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HOW I DO IT

Rectal atresia and anal stenosis: the difference in the operative technique for these two distinct congenital anorectal malformations

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Abstract Rectal atresia and anal stenosis are rare forms of anorectal malformations. The aim of the definitive surgical repair in such cases is to preserve the anal canal, the dentate line, and the sphincter complex. We present a case of rectal atresia and anal stenosis to demonstrate the differences in the operative repair. The techniques described leave the anterior wall of the very distal anal canal untouched in both rectal stenosis and anal atresia; however, the dissection of the rectum differs. The atretic rectum in rectal atresia is mobilized and sutured to the anal canal circumferentially. In anal stenosis, the posterior rectum is mobilized in the form of rectal advancement, and the posterior 180° is anastomosed directly to the skin (as in a standard PSARP) with preservation of the anal canal as the anterior 180° of the final anoplasty. These patients have an excellent prognosis for bowel control and fecal continence, and therefore, complete mobilization and resection of the anal canal must be avoided.

Keywords Rectal atresia · Rectal stenosis · PSARP · Pre-sacral mass · Anorectal malformation · Imperforate anus

Introduction

Rectal atresia and anal stenosis are rare forms of anorectal malformations representing only 1 % of cases seen [1, 2]. Various operative techniques for the repair of these malformations have been described in the literature including a transanal approach [3], reconstruction via a posterior sagittal incision [4], and using magnets [5]. Children with this type of anorectal malformation have an excellent prognosis in terms of fecal continence because they often have all the anatomical elements required for bowel control, most importantly a normal anal canal and good sphincters. Therefore, the standard PSARP with dissection starting at the distal anal opening is not ideal. The surgeon must appreciate that these malformations are also associated with the presence of a pre-sacral mass and sacral anomalies [4], and when this combination of features exists, it is known as Currarino's triad. The aim of the definitive surgical repair in such cases is to preserve the anal canal, the dentate line and the sphincter complex, and where necessary provide access to the pre-sacral space to allow for excision of the pre-sacral mass.

It became clear to the authors after the publication of 'Rectal atresia and stenosis: unique malformations' [4] that the key principles of anal canal preservation and the fundamental differences in the operative techniques for the two distinct abnormalities were not well understood. This article is intended therefore to address these technical issues through a series of operative photographs illustrating

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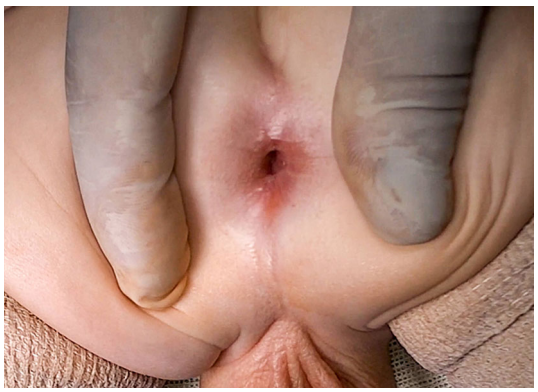

Fig. 1 Examination of a male infant with rectal atresia in the prone position. Normally sited anus centered within the sphincter mechanism. The anal caliber is normal, but blind ending at a depth of 2 cm

the key differences in the repair of rectal atresia and anal stenosis.

Rectal atresia and anal stenosis

In both conditions, the initial newborn examination typically reveals a normal anal opening within the sphincter complex with an apparently normal anal canal (see Figs. 1, 8). On further examination of the infant that has failed to pass meconium, the diagnosis of rectal atresia becomes apparent when there is an inability to pass a Hegar dilator. The diagnosis of anal stenosis is often delayed as there is usually normal passage of meconium in the immediate newborn period, but the child often goes on to develop difficulty stooling and abdominal distension. On examination of the anus, it is narrowed and may appear skin lined and 'funnel like.'

