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Meconium Plug Syndrome

One of the most significant sign of large bowel obstruction during the neonatal period is failure to pass meconium during the first day of life. The differential diagnosis to consider includes Hirschsprung's disease, anorectal malformations, meconium plug syndrome, small left colon syndrome, hypoganglionosis, neuronal intestinal dysplasia and megacystis-microcolon-intestinal hypoperistalsis syndrome. Meconium plug syndrome (MPS) was first described by Clatworthy in 1956 as a transient form of distal colonic or rectal obstruction in newborns caused by an inspissated, immobile meconium. The plug is white and chalky and rarely involves the small bowel. Clinical manifestations include progressive abdominal distension, vomiting (sometimes is bilious) and failure to pass meconium during the initial two days of life. Though most cases are idiopathic, MPS has been associated with prematurity, hypotonia, hypermagnesemia (reduces acetylcholine release with subsequent myoneural depression), diabetic mother, Hirschsprung's disease and cystic fibrosis. Colonic contrast study suggests the diagnosis (filling defect) and can be therapeutic in most cases to relieve the obstruction. Gastrografin instillation is highly effective in moving the obstructing long, thick plug, even in tiny premature infants with MPS. Suction rectal biopsy to exclude the diagnosis of Hirschsprung's disease along with cystic fibrosis screening is warranted in all cases of MPS. The diagnosis of MPS is made after all the above causes are excluded. Need for surgery is extremely rare.

References:

- 1- Loening-Baucke V, Kimura K: Failure to pass meconium: diagnosing neonatal intestinal obstruction. Am Fam Physician 60(7):2043-50, 1999
- 2- Clatworthy HW, Howard WH, Lloyd J: The meconium plug syndrome. Surgery 39:131-142, 1956
- 3- Hen J, Dolan TF, Touloukian RJ: Meconium Plug Syndrome associated with Cystic Fibrosis and Hirschsprung's Disease. Pediatrics 66 (3): 466-468, 1980
- 4- Krasna IH, Rosenfeld D, Salerno P: Is it Necrotizing Enterocolitis, Microcolon of Prematurity, or Delayed Meconium Plug? A Dilemma in the Tiny Premature Infant. J Pediatr Surg 31(6): 855-858, 1996
- 5- Olsen MM, Luck SR, Lloyd-Still J, Raffensperger JG: The spectrum of meconium disease in infancy. J Pediatr Surg 17(5):479-81, 1982
- 6- Rosenstein BJ: Cystic fibrosis presenting with the meconium plug syndrome. Am J Dis Child 132(2):167-9, 1978

Granuloma Annulare

Granuloma annulare is a rare, benign, self-limiting, subcutaneous nodule that can appear in the scalp and/or extremity of infants and children. The nodule is painless, non-mobile, characterized by rapid growth usually identified in the occipital region with absence of

bone involvement. Mean age at presentation is four years. Lesions are most commonly located about the elbow, knee, and scalp. The erythrocyte sedimentation rate could be elevated attesting to the inflammatory nature of the lesion. Otherwise, no ancillary test is specific for this disorder. Excisional biopsy can establish the diagnosis by showing lesions that resembles rheumatoid nodules, consisting of acellular central areas surrounded by palisading histiocytes. Between 20 and 50% of children have local recurrence or distant development of new lesions. Most patients will not progress to any recognized systemic illness or connective tissue disorder. The clinical course is characterized by spontaneous regression. Parental reassurance is warranted.

References:

- 1- Challa VR, Weidner N, Bell WO, Prichard RW: Granuloma annulare: a rare occipital lesion in infants and children. Surg Neurol 28(3):211-4, 1987
- 2- Davids JR, Kolman BH, Billman GF, Krous HF: Subcutaneous granuloma annulare: recognition and treatment. J Pediatr Orthop 13(5):582-6, 1993
- 3- Felner EI, Steinberg JB, Weinberg AG: Subcutaneous granuloma annulare: a review of 47 cases. Pediatrics 100(6):965-7, 1997
- 4- Trobs RB, Borte M, Voppmann A, Weidenbach H, Thiele J: Granuloma annulare, nodular type--a subcutaneous pseudorheumatoid lesion in children. Eur J Pediatr Surg 7(6):349-52, 1997
- 5- McDermott MB, Lind AC, Marley EF, Dehner LP: Deep granuloma annulare (pseudorheumatoid nodule) in children: clinicopathologic study of 35 cases. Pediatr Dev Pathol 1(4):300-8, 1998
- 6- Grogg KL, Nascimento AG: Subcutaneous granuloma annulare in childhood: clinicopathologic features in 34 cases. Pediatrics 107(3):E42, 2001

Cervical Teratoma

Germ cell tumor (Teratoma) arising in the neck region of an infant is an unusual lesion encompassing between three and 5% of all teratomas found in children. These lesions are histologically benign, usually large and can cause airway obstruction during birth. Cervical teratomas are true neoplasm composed of foreign tissue to the anatomic site of origin with all three germ layers represented. Neural tissue predominates. Fine calcifications can be seen on simple films. Newborns might require intubation within the first few hours after birth due to respiratory distress. The tumor is firm, frequently mobile, multilobular, cystic and well encapsulated. Occasionally the tumor is incorrectly diagnosed as cystic hygroma. Prenatally this tumor can be associated with polyhydramnios, pulmonary insufficiency and fetal demise. Prenatal diagnosis by ultrasound gives the clinician the opportunity of maintaining the materno-fetal circulation until the airway is properly secured during birth (EXIT procedure). Cesarean section is recommended for all tumors larger than five cm in size. After birth and stabilization management consists of prompt surgical excision. Total excision is essential to avoid local recurrence and malignant degeneration. Postoperative monitoring for recurrences should include Alpha-fetoprotein levels in difficult cases.

References:

- 1- Gundry SR, Wesley JR, Klein MD, Barr M, Coran AG: Cervical teratomas in the newborn. J Pediatr Surg 18(4):382-6, 1983
- 2- Jordan RB, Gauderer MW: Cervical teratomas: an analysis. Literature review and proposed classification.

- J Pediatr Surg 23(6):583-91, 1988
- 3- Langer JC, Tabb T, Thompson P, Paes BA, Caco CC: Management of prenatally diagnosed tracheal obstruction: access to the airway in utero prior to delivery. Fetal Diagn Ther 7(1):12-6, 1992
- 4- Azizkhan RG, Haase GM, Applebaum H, Dillon PW, Coran AG, King PA, King DR, Hodge DS: Diagnosis, management, and outcome of cervicofacial teratomas in neonates: a Childrens Cancer Group study. J Pediatr Surg 30(2):312-6, 1995
- 5- Larsen ME, Larsen JW, Hamersley SL, McBride TP, Bahadori RS: Successful management of fetal cervical teratoma using the EXIT procedure. J Matern Fetal Med 8(6):295-7, 1999
- 6- Elmasalme F, Giacomantonio M, Clarke KD, Othman E, Matbouli S: Congenital cervical teratoma in neonates. Case report and review. Eur J Pediatr Surg 10(4):252-7, 2000

* 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

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