Posterior rectal advancement with fistula preservation in patients with anorectal malformation. A multicenter study

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ABSTRACT

Background. Anorectal malformations (ARM) with rectoperineal fistula are mainly repaired with a posterior sagittal anorectoplasty (PSARP), which can be challenging given the proximity of the fistula to the vagina and urethra. The posterior rectal advancement anoplasty (PRAA), preserves the anterior wall of the fistula without leaving an anterior or a posterior sagittal incision. It is indicated for selected cases of ARM with rectoperineal fistula in which the anterior aspect of the fistula is partially surrounded by sphincter complex.

Methods. Multicentre and retrospective study of patients with rectoperineal fistula treated with PRAA. We analysed gender, associated malformations, age, operative time, short and long-term results, and complications.

Results. 18 patients aged 93.5 (1.75-312) days underwent PRAA. Surgical time was 35 (25-45) minutes and feeding was started at 24 (5-48) hours postoperatively. There were no vaginal or urethral injuries, no wound infections or dehiscenses. Throughout the 38 (12.75-45.50) months of follow-up there were no anal strictures. All patients are passing stool, 11 (61%) of them with the need of a low dose stool softener.

Conclusion. Selected patients with rectoperineal fistula can be treated with PRAA with a shorter surgical time and hospital stay. This technique provides good results and lower risk of injury to neighbouring structures.

KEY WORDS: Anorectal malformations; Posterior sagittal anorectoplasty; Posterior rectal advancement anoplasty.

ANOPLASTIA DE AVANCE RECTAL POSTERIOR CON CONSERVACIÓN DE LA FÍSTULA EN PACIENTES CON MALFORMACIÓN ANORRECTAL. ESTUDIO MULTICÉNTRICO

RESUMEN

Introducción. Las malformaciones anorrectales (MAR) con fístula rectoperineal suelen repararse mediante anorrectoplastia sagital posterior (ARPSP), que dada la proximidad de la fístula a la vagina y la uretra, puede resultar dificultosa. La anoplastia de avance rectal posterior (AARP) conserva la pared anterior de la fístula sin dejar una incisión sagital anterior o posterior. Está indicada en casos concretos de MAR con fístula rectoperineal en los que el aspecto anterior de la fístula se encuentra parcialmente rodeado de complejo esfinteriano.

Material y métodos. Estudio retrospectivo multicéntrico realizado en pacientes con fístula rectoperineal tratada con AARP. Se analizaron el género, las malformaciones asociadas, la edad, el tiempo operatorio, los resultados a corto y largo plazo, y las complicaciones.

Resultados. Se realizó AARP en 18 pacientes con una edad de 93,5 (1,75-312) días. El tiempo operatorio fue de 35 (25-45) minutos, iniciándose la alimentación una vez transcurridas 24 (5-48) horas desde la intervención. No se registraron lesiones vaginales ni uretrales, infecciones de la herida o dehiscencias. En los 38 (12,75-45,50) meses de seguimiento, no se han observado estenosis anales. Todos los pacientes defecan con normalidad, 11 (61%) de ellos con necesidad de una pequeña dosis de ablandador de heces.

Conclusión. Algunos pacientes con fístula rectoperineal son aptos para AARP, intervención que conlleva un menor tiempo operatorio y una estancia hospitalaria más reducida, con buenos resultados y un menor riesgo de lesión en las estructuras adyacentes.

PALABRAS CLAVE: Malformaciones anorrectales; Anorrectoplastia sagital posterior; Anoplastia de avance.

INTRODUCTION

Anorectal malformations (ARMs) are a group of congenital anomalies of the distal gastrointestinal and urogenital tracts that can affect both males and females. They have an incidence of 1 in every 5,000 live births[(1)]. Of these, rectoperineal fistula is the most common subtype,

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accounting for up to 40% of cases[2]. It is a benign subtype that can be surgically treated within the first 24 hours of birth, with good results in terms of faecal continence.

Prior to 1985, the cut-back perineal anoplasty and translocation anoplasty were the treatments of choice for the low anomalies[3]. The cut-back was employed as a primary and definite procedure for perineal fistulas in males and females and also in vestibular or vulvar fistulas in females[4]. It provided an early and adequate outlet for stool and prevented colonic obstruction and hypertrophy with the advantage of having no anterior dissection or posterior wound. However, this technique lost popularity due to its over-indication, such as in rectovestibular fistulas with the anterior aspect of the fistula outside the sphincter complex. In the 80s this surgical approach was replaced by the posterior sagittal anorectoplasty (PSARP), involving anterior and posterior dissection and mobilization of the fistula and distal rectum. This approach was adopted as the procedure of choice for the correction of the entire spectrum of anorectal malformations. However, it can pose a challenge given the proximity of the vagina and urethra in males and females respectively[5,6]. The technique implies the removal of the last part of the rectum due to the complete mobilization of the fistula, which some authors believe to have value in the continence mechanism, and may be involved in residual faecal incontinence in these patient[7].

