

Surgical Treatment of High Urogenital Sinuses Using the Anterior Sagittal Transrectal Approach: A Useful Strategy to Optimize Exposure and Outcomes

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Abbreviations and Acronyms

ASTRA = anterior sagittal transrectal approach

CAH = congenital adrenal hyperplasia

CC = common channel

CIC = clean intermittent catheterization

FG = feminizing genitoplasty

UGS = urogenital sinus

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Purpose: Surgical management of the high urogenital sinus remains challenging. The anterior sagittal transrectal approach provides optimal exposure, facilitates vaginal dissection and separation from the urethra, and allows reconstruction of the bladder neck musculature. In this study we report our initial experience with this technique.

Materials and Methods: We performed a retrospective review of a 6-year multi-institutional experience treating patients with a urogenital sinus anomaly using the anterior sagittal transrectal approach without preoperative colostomy or prolonged postoperative fasting. Variables analyzed included patient age, associated malformations, the need for additional procedures and surgical outcomes.

Results: A total of 23 children with a mean age of 2.3 years (range 3 months to 17 years) who underwent surgery between 2003 and 2010 were included in the study. Mean followup was 3.4 years (range 14 months to 7 years). All children had a high urogenital sinus with (16) or without (7) congenital adrenal hyperplasia. There were 3 isolated cases treated with additional procedures. Only 1 anterior sagittal transrectal approach related complication was encountered when a perineal infection developed in a child and required temporary diverting colostomy without compromising the repair. There were no postoperative urethrovaginal fistulas. All toilet trained patients were continent for feces and most were voiding normally per urethra (21), except for 2 with associated urological malformations. There were 15 patients who underwent followup examination under anesthesia, and demonstrated separate urethral and vaginal openings.

Conclusions: The anterior sagittal transrectal approach provides excellent exposure for the management of a high urogenital sinus, facilitating the separation of urogenital structures. Good outcomes in terms of urinary/fecal continence as well as the absence of urethrovaginal fistulas were achieved in the majority of cases, supporting its consideration for the surgical management of this congenital abnormality.

Key Words: urogenital abnormalities, patient positioning, child

UROGENITAL sinus persistence can occur as an isolated abnormality or secondary to in utero virilization. Defects are usually classified according to the

length of the CC or, preferentially, based on urethral length and relationship of the confluence with the bladder neck. Surgical management is



Figure 1. Patient is placed in prone position, with legs separated allowing surgeon to operate under centralized vision of field.

then tailored according to the anatomical characteristics of the individual case as well as the presence of coexisting genitourinary anomalies. Low UGS can be successfully treated with a simple flap vaginoplasty. Intermediate UGS, characterized by a higher confluence of the urethra and vagina, but with acceptable urethral length (greater than 1.5 cm), is suitable for en bloc or total mobilization without jeopardizing urinary continence.^{1,2} In contrast, the application of this technique for high UGS (1.5 cm or less) is controversial. As an alternative some favor separating the vagina from the UGS confluence and pulling it through to the perineum.³ Although appealing, the exposure of the urethrovaginal junction in such cases can be difficult and associated with significant complications (including strictures, diverticula, stenosis, incontinence and urethrovesical fistulas).^{3,4} In addition, the required aggressive mobilization can be associated with inadvertent injury to adjacent structures (such as the bladder neck, sphincter complex and ureters). Due to these technical difficulties Peña et al proposed an alternative approach based on posterior sagittal access for the management of persistent cloaca, with exposure obtained by midline sagittal division of posterior and anterior anorectal walls along with the corresponding sphincteric musculature.⁵

Adapting the proposal of Peña et al and considering the theoretical risks involved with complete division of the anorectal sphincter, Dòmini et al introduced a modification that limits the rectal incision to its anterior wall (better known by the acronym ASTRA, the anterior sagittal transrectal

approach).⁶ This technique has drawn increasing interest as it preserves the excellent exposure of the patient in the prone position while decreasing trauma to the rectal wall and musculature. There is a paucity of literature reporting outcomes on the use of ASTRA in the treatment of children with a high UGS. It is our hypothesis that this novel technique is associated with favorable results. Thus, we report our initial multi-institutional experience with ASTRA for this patient population, focusing on postoperative complications and outcomes while sharing key technical details learned thus far.

