Female Epispadias Repair

Female epispadias repair: a new 1-stage technique

Tom P.V.M de Jong, Pieter Dik and Aart J. Klijn

Abstract

Purpose: Female epispadias is a rare anomaly. According to the literature it is usually treated with staged procedures, including bladder neck reconstruction, to achieve continence.

We developed a 1-stage surgical technique that offers the possibility of achieving continence and a cosmetically normal appearance of the vulva.

Materials and Methods: We treated 4 patients 4 months to 8 years old. The main point of the technique is to free the urethral plate and bladder neck completely from surrounding tissues. After tubularizing the urethral plate into a urethra modified needle suspension brings the bladder neck and proximal urethra into the intra-abdominal position.

The pelvic floor is then reconstructed between the anterior vaginal wall and urethra. Thus, continence may be attained by intra-abdominal positioning of the bladder neck and proximal urethra as well as by pelvic floor reconstruction.

Results: Of our 4 consecutive cases of primary untreated epispadias, the technique proved successful in 3, while followup is too short in 1. One patient is completely dry and voids without a further procedure. Postoperatively 2 patients with 5 years or more of followup required injection of a bulking agent at the bladder neck level to achieve continence, including 1 who is damp during the day without the need to change clothes and 1 on clean intermittent catheterization twice daily because post-void residual urine volume causes recurrent urinary tract infection.

Conclusion: The described technique is promising for treating this disabling anomaly.

Introduction

Female epispadias is a rare anomaly, occurring in 1/150,000-300,000 live female births. In the Dutch population this value predicts 1 newborn patient every 2 to 3 years. The anomaly is a type of exstrophy-epispadias complex. Generally patients present because of abnormal external genitalia with a wide anterior gap between the labia majora and a 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 (fig 1). No urethra and no sphincteral mechanism is present, resulting in complete incontinence that is often accompanied by bladder capacity that is relatively small for age. When overlooked, after birth, female epispadias may present as severe incontinence. Sometimes the condition is wrongfully considered a manifestation of intersex, resulting in a delayed diagnosis and unnecessary psychological distress for the family.

Historically, treatment consists of staged repairs with urethral and vulvar reconstruction at stage 1, followed by bladder neck reconstruction at a later age. These procedures often do not have optimal results with respect to urinary continence. Moreover, clean intermittent catheterization and/or a continent catheterizable stoma on the often augmented bladder

Figure 2: Female epispadias. A normal hymenal ring, anteriorly the bladder neck, a bifid clitoris and the urethral plate spread out anteriorly between the halves of the clitoris.
is needed in many cases. We developed a 1-stage procedure that results in a competent urethra, functioning sphincteric mechanism and intra-abdominal bladder neck, providing an optimal chance for primary continence and normal voiding.

Patients and Methods

During an 8-year period corrective surgery was performed in 4 females 4 months to 8 years old with primary complete epispadias, including 3 who presented with intersex and 1 with therapy resistant incontinence. Preoperatively all 4 patients underwent ultrasound of the urinary tract, voiding cystourethrography and a urodynamic study. One patient had bilateral vesicoureteral reflux and 1 underwent emergency surgery at the age of 4 months because of recurrent bladder prolapse. In 3 cases the timing of surgery was elective, while in 1 referred immediately after birth we chose to perform the operation at the age of 10 months. Two girls referred late underwent surgery after urinary tract evaluation. The procedure began with cystoscopy and colposcopy, followed by excision of the urethral plate between the clitoral halves, leaving the urethral plate attached to the bladder neck (fig 2, A). The bladder neck was then completely freed from attachments to the anterior vaginal wall and the symphysis up to the level of the trigone, resulting in complete mobility of the bladder outlet and attached urethral plate. The urethral plate was tubularized into a urethra (fig. 2, B and C). The bladder neck and proximal urethra were brought into the intra-abdominal position by a modified needle suspension technique. Four 16 gauge intravenous cannulas were passed from 2 small inguinal incisions retropubically into the operating field. A polypropylene suture was fixed at the level of the bladder neck at the 3 and 9 o’clock positions. Each end of the suture was passed upward through the intravenous cannulas into the inguinal region. The intravenous cannulas were withdrawn, and the sutures tightened and knotted at the external rectus abdominis fascia, bringing the bladder neck and proximal urethra into the intra-abdominal position with the extended neourethral orifice at the anterior side of the hymenal ring. Subsequently
Figure 2: A. The urethral plate is freed from surrounding skin. The plane between the anterior vaginal wall and the bladder is developed. The bladder neck is freed completely from surrounding tissues, especially the pelvic floor, to ensure free mobility. B. The urethral plate is tubularised around a 10 Fr. silastic tube with interrupted absorbable sutures. C. Sagittal view of the situation after tubularisation of the urethra and mobilisation of the bladder neck. D. Intravenous cannulas, 2 on each side (or 1 with adapted technique) are passed through 2 small suprapubic incisions to the perineal wound, taking care to pass through the gap made in the pelvic floor without penetrating the bladder. After endoscopic inspection to make sure that the bladder has not been perforated by the cannulas, a suture is passed through the bladder neck and both ends are back-fed through the cannulas. Subsequently the cannulas are removed. By pulling the sutures the bladder neck is brought into an intra-abdominal position. The sutures are knotted upon the fascia of the external abdominal muscle. E. Sagittal view. The pelvic floor is closed between the anterior vaginal wall and the urethra. When necessary, in cases with a split symphysis, the pelvic floor is closed anteriorly as well. These sutures are very important. This is what makes the patient continent. F and G The urethra is sutured to the hymenal ring posteriorly, to the surrounding vulvar tissues medially and anteriorly. The clitoridal halves are denuded medially and fixed together with 7.0 absorbable sutures. The abundant tissue between the labia majora is excised, bringing the labia together in the mid-line.
the pelvic floor was identified visually left and right of the urethra, and closed in the midline between the anterior vaginal wall and the urethra (fig 2, D and E). These sutures make continence possible. In 1 patient with a large gap between the pubic bones complete approximation of the pelvic floor was not possible in the midline behind the bladder neck, resulting in a 5 mm. gap between the muscles. 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 external genital reconstruction. The clitoral halves were fused by denuding them of medial epithelium and connecting them with 7 or 8-zero polyglycolic acid sutures. The anterior distance of the labia majora was decreased by excising a lozenge of skin between the labia and suturing the defect longitudinally (fig. 2, F and G). Postoperatively bladder drainage was maintained for 10 days with a 10Fr transurethral single lumen catheter without a balloon. All 4 patients were on antibiotic prophylaxis during the 10-day period, consisting of 2 mg./kg. trimethoprim once daily and 0.4 mg./kg. oxybutinin daily given in 3 doses. One patient with bilateral grade 2 to 3 vesicoureteral reflux underwent simultaneous ureteral reimplantation through a separate transverse incision. Blood loss was negligible in all cases.

