

The role of laparoscopy in the management of urachal anomalies in children

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Objectives Management for urachal anomalies (UAs) is controversial. Although traditional treatment of UAs has been surgical excision, recent literature report also a conservative approach. We reviewed our experience to define the role of laparoscopy in the management of UAs in children.

Patients and methods From July 2005 to July 2015, 23 children underwent 24 interventions for the treatment of UAs. In four patients, the technique was a laparoscopic-assisted removal of the anomaly, in two patients a laparoscopic-assisted drainage of an urachal abscess, and a pure laparoscopic technique was started in 17 patients.

Results Laparoscopic-assisted removal of the UAs was achieved in five cases. In two cases a laparoscopic-assisted drainage of an urachal abscess was performed. The remaining patients were treated by pure laparoscopic technique. Operative or postoperative complications did not occur. Follow-up ranged from 6 months to 10 years and 6 months.

Introduction

Urachal anomalies (UAs) represent an incomplete regression of the urachus. The reported incidence ranges from 1:150.000 in infants to 1:5000 in adulthood [1–3], but the true prevalence is still unknown [4].

UAs may be symptomatic and are present especially in young children [2–4].

Historically, the suggested management of UAs was their surgical excision to avoid recurrence of symptoms and to preclude a possible malignant transformation later in life [2] even if recent literature also reports a conservative management [5]. In patients younger than 6 months, UAs are likely to resolve with a nonoperative management, but in those with recurrent infections surgical treatment is advocated [6]. It remains unclear whether asymptomatic UAs require surgical excision to avoid a malignant degeneration because it is not possible to predict this event.

In recent years, laparoscopic surgery had a wide diffusion as an alternative approach to open surgery for the complete excision of UA. This technique appears simple and feasible, enabling both sufficient ligation of the urachus and thorough excision of the anomaly.

Here, we report our experience with the laparoscopic management of UAs and we reviewed literature to suggest whether to perform surgical excision or conservative management for UAs, in particular for those without symptoms.

Patients and methods

From July 2005 to July 2015, 23 children underwent 24 interventions for management of UAs. Diagnosis of UAs was always obtained by ultrasound (Fig. 1).

Conclusion The pure laparoscopic approach to UAs appears safe and effective in most urachal remnants. Laparoscopic-assisted excision is an alternative approach that is easier to perform in infants. The decision to remove the UAs must be taken after an accurate informed consent of the parents, especially in cases of asymptomatic anomalies. *Ann Pediatr Surg* 13:85–90 © 2017 Annals of Pediatric Surgery.

Annals of Pediatric Surgery 2017, 13:85–90

Keywords: laparoscopy, management, urachal anomalies

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Received 17 May 2016 accepted 4 February 2017

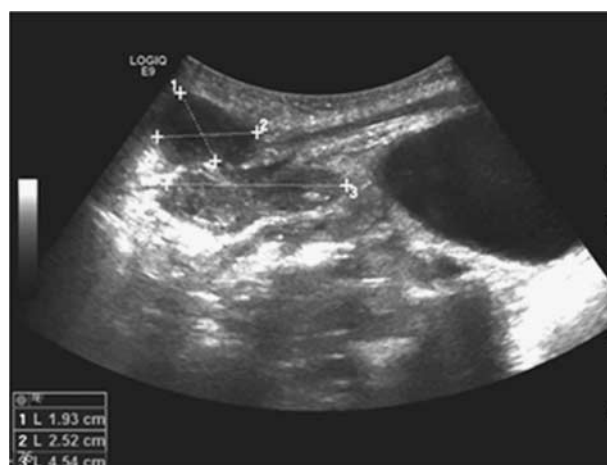
In six patients, the UA was asymptomatic, whereas 17 patients were symptomatic.

In the case of asymptomatic patients, diagnosis was carried out incidentally with an ultrasonography performed for other reasons.

The technique used in four patients was laparoscopic-assisted excision of the remnant [7].

In our first case, a laparoscopic-assisted excision of an urachal cyst was performed using a 10-mm umbilical port and two 5-mm operative ports in the right and left flanks, respectively. The first step of the intervention was the

Fig. 1



Ultrasonographic image of an urachal abscess.

dissection of the infected UA, and then we exteriorized it through the umbilical incision and its excision was performed.

