



## UROGENITAL SINUS WITH SINORECTAL H-FISTULA – A RARE VARIANT

## Paediatric Surgery

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## ABSTRACT

Urogenital Sinus is a rare form of congenital Anomaly in female which presents with conjoint urethra and vagina for variable length. The ano-rectum usually remains intact and complete. This case is a rare variant of UGS with H-Fistula between rectum and the sinus.

## KEYWORDS

Urogenital Sinus, H-Fistula

## INTRODUCTION

Urogenital Sinus is a rare form of congenital Anomaly in female which presents with conjoint urethra and vagina for variable length<sup>1,3</sup>. It occurs due to failure of differentiation of urinary and genital tract during embryonic development<sup>1,2,3</sup>. The ano-rectum usually remains intact and complete. This case is a rare variant of UGS with H-Fistula between rectum and the sinus. Only two such cases have been reported till date<sup>2,3,6,7</sup>.

## Case History

A 3 month old girl child was referred to our OPD with history of stool coming out from vagina and anus. 4th day after birth, mother noticed stool coming out from anus and vagina. On examination urethral opening could not be delineated separately. Anus normally placed. Routine investigations revealed no other congenital anomalies. Skin bridge loop sigmoid colostomy done at 3months age. Examination under GA f/b CYSTOGENITOSCOPY revealed a Persistent Urogenital Sinus, Common Channel of less than 3cm, normal placed anal opening, cervical opening seen, but Bladder opening could not be delineated.

MRI Pelvis revealed small uterus with endometrial stripe and normal cervix. B/L Ovaries not visualized (? Hypoplastic/Agensis). Chromosome analysis showed 46 XX. Diagnostic Laparoscopy revealed rudimentary uterus with right sided ovary suspicious of being streak Gonad and normal left ovary. Fistula not visualized.

Total Urogenital Mobilization with ligation of Sino-Rectal H-Fistula was performed. Intraoperative examination was performed and sino-rectal H-Fistula opening was delineated. Dissection of the fistula from the sinus along with urogenital mobilization was done. Point of differentiation of vagina and urethra was reached and urinary catheter was placed followed by ligation of fistula.



Figure 1: Testing data- load current (amperes) Figure 1a:

intraoperative examination, 1b: Delineation of H-fistula, 1c: Point of differentiation of vagina and urethra reached. Urethra catheterised f/b excision of H-fistula., 1d: Post operative status.

## DISCUSSION

Urogenital Sinus occurs due to failure of differentiation of urinary and genital tract during embryonic development<sup>1,2,3</sup>. Most common presentation is Persistent Urogenital Sinus (PUGS) with genital ambiguity. Other common presentations being Persistent Cloaca, Female extrophy, and PUGS without genital ambiguity. This case is a rare variant of UGS with H-Fistula between rectum and the sinus. Only two such cases have been reported till date<sup>2,3,6,7</sup>. The most common etiology of PUGS is Congenital Adrenal Hyperplasia (CAH)<sup>9</sup>. Urogenital sinus anomalies commonly have associated abnormalities of external genitalia<sup>8</sup>. Other associations are Disorders of Sexual Differentiation, Anorectal Malformation, Vertebral defects, Hydrocolpos, hydrometrocolpos. Commonly used method of classification are Powell and Prader. Immediate management of patients include fluid and electrolyte imbalance correction and ruling out salt wasting due to 21-Hydroxylase deficiency. Detailed history including family history and physical examination should be followed by investigations like UGS, MRI and contrast genitography. Cystoscopy and Diagnostic Laparoscopy helps rule out any anatomical abnormality. Other studies include Hormone stimulation test, hormone levels and gonadal biopsy. Goal of treatment is achieving voiding control and normal sexual function. Surgical treatment include reconstructive surgeries like clitoroplasty, labioplasty, vaginoplasty.

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