MATERIALS AND METHODS

The experience of 4 surgeons managing primary cases of high UGS during a 7-year period (between 2003 and 2010) was retrospectively reviewed. All children with a high UGS approached through ASTRA at the participating institutions with minimal followup of 14 months were included in analysis. A high UGS was strictly defined as the presence of a urethral segment proximal to the vaginal confluence measuring less than 1.5 cm (with common channels measuring more than 3 cm and 2 cm or greater for patients with CAH and an isolated urogenital sinus, respectively). Patients with cloacal malformations were not included in the study. Variables captured included age, underlying diagnosis, the presence of associated malformations, endoscopic findings, performance of additional simultaneous procedures, perioperative and postoperative complications, and surgical outcomes.

Preoperative full bowel preparation and prophylactic antibiotics were uniformly prescribed. None of the patients had protective colostomy. As a first step, diagnostic cystoscopy and vaginoscopy were conducted, with close attention paid to confluence location, proximal urethral extension, CC length, vaginal length and elasticity. A

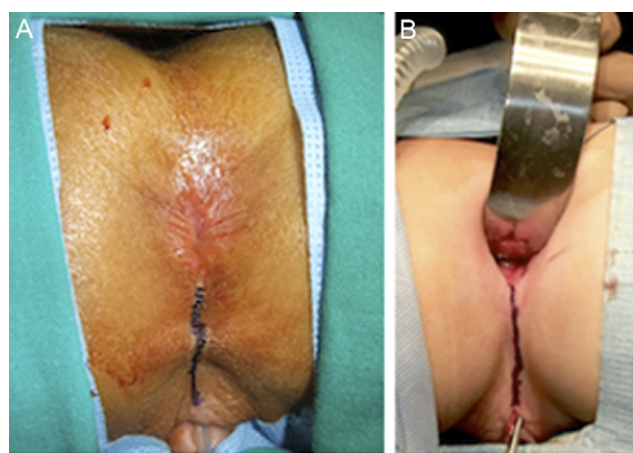


Figure 2. A, delineation of ASTRA incision. B, retraction of posterior rectal wall with Deaver retractor to maximize exposure of pelvic-perineal structures.

balloon catheter was placed in the vagina to assist with its identification during transrectal exploration. Patients were subsequently positioned prone with gentle lower extremity abduction in older children to allow the surgeon access by being positioned between the legs, facing the child's perineum (fig. 1).

A midline perineal incision is used, spanning the anterior anal border and the perineal opening of the UGS. The anterior rectal wall is incised with electrocautery while the posterior wall is posteriorly retracted with a Deaver retractor (fig. 2). The incision can be subsequently tailored by extending it toward the area

where the vaginal balloon is felt by palpation (fig. 3). The posterior vaginal wall is then exposed and incised as close as possible to the junction with the urethra. Next the vagina is mobilized posteriorly and laterally, and then dissected from the urethra. Injection of a 1:100,000 epinephrine solution in the urethrovaginal wall aids with hemostasis and helps delineate the discrete plane between these structures (fig. 4). After this is achieved, the catheter is removed from the vagina and advanced into the bladder, allowing gentle traction of the bladder neck inferiorly, a maneuver that further facilitates dissection. Once separation is complete, the

