Female Epispadias: A Case Report and Review of the Literature

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Isolated female epispadias without exstrophy is an extremely rare syndrome. The symptoms of female epispadias are primary urinary incontinence and abnormal anatomical features. A 12-year-old girl presented with primary urinary incontinence. On physical examination, bifid clitoris and labia minora were seen. The vagina and hymen were normal. Voiding cystourethrogram showed no reflux. With the diagnosis of isolated female epispadias, one-stage reconstruction of the urethra, bladder neck and labia minora and clitoris was performed.

Key Words: epispadias, female, urethra, urinary abnormality
(*Kaohsiung J Med Sci* 2009;25:613–6)

The embryological development of the female genitourinary system is quite complex. Congenital abnormalities commonly involve the urinary and genital tracts [1]. Urinary system anomalies are the third most frequent congenital deformities. Anomalies associated with the urethra such as anterior urethral valves, syringocele, megalourethra and duplicated urethra are generally seen in males [1–3]. While those conditions are seen in male patients, isolated female epispadias without extrophy is an extremely rare syndrome.

In this case report, we present a 12-year-old girl with isolated female epispadias who underwent one-stage operation to correct the abnormalities.

Case Presentation

A 12-year-old girl was admitted to our clinic with symptoms including total incontinence and skin rashes in bilateral inguinal regions. The patient's family also reported history of recurrent urinary tract infections. The patient's past medical history was normal. However, bifid clitoris and labia minora were detected on physical examination (Figure 1). Although routine biochemical parameters were within normal ranges, urinalysis revealed more than 15 red blood cells and 7–8 white blood cells per high power field. There were no abnormalities on intravenous urogram or urinary ultrasonographic examinations. A voiding cystourethrogram showed a normal-appearance bladder and no evidence of vesicoureteral reflux.

A urodynamic study revealed a bladder capacity of 107 mL with an abdominal leak point pressure of 20 cmH_2O. Urethrocystoscopy showed a short, wide urethra, approximately 1 cm long, with splaying of the roof of the urethra and a wide bladder neck. The ureteric orifices were situated 2 cm from the bladder neck and were of normal caliber. The rest of the bladder was cystoscopically normal.

Surgical technique

With the diagnosis of female epispadias, a de Jong surgical procedure was performed as previously described [4] for reconstruction. The procedure began with cystoscopy, followed by excision of the urethral plate between the clitoral halves and the urethral plate was left attached to the bladder neck. The

Received: Mar 12, 2009 Accepted: Apr 17, 2009
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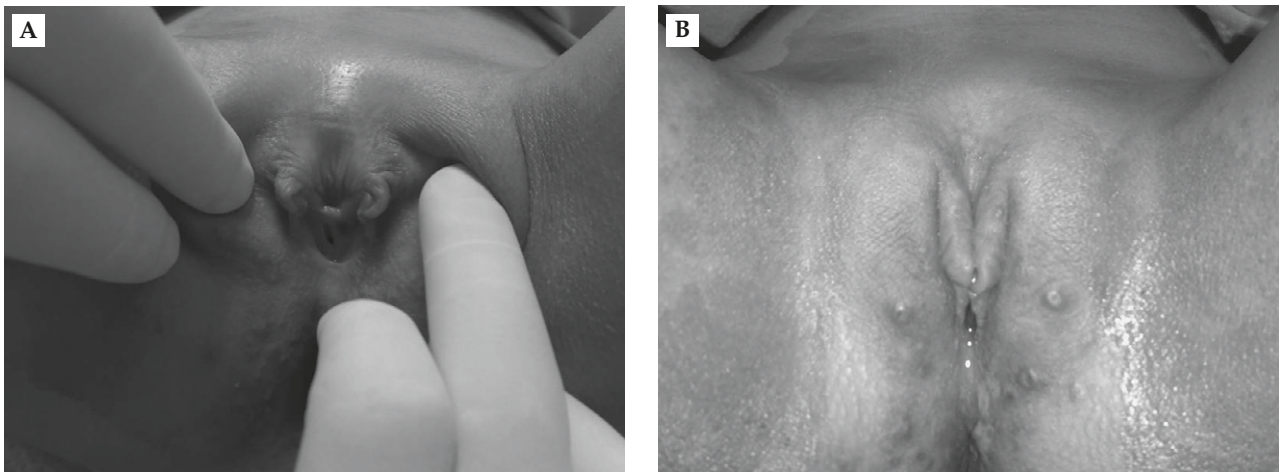

Figure 1. *(A, B) Preoperative appearance of the external genitalia shows female epispadias with a normal hymenal ring, bifid clitoris and urethral plate spread anteriorly between clitoral halves.*

attachments of the bladder neck to the anterior vaginal wall and the symphysis pubis were released by careful dissection up to the level of the trigone, resulting in complete mobility of the bladder outlet and attached urethral plate. The urethral plate was then tubularized into a urethra. The bladder neck and the proximal urethra were moved to the intra-abdominal position by a modified needle suspension technique. In this technique, a 2-0 non-absorbable polypropylene suture was first passed from the periurethral tissue at the 9 o'clock position at the level of the bladder neck. The free end of the suture was carried to the posterior (abdominal) side of the urethra and a second pass of the suture was performed at the 3 o'clock position. Both ends of the suture were then advanced and fixed to the posteroinferior border of the pubic symphysis bilaterally. The pelvic floor defect was then closed in the midline position between the anterior vaginal wall and urethra. These sutures made continence possible. The urethral end was sutured posteriorly at the level of the hymenal ring laterally and anteriorly at the labia minora. The procedure was completed with cosmetic reconstruction of the external genitalia. The clitoral halves were fused by denuding them of medial epithelium and connecting them with 7/0 polyglycolic acid sutures. The anterior distance of the labia majora was decreased by excising the skin between the labia and suturing the defect longitudinally. Postoperatively, bladder drainage was maintained with a 10-Fr transurethral catheter for 14 days. Antibiotic prophylaxis consisting of 2 mg/kg trimethoprim was given once daily and 0.4 mg/kg oxybutynin

