



PEDIATRIC SURGERY Update ©

Vol. 45 No. 02 AUGUST 2015

ACNES

Anterior Cutaneous Nerve Entrapment Syndrome (ACNES) is a recent identified cause of chronic abdominal pain in children and adults. The superficial end branches of intercostal thoracic nerves twigs are somehow trapped at the level of the rectus abdominal muscle leading to pain, and in a few occasions vomiting. The exact pathophysiology of ACNES is unknown, but is probably related to traction or compression of the anterior portions of intercostal thoracic nerves, usually involving intercostal thoracic nerve VIII to XII. Pregnancy, abdominal trauma, and chronic nerve root compression caused by thoracolumbar or orthopedic conditions are occasionally identified as predisposing factors. Diagnosis of ACNES is suspected by a constant nagging localized abdominal pain that increases during play or sport associated with a positive Carnett sign. Carnett test is performed as follows: the physician localizes the point of maximal pain with his index finger, and the child is then asked to lift the head or upper torso or the legs while the palpating index finger remains on the painful spot. If this lifting aggravates the pain, its origin is probably located in the abdominal wall. Hypoesthesia, hyperesthesia/algnesia or sometimes even allodynia may be found by comparison to the normal contralateral abdominal side. A positive pinch test is a sensitive and highly underrated sign reflecting the presence of a neuropathic pain syndrome such as ACNES. Attenuating the pain using trigger point local anesthetic infiltration support the diagnosis. These children undergo extensive laboratory and imaging workup which are usually negative and expensive. Management consists of pain medication. If pain persists, US-guided transverse abdominis plane technique blocking the entrapped nerve using subfascial lidocaine with steroid infiltration is utilized. With recurrent or recalcitrant pain an anterior cutaneous neurectomy will be needed. The neurectomy removes the affected nerve end twigs. This type of surgical procedure is highly successful in children and adults.

References:

- 1- Scheltinga MR, Boelens OB, Tjon A Ten WE, Roumen RM: Surgery for refractory anterior cutaneous nerve entrapment syndrome (ACNES) in children. *J Pediatr Surg.* 46(4):699-703, 2011
- 2- Boelens OB, Scheltinga MR, Houterman S, Roumen RM: Management of anterior cutaneous nerve entrapment syndrome in a cohort of 139 patients. *Ann Surg.* 254(6):1054-8, 2011
- 3- Boelens OB, van Assen T, Houterman S, Scheltinga MR, Roumen RM: A double-blind, randomized, controlled trial on surgery for chronic abdominal pain due to anterior cutaneous nerve entrapment syndrome. *Ann Surg.* 257(5):845-9, 2013
- 4- Akhnikh S, de Korte N, de Winter P: Anterior cutaneous nerve entrapment syndrome (ACNES): the forgotten diagnosis. *Eur J Pediatr.* 173(4):445-9, 2014
- 5- Nizamuddin SL, Koury KM, Lau ME, Watt LD, Gulur P: Use of targeted transversus abdominis plane blocks in pediatric patients with anterior cutaneous nerve entrapment syndrome. *Pain Physician.* 17(5):E623-7, 2014
- 6- Bairdain S, Dinakar P, Mooney DP: Anterior Cutaneous Nerve Entrapment Syndrome in Children. *J Pediatr Surg* 50(7): 1177-1179, 2015

POEM

Achalasia is a rare motility disorder of the esophagus due to absence of peristalsis in the esophageal body impairing relaxation of the lower esophageal sphincter during swallowing.

The etiology of achalasia is likely to be affected by various factors causing immune-mediated ganglionitis which results in degeneration of the myenteric nerve plexus of the esophageal wall. Achalasia causes regurgitation, dysphagia to solids, retrosternal pain and weight loss. The diagnosis of achalasia is first suggested in a barium swallow and corroborated using esophageal manometry studies. Achalasia is either managed by esophageal balloon dilatation and most effectively with either open or laparoscopic anterior surgical esophageal myotomy. Peroral endoscopic myotomy (POEM) is a novel procedure developed recently which has ameliorated the symptoms of patients with achalasia. The endoscopic procedure, performed in the operating room under general anesthesia, consists of entry incision on the mucosa at the level of the gastroesophageal junction, establishing a tunnel on the submucosa, performing the myotomy followed by sealing of the entry incision. The myotomy is primarily performed for the circular muscle fibers or both the circular and longitudinal muscle fibers. Leak through the mucosal seal can cause peritonitis and/or mediastinitis. Compared with the surgical approach to achalasia, POEM has the advantage of minimally invasiveness. POEM can significantly improve esophageal motility by decreasing upper esophageal sphincter and lowering esophageal sphincter pressure. POEM has no effect on esophageal body peristalsis. After POEM incomplete myotomy and gastroesophageal reflux can become a problem. The role of POEM in the treatment of other esophageal motor disorders such as diffuse esophageal spasm, non-relaxing hypertensive LES and nutcracker esophagus is still debatable. POEM is a safe and effective technique for treating pediatric achalasia.

