Total and Partial Urogenital Mobilization: Focus on Urinary Continence

Blake W. Palmer,* Brandon Trojan, Katie Griffin, William Reiner, Amy Wisniewski, Dominic Frimberger and Bradley P. Kropp

From the Department of Pediatric Urology, University of Oklahoma, Oklahoma City, Oklahoma

Abbreviations and Acronyms
CAH = congenital adrenal hyperplasia
DSD = disorder of sex development
PUM = partial urogenital mobilization
TUM = total urogenital mobilization
UGS = urogenital sinus

Submitted for publication July 28, 2011. Study received institutional review board approval.
* Correspondence: Department of Pediatric Urology, University of Oklahoma, 920 Stanton L. Young Blvd., WP 3150, Oklahoma City, Oklahoma 73104 (telephone: 405-271-6900; FAX: 405-271-3118; e-mail: blake-palmer@ouhsc.edu, brad-kropp@ouhsc.edu).

Purpose: Total and partial urogenital mobilization procedures are the most common contemporary vaginoplasty surgeries for patients with congenital adrenal hyperplasia, urogenital sinus and cloacal anomalies. There is controversy regarding the urinary continence outcomes of these procedures. We reviewed the urinary continence outcomes of children who underwent total or partial urogenital mobilization at our institution and reviewed the literature to determine the continence rates of these procedures.

Materials and Methods: We retrospectively reviewed 25 patients who underwent total or partial urogenital mobilization with a focus on postoperative continence status. Continence was defined as parental report of full toilet training with no accidents during the day and rare accidents (fewer than 2 per month) at night after age 3 years.

Results: A total of 14 congenital adrenal hyperplasia, 5 urogenital sinus and 6 cloacal anomaly cases were managed by total (18) or partial (7) urogenital mobilization procedures with a mean followup of 4.41 years (range 0.21 to 12.1). In our cohort 21 of 22 patients (95.5%) were continent by age 3 years and there were no urinary complications. A total of 111 patients were identified in the literature with congenital adrenal hyperplasia or urogenital sinus, with 107 in 7 studies being continent (96.4%) by age 3 to 4 years. In 4 studies 32 patients were identified with cloacal anomalies who underwent total or partial urogenital mobilization, of whom 28 (87.5%) were continent by age 3 to 4 years.

Conclusions: There was no significant difference between total and partial urogenital mobilization procedures regarding postoperative urinary continence in our cohort and the literature. The urinary continence rate was 96% in the congenital adrenal hyperplasia/urogenital sinus group and 89.5% in the cloacal group.

Key Words: adrenal hyperplasia, congenital; cloaca; urogenital abnormalities; urogenital surgical procedures

The 2002 joint Lawson Wilkins Pediatric Endocrine Society and European Society for Pediatric Endocrinology consensus statement for congenital adrenal hyperplasia due to 21-hydroxylase deficiency proposed that the goals of feminizing genitoplasty were to create a female typical external genital appearance, create unobstructed urinary emptying free of incontinence and urinary tract infections, and allow for adult sexual and reproductive function. A more recent set of guidelines by the CARES (Congenital Adrenal Hyperplasia Research, Education and Support) Foundation concerning sur-
gical treatment of congenital adrenal hyperplasia refers to the likelihood of incontinence after vaginoplasty as a factor to consider in decision making concerning vaginoplasty procedures. Contemporary vaginoplasties for patients with congenital adrenal hyperplasia, urogenital sinus or cloacal malformations are often performed using a total or partial urogenital mobilization technique. However, outcomes studies of these procedures are lacking. We assessed urinary continence in patients who underwent total or partial urogenital mobilization vaginoplasties.

The TUM procedure was developed for cloacal repairs and simplified the reconstructive procedure for creating a vagina by eliminating the challenging step of dividing the urethra from the vagina. TUM was then adopted for use in patients with a persistent urogenital sinus. However, the extent of the anterior dissection past the pubic bone to the level of the bladder neck has been a cause of concern regarding the possible development of incontinence in patients who undergo this procedure. This concern resulted in modification of the procedure (PUM), which limits dissection to the pubic bone.

To determine how successful TUM and PUM are at achieving the cosmetic, sexual function and reproductive goals outlined in the 2002 joint consensus statement, long-term followup is required. However, continence status can be evaluated much sooner in younger patients. Limited data exist to support or refute concerns regarding incontinence in patients who have undergone TUM or PUM. Thus, there is a need to establish continence outcomes for girls and women who have undergone these types of vaginoplasties.

