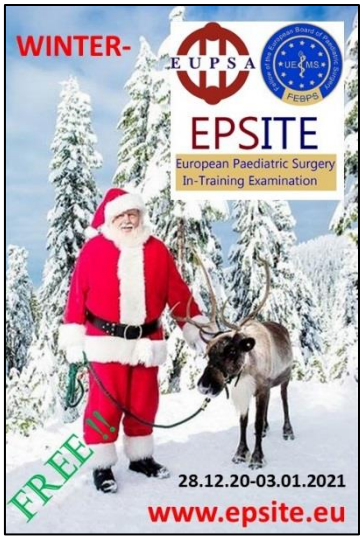

Answers

1. This is a picture of **diastasis recti**. There is a bulge superior to the umbilicus when the abdominal muscles are tensed. It is attributed to a weakness in the linea alba and usually resolves by itself.

2. This patient has Prune-Belly-Syndrome (PBS) or also known as Eagle-Barret-Syndrome (more politically correct). This syndrome is characterized by **poor abdominal muscle tone**, **undescended testicles** in males, and **urinary tract abnormalities** (such as hydronephrosis). The other findings are not typical of EBS.

3. This is a typical picture of a hernia to the chord "**chord hernia**". Umbilical cord hernia was first described in the literature by Hempel-Jorgensen in 1929. It is thought to be under-reported and often misdiagnosed as an 'omphalocele minor' due to similarities in appearance. Clinically, it can be distinguished from an omphalocele by the umbilical cord insertion site. In an umbilical cord hernia, there is a normal cord insertion with intact skin covering the umbilical ring. An omphalocele, on the other hand, is characterized by a large defect in the umbilical ring involving the skin and muscle, often covered by amniotic membrane that may contain bowel and other viscera such as the liver, with an umbilical cord insertion on top of the herniated sac. Also it is usually not associated with genetic defects. Postnatal, umbilical cord hernias are usually managed with primary surgical closure. Gastroschisis is usually to the right side of the umbilicus and the bowel loops are exposed. Cantrell's syndrome also includes other more serious malformations such as ectopia cordis.

4. The picture shows a fistula between the anus and the vaginal vestibule. These fistulae are typical of **Crohn disease**. Patients with ulcerative colitis do not present with fistulae, nor do the other diagnoses.

5. This baby has a myelomeningocele, which is associated with **neurogenic bowel and bladder**. Maternal serum AFP levels are usually elevated in neural tube defects, such as this one. The MOMS trial showed that prenatal intervention lead to lower rates of later ventriculoperitoneal shunting and better neurologic outcome. Due to the success of the MOMS trial, fetal surgery is now considered to be a treatment option for some cases of myelomeningocele.

6. Studies have shown that children with solid organ injuries who **present to pediatric trauma centers have a lower rate of being operated**, lower rates of undergoing embolization, and equivalent or better outcomes than those pediatric patients treated by general surgeons or in adult trauma centers. Splenic lacerations rarely require operative intervention, even grade 4 lesions. Newer data has shown that 1-2 days of bed rest and 1-2 weeks of refraining from athletic activities is sufficient without increasing the odds of a recurrent hemorrhage. Transfusion is only indicated in symptomatic children, not purely on the basis of a hematocrit. Most pancreatic injuries can be treated non-operatively. In cases with complete dissection, operative distal pancreatectomy can shorten hospital stay, but ultimate outcome is the same.

7. Pure tracheoesophageal fistulae without atresia are typically located at the thoracic inlet, which implies that they can be addressed via a cervical incision or by thoracoscopy. It is hard to reach the fistula by a conventional thoracotomy, since they are so high up in the chest. The fistula usually **passes from the proximal trachea to the distal esophagus**, which means that they can best detected by bronchoscopy and usually not by esophagoscopy.
8. **Alpha-fetoprotein (AFP)** is the most relevant tumor marker for hepatoblastoma. Vanillylmandelic acid (VMA) and Homovanillic acid (HVA) are elevated in the urine of children with neuroblastoma. Carcinoembryonic antigen is a tumor marker of epithelial tumors, such as adenocarcinomas of the colon and others.

