PEDIATRIC SURGERY *Update* ©
Vol 17 No 06 DECEMBER 2001

Gastrinoma

Gastrinomas, the cause of the Zollinger-Ellison syndrome, are gastrin producing tumors that result in fulminant peptic ulceration and diarrhea. In children, half the cases are associated with malignancy, metastasis and multicentricity. Epidemiologically 75% gastrinomas are sporadic and 25% associated with MEN type 1. Diagnosis of gastrinoma is confirmed after documenting fasting serum hypergastrinemia (> 200 pg/ml), markedly elevated gastric output, a positive secretin-stimulated gastrin analysis or histologic confirmation of a neuroendocrine tumor. Most tumors are found in the head, body and tail of the pancreas, followed by duodenum, liver and lymph nodes. Localization studies include CT-scan, MRCP, octreotide scan (specially indium-labeled pentetreotide), endoscopic US, and Imamura test (selective arterial secretin injection test). Radical resection of the gastrinoma to achieve biochemical cure even when tumor is extrapancreatic and in lymph nodes is the cornerstone of therapy. Unresectable or metastatic cases are managed with pharmacologic therapy using proton pump inhibitors and octreotide. Total gastrectomy is reserved for absolute failure of both medical and surgical management. Gastrinomas in children are slow growing, indolent, and compatible with long life. Prognostic factors correlating with long-term survival are small tumor size, absence of metastatic disease and non-pancreatic location of primary tumors. Factors that do not affect survival include age at diagnosis, sex, presence of lymph node metastases, associated multiple endocrine neoplasia, and method of ulcer treatment.

References:

1- Bonfils S, Landor JH, Mignon M, Hervoir P: Results of surgical management in 92 consecutive patients with Zollinger-Ellison syndrome. Ann Surg 194(6):692-7, 1981
2- Wilson SD: The role of surgery in children with the Zollinger-Ellison syndrome. Surgery 92(4):682-92, 1982
3- Imamura M, Takahashi K, Isobe Y, Hattori Y, Satomura K, Tobe T: Curative resection of multiple gastrinomas aided by selective arterial secretin injection test and intraoperative secretin test. Ann Surg 210(6):710-8, 1989
4- Wilson SD: Zollinger-Ellison syndrome in children: a 25-year follow-up. Surgery 110(4):696-702, 1991
5- Farley DR, van Heerden JA, Grant CS, Miller LJ, Ilstrup DM: The Zollinger-Ellison syndrome. A collective surgical experience. Ann Surg 215(6):561-9, 1992
6- Zimmer T, Stolzel U, Bader M, Koppenhagen K, Hamm B, Buhr H, Riecken EO, Wiedenmann B: Endoscopic ultrasonography and somatostatin receptor scintigraphy in the preoperative localization of insulinomas and gastrinomas. Gut 39(4):562-8, 1996

Epigastric Hernias

Congenital epigastric defects occur anywhere in the linea alba from the navel to the xiphoid process. They represent almost 5% of all hernias defect that presents in children. Most epigastric hernias occur in the midline, are small (15-25 mm), asymptomatic and

reducible. Multiple fascial defects can also be present in 20% of all cases. The defect might arise congenitally from an abnormally wide orifice of a blood vessel during development of the linea alba. The bump is the result of a piece of preperitoneal fat stuck through the fascial defect. Tenderness is an unusual symptom while growth of the defect occurs with time. Most surgeons recommend repair of the defect at the time of presentation. Repair is an outpatient procedure done under general anesthesia with low morbidity and risk of recurrence. Voluminous epigastric hernia (5-10 cm) with a sac that contains epiploic appendages or viscera (ileum loops, stomach) has also been rarely reported in infants.