**METHODS**

After institutional review board approval we retrospectively reviewed consecutive vaginoplasty procedures performed at our institution and identified 25 patients with a diagnosis of CAH, UGS or cloacal anomaly who underwent primary total or partial urogenital mobilization and were old enough for continence to be assessed. Preoperative evaluation, operative technique and followup data were collected. Information obtained included age at surgery and continence status preoperatively and at age 3 years. Age 3 years was selected because this cutoff is commonly used for assessing continence in the literature after performing TUM and PUM procedures, and it is also an age by which toilet training should be achieved. Continence was defined as parental report of patients being fully toilet trained with no accidents during the day and rarely (fewer than 2 per month) at night after age 3 years.

For the procedures patients are admitted to the hospital on the day prior for bowel preparation to prevent stool contamination of the field during urogenital mobilization and to allow ease of dissection. The TUM and PUM procedures are approached by first performing a full lower body povidone-iodine preparation. Cystoscopy and placement of balloon catheters in the bladder and vagina are done at the beginning of each procedure. The length of the common sinus and location of the external urethral sphincter in relation to the confluence of the vagina and urethra are noted endoscopically, as well as the location of the bladder neck. Patients with a cloaca also have a catheter placed in the colon when possible. Patient positioning, prone or supine, on the table depends on several factors. Cloacal anomaly, length of the UGS and limited space in the perineum are some of the reasons we would position the patient prone with knees and hips flexed on a sterile towel roll to increase exposure under the pubic bone. The full lower body preparation allows us to reposition during the case as needed for best exposure.

The anal sphincter complex is identified with a muscle stimulator and marked for the target of the rectal pull-through portion after the rectum is separated from the urogenital complex. After the rectum is separated patients with a cloacal anomaly are sterilely repositioned supine for the en bloc mobilization of the common urogenital sinus. The importance of staying close to the common sinus/urethra-vaginal complex is emphasized to minimize dissection of the pelvic floor. The clitoris is degloved and clitoroplasty is performed when requested by the patient and/or parent after appropriate preoperative counseling. An omega incision is routinely made in the perineum posteriorly, which allows for better exposure than a simple midline incision. We find it is unnecessary to use this as a posterior based flap, as the mucosal lined common sinus is almost always able to cover this distance even in patients with a high confluence or cloaca.

If an adequate amount of mobilization is achieved without dissecting under the pubic bone and up to the bladder neck for a tension-free anastomosis to the perineum, the dissection is complete and the procedure is stopped as a PUM. However, routinely TUM is required to position the urethra in an orthotopic position on the perineum. In these cases the dissection continues under the pubic bone through the pubourethral ligaments and to the level of the bladder neck. The catheter balloons can be palpated to identify the anatomical position of the bladder neck. The common sinus is divided in a manner that will allow for maximal use of the mucosal lined common sinus tissue flap to complete the vagina and urethral positioning on the perineum in an orthotopic position. This step can be accomplished by splitting the common sinus anteriorly and posteriorly or laterally. Perineoplasty is performed when necessary, and local skin flaps are used to reconstruct the external genitalia and place (or create) the labia minora more posteriorly lateral to the neovaginal introitus. Y-V plasty can also be used to place the labia majora more posteriorly.

A Foley catheter is left in the urethra and antibiotic ointment gauze is placed in the vagina overnight. Routinely the legs are secured to prevent incidental wide separation during the first week postoperatively. Pain control is achieved with nonnarcotic anti-inflammatory medications with good success, and patients are discharged home on postoperative day 1 or 2. Patients with CAH are followed when admitted by the pediatric endocrinology service per a collaborative protocol.
We reviewed all PubMed searchable articles using the search terms “urogenital mobilization,” “vaginoplasty” and “feminizing genitoplasty.” Articles were reviewed for description of diagnosis and indication for vaginoplasty, procedure used, age at surgery, description of continence achieved and at what age, as well as a description of urinary complications. Articles were excluded that did not specify continence as an outcome measure, that described a vaginoplasty procedure other than TUM/PUM or that had an indication for surgery other than CAH, urogenital sinus, cloacal malformation, 46,XY DSD or ovotesticular DSD. The numbers of patients from each article included in the review were grouped by diagnosis and procedure, and continence outcome and complications were summarized.

