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Unexpected megarectum: A potential hidden source of complications in patients with anorectal malformation

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ABSTRACT

Background: Primary posterior sagittal anorectoplasty is recommended to repair anorectal malformations with rectoperineal or rectovestibular fistula. The aim of this study was to identify the impact of the presence of megarectum on the relative frequency of complications related to posterior sagittal anorectoplasty.

Methods: We performed a cross-sectional retrospective study including patients with anorectal malformation, preoperative rectogram and surgically treated with primary or staged posterior sagittal anorectoplasty. Only complications related to anorectoplasty were analyzed and compared with the presence of megarectum.

Results: Thirty patients aged 1 day to 7 years were included, 60% had megarectum. Sixteen patients had primary repair: 6 with megarectum and 10 without megarectum; complications occurred in four of the six with megarectum, 66.7%, and no complication were observed in the 10 patients without megarectum ($F p = 0.008$). Fourteen patients had staged repair and no complications related to posterior sagittal anorectoplasty occurred in these patients.

Conclusions: Comprehensive preoperative evaluation in patients with anorectal malformation with rectoperineal or rectovestibular fistula could include a rectogram. Awareness of the presence of megarectum could be useful information in the decision to create a colostomy or perform a primary posterior sagittal anorectoplasty.

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1. Background

Guidelines for surgical decision making in patient with anorectal malformation during the neonatal period depend on the type of defect; a posterior sagittal anorectoplasty without colostomy (primary repair) is recommended for newborns with rectoperineal fistula or rectovestibular fistula instead of colostomy [1–3]. A successful primary repair reduces the number of surgical procedures and cost, and provides better cosmetic results, compared when these malformations are surgically treated with three operations: colostomy, posterior sagittal anorectoplasty and colostomy closure (staged repair).

Some patients born with anorectal malformation with rectoperineal fistula or rectovestibular fistula are diagnosed days or months after birth, or even years later, and although primary repair is feasible at any age, a moist perineum, contamination of the wound with feces, nonsterile mucus and urine are risk factors for wound infection, dehiscence, or both. There are no generally accepted guidelines for patients with rectoperineal fistula or rectovestibular fistula diagnosed after the

neonatal period, and a different constellation of complications can occur after a primary repair [4–7] therefore, the decision to perform primary repair or protective colostomy becomes a challenge.

In our experience, while performing primary posterior sagittal anorectoplasty in some patients with rectoperineal fistula or rectovestibular fistula, a very dilated and hypertrophic rectum (megarectum) was an unexpected finding, even in some neonates. In these patients, primary posterior sagittal anorectoplasty became a technically demanding procedure, with an increased risk of injury to urethra and vagina, and a greater susceptibility to dehiscence and wound infection. After an analysis of these patients in our morbidity and mortality session of December 2010, we decided to investigate preoperatively the presence of megarectum, thus we began in January 2011 to include in our preoperative work up a contrast radiologic study of the rectum.

The purpose of the present study was to determine the complication rate related to the posterior sagittal anorectoplasty in patients with anorectal malformation with rectoperineal fistula or rectovestibular fistula, and correlate the complications with the presence of a megarectum.

2. Methods

A cross-sectional retrospective study approved by our institutional review board was performed. We include all patients with rectoperineal fistula and rectovestibular fistula surgically treated with posterior

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sagittal anorectoplasty [8] admitted from January 2011 to December 2013 at the colorectal centers of two institutions (*Hospital para el Niño Poblano, Puebla, Mexico* and *Instituto Nacional de Pediatría, Mexico City, Mexico*). Since January 2011 in our institutions all patients with these type of anorectal malformations have a preoperative rectogram made when the malformation is diagnosed. All patients with primary repair were submitted to our standard postoperative protocol, which is focused on keeping the wound clean and dry for 7 to 10 days. The protocol consists of following components: clean the wound as needed; gentle padding of the suture line with wet gauze; no scrubbing of the suture line, nothing by mouth, total parenteral nutrition, intravenous antibiotics and urinary catheter.

