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Urachal anomalies in children: Surgical or conservative treatment?

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Abstract *Objective:* To shed light on the current controversy regarding the best treatment option for managing urachal anomalies in children.

Patients and methods: A retrospective follow-up of a case series comprising 13 children who were diagnosed with urachal anomalies was performed. All cases were diagnosed between 2000 and 2011 and followed up at the Pediatric Urology Unit of San Cecilio University Hospital in Granada (Spain). Information about the baseline and follow-up variables was collected from clinical records.

Results: Nine of the 13 patients were symptomatic (6 patients with urachal cysts and 3 patients with urachal persistency). Conservative management was originally used in all but one case. During follow-up, reinfection appeared in two cases, and these patients were treated surgically. Spontaneous resolution was achieved in eight cases (61.5%). Two children with persistent urachal cysts are still being followed (4 and 6 years after the diagnosis), although ultrasound monitoring reveals a gradual reduction in the size of the cysts. The median time between diagnosis and resolution was 16.5 months.

Conclusion: With the exception of cases in which there is a clear indication for surgery (i.e. reinfection), a conservative approach based on regular monitoring may be useful.

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Introduction

The urachus is a structure that connects the dome of the bladder to the anterior abdominal wall at the level of the umbilicus. It is an embryonic remnant that is derived from the involution of the allantois and forms a fibromuscular cord. The pathologies associated with urachus may be congenital or acquired. Congenital urachal pathologies include patent urachus, urachal sinus, urachal diverticulum, and urachal cyst [1]. Acquired urachal pathologies include repermeabilization, infection, and malignancy, the last of which is the most serious pathology, although urachal tumors constitute less than 1% of all bladder tumors [2,3]. A non-negligible proportion of urachal anomalies is incidentally diagnosed during investigation for other diseases. Considering the risk of the aforementioned acquired urachal pathologies, it is necessary to develop the most effective therapeutic strategy for cases of urachal anomaly. However, the existing literature on this topic is limited. Indeed, urachal anomalies are rare, and their incidence at birth is estimated to be 1 in 5000–8000 live births [4,5]. This finding may explain why there are a limited number of published case series [4,6,7] and why most publications on this condition are isolated case reports [5,8–11]. Furthermore, most of the published reports describe cases with complications due to malignancy in adulthood [1,3,12]. Very few reports provide guidelines for diagnostic (symptomatic or incidental) or therapeutic approaches in children, and hence these approaches remain controversial [7]. The question arises whether all cases should undergo surgical intervention or whether conservative therapeutic approaches are preferable.

Since 2000, 13 cases of urachal anomalies have been diagnosed, treated, and followed up at the Pediatric Urology Unit of San Cecilio University Hospital in Granada, Spain. The relatively large size of this case series, the variety of diagnoses, and the employed therapeutic strategies warrant a description of the results to help clarify the controversy regarding the most suitable therapeutic approaches for the management of urachal anomalies that are diagnosed in childhood.

Methods

A retrospective follow-up of a case series comprising 13 children who were diagnosed with urachal anomalies was performed. All of the cases were diagnosed between 2000 and 2011 and followed up at the Pediatric Urology Unit of San Cecilio University Hospital in Granada, Spain. The information about each case was obtained from medical records. The information consisted of the following two groups of variables: baseline variables (date of diagnosis, age, sex, diagnostic procedures, reason for diagnosis [symptomatic or incidental], type of anomaly, and initial treatment), and follow-up variables (additional diagnostic procedures, secondary treatments, final outcome, and time after diagnosis). In conservative management, follow-up was based on ultrasound explorations, which were performed every 6 months in the first 2 years and then annually up to 5 years of follow-up (except for unresolved cases, which were followed up indefinitely). Spontaneous

resolution of the lesion was documented through ultrasound in patients with cysts, and through ultrasound plus cystography in patients with a fistula. The indications for surgery were restricted to reinfection and, in one case, a large fistula.

