



PEDIATRIC SURGERY UPDATE ©

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Hirschsprung's Associated Enterocolitis (HAEC)

Enterocolitis is the most common complication (incidence 15%), and a major cause of death in children (30%) with Hirschsprung's disease (HD). Can occur before, at the time of, or after definite surgical treatment and is associated to increased morbidity and extended hospitalization. Clinical characteristics are: onset of fluid stools, abdominal distension, and fever in a child with proven HD. HAEC can be classified as: (1) inflammatory- most common type, mucosa affected only, or (2) ischemic- rare, most serious, affects the entire wall with gangrenous bowel. Risk factors identified in the development of HAEC are: (1) delay in diagnosis beyond one week of age; (2) presence of trisomy 21; (3) post-operative anorectal stricture. Histological changes of enterocolitis occur in both the ganglionic and aganglionic bowel. Management consists of: decompressive enterostomy above the level of aganglionosis, fluid resuscitation, NG decompression, and broad spectrum antibiotherapy. Pathogenesis is associated to: (1) proximal colonic dilatation with mucosa ischemia and bacterial invasion; (2) hypersensitivity reaction to bacterial antigens; (3) infection with *C. Difficile* enterotoxin (there is pathological confirmation that some colitis can be of the pseudomembranous type); (4) deficient mucosal transfer and defense of secretory IgA in GI tract; (5) a triggering event in bowel alters the composition of mucin leaving the colonic mucosa susceptible to infection by enterocyte-adherent organism producing toxins which elicit an inflammatory response.

Congenital Diaphragmatic Hernia: Intrauterine Repair

The mortality of CDH is directly related to the degree of lung hypoplasia associated. Prospective studies of prenatally diagnosed fetus prior to 25 wk. gestation has shown that 60% will die despite optimal postnatal care. This unsolved problem has prompted investigators to develop new treatment options such as preoperative stabilization, jet-frequency ventilation, and ECMO. Another area of development is intrauterine fetal surgical repair. To achieve success fetal surgery should: (1) pose no risk to the mother (innocent bystander) or her future reproductive capacity; (2) tocolytic therapy in the post-op weeks should proved effective to avoid prenatal stillbirths; and (3) the

procedure should be superior to conventional therapy. Intrauterine repair has meet with limited success due to herniation of the fetal liver into the chest through the defect. Disturbance of the umbilical circulation during or after liver reduction causes fetal death. Positive-pressure ventilation after birth reduces the liver before the baby comes for surgical repair. Dr. Harrison (USFC Fetal Treatment Center) has devised separate fetal thoraco-abdominal incisions to deal with this problem ("two-step dance"), reducing or amputating the left lateral segment of the liver. Another less invasive approach is enlarging the hypoplastic lungs by reducing the normal egress of fetal lung fluid with controlled tracheal obstruction called PLUGS (Plug Lung Until it Grows).

Did You Know...

Is estimated that medical students are expected to learn 50,000 facts during their first two years at school. They generally prefer to learn concepts.

*Edited by: **Humberto L. Lugo-Vicente, MD, FACS, FAAP**

P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico 00922-0426.

Tel (787)-786-3495 Fax (787)-720-6103

E-mail: titolugo@coqui.net Internet Address: <http://home.coqui.net/titolugo>