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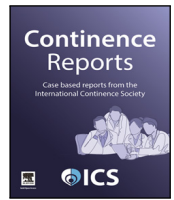
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Successful management of high urogenital sinus in an adult female: Case report

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ABSTRACT

This case report presents the successful management of high urogenital sinus (UGS) in a 21-year-old female using a modified Anterior Sagittal Transrectal Approach (ASTRA). The patient, with a history of painful cyclic hematuria and delayed menarche, was initially misdiagnosed with a transverse vaginal septum. Diagnostic clarity was achieved through cystourethroscopy, revealing a single external meatal opening indicative of UGS. The modified ASTRA technique, including a rectum-sparing approach, was employed to correct the anomaly. Preoperative administration of leuprolide acetate helped reduce the risk of postoperative infections and complications. Postoperative care included serial vaginal dilations to prevent stenosis, resulting in no complications or need for further surgical interventions at 12-month follow-up. This case highlights the importance of a multidisciplinary approach and the potential of the modified ASTRA technique in managing high UGS in adult patients. Further research to validate these findings in a larger cohort could be difficult because of rarity to miss the diagnosis in adult females.

Case Presentation

A 21-year-old female patient with BMI of 34.8 presented to our institution with a history of painful cyclic red urine and delayed menarche. Initial onset of symptoms occurred at the age of 12 years, characterized by cyclic hematuria, prompting the patient to seek medical attention. Despite multiple consultations, a conclusive diagnosis remained elusive, resulting in a misdiagnosis of a Transverse Vaginal Septum.

The patient's past medical history revealed the incidental discovery of bilateral hydronephrosis in 2005. Subsequently, she underwent bilateral ureteric Cohen's ureteroneocystostomy, along with additional surgical procedures including left Boari's bladder flap, Psoas Hitch, right ureteric (JJ) stent insertion, and repair of incisional hernia.

Further diagnostic interventions included a diagnostic cystoscopy in 2021, followed by an attempted percutaneous, perineal opening of a transverse septum in 2022, followed by spontaneous closure.

Physical examination disclosed unremarkable general and systemic findings, while local examination revealed external female genitalia

with a single opening of the common channel at the site of the external meatus (Fig. 1).

Diagnostic investigations involved cystourethroscopy with Foley's catheter insertion into the vagina to identify the vaginal opening. Surgical intervention employed a Modified Anterior Sagittal Transrectal Approach (ASTRA) technique, sparing the rectum. The stepwise approach involved identifying the vaginal opening to the urethra confluence point, Figure, locating the fistulous tract, closing the fistula, mobilizing the vaginal canal, sparing the rectum, and using skin flaps to lengthen it. Prior to surgery, the patient received a dose of 11.25 mg of leuprolide acetate to suppress GnRH and prevent menses for 3 months, reducing the risk of infection and complications.

Postoperatively, the patient exhibited no surgical site infections or incontinence. Nevertheless, serial vaginal dilatation was required to counter the risk of vaginal stenosis. It began 21 days post-operation and gradually increased the diameter. After 1, 6, and 12 months of follow-up the patient exhibited no vaginal stenosis with up to 76 Fr (25.33 mm) dilator as shown in Fig. 2. She did not require any additional intervention.

