CASE REPORT

Outcomes of transanal endorectal pull-through for rectal atresia

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SUMMARY
Rectal atresia is a rare anorectal malformation, and it has been reported to represent 1%–2% of all anorectal malformations. We report three newborns who were admitted to the neonatal intensive care unit for abdominal distention, bilious vomiting and failure to pass meconium. The external anus and genitalia were normal and well formed. Digital rectal examination showed a blind-ending anal canal. All three infants were initially managed with diverting colostomy and then transanal resection of the rectal atresia with primary anastomosis, followed by colostomy closure. All patients eventually developed normal bowel habits and gained complete bowel control at 3–5 years of age, with mild constipation managed with laxatives. Contrast enema in a newborn with distal bowel obstruction is helpful to delineate the anatomy to show the gaps and to facilitate the procedure. In conclusion, transanal endorectal pull-through is a feasible and safe procedure with satisfactory clinical outcomes.

BACKGROUND
Rectal atresia is an extremely rare condition, and it represents 1%–2% of all congenital anorectal malformations.1 It usually presents with normal appearance of the anus and normal formation of the anal muscles which can make its diagnosis challenging. The anal canal and rectum are well developed, with an atretic segment between them. Typically, rectal atresia is diagnosed with failure to insert a catheter or thermometer more than 1–2 cm into the anal canal from the anal verge. Over 13 types of procedures have been reported for the management of this rare disease, and all of these approaches show variability in management, with good outcomes. Here, we describe our experience of three cases of rectal atresia managed with transanal endorectal pull-through.

CASE PRESENTATION
Case 1
The patient was a 34-week preterm male baby born through uneventful normal spontaneous vaginal delivery referred from another hospital. At the age of 2 days, he was transferred to the neonatal intensive care unit (NICU) because of abdominal distention, bilious vomiting, feeding intolerance and failure to pass meconium. On rectal examination, a normal anal opening was noted, and there was no meconium. He did not have cardiac or urinary malformations.

Investigation
On initial investigation, abdominal radiography showed dilated bowel loops with no gas in the distal bowel (figure 1). The patient was on bowel rest and antibiotics. Contrast enema showed failure of the contrast to pass more than 2 cm above the anal verge with no fistula (figure 2).

Treatment
Laparotomy was performed at the age of 3 days. The large bowel was massively dilated, and a rectal web was identified and opened through enterotomy in the rectum. Right transverse colostomy was performed. Postoperatively, distal-loop wash-out was performed once every week through the mucous fistula to avoid faecal accumulation and obstruction of the distal segment. Follow-up distal loopography revealed persistent distal obstruction (figure 3). Thus, transanal resection of the atretic rectum with primary anastomosis was performed at 7 months of age.

Outcome and follow-up
Follow-up distal loopography performed 2 months after the procedure confirmed continuity of the bowel loops.
distal colon without stricture (figure 4). Colostomy closure was performed at 1 year of age. The procedure for sequential dilations using Hegar’s dilators was instructed to the family in order to secure anal patency. As a tertiary care hospital, the patient was referred to his referring hospital after we followed him up to 4 years of age where he was then toilet trained with full bowel function and no soiling despite intermittent constipation managed with laxatives.

Case 2
The patient was a 36-week preterm male baby who was transferred to the NICU at the age of 3 days with progressive abdominal distention and failure to pass meconium. On clinical examination, he was clinically stable and well hydrated, and he had a hugely distended loopy abdomen that was soft and not tender. A rectal catheter could not be inserted more than 2 cm above the dentate line. However, the anus appeared normal. On echocardiography, he was diagnosed with a ventricular septal defect (VSD) which was managed at 2 months of age. No other congenital anomalies were noted.

Investigation
On initial investigation, abdominal radiography showed massively dilated bowel loops with no gas in the pelvis (figure 5). Contrast enema showed failure of the contrast to pass more than 2 cm above the anal verge (figure 6). Examination was performed under anaesthesia, and diverting colostomy was performed at the age of 4 days. During the colostomy, the sigmoid was hugely distended, and it appeared transparent and dusky in colour with signs of impending perforation.

