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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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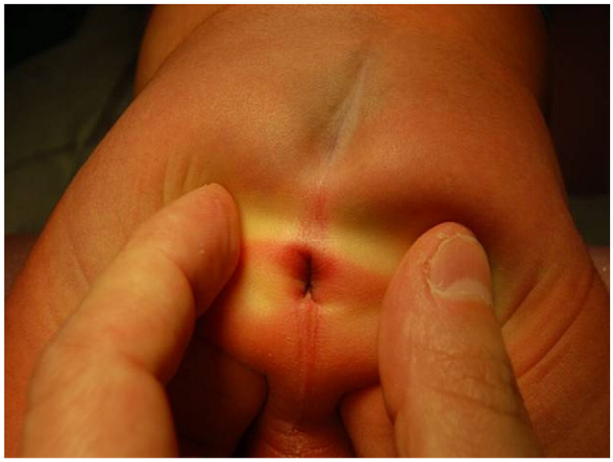

Fig. 1 Funnel anus.

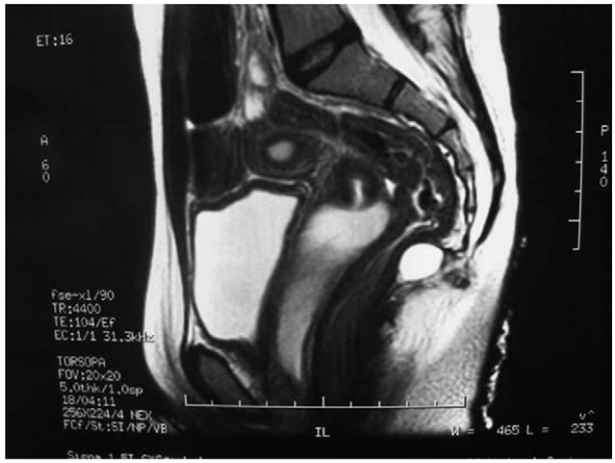

Fig. 2 Magnetic resonance imaging revealing the presence of a rectal duplication.

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

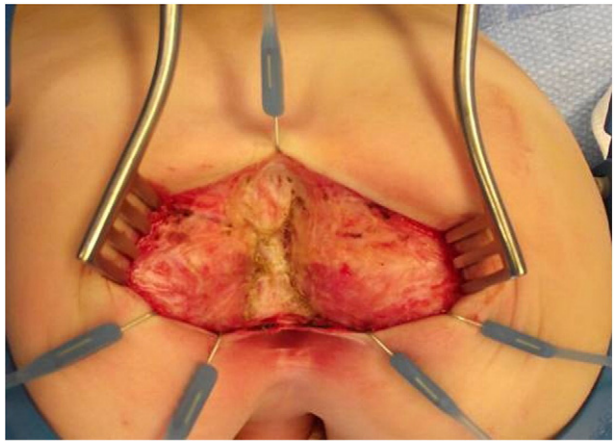

Fig. 3 Opening the posterior midline.