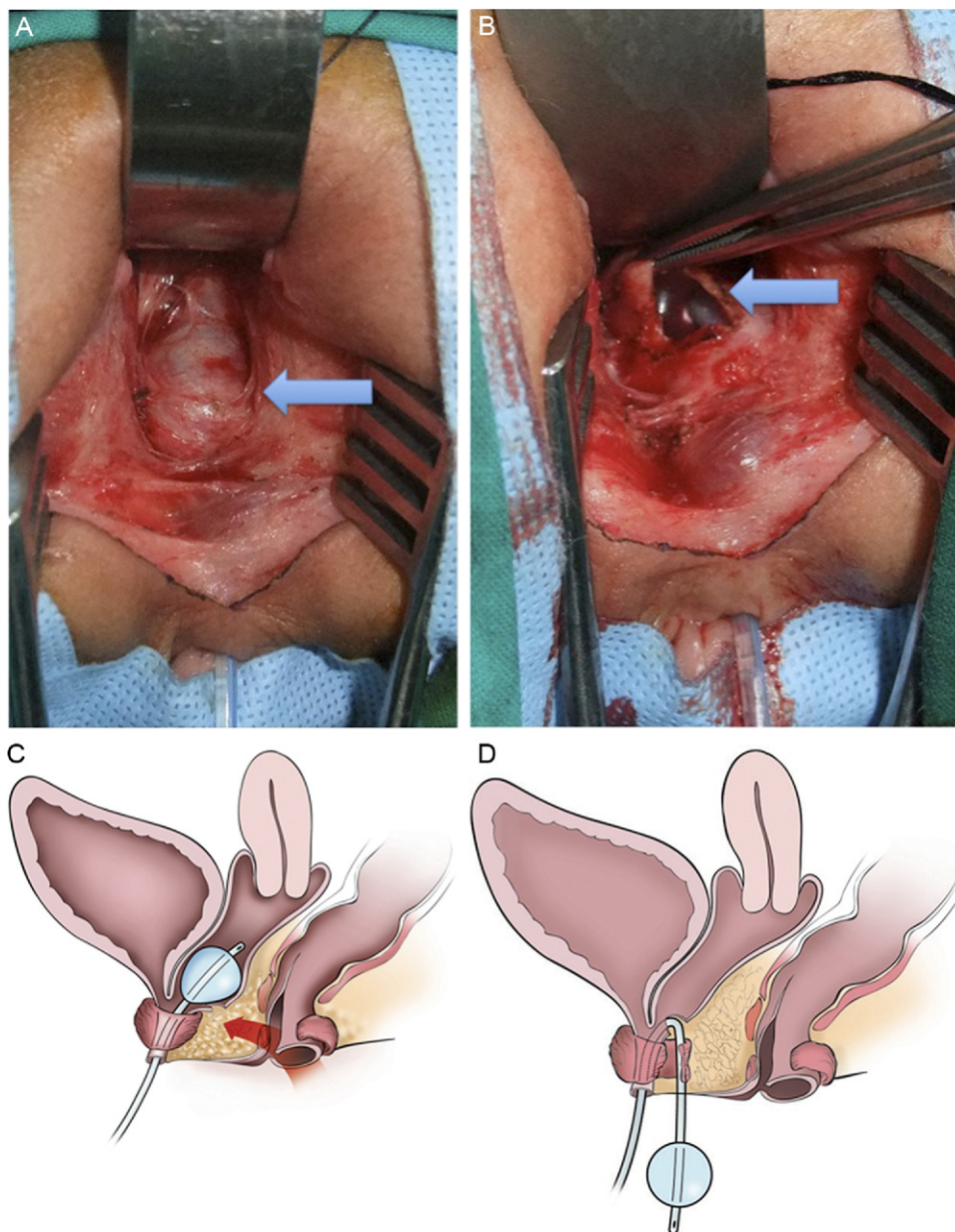


Figure 3. A, anterior rectal wall and perineal body have been incised, and posterior wall of vagina (with balloon catheter previously inserted) is inspected and palpated. Arrow points to vagina. B, posterior wall of vagina has been opened and catheter exteriorized. Arrow points to vagina. C, diagram of ASTRA incision with indwelling vaginal catheter, with indication of transverse incision to be performed in most distal aspect of vagina inserting in UGS. D, diagram of incision of posterior vaginal wall and exteriorization of catheter. Muscular complex around UGS and bladder neck have been incised to allow exposure of vagina.

