



Case Report

Managing a complex case of hypospadias cripple and chordee

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Abstract

Objectives. Hypospadias is a congenital affliction in which the urethral meatus is located on the underside of the penis. These cases are usually treated during childhood by pediatric surgeons, but more complex forms tend to reoccur, and after several failed attempts end up as hypospadias cripple in the care of urologists at an adult age. The aim of this paper is to present the management of a complex case of hypospadias cripple associated with penile curvature (chordee).

Materials and Methods. A 24 year old patient presented in our clinic with hypospadias cripple and ventral penile curvature. At the physical examination we found a penoscrotal urethral meatus, heavy scarring of the distal ventral penis and a ventral chordee of approximately 90 degrees in erect state. A two stage “Bracka” repair technique with buccal mucosa graft was chosen. The first stage of the surgery consisted of removing the scarred and defective distal urethral plate and the fibrotic tissue responsible for the penile curvature, preparing the corpora cavernosa for the graft, clefting the glans, harvesting two buccal mucosa grafts from both inner cheeks, and finally quilting the grafts on the defect. The second stage of the repair was performed after a period of about six months, and consisted in the tubularization of the matured graft and glans plasty.

Results and Conclusions. No immediate or late complications occurred after any of the two stages of the surgery; no significant ventral chordee was observed and no urethral fistula occurred till the one year follow-up. A two stage “Bracka” repair technique is best suited for treating these patients, correcting ventral chordee in the first session.

Keywords: hypospadias, cripple, chordee, penile curvature



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Introduction

The hypospadias is a congenital disease of the urethral and penile development in which the urethral meatus is ectopically located on the underside of the penis instead of its normal place at the tip of the penis. The term originates from the Greek words hypo, meaning below, and spade, meaning orifice, being first used by Galen in the 2nd century AD. And the first modern principles for the surgical treatment of hypospadias were defined in the mid-1800s by the German Thiersch and the French Duplay.

Hypospadias is the second most common birth defect in men. It has an incidence rate of about one case in 250 births (1). And in about 10% of the times it can be associated with other congenital defects (2, 3). The most frequent abnormality coupled with hypospadias was observed to be a ventral penile curvature, of various degrees, found in 14% of situations (4, 5). Another fairly common defect is an undescended testis, associated in approximately 3.7% of cases (6). The cause of this affliction is a failure of the urinary channel to mature and completely tabularize at the distal part of the penis, which occurs during the 13th week of gestation (7).

Depending on the level of the urethral opening, there are several types of hypospadias (Figure 1). The most frequent is the distal form, which represents 90% of the situations, and has the urethral meatus somewhere along the penis. The distal hypospadias can be further divided into anterior (60%) that comprises of glanular or subcoronal openings, and middle hypospadias (30%) which can be distal penile, midshaft or proximal penile. The rest of the 10% is represented by the proximal or posterior hypospadias, which consists of the penoscrotal, scrotal or perineal openings (1).

The treatment for hypospadias is surgery, meant to correct the defects and achieve a normal aspect and function of the penis. But there are as many as 300 to 400 different techniques and variations envisioned, so choosing the right method can be sometimes difficult. Therefore the decision should be taken on a case by case basis. This usually falls into the hands of pediatric surgeons, as the interventions are performed during childhood. But these physicians are supposed to be able to treat any kind of surgical problems of children, so they can be often untrained for the complex reconstructive operations required by more severe forms of hypospadias. This leads to a high reoccurrence rate and other often other complications, like urethral fistulae, urethral strictures or diverticulum, or penile curvature.

