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MMIHS

First described by Berdon in 1976, the Megacystis, Microcolon Intestinal Hypoperistalsis Syndrome (MMIHS), represents a rare lethal form of neonatal intestinal obstruction. It is seen in newborn girls showing complete intestinal obstruction (absent meconium output and bilious vomiting), large bladder causing a distended abdomen (dilated urinary and upper gastrointestinal tracts without distal anatomic obstruction), microcolon, absent peristaltic activity, lax abdominal musculature, and malrotation. Positive family history suggests autosomal recessive inheritance. Usual diagnostic studies display: KUB (lacking gas shadow with ground glass appearance), barium enema (malrotated, unused microcolon), cystogram (dilated bladder without distal obstruction), IVP (hydronephrosis and hydronephrosis), UGIS (foreshortened midgut), and suction rectal biopsy (ganglion cell present). Surgical and postmortem specimens describe thin longitudinal muscle coats with vacuolation and degeneration of smooth muscle cells of bowel and bladder, increase connective tissue between them, and abundant mature ganglion cells (referred as hollow visceral myopathy). Initial management is gastrointestinal decompression, and parenteral nutrition (TPN). MMIHS is not a surgical remediable condition. The outcome is generally fatal.

Intestinal Atresias

Intestinal atresias are the product of a late intrauterine mesenteric vascular accident as attested by Louw and Barnard in 1955. They are equally distributed from the ligament of treitz to the ileocecal junction. There is proximal bowel dilatation, with distal (unused) micro-bowel. The diagnosis is suspected with maternal history of polyhydramnios (the higher the atresia), bilious vomiting, abdominal distension and obstipation. KUB shows "thumb-size" dilated bowel loops, and barium enema a microcolon of disuse. Louw classified them into: Type I: an intraluminal diaphragm with seromuscular continuity. Type II: cord-like segment between the bowel blind ends. Type IIIA: atresia with complete separation of blind ends and V-shaped mesenteric defect (see figure), the most commonly found. Type IIIB: jejunal atresia with extensive mesenteric defect and distal ileum acquiring its blood

supply entirely from a single ileocolic artery. The distal bowel coils itself around the vessel, giving the appearance of an “apple peel” deformity. Type IV: multiple atresias of the small intestine. After preoperative stabilization, treatment consists of exploratory laparotomy, resection of proximal dilated intestine, and end to oblique anastomosis in distal jejuno-ileal atresias. Tapering jejunoplasty with anastomosis is preferred in proximal defects.

OK-432

In 1986, Ogita, et al. observed that intracystic injection of OK432 (lyophilized product of *Streptococcus pyogenes*) caused cystic (hygromas) lymphangiomas to become inflamed and led to subsequent cure of the lesion without side effects. The number of patients treated with OK432 now stands at more than 70. Results of a cooperative study for OK432 therapy in Japan were excellent. OK432 caused inflammatory reaction, but did not damage the overlying skin or lead to scar formation. Total or near-total shrinkage of the lesions, without serious complications, was noted in more than 90% of cystic type lymphangiomas (1 to 7 injections; mean, 1.8). On the other hand, this was noted in 50% of cavernous type lymphangiomas (1 to 18 injections; mean, 6.3). Side effects occurring during treatment were: fever, swelling and reddening of the tumor, increased white cell count, and increased CRP level. None of these reactions were serious. Swelling of the lymphangioma was usually insignificant. Depending on location, this could be a risk. No anaphylactic or other reactions to treatment were observed. In patients who were cured, there has been no recurrence throughout the followup period (6 months to 9 years after the last injection of OK432). (Written by: Shuhei Ogita, MD, Kyoto Pref Univ of Med, Japan)

*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>