Urachal Anomalies in Children: Experience at One Institution

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Background: The embryological and anatomical features of urachal anomalies have been well defined. Because of the variable clinical presentations, uniform guidelines for evaluation and treatment are lacking. In an attempt to establish an optimal diagnostic and treatment modality, we report our experience with urachal anomalies at a single institution over a 10-year period.

Methods: The records of 20 patients with urachal abnormalities were reviewed. These included 12 males and 8 females with ages from 1 day to 12 years (average, 3 years). The evaluation included symptoms and signs, and results of fistulography, sonography, and voiding cystography. Postoperative conditions were also reviewed.

Results: The presenting complaint was umbilical discharge in 14 patients, umbilical discharge with marked umbilical granulation tissue in 2, periumbilical erythema in 3, and abdominal pain in 1. Diagnostic evaluation included fistulography in 5 cases, sonography in 13, and voiding cystourethrography in 3. The 3 variants of urachal anomalies included a patent urachus in 4 patients (20%), urachal sinus in 13 (65%), and an infected urachal cyst in 3 (15%). Treatment consisted of primary excision with a cuff of the bladder in 3, excision with ligation in 1, excision of the sinus in 13, incision and drainage in 3, and secondary excision in 1. There was 1 postoperative wound infection.

Conclusion: Diagnosis and treatment of urachal anomalies can be made with certainty if a good physical examination and proper imaging study are performed. Voiding urethrocystography might not be required in view of the fact that none of the patients studied had an associated urinary tract anomaly.

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Key words: **urachal sinus, patent urachus, urachal cyst, fistulography, sonography.**

The urachus is a normal embryonic remnant of the primitive bladder dome.[1,2] It generally exists as a fibrous cord extending from the dome of the bladder to the umbilicus. It also occupies the potential midline space between the peritoneum and the transversalis fascia.[3] The urachus is present in children at birth and gradually regresses afterwards. It is found in only 1/3 of adults.[4] Its failure to obliterate results in various anomalies, some of which are clinically symptomatic. Urachal remnants often give rise to such problems as fatal infection and late malignant change.[5] Acknowledging the anatomy and embryology and having a high degree of suspicion are required to establish a correct diagnosis.

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Because of the variable clinical presentations, uniform guidelines for evaluation and treatment appear to be desirable. In an attempt to establish the best diagnostic and treatment modality, we report our experience in dealing with these anomalies at a single institution over a 10-year period

METHODS

The medical records of patients with urachal anomalies at Chang Gung Children's Hospital were reviewed for the 10-year period between 1992 and 2002. The diagnosis was confirmed at surgery in all patients. Urachal anomalies were identified in 20 patients, including 12 males and 8 females, aged 1 day to 12 years (average, 3 years).

The evaluation included symptoms and signs, and results of fistulography, sonography, and voiding cystourethrography. Treatment consisted of primary excision, incision and drainage, and delayed excision after a period of antibiotic administration with an initial drainage procedure. Postoperative conditions were also reviewed.

RESULTS

A patent urachus was diagnosed in 4 patients (3 neonates and 1 preschooler). Two neonates with a patent urachus presented an obvious wet umbilicus with surrounding granulation tissue (Fig. 1). One neonate presented intermittent urinary drainage that was accentuated with crying or voiding. One child presented with a wet umbilicus. Fistulography revealed a patent urachus in 3 of the 4 patients (Fig. 2). Three of the 4 patients also showed communication of the patent urachus with the bladder with no urinary tract anomaly documented by voiding cystourethrography. Three neonates had a big lumen which was excised with a cuff of the bladder. Another child had a small lumen, which was diagnosed at the time of surgery, and ligation of the urachus with transection at the level of the bladder was performed.

