
Use of the Mobilized Sinus With Total Urogenital Mobilization

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Purpose: The surgical treatment of urogenital sinus anomalies has undergone significant advances in recent years. Total urogenital mobilization, which mobilizes the urogenital sinus, vagina and urethra en bloc toward the perineum, represents one of these advances.

Materials and Methods: We have improved our results with total urogenital mobilization by incorporating the mobilized urogenital sinus tissue into the repair rather than discarding it, as described originally. We have found this a readily available, easily manipulated and well vascularized flap that is a significant aid to reconstruction.

Results: We present our 3 favored means of using the mobilized sinus tissue to create a mucosa lined vestibule, a posterior vaginal wall flap and an anterior vaginal wall flap.

Conclusions: We believe that our techniques result in a further advancement in the cosmetic and surgical outcomes in these patients, and are beneficial in the reconstructive surgery armamentarium.

Key Words: urogenital abnormalities; congenital adrenal hyperplasia; review; diagnostic techniques, surgical

Urogenital sinus abnormalities are one of the most challenging problems that the pediatric urological surgeon faces. The anatomy is complex and varied, and adequate surgical exposure may be difficult. In these patients there remains a persistent communication between the urethra and vagina, which results in a common urogenital perineal channel, the UG sinus.

Most patients are identified at birth with genital ambiguity, most commonly secondary to CAH. However, it may also occur as a pure UG sinus anomaly with normal appearing external genitalia.¹ In patients with cloacal anomalies the rectum also enters this common channel, thereby adding another degree of complexity. Pure UG sinus and cloaca cases frequently involve other severe organ system anomalies.^{2,3} In addition, some investigators believe that female exstrophy is a form of UG sinus anomaly, and a similar surgical approach has been applied in its reconstruction.^{1,4,5}

Surgery in young females with genital ambiguity is controversial at this time. Some believe that there should be a moratorium on reconstruction until further data are available or until the child is old enough to participate in the decision making.⁶ However, many believe that surgery is appropriate in these children.^{7,8} All families should be counseled regarding the controversies and treatment options, including watchful waiting. For those who elect surgery there are further questions regarding the timing of surgery and the specific procedure to be used.

Fundamentally, the operation is completely dependent on patient anatomy. Traditionally, these patients are classified as having either a low or a high confluence (fig. 1). Those with a "low confluence" have a short UG sinus, and the

junction of urethra and vagina is relatively close to the perineum. Conversely, those with a "high confluence" have the vagina entering near the bladder neck, and the common channel in this group is usually longer.

We believe that this confluence occurs over a spectrum, rather than simply as "high" or "low." From a surgical standpoint we believe that the most important factor is the distance from the bladder neck to the confluence, rather than the length of the common sinus.⁹ A short urethra with the vagina entering at or near the bladder neck is a much more challenging operation, regardless of the length of the common sinus, not only because of the degree of mobilization required, but also because adequate exposure is difficult and there is a much greater risk of bladder neck or sphincter injury. However, surgically, a long sinus may at times be advantageous, since more sinus tissue is available to incorporate into the repair.

Patients with a low confluence can usually be treated with "flap vaginoplasty,"^{1,10} as originally described by Fortunoff.¹¹ In this procedure a perineal flap of skin is anastomosed to the ventrally opened sinus. Unfortunately, for high confluence this approach would result in several unattractive features, including a severely hypospadiac urethra with urine pooling in the vagina, and potential disruption of the continence mechanism. Furthermore, with flap vaginoplasty the posterior aspect of the vagina is entirely made of skin, which may be hair bearing or prone to vaginal stenosis.

The landmark "pull-through" vaginoplasty described by Hendren and Crawford in 1969 overcame many of these deficiencies but was not without some technical problems.¹² The original description of separating the vagina from the urethra was difficult and tedious, and often left the vagina as an isolated perineal opening separate from the labia and urethra.

