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Anorectal malformation with rectobladder neck fistula: A distinct and challenging malformation

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ABSTRACT

Background: Rectobladder neck fistula is the highest and most complex anorectal malformation in boys and the only one that requires an abdominal approach, open or laparoscopic, for repair. The aim of this study was to describe the unique characteristics of rectobladder neck fistulas that warrant special attention and to describe the associated anatomic variants in the genitourinary tract.
Methods: The database of a tertiary medical center was retrospectively reviewed for all patients treated for rectobladder neck fistula, by our team in 1980–2011. Data on surgical history, associated and functional defects, treatment and outcome were collected by chart review.
Results: The study group included 111 patients. The most common anatomic urologic defect was a single kidney in 37 patients (33.3%) and the most common functional urologic defect was vesicoureteral reflux in 40 patients (36%), including 11/37 patients with a single kidney (29.7%). Of the 40 patients who underwent cystoscopy, 16 (40%) had a higher than normal location of the verumontanum. Follow-up ranged from 2 to 290 months (median 59). Urinary continence was achieved in 40 of the 61 patients (65.5%) for whom data were available, and fecal continence was achieved in 9 of the 69 patients (13%) for whom data were available. A sacral ratio of 0.4 or less was associated with lower rates of urinary control (23%) and fecal control (0%), relative to higher ratios. Twenty stomas (18%) were found to be located too distally, limiting the availability of the bowel for a pull through.
Conclusions: Rectobladder neck fistula carries a poor prognosis for bowel control and is associated with a high rate of urinary malformations that require long-term care. Pediatric surgeons need to be aware of these complications in order to provide proper treatment and parental counseling. Intra-vesical verumontanum is found in a surprisingly high percentage of patients. The combination of a single kidney with vesicoureteral reflux is common and should be closely followed to avoid renal deterioration. Special attention should be given to colostomy construction to avoid complications and unnecessary procedures. A sacral ratio of 0.4 or less is an indicator of poor fecal and urinary control.

Anorectal malformations comprise a wide spectrum of congenital defects ranging from those with an excellent functional prognosis, to complex, and difficult-to-manage anomalies, often associated with other malformations, and a poor functional prognosis [1]. The traditional classification, still in widespread use, categorizes anorectal malformations as low, intermediate, or high depending on the position of the distal rectum in relation to the levator muscle and pelvic floor [2]. However, this method fails to distinguish among different types of defects, which may be included in the same category even though they require different surgical approaches and, more importantly, have a different functional prognosis. Therefore, a more practical classification has been proposed, based on the anatomic features of the malformations [3] with implications for the surgical approach and long-term outcome. Rectobladder neck fistula is the "highest" and most complex anorectal malformation in boys. It is imperative to identify this defect as a special and separate entity since it carries a poor functional prognosis for bowel control and is the only defect in the spectrum of anorectal malformations in boys that requires an abdominal approach for repair. Pediatric surgeons need to be alert to the high rate of associated urologic malformations and coordinate care of these patients with the pediatric urologist.

The aim of this study was to describe the distinct characteristics of anorectal malformations with rectobladder neck fistula that warrant

Abbreviations: ASD, atrial septal defect; CIC, clean intermittent catheterization; PSARP, posterior sagittal anorectoplasty; SR, sacral ratio; VBM, voluntary bowel movements; VM, verumontanum; VUR, vesicoureteral reflux.

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Table 1
Urologic and non-urologic anomalies in patients with rectobladder neck malformation*.

