



PEDIATRIC SURGERY Update ©

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Long Gap Esophageal Atresia

Long gap in esophageal atresia (EA), a major surgical challenge considered when an anatomic gap of more than three centimeters (or three vertebral bodies) is identified between the two stumps of the esophagus precluding a safe anastomosis. Long gap is more commonly seen in pure esophageal atresia not associated with a tracheoesophageal fistula. Surgeons have also used this term in cases of EA with distal tracheoesophageal fistula when an anastomosis is not possible. No single technique to accomplish this goal is effective in all patients. When confronted with this situation surgeons should reproduce several alternatives of management with the central caveat that the baby own esophagus should be preserved and is far superior to an esophageal replacement technique. In cases of long gap with pure EA initial management consist of a gastrostomy for two purposes: enteral feeding and future imaging studies to determine the esophageal gap. Should the gap be less than 2 cm a primary anastomosis can be accomplished safely. Some of the commonly used technique for long gap approximation includes proximal esophageal dilatation with distal gastric bolus feedings in an effort to exert pressure on both stumps for spontaneous lengthening, a technique that takes to much time to accomplish its goal. This has prompted surgeons to developed faster technique such as sequential proximal pouch elongation (Kimura advancement) and mechanical stretching of both stumps with sutures (Foker). Still other techniques when the distal stump is extremely small encompassed isoperistalsis tubularization of a piece of stomach (Scharli) to include the esophageal stump, and creating concomitant antireflux valve procedures (Collis-Nissen). Finally, replacement of the defect with stomach (gastric transposition), small bowel or colon is a last resort technique when all other means fails.

References:

- 1- Stringel G, Lawrence C, McBride W: Repair of long gap esophageal atresia without anastomosis. *J Pediatr Surg.* 45(5):872-5, 2010
- 2- Nakahara Y, Aoyama K, Goto T, Iwamura Y, Takahashi Y, Asai T: Modified Collis-Nissen procedure for long gap pure esophageal atresia. *J Pediatr Surg.* 47(3):462-6, 2012
- 3- Friedmacher F, Puri P: Delayed primary anastomosis for management of long-gap esophageal atresia: a meta-analysis of complications and long-term outcome. *Pediatr Surg Int.* 28(9):899-906, 2012
- 4- Nasr A, Langer JC: Mechanical traction techniques for long-gap esophageal atresia: a critical appraisal. *Eur J Pediatr Surg.* 23(3):191-7, 2013
- 5- Miyano G, Okuyama H, Koga H, Okawada M, Doi T, Takahashi T, Nakamura H, Suda K, Lane GJ, Okazaki T, Yamataka A: Type-A long-gap esophageal atresia treated by thoracoscopic esophagoesophagostomy after sequential extrathoracic esophageal elongation (Kimura's technique). *Pediatr Surg Int.* 29(11):1171-5, 2013

- 6- Bairdain S, Ricca R, Riehle K, Zurakowski D, Saites CG, Lien C, Anderson GF, Wahoff DC, Linden BC: Early results of an objective feedback-directed system for the staged traction repair of long-gap esophageal atresia. *J Pediatr Surg.* 48(10):2027-31, 2013
- 7- Beasley SW, Skinner AM: Modified Scharli technique for the very long gap esophageal atresia. *J Pediatr Surg.* 48(11):2351-3, 2013

Embryonal Liver Sarcoma

Undifferentiated embryonal sarcoma of the liver (UESL) is an uncommon malignant hepatic tumor of mesenchymal origin in children. UESL accounts for almost 5% of all pediatric malignant hepatic tumors occurring mainly between five and 10 years of age without a gender predilection. Clinical presentation is typically an abdominal mass that may be accompanied by pain and systemic symptoms such as fever, weight loss or vomiting. Due to its high malignant potential UESL often metastasizes to the lung, peritoneum and pleura resulting in a poor prognosis. The tumor presents as a large (above 10 cm) solitary solid mass mainly localized in the right hepatic lobe with a solitary clear boundary. US reveals a mixed echoic mass containing an irregular anechoic region with multiple small capsular spaces. CT reveals cystic lesions due to hemorrhage, necrosis and liquefaction. Diagnosis requires biopsy. Microscopically the tumor consists of rather loose foci of pleomorphic, stellate or giant cells with hyperchromatic nuclei, and numerous mitosis. Immunohistochemistry of the tumor shows positive expression of SMA, α -ACT, desmin, vimentin and actin, while Alpha fetoprotein, CEA, CA 19-9 and cytokeratin are negative. Rarely UESL is misdiagnosed as hepatoblastoma. The key management of UESL is complete surgical resection followed by postoperative multiagent chemotherapy. Very large tumors can be managed with preop chemotherapy followed by surgery. Hepatic transplantation is an effective therapeutic method for UESL patients whose tumor unresectable or those who have postoperative recurrence of the tumor. Prognosis has improved significantly over the years.

