A Comprehensive Review of Pediatric Urachal Anomalies and Predictive Analysis for Adult Urachal Adenocarcinoma

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Abbreviations and Acronyms

CER = control event rate

CT = computerized tomography

EER = experimental event rate

MRI = magnetic resonance imaging

NNT = number needed to treat

VCUG = voiding cystourethrography

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Study received institutional research and ethics board approval.

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Purpose: We examined the presentation, diagnosis and management of radiologically detected pediatric urachal anomalies and assessed the risk of malignant degeneration.

Materials and Methods: Our radiology database (2000 to 2012) was queried for all children younger than 18 years who were diagnosed with a urachal anomaly radiographically, and the operative database was used to determine those who underwent excision. Data collected included demographics, presenting symptoms, imaging modality and indication for excision. These data were compared to the Ontario Cancer Registry to determine the risk of malignancy.

Results: A total of 721 patients were radiographically diagnosed with a urachal anomaly (667 incidentally), yielding a prevalence of 1.03% of the general pediatric population. Diagnoses were urachal remnants (89% of cases), urachal cysts (9%) and patent urachus (1.5%). Ultrasonography was the most common imaging modality (92% of cases), followed by fluoroscopy/voiding cystourethrography (5%) and computerized tomography/magnetic resonance imaging (3%). A total of 61 patients (8.3%) underwent surgical excision. Indications for imaging and treatment were umbilical drainage (43% of patients), abdominal pain (28%), palpable mass (25%) and urinary tract infection (7%). Mean age at excision was 5.6 years and 64% of the patients were male. Based on provincial data, the number needed to be excised to prevent a single case of urachal adenocarcinoma was 5,721.

Conclusions: Urachal anomalies are more common than previously reported. Children with asymptomatic lesions do not appear to benefit from prophylactic excision, as the risk of malignancy later in life is remote and a large number of urachal anomalies would need to be removed to prevent a single case of urachal adenocarcinoma.

Key Words: pediatrics, urachal adenocarcinoma, urachus

Symptomatic urachal anomalies in children have traditionally been removed to alleviate the symptoms. However, the recent literature suggests excision of even incidentally discovered urachal anomalies to prevent future problems.[1] The most serious issue encountered is development of urachal adenocarcinoma, which, although extremely rare, carries significant morbidity and mortality. The literature is inconclusive on how to manage pediatric urachal lesions, especially those discovered incidentally. Removal of asymptomatic urachal remnants is often recommended systematically after diagnosis to prevent future issues or if present on repeat imaging

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after age 6 months.[2] Others advocate removing only lesions that present with symptoms. Still others advocate nonoperative management as a reasonable approach in asymptomatic and a subset of symptomatic lesions, including those presenting as infected cysts.[3] Divergent views on management clearly highlight a lack of consensus and an ill defined therapeutic or prophylactic value of surgical resection.

The true incidence of urachal anomalies in children is unknown, as is the risk of future malignant degeneration. Given the rarity of both conditions, we postulated that the preventive value of systematic resection of asymptomatic lesions detected in childhood is minimal and the procedure is likely unwarranted. We examined the prevalence, presentation, diagnosis and management of radiologically identified pediatric urachal anomalies at a large tertiary pediatric center, correlating the results with previous publications dealing with urachal neoplasms in the same geographic region to assess the likelihood of a urachal anomaly degenerating into a malignancy later in life.[4]

MATERIALS AND METHODS

Our institution principally serves the province of Ontario, a geographic region roughly the size of western Europe, containing a population of approximately 10 million. After receiving approval from our institutional research and ethics board we retrospectively queried the electronic radiology database of our regional referral center for all children younger than 18 years examined between January 2000 and December 2012 undergoing abdominal evaluation via 4 distinct modalities, ie ultrasound, fluoroscopy/VCUG, CT and MRI. Imaging type was the denominator in the calculation of the prevalence of urachal anomalies. We then parsed radiology reports for "urachus" or "urachal." The charts of those patients were then individually reviewed to confirm the diagnosis, excluding those in whom "urachus" or "urachal" was not associated with an identified urachal lesion. Type of study used to diagnose the urachal anomaly, radiographic findings, and patient age and gender were noted. The resulting filtered list was also cross-referenced to any operative procedures performed in the same time range to identify patients who underwent surgical intervention for the identified urachal lesion. Patient demographics, presentation and histopathological data were collected, as well as indication for intervention.

Results were cross-referenced with previously published data from the Ontario Cancer Registry on the yearly incidence of urachal adenocarcinoma in the same geographic catchment area (0.18 of 100,000 individuals yearly)[4] to estimate NNT to prevent a single case of urachal adenocarcinoma, where "treat" is defined as surgical excision. This calculation was based on the assumptions that 1) all urachal adenocarcinomas develop in patients with urachal anomalies, 2) urachal excision in childhood eliminates the risk of subsequent urachal adenocarcinoma and 3) urachal anomalies do not spontaneously involute or lose malignant potential if not removed. The calculation sequence for estimating NNT to prevent a single case of adenocarcinoma is, absolute risk = annual incidence of urachal adenocarcinoma in the general population = 0.18/100,000; CER = annual incidence of urachal adenocarcinoma in those with urachal anomalies; EER = annual incidence of urachal adenocarcinoma in patients following surgical excision of the urachal anomaly (therefore = 0); RRR = CER/(CER - EER) = incidence reduction in event rate following intervention, ie excision of urachal lesion (therefore = 1); absolute risk reduction = CER - EER = CER, and NNT = 1/absolute risk reduction.

RESULTS

During the 13-year period 64,803 patients underwent at least 1 abdominal imaging study at our institution. Of those patients 721 were radiographically diagnosed with a urachal anomaly (667 incidentally). Mean age at diagnosis was 6.2 years (interquartile range 1.7–9.4). Radiological data are summarized in the table. Specific imaging diagnoses were persistent urachal remnants, urachal cysts, sinus tract/patent urachus and urachal diverticulum. Ultrasonography was the most commonly used imaging modality, followed by fluoroscopy/VCUG, CT and MRI. Figure 1 illustrates a urachal remnant on ultrasound and a large urachal cyst on CT. Figure 2 illustrates the large urachal cyst intraoperatively.

Of the study population only 60 patients (8.3%) underwent surgical excision. Six cases (10%) were excised laparoscopically and 54 (90%) in an open manner. Indications for imaging and treatment in the surgical group were umbilical drainage in 26 patients (43%), abdominal pain in 17 (28%), palpable mass in 15 (25%) and urinary tract infection in 4 (7%). However, 6 of the cases excised (10%) were incidentally diagnosed, and prophylactic excision was undertaken because of recent recommendations made in the literature, as noted in the medical charts. Mean age at excision was 5.6 years (range 3 days to 17.1 years) and 64% of patients were male. No complications were reported in those undergoing simple excision, and all symptomatic patients were cured of the presenting symptoms.

Summary of radiological data

No. imaging diagnosis (%):	
Urachal remnant	640 (89)
Urachal cyst	66 (9)
Sinus tract/patent urachus	11 (1.5)
Urachal diverticulum	4 (0.6)
No. imaging modality (%):	
Ultrasound	665 (92)
VCUG/fluoroscopy	38 (5)
CT	13 (2)
MRI	5 (0.7)