Appendicular Duplication – Variations in Anatomy and Associations

Asjad Karim Bakhteyar[1], Ramjee Prasad [1], Digamber Chaubey[1], Ramdhani Yadav[1], Pranay Kumar[1], Saurav[1], Sandip Kumar Rahul[1*]

[1] Department of Pediatric Surgery, Indira Gandhi Institute of Medical Sciences, Patna

***Address for Corresponder:** Dr Sandip Kumar Rahul, Department of Pediatric Surgery, Indira Gandhi Institute of Medical Sciences, Patna (email: **sandeep.rahul65@gmail.com**)

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Abstract

Introduction: Although Duplication anomalies are common in the Gastro-intestinal tract, appendicular anomalies are rare with variable anatomy. we describe a series of appendicular duplication cases managed at a tertiary care center.

Materials and Methods: Data regarding the clinical features, associated anomalies and management of cases of appendicular duplication at a tertiary care center from January, 2019 to December, 2020 were collected retrospectively and analyzed.

Results: Four children with appendicular duplication were managed during this period; three neonates presented with high anorectal malformation and type two pouch colons with a large colovesical fistula. They had a single caecum with two separate appendices symmetrically on either side (type B1).

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They were managed by division of colo-vesical fistula, mobilization of colonic pouch after limited pouchoplasty and anoplasty as a single stage procedure. None of the appendix was removed. In one of these cases, bilateral ureters were dilated. The fourth case presented as a 3-year-old with pain abdomen and during surgery for suspected appendicitis, partial duplication of inflamed appendix was found (type A). Appendicectomy relieved this child of his symptoms. All patients are doing well on follow-up.

Conclusion: The position, anatomy and associated anomalies of appendicular duplication can be variable complicating its presentation and management.

Keywords

- anorectal malformation
- appendix
- duplication
- pouch colon

Introduction

Gastrointestinal (GI) duplications are common anomalies in pediatric surgical practice with a wide range of presentation depending not only on the site and size of the lesion but also on the inherent qualities like presence of ectopic mucosa and associated anomalies. By contrast, duplication of the Vermiform appendix is extremely rare with an incidence of 0.004% to 0.008% in different appendicectomy series.[1- 3] Appendicular duplication (AD) itself is heterogeneous with different types and several associated anomalies of the GI tract, genito-urinary system and vertebral column. Modified Cave-Wallbridge classification describes the anatomical relation of the duplicated appendices with each other and with the Caecum.[4] Several other anomalies of Appendix other than AD have been described which include agenesis, triplication, anomalously located solitary appendix and the more recently described horseshoe appendix.[5- 7] Most often these lesions are encountered during operative procedure for some other pathology and sometimes due to involvement of appendix itself (like appendicitis, perforation, and tumor). This study summarizes the presentation, clinical feature and proper management of AD and

associated anomalies managed at our center.

Materials and Methods

A retrospective, descriptive, observational study was conducted on all patients of AD managed in the department of Pediatric surgery at a tertiary center after approval from the Institutional Ethics Committee with approval letter No (1694/IEC/2020/IGIMS). All patients of AD found at the time of surgery, when surgery was done for some related symptoms or for appendicitis were included in this study. The study period was from January 2019 to December 2020 (2years). Consent of the parents was taken for inclusion of the clinical details and pictures of their children in this study and its presentation. The clinical records including intra-operative findings of all cases of AD were collected to get detailed history, demographic details (age, sex, and residence), presenting symptoms, examination findings, investigation results (blood and radiological) and intra-operative findings, histopathological examination (HPE) results (if any), and follow-up details. Collected data was analyzed.

Result

Four patients of appendicular duplication were managed during this study period**.** Age of the patients varied from one day to three years. All the patients of AD associated with anorectal malformation (ARM) presented as neonates, while the one presenting with Appendicitis presented at the age of three years. All patients were males. **(Table 1)** summarizes the clinical features, type, associated anomalies and management of these patients.

Table 1: Clinical features, type and management of AD found in the study

S.no	AD type*	Anatomy& associated anomalies	Clinical features	Management
1	B1	2 separate appendix symmetric on either side of a single caecum; High ARM with type 2 Pouch colon & poucho-vesical fistula	found intraoperatively in a neonate being operated for high ARM with pouch colon	division of the Poucho-vesical fistula+mobilization of the pouch and completion of anoplasty (both appendix preserved as such)
2	B1	2 separate appendix symmetric on either side of a single caecum; High ARM with type 2 Pouch colon & poucho-vesical fistula with sacral deformity	found intraoperatively in a neonate being operated for high ARM with pouch colon	division of the Poucho-vesical fistula+mobilization of the pouch after pouchoplasty and completion of anoplasty (both appendix preserved as such)
3	B1	2 separate appendix symmetric on either side of a single caecum; High ARM with type 2 Pouch colon & poucho-vesical fistula; bilateral ureters dilated	found intraoperatively in a neonate being operated for high ARM with pouch colon	division of the Poucho-vesical fistula+mobilization of the pouch after pouchoplasty and completion of anoplasty (both appendix preserved as such); patient managed for neurogenic bladder in follow-up period
4	A	partial duplication of appendix on a single caecum	Presented with appendicitis; found intraoperatively	Appendicectomy done