References:
1- Coats RD, Helikson MA, Burd RS: Presentation and management of epigastric hernias in children. J Pediatr Surg 35(12):1754-6, 2000
2- Askar OM: Aponeurotic Hernias: Recent observations upon paraumbilical and epigastric hernias. Surg Clin North Am 64: 315-333, 1984
3- Corsale I, Palladino E: Diagnosis and treatment of epigastric hernia. Analysis of our experience. Minerva Chir 55(9):607-10, 2000
4- Ameh EA: Giant congenital epigastric hernia. West Afr J Med 18(2):151-2, 1999

Necrotizing Fasciitis

Necrotizing fasciitis (NF) is a rare, severe, rapidly progressive soft tissue infection involving the skin, subcutaneous tissue and superficial fascia. Most cases occur during the newborn period. In newborns NF is most frequently associated with omphalitis, necrotizing enterocolitis, balanitis, urachal anomalies and fetal monitoring. In older children NF can be seen as a postoperative complication (appendectomy, gastrostomy), in neutropenia after chemotherapy and secondarily to varicella infestation. Most frequent sites of initial involvement are the abdominal wall, followed by thorax, back, scalp, and extremity. Infection is usually polymicrobial containing both gram positive (beta-hemolytic Streptococcus and Staphylococcus aureus) and gram negative bacteria (E. Coli and Pseudomonas). Important clinical findings in infants with NF are tachycardia, abnormal white blood cell count, systemic toxicity, severe edema, and, in older children pain out of proportion to the apparent degree of infection. The child can also develop wound crepitation and radiographic evidence of subcutaneous gas. An ominous sign indicative of the need for immediate radical debridement is the appearance of a patch of dusky or gangrenous skin. Initial management consists of antibiotics, intravenously afluids, blood transfusions, calcium replacement (due to saponification) and general patient support. Neutropenic patients benefit from granulocyte transfusions. Surgical procedures include extensive debridement as soon as possible and as needed for continued necrosis, secondary closure and skin grafting. Mortality ranges from 10 to 60%. Improved survival requires early diagnosis followed by prompt aggressive surgical debridement.

References:
1- Kosloske AM, Cushing AH, Borden TA, Woodside JR, Klein MD, Kulasinghe HP, Bailey WC: Cellulitis and necrotizing fasciitis of the abdominal wall in pediatric patients. J Pediatr Surg 16(3):246-51, 1981
2- Farrell LD, Karl SR, Davis PK, Bellinger MF, Ballantine TV: Postoperative necrotizing fasciitis in children.

Pediatrics 82(6):874-9, 1988
3- Samuel M, Freeman NV, Vaishnav A, Sajwany MJ, Nayar MP: Necrotizing fasciitis: a serious complication of omphalitis in neonates. J Pediatr Surg 29(11):1414-6, 1994
4- Murphy JJ, Granger R, Blair GK, Miller GG, Fraser GC, Magee JF: Necrotizing fasciitis in childhood. J Pediatr Surg 30(8):1131-4, 1995
5- Moss RL, Musemeche CA, Kosloske AM: Necrotizing fasciitis in children: prompt recognition and aggressive therapy improve survival. J Pediatr Surg 31(8):1142-6, 1996
6- Hsieh WS, Yang PH, Chao HC, Lai JY: Neonatal necrotizing fasciitis: a report of three cases and review of the literature. Pediatrics 103(4):e53, 1999
7- Abbott RE, Marcus JR, Few JW, Farkas AM, Jona J: Necrotizing fasciitis in infancy: an uncommon setting and a prognostic disadvantage. J Pediatr Surg 34(9):1432-4, 1999

---

* Edited by: **Humberto L. Lugo-Vicente, MD, FACS, FAAP**
Professor/Associate Director of Pediatric Surgery, University of Puerto Rico School of Medicine and University Pediatric Hospital, Rio Piedras, Puerto Rico.
Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426.
Tel (787)-786-3495 Fax (787)-720-6103 E-mail: *titolugo@coqui.net*
Internet: http://home.coqui.net/titolugo

© *PSU 1993-2001*
ISSN 1089-7739