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Patent urachus and bladder outflow obstruction—chance or consequence? A study of a cohort of patients with complete patent urachus presenting to a tertiary urological center and a review of literature

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Abstract

Background: To explore the association between patent urachus and bladder outflow obstruction (BOO). A retrospective review of patient records over a 35-year period (1983–2018) with complete patent urachus was performed. Antenatal ultrasound findings were noted, and postnatal investigations included ultrasound (US), micturating cystourethrogram (MCUG), functional nuclear medicine scans (MAG3, DTPA, and DMSA), and serum creatinine. Associated anomalies and management in all patients were analyzed.

Results: Sixty-six patients with all types of urachal remnants were identified of whom only 16 had a patent urachus. All presented clinically with a discharging umbilicus, 10/16 confirmed on MCUG and 4 had umbilical cord cysts on antenatal US. Twenty-five percent had associated bladder outlet obstruction (BOO): etiologies included atresia of posterior urethra, congenital urethral hypoplasia, urethral atresia with prune belly syndrome, and sacrococcygeal teratoma. Vesicoureteral reflux (VUR) was confirmed in 37%, and four of them had bladder outlet obstruction (BOO).

Conclusion: With patent urachus, bladder outflow obstruction occurs in the minority. Based on our findings, we commend US and cystogram to document VUR. The isolated PU should be treated nonoperatively up to a year of age. Renal function should be checked with the finding of VUR. The etiopathogenesis of the condition remains uncertain.

Keywords: Patent urachus, Bladder outlet obstruction, Vesicoureteral reflux, Conservative management, Micturating cystourethrogram, Umbilical cord cyst

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Background

Patent urachus (PU) is a rarity, the quoted incidence in literature being quite variable ranging between 1 and 2.5/100,000 and 1/7610 [1–3]. It has only been recognized and diagnosed prenatally since 1988 [1]. The prenatal appearance of a patent urachus is that of a true allantoic cyst in the umbilical cord of the fetus with a prevalence of 0.4–3% at around 8–12 weeks of gestation [2].

Does obstruction cause persistence of the urachal channel? Bureau and Bolduc tried to answer this question in their case report of patent urachus and posterior urethral valves and referred to the article by Ross et al. which was a prospective study and looked at the prevalence of first trimester cysts, and among 29 cysts found in 859 women screened, and only one was associated with obstructive uropathy [4]. The association of bladder outflow obstruction (BOO) and the persistence of urachus have been referred to in published literature however evidence is lacking to suggest there is a strong association; in fact, it is quite a rare event in obstructive uropathy [5, 6].

Our hypothesis is that there is probably another contributing factor in the causation of this abnormality similar to a patent vitello-intestinal tract.

Methods

We performed a retrospective case note analysis. Patients were identified through the electronic database medical records for the period between 1983 and 2018. Search criteria included the word "urachus." Medical notes and investigations were reviewed to select carefully patients who fulfilled the criteria of a patent urachus. We excluded all other variations of urachal remnants and included only complete patent urachus in the study. Antenatal ultrasound (US) findings were noted, and postnatal investigations included ultrasound (US), micturating cystourethrogram (MCUG), functional nuclear medicine scans (MAG3, DTPA, and DMSA), and serum creatinine. Associated anomalies and management in all patients were analyzed.

Results

We identified 66 patients from the database with the search term "urachus." Only 16 patients had a patent urachus. All the patients presented clinically with a discharging umbilicus, 10/16 confirmed on MCUG and 4 were antenatally diagnosed with umbilical cord cysts. The median age of presentation was 16 days (1 day–8 years); only one patient presented beyond the age of toilet training. The study included twelve males and four females.

Postnatal US was the most common radiological investigation 15 patients (93.8%) followed by MCUG 10 patients before surgery (62.5%) and one performed after the surgery of the patent urachus for a not related subject, DMSA 6 patients (37.5%), MAG3/DTPA in four patients (25%), and CT renogram in one patient.

Nine of the 13 patients who had antenatal US had abnormalities, four had umbilical cord cysts, and one of them had left hydronephrosis as well (Table 1). One patient had an antenatal diagnosis of a possible exomphalos, and another exomphalos with bladder exstrophy; both were later confirmed postnatally as an isolated patent urachus and exomphalos associated with a patent urachus, respectively. One patient had bilateral hydroureteronephrosis. One patient was a conjoint twin MCMA (monochorionic monoamniotic) and had a connection at the level of the bladder.

