Total and Partial Urogenital Mobilization: Focus on Urinary Continence

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Abbreviations and Acronyms

CAH = congenital adrenal hyperplasia

DSD = disorder of sex development

PUM = partial urogenital mobilization

TUM = total urogenital mobilization

UGS = urogenital sinus

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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.[1] A more recent set of guidelines by the CARES (Congenital Adrenal Hyperplasia Research, Education and Support) Foundation concerning sur-

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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.[2] 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.[3] TUM was then adopted for use in patients with a persistent urogenital sinus.[4] 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.[4–8] This concern resulted in modification of the procedure (PUM), which limits dissection to the pubic bone.[9]

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.[3,9,10] 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.