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Research Article

URACHAL ANOMALIES IN CHILDREN: OUR EXPERIENCE

***Yogender S. Kadian[1], Rattan K.N. [1], and Parihar D.[2]**
[1]Deptt. of Paediatric Surgery, Pt. B.D. Sharma PGIMS, Rohtak & [2]Department of Surgery, B P S, Government, Medical College for Women, Khanpur Kalan, Sonepat
**Author for Correspondence*

ABSTRACT
The urachus is a tubular structure in intrauterine life and the failure of obliteration of its lumen can lead to various anomalies in post natal life namely; patent urachus, urachal sinus, urachal cyst or urachal diverticulim. These urachal anomalies may have symptomatic presentation such as watery umbilical discharge, swelling in the umbilical area or remain asymptomatic to be detected incidently on imaging for other urinary symptoms. The mainstay of diagnosis of rests on history and physical examination and ultrasonography. C T Scan or voiding cystourethrography are rarely needed for reaching a diagnosis of urachal anomalies. These anomalies once detected should always be excised surgically for the fear of malignant potential of the transitional epithelium which they contain.

Key Words: *Urachus, Anomalies, Umbilical Discharge*

INTRODUCTION
The urachus is a tubular structure which connects the allantois at the umbilicus to the dome of the bladder during fetal development (Atala and Retik, 1993). The lumen normally closes at about the twelfth week of gestation and obliterates completely. The failure of complete obliteration of the lumen during gestation results in urachal anomalies in infancy and older children (Gearhart and Jeffs, 1998). The presentation of these anomalies can be in one of the following variants: patent urachus (entire tubular structure is intact), urachal sinus (the umbilical end fails to close), urachal diverticulum (the bladder end fails to close), urachal cyst(both ends close but the central lumen remains open (Moore, 1982). The reported incidence of the different variants of this anomaly is different in various series; some authors report urachal sinus or patent urachus as more common and others report urachal cyst (Cilento *et al.,* 1998; Mesrobian *et al.,* 1997). The common presenting symptoms in children with urachal anomalies are umbilical drainage (clear, serous, purulent, or bloody), or a mass with or without pain. These abnormalities are a frequent concern in newborns with umbilical drainage that persists beyond a few weeks. Moreover, urachal anomalies may be incidentally discovered during radiographic examinations during the evaluation of children with urinary tract infections or hydronephrosis (Blichert-Toft and Nielsen, 1971). In this report we are presenting our experience of managing the urachal anomalies in children.

MATERIALS AND METHODS
From January 20009 to December 2012, five cases of urachal anomaliest were managed in department of pediatrics surgery PGIMS rohtak, Haryana (India). In a retrospective review, case notes, including presentations, imaging studies, operative details, pathological findings and postoperative outcome were studied. Purpose of this study was to have a outlook of these anomalies regarding the presentation, methods for diagnosis and surgical management of these anomalies.

RESULTS
Out of five cases four were male and one was female, their age ranging from 6 months to 13 years, with a mean of 3.4 years. The patients (3) presentation was , one female child presented with a pouting umbilicus and there was history of passage of clear fluid (urine) from the umbilicus during the act of micturition and in two male patients there was history of passage of watery discharge from umbilicus sometimes mixed with blood since infancy. On ultrasonography of the urinary system a tubular connection between the urinary bladder and umbilicus could be demonstrated and a diagnosis of patent

urachus was made (Fig. A and B) . Routine blood and urine examination was done and was normal in these patients. Surgical exploration of the umbilical area by a small transverse and a vertical incision (reverse T shape) was done and the patent urachus excised completely. Histopathological examination revealed lining of the patent tract was of transitional epithelium and confirmed the diagnosis of patent urachus. Another patient, 5 year male child attended the paediatric surgery outpatient with recurrent history of pain abdomen and ultrasonography (USG) of abdomen and pelvis could localize a small cystic structure with well defined wall and fluid content in it near the dome of bladder and a possibility of urachal cyst was kept. Surgical excision of the cystic structure along with the a cuff of bladder dome was done and histopahological examination revealed it to the urachal cyst. Lastly a 13 year old male child admitted in the paediatric surgery ward with the diagnosis of primary urinary bladder stone. During the procedure of open cystolithotomy the urachus was found to be quite thick walled and its lower portion was dilated and opened into the bladder and a possibility of urachal diverticulum was kept (Figure C and Figure D). The bladder stone was removed and the diverticulum was excised along with a portion of dome of bladder. In postoperative period an indwelling urinary cathter was kept for one week and the histopahological examination was consistent with diagnosis of the urachal diverticulum.

