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Rectal atresia and stenosis: unique anorectal malformations

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Abstract

Introduction: Rectal atresia/stenosis is a rare disorder in the spectrum of anorectal malformations and is particularly associated with a presacral mass. These patients are born with a normal anal canal but have a stricture or complete atresia located a few centimeters proximal to the dentate line. We present a surgical technique for the management of these patients, as well as their unique clinical concerns and outcomes.

Methods: We reviewed the records of 14 patients with rectal atresia and 3 with rectal stenosis. We describe a novel technique that we have developed for the preservation of the anterior dentate line that was performed in the last 13 patients.

Results: Rectal atresia/stenosis was associated with a presacral mass in 5 patients (29%). Definitive repair was completed using a circular rectorectal anastomosis in the first 4 patients and an anterior dentate line sparing procedure in the last 13. All patients older than 3 years have demonstrated the ability to have voluntary bowel movements.

Conclusion: With the largest reported series of rectal atresia/stenosis, we have demonstrated a safe and effective technique for repair. Preoperative evaluation must be thorough because a significant number of these patients will have an associated presacral mass.

Both congenital rectal atresia and rectal stenosis are rare disorders comprising only 1% of all anorectal malformations [1,2]. In both conditions, the initial evaluation of the newborn reveals a normal anus and what appears to be a normal anal canal, appropriately located in the midline and within the sphincter mechanism. This may sometimes appear skin lined known as a "funnel anus" (Fig. 1) but usually appears like a normal dentate line. Typically, the condition is diagnosed when a thermometer fails to pass easily into the rectum. The blind ending pouch with a dilated rectum above is typically 2 cm from the anal verge, at the junction of the rectum and anal canal. Stenosis differs from atresia in that there is a narrow, patent channel connecting the rectum with the anal canal. The infant, in cases of stenosis, may pass meconium, and the diagnosis can therefore be delayed.

There are many reported operative approaches for the management of rectal atresia [3-7]. In these repair techniques, a circumferential rectorectal or rectoanal anastomosis is fashioned, or complete removal of the native anal canal with pull-through of the proximal rectum is performed [8]. Although these approaches have provided satisfactory outcomes, most are presented as case reports and have not, therefore, been reproduced in numerous patients. We feel that a technique that preserves at least a portion of the very sensitive anorectum is important for successful outcomes in regard to bowel control.

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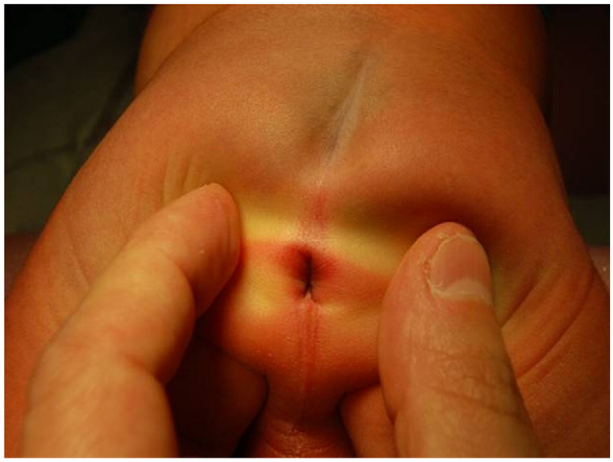

Fig. 1 Funnel anus.

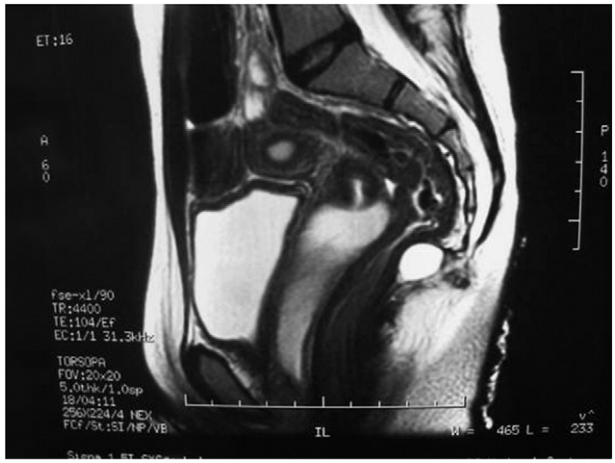

Fig. 2 Magnetic resonance imaging revealing the presence of a rectal duplication.

