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Case Review

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Congenital anterior urethrocutaneous fistula: Variable clinical features and their management

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Abstract

Anterior congenital urethrocutaneous fistula is a rare urethral anomaly with only a few cases and short case series being reported in the literature. The anomaly can present with variable clinical features and other associated anomalies. It is important to have a good understanding of the anomaly for an optimal outcome. We reviewed our successfully managed three cases of congenital anterior urethrocutaneous fistula with variable presentation requiring different treatment strategies.

Level of evidence: Not applicable for this multicentre audit.

Keywords

Paediatrics, Reconstruction, Congenital, Urethrocutaneous, fistula

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Introduction

Congenital anterior urethrocutaneous fistula (CAUF) is a rare congenital anomaly of the penile urethra. It is a localized epithelial lined defect in the ventral urethra in subcoronal, penile or scrotal areas, along with a normal meatus.[1–3] The problem has usually been described as an isolated anomaly and also in association with other genitourinary abnormalities and anorectal malformation.[3,4]

CAUF is rare compared to the posterior/perineal urethrocutaneous fistula (UCF) which is usually classified as urethral duplication or accessory urethra and is sometimes associated with anorectal malformation.[4,5] The surgical management of the condition is individualized based on various clinical features such as the calibre of the distal urethra, the site and the size of the fistula, as well as other associated anomalies. Here, we report our experience with three cases of CAUF with variable clinical presentations and management.

Case reports

Case 1

A two-year-old boy presented with a subcoronal ventral opening in the urethra. He was passing most of the urine from the fistulous opening. As per his history and pictures provided by his parents, at the time of birth, the boy had a ventral thin-walled cystic swelling at the fistulous site (Figure 1(a)) and had a normal urinary stream from a normally positioned urethral meatus. The swelling subsequently burst, leading to the formation of a urinary fistula. On examination of the genitalia, the preputial foreskin and glans were normal. The meatus was wide and normally placed. There was a large epithelial-lined UCF measuring 10 mm×6 mm in the subcoronal area, extending up to the mid-penile area (Figure 1(b)). There was no chordee, and both testes were descended. Abdominal ultrasound (US) revealed no other associated abnormality. Surgery was planned, and urethroscopy was done, which revealed a normal urethra proximal to the fistula which could be easily cannulated

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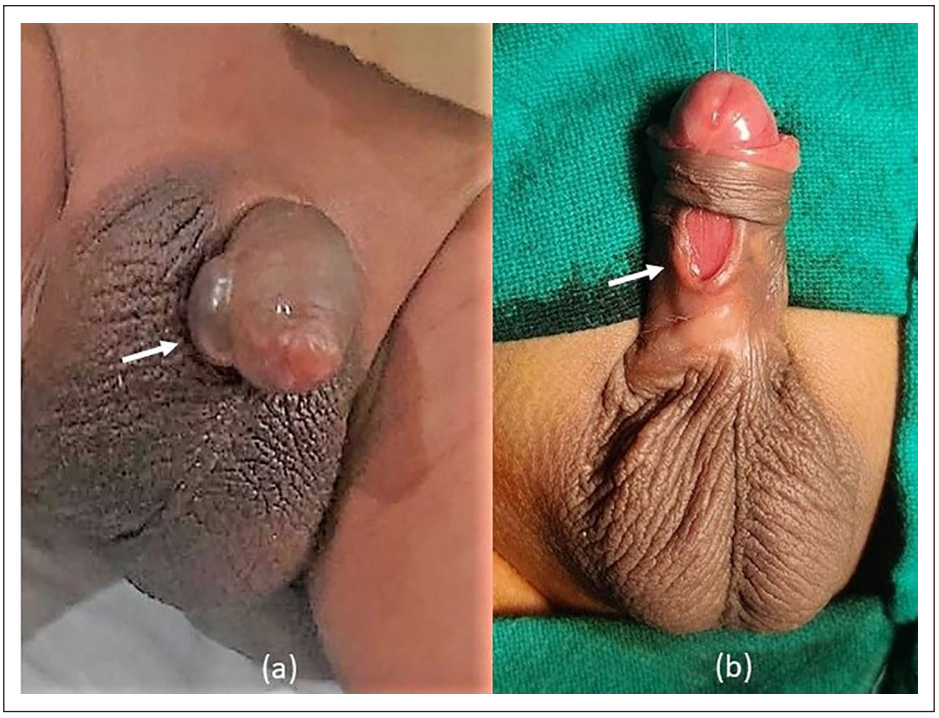

Figure 1. (a) Cystic swelling on the ventral aspect of the penis. (b) Congenital anterior (subcoronal) urethrocutaneous fistula (CAUF) following rupture of cystic swelling.

