

Rectal atresia is a very unique malformation that deserves a special description. It happens in our experience, in about 1 % of all cases of anorectal malformations. In this defect, the anus seems to be completely normal, including the quality of the sphincter and the location of the anal orifice. However, deep inside the anus, just at the junction of the anal canal with the rectum, there is an atresia or narrowing (stenosis) (Fig. 14.1). Occasionally, we see atresias or stenosis located at a different level. The space that separates the dilated blind rectum, from the anal canal, is represented by a septum that sometimes is extremely thin and can be perforated, and other times it is very thick. In some unusual cases, there is a significant separation between the blind upper rectum and the lower anal canal.

Interestingly, the sphincter mechanism is excellent in most cases. There is one particular malformation similar to this one that is represented by a stricture or by atresia of the rectum, associated to a presacral mass and a sacral defect (see Chap. 8, Sect. 8.2), which is a completely different type of defect. The only thing they have in common is the fact that the rectum is narrow or atretic.

We believe that rectal atresia with normal sacrum and no presacral mass is unique, because the sphincter mechanism is normal and also because these patients do not have the typical association with all the defects that we see in other anorectal malformations. As a consequence, the prognosis for these patients is excellent, in terms of bowel control. They have a

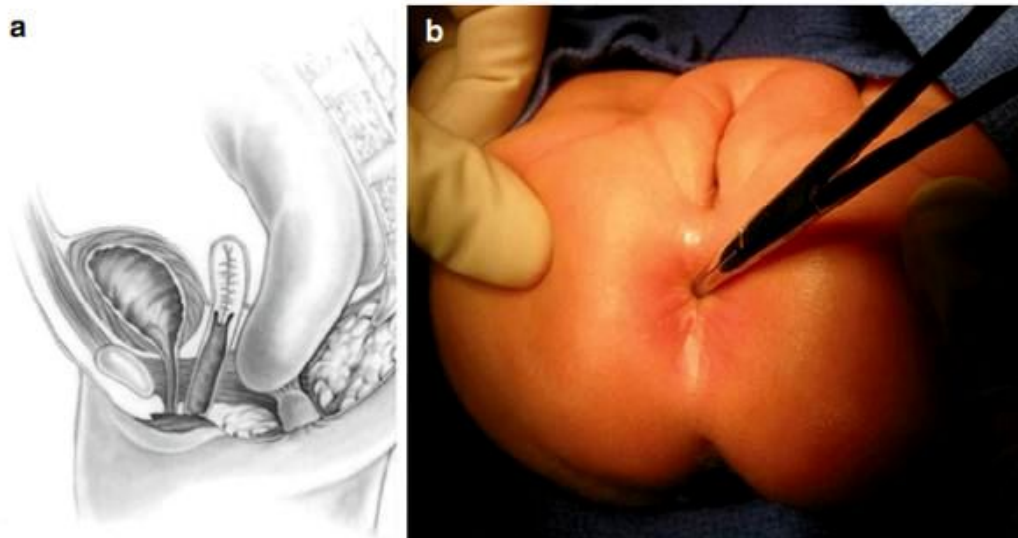


Fig. 14.1 Rectal Atresia. (a) Diagram. (b) External appearance

significant tendency to suffer from severe constipation because they are born with a blind, very dilated rectum. These malformations have been previously described in the literature [1–5].

Rectal atresia has been traditionally described in the old textbooks. The baby is born with a normal-looking anus, and the nurse or the pediatrician tries to pass a thermometer through the anus and finds an obstruction. In fact, part of a routine examination of every “normal” newborn is to check the patency of the anus, unless the baby is already passing meconium.

14.1 Treatment

If one could think in an ideal indication for a posterior sagittal approach, this would be the malformation which seems to be more indicated. The defect is easily repaired through a posterior sagittal incision. In our initial cases, we simply remove the septum that separates the upper rectum from the anal canal and created an end-to-end anastomosis (Figs. 14.2 and 14.3). Subsequently, we found some cases in which the size discrepancy between the upper blind rectum and the small anal canal was very severe, and in order to expand the size of the anal canal, we introduced a techni-

cal modification maneuver [5] (Fig. 14.4). Most of the patients that we operated on came to us already with a colostomy in place. Since the patient has a colostomy, one can perform a distal colostogram and simultaneously introduce a metallic dilator in the anal canal to have a lateral image of the atresia and estimate the distance between the upper pouch and the anal canal. If we could make the diagnosis early in an otherwise healthy newborn baby, we would recommend to do the operation without a colostomy.

