in a 6-year-old child.

Case Report
A 6-year-old child presented with vomiting and right lower quadrant (RLQ) abdominal pain. His physical examination revealed tenderness in the right iliac fossa. Laboratory investigations, including serum electrolyte levels, complete blood count, and C-reactive protein were within normal limits. Ultrasound of the abdomen showed a normal appendix. In the face of persistent abdominal pain, a computed tomography scan was performed revealing a blind-ending tubular structure arising from the ileal loops. Meckel’s diverticulum scintigraphy was negative. Laparoscopy revealed a double appendix. The two appendices were macroscopically normal. There were no other noteworthy surgical findings and the remaining abdominal viscera were normal. The appendices were removed and sent for histopathological examination, which confirmed the diagnosis of double appendix. Inflammatory changes were found in one appendix; the other appendix was normal.

Conclusion
Duplication of the appendix should be considered in the differential diagnosis of lower abdominal pain, even if the patient reports a previous appendectomy. Failure to recognize this condition may have serious clinical and medico legal consequences.
Introduction

Conjoined twin separation is one of the most challenging surgical exercise. We successfully separated one such case. We detail the surgical challenge that we faced in the management of the case especially in a developing institute.

Case Report

A primigravida mother delivered a full term delivery by LSCS with a combined birth weight of 4.6 Kgs. The twins were referred to our Institute on day 63 of life. Both babies were joined on the ventral surface with a common thorax and abdomen with a small omphalocele. The babies were investigated extensively and was found to have a fused lower chest (with two separate hearts), fused abdominal wall, common pericardial sac with joined liver.

Separation was done at 3 months of life. We discuss the challenges and preparation that were faced in a still developing institution in the separation including dealing with manpower crunch, creation of separate teams, mock preoperative drills, adequacy of equipment and dealing with the intense media scrutiny.
PW24-CR04
A TRAIN CAN HIDE ANOTHER: VOLVULUS WITH INTESTINAL MALROTATION HIDING A PANAGANGLIONNOSIS

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Introduction

Total colonic aganglionosis occurring together with malrotation is a rare occurrence and may pose diagnostic and management dilemmas for the paediatric surgeon.

Case report

A new born, was admitted at the third day of life because of bilious vomiting and stopping transit. On physical examination, the abdomen was flat and sensitive. The newborn was hemodynamically unstable. Surgical exploration found an incomplete common mesenteric volvulus with Ladd band and mesenteric fusion. Ladd’s procedure was performed and classical bands were divided. Postoperatively, we noticed persistent abdominal distension and a meconium emitted on day 10 in a very small quantity. Subsequently, emission of a small amount of meconium was observed every 4 to 5 days. The barium enema showed a non-functional microcolon. Surgical revision on the 24th day found an ileo-ileal size discrepancy located 60 cm from the angle of Treitz. Extemporaneous suction biopsies confirmed the diagnosis of total colonic Hirschsprung disease, which extended to the small intestines. We performed an ileostomy 40 cm from the angle of Treitz. Postoperatively, the patient was transferred to a neonatology department for better nutritional management, while waiting for a radical cure of her condition.

Conclusion

This case was a challenging scenario for us where a diagnosis of malrotation had obscured the hirschsprung disease. The obstructive symptoms at presentation were due to the volvulus. Postoperatively these symptoms were due to the extended form of hirschsprung disease. Persistent intestinal obstructive symptoms following a Ladd procedure is a red flag and must search for an associated Hirschsprung disease.
PW24-CR05
COMPLETE STOMACH HERNIATION IN A NEONATAL MARFAN SYNDROME

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AIM

The Neonatal Marfan Syndrome is an uncommon early clinical expression of this disease and implies a poor prognosis due mainly to cardiovascular disorders. We report a case of a hiatal hernia with a complete intra-thoracic stomach as a rare presentation of this entity in a child.

CASE PRESENTATION

A 1-month-old infant with suspected neonatal Marfan syndrome due to a strong familiar history was referred to our center with vomits and failure to thrive and with an image of a complete intra-thoracic stomach in the contrast study. He had a mild dilatation of the ascendant aorta with no cardiovascular symptoms. At admission, a laparoscopic reduction of the herniated stomach and part of the colon and a Nissen-Rossetti fundoplication was performed, taking special care in closing the hiatus properly. The postoperative course was uneventful but in the contrast study at 1-month post-op a mild migration of the fundoplication was noted. The patient reports no symptoms to date, so a REDO surgery is still not proposed.

