PEDIATRIC SURGERY *Update* ©
Vol. 41 No. 01 JULY 2013

Acquired Jeune's Syndrome

Jeune's syndrome, also known as asphyxiating thoracic dysplasia, is a type of thoracic dystrophy with severe narrow thorax leading to respiratory distress and even death in its more severe form. Jeune's syndrome either is congenital (autosomal recessive) or acquired. The acquired form of Jeune's syndrome was described in 1996 in children who had undergone repair of pectus excavatum chest wall deformity utilizing the traditional open (Ravitch) approach with subperichondrial resection of deformed cartilages and transverse osteotomy performed too early an age (less than four years of age). Permanent impairment of normal chest wall growth and subsequent restriction of lung expansion during respiration creates this type of acquired form of the disease. Years later after the primary procedure for pectus these children developed progressive dyspnea with mild exertion associated to restrictive pulmonary function tests with forced vital capacity (FVC) of 30-50% and forced expiratory volume in one second (FEV1) of 30-60% of predicted values. This acquired restrictive thoracic dystrophy is due to an aggressive resection of the involved deformed cartilages including the second costal cartilage. This complication does not occur using the actual minimally invasive repair of pectus excavatum (Nuss technique). Diagnosis is done using pulmonary function tests and three-dimensional CT reconstruction of the chest. Management of acquired Jeune's syndrome includes displacing the sternum forward with a splint or median sternotomy with interposition of autologous rib grafts to increase the chest wall diameter (Weber technique). Substantial improvement in PFT and clinical symptoms can be achieved with the sternal split technique though long-term evaluation is awaiting results.

References:

1- de Vries J, Yntema JL, van Die CE, Crama N, Cornelissen EA, Hamel BC: Jeune syndrome: description of 13 cases and a proposal for follow-up protocol. Eur J Pediatr. 169(1):77-88, 2010
2- Weber TR, Kurkchubasche AG: Operative management of asphyxiating thoracic dystrophy after pectus repair. J Pediatr Surg. 33(2):262-5, 1998
3- Fokin AA, Robicsek F: Acquired deformities of the anterior chest wall. Thorac Cardiovasc Surg. 54(1):57-61, 2006
4- Weber TR: Further experience with the operative management of asphyxiating thoracic dystrophy after pectus repair. J Pediatr Surg. 40(1):170-3, 2005
5- Phillips JD, van Aalst JA: Jeune's syndrome (asphyxiating thoracic dystrophy): congenital and acquired. Semin Pediatr Surg. 17(3):167-72, 2008
6- Lopushinsky SR, Fecteau AH: Pectus deformities: a review of open surgery in the modern era. Semin Pediatr Surg. 17(3):201-8, 2008

Metal Allergy

Jewelry, dental and surgical implants from craniofacial, orthopedic, neurosurgical and pediatric surgery physicians can lead to metal allergy in children. As many as 13% of patients are sensitive to nickel, cobalt or chromium. Metal allergy from nickel is the most common contact allergy in the United States and Europe. The classical symptom of dermatitis caused by nickel is a rash in the earlobes, periumbilical region or wrist resulting from contact with costume jewelry, buttons and zipper. Metal allergy is a typical delayed type IV hypersensitivity reaction caused by T-lymphocytes reaction. CD8 and CD4 cells cause cytotoxic and inflammatory response to the metal. Children with metal allergy usually elicit a past history of atopy including allergic rhinitis, asthma, eczema and urticarial rash. Metal allergies are frequently misdiagnosed as surgical infections. Symptoms of inflammation such as pain, warmth, erythema and swelling can be seen over the implant, including pericarditis and pleural effusion in those in a thoracic position. As a screening measure to determine if a child can or might develop metal allergy to an implant the following should be evaluated: 1- history of allergy to jewelry, orthodontic braces, metal buttons on clothing and food. 2- History of previous atopy and eczema. If any of the above indications are found, a dermal patch test should be performed. This patch test contains 23 allergens and allergen mixes that cause up to 80% of allergic contact dermatitis cases. Should the child be found to have metal allergy implants of titanium should be considered, since titanium does not produce allergic reactions but are more expensive.

