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Surgery Illustrated – Surgical Atlas
Urogenital mobilization for urogenital sinus repair

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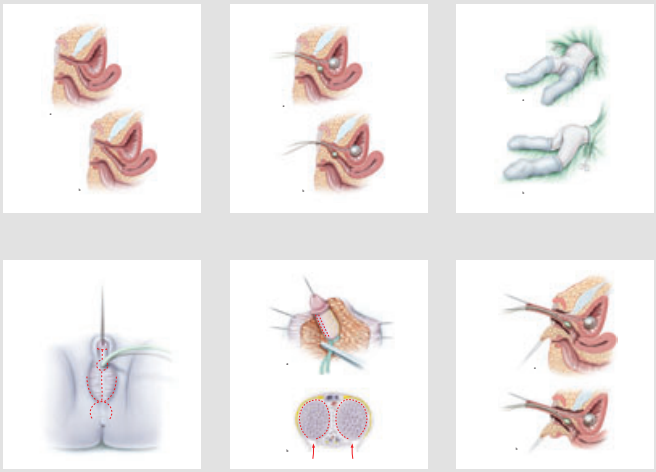
ILLUSTRATIONS by STEPHAN SPITZER, www.spitzer-illustration.com



INTRODUCTION

The care of the patient with a persistent urogenital sinus (UGS) is complex, challenging and controversial. Incomplete separation of the urethra and vagina during fetal development results in these two structures sharing a common channel (UGS) to the perineum. The vagina and urethra might join anywhere along a spectrum from the bladder neck, with a long common channel (high confluence; Fig. 1a) to those that join near the perineum with a short common UGS (low confluence; Fig. 1b). The higher the confluence, the more complex the reconstruction and the greater the risks. The surgical management requires meticulous attention to detail with careful tissue handling.

INDICATIONS

The vast majority of patients with UGS abnormalities have genital ambiguity and are thus identified as neonates. This ambiguity is most commonly secondary to congenital adrenal hyperplasia (CAH). There are patients with 'pure' UGS abnormalities where there are no obvious external genital abnormalities, and these patients might not be identified until puberty when they present with problems with menstruation. At the severe end of the spectrum is the child with a persistent cloaca, where the rectum also joins this common urogenital channel.

In this article we focus on reconstruction for patients with CAH, as they overwhelmingly make up the largest group, but the surgical steps are applicable to any child with a UGS anomaly. The indications for surgery in these

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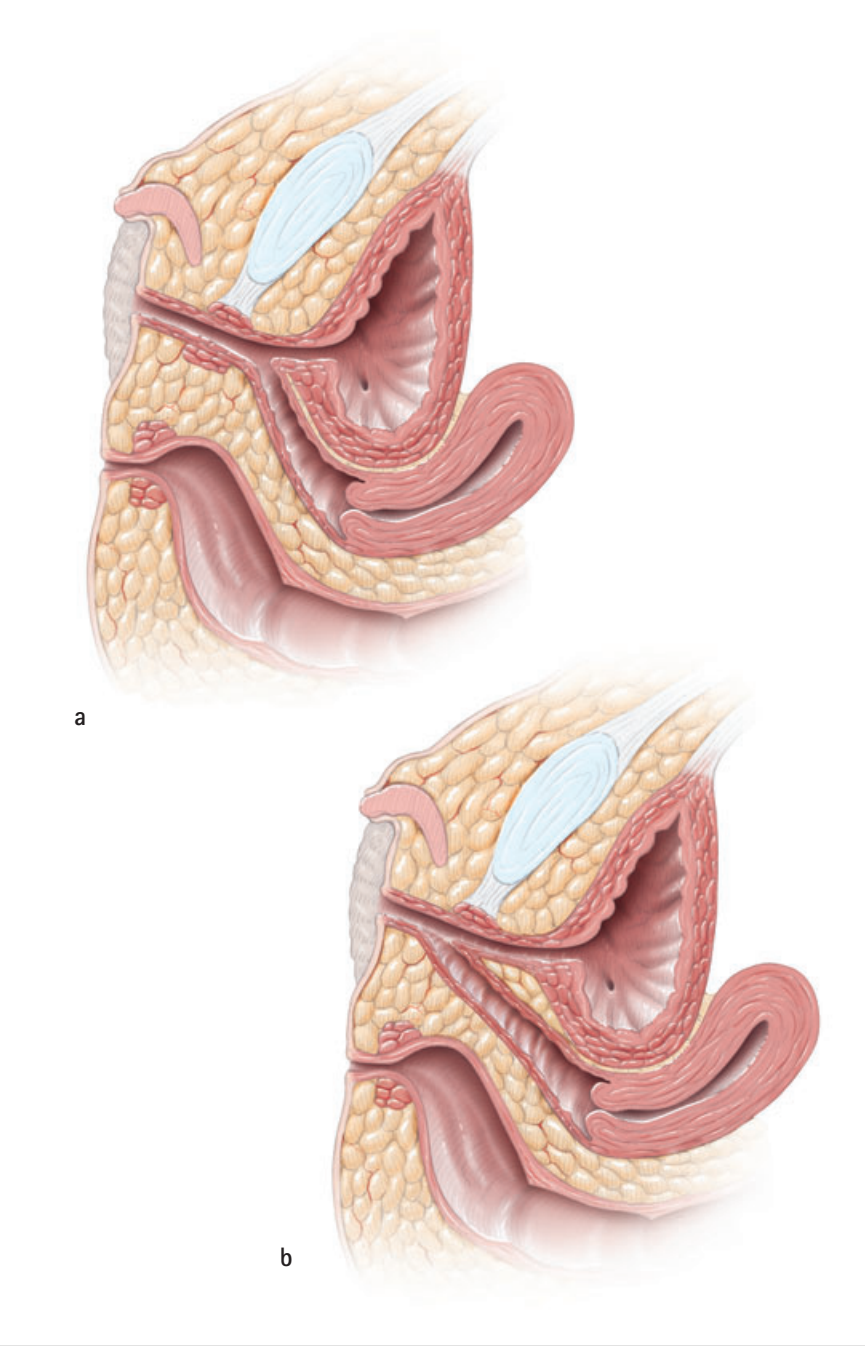
virilized girls are controversial and beyond the scope of this surgical article, but it is imperative that these children be identified and evaluated immediately after birth to manage the potentially life-threatening fluid and electrolyte changes, and to make the appropriate gender assignment. All children with genital ambiguity must be managed promptly by a multidisciplinary team which includes a neonatologist, endocrinologist, paediatric urologist (surgeon) and psychologist, who work in concert with the parents. All issues and differing viewpoints about gender assignment and surgical reconstruction should be shared with the parents, including those of advocacy groups such as the Congenital Adrenal Hyperplasia and Education Support or the Intersex Society of North America. The parents should be allowed to make an informed decision about the surgical management for their child. For purposes of this article it is assumed the family and multidisciplinary team want to proceed with surgical management.

PATIENT PREPARATION

All children with a UGS abnormality should be assessed by pelvic ultrasonography. Those with pure UGS anomalies often have associated upper urinary tract or spinal cord abnormalities and should have renal ultrasonography. The spine should be evaluated by ultrasonography in the neonate or MRI in the older patient. Those with CAH should be well managed by their endocrinologist and receive stress-dose steroids preoperatively. The anatomy should be defined before surgery by genitography and cystoscopy noting the normalcy of the bladder, urethra and the number and location of the vagina(s). The relationship of the vagina to the bladder neck is the most critical determining factor in the type of vaginoplasty to be performed. Cystoscopy can be done as part of the initial diagnostic evaluation in complex UGS abnormalities, but most often can be done at the time of reconstruction in those with CAH, avoiding additional anaesthesia.

Historically, a full bowel preparation has been given before surgery; we now reserve this for those with a very high confluence UGS. Most patients have a preoperative enema and all receive broad-spectrum i.v. antibiotics. The rectum is irrigated with 0.25% neomycin solution in the operating room before the procedure. Because of the virilization associated with CAH patients will also have virilized external genitalia, including clitoromegaly, absence of labia minora and anteriorly placed fused labia majora. The operative correction therefore usually involves three steps, i.e. clitoroplasty, labioplasty and vaginoplasty.