Treatment
Transanal endorectal anastomosis was performed at the age of 6 months, and there were no intraoperative complications.

Outcome and follow-up
Colostomy closure was performed at 1 year of age. After closure, the patient was fine, and there was no clinical occurrence of obstruction. He gained complete bowel control at around 5 years of age with intermittent constipation that was controlled well with laxatives. He started to follow up with paediatric urology at the age of 1 year after he presented with a febrile urinary tract infection episode and found to have grade 2 vesicoureteral reflux bilaterally on renal ultrasound. On further follow-ups, he was diagnosed to have neurogenic bladder disorder with cystourethrogram test that showed dysfunctional posterior urethral valves. A spinal MRI was unremarkable for sacral agenesis. Then he was started on prophylactic antibiotics, clean intermittent bladder cauterisation (CIC) and anticholinergic medication. The last follow-up was at the age of 12 years, and the patient was off prophylactic antibiotics and CIC with good urination stream, and no episodes of incontinence during the day and night. A repeated cystourethrogram test showed regression
of vesicoureteral reflux and improvement of the neurogenic bladder. We still see the patient every 6 months to reassess the constipation.

Case 3
The patient was a full-term male baby born through spontaneous normal vaginal delivery, with a weight of 3.17 kg. He had congenital heart disease (dextrocardia, atrial septal defect, VSD, patent ductus arteriosus and bicuspid aortic valve), minor omphalocele, hypospadias and undescended testes. The omphalocele was repaired at the age of 1 day.

Investigation
On day 3 after the operation, the patient experienced distal bowel obstruction with dilated bowel loops and no gas in the rectum, and a rectal tube could not be inserted (figure 7).

On examination, the anus looked normal. Laparotomy with diverting colostomy was performed, and the diagnosis of rectal atresia was confirmed during laparotomy and examination under anaesthesia as a normal anal opening with obliteration 1 cm from the anal verge. Distal loopography performed 1 month after colostomy showed complete obstruction (figure 8).

Treatment
The patient underwent transanal repair of the rectal atresia at the age of 10 months. Postreconstruction distal loopography findings are presented in figure 9.

Outcome and follow-up
Colostomy closure was performed at 2 months after the repair. The patient gained complete bowel control at around 4 years of age, with intermittent constipation that was controlled well with laxatives. The patient is still following up with us every 6 months.

DISCUSSION
Rectal atresia is rare, and it represents 1%–2% of all anorectal malformations. The lumen of the rectum is interrupted either totally as in atresia (thin membrane or dense fibrous tissue) or partially as in rectal stenosis. The clinical presentation has been reported previously and includes abdominal distention and failure to pass meconium by the second day of life, with normal-looking perineum and genitalia and a normal anus, as in our cases. Other differential diagnoses that have a similar presentation include Hirschsprung’s disease, meconium plug syndrome, meconium ileus, small left colon syndrome, hypoganglionosis, neuronal intestinal dysplasia types A and B, and megacystis microcolon intestinal hypoperistalsis syndrome.

