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Retrovesical cysts in boys—case series and literature review

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Abstract

Background: Retrovesical cysts present with varying etiologies. We initiated this study to provide insights into retrovesical cysts in boys through a case series and systematic literature review and to aid in treatment alternatives.

Materials and methods: Case series of seven patients from the in-hospital register with a systematic literature review on retrovesical cysts in children.

Results: We identified seven patients from the in-hospital register during 2005–2020 and eighty-two patients from the literature review. The literature review showed that children's retrovesical cysts are mainly asymptomatic before puberty. Those detected earlier in childhood present mostly with renal abnormalities. In our series, retrovesical cysts derived from three distinct etiologies with different treatment modalities. Asymptomatic cysts do not need excision and are to be followed up through puberty, but in the case of an ectopic ureter, nephroureterectomy is optional. Symptomatic retrovesical cysts may demand surgical excision. Ultrasonography is usually sufficient for diagnosis, but MRI provides better anatomical delineation and aids in surgical planning.

Conclusions: Retrovesical cysts in boys are benign conditions associated with abnormal development of the ureter and kidney. Ultrasonography is sufficient for diagnosis, with MRI giving further detail for surgical planning. Treatment consists of cyst excision, heminephrectomy/nephrectomy, transurethral canalization, or excision of prostatic utricle, depending on etiology. Asymptomatic cases are to be followed up through puberty, but in cases of an ectopic ureter, nephroureterectomy is preferred.

Keywords: Retrovesical cyst, Zinner syndrome, Ectopic ureter, Development

Background

Retrovesical cysts in boys are uncommon entities of varying etiology. Retrovesical cysts arise from Müllerian duct remnants or abnormal development of Wolffian ducts, with an estimated prevalence of 1–4% and 0.005%, respectively [1, 2]. Embryologically, the urogenital system derives from the mesonephric duct, explaining why retrovesical cysts may be associated with renal malformations [3]. The association of congenital seminal vesicle cysts and ipsilateral upper urinary tract abnormalities has been termed Zinner syndrome [4].

Clinical and radiological distinctions between different types of retrovesical cysts may prove difficult. While symptomatic cysts are treated surgically, most cysts are believed to be asymptomatic and found incidentally [5]. In association to renal malformations, surgical interventions are related to renal surgery, including nephrectomy, heminephrectomy, and cysts marsupialization. Cyst excision is needed occasionally [6–11]. Surgical cyst excision presents challenges due to anatomical location, with ductus deferens, pelvic nerves, rectum, and ureters posing further reasons for caution. Mini-invasive techniques have emerged as viable options for surgical treatment, with less procedural morbidity and shorter length of stay, but present technical challenges [6–8]. Robot-assisted surgery provides advantages over traditional laparoscopy, and reports of robot-assisted management of retrovesical

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cysts are emerging [9–11]. Most reports of retrovesical cysts are case reports, with only a limited number of case series presented.

We present a case series of seven patients with retrovesical cysts at our institute with data on presentation, etiology, imaging, and treatment during 2005–2020. Further, we present data from a current literature review on patients identified on PubMed.

Patients and methods

Data from a cohort of seven patients treated with retrovesical cysts are presented. Patients were identified from the in-hospital pediatric urological diagnosis register database. Patient records were dissected for the identified patients regarding reasons for presentation, imaging studies, operation notes, and follow-up data. Attendance and adherence were 100%. In all cases, ultrasonography (US) was sufficient for diagnosis. Magnetic resonance imaging (MRI) was utilized for better anatomical delineation and surgical planning (Fig. 1). For all patients with anomalous kidneys, renal scintigraphy was done. One patient had 9% differential renal function on the ipsilateral side, and all others had nonfunctioning kidneys. All patients underwent cystoscopy. Voiding cystourethrography was utilized for five patients based on individual clinical indications.

Cystoscopy was performed with patients under general anesthesia. Pediatric cystoscopes were utilized with a working channel and a 0-degree lens. For per urethral canalization, we used progressive dilatation with Ch3 to Ch5 ureter catheters introduced via the natural orifice. In the case of the prostatic utricle, the cystoscope was further inserted for dilatation after dilatation with Ch3 to Ch5 catheters.