In another two cases of urachal sinus, excision was performed by a 5-mm telescope placed at the right midclavicular line at the subcostal level. After the individuation of the whole UA, we proceeded with its open excision of both through a 10-mm subumbilical incision.

In a case of patent urachus associated with a patent omphalomesenteric duct in a 28-day-old boy, a left flank position was used to insert a 5-mm camera. After the individuation of both remnants as previously suspected by ultrasound, we proceeded with an open excision through a 10-mm subumbilical incision.

The telescope was placed at the right midclavicular line at the subcostal level also for the laparoscopic-assisted drainage of two urachal abscesses.

Pure laparoscopic technique was started in 17 cases. Patients were placed in Trendelenburg's position. We used a particular trocar's position [8]. The camera port was inserted at the midclavicular subcostal right space. Carboperitoneum was established from 6 to 10 mmHg according to the patient's weight. Another two trocars (from 3 to 5 mm) were inserted in the right flank and in the superior left quadrant (Fig. 2).

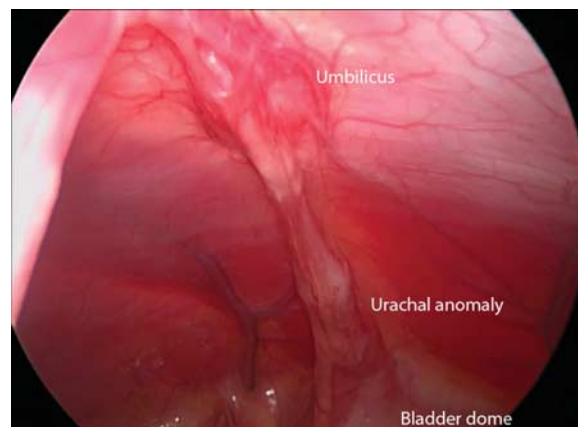
Owing to this camera position, a clear vision of the whole UA in its whole length was obtained (Fig. 3). The dissection of the entire remnant was accomplished from the umbilicus level to the bladder dome. The remnant was closed caudally with a 2.0/3.0 endoloop ensured with a 2.0/3.0 transfixed suture. A minimal excision of the bladder dome was performed to avoid possible recurrences. Bladder sealing was then confirmed by filling it with methylene blue under direct vision.

Fig. 2



Trocar positioning for laparoscopic excision of urachal anomaly.

Fig. 3



The picture shows the laparoscopic vision of the whole urachal anomaly through the camera inserted at midclavicular subcostal right level.

The specimens were then extracted in all cases from the abdomen through one 5-mm port incision (Fig. 4). In all cases, if a macroscopic umbilical fistula was found, an umbilical fistulectomy was performed.

This retrospective study was approved by the Ethics Committee of S. Maria della Misericordia Hospital of Perugia, Italy, and was conducted in accordance with Declaration of Helsinki.

Results

Twenty-three children (median age: 5.1 years, range: 40 days–14 years) (M:F 4.7:1) with UAs underwent 24 laparoscopic procedures. Seventeen patients were symptomatic, whereas six were asymptomatic. The manifestations of symptomatic UAs in our series were omphalitis, umbilical discharge, abdominal pain, and urinary symptoms. Table 1 shows symptomatology associated with different types of UA.

All UAs were diagnosed preoperatively by ultrasonography and confirmed by laparoscopy. In all cases, laparoscopy showed the right preoperative diagnosis obtained with ultrasonography except in one case. In a patient with a urachal sinus associated with a urachal diverticulum, ultrasonography showed only the sinus while a urachal diverticulum was associated. In two cases, an additional computed tomography and an MRI were performed, respectively, for two infected urachal cysts. Associated anomalies were present in 9/23 (39.1%) patients, and the most frequent was inguinal hernia (6/23) (26%) in one case associated with hypospadias diagnosed also in another patient with undescended testis.