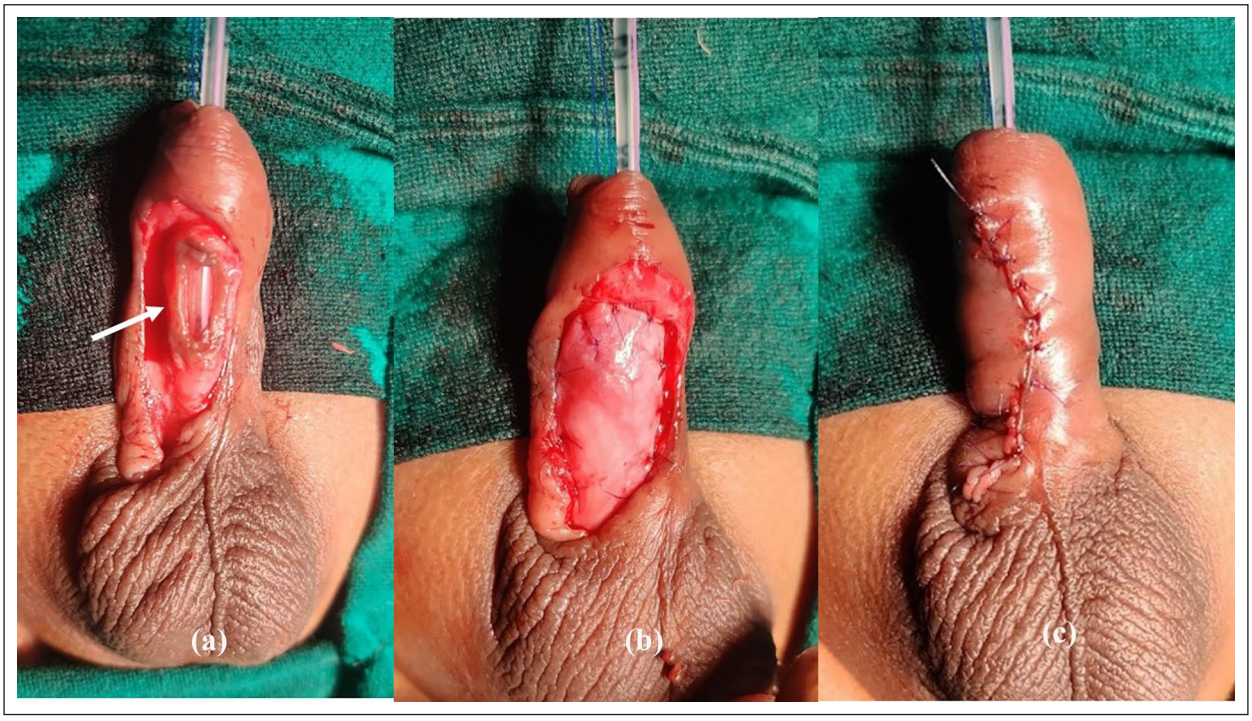

Figure 2. (a) Circumferential incision of CAUF over feeding tube no. 8. (b) and (c) Repair of fistula reinforced with tunica vaginalis flap.

with an 8 Fr feeding tube. An incision encircling the fistula was made to mobilize the edges. Local degloving was done, and the fistula was repaired along with the spongiosa. The suture line was further reinforced with a tunica vaginalis cover (Figure 2(a)–(c)). The catheter was removed on the 10th postoperative day. After a follow-up of one year, the child had good aesthetic results (Figure 3) and normal voiding.