9. Chilaiditi syndrome is defined as the **colon being located between the liver and the diaphragm**. It can mimic free air. Normally this causes no symptoms. The sign can be permanently present, or sporadically.

10. This is a typical picture of a boy with an imperforate anus and a **rectoperineal fistula**. Meconium passes through the fistula that is situated close to the scrotal raphe and escapes at the base of the penis. Rectovestibular fistula is reserved for females, a rectoprostatic fistula would not explain the meconium at the base of the penis unless there is a hypospadias (which is not the case here).

11. Up to 70% of all neurologically impaired children have gastroesophageal reflux disease (GERD). The outcome of complete (Nissen) fundoplication is not different from partial (Toupet, Thal) fundoplication. The positive effects of a fundoplication are not life-long and recurrence of symptoms very frequent, up t 43% after 10-15 years (Mauritz, FA Ann Surg 259:388–393). After failed fundoplication, Total Esophagogastric dissociation (TEGD) shows inferior results compared to redo-fundoplication (Lansdale N, J Pediatr Surg 50:1828–1832). **Laparoscopic** fundoplication has a **higher recurrence rate** compared to open fundoplication (Fyhn, TJ, Ann Surg 261:1061–1067).

12. The **presence** of ganglion cells consistently **correlates with calretinin-positive** thin nerve fibrils in the lamina propria, muscularis mucosae and superficial submucosa. Therefore calretinin immunohistochemistry offers additional diagnostic value in specimens with inadequate

amount of submucosa and rarely seen ganglion cells. Normal nerves do not stain for AChE, but increased AChE expression is associated with the hypertrophied extrinsic nerve fibres of the aganglionic segment in HSCR. In Hirschsprung disease there is an **increased acetylcholinesterase (AChE)** expression in the affected tissue. **Hypoganglionosis**, also known as intestinal hypoganglionosis, is a disorder causing a reduced number of nerves in the intestinal wall. Intestinal hypoganglionosis can **mimic** Hirschsprung disease; patients with both conditions may present with chronic constipation, intestinal obstruction, and enterocolitis (inflammation of the intestines. In Hirschsprung's disease the pathologist usually finds absent ganglion cells and submucosal nerve hypertrophy (not vascular or muscular hypertrophy. The submucosal nerve hypertrophy is found especially in the transition zone.

13. Bilious emesis in this age group is suspicious for midgut volvulus until proven otherwise. Most infants (93%) having midgut volvulus present with bilious emesis that is otherwise unexplained. Abdominal ultrasound can diagnose midgut volvulus based on abnormal positioning of the superior mesenteric artery (SMA) and vein (SMV). On ultrasound with doppler, reversal of the normal orientation of the mesenteric vessels is diagnostic of malrotation. With abnormal rotation, the SMA will appear on the right, and the SMV will appear on the left. Specific radiologic findings associated with midgut volvulus include a corkscrew appearance of the duodenum and proximal jejunum. Additionally, "whirlpool" appearance of the SMV and mesentery around the SMA may be seen on ultrasound in association with midgut volvulus. However, most pediatric surgeons prefer an upper GI to diagnose malrotation, though contrast may not pass beyond the point of obstruction. On upper GI contrast study, the duodenojejunal junction will fail to cross midline to the left, and will lie inferior to the duodenal bulb. Additionally, the second and third portion of the jejunum will not be located in the normal retroperitoneal location (Dahmer JJ, Chapter 16; "Pearls and Tricks in Pediatric Surgery" Lacher, Martin, St. Peter, Shawn D., Zani, Augusto (Eds.), Springer, 2020) https://www.springer.com/gp/book/9783030510664

14. Withdrawal of treatment is not an option here as the prognosis is not fatal. This is a neonatal emergency; therefore waiting 24-48 hours is not an option. An upper GI would not be feasible as this is most likely a TEF Type C (Gross classification) with a blind ending upper esophagus. The placement of a gastrostomy would not solve the problem of this child. The baby is in trouble because the air is not flowing into the lungs but also via the TEF into the stomach. Also potential bilious reflux (aspiration) via the TEF into the lungs may happen. In case the TEF is at the level below the ET tube (e.g. at the carina = "trifurcation") the mechanical ventilation may increase the air flow via the TEF into the stomach. **A thoracotomy with closure of the fistula would both solve the problem** (facilitate ventilation into the bronchi. prevent reflux) and give time to recover and ultimately survive. The reconstruction (anastomosis) of the esophagus can then be done later, even beyond the neonatal period.