Results

Table 1 presents the main information collected about each case in the series and is organized according to the age of the patient at diagnosis. Regarding the baseline variables, most of the cases (11 of 13 cases) occurred in males, and the age at diagnosis ranged from 1 day to 14 years (median age 6 years); four cases were diagnosed in the neonatal period, and the remaining nine cases were diagnosed between 6 and 14 years of age. The flowchart in Fig. 1 shows the two initial subgroups of patients defined (4 incidental cases and 9 symptomatic ones), and the management of each subgroup. Regarding incidental, asymptomatic cases (urachal cysts), the diagnosis was based on ultrasound exploration secondary to concurrent pathology (2 cases were diagnosed during work-up for enuresis, 1 case was diagnosed during follow-up for a previous diagnosis of urethral polyp, and 1 case was diagnosed in the setting of grade II vesicoureteral reflux). In the nine symptomatic cases (3 fistulas and 6 cysts), symptoms were mostly due to infection (7 cases). Cystography was performed in all symptomatic cases, and a computed tomography (CT) scan was performed in one child.

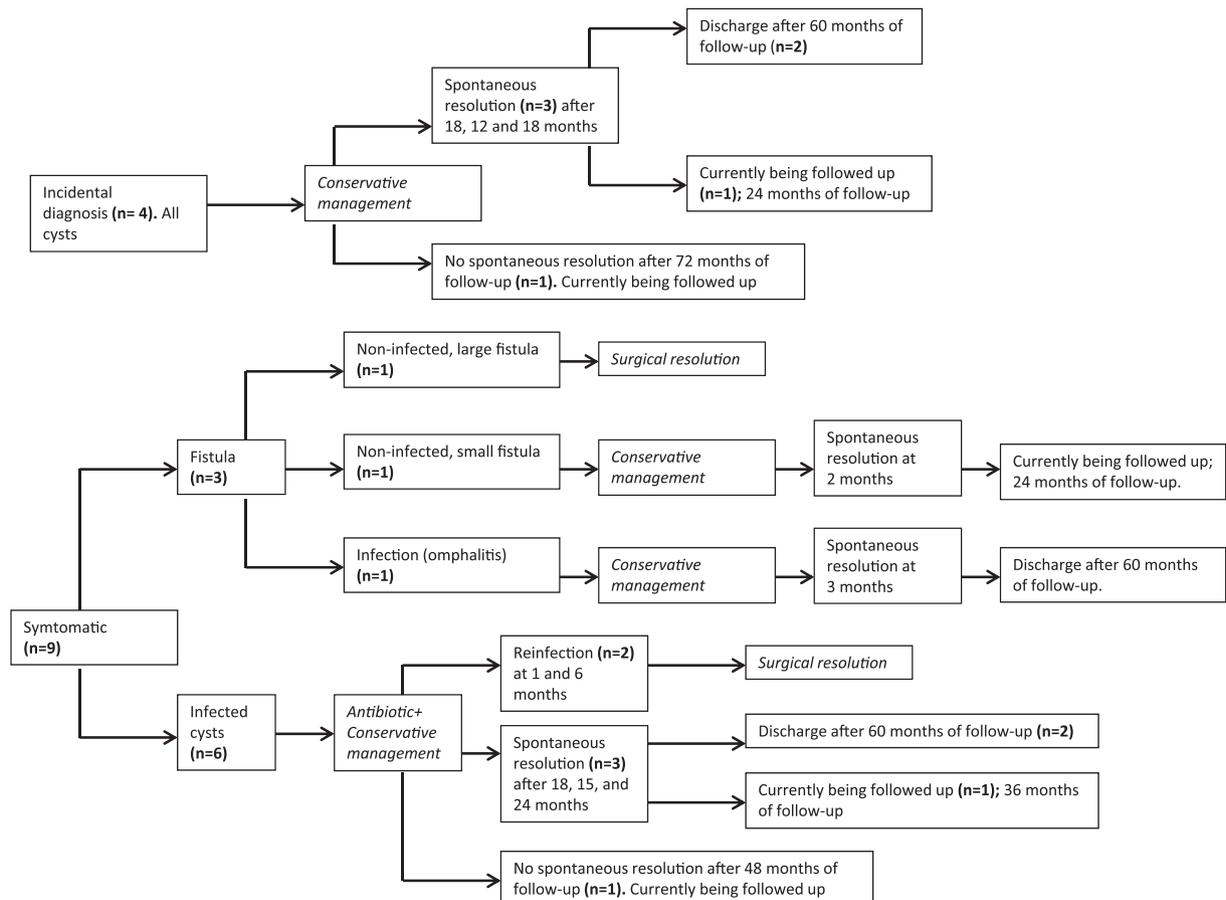
Regarding the initial therapeutic options, the four cases detected incidentally received only primary treatment for their disease. In three of these cases, spontaneous resolution (disappearance of the cyst) was achieved after 18 months in two cases and after 12 months in the other one. One case had not resolved after 72 months of follow-up, although ultrasound monitoring showed a gradual reduction in the size of the cyst. (This patient is currently 20 years old and remains in follow-up).