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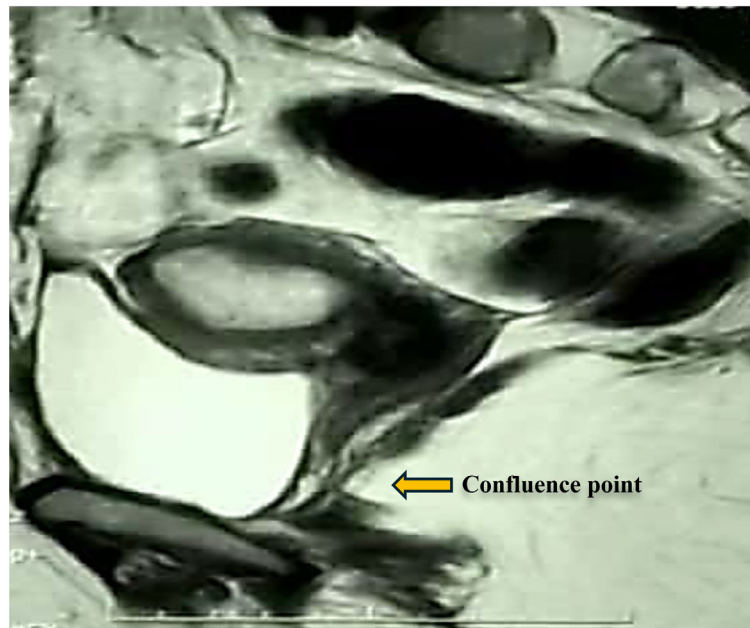
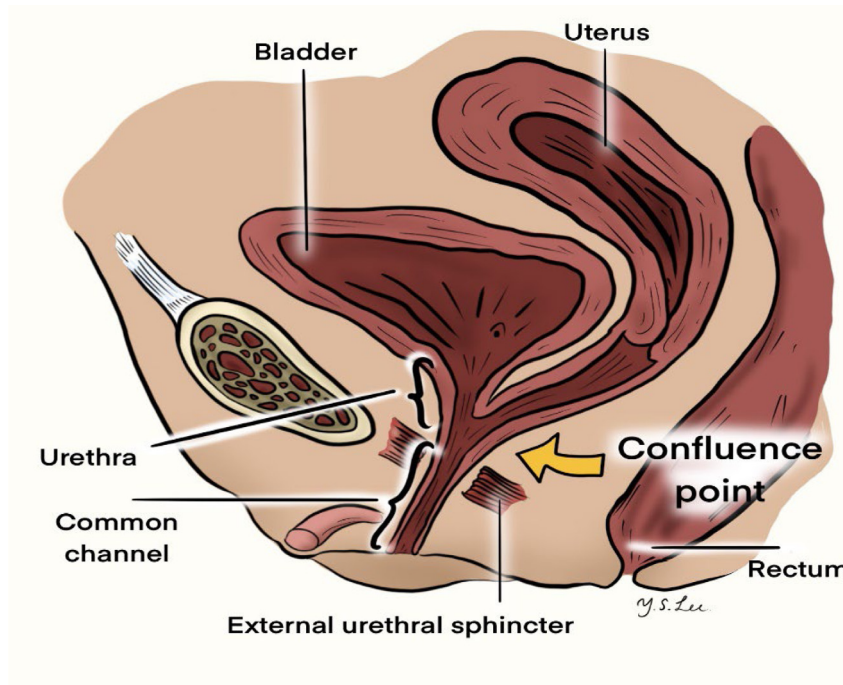


Fig. 1. Showing the confluence point and how high the urogenital sinus in the presented case was.

1. Introduction

Urogenital sinus malformation (UGS) is a rare condition in which the vaginal canal fails to open into a separate vulva and joins the urethra into a common canal. The confluence could be close to the bladder neck (high UGS) or close to the external meatus/sinus ostium (low UGS) [1]. The management of UGS is not well-established, and there are no standardized guidelines on when to perform surgery and how to follow up with patients over the long term.

High UGS in adult females is a further rare congenital anomaly impacting genitourinary function and quality of life. The embryological complexities leading to high UGS involve disruptions in urogenital differentiation, particularly during the critical stages of urogenital sinus development. Adult females rarely experience high UGS due to

the intricate nature of normal urogenital development [2,3]. Surgical intervention is the current approach and usually performed early in life.

Despite advancements, knowledge gaps persist in understanding the etiology, optimal surgical approaches, and long-term outcomes of high UGS. Collaborative studies involving larger patient cohorts, standardized outcome measures, and multicenter collaborations are crucial for this patient population. Exploring genetic factors, refining surgical techniques, and investigating psychosocial aspects will further enhance our understanding and management of high UGS.

