



Ectopic ureters in anorectal malformations

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Abstract

Purpose The literature about ectopic ureters in anorectal malformations is limited. Repair of an anorectal malformation may require dissection near the normal or abnormal insertion of the ureters. Knowledge of the presence and location of ectopic ureters may prevent intraoperative injury. We aim to describe the incidence and location of ectopic ureters in patients with anorectal malformations and to characterize associated renal anomalies.

Methods This is an IRB-approved retrospective study of patients with anorectal malformations and ectopic ureters identified in our colorectal database.

Results Of 2283 patients with anorectal malformation, 79 (3.5%) had ectopic ureter(s). Of those, 29% had bilateral ectopic ureters. Nearly all (87%) of bilateral ectopic ureters occurred in females. Ectopic ureters most commonly inserted into the bladder neck (33%), vagina (15%), or urethra (13%). Renal dysfunction was noted in a high proportion of patients. The majority (59%) of ectopic ureters were associated with dysfunction of the ipsilateral kidney. Interestingly, 29% of patients with unilateral ureteral ectopia had an abnormal contralateral kidney. Only 22 patients (28%) had two normal kidneys, and 5 (6%) had documented renal failure with 2 (2.5%) requiring renal transplant. This compares to a transplant rate of 0.6% among anorectal malformation patients without ectopic ureter.

Conclusions The incidence of ectopic ureter is 3.5% among anorectal malformation patients. Cloaca and recto-bladder neck fistula are the types of anorectal malformation with higher incidence. Ureteral ectopia seems to confer an increased risk of renal failure. Identifying ectopic ureters is important for surgical planning and monitoring renal function vigilantly.

Keywords Ectopic ureter · Cloaca · Anorectal malformation · Hydronephrosis · Renal failure

Introduction

Up to 50% of children with anorectal malformations have associated genitourinary problems. The incidence and severity of urologic anomalies tend to correlate with the severity of the anorectal malformation [1–5]. One urologic anomaly that has not been well characterized is the ectopic ureter. An ectopic ureter inserts caudal to the normal location in the bladder trigone along the pathway of the developing mesonephric system. In boys, ectopic ureters may open into the bladder neck, posterior urethra, or mesonephric duct

derivatives (vas deferens, seminal vesicle, or epididymis). In girls, ectopic ureters may insert into the bladder neck, upper urethra, vagina, vestibule, or less commonly the cervix or uterus. The surgical repair of anorectal malformations requires dissection of the posterior urethra in males, or separation of the bladder from the vagina in females, thus suspecting ectopic ureters and knowing their common locations may prevent iatrogenic ureteral damage.

The purpose of this study is to characterize the incidence and location of ectopic ureters in patients with anorectal malformations. Furthermore, we aim to identify any predictive or prognostic factors to guide pre-operative counseling, surgical planning and postoperative management.

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Methods

This is an IRB-approved (19-0417) retrospective study of children with anorectal malformations and ectopic ureters identified within our colorectal database. This database, which comprises the records of patients operated on by the senior authors, has been prospectively maintained since 1980.

Results

Incidence of ectopic ureter

Of 2283 patients with anorectal malformations, 79 (3.5%) had ectopic ureter(s). Of those, 29% had bilateral ectopic ureters. Nearly all (87%) children with bilateral ectopic ureters were female. The highest incidence of ectopic ureter was seen in patients with more severe anorectal malformations,

most notably cloacae with long common channels (> 3 cm) and recto-bladder neck fistulae. Two-proportion Z test comparing patients with anorectal malformations with and without ectopic ureter(s) revealed that patients with long common channels or recto-bladder neck fistulae were indeed more likely to have an ectopic ureter. Other malformations were not statistically over-represented in the subset of patients with ectopic ureters (Table 1A).

Location of ureteral ectopia

Ectopic ureters in our cohort most commonly inserted into the bladder neck (33%), vagina (15%), or urethra (13%) (Table 2).

Associated renal anomalies

Renal dysfunction was noted in a high proportion of patients with ectopic ureter(s). The majority (59%) of ectopic ureters were associated with dysfunction of the ipsilateral kidney

Table 1 Anorectal malformations associated with ectopic ureter

Anorectal malformation	Ectopic ureter(s) (%)	Z statistic	p value
A			
Cloaca	32 (40.5)	2.88	0.004
Long (> 3 cm)	20 (25.3)	4.69	< 0.0001
Short (≤ 3 cm)	8 (10.1)	0.51	0.610
Unknown	4 (5.1)	0.24	0.810
Bladder neck fistula	8 (10.1)	1.97	0.049
Prostatic fistula	6 (7.6)	−1.00	0.317
Bulbar fistula	3 (3.8)	−1.96	0.050
Vestibular fistula	7 (8.9)	−1.44	0.150
No fistula	1 (1.3)	−1.31	0.190
Other	22 (27.9)	−0.03	0.976
Anorectal malformation			Incidence of ectopic ureter(s) (%)
B			
Cloaca			5.3
Long (> 3 cm)			8.9
Short (≤ 3 cm)			2.9
Unknown			3.8
Bladder neck fistula			6.7
Prostatic fistula			2.4
Bulbar fistula			1.2
Vestibular fistula			2.2
No fistula			1.1
Other			3.4

Bolded *p* values are statistically significant

A Of patients with ectopic ureter, classification by type of anorectal malformation and statistical comparison to patients without ectopic ureter