The systematic literature review was conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. PUBMED was queried for retrovesical cysts in boys with the search string "retrovesical cyst children" with 39 results. Additional searches were undertaken to identify articles related to "seminal vesicle cyst children (83 results)" and "Müllerian duct cyst children (117 results)" (see Fig. 2 for flowchart). Reference lists of identified articles were searched to broaden the initial search strategy. Snowballing was further utilized for extended coverage [12]. All articles were read in full, with relevant and applicable data collected and processed. The date of PUBMED access was 2 September 2021. Articles included in the review consisted of case reports, case series, retrospective reviews, and review articles focusing on retrovesical cysts in boys. Echinococcal cysts were excluded, as well as non-English articles or articles not found online.

Data was extracted manually in several increments, starting from title and abstract survey, followed by full-text analysis given relevant inclusion criteria. Outcomes included diagnosis, presentation, imaging, intervention, and additional findings.

Results

Seven patients were identified during 2005–2020 from the in-hospital pediatric urological diagnosis register database. The individual patients are described in Table 1. The patients presented with three different etiologies for cyst formation, consisting of the following diagnostic entities: ectopic ureter, Zinner syndrome, and

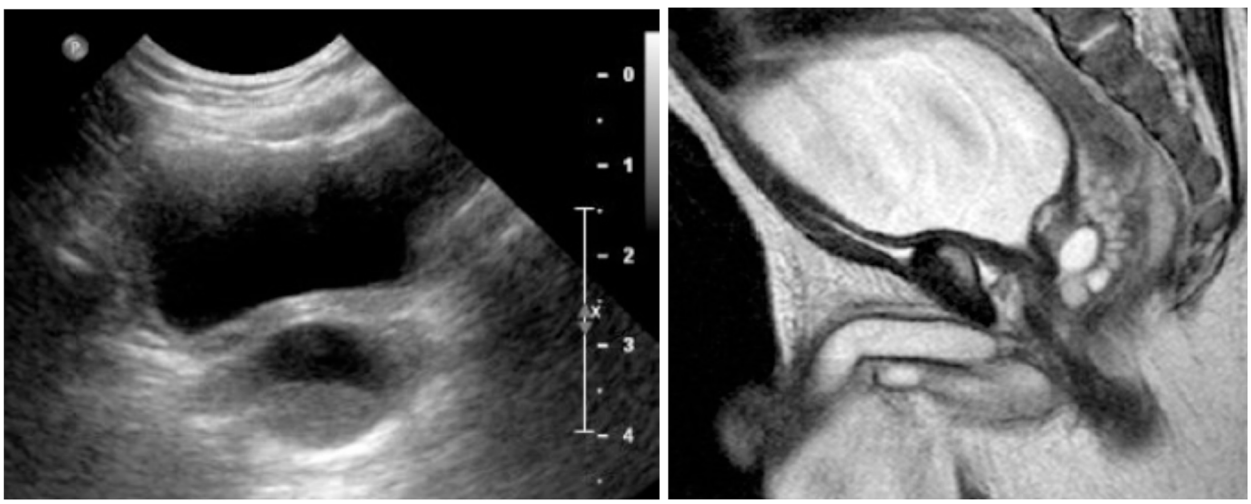

Fig. 1 Ultrasound picture of prostatic utricle and MRI of retrovesical cyst

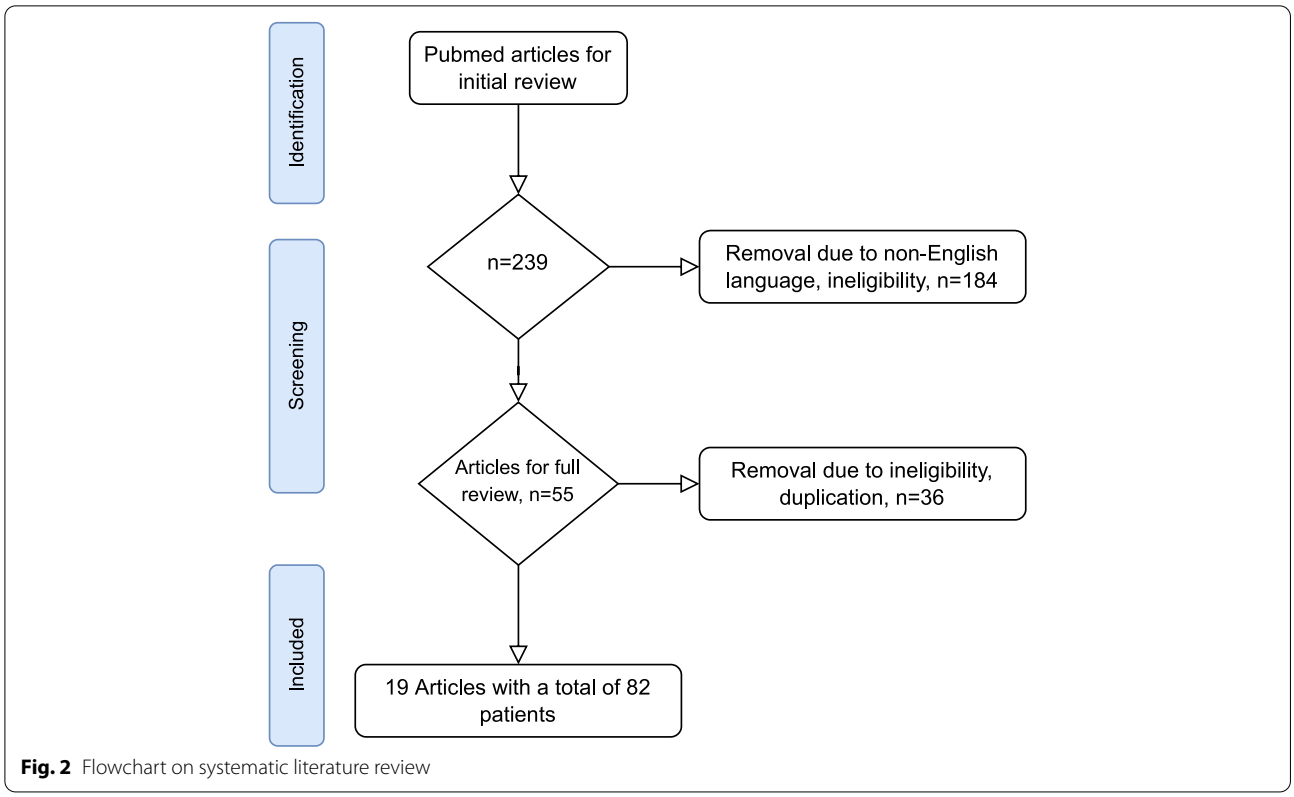

Fig. 2 Flowchart on systematic literature review

Table 1 Patient characteristic, interventions, and outcomes (MCDK, multicystic dysplastic kidney)

Patient nr	Age at diagnosis	Symptoms	Kidneys	Cysts	Surgery	Follow-up (years)	Outcome
1	Antenatal	Asymptomatic	MCDK right	Multiple vesicle cysts	Nephrectomy right	2	No cyst
2	6 months	Left sided epididymitis	Normal	Prostatic utricle	Dilatation of orifice	7.3	Urinary tract infection 6 months post-op
3	Antenatal	Asymptomatic	MCDK left	Multiple vesicle cysts	Nephrectomy left	2.5	13mm cyst behind the bladder
4	15 years	Dysuria	Agenesis left	Single vesicle cyst	Per-urethral canalisation	3	47x29mm cyst, asymptomatic
5	Antenatal	Asymptomatic	MCDK left	Multiple vesicle cysts	-	2	Multiple small cysts
6	Antenatal	Asymptomatic	Ectopic dysplastic left	Single vesicle cyst	-	2	No cyst
7	Antenatal	Asymptomatic	MCDK left	Multiple vesicle cysts	Nephrectomy left	1.2	No cyst

