



Minor And Major Challenges in Hypospadias Surgery: How to Face Them Boldly?

Abhinav Singh^{1*}, Malika Singh², Rohini Singh³ and RB Singh⁴

¹Department of Burns, Plastic and Reconstructive Surgery, Mahatma Gandhi Medical College, Jaipur, Rajasthan, India

²Department of GI Surgery, Amrita Institute of Medical Sciences (AIMS), Kochi, Kerala, India

³Department of Dermatology, Mahatma Gandhi Medical College, Jaipur, Rajasthan, India

⁴Ex. Head of the department of Burns & Plastic Surgery, Postgraduate Institute of Medical Sciences (PGIMS), University of Health Sciences, Rohtak (UHSR), Haryana, India

Abstract

Purpose: To create awareness about challenges, difficulties, problems and complexities of hypospadias surgeries faced by Hypospadiologists, hypospadiacs and their parents.

Aims and Objectives: To overcome these problems and challenges of hypospadias surgery efficiently in a scientific manner in longer-term of follow-ups for better post-operative results.

Material and Methods: The challenges, difficulties, problems and complexities detailed in this study were those that were faced by the authors during the period 1984 to 2016 while managing hypospadias patients to start with and thereafter looking after the Hypospadias and Vesico-Vaginal Fistulas (VVF) Clinic at Postgraduate Institute of Medical Sciences (PGIMS), Rohtak.

Observations: The challenges, difficulties, problems and complexities observed by the authors during management of hypospadias patients started from the day one of having seen them till their successful repair and there after ensuring them to remain complication free in longer-term of follow-ups, marital relations and reproduction.

Conclusion: For successful management of challenges, difficulties, problems and complexities of hypospadias surgeries in a scientific manner to an acceptable level of satisfaction, that too in the absence of availabilities of usable healthy and dispensable tissues, requires a brain-storming exercise and planning by the surgical team to achieve the required goals.

Keywords: Hypospadias; Challenges; Problems; Complexities; Hypospadiologists

Abbreviations

UCF: Urethro-Cutaneous Fistula; UCFs: Urethro-Cutaneous Fistulas; DTAP: Dorsal Tunica Albuginea Plication; BXO: Balanitis Xerotica Obliterans; TV: Tunica Vaginalis; STRIFs: Soft Tissue Reinforcement Interposition Flaps; IPFTSG: Inner Prepuceal Full Thickness Skin Graft; OPFTSG: Outer Prepuceal Full Thickness Skin Graft; DNVB: Dorsal Neurovascular Bundle

Introduction

Each step in the management of hypospadias is full of challenges from the beginning till longer-term of follow-ups, marital relations and reproduction. Assigning gender for a baby born with ambiguous genitalias could be the first challenge in front of the Hypospadiologists. On the other hand, management of hypospadias cripples created by occasional and unexperienced operators might require a brain storming exercise and planning to achieve the required goals to the satisfaction of patients, parents and the operating Hypospadiologists. The authors have thought worth to share their experience of different challenges, difficulties, problems and complexities faced during management of hypospadiac patients starting from the day one of having seen them to till their successful repair, marital relationship and reproduction. The challenges could be minor or major for the affected patients, parents and the surgeons. The challenges for the treating surgeons are to plan appropriate surgical techniques for the given case not only to minimize longer-term complications but also to decrease the incidence of re-doing for recurrences of complications. The challenges for the affected

OPEN ACCESS

*Correspondence:

Abhinav Singh, Department of Burns, Plastic and Reconstructive Surgery, Mahatma Gandhi Medical College, Jaipur, Rajasthan, India, Tel: 7225899112;

E-mail: abhi2889@gmail.com

Received Date: 30 Aug 2021

Accepted Date: 20 Sep 2021

Published Date: 28 Sep 2021

Citation:

Singh A, Singh M, Singh R, Singh RB. Minor And Major Challenges in Hypospadias Surgery: How to Face Them Boldly?. *J Plast Surg.* 2021; 1(1): 1002.

Copyright © 2021 Abhinav Singh. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

children are that they have to bear all types of burden of each surgical procedure like the pain and discomfort, down time, adversely affected working and studies, financial constraints, additional morbidities at donor or recipient sites, repeated failures and psychological impacts. Challenges for the parents are that they keep on thinking about the manhood of their affected children, schooling, marital relationship, sexual activities and reproduction including adverse impacts on the siblings. Parents need to manage repeated hospitalizations and finances. The challenges of the Hypospadiologists never end even till after the marriage, potency and reproduction.

Material and Methods

The study has incorporated all the challenges, difficulties, problems and complexities faced by the authors while managing different hypospadiacs during the period 1984 to 2016 at Postgraduate Institute of Medical Sciences (PGIMS), Rohtak, and explained the ways and means to face them boldly in a scientific manner to achieve the desired goals of recurrent complications-free repaired hypospadiacs in longer terms of follow-ups.

Observations

Various challenges, difficulties, problems and complexities faced by the authors are described below in a step-wise manner.

Assigning gender in ambiguous sex (indeterminate sex): When a child is born with an ambiguous sex or indeterminate sex, where the external genitalias are so much deformed and mal-developed to the extent that (i) neither the child can be straight forward accepted as male nor as female; (ii) the parents are psychologically disturbed as to whether to rear the baby as male (boy) or female (girl); (iii) the treating surgeon finds difficulty in getting different non-radiological, radiological and hormonal investigations; (iv) laparoscopic examination and biopsy are traumatizing; (v) chromosomal testing and bar-bodies are difficult to carry out in small babies, (vi) fear of the society considering the baby as transgender; and, (vii) the surgical procedures are difficult to carry out and there is great confusion whether to convert the baby in to male or female, depending upon the characteristics of such deformed external genitalias, however, the decision under such circumstances would depend upon the collective consensus of parents, family members, pediatric surgeon, pediatric urologist, endocrinologist, obstetrician and gynecologist, plastic surgeon and the treating Hypospadiologist. It also needs to be ascertained whether the baby is true-hermaphrodite or pseudo-hermaphrodite, because it will have direct impact on taking decision of conversion of gender. The conversion to female gender is technically easy as compared to conversion in to a male with fully functional penis to meet out all sexual desires and reproduction.