CONCLUSION

Hiatal hernia with intrathoracic stomach is a potentially severe complication in children with connective tissue disorders and the risk of gastric volvulus can jeopardize the patient’s life. It is important to make an early diagnosis and offer a surgical treatment. A very close follow up is highly recommended as the relapse of the herniation may rapidly occur.
Abstract
High-flow, non-ischemic priapism is a rare condition especially in children, with which many urologists and andrologists are unfamiliar.

There are three types of high-flow priapism: traumatic, neurogenic and post-shunting. We report the case of a 10-year-old boy who presented with a non-painful priapism with a history of two weeks permanent erection after suffering a perineal trauma.

Colour Doppler ultrasound detected an arteriocorporal fistula with a turbulent flow between the cavernosal artery and the corpus cavernosum. A cavernosal blood Gas confirmed the high-flow character. A superselective embolization with Gelatin sponge was safe and effective to treat the high-flow priapism. The use of gelatin sponge usually results in a temporary embolization with long-term effect of penile detumescence and preservation of erectile function.
LONG GAP ESOPHAGEAL ATRESIA, IS THERE A PLACE FOR A NEW INNOVATIVE MINIMALLY INVASIVE TECHNIQUE FOR GAP MEASUREMENT?

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Introduction:
The gap between two esophageal pouches is considered to be an important factor in outcomes in cases of esophageal atresia (EA). Previous descriptions of gap measurement techniques includes: contrast injected into distal esophageal pouch via gastrostomy, dilators or flexible endoscope to define distal esophageal pouch. We present a case with the application of a innovative technique.

Case report:
We report a case of a female newborn born in our institution with postnatal diagnosis of VACTER and Down syndrome presenting EA without TEF. The first day of life was performed colostomy and after conducting all complementary studies (with intravenous nutritional support), the patient was taken to the operating room.

First, a catheter is placed inside proximal esophageal pouch. Then, under laparoscopically view we performed a percutaneous gastric puncture with an epidural catheter (figure 1) and through it we infused a bolus of water-soluble contrast media to draw distal esophageal pouch under fluoroscopy control. This facilitated the measurement and decision making. The gap was measured in 3 vertebral bodies (figure 2), therefore it was decided to successfully perform a primary esophageal anastomosis using a thoracoscopic approach without need for gastrostomy. The follow-up has been uneventful (10 months).

Conclusions:
Based on our preliminary observations, our new minimally invasive approach for gap measurement in long gap esophageal atresia was done safely and reduced need for a gastrostomy. It may be an ideal procedure for reference centers with experience in thoracoscopically repair to avoid the need of an invasive method of gap measurement.
COSTELLO SYNDROME AND UMBILICAL LIGAMENT RHABDOMYOSARCOMA

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Introduction: Costello Syndrome (CS) is a rare congenital anomaly, characterized by prenatal overgrowth, postnatal failure to thrive, classic facial features and multisystem involvement. These patients have a strong predisposition for childhood cancer, most commonly rhabdomyosarcoma (RMS), with a cumulative risk of 15% by 20 years. We present three consecutive cases of umbilical ligament RMS in CS.

Methods: We reviewed the clinical charts of patients with CS and RMS. Focus was aimed at clinical presentation, diagnosis work-up and surgical treatment.

Results: CS was diagnosed between 5-6 months in all patients, and prior to RMS diagnosis. Tumor screening consisting of abdominal physical exam and ultrasounds were performed every 3-6 months. Two patients presented with vague abdominal pain prior to the identification of a large >5cm mass. All tumors derived from the umbilical ligament, making clinical diagnosis challenging. Biopsy confirmed the diagnosis of embryonal RMS in all three. After evaluation and stratification into standard (2) and high risk (1), treatment was initiated according to EppSG RMS 2005 protocol. Surgical resection was complete in 1 out of 3 patients, with minimal and focal invasion of peripheral margins in two patients, that required adjuvant radiotherapy. After a follow-up period of 2, 24 and 28 months respectively, the 3 patients are disease and event free.

