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Studies on Female Urethral Anomalies in Pediatric Age Group in India

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ABSTRACT

Female urethral anomalies such as epispadias and hypospadias have been rarely reported in the literature. Clinical diagnosis of female urethral anomalies is significant for management. Diagnosis may be missed leading to mismanagement of the clinical condition.

Keywords: Female epispadias; congenital anomalies; female hypospadias; urethral anomalies.

1. INTRODUCTION

Anomalies of the urogenital tract are among the most common organ system anomalies found in the fetus or neonate. Urethral anomalies in females are uncommon. External genitalia have varied appearance as classified by Davis ranging from lesser degrees with patulous urethral orifice to intermediate cases with urethra dorsally split along most of its length to the most severe cases which involve the entire length of urethra and bladder neck rendering the sphincteric mechanism incompetent [1]. The different female urethral anomalies are hypospadias, epispadias, urethral diverticulum, urethral duplication, urethral prolapse, urethral stenosis.

Female hypospadias is an abnormality of the urethra, with the urethral meatus located at any site on the anterior vaginal wall from just above the introitus up to the vaginal fornix. Its incidence is difficult to assess because many cases are labelled as urogenital sinus [2].

Female epispadias is a very rare malformation occurring in 1:480,000 female births. Male patients outnumber female patients with epispadias by a ratio of 3:1 or 4:1 [3]. In most patients the defect involves the entire urethral length and bladder neck, and produces incontinence.

Female urethral diverticulum is an uncommon pathologic entity and can manifest with a variety of symptoms involving the lower urinary tract. Female Urethral Diverticulum is a rare condition affecting less than 20 per 1,000,000 women [4]. Small diverticula in female infants are missed and recognized only when they are rendered symptomatic by trauma, infection, or increase in size later in life [5].

Urethral duplication in girls is very rare but should be considered in girls who have complete incontinence, perineal discharge, and perineal, suprapubic, or paraurethral sinuses [6]. This anomaly is often associated with duplication of bladder in females. In females, urethral duplication is classified according to plane (frontal or sagittal) of duplication into different types: (1) Double urethra and double bladder, (2) double urethra with single bladder, (3) accessory urethra posterior to the normal channel, (4) double proximal urethra and single distal urethra, and (5) single proximal urethra and duplicated distal urethra [7].

A distal urethral ring and urethral meatal stenosis in girls were once believed to be significant pathological lesions, especially when associated with urinary tract infections [8]. Now, however, it is known that urethral obstruction in girls is exceedingly rare. Infravesical obstruction results in bladder

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changes which relate to the degree and duration of the obstructive process, and to the presence or absence of urinary infection [9]. In girls there are three distinct morphological kinds of congenital distal urethral stenoses in the broad sense. Incomplete meatal stenoses are the most frequent of these [10]. Prolapse of the urethral mucosa in girls is an uncommon condition. It appears as an edematous, friable rosette of bright red or cyanotic tissue that can become infected, ulcerated, necrotic, or even gangrenous and can enlarge to cover the entire vulva [11]. Prolapse occurs most frequently in either very young premenarche girls or postmenopausal women. Because of its rarity, the diagnosis frequently is mistaken. The etiology remains unknown, and symptomatology is characterized generally by painless vaginal bleeding and, rarely, by micturition disturbances.

Persistent urogenital sinus is an anomaly of the cloaca [12]. Reported incidence is 0.6 in 10,000 female births. Urogenital sinus is defined as a common channel through which drain the urinary and genital tracts [13]. A cloaca is a common channel through which drain the genital, urinary and alimentary tracts. An important group of girls with urogenital sinus anomalies are those female infants with the adrenogenital syndrome. The vaginal wall gets edematous and swollen and thus may cause backpressure changes leading to hydronephrosis.

2. CASE REPORTS

2.1 Case 1

A 5-year-old girl presented to us with complaints of urinary incontinence. History did not reveal any evidence of urinary tract infection, trauma or operative interventions. The child had normal developmental milestones. There were no other urological complaints. The child was voiding good quantity of urine. To quantify her incontinence, it was around 4–5 pads/day. On examination of external genitalia revealed bifid clitoris [Fig. 1]. Vagina and hymen were normal. The urethra was splayed and directed cranially with continuous dribbling of urine. Neurological examination was normal. Blood, urine routine and renal function tests were normal.