References:

- 1- Bisogno G, Pilz T, Perilongo G, Ferrari A, Harms D, Ninfo V, Treuner J, Carli M: Undifferentiated sarcoma of the liver in childhood: a curable disease. *Cancer.* 94(1):252-7, 2002
- 2- Chowdhary SK, Trehan A, Das A, Marwaha RK, Rao KL: Undifferentiated embryonal sarcoma in children: beware of the solitary liver cyst. *J Pediatr Surg.* 39(1):E9-12, 2004
- 3- Wei ZG, Tang LF, Chen ZM, Tang HF, Li MJ: Childhood undifferentiated embryonal liver sarcoma: clinical features and immunohistochemistry analysis. *J Pediatr Surg.* 43(10):1912-9, 2008
- 4- Plant AS, Busuttill RW, Rana A, Nelson SD, Auerbach M, Federman NC: A single-institution retrospective cases series of childhood undifferentiated embryonal liver sarcoma (UELS): success of combined therapy and the use of orthotopic liver transplant. *J Pediatr Hematol Oncol.* 35(6):451-5, 2013
- 5- Gao J, Fei L, Li S, Cui K, Zhang J, Yu F, Zhang B: Undifferentiated embryonal sarcoma of the liver in a child: A case report and review of the literature. *Oncol Lett.* 5(3):739-742, 2013
- 6- Geel JA, Loveland JA, Pitcher GJ, Beale P, Kotzen J, Poole JE: Management of undifferentiated embryonal sarcoma of the liver in children: a case series and management review. *S Afr Med J.* 103(10):728-31, 2013
- 7- Ismail H, Dembowska-Baginska B, Broniszczak D, Kalicinski P, Maruszewski P, Kluge P, Swieszkowska E, Kosciesza A, Lembas A, Perek D: Treatment of undifferentiated embryonal sarcoma of the liver in children--single center experience. *J Pediatr Surg.* 48(11):2202-6, 2013

Trachea Diverticulum

Tracheal diverticulum is a very rare condition with a congenital or acquired origin. Congenital tracheal diverticulum is even rarer, smaller, located approximately 4–5 cm below vocal cords or just above the carina, is more common in males and can be considered as a supernumerary malformed branch of the trachea. Most reported cases of tracheal diverticulum are acquired after esophageal or tracheal surgery, orotracheal intubation or increased intraluminal pressure through a weak area of tracheal wall. It occurs more often after surgery for tracheoesophageal fistula when the fistula is not divided sufficiently close to the trachea. In this case the diverticulum becomes a remnant of esophageal epithelium created at the time of repair of the TEF. In either congenital or acquired cases the diverticulum acts as a reservoir for pooling of secretions that may spill over into the tracheobronchial tree predisposing affected patients to cough, dyspnea, stridor and chronic chest infection. CT-Scan imaging with thin slices or bronchoscopy is diagnostic. Surgical treatment of congenital tracheal diverticulum is rarely advocated. Excision of the acquired diverticulum may be needed in patients with severe respiratory symptoms, either a large diverticulum and airway compression or a suspected recurrent fistula. Another less invasive and effective alternative management is endoscopic carbon dioxide laser fulguration of the tracheal diverticulum mucosa.

References:

- 1- Sharma BG: Tracheal diverticulum: a report of 4 cases. *Ear Nose Throat J.* 88(1):E11, 2009
- 2- Berlucchi M, Pedruzzi B, Padoan R, Nassif N, Stefani S: Endoscopic treatment of tracheocele in pediatric patients. *Am J Otolaryngol.* 31(4):272-5, 2010
- 3- Gaisert HA, Grillo HC: Complications of the tracheal diverticulum after division of congenital tracheoesophageal fistula. *J Pediatr Surg.* 41(4):842-4, 2006
- 4- Johnson LB, Cotton RT, Rutter MJ: Management of symptomatic tracheal pouches. *Int J Pediatr Otorhinolaryngol.* 71(4):527-31, 2007
- 5- Cheng AT, Gazali N: Acquired tracheal diverticulum following repair of tracheo-oesophageal fistula: endoscopic management. *Int J Pediatr Otorhinolaryngol.* 72(8):1269-74, 2008
- 6- Shah AR, Lazar EL, Atlas AB: Tracheal diverticula after tracheoesophageal fistula repair: case series and review of the literature. *J Pediatr Surg.* 44(11):2107-11, 2009

* 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>

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