Conclusion: Umbilical ligament RMS diagnosis is usually made in advanced stages and carries poor prognosis. The high predisposition for this type of RMS in patients with CS warrants an aggressive screening protocol in order to improve disease-free and survival rates.
Case-Presentation

A 15-year-old boy was admitted to our hospital in critical general condition with increasing abdominal pain over 10 days. No fever or vomiting. The abdomen was tender. Laboratory examinations revealed CRP>300mg/dL and WBC 18.000/uL. CT-scan showed free fluid, intestinal wall thickening and diffuse mesenteric lymphadenitis. The appendix was not visualized. Bowel perforation was suspected.

An immediate explorative laparotomy was performed; clear ascites and distended bowel loops were found without any signs of other intestinal pathology. Post-operatively the patient developed severe SIRS. Serology and microbiology revealed no evidence of intestinal pathogens. From the 8th postoperative day on he developed pleural effusion, fever and night sweats as well as anemia and thrombocytopenia. Enlarged axillary lymph-nodes were palpable. Lymph-node biopsy, bone marrow and diagnostic pleural fluid aspiration were performed. Histology revealed multicentric Castleman’s disease. Treatment with monoclonal antibody for IL-6 receptor was successful.

Castleman’s disease is a rare lymphoproliferative disorder of the young adult. Typically, giant lymph-node hyperplasia is found. Symptoms are unspecific and may mimic neoplastic, rheumatologic or infectious diseases. Activation of the interleukin-6 cascade is typical for Castleman’s disease. It results in a massive
pro-inflammatory response and explains peritonitis and high infectious parameters in our case. Multicentric Castleman’s disease is life-threatening and extremely rare in pediatric patients. Presentation as surgical acute abdomen resulting in laparotomy has not been described earlier.

**Conclusion**
Castleman’s disease may present as surgical acute abdomen. In cases of unclear peritonitis and mesenteric adenopathy it should be included in the differential diagnoses.
ARTERIAL TORTUOSITY SYNDROME AND ITS RELEVANCE IN PEDIATRIC SURGERY

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Introduction: Arterial Tortuosity Syndrome (ATS) is a very rare connective tissue disorder, with less than 100 cases described in literature. ATS is characterized by an abnormal tortuosity an elongation of major arteries and other connective tissue disorders such as multiple hernias. The onset of the disease occurs in the childhood, with unknown prevalence. We present a case ATS with difficult surgical management.

Case Report: Male term newborn with no prenatal findings and postnatal diagnosis of mayor vessel malformation including hypoplasia of the pulmonary arteries, aortic arch and supra-aortic trunk. Clinical manifestations included pulmonary hypoplasia and a dissociation of arterial blood pressure between upper and lower extremities with no evidence of aortic coarctation. In addition, it had a giant hiatal hernia with the stomach completely lodged in the thorax and bilateral inguinal hernia. Genetic study confirmed ATS with two heterozygous mutations of SLC2A10 gene. At age 3-months surgical repair of the hiatal hernia was performed with a Belsey-Mark IV procedure. During follow-up, hiatal recurrence was evidenced with clinical symptoms of reflux and malnutrition requiring redo surgery. The inguinal indirect hernias also required multiple interventions due to recurrence. In the fourth closure attempt a bilateral polypropylene mesh was used.

Conclusions: ATS is a rare connective tissue disorder that affects mayor vessels with a high mortality rate due to cardio-respiratory insufficiency and ischemic events. The association with multiple, highly recurrent hernias of difficult management should encourage pediatric surgeons to consider the use of prosthetic material for its definitive closure.
PW24-CR12
APPRAISAL OF SACRAL NERVE INJURY BY PELVIC NEUROTACTOGRAPHY AFTER ROBOTIC SURGERY FOR SACRAL NEUROFIBROMA IN A 12 YEARS OLD GIRL

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Aim of the study: Diffusion tensor MRI allows non-invasive representation of central nervous anatomy and is increasingly included in image guided neurosurgery methods. This radiological tool, along with associated neurotractography algorithms, may be used to evaluate the peripheral nervous system, such as pelvic sacral plexus, helping pre-operative planning and post-operative evaluation of nerve injury.

