



PEDIATRIC SURGERY Update © **Vol. 39 No. 01 JULY 2012**

Intrathoracic Kidney

Congenital intrathoracic kidney is a very rare ectopic anomaly of the kidney. Most ectopic kidneys are found in the pelvic or lower lumbar region. Delayed closure of the pleuroperitoneal membrane as it occurs in diaphragmatic hernia permits excessive cranial migration of the kidney. Other postulates that delayed ingrowth of the ureter into the metanephros results in prolongation of the ascending process involving a long ureter and prolonged migration. In either case the kidney assumes a thoracic position. Intrathoracic kidneys do not display dysplastic parenchymal architecture and the kidneys are usually functional. Thoracic kidneys can be classified as thoracic renal ectopia with closed diaphragm, eventration of the diaphragm, diaphragmatic hernia (congenital diaphragmatic defects or acquired hernia such as Bochdalek hernia), and traumatic rupture of the diaphragm with renal ectopia. Conditions that affect the normal kidney such as ureteric stones, hydronephrosis, and renal cell carcinoma has also been reported to occur in intrathoracic kidneys later in life. Intrathoracic kidney can be found as a unique anomaly or associated with a diaphragmatic hernia defect, respiratory symptoms and bowel herniation within the thorax. Diagnosis is confirmed with CT or MR imaging. When presenting as a unique anomaly not associated to a bowel herniation or respiratory symptoms they can be managed conservatively with regular follow-up since normal growth and development of the affected kidney occurs. Surgical treatment has been reserved for children with associated bowel herniation or respiratory compromise. In such situation repair of the diaphragmatic defect and nephropexy is in order.

References:

- 1- Ho HF, Liaw SB, Chang PY: Delayed manifestation of congenital diaphragmatic hernia with intrathoracic kidney: report of one case. *Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi.* 38(1):61-4, 1997
- 2- Oon PC, Shen HN, Yang PC: Intrathoracic kidney. *J Formos Med Assoc.* 104(2):120-2, 2005
- 3- Panda B, Rosenberg V, Cornfeld D, Stiller R: Prenatal diagnosis of ectopic intrathoracic kidney in a fetus with a left diaphragmatic hernia. *J Clin Ultrasound.* 37(1):47-9, 2009
- 4- Rouanne M, Le Mandat A, Dorgeret S, Philippe-Chomette P, El Ghoneimi A: A rare case of ectopic intrathoracic kidney in a 1-year-old child. *Urology.* 76(1):57-9, 2010
- 5- Fiaschetti V, Velari L, Gaspari E, Mastrangeli R, Simonetti G: Adult intra-thoracic kidney: a case report of bochdalek hernia. *Case Report Med.* 2010. pii: 975168, 2010
- 6- Murphy JJ, Altit G, Zerhouni S: The intrathoracic kidney: should we fix it? *J Pediatr Surg* 47: 970-973, 2012

Port Inversion

Ports connected to central venous catheter are necessary for a variety of reasons such

as management of children who require chemotherapy, prolonged antibiotic administration, frequent blood sampling, parenteral nutrition and transfusion of blood and blood products. Totally implantable access ports improve the quality of life and have the advantage of not requiring an external dressing. Ports are inserted as totally implantable devices underneath the subcutaneous tissue infraclavicularly most commonly or under the pectoralis fascia. The most common complications of port application and maintenance include occlusion, vein thrombosis, port pocket infection, catheter related sepsis, device rotation or dislodgement, catheter migration, skin necrosis, pocket hematoma and exposed port. Port inversion occurs when the port completes a 90-degree rotation due to mobility and the caretaker cannot cannulate the port diaphragm. A simple lateral chest film will reveal the problem of port inversion. Port inversion is seen more commonly after subcutaneous placement than subpectoral fascia placement. Port implantation in the subpectoral fascia pocket has been found to have a lower rate of skin complications than the subcutaneous pocket implantation site. Whether to fix the port with sutures to avoid rotation or inversion is a matter of debate, since suturing will need more extensive dissection during removal. Port inversion will need revision.

References:

- 1- Dillon PA, Foglia RP: Complications associated with an implantable vascular access device. *J Pediatr Surg.* 41(9):1582-7, 2006
- 2- Charles HW, Miguel T, Kovacs S, Gohari A, Arampulikan J, McCann JW: Chest port placement with use of the single-incision insertion technique. *J Vasc Interv Radiol.* 20(11):1464-9, 2009
- 3- McNulty NJ, Perrich KD, Silas AM, Linville RM, Forauer AR: Implantable subcutaneous venous access devices: is port fixation necessary? A review of 534 cases. *Cardiovasc Intervent Radiol.* 33(4):751-5, 2012
- 4- Rouzrokh M, Shamsian BS, KhaleghNejad Tabari A, Mahmoodi M, Kouranlo J, Manafzadeh G, Arzanian MT, Fallah F, Anoush M, Abdollah Gorji F: Totally implantable subpectoral vs. subcutaneous port systems in children with malignant diseases. *Arch Iran Med.* 12(4):389-94, 2009

Idiopathic Megaduodenum

Idiopathic megaduodenum is a rare congenital condition defined as a massive dilatation of the duodenum without evidence of mechanical extraluminal or intraluminal obstruction. The etiology is unknown. It may be caused by neuromuscular disorder or abnormal gut collagen and interticium. Duodenal dilatation and stasis may be caused by decreased peristalsis and accumulation of motilin in the duodenal cavity. Children with idiopathic megaduodenum (ID) present with recurrent abdominal distension, nausea vomiting, diarrhea and malnutrition. Diagnosis is obtained with upper GI series. Management is surgical and should achieve drainage of the duodenum, reduce the capacity of the duodenum and cut off the forceful feeding of the duodenum. This can be accomplished more physiologically with tapering duodenoplasty and side-to-side gastrojejunostomy, duodenojejunostomy or Roux-en-Y duodenojejunostomy. A familiar variety of this condition has been described in the literature. Also, children with chronic intestinal pseudoobstruction can present with idiopathic megaduodenum.

References:

- 1- McClenahan JE, Fisher B: Idiopathic megaduodenum; report of a case. *Am J Dig Dis.* 15(12):414-6, 1848
- 2- Eaves ER, Schmidt GT: Chronic idiopathic megaduodenum in a family. *Aust N Z J Med.* 15(1):1-6, 1985
- 3- Cheng SC, Sanderson CR, Way NJ, Yoong PM: Familial idiopathic megaduodenum. *Aust N Z J Surg.* 57(11):879-82, 1987
- 4- Svartsja B, Sulonen H, Leino R: [Megaduodenum as a first sign of chronic idiopathic intestinal pseudo-obstruction]. *Duodecim.* 106(7):578-81, 1990
- 5- Nichol PF, Stoddard E, Lund DP, Starling JR: Tapering duodenoplasty and Roux-en-Y duodenojejunostomy in the management of adult megaduodenum. *Surgery.* 135(2):222-4, 2004
- 6- Zhang XW, Abudoureyimu A, Zhang TC, Zhao JR, Fu LB, Lin F, Qiu XH, Chen YJ: Tapering duodenoplasty and gastrojejunostomy in the management of idiopathic megaduodenum in children. *J Pediatr Surg.* 47(5):1038-42, 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>

© PSU 1993-2012
ISSN 1089-7739