23 Management of Hypospadias in Patients with Disorders of Sexual Development (DSD)

P. Ashwin Shekar and Amilal Bhat

23.1 Introduction

Though most hypospadias cases occur as an isolated anatomical defect, in a small subset of patients, the hypospadias may be part of a more complex anomaly of a disorder of sex development (DSD) [1, 2]. The HP in this group of patients tends to be severe with a more proximal division of corpus spongiosum and prominent hypoplasia of ventral tissue distal to it, leading to a more proximal urethral opening and a higher incidence of penile curvature [2, 3]. Besides, the presence of other associated anomalies like a bifid scrotum, penoscrotal transposition, micropenis, undescended testis (UDT), or a prostatic utricle in these patients that needed to be addressed along with HP repair necessitates the formulation of individualized surgical strategies on a case to case basis [4].

23.2 When to Suspect DSD in a Child with Hypospadias and How to Evaluate it?

One of the most important questions that arise in the mind of a treating surgeon when dealing with hypospadias is, "Should I evaluate for DSD in this patient of hypospadias?" Unfortunately, there is still no evidence-based consensus on the best answer for this perplexing question to date [3, 5]. Presently, recommendations are based mostly on the experience of individual surgeons or individual centers with marked heterogenicity in the diagnostic criteria and investigation protocols used, which make generalization of such recommendations difficult [2].

Generally, any patient with a proximally located urethral meatus and severe ventral curvature with an associated bifid scrotum and undescended testicle needs evaluation for DSD by a multidisciplinary team that includes the treating surgeon. Specifically, assessment of gonadal position is a crucial aspect of the initial assessment, which influences the direction of further investigations [6]. Additional useful clinical markers that may indicate the need for DSD eval-

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A. Bhat (ed.), *Hypospadiology*, https://doi.org/10.1007/978-981-16-8395-4_23

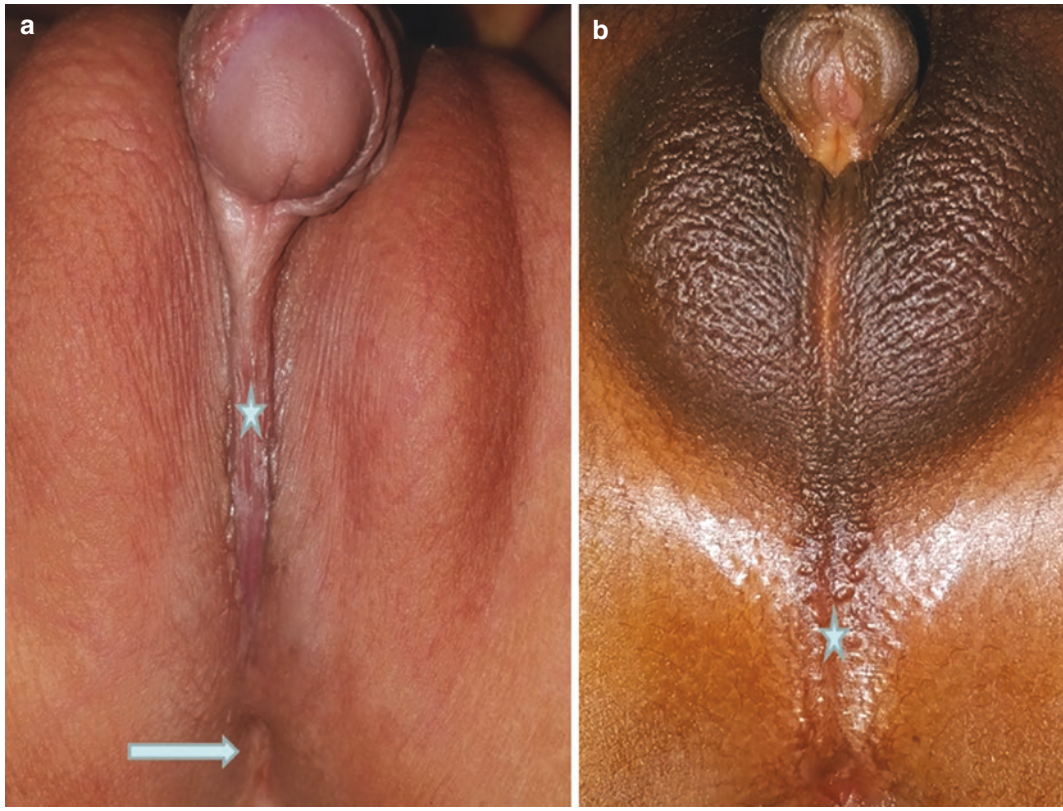

Fig. 23.1 (**a** and **b**) Clinical picture showing bifurcation of the median raphe (star) and a dimple in the perineum (arrow)