14.2 Surgical Repair

The patient is placed in the prone position and we approach the malformation posterior sagittally. We go through the skin, subcutaneous tissue, parasagittal fibers, ischiorectal fossa, and the entire sphincter mechanism to expose and open completely the anal canal and the upper blind rectum. One can see in most cases the pectinate line, at the same location as the atresia (Fig. 14.2a).

Unfortunately, we still see some of these patients, previously operated in whom the surgeon considered that the little anal canal was useless and therefore decided to resect it and pulled down the dilated piece of rectum. That is rather

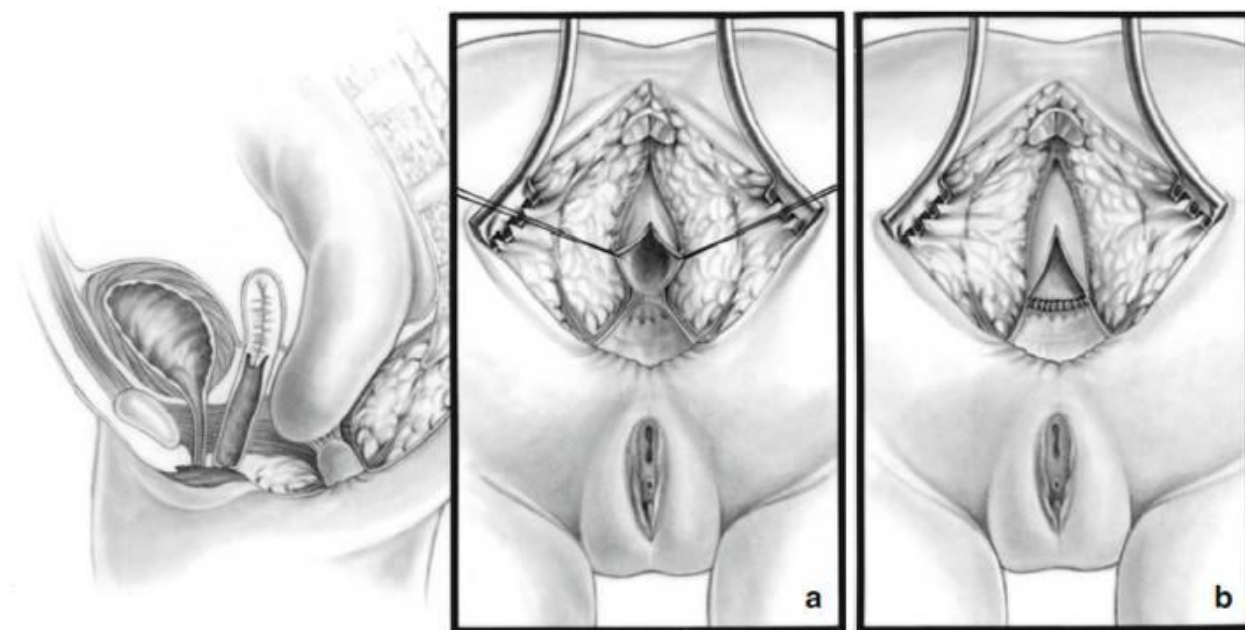


Fig. 14.2 Repair of a Rectal Atresia. (a) Incision, exposed defect, open upper rectum, and anal canal. (b) Anastomosis of the upper rectum to anal canal