In 1997 our group described a posterior prone approach, which simplified the dissection.¹ That same year

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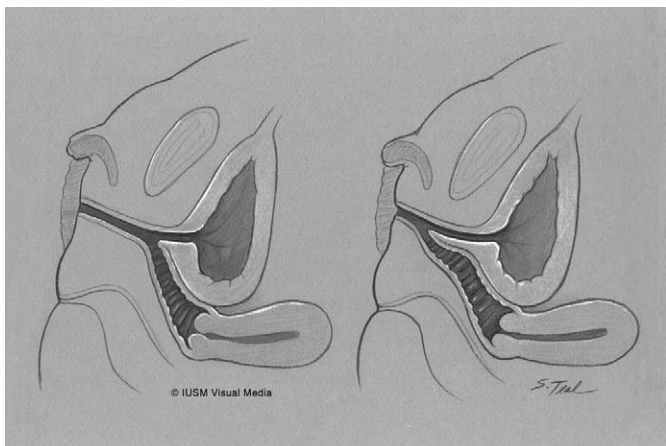


FIG. 1. Left, example of “high confluence.” Vagina inserts close to bladder neck, leaving little true urethra. Right, example of “low confluence.” Proximity to perineum and more normal urethra make this surgery much simpler.

Pena described TUM, which was another significant advancement.¹³ He described a procedure whereby the entire sinus, as a single unit, was circumferentially dissected from the perineum. By treating the UG sinus as a single unit the technically demanding and time consuming separation of urethra from vagina was avoided, and Pena was able to show that the time of dissection and blood loss were decreased.¹³ In this procedure after creation of a perineal based flap the dissection is begun along the posterior UG sinus (fig. 2). Attention is then turned to separating the anterior sinus from the corpora and pubis (fig. 3, A). This separation results in mobilization of the intact urogenital sinus, which, when completely free, usually reaches the perineum. Pena then described sewing the normal caliber vagina to the posterior flap.¹³ Therefore, the urethra, since it had been mobilized en bloc with the vagina, was left in an orthotopic position (fig. 3, B).^{13,14}

Although this procedure involved discarding the mobilized sinus tissue, we strongly believe that this tissue is valuable in the reconstruction, and can be used to improve cosmesis and avoid the problems associated with skin flaps.^{1,14–16} This hypothesis is the basis of this report.

We have used the mobilized sinus in 3 distinct ways, each for a unique application. In flap vaginoplasty the mobilized sinus is split ventrally, and is used to create a mucosa lined vestibule. If the vagina requires separation from the sinus (pull-through vaginoplasty), it can be split dorsally and used to create an anterior vaginal wall, similar to that described by Passerini-Glazel.¹⁷ Finally, we report a novel method of a laterally split spiral flap, which is used to create a posterior vaginal wall and eliminates the need for the Fortunoff flap.¹⁵ Gosalbez has presented an additional technique using the sinus to avoid the Fortunoff flap.¹⁶

We describe the indications and surgical approach for each of our applications in the use of mobilized sinus tissue for vaginoplasty. Most UG sinus cases are secondary to CAH, with masculinized external genitalia. Clitoroplasty and labioplasty may be combined with the vaginoplasty as a single surgical procedure.

MATERIALS AND METHODS

Surgical Technique for TUM

For all 3 techniques the initial surgical approach is similar. Cystoscopy is carried out before all major genital reconstruction, and a Fogarty catheter is placed in the vagina and a Foley catheter in the bladder. The patient undergoes full body preparation, from the feet to the thorax, thereby allowing access to the abdomen and perineum, and rotation to the prone position when necessary. The omega-shaped perineal flap is developed extending from the proposed perineal body to near the meatus of the sinus (fig. 2).¹⁸ Clitoroplasty, if indicated, is performed at this time. Following its completion the dissection is started in the midline posteriorly, by dividing the attachments between the UG sinus and the rectum. With the rectum retracted from the sinus the entire posterior aspect of the sinus and vagina may be exposed.

The sinus is then carefully separated from the phallus and dissected to the level of the pubis. The ligaments attaching the sinus to the pubis are divided and the entire sinus is freed (fig. 3). At this time the Fogarty catheter is palpated and the vaginal confluence is identified. The vagina is opened posteriorly over the Fogarty catheter (fig. 4). It should be emphasized that this dissection must proceed until normal vaginal caliber is encountered, as the distal third of the vagina is often narrow and may be prone to stenosis. There is usually a pronounced demarcation where the narrow UG sinus ends and the wider caliber vagina opens up. All tissue distal to this ridge will be considered UG sinus tissue and the wider proximal tissue is defined as vagina.