uation include an incomplete fusion of the median raphe and a dimple in the perineum (Fig. 23.1).

Investigations as part of the DSD workup would typically include

- Serum electrolytes immediately to rule out CAH
- Karyotype
- Pelvic ultrasound
- Hormonal studies
  - 17-OH progesterone
  - Serum testosterone, Dihydrotestosterone (if age <3 months or after HCG stimulation, if older).
  - Serum luteinizing hormone (LH) and follicle-stimulating hormone (FSH).
- Imaging studies (Ultrasound/MRI abdomen and pelvis)—to assess Mullerian structures/gonads, if impalpable.
- Genitogram
- Diagnostic laparoscopy—to assess gonadal status when impalpable.
- Cystourethroscopy
- Genetic studies—if available, for specific genetic defects.

If the investigations mentioned above are normal, the patient is considered to have idiopathic proximal hypospadias. If abnormalities in the chromosome or hormonal profile are identified, further study for a specific diagnosis should be done by a multidisciplinary DSD team. This would make it easier to give the family and caregivers the best information available that addresses specific concerns like the need for pubertal hormone supplementation, available surgical options, later fertility potential, cancer risk, and stability of sex of rearing with aging [7]. Also, identifying a specific diagnosis will help the clinicians better understand the associated

pathophysiology and, more importantly, its correlation with surgical outcomes [7].

23.3 Gender Assignment Considerations

Another unique situation that the treating clinician will frequently face is the issue of gender assignment in babies born with ambiguous genitalia, where the hypospadias appears to be a part of the spectrum of DSD. Again, this is a sensitive issue and requires to be handled with the utmost care and gentleness. Most families are under considerable distress due to the atypical appearance of the patient's genitalia.

Some of the significant factors that have to be considered here are the external phenotype, the internal karyotype, ease of reconstruction, and parental expectations/attitudes. In the past, when the investigations were limited, the "nurture vs nature" approach was used commonly, leading to the assignment of female gender in many of these patients due to ease of reconstruction; however, the knowledge of brain imprinting in the last few decades and the incidence of gender dysphoria in these patients reared as females in later life, has made this approach questionable [6]. Nowadays, with better investigations, a multidisciplinary effort must be made to come to an accurate diagnosis in all these patients, which will help in offering the best possible advice on gender assignment. Due to this more scientific approach, over the past three decades, there has been a substantial increase in male gender assignment, especially in the group of under-virilized XY males with disorders of androgen synthesis or a partial androgen insensitivity syndrome (PAIS) profile who have both testicular and phallic tissue present and also demonstrates an adequate response to stimulation of the pituitary–gonadal axis [8–10].

23.4 Timing of Re-evaluation and Surgery for Hypospadias

Surgical intervention is not typically required in these children diagnosed at birth during the neonatal period since the proximal urethral opening does not usually cause any voiding symptoms; however, it is prudent to schedule a visit to the surgeon at around 3–6 months. This time period would allow the parents to process all information received in the postnatal period with respect to the further investigations required in DSD and also allow the parents to establish a strong bond with their child, which ultimately would lead to a more focused discussion on the further course of action including surgery [6]. Also, many of these patients will have associated undescended gonads, and this would give enough time for spontaneous descent to happen [11]. As per present guidelines since infants benefit from better wound healing and therefore decreased complications in comparison to older children, HP repairs irrespective of the association with DSD has been recommended between the ages of 6 and 18 months [12]. Another purported advantage of this approach is that early reversion to normal-appearing genitalia significantly relieves parental distress and improves parent–child attachment. However, strong evidence for this belief is lacking [12, 13].

