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Recto-vestibular fistula with colonic duplication: report of three cases and review of literature

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Abstract

Background: Anorectal malformations associated with colonic duplications are a rare condition. Double terminations of tubular colonic duplication in the perineum are even rarer. Colonic duplication may have different presentations according to its location and size and incidence of less than 15% of all gastrointestinal duplications. We hereby report three such cases of recto-vestibular fistula along with underlying tubular colorectal duplication over a period of 10 years (2010–2020) managed successfully by different surgical approaches. These cases also illustrate the diagnostic challenge and variable presentation of colonic duplications in female patients with recto-vestibular fistula.

Case presentation: Case 1 is a patient with flank heteropagus with absent anal opening and recto-vestibular fistula, while cases 2 and 3 presented as suspected H-type recto-vestibular fistula with normal anal opening. All the cases were however managed with a specific surgical approach on case basis. Duplicated colon was excised in case 1, but in case 2 and case 3, duplicated colon was converted into single channel using proximal and distal staplers. Further definitive repair of female recto-vestibular fistula was done.

Conclusions: Colorectal duplication is a rare congenital malformation, and their association with anorectal malformations is even rarer. This association can have varied presentation, and surgical approach should be according to the congenital anatomical variations on case-to-case basis. Colonic duplications should always be ruled out in a female newborn child with history of passage of stools both from the vestibule and anus.

Keywords: Female anorectal malformations, H-type recto-vestibular fistula, Intestinal duplication, Colostomy

Background

Alimentary tract duplications are rare congenital malformations of the gastrointestinal tract, which may be cystic or tubular and occur anywhere from the mouth to the anus with an incidence of 1:4000–5000 [1]. More than 80% of cases are present before the age of 2 years as an acute abdomen or bowel obstruction [2, 3]. Colonic duplications are rare, accounting for 6–13% of all gastrointestinal duplications, commonly located in the cecum

[1, 4]. Although perineal canal or H-type congenital fistula is a relatively common anomaly in Asian countries, its association with Y-shaped tubular colonic duplication is uncommon [5].

Case presentation

We report three such cases that were incidentally detected with this infrequent association. All the three cases were associated with rare colonic duplication along with female anorectal malformation. They had varied clinical presentation on examination. Case 1 was a patient with flank heteropagus with absent anal opening and recto-vestibular fistula, while cases 2 and 3 presented as suspected H-type recto-vestibular fistula with normal anal opening. All cases were serially investigated and

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confirmed both on investigations and during surgery to be associated with colonic duplication. Initial colostomy was done in each patient. Further definitive surgical procedure was done specific to the case as shown in Figs. 1, 2 and 3.

