



## **PEDIATRIC SURGERY Update** ©

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### **Gastro-Esophageal Reflux Disease**

Gastro-esophageal reflux disease (GERD) has two distinct forms in children. In infants, reflux causes delayed growth & development, recurrent respiratory infections and life-threatening situations. Most symptoms resolve when the valvular competence of the cardia develops in the second year of life. Older children manifest symptoms of dysphagia and substernal burning. We manage most children medically. Indications for surgery consist of failure to thrive, esophagitis, stricture, chronic aspiration pneumonia, life-threatening events and Barrett metaplasia changes. Children referred for surgery should have an esophagogram, endoscopy, and evaluation of gastric emptying mechanisms to document: magnitude of reflux, presence of pharyngeal incoordination, dysmotility, strictures, malrotation, and grade of esophagitis. pH studies and milk scans may further find a cause and effect relationship between reflux and respiratory problems. Surgical options consist of partial (Thal, Boix-Ochoa, Toupe) or complete wrap (Nissen) fundoplasty reconstruction. The procedure can be done open or laparoscopic. Those kids with delayed gastric emptying will benefit from a gastric emptying procedure (pyloroplasty, antroplasty). Neurologically impaired children referred for feeding gastrostomy should undergo a similar work-up to identified potentially dangerous reflux problems. Alternatively the gastrostomy can be constructed percutaneously and the problems of GERD be assessed later in life. Neurologic status and gastric emptying are major predictors of operative success.

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### **Laryngo-Tracheal Clefts**

Laryngo-tracheal clefts (LTC) are rare congenital anomalies that can involve the larynx or the laryngo-tracheal and esophageal wall. Subtypes of LTC occur between the aerodigestive system and can be limited to the larynx up to involve all the way to reach the

carina or the right main bronchus (Subtypes: type 1 to the cricoid, type 2 involving the cricoid, type 3 in cervical trachea and type 4 into the thoracic trachea). LTC arises from errors in chondrification and fusion of the laryngeal supporting cartilage or tracheo-esophageal folds. LTC are associated to the "G" and Pallister-Hall syndrome, to esophageal atresia and to anal malformations. As neonate they present a hoarse cry, inability to handle secretions, cyanosis, choking, coughing, stridor and recurrent pneumonia depending on the length of the cleft. Diagnosis of LTC is made by endoscopy. Management of type 1 is conservative or endoscopically depending on symptoms. For type 2 to 4 defects initial tracheostomy for securing airway and gastrostomy for feeding is needed. This is followed by repair of the LTC using an anterior laryngeal approach for type 2 & 3 and combined cervico-thoracic approach for type 4 and those associated with esophageal atresia. Morbidity (leaks, pharyngoesophageal incoordination and reflux) and mortality is very high. Early suspicion and diagnosis are crucial.

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### Wilms Genetics

Wilms tumor (WT) development involves at least three genes. The first of these identified as WT1 is a suppressor gene mapped to a deletion of chromosome 11p13 found in sporadic and hereditary cases. WT1 required for normal renal development encodes a zinc finger binding protein that is important in regulating the formation of the early nephron. Mutations of WT1 are found in WT associated with aniridia, genitourinary defects (hermaphroditism) and mental retardation. The second WT suppression gene is WT2, known to be involved in the Beckwith Wiedemann locus located in the 11p15 region. The WT3 locus is likely to be found in the long arm of chromosome 16q and is suspected of tumor progression rather than initiation. WT like mutations will help determine if they are additional indicators of clinical behavior and outcome.

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