Four patients underwent a laparoscopic-assisted intervention, and two patients had laparoscopic drainage of an urachal abscess. One of the patients treated for an abscess was then submitted to a delayed laparoscopic excision of an urachal cyst. This patient was also submitted to a laparoscopic right inguinal hernia repair during the laparoscopic UA removal. The remaining patients were treated by pure laparoscopic technique, but in one case laparoscopy was converted to a laparoscopic-assisted

Fig. 4



The excised urachal anomaly.

excision of the remnant. Table 2 shows details of sex and age of patients, preoperative diagnosis of UA type, operative technique adopted, type of UA found at laparoscopy, and operative time.

Overall operative time ranged from 20 to 120 min (median: 64.6 min).

Operative time for laparoscopic-assisted excision ranged from 40 to 120 min (median: 84 min), whereas for the laparoscopic procedures the operative time ranged from 40 to 90 min (median: 60.2 min), respectively. In the case of two laparoscopic-assisted drainage of urachal abscess, the operative time was 20 min for both procedures. Postoperative hospital stay ranged from 2 to 5 days (median: 3 days).

In all cases, operative or postoperative complications did not occur, and the cosmetic results were good. Postoperative follow-up ranged from 6 months to 10 years and 6 months (median: 3 years and 9 months), and it was performed by scheduled ultrasonographies at 1, 6, 12 months, and then yearly, which showed complete excision of the UA in all cases.

Discussion

The urachus is a cord-like structure between the bladder and the umbilicus representing the embryologic remnant of the allantois. In the case of the presence of UAs in

children, an incomplete regression of the urachus occurs and may be found at various levels. Therefore, this rare congenital anomaly may differentiate into different types, including cyst, sinus, diverticulum, and patent urachus, and we must consider also the possibility of association among these four types of anomalies. Although obliteration of the urachus was originally thought to be a prenatal occurrence [9], recent literature suggests that this process may occur in the postnatal period, especially during the first year of life [6,10].

UAs may be associated with other urogenital anomalies [11], and in our experience the most frequent was inguinal hernia (26%).

Most UAs in the pediatric age group present symptomatically, but occasionally asymptomatic remnants are incidentally discovered on radiographic imaging [6,12].

In all patients, preoperative ultrasound showed the right diagnosis of UA, except in one case in which together with the diagnosed sinus a urachal diverticulum was present (95.6%).

In our experience, 17 out of 23 treated patients were symptomatic, whereas six were asymptomatic with an incidental diagnosis obtained during an abdominal ultrasonography performed for other reasons. Asymptomatic patients were all affected by urachal cysts.

Acute infections of urachal cysts can be treated mostly by antibiotic therapy and drainage [13].

The laparoscopic-assisted removal of the UA was performed in three cases of sinus (including the patient in whom conversion occurred), in one case of cyst, and in a 28-day-old patient with associated patent urachus and patent omphalomesenteric duct. In this case, a left flank position was used to insert a 5-mm camera. This position for the camera was chosen because of patient's age, as the liver exceeded the right lower costal line, a typical situation at that age. After the individuation of the whole remnants, we proceeded easily with an open excision of both through a 10-mm subumbilical incision. In our opinion, the laparoscopic-assisted technique was easier to perform in infants because of the short length between the umbilicus and bladder dome. In fact, when this technique was performed in older patients, it appeared more difficult to accomplish.

We treated three cases of UAs with laparoscopic-assisted technique at the beginning of our experience. In another case, we converted the laparoscopic intervention to a

Table 1 Symptomatology of patients associated with different types of urachal anomalies

Symptoms	Cyst	Sinus	Cyst + sinus	Sinus + diverticulum	Patent urachus	Total [n (%)]
Acute abdominal pain and stranguria	2	–	–	–	–	2 (8.7)
Acute abdominal pain, stranguria, and omphalitis	–	1	–	–	–	1 (4.3)
Umbilical discharge and omphalitis	–	3	1	–	–	4 (17.4)
Umbilical discharge	–	1	1	1	1	4 (17.4)
Omphalitis	–	2	–	–	–	2 (8.7)
Urinary tract infections	1	–	–	–	–	1 (4.3)
Abdominal pain	3	–	–	–	–	3 (13)
Asymptomatic	6	–	–	–	–	6 (26)
Total	12	7	2	1	1	23

