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The Hypospadias Cripple: Review of Concepts and a Systematic Approach to Management

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Introduction

Ever since the first description by Celsius (25 AD) and Galen (second century AD), hypospadias has posed itself as a continuous challenge to the general surgeons, pediatric surgeons, plastic & reconstructive surgeons and urosurgeons through the generations.[1] The promulgation of innovative techniques and modifications of the existing ones have extended the domain of hypospadias repair from extending the urethra upto the tip of the glans while correcting the associated ventral chordee to restoration of near normal function and appearance. With the advances in the operative skills, availability of technical devices and focus on 'lateral' areas such as dressing and post-operative care, the faculty of hypospadiology has undergone revolutionary advances over the past couple of decades. However, the overall complication rate still ranges between 10% to 30% and there continues to be a distinct subgroup of patients who are not cured even after multiple attempts at hypospadias repair and are left with severe deformity and/or stricture with or without penile skin deficiency.

Horton and Devine used the term "hypospadias cripple" to describe this subgroup of patients to signify the presence of residual significant penile deformity even after multiple surgeries.[2] A hypospadias cripple may present with any or a combination of hypospadiac meatus, urinary fistula, residual chordee, extensive scarring of penile skin, urethral stricture or diverticulae, meatal stenosis, or glanular dehiscence. There is no standard definition of a hypospadias cripple; a working description would include any patient of hypospadias with residual anatomic deformity or functional complication(s) after multiple attempts at surgical correction. The term has occasionally been used to define less severe cases of hypospadias as well. 'Hypospadias cripple' is not preferred currently and most experts refer to this situation as *'salvage hypospadias repair'*.

Problems with Salvage Hypospadias Repair

Reoperative urethroplasty in this subgroup of patients represent one of the most challenging situations. They require extensive repair amidst scarred and devitalized tissue. These patients have a paucity of healthy, unscarred penile and preputial skin due to multiple failed prior phallic reconstructions. In fact, excision of the excess foreskin at the time of primary hypospadias reconstruction is a routine in some countries reflecting their medical, cultural and religious bias towards circumcision. Even, the scrotal skin may have been used up in the previous surgeries. The penile skin that is present is typically immobile, scarred and has a variable and poorly defined blood supply. The dense fibrous tissue with poor vascularity causes difficulty in wound healing and increases the incidence of surgical complications. Lack of well-vascularized skin precludes the creation of flaps and the lack of a good vascular bed hinders the success of free grafts. The previous surgeries also result in loss of tissue planes and subsequent degloving of the penis may be associated with significant bleeding, tissue trauma, devitalization of the penile skin and healing with enhanced fibrosis.

The urethral plate may have been resected in the previous attempts at repair or may be scarred. An otherwise normal looking urethral plate may not be supple enough to be re-tubularized. Presence of multiple urethral fistulae separated by thin skin bridges may necessitate lay-open of the entire region thereby converting it into a large fistulous opening. Preputial dartos may not be sufficient to provide a 'water-proofing' cover to the neourethra.

More often than not, the parents of the patients have a tendency to change the surgeon after facing multiple failures. Lack of proper past surgical history and non availability of baseline data about the initial degree of hypospadias or the surgical procedures carried out may not be indispensable for planning the future repair but it does affect the understanding of the true long-term outcomes of earlier types of surgeries.[3]

Psychological, Psychosocial and Psychosexual Consequences of Multiple Failed Repairs

A significant proportion of these patients have grown into adolescent or crossed puberty. The psychological impact on the patients and the parents as a result of multiple failures may be severe. This may be related to

urinary dysfunction, unacceptable cosmesis and even sexual dissatisfaction in the elderly patients. These patients develop a negative appraisal of their genital organs.[4,5] This may act as a hindrance to the development of social competencies and social relations. The situation may be further aggravated by the parental apprehensions about the future masculinity of their child which can be transferred vertically and may affect the child's capacity to accept his bodily difference.[6] The peer pressure acts as yet another factor which makes a child sensitive to the difference. They develop a fear of appearing ridiculous in front of the friends. The same fear gets converted later in life into a fear of appearing ridiculous in front of their partner if he or she ever discovered the difference in their genital appearance. It has been observed that these patients even hide their genitals in situations where it is conventional to be exposed such in public toilets or public showers.