Counseling of parents and family: The psychologically disturbed parents need (i) detailed counseling regarding handling of such a deformed child who cannot be reared as male or female so easily, (ii) to be explained about the sex of the child only after assessing all the investigations, (iii) not to be guaranteed about the potency and fertility of the child, (iv) to be explained the need of conversion of sex of such a child and the different procedures described for that purpose, (v) to be informed about the early or late development of the secondary sex characters like the distribution of hair on the face, in the axilla and pubic region, the development of penis or the breast, body built and the muscle mass, characteristics of speech and behavioral changes, which might be affected by hormonal status of the child and the rearing of the child as male or female.

Psychological and psychiatric concerns: The baby born with ambiguous sex remains a constant source of worry till a definitive solution has been worked out and it might take some time. There is mental tension about the future of the newly born child, and this may trigger psychiatric illness. Only a detailed counseling can prevent such trauma to the parents and family. All the non-surgical and surgical options are explained to the parents after receiving all investigations and thorough discussion among all the members of the treating team.

Rearing a child with ambiguous sex: On the advice of the treating team and after consultation with parents and family, a unanimous decision is taken as to whether the child is to be reared as male or female. A final decision regarding gender will only be possible after confirmation on biopsy. Once the gender has been assigned then the rearing might have to be changed and the surgical planning is done accordingly to correct the external and internal genitalias. The abnormal development of breast, clitoris and penis will also need additional surgical interventions. The Hypospadiologist or the pediatric urologist comes in to picture for correction of penile deformities, only once the child with ambiguous sex has been assigned as male gender, and this might require multi-staged correction.

Reproductive capabilities: The parents are worried about the potency and fertility of the child. To answer this question to the parents and the family is not that simple, because nothing can be said about the functions of the internal and external sex organs, and it is only possible at the time of onset of puberty, i.e., whether the female-assigned gender has onset of regular menstrual cycle, development of secondary sex characteristics and on investigations, the ovary, tubes, uterus, cervix and vagina are anatomically and functionally normal, or whether the male-assigned gender has normal functioning penis and testes with potency and spermatogenesis. Therefore, an uncertainty prevails regarding productive activities, both in males and females assigned gender. The Hypospadiologist or the treating surgeon should never guarantee the potency and fertility at any point of time.

Marital concern: After having decided the gender of the child with ambiguous sex whether male or female, the next question is regarding marital issues, i.e., the marital compatibility, success, potency, fertility and reproduction, and also whether there are risks of developing such congenital anomalies in the siblings. Various surgical procedures produce scarring in different regions, and this could be a potential reason of marital disharmony. To prevent post-marital disputes, the scarring should be brought in to the knowledge of the other partner (inspection in privacy, through the treating team member, by way of photographs or through mutual discussion and understanding). The treating team can only certify about the structure of primary and secondary sex organs, but should not comment about potency and fertility. Whether one should marry or not is the entire and final decision of the person concerned including the family members. Nothing should be kept hidden and secrete about the status of sex organs. Where the anatomy and functions of the sex organs is not normal, the chances of pregnancy could be compromised or totally absent, therefore, under such circumstances, one should either not marry or marry with the clear understanding that the only solution to have siblings could be the adoption or may be *in-vitro*-Fertilization (IVF). Such clear thoughts and understanding before the marriage, though difficult, but will help in prevention of subsequent separations, divorce or break-down of the family or litigations. Further, the risks of psychological and psychiatric problems are also minimized.

Timings of repair of hypospadias: Once the male gender has been assigned or the male sex has been proved after investigations, the Hypospadiologist decides about the timings of start of repair of hypospadias, either in single-staged or in multi-staged manner depending upon the characteristics of the hypospadias, severity of deformities of genitalia and the expertise of the surgeon. As the child grows-up, the parents are becoming more and more worried about his (i) schooling, (ii) exposure to other class students, (iii) psychological impacts due to his deformed, un-corrected or scarred penile shaft, (iv) participation in ground or water sports and (v) hesitation, shyness and inferiority in class. Therefore, all the corrections of his deformed penis will have to be completed successfully to the satisfaction of the parents before the child is sent for schooling. Hence, the planning has to be done in such a manner that child is made a pointer (able to urinate in standing position along with his fellow-colleagues during outings for picnic etc.), has straight penis without much of obvious scarring and the penile appearance is almost similar to like that of his other friends and class-mates. The only purpose of such a planning is to prevent any psychological or psychiatric trauma to the growing child, as this is difficult to remove from the brain of the child once deeply invaginated. Severe deformities might need three to four stages with an interval in between varying from 3 to 6 months, like (i) correction of the chordee (orthoplasty) and re-construction of the neo-urethral plate, (ii) tubularization of the neo-urethral plate to re-construct neo-urethra, (iii) closure of urethro-cutaneous fistulas (UCFs), (iv) scrotoplasty and sometime one might have to correct residual chordee before proceeding to neo-urethroplasty or the neo-urethral plate might not be ideal or stenosis or strictures get formed, thus requiring further stages for corrections. Even there could be associated inguinal hernia or undescended testis, and both these need treatment on priority. The distal hypospadias might be delayed or postponed for correction, but proximal hypospadias would need correction due to chordee and urination in sitting like a girl. Minor corrections of scarring, tracts and sinuses can be taken care later on.