Case 1: Rectal atresia

The first case demonstrates the operative repair of rectal atresia through a posterior sagittal incision. Figure 1 shows the examination findings of a male infant (10 months old, weighing 6.6 kg) in the prone jackknife position, demonstrating a normal anal canal within the sphincter mechanism, calibrating to a size 12 Hegar dilator which is normal for age and weight. In the newborn period this infant failed to pass meconium, and failure to pass a Hegar beyond 2–3 cm revealed the diagnosis and a colostomy was fashioned. The sacrum was normal, and there was no evidence of a pre-sacral mass on MRI. The distal loop colostogram (Fig. 2) did not reveal any communication to the urinary tract and also demonstrated that the rectum could be reached via a posterior sagittal incision.

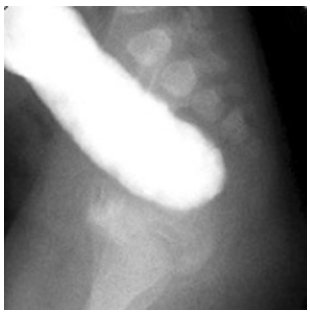

Fig. 2 Distal loop colostogram

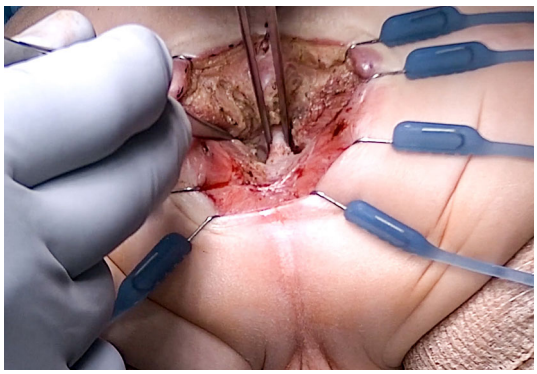

Fig. 3 Atretic segment

Operative technique

The patient has a urinary catheter inserted pre-operatively. A posterior sagittal incision is made from the coccyx to the anus, with the anus and dentate line opened in the posterior midline (12 o'clock position) and laid open like a book. Careful dissection continues until the white fascia of the rectum is seen. The atretic segment is identified as demonstrated in Fig. 3 with the dissecting forceps. This segment is excised taking care to leave the very distal (patent) anal opening and dentate line intact. The more proximal blind ending rectum is then identified. In this case adequate exposure required excision of the coccyx. Figure 4 shows that the blind ending rectum has been opened and that silk sutures have been placed circumferentially around the enterotomy. The rectum is mobilized circumferentially, as in a standard posterior sagittal anorectoplasty, to create adequate length for the rectum to reach the perineum without tension. Care is taken in dissecting the anterior rectal wall, so as not to injure the urethra. Figure 5 demonstrates the beginning of the anastomosis. The mobilized rectum is sutured to the anal canal (above the preserved dentate line). In Fig. 6, the diagram illustrates that the original anal canal (A-blue circle) is laid open with the original 12 o'clock position (orange dot) currently split in half and now lying in both the 3 o'clock and 9 o'clock