Results

Convalescence was unremarkable in all patients. When voiding, 2 patients had, temporarily had insufficient bladder capacity for age, producing urge, stress and overflow urinary incontinence. They eventually required bladder neck injection therapy with silicone granules in povidone. Technically bladder neck injection with 2 ml. of bulking agent at the 6 o’clock position was not more difficult than injection therapy for other indications. We routinely position the needle in the bladder neck by suprapubic puncture under direct vision via the transurethrally introduced cystoscope. Each patient achieved sufficient bladder capacity for continence. One girl is on clean intermittent catheterization twice daily to prevent urinary tract infection and 1 suffers dampness during sports without the need to change clothes. One patient who underwent surgery at age 10 months is
continent and voids normally since the operation. Urodynamic and radiological studies of the lower urinary tract were completely normal. The remaining patient has a follow-up too short to be conclusive at age 10 months. Three patients had no morbidity after surgery, while 1 had recurrent urinary tract infections. The tabel shows urodynamic results.

Vesicoureteral reflux was graded on a scale of 0 to 5, and compliance was considered normal when bladder capacity in ml. divided by bladder pressure in cm. water was greater than 10 at the end of the filling phase. Cosmesis is excellent in all cases (fig. 3). Followup ranges from 6 months to 6 years.

**Figure 3:** Photograph 6 months after reconstruction. The result is a completely normal appearance of the external genitalia, a competent bladder neck and urethra in an intra-abdominal position and a functioning pelvic floor surrounding the proximal urethra.
Discussion

Few reports exist in the literature of relatively large series of female epis-padias with 6 to 12 patients collected for many years. Welch included 50 females in a review article. Kramer and Kelalis described 12 patients, including 10 who were completely continent after bladder neck recon-struction. Hendren reported on 6 patients treated with an abdominoperineal approach that included bladder neck reconstruction, of whom 3 were continent. Gearhart et al described 11 females who underwennt staged reconstruction, including bladder neck reconstruction in the major-ity, in 3 of whom previous procedures failed. Five patients were completely dry, 3 were dry up to 3 hours during the day and 1 was dry for 1 to 3 hours, resulting in an overall daytime continence rate during of 87.5%. Mollard et al reported on 10 patients, including 8 with complete continence and some leakage in 2. Four patients under-went staged repair and 6 an unstaged procedure consisting of genitoplasty combined with bladder neck reconstruction.

To achieve continence an intra-abdominal position of the bladder neck and proximal urethra is mandatory. The previously described techniques from the literature achieve this goal by bladder neck reconstruction that extends the urethra into the bladder. These methods do not use the complete length of the urethral plate to construct a competent urethra. The 1-stage technique that we developed has certain advantages. No major bladder neck surgery or ureteral reimplantation is needed in the absence of vesicoureteral reflux. The bladder neck and proximal urethra are brought into the intra-abdominal position by a relatively simple procedure that provides the optimal chance for primary continence without hamper-ing possible subsequent bladder neck reconstruction, when needed. However, long-term results cannot be predicted in this extremely rare anomaly with severe consequences to the continence mechanism. The rar-ity of the abnormality prohibits evaluating the efficacy of surgical inter-vention in large groups of patients.
Conclusion

Our newly developed procedure involving 1-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 promising for treating female epispadias. More experience is needed to prove the superiority of this technique for this rare anomaly. Recently this method was adopted in several pediatric urological centers, helping to prove its value for treating these difficult cases. Moreover, the technique has been tried by others as a secondary procedure in girls with epispadias after primary exstrophy closure. The results of these cases are not yet available.
Literature