PEDIATRIC SURGERY *Update* ©
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## Dermatofibrosarcoma Protuberans

Dermatofibrosarcoma protuberans (DFSP) is a rare, low grade malignant soft tissue tumor of the skin and subcutaneous tissue with a high propensity for local invasion and recurrence. Though mostly seen in middle age, a few cases have been described in infants and children between the ages of 14 months and 12 years. Clinically, DFSP presents as an asymptomatic raised, firm nodular lesion fixed to the skin but mobile over the deep fascia, with a pattern of slow, progressive growth. Diagnosis is established after excisional biopsy or punch biopsy. The most common location of this tumor is the trunk followed by the extremities. CT-scan is useful to determine the tumor extent of penetration. DFSP has two histologic variants: the more typical low-grade tumor, and a high-grade rare fibrosarcomatous variant demonstrating necrosis, high mitotic rate (> 10 mitoses high-power fields) and presence of pleomorphic areas. This last variant is associated with a poor clinical outcome. Metastasis is rare. Lung metastasis are most common, while lymph node metastasis is exceedingly. Rare. Wide surgical excision using a margin of three centimeters with inclusion of superficial fascia is currently the standard therapy in children. Mohs micrographic surgical excision has wide acceptance among adults cases. DFSP is a radiosensitive tumor.

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## Congenital Cystic Adenomatoid Malformation

Congenital Cystic Adenomatoid Malformation (CCAM) is a rare lung bud lesion characterized by the presence of a multicystic mass of immature pulmonary tissue due to abnormal development of lobar and segmental bronchi. Embryologic origin is cessation of bronchiolar maturation with mesenchymal overgrowth occurring in the late 5th or 6th weeks of gestation. CCAM is divided into Type 1 (large, > 2 cm, irregular widely space cysts; the most common type), Type II (smaller, < 1 cm, closer together; more numerous cysts resembling dilated bronchioles), and Type III (large, solid appearance with small unrecognizable cyst). Due to communication between bronchiole-like structure and cyst significant air trapping can occur causing acute life-threatening respiratory distress (tachypnea, cyanosis and chest wall retraction) in one-third of cases. The other clinical presentation is late-onset pulmonary infection (fever, cough and dyspnea). Most cases have unilobar involvement. Bad prognostic factors associated with CCAM are fetal hydrops, polyhydramnios, increase mediastinal shift, large size of the mass and presence of pulmonary hypoplasia. Postnatal diagnosis is confirmed with chest CT-Scan. Symptomatic infants with CCAM should undergo immediate resection of the lung lesion (usually lobectomy). Limited lung resection (segmentectomy) preserves more functioning lung parenchyma and represents an alternative in bilobar disease. Since asymptomatic lesions can undergo spontaneous regression, a policy of watchful observation with periodic imaging followed by elective resection is justified. Children with large lesions (> 50%) of the hemithorax) has a significant chance of requiring surgical intervention. Better height and weight growth has been found in patients operated upon in later childhood compared with those operated in infancy.

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## Retching

Retching, choking and gagging are severe, debilitating oral-motor dysfunctional complex symptoms seen rarely after fundoplication and gastrostomy placement in neurologically impaired children. Retching is not a symptom of reflux, but the first part of the ejection phase of the emetic reflex consisting of rhythmic contraction of the entire diaphragm and anterior abdominal wall muscles. The etiology of this complex symptom is still not known. It is argued that following fundoplication gastric dysrhythmia ensues due to vagal afferent damage, gastric mucosal injury, altered morphology or increased gastric filling. Initial management of postoperative retching includes antiemetics (dopamine or hydroxytryptamine receptors antagonists) such as domperidone or ondansetron. If symptoms persist, the child can be paced in continuous gastric feedings. With no improvement jejunal feedings bypassing the stomach to minimize gastric vagal afferent stimulation results in significant relief. Children who retch preoperatively or have a hypersensitive emetic reflex has three times the risk of retching after fundoplication. Retching can cause wrap migration or disruption. Development of a paraesophageal hernia from failed crural repair is a surgical correctable cause of postoperative retching after fundoplication.

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