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Primary congenital bladder diverticula: Where does the ureter drain?

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ABSTRACT

Background: Primary congenital bladder diverticulum (PCBD) is related to a deficient detrusor layer allowing out-pouching of the bladder mucosa through the inadequate muscularis wall. We aimed to review our experience with symptomatic PCBD in order to correlate clinical findings with anatomical aspects and to present late outcome. **Materials and Methods:** We reviewed all patients operated in our institution since 2004. We evaluated the charts for complaints, radiological exams, method of treatment, complications and length of follow-up. **Results:** We treated 10 cases (11 renal units - [RU]), predominantly males (9/10), mean age at surgery of 5.3 years. All patients had significant urological complaints presenting either with antenatal hydronephrosis (4) or febrile urinary tract infection (5) and urinary retention in one. The ureter was found implanted inside the diverticulum in 8/11 RU. An extravesical psoas-hitch ureteroneocystostomy and diverticulum resection was performed in 10/11 cases, whereas 1 case was treated intravesically based on surgeon's preference without performing cystoscopy. Mean follow-up was 34.1 months (1-120) without complications. **Conclusions:** PCBD is an uncommon diagnosis and has a high probability of drainage inside the diverticulum (72.7%). We recommend the extravesical approach associated with diverticulectomy and ureteroneocystostomy as the preferred technique to treat this abnormality.

Key words: Bladder, diverticula, infection, paediatric, voiding dysfunction

INTRODUCTION

The definition of primary congenital bladder diverticulum (PCBD) is one that occurs in the absence of obstructive factors such as posterior urethral valves (PUV) and neurogenic bladder and is related to a deficient detrusor layer that is abnormally thin or partially developed, allowing out-pouching of the bladder mucosa through the inadequate muscularis wall.[1] Interestingly, the ureteral orifice can be incorporated into the diverticula producing vesicoureteral reflux (VUR) and urinary tract infection (UTI). A large diverticula may compress the bladder neck and cause urinary retention. Alternatively, it can be adjacent to an obstructive ectopic ureter presenting as megaureter.

Treatment of PCBD can be controversial, especially when diagnosis is made incidentally. On the other hand, most cases are diagnosed after significant symptoms such as recurrent UTI, bladder outlet obstruction (BOO) or as part of investigation of antenatal hydronephrosis.

We aimed to review our experience with symptomatic PCBD excluding the Hutch-saccules, in order to correlate clinical findings with anatomical aspects such as the site of ureteral implantation.

MATERIALS AND METHODS

We reviewed all patients with congenital primary bladder diverticulum operated in our institution since 2004. We had also access to a video-bank of images of all exams and photographies at surgery. The radiological evaluation included sonography, voiding

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cystourethrography (VCUG) and renal scintigraphy by all patients.

We evaluated the charts for age, sex, complaints, voiding dysfunction, UTIs, method of treatment, objective and subjective improvement, length of follow-up and evidence of haematuria. Voiding dysfunction was defined as significant symptoms of frequency, dysuria in the absence of infection, incomplete emptying, urgency, constipation or incontinence/enuresis. Follow-up proposed after surgery was sonography every 6 months in the 1st year and then once a year. If last appointment was superior to 6 months, patients and families were contacted by telephone and interviewed regarding recent infections or symptoms of voiding dysfunction. Operative records were reviewed to determine if additional procedures had to be performed in addition to diverticulum repair such as ureteral reimplantation.

RESULTS

We treated 10 cases from 2004 to 2014. The ratio of male/female was 9:1; mean age at surgery was 5.3 years (0.9-13). All findings are summarised in Table 1. Diagnosis was made through investigation of antenatal hydronephrosis in 4 cases, 5 after febrile UTI evaluation protocol and 1 patient presented with BOO and voiding complaints. Voiding dysfunction and enuresis were found in 2 cases. All patients were referred to our facility after initial paediatric assistance and initial treatment.

