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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.