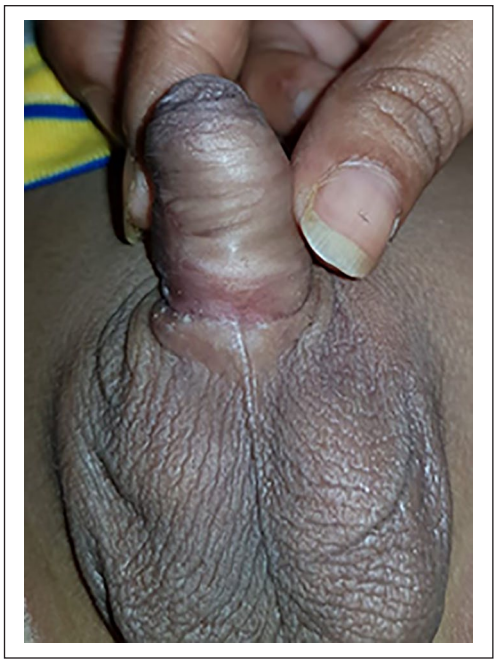

Figure 3. A good aesthetic outcome after follow-up of one year.

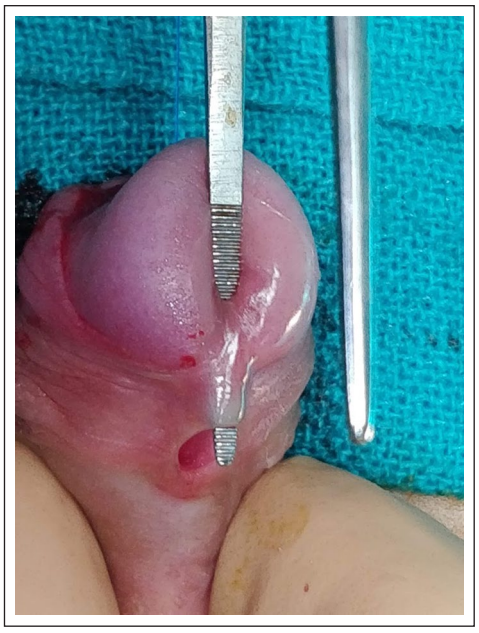

Figure 4. Subcoronal CAUF with chordee and thin bridge between fistula and normally positioned meatus.

Case 2

A boy aged one year and 7 months presented with a complaint of passing urine from a subcoronal urethral opening. On examination, he had a normally placed meatus with well-developed glans. A fistulous opening measuring 5 mm×5 mm was present in the subcoronal site separated by a thin bridge from the normally placed meatus. The prepuce was deficient ventrally with mild ventral chordee (Figure 4). The urethra could be easily cannulated with a 8 Fr feeding tube. Because of the presence of a thin bridge between the fistula and the normal meatus and associated chordee, Snodgrass urethroplasty was performed after laying open the urethra distal to the fistula. A double dartos cover was given to reinforce the suture line. After three years of follow-up, the patient is having normal voiding and insignificant residual urine on post-void US.

Case 3

A seven-year-old boy presented with a history of recurrent urinary tract infection. In addition, since birth, he had been passing urine from two abnormal openings: one in the subcoronal area from where he was passing urine in good stream, and another one in the perineum from where he was passing urine in drops. On examination, he had a normal foreskin, glans and meatus. A subcoronal epithelial lined fistula was present, measuring 7 mm×5 mm, through which the child was passing urine. A small pinpoint opening could be identified in the perineum 1 cm anterior to the anal verge through which he was passing urine in drops (Figure 5). The opening could not be cannulated. Renal

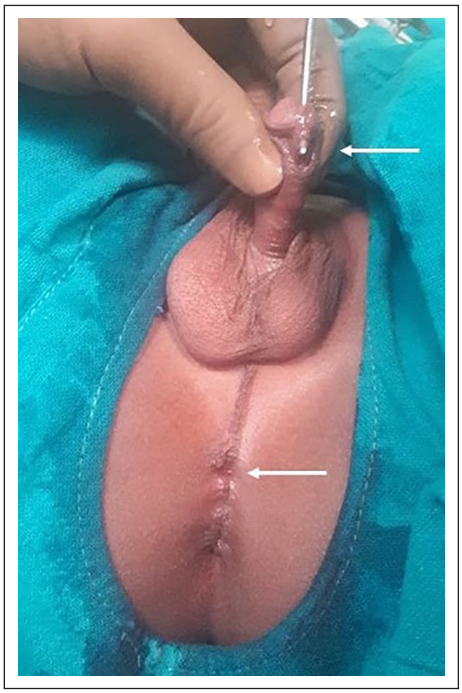

Figure 5. Subcoronal CAUF with normal meatus and associated perineal UF.