RESULTS

Retrospective review demonstrated that 18 patients underwent TUM and 7 underwent PUM. Of the patients who underwent PUM 4 had a diagnosis of CAH, 2 UGS and 1 cloacal anomaly. Of the patients who underwent TUM 10 had a diagnosis of CAH, 3 UGS and 5 cloacal anomaly. Mean followup was 4.41 years (range 0.21 to 12.1). Median age at surgery was 1.25 years (range 0.48 to 17.6), with 21 operations being performed in prepubertal and 4 in postpubertal patients. Preoperative endoscopy revealed the common channel length was distal to the external urethral sphincter, and less than 3 cm in 13 patients (52%) and greater than 3 cm in 12 (48%). Average urogenital sinus length was 2.62 cm (range 0.5 to 4) in the 18 patients undergoing TUM and 2.21 cm (1 to 4) in the 7 patients undergoing PUM. Average urethral length was 1.67 cm (range 0.5 to 4) in patients undergoing TUM and 1.5 cm (0.5 to 2.5) in those undergoing PUM.

Postoperatively no patient had a urethral or bladder complication. Urinary continence was evaluated in 22 patients who were older than 3 years. Three of the patients were younger than 3 years and thus incapable of being evaluated for successful toilet training. Of those children eligible for evaluation 21 (95.5%) were continent day and night. Five patients (22.7%) who underwent TUM had a history of nocturnal enuresis lasting more than a year after achieving daytime dryness with toilet training (3 with CAH, 2 with UGS).

One patient with UGS who underwent TUM had persistent episodes of frequency and urge urinary incontinence daily that was infrequent after toilet training and surgery at age 2 but had worsened in the last 1 to 2 years after age 12. She experienced nocturnal enuresis at age 12 after being dry at night for approximately 10 years following vaginoplasty. She was absent from clinical followup for many years but was seen again with these new symptoms and a history significant for constipation and dysfunctional elimination. Urological examination was normal, and she is currently being treated with behavioral modification (timed voiding) and deconstipation.

We reviewed articles from 1996 to 2011 and identified 7 describing total or partial urogenital mobilization procedures in patients with CAH, UGS, cloacal malformations, 46,XY DSD or ovotesticular DSD, including information about postoperative continence (tables 1 and 2).10 A total of 116 patients were evaluated, and 107 of 111 (96.4%) achieved continence by age 3 to 4 years. Patient age at vaginoplasty ranged from 3.8 to 41.4 months, and followup was 2.1 to 5 years.

Four peer-reviewed articles from 1996 to 2011 were identified that determined success with TUM or PUM in the cloacal population and described outcomes of 41 patients.8,10 Of the 32 patients older than 3 to 4 years 28 (87.5%) achieved continence. In these articles patient age at TUM/PUM was 3.8 to 14 months and followup was 2.5 to 8.5 years.

Chi-square analyses showed no difference in continence outcome between patients who underwent

<table>
<thead>
<tr>
<th>References</th>
<th>Diagnosis</th>
<th>No. Surgery</th>
<th>Median Mos Age at Surgery (range)</th>
<th>Median Yrs Followup (range)</th>
<th>No. Continent/Total No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Current study</td>
<td>CAH/UGS</td>
<td>13 TUM, 6 PUM</td>
<td>15 (10–21) for TUM, 12 (9–15) for PUM</td>
<td>5.8 (0.2–11) for TUM, 4.7 (0.3–12) for PUM</td>
<td>11/12 (91.7) for TUM, 4/4 (100) for PUM</td>
</tr>
<tr>
<td>Cammarini et al13</td>
<td>CAH</td>
<td>6 TUM</td>
<td>10 (8–34)</td>
<td>5 (3–6)</td>
<td>5/5 (100)</td>
</tr>
<tr>
<td>Leslie et al14</td>
<td>CAH</td>
<td>18 TUM, 26 PUM</td>
<td>13 (3–174) for TUM, 12 (4–149) for PUM</td>
<td>Not reported</td>
<td>19/18 (100) for TUM, 26/26 (100) for PUM</td>
</tr>
<tr>
<td>Savanelli et al15</td>
<td>CAH</td>
<td>14 PUM</td>
<td>16 (6–48)</td>
<td>Not reported</td>
<td>14/14 (100)</td>
</tr>
<tr>
<td>Braga et al16</td>
<td>CAH</td>
<td>24 PUM</td>
<td>41.4 (1–192)</td>
<td>2.1 (not reported)</td>
<td>18/20 (90)</td>
</tr>
<tr>
<td>Gosabal et al17</td>
<td>CAH/UGS</td>
<td>9 TUM</td>
<td>3.8 (3–156)</td>
<td>2.5 (0.25–5)</td>
<td>9/9 (100)</td>
</tr>
<tr>
<td>Kryger and Gonzalez18</td>
<td>CAH/mixed gonadal dysgenesis/pure gonadal dysgenesis/true hermaphrodite</td>
<td>13 TUM</td>
<td>Not reported (5–216)</td>
<td>3.4 (0.4–4.8)</td>
<td>11/13 (84.6)</td>
</tr>
<tr>
<td>Jenak et al17</td>
<td>CAH/UGS/tru hermaphrodite</td>
<td>6 TUM</td>
<td>Not reported (4.5–234)</td>
<td>Not reported (0.8–7.5)</td>
<td>6/6 (100)</td>
</tr>
</tbody>
</table>

