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Epispadias in boys with an intact prepuce



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Abstract *Objective:* To present an overview of the clinical presentation and pathological anatomy, and the results of surgical correction of 7 cases of epispadias with intact prepuce; a rare condition that has only occasionally been reported in literature.

Patients and methods: A retrospective search was performed in the surgical and diagnoses database between 1991 and 2011. Seven cases of epispadias with intact prepuce were identified. Five presented as a webbed and buried penis, 1 as phimosis and 1 with suspicion for congenital anomaly of the genitalia.

Results: In 3 of 7 cases, epispadias was suspected or diagnosed at first presentation and could be surgically corrected in the first intervention. In the other 4 cases, epispadias was discovered during surgery, requiring an additional intervention to perform epispadias repair in 3 cases. One boy was diagnosed with glandular, 3 with coronal, 1 with shaft and 2 with penopubic epispadias. Epispadias repair was successful with regard to cosmesis and erectile function. Five patients developed normal continence after surgery, 1 after intensive urotherapy. An under average penile length was the main reported problem during follow-up.

Conclusion: In the diagnostic process for a concealed penis, the possibility of epispadias should be considered. If epispadias is suspected or confirmed, epispadias repair can occur in the first intervention, reducing the number of additional interventions. Epispadias with intact prepuce appears to have a better prognosis concerning urinary continence compared to classical epispadias.

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Abbreviations: VUR, vesicoureteral reflux; VUDS, video urodynamic study; Pt. no., patient number.

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Introduction

Epispadias is part of the exstrophy–epispadias complex, a spectrum of diseases including exstrophy variants, cloacal exstrophy, classic bladder exstrophy and epispadias [1]. Isolated epispadias without the presence of exstrophy is considered the least severe defect of the exstrophy–epispadias complex. It is a rare congenital anomaly and is seen in approximately 1 in 120,000 male births [2]. Epispadias does not involve the body of the bladder or the hindgut but does affect the urethra, and can affect the bladder neck, sphincter function and the pubic symphysis [3]. Bladder function may be affected because of low urethral resistance during early development.

Isolated epispadias can be differentiated into glandular, coronal, shaft or penopubic epispadias. In distal epispadias, incontinence is rarely seen. In penopubic epispadias, the urethral meatus extends to the membranous urethra and bladder neck; sphincter insufficiency often may coexist [4].

In cases of isolated epispadias the prepuce is usually absent on the dorsal side of the penis and hangs as a tag of redundant tissue on the ventral side, leaving the glans uncovered [5,6]. It is very uncommon for epispadias to present with an intact prepuce; to date only 8 cases have been reported in 7 literature reports [5–11]. At first presentation, the diagnosis is easily overlooked, as the epispadias is not directly visible. Based on specific clinical signs, such as a broad, spade-like glans and a dorsally directed preputial opening and urinary stream, suspicion for epispadias may persist. In addition, a gap between the corpora cavernosa may be palpated. Furthermore, dorsal chordee and abnormalities of the penile raphe have been reported.

In our center, 7 of such cases with intact prepuce presented between 1991 and 2011. In this article we provide an overview of the clinical presentation and pathological anatomy, and the results of surgical correction.

Methods

A retrospective search for epispadias in males in the surgical and diagnoses databases of 2 large academic medical centers identified 25 cases of isolated epispadias. Seven of these cases presented with an intact prepuce. Epispadias was classified according to the position of the urethral meatus; glandular, coronal, on the shaft of the penis or penopubic.

Surgical technique

Repair of epispadias aimed to reconstruct the epispadiac urethra and glans, and straighten the penis with correction of dorsal chordee. Outcome parameters were reconstruction of the penis with a satisfactory appearance, maintenance of erectile function and creation of urinary continence.

