



# **PEDIATRIC SURGERY Update** ©

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### **Fowler-Stephen Orchidopexy**

Undescended testis (UT) is the most common congenital anomaly of the genitalia of newborn males. It is more common in prematurely born infants. It is estimated that 20% of undescended testis are non-palpable. Non-palpable UT might be intraabdominal, canalicular, atrophic or absent. Orchidopexy in UT should be performed before the age of 18 months to avoid damage to spermatogonia. Diagnostic laparoscopy is the gold standard maneuver to determine localization and eventual management of non-palpable UT. Laparoscopy is better than CT-Scan, MRI, gonadal arteriography or venography in localizing the presence, distance and size of a nonpalpable UT. Blind ending vessels without a distal testis determine absence and no further management is required. If vessels and vas enter the internal spermatic ring, the child probably has a canalicular testis or nubbin in either case it should be moved and fixed to the scrotum or removed respectively using an inguinal incision. Atrophic or hypoplastic testes are removed during the diagnostic procedure. Intraabdominal testes are either brought down to the scrotum and fixed if the vascular pedicle permits such maneuvers (laparoscopic assisted orchidopexy) or staged its descent using Fowler-Stephen technique (FST). Two-stage FST is performed if the testis is at a high position estimated as more than 2 centimeters between the manipulated testis and the internal ring. The technique consists of clamping and dividing the vascular supply to the testis and leaving it alone so that collateral blood supply develops from the vas deferens. FST can be performed one or two staged procedure. The one-stage FST preserved the gubernaculum for additional collateral blood supply. In the second stage of FST orchidopexy is performed six months later descending the testis with vas deferens-based circulation by fixing it to the scrotum using laparoscopy and inguinal surgery. Success rate for one stage FST can approach 70% which are encouraging. The most common complication after performing either FST is testicular atrophy.

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## Epiphrenic Esophageal Diverticulum

Epiphrenic esophageal diverticulum (EED) is a rare out pouching of the lumen of the esophagus originating in the distal end of the esophagus typically four to 10 cm above the cardia and usually projecting from the right posterior wall. EED is a false pulsion diverticulum induced by high intraluminal pressure on the lower esophagus. Herniation of the mucosa and submucosa through the muscle layers of the esophageal wall occurs. Only 15-20% of patient who harbor an EED have symptoms. EED predominant symptoms include dysphagia followed by regurgitation, chest pain, heartburn, weight loss and less frequent atypical respiratory symptoms. The diagnosis of EED should be made performing barium swallow, upper endoscopy and manometry. The size of the diverticulum influences the severity of symptoms. The average size of the diverticulum is 5.5 cm and is associated with an esophageal motility disorder such in 60% of all cases, achalasia leading the list followed by nutcracker esophagus and nonspecific esophageal motor disorder. The etiology of epiphrenic esophageal diverticulum involves an increased lower esophageal pressure and congenital weakness of the esophageal wall associated with a motor esophageal disorder such as achalasia or diffuse esophageal spasms. The management of an EED consists of laparoscopic diverticulectomy using a linear stapler, long myotomy on the contralateral side of the diverticulum extended to the gastric wall and an anterior Dor partial fundoplication. Complications include leakage at the diverticulum stapled base, empyema, paraesophageal hernia, pneumothorax, atelectasis, pleural effusion and bleeding. Most patient improves significantly with the proposed plan of management.

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## Icteropyloric Syndrome

Hypertrophied pyloric stenosis (HPS) is the most common surgical cause of vomiting in the first two months of life occurring mostly in males infants. Management consists of open or laparoscopic pyloromyotomy after adequate hydration from persistent vomiting. Along with HPS some infants develop concomitant jaundice, referred as the icteropyloric syndrome (IPS). The prevalence of icteropyloric syndrome ranges from 8-10% of children with HPS. Initially it was thought that jaundice in the setting of HPS was due to dehydration, mechanical obstruction, decreased carbohydrate intake, decreased hepatic perfusion or a combination of such factors. Later it was believed that these children had Gilbert syndrome genotype. Infants with IPS are younger than infants with HPS suggesting a role for physiologic jaundice. Although prolonged physiologic jaundice could represent a manifestation of Gilbert syndrome, it does not persist into the second month of life. Children with IPS have a significant higher serum bicarbonate and lower serum chloride levels than non-jaundice infants due to greater losses of hydrochloride acid. Metabolic stress potentiates the manifestation of jaundice in infants with IPS along with hepatic levels of residual enzyme activity, bilirubin load and duration of fasting. A Gilbert syndrome genotype is found in 45-65% of infants with IPS. The risk of having Gilbert syndrome genotype is four times higher in IPS than HPS suggesting Gilbert syndrome plays a role in infants with IPS due to molecular defects within the gene promoter. Children with IPS should have genetic workup for Gilbert syndrome mutations.

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