FEMALE EPISPADIAS: A CASE REPORT

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Female epispadias without bladder exstrophy is an extremely rare anomaly occurring in 1:480.000 girls. It presents typical features and can be diagnosed immediately at birth. Early surgical reconstruction of the bladder neck, urethra, and external genitalia within the physiological phase for the development for continence, is relevant towards establishing urinary continence and to reduce the psychological impact on the parents and the child. In this case report we present a 3-years-old girl with isolated female epispadias who underwent total reconstruction at a single procedure with a follow-up of 6 months.
Key words: female epispadias, one-stage epispadias reconstruction, bladder exstrophy.

INTRODUCTION

Isolated female epispadias without exstrophy is a rare anomaly that forms part of the extrophy-epispadias-complex occurring in 1:480,000 girls. The condition is often missed at first examination but should be diagnosed immediately at birth. At informed routine examination of the external genitalia the wider central space between the labia majora, the associated diastasis of the pubic symphysis, the anomalous 'open' aspect to the dorsal urethra, and the divided hemiclitori lying to each side are obvious telltale indicators. Most cases are associated with an incomplete urethral sphincter, an open bladder neck, and a small bladder capacity with virtually absent bladder filling. Careful observation will confirm constant dribbling of urine and the absence of an intermittent urinary stream. Early diagnosis and surgical reconstruction of the bladder neck, urethra and external genitalia (1,2) within the physiological time frame are relevant to improve the chance for urinary continence and to reduce the psychological and psychosocial problems for the parents and the child.
In this case report we present a 3-year old girl with isolated female epispadias with total urinary incontinence, who underwent total reconstruction at a single procedure, with a followup of 6 months. The child is now 4 years old and is continent, voiding at-will with no interim wetting, with a good aesthetic appearance to the vulva and easy access to the urethra and vagina.

CASE REPORT

A 3-year old female, born by normal vaginal delivery at 40 weeks gestation, was admitted to our clinic for evaluation of 'genitalia abnormalities' and constant wetting in the context of otherwise normal developmental milestones. She was the product of a non-consanguineous marriage with no family history of a similar problem. A younger male sibling was normal. There was no significant past medical history except for occasional 'pyrexia of unknown origin' for which a urinary origin had been excluded by urine culture and analysis. Although the correct diagnosis was suspected when she was 1 year old, she was referred to us at 3 years of age when her weight at 10 kg and her height at 84 cm were under the 5th percentile. Informed examination of the external genitalia revealed dribbling incontinence with typical features of complete epispadias without exstrophy. There was a depressed mons pubis with a palpable wide diastasis of the pubic symphysis (confirmed at x-ray) that was covered by thin skin extending in the midline between the labia majora. Separation of the labia revealed a hemiclitoris and a small labium minor on either side. The central urethra lying above the vagina, was short and widely open dorsally, communicating with an open bladder neck that was incontinent of urine (Fig. 1). Renal function studies, routine blood evaluations, urinalysis and urine cultures were normal. Renal ultrasound revealed two normal kidneys with no ureteropelvic dilatation and an empty bladder. A micturating cystourethrogram showed a small bladder capacity and left grade I vesico ureteric reflux. Urodynamic studies were not performed because of the age of the child. X-ray confirmed a 2 cm symphyseal diastasis. Cystourethroscopy confirmed the short wide open urethra and the wide in-

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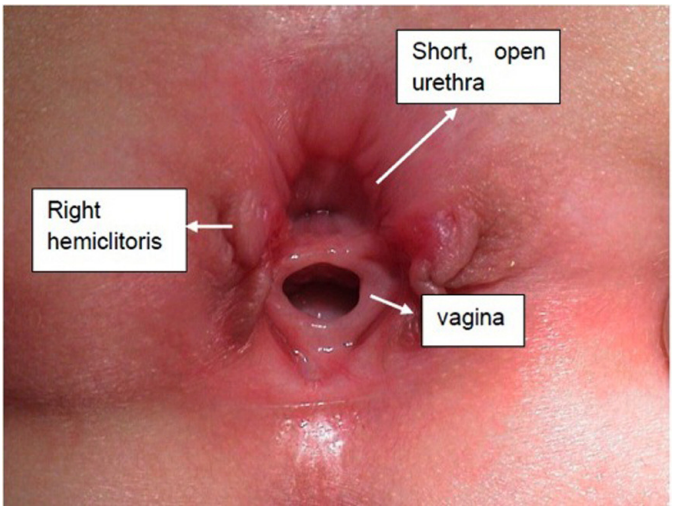