Timing for surgery is another area that is controversial, with two main opposing but rational views. Some think that all three procedures should be done in combination when the patient is a neonate. Others think that delaying these procedures until puberty or until the child can participate in the decision for or against surgery is appropriate. There is a hybrid of these two, whereby the clitoroplasty and labioplasty are performed as a neonate and the vaginoplasty is delayed until puberty. The arguments for these viewpoints have been well addressed elsewhere and will not be repeated in this illustrated surgical text, but the surgeon should be familiar with the pros and cons of each approach. The consensus statement on CAH by the Lawson-Wilkins Paediatric Endocrine Society and the European Society for Paediatric Endocrinology recommends surgery when the patient is 2–6 months old for those with a high confluence, and surgery between 12 months and adolescence is not recommended. Regardless of timing, the following steps are applicable.



SURGICAL STEPS

Figure 2

With the child in the lithotomy position, endoscopy is used to assess the common UGS. The distance from the bladder neck to vagina (level of confluence) and the vagina to the perineal meatus (length of common channel) is measured. A Fogarty catheter is placed in the vagina with the balloon inflated, and a Foley catheter is placed in the bladder. The surgery can be done in this position but we prefer to keep the catheters sterile while returning the child to supine.

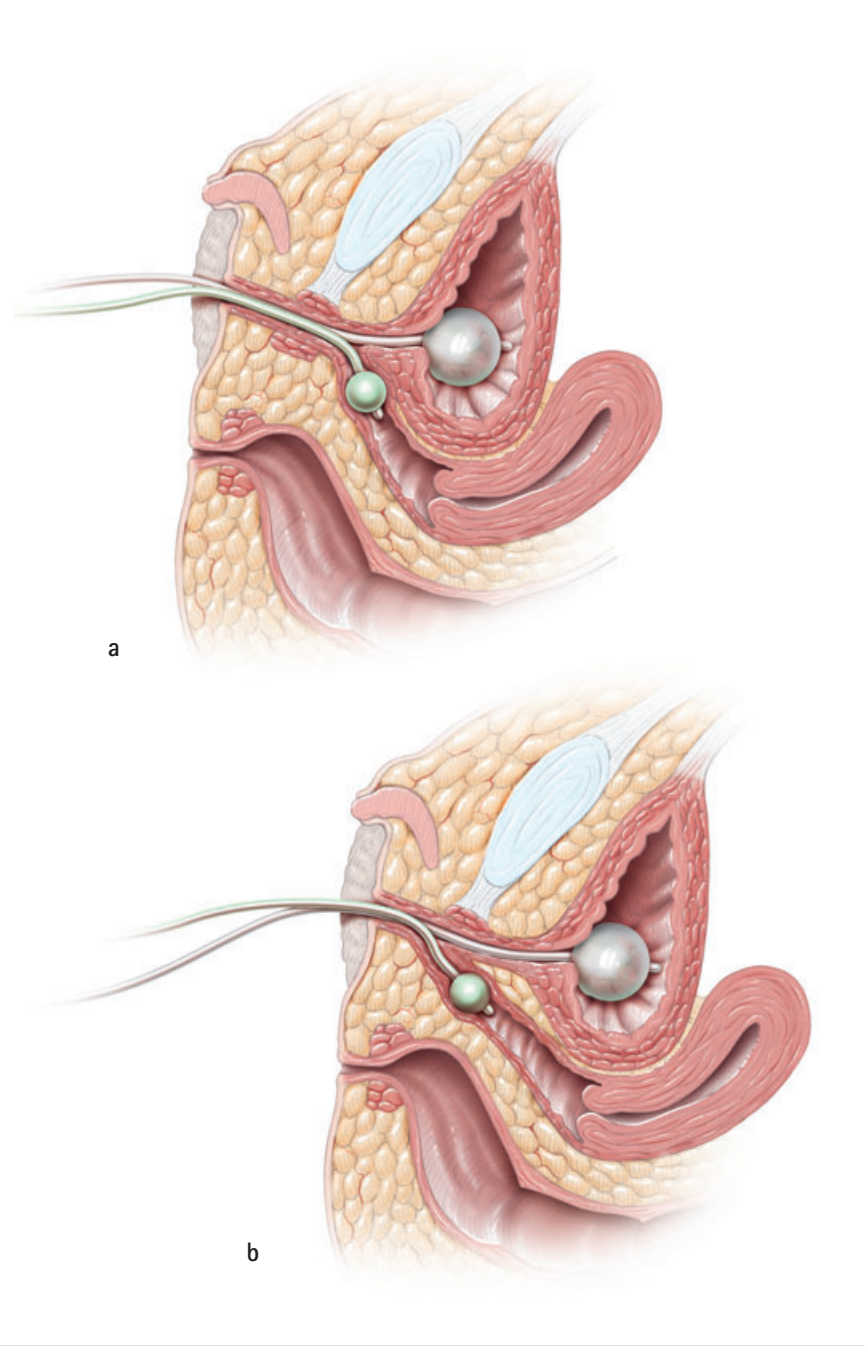
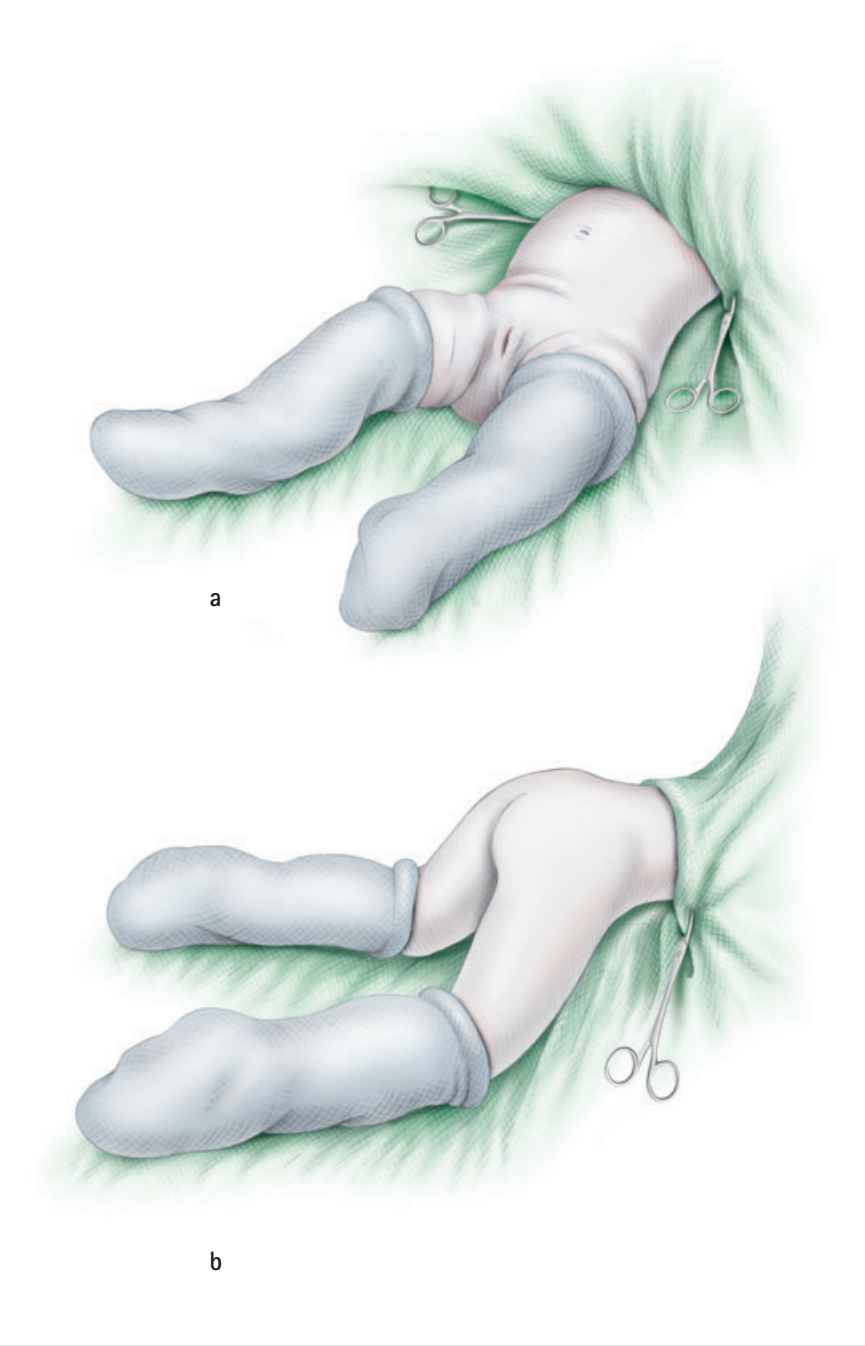


Figure 3

We then prepare the entire lower body, nipples to toes, front and back, with povidone iodine solution. The legs are wrapped and the entire lower body is passed through the aperture in the drapes. The buttocks are elevated on sterile towels giving access to the perineum and abdomen. This also allows us to rotate the child to prone when necessary. Furthermore, with this preparation there is much better vision for the surgical assistants/trainees.