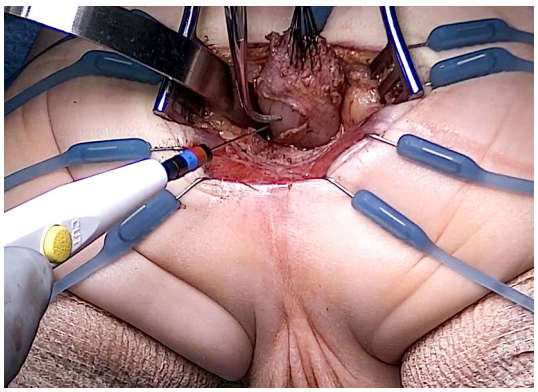

Fig. 4 Blind ending rectum

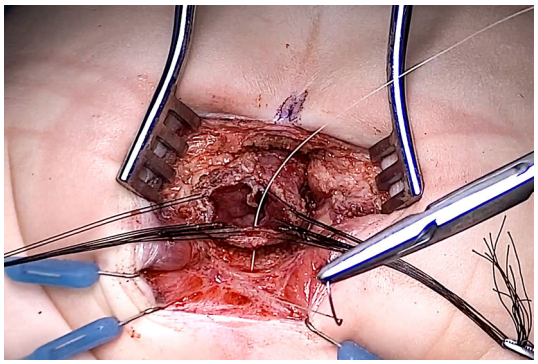

Fig. 5 Beginning of the anastomosis

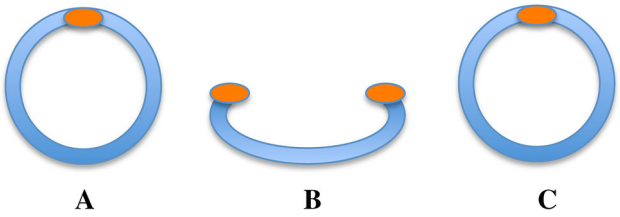

Fig. 6 Anus normal caliber anus (**a**) is laid is open as shown in (**b**). This is then re-established after the anastomosis as in (**c**)

positions (B). The first stitch is placed at the 6 o'clock position. The anastomosis is completed around the 360° circumference, and the 12 o'clock position is re-established (Fig. 6c). The posterior sagittal incision is closed with absorbable sutures (Fig. 7).

Case 2: Anal stenosis

Figure 8 shows the clinical examination findings of a female infant (13 months old, 8.1 kg) in the prone jack-knife position. Here you can see the 'funnel-shaped' anus. In this patient the anus is stenotic allowing passage of only a size 6 Hegar. The AP radiograph of the sacrum (Fig. 9a) demonstrated a scimitar sacrum and the pre-operative MRI (Fig. 9b) revealed a pre-sacral mass, therefore collectively

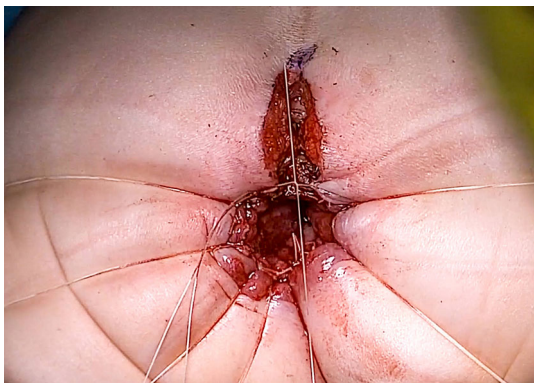

Fig. 7 Anastomosis is completed around the 360° circumference and the 12 o'clock position is re-established

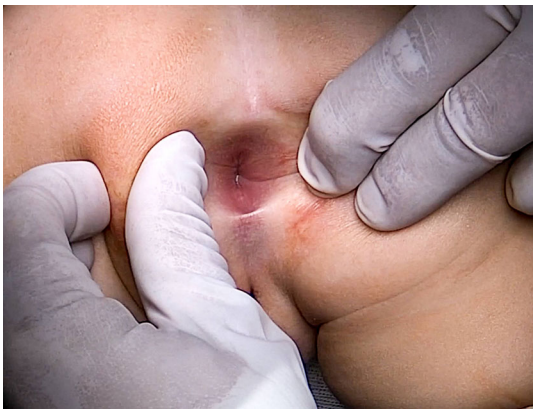

Fig. 8 Anus is 'funnel' shaped and skin lined

forming the Currarino's triad. (This child's mother was also known to have a pre-sacral mass and sacral defect). The alpha-1-fetoprotein level pre-op was 4.3 ng/mL.