However, over the last decade, the ideal timing of surgical intervention in patients with hypospadias has become a subject of debate from an ethical perspective, more so in children with associated DSD [14–16]. Though there are obvious benefits of early intervention with respect to functional improvement in voiding and positive adaption in these children, arguments against an early age of intervention have revolved around the issue of consent of patients with this condition, with child rights activists advocating that only the patient himself can give consent to surgical intervention on their genitalia and any intervention should be deferred until the patient has developed a stable gender identity, to avoid the possibility of gender dysphoria at adulthood leading to dissatisfaction and even necessitating a sex reversal [14–16].

It is clear now that this issue of timing and need for surgical intervention is a sensitive one. The treating clinician must present both sides of the argument to the parents and caregivers in a very unbiased way and assist in their decision-making process on whether to proceed with surgery or not [6]. Ultimately, both the patient's physical

status and the parents' psychosocial preparedness have to be considered when deciding on the timing of surgery to ensure the best possible results for the child and the family [17, 18].

23.5 Preoperative Evaluation and Goals for Surgical Intervention

Thought the primary aim of a masculinizing genitoplasty done in childhood is the reconstruction of the external genitalia to better correspond with male sex of rearing and prevention of complications like infection and malignancy, a good reconstruction should also allow for future sexual activity and optimize the fertility potential [8, 19].

When preparing for a masculinizing genitoplasty in these children, the physical examination should focus on the dimensions of the phallus, including length and girth, urethral meatus location, and presence of an orifice suggestive of a potential vaginal cavity. Careful palpation for gonads that can be present in the inguinal region, labioscrotal folds or scrotum, is crucial for clinical examination, which cannot be overemphasized [1, 19]. Regarding the scrotal appearance, patients can present in a spectrum ranging from a normally formed scrotum to a bifid scrotum with separated labioscrotal folds or varying degrees of penoscrotal transposition, depending on the degree of under-virilization [19]. Another clinical finding which can be used as a surrogate marker of abnormal male reproductive tract masculinization and sexual dimorphism is a reduced anogenital distance (AGD) which, along with penile length, are now considered as accessible end-point markers for male reproductive health. An essential part of the workup which should be done in all cases of the suspected DSD is a genitogram, which will delineate the length of common confluence and presence of vagina, prostatic utricle, uterine and fallopian tubes, and their size, location, and relation with the urethra (Fig. 23.2a–b). Patients with a common perineal opening will also need an urethrocystoscopy to assess for a potential vaginal cavity/prostatic utricle (Fig. 23.3a–b).