In the western culture, the penis represents activity, self-affirmation, social success, strength and masculine power. A deficiency in the penis leads to a psychological deficiency resulting in fear of incapacity or incompetence extending beyond the purely physical functions of urination and sex. The extension of these fears into the domains of psychological function, social relations and sexual behavior may affect their psychological, psychosexual and psychosocial development.[7]

Approach to A Patient with Multiple Failed Repairs

Each case should be treated as different and should be evaluated individually. The salvage hypospadias repair should ideally address the following issues:

1. Elimination of penile chordee: intrinsic or iatrogenic
2. Construction of urethra to restore the continuity upto the tip of the glans while maintaining adequate calibre
3. Cosmetic skin coverege to the penile shaft

Assessment of Urinary Meatus for Need for Meatotomy

In the very first visit, the patient should be assessed for the adequacy of the urinary meatus. If the meatus appears to be stenotic, a meatotomy should be contemplated. A 'cut-back urethrotomy' may be necessary to get rid of the obstruction.

Role of Testosterone in Salvage Hypospadias Repair

Pre-operative use of testosterone for temporary penile stimulation results in a larger and more vascularised organ. It improves the texture, vascularity and pliability of penile skin.[8] Koff and Jayanthi observed that hCG pretreatment in infancy produces disproportionate penile enlargement which advances the meatus distally to decrease the severity of hypospadias and chordee.[9] Monfort and Lucas reported a mean increase in penile length and circumference by 50% after local penile stimulation with 5% dihydrotesterone cream before hypospadias repair.[10] Ahmad *et al* observed a mean increase in penile length, transverse preputial diameter and diameter at the base of penis without any change in the serum testosterone level after parenteral testosterone administration.[8] Use of testosterone in hypospadias cripples will help in penile augmentation and enhanced vascularity of the penile skin which may affect the post-operatively results favourably.

Residual Chordee

Persistent chordee may result from tethering of the penile skin ventrally, foreshortened urethra or from corporal disproportion.[11] The penile curvature is assessed clinically at the time of first presentation and any suspicion of residual chordee is confirmed intra-operatively.

The penile shaft must be degloved evenly at Buck's fascia upto the penoscrotal junction. Care must be taken to identify and protect the existing urethra with a small catheter in the urethra. Artifical erection is induced by injection of saline into the lateral aspect of one of the corpora with a tourniquet at the base of penis. Pharmacological erections are not usually recommended due to lack of accurate dosing regimen in children, lack of response or fear of prolonged erections (priapism), additional cost and need of phenylephrine for reversal.[12] The urethra should be inspected to determine if it is supple. When the urethral plate is normal, dorsal plication should be sufficient. A 'bowstring' urethra is indicative of fibrosis and merits transaction followed by resection of fibrous tissue. Persisting curvature even after urethral transaction is usually indicative of corporal disproportion and calls for ventral corporotomy with grafting (skin, small intestine submucosa or tunical vaginalis flaps or grafts) or dorsal plications.

Re-tubularization of the Urethral Plate: Tubularized Incised Plate Urethroplasty (TIP Repair)

TIP concept was described as a salvage procedure in 1987 by Orkiszewski.[13] It is the preferred technique of redo hypospadias surgery in the authors' hands. However, it is important to assess the quality of the urethral plate prior to taking a decision. Tubularization of the urethral plate may be attempted in those patients in whom the urethral is supple and there is no scarring or any evidence of incision in the past. Previous incision adversely affects the vascularity of the urethral plate and the results of re-tubularization. Not only this, a previously failed TIP urethroplasty may be an indirect evidence of an intrinsic abnormality of the urethral plate or the corpus spongiosum. A second attempt at urethroplasty in these patients by the same technique may not be wise. The urethral plate may be incised at the time of re-operation but must be reassessed for suppleness after incision. A 'water-proofing' barrier layer is absolutely necessary especially in redo cases. While outlining the urethral plate, the surgeon may 'borrow' the adjacent skin from both sides of the urethral plate to facilitate tension-free closure.[14] If the plate is fibrosed or non-conducive towards a healty tension free urethroplasty, alternative surgical techniques must be considered.

Nguyen and Snodgrass made observations in 31 consecutive patients undergoing tubularized incised plate reoperations. 18 (58%) in this group had undergone primary repairs that involved midline plate incision. The complication rate in this subgroup was 22% as against 23% in those patients in whom the urethral plate had not been incised in the previous surgery.[15] Overall, they realized that complications are low despite previous urethral plate incisions if there is no apparent scarring of the urethral plate. Besides, TIPS also results in a functional neourethra with a vertical slit meatus and fistulas are less likely when a flap is interposed between the neourethra and skin. Similar observations have been published in other reports also.[16,17,18,19,20]

