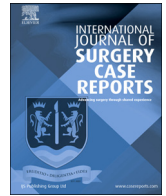




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A case report of complete appendiceal duplication on the normal site of a single caecum: A new variant?

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ABSTRACT

INTRODUCTION: Duplicated appendix is an uncommon entity, typically discovered as an incidental finding during surgery for appendicitis or other abdominal pathologies. It may be associated with other congenital malformations. We report a case of a male neonate incidentally discovered to have an unrecognized variant of duplicated appendix during a laparotomy plus diversion colostomy for imperforate anus at 4 days of age.

PRESENTATION OF CASE: A baby delivered at home from an unbooked pregnancy at term, was referred from a primary care clinic to a specialist referral hospital, with a fever and suspected neonatal sepsis on day 1 of life. The patient had not passed meconium and physical examination revealed an imperforate anus. After initiating treatment for sepsis, the patient underwent a laparotomy where a situs inversus totalis and complete appendiceal duplication was found, with both appendices on the normal site of a single caecum. The appendices were left in situ and a diversion colostomy was performed. The patient did well following surgery and was discharged on postoperative day 10 to await definitive surgery.

DISCUSSION: Appendiceal malformations have been reported either in isolation or in association with other congenital anomalies. Duplicated appendix occurs rarely and the pathogenesis is not fully understood. This case adds more evidence that the classification of appendiceal abnormalities should continue evolving as newer types are described.

CONCLUSION: Surgeons operating on patients with congenital anomalies must exercise extreme vigilance to identify and document other rare pathologies that may later pose challenges thus avoid morbidity, mortality and potential medicolegal pitfalls.

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1. Introduction

Duplication of the appendix is uncommon and is typically discovered as an incidental finding during surgery for appendicitis or other abdominal pathology [1]. In the neonates, it may be associated with gastrointestinal and/or genito-urinary anomalies such as imperforate anus [2]. Sometimes a missed duplicated appendix in a patient with an appendectomy becomes inflamed confounding the clinicians as to the plausible cause and there are reported cases of litigation involving such missed cases [3]. Since the appendix is considered to be the most frequent cause of the acute abdomen, it becomes even more important to be wary of variations which may later confound the clinician and cause unnecessary morbidity and mortality to the patients [4]. We present a case of a 4 day old male neonate who underwent laparotomy plus diversion colostomy for imperforate anus and was incidentally discovered to have an unclassified type of duplex appendix and situs inversus totalis among other congenital anomalies. This work has been reported in line with the Surgical Case Report (SCARE) criteria [5].

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2. Case report

A 1-day-old baby boy delivered at home from an unbooked pregnancy, was referred from a local urban maternity clinic to our specialist referral institution with a pyrexia and provisional diagnosis of neonatal sepsis. His birth weight was 2830 g at term and the baby was noted to have cried at birth but no Apgar scores were given as the baby was born at home. On admission by the Paediatricians, the neonate was noted to have a fever of 38 °C, with a distended abdomen, no vomiting and no passage of meconium after hours of life. There was no family history of malformations,

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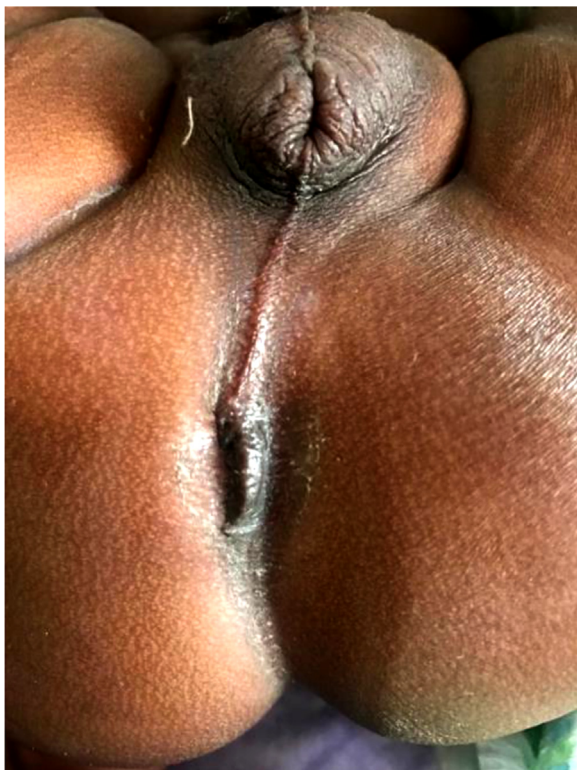


