



# **PEDIATRIC SURGERY Update** ©

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### **Chylothorax**

Effusion of lymph (chyle) into the pleural cavity is known as chylothorax. Chyle is clear-milky fluid with an elevated total protein and albumin level, a specific gravity above 1.012, the presence of WBC with lymphocyte predominance (80%), and elevated triglyceride (chylomicrons). In children is a potentially life-threatening disorder that has profound respiratory, nutritional (hypoalbuminemia), electrolyte (hyponatremia) and immunologic (lymphopenia, hypogammaglobulinemia, T-cell depletion) effects. Chylothorax has a congenital (mediastinal lymphangiomatosis), acquired or idiopathic origin. Acquired chylothorax is most commonly found; the result of a direct lesion of the thoracic duct or lymphatic vessels by trauma (thoracotomy, central venous catheters or chest tubes insertions), during cardiac surgery, mediastinal malignancy (neuroblastoma) or infection, repair of a diaphragmatic hernia or associated with superior vena cava obstruction (thrombosis). Initial management consists of: 1- chest tube drainage after failed thoracentesis (pleural space tamponade), 2- medium-chain triglyceride enriched formula for a week (lymphatic decompression), 3- TPN if chylothorax increases or persists. More protracted course (4 week medical tx) will require surgery to locate and suture ruptured subpleural lymphatics, ligate the thoracic duct, do chemical pleurodesis or place a pleuroperitoneal shunt. Those associated with venous obstruction or increase right sided cardiac pressure produce more volume, persist longer and are more difficult to manage.

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## Multiple Endocrine Neoplasia Type 1

Multiple endocrine neoplasia type 1 (MEN-1) is an autosomal dominant disorder characterized by the combined occurrence of parathyroid, pancreatic islet and anterior pituitary tumors. A single inherited locus on chromosome 11, band q13, causes MEN-1. Primary hyperparathyroidism (HPT) is the most common lesion of MEN-1. The albumin corrected total calcium is usually normal, but the ionized calcium and PT hormone is elevated. Once elevated the patient develops insidious complication from hypercalcemia (pancreatitis, peptic ulcer disease, muscle pains, weakness, neuropsychiatry disorders and nephrolithiasis). Surgical management of primary HPT in MEN-1 is controversial. The salient features of HPT in MEN-1 are a high incidence, if not universal occurrence of multiglandular disease, an operative failure rate because of failure of both to identify all four glands and to perform a radical resection, and a significant incidence of recurrent disease, sometimes cause by supernumerary or ectopic gland involvement. Surgical principles should be (1) identification of all four glands, (2) subtotal resection to ensure cure and facilitate possible reoperation, and (3) excision of supernumerary thymic glands. Extirpation of a single gland as a general primary procedure is inadequate causing recurrence. Although many patients with primary HPT and MEN syndrome have multiple abnormal parathyroid glands, two populations of patients exist; one population has solitary or double adenomas and recurrence is uncommon, whereas the other population of patients has hyperplasia and persistent or recurrent disease is common.

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## Ranula

The word ranula comes from the Latin: *rana*, frog. Ranula is a large sessile cyst of the sublingual salivary gland in the floor of the mouth under the tongue. The lesion can be as small as a pea-sized cyst to one side or the other of the frenulum, or an enormous blue-gray translucent swelling that fills the mouth and cause respiratory problems. Two ranula varieties are described: a superficial, epithelial lined cyst resulting from ductal obstruction, and a cervical pseudocyst without epithelial lining resulting from extravasation of saliva (plunging) that dissects through the tissue planes of the neck and appear as a neck mass. In both cases management consists of excision of the thin-walled sac and sublingual gland

if possible, or marsupialization to the oral cavity.

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