Congenital Urethrocutaneous Fistula—Our Experience with Nine Cases

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Abstract Congenital urethrocutaneous fistula is a very rare anomaly with about 40 odd cases reported in literature till 2008. We present here 9 such cases all of whom were uncircumcised at presentation. 7 out of 9 cases had a fistula in the distal shaft and the rest 2 cases had a fistula in the mid-shaft of the penis with an associated chordee. Associated congenital anomaly was present in only one case which had an associated imperforate anus. When the fistula was present distally, we did a primary repair of the fistula which was reinforced by a Bayer's Flap. When the fistula was present in the mid shaft we did a Primary repair of the fistula & reinforced it by a Tunica Vaginalis Flap.

Keywords Penis · Congenital · Urethrocutaneous · Fistula · Urethra

Introduction

Congenital anterior urethra cutaneous fistulae is a very rare anomaly that may present as an isolated abnormality or in association with other abnormalities such as imperforate anus [1, 2]. The cause is not clear and some have even questioned the validity of its diagnosis, claiming that this

entity may be the result of urethral injury during circumcision [3, 4]. However, they most probably reflect a focal defect in the urethral plate that prevents fusion of the urethral folds [5].

In this study, we have added nine of these very rare cases to about 40 odd cases, which have been already reported in the literature till 2008 [2, 5–11]. None of the cases in our series were circumcised.

Materials and Methods

We have treated nine patients with congenital anterior urethrocutaneous fistula over a period of 20 years from 1981 to 2011. In seven cases, the fistula was present in the distal shaft (Fig. 1) and the distal urethra or spongiosum was completely intact. In the rest two cases, the fistula was present in the midshaft and these two patients also had an associated chordee.

Congenital anomalies were present in only one case in our series where there was an associated imperforate anus. No patients had a urethral stricture distal to the fistula or meatal stenosis.

Results

The type of repair that was done was dictated by the position of the urethrocutaneous fistula. When the distal urethra or spongiosum was intact, as was present in seven of our cases, a primary fistula closure was done, which was reinforced by Bayer's flap. In the midpenile fistula, the primary fistula repair was reinforced by a Tunica Vaginalis flap. In five out of nine cases urethral dilatation was recommended 2 weeks after urethroplasty. Follow-up after 1 year showed that all patients had a good urinary stream through the normal urethral opening (Fig. 2).

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Fig. 1 Anterior urethra cutaneous fistula at the subcoronal level with intact distal urethra, no chordee

Discussion

Developmentally congenital urethral fistula can be grouped into three sets [12, 13]. The commonest one is rectourethral fistula seen in association with anorectal malformation. The second set is posterior urethroperineal fistula, usually classified as accessory urethra or Y type of duplication. The third and rarest set is congenital fistula of the distal urethra that has a direct continuity with the distal urethra. The urogenital system of the male embryo develops during weeks 8 to 14 of the gestational age [14]. At 8 weeks, the external genitalia of both male and female embryos are indistinguishable. Both have a midline genital tubercle just above a urogenital membrane flanked on each side by outer genital swellings and inner urethral folds [15]. Masculinization is an androgen-driven process, beginning under the influence of human chorionic gonadotropin, which stimulates the Leydig cells of the fetal testes to produce testosterone [16]. Testosterone is then converted to the more active dihydrotestosterone (DHT) by the enzyme 5- α reductase type II. For DHT to effect masculinizing action on the developing genitalia, it must bind to androgen receptors located in the genital tissues [16]. For this to occur, cellular signaling through the androgen receptors must be intact [17].

Early effects of testosterone are to increase the distance between the anus and genital structures, followed by elongation of the genital tubercle, which will become the penile shaft and glans [18]. The genital swellings, also called labioscrotal

Fig. 2 Normal urinary stream 6 months after repair of the anterior urethra cutaneous fistula

folds, migrate caudally and start to fuse, forming the scrotum [14]. A urethral groove developing on the underside of the penis becomes the urethra [18]. Folds of tissue, called urethral folds, which frame the lateral walls of this groove have inner (endodermal) and outer (ectodermal) edges [14, 18].

As the urethral groove develops from the posterior to anterior surface, it is soon enclosed by fusion of the endodermal folds to form a tubular penile urethra [17, 19]. The fusion of the endodermal edges creates an epithelial seam, which is subsequently reabsorbed [20]. The ectodermal edges then fuse over the urethra to fashion the penile shaft skin, leaving behind the median raphe [18]. The distal or glanular urethra develops last by one of two possible mechanisms [17]. The classic theory is that the distal portion of the urethra develops as an ingrowth of the ectoderm from the tip of the penis until it joins the proximal tubular urethra [14, 21]. Recent evidence, however, suggests that the entire urethra, from base to tip, is formed by continuous extension and fusion of the endodermal urethral groove [19, 22].

The embryological basis of congenital anterior urethrocutaneous fistula can be explained by the theory put forward by Glenister [23]. As the penile and glanular urethra develop from different sources, a fistula can develop in between due to misalignment of glanular and penile urethra. Olbourne [4] suggested that formation of a congenital urethrocutaneous fistula was due to a focal or temporary defect in the urethral plate, which prevented fusion of urethral folds and resulted in arrested distal migration of the urethral plate, or a localised deficiency of a portion of the plate. A deficiency of spongiosum with complete canalization of the distal glanular urethra may represent an abnormality, primarily of the anlage of corpus spongiosum, which is derived from inner genital folds [2]. In addition, this can also result from a blowout phenomenon of urethral diverticulum, as suggested by Campbell [24] and may represent a corpus spongiosum defect or incomplete migration of the lateral spongy tissue of the inner genital folds with later erosion of the overlying skin. This theory has been substantiated by Caltamone et al. [2] who noted a ventral penile cyst before the spontaneous development of urethral fistula, a similar finding was also noted by Knapp (1935) who noted six cases of cysts along the mediogenital raphe at the frenulum [24]. Coplen also documented prenatal identification of a penile cystic structure in a patient with postnatal anterior urethrocutaneous fistula [25]. However, we in our series of nine cases did not notice any ventral penile cyst. Similarly a defect in DHT, 5- α reductase or a localised androgen receptor defect may be a causative factor—these are the same reasons as suggested to be the etiology for hypospadias.

A review of the literature and a study of our nine cases suggest that there can be two types of anterior urethra cutaneous fistula. These are related to the presence of the fistula either as an isolated abnormality or in association

with hypospadias-like characteristics like presence of chordee, a dorsal hood, with or without a distal urethral or spongiosal defect and/or deficient foreskin. In our series, only in two cases the distal urethra was poorly developed with inadequate spongiosum. Only in one case there was an associated anorectal malformation.

The surgical approach to a congenital urethra-cutaneous fistula depends on whether the fistula involves an isolated abnormality or a hypospadias-like defect. One may consider primary fistula closure if the distal urethra/spongiosa is normally formed [2], as was present in seven out of our nine cases. This fistula closure was reinforced by Bayer's flap. With the presence of a distal urethral/spongiosal defect, and presence of chordee, as in our two cases, we did a primary fistula closure, which was reinforced by a Tunica Vaginalis flap.

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