The seven patients with infection (1 case of fistula with omphalitis and 6 infected cysts) received antibiotic treatment and were followed up. Two patients had reinfection and were then treated surgically. In four patients, spontaneous resolution was achieved after 3, 15, 18, and 24 months. In one patient (currently aged 17 years), resolution had not been achieved after 48 months of follow-up, although the size of the cyst had decreased; follow-up continues at the time of writing. In the remaining two symptomatic cases (fistulas), one patient underwent surgery (see Methods); the other one was managed conservatively, and spontaneous closure of the fistula was achieved after 2 months of follow-up.

In summary, in eight patients (61.5%) spontaneous resolution was achieved. In these cases, the time between diagnosis and resolution ranged from 2 to 24 months (median 16.5 months). Two patients with persistent non-infected cysts are still being followed (48 and 72 months after diagnosis). In both cases, surgical resolution was proposed after the patients' parents were informed about the risk of malignant transformation in adulthood, but this option was declined because of the favorable course during follow-up.

Table 1 Characteristics of the patients with urachal anomalies in our case series.

Case	Sex	Age	Anomaly	Diagnosis	Initial treatment	Outcome (time after diagnosis)
1	Male	1 day	Fistula	Umbilical urinary discharge	Surgical resection (large fistula)	Surgical resolution (2 months)
2	Male	11 days	Fistula	Umbilical secretion, periumbilical swelling	Under observation	Spontaneous resolution (2 months)
3	Male	13 days	Double cyst	Purulent umbilical secretion	Antibiotics	Reinfection: surgical resolution (6 months)
4	Male	25 days	Cyst	Fever, serous secretion, omphalitis	Antibiotics	Spontaneous resolution (18 months)
5	Female	3 years	Cyst	Incidental	No specific treatment	Spontaneous resolution (18 months)
6	Female	6 years	Sinus/cyst	Incidental	No specific treatment	Spontaneous resolution (12 months)
7	Male	6 years	Cyst	Periumbilical pain, fever, urinary symptoms	Antibiotics	Re-infection: surgical resolution (1 month)
8	Male	6 years	Cyst	Incidental	No specific treatment	Spontaneous resolution (18 months)
9	Male	11 years	Fistula	Omphalitis	Antibiotics	Spontaneous resolution (3 months)
10	Male	13 years	Cyst	Omphalitis, abdominal pain and vomiting	Antibiotics	A smaller sized cyst on the annual ultrasound scan (48 months)
11	Male	14 years	Cyst	Umbilical secretion, low-grade fever, leukocytosis	Antibiotics	Spontaneous resolution (15 months)
12	Male	14 years	Cyst	Protruding infraumbilical lesion and abdominal pain	Antibiotics	Spontaneous resolution (24 months)
13	Male	14 years	Cyst	Incidental	No specific treatment	A smaller cyst on the annual ultrasound scan (72 months)

**Figure 1** Flow-chart describing the presentation, management, follow-up and resolution of the 13 cases of urachal anomalies.