The posterior rectal advancement anoplasty (PRAA) first described by Halleran et al in 2021[8] is a technique that preserves the anterior wall of the fistula and does not leave an anterior or posterior sagittal incision. PRAA is only indicated in patients with rectoperineal fistulas in whom the anterior part of the fistula is, at least, partially surrounded by sphincter mechanism. We chose to utilise this technique in our patient population because of the technical advantage of avoiding any dissection near the urethra or vagina, the avoidance of dehiscences because of the limited incision and avoidance of the perineal body altogether, and the potential value of the distal rectum for continence.

MATERIALS AND METHODS

We performed a multicentre and retrospective study on 18 patients diagnosed with rectoperineal fistula and surgically treated with posterior rectal advancement anoplasty between the years 2019 and 2023 in 3 tertiary paediatric surgery departments. The procedure was offered to patients in which the anterior aspect of the fistula was partially surrounded by the sphincter complex. The fistula was defined as a perineal orifice smaller than what is considered normal calibre for a full-term anus (Hegar 10-12). A normally sized anus positioned on the anterior limit of the sphincter would be considered an anterior anus, not requiring surgical repair. We analysed associated malformations, age at the time of the intervention, operative time, short and long-term results, and complications. All patients underwent VACTERL screening to exclude associated malformations.

We excluded patients with rectoperineal fistula in whom the anterior aspect of the fistula was completely outside the sphincter complex thus who required a complete mobilization of the anterior part of the fistula. In those patients a classic mini-PSARP was performed with circumferential dissection of the fistula.

The PRAA operative steps are shown in figure 1.

After identifying the elliptical sphincter complex with the use of a muscle stimulator, a midline sagittal incision is made from the posterior aspect of the fistula to the edge of the posterior aspect of the identified anal sphincter. Traction stitches are placed on the posterior hemi-circumference of the anus and the rectum is dissected down to its posterior wall. When sufficient length of the rectum is obtained, the posterior edge is incised for 1-2 mm, and the posterior rectal edge is advanced to skin level at the posterior border of the sphincter, performing an advancement flap. A hemi-anoplasty is performed, leaving the anterior part of the anus undissected and the posterior part reconstructed, entirely surrounded by the sphincter.

Clinical data was entered into a database (Microsoft Excel 2019; Microsoft Corporation, Redmond, WA, USA), and statistical analysis was performed using SPSS for MAC OSX (version 22; IBM Corp., Armonk, NY, USA). Qualitative variables are reflected as absolute numbers or percentages, while quantitative variables are shown as medians and 25th and 75th percentiles given that none followed a normal distribution when tested with the Kolmogórov-Smirnov test.

RESULTS

18 patients underwent a PRAA with a median age of 93.5 (1.75-312) days of life. Only 1 (5%) patient was diagnosed prenatally, with confirmation at birth. Seven (38%) patients were treated in the neonatal period and 11 (61%) in a delayed fashion, between 1 and 29 months of age. 3 (16%) patients underwent a stoma which was later closed uneventfully.

11 (61%) patients presented associated congenital malformations, of which 3 (16%) had at least 3 diagnostic malformations for VACTERL association. Another 5 (27%) patients did not meet the criteria for VACTERL but presented another associated anomaly.

The most frequent associated malformations were cardiac in 6 (33%) cases, followed by renal in 4 (22%) and vertebral in 3 (16%). No patient had an oesophageal atresia or a limb malformation. The characteristics of the associated malformations are showed in Table 1.

The median surgical time was 35 (25-45) minutes. Feeding was started at a median 24 (5-48) hours postop-