The diagnosis of rectal atresia can be challenging as the patient shows a normal anus. The inability to insert a firm rectal tube or thermometer more than 2 cm from the anal verge was the most important indicator of rectal atresia in our cases. Contrast enema has a valuable role in the evaluation and diagnosis of distal bowel obstruction. In a previous report, the diagnosis of rectal atresia was missed, and laparotomy was performed for an undetermined bowel obstruction. In our third case, the diagnosis was made in the operating room after performing examination under anaesthesia.
Since 1950, over 13 distinct surgical procedures have been described for the treatment of rectal atresia. We have presented these operative techniques in Table 1. All the procedures appeared to have good outcomes for the patients, as the optimal goal was to achieve continence with normal bowel habits. Diverting colostomy is performed and recommended in almost all patients born with rectal atresia, and it was performed in all our patients. Colonography is a critical diagnostic tool prior to definitive repair and colostomy closure. Our procedure technique to all the cases started by positioning the patient in lithotomy position. The anal canal area then was accessed with Lone Star retractor with gentle dilation. Then we started the procedure by dissecting the rectum 0.5 cm above the dentate line in a circular fashion, distal to the atretic segment. Subsequently, we used electrocautery starting posteriorly, the mucosa was held by stay sutures of 4–0 silk. A continuous circular fashion dissection was carried out of the submucosa on both sides of the rectum and anteriorly exposed the rectal muscle cuff. The submucosa then can be stripped from the rectal muscle cuff by using a peanut. The rectal cuff then is dissected in a circular fashion allowing the full thickness of the bowel to be mobilised. After complete and uneventful dissection of the distal rectum and atretic segment, the bowel was freely mobile and easy to be pulled down. From the contrast imaging study, we knew that the atretic segment was not too long or high. By pulling the bowel down, we then resected the cord like atretic segment gradually while establishing our anastomosis sutures between the proximal bowel and the native rectum, preserving the normal anatomy of the anal canal with surrounding sphincters. The rectoanal anastomosis was fashioned with interrupted stitches around 0.5 cm above the dentate line with absorbable suture material (polydioxanone 4–0); we started with four sutures at 12, 3, 6, and 9 o’clock positions. Then, we applied more sutures in between until we reached eight sutures in total. After accomplishing the anastomosis, we did a rectal examination by inserting a 12-French Hager dilator, and it appeared to have a good patent lumen. This procedure is not associated with major wound dehiscence, presacral abscess and fistulas because there is no large incision compared with posterior sagittal anorectoplasty.

In our opinion, according to the unique anatomy of rectal atresia pathology, it can be managed by transanal endorectal pull-through, posterior sagittal anorectoplasty or other new techniques listed in Table 1. Due to the lack of bowel continuity between the two rectal ends, Soave and Duhamel are not suitable for these cases although they have been discussed in the literature. These procedures are well...
known in the management of Hirschsprung disease where there is a good continuity of the bowel. In Duhamel, posterior portion of the defective colon segment is resected then a side-to-side anastomosis is performed. In Soave technique, a circumferential dissection through the muscular coat of the defective segment is performed. Then the proximal normal colon is pulled through with retained muscular sleeve with telescoping anastomosis of the normal colon to the anal canal.

Generally, the prognosis of rectal atresia is promising with a high satisfaction rate and few manageable complications. All our patients experienced constipation, and this is consistent with the fact that our patients presented with low anorectal malformations which have the highest incidence of constipation. A previous report mentioned that more than 50% of patients with anorectal malformations have other concurrent congenital anomalies. The associated anomalies were classified based on the major organ systems involved, and genitourinary anomalies were the most common (49%–81%), followed by musculoskeletal anomalies (43%–45.5%), cardiovascular anomalies (28%), gastrointestinal anomalies (18%) and central nervous system anomalies (12%). In our second case, the patient had a congenital heart defect (VSD), and he was treated accordingly. In our third case, the patient showed constipation that may need long-term balloon dilatation.

CONCLUSION

Transanal endorectal pull-through for rectal atresia is a feasible and safe procedure with satisfactory long-term outcomes. Owing to the normal appearance of the anus in patients with rectal atresia, the diagnosis of rectal atresia can be challenging. All newborns with a picture of distal bowel obstruction and a normal-looking anus necessitate a passage of soft catheter to rule out rectal atresia. Inability to insert a soft rectal tube is the definitive diagnosis while findings of contrast enema might delineate the anatomic features. Patients with rectal atresia tend to have constipation that may need long-term laxative use.

Learning points

- Inability to insert a soft rectal tube or thermometer should raise suspicion of rectal atresia.
- Failure to pass meconium, abdominal distension and bilious vomiting are indicators of neonatal obstruction.
- Patients with rectal atresia should be investigated for other concurrent congenital anomalies.

Contributors

MG performed the literature review about the subject and wrote the proposal. NK wrote the first draft and the final paper. MN revised the final article. SJ collected the patients’ data and revised it.

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