Situs Inversus

Situs inversus is a defect of lateralization that results in mirror image positioning of abdominal and thoracic viscera. Complete situs inversus is estimated to occur in one per 8000 persons and may be isolated, associated with cardiac or alimentary tract malformations (duodenal obstruction and midgut volvulus), or occur as a component of Kartagener’s syndrome, with autosomal recessive inheritance and associated disturbance of ciliary function, causing bronchiectasis, chronic sinusitis and male infertility. Situs inversus can present with dextrocardia, levocardia or partial heterotaxia. Children with situs inversus and no cardiac lesion are older than two months when diagnosed presenting vague symptoms of intermittent vomiting, abdominal pain, constipation, diarrhea or failure to thrive. The anomaly causes a rotational defect which can cause midgut volvulus. Situs inversus can be diagnosed from prenatal ultrasound. An UGIS and barium enema will help diagnosed the intestinal rotation and fixation anomaly. Liver/spleen scans will determine the presence or absence of the spleen. A prophylactic Ladd procedure has proven to be both safe and effective in preventing intestinal volvulus or obstruction from congenital bands. Some workers advocate use of laparoscopy in malrotation associated with situs inversus since it is minimally invasive, accomplish the same surgical act and is associated with fewer postoperative adhesions.

References:

Pre-duodenal Portal Vein Revisited

Pre-duodenal portal vein (PDPV) is a rare congenital vascular anomaly which is often symptomless leading to duodenal intestinal obstruction requiring surgical correction. PDPV is frequently associated with other congenital malformations, among which, and in order of
frequency, are duodenal stenosis and atresia, abnormalities of the biliary tract (atresia and duplications), annular pancreas, and malrotation (intestinal, situs inversus). The duodenal atresia associated with PDPV is intrinsic and not due to the external pressure of the anomalous vein on the duodenum. PDPV occurs because of persistence of a preduodenal vitelline communicating vein. Most children with PDPV presents with bowel obstruction and two-thirds are detected in the first two weeks of life. Identification of PDPV is rarely made preoperative. Prenatal Ultrasound has demonstrated the unusual position of the portal vein in cases of PDPV. Presence of PDPV complicates any duodenal surgery since the integrity of the vessel must be preserved to avoid portal vein thrombosis. Management consists of duodenoduodenal bypass anastomosis anterior to the portal vein. Another alternative is gastrojejunostomy with truncal vagotomy.

References:

Congenital Melanocytic Nevus

Skin nevus found in infants and children can be a source of concern to both parents and physician dealing with pediatric patients. Congenital melanocytic nevus (CMN) is a characteristic pigmented nevus with mild raised borders and a strong skin discoloration caused by melanin deposits. Incidence of CMN is 0.2% with more than 90% small nevi (< 1.5 cm). Congenital melanocytic nevus may cause cosmetic defects and represent a risk of malignant transformation, namely melanoma. The incidence of developing melanoma is associated with location and size of the CMN; those identified in the face and neck or covering large areas of the body (larger than 5 cm) has a higher incidence of malignant transformation. Not always there is a correlation between pre and postoperative diagnosis after prophylactic excision of a nevi. Digital videomicroscopy using polarize light can help increase the diagnostic yield of CMN. A high index of suspicion for cutaneous melanoma is needed by clinicians assessing melanocytic lesions in children and adolescents for early diagnosis taking into consideration change in color, growth rate, and bleeding. For large lesions and those in exposed areas of the body, surgical excision is the treatment of choice. Medium and small lesions can be managed with Ruby laser treatment. Ruby laser treatment does not result in scarring, mutilation, or functional impairment.

References:
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