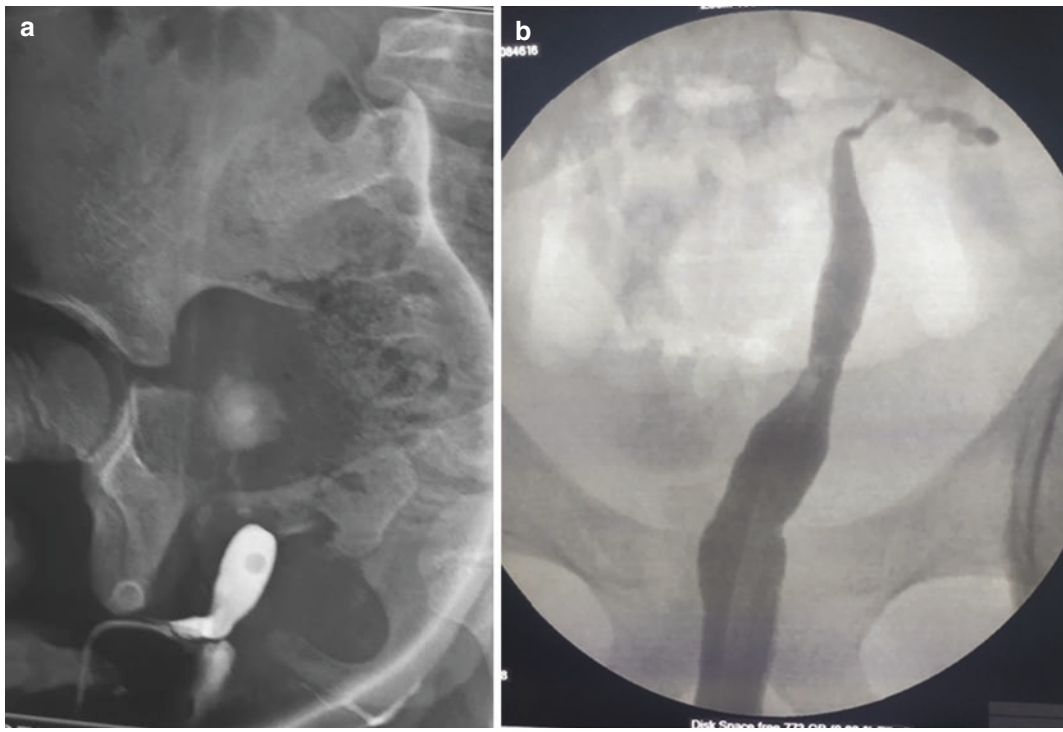

Fig. 23.2 Genitogram showing (**a**) Well-delineated vagina and common urethral canal, (**b**) Vagina, hypoplastic uterine cavity, and fallopian tube

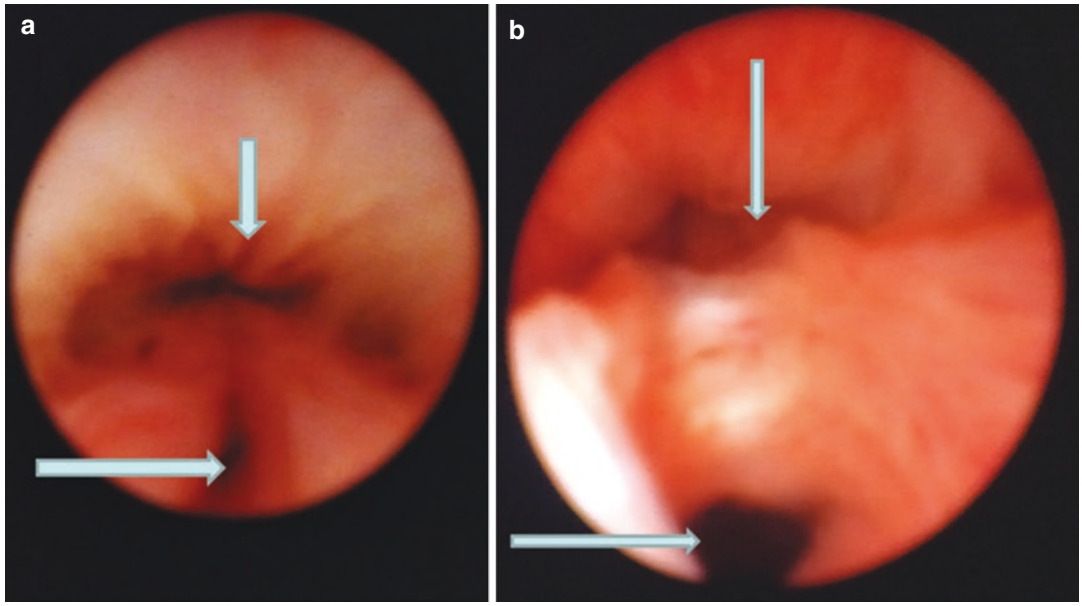

Fig. 23.3 (**a** and **b**) Cystoscopic view showing the double opening in the urethra (upper urethral and lower vaginal opening)

23.6 Preoperative Administration of Testosterone

Another controversial area in hypospadias surgery that has divided hypospadiologists over the years has been the role of preoperative testosterone administration before hypospadias surgery. The present consensus is that testosterone has a role in children with a small penis, more so in those children with small glans and severe curvature, where it can improve the surgical outcomes by making tissues more robust for surgical handling [6, 20]. However, there are marked variations among different surgeons and centers on the timing of testosterone administration, dose and route of administration (intramuscular vs topical application). In our center, we prefer the intramuscular route for testosterone administration given at a dose of 2 mg/kg/month for two consecutive months followed by surgery after a month of the second dose (Fig. 23.4a—pre- and, b—post-testosterone). It has to be noted that in children with significant androgen insensitivity, comparatively higher testosterone doses may be needed of testosterone to demonstrate the desired response [6].

