



Case Report

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Congenital Prepubic Sinus (Type 2 Stephens Variant of Epispadiac Dorsal Urethral Duplication): An Uncommon Anomaly

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Abstract

Variants of congenital prepubic sinus have been reported rarely, and because the anatomic features often differ from each other, a consensus concerning the embryology and classification was not achieved yet. Various names including congenital prepubic sinus, sub pubic fistula, prepubic dermoid sinus, and suprapubic dermoid sinus, were used to identify these lesions, and, among the classifications available, none seems to clearly describe this entity. We present a 2-year old boy with a case of epispadiac variant of urethral duplication in which the duplicated urethra presented as a prepubic sinus. We report this uncommon anomaly and review the scattered published reports to improve the global understanding of this uncommon congenital lesion.

Keywords: Urethral duplication; Epispadias; Congenital prepubic sinus; Sinus tract

Introduction

Congenital prepubic sinus (CPS) is a rare anomaly of uncertain etiology. The sinus usually presents as a small tract, commencing on the skin overlying the penis or prepubic area, and extending toward the anterior bladder wall or umbilicus [1,2]. The anatomic and pathologic features of this disorder have been documented, but controversies over its embryologic basis are ongoing.

Case Report

A 2-year-old male child was presented with congenital opening over the dorsal surface of the penis. He was asymptomatic except for occasional clear discharge from the opening. Child was passing urine through the normal urethral opening. Local examination revealed a deformed penis with ventrally hooded prepuce and 8mm midline epispadiac opening over the dorsal surface of the penis. A number 8 infant feeding tube could be easily passed in the opening for a distance of 3cm and could be felt going under the pubic symphysis (Figure 1a & 1b). Fistulography was showing blind ending tract around 2.5cm long going up towards the prevesical space without any communication with bladder (Figure 2a & 2b). At operation, the tract was dissected and was found to be going under the pubic symphysis (Figure 3a & 3b). The tract was traced up to prevesical space, transfixated, ligated and divided. Specimen (2.5cm sinus tract) was sent for histopathology

which showed tract lined by transitional epithelium and focal inflammation (Figure 4).

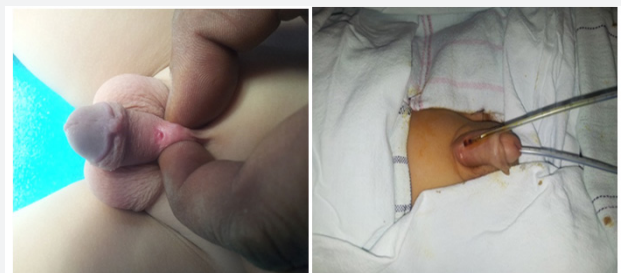


Figure 1 a & b: Sinus opening over the prepubic area and a ventrally hooded prepuce.

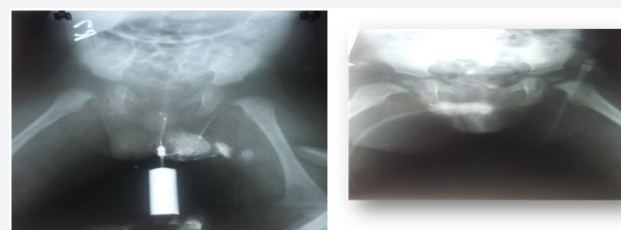


Figure 2 a & b: Fistulography showing tract ending blindly without any communication with bladder.



Figure 3 a & b: Intraoperative findings of sinus tract.



Figure 4: Histopathological specimen (2.5cm sinus tract).