Prepuceplasty (re-construction of prepuce): The sole purpose of operating the child at an early age is that the child should not be aware of (i) his deformed external genitalia, (ii) his private parts having been traumatized multiple times, (iii) his feeling of inferiority to his colleagues, (iv) his pre-operative and post-operative deformities to adversely affect his psychology and (v) having ever been operated for his deformed external genitalias. This can be possible by re-constructing a prepuce which will give an appearance of normal penile shaft having never been operated or traumatized or there could be fully developed prepuce in a concealed distal penile hypospadias (Figure 1). The re-constructed prepuce will (i) provide cosmetic look to an operated penis, (ii) because concealment of scarring and deformed glans, (iii) play valuable role during sexual activities and (iv) prevent constant exposure of glans to get hypoesthesia, depigmentation, recurrent trauma, meatal stenosis or frequent bleeding due to constant friction caused by under-garments. Therefore, the prepuce re-construction has cosmetic and functional roles. Presence of prepuce will help in future correction of stricture, UCFs, diverticulum, short or the hairy urethra by providing healthy, non-hairy, usable and dispensable ideal tissue of genital region.

Rehabilitation: A timely rehabilitation has great impact on the overall behavior and adjustment with his class-mates, friends, social circle and family members. Timely rehabilitation will let the child become shy-less, fearless, free from hesitation and inhibition, without inferiority in any aspect and also fully involved in all social,



Figure 1: Well-developed and long prepuce in a concealed distal hypospadias. Nothing would be visible after its repair. No psychological disturbances.

sport and cultural activities, and this is important for overall normal development of the child psychologically, socially, physically and mentally. Long standing depression could result in overall defective growth of the child. All assurance is given to the child by parents and treating surgeon to the effect that his external genitalias are normal anatomically as well as functionally.

Correction of chordee (orthoplasty/straightening of curved penile shaft): After having assigned the gender to a child born with ambiguous sex, the next challenge for the Hypospadiologist is the correction of severe ventral penile curvature (ventral chordee), especially when (i) the penis is not fully developed (micropenias) in its length and girth, (ii) Dorsal Tunica Albuginea Plication (DTAP) under such circumstances will have extremes of detrimental effects on the future growth of the penis due to its de-skeletonization done while correcting severe chordee to straighten out the severely bent penis, (iii) the pre-existing short length of penile shaft is being further shortened following DTAP, (iv) there appears buckling of penile shaft at the site of DTAP, (v) there is hypoesthesia due to accidental suturing of the dorsal nerves of penis in DTAP sutures, (vi) there occurs complete loss of sensations in the glans by suture ligation of dorsal neurovascular bundle of penile shaft and (vii) the sutures are felt on the penile shaft while masturbating or performing sexual intercourse. The alternate option with the treating surgeon is to lengthen the penile shaft in micropenias by way of ventrally incising the half circumference of tunica albuginea at the site of maximum ventral curvature and filling the resultant gap of the tunica albuginea by dermal graft or the graft from parietal layer of tunica vaginalis. However, it will produce further scarring in the scrotum or the groin for harvesting the free graft. Even the operating surgeon in consultation with the parents might plan urethroplasty without correction of chordee, and the left over penile chordee will be corrected only after having the penis developed in its length and girth at or after the onset of puberty. Correction of severe chordee in micropenias is really a challenge for all Hypospadiologists, and needs to be handled very carefully keeping in mind not to shorten the already short and under developed penis. Rather all efforts are made to lengthen the micropenias.

Reconstruction of ideal neo-urethral plate: After adequately straightening of the penile shaft in proximal hypospadias with severe chordee, the challenge in front of the Hypospadiologist is to



Figure 2: An ideal re-constructed neo-urethral plate using IPFTSG, which is smooth, shiny, non-keratinized and non-hairy.



Figure 4: Tunica vaginalis flap is displayed for its use in water-proofing of the re-constructed neo-urethra.



Figure 3: Planning to harvest buccal mucosal graft to re-construct neo-urethral plate. Genital skin was not available due to its use in previous surgical interventions.



Figure 5: Deep and dense adhesions between penile skin and deeper penile shaft. Penile erection and sexual intercourse become painful.

re-construct an ideal neo-urethral plate having its all characteristics similar to native urethral plate or the native urethra (Figure 2). Another associated challenge could be the deficient prepuccial hood, under developed prepuccial hood or the circumcised penis on religious grounds. Under such circumstances, the surgeon will have to search another tissue for re-construction of ideal neo-urethral plate like the (i) use of graft from bladder urothelium, (ii) buccal mucosal graft (Figure 3) or (iii) tunica vaginalis-supported split skin graft. Therefore, in the absence of adequate prepuccial skin, arrangement of alternative tissues planned will be able to (i) withstand constant exposure of urine without getting transmural inflammation, destruction, breakdown, fistulizations, hair growth, diverticulization, metaplasia, stones, stricture or the carcinoma.

Reconstruction of neo-urethra: After having re-constructed an ideal neo-urethral plate and permitting to get it fully matured and vascularized, the next challenge is to re-construct an ideal neo-urethra having all the characteristics of a normal native urethra, i.e., its epithelial lining resistant to adverse effects on constant urinary soiling, non-hairy, resistant to Balanitis Xerotica Obliterans (BXO), uniform diametered, free of stricture, fistula, diverticulum, desquamation, debris and stone formation, metaplasia or the cancer formation. Here the surgeon will have to symmetrically mark the neo-