23.7 Surgical Considerations

Hypospadias surgery in patients with concomitant DSD is no different from those without DSD in the surgical technique used. The basic principles of hypospadias surgery remain the same, which involves the following main steps.

23.7.1 Surgical Technique

Cysto-urethroscopy is done after inducing the general anesthesia. This helps assess the length of the common urethrovaginal canal, the orifice of the urethra and vagina (Fig. 23.3a and b), orifice of vaginal opening and length of the vaginal cavity, prostatic utricle, bladder size, and any other abnormality of ureteric orifice/bladder. Circumferential circumcoronal incision is given after injecting the 1:100000 solution of adrenaline, and penile degloving is done while dissecting at the level of Buck's fascia. Gittes test is done to assess chordee (Fig. 23.5b). If the chordee still persists, then the proximal urethra is mobilized (Fig. 23.5c and d, 23.6b). Mobilization of the urethra is done along with

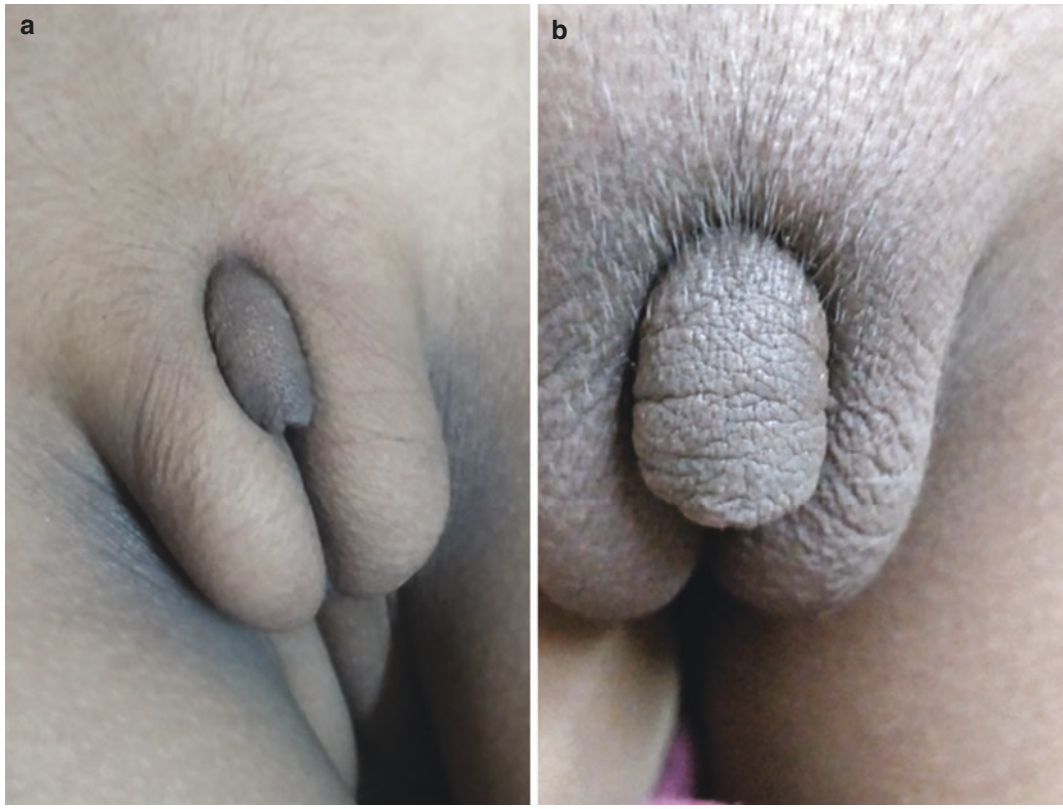

Fig. 23.4 (**a**) The appearance of phallus and scrotum before (**a**) and after (**b**) testosterone therapy