Re-tubularization of the Urethral Plate: Snodgraft Repair

Sometime, the axial integrity of the urethral plate can be maintained and requires only circumference augmentation. This is especially true for situations when the urethral plate is supple but is not wide enough to facilitate tension-free urethral closure around stent adequate for the age of the patient. Using a smaller sized stent or making too tight an anastomosis would result in stricture or dehiscence. However, adequate size of the urethral stent and the neourethra has not been clearly defined. Snodgrass clearly cited that the minimum size of neourethra should be 10Fr.[21] Yang *et al* have suggested that the ideal size of the urethral stent should be decided on the basis of the age of the patient and the size of the proximal urethra. In boys younger than 6 years, the maximal width of the separated urethral plate should be 12mm or larger.[22] In older patients, it is wise to perform urethral calibration before urethroplasty and the calibre of the neourethra should be 80 to 90% of the size of the proximal urethra. Elder *et al* made an observation from the then published reports that the meatal size in boys aged 0 to 3, 4 to 10 and 11 to 12 years were usually not smaller than 10, 12 and 14 Fr, respectively.[23] The urinary meatus is the narrowest part of the male urethra and the limiting factor; the size of the urinary meatus can be regarded as the minimal size of the urethra. They also recommended a 2 to 4 Fr smaller sized stent than the maximal width of the urethral plate separated after incision.

'Snodgraft' repair is a modification of the TIP urethroplasty in which the defect from the dorsal wall releasing incision is closed by quilting in a free graft such as the preputial skin or the buccal mucosa rather than being left open to re-epithelialise spontaneosly. The term 'Snodgraft' was coined by Hayes and Malone who first described the use of buccal mucosa graft augmented TIP repair.[24] This procedure has reduced the need for two-stage repairs in many salvage hypospadias repairs. Due to fibrosis, the urethral plate in redo cases in not a good source of new epithelium. More often than not, the glans groove is poor in these cases and it becomes mandatory to extending the dorsal midline clefting incision beyond the urethral plate into the body of the glans. Unless this extended incision is grafted, it is likely to heal back together and may result in meatal stenosis.

Use of Adjacent Genital Tissues in Cases with Poor Urethral Plate

Use of vascular adjacent genital tissues may be contemplated depending on their availability if the urethral plate is not usable. A Thiersch Duplex tubularization may be attempted if adequate penile skin is available. A significant increase in the incidence of postoperative fistulas, strictures and glansplasty may be expected when adjacent skin is used for repeat hypospadias. This may be related to local scarring and vascular compromise from multiple prior procedures. Scrotal skin is often very well vascularised but its use is limited due to its hair bearing nature. When the scrotal skin in incorporated into the urethroplasty inadvertently, it may result in repeated episodes of urinary infections, calculi and trichobezoar (hair ball). Preemptive epilation has been suggested as a remedy but electrocoagulation of the dermal papilla is usually ineffective due to poor depth of

penetration. Nd:YAG laser has 5 mm depth of penetration and destroys hair follicles more effectively. Previously incorporated hair bearing skin may be managed with endoscopic removal of hair follicles with laser coagulation of the base. Urethrotomy may be necessary for abundant hair growth followed by epilation or replacement of the hair bearing segment with a nonhair-bearing graft. Regrowth of hair may be managed with chemical de-epilation using thioglycolate.[25]

Patel *et al* suggested the use of split onlay skin (SOS) flap for salvage hypospadias repair to improve the outcome. The technique is useful in patients where after multiple failed surgeries there is absence of penile foreskin and shortage of ventral skin. The SOS flap aims to transfer healthy dorsal tissue to the ventral surface of the penis for onlay salvage urethroplasty. The technique entails mobilization of a transverse island of penile shaft skin on its vascular pedicle. The flap is rotated onto the ventral surface of penis and is transacted transversely. One half of the flap is used as 'onlay' to repair the urethra. The other half of the flap provides additional skin coverage where needed on the penile shaft. In this way, a redo urethroplasty is completed with an onlay island flap using residual penile skin and adequate skin is rotated for ventral penile skin coverage.[26]

Two Stage Repair: Grafts

Two stage repair comes into picture when it is necessary to transect the urethra or the urethral plate and a full circumference replacement is mandatory. Various indications include severe persistent chordee, severly scarred urethral plate, balanitis xerotica obliterans (BXO) or to replace a hairy segment of urethra. Flaps may not be feasible in redo cases because of paucity of skin locally and dense fibrosis.

