Two Stage Perineal Hypospadias Cripple Repair – case report

C. Gingu1,2, S. Ianiotescu1, A. Dick1, C. Baston1,2, M. Crasneanu1, A. Andresanu1, O. Himedan1, Mihancea A.1, L. Domnisor1, I. Sinescu1,2

1 Center of Urological Surgery and Renal Transplantation, Fundeni Clinical Institute, Bucharest, Romania
2 University of Medicine and Pharmacy Carol Davila, Bucharest, Romania

Abstract

Introduction and Objectives. Hypospadias, one of the most prevalent birth malformation of the male genitalia, consists of the insertion of the urethral meatus on the ventral side of the penis instead of the tip of the glans common associated with other penile defects and ventral chordee. It can vary in location from the glandular region (distal hypospadias) towards the perineum (proximal hypospadias). The later, although far less common, it is a lot more difficult to treat as it requires the reconstruction of the whole circumference and the full length of the urethra. The aim of this paper is to present the surgical management of a difficult case of perineal hypospadias cripple.

Materials and Methods. A 21 years old patient presented in our clinic with a perineal hypospadias, inadequately treated during his infancy and early childhood at a pediatric department, when the perineal segment of the urethra was created using a scrotal skin graft, while the penile segment was reconstructed from a prepuce and penile skin flap and left “a plat”. The physical examination revealed a perineal urethral meatus, with urethral calculi formed on hair grown inside the perienal segment. No ventral chordee was observed in this case. We decided on a two stage repair technique, spaced over a six months period. During the first stage the defective perineal urethra was completely removed, and two long buccal mucosa segments were harvested from both inner cheeks and grafted to the corpora cavernosa to replace the old scrotal skin graft. After the wait period of six months the matured buccal mucosa graft was tubularized in continuity with the prepuce flap and the neourethra was created. A 16 Fr Foley catheter was left in place for about three weeks.

Results. No immediate or late complications were observed after any of the two stages of the intervention. The patient was discharged 5 days after the grafting and 6 days after the tubularization. The Foley catheter was removed after 21 days and normal micturitions using the neomeatus were possible, voiding at 18 ml/sec.

Conclusions. Although hypospadias remains one of the most challenging problems of the urologist, even the more severe perineal cases can be successfully treated. A two stage repair is a versatile technique as it allows the use of buccal mucosa grafts in combination with prepuce and penile skin flaps to reconstruct of the whole circumference and full length of the urethra.

Key-words: hypospadias, cripple, perineal, urethra

Correspondence to: Assist. Prof. Dr. Catalin Baston M.D., Ph.D.
Fundeni Clinical Institute, Urology and Renal Transplantation Center
258 Fundeni Str., sector 2, code 022328, Bucharest, Romania
Tel/Fax: +40213007570
e-mail: drc_baston@yahoo.com
Introduction And Objectives

After undescended testis, hypospadias is the second most frequent congenital malformation of the male genital system. It has an incidence rate of about one case in every 250 infant boys. And in about 10% of the times it can be associated with other birth defects.

The etiology of the hypospadias is not exactly known. There are many hypotheses that include: genetic predisposition, environmental influence, maternal-placental factors and inadequate hormonal stimulation prenatally. Therefore it is possible that the etiology of hypospadias to be multifactorial.

The genital tubercle elongates to become the penis under the influence of fetal testicular androgens. A portion of the cloacal folds becomes the urogenital folds, which laterally bound the urogenital ostium with the labioscrotal folds developing laterally. The labioscrotal folds fuse in the midline to form the scrotum. As the solid epithelial urethral plate elongates towards the tip of the genital tubercle, it canalizes to form a groove on the ventral surface of the genital tubercle bounded by urethral folds. These urethral folds fuse in the midline converting the urethral groove into the penile urethra. Failure of fusion of the penile urethral folds from embryonic weeks eleven to sixteen results in hypospadias.

The urethral opening tells us what type of hypospadias we are dealing with. Hypospadias is categorized in several ways, the most classical one being first degree (urethral opening on the ventral glans), second degree (urethral opening on the ventral shaft), and third degree (urethral opening on the perineum). Second degree hypospadias is frequently accompanied by ventral chordee of the penis, which should be coded separately. The widely used Duckett classification categorizes hypospadias into glandular, coronal, distal penile shaft (or “anterior”) subgroup; mid-shaft (or “middle”) subgroup; and proximal penile shaft, penoscrotal, and perineal (or “posterior”) subgroup. Nearly 70% of hypospadias are either glandular or distally located on the penis and are considered a mild form of this disorder, whereas the remainder are more severe and complex.

The proximal hypospadias, although far less common, it is a lot more difficult to treat as it requires the reconstruction of the whole circumference and the full length of the urethra.