* Patients with CAH/UGS/cloacal anomalies were not separated and, therefore, reported median age at surgery and median followup are for total population.
TUM vs PUM (1.17, p > 0.05) in our study or in the collective literature evaluation of these procedures. There was also no difference when only patients with CAH/UGS were compared. In our cohort of 25 patients none had a urethral or bladder complication intraoperatively or at any point during followup. Overall urethral/bladder complication rate associated with TUM/PUM for the 9 articles reviewed was 2.83% (3 of 106 patients).

DISCUSSION

Urogenital mobilization represents a significant advancement in the surgical management of cloacas and common UGS, and its use has become widely accepted. The original TUM allowed for safer surgical dissection, avoiding the difficult separation of the urethra from the vagina. Data from our cohort coupled with the low complication rates reported by others confirm that TUM/PUM procedures are effective in allowing patients to achieve urinary continence. Thus, the goal of allowing for unobstructed urinary flow outlined in the 2002 joint consensus statement is possible with PUM and TUM. A previous report also demonstrated that patients with CAH who underwent feminizing genitoplasty, regardless of type of procedure or age at surgery, did not have an increased risk of urinary tract infection.

Since the introduction of TUM, there have been concerns regarding the impact of this procedure on urinary continence, given the aggressive surgical dissection around the urethra and up to the level of the bladder neck. Review of our patients as well as the overall literature suggests that the ability to achieve continence is preserved when a patient with CAH, UGS or cloacal anomaly undergoes total or partial urogenital mobilization. TUM does not confer an increased risk of incontinence compared to the limited dissection of PUM. The literature is sparse regarding reports of urinary continence following vaginoplasty procedures other than TUM/PUM. However, a survey conducted in adults with CAH who underwent genitoplasty in childhood before the current surgical practices revealed a 68% incidence of urge incontinence and 47% incidence of stress incontinence. This outcome shows a dramatic improvement with the TUM/PUM procedures in our cohort, with more than 90% of those treated at our institution achieving continence and more than 85% of those treated elsewhere achieving continence. Urinary function studies should be extended to women who underwent TUM/PUM as children to assess long-term continence following these procedures.

The remaining goals of the 2002 joint consensus statement regarding the ability to create a feminine external genital appearance and to allow for satisfactory adult sexual and reproductive function are important aspects of TUM/PUM that remain to be investigated. Cosmesis following TUM/PUM is reported as good or acceptable by some surgeons. However, the ultimate assessment should come from patients themselves. The same can be said for sexual functioning following these procedures. Long-term followup and standardized evaluations in adulthood are needed to evaluate the ultimate success of these procedures.

Limitations of this study include the heterogeneous nature of the groups (CAH/UGS vs cloacal anomaly) in terms of pelvic anatomy. Patients with CAH/UGS typically have established perineal musculature and normal neurospinal anatomy, whereas those with cloacal anomalies have an anteriorly rotated pelvic floor and associated lumbosacral spinal malformation. Despite these differences, our data and the literature in general demonstrate success from a urinary standpoint for TUM and PUM procedures. Another limitation is that continence was subjectively evaluated by parental interview and questioning in the clinic. Of the other reports in the literature only 1 included a bladder diary and 2 included urodynamics, while none used an objective questionnaire or other method to assess continence. Finally, the method to determine continence was not stated in more than half of the reports.

CONCLUSIONS

In our experience PUM is sufficient for patients who do not require urethral mobilization, but TUM is required if urethral mobilization is necessary due to a long/high common channel or short urethra. Commonly the degree of difficulty and prognosis are determined by the level of the confluence. However,
most important is the length of the urethra itself. One report describes the importance of a urethral length of at least 1.5 to 2 cm.\textsuperscript{10} In our study the mean urethral lengths were 1.67 cm in the TUM group and 1.5 cm in the PUM group, which did not differ significantly or have an impact on continence. There are few if any data to support claims of 1.5 to 2 cm urethral length necessary for urinary continence, and in the reports cited here only 2 describe the urethral length in their cohorts.\textsuperscript{10,18} These facts illuminate how few data are available to support previous claims of worse continence outcomes and also our inability to predict who is at risk for incontinence after vaginoplasty by TUM/PUM. Standardized reporting and long-term evaluation with better objective measures will ultimately determine the success of the TUM/PUM technique, although to date it is superior to historical procedures.\textsuperscript{12}

REFERENCES