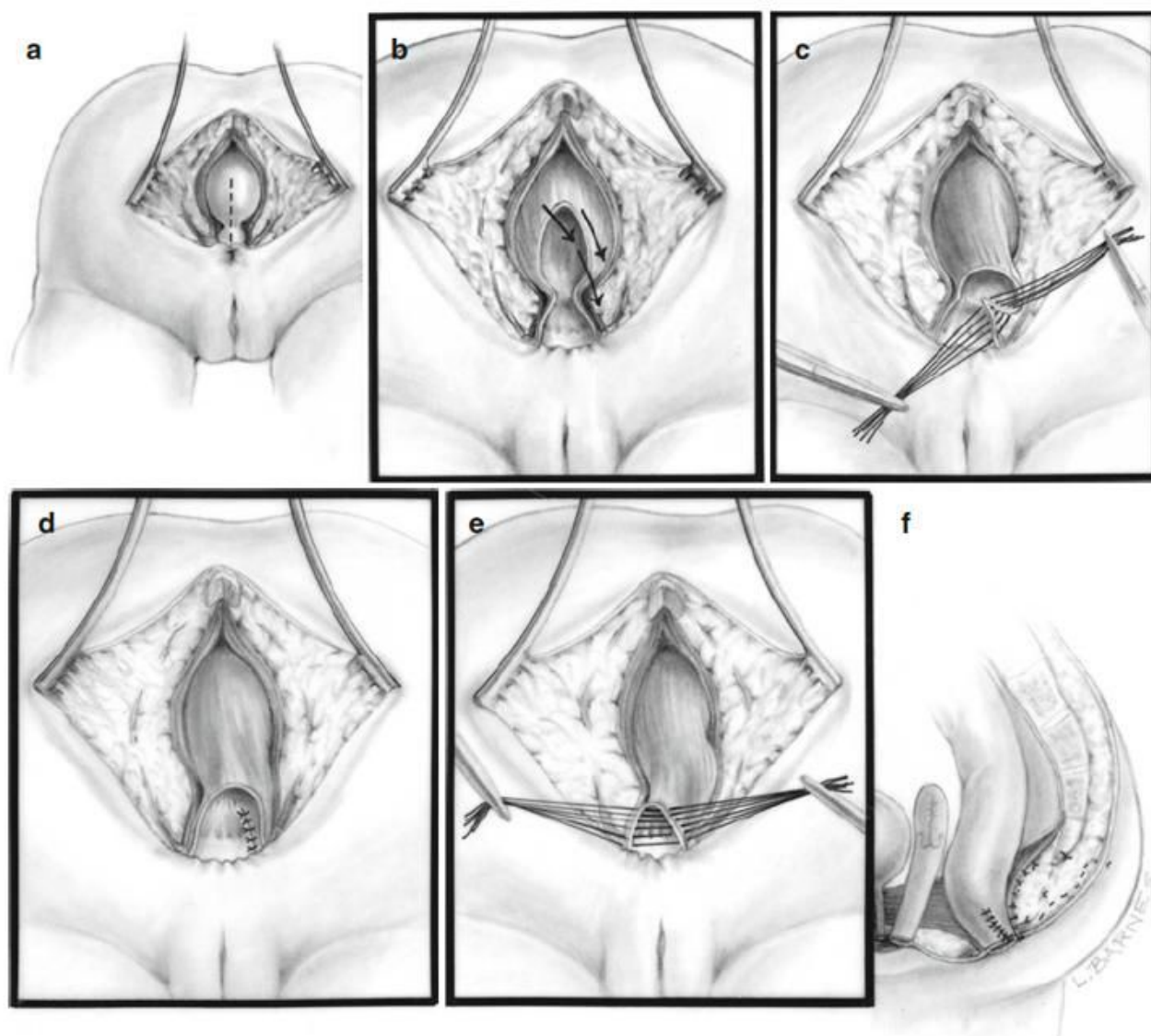


Fig. 14.4 Technical variant to expand the size of a very small anal canal. (a) Incision. (b) Open rectum. Arrows show the portion of the rectum to be mobilized. (c) Sutures on one side of anal canal and rectum. (d) Sutures tied down. (e) Same maneuver, opposite side. (f) Finished operation

References

1. Dias RG, Santiago Ade P, Ferreira MC (1982) Rectal atresia: treatment through a single sacral approach. *J Pediatr Surg* 17(4):424–425
2. Upadhyaya P (1990) Rectal atresia: transanal, end-to-end, rectorectal anastomosis: a simplified, rational approach to management. *J Pediatr Surg* 25(5):535–537
3. Kisra M, Alkadi H, Zerhoni H, Ettayebi F, Benhammou M (2005) Rectal atresia. *J Paediatr Child Health* 41(12):691–693. doi:[10.1111/j.1440-1754.2005.00763.x](https://doi.org/10.1111/j.1440-1754.2005.00763.x)
4. Nguyen TL, Pham DH (2007) Laparoscopic and transanal approach for rectal atresia: a novel alternative. *J Pediatr Surg* 42(11):E25–E27. doi:[10.1016/j.jpedsurg.2007.08.049](https://doi.org/10.1016/j.jpedsurg.2007.08.049)
5. Hamrick M, Eradi B, Bischoff A, Loudon E, Pena A, Levitt MA (2012) Rectal atresia and stenosis: unique anorectal malformations. *J Pediatr Surg* 47(6):1280–1284. doi:[10.1016/j.jpedsurg.2012.03.036](https://doi.org/10.1016/j.jpedsurg.2012.03.036)

