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Surgery

HINDGUT DUPLICATIONS IN CHILDREN; SURGICAL CHALLENGES AND LESSONS LEARNT.

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ABSTRACT

Background Hind gut duplications account for 8% of all alimentary canal duplications. The varied presentation and the altered anatomy of the colon and its frequent association with other organ systems makes the management of theses rare anomalies a challenge.

Materials and methods The authors have retrospectively reviewed their experience of 7 cases of children who were diagnosed as colonic duplication on clinical and radiological evaluation. Diagnostic modalities, findings at laparotomy and surgical management has been described.

Results Successful surgical outcome was seen in six out the seven cases.

Conclusion The surgical management of the colon duplication is challenging. The treatment has to be tailored for each of the patient depending on the anatomical variation and the associated congenital anomalies.

KEYWORDS : Tubular Colon Duplication, Double rectourethral fistula, Rectovestibular Fistula, Caudal Duplication

Introduction

Alimentary canal duplications are uncommon congenital anomalies, with a reported incidence of 1:18000 live births [1]. They can present from oropharynx to anal canal and common wall and blood supply. They are either cystic or tubular. Alimentary canal duplications are commonly seen in ileum or esophagus. Duplications of colon account for 8 to 13 % of total gastrointestinal (GI) duplications [2]. The presenting symptoms of cystic duplications depends on the site and size, while, the tubular duplication of colon may remain asymptomatic or present with cosmetic issues due to double termination of duplicated colon. Tubular colon duplications are often associated with anomalies of other systems.

We describe our experience with 7 cases of colorectal duplications. Three children had cystic variety one in transverse colon and two in rectum. Three patients had anorectal malformations along with duplicated colon with double termination. In one of the child the tubular duplication of colon was a part of caudal duplication syndrome.

Material methods

Patients

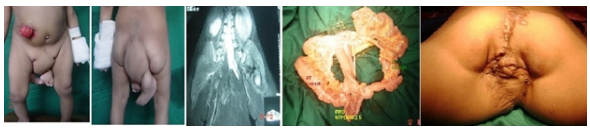
This is a retrospective descriptive study. The cases files of all children who diagnosed as colorectal duplication between 2004 and 2016 were reviewed. A total of 7 cases of colorectal duplications, aged between 8mths to 14 years were identified and evaluated. There were 4 boys and 3 girls. Two girls presented with rectoperineal fistula along with a normal anus. One girl had caudal duplication with two introituses placed coronally discharging the stools and urine. Two infants presented with intractable constipation were found to have cystic rectal duplication, one four years old boy with abdominal pain and mass and cystic transverse colon duplication and fourth boy is five years with 'Y'

Table 1: Summary of the cases.

Age/ sex	Clinical presentation	Working diagnosis	Intraoperative anatomy	Surgical Procedure	Complications	Outcome/ Follow period
8m/F	Passing stools from the introitus and from the normal anus	Rectovaginal fistula	Complete duplication of colon with Separate mesentery	Excision of duplicated colon	None	Good 18 months
1y/F	A bifid spine, central mass with rudimentary feet with duplicated intoituses and vestibular anii, two urethra	Caudal Duplication	Complete colon duplication with 10 cm of duplicated ileum, two appendices separate mesentery.	Excision of duplicated right colon	Left colon underwent delayed torsion, was resected and ileoanal anastomosis done	Not satisfactory 8 years
5y/M	Colostomised ARM with passage of stools from the urethra.	High Anorectal Malformation	'Y' duplication sigmoid colon with double termination in posterior urethra	Excision of Duplicated Sigmoid colon and Pull through of the Descending Colon	None	Fair Incontinent for stools 2 years
14y/ F	Passing stools from the introitus	Rectovestibular Fistula with Normal Anus	Duplication of Complete colon distal ileum, & appendices. Single mesentery with common blood supply	Fenestration of common wall and extirpation of residual mucosa	None	Good 3 years
1y/M	Intractable constipation	Anterior sacral Meningocele	Retrorectal duplication cyst shared common wall with rectum	Posterior saggital approach and complete excision of cyst	None	Good 1 year
2y/M	Intractable constipation	Cystic Rectal Duplication	Retrorectal cyst shared common wall with rectum	Laparotomy and excision and extirpation of residual mucosa	none	Good 2 years
4y/M	Abdominal pain and epigastric Mass	Duplication cyst of transverse colon	Cystic duplication of Transverse colon	Excision of the cyst with the adherent colon	None	Good 2 years

duplication of sigmoid colon and high anorectal malformation. [table1]