One of the more challenging aspects of this surgery is now encountered, as the surgeon must decide how the final reconstruction will occur. In most children the va-

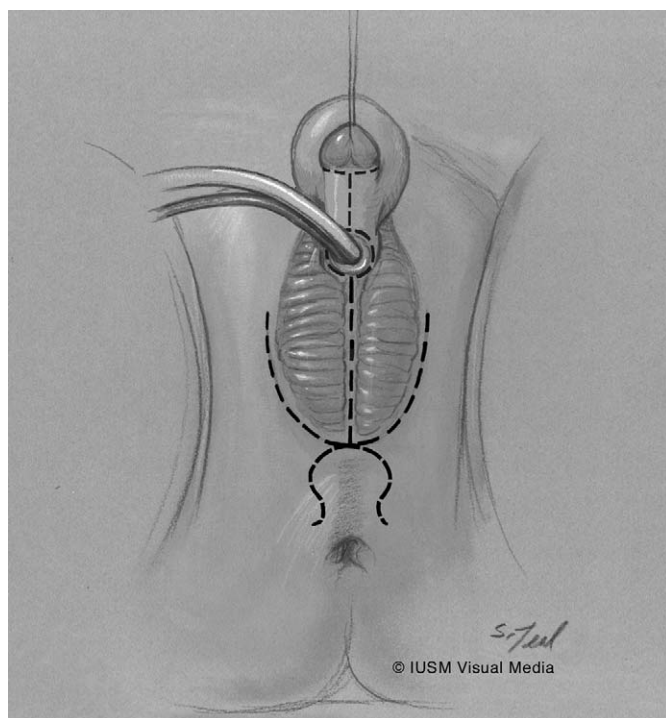


FIG. 2. Planned incision. Modified Fortunoff flap is omega-shaped, as described by Freitas Filho et al.¹⁸

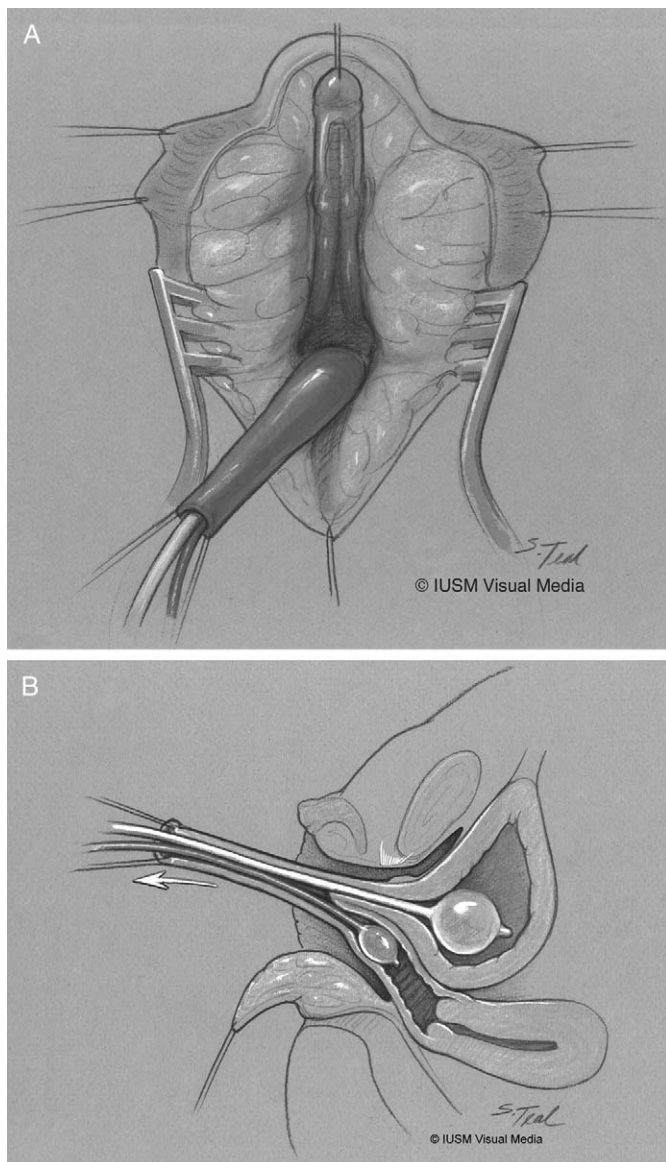


FIG. 3. A, anterior dissection frees sinus from corpora, and continues beneath pubis. B, after dissection has been completed sinus is mobile, and most will easily reach perineum.