Urachal sinus was diagnosed in 13 patients (3 infants, 6 preschoolers, and 4 school-aged children). Twelve of the 13 patients presented with periumbilical drainage, while the other had abdominal pain. Fistulography revealed the presence of a fistula in 2 cases. Eleven of the 13 patients were diagnosed by sonography (Fig. 3). For lesions that did not communicate with the bladder, only the umbilical tract to the urachal portion of the sinus was excised. A urachal cyst was diagnosed in 3 patients, all of whom presented with infection and developed an abscess. Sonography revealed abscess formation,

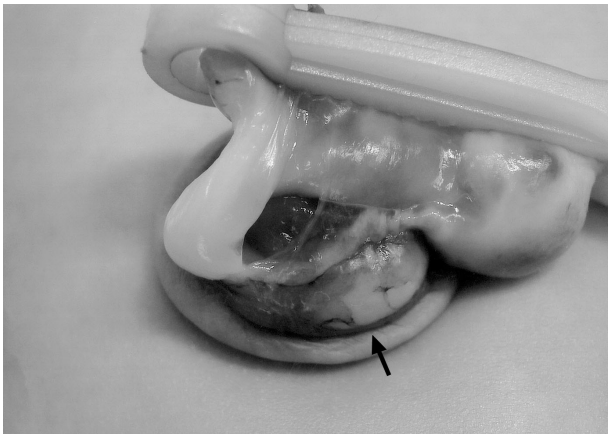

Fig. 1 Large, everted umbilicus with visible mucosa (arrow) and fluid discharge.

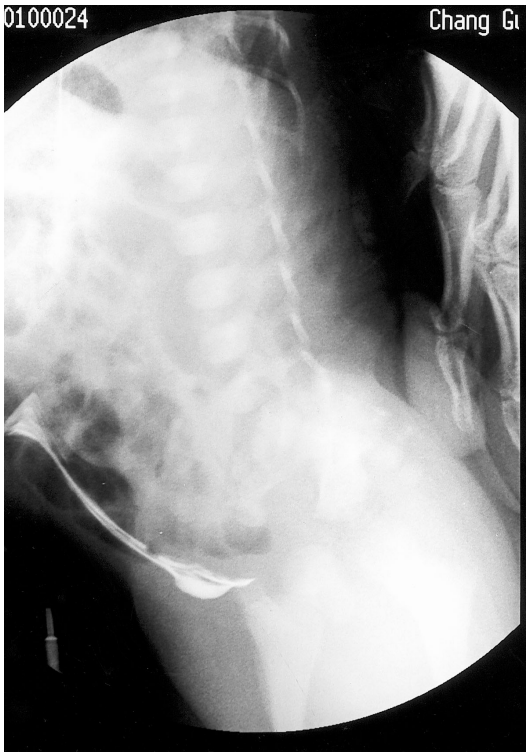

Fig. 2 Fistulography showing a patent urachus with communication with the bladder.

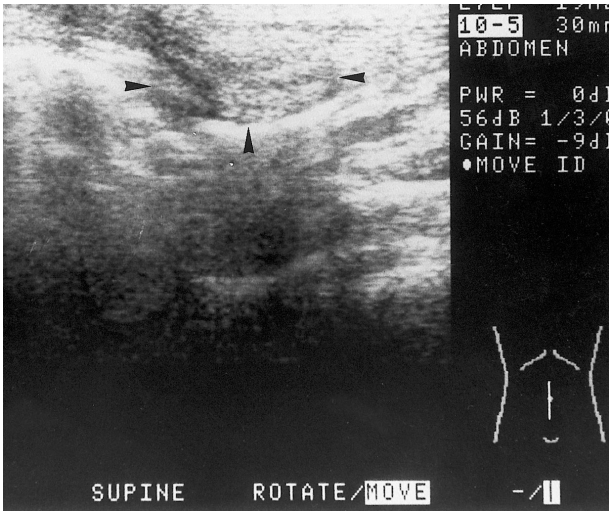

Fig. 3 Sonography revealing a urachal sinus (arrowhead).

and an infected urachal cyst was suspected. Two of the 3 patients received incision and drainage only. Another received delayed excision of the residual cyst after initial antibiotic treatment. *Staphylococcus aureus* was the predominant organism isolated from the pus. Postoperative wound infection occurred in 1 patient. None developed any long-term sequelae.

DISCUSSION

Urachal anomalies are rare, and most present in early childhood.[6,7] Four clinical urachal anomalies have been described: a patent urachus, urachal cyst, urachal sinus, and vesicourachal diverticulum.[8-10] A urachal sinus was the most common diagnosis in our series. It occurs in infancy or childhood.[11,12] Sonography was the most accurate modality for diagnosis in these patients. The small fistula tract and sinus can be clearly be seen from the ultrasound image. This may be because the preperitoneal space is free of interfering bowel gas.[13-15] An extraperitoneal excision is the treatment of choice for patients with urachal sinus. No postoperative complication was noted in this series.