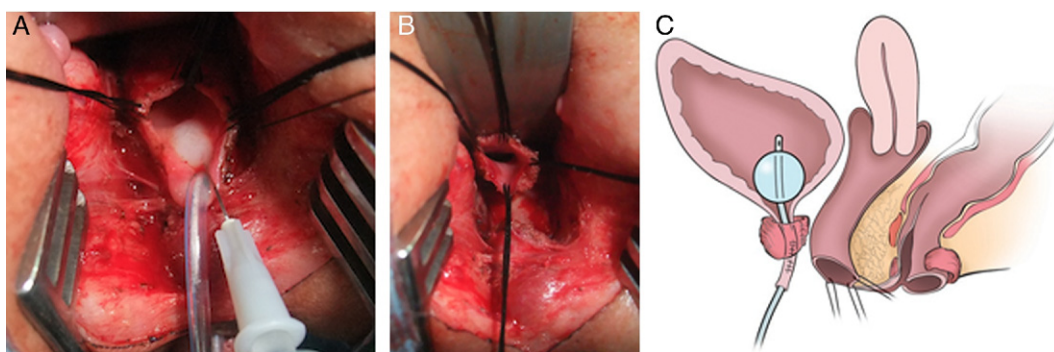


Figure 4. A, anterior vaginal wall is injected with 1:100,000 epinephrine solution to minimize bleeding. B, vaginal dissection and separation from urethra and bladder neck. C, diagram of vaginal pull-through and reconstruction of muscle complex around bladder neck.

urethral opening is closed with interrupted sutures and the bladder neck reinforced posteriorly in the midline, reconstructing the horseshoe-shaped musculature defect left in the dorsal aspect (fig. 5). This step is believed to be important to avoid postoperative incontinence. In infants there is often no need for perineal skin flaps to reach the perineum if adequate perivaginal dissection is performed and if the vagina measures more than 3 cm. In older children with short vaginas or in those with significant vaginal tethering, perineal skin flaps (suturing 2 halves of an omega-shaped segment) may be necessary to obtain a tension-free anastomosis. The anorectal wall is then closed in 2 layers. The anorectal sphincter and perineal body are subsequently reconstructed. The patient is then turned supine in the lithotomy position if FG is deemed necessary (fig. 6).

None of the patients were subjected to prolonged postoperative fasting and were fed as soon as they re-

covered from the anesthetic. An indwelling urethral catheter was kept in place for approximately 7 days. Routine vaginal dilations were not routinely performed. After parental agreement 15 patients underwent cystoscopy, vaginoscopy and vaginal calibration 3 to 6 months postoperatively.

RESULTS

A total of 23 children with a mean age of 2.3 years (range 3 months to 17 years) were identified. Only 2 girls were of postpubertal age at the time of the procedure. High UGS was associated with CAH in 16 girls and the other 7 presented with isolated UGS without virilization. All but one of the CAH associated cases had simultaneous FG. The exception was a 17-year-old adolescent who had undergone prior FG without vaginoplasty during infancy. Followup ranged between 14 months and 7 years (mean 3.4 years).

Only children diagnosed with isolated UGS had associated genitourinary malformations. These included 1 patient with bilateral ectopic ureters draining into the UGS confluence, 1 with a solitary kidney and an ectopic ureterocele also draining into the sinus confluence, and 4 with hydrocolpos. These cases were previously treated with vesicostomy, endoscopic incision of the whole urethra misdiagnosed as a vaginal septum, percutaneous vaginostomy and open vaginostomy. In 1 child with prior open cutaneous vaginostomy, transabdominal mobilization was required to release all perivaginal adhesions. Three patients underwent concurrent urological procedures, including appendicovesicostomy in 2 with preoperative bladder dysfunction (isolated UGS), ureteral reimplantation in 2 with ectopic ureters/ureterocele, and complete bladder neck repair in 2 performed with the patients in the prone position using the ASTRA (in 1 with bilateral ectopic ureters and in the other with ectopic ureterocele at the level of the UGS).



Figure 5. Muscle complex is shown just before being approximated to wrap around bladder neck.

