

## Case Report

# Congenital prepubic sinus: a series of six cases of extremely rare congenital anomaly

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### ABSTRACT

Congenital prepubic sinus (CPS) is a very rare congenital anomaly. The baby generally present with discharge from an opening situated in midline below umbilicus to root of penis in male or clitoris in female child. The etiology of this congenital anomaly is not exactly known. There is debate in etiology as one theory support anomalous anterior abdominal wall closure and another support variant of dorsal urethral duplication. Here we are presenting the six cases of CPS with their management and brief discussion on theories that have been proposed for their etiology.

**Keywords:** CPS, Dorsal duplications of urethra

### INTRODUCTION

CPS is a very rare congenital anomaly of uncertain etiology affecting both sexes.<sup>1</sup> The main complaint of tiny opening in skin overlying supra-pubic and pre-pubic skin with watery or some time purulent discharge with varying degree of skin abnormality due to local skin insult by secretions. We are reporting series of 6 cases with such anomaly and brief review of literature.

### CASE REPORT

This is a retrospective review of cases presented to our center in last 10 years, six cases of age range 2 months to 3 years, two female and rest male baby presented with discharging opening situated as shown in Table 1.

In all the cases ultrasonography of kidney and bladder and micturating cystourethrography were normal. Sinogram of all the cases showed a blind ended tract traversing towards pubic symphysis (shown in Figure 1 and 2 for case no 3 and 6). In all the cases excision of tract were done under general anesthesia.

Histopathological finding of all cases of excised tract were showed in Table 1. All the cases were doing well in follow up of 6 months-3 years. In none of the cases recurrence of sinus or any other complications occurred.

### DISCUSSION

CPS is a rare congenital anomaly of lower abdominal wall and external genitalia of unknown etiology with almost 1:1 incidence in both male and female till date reported cases.<sup>2</sup> First case of CPS was described by Campbell et al. in 1987.<sup>3</sup>

On through search of literature only around 40 cases of CPS have been found till date. Though urethral duplication is the most frequently reported anomaly, dorsal duplications of urethra is rare and classified in to 3 types by stephens;

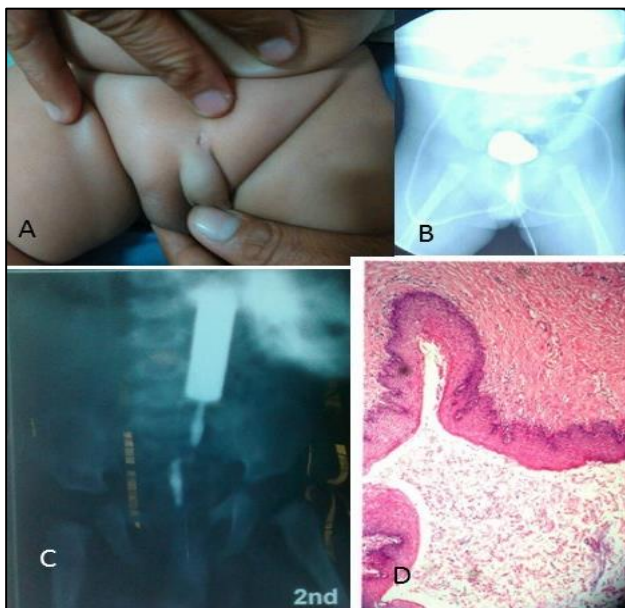
- A complete or incomplete tandem channel from the glans to the bladder
- An epispadiac type; and
- A dermoid sinus that simulates an accessory urethra, external opening of CPS may lie anywhere in the

skin overlying the symphysis pubis in the midline from lower abdominal crease to dorsal radix of

phallus and the non-communicating tract ending towards pubic symphysis.<sup>4</sup>