The diverticulum was mostly seen on the right side (60%), whereas 3 patients had it on the left side. One patient presented with both sides diverticula and interestingly, this patient had additionally bilateral obstructive megaureter, decreased renal function on dimercaptosuccinic acid (DMSA) scan and a history of breakthrough UTI [Figure 1]. Only 1 patient had duplicated system. VUR was found in 4 renal units (RU) (VUR: 4/11: 36.4%).

Patients those presented with antenatal hydronephrosis (n = 4) were operated due to recurrent febrile UTI associated with worsening of renal function on DMSA scan and 2 of them had also VUR.

The ureterovesical junction was assessed by extravesical approach in 10/11 RU (9 patients) and only 1 case was treated by intravesical approach based on surgeon's preference. The diverticula could be more easily identified by the extravesical approach. We found that in 8/11 RU the ureter implanted inside the diverticulum (72.7%) [Figure 2]. A psoas-hitch ureteroneocystostomy

Table 1: Diverticula cases

Case	Antenatal hydronephrosis	UTI	Bladder outlet obstruction	SS or DS	Side of diverticula	VUR	Age at surgery	Surgery	Ureter inside diverticulum	Mean follow-up	Complications
1	Yes	No	No	SS	Right	Yes	1-year	DEV/PHU	No	35 months	No
2	No	No	No	SS	Right	Yes	6.3 years	DEV/PHU	Yes	62 months	No
3	Yes	No	No	SS	Left	Yes	3.3 years	DEV/PHU	Yes	20 months	No
4	Yes	Yes	No	DS	Right	No	0.9 years	DEV/PHU	Yes	37 months	No
5	No	No	Yes	SS	Right	No	6.2 years	DEV/PHU	Yes	29 months	No
6	No	Yes	No	SS	Left	No	13 years	DEV/PHU	Yes	1-month	No
7	Yes	No	No	SS	Right	No	3 years	DEV/PHU	Yes	120 months	No
8	No	Yes	No	SS, both sides	Both sides	No, both sides	2 years	DEV/PHU, both sides	No, both sides	4 months	No
9	No	Yes	No	SS	Right	No	7 years	DIV	Yes	23 months	No
10	No	Yes	No	SS	Left	Yes	10 years	DEV/PHU	Yes	10 months	No
Total	40%-yes /60%-no	50%-yes /50%-no	10%-yes/90%-no	90.9%-SS /9.1% DS*	60%-right/30%-left/10% both sides	36.4%-yes/63.6%-no*	5.3 years	90.9%-extravesical /9.1%-intravesical*	72.7%-yes /27.3%-no*	34.1 months	100%-no

*This mark calculations that involves each diverticulum, not isolated cases. DEV: Diverticulectomy, extravesical approach, DIV: Diverticulectomy, intravesical approach, PHU: Psoas-Hitch ureteroneocystostomy, SS: Single system, DS: Double system, UTI: Urinary tract infection, VUR: Vesicoureteral reflux

was performed and the diverticulum was resected in 10/11 cases. Only 1 patient was treated by the intravesical route with diverticulum excision and reimplantation. Mean follow-up is 34.1 months (1-120). We had no complications related to the surgery, no patient presented UTI thereafter. Voiding returned to normal with complete bladder emptying in all patients. No patient had recurrence of the diverticulum or post-operative VUR.

DISCUSSION

Forsythe and Smyte in 1959, and Johnston in 1960 were the first to describe primary diverticula in non-obstructed children. From then on, it has been necessary to establish a clear differentiation between primary diverticula and diverticula secondary to PUV, neurogenic bladder and so forth, which should be treated before treating the diverticula.