A twelve months old girl diagnosed as caudal duplication presented with a large mass with two rudimentary feet complete with toes and finger nails between lower limbs. [Fig 1, 2]The child had two introituses through which she passed urine and stools from each of them. The right introitus had one urethra and one vestibular anus discharging the right duplicated colon, while the left introitus had one urethra, a hemi vagina and a vestibular anus discharging left duplicated colons. Contrast enema showed a completely duplicated colon and cystogram should two urinary bladders draining their respective kidneys. Magnetic Resonance Imaging (MRI) confirmed the presence of complete duplication of spine, colon, urinary bladder and presence of left hemivagina and left hemi uterus. [Fig 3] The two colons had separate mesentery with their own vascular supply originating from a single midline superior mesenteric artery and they were mobile freely in the abdomen [fig 4]. The right colon was excised completely. The midline mass was excised. Child under went multiple procedures for pelvic alignment. Four months after the colon excision the child underwent emergency laparotomy where in the entire left colon underwent torsion and necrosed. The surviving ileum was anastomosed to the left rectal stump. The child is fully continent for urine and stools. The child is on regular follow up and needs regular support to manage the liquid stools [Fig 5].

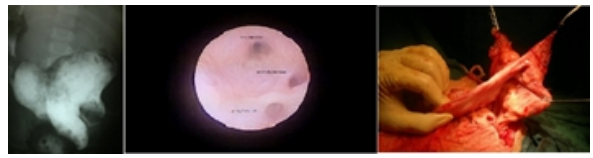


Figures 1-5: Caudal duplication two introituses discharging urine and stools, duplicated colons, bifid spine and post-operative status.

The second case, MRI revealed complete tubular duplication of colon with double termination along with the duplicated urinary bladder and urethra. One arm of the duplicated colon opened in between the two hemi vagina and the other arm open into the normal anal canal. The duplicated colon terminating into the introitus was completely excised. The mucosa from the residual fistula was extirpated completely from the perineum and defect was closed. The mobile orthotopic colon was fixed to posterior abdominal wall using non absorbable sutures. One year of follow up she symptoms free.

The third case is a thirteen years old girl's contrast study and MRI showed a complete duplication of the colon and distal ileum. At laparotomy, smaller caliber duplicated colon was loaded with hard inspissated stools, hence Ileostomy was done and the child was put up on distal stoma washouts. Six months later two fenestrations were created one at the cecum level and the second at the distal sigmoid and the rectum level. The mucosa of the distal duplicated colon was extirpated completely before closing the muscular defect. The ileostomy was closed after six weeks. At two years of follow up the child in symptom.

A five years old boy who referred to us with a stenosed and retracted colostomy done in neonatal period for high anorectal malformation[ARM]. The child passed small quantity of stools from the stenosed colostomy and passed good quantity of stools from per urethra, due to recurrent urinary tract infections both the kidneys were scarred and he had elevated renal parameters. Ultrasonography (USG) of abdomen showed bilateral echogenic kidneys. Dimercaptosuccnic Acid (DMSA) scan showed poorly functioning bilateral renal parenchyma with multiple photopenic areas. MRI showed multiple hemi-vertebrae in the lumbar region and an incompletely formed sacrum. The pelvic musculature was poorly developed. Contrast study and Voiding Cystourethrogram (VCUG) was done in the same sitting revealed 'Y' duplication of sigmoid colon. The rectum was separate and was dorsal to the hugely dilated bowel. [Fig 6]The cystoscopy which showed two ectopic openings in the posterior urethra. The cranial opening in the prostatic urethra opened into hugely dilated bowel and the distal opening opened into the normal sized rectum containing stools [fig7]. The 'Y' duplication of sigmoid colon was excised and a descending end colostomy was done [fig 8]. Two months later child underwent abdomen assisted Posterior Saggital Anorectoplasty (PSARP) and the descending colon was brought through the poorly developed sphincter muscle complex. Child at one year follow up has poor continence for stools. There no urinary tract related issues. Malone's Aantegrade Continent Enema is planned for the child's stool incontinence.