Discussion

Congenital prepubic sinus is a tract originating in the skin overlying the symphysis pubis, superior to the base of the penis or clitoris, and extending to, but not communicating with, the anterior bladder wall [3]. There are four generally proposed theories for the etiology of Congenital prepubic sinus: First anomaly of abdominal wall closure [4]; and second urethral developmental anomaly, a variant of dorsal urethral duplication [1,2,5-8]. The third theory is that it is a congenital fistula of the primitive urogenital sinus, with three anatomic subtypes depending on the direction of the sinus tract: high, toward the urachal remnant; middle, toward the bladder; and low, toward the prostatic urethra [9]. Fourth theory suggests that it is a remnant of the cloaca [10]. Of all these theories, reports supporting the anomaly of dorsal urethral duplication predominate [1,3,5-8,11]. As these theories

cannot explain all the varied presentations of CPS, Tsukamoto et al. postulated recently that CPS may be caused by a residual cloaca membrane and umbilicophallic groove, and that the depth may determine the position of the end of the sinus tract [12]. However, Huang et al. [1] used an immunohistochemical staining technique of the excised sinus to reinforce the theory of dorsal urethral duplication in a report of five patients with congenital prepubic sinus, when they found the presence of transitional epithelium in the proximal part of the sinus with surrounding smooth muscle bundles [1]. Balster et al. [11] in 2003 also supported this assumption with an immunohistochemical study on the excised sinus tract of a 2-year-old boy with a skin fistula on the dorsal side of the penis [11]. Urethral duplication remains a rare and confusing problem, more so when it presents as a prepubic sinus. Urethral duplication does not represent a uniform entity making it difficult to find an unequivocal and comprehensive classification. Stephens described three types of dorsal urethral duplication according to the anatomy [13]. Type 1 is a complete or incomplete channel that runs parallel to the normal urethra from the glans to the bladder, which may join the urethra or ends blindly. Type 2 is an epispadiac type of channel from the dorsum of the penis to the bladder or one that joins the urethra at some point. Type 3 is a dermoid sinus that simulates an accessory urethra but tracks from the base of the penis in front of the pelvic urethra and bladder behind the pubic symphysis to or towards the umbilicus. We found only 7 reports of 9 cases of epispadiac variant of dorsal urethral duplication in the English literature (Table 1) (6,8,14-18). The similarity of the anatomy of our case to the type 2 variant of the Stephens classification favours the theory that Congenital prepubic sinus is a variant of dorsal urethral duplication. The presence of transitional epithelium in the lining of the sinus in this patient reinforced this theory. Although the tract ended blindly toward the anterior bladder wall, the presence of dorsal chordee, a ventrally hooded prepuce as well as penile torsion supports an epispadiac variant.

Table 1: Published cases of congenital prepubic sinus with epispadiac variant of dorsal urethral duplication.

Studies	Number of Patients/ Sex	Histopathology	Diagnosis
Campbell et al. [6]	1M, 2F	Proximal transitional; distal squamous surrounded by smooth muscle in 2; stratified squamous in 1	Variant of epispadiac duplication of the urethra (Stephens type 2)
Gonzalez et al. [14]	1M	Stratified squamous surrounded by smooth muscle	Variant of dorsal urethral duplication (intermediate between Stephens type 2 and 3)
Mostin et al. [17]	1M	Proximal transitional; distal Squamous epithelium	Epispadiac duplication of the urethra
Lopez Aramburu et al. [15]	1M	Proximal transitional; distal squamous, surrounded by smooth muscle	Variant of epispadiac duplication of the urethra (Stephens type 2)
Green et al. [8]	1F	Stratified squamous epithelium	Variant of epispadiac duplication of the urethra (Stephens type 2)
Al-Wattar et al. [16]	1M	Proximal transitional; distal Squamous epithelium, surrounded by smooth muscle	Variant of epispadiac duplication of the urethra (Stephens type 2)

Abdulrasheed A. Nasir et al. [18]	1M	Transitional epithelium	Variant of epispadiac duplication of the urethra (Stephens type2)
Current case	1M	Tract lined by transitional epithelium and focal inflammation.	Variant of epispadiac duplication of the urethra (Stephens type2)

Patient with Congenital prepubic sinus should be thoroughly evaluated because of its variable presentation, come to medical attention because of the opening or because of persistent discharge. Diagnosis is mainly clinical but imaging techniques such as micturating cystourethrogram and sinogram could help to outline the direction of the tract, and whether it is blind ending or communicating with the urinary tract. Treatment is individualized depending on the anatomy and severity of the anomaly, and usually consists of excision of the non dominant urethra or sinus tract, usually the dorsal one. Excision is usually curative.

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