



# **PEDIATRIC SURGERY Update** © **Vol. 31 No. 03 SEPTEMBER 2008**

## **Jeune Syndrome**

The two most common chest wall deformities in pediatric surgery are pectus excavatum and pectus carinatum. Pectus excavatum manifests itself during early childhood, while pectus carinatum is seen more commonly in early adolescent ages. Jeune syndrome is a rare autosomal recessive chest wall skeletal dysplasia also known as asphyxiating thoracic dystrophy or insufficiency. Almost all cases of Jeune syndrome has been described in children. The syndrome is characterized by respiratory distress, osseous dysplasia, and short stature. Patients generally die during the first months of life since the thoracic chest wall will not grow creating entrapment of lung and asphyxia. Besides a small thorax, varying degrees of rhizomelic brachymelia, polydactyly, pelvic abnormalities, renal anomalies, cystic lesions of the pancreas, retinal anomalies and hepatic fibrosis are also present in the syndrome. Diagnosis can be made prenatally. The life-saving procedures to expand the chests of infants born with Jeune asphyxiating thoracic dystrophy provide a static solution incapable of responding to the growth demands of thriving patients. Actual management consist either of dynamic lateral thoracic expansion with titanium struts or vertical expandable prosthetic titanium rib thoracoplasty creating additional chest wall that is formed of autologous tissue.

### **References:**

- 1- Giorgi PL, Gabrielli O, Bonifazi V, Catassi C, Coppa GV: Mild form of Jeune syndrome in two sisters. *Am J Med Genet.* 35(2):280-2, 1990
- 2- Chen CP, Lin SP, Liu FF, Jan SW, Lin SY, Lan CC: Prenatal diagnosis of asphyxiating thoracic dysplasia (Jeune syndrome). *Am J Perinatol.* 13(8):495-8, 1996
- 3- Kaddoura IL, Obeid MY, Mroueh SM, Nasser AA: Dynamic thoracoplasty for asphyxiating thoracic dystrophy. *Ann Thorac Surg.* 72(5):1755-8, 2001
- 4- Yerian LM, Brady L, Hart J: Hepatic manifestations of Jeune syndrome (asphyxiating thoracic dystrophy). *Semin Liver Dis.* 23(2):195-200, 2003
- 5- Davis JT, Long FR, Adler BH, Castile RG, Weinstein S: Lateral thoracic expansion for Jeune syndrome: evidence of rib healing and new bone formation. *Ann Thorac Surg.* 77(2):445-8, 2004
- 6- Waldhausen JH, Redding GJ, Song KM: Vertical expandable prosthetic titanium rib for thoracic insufficiency syndrome: a new method to treat an old problem. *J Pediatr Surg.* 42(1):76-80, 2007

## **Pectus Excavatum**

Pectus excavatum (funnel chest) is the most common chest wall deformity in children. The depression in the sternum in relation to the costal cartilages causes a distressful cosmetic defect with mild to no changes in pulmonary and cardiac function. Symptoms of lack of endurance, shortness of breath with exercise, or chest pain are frequent. Indications for surgical treatment includes severe, symptomatic deformity; paradoxical respiratory chest wall motion; CT scan with a pectus index greater than 3.25, cardiac

compression/displacement, pulmonary compression, pulmonary function studies showing restrictive disease, or other cardiac pathology secondary to compression of the heart. Originally repaired open using the Ratvich technique of costal cartilage removal, this has fallen into disuse and replaced with a minimally invasive technique called Nuss procedure where a titanium bar is placed behind the sternum and fixed to each side of the thoracic wall for two years before removal. Thoracoscopy while placing the bar behind the sternum helps avoid significant complications. Though the complications reported with the Nuss procedure are minimal, a few of them are significant such as laceration of the internal mammary artery, hemopericardium, recurrence of the pectus deformity after bar removal and bar displacement. There is a 1.5% incidence of postoperative wound infection. Surgical repair of the pectus excavatum improves cardiovascular function but there is no significant improvement in pulmonary function. The value of routine testing of pre- and postoperative lung function in patients with pectus excavatum is questionable. Good sternal elevation as measured by preoperative and postoperative CT scans can be achieved with the Nuss procedure regardless of the severity of chest depression or age. Results are good with excellent cosmetic results. Minimally invasive repair of pectus excavatum procedure and removal of the pectus bar should only occur in specialized institutions with wide experience in thoracic surgery.