15. A testicular descent beyond 6 months of life is very unlikely, therefore waiting another 6 months does not make sense. The gold standard for a non-palpable and possible abdominal testis is diagnostic laparoscopy as it is both diagnostic as well as therapeutic. Therapeutic options include lap-assisted orchidopexy, 1 or 2-stage Fowler-Stevens orchidopexy or Shehata orchidopexy. The sensitivity of an MRI is lower than laparoscopy, also an MRI requires anesthesia in this age group. Endocrine referral with hormonal evaluation is only indicated in bilateral cryptorchidism.

16. The lung is the most common site of relapse from osteosarcoma. Several studies revealed that the patients with pulmonary metastases of osteosarcoma have a poor prognosis. Outcome of the patients with pulmonary metastases at initial presentation is not a dismal when the appropriate chemotherapy and thoracotomy are performed. As a treatment factor, complete surgical resection of the pulmonary metastases is a most important predictor for survival of those patients, although the benefit of video-assisted thoracoscopic surgery or multiple thoracotomy is still controversial. The number of published data available regarding the efficacy of radiotherapy in the treatment of pulmonary metastases is limited. Emerging techniques including stereotactic body radiotherapy and radiofrequency ablation could be an effective option of the treatment for unresectable metastatic lesion.

17. The term vascular ring refers to congenital vascular anomalies of the aortic arch system that compress the esophagus and trachea, causing symptoms related to those two structures. The most common vascular rings are double aortic arch and right aortic arch with left ligamentum. The symptoms are related to the compression of the trachea, esophagus or both by the complete vascular ring. Presentation is often within the first month (neonatal period) and usually within the first 6 months of life. **Starting at birth an inspiratory and expiratory stridor (high pitch noise from turbulent airflow in trachea) may be present often in combination with an expiratory wheeze.** Secondary to compression of the esophagus babies often feed poorly. They may have difficulties in swallowing liquids with choking or regurgitating and increased respiratory obstruction during feeding (**less** frequent than respiratory symptoms). Older patients might refuse to take solid food, although most infants with severe symptoms nowadays are operated upon before they are offered solid food.

18. This 3 month old infant is 6 weeks post ileostomy takedown and now having a distended abdomen with poor feeding as well as failure to thrive. A typical feature of NEC are colonic strictures. Therefore prior to any reversal of an ileostomy a stricture of the colon needs to be ruled out by an antegrade contrast study. An anastomic stricture would become symptomatic immediately and not 6 weeks later, the malrotatin of the midgut presents with (bilious) vomiting. This case is not suspicious for Short Bowel Syndrome as there is no information that large parts of the small bowel were resected during the initial surgery. The child is 3 months old already, therefore not being at a typical age-group for recurrent Necrotising Enterocolitis.

19. This is a term boy with no visible anal opening. The standard protocol is to wait 24 hours in order to observe whether a fistula is visible thereafter (e.g. rectoperineal fistula). A colostomy is certainly a good next step if after this time period there is no fistula. As there are no signs for respiratory distress, therefore intubation and mechanical ventilation is not indicated. An Magnetic resonance imaging (MRI) of the abdomen may help to define the level of the fistula in a few months of life. However, this study requires general anesthesia which should be avoided in the neonatal age unless there is no other option. A neonatal Posterior sagittal anorectoplasty can be performed for lower types of ARM in experienced hands. However, also a PSARP is done only after observation for 24 hours.

20. This 14-yo boy has blunt abdominal trauma. He is pale & tender in the right upper quadrant, tachycardic (HR: 109/min) and his Hemoglobin/hematocrit of 10.1/31 is decreased. The initial management is 20cc/kg cristalloids (Ringer, normal saline 0,9%). See ATOMAC guidelines (Notrica DM, J Trauma Acute Care Surg. 2015 Oct;79(4):683-93).