PEDIATRIC SURGERY *Update**
Vol. 56 No. 04 APRIL 2021

Abdominal Wall Defects and Undescended Testes

The most common congenital abdominal wall defects (AWD) in children affecting testicular descent include gastroschisis, omphalocele and prune belly syndrome. Gastroschisis is more common than omphalocele or prune belly syndrome. Undescended testes are more common among AWD patients than the general population, with the highest prevalence in omphalocele. Testicular descent is known to be related to intraabdominal pressure and the gubernaculum. Without higher pressures the forces that encourage testicle migration are absent. Failure of appropriated abdominal pressure and/or sudden disruption of the gubernaculum at the time of formation can lead to abdominal undescended testes. Undescended testes are a concomitant birth anomaly associated with abdominal wall defects in almost 20 to 40% of all males patients. Undescended testes in children with abdominal wall defects may be found within or outside the abdominal cavity. The majority of undescended testes are found at the internal spermatic ring. Gestational age and birth weight do not appear to be a significant factor associated with testicular maldescent in several series. Closure of the abdominal wall defect using either mesh or primary closure is initially warranted. After closure of the abdominal wall defect and in the ensuing next 12 months, more than half of all undescended testes migrate to the appropriate position at follow-up requiring no intervention. Migration of the testes into the scrotum in cases of left-sided undescended testes born with gastroschisis, along with most in omphalocele is less likely. Of the testes that do not spontaneously descend into the scrotum, nearly half (43%) migrated into the inguinal canal. In cases where the testes remained in the abdominal cavity (non-palpable), laparoscopy is successfully performed to localize and remove or reposition the testes. Early conservative management allows normal spontaneous descent in most testes. Testes that are extra-abdominal at birth appear to be less likely to spontaneously migrate into the scrotum compared with those that are intraabdominal at birth. Extra-abdominal testes seem to have a greater incidence of atrophy and need for orchiectomy. Babies with prolapse testes out of the abdominal cavity should undergo manual repositioning placing the testis as near as possible to the internal spermatic ring. It is important to record the position of the intraabdominal undescended testes at the time of abdominal wall repair because future diagnostic laparoscopy or exploration is likely to be difficult. The majority will descend on their own without any need for surgical intervention. Orchiopexy in a newborn with fragile testicular vessels and the risk of compromised blood flow in the presence of temporarily raised intracelomic pressure after primary closure may jeopardize testicular viability. Other authors believe that early mobilization and fixation can improve the outcome of the undescended testis. Surgical intervention for an undescended

testis after closure of AWD is challenging, characterized by adhesions and short gonadal vessels necessitating staging the descent.

References:
1- Lawson A, de La Hunt MN: Gastroschisis and undescended testis. J Pediatr Surg. 36(2):366-7, 2001
2- Chowdhary SK, Lander AD, Buick RG, Corkery JJ, Gornall P: The primary management of testicular maldescent in gastroschisis. Pediatr Surg Int. 17(5-6):359-60, 2001
3- Hill SJ, Durham MM: Management of cryptorchidism and gastroschisis. J Pediatr Surg. 46(9):1798-803, 2011
4- Yardley IE, Bostock E, Jones MO, Turnock RR, Corbett HJ, Losty PD: Congenital abdominal wall defects and testicular maldescent--a 10-year single-center experience. J Pediatr Surg. 47(6):1118-22, 2012
5- Raitio A, Syvanen J, Tauriainen A, et al: Congenital abdominal wall defects and cryptorchidism: a population-based study. Pediatr Surg Int. 2021 Jan 31. doi: 10.1007/s00383-021-04863-9.
6- Berger AP, Hager J: Management of neonates with large abdominal wall defects and undescended testis. Urology. 68(1):175-8, 2006

Congenital Anal Stenosis

Congenital anal stenosis is a rare disorder classified as a low anorectal malformation, mainly a short stenosis in most cases, but sometimes the child has a funnel shape long stenosis associated with a presacral mass, bony sacral defect, anterior meningocele or other anomaly. The type of sacral dysplasia is scimitar and the presacral tumor is a mature teratoma in most of the cases. Children with congenital anal stenosis suffer from chronic intractable constipation, soiling, encopresis and mega rectosigmoid. Constipation is the most common early functional problem in children with anal stenosis in more than 40% of the cases. Recalcitrant constipation is more common in children with a delayed diagnosis and treatment of anal stenosis. Congenital anal stenosis (CAS) can also be associated with Mayer-Rokitansky-Küster-Hauser syndrome. On physical examination, children with CAS have a stenotic opening at the normal anal site barely admitting a small Hegar dilator. Palpable fecalomas can be found upon abdominal examination. In this malformation, the anal canal is usually located at least partially inside the voluntary sphincter funnel. Occasionally the diagnosis of anal stenosis is delayed to later infancy, especially in cases where the bowel outlet is stenotic but at or near the proper anal position. Newborns with anal stenosis usually pass meconium in the first 48 hours after birth. Children born with CAS should undergo an active search for associated malformations including echocardiogram, ultrasound of the spinal cord and kidneys, cystourethrogram and imaging of the entire spine including the sacrum (MRI). CAS can be managed with gradual Hegar dilatations usually without the need for anesthesia. Serial dilatations are started with a Hegar dilator that is easily fitted into the anal opening, usually with a size between six- and 8-mm. Dilatations are taught to parents to continue management increasing to the next number on a weekly basis, continued for six weeks after the age-appropriate size of the anus is reached. Currarino syndrome, dysganglionosis including Hirschsprung's disease and chromosomal defects commonly occurs in children with funnel anus. The mortality of patients with low anomaly as CAS, is about three times lower than that of patients with high anomalies, and usually associated to cardiac defects. Long-term results of low malformations are usually good in most patients. Poor results are usually associated to neurological damage, mental retardation or insufficient care of patients. In severe cases of

anal stenosis, the posterior rectum is mobilized in the form of rectal advancement, and the posterior 180 degrees is anastomosed directly to the skin with preservation of the anal canal as the anterior final anoplasty. These patients have an excellent prognosis for bowel control and fecal continence, and therefore, complete mobilization and resection of the anal canal must be avoided. Those children with CAS and stenosis involving only the skin-level can be managed with a Heineke-Mikulicz anoplasty with very good results.