PROCEDURE

With the child placed supine, we place a moistened radio-opaque sponge in the rectum with a suture attached so that it is easily retrieved at the end of surgery. We now approach all children with the same initial plan, regardless of the location of the vagina.

Figure 4

A traction suture is placed through the clitoris in those with CAH. The proposed incision lines are as shown in Fig. 4. An omega-shaped perineal flap is drawn. The inferior aspects of the labia majora are outlined. A circumferential incision is marked around the meatus, and depending on the location of the meatus parallel incisions are made along either side of the urethral plate. Along the proposed incisions, 0.5% lidocaine with 1 : 200 000 adrenaline is injected s.c. to aid in haemostasis. In children with CAH with significant clitoral hypertrophy, the clitoroplasty is then performed, avoiding any risk to the neurovascular bundles or tunics.

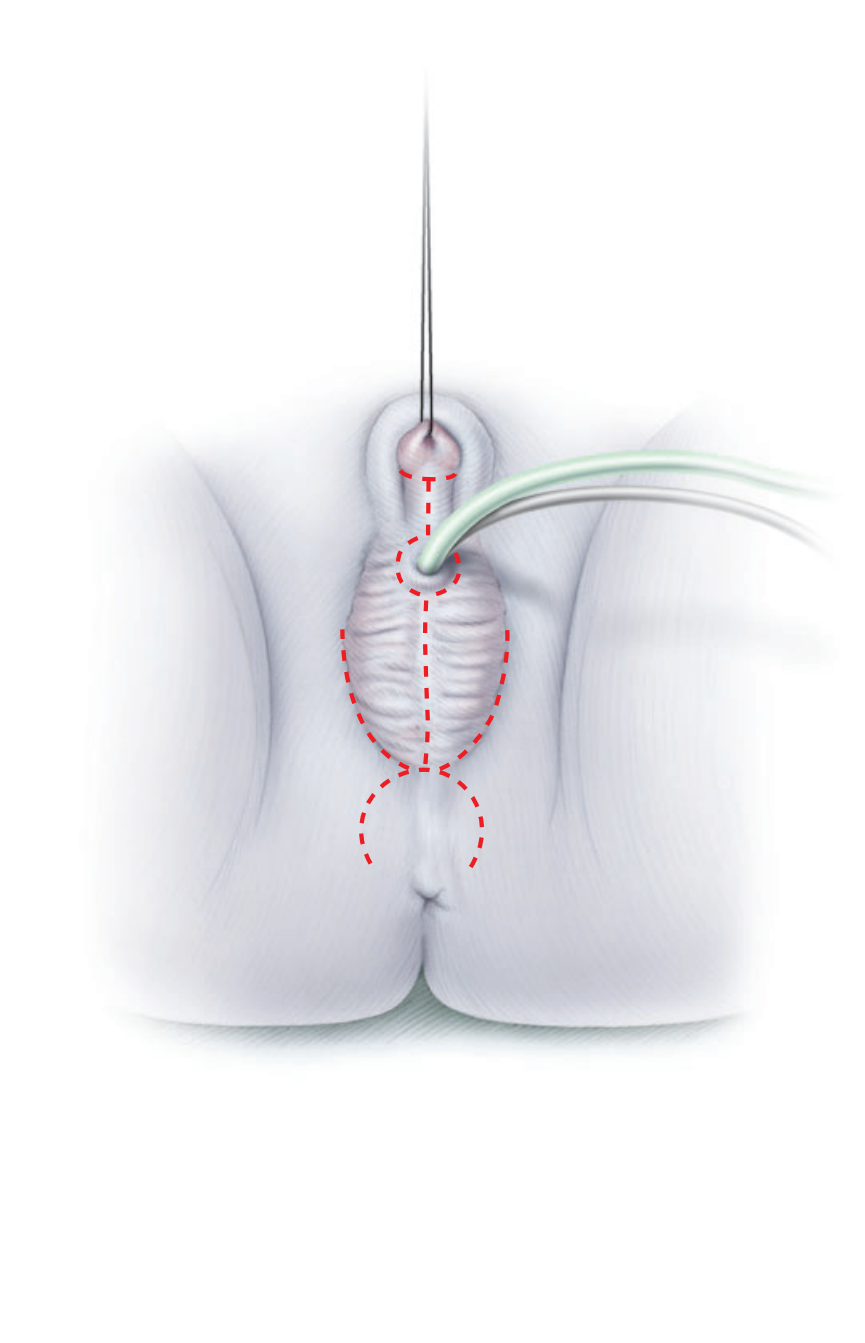
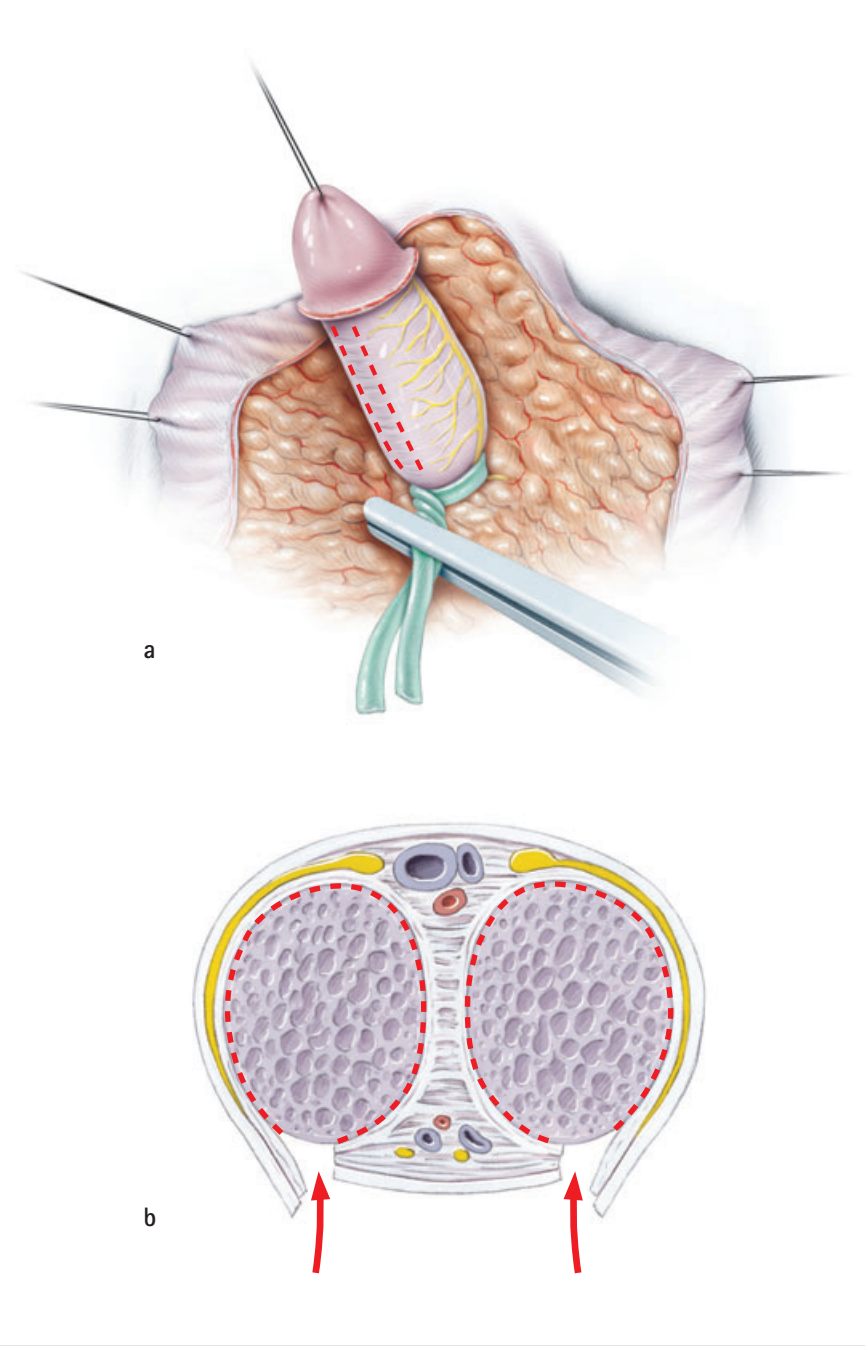


Figure 5

The corporal tissue is removed through ventral incisions. The surgeon must not only provide an excellent cosmetic outcome but also retain all clitoral innervation for optimal sexual function. This portion of the procedure has been well described elsewhere and will not be detailed here. However, importantly, the phallic skin is preserved, which at times can be beneficial for use in repairing the UGS.