The severity of the epispadias determined the technique that was used to reconstruct the penis. In cases of coronal, shaft or penopubic epispadias, the urethra was positioned between the 2 corpora cavernosa using the Cantwell–Ransley technique. The penile shaft skin was degloved from the coronal sulcus up to the base of the

penis in penopubic epispadias, or in distal epispadias, depending on the position of the urethral meatus. The urethral plate was dissected from the corpora cavernosa, leaving the distal end attached to the glans. Both corporal bodies were dissected free from the pubic bone to the glans. The urethral plate was tubularized over a 12 or 14 Fr catheter. The corpora were brought together over the urethra, placing the urethra in a ventral position, and were joined dorsally by non-absorbable sutures. Dorsal chordee, if still present, was corrected with plication of the tunica opposite to the curvature. In cases of glandular epispadias, the so-called Ipgam procedure was sufficient to achieve a cosmetically good result [3].

Penile lengthening was performed in cases where fixation of the corpora to the anterior side of the pubic bones appeared to be insufficient. Surgical procedure consisted of stretching the penis and fixation of the corpora cavernosa to the anterior side of the pubic bones with non-absorbable sutures, left and right of the symphysis. For cosmesis, redundant presymphyseal fat was removed and dorsal skin lengthened.

Urodynamic study

To diagnose vesicoureteral reflux (VUR) and to have insight into bladder function, sphincter function, bladder capacity and patency of the bladder neck, our routine is to perform a video urodynamic study (VUDS) in most cases of epispadias.

Case reports

Patient characteristics concerning interventions, age, urodynamic findings and the results of surgical correction are listed in Table 1. In addition to Table 1, several notable characteristics are described below.

Additional remarks

Patient number (Pt. no.) 1 was diagnosed with glandular epispadias. Although a distal epispadias was present, this was the only patient with insufficient bladder and sphincter function at VUDS. Following epispadias repair, the patient was constipated and incontinent for urine, mainly during stressful situations. Subsequently laxatives as well as cognitive and biofeedback urotherapy were prescribed. Bladder neck reconstruction was not indicated.

Pt. no. 2 was diagnosed with coronal epispadias. It was decided not to perform VUDS in this patient, since continence was achieved before the age of 4 years and the bladder neck appeared to be closed on ultrasound images.

Pt. no. 3 and pt. no. 6 were diagnosed with penopubic epispadias. A minor sphincter abnormality was seen in both patients during cystoscopy (performed during epispadias repair). It is notable that the bladder neck was considered sufficient at VUDS in these patients. Pt. no. 3 was potty-trained and became continent, while pt. no. 6 is under the age of 5 years and is being potty-trained. Figs. 1 and 2 show the aspect of the penis of pt. no. 3 before surgery and after retraction of the prepuce.

Pt. no. 5 was diagnosed with shaft epispadias and was referred to our center at the age of almost 4 years.

Table 1 Patient characteristics, clinical presentation, diagnosis, results of surgical epispadias repair and results of video urodynamic study in males with intact prepuce.

	Pt. no. 1	Pt. no. 2	Pt. no. 3	Pt. no. 4	Pt. no. 5	Pt. no. 6	Pt. no. 7
Clinical presentation	Buried penis	Webbed penis	Webbed and buried penis, ballooning of prepuce during micturition	Buried penis, phimosis, suspicion of epispadias	SE	Phimosis, GE	Webbed and buried penis, dorsal chordee
Diagnosis	GE	CE	PE	CE	SE	PE	CE
Postoperative FU in y	8.4	1.8	3.9	13.2	0.3	1.3	2.1
Epispadias repair in 1st intervention	No	No	No	Yes	Yes	Yes	Yes
Indication for add. interventions during LTFU	Penile length, stress incontinent (dysfunctional voiding)	Wide urethral meatus, penile length, webbed penis	Penile length	Buried penis	Recently operated, LTFU NA	None	Buried penis
Surgical interventions (age in y)	1. Intended circumcision (1.3) 2. Epispadias repair and Z-plasty (1.7) 3. Z-plasty dorsal shaft skin (5.8)	1. Preputioplasty (3.5) 2. Epispadias repair (4.9) 3. Urethral meatoplasty, penile lengthening procedure, correction of scrotal webbing (6.2)	1. Intended preputioplasty (0.6) 2. Epispadias repair (0.9)	1. Epispadias repair (1.2) 2. Ventral skin plasty (4.9) 3. Penile lengthening procedure, ventral skin plasty (14.3)	1. Epispadias repair (6.2)	1. Epispadias repair (1.4)	1. Epispadias repair (1.1) 2. Penile lengthening procedure, circumcision (1.9)
No of surgical interventions	3	3	2	3	1	1	2
Pubic diastasis (in cm)	Closed	Closed	Closed	0.5	Closed	0.7	Closed
VUDS	Open bladder neck and insufficient bladder function, VUR R0L1	Not performed	Normal bladder and bladder function, no VUR	Normal bladder neck and bladder function, no VUR	Normal bladder neck, overactive bladder, VUR R1L1	Normal bladder neck and bladder function, VUR R2L3	Normal bladder neck and bladder function, VUR R2L2
Bladder capacity in ml (PBC based on age in %)	275 (92)	NA	40 (80)	205 (205)	182 (85)	95 (95)	135 (169)