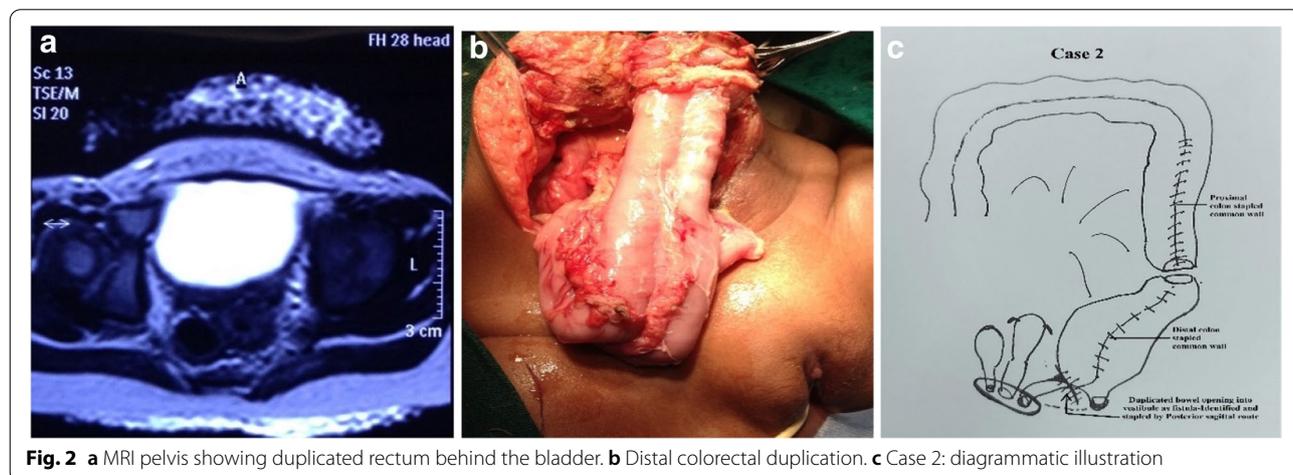
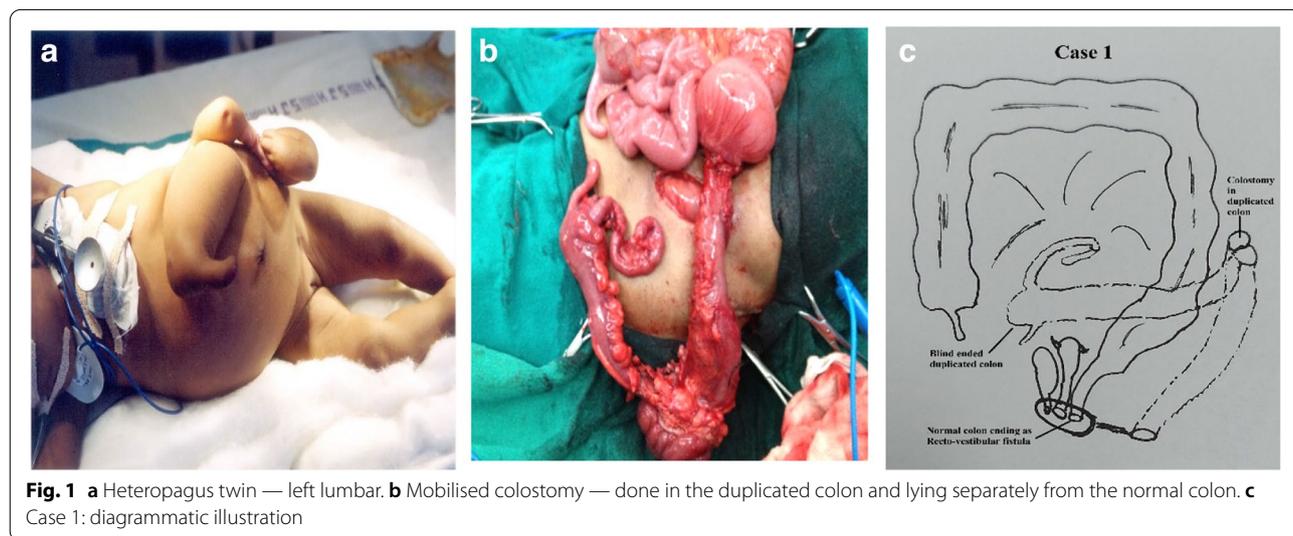
Case 1

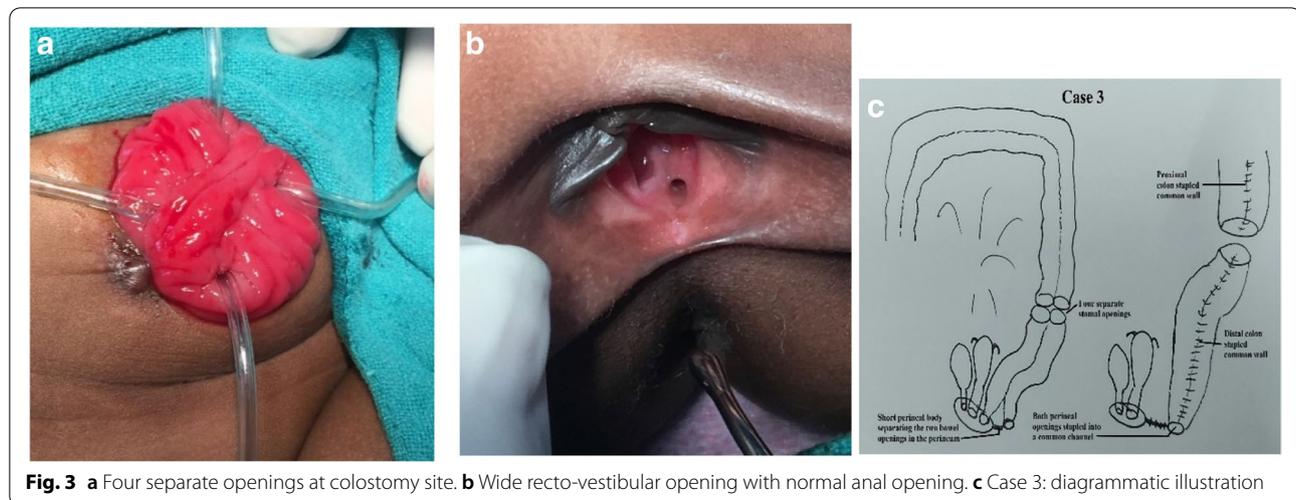
A newborn female child presented with flank heteropagus (Fig. 1a) with abdominal distension since birth along with recto-vestibular fistula. X-ray of the abdomen was suggestive of foetus in foetu. A diagnosis of foetus in foetu with recto-vestibular fistula was made. The child underwent colostomy in emergency at birth on day 2, but that did not function. USG of the abdomen was done, and it suspected an underlying intestinal duplication. Therefore, a redo surgery was planned, and on exploratory laparotomy, colostomy was found to be done on the duplicated colon that was lying separately from the

normal colon which was ending blindly with a short ileal segment (Fig. 1b). The functioning normal bowel was open as fistula in the vestibule with meconium drainage, while the duplicated bowel ended as the normal anal opening. Complete excision of the duplicated colon and heteropagus twin segment was done along with colostomy in the functioning sigmoid colon. Three months later, a posterior sagittal anorectoplasty (PSARP) was done for recto-vestibular correction. Diagrammatic representation of the case findings is shown in Fig. 1c.