Rectogram was performed using water-soluble iodinated contrast and a 5Fr or 8Fr regular feeding tube. Contrast was administered through the fistula. To avoid false rectal dilatations from high pressure, the contrast was administered using hand injection. Megarectum was defined when a rectopelvic ratio was greater than 0.61 [9,10] (Fig. 1).

Gender, age at diagnosis of the anorectal malformation, type of anorectal malformation, type of repair (primary or staged), presence of megarectum, and complications related to the sagittal approach in the following 30 postoperative days were recorded. Three pediatric colorectal surgeons performed all the procedures and the decision to perform primary or staged repair was based on their own clinical judgment.

Complications were correlated with the type of repair and the presence of megarectum. Statistical analysis was done with the Statistical Program for the Social Sciences (SPSS®) version 20.0, and included descriptive statistics for the demographic variables and exact Fisher's exact test for the categorical variables.

3. Results

Thirty patients were included, 17 girls and 13 boys. Age at diagnosis of the anorectal malformation ranged from 1 day to 7 years, median 2 months. Twelve patients had rectovestibular fistula and eighteen had rectoperineal fistula. None of the patients had an associated malformation making it necessary to perform a colostomy.

3.1. Patients with megarectum at diagnosis

Eighteen patients (60%) had megarectum at the time of diagnosis, 11 had rectoperineal fistula, and 7 had rectovestibular fistula. Six patients had primary repair and twelve had staged repair. Four patients with megarectum and primary repair had complications (66.7%): three had partial dehiscence of the anorectoplasty that were resolved with local wound care, and one patient had total dehiscence of the anorectoplasty

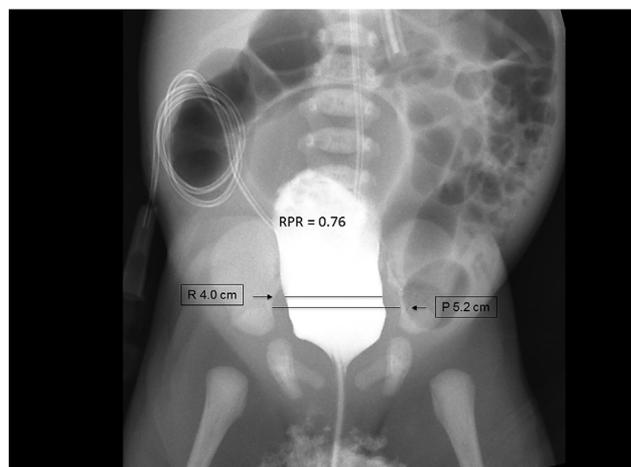


Fig. 1. Megarectum in a newborn girl with rectovestibular fistula. RPR = R/P (16,17). RPR: rectopelvic ratio, R: diameter of the rectum, P: maximal pelvis diameter.

Table 1

Characteristics of 18 patients with anorectal malformation and associated megarectum.

Type of anorectal malformation	Age at diagnosis (months)	Rectopelvic ratio	Type of repair	Complication
Perineal	0	0.7	Staged	
Perineal	0	0.73	Staged	
Perineal	9	1.12	Staged	
Perineal	8	0.68	Staged	
Perineal	13	0.61	Staged	
Perineal	2	1.29	Staged	
Perineal	16	0.67	Staged	
Vestibular	0	0.62	Staged	
Vestibular	3	0.76	Staged	
Vestibular	0	0.76	Staged	
Vestibular	0	0.76	Staged	
Vestibular	1	1	Staged	
Perineal	0	1	Primary	
Perineal	36	0.68	Primary	PRESENT
Perineal	2	0.73	Primary	
Perineal	9	0.68	Primary	PRESENT
Vestibular	2	0.73	Primary	PRESENT
Vestibular	6	0.71	Primary	PRESENT

requiring a diverting colostomy. No complications were observed in the 12 patients with staged repair (Table 1).