Table 2 Details of sex and age of patients, preoperative diagnosis of urachal anomalies type, operative technique adopted, type of urachal anomalies found at laparoscopy, and operative time

Case nos	Sex	Age	Preoperative US diagnosis	Treatment	Conversions	UA type	Operative time (min)
1	M	7 years	Cyst	Lap.-assisted removal		Cyst	120
2	M	1 year	Sinus	Lap.-assisted removal		Sinus	60
3	F	4 years	Sinus	Lap.-assisted removal		Sinus	40
4	M	11 years	Cyst	Lap. removal		Cyst	70
5	M	12 years	Cyst	Lap. removal		Cyst	70
6	M	4 years	Cyst	Lap. removal		Cyst	60
7	M	4.5 years	Cyst	Lap. removal		Cyst	65
8	M	9 months	Sinus	Lap. removal	Lap.-assisted removal	Sinus	110
9	M	2.6 years	Cyst	Lap. removal		Cyst	65
10	M	3 years	Cyst	Lap. removal		Cyst	60
11	M	1 year	Sinus	Lap. removal		Sinus	40
12	F	13 years	Sinus + cyst	Lap. removal		Sinus + cyst	90
13	F	2.5 years	Unidentified urachal abscess	Lap.-assisted drainage	Lap. removal + lap. right inguinal herniorrhaphy	Infected cyst/urachal abscess	20
14	M	2 years	Infected sinus/urachal abscess	Lap.-assisted drainage		Infected sinus/urachal abscess	70
15	M	6 years	Cyst	Lap. removal		Cyst	20
16	M	6.5 years	Cyst	Lap. removal		Cyst	65
17	M	5.5 years	Cyst	Lap. removal		Cyst	55
18	M	6 years	Sinus + cyst	Lap. removal		Sinus + cyst	60
19	M	1.5 years	Sinus	Lap. removal		Sinus + diverticulum	60
20	F	14 years	Sinus	Lap. removal		Sinus	45
21	M	1 year	Sinus	Lap. removal		Sinus	40
22	M	1 month	Patent urachus + suspected patent omphalomesenteric duct	Lap.-assisted removal		Patent urachus + patent omphalomesenteric duct	90
23	M	8 years	Cyst	Lap. removal		Cyst	50

F, female; Lap., laparoscopic; M, male; UA, urachal anomaly; US, ultrasound.

laparoscopic-assisted technique because of the important postinflammatory situation. In the case of the association between patent urachus and patent omphalomesenteric duct after the initial individuation of the anomalies a single subumbilical incision in a newborn was sufficient to remove both anomalies and cosmetically comparable with scars obtained with laparoscopy.

We treated two patients with urachal abscess by laparoscopic-assisted drainage. We did not perform drainage of abscess under ultrasonographic guidance because when grossly infected UAs can break into the peritoneal cavity causing peritonitis [14,15]. With laparoscopic drainage of urachal abscesses, it is possible to check a possible peritoneal contamination and to perform a peritoneal toilette if necessary.

The remaining patients were treated by pure laparoscopic technique. In one patient during the removal of the UA, a laparoscopic right inguinal hernia repair was also performed [16].

Although in literature a surgical approach is suggested for the treatment of these anomalies for their potential recurrent inflammation and for the risk of malignant degeneration, in recent years a conservative management has been advocated from some authors.

In the case of symptomatic UAs, Lipskar *et al.* [5] in a retrospective study affirmed that nonoperative

management is a reasonable approach because in some patients spontaneous resolution could be expected. The authors affirm also that this conservative approach may be extended to infected urachal cysts after initial drainage.

Sun *et al.* [13] showed instead a series of recurrent infections after surgical drainage of UAs due to urachal sinus. In our experience, the opposite of what the last two authors claim occurred. We performed two laparoscopic-assisted drainages of urachal abscess, respectively, because of an urachal cyst and an urachal sinus. In the case of urachal sinus, a progressive obliteration leading to the complete disappearance of the anomaly occurred. On the other hand, an urachal cyst in a young female only partially reduced in dimensions and did not improve with time, maintaining the same size at 3 and 6 months postoperatively. In this last case, we decided to remove the UA, with the parents' consent.