Anomaly	No. of patients	Anomaly	No. of patients
Associated urologic malformations		Associated gastrointestinal anomalies	
Vesicoureteral reflux	40	EA	15
Absent kidney	37	Malrotation/Incomplete rotation	4
Hydronephrosis	33	Meckle diverticulum	2
Undescended testis	30	Congenital short colon with cystic dilatation	1
Bifid scrotum	29	Ileocolic atresia	1
Hypospadias	20	Annular pancreas	1
Intravesical verumontanum	16	Pyloric stenosis	2
Urethral stenosis	14		
Neurogenic bladder-congenital	12	Sacrovertebral defects	
Megaureter	10	Hemivertebra	25
Megaurethra	7	Tethered cord	17
UVJ obstruction	6	Missing sacral vertebrae	9
Double collecting system	4	Syrinx	4
Horseshoe kidney	3	Hemivertebrae	25
UPJ obstruction	2	Other	5
Penoscrotal transposition	2		
Paraureteral diverticulum	1	Extremity anomalies	
Bladder diverticulum	1	Polydactyly	5
Associated cardiac anomalies		Equino varsus	4
Patent ductus arteriosus	12	Abdominal wall defects	
Ventricular septal defect	10	Prune belly syndrome	3
Atrial septal defect	9	Omphalocele	1
Patent foramen ovale	5		
Tetralogy of Fallot	3		
Pulmonic stenosis	2		
Tricuspid regurgitation	2		
Other	7		
Other vascular anomalies			
Single umbilical artery	5		
Duplication of inferior mesenteric artery	1		

* Some patients had more than one.

special attention and to describe the associated anatomic variants in the genitourinary tract.

1. Materials and methods

The database of a tertiary medical center was retrospectively reviewed for all patients treated for rectobladder neck fistula from December 1980 to August 2011.

Data were collected by chart review as follows: associated malformations, type and characteristics of the colostomy created at birth, type of main repair surgery, and outcome. Sacral ratio was calculated as described by Peña [3].

Findings were analyzed by descriptive statistics and are represented as medians and means.

This study was approved by the local institutional review board (approval no. 2009–0905).

2. Results

The study cohort included 111 patients with rectobladder neck fistula, of whom 98 underwent primary repair by our team and 13 underwent reoperation after initial surgery performed elsewhere.

The patients accounted for 10% of the total 1069 male patients operated on by the authors for anorectal malformation during the study period. The duration of the follow-up ranged from 2 to 290 months (median 59).

2.1. Associated urologic malformations

An anatomic or functional urologic malformation was identified in 99 patients (Table 1) (89.1%). The most common associated anatomic malformations was a single kidney, in 37 (33.3%) of the patients, followed by hydronephrosis, in 33 patients (29.7%), including 9 with single kidney (27.2%). The most common functional disorder was vesicoureteral reflux (VUR), in 40 patients (36%), including 11/37 patients with a single kidney (29/7%) and 16/33 patients with hydronephrosis (48.4%). VUR usually occurred on the left side; no side dominance was noted for a single kidney. Other anatomic urologic malformations were undescended testis (UDT) in 30 patients (27%), twice as common on the left side and bifid scrotum, in 29 patients (26.1%). Forty-two of the 99 patients with urologic malformations (42%) had also non-urologic anomalies.

2.2. Ectopic verumontanum

In 16 patients (14.4%), the VM was located in a higher-than-normal anatomic position: at the trigone in 5 (31.2%) patients, bladder neck in 9 (56.2%)) and immediately distal to bladder neck in 2 (12.5%). This group accounted for 40% of the patients who underwent cystoscopy (which was routinely performed in cases operated on since 2001). Patients age at the time of follow up ranged from 1 to 26 years (mean 5.5 years). Three (18.7%) patients had episodes of orchioepididymitis during follow-up. Three patients had reached puberty by the time of the last clinic visit. One complained of severe pain during ejaculation as well as episodes of prostatitis and epididymitis, and another presented at age 24 years with failure to ejaculate during intercourse, which was subsequently attributed to an ectopic VM at the trigone.

2.3. Associated non-urologic malformations

Forty-seven patients (42.3%) had non-urologic anomalies, of whom 23 patients (20.7%) had more than one associated anomalies (Table 1). Twenty-nine (26%) had cardiac anomalies, mainly patent ductus arteriosus, ventricular septal defect, arterial septal defect, and patent forman ovale. Five of the patients with vascular anomalies had a single umbilical artery; 2 of them had also kidney agenesis. Gastrointestinal malformations occurred in 25 (22.5%) patients, including esophageal atresia in 15.

Table 2
Technical modifications to correct problems caused by a too-distal colostomy.

Surgical modification	No. pts
Mucous fistula mobilization	9
–Mucous fistula closed as a Hartmann's pouch	6
–Mucous fistula taken down and relocated	2
–Mucous fistula taken down and anastomosed to a proximal colonic segment (colostomy closed) to reach perineum	1
Pull-through of colostomy with resection of distal bowel	3
Laparoscopic procedure converted to open	1
Revision of stoma before PSARP	4

PSARP, posterior sagittal anorectoplasty.