(continued on next page)

Table 1 (continued)

	Pt. no. 1	Pt. no. 2	Pt. no. 3	Pt. no. 4	Pt. no. 5	Pt. no. 6	Pt. no. 7
Continence	Continent (After hospitalized urotherapy)	Continent	Continent	Continent	Continent during day, incontinent at night	< 5 y is being potty-trained	Continent
Penile length in cm (age in y)	5 (5.5) Treated with dihydrotestosterone gel	4.5 (4)	5 (4.2) Treated with dihydrotestosterone gel	6 (14.3)	6 (6.2)	3.5 (1) Treated with dihydrotestosterone gel	5.5 (3.3)
Penile length (SD) [12]	-1 SD	< -1 SD	-0.5 SD	< -2.5 SD	0 SD	< -1 SD	0 SD

Abbreviations: Pt. no.: patient number, GE: glandular epispadias, CE: coronal epispadias, SE: shaft epispadias, PE: penopubic epispadias, FU: follow-up, y: years, LTFU: long-term follow-up, NA: not available, VUDS: video urodynamic study, VUR: vesicoureteral reflux, R: right, L: left, PBC: predicted bladder capacity in ml, based on the formula (patient age × 30) + 30, SD: standard deviation.



Figure 1 Aspect of the penis before surgery in pt. no. 3.

For psychological reasons, surgery was postponed until the age of 6 years. Surgery has recently been performed; therefore no long-term follow-up data is available in this patient and continence has not yet developed. A normal bladder neck was seen at VUDS; however during cystoscopy (performed during epispadias repair) the bladder neck was observed to be open. Figs. 3 and 4 show the aspect of the penis of pt. no. 5 before surgery and after retraction of the prepuce.

Results

In 3 of 7 patients described in this series epispadias was suspected or diagnosed at first presentation. In 4 of 7 cases epispadias was discovered during surgery, and in 3 of these reconstructive surgery was postponed for logistical reasons such as operative scheduling and the requirement for informed consent, and a reconstruction was planned electively (pt. no. 1, 2, 3). The primary indication for surgery is described in Table 1 in the Surgical interventions section. In the cases with suspected or diagnosed epispadias, the epispadias repair could take place in the first intervention

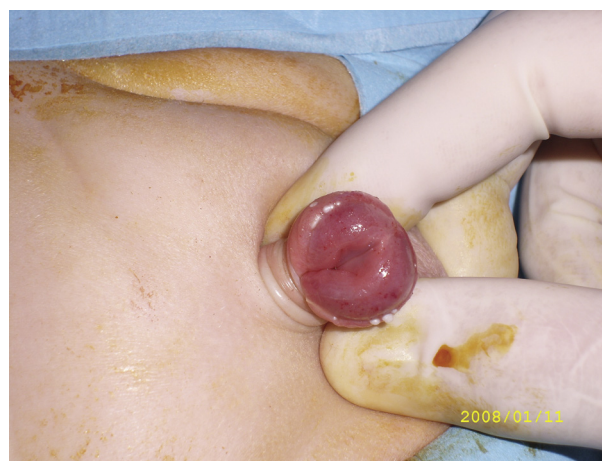


Figure 2 Penopubic epispadias was seen after retraction of the prepuce in pt. no. 3.