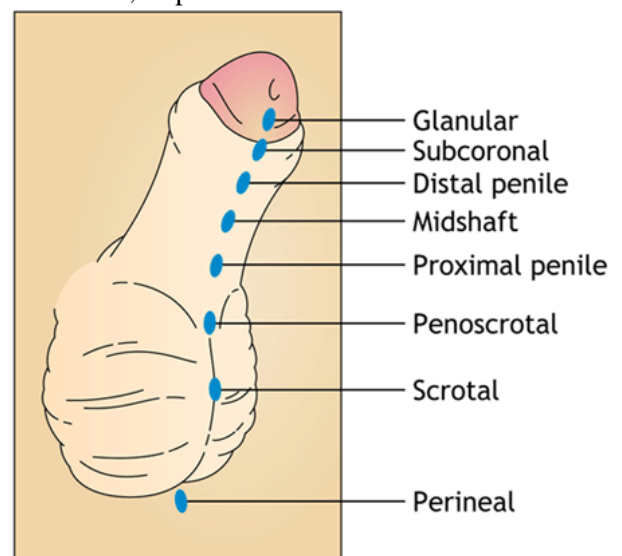


Figure 1. Types of hypospadias (8)

Therefore, in 1970 Horton defined the term of hypospadias cripple, referring to complex cases of hypospadias associated with various penile deformities, either from the get go, or after multiple failed repair attempts. These cases are usually treated by urologists skilled in penile and urethral reconstructive surgery.

The aim of this paper is to present the management of a complex case of hypospadias cripple associated with ventral penile curvature.

Case Report

A 24 year old patient presented in our clinic with hypospadias cripple and ventral chordee. He had already undergone six failed interventions to correct the problem during his early childhood, in different pediatric surgical clinics, and was only now referred in our care. At the physical examination we found a penoscrotal urethral meatus, heavy scarring of the distal ventral penis and a ventral curvature of approximately 90 degrees in erect state (Figure 2). A two stage “Bracka” repair technique with buccal mucosa graft was chosen, because we were dealing with a defective urethral plate (8, 9).



Figure 2. Hypospadias and false meatus

The first procedure was performed immediately, under general anesthesia, with the patient in a dorsal decubitus. In the beginning a tourniquet was placed at the base of the penis in order to reduce the blood loss (Figure 3). Then the urethral plate distal to the original meatus was dissected and the scarred and fibrotic tissue responsible for the penile curvature was removed (Figures 4, 5). And the glans was clefted on the ventral midline. This left a good grafting bed on the corpora cavernosa, about 6 cm in length. Two

buccal mucosa grafts were needed, harvested from both inner cheeks (Figures 6, 7). The grafts were then dorsally quilted on the corpora cavernosa, and a tie-over gauze was used to firmly hold them in place (Figures 8, 9).



Figure 3. Penile curvature - approximately 90 degrees



Figure 4. Dissecting the urethral plate and removing the fibrotic tissue



Figure 5. Reduced curvature in artificial erection

An 18 Fr Foley catheter was inserted, and suppressed after seven days, simultaneously with the tie-over gauze. The patient was discharged on the 8th day after surgery (Figure 10).

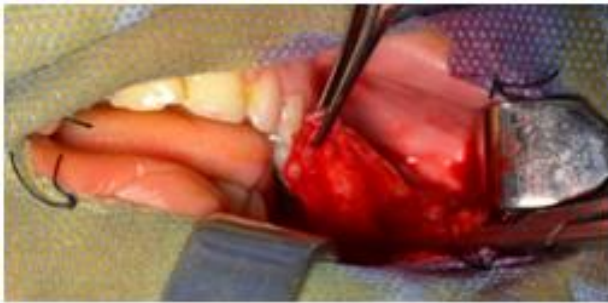


Figure 6. Harvesting the grafts from the inner cheeks



Figure 7. Preparing the grafts for placement



Figure 8. Placing the 2 buccal mucosa grafts

The second stage of the repair was performed after a wait period of about six months, in order for the graft to mature (Figure 11). It was also under general anesthesia, with the patient in a dorsal decubitus. A tourniquet on the base of the penis was again necessary to avoid unwanted blood loss. The matured graft was then dissected on the edges (Figure 12), and tubularized on an 18 Fr Foley catheter (Figures 13,

14). The suture line was covered with portions of the dartos fascia, and a glansplasty was carried out (Figure 15). At the end the skin was closed laterally to the neourethral suture, in order to minimize the risk of fistulae (Figure 16).