3.2. Patients without megarectum at diagnosis

Twelve patients (40%) did not have a megarectum at the time of diagnosis of the anorectal malformation. Seven of these patients had rectoperineal fistula and five had rectovestibular fistula. Ten of these 12 nonmegarectum patients had a primary repair, and two patients had a staged repair. No complications were observed in any nonmegarectum patients (Table 2).

3.3. Primary or staged repair

Sixteen patients had a primary repair, six of whom had an associated megarectum, and ten of whom did not have an associated megarectum; complications were observed only in 4 patients with an associated megarectum ($F p = 0.008$) (Table 3). Fourteen patients had a staged repair, 12 with megarectum and 2 without megarectum. No complications related to the posterior sagittal anorectoplasty were observed in these patients.

4. Discussion

Standard guidelines for surgical decision making in newborns with anorectal malformation with rectoperineal fistula or rectovestibular fistula would recommend primary repair [1–3]. Primary posterior anorectoplasty possesses great advantages compared with the staged

Table 2

Characteristics of 12 patients with anorectal malformation and without associated megarectum.

Type of anorectal malformation	Age at diagnosis (months)	Rectopelvic ratio	Type of repair
Perineal	0	0.58	Staged
Vestibular	3	0.59	Staged
Perineal	0	0.58	Primary
Perineal	0	0.41	Primary
Perineal	0	0.60	Primary
Perineal	84	0.47	Primary
Perineal	7	0.57	Primary
Perineal	5	0.59	Primary
Vestibular	0	0.57	Primary
Vestibular	0	0.56	Primary
Vestibular	0	0.58	Primary
Vestibular	8	0.38	Primary

Table 3

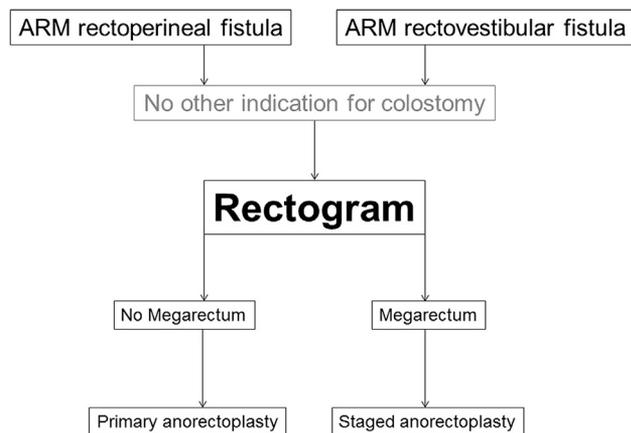
Comparison of frequency of complications related to posterior sagittal anorectoplasty, and the presence of an associated megarectum in 16 patients with primary repair.

		Complications	
		Yes	No
Megarectum	Yes	4 ^a 66.7%	2 ^b 33.3%
	No	0	10
			100%
Fisher's exact test p = 0.008			

^a Age of primary repair and type of malformation: 2 months (vestibular), 6 months (vestibular), 9 months (perineal) and 3 years (perineal).

^b Age of primary repair and type of malformation: 1 day (perineal) and 2 months (perineal).

repair: reduces the number of operations from three to one, it avoids the inherent risks of two operations, the cost is much less and provides the best cosmetic results. Good anatomical postoperative results conserve the sphincter mechanisms that are usually adequate in these types of defects. However, different circumstances such as the specific characteristics of this wound (location, risk of contamination with urine, mucus and feces not sterile, and humidity) can facilitate the development of wound complications resulting in some case in anatomical damage to the sphincters, anal strictures, acquired atresias, misalignment of the anus, and recurrent fistulas requiring reoperation [11,12].



ARM. Anorectal malformation
Megarectum is defined with a RPR > 0.61

Fig. 2. Decision algorithm for patients with anorectal malformation with rectoperineal or rectovestibular fistula used in the Colorectal Center of Mexico.