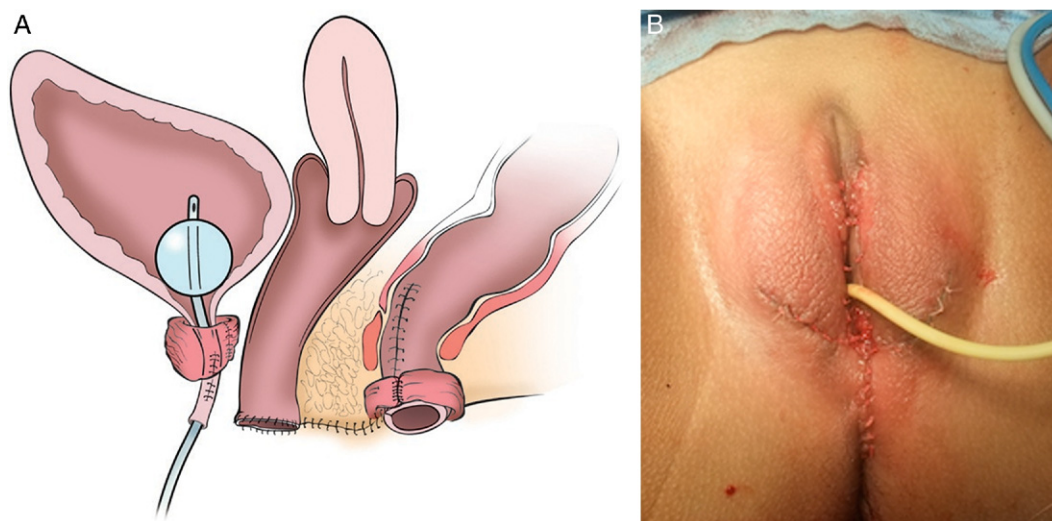


Figure 6. A, illustration of final aspect of reconstruction. B, final aspect of genitalia after completing FG.

There was only 1 postoperative complication, and it occurred in a 5-year-old child with CAH who presented with an early perineal infection. The patient was treated with a diverting colostomy which was closed after 2 months. This complication did not compromise the UGS repair.

At last followup none of the children presented with evidence of urethrovaginal fistulas, toilet trained children remained continent for feces and 21 patients were voiding voluntarily with good urinary control. The 2 girls with preoperative bladder dysfunction and associated urological malformations were conducting CIC through Mitrofanoff channels. However, only 1 remains dependent on catheterization while the other has had progressive improvement of bladder emptying, using the appendicovesicostomy conduit only for measurement of post-void residuals. The child with isolated high UGS and bilateral ectopic ureters remains incontinent, and is currently awaiting further treatment.

In all patients the vagina could be comfortably mobilized to the point that there was no need for the use of a common channel flap to bridge the anterior wall. Nevertheless, we did use this tissue for the introitoplasty. Since the posterior wall of the vagina was systematically spatulated to avoid stenosis, in 5 cases we fashioned a Fortunoff-type flap joining 2 hemi-flaps in an omega configuration and anastomosing to the vertex of the posterior vaginal wall incision.

Fifteen children underwent postoperative vaginotomy and cystoscopy under anesthesia 3 to 9 months after surgery (mean 5). None of these cases had endoscopic evidence of vaginal stricture, stenosis/contracture or urogenital fistulas. In all cases the

vagina and urethral tissues appeared supple and elastic. On postoperative physical examination a satisfactory cosmetic and anatomical result was reported in all girls, with separate orthotopic vaginal and urethral orifices. Importantly, to date, there has been no evidence of vaginal retraction, hypospadias or significant introital stenosis.