Fig.1: preoperative appareance of external genitalia shows female epispadias without bladder exstrophy

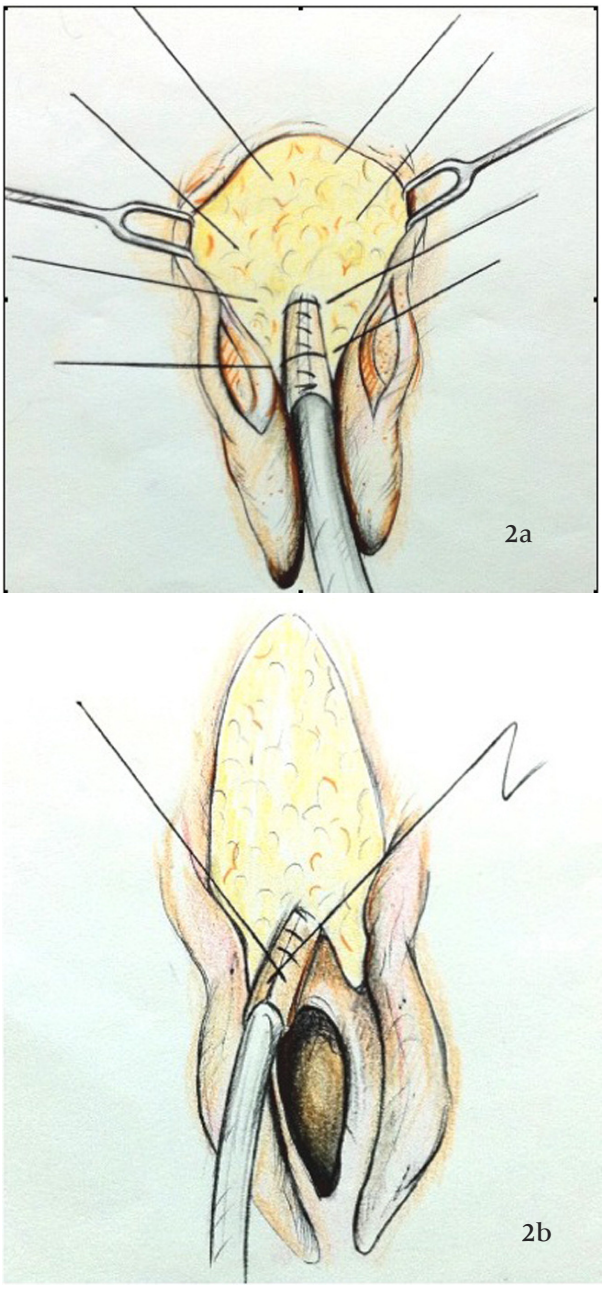

Fig.2(a-b): classical surgery

competent bladder neck. The small capacity (60-70 cc) non-inflamed bladder demonstrated a single normally placed ureteric orifice on each side. There were no other anomalies and all other systems were normal

At a single stage reconstructive procedure, the urethral plate, bladder neck and bladder were carefully dissected free and mobilized at a natural plane close to the pubic bones, liberating all available possible sphincteric muscle anteriorly and laterally. The urethral plate and bladder neck were tubularized with interrupted 5.0 Vicryl absorbable sutures around an 8 Fr silicone catheter, and the bladder neck was further reinforced by approximating the available 'sphincteric muscle' in the midline (Fig 2 a,b). The hemiclitori (glans and corpora) were freshened medially and approximated in the midline at the normal site above the urethral orifice. The procedure was completed with a cosmetic reconstruction of the external genitalia. A supra-pubic cystostomy was placed, and maintained with antibiotic cover for 3 weeks. A pelvic osteotomy was not performed.