Figure 3 Aspect of the penis before surgery in pt. no. 5.

(pt. no. 4, 5, 6). One boy with unsuspected epispadias underwent epispadias repair in the first intervention due to consultation with his parents during surgery (pt. no. 7).

During surgery, following rotation and fixation of the corpora, 6 out of 7 operated cases had a straight penis during artificial erection. One patient required plication of the corpora because of asymmetry of corpora and persistent curvature after rotation (pt. no. 7). Shortly after surgery, no complications occurred; there were no fistulas, excessive chordee or erectile dysfunction.

Four boys had low-grade VUR at VUDS requiring no specific treatment; reflux was bilateral in 3 boys and unilateral in 1 boy.

The functional results of epispadias repair, in terms of reconstruction of the penis with maintenance of erectile function (by parental observation) and development of urinary continence, were considered to be successful in all patients. No information is available about sexual functioning and erectile function after puberty since all patients are yet to complete puberty. Five boys developed normal continence and were able to urinate while standing, one patient after additional urotherapy (pt. no. 1).



Figure 4 Shaft epispadias was seen after retraction of the prepuce in pt. no. 5.

Continence has not yet been achieved in one patient who has recently been operated (pt. no. 5), and one patient who is under the age of 5 years (pt. no. 6).

At long-term follow-up, most patients were dissatisfied with cosmesis, mainly based on short penile length according to parents' or patient's opinion. Four patients required additional surgery after epispadias repair (pt. no. 1, 2, 4, 7) to improve the aspect of the penis because of small size. Two patients did not require further surgery after epispadias repair (pt. no. 3, 6). Three patients were treated with dihydrotestosterone gel to enhance penile length (pt. no. 1, 3, 6). Long-term follow-up data was not available for pt. no. 5, since epispadias repair has been performed recently.

Discussion

The understanding of embryology of the male urethra and prepuce is not complete. However, there appears to be a correlation between the development of the urethra and the prepuce. When a developmental defect of the urethra occurs, for example in cases of hypospadias or epispadias, the prepuce usually fails to develop on the corresponding side [5]. The pathogenesis of malformations of the exstrophy–epispadias complex is frequently explained by defective development of the cloacal membrane (e.g. premature rupture or wedge effect). According to this explanation, one would expect to find a bifid penis or clitoris in every affected baby (Männer J 2012, pers. comment, 14 June). In our patients, the prepuce was developed completely albeit not normally; 6 out of 7 patients presented with a concealed penis or with phimosis. Similarly, subtypes of hypospadias with complete prepuce have been reported in the literature and approximately 1% of hypospadias cases appear as a megalomeatus or a coronal meatus with intact prepuce.

The embryogenesis of the urethra is said to begin during the second month of intrauterine life when the cloaca subdivides into a posterior portion, the anorectal canal, and an anterior portion, the primitive urogenital sinus [1]. The urogenital sinus is the precursor of the bladder, proximal prostatic urethra and the membranous urethra. The most caudal aspect of the cloaca, the phallic cloaca, extends distally through the developing genital tubercle. Proliferation of the genital tubercle displaces the cloaca in a way that it is situated on the caudal aspect of the developing glans. Failure to complete this step results in epispadias [13].

The formation of the prepuce begins during the third month of intrauterine life as the genital tubercle is proliferating. Following median cleavage of the urethral plate, two sets of tissue folds develop on the ventral surface on either side of the urethral groove. The medial endodermal folds fuse in the ventral midline to form the urethra. The more lateral ectodermal folds fuse over the developing urethra to form the penile shaft skin and the prepuce. A ring of ectoderm forms just proximal to the developing glans penis. This skin advances over the corona of the glans and eventually covers the glans entirely as the prepuce or foreskin [10]. Others say that the prepuce is formed by a combination of preputial folding and the ingrowth of a

cellular lamella. This ingrowth creates the prepuce, glans, corona and coronal sulcus mucosa [14]. Only a limited number of articles have been published on the development or embryology of the penile prepuce [15–19]. The embryology of the penile prepuce remains controversial ever since the first theories were reported, particularly in conditions that do not fit these theories, such as epispadias or hypospadias with intact prepuce.