function tests were normal. US revealed right hydroureteronephrosis. On voiding cystourethrogram, right grade 5 vesicoureteric reflux was present. The urethra was normal, but no definite perineal fistulous connection could be delineated. A dimercaptosuccinic acid scan revealed a right non-functioning kidney. The child was planned for

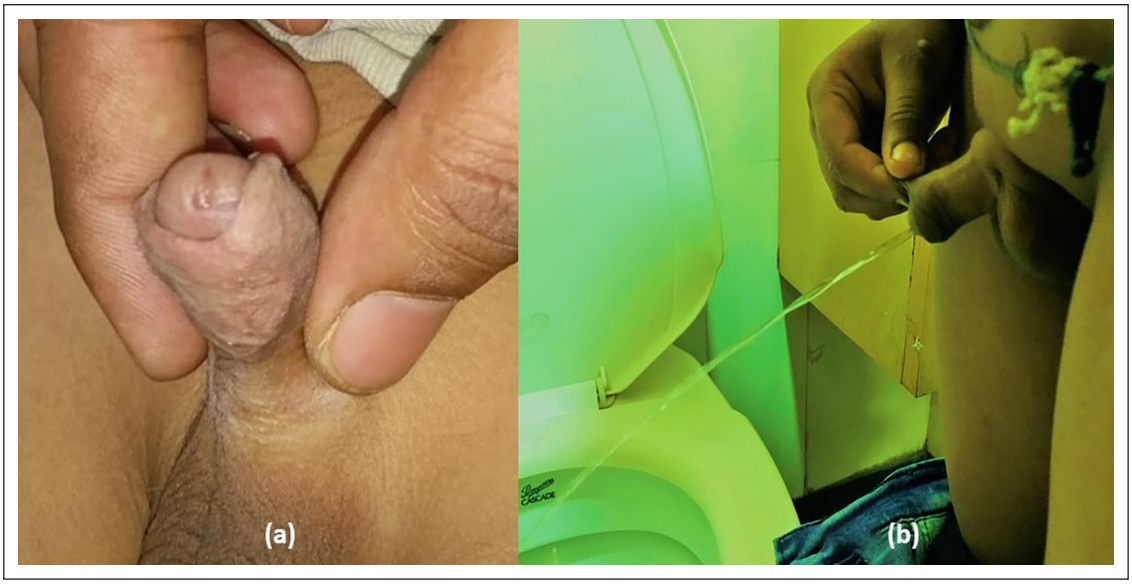

Figure 6. (a) and (b) Image after three years of follow-up with good cosmetic results and normal voiding.

urethroscystoscopy and right laparoscopic nephrectomy. On urethrocystoscopy, the urethra was normal except for a small fistulous opening identified adjacent to the verumontanum in the posterior urethra. The opening could not be cannulated. The leakage could be identified from the perineal fistulous opening, suggestive of perineal UCF.

After one month, surgery was planned for the closure of the anterior UCF and the perineal fistula. The anterior UCF was repaired as described in case 1. The perineal fistula was excised through a trans-perineal approach. Both repairs were reinforced utilizing a bilateral tunica vaginalis flap. After three years of follow-up, the patient is having normal voiding without any fistula recurrence (Figure 6(a) and (b)) and insignificant post-void residual urine on US.

Discussion

CAUF is a rare entity, with around 63 cases have been described in the literature.[6] Based on clinical characteristics, two varieties of CAUF have been described: one with a normal foreskin, intact distal urethra and normal spongiosum, and the other with characteristics of hypospadias-like features, with associated chordee, dorsal hood and spongiosal defect. This latter type comprises about one third of all the cases of CAUF.[7,8]

The site of anterior UCF can vary from subcoronal to the penoscrotal area.[3,6] In a detailed review of 63 cases by Lin *et al*.,[3] subcoronal fistulas were the most common, detected in 29 patients. A similar observation was made by User *et al*.[6] All three of our patients had subcoronal fistulas.