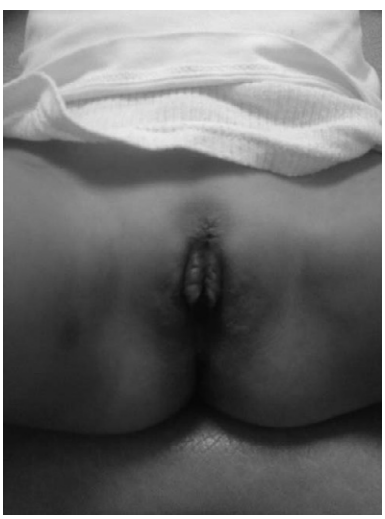

Figure 2. *Appearance of the external genitalia 3 months after reconstruction. The neourethra is in the normal position with a normal hymenal ring. The bifid clitoral halves are attached and the gap at the anterior labial junction is straightened.*

was given daily in three doses during the catheterized period. The postoperative cosmetic appearance was acceptable (Figure 2).

A postoperative control voiding cystourethrogram 3 months after the operation revealed a normal bladder neck and a lengthened urethra, with no reflux. There was no residual urine and the patient achieved continence within 2 weeks after the operation. A postoperative urodynamic study in the 3rd month after the operation showed that the bladder capacity was 175 mL with an abdominal leak point pressure of 44 cmH_2O. After an uneventful postoperative period

at 6 months, micturition and the cosmetic appearance of the patient were normal.

Discussion

The development of the female genitourinary system is complex. The wide spectrum of anomalies of female embryogenesis result in problems related to the development of the urinary and genital tracts [5]. Numerous anomalies of the urethra exist, either as isolated anomalies or in combination with other disorders. Female epispadias is a rare congenital anomaly with an incidence of 1/480,000 female births [6]. Patients are generally diagnosed before adolescence, as in the present case. However, diagnosis might be missed unless the labia are separated and examined. Generally, patients present with abnormal external genitalia and a wide anterior gap between the labia majora and bifid clitoris. The urethral plate extends between the clitoral halves. In most cases, the bladder neck is located at the level of the hymenal ring. No urethra and no sphincteric mechanism is present, resulting in complete incontinence, which is often accompanied by a bladder capacity that is relatively small for age. In contrast, the vagina and internal genitalia are usually normal [7,8]. The ureters open laterally in a more-or-less straight course, with refluxing in 30–75% of cases. There may be history of recurrent urinary tract infection [8].

The aims of management are achievement of continence, preservation of the upper tract and reconstruction of functional and cosmetically acceptable genitalia. The anomaly can be corrected by surgical reconstruction of the bladder neck, urethra and external genitalia. To achieve continence, an intra-abdominal position of the bladder neck and proximal urethra is essential [1,9,10]. Although, historical treatment of epispadias consists of staged repairs with urethral and vulva reconstruction at stage 1 at 1–1.5 years, followed by bladder neck reconstruction at 4–5 years, these procedures often have suboptimal results with respect to urinary continence. In addition, staged procedures require multiple sessions of surgery and anesthesia, and are associated with relatively higher morbidity [11]. Moreover, clean intermittent catheterization and/or a continent catheterizable stoma of the augmented bladder is needed in many cases. Persistent incontinence may be treated with collagen and/or dextranomer hyaluronic acid implantation at the bladder neck. More than 50 years ago, Young recognized the need to radically alter the urethra and bladder neck to achieve continence in these cases [12]. Various modifications have since occurred. Hendren's combined abdominal-perineal single-stage reconstruction is one of the most commonly performed procedures and is associated with good continence results [13]. If the ureters have high-grade reflux, they should be reimplanted. Reimplantation is also required if the ureters are so distal in the trigone that they interfere with upward tubularization of the urethra and bladder neck. In single-stage procedures, the urinary continence rates are between 60% and 87.5% [11,13]. Recently, de Jong et al treated four patients using urethral and pelvic reconstruction with bladder neck suspension via a transperineal approach [4]. They described satisfactory continence in three of the four patients by bringing the bladder neck and posterior urethra into an intra-abdominal position. We used a similar technique in this case. Although epispadias is frequently observed in boys, isolated female epispadias is a rare condition presenting with specific therapeutic problems.

One-stage urethral reconstruction, intra-abdominal positioning of the bladder neck and proximal urethra with a modified needle suspension technique, and pelvic floor reconstruction appears to be a promising approach for treating female epispadias. Although we had very good results, both functionally and cosmetically, our case series is too small to reach a dogmatic conclusion. More experience is required to confirm the advantages of this technique. We advocate this procedure as a first choice in all patients with female epispadias, because it is simple, safe and effective for continence. More experience is needed to confirm the superiority of this technique for this rare anomaly.

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