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KTS

The Klippel-Trenaunay Syndrome (KTS) is a congenital angiodysplasia described in 1900 by two French physicians consisting of the following triad: 1) soft tissue and bone hypertrophy with overgrowth of the extremity, 2) hemangiomas a/o lymphangiomas, and 3) venous varicosities. A mesodermal abnormality during fetal development leads to arterio-venous communication in the limb bud producing the resulting KTS. Overgrowth of the unilateral lower extremity is commonly found. Hemangiomas are capillary or port-wine nevus (diffuse telangiectasias of the superficial vessels of the dermis). Pelvic extension of hemangiomas may lead to rectal bleeding or hematuria. Varicosities are atypical, occurring in the lateral extremity aspect due to persistence of embryological (sciatic vein) venous channels. Additional anomalies: syndactylia, spina bifida, and equinovarus. Diagnostic work-up includes roentgenograms to document limb length discrepancy, non-invasive arterio-venous evaluation, venography, and MRI. Management is predominantly conservative such as elastic support for varicosities. Surgery is done selectively for cosmetic reasons, marked leg discrepancy, and complications of the hemangiomas or venous insufficiency.

Morgagni Hernias

First described in 1769, Morgagni Hernias (MH) are rare congenital diaphragmatic defects close to the anterior midline between the costal and sternal origin of the diaphragm. They occur retrosternally in the midline or more commonly on either side (parasternally) of the junction of the embryologic septum transversum and thoracic wall (see the figure) representing less than 2% of all diaphragmatic defects. Almost always asymptomatic, typically present in older children or adults with minimal gastrointestinal symptoms or as incidental finding during routine chest radiography (mass or air-fluid levels). Infants may develop respiratory symptoms (tachypnea, dyspnea and cyanosis) with distress. Cardiac tamponade due to protrusion into the pericardial cavity has been reported. The MH defect contains a sac with liver, small/ large bowel as content. Associated conditions are: heart defects, trisomy 21, omphalocele, and Cantrells'

pentology. US and CT-Scan can demonstrate the defect. Management is operative. Trans-abdominal subcostal approach is preferred with reduction of the defect and suturing of the diaphragm to undersurface of sternum and posterior rectus sheath. Large defects with phrenic nerve displacement may need a thoracic approach. Results after surgery rely on associated conditions.

GD

Gallbladder Dyskinesia (GD) is uncommonly reported in the pediatric age group. Portrays a motility disorder of the biliary system characterized by poor contractility of the gallbladder causing symptoms similar to those of gallstone disorders, but with a more protracted clinical course. Believed to be caused by spasm of the sphincter of Oddi associated to either a hypersensitivity of the gallbladder or hyposensitivity of the sphincter of Oddi to cholecystokinin (CCK). The result is a gallbladder contracting against a closed biliary system. Diagnosis of GD is considered when the ejection fraction of the gallbladder content is less than 35% during a hepatobiliary scan (DISIDA) stimulated with CCK. Twelve children (14%) out of 85 underwent laparoscopic cholecystectomy (LC) at San Pablo Medical Ctr. during a sixty-six-month period for GD. Mean age was 14 years and classic biliary symptoms predominated (RUQ pain and fatty food intolerance). Mean ejection fraction was 16.8%. Pathology specimens showed ten cases with mild to moderate chronic cholecystitis (83%), and two unremarkable. These changes correlated with the mean duration of symptoms. Clinical improvement after surgery was seen in most cases. We believe that LC should be offered to symptomatic children with low ejection fractions if thorough work-up fails to show other GI disorder.

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