Given the normal anal canal and normal sphincter mechanism, functional outcomes for these patients should be excellent. We attempted to identify a reproducible method of repair resulting in minimal short- and long-term morbidity, as well as a good functional result. We describe a technique that uses a posterior sagittal approach to repair rectal atresia/stenosis and leaves the anterior portion of the anal canal preserved.

1. Materials and methods

After institutional review board approval, we reviewed the records of all patients who were referred to our facility with the diagnosis of congenital rectal atresia or stenosis (institutional review board no. 2010-2002). At birth, each patient was noted to have a normal-appearing anus, identified in the correct location in the perineum. Most of these patients were diagnosed in the early neonatal period when they failed to pass meconium or when it was found that a thermometer would not pass into the rectum.

Our evaluation included a thorough history and physical examination along with an evaluation for associated anomalies including spinal ultrasound in the newborn period, or magnetic resonance imaging (MRI) after 3 months of age, to assess for tethered cord and for a presacral mass (Fig. 2). We routinely perform pelvic MRIs for patients with rectal atresia or stenosis to rule out an associated presacral mass because ultrasound can sometimes miss smaller lesions. If an initial colostomy is performed, patients are studied with a distal colostogram to confirm the length and location of the distal colon as well as to rule out the possibility of an associated fistula.

1.1. Surgical technique

Patients were admitted to the hospital the day before the planned operation for irrigation of their mucus fistula and intravenous fluid administration if needed. Our preferred operation, performed in the last 13 patients, starts with the patient positioned in the classic prone jack-knife position. A posterior sagittal incision is opened from posterior near the coccyx to the edge of the anus. Meticulous dissection is carried down until the white fascia of the posterior rectum is identified (Fig. 3). Multiple silk sutures are placed along the posterior aspect of the distal anal canal, and the posterior wall of the rectum is mobilized, continuing to preserve the anterior anal canal and dentate line, avoiding dissection of the anterior rectal wall (Fig. 4). The distal anal canal and rectum are opened posteriorly, extending from the skin edge through the level of atresia or stenosis, thus preserving the anterior 180° of the dentate line (Fig. 5). In all patients, the proximal rectal pouch was immediately adjacent to the distal anal canal. Then, with the posterior rectal wall adequately mobilized, it can be brought down and anastomosed to the anoderm along the posterior 180° (Fig. 6). The posterior wall of the rectum is sutured to the posterior edge of the muscle complex, and the posterior sagittal incision is closed. Patients were considered for ostomy reversal when their wound was

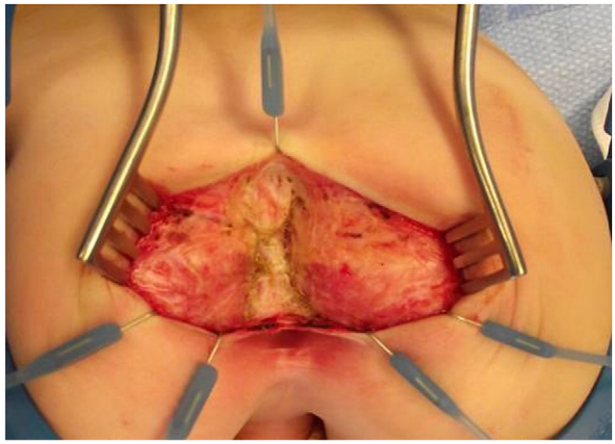

Fig. 3 Opening the posterior midline.