Methods: A 12-years-old girl with neurofibromatosis was diagnosed with a left sacral neurofibroma and scheduled for surgery because of the slow growth of the lesion on pelvic MRI (8x8x3 cm) and PET scan characteristics (SUV = 2.83), that suggested high risk of malignant transformation. Diffusion tensor pelvic MRI with neurotractography was performed before and after tumoral resection.

Main results: Pre-operative neurotractogram showed a tumor in close contact with the left sacral plexus that seemed to arise from S2 sacral root. The tumor was resected by robotic assisted laparoscopy, fragmented in a plastic bag and extracted through the umbilical incision, with an operating time of 150 minutes, without blood transfusion. Post-operative course was uneventful and hospital discharge was done at day 3. Paresthesia and dysesthesia of the left leg calf and of the plantar vault, without any motor deficit, appeared in the following weeks. Post-operative neurotractogram confirmed interruption of left S2 sacral root, consistent with clinical symptoms.

Conclusions: This case validates the accuracy of the pelvic network anatomy delivered by pelvic neurotractography. It may be a useful tool in pelvic surgery management, with perspectives of real time image guided surgery to improve outcome.

Figure 1: Pre-operative MRI and tractography included in 3D model. Sequence: coro T2 cube. Arrow: Left sacral neurofibroma
Figure 2: Pelvic 3D model with pre-and post-operative neurotractography. Arrow: Left S2 root defect
SCIENTIFIC SESSION XII: ONCOLOGY

Saturday June 23rd

SCXII-ON01:
PREOPERATIVE INTERVENTIONAL RADIOLOGY FOR PAEDIATRIC SOLID TUMOURS:
A SYSTEMATIC REVIEW AND META-ANALYSIS FROM THE GROUP OF YOUNG SURGEONS OF EUROPE

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Aim: Interventional radiology (IR) in the management of solid malignant tumours in paediatrics has increased over the last 3 decades. Nowadays, IR clinical application is mainly addressed to unresectable tumours, resulting in preoperative tumour shrinkage and sometimes near total ablation. Our aim was to review the effectiveness of IR procedures in the treatment of malignant lesions in children.

Methods: Using a defined search strategy, we identified studies reporting the effectiveness of embolization/ablation techniques in paediatric solid malignant tumours between 1980 and 2017. Reports with less than 3 patients, review and opinion articles were excluded. Data included factors related to patient, tumour or IR technique. We analysed dichotomous and continuous variables by appropriate statistical methods. The meta-analysis was conducted using RevMan5.3.

Results: Of 567 articles screened, we analysed 21 papers (12 retrospective, 7 prospective, 2 randomised-control-trials). IR-guided ablations were mainly rescue procedures to treat primary unresectable tumours, local recurrences, metastases. Inclusion-exclusion criteria and success definition were not specified in most reports. Major side effects were documented in 17/286 (6%) infants, minor side effects were self-limiting in most patients.
Meta-analysis: 6 studies compared embolization in kidney and liver (127 infants) to control (113 infants, surgery alone/chemotherapy/nothing). IR group vs controls had lower mortality (Table1) and surgical time for resection, higher 2-years tumour-free-survival and better tumour-specific markers (p<0.001).

Conclusions
Ablative techniques are feasible and promising treatments for paediatric tumours. However, specific prospective trials should focus on IR in paediatric solid tumour oncology to clarify efficacy, especially in early-stages tumours.
SCXII-ON02
CAN WE AVOID RADIATION EXPOSURE FOR CENTRAL VENOUS PORT SYSTEM PLACEMENT IN CHILDREN?

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Introduction/ Aim: Central venous port (CVP) placement is traditionally performed under fluoroscopy guidance. However, efforts for radiation-dose reduction in children have allowed the introduction of ultrasound-guidance and anatomic landmarks as an alternative technique for CVP placement. The aim of this study is to determine whether intraoperative fluoroscopy (IF) is required to confirm the correct position of the catheter tip in children.