References:

1- Mortz CG, Lauritsen JM, Bindslev-Jensen C, Andersen KE: Nickel sensitization in adolescents and association with ear piercing, use of dental braces and hand eczema. The Odense Adolescence Cohort Study on Atopic Diseases and Dermatitis (TOACS). Acta Derm Venereol. 82(5):359-64, 2002

2- Dotterud LK, Falk ES: Metal allergy in north Norwegian schoolchildren and its relationship with ear piercing and atopy. Contact Dermatitis. 31(5):308-13, 1994

3- Kalimo K, Mattila L, Kautiainen H: Nickel allergy and orthodontic treatment. J Eur Acad Dermatol Venereol. 18(5):543-5, 2004

4- Katting B, Brehler R, Traupe H: Allergic contact dermatitis in children: strategies of prevention and risk management. Eur J Dermatol. 14(2):80-5, 2004

5- Rushing GD, Goretsky MJ, Gustin T, Morales M, Kelly RE Jr, Nuss D: When it is not an infection: metal allergy after the Nuss procedure for repair of pectus excavatum. J Pediatr Surg. 42(1):93-7, 2007

6- Thyssen JP, Jakobsen SS, Engkilde K, Johansen JD, SA¸balle K, Menna T: The association between metal allergy, total hip arthroplasty, and revision. Acta Orthop. 80(6):646-52, 2009

PRETEXT

The PRETEXT (PRE Treatment EXTent of disease) system was designed by the International Childhood Liver Tumor Strategy Group (SIOPEL) for staging and risk stratification of liver tumors, namely hepatoblastoma, hepatocellular carcinoma and epithelioid hemangioendothelioma. PRETEXT describes tumor extent before any therapy allowing different groups to have a more effective comparison in future studies. PRETEXT staging is based on Couinaud's liver segmentation grouping the liver into four

sections: segment 2 and 3 (left lateral section), segment 4a and 4b (left medial section), segments 5 and 8 (right anterior section) and segments 6 and 7 (right posterior section). The PRETEXT number is derived by subtracting the highest number of contiguous liver sections that are not involved by tumor from four. PRETEXT also utilizes other criteria such as involvement of the caudate lobe (designated C), involvement of the inferior vena cava or hepatic veins (V), involvement of the portal veins (P), extrahepatic abdominal disease (E) and distant metastasis (M). Other high risk criteria include tumor rupture or intraperitoneal hemorrhage at diagnosis (H1) and alpha fetoprotein levels below 100 ug/L. In PRETEXT I one section is involved and three are free. This group includes only a small portion of all cases. In PRETEXT II one or two sections re involved, but two adjoining sections are free. They are limited to the right lobe or left lobe of the liver. In PRETEXT III two or three sections are involved and no two adjoining sections are free. The unifocal tumors in this category spare only the left lateral or right posterior section. This group is relatively common. In PRETEXT IV all four sections are involved. Involvement of the caudate lobe is a potential predictor of a poor outcome. Extrahepatic disease refers to diaphragm involvement, peritoneal seeding, ascites and abdominal lymph node metastasis. Distant metastasis is manly to the lung.

References:

1- Brown J, Perilongo G, Shafford E, Keeling J, Pritchard J, Brock P, Dicks-Mireaux C, Phillips A, Vos A, Plaschkes J: Pretreatment prognostic factors for children with hepatoblastoma-- results from the International Society of Paediatric Oncology (SIOP) study SIOPEL 1. Eur J Cancer. 36(11):1418-25, 2000

2- Perilongo G, Shafford E, Plaschkes J; Liver Tumour Study Group of the International Society of Paediatric Oncology: SIOPEL trials using preoperative chemotherapy in hepatoblastoma. Lancet Oncol. 1:94-100, 2000

3- Roebuck DJ, Aronson D, Clapuyt P, Czauderna P, de Ville de Goyet J, Gauthier F, Mackinlay G, Maibach R, McHugh K, Olsen OE, Otte JB, Pariente D, Plaschkes J, Childs M, Perilongo G; International Childrhood Liver Tumor Strategy Group: 2005 PRETEXT: a revised staging system for primary malignant liver tumours of childhood developed by the SIOPEL group. Pediatr Radiol. 37(2):123-32, 2007

4- Roebuck DJ: Assessment of malignant liver tumors in children. Cancer Imaging. 9: S98-S103, 2009

5- Meyers RL, Czauderna P, Otte JB: Surgical treatment of hepatoblastoma. Pediatr Blood Cancer. 59(5):800-8, 2012

6- Tajiri T, Kimura O, Fumino S, Furukawa T, Iehara T, Souzaki R, Kinoshita Y, Koga Y, Suminoe A, Hara T, Kohashi K, Oda Y, Hishiki T, Hosoi H, Hiyama E, Taguchi T: Surgical strategies for unresectable hepatoblastomas. J Pediatr Surg. 47(12):2194-8, 2012

---

* Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP
Professor of Pediatric Surgery, University of Puerto Rico - School of Medicine,
Rio Piedras, Puerto Rico. Director - Pediatric Surgery, San Jorge Childrens Hospital.
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

ISSN 1089-7739