



# **PEDIATRIC SURGERY Update** © **Vol. 46 No. 01 JANUARY 2016**

## **Congenital Paraesophageal Hernia**

Congenital paraesophageal hernia (CPH) is the rarest of all diaphragmatic hernia of childhood. As the name implies, the patient is born with the defect. Most paraesophageal hernias are acquired after procedures performed in the GE junction (antireflux procedures). In CPH the defect can go unnoticed for several years or present with recurrent chest infections or vague gastrointestinal symptoms. Left alone CPH can cause high morbidity or even mortality hence the importance of an early diagnosis. CPH occurs because of absent or abnormally lax anatomic anchors of the stomach and gastroesophageal junction associated to a paraesophageal defect. The fundus is pushed through the defect by increase intraabdominal pressure entering the thoracic cavity causing lung compression and mediastinal shift. If this progress the antrum also enters the thorax and organoaxial volvulus of the stomach occur causing gastric obstruction and esophageal dilatation. With enlargement of the defect bowel and omentum can herniate. There is absence of pulmonary hypoplasia. Clinically PEH can present acutely with incarceration, obstruction, gangrene, perforation, bleeding, anemia, as well as acute respiratory complications. Other times the child has chronic chest infections with intermittent vomiting. Initial work-up should include chest film, upper gastrointestinal series and abdominal CT-Scan to determine herniated content and detect complications of affected lungs. Associated anomalies with CPH include short esophagus and microgastria. Surgical repair should be done promptly to avoid serious complications. The procedure can be performed open or laparoscopically and aims to resect the hernia sac, reduce the herniated content, close the hiatal defect, anchor the stomach or do a partial antireflux procedure, and check for associated malformations.

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## **Intradiploic Dermoid Cysts**

Dermoid cysts are congenital benign neoplasms derived from both ectoderm and mesoderm tissue layers. They typically occur in childhood during the first or second decade of life. They are composed of a keratinizing squamous epithelium present together with dermal derivatives such as hair follicles, smooth muscle, sweat gland, sebaceous gland and fibroadipose tissue. One-tenth of all dermoid cysts occurs in the region of the head and neck in locations such as periorbital, nasal, frontotemporal, submental and suprasternal. Dermoid cysts of the head and neck are thought to be the congenital inclusion type. The periorbital region is the most commonly reported site for dermoid cysts of head and neck, most commonly near the zygomaticofrontal suture, laterals to the lateral canthus. Some dermoid cysts exhibit intermittent discharge of sebaceous material or recurrent local infection. In the temporal bone dermoid cysts can be mistakenly labeled as a first branchial cleft or pouch anomalous cyst. Lesions in the midline of the cranium are more concerning for intracranial extension. Dermoid cysts that are located in the nasal bridge or temporal area, produce recurrent infection or exhibit a draining sinus should alert the physician toward the presence of intracranial extension warranting further imaging with CT-Scan or MRI to plan excision. Intradiploic dermoid cysts are a rare subtype of the frontotemporal dermoids occurring most commonly at the bregma. They may appear as a lytic defect in the skull. The overwhelming majority of periorbital and frontotemporal dermoids are superficially located and amenable to treatment with simple excision. Complete en bloc surgical resection with careful follow-up is the treatment of choice for dermoid cysts.

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## **Esophageal Atresia with Right Aortic Arch**

Almost 5% of children with esophageal atresia (EA) with or without an associated tracheoesophageal fistula have a right aortic arch (RAA). Preoperative localization of the aortic arch position previous to repair of EA is of utmost importance to avoid an extremely difficult right thoracotomy. Preoperative localization of a RAA using echocardiography correctly identifies the arch position in two-thirds of patients. The surgeon needs to reliably identify the position of the aortic arch, the laterality of the

descending thoracic aorta and the nature of any associated congenital heart disease. If an aortic arch passes over the right mainstem bronchus, it will descend through the right side of the chest 95% of the time, and there will be complete absence of any retro-esophageal component to the aorta. Most will have mirror-image branching of the innominate and subclavian arteries. Infants with EA with RAA have more vascular rings, cardiac anomalies and operative complications compared with those with LAA. Up to 50% of children with EA and RAA have associated cardiac malformations, especially ventricular and atrial septal defects. RAA is found in 25% of cases of Tetralogy of Fallot. Should the suspicion of a RAA in EA arise, further diagnostic evaluation using CT-angio or MRI will elucidate the exact anatomic variant. When RAA is associated with EA, the gap length between the esophageal pouches is usually increased. A left thoracotomy in the face of RAA allows one to gain better exposure to the esophagus and tracheoesophageal fistula without the obstruction of the aorta. In addition, owing to the type of vascular ring that can be encountered the left-sided thoracotomy allows access to ligate the PDA or manage the vascular ring. Should a RAA be unexpectedly encountered during a right thoracotomy and the anastomosis is not technically feasible a delayed left thoracotomy permits more accurate evaluation of the vascular anatomy.

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