Figures 6-8: contrast study from stoma, cystoscopy showing double termination, intraoperative 'Y' duplicated colons.

Two infants one and two years were investigated for intractable constipation. A space occupying retrorectal lesion was seen on contrast enema in both cases. [fig 9] MRI confirmed the rectal duplication cyst. Both underwent successful excision of cyst and complete extirpation of mucosa of the common wall. The raw surface was fixed to presacral area. The raw surface was fixed to presacral space. On follow up the both children is completely free of symptoms.

A four years old boy was diagnosed as enteric duplication cyst shared a common wall with the transverse colon, the cyst was excised along with the part of colon. Histology confirmed the duplication cyst. None of the cysts in our series had ectopic mucosa.

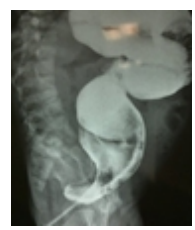


Figure 9: Contrast study showing compression of rectum by duplication cyst.

Results

The cystic variety of colon duplication was common in boys (3 out of 4) while complete tubular duplication of colon was common in girls (3 out of 4) and one boy had 'Y' duplication of sigmoid colon. Six out of the seven children had other organ system anomalies [chart1]. The girl who has caudal duplication had major catastrophe due to torsion of the colon. She had frequent issues with liquid stools and perianal excoriation. Her outcome was unsatisfactory. She has a waddling gait. The boy with 'Y' duplication of colon has incontinence for stool. He is free from urinary tract infections. All other patients are symptoms free in the follow up.

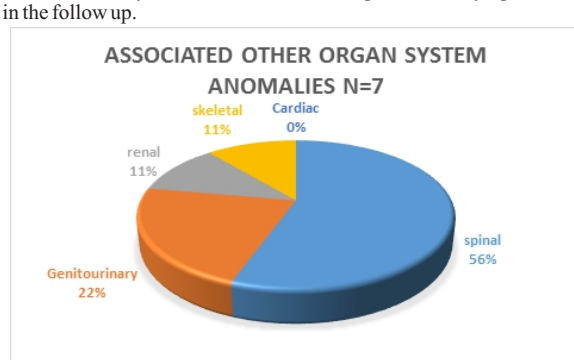


Figure 10: Pie diagram showing associated other system anomalies.

Discussion:

Gastrointestinal (GI) Duplications are uncommon anomalies, detected in 1 of every 4,500 autopsies. In one of the largest reported of children with 101 gastrointestinal (GI) Duplications [2], the commonest site of GI duplication is the ileum (30%), followed by the esophageal(30%), jejunum (8%), colon (6%–7%), and rectum (5%) [3]. The anatomical subtypes of GI duplications are cystic type (90%) and the tubular type (10%). The tubular type includes the double-barreled type (80%) and the Y- or T-shaped type (20%) [4].

Hindgut duplications account for 8-13% of all duplications [5]. Four varieties of hindgut duplications have been described viz, appendiceal,

cystic colorectal duplication, tubular colorectal duplication and anal canal duplication[5,6,7] Colorectal tubular duplications is further classified into five types: type 1, two separate perineal anii; type 2, female with a recto-vaginal, recto-vulvar or recto-perineal fistula; type 3, male with recto-vesicle or recto-urethral fistula; type 4, duplication with imperforate anus of either or both the tubular duplication and native rectum; type 5, single anus with communicating duplication proximally and distally, mostly seen in females[5].Both the female tubular duplication of colon in our series are type 2 and the 'Y' duplication of the male Anorectal Malformation falls into type 4, despite extensive literature search we did not find a case of tubular duplication where both duplicated colon terminate in the posterior urethra.

The proposed embryonic theory to explain the association of the tubular duplication of the colon and the spinal anomalies is split notochord syndrome by Bentley [8]. According to this the theory nueroenteric canal connecting the yolk sac and the amnion is responsible for the associated spinal defects [9]. During development of the split notochord, an endodermal-ectodermal adhesion between the cord leads to the persistence of the endomesenchymal tract between the amnion and the yolk sac. This endomesenchymal tract formed is responsible for the duplication of a part or of the entire gastrointestinal tract.[1]

Colorectal duplications are twice as common in girls [5, 8]. They are frequently associated with anomalies in other systems. About 80% have had anomalies in at least one other system, and 55% in three or more [10]. In our series has five out of seven had multiple organ system involvement (85%), none of our cases had cardiovascular involvement [Chart 1]. The clinical presentations are related to the location and size of the duplication, the anatomical type such as cystic or tubular type and the presence of ectopic mucosa. [11]. Cystic duplications may present with chronic pain, mass in abdomen, acute bleeding and rarely malignant change. Rectal duplications may cause mass effect on the rectum causing constipation, they may prolapse through anus or present as chronic anal fistula [12,13].

