



# **PEDIATRIC SURGERY UPDATE ©**

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### **Intestinal Neuronal Dysplasia**

Intestinal Neuronal Dysplasia (IND) is a colonic motility disorder first described in 1971 by Meier-Ruge associated to characteristic histochemical changes of the bowel wall (hyperplasia of submucous & myenteric plexus with giant ganglia formation, isolated ganglion cells in lamina propia and muscularis mucosa, elevation of acetylcholinesterase in parasympathetic fiber of lamina propia and circular muscle, and myenteric plexus sympathetic hypoplastic innervation), also known as hyperganglionosis associated to elevated acetylcholinesterase parasympathetic staining. The condition can occur in an isolated form (either localized to colon or disseminated throughout the bowel), or associated to other diseases such as Hirschsprung's (HD), neurofibromatosis, MEN type IIB, and anorectal malformations. It is estimated that 20-75% of HD cases have IND changes proximal to the aganglionic segment. Clinically two different types of isolated IND have been described: Type A shows symptoms of abdominal distension, enterocolitis, bloody stools, intestinal spasticity in imaging studies (Ba Enema) since birth, is less common and associated with hypoplasia of sympathetic nerves. Type B is more frequent, symptoms are indistinguishable from that of HD, with chronic constipation, megacolon, and repeated episodes of bowel obstruction. Management depends on clinical situation; conservative for minor symptoms until neuronal maturation occurs around the 4th year of life, colostomy and resectional therapy for life threatening situations.

### **Prenatal Cystic Hygroma**

Cystic hygroma (CH) is an uncommon congenital lesion of the lymphatic system appearing as a multilocular fluid filled cavity most commonly in the back neck region, occasionally associated with extensive involvement of airway or vital structures. The etiology is intrauterine failure of lymphatics to communicate with the venous system. Prenatal diagnosis can be done during the first trimester of pregnancy as a huge neck tumor. Differential diagnosis includes teratomas, encephalocele, hemangiomas, etc. There is a strong correlation between prenatal dx and Turner's syndrome (> 50%), structural defects (Noonan's syndrome) and chromosomic anomalies (13, 18, 21). Early

diagnosis (< 30 wk gestation) is commonly associated to those anomalies, non-immune hydrops and dismal outcome (fetal death). Spontaneous regression is less likely but can explain webbed neck of Turner and Noonan's children. Prenatal dx should be followed by cytogenetic analysis: chorionic villous sampling, amniocentesis, or nuchal fluid cell obtained from the CH itself to determine fetal karyotype and provide counseling of pregnancy. Late diagnosis (>30 wks) should be delivered in tertiary center prepare to deal with dystocia and postnatal dyspnea of newborn. The airway should be secured before cord clamping in huge lesions.

### **GER and RS**

Although a fundoplication is effective in controlling gastroesophageal reflux (GER), it could be less effective in curing respiratory symptoms (RS) such as aspiration, apnea, choking, chronic cough and wheezing attributed to reflux in children. The problem is documenting a cause-effect relationship between GER and RS. Objective parameters used are the ZMD (mean duration of reflux episodes during sleep by 18-24 hr ph-study), isotopic tracing in lung during milk scintigrams and sx preceded by a drop in ph. A ZMD > 4 min can be a reliable preop sign the antireflux surgery will cure the RS. Reversible obstructive airway disease pts. are more prone to suffer from GER secondary to RS with less chances of being relieved by fundoplication. Neurologically impaired child with RS has highest rate of failure. Medical tx and transgastric jejunostomy feeding are effective for poor operative risk pts.

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