Operative technique

A posterior sagittal incision is made (Fig. 10) from the coccyx down to the level of the anus, with the plan to split the anal canal in the posterior midline. Dissection continues to the level of the white rectal fascia. The pre-sacral mass has been excised (Fig. 11) and the coccyx removed. Now the rectum is mobilized on the posterior 180° only, to allow for posterior advancement of the rectum (Fig. 12). No anterior rectal wall dissection is needed. The anal canal is opened in the posterior midline with cautery (Fig. 12) through the stenotic segment into the normal caliber lumen to allow passage of a size 15 Hegar (Fig. 13). The original dentate line is once again laid open like a book (Fig. 14), but there is a distinct difference at this point with regards to the anastomosis.

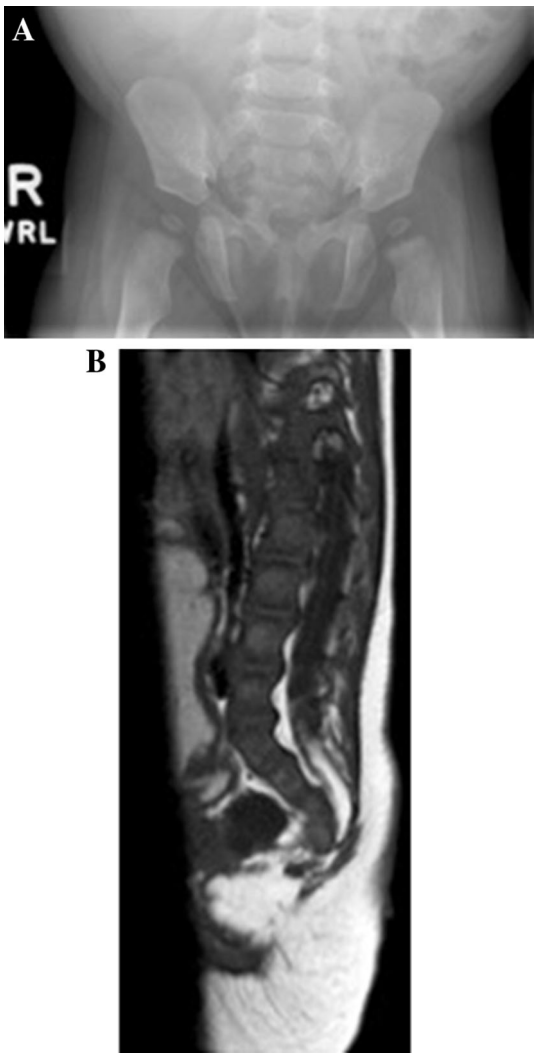

Fig. 9 **a** AP radiograph sacrum demonstrating scimitar sacrum, **b** MRI spine demonstrating a pre-sacral mass