urethral plate boundaries of sufficient width to tubularize it to produce neo-urethra of uniform caliber for smooth resistance and turbulence free flow of urinary stream. Each suture bite has to be appropriate to avoid strangulation of tissues and eversion of epithelial lining. The use of non-absorbable suture is not advised for fear of permanent knots being felt manually as well as during sexual intercourse. Special precautions are taken to avoid anastomotic stricture as well as meatal stenosis. Even after two layered tubularization of neo-urethral plate, the inner continuous and outer interrupted, there is need of re-strengthening of the neo-urethra by covering it with one or two water-proofing flaps depending upon the availability, i.e., whether a dartos fascial flap can be designed from the well-developed prepuccial hood or a distant Tunica Vaginalis flap (TV flap) is to be harvested from the parietal wall of tunica vaginalis (Figure 4). Harvesting of water-proofing flap from the prepuccial hood prevents prepuccioplasty and the harvesting of TV flap will require additional time and surgical procedure. Designing and harvesting of a water-proofing flap might become a challenge in those proximal hypospadiacs that have scarcity of tissues due to under-developed prepuce and bilateral undescended testes. The successfully operated hypospadiacs should be able to pass urine in a single compact and thick projectile stream of urine without any splaying or holding of urine. Further, the challenge to the Hypospadiacologist is to produce a neo-urethra having contractile power as seen in a native urethra covered all around by corpus

spongiosum, but such tissue is not available that can make the neo-urethra contract to expel the urine and ejaculate following sexual intercourse. A little bit of contractile power could be provided to the re-constructed neo-urethra by a water-proofing flap of the tunica vaginalis that has cremasteric muscle fibers with its neurovascular bundle. The other option could be the use of gracilis muscle cover to the re-constructed neo-urethra which will serve dual purpose, i.e., to increase penile girth and also to provide contractile power to the newly re-constructed neo-urethra.

Correction of scarring, un-even surface and girth, pigmentary disorders, deeper tissue adhesions and angulation: Even after correction of hypospadias and having got straight and functional penis, the child poses further challenge for beautification of the repaired penile shaft. All such scars, un-evenness and discolorations can be tackled on the similar pattern as adopted for elsewhere in the body like revision of scars, excision of bulging tissues, auto-grafting in the region of depressions, tissue-filling in the region of narrowness, excision of discolored skin and grafting it with appropriate colored skin graft, tattooing and dye application where ever indicated, the adherent penoprepuccial skin is freed from deeper adhesions and thereafter massaged manually to prevent re-formation of adhesions (Figure 5). The significant angulation is exposed and corrected by either removal of inappropriately applied sutures or by doing tunica albuginea plication on the opposite side of the angulation to be corrected.

Correction and prevention of recurrences of UCFs, urethral stricture, urethral diverticulum, anastomotic stricture, meatal stenosis, hairy urethra, stone formation, metaplasia and malignancy in the re-constructed neo-urethra: The real challenges for the treating surgeon start while managing the above-mentioned complications, because each of these complications will require special measures not only to correct it but also to prevent its recurrence. In extreme of the worst complications, all these might be present in one repaired hypospadias, and the challenge is of complete excision (urethrectomy) of so badly affected neo-urethra and re-doing for the re-construction of neo-urethral plate and the neo-urethra. It is known that repeated failures are associated with increasing incidence of recurrences of such complications; therefore, the surgeon is under extreme of pressure to use such tissues and techniques that will prevent such recurrences in longer-term of follow-ups.

(i) Urethro-Cutaneous Fistulas (UCFs): Although the most common etiology of UCFs formation is use of avascular or is chemist issues during the re-construction of neo-urethra associated with failure to use water-proofing flap for the re-strengthening of the re-constructed neo-urethra. There could be formation of multiple UCFs (Figure 6). All the fistulas are converted in to one larger fistula before repair. The water proofing flaps have dual role in preventing UCFs formation due to their mechanical and biological properties. The re-constructed neo-urethra needs to be examined for the presence of the associated urethral stricture, meatal stenosis, hairy neo-urethra, diverticulum, stone formation, and metaplasia and cancer formation, un-supported or attenuated neo-urethral wall. The UCFs are not to be simply freshened and closed, rather these are better closed with the help of a flap (fistuloplasty) from scrotal skin or penile skin to provide further strength to the repaired fistula against its recurrence (Figure 7, 8). The Hypospadiologist will have to find suitable donor sites for harvesting Soft Tissue Reinforcement Interposition Flaps (STRIFs) to work as water-proofing flap [1]. The real challenge is when there



Figure 6: Before attempting repair of multiple UCFs, all are converted in to one larger UCF by dividing their intervening skin bridges; else failures will be high if individual fistula is repaired separately.

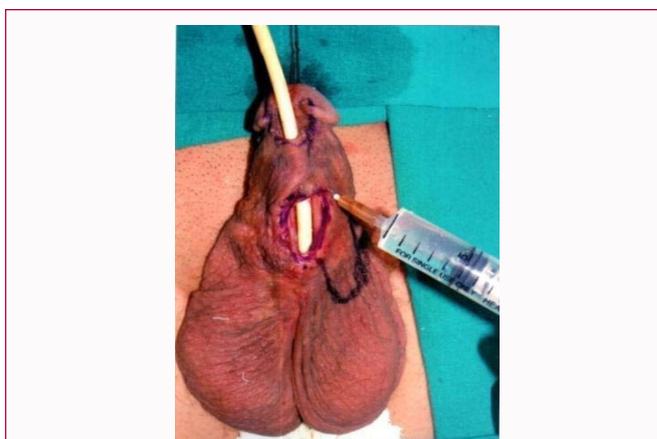


Figure 7: Scrotal skin flap closure of UCF (fistuloplasty).



Figure 8: Penile skin flap closure of UCF (fistuloplasty).

is scarcity of donor tissue that can be sacrificed without producing additional morbidities. In extreme scarcity of expendable tissues, the surgeon might have to think of using scrotal skin (Figure 9) and its dartos as described by for multi-staged Cecil-Culp technique [2]. The designing of tunica vaginalis flap is another alternative to meet-out such circumstances [3]. The prepucial skin is also used to design dartos fascial flap for water-proofing [4]. Before directly repairing the UCFs, all other associated morbidities of the neo-urethra (Figures 10-12) as described above must be corrected; else the recurrences of



Figure 9: Cecil-Culp technique is used to provide scrotal skin cover to the ventrum of penile shaft in those having scarcity of dispensable healthy peno-prepuccial skin.