Fig. 1. Imperforate Anus.

consanguinity and no history of potentially teratogenic medication during pregnancy. No antenatal visits or ultrasound examination had been done since the pregnancy was unbooked. Physical examination revealed an abdominal distension and an imperforate anus. (Fig. 1) The patient was noted to be passing meconium stained urine however the appearance of the external male genitalia was normal. Routine complete blood count tests revealed an anaemia of 9.4 g/dl and a raised white cell count of 12250 cells/ μ l while biochemical parameters were all normal. Due to lack of radiological support at the time of presentation, no x-rays were done and a decision to

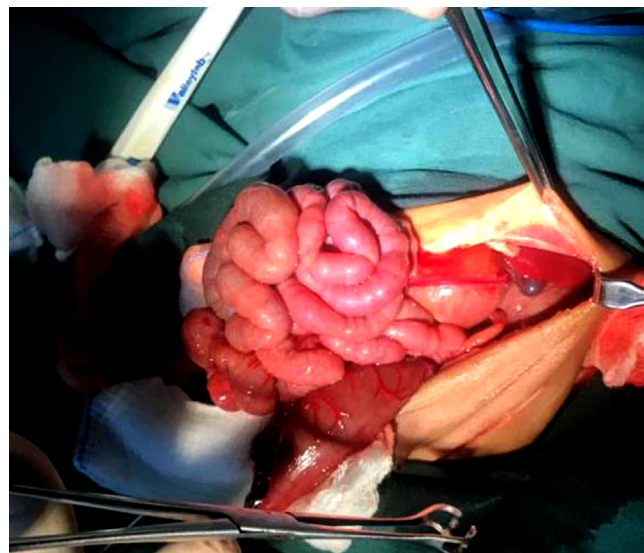


Fig. 2. Left sided liver and Gall Bladder.

proceed with operative management was taken. The patient was admitted into neonatal intensive care unit (NICU) and put on intravenous fluids, antibiotics and blood transfusion. The patient was kept nil per mouth with a nasogastric tube for drainage.

After optimising for surgery, the patient was taken to theatre on day 4 of life. Initially, a small abdominal incision had been done with the intention to fish out the colon for faecal diversion but this proved difficult necessitating conversion to a full transverse laparotomy. At laparotomy, the neonate was noted to have transposed terminal ileum on the left side, with the gallbladder and the bulk of the liver also on the patient's left (Fig. 2). There was a single mobile caecum associated with two appendices originating from a common base at the normal site (Fig. 3). Both appendices shared an appendicular artery arising from the ileocolic artery (see illustration in Fig. 5A2). The rest of the large intestine appeared shortened, and there was a vascular anomaly noted. No other obvious intra-

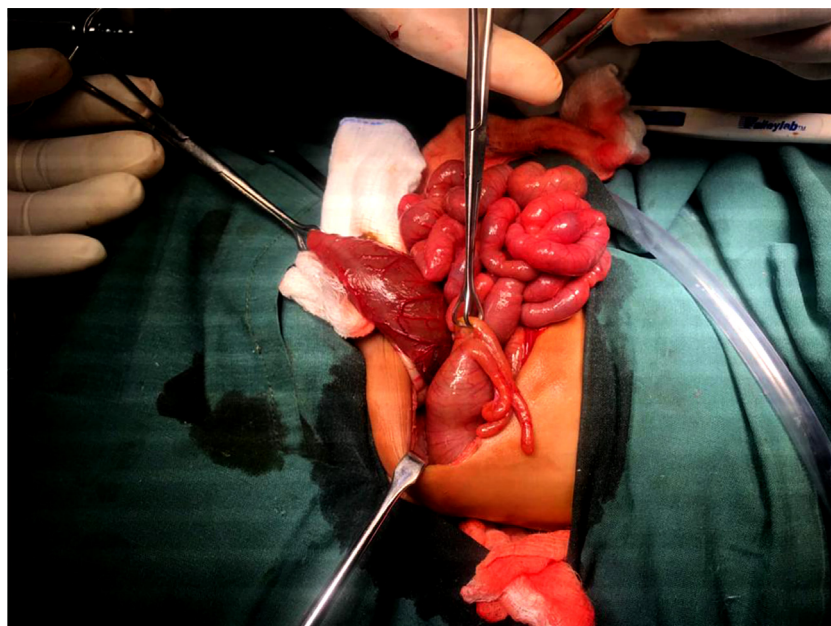


Fig. 3. Complete duplication of appendix on single caecum.



Fig. 4. Chest X-ray showing dextrocardia.