gina will easily reach the perineum. It then may be sewn flush to the perineum but we prefer a posterior flap, which will widen the introitus. In this situation the mobilized sinus tissue may be used in 1 of 2 ways. It can be split ventrally to create a mucosa lined vestibule with a Fortunoff flap to augment the posterior wall, or it can be split laterally to create the posterior flap and avoid the use of the Fortunoff flap.

In other children, even after TUM, the vagina will remain a significant distance from the perineum. This condition will dictate that the vagina be separated from the urethra, and a pull-through type procedure performed. This approach is much less common but in this scenario the mobilized sinus can be split dorsally and used to create the anterior vaginal wall. This procedure allows for a vascular flap to be placed between the urethra and vagina, decreasing the risk of fistula as well as improving cosmesis.

Flap Vaginoplasty

Mucosa lined vestibule. If, after the vagina is opened over the Fogarty catheter, the posterior wall is able to reach the perineum easily with a minimum of tension, there are 2 options. The vagina may be sewn flush to the perineum as described by Pena,¹³ or a flap vaginoplasty may be performed.

If a perineal flap is selected, the posterior aspect of the mobilized sinus is opened posteriorly in the midline, and the omega-shaped flap is anastomosed to the normal caliber vagina. The incision is carried out only to the level of the planned introitus, and not into the vagina proper. In this situation the redundant mobilized sinus can be used to improve cosmesis. This redundant sinus tissue is opened in the midline “ventrally,” and the tissue is folded up to create a mucosa lined vestibule. This tissue will significantly improve the cosmetic appearance and prevent complications from excessive skin flaps. In the case of a virilized female the split phallic skin is then sutured alongside the opened sinus to create the labia minora (fig. 5). It must be reinforced that this technique only applies to the low to midlevel confluence, since it is not generally applicable to the high confluence, as noted previously.

Posterior vaginal wall sinus flap. As mentioned previously, we have some concerns about the use of a perineal skin flap for the posterior wall of the vagina. If, after opening the vagina over the Fogarty catheter, the posterior vagina is unable to reach the perineum easily, yet is not so high as to require separation and a pull-through, we use a laterally based flap of sinus tissue. The entire sinus is opened along its “lateral” aspect, and the exposed urethral meatus is anastomosed to the perineum (fig. 6,

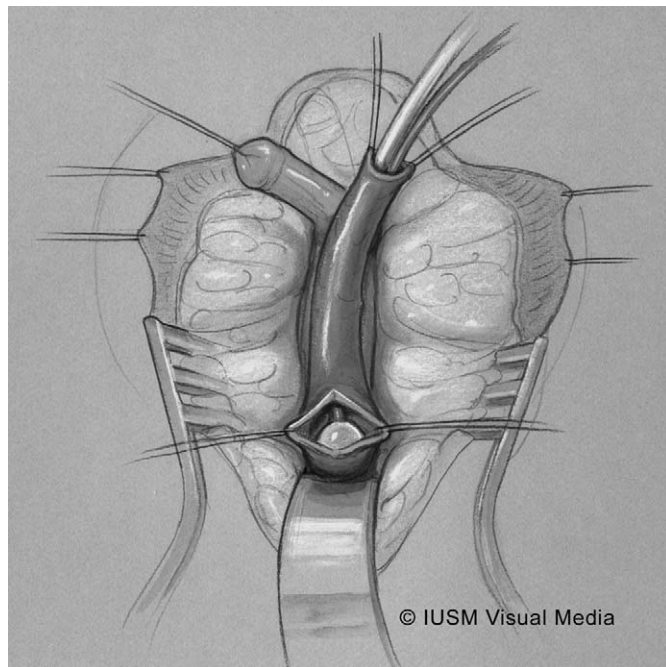


FIG. 4. After posterior vagina is dissected off rectum Fogarty catheter is palpable in vagina just above confluence with urethra. Stay sutures are placed lateral to planned vertical incision. Proximity of this opening to perineum will determine which flap is most appropriate to use.