DISCUSSION

Our series provides evidence supporting the successful surgical management of high UGS using the ASTRA with few postoperative complications. Most patients underwent surgery during infancy and had concurrent FG, reflecting our treatment philosophy and respecting parental preferences. Despite the young patient age, reconstruction was straightforward and relatively easy. Although only 2 cases were postpubertal, in our limited experience with this age group dissection was subjectively more tedious due to bleeding and difficulty defining the correct planes of dissection. Therefore, despite recognizing all the controversies surrounding this issue,^{7,8} we tend to favor performing UGS correction with vaginoplasty in infancy, particularly in cases that are being considered for early FG. We recognize that the vagina can be thin and friable in some cases, which is why we have been applying preoperative local estrogen cream in an attempt to achieve a more robust vaginal structure.

Careful endoscopic assessment of vaginal dimensions and urethral length should be done preoperatively. We concur with others that the most crucial decision making factors are urethral length and the characteristics of the vagina, although some prefer to measure the length of the CC. In this regard the

classification proposed by Rink et al is useful as it includes all essential elements for adequate decision making.⁹ In particular, the length of the urethra helps in the decision of whether UGS mobilization or ASTRA with vaginal pull-through is necessary. In our opinion a high UGS (with urethral length less than 1.5 cm) should be managed with vaginal pull-through because in such cases UGS mobilization may theoretically place the child at future risk for urinary incontinence by bringing the bladder neck too low.^{10,11} An important caveat is that a hypoplastic vagina (ie less than 3 cm in length) may not reach the perineum even after extensive mobilization. These children are at higher risk for vaginal devascularization and ischemia or for requiring interposition with a segment of intestine to reach the desired anatomical location of the introitus. In such cases we support postponing the procedure until puberty as local care (ie dilations and irrigation) is often necessary and difficult to impose in childhood. In addition, this strategy may have the advantage of allowing some vaginal growth to occur after hormonal stimulation, menarche and accumulation of menstrual fluid. However, this approach demands close observation because some girls may experience complications related to müllerian obstruction and retrograde menstruation such as endometriosis.¹²

In our collective experience fecal continence has been universally preserved, which was also reported in other ASTRA series to date.⁶ Following experience with other colorectal procedures in pediatric surgery we purposefully avoided the creation of temporary diverting colostomies. Although most patients in this series fared well under this protocol, 1 case of perineal infection at the beginning of our experience stands out. To be safe we elected to create a temporary fecal diversion while providing wound care and administering adequate antibiotic coverage.

The transrectal approach allows excellent exposure of the proximal urethra and bladder neck, ensuring safe dissection and optimal separation of the vagina from adjacent structures. The balloon catheter kept under light traction facilitates transoperative identification of the vagina proximal to the confluence. In some cases the catheter can be easily oriented to the vaginal lumen. In others, particularly when the confluence is narrow, placement of an open-ended Foley over a guidewire permits accurate advancement. Injection of epinephrine solution in the common urethrovaginal wall facilitates dissection, allowing delineation of the plane while minimizing bleeding. The resultant mobilization often spares the need for cutaneous flaps to bring the vagina down to the perineum in a tension-free fashion.

Summary of published cases of high UGS treated with ASTRA

References	Total No. Cases	Age Range	Reason for Treatment	Followup	Preop Colostomy	Accessory Surgery	Postop Continence	Results	Postop Dilation
Dòmini et al ⁶	10	1–11 Yrs	CAH 10, reoperations 3	2–4 Yrs	10/10	FG 7/10	Normal, 8/10 toilet trained	Good 10/10	No
Di Benedetto ¹⁷	2	5–2 Yrs	Denys-Drash syndrome (female sex or rearing) 1, CAH 1	9–10 Mos	2/2	FG 2/2	Normal 2/2, (voiding cystourethrography, urodynamics, anorectomanometry)	Good 2/2	No
Rink et al ^{13*}	8	6 Mos–25 Yrs	Isolated high UGS 4, CAH 3, UGS + genital transposition 1	6 Mos–5 Yrs	None	Abdominal transtrigonal vaginal mobilization 1, FG 4	No citation	Good 8/8, cystoscopy 6/8, asymptomatic urethrovaginal fistula 1, vaginal stenosis 1	4/8
Arena et al ¹⁸	3	6, 8 & 6 Mos	Isolated high UGS	Mean 8 Mos	3/3	None	Normal 3/3 (urodynamics, anorectomanometry)	Good 3/3	No
Present series	23	3 Mos–17 Yrs	CAH 16, isolated high UGS 7	14 Mos–7 Yrs	None	FG 11/12, Mitrofanoff 2, reimplants 2, bladder neck repairs 2	100% Toilet trained pts, CIC 2/19	Good 23/23 perineal infection without compromising repair 1, examination under anesthesia with cystoscopy 11/19	No
Overall	46	3 Mos–25 Yrs	CAH 26, isolated high UGS 14, other disorders of sex development 2	6 Mos–7 Yrs	15/42	FG 24/42, transabdominal vaginal mobilization 1, urological bladder procedures 6	Most preserved	Good 42/42 complications 3/42 (urethrovaginal fistula 1,* vaginal stenosis 1,* perineal infection 1)	4/46