References:

- 1- Allaix ME, Patti MG: Endoscopic Dilatation, Heller Myotomy, and Peroral Endoscopic Myotomy: Treatment Modalities for Achalasia. *Surg Clin North Am.* 95(3):567-578, 2015
- 2- Zhang Y, Ling-hu E, Zhai Y, Peng L, Wang X: Peroral endoscopic myotomy plus balloon shaping for achalasia: a preliminary study. *Hepatogastroenterology.* 62(137):82-6, 2015
- 3- Tang X, Gong W, Deng Z, Zhou J, Ren Y, Zhang Q, Chen Z, Jiang B: Usefulness of peroral endoscopic myotomy for treating achalasia in children: experience from a single center. *Pediatr Surg Int.* 31(7):633-8, 2015
- 4- Kumbhari V, Khashab MA: Peroral endoscopic myotomy. *World J Gastrointest Endosc.* 7(5):496-509, 2015
- 5- Yao S, Linghu E: Peroral endoscopic myotomy can improve esophageal motility in patients with achalasia from a large sample self-control research (66 patients). *PLoS One.* 10(5):e0125942, 2015
- 6- Ramchandani M, Reddy DN, Darisetty S, Kotla R, Chavan R, Kalpala R, Galasso D, Lakhtakia S, Rao GV: Peroral endoscopic myotomy for Achalasia Cardia: A single center experience of over 200 consecutive patients: treatment analysis and follow up. *Dig Endosc.* May 27, 2015

Lymphorrhoea

Lymphorrhoea is an abnormal flow of lymph draining externally from disrupted lymphatic vessels or previously retained within a wound. In children lymphorrhoea usually occurs following a surgical procedure for a lymphatic anomaly, following cardiothoracic surgery, in penetrating trauma to the neck or during removal of lymph nodes for diagnostic or therapeutic purposes. Complications of uncontrolled lymphorrhoea can lead to subcutaneous collection of lymphatic fluid or a draining fistula. High output lymphatic leakage is associated with increased mortality when intervention is not initiated in a timely fashion due to critical loss of fluids, proteins and electrolytes causing lymphocyte and antibody depletion. Most cases of lymphorrhoea in children occur after surgery for lymphangioma or cystic hygroma, a benign condition of the lymphatic system consisting of cysts of varying size. Initial management of lymphorrhoea consists of diet modification, drainage, pressure dressings, and reoperation to stop lymphatic drainage. Octeotride, a somatostatin analog has been used successfully to manage post-operative lymphorrhoea in children after removal of large lymphangioma in the cervical and axillary region. Midodrine,

an oral selective alpha-adrenergic drug, can affect contraction of smooth muscle and decrease lymphatic flow. Sclerotherapy using either doxycycline, ethanol, bleomycin or minocycline has also been successfully used to manage lymphatic leakage. Surgical exploration to occlude lymphatic vessels with the use of fibrin sealant has also produce benefit in selected patients.

References:

- 1- Suver DW, Perkins JA, Manning SC: Somatostatin treatment of massive lymphorrhea following excision of a lymphatic malformation. *International J Pediatr Otorhinolaryngology*. 68: 845-850, 2004
- 2- Carcoforo P, Soliani G, Maestroni U: Octeotride in the Treatment of Lymphorrhea after Axillary Node dissection. A prospective randomize controlled trial. *J Am Coll Surg*. 196:365-369, 2003
- 3- Lemaitre J, Segers B, Lebrun E: [The use of lanreotide in the management of lymphorrhea after an aortic valve replacement]. *Interactive Cardiovascular and Thoracic Surgery* 15:762-763, 2012
- 4- Friedmann MD: Evaluation and Management of Lymphadenopathy in Children. *Pediatrics in Review*. 29(2): 53-60, 2008
- 5- Miloundja J, Manfoumbi Ngoma AB, Mba Ella R, Nguema Edzang B, N'Zouba L: [Cystic cervicofacial lymphangioma in children in Gabon]. *Ann Otolaryngol Chir Cervicofac*. 124(6):277-84, 2007
- 6- Cuervo JL, Galli E, Eisele G, Johannes E, Fainboim A, Tonini S, Joaquin W, Viola B, Nazar M: [Lymphatic malformations: percutaneous treatment with bleomycin]. *Arch Argent Pediatr*. 109(5):417-22, 2011

***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)-999-9450 Fax (787)-720-6103 E-mail: *titolugo@coqui.net*
Internet: <http://home.coqui.net/titolugo>