2. Discussion

UGS presents a spectrum of clinical challenges. These congenital conditions involve the confluence of the urinary and genital tracts into a common channel, often leading to significant diagnostic and

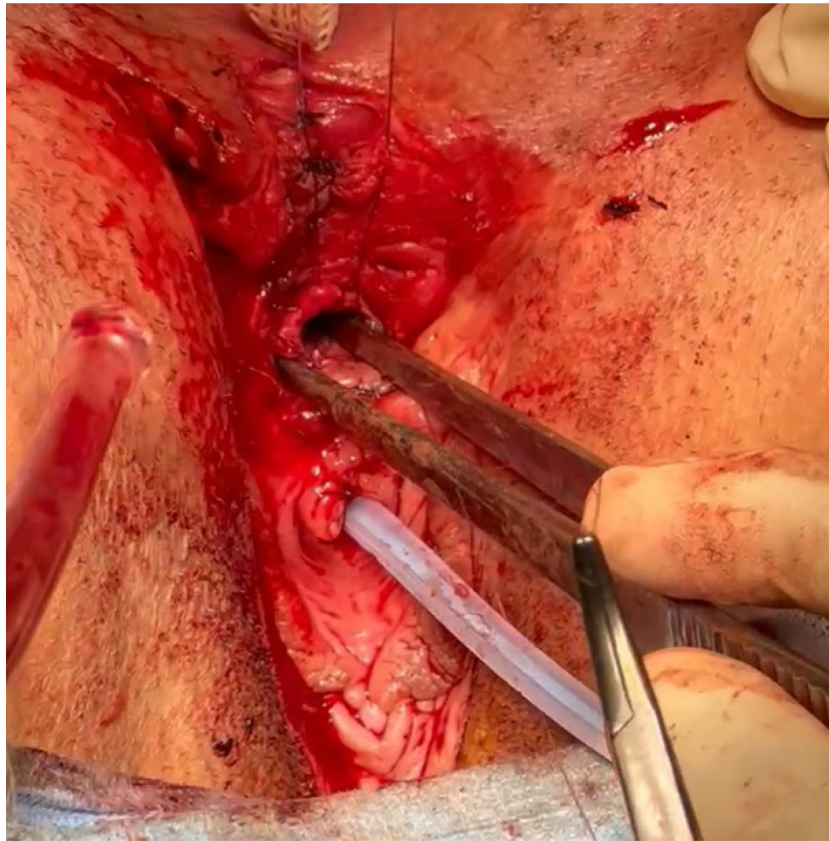


Fig. 2. Case Surgical Details and Follow-up The distal vaginal stump is freed and pulled down to meet the skin flaps without tension.

Surgical Steps:

Step 1: Cystourethroscopy with F10 Foley's catheter insertion into the vagina at the confluence orifice.

Step 2 (Sparing the rectum)

The patient's position is then switched to prone knee-chest. (Jack-knife) position with abduction of the lower extremities. An anal pack soaked in betadine is inserted into the rectum. Midline sagittal incision is made 1cm below the anterior margin of the anus to 1cm above the common channel opening. The posterior vaginal wall is dissected from the anterior rectal wall while sparing the ano-rectal sphincter and anterior rectal wall. Dissection is carried out up till the cervix.

Step 3 (most difficult step)

The anterior vaginal wall is separated from the posterior urethral wall and bladder neck/trigone area. The fistulous track is identified, and the F10 Foley's catheter is replaced with a feeding tube which is passed antegrade from the fistulous track through the common channel to the outside. Total mobilization (TM) of the vaginal stump downwards is performed without tension.

Step 4 (Fistula Closure)

The fistulous track is identified, then both, posterior urethral wall and anterior vaginal wall are closed separately in two overlapping layers each using 3-0 PDS.

Step 5 (Flaps to elongate the vaginal canal)

Two small flaps from the common channel opening skin are stitched to the spatulated vaginal wall anteriorly. Two perineal skin flaps are raised and anastomosed to the posterior vaginal wall.