the mobilization of the vaginal cavity. Care is taken not to damage the urethra and bladder while mobilizing the vagina. This mobilization of the vagina releases the urethral attachments and adds length, which helps in the correction of chordee (Fig. 23.6c, d and e). The Gittes test is repeated again to confirm chordee correction (Fig. 23.5e). The vaginal cavity is closed in two to three layers according to the depth of the cavity after resection of the vagina (Fig. 23.6f). The urethral plate is tubularized with or without incision and dorsal inlay, followed by spongioplasty (Fig. 23.5f and g, 23.6g). Dorsal dartos flap flaps are mobilized and used to cover the urethra in a single or double-layer as an interposing healthy tissue (Fig. 23.5h). Glanuloplasty is done in two layers to bring the meatus to the tip, and a conical glans is refashioned. Midline scrotal tissue is mobilized, scrotoplasty is done in layers, and finally skin closure is done (Fig. 23.5i). If parents choose preputioplasty, the tunica vaginalis can be used to interpose healthy tissue, and the spared prepuce is utilized for preputioplasty (Fig. 23.6h).

In cases with severe curvature, the urethral plate is transected at the corona, and the urethral plate with spongiosum is mobilized at the level of tunica albuginea (Fig. 23.7a, b and c). Chordee correction is then confirmed. If the chordee persists still, then the proximal urethra alone or the urethra with the vagina is mobilized to correct the chordee (Fig. 23.7d). The mobilized urethra is then brought up to the penoscrotal junction. The prepuce is brought ventrally either as a flap by Byars technique or as a preputial graft and fixed to the shaft to create a urethral plate for second-stage urethroplasty followed by scrotoplasty (Fig. 23.7e).

The three main considerations in these hypospadias repairs are:

1. *Correction of ventral curvature/Chordee*

This involves degloving of the penis to passively assess the severity of the hypospadias based on the level of division of the corpus spongio-

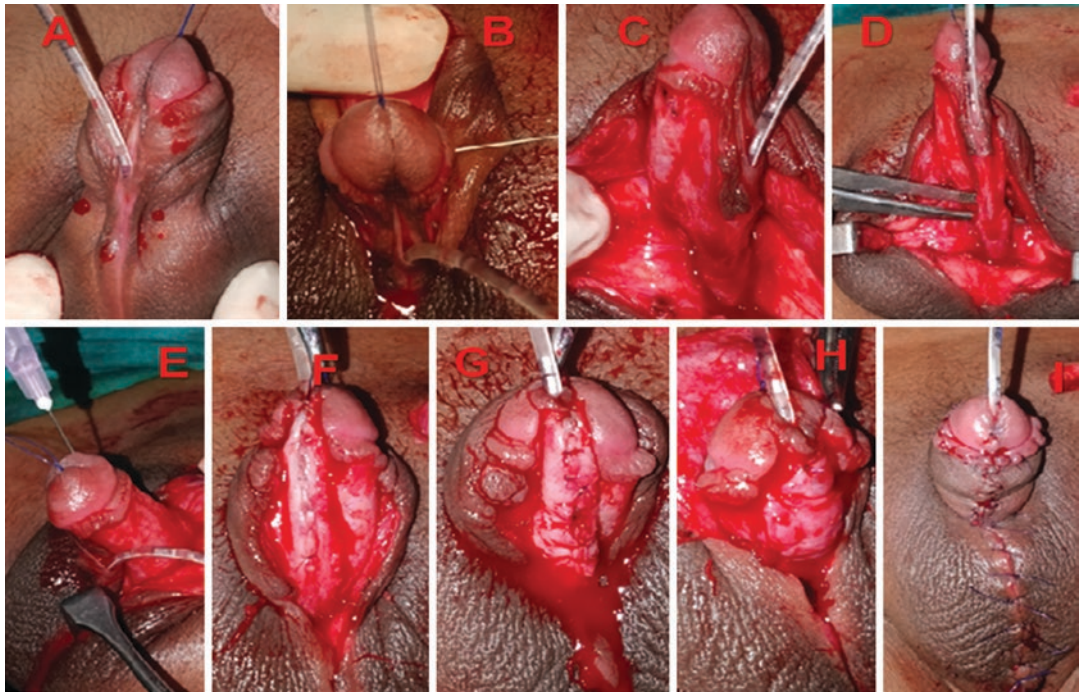

Fig. 23.5 Intraoperative pictures showing the steps of tubularized urethral plate urethroplasty. (**a**) Proximal penile hypospadiac opening. (**b**) Penile degloving and Gittes test showing chordee. (**c** and **d**) Proximal urethral mobilization. (**e**) Repeat Gittes test confirming chordee correction. (**f**) Tubularization of the urethral plate. (**g**) Spongioplasty. (**h**) Dorsal Dartos cover. (**i**) Glanuloplasty and skin closure