X-ray pelvis showed a defect in symphysis pubis [Fig. 2]. Ultrasonography was done to rule out upper tract disease and was normal. Micturating voiding cystourethrogram revealed good capacity bladder. Cystourethroscopic assessment revealed normal appearing ureteric orifices and intact bladder neck.

A vertical parallel incision was taken along the urethra [Fig. 1]. The urethra was mobilized to both the sides. Urethral reconstruction was done over a 10 French Foley's catheter. The redundant urethra was excised. The triangular area over the bifid clitoris was marked, and the rawarea was sutured to obliterate the dead space. Postoperative period was uneventful. The catheter was removed after 9 days. The child was continent and could void well with no complaints. Postoperative ultrasonography was done at 6 weeks and 12 weeks with bladder capacity of 140 ml and 160 ml, respectively.

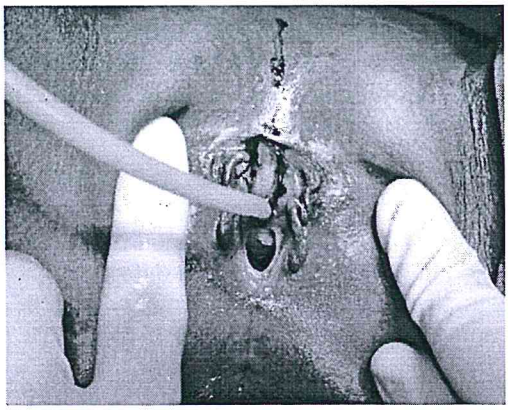

Fig. 1. Parallel incision taken for reconstruction of urethra. Note the bifid clitoris with deficient dorsal wall

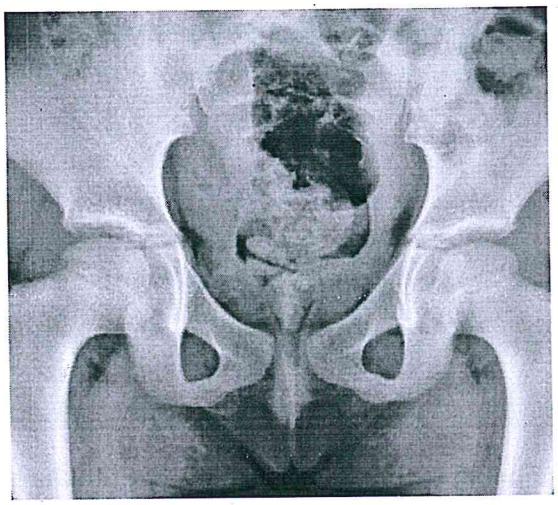

Fig. 2. X-ray pelvis shows defect in the symphysis pubis

2.2 Case 2

A 7-year-old girl presented to us with complaints of only true urinary incontinence. Her developmental milestones and higher motor functions were normal. There were no neurological deficits. Hemogram and urine routine was normal. On examination of external genitals, urethral meatus was caudally directed, and continuous leaking of urine was noted. The vagina was normal for the child's age on genitoscopy. Vulva and hymenal ring were normal. Cystourethrogram revealed short urethra. On cystourethroscopic assessment was done prior toa definitive procedure revealed short urethra (<1 cm). Urethral opening was seen almost close to the bladder neck. Both the ureteric orifices were normal, and bladder was normal with a capacity around 120 ml. There was no evidence of trabeculations. On insertion of 12 French Foleys catheter, the bulb of the catheter could be easilyvisualized [Fig. 3]. The patient underwent primary urethral reconstruction over a 12 French Foleys catheter. The wide-open urethral plate above the meatus and below the clitoral opening was mobilized by taking an inverted "U" incision [Fig. 3]. After careful mobilization of the urethra, it was tubularized over 12 French Foleys catheter using absorbable suture [Fig. 4]. The neomeatus formed after the reconstruction was sutured to the vagina after mobilization of the periurethral tissue. Catheter was kept for 11 days. The child was continent and voided adequate amount of urine with good flow. Postoperativeultrasonography was done at 12 weeks with bladder capacity of 220 ml.