prostatic utricle-derived cysts. Of the seven patients, two were symptomatic: one patient presented with a prostatic utricle and epididymitis associated with urinary tract infection (UTI), and one adolescent patient with Zinner syndrome had dysuria. Six patients had anomalous kidneys associated with the etiology of cysts. In five patients, the diagnosis was made antenatally. Four had ipsilateral a nonfunctioning multicystic dysplastic kidney (MCDK) with ectopic ureter to seminal vesicle. Five patients had voiding cystourethrography performed, in which one had reflux to an enlarged prostatic utricle that had looked like a cyst, and one had a wide posterior urethra. The remaining three patients had no specific findings in the voiding cystourethrographies. The surgical interventions and

outcomes are presented in Table 1. Of the patients with MCDK, three underwent nephroureterectomy because the ipsilateral ureter coalesced ectopically to seminal vesicles, and one of these had an endoscopic dilatation of the cyst tract simultaneously. One of the patients with MCDK was followed up conservatively with the kidney becoming undetectable on ultrasonography. Three patients presented with solitary cysts. One was diagnosed with Zinner syndrome and one with a prostatic utricle with a narrow orifice; both were treated with transurethral dilatation of the cystic tract. One of the operative patients suffered from extended-spectrum beta-lactamase-producing *Escherichia coli* (ESBL) sepsis postoperatively but subsequently has not presented with any symptoms. In the other symptomatic patient with Zinner syndrome, dysuria resolved after urethroscopic canalization that was performed twice. However, 4 months after the latest procedure, dysuria symptoms, and the cyst reappeared. After that, the patient has been in follow-up for two years in adult urology without additional interventions as the symptoms have been intermittent.

All patients have been asymptomatic at follow-up visits, except for those with Zinner syndrome and the patient with the prostatic utricle, as described previously. The median follow-up time was 2 years (range 1.2–7.3 years).

The literature review identified 82 cases (Fig. 2) [2, 3, 5, 8, 9, 13–26]. In most cases, the diagnosis was made with ultrasonography and magnetic resonance imaging, and voiding cystourethrography was utilized in select cases. The largest group consisted of seminal vesicle cysts. The second-largest reported diagnosis was retrovesical cysts. The remaining were made up of Zinner syndrome. The age of onset ranged from birth to puberty. For symptomatic cases, surgical intervention was most common. The most common mode of operation was mini-invasive excision, either laparoscopic or robot-assisted. There were no cases of malignancy during follow-up. No predilection for laterality was detected. For asymptomatic cases, follow-up was unanimous in all, with ultrasonography as a method of investigation. Follow-up was not complicated by conversion towards symptoms in these reports. In 63% of cases, the finding was not limited to the cystic structure but included the ipsilateral upper renal tract, with the most common mode a dysplastic kidney.

Discussion

We present here a cohort of seven pediatric patients with retrovesical cysts. Five had antenatally detected cysts behind the bladder or unilaterally abnormal kidneys. In four cases, the presentation was multiple cysts because the ectopic ureter filled the vesicles with urine. In three of them, the cystic outlook of vesicles disappeared after removing the multicystic dysplastic kidney, and in the remaining case, no operation was necessary, as the size of the cystic area was petite, and the multicystic kidney disappeared. In two other cases, the vesicle cyst was single; the other was associated with unilateral renal agenesis (Zinner syndrome) and the other with a small dysplastic ectopic kidney. In the first case, the cyst was drained periurethrally with partial success, and in the other case, the cyst disappeared by itself. In the remaining seventh case, the cyst-like structure was an enlarged prostatic utricle which was treated by dilatation of the orifice to the urethra. The systematic literature review highlighted that in almost two thirds of patients with retrovesical cysts identified, the ipsilateral upper renal tract was also affected.

In boys, congenital retrovesical cysts result either from aberrant development of Wolffian or Müllerian duct cysts [3]. Wolffian duct cysts are located paramedial (seminal vesicle, vas deferens, ejaculatory duct), and Müllerian duct cysts are located in the midline with normal seminal vesicles bilaterally and intact ejaculatory ducts. Acquired seminal vesicle cysts can be associated with inflammation and ejaculatory duct obstruction. Congenital cysts are believed to arise from a malformation of the ejaculatory duct, explaining the scarcity of reports in young children [18].