Figure 12: The whole of the re-constructed neo-urethra is seen dilated to form diverticulum.



Figure 10: Urethral stricture, diverticulum and UCF. Methylene blue dye is injected through external urethral meatus after perineal compression to occlude proximal urethra.



Figure 13: IPFTSG is being harvested for re-constitution of neo-urethral plate, which will be tubularized to re-construct neo-urethra after its full maturation.



Figure 11: Healing of a UCF by secondary intension to cause further scarring and constriction of urethra to result in to formation of severe urethral stricture.

UCFs are un-avoidable. Extreme of associated scarring around the Urethro-Cutaneous Fistula (UCF) might need urethrectomy. Such UCFs are never repaired in the same sitting, rather adequate time is given for the tissues to get fully matured to become soft, supple and revascularized, and this period might range from 3 to 6 months

or even a year in cases of repeated recurrences. The fistulas are laid open (fistulectomy) or excised (fistulectomy) or complete excision of the affected neo-urethra (urethrectomy) is done, and the resultant raw area is re-surfaced by appropriate epithelial lining like the Inner Prepuccial Full Thickness Skin Graft (IPFTSG) (Figure 13) or Outer Prepuccial Full Thickness Skin Graft (OPFTSG) or buccal mucosal graft or the bladder urothelium or the TV flap-supported Split Skin Graft (SSG) is also an option. The re-constructed neo-urethral plate is tubularized after 3 to 6 months to re-construct the neo-urethra or after its having been fully matured.

(ii) Stricture urethra: The most common location of the neo-urethral stricture is the site of anastomosis between the native urethra and the re-constructed neo-urethra, and it is the result of technical fault due to (a) continuous suturing of the distal end of the native urethra and the proximal end of the re-constructed neo-urethra to produce purse-string effects at the anastomotic site or (b) anastomosing the ischemic ends of native and re-constructed neo-urethra. Such stricture produces hydroureterosis and hydronephrosis in proximal hypospadiacs. However, the stricture along the course of re-constructed neo-urethra could be the result of infection, ischemia, trauma, electro-cautery burns, and such strictures would produce breakdown of proximal neo-urethra to a variable extent like the formation of ultra-micro urethro-cutaneous fistulas, micro



Figure 14: Urethrogram showing urethral diverticulum.



Figure 15: Neo-urethral diverticulum as a result of distal urethral stricture.



Figure 16: Laid-open neo-urethra showing its hairy lining following use of extra-genital hairy skin from groin for re-construction of neo-urethra about 3 years back following its loss in a road traffic accident.



Figure 17: Stones in the re-constructed neo-urethra of extra-genital hairy skin in origin.

urethro-cutaneous fistulas, macro urethro-cutaneous fistulas, single or multiple fistulas including complete or near complete disruption of the neo-urethra. The tight stricture might produce urethral dilatation or urethral diverticulum (Figure 14). The treatment of urethral stricture is a surgical challenge to Hypospadiologist, because different maneuvers and surgical procedures would be required depending upon the characteristics of stricture, and these could range from repeated urethral dilatations, internal optical urethrectomy, excision of stricture (stricturotomy), laying open the stricture (stricturotomy), excision of strictured neo-urethra (urethrectomy) and interposition urethroplasty or complete urethrectomy and re-construction of neo-urethral plate, which will be tubularized after its full maturation to have become soft and supple with adequate mobility.

(iii) Urethral diverticulum: This could be due to distal urethral stricture (Figure 15) or simple ballooning of an attenuated neo-urethra due to normal pressure of urinary stream. Another potential reason could be the use of extra-genital hairy skin in the re-construction of neo-urethral plate or the neo-urethral tube. Such extra-genital skin gets damaged by its constant exposure of urine by way of transmural inflammation and cellular infiltration, thus causing break-down of dermal component and weakness, which ultimately balloons out to form diverticulum. Simple excision of diverticulum is not the treatment, rather it is best treated only after knowing its etiology and then to correct the causative factor, so that no recurrences are seen in longer-term of follow-ups. One might have to (i) treat associated

stricture, (ii) strengthen the attenuated thin-walled neo-urethra and (iii) replace hairy part of the re-constructed neo-urethra.

(iv) Hairy neo-urethra: This is a serious challenge to the Hypospadiologists, because the assured procedure for such condition is complete replacement of the hairy lining of the re-constructed neo-urethra, and it invariably ends in to urethrectomy of its affected part and re-do neo-urethroplasty either in single stage (interposition urethroplasty) or multi-staged (excision of hairy part and re-construction of neo-urethral plate using appropriate epithelial lining like the genital skin or bladder or buccal mucosa depending upon the availability of ideal donor site. Endoscopic depilation is not possible and electrocautery is not advisable because there remains risk of residual hairs as well as recurrences or post-burn urethral stricture. The commonest cause of hairy neo-urethra is use of extra-genital skin for re-construction of neo-urethral plate and neo-urethra (Figure 16).

(v) Stone formation: The basic factor responsible for stone formation is the use of extra-genital hairy skin for the re-construction of neo-urethral plate or the neo-urethra. Extra-genital skin is not accustomed to constant exposure of urine, that causes intra-mural inflammation and de-squamation, debris formation, slow destruction of the neo-urethral wall to result in to its diverticulization (Figure 17). There is stagnation of urine and debris in to the diverticulum, which in the presence of hairs will collectively be responsible for stones or



Figure 18: Crippled hypospadias following multiple failed surgical repairs. Scarring, absence of healthy usable tissues, breakdown of repair, irregular shaft and surface, discoloration, stricture, fistulizations and no urethral plate.