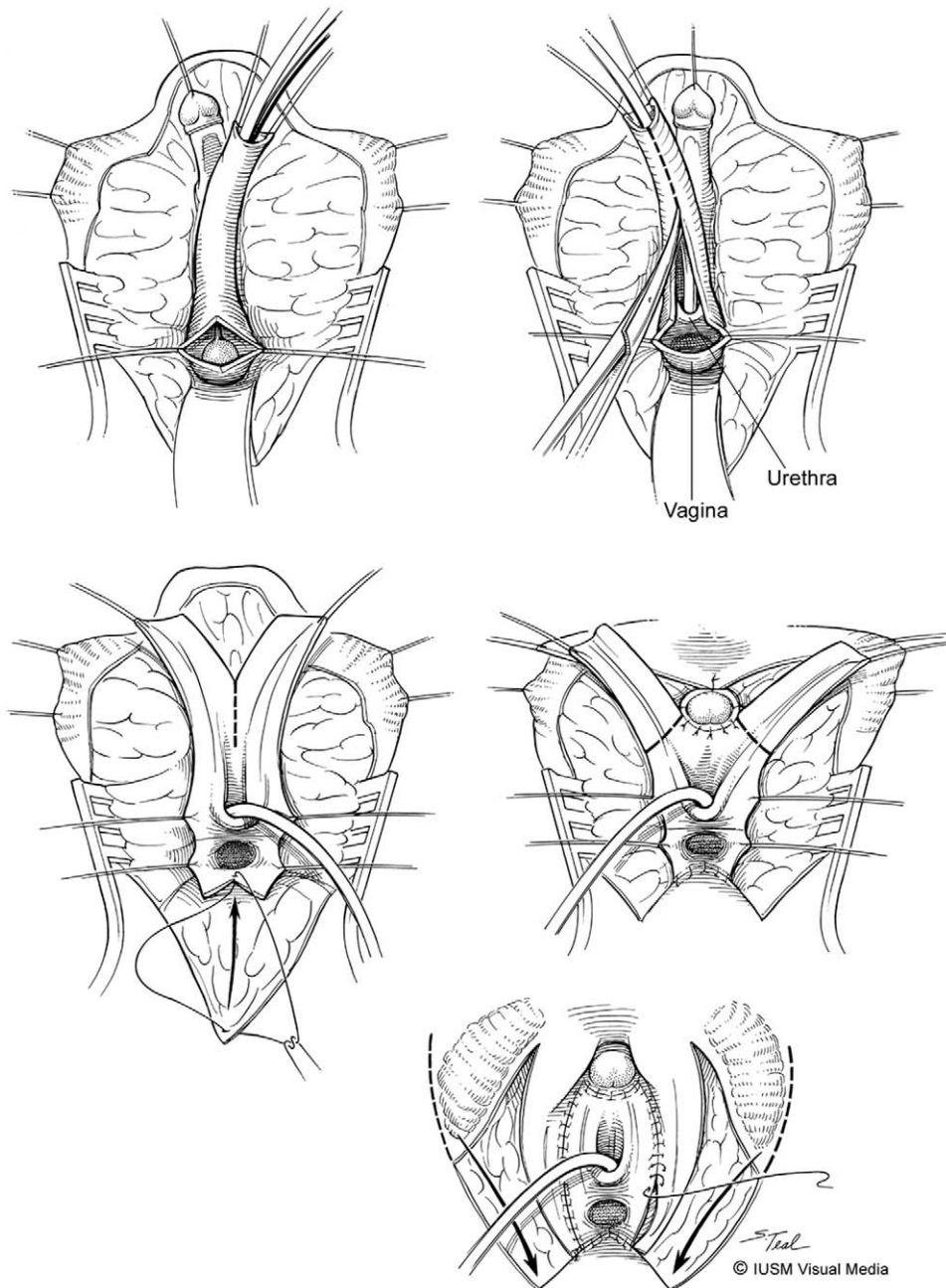


FIG. 5. Ventrally split flap can be rotated anteriorly so it lines vaginal vestibule. Urethral meatus is brought to its orthotopic position beneath clitoris, and sinus tissue is anastomosed to clitoris and labia minora, which have been fashioned from preputial flaps.

