Urachal Remnants

Urachal remnants are uncommon anomalies that present symptoms in infancy or early childhood. The urachus is formed in early embryonic life as a tubular connection between the dome of the bladder and the allantoic stalk. This fistulous tract obliterates into a medial ligament. Rare persistence of this remnant after birth can lead to a completely patent urachus, a sinus (opening to the navel), a diverticulum (opening to the bladder), a cyst (both end close but central portion remains open) or alternating sinus (cyst-like structure can drain to either the bladder or umbilicus). Neonatal patent urachus can undergo spontaneous involution. Otherwise, the transitional epithelium is replaced by granulation tissue and the main symptom of a patent urachus is urine discharge from the umbilicus associated with cellulitis, pain and swelling. A cyst or diverticulum can present as a midline, localized, painful abdominal mass associated with fever and leucocytosis. The diagnosis can be done with ultrasound (cystic process is shown), cystography (the ventral position of urachal diverticulum and fistula is seen, and a bladder-neck obstruction diagnosed) or fistulography (a connection with the bladder is demonstrated). If infected initial management consists of antibiotics, otherwise cure is accomplished with excision of the urachal remnant with a cuff of bladder (to avoid recurrent infection, stone formation and later development of carcinoma), double layer closure and drainage.

References

Medullary Thyroid Carcinoma

Medullary thyroid carcinoma (MTC) is a rare, solid, thyroid neoplasm with amyloid stroma that arise from the parafollicular C-cells, is inherited as autosomal dominant, and may develop in childhood sporadically or associated with a multiple endocrine neoplasia (MEN) or Familial syndrome. In sporadic cases the presentation occurs in adolescence as a thyroid nodule. Production of thyrocalcitonin by MTC and its precursor (C-cell hyperplasia) permits diagnosis and follow-up, though a significant number of these children are not
cured by surgery due to extensive disease at diagnosis. DNA testing has found that the RET proto-oncogene mutation is associated with MCT development in kindreds of sporadic cases, MEN or familial MTC syndromes. This has permitted early (prophylactic) gland removal in infancy (MEN IIB) or early childhood (MEN IIA) before biochemical or clinical MCT develops. MCT metastasizes to local lymph nodes followed by lungs, bone and liver. Management consists of total thyroidectomy and central lymph node sampling with general sampling if enlarged.

References

Gastric Perforation

Gastric perforation (GP) is a rare abdominal catastrophe seen chiefly in premature infants. GP occurs primarily (spontaneously) after selective ischemia with blood shunting during periods of neonatal asphyxia. Secondary GP is caused by mechanical disruption (excessive distension or instrumentation) as observed in situations such as: aggressive mask resuscitation, duodenal atresia, esophageal atresia with TE fistula, volvulus of the stomach, esophageal intubation, mechanical ventilation and nasogastric tubes (iatrogenic). Boys are more commonly affected than girls. Sudden onset of abdominal distension, feeding intolerance, respiratory distress, metabolic acidosis, shock and hypoactivity within the first week of life is characteristic. Pneumoperitoneum is seen in plain abdominal films. Most GP occurs along the greater curvature of the stomach between the smooth muscle layers. With severely sick infants, temporary peritoneal drainage with lavage removes gas and acid, decompresses the abdomen improving ventilation (abdominal compartment syndrome), and grants time to stabilized the sick infant (improve acidosis, shock and coagulopathy) in preparation for surgical closure of the perforation. GP carries a high mortality rate.

References
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