Linke and Mongiat-Artus, suggest that the diverticulum originates from areas of weakness of the wall which occurs due to areas of weak embryological junctions where muscle fusion is incomplete.[1,2] The trigone and bladder dome would be the local election because they have embryologically distinct area.[2] Reinforcing the theories above, approximately 90% of primary diverticula are located near the ureteral orifice. The same authors classified the diverticula as their location relative to the ureteral ostium and the relationship with VUR and renal dysplasia. Diverticula in which the mouth of the ureter occurs within, are associated with VUR and in 15-33% of cases there is some degree of renal dysplasia.[1]

The associated abnormal renal development is attributed to either the inability of the ureteral bud to ascend normally or the association with an obstructive process, which may disrupt normal renal organogenesis. Tokunaka *et al*. believe that the distinction of paraureteral versus periureteral diverticula could be useful in both denoting location with respect to ureteral orifices and distinguishing potentially different aetiologies, including embryological weakness in the detrusor muscle, hypoplasia of the muscularis, absence of the muscularis and a non-functional detrusor muscle.[3]

Incidental finding after a US is also possible. The posterolateral diverticula, which represent 10% of BD, have a quite peculiar clinical presentation. They often grow large and patients can present with urinary retention, urinary stasis, infection and stone formation. We had only patient presenting with BOO and it was exactly the one with larger diverticula situated posteriorly [Figure 3].

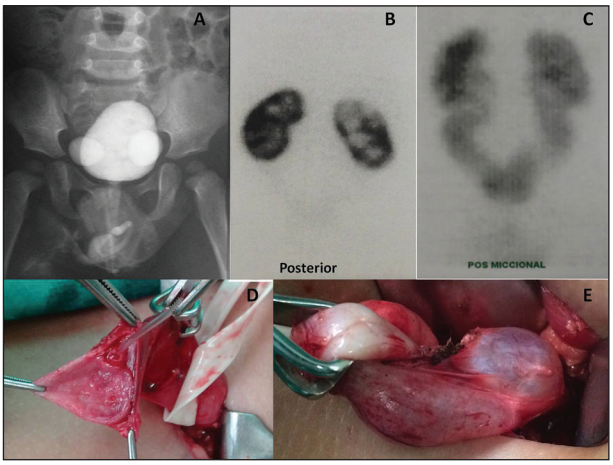

Figure 1: Unusual bilateral congenital bladder diverticula associated with bilateral ureteral obstruction

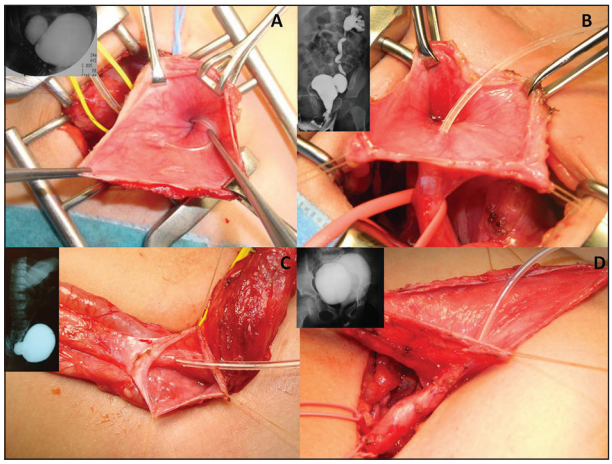

Figure 2: Ectopic placement of the ureter inside the diverticula

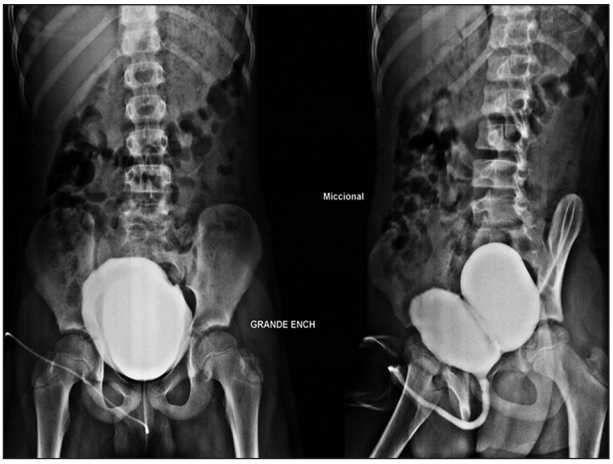

Figure 3: Large posterior bladder diverticula associated with bladder outlet obstruction

One of our most interesting findings was the high incidence of ureteral implantation inside the diverticulum (72.7%). We believe that the extravesical approach allows better access to the

region and consequently clear identification of the structures, which may influence on achieving a better outcome. To our knowledge, such a high figure of ureteral drainage inside the diverticulum has not been outlined, although only 1 patient was treated intravesically, the repair of this particular case was much more difficult. In addition to that, extravesical approach enables safe ureteral reimplantation in cases of association with obstructive megaureter, such as in the bilateral case. We acknowledge that in this case we performed reconstruction in two settings.

