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Spontaneous bladder rupture in non-augmented bladder exstrophy

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Summary

Objective
Bladder perforation is not commonly described in bladder exstrophy patients without bladder augmentation. The goal of this study was to identify the risk factors of spontaneous perforation in non-augmented exstrophy bladders.

Methods
The study was a retrospective multi-institutional review of bladder perforation in seven male and two female patients with classic bladder exstrophy–epispadias (E–E).

Results
Correction of E–E was performed using Kelly repair in two and staged repair in seven (Table). Bladder neck repair was performed in eight patients at a mean age of 6 years. Three patients had additional urethral surgery. Before rupture, six patients were voiding only per urethra. Two patients were voiding urethrally but were also performing occasional CIC via a Mitrofanoff. One patient was performing CIC 3 hourly per urethra. Six were dry during the day. Six of the patients had lower urinary tract symptoms: five had frequency and four were straining to void. Two had suffered episodes of urinary retention. Pre-rupture ultrasound showed that the upper urinary tract was dilated in four patients. Micturating cystourethrogram was performed in six showing vesico-ureteral reflux in five. Two had urethral stenosis. Nuclear medicine was done in three patients with two abnormal differential function. Urodynamics was performed in two patients with low capacity (100 mL) and hypocompliant (<10) bladders. Both had high leak point pressures: 60 cmH_2O at 100 mL. The mean age at rupture was 11 years, with a range of 5–20 years. Patients presented with abdominal pain, associated with signs of intestinal obstruction in seven and fever in two. Eight patients underwent laparotomy and one prolonged drainage via SPC. Simple closure was performed in seven and bladder neck closure in one, because of extension of the rupture inferiorly. All patients recovered well. Following rupture, five underwent augmentation and Mitrofanoff. One of these suffered a recurrent rupture. Two other patients refused augmentation and Mitrofanoff and one of these has since had a subsequent rupture.

Conclusions
The limitations of this series include the small number of patients and its retrospective nature, without knowledge of the incidence. Bladder rupture is a risk even in non-augmented bladder exstrophy. It is potentially life-threatening and most often requires laparotomy. Rupture occurs because of poor bladder emptying and/or high pressure. Urodynamics may identify those at risk. CIC with or without augmentation should not be delayed once poor bladder emptying and/or high pressure are identified.

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Table Patients with bladder perforation.

Pt	Pre-rupture imaging	Pre-rupture continence	Age, years	Management	Outcome
1	Small right kidney with high-grade VUR and high PVR	90 min voiding with episodes of retention	13	Laparotomy IDC 3 weeks	Refused augmentation and Mitrofanoff. Recurrent bladder rupture
2	Small left kidney (15% df) with high-grade VUR and high PVR. Poorly compliant bladder on urodynamics	Straining to void with frequency	8	Laparotomy and bladder neck closure with SPC	Augmentation and Mitrofanoff
3	Trabeculated bladder	Straining to void. High PVR. Incontinent	5	Laparotomy SPC 3 weeks	Refused Mitrofanoff. Ongoing straining and lost to f/u
4	Trabeculated bladder, low-grade VUR bilaterally and urethral stenosis	2—3 hourly voiding with straining. High PVR	20	SPC with prolonged drainage	No further surgery or recurrences known
5	Bilateral HUN	3 hourly voiding with high PVR	15	Laparotomy IDC 3 weeks	No further surgery or recurrences known
6	Normal USS	Voiding and dry on desmopressin and oxybutynin	11	Laparotomy IDC 3 weeks	Augmentation with Mitrofanoff
7	Bilateral HUN with high-grade VUR. Poorly compliant bladder on urodynamics	CIC 3 hourly	9	Laparotomy SPC	Augmentation with Mitrofanoff. Bladder stone post augment
8	Bilateral HUN with high-grade VUR and reduced function on left	Voiding with day and night wetting. Occasional CIC via Mitrofanoff	8	Laparotomy. Drainage via Mitrofanoff	Augmentation. Recurrent rupture near the anastomosis of the augment
9	Normal USS. Bladder capacity 100 mL. Low compliance	Voiding with day and night leaking. Occasional CIC via Mitrofanoff	10	Laparotomy. Drainage via Mitrofanoff	Augmentation, redo Mitrofanoff (Monti)

CIC, clean intermittent catheterization; HUN, hydroureteronephrosis; IDC, indwelling catheter; Pt, patient; PVR, post void residual; R, rupture; SPC, suprapubic catheter; USS, ultrasound; VUR, vesicoureteric reflux.

Introduction

Bladder rupture is a well-recognised complication following bladder augmentation in exstrophy but is uncommon in non-augmented patients [1,2]. The goal of this series was to identify the risk factors of spontaneous perforation in non-augmented bladder exstrophy patients.

Patients and methods

Nine cases of spontaneous intra-peritoneal bladder rupture were identified between 1991 and 2012 across four centres in France and Australia. These cases were reviewed locally and data were collected with proforma.

Results

All of the cases were classic exstrophy—epispadias (E—E) (Table 1). Two patients were female. Both had Mullerian abnormalities (duplications), one with a fistula between the bladder plate and the left hemivagina. The remaining seven male patients had no other congenital anomalies. All nine cases were born with normal upper tracts.

Repair was performed using Kelly repair in two and staged repair in seven (six with osteotomy). Initial repair

Table 1 Patient diagnosis and initial management.

Pt	Diagnosis	Sex	Repair	Timing	Osteotomy
1	Classic E—E	M	Kelly	6 Weeks	−
2	Classic E—E	M	Kelly	Day 2	−
3	E—E, Mullerian duplication, vesicovaginal fistula	F	Staged	Day 2	+
4	Classic E—E	M	Staged	Day 6	−
5	Classic E—E	M	Staged	1 year	+
6	Classic E—E	M	Staged	Day 1	+
7	Classic E—E	M	Staged	Day 2	+
8	Classic E—E, Mullerian duplication, left hemivaginal agenesis	F	Staged	Day 17	+
9	Classic E—E	M	Staged	Day 2	+

E—E, exstrophy—epispadias; Pt, patient.