

PEDIATRIC SURGERY Update © Vol. 28 No. 02 FEBRUARY 2007

Gonadoblastoma

Gonadoblastoma is a sex cord gonadal tumor that contains both germ cell and sex cord stromal elements. It occurs almost exclusively in sexually abnormally individuals with gonadal dysgenesis and Y-containing cells, while other cases occur in children with mixed gonadal dysgenesis (mosaic 45XO/46XY). The combination of the Y chromosome with a dysgenetic gonad is all that is needed for a gonadoblastoma or dysgerminoma to develop. The tumor is usually quite small and calcifications are common. Almost 40% of all gonadoblastomas are bilateral. The germ cell component may outgrow the stromal elements and result in the formation of a dysgerminoma. Most cases will appear in young female adults with history of primary amenorrhea during teenage years and virilization. Management of gonadoblastoma consists of removal of both dysgenetic gonads irrespective of the bilaterality of the lesion. Because these tumors occur in up to 50% of patients with gonadal dysgenesis early bilateral prophylactic gonadectomy should be performed. Gonadoblastomas can exhibit either benign or malignant features, though most cases are benign tumors that have a good prognosis after excision. Gonadectomy can either be done open or laparoscopically. With the presence of malignant germ cell elements, chemotherapy will be needed. Other children at risk to develop gonadoblastoma later in life include those with Turners and androgen insensitivity syndrome.

References:

1- Olsen MM, Caldamone AA, Jackson CL, Zinn A: Gonadoblastoma in infancy: indications for early gonadectomy in 46XY gonadal dysgenesis. J Pediatr Surg. 23(3):270-1, 1988

2- Gibbons B, Tan SY, Yu CC, Cheah E, Tan HL: Risk of gonadoblastoma in female patients with Y chromosome abnormalities and dysgenetic gonads. J Paediatr Child Health. 35(2):210-3, 1999

3- Gravholt CH, Fedder J, Naeraa RW, Muller J: Occurrence of gonadoblastoma in females with Turner syndrome and Y chromosome material: a population study. J Clin Endocrinol Metab. 85(9):3199-202, 2000 4- Uno T, Kazui T, Muhammad BA: Laparoscopic surgery for gonadal dysgenesis in children. Surg Laparosc Endosc Percutan Tech. 9(2):151-5, 1999

5- Mazzanti L, Cicognani A, Baldazzi L, Bergamaschi R, Scarano E, Strocchi S, Nicoletti A, Mencarelli F, Pittalis M, Forabosco A, Cacciari E: Gonadoblastoma in Turner syndrome and Y-chromosome-derived material. Am J Med Genet A. 135(2):150-4, 2005

6- Templeman CL, Fallat ME: Bening Ovarian Masses. Semm Pediatr Surg. 14(2): 93-99, 2005

7- Bianco B, Lipay MV, Melaragno MI, Guedes AD, Verreschi IT: Detection of hidden Y mosaicism in Turner's syndrome: importance in the prevention of gonadoblastoma. J Pediatr Endocrinol Metab. 19(9):1113-7, 2006

Encopresis

Encopresis refers to the involuntary loss of formed, semiformed, or liquid stools into the child's underwear in the presence of constipation Solid fecal material accumulated in the distal rectum unable to be discharged appropriately produces seepage of more proximally

fecal fluid which escapes unconsciously into the cloths of the child. It is a very difficult social and physical problem to manage satisfactorily in the child. Encopresis is a complex abnormal motility disorder, requiring a multidisciplinary approach. The most common causes associated with encopresis consist of slow transit functional constipation, Hirschsprung is disease and anorectal malformations. Severely constipated children with encopresis in whom outpatient management has failed frequently require several days of hospitalization, as well as conventional treatments involving cathartics and enemas. A balanced electrolyte solution of the nonabsorbable polymer polyethylene glycol (GoLytely) offers a safe and efficient method for clearing the intestine in such cases. Children with encopresis have normal functioning internal sphincter and can acquire normal bowel control using biofeedback therapy to correct the abnormal defecation dynamics. A continent appendicostomy (Malone procedure) is a promising treatment that completely cleanses the colon, increases the child's autonomy, and decreases the chance of soiling in intractable cases of encopresis with pseudo-incontinence.

References:

1- Ingebo KB, Heyman MB: Polyethylene glycol-electrolyte solution for intestinal clearance in children with refractory encopresis. A safe and effective therapeutic program. Am J Dis Child. 142(3):340-2, 1988

2- Bulut M, Tekant G: Encopretic children: experience with fifty cases. Turk J Pediatr. 33(3):167-72, 1991
3- Loening-Baucke V: Encopresis and soiling. Pediatr Clin North Am. 43(1):279-98, 1996

4- Iwai N, Iwata G, Kimura O, Yanagihara J: Is a new biofeedback therapy effective for fecal incontinence in patients who have anorectal malformations? J Pediatr Surg. 32(11):1626-9, 1997

5- Hutson JM, McNamara J, Gibb S, Shin YM: Slow transit constipation in children. J Paediatr Child Health. 37(5):426-30, 2001

6- Di Lorenzo C, Benninga MA: Pathophysiology of pediatric fecal incontinence. Gastroenterology. 126(1 Suppl 1):S33-40, 2004

Torsion Fallopian Tubes

Torsion of a fallopian tube is a very rare event presenting in premenarchal girls and postmenarchal teenagers. Factors associated with torsion include abnormally long tube and mesosalpinx, adnexal venous congestion and abnormal peristalsis. Other times extrinsic factors such as pelvic masses or trauma are the principal cause of torsion. Torsion is more common in the right fallopian tube. Diagnosis is difficult due to nonspecific symptoms. Primordial symptoms consist of sudden abdominal pain, nausea, and vomiting. Other times the child will develop abdominal tenderness with peritonitis. Pelvic ultrasound with color Doppler can identify an elongated cystic mass with variable septs and scattered internal echoes. CT-Scan can demonstrate thickened of the affected tube with hemorrhage. The gold standard of diagnosis and management consists of operative laparoscopy. With isolated tubal torsion the tube can be untwisted. If torsion occurs due to a pelvic mass, the mass should be fixed to the peritoneum or cul-de-sac. Unless a high index of suspicion is maintained for torsion of the fallopian tube in adolescent females, this disorder may not be detected until after tubal destruction.

References:

1- Evans JP: Torsion of the normal uterine adnexa in premenarchal girls. J Pediatr Surg. 13(2):195-6, 1978

2- Hockberger RF, Sternbach G: Torsion of the fallopian tube. JACEP. 7(8):315-7, 1978

3- Ghossain MA, Buy JN, Bazot M, Haddad S, Guinet C, Malbec L, Hugol D, Truc JB, Poitout P, Vadrot D: CT in adnexal torsion with emphasis on tubal findings: correlation with US. J Comput Assist Tomogr. 18(4):619-25, 1994

4- Rizk DE, Lakshminarasimha B, Joshi S: Torsion of the fallopian tube in an adolescent female: a case report. J Pediatr Adolesc Gynecol. 15(3):159-61, 2002

5- Perlman S, Hertweck P, Fallat ME: Paratubal and tubal abnormalities. Semm Pediatr Surg 14(2): 124-134, 2005

6- Pinkert M, Klein Z, Tepper R, Beyth Y: Hydrosalpinx with adnexal torsion in an adolescent virgin patient--A diagnostic dilemma: case report and review of the literature. J Pediatr Adolesc Gynecol. 19(4):297-9, 2006

* Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP

Professor /Academic Director of Pediatric Surgery, University of Puerto Rico - School of Medicine, 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* 1993-2007 ISSN 1089-7739