2.4. Sacro-vertebral anomalies

The most common sacro-vertebral anomaly was hemivertebra, found in 25 patients: 10 sacral, 6 lumbar, 6 thoracic, 1 cervical and 2 unspecified (Table 1). The average sacral ratio, calculated from the available data on 95 patients, was 0.59 (range 0–1). Tethered cord was identified in 17 of the 52 patients (32.6%) who underwent screening for this condition.

2.5. Colostomy opening

All patients but one underwent colostomy at birth, at other institutions. In the outstanding patient, the operative report said the surgeons had created a colostomy but in fact they created an ileostomy. In 20 patients (18%), the stoma was located too distally, limiting the availability of the bowel for a pull through. Four of them underwent revision before the main repair and 13 required special technical maneuvers during the main repair. These included mobilization of the mucous fistula to a lower location in the abdominal wall or closure as a Hartmann's pouch (9), resection of an exceptionally short piece of distal bowel and pull-through of the proximal colostomy, with loss of the rectum (3), and conversion of a laparoscopic to an open procedure because of a short distal bowel segment (1 patient) (Table 2).

2.6. Surgical approach

The main repair in the 98 patients operated at our institution consisted of posterior sagittal anorectoplasty (PSARP) with laparotomy (80 patients), laparoscopic assisted PSARP (8 patients) and PSARP alone (10 patients). Three of the laparoscopic procedures were converted to laparotomy to achieve better rectal mobilization, perform rectal tapering or define the vascular supply (1 patient each). The other 13/111 patients underwent primary surgery at other institutions and were reoperated at ours. The procedures performed and indications for reoperation are described in Table 3.

2.7. Functional results

The functional results were assessed by the operating surgeon at the last clinic visit. Of the 98 patients who underwent primary surgery at our institution, 12 were lost to follow-up and 13 were less than 3 years old and were excluded from continence evaluation as we consider this age the time of toilet training. There were no data in the medical charts on fecal control for 3 patients and for urinary control, for 11 patients. In one patient, the colostomy was still open at the parent's request owing to poor functional prognosis. One patient had a vesicostomy.

2.7.1. Fecal control

Data on fecal control were available for 69 patients. Nine patients (13%) had fecal control, defined as voluntary bowel movements without soiling; 8 (11.5%) had partial fecal control, defined as voluntary bowel movements with soiling; and 52 (75.3%) had no fecal control (incontinence) defined as absence of voluntary bowel movements. The patients without fecal control required bowel management with enemas.

2.7.2. Urinary control

Data regarding urinary control were available for 61 patients. Forty patients (65.5%) had full urinary control, both diurnal and nocturnal; 3 (4.9%) had partial control, defined as spontaneous voiding with dribbling between voiding episodes; and 18 (29.5%) had no urinary control and required clean intermittent catheterization. Of the 13 patients who underwent reoperation, 11% achieved fecal control and 23%, urinary control.

2.8. Relationship between sacral ratio and outcome

Sacral ratio (SR) was calculated as described by Peña [3] and the results were divided into 3 groups: 0–0.4, 0.41–0.69 and ≥0.7 (normal).

2.8.1. Fecal control

Of the 69 patients for whom we had data on fecal control, 12 had SR of 0–0.4, 26 had a ratio of 0.41–0.69, and 21 had a ratio of ≥0.7. In 10 patients, the ratio could not be calculated. Of 12 patients with SR 0–0.4;

Table 3
Reoperations of primary repair procedures in patients with rectobladder neck fistula.

