Urogenital sinus (UGS) and cloacal malformations are the spectra of disease affecting mainly females, resulting in an unusual confluence of the genital and urinary tract with or without the involvement of the gastrointestinal tract. Successful reconstruction of these anomalies depends on the accurate preoperative delineation of abnormal anatomy with the help of cross-sectional and other contrast studies like genitourogram along with cistourethroscopy wherever indicated. We hereby report a case of a 14-year-old female who presented with irregular cyclical hematuria and was diagnosed with persistent UGS with urethral duplication. After a thorough evaluation, the patient was successfully managed with surgical reconstruction, described in this study. Persistent UGS is a complex developmental anomaly. Complete characterization of anomaly requires a thorough evaluation such as hormonal assessment, endoscopy, cross-sectional, and radiological contrast study. Surgical reconstruction needs individualization and may need clitoroplasty, labioplasty, and urethral and vaginal mobilization. Morphological and functional outcome is satisfactory in a well-planned surgical reconstruction.

**Keywords:** Ambiguous genitalia, urethral duplication, urogenital sinus

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**INTRODUCTION**

Persistent urogenital sinus (UGS) with urethral duplication is a rare congenital anomaly of the genitourinary tract, and only a few cases have been reported in the literature. Developmental anomalies such as a defect in the development of UGS, incomplete mesodermal fusion, ischemic events in embryogenesis, or abnormal Mullerian ducts may lead to the formation of urethral duplication. There are several clinical variants of UGS and their management depends on the type and presentation. To get good results in the treatment of persistent UGS with urethral duplication, a precise clinical and radiological diagnosis must be made, and appropriate surgical techniques should be chosen. Here, we report a case of persistent UGS of low confluence vaginal type with ambiguous genitalia and inverted ‘Y’ type urethra in a female patient successfully managed by surgical reconstruction.

**CASE REPORT**

A 14-year-old female presented with cyclical hematuria for 2 years. She attained menarche at 12 years of age, and thereafter, she had complained of cyclical hematuria with irregular menstruation at an interval of 5–6 months. The patient was passing urine mainly through a common opening of UGS. She was also passing a minimal amount of urine through an opening at the tip of the enlarged clitoris occasionally. There was no history suggestive of recurrent urinary tract infection (UTI) or voiding difficulty or family history of consanguineous marriage.

She had average built with well-developed secondary sexual characters. On local examination, a clitoromegaly

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with a formed urethral opening was found at its tip. Phallus length was 3.5 cm in length and 1.5 cm in width, and the clitoral index was 5.25 cm². There was a partial fusion of the labioscrotal fold with no rugosities and a common urogenital opening. There was no palpable gonad. The anal orifice was normal in the position [Figure 1].

Investigations
Routine blood and urine investigations were within the normal limits. No musculoskeletal abnormality was found in radiological studies. Ultrasound abdomen showed orthotopically located kidneys with a bicorticate and hypoplastic uterus. The endocrine evaluation revealed normal luteinizing hormone, follicle-stimulating hormone, cortisol, testosterone, estradiol, inhibin B, and 17-OH progesterone, which ruled out congenital adrenal hyperplasia. The karyotype of the patient was 46 XX. Magnetic resonance imaging pelvis showed clitoromegaly, but anatomy at the confluence of the urogenital tract was not vivid [Figure 1]. Genitourogram showed a pooling of contrast in the vagina but was inconclusive in determining the actual anatomy. Cystourethroscopy and gonioscopy revealed urethra traversing through the clitoris was bifurcating proximally, and it’s one limb was blind, and another end was opening in the distal end of the vagina (Inverted Y), and the urethra starting from the bladder neck was opening in the distal end of the vagina.

Treatment
After preoperative parent counseling and preparation patient was planned for urogenital reconstruction. The patient underwent excision of the clitoral part of the urethra at its proximal portion with circumferential mobilization of the urethra and vagina to bring it to the perineal skin surface to fix it there. Clitoroplasty and labioplasty were done. [Figure 2]. There were no postoperative complications.

In the beginning, a suprapubic cystostomy was performed. A guidewire introduced from a suprapubic cystostomy was pulled out through the clitoral opening with the help of a pediatric cystoscope. A circumcoronal and a midline ventral incision were given on the enlarged clitoris, and the clitoris was degloved. The circumcoronal and midline incision was deepened through Buck’s fascia. Spongiosal and bilateral corporal tissue were excised starting from under the surface of the glans clitoris till the level of inferior pubic ramus leaving behind the glans along with its intact neurovascular bundle in between Buck’s fascia and tunica albuginea. Under the inferior pubic ramus, the urethra and distal end of the vagina were mobilized circumferentially for 2 cm. Excess of clitoral skin was excised and mobilized distal urethra and vagina were fixed with suture to the cutaneous tissue of labia minora.

Follow-up
At 12 months of follow-up, the appearance of external genitalia was having acceptable cosmesis, and she was continent with normal voiding.