UGS

Figure 6

Currently we use UG mobilization in nearly all children. Pena [1] initially described as 'total urogenital mobilization' (TUM) in 1997. In this procedure, the sinus is dissected circumferentially and mobilized. The dissection is carried through the pubourethral ligament anteriorly behind the pubic bone and posterior to the vagina allowing the entire UG complex (sinus, urethra, bladder, vagina) to move toward the perineum. Pena amputated the redundant mobilized sinus. We have shown this mobilized tissue to be very valuable in the repair, and it can be used to create a mucosal-lined vestibule, a posterior flap for the vagina or the anterior wall of the vagina [2]. Furthermore, we showed that the vast majority of patients do not need the more aggressive complete mobilization but can be managed by partial UM (PUM) stopping the dissection at the pubourethral ligament [3].

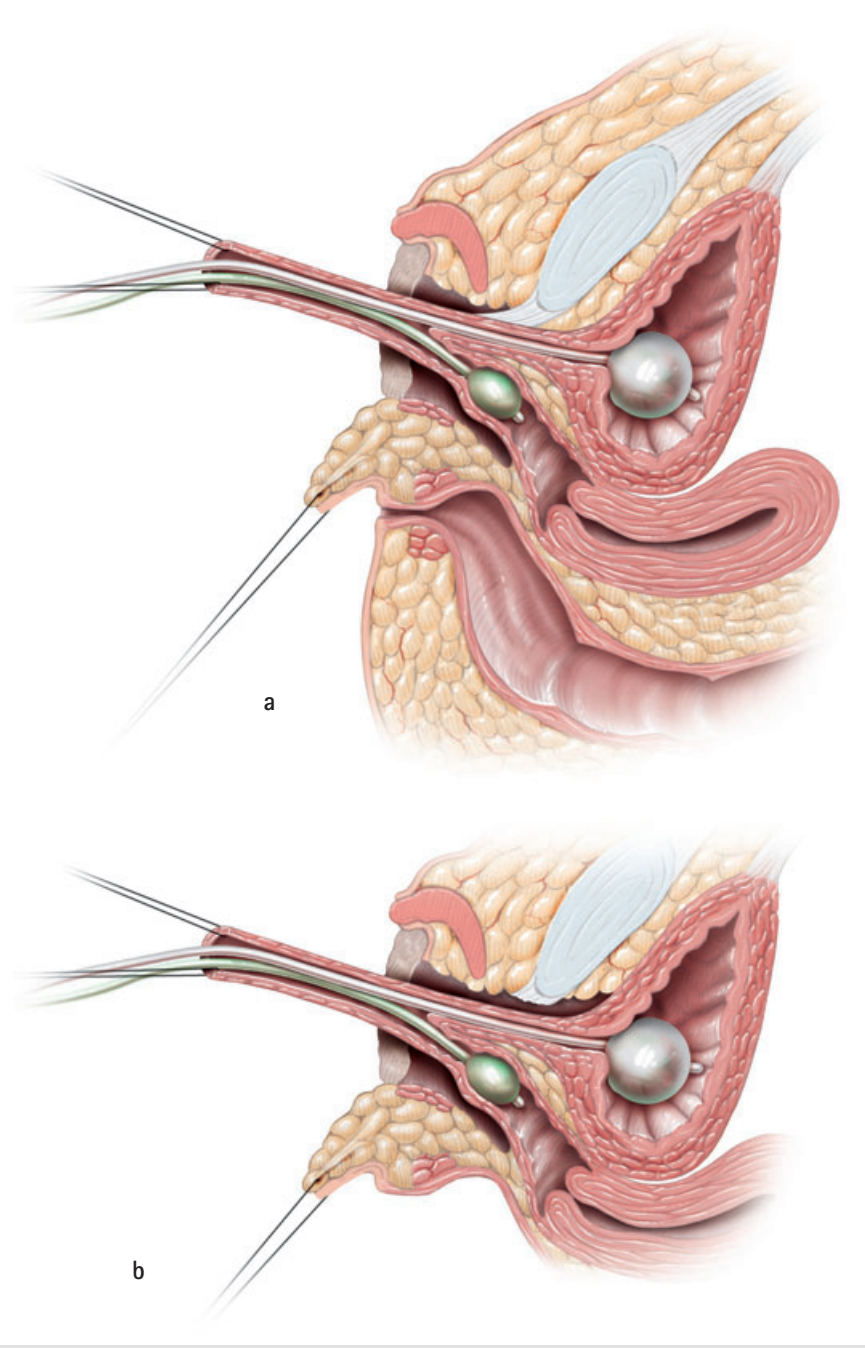


Figure 6a

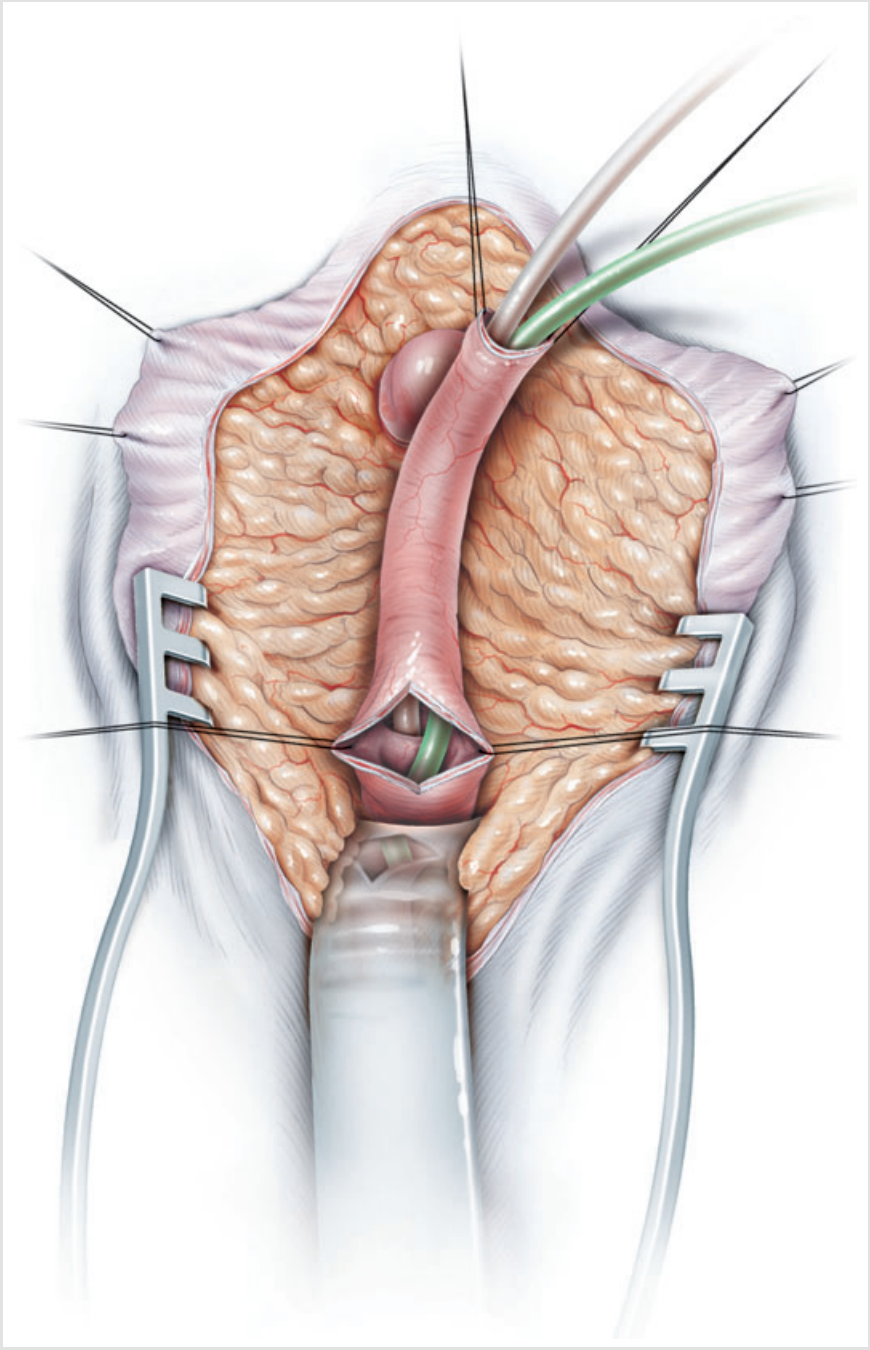
The UGS is dissected from the corporal bodies. In the very virilized patient, this is similar to mobilizing the urethra from the corpora, and in the less virilized the urethral plate is mobilized to the level of the meatus and then the entire urogenital sinus mobilization is continued to beneath the bifurcation of the corporal bodies. In all patients initially, we stop our dissection at this point at the level of the pubourethral ligament (i.e. a PUM).