The presenting symptoms varied depending on the associated anomalies in three and features of appendicitis in one patient. Three neonates presented with high anorectal malformation (ARM) and type two congenital pouch colons (CPC) with large poucho-vesical fistula. On preoperative imaging, a large pouch occupying more than half of the abdominal diameter could be appreciated in all the three cases suggesting the presence of pouch colon **(Figure 1).** They had a single caecum with two separate appendices symmetrically on either side (type B1) **(Figures 2 and 3).** They were managed by division of the colo-vesical fistula, mobilization of colonic pouch after limited pouchoplasty and anoplasty as a single stage procedure. None of the appendix was removed. In one of these cases, bilateral ureters were dilated. In the post-operative period, although the dilatation lessened, on follow-ups, this child showed evidence of neurogenic bladder and was managed for the same.

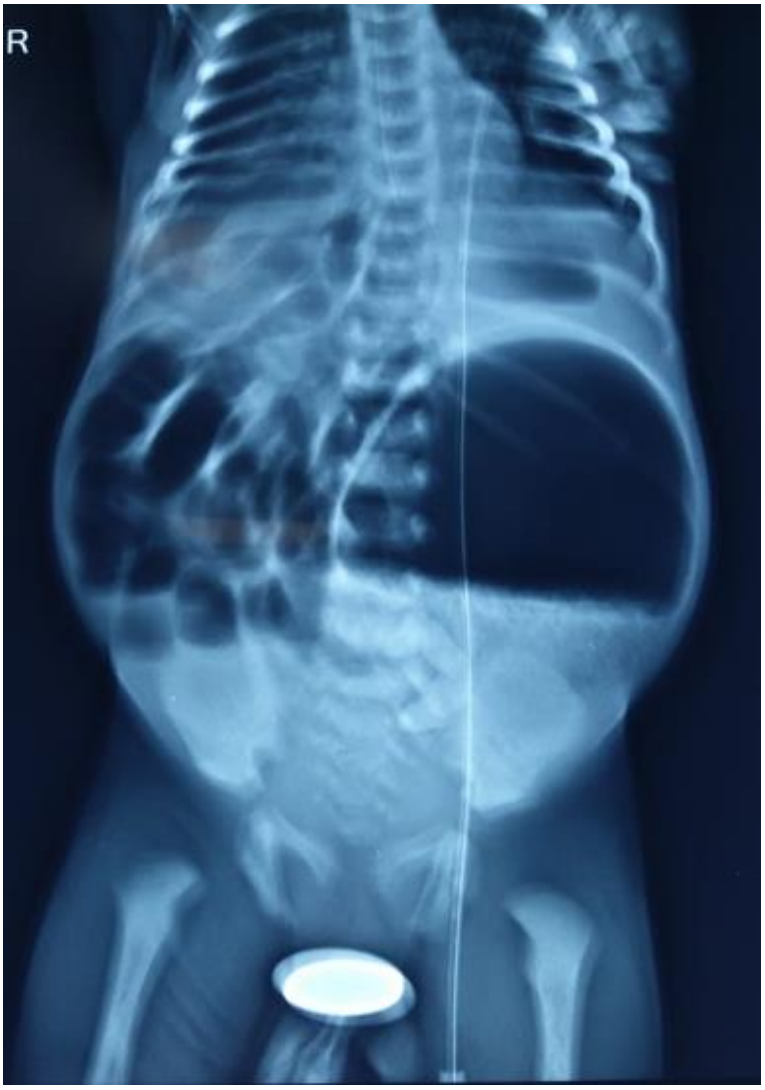

Figure 1: X-ray abdomen & Pelvis (erect) showing a large pouch colon occupying more than half of abdomen