- Inner preputial skin is the ideal substitute for the native urethra. It is thin, flexible, hair-free, designed to be moist and shows a good uptake. The donor site is convenient. However, the preputial skin is not available for redo cases especially those labelled as 'cripples'.
- External preputial skin is not ideal as it is not moist naturally and becomes hairy in some after puberty.
- Oral mucosa is probably the most commonly graft in use. It may be harvested from teh buccal, labial or lingual donor site. It is an excellent substitute for native urethra. It is hair-free, designed to be moist and leaves no visible scars at the donor site. Buccal mucosa has a good tensile strength and a very vascular submucosal plexus that favours graft uptake. However, it is bulky and requires aggressive thinning. Labial mucosa is thinner but limited in width. Lingual donor sites are yet under evaluation.
- The Post Auricular Wolfe Graft (PAWG) is harvested from the back of the ear or posterior auricular sulcus. They are similar to buccal mucosal grafts in thickness and designed to be moist. Although fine lanugo hair growth may be present, coarse hair is never a problem provided the donor area is close to the mastoid. However, the donor site sometimes develops keloid scars. Junctional keloids with reconstructed urethra necessitate replacement of post-auricular skin with buccal mucosa.
- Bladder mucosal urethral replacement was popular in mid 1980s. The idea was theorized on the transitional epithelial lining of the urinary bladder which is normally exposed to urine. The need for lower abdominal incision and the associated urethral meatal prolapsed led to the use of other graft materials.

A graft must derive vascularity from the recipient bed which is usually scarred in these patients. It is therefore more reliable to do a stage procedure because the corporal bodies are likely healthier than the skin of the penile shaft which would be important in case of an onlay tube or graft (single stage). A tunnelled placement of a tube graft is less likely to get disrupted due to vascular insufficiency. The authors advocate buccal graft urethroplasty using the staged technique as described by Bracka.[27,28,29,30] During the first stage, the graft is secured to the corporal bodies from the current urethral meatus to the tip of the glans. The graft is quilted onto the corpora and covered with a compression dressing to prevent hematoma formation. The uptake is better as the graft is dependent on the corporal bodies and not upon the penile skin which is fibrosed and hypovascularized. The grafted site should be reviewed every month and any focus of scarring or necrosis should be regrafted. Tubularization is to be done after at least 6 months. The Mainz group recommends keeping the urinary neomeatus at the corona to prevent generation of high pressure at the tip which may encourage blowout proximally.

Tissue Expanders in Salvage Hypospadias Repair

A hypospadias cripple has minimal residual skin available for additional reconstructive efforts. The argument favouring tissue expansion over grafts is related to the pigment and texture-matching of the penile skin. Mir *et al* reviewed their experience with tissue expanders in 6 patients of hypospadias after multiple failed repairs (5-8 failed surgeries). The injection was positioned in the suprapubic region. The tissue expander was positioned

under the skin on the dorsum of the penile shaft and inflated slightly in the operating room. Subsequent inflations were made at intervals of 2 weeks upto 12-16 weeks. After maximum tissue expansion, the expander was removed and urethral reconstruction was attempted. Successful phallus resurfacing could be achieved in all 6 patients. Urethrocutaneous fistula and meatal stenosis were seen in 1 patient each which was successfully managed.[31]

Mathews *et al* made their observations in a group of 18 patients with multiple failed phallic reconstructive efforts. Of these, 7 patients were hypospadias cripples. Tissue expanders could be placed and successfully inflated during a 6-week period in 17 patients. 8 patients had good surgical results with no need for additional surgical procedures. 2 patients required repeat tissue expansion; one of these had a good clinical outcome after repeat expansion followed by supplemental pedicle grafting. However, the other patient had persistent urethrocutaneous fistulae and required forearm graft. Extrusion was seen in 3 patients requiring graft replacement. The graft was removed for malfunction, erosion and infection in 1 patient each.[32]

Urethral Stricture After Hypospadias Repair

Stricture is another common problem after repeat urethroplasties. They may be related to faulty technique, vascular compromise in the tissues, post-operative wound infections, traumatic stent removal or balanitis xerotica obliterans. Stictures may develop even long after an apparently successful hypospadias repair which may be secondary to failed growth at puberty or BXO. Direct Vision Internal Urethrotomy with or without stenting has been used as a remedy but with poor results which are probably related to lack of spongiosal layer around the urethra. The extravasation of fluid, urine or blood into the periurethral tissues is not contained and results in a significant inflammatory response and stricture progression.[33] Barbagli *et al* suggested a single-staged dorsal free graft urethroplasty for these cases.[34] A free graft is doomed to fail in patients with BXO and buccal mucosa should be used as the graft material in these cases. Gupta NP and Sripathi *et al* independently suggested that the stricture be approached through a ventral sagittal urethrotomy, division of stricture and the buccal mucosal inlay grafting on the dorsal aspect.[14,35] A ventral approach to widen the stricture with a tunica vaginalis patch on a vascular pedicle has also been reported.[14]

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