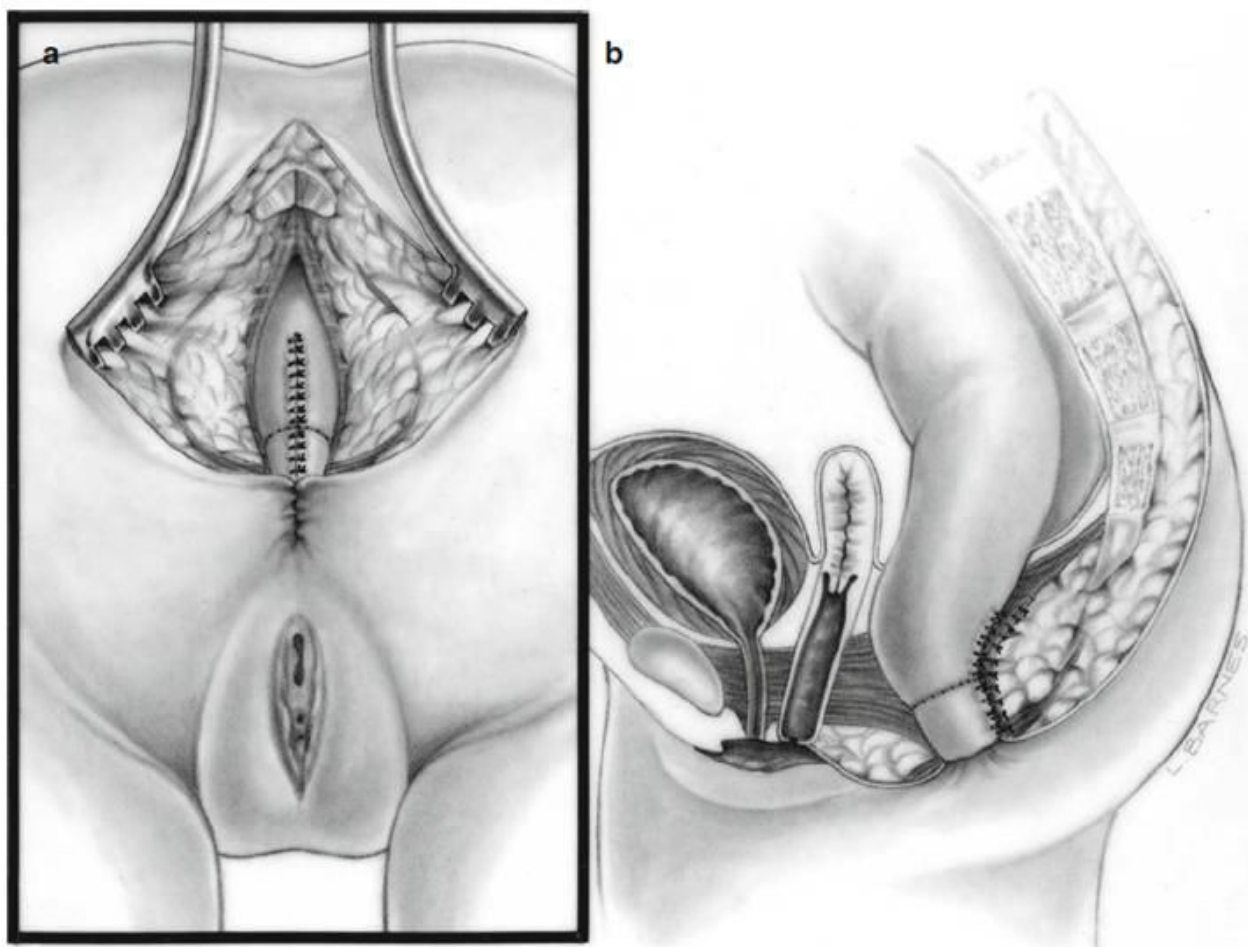


Fig. 14.3 Diagram showing the repair of rectal atresia. (a) Rectum repaired, (b) Sagittal view of the finished operation

regrettable, because the anal canal, as we know, represents the area of sensation that will provide bowel control to these patients. It is, therefore, very important to preserve that little anal canal. Sometimes the size of the anal canal is too small. For that, we introduced a technical modification [5], consisting in mobilizing the posterior rectal wall, down to the skin of the anus (Fig. 14.4), enlarging the circumference of the anus. We realize that by doing that, the posterior aspect of the anus will no longer be a real anal canal, but rather a rectal wall. However we manage to preserve most of the circumference of the original anal canal, which will provide enough sensation to have bowel control. We must keep in mind that after we finish this procedure, the anastomosis that we created between the upper dilated rectum

and the anal canal is going to be permanently collapsed by the effect of the sphincter mechanism that keeps the anal canal closed all the time, except during defecation; therefore, these babies must be subjected to the same protocol of anal dilatations that we already described.

Some surgeons [4] went as far as to perform a “laparoscopic transanal approach” to repair this malformation. To demonstrate that something can be done does not mean that it must be done. We cannot justify to change a limited, painless, bloodless, quick, minimally invasive, non-laparoscopic procedure for a laparoscopic invasive operation that includes an unnecessary total rectal dissection.

Our experience includes 11 cases and has been previously published [5].