



## **PEDIATRIC SURGERY** *Update* 8 **Volume 24 No. 03 MARCH 2005**

### **STEP**

Short bowel syndrome is a very serious gastrointestinal disorder characterized by the absence of significant length of bowel capable of normal digestion and absorption. It is estimated that more than 70% of small bowel length must be lost to develop a short bowel syndrome. The three most common causes of short bowel syndrome in the pediatric age are necrotizing enterocolitis, midgut volvulus and gastroschisis. Intestinal adaptation can occur when the neonate is left with more than thirty (30) centimeters of small bowel with an intact ileo-cecal valve. Though the prospect of bowel transplant continues to develop better forms of avoiding acute rejection, still the median survival after transplantation is short (mean of 15 months). Recently, a novel experimental procedure, has attained the attention of surgeons managing this devastating disease complication. The operation is termed serial transverse enteroplasty (STEP) procedure. After short bowel ensues the process of adaptation includes mucosal hyperplasia and bowel dilatation. The STEP procedure is based on the anatomic principle that the blood supply to the bowel comes from the mesenteric border traversing along the perpendicular long axis of the bowel. Multiple stapler lines are placed perpendicularly alternating the direction of the stapler creating a channel of bowel smaller in diameter and longer in length than the original bowel. Advantages of STEP: easy to do, no anastomosis needed, does not result in intestinal obstruction, mesentery is not jeopardized, the length is almost double, the tapering is customizable, and can be performed in sequence after a successful Bianchi procedure. STEP could become the lengthening bowel procedure for short bowel syndrome.

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### **Peritoneal Gliomatosis**

Peritoneal Gliomatosis (PG) is a rare complication of solid mature or immature ovarian teratomas. Occurs with rupture of the tumor capsule causing multiple mature glial implantation (neural tumor tissue) on the parietal, visceral peritoneum, omentum or space of Douglas. GP is found in childhood, adolescence, as well as in young women. Peritoneal gliomatosis can be considered to be implantation metastases. The nodules may vary in size, but are usually around 3 mm in diameter. Peritoneal gliomatosis is a benign condition.

Mean age at time of diagnosis is eleven years. Most cases described are occurs with mature ovarian teratoma. The prognosis depends chiefly on the degree of maturity of the implants. In mature GP, usually no additional chemotherapy is necessary; in immature GP, chemotherapy can induce maturation of the implants. The fate of the glial tissue is still not clear, but the nodule can persist without detectable changes after more than fifteen years of the initial operation, can undergo fibrosis and disappear, or may transform into malignant glial or teratomatous tissue. Close follow-up of these patients is mandatory. The few cases originating from immature teratoma can developed into malignant transformation. Gliomatosis peritonei in extremely rare circumstances has been reported due to transport of glial tissue from the cerebrospinal fluid into the peritoneal cavity via a ventriculo-peritoneal shunt.

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## **Nevus Sebaceous**

Nevus sebaceous of Jadassohn is a congenital hamartomatous skin lesion occurring mostly on the scalp, face and neck. At physical exam the lesion is well circumscribe with a yellow-orange smooth plaque appearance. Borders become irregular with puberty and hormonal changes. Malignant transformation has been reported in 10% of cases occurring only after puberty. The nevus sebaceous can transformed into a basal or squamous cell carcinoma. The lesion will not go away spontaneously. It is uncommon for malignancy to develop in a sebaceous nevus before puberty. Due to the risk of malignant transformation and the difficulty in follow-up of this children with time, early complete excision for prophylaxis is recommended in cases of nevus sebaceous. Excision must encompassed clean deep and lateral margins of resection to be effective. Large lesion will benefit from use of tissue expanders.

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