Post-operative care:

Vaginal pack removed in 48 hours Received LHRH agonist to temporarily stop menses. Serial vaginal dilatations (as outlined).

therapeutic dilemmas. Our case of a 21-year-old female with a BMI of 34.8 who experienced cyclic hematuria and delayed menarche from the age of 12 exemplifies the complexities in diagnosing and managing such anomalies.

The patient's history of cyclic red urine and delayed menarche initially suggested a gynecological etiology. However, despite multiple consultations and misdiagnoses, a definitive diagnosis remained elusive. Diagnostic clarity was eventually achieved through cystourethroscopy, revealing a single external meatal opening and confirming the presence of a urogenital sinus. The Modified ASTRA technique was employed for surgical correction, emphasizing the importance of a rectum-sparing approach to minimize complications. Preoperative administration of leuprolide acetate to suppress menses helped reduce the risk of postoperative infections and complications.

2.1. Clinical features and diagnosis

The general clinical presentation included cyclic hematuria, amenorrhea, and genitourinary symptoms. Diagnostic modalities such as

cystourethroscopy and magnetic resonance imaging (MRI) aid in visualizing anatomy and confirming the diagnosis. Distinguishing high UGS from persistent urogenital sinus (PUGS) and cloacal malformations is crucial for accurate diagnosis and tailored management. The review by Smith et al. emphasizes the role of diagnostic imaging in the clinical management of persistent (PUGS). For adult females, accurate imaging is essential to delineate the complex anatomy and plan the surgical approach. Advanced imaging techniques could significantly enhance preoperative planning and postoperative assessment, ensuring better outcomes.

2.2. Management: Anterior Sagittal Transrectal Approach (ASTRA)

This technique, highlighted in multiple studies, offers significant advantages in terms of exposure and dissection. The initial experience reported by Braga et al. suggests that ASTRA can optimize outcomes by providing better access to the urogenital structures, facilitating precise vaginal dissection, and allowing effective reconstruction of the bladder neck musculature. Although primarily tested in children, this approach

proved beneficial for an adult female due to the similar need for extensive exposure and careful dissection in high UGS cases [4–9].

We were mainly wary of a few aspects during the procedure: identifying the vaginal opening at the point of confluence, which is essential for accurate dissection and subsequent repair. To avoid rectal injury and its associated complications, we also aimed for rectal sparing approach. We then identified and proceeded to close the fistulous tract, preventing recurrent urinary infections and improving continence. We also mobilized the vaginal canal to ensure functional vaginal length and patency and used flaps to elongate the vaginal canal, enhancing vaginal functionality and reducing the risk of stenosis.

Further studies are however necessary to confirm its efficacy and safety in adult patients. In our case, this approach has been utilized and proven successful after 1-year follow up with no complications and no need for further vaginoplasty and hymenoplasty.

2.3. Postoperative care and outcomes

Postoperative care involves vigilant monitoring to detect and address complications promptly, ensuring optimal patient outcomes. The postoperative period was managed meticulously to prevent complications such as vaginal stenosis. The patient underwent serial vaginal dilations starting 21-days post-surgery, a strategy that proved effective in maintaining vaginal patency. At follow-ups at 1, 6, and 12 months, the patient showed no signs of vaginal stenosis, with successful dilation up to 76 Fr (25.33 mm).

Despite the initial plan for potential future interventions like vaginoplasty with buccal mucosal graft and hymenoplasty, these were rendered unnecessary due to the successful primary surgery and postoperative management.

3. Conclusion

Our peculiar case underscores the diagnostic challenges and the importance of a tailored, multidisciplinary approach to managing high UGS in an adult female. The successful outcome in this patient highlights the efficacy of the modified ASTRA technique to spare the rectum and the critical role of preoperative hormonal suppression and postoperative care in optimizing surgical results. Future management of similar cases should consider these strategies to enhance patient outcomes and quality of life. Further research to validate these findings in a larger cohort could be difficult because of rarity of the disease especially in adult females.

Ethical considerations

Ethical approval was not obtained.

Consent to participate

Obtained.

Consent for publication

Obtained.

Funding statement

None.

Declaration of competing interest

None.

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Data availability

The data that support the findings of this study are available on request from the corresponding author.

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