Congenital urethral hypoplasia with urethral fistula without imperforate anus: Report of two cases

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ABSTRACT

Two male children with ano/recto-urethral fistula without anorectal malformation were managed in our institute. These patients had variable clinical manifestations and associated anomalies including congenital urethral hypoplasia, renal agenesis and patent urachus. Both were treated successfully according to their status of disease-complexes. This is to discuss these rare cases and its management with review of the literature.

Key words: Ano/recto-urethral fistula, patent urachus, urethral hypoplasia.

INTRODUCTION

Congenital abnormal fistulae between anorectum and urethra or vagina are usually associated with imperforate anus. Fistulae associated with normal anal anatomy have been reported almost exclusively in females.[1] Congenital ano/recto-urethral fistulae without anorectal malformations are extremely rare in male and have been called N-type or H-type fistulae.[2] We are reporting two cases of these rare anomalies and its management with review of the literatures.

Case 1

A three year old male child weighing 9 kg presented with complaints of excessive straining and crying during micturition, and discharge of serous fluid through umbilicus for few months after birth. He had history of recurrent urinary tract infection and passing of watery stool since birth. On examination child was malnourished with patent urachus, extensive excoriation and discharge around the umbilical region and periumbilical abscess. Genitalia were normal with bilateral palpable testicles. Although the urethral meatal opening appeared normal, [Figure 1] even a fine probe could not be negotiated into penile urethra beyond 1 cm. Anal opening was adequate with normal mucosa, and did not communicate with the urethral fistula. Blood urea and serum creatinine results were within normal limits; ultrasonography of the abdomen showed bilateral normal kidney with parietal wall abscess.

Drainage of parietal wall abscess was done. At the same time, urinary bladder was catheterized through patent urachus. After four weeks, cystoscopy and rail-road catheterization was tried but it failed. Ligation of patent urachus with suprapubic cystostomy was performed. [Figure 2] Two weeks latter, antegrade voiding cystourethrogram was done which showed recto-urethral fistula without ureteric reflux. Through anterior perineal approach, urethral fistula, situated about 2 cm proximal to dentate line, was separated from anterior wall of rectum and perineal urethrostomy with anoplasty was performed. Colonic irrigation by normal saline was done one day before surgery. On the second post-operative day, the urethral catheter was dislodged accidentally, which necessitated a protective divided sigmoid colostomy. After four week, colostomy was closed. Urethroplasty was affected in two stages. [Figure 2] He developed an urethro-cutaneous fistula, [Figure 1-3] which was treated satisfactorily. In the last six months, the boy suffered two episodes of urinary retentions with infections that needed urethral dilatation and antibiotics. He is presently passing urine and stool normally.

Case 2

A four year old male child weighing 12 kg presented with complaint of passing urine through the anus and dribbling urine per urethra since birth. On examination, urachus was obliterated and genitalia were normal with bilateral palpable testes in the scrotum. External urethral meatal opening was normal but catheter could not be negotiated into penile urethra beyond 3 cm. Anal opening was adequate and normally situated. A

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Figure 1: Ano-urethral fistula showing spurt of urine

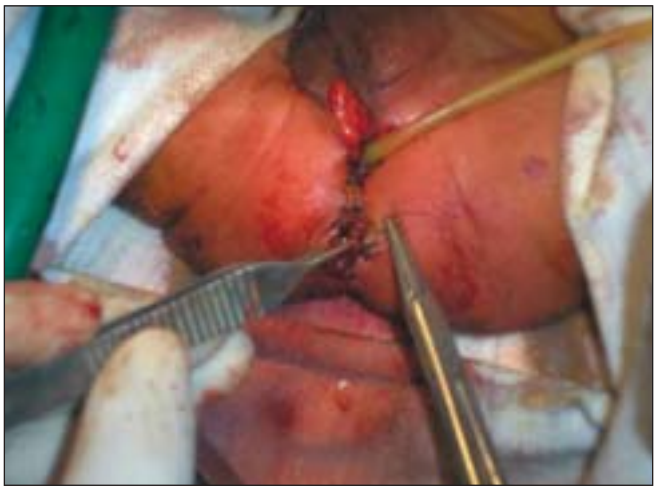


Figure 2: Operative photograph showing perineal urethrostomy and anoplasty



Figure 3: Urethroplasty for perineal hypospadias and cnnulated penile urethra with penoscrotal urethrostomies