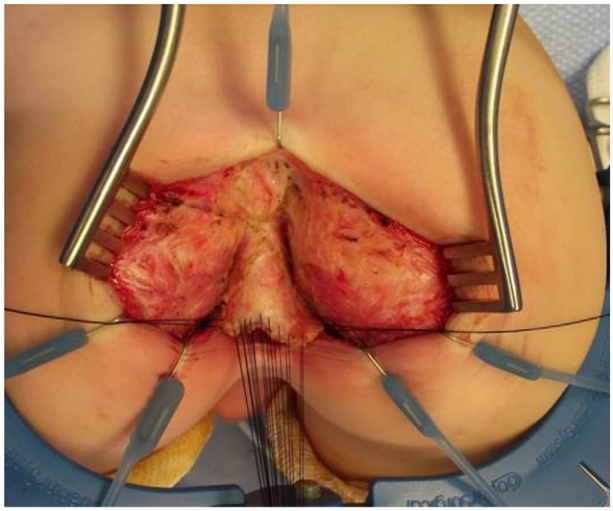

Fig. 4 Silk sutures along the posterior aspect of the distal anal canal and the posterior wall of the rectum being mobilized.

well healed and anal size was appropriate for age. We began anal dilations at 1 month postoperatively.

2. Results

Fourteen patients were referred to our care for correction of rectal atresia and 3 for rectal stenosis. Ten boys and 7 girls comprised our data set. Eleven out of the 14 patients with rectal atresia underwent creation of a diverting colostomy early in life, 2 underwent primary repair without diversion, and 1 was diverted at the same time as definitive repair. Two patients with rectal stenosis were dilated at other institutions with no diversion performed and were referred for definitive repair. The third patient with rectal stenosis underwent diverting colostomy at the time of the definitive repair.

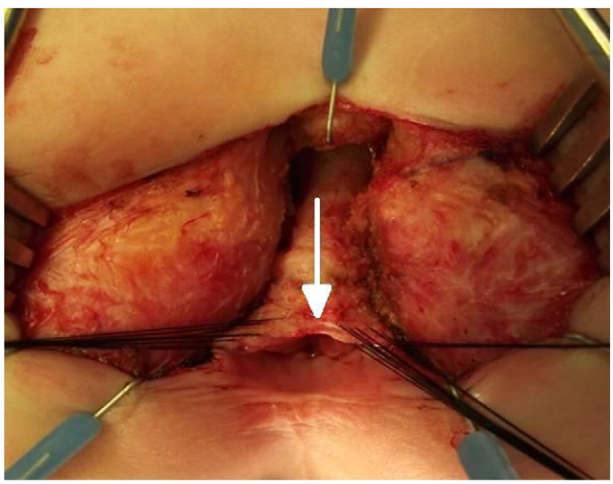

Fig. 5 Splitting of the posterior rectum through the level of the stenosis (denoted by the white arrow).

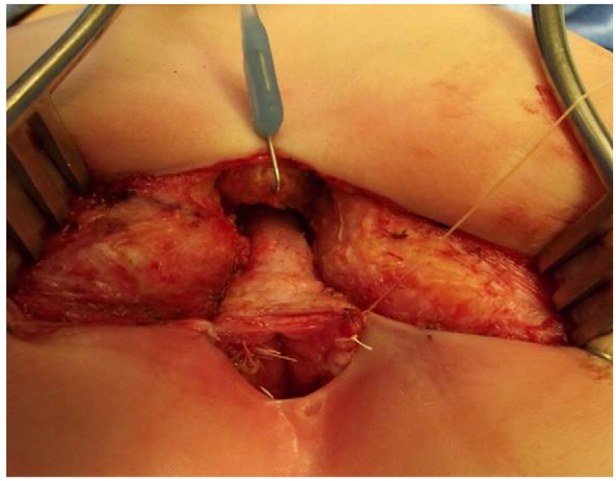

Fig. 6 Distal rectum anastomosed to the posterior anoderm.