The child healed well without any postoperative complications. Assessment under anaesthesia 2 weeks postoperatively confirmed easy passage of a 6 and 8 F Foley catheter through a good length patent urethra and bladder neck with no stenosis or dehiscence. On the 20th postoperative day an US study with a clamped suprapubic catheter showed bladder filling and voluntary micturition with no recordable residual volume and no uretero-pelvicalyceal dilatation. The suprapubic catheter was removed 24 hrs later. The child was discharged one month after operation, at which time she was observed to be completely dry for periods of up to 70 mins. Her mother reported that the child would express a sensation of a full bladder and a desire to micturate, with an ability to hold her urine and to initiate micturition voluntarily with complete bladder emptying and no residual urine. At 6 months follow-up her continence had improved and she could be dry for at least 2 hours. She had been potty trained and was using knickers by day and the nappy by night. Her bladder capacity was assessed by US and clinically at about 100 ccs. There had been no urinary infections and there was no upper tract dilatation. The cosmetic appearance of the vulva was considered to be acceptably normal.

DISCUSSION/CONLUSIONS

Although epispadias lies at the less severe end of the exstrophy-epispadias complex, it is nonetheless a fundamental and complicated developmental anomaly often associated with complete urinary incontinence. The condition is commoner in the male, and female epispadias is indeed rare, with an incidence of 1:480,000 girls. The aetiology of this developmental anomaly of the urethra and

bladder neck remains uncertain and is considered possibly to be the result of a combination of several genetic and environmental factors (3). Associated anomalies are commonly confined to the urinary tract with an incidence of vesico-ureteral reflux at 30%-75% (3), to the pelvis, the pelvic floor, and the abdominal wall but it is relevant also to assess the spine and the anus (4).

The physical findings in female epispadias are characteristic and well described such that the anomaly can and should be diagnosed immediately at birth on routine informed clinical genital examination, but is often missed presenting at a later age with failure to potty train and constant wetting. Early diagnosis allows early parental counselling and the option of a planned surgical reconstructive procedure preferably within the natural time for the physiological development of urinary continence. The issue and relevance of pelvic osteotomy for isolated epispadias remains controversial however early diagnosis allows easier and more manageable pelvic surgery that is possible within the first months of life but becomes more complex at an older age and once the child is attempting to stand.

The main objective behind surgery for epispadias is voluntary urinary continence, and to this end it is particularly relevant to mobilize carefully all available tissue anteriorly and laterally, minimizing injury to nerve plexi and potential sphincteric muscle in the area. The urethral plate and muscle, and the bladder neck and sphincteric muscles are tubularized and united without tension in the midline anteriorly to create a normal length urethra and bladder neck of normal diameter. Effective suprapubic diversion is relevant to avoid pressure disruption of healing tissue in the first days after surgery. Although multistage procedures were favoured historically, it is now considered preferable to attempt correction of the associated vulval and pubic anomalies at the same time within a single reconstructive event. At first operation there is the advantage of clean surgical dissection planes without scarring from previous surgery and therefore a greater likelihood of avoiding nerve and sphincteric muscle injury. The hemiclitori can be carefully mobilized and brought closely together, also reconstructing a prepucial hood and achieving closer apposition of the labia majora and the tissues of the mons pubis towards a more normal appearance of the vulva. Indeed the literature suggests that single stage reconstruction is safe and aesthetically acceptable, with a 60% and 87.5% (4,5) chance for urinary continence.

In our management of this child we have attempted to replicate the best practice of the day, reconstructing the urethra and bladder neck with the least possible injury to surrounding structures, and with vulval reconstruction for a good aesthetic appearance. We were gratified by good primary healing without complications, and particularly with the early suggestion of good bladder sensation and urinary continence. Interestingly within the first weeks of surgery the mother reported that the child expressed a sensation of bladder fullness and a desire to pass urine. She demonstrated from early on an ability to hold her urine, to initiate micturition and to void without leaving any residual urine. Although this is only a single case report with a relatively short follow-up, the immediate result, together with the absence of urinary infection or uretero-pelvicalyceal dilatation, supports the concept of a careful single stage reconstruction with mobilization and least possible trauma to crucial nerves and sphincteric muscle around the urethra and bladder neck. The child's response with early bladder sensation and the ability to hold and voluntarily void completely, have been gratifying and would suggest the likelihood of long term effective urinary control and normal continence.

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