It is helpful to place traction sutures around the sinus meatus. The perineally based omega-flap incision is made. It is beneficial to leave some subcutaneous fat on this flap to prevent compromising the flap vascularity. This exposes the posterior aspect of the UGS. It is imperative that the perineal flap be long enough to easily reach the normal calibre of the vagina with a tension-free anastomosis. One of the more difficult aspects of the procedure is separating the posterior aspect of the sinus from the urorectal musculature. It is important to stay in the midline posteriorly, with care not to injure the rectum or enter the sinus. Once the musculature has been separated the entire posterior aspect of the vagina can be easily recognized by its white appearance, and the remaining attachments to the posterior aspect of the vagina are then easily swept toward the rectum. It is helpful to place a small malleable retractor to retract the rectum from the sinus and posterior aspect of the vagina. The Fogarty balloon lying within the vagina is palpated.

Figure 6b

In those with a higher urogenital confluence a TUM might be required. In this procedure the pubourethral ligament is divided, allowing more mobilization of the entire UG complex.

Figure 7



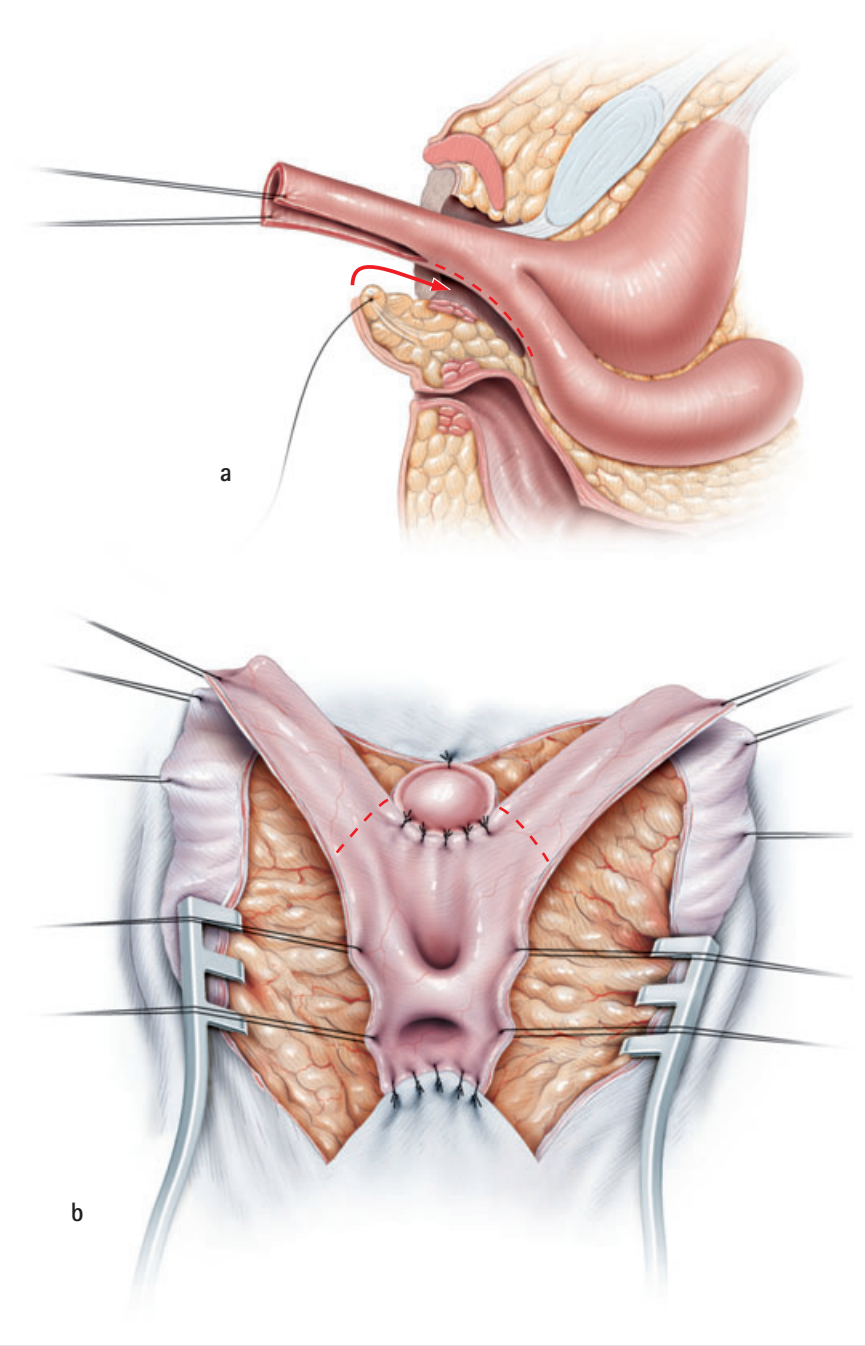
The posterior wall of the vagina is opened over the Fogarty catheter between stay sutures. It is at this point that the most difficult decisions are made. The surgeon must determine whether the vagina will easily reach the perineum. If so, we would do a flap vaginoplasty. If the vagina remains far from the perineum, then the surgeon must consider proceeding with further dissection anteriorly through the pubourethral ligament i.e. a TUM, or doing a 'pull-through vaginoplasty', where the vagina is completely separated from the UGS, or a combination of these two. Regardless of the decision made, it is imperative to realise that the distal third of the vagina is usually narrowed and must be opened into its normal calibre to prevent postoperative stenosis.

VAGINOPLASTY FOR THE LOW OR MODERATE LEVEL CONFLUENCE

PUM as described above is nearly always adequate for the low or moderate confluence UGS. We have found that the mobilized sinus tissue is very helpful in achieving improved cosmetic and functional results, allowing us to use true UGS tissue in the reconstruction of the introitus and vagina rather than skin flaps, which have a propensity to stenose.