B Of all patients with an anorectal malformation, incidence of ectopic ureter by malformation type

Table 2 Location of ectopic ureter(s)

Laterality	N (%)	Insertion	N (%)
Right	29 (37)	Bladder neck	28 (33)
Left	21 (27)	Vagina	13 (15)
Bilateral	23 (29)	Urethra	11 (13)
Unknown	6 (8)	Caudally displaced	9 (11)
		Vas deferens	2 (2.4)
		Vestibule	2 (2.4)
		Uterus	1 (1.2)
		Other	7 (8.3)
		Unknown	11 (13)

(absent kidney 6%, dysplasia 20%, hydronephrosis 33%). Interestingly, 29% of patients with unilateral ureteral ectopia had an abnormal contralateral kidney (absent kidney 12%, dysplasia 7%, hydronephrosis 10%). Only 22 patients (28%) had two normal kidneys, and 5 (6%) had documented renal failure with 2 (2.5%) requiring renal transplant. This compares to a transplant rate of 0.6% among patients with anorectal malformation without ectopic ureters (z statistic 2.10, $p=0.036$) (Table 3).

Discussion

The high incidence of urologic anomalies in children with anorectal malformations has been known, albeit underappreciated, for half a century [5–7]. Higher rectal fistulae are associated with a higher incidence of urologic anomalies, with approximately 90% of patients with cloaca or recto-bladder neck fistula having some genitourinary anomaly [2]. A priori knowledge of the urologic anatomy is important for both surgical planning and preserving renal function.

Our findings demonstrate, perhaps unsurprisingly, that patients with more severe anorectal malformations are more likely to have ectopic ureters. Girls with long common channels and boys with recto-bladder neck fistulae are most at-risk for ectopic ureter. Nearly half of ectopic ureters inserted at the bladder neck or posterior urethra, a critical area of dissection rife with opportunity to damage urinary, ejaculatory, and erectile function. This is due to the lack of a natural

plane between the anterior rectum and posterior bladder and urethra, the confluence of the ejaculatory and urinary tracts, and the proximity of the prostatic nerve plexus. Iatrogenic injury to the genitourinary tract is described in the anorectal surgical literature [8–10]. Both meticulous dissection and a high index of suspicion are necessary to identify and preserve ectopic ureters intraoperatively as they are not reliably identified on renal ultrasound, cystoscopy or vaginoscopy. Cystoscopy demonstrating any finding other than two orthotopic ureteral orifices on the intertrigonal ridge should raise suspicion for an ectopic ureter, though apparently normal findings do not definitively rule out ectopic ureter.

On their own, ectopic ureters are a relatively rare congenital anomaly. In the absence of anorectal malformation, approximately 80% of ectopic ureters are associated with renal duplication. In children with anorectal malformations, single-system ectopic ureters are thought to be more common and can be associated with renal dysplasia or agenesis, hydronephrosis, and renal ectopia [5]. These findings should be noted on initial renal ultrasound, and any abnormality should prompt both a complete nephro-urological workup and a higher index of suspicion for ectopic ureter. High rates of kidney failure in the setting of severe anorectal malformations have been previously reported [11–13]. In the largest study dedicated to studying renal outcomes in cloaca patients, 50% of patients were noted to have chronic kidney disease with 17% progressing to end stage renal failure and 6% dying from renal disease [11]. Over 80% of these patients had underlying renal structural anomalies of the urinary tract. This serves as a reminder that nephro-urologic complications are a leading cause of death in patients with anorectal malformation, and that preserving and correcting urinary tract function is of utmost importance.

Limitations

There are limitations to our study that prevent us from drawing more detailed insights. Our database is intrinsically limited. Many older records are incomplete, inconsistent, and contain only abstracted information without full imaging reports, lab data, or operative notes. The study population represents complex patients seeking tertiary and quaternary

Table 3 Renal anomalies in anorectal malformation patients with ectopic ureter(s)

Kidney function	N (%)	Kidney with ectopic ureter	N (%)	Kidney with orthotopic ureter	N (%)
Bilateral normal	22 (28)	Normal	36 (41)	Normal	41 (71)
Solitary normal OR unilateral abnormal	34 (44)	Absent	5 (6)	Absent	7 (12)
Solitary abnormal OR bilateral abnormal	17 (22)	Dysplastic	17 (20)	Dysplastic	4 (7)
Renal failure and/or transplant	5 (6)	Hydronephrosis	29 (33)	Hydronephrosis	6 (10)

care, thus our findings may not be generalizable. Moreover, many patients who sought surgical care from the senior authors received their pre- and post-operative care locally, thus the patterns of care and quality of follow-up data are variable. It is quite likely that some patients went on to develop significant renal failure that was not captured in our database. Still, we feel this review of a large cohort of patients provides valuable characterization of an uncommon but important anatomic anomaly that is critical to recognize intraoperatively.

Conclusion

The overall incidence of ectopic ureter in patients with anorectal malformations is 3.5%. Girls with long common channel cloacae and boys with recto-bladder neck fistulae have a higher incidence. Surgeons operating on these patients must have a high index of suspicion for ectopic ureter. Ureteral ectopia seems to be associated with an increased risk of renal failure. Identifying ectopic ureters is important for surgical planning and patients with ectopic ureter must have their renal function monitored vigilantly.

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