A). The sinus is then rotated laterally and posteriorly, and anastomosed to the posterior vaginal wall (fig. 6, B). This approach results in the extension of the vaginal wall to the perineum with sinus tissue and avoids the use of skin flaps. In essence, this procedure remains a “flap vaginoplasty” but the flap is constructed from sinus tissue rather than perineal skin.

Pull-Through Vaginoplasty (Anterior Wall)

After the vagina is opened over the Fogarty balloon it may be too high to reach the perineum easily without tension. This situation requires formal separation of the vagina from the urethra, and is based on the “pull-through” procedure originally described by Hendren and Crawford.¹²

We believe that this technique is best accomplished with the patient in the prone position and is facilitated by the mobility provided by TUM, which allows improved access to the confluence of urethra and vagina.¹ After separation the area of the previous confluence is closed in several layers, and the common sinus now forms the urethra (fig. 7, A). The previous mobilization allows the urethra to reach the perineum easily, leaving a significant amount of residual healthy sinus tissue. The anterior vaginal wall is formed by first splitting this redundant sinus in the midline “dorsally” (fig. 7, B). The tissue can then be folded back on itself and anastomosed to the distal vagina, creating a normal appearing anterior vaginal wall (fig. 7, C). This tissue also serves to cover the closed sinus and helps

