



PEDIATRIC SURGERY UPDATE ©

Vol 06 No 03 MARCH 1996

SVCS

Superior vena cava syndrome (SVCS), first described by Hunter in 1757, refers to a constellation of signs and symptoms caused by severe reduction in venous return from the head, neck, and upper extremity. The pathogenesis relate to either extraluminal (tumor, mass) or intraluminal (thrombosis) compression of the superior vena cava. The term superior mediastinal syndrome (SMS) is use interchangeably when tracheal compression causing extrinsic respiratory obstruction manifests. In children the most common initial symptom is cough, followed by wheezing, tachypnea, dyspnea, and orthopnea that aggravates lying down (supine) or bending forward. Face, neck and arm swelling may be present. Severity of the SVCS depends on the rapidity of occlusion and collateral vessel development. Most fear complications are cerebral and laryngeal edema or tracheal compression. Non-Hodgkin's mediastinal lymphoma is the leading etiology of SVCS in the pediatric age, followed by cardiac surgery, histoplasmosis mediastinal fibrosis and indwelling venous catheters. CT evaluation estimates the degree of tracheal compression. The strategic approach toward mediastinal tumors causing SVCS/SMS is controversial; the result of the risk of unexpected tracheo-bronchial obstruction during muscle relaxant use. Tissue diagnosis is needed to institute correct therapy. Least invasive diagnostic procedure such as bone marrow aspiration, peripheral lymph node biopsy (IV ketamine with local infiltration), diagnostic thoracentesis (surface marker analysis), or CT guided percutaneous biopsy avoiding profound sedation should be tried first. For open biopsy spontaneous ventilation in sitting position should be preserved, lower extremity IV lines secure, muscle relaxant avoided, if possible, and bronchoscopic instrumentation available.

Chylous Ascites

Chylous ascites (CA) is a rare clinical entity, the result of either intrinsic/extrinsic obstruction of lymphatic drainage or traumatic rupture of lymphatic channels in the mesentery. Chyle leaks through a fistulous tract, exudate through the wall of retroperitoneal lymphatic or from dilated lymphatic in the wall of the bowel or mesentery with proximal obstruction at the base of

the mesentery, cisterna chyli or thoracic duct. Characteristically the fluid has: a milky appearance, separates on standing, fat content > 1 gm/dl, total protein > 3 gm/dl, high content of chylomicrons, triglycerides and lymphocytes, and specific gravity above 1.012. Most cases in children are congenital in nature (lymphangiectasia, cystic hygroma of the abdomen, lymphatic dysplasia, etc.). Others have an inflammatory, traumatic, surgical, or neoplastic etiology. Infants are more commonly affected and boys outnumber girls 2:1. CA generally develops insidiously with painless abdominal distension (ascites), weight loss, increase abdominal girth, hypoproteinemia and inanition. Other patients develop acute abdominal symptoms when associated to intestinal obstruction, intussusception or volvulus. Diagnosis relies on the character of the fluid on paracentesis. Lymphangiography can delineate the retroperitoneal lymph pathways but has no direct access to mesenteric or bowel wall lymphatics. Management depends on effective alleviation of etiologic factors. If after extensive work-up the cause is unknown medical therapy using low fat-high protein diet, bed rest, diuretics and repetitive paracentesis should be tried for four weeks. With no improvement, exploratory laparotomy should follow to establish a diagnosis. Sudan III dye given by mouth six hours before surgery helps identify leaking ducts. Intractable ascites have been managed using peritoneo-venous shunting (LeVeen) with mix results. Mortality is related to the specific underlying disease and has been cited as 24% in some series.

NET Address!

Since January 1996 'Pediatric Surgery Update' has an Internet homepage. See the address below and come visit us.

*Edited by: **Humberto L. Lugo-Vicente, MD, FACS, FAAP**

P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico 00922-0426.

Tel (787)-786-3495 Fax (787)-720-6103

E-mail: titolugo@coqui.net Internet Address: <http://home.coqui.net/titolugo>