DISCUSSION

The reported incidence of urachal anomalies is low. Drawson *et al.,* (1994) reported 2 cases in 300,000 hospital admissions and Nix *et al.,* (1958) reported 3 cases in 200,000 hospital admissions in Boston and 3 cases in 1,168,760 hospital admissions in New Orleans (Drawson *et al.,* 1994; Nix *et al.,* 1958). In our institute the exact incidence of this anomaly is difficult to assess but we have managed a total of five patients in four years. The low incidence may be because of the urachal anomalies are not easily encountered in clinical cases as they are frequently asymptomatic. The reported incidence rate in males is twice as high as in females (Choi *et al.,* 2006) but in our series this ratio is 4:1 in favour of males. Urachal anomalies are more common in infancy ranging from one day to two years of age. The majority of urachal anomalies can be classified into one of the following groups: patent urachus, in which the entire tubular structure is intact; urachal sinus, in which the umbilical end of the structure fails to close; urachal diverticulum, in which the bladder end of the structure fails to close; urachal cyst, in which both ends close but the central lumen remains open.

Combining data from various series in the shows the most common type of urachal anomaly to be cysts (45%), followed by sinuses (37%), then patent urachuses (16%) (Cilento *et al.,* 1998; Mesrobian *et al.,* 1997). However in other series the most common type of urachal anomalies is patent urachus (most common, 48%) followed by urachal cyst (31%), urachal sinus (18%) urachal diverticulum (3%) (Blichert-Toft and Nielsen, 1971). In our series 3/5patients were patent urachus, one each of urachal cyst and urachal diverticulum. However in this small series it is difficult to comment on the exact incidence of various anomalies.

The common way of presentation of these anomalies is periumbilical drainage. Other presentations include abdominal pain, periumbilical mass, periumbilical erythema and urinary symptoms. In our series; one of our patient presenting symptom was a urinary stream via urachus at the time of micturition which is a very rare mode of presentation (for patent urachus) and other anomaly, urachal diverticulum, was detected during the surgery for bladder stone. The differential diagnosis for this constellation of presenting symptoms includes anomalies of the vitelline ducts, appendicitis, omphalitis, and granulation tissue from an umbilical stump. In the differential diagnosis of abdominal pain, appendicitis is relatively common, while anomalies of the vitelline ducts are less common with a 2% overall incidence (Atala and Retik, 1993).

A thorough history and physical examination is necessary for the diagnosis of urachal anomalies. The imaging modalities for confirmation of the diagnosis includes ultrasonography, computed tomography, voiding cystourethrography and fistulogram (Ueno *et al.,* 2003; Yu *et al.,* 2001). Among these,

ultrasonography and computed tomography are reported as the best diagnostic tools. Ultrasonography is usually performed first, and if not definitive, computed tomography may subsequently be performed.