Discussion

The results presented in this study are consistent with those in the reviewed literature because a greater incidence of urachal anomalies has been noted in males [10] and urachal cysts have been confirmed as the most common anomaly [5]. The symptoms observed in this study are also consistent with those in previous reports: the presence of umbilical urinary discharge with periumbilical inflammation associated with the existence of a patent urachus, and the clinical manifestations of an infected urachal cyst, such as fever, peri-umbilical abdominal pain, and urinary symptoms with or without infection that was occasionally accompanied by a palpable suprapubic mass [1,9]. It is well known that of all of the possible complications of urachal cysts (such as progressive growth, infection, stones, intracystic bleeding, intraperitoneal rupture, bowel fistula, bowel obstruction, urinary tract infections, Reiter's disease, and malignancy) [5,8,13], infection is the most common complication, and it is usually caused by the migration of bacteria, particularly *Staphylococcus aureus*, from the umbilicus.

Regarding the chosen therapeutic option, there are opposing opinions on whether prophylactic surgery should be used in all patients with urachal anomalies. Several authors suggest the systematic excision of urachal lesions that are detected in infancy to prevent infection, reinfection, or other problems in adulthood [1,5,14]. However, other authors propose conservative management, even in cases of infected anomalies [2,7]. Furthermore, Copp et al. [6] suggest that surgery should be performed only in cases with unfavorable histology that is suggestive of malignancy (i.e. cases with transitional epithelium, squamous metaplasia, intestinal metaplasia, or mixed metaplasia).

In our case series, we preferred to adopt a conservative strategy, restricting surgery for the removal of a persistent patent urachus and reinfection of unresolved urachal cysts in two cases. Although laparoscopic surgery is currently the most recommended surgical approach [15–17], we opted for open surgery because of the location and characteristics of the lesions. Otherwise, antibiotic treatment was chosen for cases of infection, reducing the inflammation to ensure the confinement of infection, as proposed by most authors [5,8,13].

Our approach (i.e. close monitoring of the lesion with periodic ultrasound and without surgical intervention) was successful in eight of the 13 cases (61.5%). In two additional cases, a reduction in the size of the cysts favored the continuation of the conservative approach, although the need for surgery should not be ruled out. Although malignancy of urachal anomalies accounts for only 0.034% of all bladder tumors [2,4,18], the risk of this complication in children with urachal anomalies is unknown, mainly because of the small number of cases and the extended follow-up that is required for making this assessment (taking into account that most bladder tumors occur from 40 years of age onward).

Conclusions

With respect to the controversy regarding the best therapeutic option for the management of urachal anomalies,

the results obtained in this case series suggest that with the exception of cases in which there is a clear indication for surgery (i.e. reinfection), a conservative approach based on regular monitoring may be useful, particularly in those cases with an incidental diagnosis. Using this approach, many lesions will spontaneously resolve, the most common complications will be prevented or treated early, and the option of surgical intervention will still be available for those cases in which the spontaneous resolution of lesions is incomplete after a pre-specified follow-up period. Recent improvements in ultrasound technology [19] support the use of this imaging technique in the follow-up of these children. Although the documented sensitivity of ultrasound is acceptably high (near 80%) [19], repeated explorations during long-term follow-up (up to 5 years in our study) seem advisable to rule out false-negative results.

Undoubtedly, our results are not conclusive: further longitudinal studies with long-term follow-up periods (ideally up to adulthood) will be required to accurately estimate the risk of malignant transformation. Cost-effectiveness analyses would also be potentially valuable.

Ethical approval

Not required.

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Conflict of interest

None.

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