Y-type congenital urethral duplication with normal dorsal urethra and small ventral fistula to perineal skin – 28th reported case

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ABSTRACT

There are numerous types of urethral duplication previously described in the literature including a Type IIA2Y where a large ventral channel branches from the hypoplastic, dorsal, orthotopic urethra. There have been 27 previously reported cases of a similar defect, called “congenital posterior urethrocutaneous fistula” (CUPF) with the only difference being that the ventral urethra is hypoplastic and the dorsal, orthotopic one normal. The difference in treatment for these 2 entities is markedly different, and pre-operative identification of the appropriate abnormality is essential. Treatment of CUPF requires only safe excision of the ventral segment, but treatment for a Type IIA2Y duplication requires resection of the hypoplastic, orthotopic urethra and transposition of the larger ventral segment into the dorsal orthotopic position. The outcome for treatment of CUPF is much better than for treatment of Type IIA2Y entities. We herein present the 28th case of CUPF in the English literature with discussion of the anatomy, appropriate diagnostic criteria, and safe method of treatment for these entities.

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Urethral duplication in children is a spectrum of abnormalities well described in 1976 by Effmann et al. [1]. His Type IIA2Y defect consisted of a large ventral channel which branched from the hypoplastic, dorsal (orthotopic) urethra with correction of the defect requiring excision of the dorsal urethra and transposition of the ventral one into the normal dorsal urethral bed. At least 27 cases of a defect similar to the Type IIA2Y defect, but definitely different with a large functional dorsal urethra and a smaller ventral defect opening onto the perineal skin, have been described [2–19]. This defect is usually referred to as a congenital posterior urethrop erineal fistula (CUPF). Since the treatment for the two defects is quite different, Bello [19] has suggested modifying the Effmann classification to include the CUPF as a “Type IIA2, Y-hypoplastic ventral urethral defect.” This presentation is the 28th case of CUPF reported in the English literature in the last 50 years.

1. Case report

A 2 year, 9 month old male was admitted to our children’s hospital because of multiple recurrent Escherichia Coli urinary tract infections (UTIs) for the past 9 months. After his second UTI, he had undergone a voiding cystourethrogram (VCUG) at an outside hospital which was read as normal, and he had undergone a normal cystoscopy. On this admission, the parents noted that the child, for the last 1.5 months, had been passing some of his urine per rectum and had been sitting on the toilet each time he micturated. A VCUG performed on admission at our hospital showed a fistula extending from the posterior, prostatic urethra into, what was thought to be, the distal rectum or anus (Fig. 1). A retrospective review of the VCUG performed at the outside hospital several months previously also showed the same fistula.

The child underwent examination under anesthesia with anoscopy. No distal fistula opening could be visualized in the perineum or inside the anus. Saline was injected into the urinary bladder through a urethral catheter and a Crede maneuver performed. Saline appeared in the perineum, but the exit point could not be clearly visualized. Methylene blue was injected into the bladder, and a repeat Crede maneuver performed. No methylene blue appeared in the anorectum, but a very small fistula opening was identified in the perineum just outside the anal sphincter muscle complex on the right side. After identifying the perineal opening, a 24 guage angiocath was inserted (Fig. 2) and a fistulogram performed which confirmed that the internal fistula opening was into the posterior wall of the prostatic urethra.