McCahill et al. explain the phenomenon of epispadias with an intact prepuce with the active growth of mesenchyme between the preputial fold and the glandular lamella, which transports the fold distally until it covers the glans completely. If these folds appear proximal to the urethral defect, they cover the defective urethra as well as the glans. Thus, epispadias will not influence the development of the prepuce in these cases [8]. However, it seems that cases of proximal epispadias (e.g. penopubic epispadias) cannot be explained by this theory.

Although various other hypotheses have been formulated, it remains difficult to explain the development of epispadias in combination with an intact prepuce. What we have seen in the described cases is that an intact prepuce can mislead the surgeon. Definitive epispadias repair was postponed in 3 of 7 cases, which made an invasive additional surgery necessary.

With regard to the outcome of epispadias repair, we believe that the combination of epispadias with an intact prepuce seems to be favorable compared to cases with classical isolated epispadias. Fewer complications, less additional interventions and better continence rates seem to be noted for the boys with epispadias and intact prepuce. In contrast to other cases of epispadias, where urinary incontinence based on bladder neck and sphincter incompetence often is an important issue, bladder neck insufficiency was seen in only 1 case in our series without persisting incontinence. Congenital curvature of the penis appears to be a relatively minor problem in epispadias with intact prepuce compared to normal epispadias. On the other hand, the results of penile lengthening procedures seem to be disappointing as a short penile length was coexistent in most cases of epispadias with concealed penis. We did not find any way to achieve better results in this regard. However, it is apparent that during the first surgery the utmost care should be taken to ensure that the corpora are firmly attached to the anterior level of the pubic bone to avoid the need for doing this in a secondary procedure.

In this series VUDS was performed in 6 out of 7 patients. In general, VUDS did not influence or change initial management; therefore one may discuss the need for such study. Low-grade VUR was seen in 4 of 6 patients in this series. Reflux is closely linked to bladder exstrophy [20]. It is also commonly seen in isolated epispadias, due to a deficient ureterovesical junction and lateralization of the ureteric orifices in the bladder so that reflux can occur more easily [21]. Incidence rates of reflux in isolated epispadias have been reported to be 30–75% in small patient groups [21,22]. It is important to monitor the degree of reflux because, in theory, VUR can aggravate post-operatively by enhancement of urethral resistance. This could accelerate damage to the upper urinary tract.

We cannot explain the remarkable number of cases of epispadias with intact prepuce, 7 patients with this condition

have been seen in 20 years. Underreporting of the condition may exist. For example, Perovic described 5 cases of epispadias in his book 'Congenital abnormalities of the external genitalia', and 2 of these cases appear to have a complete prepuce [23]. Epispadias with an intact prepuce is possibly not as uncommon as has been stated in the literature. This anomaly would not be easily overlooked in areas where the majority of males are circumcised; however in countries where most males are not circumcised, it may go undetected.

Furthermore, although the prepuce cannot always be retracted in the case of an intact prepuce and epispadias, this should not delay the diagnosis of epispadias. Certain clinical signs may assist the diagnosis of epispadias in concealed penises, as described in the introduction. Therefore, a thorough physical examination is required to reduce the number of additional surgical interventions and to enhance the outcome of surgical epispadias repair. This series demonstrates that patients undergoing epispadias repair in the first intervention did have a smaller number of open interventions (mean number of open interventions, 1.75) in contrast to those who did not undergo epispadias repair in the first intervention (mean number of open interventions 2.67).

Conclusion

In the diagnostic process for a concealed penis or phimosis, the possibility of epispadias should be considered. If epispadias is suspected or confirmed, epispadias repair can take place in the first intervention, reducing the number of additional interventions.

Conflict of interest statement

The authors have no conflict of interest or financial relationships to declare. No external funding was utilized.

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