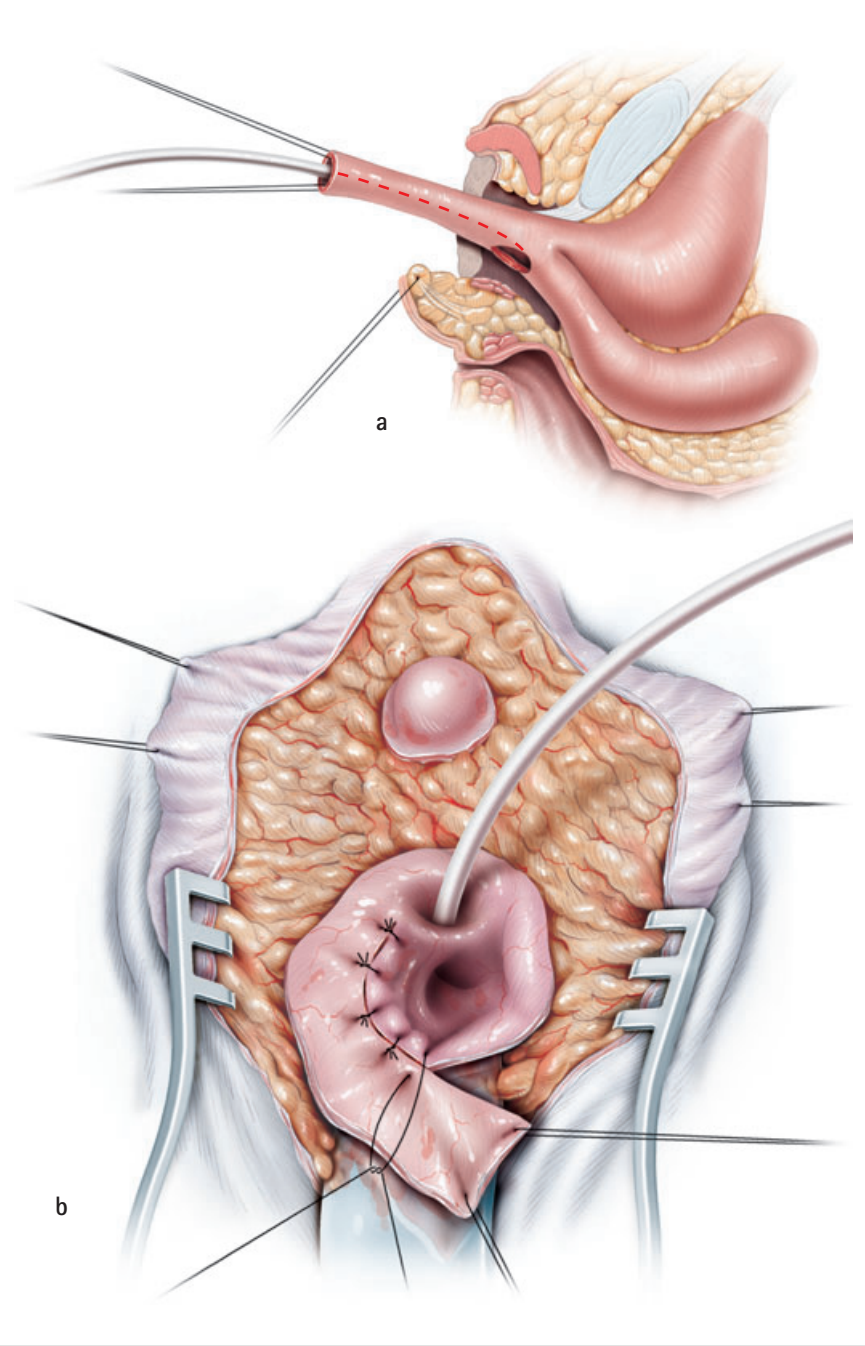
Figure 8

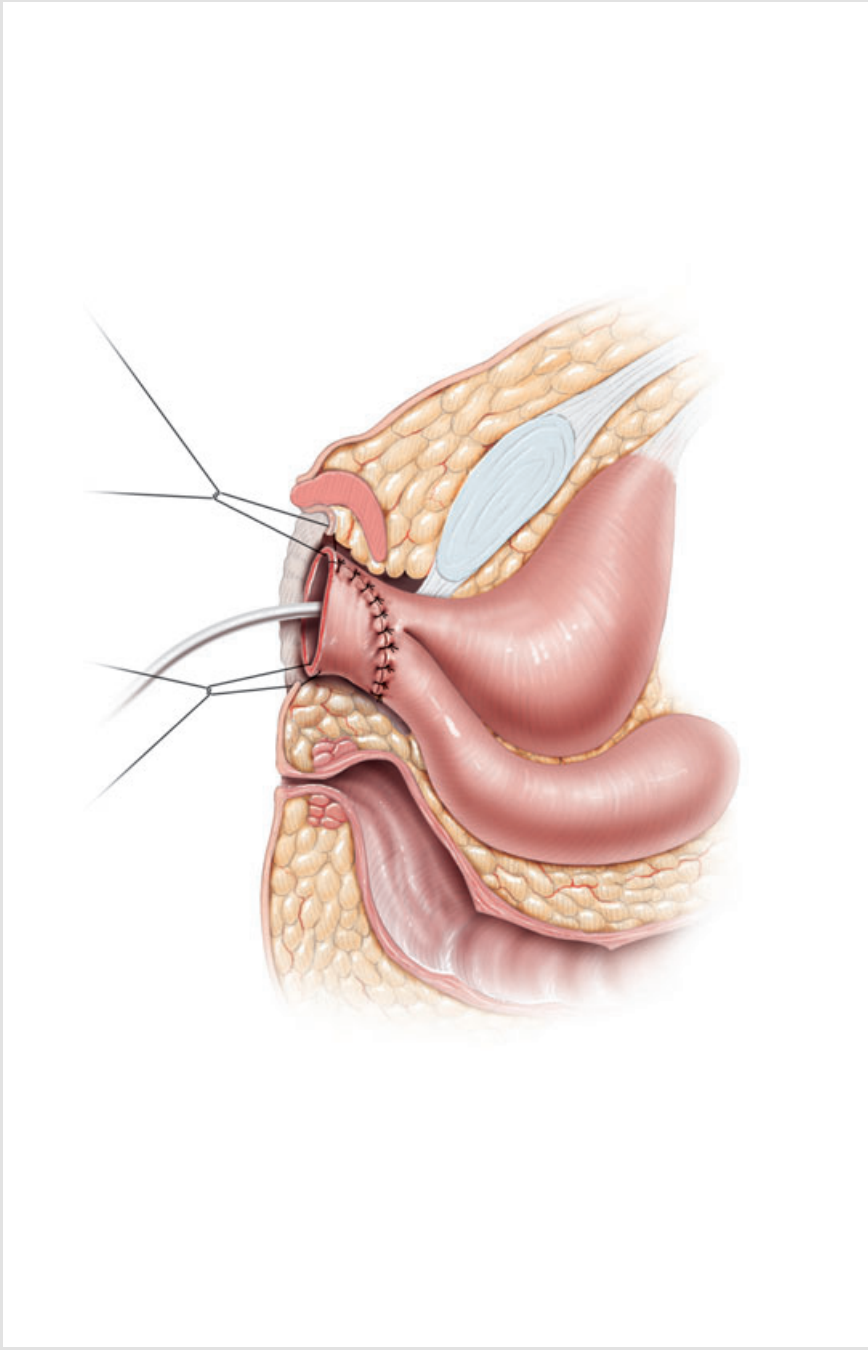
In the classic 'flap vaginoplasty', once the vagina has been opened widely posteriorly into the normal-calibre vagina, the perineal flap skin is sutured in place with interrupted absorbable sutures, to complete the flap vaginoplasty. In this situation the redundant mobilized sinus tissue is split ventrally to create a mucosa-lined vestibule.



Figures 9 and 10

More recently, we have described opening the sinus laterally allowing the sinus to rotate to create a posterior wall of the vagina, eliminating the need for the posterior skin flap. This latter manoeuvre requires the mobilized sinus to be long and well vascularized.





VAGINOPLASTY FOR HIGH CONFLUENCE

If the sinus confluence is very high near the bladder neck and does not readily reach the perineum with a PUM, there are two options.

Figure 11

Historically, traction sutures have been placed around the edge of the sinus and the anterior wall of the vagina is separated from the UGS in what is known as a 'pull-through' vaginoplasty.