**Table 1: Clinical and histopathological finding of all six cases.**

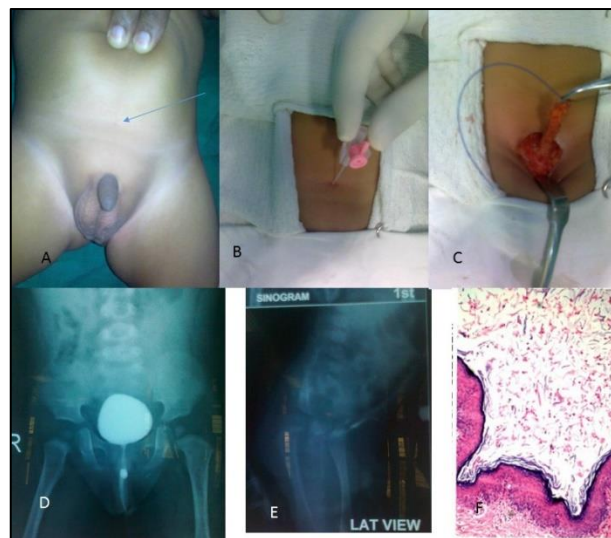
Age (months)	Sex	Site of external opening	Histopathological finding
8	Male	Prepubic	Sinus tract lined by keratinizing squamous epithelium
2	Female	Prepubic	Sinus tract lined by keratinizing squamous epithelium
5	Male	Dorsal root of penis	Distal Sinus tract lined by keratinizing squamous epithelium and proximal sinus tract lined by transitional epithelium
24	Male	Prepubic	Sinus tract lined by keratinizing squamous epithelium
16	Female	Above clitoris	Distal Sinus tract lined by keratinizing squamous epithelium and proximal sinus tract lined by transitional epithelium
36	Male	Above pubis	Sinus tract lined by keratinizing squamous epithelium



**Figure 1: Clinical, radiological, and histopathology of case no 3. A) Clinical picture showing a tiny opening at root of penis; B, C) Contrast study showing blind tract going towards pubic symphysis; D) Histopathology of excised tract showing tract lined by keratinizing squamous.**

Diagnosis is generally done by clinical finding, ultrasonography, sinogram and / or cystogram, as done in our cases. Magnetic resonance imaging may be useful to delineate precise anatomy of the sinus. The exact etiology of CPS is still debated. Embryological explanation is still moving around two theories: including an anomalous

anterior abdominal wall closure or variant of dorsal urethral duplication. This theory of localized failure of midline fusion of anterior abdominal wall was given by Campbell et al.<sup>3</sup>



**Figure-2, Clinical, radiological, intraoperative and histopathology of case no 6. A) Clinical picture showing a tiny opening in mid line above pubic symphysis showed by arrow; B, C) intraoperative picture showing tract going towards pubic symphysis; D, E) Contrast study showing blind tract going towards pubic symphysis; F) Histopathology of excised tract showing tract lined by keratinizing squamous.**

As per this theory closure of the anterior abdominal wall is complete after the cloacal membrane has moved downwards from the umbilical cord and fusion of the bilateral folds has occurred at the 4-week-old fetus. Any defect at this stage will cause anomalies, including omphalocele, bladder exstrophy and epispadias. Umbilicophalic groove normally disappears in the 9th week, persistence of the cloacal membrane above the genital tubercle will interrupt complete fusion causing various anomalies, including widening of the pubic symphysis, bifid clitoris, separation of the corpus cavernosum and epispadia glands. The presence of stratified squamous epithelium in the entire tract in four of our cases supports the theory of midline closure defect. The other theory is that CPS is a variant of dorsal urethral duplication.<sup>5</sup> The epithelium of the entire female urethra and most of the male urethra develops from the endoderm of the urogenital sinus.

However, the distal portion of the male urethra is derived from the ectodermic glandular plate, which grows backwards from the tip of glans penis to meet the urogenital sinus. The distal penile urethra is lined by stratified squamous epithelium and proximal urethra by transitional epithelium. However, the first theory does not clearly explain the presence of transitional epithelium that is seen in some cases.

In four of our cases the external opening of sinus was prepubic and above pubic, histopathologies of tracts in these cases were lined by keratinizing squamous epithelium. In rest of two cases where external opening of sinus tract was at root of penis and root of clitoris, the histopathology of sinus tract in these cases were distal sinus tract lined by keratinizing squamous epithelium and proximal sinus tract lined by transitional epithelium. Based on these finding we proposed that in cases where

external openings were prepubic and above pubic were due to localized failure of midline fusion of anterior abdominal wall, and cases where external opening were near urethra (root of penis or clitoris) were a variant of dorsal urethral duplication.

Whatever the etiology, these lesions can be easily and completely cured by a simple excision.

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