The last patient was diagnosed with a sacrococcygeal teratoma. Four of 13 patients had normal antenatal scans; however, two were identified to have urinary tract and other system anomalies postnatally.

Four patients (25%) had associated BOO clinically diagnosed with retention of urine, interrupted urine stream and confirmed with MCUG. The BOO could be partial or complete obstruction of the urine outflow from the bladder through the bladder neck and/or urethra. The obstruction could be intrinsic such as atresia of posterior urethra, congenital urethral hypoplasia, urethral atresia with prune belly syndrome (PBS) or extrinsic such as sacrococcygeal teratoma (Altman type III) in our series.

The diagnosis of BOO was based on postnatal clinical examination and postnatal US where hydroureteronephrosis was evaluated by MCUG. The MCUG was performed on 10/16 patients. On the 6 patients, it was not performed as they did not have ureteral hydronephrosis on the postnatal scan.

In patients with BOO, 1 had an indwelling catheter for a week following excision of the sacrococcygeal teratoma and the PU spontaneously closed after teratoma removal. In the remaining three patients, the urachus was initially converted to a vesicostomy to optimize drainage before further definitive surgery.

All patients with isolated patent urachus had surgical closure with a good outcome. The urachus was circumscribed and followed down to the bladder. Bladder closed with vicryl 4/0, closure of umbilical defect vicryl 4/0, excision of granulomatous tissue, and skin closure.

In six patients out of the 16 with PU (37%), vesicoureteral reflux (VUR) was confirmed among those investigated and four of them had BOO as well, one with prune belly syndrome and an anterior urethral atresia who had left-sided reflux into a dysplastic kidney and progressive worsening hydronephrosis of the right side despite drainage; the initial cystourethroscopy revealed no urethral meatus impossible to introduce any scope

Table 1 Patients with patent urachus associated with/without BOO showing antenatal US findings, associated anomalies, management, and outcomes. Outcome after surgery of the PU as it is. In the patients with another pathology associated, the outcome includes renal function

Diagnosis	BOO	Antenatal us findings	Associated anomalies	Management	Outcome
Sacrococcygeal teratoma	Present	Sacrococcygeal teratoma	Bilateral hydronephrosis/ neuropathic bladder and bowel	Indwelling catheter	Good renal function, incontinence fecal, and urinary due to neuropathy
Atretic posterior urethra	Present	Normal (1987)	Fallot tetralogy/bilateral hydronephrosis/left undescended testes	Cystofix > vesicostomy > final repair	Good renal function, resolution of upper renal tract dilatation
Atretic anterior urethra/PBS	Present	Normal (1983)	Prune belly syndrome/bilateral VUR/bilateral undescended testes	Vesicostomy > mitrofanoff + augmentation	CKD stage 2 Hypertension
Urethral hypoplasia	Present	Cord cyst Left hydronephrosis and megacystis	VACTERL/left VUR in dysplastic left kidney	Vesicostomy	CKD stage 2
Exomphalos	Absent	Bladder exstrophy or patent urachus or exomphalos	Exomphalos	Repair of exomphalos and closure of PU	Good
PU	Absent	Normal	Right grade III VUR	PU excision and antibiotics	Good
PU	Absent	Cord cysts	Right duplex kidney with VUR into the lower moiety	PU excision and antibiotics	Good
PU	Absent	Normal	Proteus syndrome/hemangioma left testes	PU excision	Good
PU	Absent	Not available		PU excision	Good + keloid
PU	Absent	Cord cyst		PU excision	Good + protuberant umbilicus
PU	Absent	Cord cyst		PU excision	Good
PU	Absent	Omphalocele		PU excision	Good
PU	Absent	Normal		PU excision	Good
MCMA twin and urogenital sinus	Absent	Conjoint twin	Two hemivaginal openings at introitus	Excision of PU and ileostomy	Good renal function
PU	Absent	Normal	Hemangioma on scalp	PU excision	Good
PU	Absent	Not available		PU excision	Good

per urethra, scope passed per vesicostomy showing a short prostatic urethra ending with a membrane. A channel was seen post-membrane but was impossible to scope into, concluding that the anterior urethra is completely atretic and the urethra posteriorly patents up to the bulbar portion. The bladder wall thickened but not trabeculated.

The second patient had duplication of the urethra, hypoplastic megalourethra, and bilateral hydronephrosis which resolved the patient initially presented with difficult catheterization, bifid scrotum, penis covered in redundant skin dorsally, and floppy redundant skin ventrally (Fig. 1a).

EUA and cystoscopy revealed a hypoplastic proximal penile and bulbar urethra with a pouch diverticulum at the bulbo-penile junction and a distal urethra (Fig. 1b).