References:
1- Suomalainen A, Wester T, Koivusalo A, Rintala RJ, Pakarinen MP: Congenital funnel anus in children: associated anomalies, surgical management and outcome. Pediatr Surg Int. 23(12):1167-70, 2007
2- Joshi M, Singh S, Vyas T, Chourishi V, Jain A: Mayer-Rokitansky-Kuster-Hauser syndrome and anal canal stenosis: case report and review of literature. J Pediatr Surg. 45(12):e29-31, 2010
3- Pakarinen MP, Rintala RJ: Management and outcome of low anorectal malformations. Pediatr Surg Int. 26(11):1057-63, 2010
4- Lane VA Wood RJ, Reck C, Skerritt C, Levitt MA: Rectal atresia and anal stenosis: the difference in the operative technique for these two distinct congenital anorectal malformations. Tech Coloproctol. 20(4):249-54, 2016
5- Halleran DR, Sanchez AV, Rentea RM, et al: Assessment of the Heineke-Mikulicz anoplasty for skin level postoperative anal strictures and congenital anal stenosis. J Pediatr Surg. 54(1):118-122, 2019
6- Brem H, Beaver BL, Colombani PM, et al: Neonatal diagnosis of a presacral mass in the presence of congenital anal stenosis and partial sacral agenesis. J Pediatr Surg. 24(10):1076-8, 1989

Appendix Duplication

The vermiform appendix is a tubular, narrow, worm-shaped part of the alimentary canal that lies near the ileocecal junction and communicates with the cecum. The appendix develops as a conical extension from the apex of the cecal diverticulum which arises from the antimesenteric border of the proximal part of the post arterial segment of the midgut. Anomalies associated with the appendix are rare. Duplication of the vermiform appendix is very rare occurring with an incidence of 0.0004% after appendectomy. Appendiceal duplication is classified into three types (Cave-Wallbridge classification). Type A consists of various degrees of partial duplication on a normally localized appendix with a single cecum. Type B consists of a single cecum with two completely separate appendixes. This type is further subdivided into a B1, bird-like type if the two appendixes are located symmetrical on either side of the cecum as usually occurs in birds, and B2 also known as tenia-coli type which has a normally located appendix arising from the cecum at the usual site and a second separate rudimentary appendix located along the line of one of the tenias. B1 or bird-like type of duplication is the most common type of appendiceal duplication (37%). Type B2 duplication can be mimicked by a solitary inflamed diverticulum found on the cecum, usually at the medial border just above the ileocecal junction. B3 if the second appendix is located along the tenia of the hepatic flexure of the colon, and B4 if the location of the second appendix is along the tenia of the splenic flexure of the colon. Type C consists of a duplicated cecum, each with an appendix. A horseshoe configuration of duplication and triple appendices have also been described. Appendiceal duplication is most commonly identified incidentally during surgery or at autopsies. The most common presentation is appendicitis with the duplication being discovered intraoperatively. Median age at presentation of these patients is adolescent years, males and females affected

equally. CT-Scan is the best mode of imaging to identify a duplicated appendix. Besides inflammation, appendiceal duplication can present with recurrent intussusception or an appendiceal mass. Surgeons have to be aware of such anomalies since a second laparotomy revealing a previously removed appendix can cause medicolegal situations. In cases with appendiceal duplication, when only one appendix is inflamed, both should be removed to avoid a diagnostic dilemma that may arise later.

References:

1- Varshney M, Shahid M, Maheshwari V, Mubeen A, Gaur K: Duplication of appendix: an accidental finding. BMJ Case Rep. 2011 Mar 8;2011:bcr0120113679. doi: 10.1136/bcr.01.2011.3679.
2- Marshall AP, Issar NM, Blakely ML: Aapendiceal duplication in children presenting as a appendiceal tumor and as recurrent intussusception. J Pediatr Surg. 48: E9-E12, 2013
3- Dubhashi SP, Dubhashi UP, Kumar H, Patil C: Double Appendix. Indian J Surg. 77(Suppl 3):1389-90, 2015
4- Bhat GA, Reshi TA, Rashid A: Duplication of Vermiform Appendix. Indian J Surg. 78(1):63-4, 2016
5- Nageswaran H, Khan U, Hill F, Maw A: Appendiceal Duplication: A Comprehensive Review of Published Cases and Clinical Recommendations. World J Surg. 42(2):574-581, 2018
6- Chew DK, Borromeo JR, Gabriel YA, Holgersen LO: Duplication of the vermiform appendix. J Pediatr Surg. 35(4):617-8, 2000

***Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP**
Professor of Pediatric Surgery, UPR - School of Medicine, UCC School of Medicine & Ponce School of Medicine.
Director - Pediatric Surgery, San Jorge Children's & Woman Hospital.
Postal Address: P.O. Box 10426, San Juan, Puerto Rico USA 00922-0426.
Tel (787) 340-1868 E-mail: *pediatricsurgerypr@gmail.net*
Internet: pedsurgeryupdate.com

**** PSU 1993-2021***
ISSN 1089-7739