Workup for associated anomalies revealed a presacral mass in 5 patients (29%), 1 of whom had a hemisacrum. Thorough screening with pelvic MRI identified 3 presacral masses (60%), which had gone previously undiagnosed at the original institution. An associated tracheoesophageal fistula was found in one patient and an ectopic kidney in another.

All 17 patients underwent a repair using a posterior sagittal approach. In the first 4 patients, a circular rectal to anal canal anastomosis was performed. In the final 13 patients, the anterior dentate line was preserved as described previously. The patient's ages at the time of definitive surgery ranged from 2 days to 6 years (median, 6 months; mean, 10 months). Of the patients who had a presacral mass, all underwent resection at the time of their definitive operation. Pathology of these masses was benign teratoma in 4 cases and a rectal duplication in 1.

Sacral ratios were normal in all patients (range, 0.75-1.00), connoting a lack of caudal regression. At the time of this report, 12 (71%) of the 17 patients are older than 3 years (mean follow-up, 4 years) and have demonstrated the ability to have voluntary bowel movements and are clean between bowel movements. Five (29%) of the 17 patients had some degree of constipation requiring laxative administration. Once constipation was adequately treated; these patients demonstrate the ability to remain clean without soiling.

There were 3 operative complications, a rectovaginal fistula and 2 presacral abscesses, all occurring in patients who underwent repair without a diverting colostomy. Interestingly, both presacral abscesses occurred in patients with an excised presacral mass. The patient who developed the rectovaginal fistula was one of the original patients who had total mobilization of the anal canal, a technique we have subsequently abandoned. In all of these patients, diverting ostomies were performed, and the complications were resolved. The patient's have subsequently had their ostomies reversed, and they have all demonstrated the ability to have voluntary bowel movements. One patient with anal stenosis was managed for a prolonged period at an outside hospital

with anal dilations, was repaired without fecal diversion, and has progressed without complication. All patients underwent postoperative anal dilations, and none developed a postoperative stricture.

3. Discussion

Rectal atresia and stenosis are very rare anorectal anomalies occurring in less than 1% of all anorectal malformations [1,2]. Over the years, many repair techniques have been described in the literature [3,4,7]. In this study, we present our experience with correction of this anomaly using a posterior sagittal approach that leaves the anterior anal orifice and anterior anal canal untouched. We open the posterior anal canal and mobilize the posterior wall of the anal canal and distal rectum, leaving the anterior anal canal untouched to become the anterior 180° of the completed anoplasty. The approach is reproducible and avoids the possible complications of rectovaginal fistula, a urethral injury, or a mislocated anus within the sphincter mechanism. The surgeon can also address the problem of a presacral mass at the same time as the definitive rectal repair.

In most patients born with rectal atresia or stenosis, management includes a diverting colostomy. A diverting colostomy is valuable because it may avoid the complication of a presacral abscess, especially in patients left with a large potential space after excision of a presacral mass. In our series, 13 patients (76%) were managed in this fashion. Postoperative complications were seen in 3 (75%) of the 4 patients who underwent repair without fecal diversion. We, therefore, recommend diversion in all such cases.

In the newborn period, a diligent workup is required to rule out associated anomalies as well as to identify the possibility of a presacral mass, which, in our small series, occurred with moderate frequency (29%). Along with the standard screening tests, a distal colostogram can confirm with certainty the absence of any fistulous communication with the urinary tract and the distance between the distal rectal pouch and anal canal. We suggest spinal ultrasound in the newborn period as well as pelvic MRI to thoroughly evaluate for an associated presacral mass.

Given the fact that these patients have a normal-appearing anus lying within a normal sphincter mechanism, it may be easy to initially overlook the diagnosis of rectal atresia. In the case of rectal stenosis, when the newborn is passing meconium, this is particularly easy to do. As in our neonatal unit, all newborns should be screened by placing a thermometer or cotton-tipped applicator at least 3 cm into the anal canal to confirm patency.