The treatment of hypospadias is surgery and its main goal is to achieve both good cosmetic and functional results. The intervention should be done during infancy because some studies have showed that young patients who do not remember the surgery are more likely to have a positive image of their body and be satisfied with their appearance then those who recall the intervention.

Usually the interventions are done by pediatric surgeons, but for different reasons some cases end up as hypospadias cripple in adult patients. The lack of experience leads to a high reoccurrence rate and other complications, like hematoma, necrosis of flap and graft, urethral fistulae, urethral strictures, penile curvature, hair in the urethra or urethral calculi.

Horton and Devine used the term of hypospadias cripple, to describe patients who had undergone multiple repair attempts with unsuccessful results. These cases are usually treated by urologists with great experience in penile and urethral reconstructive surgery.

When dealing with a patient with hypospadias cripple the physician should first try to understand what went wrong in the prior interventions, what were the complications and, if possible, operative reports should be obtained from prior repairs.

The aim of this paper is to present the surgical management of a difficult case of perineal hypospadias cripple.

Materials And Methods

A 21 years old patient presented in our clinic with a perineal hypospadias, inadequately treated during his infancy and early childhood at a pediatric department, when the perineal segment of the urethra was created using a scrotal skin graft, while the penile segment was reconstructed from a prepuce and penile skin flap and left ‘a plat’. The physical examination revealed a penoscrotal urethral meatus, with urethral calculi formed on hair grown inside the bulbar segment.
We decided on a two-stage repair technique, spaced over a six months period.

The first procedure was performed under general anesthesia with nasotracheal intubation. The patient was set in a dorsal decubitus. A tourniquet was placed at the base of the penis in order to reduce blood loss. The defective bulbar urethra was carefully dissected and completely removed.

Due to the large defect two buccal mucosa grafts were needed, which were harvested from both inner cheeks. The grafts were then dorsally placed on the corpora cavernosa, to replace the old scrotal skin graft, and a tie-over bandage was used to firmly hold them in place. We created a perineal urethrostomy and a 16 Fr Foley catheter was inserted. After four days the catheter was suppressed simultaneously with the tie-over bandage. The patient was discharged on the 5th day after surgery, voiding through the perineal urethrostomy.

After a period of six months the matured oral mucosa graft was tubularized in continuity with the prepuce flap and the neourethra was created up to the tip of the glans. The second stage of the repair was performed also under general anesthesia, with the patient in a dorsal decubitus. The matured graft was then dissected on the edges, and tubularized on a 16 Fr Foley catheter. The suture line was covered with the dartos fascia, and a glansplasty was carried out. At the end the skin was closed laterally to the neourethral suture, in order to minimize the risk of fistulae. The 16 Fr Foley catheter was left in place for three weeks.

Results

No immediate or late complications were observed after any of the two stages of the intervention. The patient was discharged 5 days after the first stage surgery and 6 days after the second stage. The Foley catheter was removed after 21 days and normal micturitions using the neomeatus were possible, voiding at 18 ml/sec.

Discussions

The first point of discussion is who should perform this type of surgery. The best moment to operate is in early infancy, starting form three months of age, or six months in premature babies, to avoid psychological trauma. The pediatric surgeon is in the first line of the treatment. But because they are often untrained enough for this type of complex urethral reconstruction, complication rate can get quite high. Therefore these patients should be referred directly to skilled urologists, that have enough experience with the urethral surgery.

The second and most important, due to the com-
plexity of proximal hypospadias, is choosing the best type of surgery technique, a single or multi-stage repair. The outcomes of both types of surgery for proximal hypospadias are comparable; no technique can be considered better than the other. Therefore we can only conclude that the surgeon should master a certain procedure and accumulate excellent experience to achieve the best results, regardless of the technique used. In hypospadias cripple situations, Bracka’s two stage repair with the excision and substitution of the old defective scared tissue is the best option, as it is reliable and ensures good cosmetic and functional results.

Conclusions

Although hypospadias remains one of the most challenging problems of the urologist, even the more severe perineal cases can be successfully treated. A two stage repair is a versatile technique as it allows the use of buccal mucosa grafts in combination with prepuce and penile skin flaps to reconstruct of the whole circumference and full length of the urethra.

References

4. Stoll, C; Alembik, Y; Roth, M P; Dott, B (1990). “Genetic and environmental factors in hypospadias”. Journal of Medical Genetics. 27 (9): 559. doi:10.1136/jmg.27.9.559. PMID 2231648
5. Calzolari, E; Contiero, M R; Roncarati, E; Mattiuz, P L; Volpato, S (1986). “Aetiological factors in hypospadias”. Journal of Medical Genetics. 23 (4): 333. doi:10.1136/jmg.23.4.333. PMID 3746833