Considering VUR, our series comprises a low-grade reflux in 2 of our 4 cases. One patient who had a higher grade VUR had also a much complex clinical scenario and the BD was only a minor aspect. This patient had also an urorectal fistula and penile urethral atresia, that we treated initially by an ASTRA surgery and a tunica vaginal two-stage urethroplasty.[4]

We reviewed all available literature of PCBD and produced a table that summarises clinical characteristics and results of 14 series[5-17] [Table 2]. Garat *et al*.[5] treated 11 children with PCBD. At diagnosis, their ages ranged between 7 months and 9 years (mean 4.6 years). Ten of them were boys and the main reason for consultation was febrile urinary infection ($n = 5$), recurrent urinary infections ($n = 3$), enuretic syndrome ($n = 1$), gross haematuria ($n = 1$) and pain in the right iliac fossa ($n = 1$). Rawat *et al*., from India, reported on 9 patients, all males with age at presentation who ranged from 6 months to 8 years (mean 3 years). All were diagnosed postnatally by ultrasound and/or VCUG and confirmed on urethrocystoscopy. Open surgical excision of diverticulum was done in all the patients. Ureteral reimplantation was simultaneously done only in 3 patients with VCUG-documented high-grade VUR.[6] These two series, similar to ours, express the clinical variability at presentation that include basically two groups: Neonates and infants, diagnosed based on antenatal hydronephrosis or after febrile UTI and a later group of scholars that have incidental findings or unspecific voiding complaints.

The Philadelphia group published in 2004 a 10-year retrospective review series that revealed 4 patients (3 boys and 1 girl) with BOO due to giant BD. Prenatal and postnatal clinical and imaging records were reviewed. The female patient (11 years) had the Ehlers-Danlos syndrome (EDS) and was the only patient who presented with UTI. All boys had progressively decreasing urinary stream and urinary retention. In each patient, VCUG showed a giant (>7 cm) BD that descended below the bladder neck and compressed the urethra during voiding. VUR was seen in 2 patients. Ultrasonography demonstrated moderate unilateral hydronephrosis in 2 patients. Bladder diverticulectomy was successfully performed in all patients, with ureteral reimplantation in 3.[7]

The EDS is commonly associated to PCBD. The EDS is a group of inherited connective tissue disorders, caused by various defects in the synthesis of collagen. EDS is known to affect men and women of all racial and ethnic backgrounds. There are six distinct types of EDS currently identified. All share joint laxity, soft skin, easy bruising and some systemic manifestations. Each type is thought to involve a unique defect in connective tissue, although not all of the genes responsible for causing EDS have been found. We found cases reported of PCBD and EDS.[1,7-11]

The largest series found in the literature comprises 22 children (21 boys and 1 girl) aged 13 days to 14 years old (median age 2 years).[12] Transvesical diverticulectomy was performed as primary treatment in 20 patients. In the remaining two who presented at the age of 2 weeks with acute retention, cutaneous vesicostomy was the primary management followed after 1-year by diverticulectomy. Diverticulectomy was combined with ureteral reimplantation in 14 ureters (four refluxing and 10 non-refluxing) which were incorporated within the diverticular sac. In two children with bladder-neck hypertrophy, a bladder neck incision was additionally performed.

CONCLUSION

PCBD is an uncommon diagnosis, but it demands reconstructive surgery in most symptomatic cases. One must be aware of the high incidence of ureteral implantation inside the diverticula. Based on our experience, the extravesical approach associated with diverticulectomy and ureteroneocystostomy should be considered as the preferred approach.