	Primary procedure	Indication for reoperation
1	PSARP	Bladder was identified as rectum and pulled down
2	PSARP with laparotomy	Massive prolapse and posterior mislocation of anus
3	Abdominoperineal pull through	Rectal stenosis
4	PSARP	Rectal stenosis and massive megasigmoid
5	PSARP with laparotomy	Anteriorly mislocated anus and a prolapse
6	PSARP with laparotomy	Anteriorly mislocated anus and a prolapse
7	Laparotomy, opening of transverse colostomy, dividing the fistula from bladder neck and resection of distal colon	Pull through of the end colostomy
8	PSARP with laparotomy	Severe rectal prolapse
9	Attempted PSARP with laparotomy	Surgeons unable to find the rectum, closed the PSARP incision and entered through abdomen
10	Attempted PSARP	Surgeons unable to pull rectum down, thought they closed the recto bladder neck fistula but this was found to be persistent in the redo.
11	Attempted PSARP with laparotomy	Surgeons entered posterior sagittal and were unable to pull rectum down as it was found to be very high. They continued through the abdomen, took down the mucous fistula and were then able to pull through the rectum. Pathology specimen showed that vas deferens and seminal vesicles tissues were resected. The patient developed anal stricture.
12	Attempted aborted PSARP	Surgeons unable to find the rectum and closed incision
13	Attempted PSARP with laparotomy	Surgeons closed the recto bladder neck fistula but were unable to pull rectum down

PSARP, posterior sagittal anorectoplasty.

none had fecal control. Of 26 patients with SR 0.41–0.69; 2 (7.6%) had fecal control, 3 (11.5%) partial fecal control, and 21 (80.7%) no fecal control. Of 21 patients with SR ≥0.7; 7 (33.3%) had fecal control, 3 (14.2%) partial fecal control and 11 (52.3%) no fecal control.

2.8.2. Urinary control

Of the 61 patients for whom we had data on urinary control, 13 had SR of 0–0.4, 22 had SR of 0.41–0.69, and 19 had a ratio of ≥0.7; in 7 patients, the ratio could not be calculated. Of 13 patients with SR 0–0.4; 3 (23%) had urinary control, 1 (7.6%) partial control and 9 (69.2%) were on CIC. Of 22 patients with SR 0.41–0.69; 18 (81.8%) had urinary control, and 4 (18.2%) were on CIC. Among 19 patients with SR ≥0.7; 13 (68.4%) had urinary control, 2 (10.5%) partial control and 4 (21%) were on CIC.

3. Discussion

This study of patients with rectobladder neck fistula revealed a very high rate (89.1%) of associated genitourologic anomalies. This value is close to the 92% reported in our earlier study in a smaller group of patients [4]. Left undiagnosed and untreated, urologic anomalies can lead to significant upper urinary tract complications [5,6] and are responsible for much of the lifelong morbidity of these patients. Therefore, careful investigation is needed during the initial work-up as well as expertise and experience in urologic management. All patients with anorectal malformations should undergo early kidney ultrasound to evaluate renal status and identify obstruction. When there is evidence of a dilated upper urinary tract or renal scarring, a voiding cystourethrogram is recommended to delineate the anatomy of the urinary system and asses the presence of VUR [7,8].

An unexpected finding of our study was an intravesical VM among 40% of the patients who underwent cystoscopy. Our search of the literature failed to yield prevalence data on ectopic verumontanum in rectobladder neck fistula. We suspect that the actual prevalence of this anomaly was likely even higher, since it did not include patients in whom cystoscopy was not performed. This anomaly may manifest as ejaculation directly into the bladder. This is distinct from the retrograde ejaculation reported by some in patients with anorectal malformations [9] which was attributed in part to congenital neurologic damage affecting the erectile center and manifesting in incomplete closure of the bladder neck. Patients present with complaints of failure to ejaculate, pain on ejaculation or recurrent prostatitis. Another potential consequence is infertility. Of the 16 patients in our study with ectopic verumontanum, only 3 had reached puberty at the last follow-up. Assisted reproduction techniques such as sperm retrieval from urine followed by intra-uterine insemination, in-vitro fertilization, and intracytoplasmic sperm injection, should be discussed with the patients.

Another common urologic malformation associated with rectobladder neck fistula is bifid scrotum. Our experience shows that the higher the malformation, the more likely the finding of bifid scrotum. Therefore, bifid scrotum may serve as a sign for the diagnosis of a rectobladder neck malformation.

