

## Case Report

# Concealed epispadias: a rare anomaly - case report and review

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### ABSTRACT

Epispadias with an intact prepuce (Concealed Epispadias) is an uncommon anomaly. The specific clinical signs in this situation are broad, spade-like glans with a dorsally directed preputial opening, gap between the corpora cavernosa may be palpable, dorsal chordee and abnormalities of the penile raphae. We report the case of a 3 year old male child with Concealed Epispadias who underwent a modified Cantwell-Ransley operation with good outcome. The relevant anatomy, embryology and surgical options are reviewed.

**Keywords:** Epispadias, Concealed, Intact prepuce, Cantwell-Ransley operation

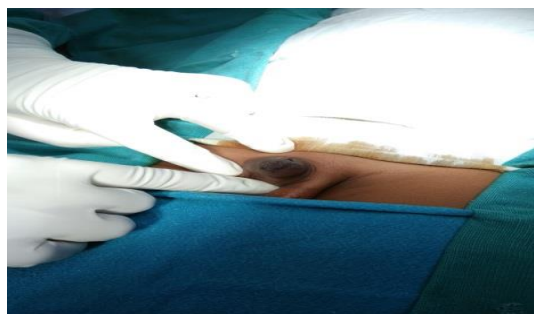
## INTRODUCTION

Isolated epispadias is the least severe defect of the extrophy-epispadias complex. It is an uncommon anomaly with incidence of 1 in 120,000 male births.<sup>1-3</sup> Isolated epispadias can be classified as glanular, coronal, shaft or penopubic epispadias. In distal variants of epispadias, incontinence is uncommon. In penopubic epispadias, the urethral meatus extends to the membranous urethra and bladder neck and sphincter insufficiency may occur.<sup>4</sup>

In isolated epispadias, the prepuce is usually absent on the dorsal side of the penis leaving the glans uncovered. Prepuce is seen as a tag of redundant tissue on the ventral side. Epispadias with an intact prepuce (Concealed Epispadias) is an uncommon anomaly.<sup>5,6</sup> The diagnosis can be missed unless looked for specifically. The specific clinical signs in this situation are broad, spade-like glans with a dorsally directed preputial opening, gap between the corpora cavernosa may be palpable, dorsal chordee and abnormalities of the penile raphae.<sup>5-7</sup> Variants of hypospadias with complete prepuce are also seen and about 1% of hypospadias present as megameatus/ coronal meatus with intact prepuce.

## CASE REPORT

A 3 year old male child was detected to have Concealed epispadias on clinical examination for non-specific urinary symptoms (Figures 1-3). The child had penopubic epispadias with intact prepuce. The phallus was short with mild pubic diastasis, but there was no urinary incontinence. There was no other anomaly on clinical examination. An Ultrasound KUB was done, which was normal. The child underwent an elective Modified Cantwell-Ransley operation for correction of Epispadias (Figures 4 and 5).



**Figure 1: Epispadias with intact prepuce (concealed epispadias).**



**Figure 2: Penopubic epispadias seen on retracting the prepuce.**



**Figure 3: Broad, spade like glans with no significant dorsal chordee.**



**Figure 4: Epispadias, post repair (modified Cantwell-Ransley operation).**



**Figure 5: Postoperative appearance of the phallus.**

The postoperative period was uneventful and silicone urethral catheter was removed on tenth day after surgery. Post operative outcome was good with no complications like residual dorsal curvature, fistula or incontinence. The child is doing well on follow up.

#### **DISCUSSION**

The development of the urethra and the prepuce are related to each other. When a developmental defect of the urethra occurs (as in hypospadias or epispadias), the prepuce usually fails to develop on the corresponding side.<sup>1,2</sup> The pathogenesis of malformations of the exstrophy-epispadias complex is explained by defective development of the cloacal membrane (premature rupture or wedge effect).<sup>1,2</sup> The embryogenesis of the urethra starts 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. 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 such a way that it is situated on the caudal aspect of the developing glans. Failure to complete this step results in the anomaly of epispadias.<sup>1,2</sup>

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.<sup>1-3</sup> 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. Another theory is 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.<sup>3-5</sup>

The occurrence of epispadias with an intact prepuce is explained by McCahill et al by the theory of active growth of mesenchyme between the preputial fold and the glandular lamella, which transports the fold distally until it covers the glans completely.<sup>4-6</sup> If these folds appear proximal to the urethral defect, they cover the defective urethra as well as the glans. Therefore, epispadias will not influence the development of the prepuce in these cases.<sup>5</sup> However, cases of proximal epispadias (penopubic epispadias) cannot be explained by this theory. Although various other hypotheses have been formulated, it is difficult to explain the development of epispadias in combination with an intact prepuce.

### **Surgical technique**

The aim of surgery is to reconstruct the epispadiac urethra and glans along with correction of dorsal chordee.<sup>7</sup> The severity of the epispadias determines the technique used for reconstruction. In coronal, shaft or penopubic epispadias, the urethra is positioned between the corpora cavernosa using the Cantwell-Ransley technique.<sup>8</sup> The penile shaft skin is 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.<sup>9</sup> The urethral plate is dissected from the corpora cavernosa, leaving the distal end attached to the glans. Both corporal bodies are dissected free from the pubic bone to the glans.<sup>7-9</sup> The urethral plate is tubularized over a 12 Fr catheter. The corpora are brought together over the urethra, placing the urethra in a ventral position, and are joined dorsally. Persistent dorsal chordee is corrected by plication of the tunica opposite the curvature. In cases of glandular epispadias, the IPGAM procedure (reverse MAGPI) is sufficient to achieve a good result. Another option is the Complete Penile disassembly technique proposed by Mitchell, where there is a need for having to create a hypospadias urethra. The result is assessed based on the cosmesis of reconstruction of the penis, maintenance of erectile function and achievement of urinary continence.<sup>7-9</sup>

Penile lengthening is done where fixation of the corpora to the anterior side of the pubic bones is insufficient. The procedure consists of stretching the penis and fixation of the corpora cavernosa to the anterior side of the pubic bones on either side of symphysis with non-absorbable sutures. The redundant pre-symphysis fat is removed and dorsal skin lengthened for cosmesis.<sup>8-10</sup> Complications include urethrocutaneous fistulas, skin or glans dehiscence, residual dorsal chordee, persistent short phallus and urinary incontinence.

Concealed epispadias may have a more favorable outcome compared to classical isolated epispadias. Fewer complications, less additional interventions and better continence rates are observed in Concealed epispadias. Compared to other variants of epispadias, where urinary incontinence due to bladder neck and sphincter incompetence, is a frequent issue, bladder neck insufficiency is rare in Concealed epispadias.<sup>7-9</sup> Congenital curvature of the penis appears to be a relatively minor problem in Concealed epispadias. Short penile length is an important issue in most cases of Concealed epispadias. During the primary surgery, it should be ensured that corpora are firmly attached to the anterior level of the pubic bone, to avoid the need a secondary procedure.<sup>8-10</sup>

### **CONCLUSION**

Concealed Epispadias may not actually be very uncommon among the anomalies of Exstrophy-Epispadias complex, but has a chance of being undetected and therefore under-reported, especially in regions where circumcision is not routine. Prompt recognition of the entity requires awareness, high index of suspicion and thorough clinical examination.

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