Total mobilization and resection of the anal canal [3] can injure the sphincter mechanism and the sensation inherent in it. It can also lead to inadvertent injury to the rectal wall, vagina, or urethra. To avoid these possible complications, the technique described here leaves the anal orifice, anterior anal canal including the dentate line, and anterior rectal wall untouched. Another benefit to performing an anastomosis in this fashion is that it may not require dilatations in the postoperative period because the anterior dentate line remains intact. Our routine is to calibrate the anus postoperatively at 1 month and continue with dilations daily for several months, but none of the patients developed a stricture, suggesting that dilations may be an unnecessary step. This could minimize trauma to the child as well as the parents.

Patients carrying the diagnosis of rectal atresia or stenosis have excellent prognosis in terms of bowel control, particularly those without an associated presacral mass. We have noted that a significant number of the patients in our series have experienced constipation. This is consistent with the fact that patients with low anorectal malformations have the highest incidence of constipation [9]. Although most of these patients can be managed with fiber and laxatives, careful follow-up is vital (as with all patients who have anorectal malformations) so that patients who need early management of their constipation can be identified. This can potentially avoid complications such as megacolon and overflow pseudoincontinence [10]. Frequent abdominal radiographs and proactive laxative administration allow for early intervention in patients with suspected constipation [10].

In our series, we had 3 complications, a rectovaginal fistula and 2 presacral abscesses. The rectovaginal fistula occurred after the patient initially underwent total mobilization of the anal canal without diversion. We believe that this represents a rare complication in this patient population but could have been avoided with our newer technique of leaving the anterior anus and anal canal untouched. Likewise, both of the presacral abscesses occurred in patients who also had an associated presacral mass and no fecal diversion. These, possibly, were precipitated by the large potential space remaining after resection of the masses and represent another reason why fecal diversion should be done in these patients. Drainage of this potential space may also be warranted. All complications resolved spontaneously after a diverting colostomy was opened.

Anal canal preservation offers a safe, easy, and reproducible technique to repair rectal atresia and stenosis. It minimizes dissection, decreases the chance of injury or anus misplacement, and may avoid the postoperative discomfort of serial anal dilatations. This approach also allows for the surgeon to address a presacral mass, which is a common associated finding. Patients undergoing this procedure who have a normal sacrum and no presacral mass should expect complete continence, although surgeons must be diligent in their long-term management of constipation.

References

[1] Klein M, Thomas R. Surgical conditions of the anus rectum and colon. 18th ed. Kleigman: Nelson textbook of pediatrics; 2007. p. 1635-40. Chapter 341.

[2] Peña A, Levitt MA. Imperforate anus and cloacal malformations. In: Ashcraft KW, Holcomb GW, Murphy JP, editors. Pediatric surgery. 4th ed. Philadelphia (Pa): Elsevier Saunders; 2005. p. 496-517.

[3] Dias RG, Santiago AP, Ferreira MC. Rectal atresia: treatment through a single sacral approach. J Pediatr Surg 1982;17:424-5.
[4] Kisra M, Alkadi H, Zerhoni H, et al. Rectal atresia. J Paediatr Child Health 2005;41:691-3.
[5] Nguyen TL, Pham DH. Laparoscopic and transanal approach for rectal atresia: a novel alternative. J Pediatr Surg 2007;42:E25-7.
[6] Pena A, Levitt MA. Anorectal malformations. In: Grosfeld JL, O'Neil JA, Fonkalsrud EW, Coran AG, editors. Pediatric surgery. Philadelphia: Mosby; 2006. p. 1566-89.
[7] Upadhyaya P. Rectal atresia: transanal, end-to-end, rectorectal anastomosis: a simplified, rational approach to management. J Pediatr Surg 1990;25(5):535-7.
[8] Luo CC, Ming YC, Chu SM, et al. Individualized management of upper rectal atresia. J Pediatr Surg 2009;44:2406-9.
[9] Pena A. Anorectal malformations. Semin Pediatr Surg 1995;4(1): 35-47.
[10] Levitt M, Kant A, Pena A. The morbidity of constipation in patients with anorectal malformations. J Pediatr Surg 2010;45(6):1228-33.