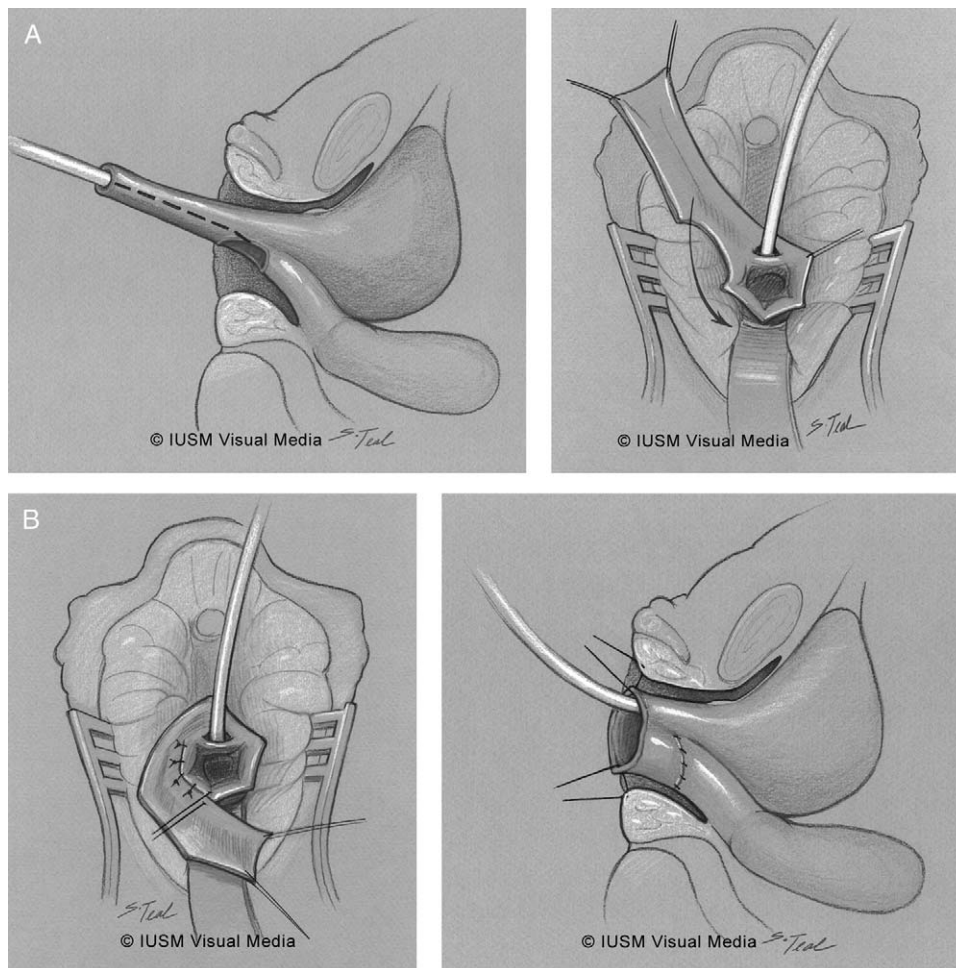


FIG. 6. *A*, to avoid excessive use of skin in posterior vagina, sinus is split along its lateral aspect and rotated to distal vagina. *B*, laterally based spiral flap brings excess UG sinus tissue to posterior wall of vagina. Side view reveals vagina is now flush with perineum, and Fortunoff flap is not required.

prevent fistula formation. The posterior vaginal wall often requires a posterior perineal flap (fig. 7, *D* and *E*).

Finally, we have found that the mobilized sinus tissue is extremely useful even if vaginoplasty is not performed at the time of mobilization. In the severely virilized patient with CAH, eg Prader 5, the vagina may be inadequate for reconstruction. In this scenario we defer the vaginoplasty until after puberty, after the accumulation of menstrual fluid has naturally expanded the tissue. However, the sinus still requires mobilization to bring the urethra to its orthotopic position, and labioplasty is also performed. Therefore, we will split the sinus ventrally and it again is used for a mucosa lined vestibule, greatly improving the cosmesis of the external genitalia.

DISCUSSION

Urogenital sinus abnormalities are extremely challenging surgically, because they require meticulous technique, prolonged operative times, and the ability to think and plan in 3 dimensions. Techniques continue to be modified, and all are based on the work of our predecessors.^{1,10,15-17,19} Current techniques, especially with the introduction of TUM, have simplified dissection and, some believe, decreased surgical complications.^{3,19,20}

The original description by Pena of TUM limited its use to UG sinuses less than 3 cm long and called for discarding the excess sinus tissue.^{3,13} However, based on our experience, the length of the urethra, as measured from the bladder neck to the confluence, is a much more important predictor of surgical complexity.⁹ Furthermore, we believe that the UG sinus tissue can be an invaluable aid in reconstruction. The sinus provides well vascularized and malleable tissue appropriate for a variety of applications.

The use of TUM and other genital surgery remains controversial. Thus far, all results are short term. It remains to be seen if the circumferential dissection around the proximal urethra will result in any injury to the sphincteric function or innervation, with subsequent incontinence. One must be especially cautious with the high confluence when TUM and pull-through vaginoplasty are required. Also, we are concerned about the long-term risk of stress urinary incontinence introduced by extensive mobilization of the bladder neck toward the perineum. Although Kryger and Gonzalez have recently written an encouraging report regarding preservation of urinary continence, their oldest patient was 22 years at last followup.²⁰ Therefore, our concern remains for results seen in late adulthood, especially in the postpartum