Table 1
Cave – Wallbridge Classification.

Type A	Single caecum and incomplete duplication
Type B1	Symmetric duplication at both sides of the ileocaecal valve
Type B2	Duplication, one normal and the other at a different localization
Type C	Duplication occurring with Caecum duplication

abdominal anomaly was noted. The appendices were left in situ and a double barrelled colostomy was fashioned in the right iliac fossa.

The patient was breastfed on day 2 after surgery and tolerated feeds with the stomas functioning well. He however developed a superficial surgical site infection due to faecal soiling of the suture line. The wound infection healed well with local wound dressings, and the patient was discharged home on postoperative day 10. The patient was reviewed as an outpatient and investigated for associated congenital anomalies. Radiographs (Fig. 4) and echocardiography confirmed a dextrocardia with an atrioseptal defect of 3.5 mm, affirming a diagnosis of situs inversus totalis, also noted were some vertebral anomalies. At the time of publication the patient was still awaiting radiological investigations to characterise the anatomy of the distal colon and the suspected recto-vesical fistula before definitive surgery.

3. Discussion

Many malformations of the appendix have been reported either in isolation or in association with other congenital anomalies [1,4,6]. Duplex appendix is a rare occurrence that is incidentally diagnosed during surgery for appendicitis or unrelated abdominal pathology and has a reported incidence of 0.004%–0.009% [7,8]. Similarly, Situs inversus totalis is also uncommon with a reported incidence of 0.1–0.6 per 10000 live births and can occur in association with other congenital anomalies [9]. For both conditions to occur in one patient as happened in our patient is extremely rare, to the best of our knowledge this is the first published case in English Literature.

The embryogenesis of the appendix is well known, however, the pathogenesis of duplication is not well documented although some theories have been put forward including the split notochord theory, median septum formation, normal regression of embryonic diverticula and the partial twinning procedure [10,11]. The classification mostly used to describe the duplex appendices is the Cave–Wallbridge Classification (Table 1), originally described by Cave in 1936 before modification by Wallbridge in 1962 to encompass other anatomic variations [12,13]. However, even the Wallbridge modification has subsequently been modified by sev-

Table 2
Classification of Appendiceal Abnormalities.

Number Anomalies	1. Agenesis: absence of appendix
	2. Duplex appendix (Cave Wallbridge Classification)
	A. *Partial duplication or complete duplication on the normal site of a single caecum.
	1. Partial duplication with both appendices sharing a common base like “Y-shaped” on a single caecum.
Shape anomalies	2. Complete duplication with both appendices side by side on normal site of single caecum.
	B. *Complete duplication of the appendix on an abnormal site of a single caecum
	1. Avian type or “bird-like appendix”: two appendices symmetrically placed on either side of the ileocaecal valve.
	2. Taenia coli caecum type: one appendix arising from the usual site of the caecum and the other arising from the caecum along the taenia
	3. Taenia coli hepatic flexure type: one appendix arising from the usual site of the caecum and the other arising from the hepatic flexure of the colon along the taenia
	4. Taenia coli splenic flexure type: one appendix arising from the usual site of the caecum and the other arising from the splenic flexure of the colon along the taenia
	C. Duplication of the caecum, each has its own appendix
	B2, B3, and B4 are not associated with other embryonic anomalies generally while B1 is found associated with intestinal and genitourinary anomalies
	3. Triplex appendix: complete triplication of appendix on the caecum
	Horseshoe appendix (also can be classified as Type 2D)
	Location of the mesentery
	• Sagittal disposal: the both bases of the appendix are along the taenia in sagittal direction
	Location of the orifice
	• Caecum-caecum
	• Caecum-ascending colon
	• Caecum-hepatic flexure of the colon
	• Frontal disposal: the bases of the appendix are not on the taenia
	Location of the orifice
	• Caecum-caecum

* The proposed changes to the classification. Table made based on these studies [11–17].

eral other authors as new variants of the anatomy were described including the horseshoe appendix, various shape anomalies and appendiceal agenesis [6,14–16]. Other modifications have also been done to include possible embryological aetiologies for the various anatomic variants [14].

In some of these publications there were records of cases that did not fit the classic classification systems [6,15–17]. This suggests that there probably will be more modifications to come and we believe that our case illustrates this point. As noted, our patient's two appendices (Fig. 1) originate abreast of each other at the normal point on a single caecum, so the case cannot be classified as either type A (Y-shaped) or any of B1 or B2. We therefore propose that this new variant be classified as type A2—where Type A1 is a Y shaped appendix and Type A2 is where both appendices are completely split from their bases but originate at the normal point in a single caecum (Fig. 5). Calota et al. had proposed a classification that included the Cave–Wallbridge classification as well as other anomalies not previously included [15]. Table 2 shows a suggested classification based on Calota's modification with our proposed variant factored in.