Case 2

An 18-month-old female child presented with history of passage of stool per vaginal area and occasional stool per normally sited anus (H-type recto-vestibular fistula) since birth. The child had undergone colostomy initially at birth at a private hospital. She was then planned for examination under anaesthesia (EUA) and further PSARP. But





on EUA, an opening was noticed in the vagina which on probing went up endlessly up to the colostomy and not felt in finger kept simultaneously in the normal rectum. Therefore, further PSARP was deferred, and an abdominal exploration was done, which revealed that the whole of the colon both proximally and distally to the colostomy was duplicated with one end terminating at anus and the other end terminating as vestibular fistula (Fig. 2b). Distal cologram and MRI pelvis (Fig. 2a) were further suggestive of colorectal duplication. After 3 months, the child underwent definitive surgery with exploratory laparotomy and colostomy mobilisation. The proximal colonic duplication common wall was stapled cut by linear cutter 100 mm, and the distal colonic complete duplication common wall was stapled cut by linear cutter 75 mm. The normal rectum was mobilised by posterior sagittal route (through a sphincter-saving approach) to delineate the part of duplicated colorectum ending into the vestibule as fistula. A wide fistulous bowel entering into vestibule was then identified and divided using stapler from below, and mucosectomy of the distal residual mucosa of the stapled fistula part was done from the vestibular opening (Fig. 2c). The child recovered well, and colostomy closure was done after 3 months.

Case 3

A 1-year-old female child presented with passage of stools from both the vestibule and per anum (H-type recto-vestibular fistula). The perineal body separating the recto-vestibular fistula and anus was short and thin on examination. The child underwent sigmoid loop colostomy which showed duplicated colon extending both proximally and distally, and therefore, a colostomy was fashioned with four separate openings

(Fig. 3a and b). Distal cologram was suggestive of an adequate anus with faecolith present in colon ending into the vestibular orifice. The child had exploratory laparotomy and colostomy mobilisation after 4 months for definitive surgery. Proximal colonic duplication common wall was stapled cut by linear cutter 100 mm, and distal colonic complete duplication common wall was stapled cut by linear cutter 75 mm as in case 2. But in this case, the thin perineal body between the fistulous opening and anus was stapled with linear cutter 75 mm to form it as a part of anal canal from below, and this common channel was mobilised through posterior sagittal approach. PSARP was then completed with neo-anus placed in the muscle complex, and a good perineal body was formed with divided colostomy done at the same level (Fig. 3c). Colostomy closure was done 3 months later.

Hence, in this rare case series, case 1 underwent complete excision of duplicated colon along with heteropagus twin, and PSARP was done at a later date, while cases 2 and 3 had similar presentation with H-type recto-vestibular fistula along with colonic duplication, but their surgical approach during corrective surgery was different. In case 2, the fistula was stapled and separated from the duplicated colon during posterior sagittal approach (Fig. 2c). However, in case 3, both the fistula and the anus were stapled into a single common channel from below as the common wall between the distal openings was very thin (Fig. 3c). All three patients recovered well in postoperative period without complications. Cases 2 and 3 had some constipation initially but later recovered well with the passage of time. On follow-up of more than 5 years in all the three patients, anal continence became normal over time, and a near-normal perineal anatomy was also restored.

Discussion

Duplications of the colon are uncommon malformations, which results from early aberration in the formation of the primitive hindgut. It is hypothesised to cause a split or twinning process of the anlage, which results in terminal gut duplication with or without duplication of the genitourinary organs [6]. Tubular colonic duplications are double-barrelled or Y-shaped. They possess a double muscular layer and epithelium similar to the rest of the colon [1, 7]. Either both lumina may be unobstructed and function normally as two perineal ani or terminate distally blindly as imperforate anus of one or both lumina. In some cases, the ventral colon may end as a recto-urinary, recto-vaginal, or vestibular fistula [7].

The presenting features are constipation, vomiting, volvulus, perforation, and, most commonly, intestinal obstruction due to compression of the normal bowel by the blind end of the duplication [8]. Associated anomalies include genitourinary duplications, skeletal anomalies, bladder exstrophy, malrotation of gut, omphalocele, and Meckel's diverticulum [1, 7, 8]. In the absence of other associated malformations or an ectopic opening, tubular duplications of the colon remain unnoticed, until its complications warrant surgical intervention.

