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CHAOS

Congenital high airway obstruction syndrome (CHAOS) is a term coined in 1994 to development of complete or near-complete obstruction of the fetal airway. Though rare, CHAOS is not always incompatible with life. CHAOS is thought to occur when the upper airway fails to recanalize around the 10th week of gestation causing the obstruction. This obstruction blocks the normal egress of fluid from the lung raising the intratracheal pressure and leading to distension of the tracheobronchial tree and proliferative lung growth. The lung expands, the diaphragm flattens and the heart is compressed in the midline. The elevated intrathoracic pressure causes decreased venous return, fetal cardiac failure with ascites, placentomegaly and hydrops fetalis. Diagnosis is performed with prenatal ultrasound and MRI with findings such as dilated airways distal to the obstruction, large echogenic lungs, flattened diaphragm, fetal ascites and hydrops. Most common cause of CHAOS is laryngeal atresia, followed by subglottic stenosis, laryngeal/tracheal webs or agenesis. The prenatal natural history and postnatal course of CHAOS depends on whether the airway obstruction is complete. Survival depends on emergency tracheostomy immediately after birth with the use of the ex-utero intrapartum treatment (EXIT) procedure. The presence of hydrops is an ominous sign. The presence of a pinpoint laryngotracheal or tracheoesophageal fistula in some fetus with CHAOS will allow airway decompression with resolution of ascites and diaphragmatic eversion.

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Thoracic Neuroblastoma

Neuroblastoma is the most common malignant solid tumor in children with almost 20% of them arising in the thorax. Thoracic neuroblastomas have a better prognosis than those occurring in other parts of the body. Most cases present before the age of one year. Mediastinal neuroblastoma becomes symptomatic earlier and can be detected in an earlier favorable stage. The cell of origin is the dorsal root sympathetic ganglion cell. In what respect biology markers, thoracic neuroblastomas have favorable biological profiles such as DNA index greater than one, significant lower N-myc amplification, LDH level < 1500 and low ferritin levels. Clinically the child might demonstrate cough, dyspnea, wheezing, neurogenic cord compression, dancing eyes' syndrome and Horner syndrome. For diagnosis the chest films suggest the presence of a posterior mediastinal mass. CT scan is sensitive predicting chest wall involvement. MRI is very sensitive for predicting lymph node extension, intraspinal extension and chest wall involvement. Management consists of complete surgical excision with adjuvant chemotherapy. Thoracoscopic resection of neurogenic tumors achieved similar local control and disease-free survival when compared with open resections. Results that were accompanied by shorter hospital stay and decreased blood loss.

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Anal Achalasia

Internal anal sphincter achalasia (IASA) is a condition with similar clinical presentation to Hirschsprung's disease, but with ganglion cells present in the rectal biopsy. Theories on pathogenesis include nitregic nerve depletion, defective innervation of the neuromuscular junction and altered distribution of interticial cells of Cajal, considered the pacemakers of the bowel. Children with anal achalasia present with severe constipation with or without soiling. Diagnosis of IASA is made with anorectal manometry which shows the absence of the rectoinhibitory reflex upon rectal balloon inflation with presence of ganglion cells and normal acethycholinesterase activity in a rectal biopsy. Management consists of posterior internal sphincter myotomy. Intrasphincteric injection

of Clostridium botulinum toxin has also been tried causing a local and six-month transient denervation of the sphincter. The majority of patients with internal anal sphincter achalasia can be treated successfully by internal sphincter myectomy. In the long term, a significant number of patients have been found to suffer from soiling-related social problems.

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