



PEDIATRIC SURGERY UPDATE ©

Vol 06 No 05 MAY 1996

Neuroblastoma

Neuroblastoma (NB) is the most common solid tumor in infants. Originates from the neural crest: sympathetic ganglion chain and adrenal medulla. 75% arise in the retroperitoneum (adrenal gland and paraspinal ganglia), 20% in the posterior mediastinum, and 5% in the neck or pelvis. NB is a solid, highly vascular tumor with a friable pseudocapsule. Most children present with an abdominal mass, and one-fourth have hypertension. Other have: Horner's syndrome, Panda's eyes, anemia, dancing eyes or vaso-intestinal syndrome (VIP). Diagnosis is confirmed with the use of simple X-rays (stipple calcifications), ultrasound, and CT-Scan. Work-up should consider: bone marrow, bone scan, myelogram (if there is evidence of intraspinal extension), and plasma/urine tumor markers level: VMA, HVA, VGA, DOPA. Management depends on stage of disease at diagnosis. Localized tumors receive surgical therapy. Partially resected or unresectable cases need chemotherapy a/o radiotherapy after establishing a histologic diagnosis. Independent variables determining prognosis are age at diagnosis and stage of disease. Young children with stage I/II have a better outcome. Poor outcome for greater stages, older patients, and those with bone cortex metastasis. Other prognostic variables are: site of primary, maturity of tumor, presence of positive lymph nodes, high levels of ferritin, neuron-specific enolase, and diploid DNA.

Duplications

Duplications of the gastrointestinal tract are considered uncommon congenital anomalies usually diagnosed or unexpectedly encountered intraoperatively during the first two years of life. The duplicated bowel can occur anywhere in the GI tract, is attached to the mesenteric border of the native bowel, shares a common wall and blood supply, coated with smooth muscle, and the epithelial lining is GI mucosa. May contain ectopic gastric or pancreatic tissue. Most are saccular, other tubular. Theories on their origin (split notochord syndrome, twining, faulty solid-stage recanalization) do not explain all cases of duplicated bowel. Three-fourth are found in the abdomen (most commonly the ileum and jejunum), 20% in the thorax, the rest thoraco-abdominal or

cervical. Symptoms vary according to the size and location of the duplication. Clinical manifestations can range from intestinal obstruction, abdominal pain, GI bleeding, ulceration, or mediastinal compression. Ultrasound confirms the cystic nature of the lesion (muscular rim sign) and CT the relationship to surrounding structures. Management consist of surgical excision avoiding massive loss of normal bowel and removing all bowel suspect of harboring ectopic gastric mucosa.

Ankyloglossia

Inferior ankyloglossia, a condition more commonly known as tongue-tie, is common among infants. It is caused by a thin velum that fixes the tongue to the floor of the mouth. Initially seen in the neonate, it can disappear spontaneously or with sucking if the frenum is sufficiently thin. Other times it will persist causing sucking or swallowing problems, speech disorders or mechanical restriction of tongue movements. Frenulectomy is curative. Indications for frenulectomy are: articulation speech difficulties, mechanical limitations (inability to leak lips, perform intraoral toilet, or play a wind instrument), and problems with either feeding or suction. Should never be done as an office procedure, but at OR under deep mask anesthesia, rapid tongue retraction and electrocoagulation; no sutures are required.

Submit!

We are inviting all of you to submit contribution of your own to the newsletter. Credit will be given to the authors keeping his or her name, institution and E-mail address on their work. A list of contributors will appear in the internet homepage. All works should be send to Dr H. Lugo-Vicente to the e-mail or address below.

*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>