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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.

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.