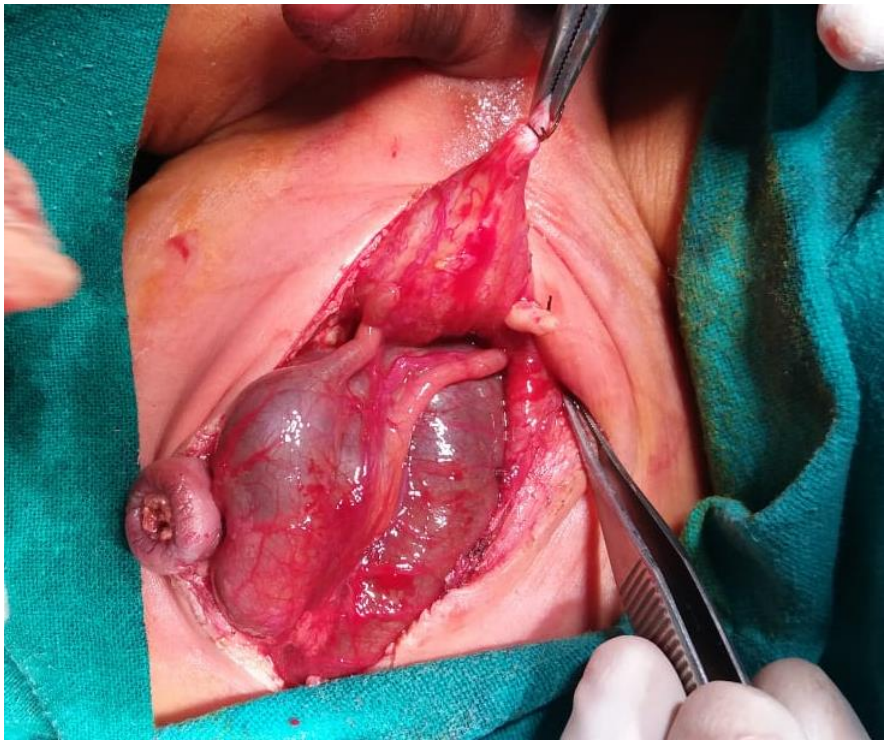

Figure 2: Caecum with large type 2 pouch colon with double symmetric appendices (Type B1)

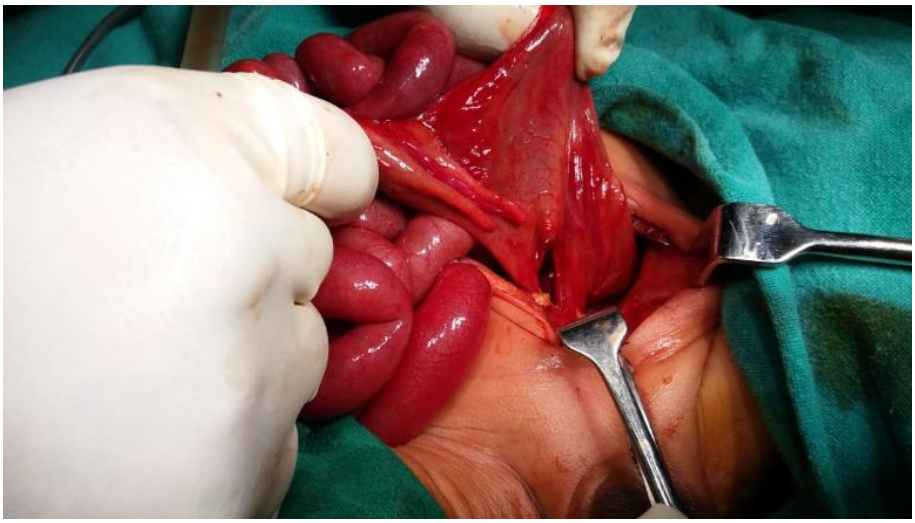

Figure 3: Pouch colon ending as a fistula to bladder and two symmetric appendices on a single Caecum (Type B1)

The fourth case presented as a 3-year-old male with migrating right lower abdominal pain (typical of Acute Appendicitis) and during surgery for suspected appendicitis, partial duplication of inflamed appendix was found (type A). Appendicectomy relieved this child of his symptoms.

Of the three neonates, two children are around 1.5 years of age and one child is of 8 months now; all of them are doing well with bowel management program consisting of serial neoanal dilatation and daily rectal washes with normal saline and glycerine. Eight months child has evidence of neurogenic bladder with vesico-ureteric reflux and is being managed as per the neurogenic bladder management protocol with clean intermittent catherization (CIC) and chemoprophylaxis. Fourth patient of acute appendicitis is four years old now and is asymptomatic. All patients are doing well on follow-up.

Discussion

Duplication of the GI tract themselves represent a heterogeneous group of anomalies with variable presentation, often associated with other anomalies.[8] Although rare with an incidence of around 0.004%, AD is notorious for variations in types so much so that a separate classification system has been proposed to address its different anatomical variations.[2, 9] Several associated anomalies involving the gastrointestinal, genito-urinary or vertebral tracts have been described.[11-13]

Appendix first appears during the 8th week of gestation as an out-pouching of the caecum and elongates thereafter with gestation. However, the embryology of AD and its variations is less well understood.[14] Cave proposed two theories to explain the embryogenesis of appendicular duplication: (i) the persistence of a transient embryological structure and (ii) incidental appendicular duplicity due to a more general affection of the primitive midgut.[15] In Modified Cave-Wallbridge classification further modified by Cocker et al, appendicular duplication has been divided into 4 categories **(Table 2)**.