During reconstruction, no erectile tissue was found in the penis. The corpora spongeosum was completely fibrotic. Duplication of the urethra was confirmed on dissection.

The ventral urethra has its opening from the grossly dilated proximal urethra just below the veru montanum.

The distal urethra starts as a long dilated megalourethra distally in the penile portion and is also tight meatal stenosis.

The ventral blind pouch was taken down and used as a free graft to augment the posterior urethra dorsally. A distal megalourethra was remodeled with redundant tissue taken down and used as a free graft to augment the membranous urethra proximally. A 10 fr. Silastic stent was left in situ.

The third patient had an atretic anterior urethra with bilateral VUR and finally. MCUG revealed moderately dilated posterior urethra with soft tissue filling defect obstructing urethral outflow separate to expected insertion of a possible rectourethral fistula (patient had anorectal malformation).

Cystoscopy showed a fleshy polyploid structure just below the bladder neck.

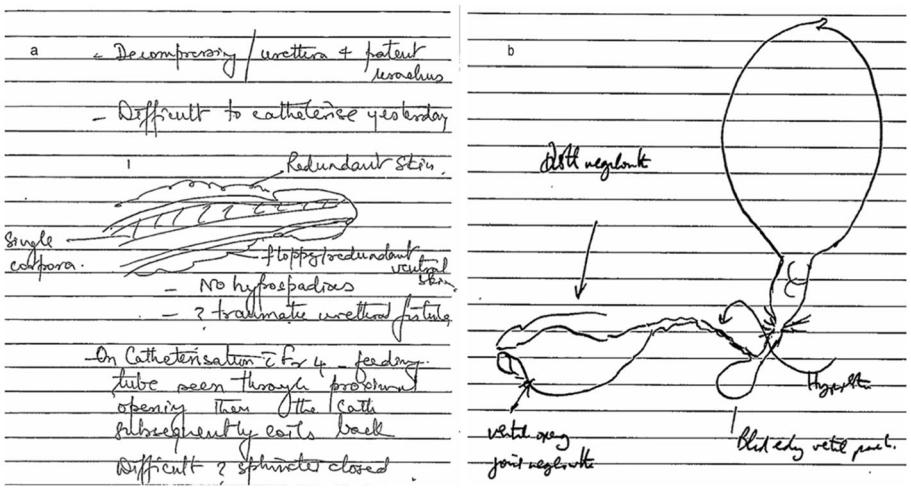

Fig. 1 **a** External appearance of urethral duplication. **b** Cystoscopic depiction of urethral duplication

The fourth patient had a sacrococcygeal teratoma who had bilateral hydronephrosis.

In the two patients with VUR and no BOO, one had right grade 3 VUR and the second had a right duplex kidney with reflux into the lower moiety.

Discussion

Urachal remnants are persistent intra-embryonic connections between the allantois and the cloaca. The allantois appears around day 16 as a tiny, finger-like outpouching from the caudal wall of the yolk sac, which is contiguous with the ventral cloaca at one end and the umbilicus at the other. The ventral portion of the cloaca develops into the bladder after cloacal division by the uro-rectal septum [3]. The descent of the bladder toward the pelvis stretches the urachus, eventually obliterating of its lumen leading to the formation of the median umbilical ligament, which is the resultant fibrous cord and runs from the umbilicus to the dome of the bladder. Failure in the obliteration of the lumen in its entirety results in a patent urachus [7]. Differential diagnosis of this condition includes patent omphaloentric duct.

Published literature on patent urachus is lacking. Most of the available literature discusses the entire spectrum of urachal anomalies collectively. Our study carried out in a tertiary referral center had the unique aim of focusing only on pure patent urachus. We limited our reference to seven other case studies and focused on complete patent urachal anomalies in those studies [3, 5–10].

In six of seven case series, US and MCUG were carried out with some additional investigations varying from one series to another, sinogram in three of the seven series [3, 5, 6], CT renogram in two of the seven series [3, 7], and MRI in one [7]. The primary aim of MCUG was to diagnose or exclude BOO. Both Yiee et al. and Huang et al. recommended that MCUG is not necessary as BOO is exceedingly rare [5, 6].

Our center dealt with around 600 patients with posterior urethral valves over 30 years; we would infer that not a single one had a patent urachus. What does that imply? Is it the timing of obstruction, abnormal abdominal wall development, the severity of obstruction, or a combination of these factors or other unrecognized factors involved in its aetiopathogenesis?