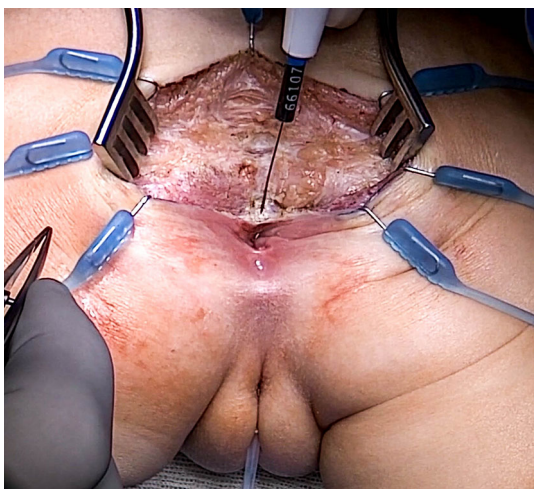

Fig. 10 Posterior sagittal incision

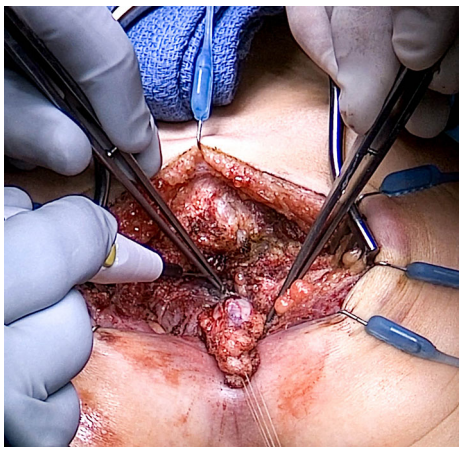

Fig. 11 Excision of the pre-sacral mass

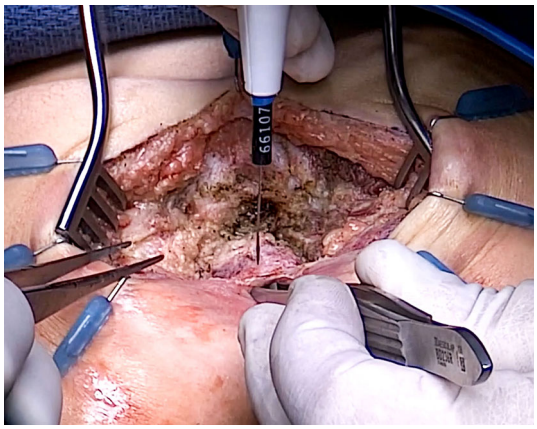

Fig. 12 Rectum is opened in the posterior midline through the stenotic segment into the normal caliber lumen

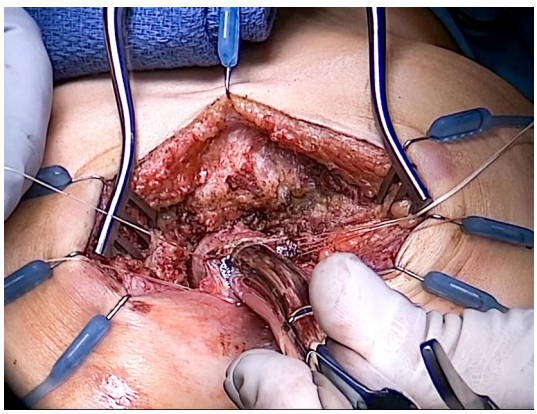

Fig. 13 Size 15 Hegar

Figure 14 illustrates the stenotic anus in (A) with the orange dot representing the 12 o'clock position once again. In diagram (B) the native anus is laid open with the 12

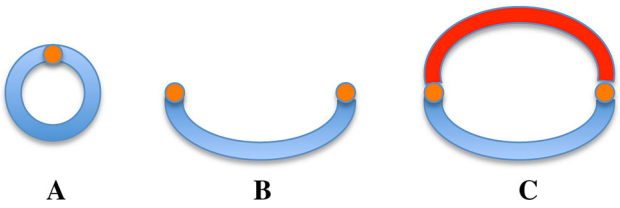

Fig. 14 Anus normal caliber anus (**a**) is laid is open as shown in (**b**). **c** demonstrates the reconstructed anus