urinary fistula was present on the anterior wall of anal canal, about 1cm proximal to anal verge with spurt of urine. Renal function was normal. Ultrasonography and IVP did not show the left kidney, but DTPA-scan revealed a left renal agenesis with normal functioning right kidney (Global GFR - 48 ml /min /0.6 sqm BSA). Cannulation of urinary fistula by guide wire was done under fluoroscopy. Contrast study showed the opening of fistula at membranous urethra with passage of dye into urinary bladder. Child was operated after colonic wash. Separation of urethral fistula from anterior wall of anal canal through anterior perineal approach and perineal urethrostomy with anoplasty was performed. Penile urethra was dilated and another urethrostomy was performed at penoscrotal region. Colostomy was not done. After regular dilatation of penile urethra for three months, urethroplasty was affected in stages. The patient is continent and passing urine and stool normally, three months after follow-up.

DISCUSSION

Congenital ano/recto-urethral fistula without imperforate anus in male is rare representing less than 1% cases of all anorectal malformations.[1] Associated urethral hypoplasia is further an extremely rare variant of anorectal malformations among boys.[2]

Congenital urethro-anal fistula with normal anus is usually associated with an atretic / hypoplastic anterior urethra and has been variously described as a variant of anorectal malformation by some authors[1,3] and urethral duplication by others.[2,4,5] Most authors [1,3,6] believed that ano/recto-urethral fistula with normal anus is a variant of anorectal malformation and thought that fistula is a result of persistence of cloacal duct. Kluth *et al*,[6] described that cloacal membrane and dorsal cloaca are essential for the normal development of the anal orifice. A defective cloaca analoge results in a missing and misplaced anal orifice and an abnormal communication between the rectum and the ventral urogenital tract.[6] The tract cannot be considered as a urethral duplication because of the squamous epithelial lining.[1] These ARMs in boys are also termed urethroanal, urethrorectal fistula or N-type / H-type fistula of anorectal region and corresponds to the anorecto-vestibular fistula with a normal anus (perineal canal) in a female.[1,3] Other authors believe that all urethro-anal fistulas are not a variant of anorectal malformation but could be due to urethral duplication which may be H-type, Y-type, N-type etc.[2,4,5] These anomalies are variably associated with other anomalies and different clinical presentations like patent urachus, pelvic/ectopic/

agenetic kidney, hydronephrosis, recurrent urinary tract infection etc.[4] The embryological basis for urethral fistula with severe urethral hypoplasia and normal anus remain speculative; different suggestions have been proposed.[2] Al-Bassam *et al*,[2] reported 31 cases of ano/recto-urethral fistula with normal anus but only 12 of the 31 patients had associated urethral hypoplasia, stenosis or atresia. We are adding these two cases of severe urethral hypoplasia, associated renal agenesis and patent urachus.

Various surgical procedures are described to treat the urethral hypoplasia with ano / recto urethral fistula,[2] involving protective colostomy, cystostomy and staged urethroplasty.

We conclude that these anomalies are extremely rare and can be present with variable clinical manifestations and associated anomalies. Management can be successful via perineal approach. Anterior urethral hypoplasia can be treated by regular urethral dilatation with minimum complications.[4] Detailed clinical history and examination to find out the exact pathology is necessary to minimize the postoperative complications. Diagnosis is usually difficult and can often be missed. The VCUG is the most useful diagnostic tool to clinch out the anomaly. Protective colostomy and even cystostomy can be avoided with manageable complications.

REFERENCES

1. Hong AR, Croitoru DP, Nguyen LT, Laberge JM, Homsy Y, Kiruluta GH. Congenital urethral fistula with normal anus: A report of two cases. J Pediatr Surg 1992;27:1278-80.
2. Al-Bassam A, Sheikh MA, Al-Smayer S, Al-Boukai A, Al-Damegh S. Congenital H-type anourethral fistula with severe urethral hypoplasia: Case report and review of the literature. J Pediatr Surg 1998;33:1550-3.
3. Lal P, Gupta A, Krishna A, Taneja K. Congenital H-type urethro-anal fistula. Pediatr Surg Int 1998;13:193-4.
4. Passerini-Glazel G, Araguna F, Chiozza L, Artibani W, Rabinowitz R, Firlit CF. The PADUA (Progressive augmentation by dilating the urethra anterior) procedure for the treatment of severe urethral hypoplasia. J Urol 1998;140:1247-9.
5. Desgrez JP, Cendron J. Incomplete duplication of the urethra with intra-rectal opening of the posterior channel. J Urol Nephrol (Paris) 1975;81:169-74.
6. Kluth D, Hillen M, Lambrecht W. The principle of normal and abnormal hindgut development. J Pediatr Surg 1995;30:1143-7.

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