Clinical Urological Outcomes Following Primary Tethered Cord Release in Children Younger Than 3 Years

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Purpose: Current practice at our institution is to recommend tethered cord release at diagnosis to prevent the onset or worsening of symptoms. Tethered cord release is frequently performed in children younger than 3 years who often have no urological manifestations. To our knowledge there are currently no long-term data on urological outcomes in this age group.
Materials and Methods: We completed a retrospective review of 475 cases of tethered cord release performed at a single institution between 1995 and 2002. Of these surgeries 173 were performed in children younger than 3 years. Clinical outcomes, and preoperative and postoperative urodynamic and radiographic studies were evaluated.
Results: A total of 79 patients met study criteria. Average age at surgery was 9.6 months and average followup was 5.2 years (range 6 months to 11.2 years). At followup 49 patients (62.1%) had no urological complaints and 30 (38%) had urological problems. A total of 20 children (25.3%) had minor problems (constipation, delayed toilet training or other) and 10 (12.7%) had major problems (need for clean intermittent catheterization, febrile urinary tract infection or reflux). Of 66 patients 30 (45.5%) had abnormal preoperative urodynamics. One of 31 patients (3.2%) had hydronephrosis on preoperative ultrasound. Statistical analysis revealed that abnormal preoperative urodynamics and ultrasound were not predictive of major urological problems. Lipomatous dysraphism and preoperative musculoskeletal symptoms positively correlated with major urological problems ($p = 0.0076$ and 0.0484, respectively).
Conclusions: The majority of children did not experience urological problems following tethered cord release. Only a small set of children had major urological problems. Children with lipomatous dysraphism and musculoskeletal symptoms were more likely to experience poor urological outcomes.

Key Words: bladder, neural tube defects, urodynamics, urination disorders, spinal dysraphism

Tethered cord is abnormal fixation of the spinal cord in patients with spinal dysraphism. It is thought that this fixation leads to stretching of the spinal cord, and subsequent ischemia and hypoxia of the spinal cord and sacral nerve roots.[1] Tethered cord syndrome can be primary or secondary. Primary tethered cord results from sacral dysgenesis and occult spinal dysraphism. Secondary tethered cord primarily occurs following surgical repair of myelomeningocele or other dysraphic entities.[2] In either case the hypoxemic damage can result in upper motor neuron deficits, which may include motor or sensory symptoms in the lower limbs and/or bladder dysfunction.

Currently the timing of TCR is controversial. This is especially true for primary tethered cord when patients have primarily cutaneous manifestations, such as a sacral dimple and minimal to no symptoms. Some groups advocate early untethering, reporting that it can slow progression or possibly prevent the onset of neurological deficits and bladder dysfunction.[3–7] Others advocate close monitoring with surgery only after upper motor neuron symptoms occur.[8]

Current practice at our institution is to recommend TCR for patients with primary tethered cord at diagnosis in an effort to prevent the onset or worsening of symptoms. These procedures are frequently performed in children younger than 3 years. At this young age urinary continence has not yet been attained and, therefore, clinical urological manifestations of tethered cord are rarely apparent. To our knowledge there are currently no long-term data on the urological outcomes of TCR in patients with primary tethered cord in this age group.

MATERIALS AND METHODS

Patient Selection

We completed a retrospective review of 475 cases of TCR performed at our institution between 1995 and 2002. Of these patients 173 were younger than 3 years. Only patients with primary tethered cord were included in the study. Patient diagnoses included L, DSM, DST, FF and MMM. Children with myelomeningocele, imperforate anus, cloacal exstrophy, previous TCR or less than 6 months of followup were excluded from study.

Urodynamic Studies

Preoperative and postoperative UDS were evaluated and classified as normal or abnormal. Studies were classified as abnormal if they demonstrated 1 of decreased compliance in the filling phase and/or hypotonia, increased post-void residual urine, DSD or increased voiding pressures in the voiding

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phase. Since uninhibited detrusor contractions are difficult to interpret and they are a frequent finding in patients with a normal bladder in this age group, UDS with isolated uninhibited detrusor contractions were not considered abnormal for study purposes.

Radiographic Studies

Preoperative and postoperative results of renal US and VCUG were reviewed. US was classified as abnormal if any degree of hydronephrosis was present. Anatomical abnormalities, such as horseshoe kidney or a duplicated system, were not considered abnormal in the absence of hydronephrosis. VCUG was defined as abnormal if any degree of VUR was present. Comments on bladder changes were also noted.