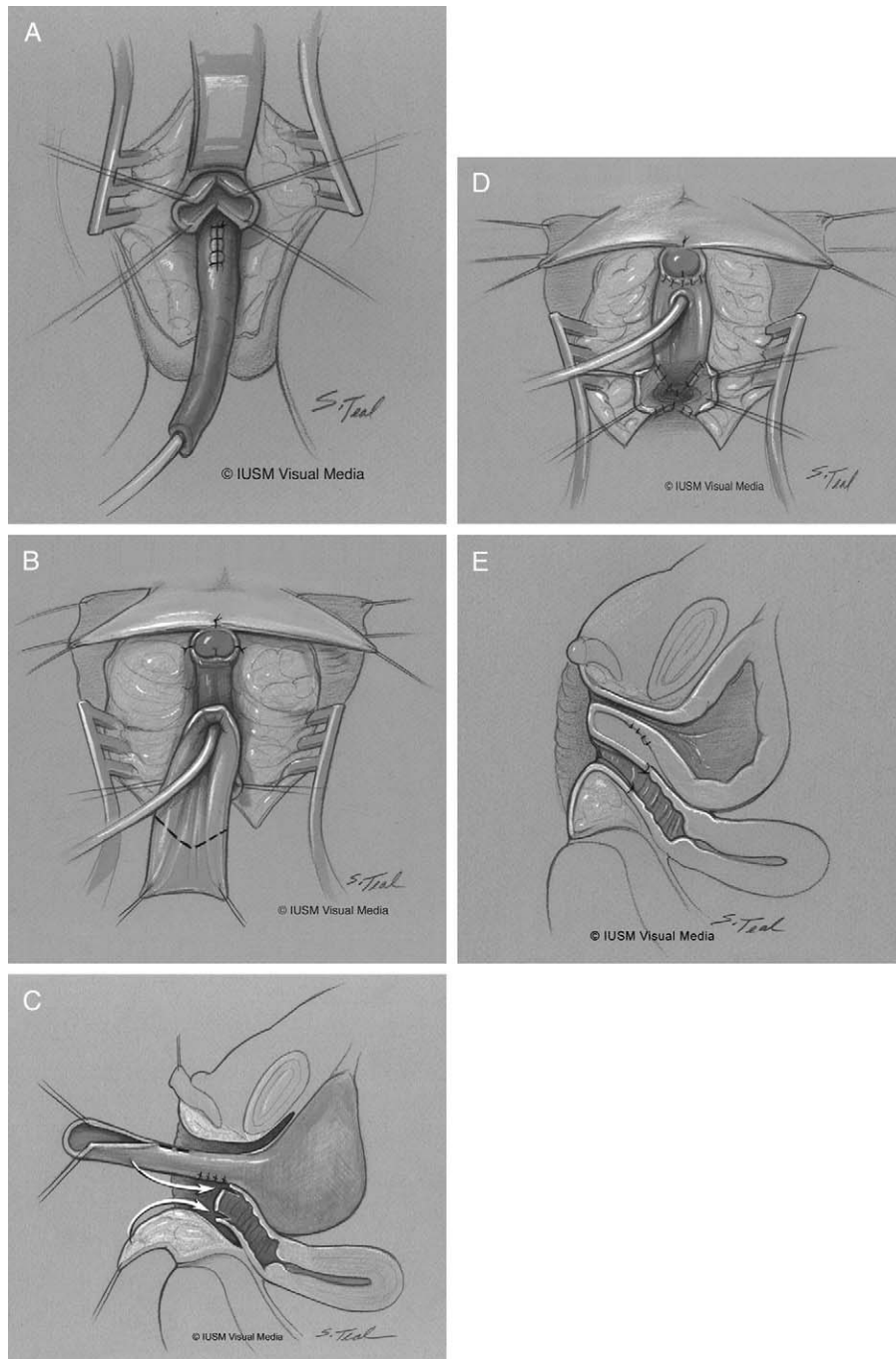


FIG. 7. *A*, prone view of mobilized UG sinus after vagina has been dissected off of UG sinus. Defect in sinus has been closed in several layers and now forms urethra. *B*, with patient in supine position urethra has been mobilized to perineum, and excess sinus is split on its dorsal aspect to create orthotopic meatus. *C*, flap is next rotated posteriorly (arrows) to create distal anterior vaginal wall and provide tissue support for urethral anastomosis. *D*, completed pull-through vaginoplasty with anterior vaginal wall formed from dorsally opened UG sinus in Passerini-Glazel fashion. *E*, sagittal view of completed anterior vaginal wall flap demonstrates coverage of urethral incision line.

population. Thus, each surgeon must address each area of concern with the family and other members of the medical team.

CONCLUSIONS

Not every patient with a persistent UG sinus needs TUM, but when this procedure is elected we believe that all sinus tissue should be incorporated into the reconstruction. We

have described our 3 techniques for vaginoplasty using the mobilized sinus. To our knowledge our spiral flap has not previously been described in the literature, and we believe that this technique will improve outcomes by reducing the reliance on skin flaps. Since most patients undergo surgery in infancy, ultimate outcomes regarding intercourse, sexual sensitivities and urinary incontinence are many years away.

Abbreviations and Acronyms

CAH	=	congenital adrenal hyperplasia
TUM	=	total urogenital mobilization
UG	=	urogenital

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