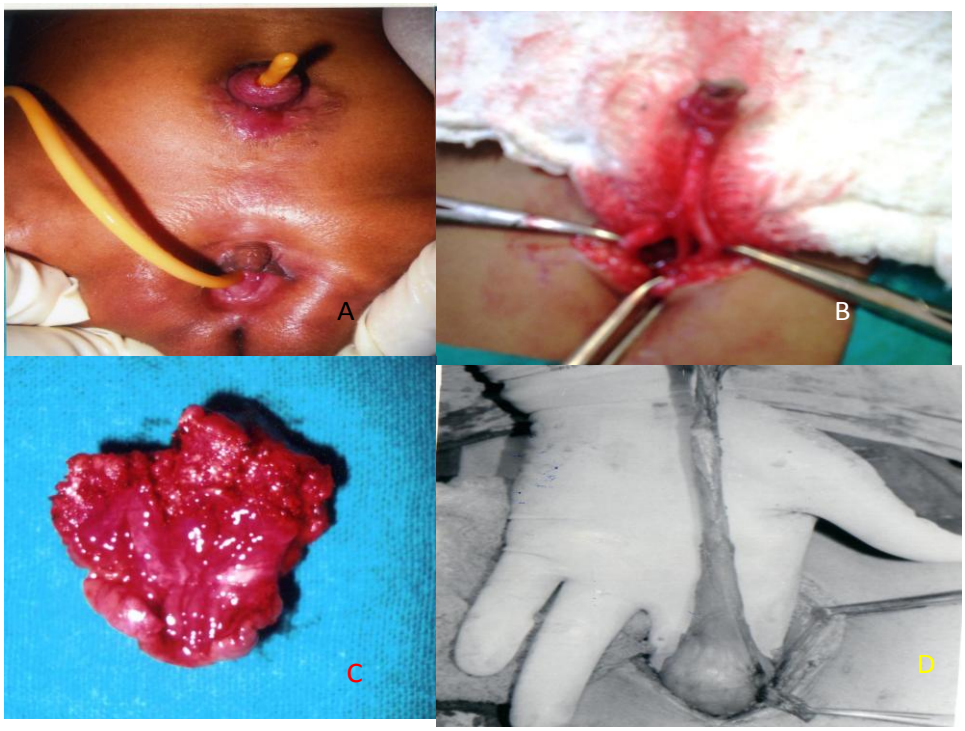

Figure 1: A- Clinical photograph showing patent urachus and Foleys Cather in situ; **B-** Operative photograph showing patent urachus; **C**- Excised urachal cyst photograph; **D**- Operative photograph of the urachal diverticulum

Although the chance of finding other associated anomalies of the genitourinary tract is low in patients with a patent urachus, the evaluation should also include a renal ultrasound to ensure the absence of hydronephrosis or other congenital kidney anomalies an urachal cyst greater in size than a few millimeters can be readily imaged by ultrasound (Holten, 1996).

They may be incidentally detected in asymptomatic children when the bladder is examined during routine ultrasonographic evaluation, such as after a urinary tract infection. In cases where there is diagnoses is uncertain, a computed tomography scan (CT) can give excellent anatomical deta. But because of risk of the radiation exposure in children the CT scan should not be considered an integral component of the routine workup. However, in cases where there is a high degree of clinical suspicion with a negative ultrasound (such as recurrent bloody umbilical drainage), a CT scan can be helpful in imaging the urachus and showing lesions which may be missed by ultrasound (Yu *et al.*, 2001). But in the present study CT scan was not required in any of the patient. A patent urachus that allows urine to drain freely through the umbilicus can also be imaged with a high degree of sensitivity with either a voiding cystourethrogram (VCUG) or sinogram. But these investigations should be used judiciously as the reported incidence of the lower urinary tract outflow obstruction in patent urachus is very low (Little *et al.*, 2005; McCollum, 2003). The incidence of concomitant kidney abnormalities has varied widely in published series, but given the lack of morbidity and risk with ultrasound, it is prudent to include imaging the kidneys as part of the work-up (Yu *et al.*, 2001).

The symptomatic urachal remnants should be treated with surgical excision. This should include complete excision of the urachus from the umbilicus to the dome of the bladder. In infants and small children complete resection of the urachus can easily be accomplished through a small incision. It can be oriented in either a transverse or vertical midline. For infants, a small 1-1.5 cm incision midway between the pubis and umbilicus will give access to the urachus and allow complete resection from the umbilicus to the dome of the bladder. However a small transverse combined with vertical incision (T shape) gives better exposure from umbilicus to the bladder and we have used this incision in the present series with good functional outcome. Recently Schaefer *et al.,* (2012) introduced a three step technique for umbilicoplasty in the surgical excision of the patent urachus and it appears promising as far as cosmetic outcome of the umbilicus is concerned (Schaefer *et al.,* 2012). Surgical excision of the urachal remnant is curative and there are no functional sequelae from its excision, as it is a vestigial remnant. The management dilemma of urachal anomalies occurs in patients who present with an asymptomatic lesion that is incidentally discovered on a workup of patients with urinary tract infections or other anomalies of urinary tract. It is better to excise the asyptomatic urachal remanents because there is always a risk of malignancy from the transional epithelium of the urachal remanents (Sheldon *et al.,* 1984; Rubin *et al.,* 1999).

To summarise, urachal abnormalities are rare. The diagnosis is mainly based on clinical presentation and the imaging study required is ultrasonography. Other imaging modalities like CT scan and fistulography should be used only in selected cases as none of the patient in this series required these investigations. Voiding cystourethrography does not seem necessary because in these anomalies the incidence of lower urinary tract obstruction is extremely low. The treatment of choice is excision of the patent urachus or the urachal cyst 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 an urachal sinus. The asymptomatic urachal anomalies detected incidentally should also be excised for the fear of malignancy from the lining epithelium.

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