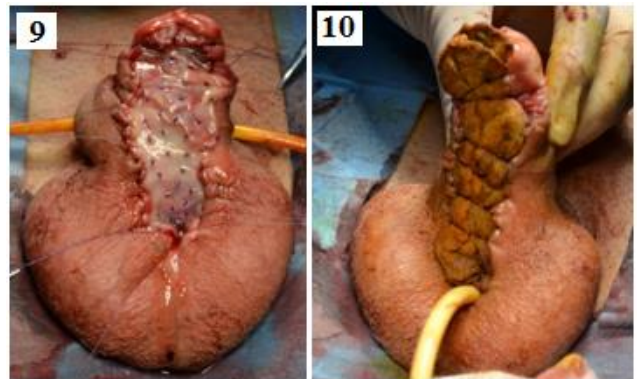


Figure 9. The grafts completely placed

Figure 10. Final aspect of the first stage - tie-over gauze



Figure 11. Hypospadias - matured aspect of the graft after 6 months

Figure 12. Dissecting the borders of the matured graft

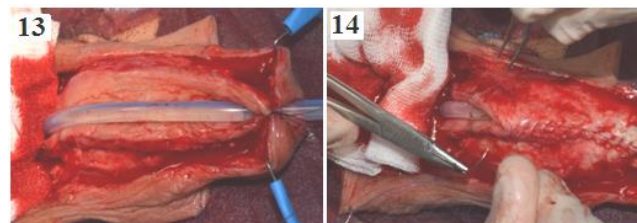


Figure 13. Inserting a Foley catheter

Figure 14. Closing the neourethra



Figure 15. Closing the dartos fascia

Figure 16. Final aspect of the second stage

Results

No immediate or late complications occurred after any of the two stages of the surgery. The patient was discharged on the 4th day, and the second 18 Fr Foley catheter was kept for three weeks after the tubularization. Afterwards the patient had normal micturition through its distal penile neomeatus.

No significant ventral chordee was observed and no urethral fistula occurred till the one year follow-up. The patient has a good quality of life, with increased self esteem and a rehabilitated sexual performance (Figure 17).



Figure 17. Voiding normally through the neomeatus

Discussions

After reviewing this case a few items should be up for debate. Firstly and most importantly is the choice of the surgical technique. The most common option in the hypospadias treatment a TIP repair technique (tubularized incised plate), which is used in distal forms of the disease, with success rates as high as 90% (10, 11). But this method can be performed only when we have a narrow but good quality urethral plate available. So it is not suited for hypospadias cripple cases, where there is no salvageable urethral plate distal to the original meatus. In these situations Bracka's two stage repair with the excision and substitution of the defective urethral plate is mandatory, as it is reliable, reproducible and versatile, and ensures good cosmetic results and a vertical slit-like meatus.

The second point of the discussions is who should perform these interventions. As the best moment to operate is during the early infancy and childhood, starting from three months of age, in order to avoid psychological trauma related to recalling the event, the pediatric surgeon is in the first line of the treatment (12). But because these physicians are supposed to be able to cure all surgical problems of children, they are often untrained enough for this type of complex urethral reconstruction. And reoccurrence can get quite high for the more difficult cases. Therefore these patients should be referred directly to skilled urologists that have enough experience with the urethral surgery.

And last but not least, we should keep in mind that hypospadias is not an isolated congenital defect. It can be associated with various other afflictions, in as high as 10% of cases (2, 3). And these numbers can be underestimating the reality because of lack of medical knowledge and information provided to the parents, and potentially no other signs or symptoms at the moment of diagnosis and treatment. Also while the most common problems associated to hypospadias are usually affecting the genitalia, and can be simultaneously solved, some conditions, seemingly unrelated, like the tetralogy of Fallot or a degree of mental retardation have been observed. So further testing of hypospadiac patients could prove in the early diagnosing and treating of other congenital afflictions (13, 14).

Conclusions

Hypospadias is a common congenital abnormality in men. Most of the cases are treated during childhood by pediatric surgeons, but the more complex forms reoccur as hypospadias cripples. As these represent the most challenging aspect of the urethral reconstructive urology, they should be handled only by specialized surgeons. A two stage "Bracka" repair technique is

best suited for treating these patients, correcting ventral chordee in the first session.

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