



PEDIATRIC SURGERY Update ©

Vol 15 No 02 AUGUST 2000

'Official Publication of the Puerto Rico Association of Pediatric Surgeons'

Carcinoid Syndrome

The carcinoid syndrome (facial flushing, diarrhea, tricuspid regurgitation, pulmonic stenosis, valvular fibrosis and wheezing) is the result of serotonin overproduction by a carcinoid tumor. Carcinoid tumors arise from enterochromaffin cells (APUD cells from the neural crests), occur in virtually every organ, could be multiple, metastatic and associated with a second malignancy. Patients are diagnosed biochemically from increased urinary excretion of 5-hydroxyindoleacetic acid (5-HIAA). Platelet serotonin levels are more sensitive for detecting carcinoids that secrete small amounts of serotonin. Jejunum-ileum, bronchus and appendix are the most common sites of origin. Carcinoid of the appendix is the most common neoplasm of the GI tract in childhood. Metastasis to liver of midgut carcinoids produce the syndrome. Tumors greater than 2 cm are more prone to metastasis needing aggressive surgical management. Octreotide scan and I-131 MIBG are useful in determination of location and extent of some carcinoid tumors, particularly those of midgut origin. A positive scan may predict the ability of Octreotide therapy to control symptoms of hormonal hypersecretion. Scans provide localization of the primary tumor that should be widely excised including lymph nodes. Higher survival rates are found for patients with midgut lesions who undergo intra abdominal debulking procedures excluding the liver. For single liver lesion resection is justified, otherwise with multiple diffuse disease hepatic artery ligation or embolization has been tried. Symptomatic metastasis should be managed with Octreotide. Prognosis is associated with the presence of liver metastasis, syndrome development and level of tumor markers (chromogranin A).

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Left Hypoplastic Colon Syndrome

Colonic obstruction in the newborn child could be the result of necrotizing enterocolitis, atresia, meconium plug syndrome, duplication cyst, Hirschsprung disease or the small left colon syndrome. The left (small) hypoplastic colon syndrome (LHCS) is a very rare cause of colonic obstruction identified in newborns with characteristic roentgenographic features resembling those of Hirschsprung's disease. Manifesting in the first 24-48 hours of life, LHCS is a functional disturbance related to immaturity of the intrinsic innervation of the colon that is especially common in low birth weight neonates or of diabetic mothers. Intestinal perforation, sepsis, hypoglycemia and death may occur. The diagnosis is suggested in a barium enema when the caliber of the left colon is small with a transitional zone at the splenic flexure. Management consists of hypoglycemia correction, antibiotics, nasogastric decompression and observation. In most babies the obstruction clears in 48-72 hours. When the clinical diagnosis is not readily apparent a rectal biopsy and sweat chloride test should be done to differentiate LHCS from Hirschsprung disease and cystic fibrosis respectively. The narrowed left colon remains narrow in follow-up.

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Beckwith-Wiedemann Syndrome

The Beckwith-Wiedemann Syndrome (BWS), first described in 1964, is characterized by the presence of macrosomia (gigantism), macroglossia, omphalocele and unusual linear fissures in the lobules of the external ear. One of the more frequent metabolic changes is transient neonatal hypoglycemia, the result of pancreas cell hyperplasia. Inheritance of the syndrome remains uncertain. Most cases are sporadic, but a number of familial cases have been reported. BWS is associated with a predisposition to embryonal tumors, most commonly Wilms' Tumor. Genetic abnormalities found in these tumors affect the same chromosome region (11p15), which has been implicated in the etiology of BWS. Routine abdominal ultrasound screening every six months up to the age of eight years is recommended for children with BWS.

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