

## Case Report

# Congenital urethrocutaneous fistula: Case report with review of literature

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### ABSTRACT

We are presenting two cases of congenital urethrocutaneous fistula on ventral penile shaft. Congenital urethral fistula is an extremely rare, but easily manageable anomaly that may be confused with hypospadias. Awareness of the entity will avoid complications. This condition may be associated with other anomalies like congenital hernias and anorectal malformations. Treatment of this entity is individualized according to site of fistula, associated anomalies and condition of the distal urethra. All the principles of hypospadias surgery should be strictly followed.

### KEY WORDS

Congenital; fistula; urethro cutaneous

### INTRODUCTION

In contrast to acquired urethrocutaneous fistula, congenital anterior urethrocutaneous fistula is a rare anomaly that may present in an isolated fashion or in association with hypospadias or chordee. In the literature, it has been described as unusual type of hypospadias, cryptospadias and congenital urethrocutaneous fistula.<sup>[1]</sup> Only 24 cases have been reported in the English literature.<sup>[1,2]</sup> There is considerable controversy regarding existence of this entity and some claim that it is a result of injury following circumcision.<sup>[3]</sup>

### CASE REPORTS

#### Case 1

A 3-year-old boy presented with a congenital, mid-penile

ventral opening with normal external meatus. There was no previous history of circumcision, trauma or urinary retention. On examination prepuce, glans and external urinary meatus were normal with absence of any chordee. Distal urethra was patent, with a thinned out ventral skin bridge between the fistula and frenulum. A fistula was present in the mid-penile region measuring less than 0.5 × 0.5 cm in size. Infant feeding tube of 6 fr was passed from the normal urinary meatus to bladder easily. The fistulous site was cannulated with 4.0 prolene suture [Figure 1]. Anorectum was normal. No other local or systemic abnormality was present. Patient underwent surgical correction under general anaesthesia. The whole of the urethra from fistula to the corona was laid open [Figure 2] and formal urethral closure was performed by using local tissue flaps in two layers. The glanular urethra and meatus were normal and hence left untouched [Figure 3]. After a 5-year follow-up, the patient is having normal voiding with no recurrence.

#### Case 2

A 5-year-old male child was referred to us with diagnosis of distal penile hypospadias. The child had

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an associated congenital left inguinal hernia. The parents of the child stated that he used to pass urine simultaneously from the glans tip and ventral openings since birth. Examination of local site revealed a small opening on the ventral side of the penis slightly proximal to corona along with a deep pit at the external urethral meatus. The glans and prepuce were normal with good quality Skin Bridge between the fistula and frenulum. The length of the penis was normal for age with absence of chordee. On calibrating the pit at the site of the normal meatus under sedation, the urethra distal to the fistulous opening was patent and hence the diagnosis of congenital urethrocutaneous fistula was made [Figure 4]. The child was able to pass urine with thin stream from the external meatus following calibration. Two weeks post-cannulation, there was complete re-obliteration of the distal urethra. In view of recurrent obliteration of the distal urethra, entire urethra including the glans was laid open and

Snodgrass repair was performed [Figure 5]. The patient is doing well without any complication after a follow-up of 2 years.

## DISCUSSION

The aetiology of congenital urethra-cutaneous fistula until now is not well established; perhaps it is multi-factorial because of varied appearance of congenital fistulas with or without associated hypospadias.<sup>[1,4]</sup> Campbell stated that congenital anterior urethro-cutaneous fistula represents embryonal urethral blow-out behind a distal congenital obstruction.<sup>[5]</sup> Olbourne theorized that a focal defect in the urethral plate results in arrested distal migration of the urethral plate or localized deficiency of a portion of the plate.<sup>[6]</sup> Similarly, the testosterone or androgen receptors may be at fault leading to the development of fistula. Coronal type of fistula may be explained by misalignment



Figure 1: Congenital fistula that is threaded



Figure 2: Lay opened urethra distal to the congenital fistula with normal corona, glans and external urinary meatus



Figure 3: Final look after formal closure of the fistula



Figure 4: The patent distal urethra after calibrating the external urethral meatus with repaired left inguinal hernia



Figure 5: Post-operative view of case 2

of the glanular and penile urethra.<sup>[7]</sup> Cook and Stephens suggested an alternative mechanism, namely pressure atrophy from the heel of the babies' foot, leading to the pressure necrosis.<sup>[8]</sup> These fistulas may also result from development of cysts along the mediogenital raphe at the frenulum during prenatal period.

There are two varieties of congenital anterior urethrocutaneous fistula. One is isolated fistula, which is associated with normal foreskin, no chordee or hypospadias and an intact distal urethra and spongiosum as in our cases. The other type is associated with hypospadias-like characteristics, that is, chordee, a dorsal hood with or without distal urethra or spongiosal defect. Congenital anterior urethrocutaneous fistula is uncommon as compared with posterior urethrocutaneous fistula, which usually represents Y-type duplication of the urethra with anorectal atretic malformations.<sup>[9]</sup>

Surgical approach to repair congenital anterior urethrocutaneous fistula depends upon the type of fistula. It is important to exclude urethral duplication, which produces Y type of fistula and is associated with anorectal malformations. Probing the fistula, radiographic dye study or cystourethroscopic examination may be required to corroborate the diagnosis. In case of an isolated fistula with intact spongiosum, repair with local flaps is sufficient, but if they are associated with deficient distal urethra or spongiosum, associated chordee, hypospadias

or stenosis of external meatus, then formal hypospadias repair is recommended.<sup>[10]</sup> Various techniques are proposed to repair these fistulas including primary closure via Thiersch-Duplay urethroplasty, turned-down flap urethroplasty and pedicled island tube or onlay urethroplasty.<sup>[11]</sup>

## CONCLUSION

In this article, we have highlighted a rare variant of hypospadias, which is often confused with coronal or distal penile hypospadias. Treatment of this entity is individualized according to the site of fistula and associated anomalies, as well as the condition of the distal urethra. All the principles of hypospadias surgery should be strictly followed.

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