Figure 19: Skin lined urethral plate, unfit for re-constructing neo-urethra. Such keratinized skin needs to be replaced by smooth non-keratinized genital skin like the IPFTSG/ OPFTSG/ buccal or bladder mucosa.

concretion formation. The treatment is not the removal of stone; rather it will require complete excision of the affected segment of neo-urethra, followed by its re-construction, either in the same sitting or after a gap of 3 to 6 months where staged correction is planned. Under such conditions, even the adjacent proximal and distal parts of the normal appearing urethra are also adversely affected and will need excision.

(vi) Metaplasia: Long standing diverticulum and the presence of debris and stones initiate process of onset of metaplasia due to constant irritation and inflammation. This is a pre-malignant condition and requires resection of the affected part of neo-urethra with normal adjacent margins, both proximal-wards and distal-wards.

(vii) Malignancy: Neglected cases of urethral malignancy might end-up with not only radical urethrectomy but also penile amputation at varying levels depending upon the extent of lesion. Advanced cases might develop secondaries in the groin, necessitating bloc dissection. Although its incidence is quite low but it is a potential risk in the presence of long-standing neglected cases of stones with diverticulum. This is a great challenge for the patients, parents and the treating Hypospadiologists. There is no perfect surgical technique that can re-construct a normal functioning penis and urethra.

Crippled hypospadias: This is another challenge in front of the Hypospadiologists, wherein multiple morbidities have occurred due to multiple un-successful surgical procedures by un-trained and occasional operators (Figure 18). The affected penile shaft is having (i) shortage of usable, dispensable and healthy unscarred tissues (no skin and prepuce), (ii) extensive scarring, (iii) significant residual chordee, (iv) multiple UCFs, (v) urethral stricture, (vi) diverticulum, (vii) stone and concretions, (viii) BXO changes, (ix) metaplasia and (x) hairy neo-urethra with partially flaccid penile shaft associated with severe psychological disturbances. The greatest challenge under such circumstance is to find out appropriate donor site for harvesting non-hairy epithelial structure (buccal mucosa, bladder mucosa, TV-supported SSG) to re-construct ideal neo-urethral plate after excising all un-healthy and scarred tissues. Even the glans could be blunt and ugly looking with its partial loss, pigmentary lesions and under-developed penile dimensions (length and girth). This challenge is difficult to face, because the treating surgeon will have to re-do the whole procedure for achieving an ideal neo-urethral plate and neo-

urethra. The technique of urethral culture is not fully developed for clinical use from where one can have such facility.

Sub-optimal neo-urethral plate and residual chordee: Despite taking all precautions for correction of chordee, the hypospadiologist finds presence of significant residual chordee on saline induced artificial erection just before the start of neo-urethroplasty, and under such conditions, the neo-urethroplasty will have to be postponed and only chordee correction is done. Similarly, the final picture of the neo-urethral plate could be un-satisfactory and this will require its redo for creation of an ideal neo-urethral plate, therefore, the neo-urethroplasty has to be postponed till an ideal neo-urethral plate is re-constructed and penile shaft is straight. The skin lined urethral plate (Figure 19) is un-ideal for use and must be excised and re-constructed by genital skin. Here the challenge for the treating surgeon is to do Dorsal Tunica Albuginea Plication (DTAP) in the presence of previous adhesions that might risk injury to the Dorsal Neurovascular Bundle (DNVB) as well as further shortening of the length of penile shaft. Further, the surgeon will have to excise the un-healthy neo-urethral plate and find suitable donor site for harvesting epithelial tissue (buccal mucosa, bladder urothelium or TV-supported SSG) that could meet all the characteristics of providing ideal neo-urethral plate.

Retarded penile growth: Extensive de-skeletonization of penile shaft in the micropeniacs with severe ventral chordee alone or in association with hormonal disturbance could have detrimental effects on future growth of penile shaft. No satisfactory procedures are available to achieve the desired characteristics and dimensions of a normal penile shaft. The patients, parents and the surgeon find difficult to face this challenge and also hard for them to get it solved up to the expectations. However, the penis might attain its normal dimensions at the onset of the puberty. The re-constructed penis is neither anatomically nor functionally normal or near normal.

Tight penile skin, sinuses, tracts, skin tags and heaped-up skin, hypertrophic scarring, keloid formation, granulomas, retention cysts at recipient or donor sites: These are minor challenges that can be solved easily with minor surgical procedures either by excision (Figure 20) or the policy of tissue re-adjustment or replacement.

Priapism: This is continuous and persistent penile erection that could occur either per-operatively or post-operatively, causing extensive bleeding and difficulty in further surgical dissection. This



Figure 20: Excision of skin tags before proceeding to neo-urethroplasty.



Figure 21: Extensive scarring of penile shaft and partial loss of the glans due to BXO and repeated failed attempts of hypospadias repair.

emergent situation of unwanted penile erection in the post-operative time could cause bleeding and disruption of repair. Sub-cutaneous or intra-venous administration of Bricanyl could cause de-tumescence. Quick de-tumescence could be caused by wide-bore needle puncture of the glans, and the bleeding from the glans will settle this problem.

Intercourse and semen deposition: This is a matter of great concern to the married couple, because the reconstructed ne-urethra is deficient of corpora spongiosum, therefore, there are no contractile activities in the reconstructed neo-urethra, thus resulting in to holding of the ejaculate in the non-dynamic neo-urethra. The retained ejaculate will have to be manually expelled by massaging from proximal to distal-wards after the act of intercourse so as to deposit it in the vagina or it has to be artificially inseminated.