The child had a normal serum creatinine, and after successful treatment of his urinary tract infection he was taken electively to
the operating room for excision of the ventral tract of the urethral duplication. After induction of general anesthesia, he underwent an on the table bowel prep in case there was rectal violation during the dissection, and then he was placed in a modified lithotomy position. After a standard surgical skin preparation and sterile draping, he underwent cystourethroscopy for the purpose of confirming that there were no strictures in the dorsal urethra. A Foley catheter was inserted without difficulty and confirmed with fluoroscopy to be in the bladder. The patient was then placed in a prone jackknife position. The external opening of the fistula was identified and the fistula catheterized with a 5-French feeding tube which was secured to the skin to serve as a palpable landmark for the dissection. A muscle stimulator was used to accurately define the outer limits of the anal sphincter muscle complex. A semicircular incision was performed from the right lateral edge of the fistula tract, just along the anterior, external edge of the sphincteric mechanism, crossing the midline and proceeding to a similar position on the contralateral side. A midline incision was made anterior to and connecting with this semicircular one (completing a Y-incision). The fistula tract was circumscribed at the skin and then carefully dissected outside the anal sphincter muscle complex up to the entry into the posterior urethra. Great care was taken to stay immediately adjacent to the fistula tract which was easily palpable with the feeding tube inside. The fistula tract was ligated just outside its entry into the prostatic urethra taking care not to damage the prostate or urethra proper. There was no evidence of damage to the urethra, prostate, rectum, or anal muscle complex. Pathology examination of the excised tract demonstrated transitional epithelium nearest the prostatic urethral connection and squamous epithelium nearest the external fistula opening.

It was planned to leave a Foley catheter in place for 1 week postoperatively and then perform a retrograde urethrogram before discontinuing the catheter. However, on the third postoperative evening the catheter malfunctioned and was removed without obtaining an urethrogram. The child voided quite well without any evidence of stricture or leakage and was discharged home at that time. He has been followed closely for the past 15 months and is voiding and stooling well with no known complications.

2. Discussion

The initial comprehensive classification of duplications of the urethra by Effmann et al., in 1976 [1] has remained without significant change since publication. It has been reproduced several times [1,8,19] and will not be reproduced here. The Effmann classification type which comes closest to describing the case presented herein is the “Type IIA2,” subsequently supplemented with the addendum “Y-type.” In this Type IIA2Y urethral duplication, the larger channel is the ventral one while the orthotopic dorsal channel going through the penis is hypoplastic. Therefore most of the urine exits through the perineum with only small drops coming through the penis. Operative correction involves resection of the dorsal, penile urethra and transposition of the larger ventral urethra into the penis [1]. A variation of the Effmann Type IIA2Y duplication in which the ventral opening into the perineum branches from the posterior prostatic urethra and is actually the smaller of the two channels while the orthotopic, dorsal urethra is the larger and more functional channel (Fig. 3), was previously published in 1966 [2]. Even though published before Effmann’s classification system, this defect, referred to as a congenital posterior urethral fistula (CUPF) and not as a urethral duplication, was not included by Effmann.

Since the initial presentation of this CUPF variant, at least 26 more cases have been published in the English literature [2–19]. Several authors have demonstrated that the ventral fistula in these cases is usually partially or completely lined with transitional epithelium, and the abnormality should be labeled as a true urethral duplication and not just as a simple urethral fistula [5,11,17–19]. The most common, current terminology for this abnormality, however, remains “congenital posterior urethroperineal fistula.” No matter what the abnormality is called, the surgical treatment for this variant is much easier and more anatomic than that for the traditional Type IIA2Y, and the results of treatment are
much better [17–19]. The most common treatment currently for CUPF, as described in this case report, is surgical excision of the ventral tract with great care taken: (a) to avoid damage to the anus and anal sphincter muscle complex and (b) to avoid damage to the prostatic urethra by leaving a small amount of the ventral tract adjacent to the prostatic urethra instead of trying to excise 100% of the ventral tract. Fulguration of these CUPF anomalies, instead of operative resection, has been attempted with some success[9] and with some failure[3]. Newer radiologic techniques may eventually change treatment modalities, but since these cases are so rare and since operative excision has such excellent success it currently remains the treatment of choice for congenital posterior urethroperineal fistula.

3. Conclusion

Approximately 27 cases of congenital posterior urethrocata

neous fistula, in which the ventral urethral channel is small and the dorsal orthotopic one normal, have been previously reported in the English literature. The 28th case is herein reported. This abnormality is different from the traditionally described Y-type urethral duplication in which the dorsal, orthotopic urethral channel is hypoplastic and the ventral, ectopic channel is large. Differences in the anatomy and in the method of treatment and outcome of treatment for both of these entities are discussed.

References