Patients and Methods: A prospective, single-center study was performed between July and December 2017. Standard venous access site was the right internal jugular vein under ultrasound-guidance. Estimated catheter length (ECL) was measured using anatomic landmarks. Ideal catheter length (ICL) was measured after placement under fluoroscopy guidance in the same patient. Age, sex, radiation dose, and complications were also analyzed. A T-test for paired samples and intraclass correlation coefficient were performed to analyze results.

Results: A total of 31 consecutive patients aged 7±2 years underwent CVP placement. The mean ECL was 17.5±1.8 cm while the ICL was 17.7±1.8 cm. The mean difference between measurements was 0.28 cm (95% CI -0.29 to 0.86; p=0.324). Intraclass Correlation Coefficient analysis showed an agreement of 0.95 (95% CI 0.91-0.98) between measurements. Mean radiation exposure during the procedure was 1.06±0.78 mGy m² during 0.34±0.6 minutes. There were no complications registered during CVP placement.

Conclusion: The correlation between IF and USG and anatomically guided catheter tip placement is optimal. These results suggest that fluoroscopy and the radiation exposure it entails can be safely avoided in selected children.
Aims of the Study: We sought to determine the clinical profile of patients with Wilms’ tumor (WT) undergoing circumferential resection of inferior vena cava (IVC) without its replacement, and to evaluate their oncologic outcomes and procedure-related morbidity.

Methods: Extensive literature review (1971-2017) and personal experience on WT patients receiving cavectomy without reconstruction as part of their surgical treatment for IVC involvement. Data collected included patient demographics, stage of disease, laterality of renal primary, level of IVC extension, histopathology, treatment received, morbidity and mortality.

Main results: 15 patients were identified, including 2 new cases from our own institution. Renal primary was right-sided in 13 and left-sided in 2 cases. Indications for cavectomy included complete IVC occlusion from tumor invasion (13) or adherence (2), and IVC resection was performed en bloc with nephrectomy (12) or as second-look intervention (3). Type of cavectomy included resection of the infrarenal portion (4), and resection of the infrahepatic portion with ligation of left (9) or right renal vein (2). Four stage IV patients died, including 1 perioperative death. The remaining 11 patients (3 stage II, 5 stage III, 2 stage IV, 1 stage V) are alive and disease-free at median follow-up of 46 months (range, 10-132). One patient experienced a transient moderate lower extremity edema.

Conclusions: In WT patients with IVC involvement, cavectomy without reconstruction is well tolerated because most patients have well-developed collaterals secondary to pre-existing IVC occlusion. Such procedure may allow for complete tumor clearance, thus obviating the need for postoperative radiation therapy.
SCXII-ON04
ADRENOCORTICAL CARCINOMA: INFLUENCE OF TP53 STATUS ON PROGNOSIS?

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Aim of the Study: Adrenocortical carcinoma (ADCC) is a rare tumor, very suggestive of Li Fraumeni syndrome (LFS), a familial predisposition to cancer, caused by a germline mutation of the TP53 gene. Germline TP53 mutations are found in half of the children with ADCC. The aim of the study was to compare clinical features of ADCC in children with or without TP53 mutation.

Methods: Clinical data concerning patients that had been addressed to our genetic laboratory for TP53 testing were retrospectively collected. TP53 molecular analysis was performed using Sanger sequencing or New Generation Sequencing and QMPSF.

Main results: 98 blood samples of patients with medical history of pediatric ADCC (31 males, 67 females) were referred to our genetic laboratory for TP53 testing. TP53 sequencing revealed 45 germline heterozygous mutations and 3 mosaicisms (49%). TP53 mutation detection rate in males and females was 38% and 55%, respectively. In males and females presenting with ADCC before 2 years of age, it increased to 54% and 72%, respectively. Sporadic tumors in mutated and wild type patients represent 42% and 78% of the tumors, respectively. Medical follow-up was available for 47 patients (mean follow-up 9 years), with a 5-year survival rate of 71% (82% in patients without mutation, 67% in mutated patients). Twelve patients, all carrying a TP53 mutation, were treated for a secondary tumor during follow-up.