Catheter and tubes related complications: The emergency situation is faced when there is epididymo-orchitis or the catheter or the tube gets retained inside the bladder. Forceful removal of such retained objects could cause disruption of the neo-urethra. Different procedures are required to be performed for removal of retained Foley catheter and the knotted infant feeding tube. Such maneuvers in the presence of immature repair could result in trauma or the infection (epididymo-orchitis, testicular abscess, atrophic testis, Azoospermia).

Meatal stenosis and blunt glans: Meatal stenosis is not a challenge because it can be corrected either by frequent gentle meatal dilatation or by doing minor surgical procedure like meatotomy or meatoplasty. Blunt glans appearance could be an extreme of psychologically disturbing problem and of great concern for sexual intercourse (Figure 21). Repeated attempts of glanuloplasty could result in a serious threat to its loss of vascularity and sensations. The glans could be destructed by BXO. Extreme care is taken while designing glans wings so as to prevent trauma to its neurovasculature as well as trauma to the apical region of the tunica albuginea, because such injury to tunica albuginea could result in de-tumescence in the distal part of the penile shaft, and this becomes an unbearable challenge.

Peno-scrotal fusion and hairy penile shaft: This is more of a psychological threat (Figure 22). This can be easily managed by depilation or excision and re-grafting of the hairy skin of penile shaft. Such condition arises where scrotal skin is used to provide skin cover to the raw areas on the penile shaft or where Cecil-Culp procedure has been done. Corrective procedures are easy with good results. Release of penile shaft from the scrotum cause penile lengthening.



Figure 22: Peno-scrotal fusion depicting virtual shortening of penile shaft. Release of penis from scrotum by simple division of this webbing would show real penile length.

Pre-mature or delayed secondary sex characters: Timely appearance of secondary sex characters is also important to become a complete human being. Even pre-mature presence of such secondary sex characters is also disturbing to the parents (Figure 23). Absence of beard, moustaches, chest hairs, axillary and pubic hairs are traumatizing to the affected child and are also equally disturbing to parents and treating surgeons. This condition might need hormonal administration at times. Most of such children would develop all this at the onset of puberty.

Graft loss, dressings and splints: Excessive loss of graft due to any reason is of great concern to the patients, parents and the surgeons, because the resultant raw area would heal by secondary intention to develop scarring or hypertrophic scarring or the contracture or the secondary chordee. The neo-urethral plate formed is of sub-optimal quality, which might require redo grafting later on to have an ideal neo-urethral plate. The streptococcus Beta-hemolyticus organisms must be excluded and eradicated, if found on culture sensitivity of the exudates from the raw areas. The dressings are time consuming and needs to be done with patience. Correct dressing and application of penile splint will enhance quick healing and maturation of the neo-urethral plate.

Non-availability of trained Hypospadiologists, presence of occasional operators, absence of tertiary care centers and



Figure 23: Psychological disturbances as a result of pre-mature appearance of secondary sex characters.



Figure 25: Eccentric incision to define urethral diverticulum so as to prevent suture-line superimposition and fistulizations.



Figure 24: Near total disruption of un-supported and attenuated neo-urethra due to not using water-proofing flap for its further re-strengthening.



Figure 26: Distal hypospadias with severe glandular chordee of 90°.

lack of tissue culture facilities: These conditions are great challenge and threat to the patients, parents and especially to the trained Hypospadiologists because these all are responsible for creating hypospadias cripples which will ultimately be handled by Hypospadiologists.

Retained catheter and infant feeding tube: The removal of such retained tubing's in an incompletely healed neo-urethra could be a challenge, because any amount of intervention could result in to disruption of repair. When Foley catheter has been used, its balloon must be fully deflated before attempting its removal. Different maneuvers could be required for deflating the balloon of the retained Foley catheter, ensuring no trauma to the recently repaired hypospadias. The intra-vesical knotted infant feeding tube might necessitate its removal through supra-pubic route, because its trans-urethral endoscopic removal will also be harmful.

Failure to use water-proofing flaps: Failure to re-strengthen the re-constructed neo-urethra is prone to disruption, which could be a near total breakdown of neo-urethroplasty (Figure 24). The wall of the re-constructed unsupported neo-urethra is attenuated and it tends to balloon out or breakdown to form UCF. Such banked soft tissue could be used in the management of recurrences of UCFs.

Superimposing incisions and closure: All the incisions be ensured that they are made eccentric (Figure 25) and closed eccentric,



Figure 27: Such long prepucio-plasty would camouflage scarring resulting from repair of distal hypospadias.

because this prevents incidence of fistulizations.

Glans chordee: Extreme of care is taken while correcting severe distal glans chordee of 90° (Figure 26) because the dissection in this region is risky due to high incidence of injury to DNVB, which might produce hypoesthesia or loss of part of glans. Extensive de-skeletonization in micropeniacs endangers retarded penile growth.

Concealment of scarring: The only way to have camouflaging



Figure 28: Operated case of high ARM with perineal hypospadias, bifid scrotum and scrotal transposition. Hypospadias repair is postponed in the presence of ARM, hernia or undescended testis, which are corrected on priority.



Figure 29: Micropenis with severe chordee and asymmetrical scrotum. De-skeletonization of so small and underdeveloped penile shaft to correct chordee would cause its subsequent retarded growth.

effects on the operated penis is by way of (i) re-constructing full length of prepuce, i.e., prepuceplasty (Figure 27) in distal hypospadiacs and (ii) by uniformly draping the penile shaft with smooth and non-hairy skin along with creation of median raphe on the ventral aspect of penile shaft and the scrotum.

Flap closure of fistulas (fistuloplasty): The best way to prevent recurrences of UCFs is their flap closure, and such skin flaps could be designed either from the prepuccial hood, penile skin, previously banked skin or the scrotal skin.