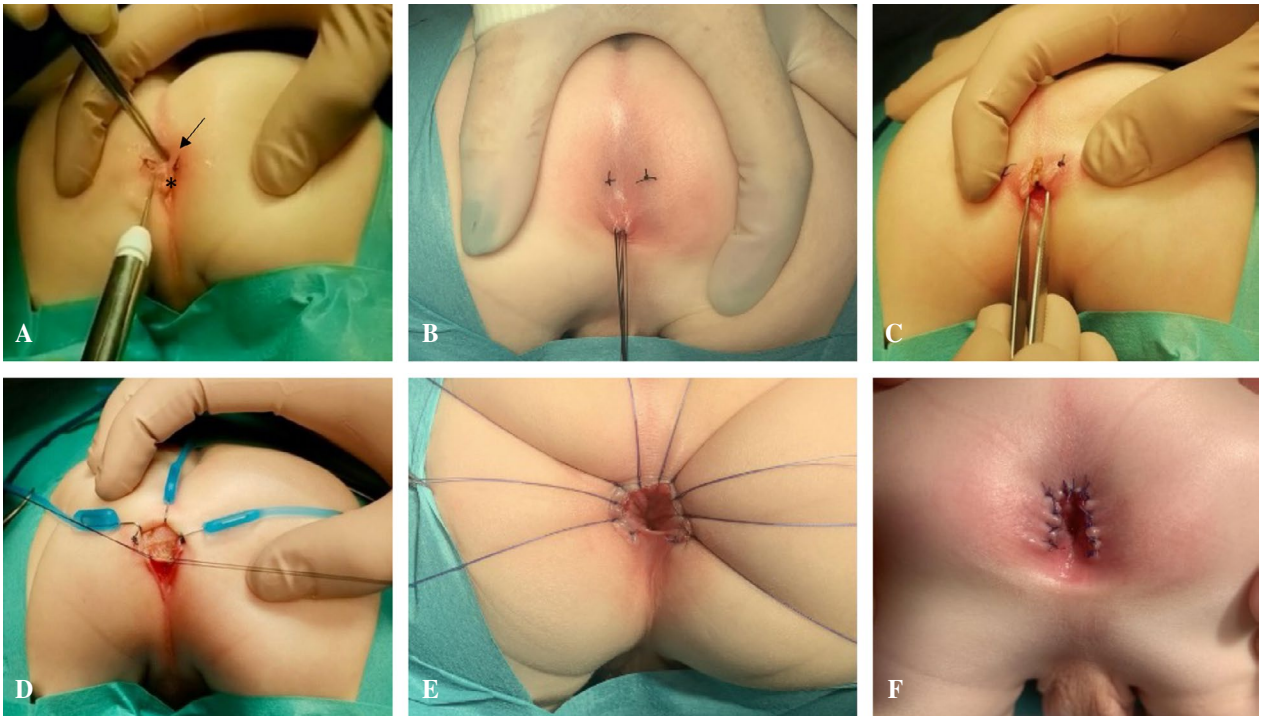

Figure 1. Operative steps of the posterior rectal advancement anoplasty. A) Identification of the sphincter complex with muscle stimulator. The arrow is pointing to the centre of the sphincter and the asterisk is marking the fistula. B) Traction stitches on the fistula and marking stitches on the posterior edge of the sphincter. C) Midline sagittal incision from the posterior aspect of the fistula to the edge of the sphincter. D) Traction stitches on the posterior hemi-circumference of the anus and rectum dissection down to the posterior wall. E) Advancement flap of the rectum to skin level at the posterior border of the sphincter. F) Hemi-anoplasty.

Table 1. Patient characteristics.

Patient	*Associated anomaly*	*VACTERL (yes/no)*
1	Hemivertebrae. Atrial septal defect and atrial septal aneurysm	Yes
2	Atrial septal defect. Aplasia cutis	No
3	Left lumbar hemi-vertebrae (L3, L5). Atrial septal defect, Noonan syndrome. Pyelocaliceal ectasia with dilatation of left ureter, hypospadias bilateral cryptorchidism. 3 pairs of ribs. Lumbosacral scoliosis	Yes
4	Atrial septal defect. Left pyelic ectasia. Pseudoarthrosis of the right clavicle	Yes
5	Atrial septal defect with mild pulmonary stenosis	No
6	None	No
7	Atrial septal defect. Glandular hypospadias. Right preauricular appendage	No
8	None	No
9	Crossed renal ectopia and left and right kidney fusion. Thymic ectopia	No
10	None	No
11	Right vesicoureteral reflux	No
12	None	No
13	None	No
14	None	No
15	Coccygeal agenesis	No
16	None	No
17	Right cryptorchidism. Small left periprostatic cyst	No
18	Mullerian remnant cyst	No

eratively. As for complications, there were no vaginal or urethral injuries, no wound infections and no dehiscences.

Throughout the 38 (12.75-45.50) months of follow-up there were no anal strictures identified. 12 (66%) patients did proactively undergo dilation of the anoplasty during the first 4 weeks after the surgery, without further need for dilation onwards. The final Hegar size was 13 (12-14). All patients are passing stool, 11 (61%) of them with the need of low doses stimulant or osmotic laxatives.