Clinical Outcomes

Clinic notes were reviewed to evaluate patient clinical status. Additionally, parents of patients were contacted and information was obtained regarding history of UTI, age at toilet training, constipation, need for CIC and other urinary complaints. Minor poor urological outcomes following TCR included a history of afebrile UTI, constipation and delayed toilet training, defined as the absence of daytime urinary continence by age 4 years. Major poor urological outcomes included the requirement of long-term CIC, history of FUTI and documented new onset or progression of VUR or hydronephrosis. Followup was calculated from the day of surgery to the last date of contact with the patient.

Statistical Analysis

Chi-square analysis was performed on variables, including sex, preoperative diagnosis, preoperative motor symptoms, and preoperative and postoperative urodynamic and radiographic studies, to determine factors predictive of poor urological outcomes.

RESULTS

Preoperative Parameters

A total of 79 patients met study criteria. The average age at which TCR was performed was 9.6 months. Mean followup was 5.2 years (median 4.8, range 6 months to 11.2 years). Table 1 lists preoperative patient characteristics. The most common diagnoses in the study group were FF in 26 of 79 patients (32.9%) and lipomatous dysraphism in 26 (32.9%). A total of 69 patients (87.3%) initially presented with a cutaneous sacral abnormality, including a dimple, mass or hemangioma. Of the patients 42 (53.2%) and 37 (46.8%) were female and male, respectively. A total of 14 patients (17.7%) presented with musculoskeletal symptoms at diagnosis, including lower extremity weakness, club foot or delayed motor milestones. Only 2 patients (2.5%) had urological symptoms at diagnosis. One patient presented in urinary retention following cardiac surgery and was found to have a tethered cord on MRI. The other patient had a sacral dimple with 2 FUTIs before TCR.

Preoperatively 36 of 66 patients (45.5%) had normal UDS and 30 (54.5%) had abnormal UDS. Table 2 lists preoperative urodynamic abnormalities.

A total of 31 patients were evaluated with preoperative US, of whom 30 (96.8%) were found to be normal, whereas 1 (3.2%) demonstrated grade I hydronephrosis. Of the patients 12 underwent preoperative VCUG, of which 1 (8.3%) was positive for bilateral grade II VUR and 1 (8.3%) revealed mild bladder trabeculation.

TABLE 1. *Preoperative patient characteristics*

	No. Pts (%)
Diagnosis:	
L	26 (32.9)
DST	18 (22.8)
DSM	5 (6.33)
FF	26 (32.9)
MMM	4 (5.1)
Sex:	
M	37 (46.8)
F	42 (53.2)
Presentation:	
Cutaneous	69 (87.3)
Musculoskeletal	14 (17.7)
Urological	2 (2.5)
Preop UDS:	
Normal	36 (54.5)
Abnormal	30 (45.5)
Preop US:	
Normal	30 (96.8)
Abnormal	1 (3.2)
Preop VCUG:	
No reflux	11 (91.7)
Reflux	1 (8.3)

Postoperative Outcomes

Table 3 lists postoperative urological outcomes. A total of 49 patients (62.1%) had no urological complaints following surgery and 30 (38%) complained of minor or major urological problems. The 20 children (25.3%) with minor urological complaints experienced constipation, delayed toilet training, afebrile UTIs and/or dysfunctional voiding. Ten patients (12.7%) had major urological problems, including the need for CIC in 6, FUTIs in 8, new onset hydronephrosis in 5, and/or new onset or progression of VUR in 5. Interestingly the 2 children who presented with urological complaints before TCR showed improvement and neither experienced major urological problems.

Risk Factors for Poor Urological Outcomes

Table 4 lists clinical characteristics, and urodynamic and imaging results for each of the 10 patients with major urological problems. Table 5 shows the statistical correlation of clinical characteristics and preoperative studies with major urological problems.

Eight children (80%) with major urological problems were female, while only 2 (20%) were male. This compares with 35 male patients (50.7%) and 34 female patients (49.3%) without major urological problems. This difference approached but did not attain statistical significance ($p = 0.0688$).

Seven of the 10 children (70%) with major urological problems were diagnosed with lipomatous dysraphism. The other 3 children with major urological problems were diagnosed with DST (2) and FF (1). Therefore, 7 of 26 children (26.9%) with lipomatous dysraphism had a major urological problem in comparison to only 3 of 53 (5.6%) with any other primary tethered cord diagnosis ($p = 0.0076$). Children with FF in our cohort were less likely to have a poor urological outcome, although this did not achieve statistical significance ($p = 0.099$).