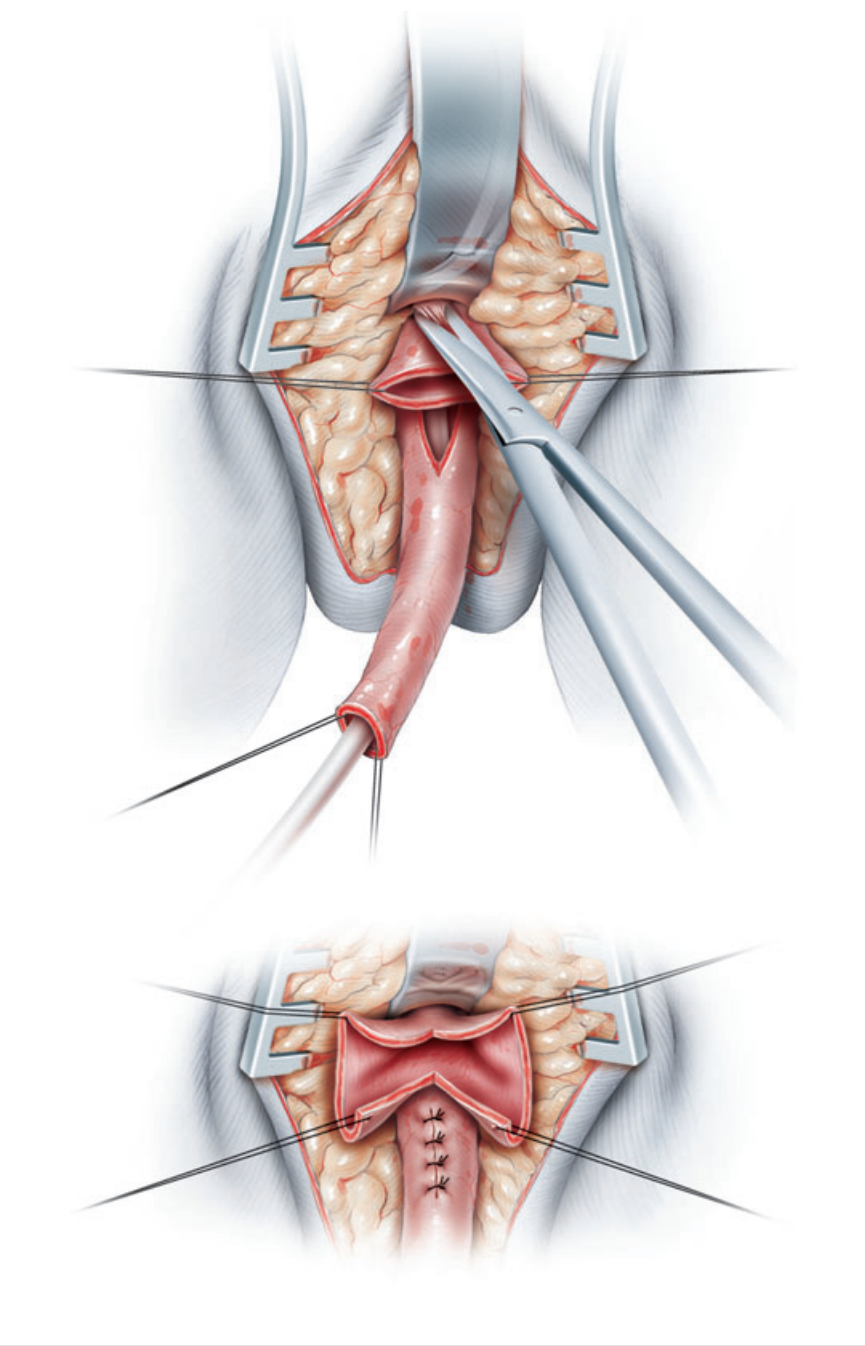


Figure 12

If decide that the vagina needs to be separated from the urinary tract, we have found this to be much easier with the child prone. We therefore will rotate the child prone within the aperture of the drape to do this part of the operative procedure. In this situation the mobilized sinus is divided dorsally and then used as a Passerini-like flap to create the anterior wall of the vagina. There are no easily identifiable tissue planes in the beginning of this dissection and the surgeon should always err toward entering the vagina rather than the urinary tract. With the vagina completely mobilized from the urogenital sinus, the opening in the UGS created by separating the vagina is closed in two layers using 6–0 monofilament polyglyconate suture. A second layer is closed over this with running 6–0 polyglyconate. We then cover these two layers with a mobilized fat pedicle to prevent fistula formation.

The other option before a pull-through vaginoplasty is to proceed with a TUM by extending the anterior dissection of the UGS through the pubourethral ligament. The dissection carries on proximally beneath the pubis and as the avascular ligaments from the pubis are divided, the entire UGS is felt to 'give' and move toward the perineum. This procedure might avoid a pull-through vaginoplasty, allowing the vagina to move out further toward the perineum so that it can be treated similarly to the above described low and mid level confluence vagina. However, if the vagina remains high, then a pull-through vaginoplasty can be done as shown in Fig. 12.

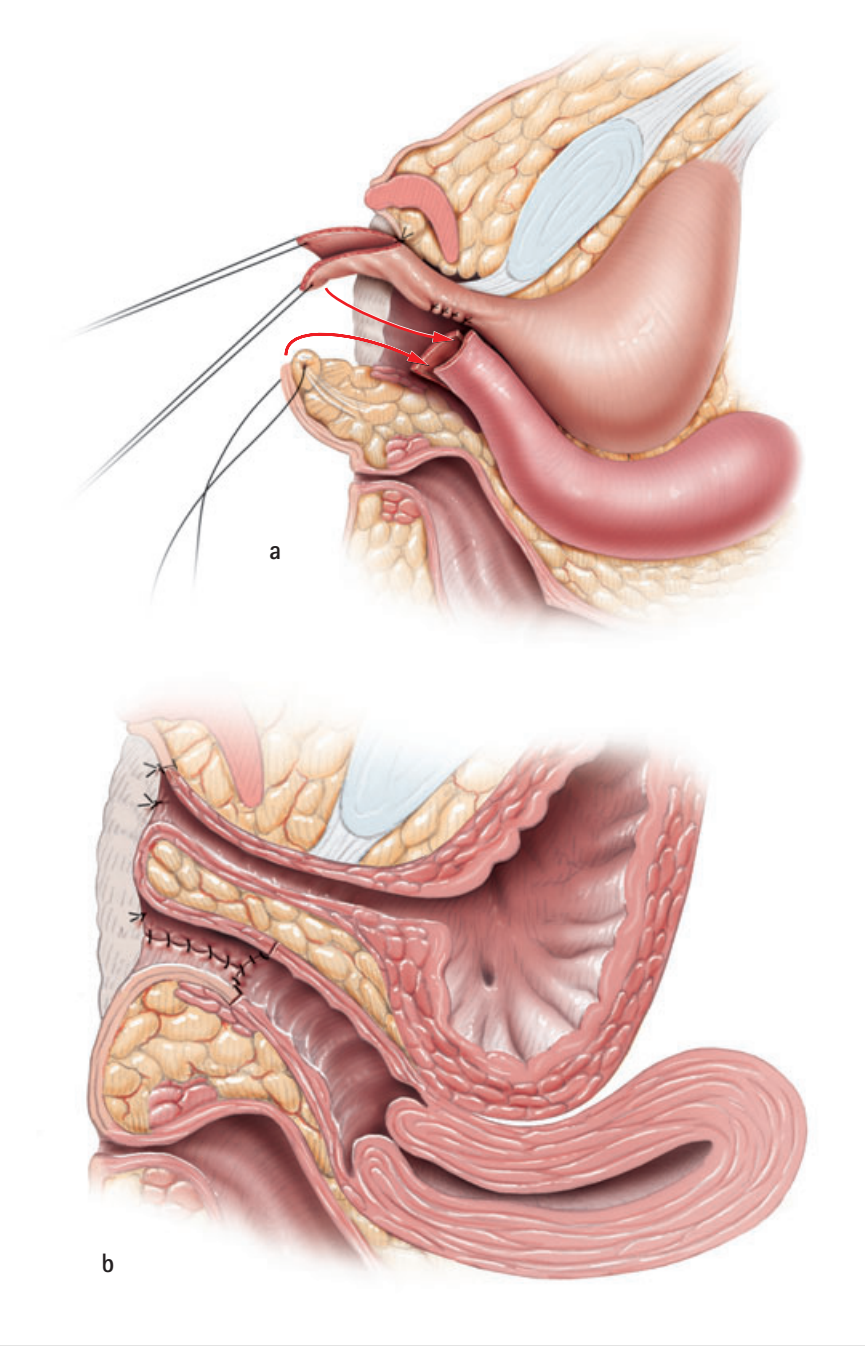
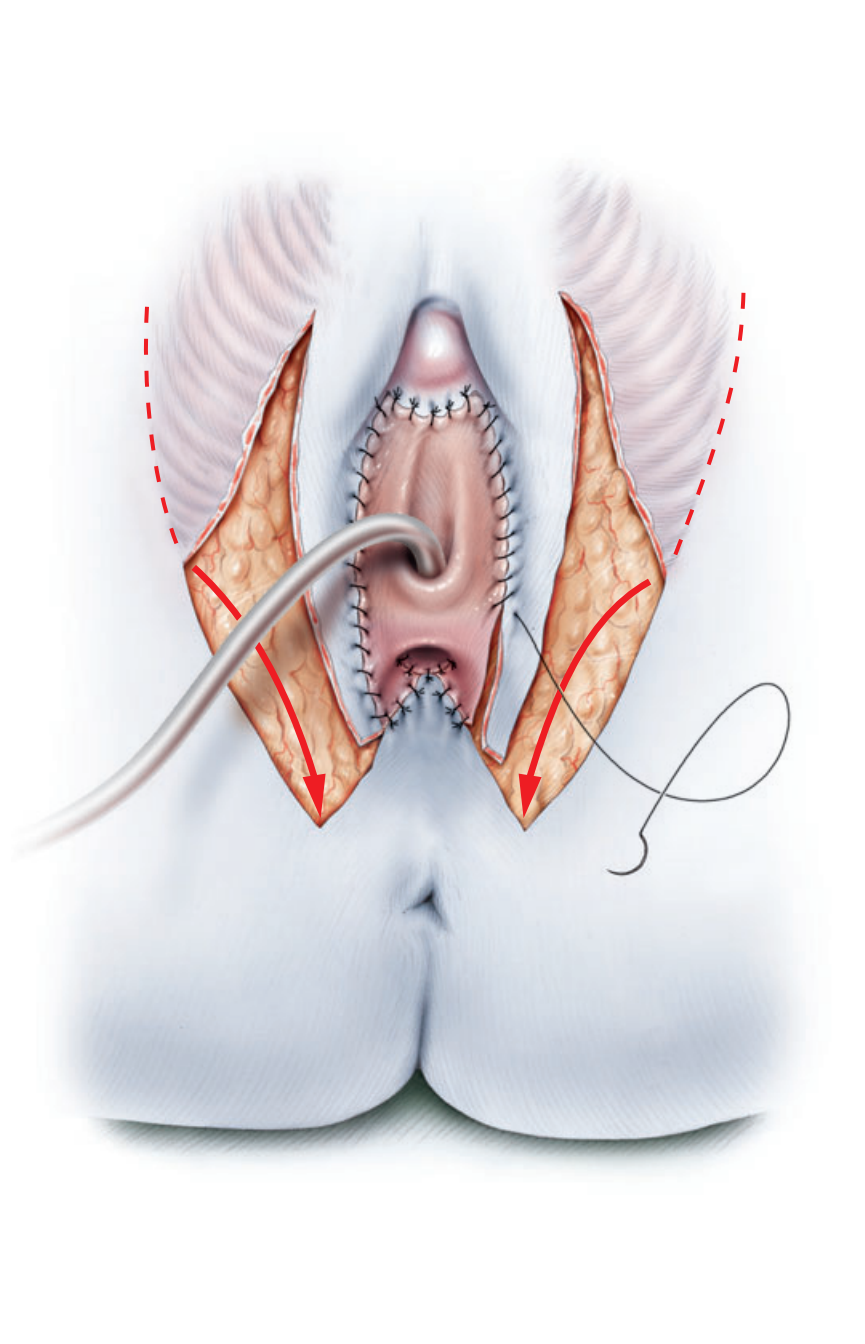