Why do children without BOO get a patent urachus? In fact, 12 of 16 in our series had no BOO. This observation goes on to infer that only a minority of these children with a patent urachus have a BOO. The real cause of the patency of urachus remains unclear, but it is probably related to an early embryological mesenchymal failure. In the classic PUV, there is a closed urachal cap and almost no incidence of patent urachus; therefore, it is logical to conclude that patency of the urachus has nothing to do with bladder outflow pressures and that the incidence of BOO in PU is random and low.

In our study, 3 patients had a conversion of the PU to a formal vesicostomy in the BOO group. All the three had urethral anomalies causing suboptimal drainage via the patent urachus resulting in worsening serum creatinine and upper tract dilatation. Conversion to a formal vesicostomy was undertaken to improve drainage and achieve stabilization before the definitive intervention could be offered. This has been reported previously in some series [11].

Of the seven case series reviewed in the literature [3, 5–10], two have raised the option of nonoperative management. Naiditich et al.'s series included 21 patients with PU, 14 of which were treated nonoperatively [7]. The second series, which has specifically addressed the

option of nonoperative management, was by Lipskar et al. comprising five patients of whom four were nonoperatively managed with success [9]. This study strongly recommended nonoperative management avoiding surgical excision under 1 year of age. The argument favoring this approach was based on the debate about the timing of obliteration of the urachus [1]. It was also stated that in normal development, the urachus obliterates by the 12th week of gestation. In the same year, a study carried out on 102 asymptomatic infants by Zieger et al. concluded that urachal involution is not complete at birth and can be followed up sonographically in the first months of life [12]. They recommended that this understanding should prompt a new strategy in young infants with discharging umbilicus instead of early surgery an ultrasound-guided conservative approach seemed reasonable. We treated all our patients with isolated PU surgically as nonoperative management was not part of our treatment protocol for patent urachus, but nonoperative management until 1 year of age can be offered as an option. All patients who underwent surgical repair for isolated patent urachus did not carry out routine urodynamics postoperative but postoperative ultrasound confirming good bladder emptying was reassuring and consequent discharged from our care.

Although our main focus was the incidence of bladder outflow obstruction in patent urachus, we observed a high incidence of VUR in our series: six of sixteen (37%) and two in those without BOO. The quoted incidence of VUR in patent urachus has been as high as 64% from the Mayo Clinic series [10]. However, this was 34% when it came to a pure patent urachal anomaly. The group recommends routine MCUG and US for those with a positive family history of VUR, febrile UTI, renal anomaly, or previous hydronephrosis [10]. The uncommon association of bladder outflow obstruction with patent urachus does not eliminate the need to carry out a cystogram as the association of VUR may be of significance, and hence, its presence or absence should be established [10].

Renal function was preserved in two of the four (50%) patients with BOO and in all without BOO. Rental function should be checked in patients with BOO or hydronephrosis. Based on our data and literature review, we noted a higher incidence of bladder outflow obstruction of up to 25% (four of sixteen patients). The quoted incidence of bladder outflow obstruction associated with patent urachus in studies carried out between 1937 and 1981 is 14% [5]. Five of the largest series between 1997 and 2007 included 167 patients; however, it did not document any bladder outflow obstruction [5].

Our study is also subject to selection bias as it is based on a select group of children who have PU and other anomalies that lead to a referral to a tertiary center. There is a possibility that more simple and isolated cases of PU may be managed nonoperatively by primary care providers and are never referred to our center. This type of selection bias may have contributed to the incidence of BOO in our study.

Conclusion

With patent urachus, bladder outflow obstruction occurs in the minority. Based on our findings, we commend US and cystogram to document VUR. The isolated PU should be treated nonsurgically up to a year of age. Renal function should be checked with the finding of VUR. The etiopathogenesis of the condition remains uncertain.

Abbreviations

BOO: Bladder outflow obstruction; US: Ultrasound; MCUG: Micturating cystourethrogram; VUR: Vesicoureteral reflux

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Authors' contributions

HT: data collection, drafting and revision of the manuscript, and operating. SR: data collection and revision of the manuscript. NS: operated on a few cases. DD: operated on cases. IM: operated on cases. PC: operated on cases. AC: senior author drafted the manuscript and operated on cases, supervised writing the manuscript. The authors read and approved the final manuscript.

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It is available and can be submitted upon request.

Ethics approval and consent to participate

The project was registered and appropriate approval was obtained from the study institution.

Consent for publication

All authors agree to the publication of this study.

Competing interests

There are no conflicts of interest.

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