CAUF usually presents as an isolated anomaly, with about one third of cases having hypospadias-like features (chordee, dorsal hood, hypospadiac meatus, spongiosal defect).[7] The clinical presentation was variable in all three cases described in this series. The first case had an isolated subcoronal fistula with intact well-developed spongiosum and distal urethra. The second case had hypospadias-like features with poor spongiosum and chordee. Although the third case had normal spongiosum and distal urethra without chordee, it was unique as it was associated with perineal urethrocutaneous fistula. None of these cases were circumcised, and two of them had a well-developed preputial foreskin. A detailed clinical examination and evaluation is required to diagnose any other associated anomalies. In a review of 51 patients by User *et al*.,[3] various external congenital anomalies such as undescended testes, inguinal hernia, duplicated urethra and so on were associated with anterior UCF in 19 patients, while various internal anomalies such as vesicoureteric reflux, single kidney and so on were present in five patients. One of our cases had the rare and unique association of CAUF with perineal fistula and vesicoureteric reflux with a non-functioning kidney. The association of anterior UCF with posterior or perineal UCF or urethral duplication is rare and has only been reported twice. Both reported cases had CAUF with 'Y' duplication of the urethra in the form of urethroanal fistula and epispadiac type urethral duplication.[9,10]

Because of its rarity and variable clinical characteristics, the exact embryology is not known.[7] Focal or temporary defect of the urethral fold, resulting in deficiency of urethral fold fusion, can lead to the formation of UCF.[11,12] Transient deficiency of androgen or impaired local effect of testosterone is another proposed theory very similar to what has been described in the formation of hypospadias.[13] Another theory of the blowout phenomenon of urethral diverticula proposed by Campbell[14] is applicable in case 2. Defective corpus spongiosum or incomplete spongiosum leads to diverticula formation, with subsequent erosion of the overlying skin leading to the formation of UCF, as observed in this case. The association of CAUF with posterior UCF or urethral duplication is difficult to explain embryologically.[9,10]

The surgical technique should be individualized based on the various clinical characteristics such as (a) the patency of the distal urethra and the development of normal spongiosa, (b) the site and size of the fistula, (c) the presence of hypospadias-like features such as chordee, preputial hood and so on and (d) other associated anomalies.[1,2,6] In the presence of a normal calibre distal urethra and spongiosa, a simple fistula repair utilizing the principle of Dennis Browne urethroplasty with spongiosal cover can be successfully used. Incision of the urethral plate can be used in case of a narrow urethral plate.[3] In cases 1 and 3, the distal urethra was patent with a well-developed spongiosa. A primary repair could be performed easily, giving a wide calibre urethra. In patients with hypospadias-like features, as seen in case 2, urethroplasty utilizing principles of hypospadias repair should be performed. This will require correction of chordee and urethroplasty using the Thiersch Duplay technique or Snodrass repair.[2]

Size should be considered as an important determinant in fistula repair.[2,6] Two of the described cases requiring primary repair were ⩽1 cm in size, and tension-free repair could be easily done with a good outcome. A large fistula can be repaired by incising the urethral plate[3] or preputial island flap[15] or reconstruction by buccal mucosa.[16]

Another important determinant in planning surgical management is the association with other urethral anomalies. Case 3 had a rare association of anterior as well as posterior UCF, and both repairs were successfully done in the same sitting.

None of our patients had any complications and had satisfactory cosmetic and functional outcomes during follow-up. A recurrence rate of 11.3% has been reported in the literature,[6] which we feel can be further reduced with the correct surgical technique and spongioplasty reinforced with a dartos muscle or tunica vaginalis flap, as was done in our cases.

To conclude, CAUF is a rare urethral anomaly. Besides awareness about the condition, a careful clinical examination, evaluation and an appropriate surgical technique are required to achieve an optimal functional and aesthetic outcome.

Conflicting interests

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Ethical approval was obtained.

Informed consent

Informed consent was obtained from the patients for publication of clinical information and photographs.

Guarantor

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Contributorship

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