Figure 13

In the patients with CAH the clitoris is repositioned and secured to the corporal bodies at their bifurcation. The redundant tunics with their neurovascular bundles are folded back into a subcutaneous pocket. Creating labia minora from the phallic skin and moving the labia majora inferiorly by Y-V plasties complete the labioplasty. This is critical to achieve more normal cosmesis, allowing the vagina to reside between the labia rather than appearing as a separate isolated hole.



POSTOPERATIVE CARE

A Foley catheter is left indwelling; it can be removed early after surgery in the low-confluence flap vaginoplasties. If a pull-through procedure has been done and the sinus has been closed to create a urethra, we would leave the Foley in place for at least a week.

A Penrose drain is placed within the vagina. We do not place any vaginal packs as we believe these can result in some vascular compromise of the healing vagina. The length of time the Penrose is left in place is based on the degree of complexity of the vaginoplasty, but it can usually be removed in 2–3 days.

We place a wound sealant (Indermil, Syneture, Norwalk, CN; or Dermabond, Ethicon Inc., Somerville, NJ, USA) over the incisions followed by a telfa pad and fluffed gauze. Foam tape is criss-crossed over the perineum from abdomen to buttocks. We then wrap the child's legs with an elastic wrap, with foam placed between the knees and ankles, for 1–2 days.

Caudal blocks done administered by our anaesthesiologists have been very helpful in postoperative pain management. For the most significant repairs an epidural catheter can be placed; this has been very beneficial in providing patient comfort, but does require careful patient monitoring.

FROM SURGEON TO SURGEON

We have now seen several children (not with CAH) with a vagina(s) that enters into the bladder neck or back wall of the bladder. The anatomy can be extraordinarily complex in these patients, with some having duplicated vaginas and/or bladders. If the anatomy cannot be understood endoscopically or via a perineal surgical approach, it is very helpful to open the abdomen. While this is virtually never required for patients with CAH or routine PUM/TUM procedures, it can be very helpful in delineating the anatomy in these challenging cases.

TECHNICAL KEY POINT

There are two technically challenging steps. The first is initially developing the plane between the UGS and urorectal musculature. This step is actually more difficult with TUM/PUM than it was previously when we opened the entire posterior aspect of the sinus. Getting into the wrong plane posteriorly can injure the rectum, while entering the spongiosal tissue around the sinus might result in significant bleeding. Once the correct plane is identified the dissection proceeds easily.

Fortunately, pull-through vaginoplasties are only occasionally required, as the most challenging step is separating the vagina from the urinary tract. The lack of tissue planes necessitates meticulous dissection. This procedure is fraught with potential for injury to innervation and continence. Again, we think that having the patient prone allows much better visualization for this portion of the vaginoplasty.

JUDGEMENT

There are also two key aspects necessitating good clinical judgement. The first is deciding on whether PUM is adequate or if further mobilization by TUM is required. In our hands we have found nearly all patients with CAH can be managed by PUM with a flap vaginoplasty. There is no clear way to define the need for TUM but if it appears that the vagina will not easily reach the perineum or will require significant skin flaps to reach the perineum, then we would proceed to TUM.

A similarly difficult decision is required for the very high vagina. The surgeon must decide whether separation of the vagina is required. If severe female hypospadias will result from 'flap' vaginoplasty we would recommend a 'pull-through' procedure. Notably, we think that there is a greater risk of vaginal stenosis with a pull-through vaginoplasty and the greatest risk of injury to innervation is in those having both a TUM and a pull-through procedure.

'HIDDEN SECRETS'

The most helpful aspect in our opinion is total body preparation, allowing easy access to abdomen and perineum, and pacing the patient prone. We have had no difficulty rotating the child within the aperture in the drapes to the prone position. We would suggest prone positioning if a pull-through vaginoplasty is necessary.

PITFALLS

The most significant preventable pitfall is inadequate opening of the narrowed distal vagina, which results in apparent stenosis. It must be opened posteriorly in the midline until the normal-calibre vagina is located. We believe it is extremely rare to ever require opening of the rectum to achieve adequate exposure. If one has difficulty identifying the rectum as the posterior dissection is done, do not hesitate to place a finger in the rectum, which allows easy identification of its wall. We have had no problems with infection with this manoeuvre.

ACKNOWLEDGEMENTS

The illustrations for this chapter are based on material provided by the authors' artist Sharon Teal.

Illustrations by Stephan Spitzer, www.spitzer-illustration.com

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Abbreviations: **UGS**, urogenital sinus; **(P)TUM**, (partial) total urogenital sinus mobilisation; **CAH**, congenital adrenal hyperplasia.