Discussion
Persistent UGS is a rare congenital anomaly characterized by abnormal communication of the urethra and vagina with an incidence of 0.6/10000 female births. In newborn females, it is found as a single external common

![Figure 1: A 14-year-old female presented with irregular cyclical hematuria, (a and b) The appearance of the external genitalia, a note was made of significant clitoromegaly, (c and d) Magnetic resonance imaging pelvis showing clitoromegaly (arrow), (e) Postvoid film of intravenous urogram showing pooling of contrast in the vagina (arrow), (f) Genitourogram showing contrast in the vagina as bladder capping (arrow)](image1)

![Figure 2: Surgical steps of urogenital sinus reconstruction, (a) Schematic diagram urogenital sinus low confluence vaginal type with ambiguous genitalia and inverted “y” type of urethra, (b and c) Clitoroplasty and labioplasty, (d) The final appearance of external genitalia](image2)
orifice with a separate rectal opening.\[^{4}\]\ UGS anomaly may be associated with intersex, bladder or vaginal agenesis, rectovaginal anomaly, and hydrocolpos.\[^{4}\]\ There are four different types of persistent UGS anomalies depending on the location of the confluence of vagina and urethra, and they are characterized by Type I labial fusion; Type II distal confluence; Type III proximal or high confluence with a long common tract, and in Type IV vagina is absent.\[^{5}\]\ Various theories exist regarding the embryogenesis of urethral duplication.\[^{4,5}\]\ Although the most commonly accepted hypothesis of Patten and Barry states that an abnormal relationship between genital tubercle (lateral fold) and cloacal membrane (ventral end) leads to complete urethral duplication, it failed to explain the development of all subtypes of urethral duplication. Genitourinary tract (33%), gastrointestinal (13%) cardiovascular system (13%) anomalies may be associated with persistent UGS anomalies.\[^{6}\]\ Urethral duplication may be complete and incomplete and associated with duplication of bladder anomalies or rectal anomalies, and vertebral anomalies. Patients with urethral duplications are usually asymptomatic but may have varied presentations such as UTI, urinary incontinence, double stream, deformed external genitalia, serous discharge from sinus, outflow obstruction, and associated anomalies. Most patients are usually presented with ambiguous genitalia, and it requires a combined approach of clinical examination, special laboratory investigations such as hormonal evaluation with karyotyping, and imaging before surgical correction.

Delineation of abnormal anatomy of congenital urogenital malformation by endoscopy and contrast study is crucial before surgical reconstruction of this challenging condition. Surgical reconstruction depends on the anatomical location of the confluence of the urethra and vagina. In low confluence type of persistent UGS anomaly flap vaginoplasty and high confluence type UGS anomaly, pull-through vaginoplasty is done.\[^{7}\]\ Total UGS mobilization helps for a more comfortable way of reconstruction of the vagina. It helps to avoid the dissection to separate between the vagina and urethra.\[^{8}\]\ Bonney \textit{et al.} described complex urogenital malformation as complete urethral duplication, clitoral hypertrophy, a vaginal urethra with fused labia minora in a girl without adrenogenital syndrome.\[^{9}\]\ In this case report, the patient was diagnosed as persistent UGS low confluence vaginal type with clitororomegaly with urethral duplication and underwent transection and ligation of the clitoral part of the urethra with partial UGS mobilization and clitoroplasty and labioplasty.

**Conclusions**

Persistent UGS with urethral duplication either isolated or associated with another anomaly should be managed by a proper surgical approach. Total UGS mobilization is a feasible and safe technique for urogenital reconstruction with a good cosmetic outcome.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Haleblian G, Kraklau D, Wilcox D, Duffy P, Ransley P, Mushtaq I. Y-type urethral duplication in the male. BJU Int 2006;97:597-602.
2. Sánchez MM, Vellibre RM, Castelo JL, Arias MP, Sarmiento RC, Costa AR. A new case of male Y-type urethral duplication and review of literature. J Pediatr Surg 2006;41:e69-71.
3. Arena S, Scuderi MG, Sanges G, Arena F, Di Benedetto V. Urethral duplication in males: Our experience in ten cases. Pediatr Surg Int 2007;23:789-94.
4. Kitta T, Kakizaki H, Iwami D, Tanda K. Successful bladder management for a pure urogenital sinus anomaly. Int J Urol 2004;11:340-2.
5. Powell DM, Newman KD, Randolph J. A proposed classification of vaginal anomalies and their surgical correction. J Pediatr Surg 1995;30:271-5.
6. Mallmann MR, Reutter H, Mack-Delefsen B, Gottschalk I, Geipel A, Berg C, \textit{et al.} Prenatal Diagnosis of Hydro (metro) colpos: A Series of 20 Cases. Fetal Diagn Ther 2019;45:62-8.
7. Valentini AL, Giuliani M, Gui B, Laino ME, Zecchi V, Rodolfino E, \textit{et al.} Persistent urogenital sinus: Diagnostic imaging for clinical management. What does the radiologist need to know? Am J Perinatol 2016;33:425-32.
8. Bouhafs A, Halim Y, El Azzouzi D, Arifi M, Belkacem R, Barahioui M, \textit{et al.} An exceptional case of urogenital sinus associated with a scrotal pouch and duplicate urethra in a female child. J Pediatr Urol 2006;2:489-92.
9. Bonney WW, Young HH 2\textsuperscript{nd}, levin D, Goodwin WE. Complete duplication of the urethra with vaginal stenosis. J Urol 1975;113:132-7.