Conclusion: In children with ADCC, presence of a TP53 mutation is characterized by earlier age of onset, poorer prognosis and a risk of secondary tumors.
SCXII-ON05
NEUROENDOCRINE TUMORS OF THE APPENDIX: AN UPDATE FROM THE TREP PROJECT

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Aim. Neuroendocrine tumours (NETs) of the appendix are the most common gastrointestinal epithelial tumor in childhood and are usually incidentally discovered after an operation for appendicitis. The treatment after diagnosis is still debated: in children, tumors are generally well-differentiated and it is still controversial whether right hemicolecotomY, traditionally recommended for >2cm-tumors, may offer more advantages in terms of overall/event free survival. The aim of the study was to confirm the choice of a non-aggressive approach in

Methods: NETs of the appendix have been prospectively registered within the Italian TREP project (Rare Tumors in Pediatric Age) since 2000. After appendectomy, the treatment suggested by the TREP guidelines are the following: no treatment in completely excised tumors, regardless of size and mesoappendix invasiveness, and caecum resection with pericolic lymphnodes biopsy, in case of positive margins.

Main Results: A total of 152 patients younger than 18 years with a diagnosis of well-differentiated NETs were registered from January 2000 to December 2017. In 138/152 the tumour was smaller than 2 cm and in 7 larger than 2 cm (unknown in 7). Excision margins were free in 142 (unknown in 7). Caecum resection or hemicolecotomY was performed in 4/152, and residual tumour was detected in 1. All 152 patients are alive in complete remission.

Conclusions. Our experience confirms that appendiceal NETs have a benign behaviour and appendectomy alone is curative for almost all patients; a more aggressive surgical approach is reserved in those cases with incompletely excised tumor.
PLEUROPULMONARY BLASTOMA: RESULTS OF A NATIONAL REGISTRY

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Aims: Pleuropulmonary blastoma (PPB) is a rare tumor divided in 3 subtypes ranging from cystic to solid. We aimed to described the management and outcomes of a national series of PPB.

METHODS: Retrospective observational study on children born between 2000 and 2017, and diagnosed with PPB in France.

RESULTS: Thirty-five children were included, 20 girls and 15 boys. Median age at diagnosis was 29 months. 12 children had a cancer family history. There were 16 type I (46%), 12 type II (34%) and 7 type III or II/III (20%). Four children presented with metastatic disease at diagnosis. Six children had surgery only. 15 had chemotherapy after surgery (3 required redo surgery) and 14 had neo-adjuvant chemotherapy followed by surgery. Three patients underwent radiotherapy. None of the children only treated with surgery went through secondary surgery. Three patients died from PPB type 2 or 3 progression. 100% of type 1 patients were alive at last clinic. The 5-year overall survival rate was 90%. Two patients, one type II and 1 type III, presented a relapse. The 5-year disease free survival rate was 83%. 15 of the 31 tested patients (48%) had a DICER1 mutation.

CONCLUSION: Cystic type I PPB has a more favorable prognosis than type II or III PPB. Our national cohort has similar outcomes than the International PPB registry.
SCXII-ON07
PANCREATIC TUMORS IN PEDIATRIC AGE: AN UPDATED OVERVIEW FROM THE TREP PROJECT

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Aim of the study
Pancreatic tumours are rare in children (0.2% of paediatric tumours). They represent a diagnostic and therapeutic challenge. The aim of this study was to review our national multi-institutional experience of pancreatic tumours in children.

Methods
We reviewed all cases of children with pancreatic tumors, prospectively enrolled into the TREP registry from 1 January 2000 to 31 December 2017. Particular attention was given to clinical features, diagnostic work-up, histology, treatment and outcome.

Main results
We identified 62 patients (24 M, 48 F). The most common symptom at presentation was abdominal pain. US scan was performed in all patients, followed by CT scan and MRI.

The first surgical approach consisted of initial biopsy in 13 patients (in 5 was the only surgical approach) and tumor resection in 49. There were 9 postoperative complications (pancreatic fistula, bleeding).

Histology: solid-cystic papillary tumors (38), neuroendocrine tumors (11), pancreatoblastoma (6), other tumors (7). Survival rate ranged from 100% for solid-cystic papillary tumors, to 50% for pancreatoblastoma. Follow-up ranged from 2 months to 16 years.