In our opinion, symptomatic UAs should be managed case by case. Indication for surgical removal should be reserved to those UAs with recurrent symptoms or in the case of persistence of UA without signs of regression after an abscess drainage.

In the case of asymptomatic UAs, the main debate between a surgical or a conservative management concerns the possibility of a malignant degeneration later in life.

Naiditch *et al.* [17] claim that conservative management of asymptomatic and incidentally identified UAs can safely be managed nonoperatively without the risk of developing complications in the short term and a high likelihood of spontaneous resolution, regardless of the type of UA identified and because complications after surgical excision of UAs are not uncommon (14.7%). On the contrary, Ashley *et al.* [18] recommended removing all recognized pediatric UAs, as the ability to predict malignant transformation is poor and the morbidity of surgical excision is low.

In our experience, asymptomatic patients were treated after a minimum of 1 year of ultrasound follow-up testifying the increasing size of the urachal cysts or the absence of regression. In these patients, parents decided to perform surgical removal after a careful informed consent about treatment options. In all these patients treated by laparoscopic excision of the UA, operative or postoperative complications did not occur.

It is unclear whether UAs should be excised to prevent future malignancy even if malignant transformation is rare but possible also in the pediatric age group [18].

In adults, urachal carcinoma is responsible for less than 1% of bladder cancer cases, but its aggressiveness and poor overall survival inferior to 50% led some authors to believe that a more proactive approach may be reasonable [19–21]. Nevertheless, Gleason *et al.* [22] in a recent publication affirm that for asymptomatic UAs in particular a large number would need to be removed (5.721) to prevent a single case of adenocarcinoma of the urachus. Therefore, they preferred a conservative attitude.

Literature shows that overall urachal cancers represent from 1–10% of all cancers of the bladder, and most of them are of the epithelial type [23,24]. UAs containing gastrointestinal or metaplastic epithelium should be those that can more easily have a potential malignant transformation, as approximately 95% of urachal cancers are of the epithelial subtype [19,20]. In fact, less than 5% of urachal tumors such as sarcomas do not have an epithelial origin [19].

Nevertheless, urachal rhabdomyosarcoma in children seems to have a poor outcome [25]. Thus, not only the presence and type of epithelium should be considered for a possible malignant degeneration but also the type of stroma.

Once it is established that UAs without epithelial tissue apparently carry little risk of malignant transformation, it is very difficult or almost impossible to predict which UA could have an epithelium, on the basis of preoperative studies.

Although in our experience surgical outcomes remain excellent, UA management remains a challenge in particular for asymptomatic cases, and the therapeutic program should be decided including and informing the family as much as possible about different managements.

Based on our experience, the laparoscopic management of UAs in the pediatric age group appears to be safe and

effective, allowing a radical excision of the remnants with all the advantages of this procedure. Furthermore, intraoperative and postoperative complications in this series were absent. In the case of decision for surgical removal, we emphasize the use of laparoscopy. Laparoscopy permits to have the final diagnosis, to accomplish the intervention in a good ranging time with good cosmetic results, and to treat other associated anomalies such as inguinal hernia with the same trocar's disposition.

Laparoscopic-assisted technique by a 10-mm subumbilical incision could be alternative minimally invasive techniques to adopt for the removal of UAs in infants. In the case of rare associated anomalies, such as patent omphalomesenteric duct, laparoscopy may be useful for diagnosis confirmation.

Recent trends in the management of UAs suggest a conservative approach especially for asymptomatic cases, because UAs with no epithelial tissue apparently have little risk of malignant transformation. Nevertheless, we cannot predict which patients do not have epithelium in the remnants. Thus, the decision whether or not to remove an asymptomatic UA must always be taken together with the children's parents by providing accurate information about a possible malignant degeneration but also concerning the possibility of a nonoperative management because of the inability to predict which UAs will undergo malignant transformation.

Conflicts of interest

There are no conflicts of interest.

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