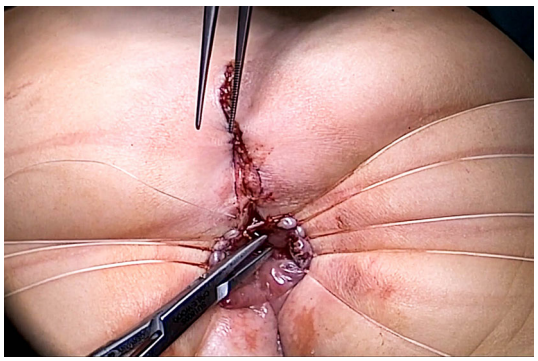

Fig. 15 Completion of the anoplasty

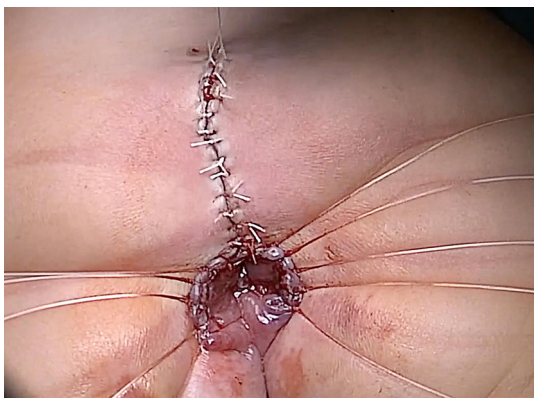

Fig. 16 Closure of the posterior sagittal incision

o'clock position now being at 3 and 9 o'clock positions. The native anus and dentate line in the case of anal stenosis, at the conclusion of the procedure, forms only the anterior 180° of the neo-anus (C). The posterior neo-anus is the posteriorly advanced rectum which is sutured directly to the skin (Fig. 14) as in a standard PSARP (as demonstrated in red).

Therefore, the anterior 180 is the original anal canal now laid open, and the posterior aspect is rectal mucosa to skin, essentially doubling the size of the lumen. The point of tension in the closure of the posterior incision is at 12 o'clock (Fig. 15). This is different from a normal PSARP closure where the 6 o'clock and 12 o'clock sutures are tension free. No sutures are required on the anterior 180° (Fig. 16).

The histology from the resected pre-sacral mass was consistent with a mature teratoma. The postoperative alpha-1-fetoprotein level was 24.1 ng/mL, and this will be followed closely observing for recurrence.

Discussion

There are several reported operative techniques for the management of rectal atresia and anal stenosis [4–9], and these include repair via a transanal approach and via a posterior sagittal incision. Our concern with the transanal only approach is that it is seems likely that the plane of dissection would be difficult to establish in cases of a long atretic segment increasing the chance of inadvertent injury to the urethra or vagina.

In addition, there is a known association with a pre-sacral mass in cases of both rectal atresia and anal stenosis [4], highlighting the fact that children with anorectal malformations need a comprehensive pre-operative work up which includes a spinal ultrasound and MRI of the spine. In our opinion excision of the mass should be expedited as the mass may be malignant or undergo malignant transformation. If there is a communication between the spinal column and the mass (or an associated tethered cord), it is our normal practice to ensure that the neurosurgical components are managed prior to the anoplasty. In this recent series the pre-sacral mass was resected at the time of the operative repair of the anorectal malformation through the posterior sagittal incision. Resection of a pre-sacral mass and removal of the coccyx are not possible via a transanal only approach.

A novel technique of 'magnamosis' in the treatment of rectal has been reported [5] in the literature. This technique is potentially suitable for cases of a web/membrane, however, unlikely to be successful in cases of a long stenotic or atretic segment.

The key principle with regards to the operative repair of rectal atresia and anal stenosis is preservation of the dentate line and the sphincter mechanism as the patient is essentially born with a normal anal canal. These patients have an excellent prognosis for bowel control and fecal continence [4, 10]. Complete mobilization and resection of the anal canal must therefore be avoided. The techniques described here leave the anterior wall of the very distal anal canal untouched in both rectal stenosis and anal atresia. However, the dissection of the rectum in these cases differs. The atretic rectum in rectal atresia is mobilized circumferentially and with the potential to damage to the urethra in males and the vagina in females. The distal rectum is mobilized and sutured to the anal canal circumferentially.

In anal stenosis, this anterior dissection of the rectum is not required. The posterior rectum is mobilized in the form of rectal advancement, and the posterior 180° is rectum anastomosed directly to the skin as in a standard PSARP, with preservation of the anal canal, but leaving it as the anterior 180° of the final anoplasty. The point of tension in the repair of anal stenosis is different from a normal PSARP or the repair of rectal atresia. The point of tension is at 12 o'clock, whereas in rectal atresia or a standard PSARP, the sutures are tension free. It is this tension and the potential pre-sacral space following excision of a mass, which has led us to recommend defunctioning with a colostomy in all cases of anal stenosis [4] and insertion of a drain if the dead space is of a significant size.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This study was approved by the Institutional Board Review.

Informed consent Informed consent was obtained from the parents for the use of photographic images and recording of the operative procedures.

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