PEDIATRIC SURGERY *Update* ©
Vol 13 No 01 JULY 1999

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

Tongue Cysts

Cysts in the tongue are rare in children. The differential diagnosis includes: mucoceles (retention cyst), cystic hygroma, thyroglossal cysts, cysts of foregut origin and teratoma. They usually presents at birth interfering with mouth closure, swallowing, normal feeding or even causing respiratory problems. Complications of the cyst include infection, hemorraghe and rupture leading to increase in size and asphyxia, inability to feed and aspiration pneumonia. Mucoceles (ranula) may appear in the base of the tongue and needs excision or marsupialization. Multiple cysts can be a cystic hygroma in the tongue that may need partial glossectomy or management with sclerosing substances. Lingual thyroglossals are found in the base of the tongue and can safely be managed with marsupialization without excision or Sistrunk procedure. Cysts of foregut origin presents in the neonatal period, can be found in the anterior part of the dorsum of the tongue needing complete surgical excision through a sagittal glossal split. Cyst aspiration is inadequate definitive treatment. A teratoma likewise needs surgical excision for cure.

References

1- Urao M, Teitelbaum DH, Miyano T: Lingual thyroglossal duct cyst: a unique surgical approach. J Pediatr Surg 31(11):1574-6, 1996

2- Surana R, Losty P, Fitzgerald RJ: Heterotopic gastric cyst of the tongue in a newborn. Eur J Pediatr Surg 3(2):110-1, 1993

3- Wiersma R, Hadley GP, Bosenberg AT, Chrystal V: Intralingual cysts of foregut origin. J Pediatr Surg 27(11):1404-6, 1992

4- Velcek FT, Klotz DH, Hill CH, Ladogana LE: Tongue lesions in children. J Pediatr Surg 14(3):238-46, 1979

Primary Immunodeficiency

The body mounts an immune response by producing antibodies (B cells), inciting cellular division (T cells), cascading complement and producing phagocytosis. Congenital absence or deficiency of this response not associated to a secondary illness is known as primary immunodeficiency (PI). PI is very rare (400 new cases/year in the USA), and is divided into deficiency of: antibody (most common), T-cell, combined T and B-cell, phagocytic or complement activity. Most cases of PI are identified in the first year of life. Primary symptom is recurrent infections with unusual organisms (low pathogenicity) that is severe, prolonged and resistant to conventional therapy. T-cell deficiency occurs before the age of six months while antibody deficiency presents clinically after this age due to transplacental protection. Phagocytic and complement deficiency is present since birth as the baby might show a lack of pus formation or delayed umbilical cord detachment. After respiratory tract, the skin is the 2nd most common site of infection, i.e., thrush, furuncles, abscess, fistulas and cellulitis followed by the GI tract. The child usually appears chronically ill with growth failure. Extensive lab testing will establish the diagnosis. Live

vaccine should be avoided. The child with PI who will undergo a surgical procedure needs: protective isolation, antibiotic prophylaxis, nutritional backup, physiologic monitoring and selective replacement of defective substance (immunoglobulins) a/o immunologic enhancing agents use. Bone marrow transplant can sometimes correct a PI.

References

1- Peerles AG, Stiehm ER: The Immunocompromised Child, In Fonkalsrud and Krummel "Infections and Immunologic Disorders in Pediatric Surgery", WB Saunders Co, 1993, pag 77-90
2- Stiehm ER: Human intravenous immunoglobulin in primary and secondary antibody deficiencies. Pediatr Infect Dis J 16(7):696-707, 1997
3- Pachman LM, Lynch PA, Silver RK, Ozog DL, Poznanski AK: Primary immunodeficiency disease in children: an update. Curr Probl Pediatr 19(1):1-64, 1989
4- Primary immunodeficiency diseases. Report prepared for the WHO by a scientific group on immunodeficiency. Clin Immunol Immunopathol 28(3):450-75, 1983
5- Pappas BE: Primary immunodeficiency disorders in infancy. Neonatal Netw 18(1):13-22, 1999

Lymphedema

The most common forms of lymphedema in children affect the lower extremity. Lymphedema in children can be classified into: congenital, precox and tarda. The underlying pathology is absence (10%) or hypoplasia (90%) of the lymphatic channels of the lower extremity. Diagnosis of lymphedema is made clinically. Congenital lymphedema appears early in life, involves more than one extremity, rarely extends above the knee and enlarges at a slower rate than body growth. The swelling becomes less pronounced with age, and no specific therapy is required in two thirds of cases. Lymphedema precox appears during adolescence, occur mostly in females, extends to the groin and is associated with yellow nails (inadequate lymphatic drainage). Lymphedema tarda occurs spontaneously in middle age people. In unilateral cases a retroperitoneal tumor or venous obstruction should be rule out. Complications of lymphedema include swelling, brawny edema, cellulitis and lymphangitis. Management consists of extremity care, compressive support stocking and leg elevation. In severe cases the option is excision of excess skin and subcutaneous lymphatics (modified Kondoleon procedure).

References

1- Fonkalsrud EW: Surgical management of congenital lymphedema in infants and children. Arch Surg 114(10):1133-6, 1979
2- Hilliard RI, McKendry JBJ, Phillips MJ: Congenital abnormalities of the lymphatic system: a new clinical classification. Pediatrics 86:988, 1990
3- Levine C: Primary disorders of the lymphatic vessels: a unified concept. J Pediatr Surg 24:233, 1989
4- Fonkalsrud EW: Congenital Malformations of the Lymphatic System. Semm Pediatr Surg 3(2): 62-69, 1994

---

* Edited by: **Humberto L. Lugo-Vicente, MD, FACS, FAAP**
Associate Professor/Administrative 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 00922-0426.
Tel (787)-786-3495 Fax (787)-720-6103 E-mail: *titolugo@coqui.net*
Internet: http://home.coqui.net/titolugo