Seminal vesicle cysts associated with ipsilateral renal agenesis were first described by Zinner, and thus far, over 200 patients have been described [3, 4, 27]. Zinner's triad is a consequence of a congenital disturbance in the Wolffian duct development and refers to the entity of seminal vesicle cysts, ejaculatory duct obstruction, and ipsilateral renal agenesis [15]. In these cases, it has been proposed that the cysts represent a residue of a dysplastic kidney [18, 28]. In contrast to renal agenesis and Zinner syndrome, most of our patients with retrovesical cysts had MCDK with ectopic ureters draining to the cysts. Previous research has shown that fifteen percent of patients with MCDK have ipsilateral genitourinary tract abnormalities [14]. The close embryologic relationship has been proposed as an explanation for the association between multicystic renal dysplasia and genitourinary malformations during development with both organ systems arising from the Wolffian duct [3, 14, 29].

Most patients with congenital retrovesical cysts are asymptomatic before puberty. Symptomatic presentation is usually in the form of hematuria, dysuria, urgency, frequency, repeated UTI, ejaculatory pain, perineal discomfort, lower abdominal pain, chronic prostatitis, and infertility and begins after puberty [3, 30, 31]. Symptomatic patients are best treated actively. However, surgical cyst excision presents challenges due to anatomical location, with ductus deferens, pelvic

nerves, rectum, and ureters posing further reasons for caution. Our patients did not need cyst excision as only one patient presented with cyst-related symptoms, and in his case, patient-reported morbidity was low. In addition, in four cases, the seminal vesicle cysts disappeared after removal or resolution of the dysplastic kidney.

Currently, data on the need for intervention in asymptomatic cases are scarce. Nevertheless, a conservative approach in asymptomatic cases will avoid possible side effects of cyst excision, as long-term follow-up has not been linked to deterioration or worse outcomes [3, 32]. More than 80% of cases reported in children have been asymptomatic without progression during follow-up [3, 5, 17]. In our series, treatment options were dictated by cyst etiology and included heminephrectomy/nephrectomy and transurethral canalization. Most asymptomatic neonates with ectopic ureters were treated by nephroureterectomy of the nonfunctioning kidney because cystic accumulations were, in fact, enlarged vesicles due to the ectopic ureter. In two asymptomatic patients, no intervention was performed since the kidney remnants were tiny and became invisible in follow-up with US. Nowadays, we prefer a conservative approach even if the cysts are larger in the initial evaluation. In the one patient, the cyst was not visible in the US at the latest outpatient clinic; in the other, the cystic accumulation remained very small. In two patients, per urethral drainage was done for patients with Zinner syndrome and a large utricle with partial success.

Our study has limitations. The material is small and heterogeneous and consists mainly of patients with ectopic ureter. The patients with enlarged cystic-looking vesicles underwent nephrectomy of the nonfunctioning kidney despite of not presenting with symptoms. Accordingly, we cannot present the natural history of those cases. In addition, the follow-up time of our patients is relatively short, but we aim to follow our patients up to puberty as recommended [5]. Our study did not include patients with hypospadias or DSD with enlarged utricles, which may also present as retrovesical cysts.

Retrovesical cysts in boys are benign conditions associated with abnormal development of the ureter and kidney. The nature of the cysts is to be diagnosed with ultrasonography and MRI initially. The cases resulting from ectopic ureter were treated with success and without complications by nephroureterectomy. Although symptomatic cases demand distinct treatment strategies based upon whether a dysplastic kidney was involved or not, most patients are asymptomatic during childhood and should be treated conservatively.

Abbreviations

ESBL: Extended-spectrum beta-lactamase-producing *Escherichia coli*; MCDK: Multicystic dysplastic kidney; MRI: Magnetic resonance imaging; PRISMA: Preferred Reporting Items for Systematic Reviews and Meta-Analyses; US: Ultrasonography; UTI: Urinary tract infection.

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Authors' contributions

NP, EM, and ST planned this study. All authors analyzed data. NP and ST drafted manuscript. All authors commented on the manuscript. The authors read and approved the final manuscript.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Institutional approval: The study was done in accordance with the Declaration of Helsinki. Due to retrospective nature of the study, institutional approval was deemed sufficient, but consent for publication was obtained from parents. Consent to participate: Obtained from parents.

Consent for publication

Yes. Obtained from the parents.

Competing interests

The authors declare that they have no competing interests.

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