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Spontaneous Pneumothorax

Most pneumothorax in children are the result of blunt/open chest trauma, mechanical ventilation (barotrauma), bronchial asthma or an infectious pulmonary process. Primary spontaneous pneumothorax (PSP) is rare in children with most cases seen in adolescent males with thin body habitus. Main presenting symptoms consist of chest pain, cough and shortness of breath. Recurrence is high in this older population of children. PSP is usually the result of: 1) a ruptured apical bleb or bullae in three-fourth cases, 2) destructive parenchymal disease (cystic fibrosis, AIDS), or 3) alveolar rupture due to proximal airway obstruction. Initial management consists of oxygen supplementation for small pneumothorax less than 15% with no tension physiology present. Chest tube drainage is needed for medium or large size pneumothorax. Recurrence or persistent pneumothorax is managed with video-assisted thoracoscopic surgery (VATS) by ablating with endoscopic stapling (Endo GIA), suturing or ligating using an endoloop technique the apical bullae followed by pleurodesis. Pleurodesis can be done chemically or surgically. Chemical pleurodesis is achieved with such agents as talc, tetracycline, bleomycin or quinacrine instillation. Mechanical pleurodesis carries a lower recurrent rate and can be achieved by abrasion or electrocoagulation. Most common complication is persistent air leak. VATS is a fast, cost-effective method of treatment for PSP with less morbidity.

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Gastrointestinal Autonomic Nerve Tumor

Gastrointestinal Autonomic Nerve Tumor (GANT), also known as plexosarcoma, is a very rare recently described aggressive malignant spindle cell tumor that arises from the autonomic myenteric plexus of the bowel wall. Tumor mostly originates in the small

intestine and stomach with a clinical course characterize by either local or distant metastases (liver). Histologically GANT is a low-grade, epithelioid or spindle-cell neoplasms that can be distinguished from the other gastrointestinal stromal tumors on the basis of its unique electron microscopy ultrastructural features. Distinction of GANT from other stromal tumors is not possible based on imaging studies. The tumor is solid and cystic, hemorrhagic, often transmural and usually involving omentum, mesentery and retroperitoneum. Signs and symptoms include abdominal pain, mass, gastrointestinal bleeding, perforation and anemia. Tumor can present with volvulus. In children they occur in females during adolescent years (mean age 12 years) mostly involving the stomach. Primary management consists of surgical resection which is curative, leaving chemotherapy for residual or recurrent tumor. Prognosis is good for younger patients.

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Allgrove Syndrome

Achalasia, Alacrima & isolated Adrenal Insufficiency is also known as triple-A or Allgrove syndrome. The high degree of consanguinity usually identified supports an autosomal recessive mode of inheritance for this disorder. Alacrima is the earliest and most consistent clinical sign of Allgrove syndrome. All affected children shows esophageal dysmotility even in the absence of symptomatic dysphagia. The adrenal insufficiency is due to adrenocorticotropic hormone (ACTH) resistance. Other associated neurological abnormalities include autonomic, sensory, and upper- and lower-motor neuropathy, deafness, and mental retardation. Genetically the syndrome has been mapped to mutations in chromosome 12q13 region near the type II keratin gene cluster. Though most cases arise in children between the ages of two and eight years, a few cases originate during early adult years. Management for alacrima consists of artificial tears replacement. Adrenal insufficiency will need steroid replacement. Achalasia is treated with a modified Heller esophagomyotomy which can be done open or laparoscopically. A few neurologically impaired children will need gastrotomy feedings.

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