Conclusions
Pediatric pancreatic tumours may represent a diagnostic and surgical challenge. Surgery is curative for solid-cystic pseudopapillary tumors, showing a good overall outcome. On the other hand, pancreatoblastomas need a multidisciplinary approach. The TREP project has been fundamental for the study of these tumors, as well as the diagnostic and therapeutic approach. Through this project, these tumors and other hystotypes are included into the prospective European cooperative study ExPERT.
SCXII-ON08
PROPHYLACTIC THYROIDECTOMY IN HEREDITARY MEDULLARY THYROID CARCINOMA IN CHILDREN. A SINGLE-CENTRE EXPERIENCE

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OBJECTIVES: To describe the complications and long-term results in patients with MEN 2A syndrome on whom prophylactic thyroidectomy was performed, in relation to the recommendations of the American Thyroid Association (ATA).

METHODS: A retrospective study of 14 patients with MEN 2A thyroidectomized between 2000 and 2017. We review demographic, clinical, analytical and radiological data. Postoperative complications and long-term follow-up were analyzed.

RESULTS: We treated 8 boys and 6 girls with a median age of 5 years (range 2-10). The predominant genetic mutation belonged to codon 634 (8/14, 57.14%). Total thyroidectomy without cervical lymphadenectomy was performed in all patients. A right upper parathyroidectomy was performed due to intraoperative suspicion of increased volume in one patient, but histology revealed no alterations. Two patients presented transient hypocalcemia postoperatively; no patient had permanent hypocalcemia or nerve damage. Pathological anatomy confirmed medullary thyroid microcarcinoma in 5/14 patients, all of them carrying codon 634 mutation; two of them with preoperative basal calcitonin levels below 40 pg/ml. After a mean follow-up of 5 years, no recurrences or metastases have been detected. A patient with codon 634 mutation developed a unilateral pheochromocytoma at 25 years old. No patient has presented hyperparathyroidism.

CONCLUSIONS: Prophylactic thyroidectomy without cervical lymphadenectomy after 5 years of age, regardless of basal calcitonin levels, is an effective preventive treatment in patients with mutation of codon 634 and safe when performed in tertiary centers with experienced surgeons.
SCXII-ON09
MINIMALLY INVASIVE SURGERY FOR WILMS TUMOURS: CAN WE SAFELY EXPAND THE SIOP UMBRELLA PROTOCOL CRITERIA?

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Aim of the study: To determine if the SIOP criteria for Minimally Invasive Surgery (MIS) in Wilms Tumours (WT) could be safely expanded.

Methods: International multicentre review of MIS resection for WT between 2006-2017. Electronic or paper medical records were retrospectively assessed. Demographic, imaging, treatment, pathologic and oncological outcomes data were analysed. Only WT were included. All sites complied with institutional ethics board guidelines.

Main results: Forty-six patients from 8 centres were included. Median age at diagnosis was 38 months (6-181). Twenty-four tumours were left-sided and three were ruptured pre-operatively. Twenty patients had a biopsy prior to treatment and all received neoadjuvant chemotherapy. Location of the tumour was upper pole (16), lower pole (13) and interpolar region of the kidney (17). Median tumour volume post-chemotherapy was 673.5 ml (17.9-2170.4), 14 were crossing the lateral border of the spine and three were crossing the midline. There were 6 conversions but no intraoperative rupture. A median of 4 lymph nodes (1-10) was sampled in 39 cases. Tumours were Stage I (25), II (16) and III (5) and histology was reported as Low (2), Intermediate (40) and High risk (4). Three patients had positive tumour margins. After a median follow-up of 69 months (2-137), there was one local recurrence and one metastatic relapse.

Conclusions: In experienced hands in both MIS and oncologic surgery, SIOP criteria can be safely expanded to tumours abutting the midline. Low threshold for conversion should however remain if difficulty arise and attention should be paid to lymph node sampling.
PERITONEAL MESOTHELIOMA IN CHILDREN: A STUDY OF 15 CASES

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Aim of the Study: Benign and malignant peritoneal mesothelioma are rare tumors in children originating from mesothelial cells. The purpose of the study was to present a retrospective national review of 15 cases of peritoneal mesothelioma in children.

Methods: We included all children identified, treated in France from 1987 to 2017, for diffuse malignant peritoneal mesothelioma (DMPM) or benign cystic peritoneal mesothelioma (BCPM).