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Nil.

Conflicts of interest
There are no conflicts of interest.

Table 2: Reviewed cases

Article	*n*	Mean age	Presentation at diagnosis	USG/intravenous urography-hydronephrosis (%)	Palpable mass (%)	VUR (%)	Treatment	Follow-up	Postsurgery
Orikasa *et al.*, 1990	2	2.8 years	UTI (50%), BOO (50%)	50	50	50	Nephroureterectomy (1/2), UNC (2/2), DLC (2/2)	—	Asymptomatic
Zia-Ul-Miraj, 1999	1	2 years	BOO (100%)	100	100	100	DLC and UNC	6 months	Asymptomatic
Schukla *et al.*, 2004	4	6.2 years	UTI (25%), haematuria (25%) and BOO (100%)	50	100	50	DLC (4/4) and UNC (3/4)	5.8 years	Asymptomatic
Evangelidis *et al.*, 2005	21	8.16 years	UTI (90.5%), VD (57.1%)	—	—	—	DLC (21/21) and UNC (15/21)	44.2 months	VD (9.5%)
Bogdanos, 2005	22	2 years	UTI (45.5%), BOO (22.7%), bacteriuria (18.2%), haematuria (4.5%) and incidental (18.2%)	—	—	18.2	DLC (20/22), UNC (14/22) and bladder neck incision (2/22)	16 years	Bladder neck stenosis (4.5%), severe haematuria and clot retention (4.5%)
Corbett *et al.*, 2007	1	7 months	UTI (100%), BOO (100%)	0	0	0	DLC and UNC	—	—
Garat *et al.*, 2007	11	4.6 years	UTI (45.5%), enuresis (9.1%), haematuria (9.1%) and AP (9.1%)	—	—	36.4	DLC (10/11) and UNC (5/11)	9.3 years	Asymptomatic
Rawat *et al.*, 2008	9	3 years	UTI (66.7%), VD (44.4%), BOO (22.2%) and enuresis (11.1%)	77.8	—	66.7	DLT (9/9) and UNC (3/9)	4 years	Asymptomatic
Meeks *et al.*, 2009	1	12 years	UTI (100%), haematuria (100%)	—	—	—	Robotic-assisted laparoscopic diverticulectomy	6 months	Asymptomatic
Singal and Chandrasekharam, 2009	7	30 months	UTI (85.7%), poor stream (28.6%)	100	85.7	71.4	DLC and UNC (5/7), vesicostomy (2/7)	26 months	Asymptomatic
Aydogdu *et al.*, 2010	51	6.4 years	UTI (100%)	—	—	61 reflexive units in 51 patients	DLT (51 patients), intravesical UNC (23 RU), subureteral injection (28 RU), extravesical UNC (10 RU)	22.1 months	Overall success rates from 79% to 91%
Bhat *et al.*, 2012	12	16.8 months	BOO (100%), UTI (41.7%)	100	50	75	DLT and UNC	26 months	Supra-pubic fistula (8.3%), VUR (8.3%), hydronephrosis (16.6%)
Khemakhem *et al.*, 2013	7	3.5 years	UTI (57.1%), UTI (42.9%) and AP (28.6%)	57.1	28.6	42.9	DLT (7/7) and UNC (4/7)	4 years	Asymptomatic
Current series	10	5.3 years	UTI (50%), VD (20%), enuresis (10%) and BOO (10%)	40	—	54.5	DEV (10/11); DIV (1/1); PHU (11/11)*	34.1 months	Asymptomatic

*This mark calculations that involves each diverticulum, not isolated cases. DEV: Diverticulectomy, extravesical approach, DIV: Diverticulectomy, intravesical approach, PHU: Psoas-Hitch ureteroneocystostomy, BOO: Bladder outlet obstruction, UTI: Urinary tract infection, VD: Voiding dysfunction, AP: Abdominal pain, DLT: Diverticulectomy, UNC: Ureteroneocystostomy, USG: Ultrasonography, VUR: Vesicoureteral reflux, RU: Renal units

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