A patent urachus was the next common diagnosis in this series. It occurred most often in neonates with a prominent everted large umbilicus along with visible mucosa and a large fistula tract. Fistulography with the use of radiopaque contrast medium was helpful in the diagnosis. The concern that a patent urachus may be secondary to an infravesical urinary tract obstruction has justified the use of voiding cystourethrography with or without subsequent endoscopy to investigate this anomaly.[16,17] Voiding cystourethrography was performed in all our cases to identify the fistula tract and, more importantly, to rule out the concomitant presence of bladder outlet obstruction or vesicoureteral reflux. In contrast to the study of Herbst,[18] none of our patients had infravesical lesions. Therefore, it is unlikely that a persistent urachus was directly associated with the obstruction. Excision of the urachal tract to the bladder through an extraperitoneal approach with or without inclusion of the bladder cuff is the treatment of choice.

Most urachal cysts in this series were infected, with pain, tenderness, erythema, and localized swelling in the infraumbilical region, when diagnosed. Sonography always revealed abscess formation,[19] and an infected urachal cyst was thus suspected. Incision and drainage comprised the initial treatment[20] in 3, and in only 1 patient did we need to remove the residual cyst remnant owing to persistent wound infection. *Staphylococcus aureus* was the most common bacterium cultured in this study.

In conclusion, urachal abnormalities are rare. The optimal diagnostic imaging study depends on the clinical presentations. Our experience suggests that in young infants and neonates suspected of having a patent urachus, fistulography should be performed. The treatment of choice is excision of the patent urachus with or without inclusion of the bladder cuff. Any child who presents with a wet umbilicus should receive a sonographic examination to rule out the possible diagnosis of a urachal sinus. Surgical excision of the sinus is the goal of treatment. Nevertheless, most urachal cysts are asymptomatic until they become infected. Incision and drainage or delayed excision with initial antibiotic therapy are the treatment of choice. Voiding cystourethrography does not seem necessary in view of the fact that none of the patients studied had an additional associated urinary tract anomaly.

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在兒童發生的臍尿管異常：一個機構的經驗

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背 景： 在胚胎學和解剖學上有關臍尿管異常的特色已被很詳細的界定，由於各有許多不同的臨床表現，所以缺乏一致性的診斷及治療模式。我們統計過去本院十年的經驗，期望能找出一致性的診斷及治療模式。

方 法： 從1992 到2002十年間共有20例兒童臍尿管異常案例，12位是男生，8位是女生，年齡從1 天到12歲 (平均年齡是3歲)，病人的臨床症狀和影像檢查是我們診斷的依據。影像檢查則包括瘻管攝影術，超音波和膀胱尿道排出攝影法，至於術後的情況也一併分析。

結 果： 在表現的症狀中，僅有肚臍分泌者有14位 (70%)，合併肚臍分泌及肉芽組織者有2位 (10%)，肚臍紅腫者有3位 (15%)，腹痛有1位 (5%)，診斷法包括瘻管攝影術5例，超音波13例，膀胱尿道排出攝影法有3例。臍尿管異常的三種表現：開放性臍尿管有4例 (20%)，臍尿管竇有13例 (65%)，感染性臍尿管囊腫有3例 (15%)，治療法包括臍尿管及部分膀胱切除有3位新生兒病人，切除並結紮有1人，切除臍尿管竇及瘻管者有13例，3例感染性臍尿管囊腫則接受切開引流，其中1位日後又再進行第二次切除手術，術後發生1例傷口感染。

結 論： 臍尿管異常的病例，如有良好的理學檢查及合宜的影像檢查則可獲得正確的診斷，進而獲得良好的治療。在這20位臍尿管異常的病例中，膀胱尿道排出攝影法並沒發現合併其他泌尿道異常，也許此種檢查並非必要。

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關鍵字： 臍尿管竇，開放性臍尿，臍尿管囊腫，瘻管攝影，超音波。

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