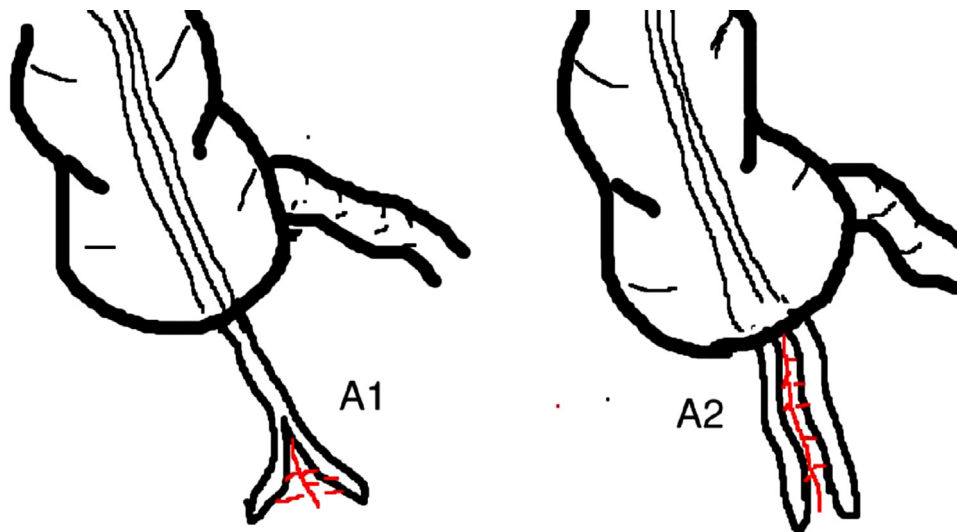


Fig. 5. Proposed change to type A Cave-Wallbridge appendix duplication.

Regardless of classification, failure to identify and document duplicated appendix, including its associated anomalies, may come back later to haunt the patient and clinicians as there is a risk of physical and medicolegal consequences respectively [3]. It is therefore important to search for other anomalies in the same sitting or afterwards where possible [18]. In patients with imperforate anus, surgeons at times perform a diversion colostomy through a limited incision precluding thorough examination of the viscera and thus may only discover the duplication during further reconstructive surgeries [2,18]. This highlights the need for vigilance when operating paediatric patients, to look out for previously undiagnosed congenital anomalies. Our patient had a situs inversus totalis which made it difficult to access the colon for diversion without an extended incision, thus we discovered the anomalies on the first sitting.

In a patient with duplicated appendix, when one appendix gets inflamed it is advised to also resect the uninvolved appendix. However, when duplication is discovered incidentally, appendicectomy need not be done, though documentation and patient education is required [19]. In our case both appendices were left in situ as they happen to be on the same site thus more easier to diagnose if inflamed.

4. Conclusion

Neonates with congenital anomalies such as imperforate anus may present with rare anomalies like duplicated appendix and situs inversus as part of a spectrum. All these anomalies may later pose a diagnostic challenge if thorough evaluation, documentation of associated abnormalities and counselling of the patients is not done. Duplicated appendix classification is evolving as more previously unclassified types are described and clinicians identifying a duplication need to carefully document their findings to improve the existing anatomical classification systems.

Declaration of Competing Interest

There is no conflict of interest.

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Ethical approval

Ethical approval for this study was exempted by our institution. Written informed consent was obtained from the parents of the patient for publication of this case report and accompanying images.

Consent

Written informed consent was obtained from the parents of the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor in Chief of this journal on request.

Author contribution

Allan Ngulube – Conceptualization, Methodology, Writing - Original draft, Visualisation, Project Administration.

Crispin Olivier Mambote Ntoto – Methodology, Visualisation, Supervision. Writing - Review & Editing.

Derek Matsika – Investigation, Writing - Original Draft.

Wedu Ndebele – Methodology, Writing - Review & Editing.

Ngqabutho Sanders Dube – Validation, Writing-Review & Editing.

Paradzai Gapu – Validation, Writing - Review & Editing.

All authors read and approved the final manuscript.

Registration of research studies

1. Name of the registry: UMIN Clinical trials registry
2. Unique identifying number or registration ID: UMIN000040517
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): <https://upload.umin.ac.jp/cgi-bin/ctr-e/ctr.view.cgi?recptno=R000046249>

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