#### **References:**

- 1- Malek MH, Berger DE, Housh TJ, Marelich WD, Coburn JW, Beck TW: Cardiovascular function following surgical repair of pectus excavatum: a metaanalysis. *Chest*. 130(2):506-16, 2006
- 2- Shin S, Goretsky MJ, Kelly RE Jr, Gustin T, Nuss D: Infectious complications after the Nuss repair in a series of 863 patients. *J Pediatr Surg*. 42(1):87-92, 2007
- 3- Aronson DC, Bosgraaf RP, Merz EM, van Steenwijk RP, van Aalderen WM, van Baren R: Lung function after the minimal invasive pectus excavatum repair (Nuss procedure). *World J Surg*. 31(7):1518-22, 2007
- 4- Vegunta RK, Pacheco PE, Wallace LJ, Pearl RH: Complications associated with the Nuss procedure: continued evolution of the learning curve. *Am J Surg*. 195(3):313-6, 2008
- 5- Nakagawa Y, Uemura S, Nakaoka T, Yano T, Tanaka N: Evaluation of the Nuss procedure using pre- and postoperative computed tomographic index. *J Pediatr Surg*. 43(3):518-21, 2008
- 6- Kelly RE Jr.: Pectus excavatum: historical background, clinical picture, preoperative evaluation and criteria for operation. *Semin Pediatr Surg*. 17(3):181-93, 2008
- 7- Nuss D: Minimally invasive surgical repair of pectus excavatum. *Semin Pediatr Surg*. 17(3):209-17, 2008

## **Trocar Hernias**

With the advent of minimally invasive procedures the trocar (port site) hernia has emerged. A trocar hernia is an abdominal defect caused by the sheath of the trocar through which omentum or even small bowel can protrude (incisional hernia). Trocar hernias occur in 3% of all laparoscopic procedures. Risk factors which increase the development of a trocar hernia are: large trocar (10 mm or larger), trocar design, use of linea alba for port placement, pre-existing fascial defect, pre-school children and skinny constitution. When small bowel protrudes a Richter's hernia develops. The usual presentation is of crampy abdominal pain with nausea and vomiting. Treatment is reduction of the bowel that is incarcerated and then repair of the fascial defect. To avoid trocar hernias fascial closure of port sites of 10-mm in adults and 5-mm in children should be accomplished whenever possible. Trocar site hernias in infants are mainly of omental protrusion and occur within

the first postoperative week.

**References:**

- 1- Holzinger F, Klaiber C: Trocar site hernias. A rare but potentially dangerous complication of laparoscopic surgery. *Chirurg.* 73(9):899-904, 2002
- 2- Boughey JC, Nottingham JM, Walls AC: Richter's hernia in the laparoscopic era: four case reports and review of the literature. *Surg Laparosc Endosc Percutan Tech.* 13(1):55-8, 2003
- 3- Tonouchi H, Ohmori Y, Kobayashi M, Kusunoki M: Trocar site hernia. *Arch Surg.* 139(11):1248-56,2004
- 4- Mahmoud Uslu HY, Ustuner EH, Sozener U, Ozis SE, Turkcapar AG: Cannula site insertion technique prevents incisional hernia in laparoscopic fundoplication. *Surg Laparosc Endosc Percutan Tech.* 17(4):267-70, 2007
- 5- Paya K, Wurm J, Fakhari M, Felder-Puig R, Puig S: Trocar-site hernia as a typical postoperative complication of minimally invasive surgery among preschool children. *Surg Endosc.* Feb 13,2008

---

\* 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](mailto:titolugo@coqui.net)  
Internet: <http://home.coqui.net/titolugo>

© PSU 1993-2008  
ISSN 1089-7739