Main results: Fifteen patients (5 boys and 10 girls) aged 2.2 to 17.5 years were included. Abdominal pain, ascites and change of general conditions were the most common symptoms. Histologically, 8 patients had epithelioid malignant mesothelioma, 3 biphasic malignant mesothelioma, and 3 patients had BCPM. For a patient there was a doubt diagnosis between a malignant mesothelioma and another aggressive tumor. Seven patients were treated in first-line by cytoreductive surgery associated with hyperthermic peritoneal chemotherapy (HIPEC), among these, 2 patients received complementary systemic chemotherapy; 5 patients were treated with systemic chemotherapy alone. For the BCPM, 2 patients were treated by surgery alone, and 1 patient with HIPEC. The mean follow-up between the diagnosis and the last consultation was 98.7 months (24-288); 4 out of 12 DMPM and 2 out of 3 BCPM had local or distant recurrence; 2 patients with DMPM died 3 months after diagnosis.

Conclusions: Peritoneal mesothelioma is very rare in children. The diagnosis is difficult and the risk of recurrence is high. Networking of patients and national or even European interdisciplinary collaboration are needed to establish treatment protocols and evaluate outcomes in children.
AIM OF THE STUDY
Reporting our experience using 3D printing models and virtual recreation, for surgical planning of complex solid pediatric tumors.

METHODS
Data were obtained from preoperative MRI. Imaging analysis and 3D virtual recreations were performed using Cella-supplied software (Cella Medical Solutions, www.cellams.com). 3D real-scale geometry models, including tumor, adjacent organs and relevant vascularization, were printed in colorimetric scale and different materials (e.g. polylactic acid, polyurethane rubber) for optimal structures discrimination.

MAIN RESULTS
Four challenging cases were selected. First one was a bilateral Wilms tumor. Total left and partial right nephrectomy were initially considered, but the volumetric reconstruction proved that bilateral nephron-sparing surgery was achievable. The second case was a Wilms Tumor’s bilateral pulmonary metastatic recurrence. 3D technology contributed to an accurate localization of the lesions. The third case was a right abdominal neuroblastoma, enclosing major vessels. Initially, it was considered unresectable. Virtual recreation and the real scale 3D printed prototypes were of major value for achieving complete resection. The fourth case is a mediastinal neuroblastoma with anatomopathological diagnosis of ganglioneuroma. The optimal approach is being planned using the 3D implements due to the intimate relations between tumor, heart and major vessels.

CONCLUSIONS
3D virtual recreation and real scale model printing is an extra tool in complex oncological cases. It contributes to a better understanding of the relationship between the tumor, non-affected organs and relevant vascularization. Complete surgical resection is prognostic; therefore using all available resources is valuable in order to get the best outcome.
Ultrasound Assisted Cannulation of the Internal Jugular Vein: A Prospective Comparison to the Open Cut-Down Technique

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In critically ill children vascular access represents the decisive factor for survival. The cannulation of internal jugular vein (IJV) is recommended. Traditionally venous access has been performed by the open cut-down technique. The importance of US-guided venous access has increased in recent years. The objective of this study was to compare the two techniques.

This prospective randomized study compares two groups: US-guided IJV cannulation, including 321 patients from 2008 to 2016 (group A) and IJV cannulation by the cut-down technique, including 341 patients from 2002 to 2006 (group B). We used 3 types of catheter: Hickman, Broviac e Port-cath. We use the US-guided technique after the 6 months of life.

There were no between-group differences in sex, weight, hospital setting where the procedure was performed or puncture site. The median age was greatest in the group A. The time to IJV localization, time to achievement of venous access and time to cannulation were all shorter in the group A. The cannulation failure rate was also remarkably lower in the group A. 15 complications were noted, 1 in the group A (kinking of the catheter) and 14 in the group B (hematoma, bleeding, dislocation of the catheter and wound infection).

The findings of this study demonstrate that US guidance is a useful adjunct for central venous access in children. US increases the efficacy of the procedure and improves its safety significantly. US-guided cannulation should be recommended as the method of choice for safe achievement of central venous access in children.
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Published in June 2018 for the “19th EUPSA Congress”
Paris, 19th-23rd June

Designed & Edited