The tubular duplicated colon may be a blind ending or may end as an additional opening in the perineum. Associated anomalies may be two anii, and a combination of functioning and an imperforated anus. The fistula may open into posterior urethra in males [14]. They may be in midline or paramedian fistulae. In girls the complete tubular colon duplication may present with two functioning anii, or a presence of rectovulval, rectovestibular or rectovaginal fistula in addition to a normally positioned functioning anus. Tubular duplication of colon are often associated genitourinary duplications, skeletal anomalies, bladder extrophy, omphalocele, and spinal dysraphisims. [15]

The child should be adequately investigated, a contrast study done from both the perineal orifices help in delineating the colorectal anatomy. Ultrasonography has a sensitivity of 55% in diagnosing duplications of intestine. Computed tomogram (CT scan) with contrast may delineate the anatomy of the colon and genitourinary system. Spine and pelvic musculature are better delineated by Magnetic Resonance. Colonoscopy, Cystoscopy and Cystovaginoscopy helps to localize the ectopic opening of the duplicated colon.

The cystic colon duplications have variable presentation. They should be excised completely for the fear of bleeding and malignant potential. It often involves excision of a part of native colon along with the cyst due to shared common wall and blood supply. In rectal cystic duplication the mucosa of the common wall should be extirpated as complete excision of cyst is fraught with difficulties. Tubular colon may be asymptomatic and only present as cosmetic issue of duplicated anus, vagina and of penis. The duplicated colon may need no further surgical management if the internal communication is adequate [16]. However the majority of the authors recommend by excision of the duplicated colon and anus. We excised the duplicated colon completely in two cases. The duplicated colon had separate mesentery with own blood supply from midline superior mesenteric artery. The mesentery and the colon are extremely mobile and has a propensity to undergo torsion. One of our child with caudal duplication syndrome developed torsion after excision of the duplicated colon. We recommend fixation of the retained colon to the retro peritoneum.

In one girl, complete excision of the duplicated colon and a part of ileum was impossible due to the shared blood supply. We created two large fenestrations between the two arms of duplicated bowel, one at the level of cecum and the other at the sigmoid colon level.

The excision of the distal ventral colon is fraught with difficulties. Both arms of the duplicated colon traverse the same puborectalis muscle sling, and any attempt to excised one disrupts the continence mechanism of the normally sited anus. Stephan and Smith in their series of double perineal anii opined that, provided there is no neurogenic element and both recti appear to lie in the same pubore ctalis muscle sling and within a single external sphincter, it may be best to accept the two anii permanently. Attempt to excise of rectum from the other may jeopardize the normal continence. [17] We extirpated the mucosa of the distal segment from within the lumen of the bowel. This prevented damage to the sphincter muscle complex and simultaneously we could address the opening in the vestibule.

Tubular Duplication of Colon with rectourethral fistula in Anorectal malformation is exceedingly rare [14]. The case with 'Y' tubular duplication of the sigmoid colon, the dilated duplicated colon opened into the prostatic urethra proximally while the normal caliber colon opened distally. Despite our extensive search we did not find and similar case where both the duplicated colon arms terminated into the posterior urethra. Multiple hemivertebrae, incompletely formed sacrum and poorly developed sphincter muscle complex contributed towards the child's incontinence.

Conclusion

Duplications of colon and rectum are rare clinical entities. The treatment of the duplications should be tailored for individual case. The association of colon duplication with anorectal malformation is exceedingly rare in male. The pathological anatomy should be carefully delineated before undertaking definitive repair of the anorectal malformation. The associated spinal and genitourinary malformation may affect the final outcome of the repair.

Complete excision of the tubular duplication is possible in cases where there is separate blood supply and the colon and its mesentery should be fixed to retroperitonium. In cases where there is shared blood supply, limited fenestration of the common wall and endoluminal mucosectomy of the distal duplicated colon restores the normal anatomy while preserving the continence mechanism.

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