In the present study, we found that an associated megarectum in primary repairs can be other factor that increases risk of complications of the wound.

Levitt reported a 58% rate of constipation and an 83% rate of fecal control in patients with rectoperineal fistula, and a 55% rate of constipation and a 64% rate of fecal control in patients with a rectovestibular fistula [2]. However, postoperative complications related to posterior sagittal anorectoplasty could clearly worsen these functional outcomes [11–15]. So, the decision as to creating most important question is whether to create a colostomy or performing a primary repair is very important. Colostomy can be considered in the presence of an associated presacral mass, any major malformation in the pelvis, such as vaginal agenesis, and megarectum (Fig. 2). Comprehensive preoperative evaluation may include a rectogram to assess the size of the rectum to detect preoperatively an associated megarectum that seems to be useful in deciding whether to create a colostomy or offer a primary repair, particularly in cases with delay diagnosis. A clinical trial will provide solid data on this hypothesis.

References

- [1] Levitt M, Peña A. Imperforate anus and cloacal malformations. In: Holcomb GW, Patrick JM, Ostlie DJ, editors. *Ashcraft's of Pediatric Surgery*. Philadelphia, PA: Elsevier Saunders; 2014. p. 492–514.
- [2] Levitt MA, Peña A. Anorectal malformations. In: Coran A, Grosfeld JL, O'Neill JA, Fonkalsrud EW, editors. *Pediatric Surgery*. Philadelphia, PA: Elsevier Saunders; 2012. p. 1289–309.
- [3] Pakarinen MP, Rintala RJ. Management and outcome of low anorectal malformations. *Pediatr Surg Int* 2010;26:1057–63.
- [4] Eltayeb A. Delayed presentation of anorectal malformations: the possible associated morbidity and mortality. *Pediatr Surg Int* 2010;26:801–6.
- [5] Kumar B, Kandpal DK, Sharma SB, et al. Single-stage repair of vestibular and perineal fistulae without colostomy. *J Pediatr Surg* 2008;43:1848–52.
- [6] Kuijper CF, Aronson DC. Anterior or posterior sagittal anorectoplasty without colostomy for low-type anorectal malformation: how to get a better outcome? *J Pediatr Surg* 2010;45:1505–8.
- [7] Sham M, Singh D, Phadke D. Anorectal malformations: definitive management during and beyond adolescence. *J Indian Assoc Pediatr Surg* 2012;17:120–3.
- [8] Peña A, deVries PA. Posterior sagittal anorectoplasty: important technical considerations and new applications. *J Pediatr Surg* 1982;17:638–43.
- [9] Meunier P, Louis Dominique JM. Physiologic investigation of primary chronic constipation in children comparison with the barium enema study. *Gastroenterology* 1984;87:1351–7.
- [10] Van Der Plas RN, Benninga MA, Staalman CR, et al. Megarectum in constipation. *Arch Dis Child* 2000;83:52–8.
- [11] Peña A, Grasshoff S, Levitt M. Reoperations in anorectal malformations. *J Pediatr Surg* 2007;42:318–25.
- [12] Chan KWE, Lee KH, Wong HYV, et al. Outcome of patients after single stage repair of perineal fistula without colostomy according to the Krickenbeck classification. *J Pediatr Surg* 2014;49:1237–41.
- [13] Griffin SJ, Parkinson EJ, Malone PSJ. Bowel management for paediatric patients with faecal incontinence. *J Pediatr Urol* 2008;4:387–92.
- [14] De Blaauw I, Midrio P, Breech L, et al. Treatment of adults with unrecognized or inadequately repaired anorectal malformations: 17 cases of rectovestibular and rectoperineal fistulas. *J Pediatr Adolesc Gynecol* 2013;26:156–60.
- [15] Wakhlu A, Kureel SN, Tandon RK, et al. Long-term results of anterior sagittal anorectoplasty for the treatment of vestibular fistula. *J Pediatr Surg* 2009;44:1913–9.