* Perineal sagittal approach without opening the rectum.

We believe the posterior bladder neck musculature defect (generated by dissection of the vagina from the urethra and bladder neck) should be reconstructed. This is not easily achieved with the patient in the supine position¹³ but is facilitated if exposure is obtained by ASTRA. Our positive initial results for urinary continence may be related to this careful reconstruction. However, we remain cautious due to the potential impact of aggressive vaginal mobilization necessary in some cases of high UGS. This dissection, which commonly advances behind the bladder, can theoretically injure perivesical innervation with resulting neurogenic incontinence. Although thus far we have not evidenced unexpected urinary incontinence in our cases, 1 patient with isolated UGS had a persistently atonic bladder and retention following ASTRA, requiring CIC through a Mitrofanoff channel. In this particular case detrusor dysfunction was detected preoperatively, which led to the concurrent creation of an appendicovesicostomy in a preemptive fashion. Nevertheless, it is plausible that the bladder dysfunction was worsened by surgical manipulation.

Peña et al proposed the posterior sagittal anorectal approach, with complete anterior and posterior transection of the anorectal sphincteric muscles, for the surgical treatment of high UGS.⁵ Whereas good exposure is achieved, it requires effectively bivalving the rectum. ASTRA has emerged as an alternative, preserving the excellent exposure provided by the posterior sagittal anorectal approach while minimizing injury to the anorectal sphincter complex, with an increasing body of evidence supporting its favorable safety and outcome profile (see table).^{14–19} Although these studies are all retrospective in design, and characterized by short followup and a limited number of patients, they confirm the rare development of complications, satisfactory exposure, and good cosmetic and functional results with ASTRA.^{17,18} Our results are in agreement with the literature.

Interesting variations of the ASTRA which avoid entering the rectum have been proposed, including a prone transperineal approach (by Rink et al¹⁹) and a pararectal approach.^{14,15,19} Both techniques are conducted through an incision limited to the perineal body, preserving the intact anorectal sphincters and the anterior anorectal wall. Reportedly good visualization is achieved while avoiding the potential risks incurred by anorectal disruption. Although appealing, there is a paucity of data supporting these benefits. Anecdotally we attempted these approaches in 2 patients, and had to convert to ASTRA in both to gain adequate exposure. This issue merits further exploration and we accept that either approach can be attempted initially, especially in infants (in whom tissue elasticity is excellent and planes are easier to dissect), followed by ASTRA if better visualization is needed.

There are important limitations that must be acknowledged, particularly those incurred by a retrospective multi-institutional review. This is a highly selected patient population with limited followup. Indeed, larger series with longer monitoring and a comparison group would be of value in better defining the role of ASTRA. Nevertheless, our results provide further evidence supporting this approach as a safe surgical technique that provides excellent exposure of the pelvic and perineal structures in female patients.

CONCLUSIONS

Our experience with ASTRA provided excellent exposure in the surgical management of cases of high UGS. These favorable results, along with adequate urethrovaginal separation and the absence of fistula formation, suggest this technique should be considered for this patient population. Larger series are needed to support these findings, and better define the role of ASTRA and alternative approaches for this condition.

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