DISCUSSION

In this article we describe the experience in three tertiary centres with a recently described surgical technique[8], a posterior rectal advance anoplasty (PRAA). We indicate this surgical approach to treat patients with rectoperineal fistulas when the fistula is located at the anteriormost aspect of the sphincteric ellipse. With this technique we aimed to leave the anterior wall of the fistula intact, to decrease the chance of urethral or vaginal injury. Contrary to the classic limited PSARP, the most distal part of the rectum sometimes called fistulous tissue is left in place and the anus is placed within the sphincter complex. We describe 18 patients with good perioperative and short-term postoperative results.

Even though rectoperineal fistula are the most benign lesion in the spectrum of anorectal malformations, they are often associated with other anomalies and should undergo a full evaluation to rule out associated malformations, as more complex anorectal malformation would[9]. In our series there were 11 (61%) patients with some sort of associated malformation, of which 3 (16%) were VACTERL association, a syndrome with an approximate incidence of 1 in 10,000 to 1 in 40,000 live-born infants[10].

Surgical decision making in patients with anorectal malformations depends on the type of defect. For rectoperineal fistula, neonatal primary repair with a PSARP is the option of choice[1]. Early diagnosis and treatment, especially given its benign nature, is important to avoid complications. When the rectoperineal fistula is diagnosed after the neonatal period, as occurred in 61% of our patients, a late diagnosis can lead to the development of megarectum or, in a more serious scenario, it can also present with abdominal distension, constipation and faecal impaction, fever, vomiting, dehydration, and sepsis[11]. In addition to these clinical complications, the surgical approach also entails higher risks. In these patients, PSARP is a demanding procedure with a high risk of injury to the neighbouring structures, whose repair should be protected with a protective colostomy. In such cases a moist perineum, contamination of the wound with faeces, nonsterile mucus and urine create an environment for a higher risk of complications such as wound infection and dehiscence[12].

In addition to the most widely used PSARP, there are a number of surgical approaches described for the treatment of ARM. The cutback anoplasty described by Bowne and intended for shot-gun perineum in females and external ectopic anus in males consists of a posterior incision of the fistula into the normal position of the anal opening, without suture of the bowel to the skin[13]. This leaves the patient without any sphincter muscle around the anterior aspect of the anal opening and an inadequacy of the perineal body in females. For this reason, Potts described the transfer anoplasty, involving the dissection of the fistula through a posterior curvilinear incision and displacing the rectum posteriorly and suturing it to the skin through a separate incision overlying the sphincter muscle[14]. There is a further cutback modification, similar to the one described by Halleran, but with a different approach to the skin incision, with the creation of a posterior cutaneous flap[4].

With these two approaches in mind, Halleran et al proposed the PRAA for perineal fistula located in the anterior aspect of the sphincter complex[8]. The technique is as we have described in our methods and is intended to create an anal opening at the centre of the sphincter complex without the need for an anterior rectal dissection and its associated risk of urethral and vaginal injury. The incidence of such injuries during the dissection of a long rectoperineal fistula adherent to the vagina or urethra has been reported as high as 5%[15], as opposed to the PRAA which in both Halleran et al and our series had no such complication. The PRAA is therefore a less invasive and a simpler technique, as well as being quicker to perform. In both Halleran et al and our series all cases were completed in under 60 minutes. Regarding the indication for PRAA, we must emphasise that it is not indicated in cases of anal stenosis (funnel anus like) associated with Currarino syndrome. In these cases, the surgical technique of choice would be a classical posterior sagittal incision with a modified anoplasty preserving half of the anal canal as described by Lane et al.[16].

Another advantage of the PRAA is the preservation of the distal rectum, which some authors have linked to an improved bowel function outcome. Ruttenstock et al showed the presence of functional anal structures within the fistula in a series of patients who underwent preoperative rectal manometry of rectoperineal or rectovestibular fistula, with no significant differences in the postoperative manometry after undergoing a complete transposition of the fistula[7]. Despite this, in our series 27% of patients are receiving low doses of stool softeners.

Although PRAA expectedly minimizes the risk of postoperative stricture by reducing the disruption of blood supply to the distal rectum and by performing a non-circumferential incision[8], some of the patients in our series underwent dilation of the anoplasty. In those cases, dilations were provided as a routine (based on centre and/or surgeons protocol) but without any actual anal strictures. This routine practice of postoperative dilations has been

called into question recently in a randomised controlled trial[17]. Given that there is no incision other than the anoplasty, there is also a reduced risk of wound infection and dehiscence[8].

Early diagnosis and surgical treatment is of great importance in the management of anorectal malformations. PRAA could become the option of choice for selected patients with rectoperineal fistula with the anterior aspect within the sphincter complex. The technique has shown a short surgical time and hospital stay, with good results and low risk of injury to neighbouring structures. Long-term follow up is still pending to determine the continence outcome of these patients but the results are promising.

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