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Cricopharyngeal Achalasia

Congenital cricopharyngeal achalasia (CCA) is an important but relatively seldom diagnosed cause of dysphagia in children caused by failure of the cricopharyngeal muscle to relax at the appropriate time during the third period of swallowing mechanisms in the absence of other motor abnormalities. Age of initial presentation ranges from birth to six months. Symptoms include failure to thrive, regurgitation of food, choking, cyanosis, nasal reflux, coughing and recurrent aspiration pneumonia. Cine-esophagogram with fluoroscopic observation of the swallowing mechanism will establish the diagnosis in most children. The characteristic sign is a round and regular posterior narrowing on the posterior wall of the esophagus at the level of C4-C5 with enlargement of the hypopharynx. Esophageal motility studies will quantify changes and also evaluate lower esophageal dysfunction not easily identified in esophagograms. Though CCA can rarely be found as an isolated condition, it is usually associated with neurologic deficit as seen in myelomeningocele and Arnold Chiari malformations. Management of CCA includes positioning, nasogastric lavage feedings, balloon dilatation of the upper esophagus or surgical myotomy of the cricopharyngeal muscle. If all other measures fails, cricopharyngeal myotomy is a safe and effective operation with excellent results. Symptomatic relief is immediate and complete with no long-term recurrence documented.

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Ganglioneuroma

Ganglioneuroma is a rare and benign neural mature crest tumor of the sympathetic nervous system. They arise wherever sympathetic tissue exists and may be seen in the neck, posterior mediastinum, adrenal gland, retroperitoneum, and pelvis. The clinical presentation of most patients is that of an asymptomatic slow growing solid mass in an older child. On many occasions the tumor is found incidentally after simple chest films or

abdominal imaging studies. Imaging studies cannot differentiate a ganglioneuroma from its malignant counterpart neuroblastoma. Rarely the child develops respiratory problems, painless spinal deformity or neurologic deficit before the mass is found. Most of these tumors are hormone silent. Histologically, ganglioneuroma is completely differentiated and composed of mature ganglion cells, Schwann's cells and neuropils. Ganglioneuromas frequently produce somatostatin and vasoactive intestinal peptide (VIP). Management consists of surgical excision whenever possible. Transperitoneal laparoscopic adrenalectomy has been safely performed when this tumor arises from the adrenal gland.

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Thoracoscopy

As we enter the 21st century, minimal invasive surgical procedure will continue to demonstrate its superiority over conventional open surgery in terms of pain control, convalescence, hospital stay, cosmesis and achieving its purpose. Thoracoscopy using video-endoscopic technique is reliantly replacing open thoracotomy in many chest conditions in children. As diagnostic aid, thoracoscopy can be use for biopsy of lung, pleural, foregut and mediastinal benign and malignant masses. Therapeutically, thoracoscopically has been utilized for excision of esophageal duplication cysts, closure of patent ductus arteriosus, pleurodesis, resection of lung (bullectomy, lobectomy or segmentectomy), thymectomy, anterior spinal fusion procedures for scoliosis, management of empyema, and resection of mediastinal cysts and tumors. Almost the entire anterior and posterior mediastinum is visible by thoracoscopy. Advantages of thoracoscopic procedure consist in less pain, less compromise of pulmonary reserve, shorter hospital stay and costs. Contraindications for thoracoscopy consist of inability to develop a pleural window with the scope and patients in high pressure mechanical ventilation in need of lung biopsy due to the high incidence of postop air leak. Once the thoracoscope is in you should always perform an evaluation of the anatomy before committing the child to an open thoracotomy.

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