The need for surgery when asymptomatic is debatable [8]. In most cases, resection of the duplicated colon may not be possible because of common blood supply of the two colons [8]. The recommended surgical procedure is excision of the duplicated colon. Although malignant changes have been reported in adults [9], colorectal duplications are essentially benign lesions, and radical surgical excision is not required. In our case series, in cases 2 and 3, both the children were asymptomatic for the proximal colonic duplication, and the duplicated colon could not be resected without resection of almost the entire colon in view of the shared vascular supply, hence dividing the septum between the two lumens of the proximal and distal colonic segment along with further definitive surgical procedure as described above.

The options to manage unresectable proximal duplicated colons include dividing the septum to convert it into a common colonic channel; long side-to-side anastomosis of adjacent duplicated bowel, transection of the rectum over the peritoneal reflection, and anastomosis of the proximal end of the neighbouring duplicated colon to the main colon and excision of the distal common septum with creation of a common channel should prevent accumulation of faeces in the false lumen or performing mucosectomy on one limb of the proximal duplication [10]. Excision of the distal duplicated segment is even more difficult, as, although, all ectopic openings require surgical intervention, but it can further compromise the continence mechanism of the normal sited anus.

To prevent unexpected findings on surgical exploration, radiological evaluation is recommended. Contrast enema and a fistulogram performed concomitantly, preferably with contrast media of differing densities, help to delineate both colons. Occasionally, upper gastrointestinal contrast follow-through studies have also been reported to locate the proximal extent of duplication [10].

Management options for the distal duplicated segment include fistula closure with excision of distal few centimetres of septum to create a common distal channel and mucosal stripping of the duplicated colon like a soave's procedure. A mucosectomy and transection of the fistulating rectum below the peritoneal reflection can be performed transabdominally, or a posterior sagittal approach can be used to excise the fistula; this prevents mucus discharge per vagina from a residual rectal pouch [7, 10]. Stephens and Smith [11] presented a series of double perineal ani and opined that, provided that there is no neurogenic element, and both recta appear to lie within the same puborectalis muscle sling and within a single external sphincter; it may be best to accept the two ani permanently. An attempt to excise one rectum from the other may jeopardise normal continence. Therefore, for the distal segment, if the colon proper ends as a fistula, a posterior sagittal approach helps in appropriate delineation and transection of the fistula and also prevent compromise of continence mechanism [12].

In cases where hindgut duplication is discovered unexpectedly at initial laparotomy for stoma formation in anorectal malformations, sigmoidostomy has proven to be well tolerated as it does not compromise subsequent reconstruction and facilitates radiological evaluation of the complex anatomy [12]. Although the management of these cases is difficult, our case series will provide different surgical options to the surgeons to deal with these uncommon and difficult case scenarios.

Moreover, the perineal canal is a rare variety of anorectal malformations (3–4%) in which a fistulous tract is noted between the anterior wall of normally formed anal canal and the perineum and is identified by different nomenclatures like H fistula, double termination of the alimentary canal, and ano-vestibular fistula [13]. However, due to the rarity of the anomaly, different methods of management and surgical repair have been tried worldwide. The spectrum of anorectal malformations including perineal canal and H-type recto-vestibular fistula has been treated by surgical techniques such as vestibule-rectal pull through [14], perineal repair, posterior sagittal anorectoplasty, and anterior anorectoplasty [15]. However, their association with colonic duplication is a very rare occurrence and needs a definitive repair of the fistulous rectal opening into vestibule along with management of distal colonic duplication.

Conclusions

Tubular colorectal duplication should be considered as one of the differential diagnoses with perineal canal in cases of vestibular fistula along with a normal anus. One should always suspect and rule out colonic duplications in these patients in order to prevent on-table surprises. Proper examination and radiologic evaluation even before exploration for a colostomy through probing, fistulogram, or a contrast enema will help to delineate the anatomy in such cases, especially in obstructed cases. Colostomy at the initial laparotomy is useful for further radiographic evaluation and future reconstruction. A colostomy should be fashioned proximal to the duplication, if possible, to decide the best surgical option in the next stage and decrease number of operations and make them technically easier. Management option for the cases of H-type recto-vestibular fistula associated with colonic duplication should be individualised and on case-to-case basis.

Abbreviations

PSARP: Posterior sagittal anorectoplasty; EUA: Examination under anaesthesia; USG: Ultrasonography; MRI: Magnetic resonance imaging.

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Authors' contributions

RP — compiled the data, drafted the manuscript, and edited and assisted and followed up the cases. SKA — edited the manuscript and assisted the first author in study design and data compilation. DB — operated the cases and edited and concluded the study. The author(s) read and approved the final manuscript.

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Consent for publication

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