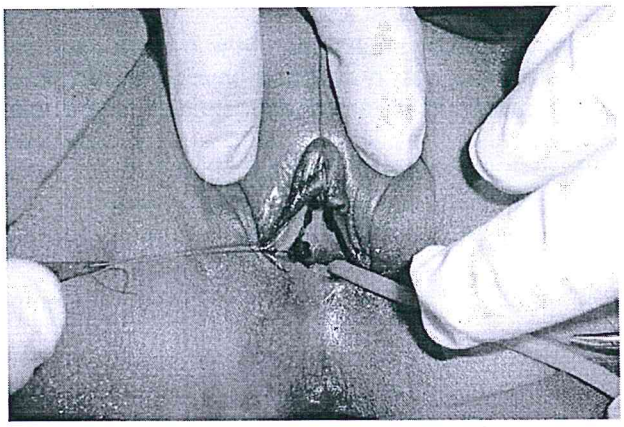

Fig. 3. Female hypospadias; inverted "U" incision is taken for reconstruction of urethra. Note: that the bulb of the catheter is visualized suggestive of short urethra

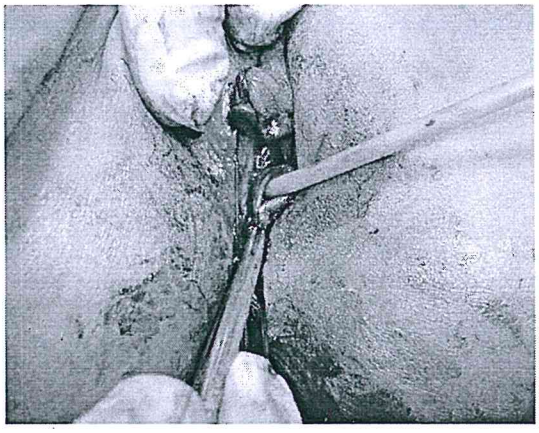

Fig. 4. Urethra being reconstructed over a 12 French Foley's catheter

3. DISCUSSION

Female urethral anomalies are rarely encountered in pediatric age group. These children present with various symptoms ranging from obstruction to urinary incontinence, posing a great challenge for pediatric urologists. High degree of suspicion and good clinical examination are paramount for accurate diagnosis and management. The aim is to provide continence and give a good cosmetic result, which will have a positive psychological impact on the child (14). Majority of the anomalies are case reports, with few centres having significant expertise. We have presented two rare anomalies which were managed successfully.

The importance of a careful examination should be emphasized, especiallyin female patients with chronic wetting of unknown cause. Davis has divided the female epispadias into three degrees: (1) Patulous urethra (milder form); (2) urethradorsally split (intermediate degree) and; (3) cleft involving the whole urethra and bladder neck (severe degree) [15].

The genital defect is characterized by bifid clitoris and mons is depressed in shape and coated by a smooth area of skin. The vagina and the internal genitalia are normal. 30–75% of epispadias are associated with vesicoureteral reflux [16]. These children should undergo a completeradiological evaluation to identify and rule out other causes of incontinence. The role of the urodynamic study is indefinite but would help to justify the poor outlet resistance. Cystourethroscopy is required to access urethral length, bladder capacity and position of the ureteric orifice.

Female hypospadias is characterized by shortening of the urethra and ectopia of the external urethral opening. This deformity is rarely diagnosed because physicians are not well-informed about female hypospadias. There are various variants of female hypospadias [17]. These include:

- Low vaginal ectopia of the external urethral opening
- High vaginal ectopia of the external opening of the urethra
- Urovaginal (vesicovaginal) fusion of the neck of the urinary bladder with vagina accompanied with enuresis
- Urogenital sinus in females (ectopia of the external urethral opening in the urogenital sinus).

All the variants of female hypospadias must be surgically corrected as transposition of the external opening of the urethra from the vagina on the perineum under the clitorisin chronic urethritis. The second case mentioned above mimics sinus urogenitalis, but is not so because the majority of children born with a urogenital sinus have genital ambiguity [18]. At the same time, in the second case there was a healthy ventral urethral plate which is suggestive that it is a case of female hypospadias rather than sinus urogenitalis.

5. CONCLUSION

Diagnosis of female urethral anomalies and its treatment possess a great challenge to pediatric urologists. Careful clinical examination plays an important role in the diagnosis of these anomalies. The objectives of surgical repair include achievement of urinary continence with preservation of the upper urinary tracts and the reconstruction of functional and cosmetically acceptable genitalia [19].

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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