During opening of a colostomy in a patient with anorectal malformation, accurate placement of the stomas is essential to prevent unnecessary complications. We advocate forming divided descending colostomies, placing the proximal stoma at the fixed part of the descending colon as it emerges from the retroperitoneum [10] and leaving enough distal bowel to the pull through. Opening the colostomy in a too-distal location, a common error found in 18% of our patients, leads to technical difficulties at the time of PSARP and the need for surgical modifications, such as mobilizing the mucous fistula, changing the surgical approach, and more critically, pulling through the proximal colostomy with loss of the rectum and consequent reduction in the potential for bowel control (Fig. 1).

Knowing the prognosis for fecal and urinary control is important in order to properly counsel parents already from the newborn period. Our data show that rectobladder neck fistula is associated with poor functional prognosis: only 13% of our patients achieved full fecal control and 11.5%, partial control. Urinary control was achieved by 65.5% of patients. The prognosis is even worse in patients who require secondary procedures following failed repairs. Only 11% of this subgroup in our study achieved fecal control, and 23% achieved urinary control. We also found that incontinence correlates with the sacral ratio. None of the patients with a sacral ratio of 0.4 or less had fecal control. Thus, surgeons need to adjust parental expectations, at birth, and encourage them to plan for a bowel management program at the age of toilet training. Studies report success with keeping children artificially cleans with enemas [11,12]. Regarding urinary continence, patients with a sacral ratio of 0.4 and less had only a 23% likelihood of urinary control, but the prognosis for patients with a ratio above that was relatively good.

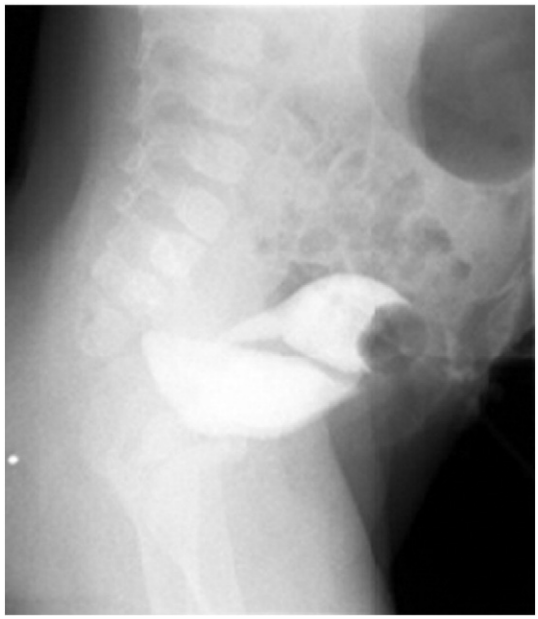

Fig. 1. Distal colostogram of a patient with recto bladder neck fistula, showing a colostomy that was opened too distal at birth. As a result the bowel segment between the mucous fistula and the rectum was too short and could not be used for the anorectoplasty. In this case we had to pull through the proximal colostomy and therefore the rectum was lost.

Our experience has shown that in some cases in which fecal incontinence is expected, the patient nevertheless has voluntary bowel movements. We attribute this finding to a good bowel movement pattern combined with bowel conditioning by regular meals at regular intervals, so that patients have 1–2 formed bowel movements per day at a predictable time. This makes it possible for the parents to sit the child on the toilet at the time when he will most likely have a bowel movement. However, when bowel control is challenged, as in the event of diarrhea, the patient has no control.

For surgical repair, we recommend an abdominal approach (laparotomy or laparoscopy) with a posterior sagittal incision. Interestingly, in 10 of our patients, we were able to find and successfully pull the rectum down via a posterior sagittal approach, without entering the abdomen. These, however, were special cases in which the distal colostogram showed a very short sacrum, which allowed us to reach the area of the bladder neck from below and, using specific maneuvers, properly position the rectum. Nevertheless, we recognize that it is a rather risky approach for this type of malformation. We consider the laparoscopic approach to be particularly indicated for this type of defect [13].

In conclusions, rectobladder neck fistula is a challenging and distinct malformation with a high rate of associated urologic anomalies. It has a poor functional prognosis in terms of urinary and especially bowel control. A sacral ratio of 0.4 or less is indicative of poor fecal and urinary continence. Intravesical verumontanum is a frequent finding, and its implications are not yet fully understood. Close urinary follow up is mandatory, and particular attention should be addressed to correct colostomy construction.

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