Discussion

The hypospadias surgeries are full of multiple challenges [5]. The minor challenges like: Isolated distal penile hypospadias; mild to moderate penile chordee in a fully developed penis with tumescence and normal spermatogenesis; isolated micro Urethro-Cutaneous Fistula (UCF) in the absence of urethral stricture, urethral diverticulum, hairy urethra, stones, BXO, metaplasia and malignancy; surgical scarring, tracts, sinuses, heaped-up penile skin, hairy penile skin cover, mild to moderate penoscrotal fusion; isolated meatal stenosis; isolated inguinal hernia or unilateral un-descended testis; un-even surfaced penile shaft; skin discoloration; partial prepuceplasty; isolated granulomas; inability to expel urine and

ejaculate spontaneously; delayed onset of secondary sex characters; presence of mild to moderate psychological disturbances; and, ease in counseling of parents and family members where the manhood of the baby, his schooling, marital relationship, reproductive activities are being questioned by the society, are all managed in routine with ease as no much time and finances are required. The major challenges are like: Ambiguous sex; severe psychological disturbances and difficulties in counseling; proximal hypospadias with severe chordee, micropenis and circumcision; bilateral undescended testes with upper urinary tract anomalies; presence of associated high anorectal malformations (Figure 28); crippled hypospadias consequent upon multiple failed surgical attempts by untrained surgeons; presence of impotency and infertility; co-existence of UCF, stricture, diverticulum, hairy urethra, stones and concretions, metaplasia and BXO changes; to achieve fully functional penis, contractile neo-urethra and normal spermatogenesis for reproduction; and, situations of separations, divorce and family breakdown are real major challenges for the Hypospadiologists who will undergo brain storming exercises to find permanent or long lasting solution despite multiple constraints for such affected children so that they can stay asymptomatic.

The trained Hypospadiologists can face these challenges boldly when (i) the prepuce is intact in full or partial, (ii) bilateral testes are fully developed and descended with normal spermatogenesis, (iii) penis is of adequate length and girth and (iv) patients and parents are cooperative and willing to consent for additional surgical procedures and resultant scarring for harvesting of buccal or bladder mucosal graft or the tunica vaginalis flap or even for complete dismantling of the crippled hypospadias and then its redo (chordee correction, re-construction of neo-urethral plate, re-construction of neo-urethra and designing of water-proofing flap). The worst situation is when there is severe scarcity of usable healthy peno-prepuccio-scrotal skin, severe chordee, multiple UCFs associated with stricture, diverticulum, stones, metaplasia and cancer, deep scarring risking injury to DNVB, short penis, scarred and hypoesthetic glans and hypogonadism with defective spermatogenesis.

Minor challenges are bound to be faced by the Hypospadiologists after successful repair of proximal hypospadias with severe chordee, even in the hands of experts, but prevention of occurrence of major challenges is possible to a great extent where:

- The treating surgeon is a trained Hypospadiologist.
- The chordee has been corrected to an acceptable level.
- Prepuccial hood has been appropriately used to re-construct an ideal neo-urethral plate.
- Fully vascular and viable water-proofing flap has been designed from the peno-prepuccial skin or the tunica vaginalis.
- Multi-staged technique has been planned instead of single staged repair.
- The neo-urethra has been re-constructed by tubularizing symmetrically defined neo-urethral plate of non-hairy skin of genital region or of the buccal or bladder mucosa or the tunica-vaginalis supported split skin graft.
- Adequate time interval is given between two successive stages, which could range from 3 to 6 months of waiting for adequate maturation and vascularization of the tissues before their subsequent intervention.



Figure 30: Repaired case of distal hypospadias. Severe distal glandular chordee obvious. DTAP in the distal region in the presence of previous scarring endangers damage to DNVB.

- Only viable tissues are used while re-constructing neo-urethra and also at the anastomotic site between the native urethra and neo-urethra.
- Complete correction of chordee is ascertained by saline induced artificial penile erection before proceeding to final neo-urethroplasty.
- Extensive de-skeletonization is avoided, especially in micropeniacs with severe chordee (Figure 29). Correction of residual or left over chordee of distal glans in the presence of adhesions (Figure 30) due to previous surgical interventions could cause damage to DNVB in the region of coronal sulcus where it is difficult to identify.
- Simple closure of UCF is avoided until other associated morbidities like stricture, diverticulum, hairy urethra, metaplasia are

excluded by thorough examination and investigations, especially in long standing and neglected cases.

- Adequate pre-operative planning is done in consultation among all the members of the surgical team to finalize the best possible procedure for the given case under consideration.

Conclusion

Trained Hypospadiologists have all the courage to face such different minor and major challenges that come on the way while managing hypospadiacs, but it requires a brain storming exercise to find solutions which will prevent occurrence and recurrences of complications to keep the operated hypospadiacs asymptomatic in longer-term of follow-ups.

References

1. Singh RB, Dalal S, Pavithran NM, Sharma B. Soft tissue reinforcement interposition flaps in hypospadias repair. *Ind J Plast Surg.* 2007;40:170-7.
2. Ehle JJ, Cooper CS, Peche WJ, Hawtrey CE. Application of Cecil-Culp repair for treatment of urethrocutaneous fistulas after hypospadias surgery. *Urology.* 2001;57(2):347-50.
3. Snow BW. Use of tunica vaginalis to prevent fistula in hypospadias. *Br J Urol.* 1986;136(4):861-3.
4. Belman AB. De-epithelialized skin flap coverage in hypospadias repair. *J Urol.* 1988;140(5 Pt 2):1273-6.
5. Misra D, Elbourne C, Vareli A, Banerjee D, Joshi A, Friedmacher F, et al. Challenges in managing proximal hypospadias: A17-year single-center experience. *J Pediatr Surg.* 2019;54(10):2125-9.