Table 2: Classification of Appendicular Duplication

Type	Features
A	A single caecum with various degrees of partial duplication
B	
1	"**Bird type**" in which the two appendices are symmetrically placed on either side of the ileocaecal valve
2	"**Taenia-coli type**" in which one appendix arises from the caecum at the usual site and a second appendix branches from the caecum along the lines of the taenia at various distances from the first.
3	First appendix is at normal site and second appendix arises from the hepatic flexure
4	First appendix is at normal site and second appendix arises from the splenic flexure
C	A double cecum each with an appendix
D	**"Horse shoe appendix"** single caecum with two separate opening of single appendix into same single caecum
Cave-Wallbridge classification of appendicular duplication, modified by cocker et al[9]	

There may be various presentations of these duplex appendices. In neonatal stage, it presents in association with other congenital anomalies but in older age group it may present as appendicitis, perforation leading to peritonitis or mass. It may remain asymptomatic throughout life in many patients.

Of the four patients in this study, three neonates presented with high anorectal malformation and type two pouch colon with a large poucho-vesical fistula. They had a single caecum with two separate appendices symmetrically on either side (type B1). The caecum was normal and two appendices were present. Both appendices

were well formed, separate and symmetrical to the ileo-caecal valve with their own separate meso-appendices. This anatomy mimics the anatomy found in birds and is therefore called the 'bird type' or 'avian type'. Type B1 and C is known to be associated with various congenital anomalies while type B2 duplication is not known to be associated with any other congenital anomalies .[16] Several investigators have found the co-existence of Appendicular duplication with type 2-CPC. [17]

Congenital pouch colon (CPC) is an unusual anomaly in which a pouch-like dilatation of a shortened colon is associated with an anorectal malformation.[18] The pouch usually terminates in a fistulous communication with the genitourinary tract. All the three neonates presented here in our study had poucho-vesical fistula. They were managed by division of the poucho-vesical fistula, mobilization of colonic pouch after limited pouchoplasty and anoplasty as a single stage procedure. CPC is of four subtypes (Types I–IV) based on the length of normal colon proximal to the colonic pouch. [18] Saxena and Mathur et. al. added a type V pouch colon to this classification.[19] Several investigators have reported type 2 pouch colons in association to AD. [17, 20]

CPC seen in many patients of high ARM itself accompanies malformations/pathologies like those involving the genitourinary system (hydroureteronephrosis, Vesico-ureteric reflux, renal agenesis, Undescended testes, Hypospadias), esophageal atresia, congenital heart disease, sacral agenesis.[21] Out of our four cases, we had bilateral dilated ureter in one case.

Reported cases of appendicitis in patients with AD are rare in the literature, however, it is most likely that second appendix may be missed simply because it was never identified. Retrocaecal appendices and type B duplications are most likely to remain unnoticed.[16]Failure to recognize this condition may have serious clinical and medico-legal consequences, if sister appendix is not removed.[22] It is therefore necessary to be aware of the different anatomical variations of AD. We removed the partially duplicate appendix in the three years-old patient with features of acute appendicitis; on the contrary, the type B1 appendices in all the three neonates were left as such because the already deficient blood supply in type 2 pouches would

further deteriorate in an attempt to ligate the two mesoappendices. Should appendicitis occur in the future in these patients, they can be diagnosed using an ultrasound keeping in mind the initial deformity and surgery can be performed if needed.

Patient with type B1-AD having type 2-CPC and bilateral hydroureteronephrosis also had features of neurogenic bladder and was managed initially on CIC and antibiotic prophylaxis. Patient with associated sacral deformity was also given guarded prognosis regarding future continence of stool.

Conclusion

The position, anatomy and associated anomalies of appendicular duplication can be variable; it may be associated with various other congenital anomalies complicating its presentation and management. Both appendices should be removed at the time of appendicitis otherwise it may have serious clinical and medico-legal consequences. If not removed, detailed abnormal anatomy should be clearly highlighted in the postoperative notes.

Limitations of the study

Given the rarity of the condition, a study with larger number of patients or of longer duration would help in better understanding and bring out a stronger evidence regarding the management of this rare anomaly.

Ethical Consideration

This study was approved by Indira Gandhi Institute of Medical Sciences, Sheikhpura Patna-14 with the code number: 1694/IEC/2020/IGIMS.

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Conflict of interests

There is no conflict of interest

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