sum, the degree of hypoplasia of the ventral tissues, and the subsequent ventral curvature and then performing a Gitte's test by injecting saline into corpora to see for the extent and location of maximum ventral curvature [8]. Correction of chordee can be done by penile degloving, mobilization of urethral plate and spongiosum, dorsal plication, urethral transaction, ventral corporotomies with or without graft, or a combination of these depending on the severity and degree of curvature in individual circumstances. Caution must be exercised while using dorsal plication for chordee correction as though this has the advantage of leaving the urethral plate intact. But it can cause penile shortening, which can be quite significant in this group of patients who already have a shorter than average penile length. Hence, overzealous usage of dorsal plication to straighten the penis to allow a one-stage procedure should be avoided. This increases the chance of recurrent curvature in later life, which will necessitate a complete redo surgery [6].

2. *Refashioning of the missing urethra (urethroplasty)*

In cases with mild curvature, a one-stage urethroplasty is feasible if the chordee is correctable by penile degloving, mobilization of the urethral plate with spongiosum, with or without dorsal plication without the need to transact the urethral plate. The usage of the popular TIP (tubularized incised plate) repair or an onlay flap procedure for proximal hypospadias is possible in these cases [21]. Preputioplasty is also feasible in some of these cases where the patients or parents opt for foreskin preservation.

In cases with severe curvature that necessitate urethral transection for chordee correction, options include a single-stage repair using preputial based flaps or a staged repair with

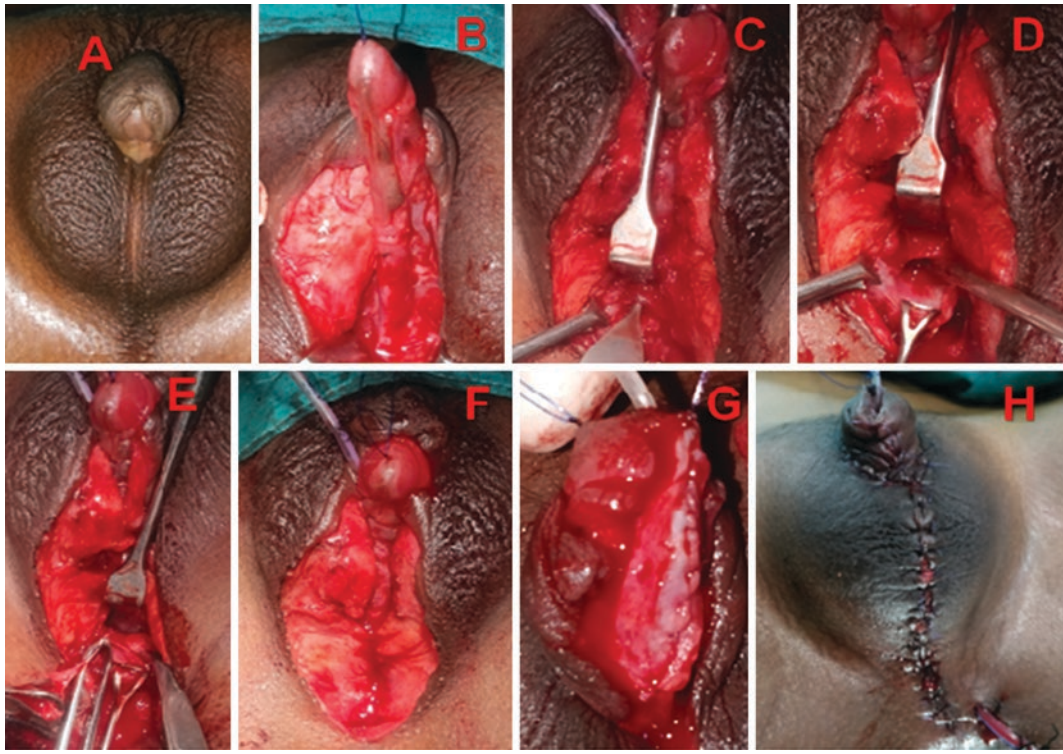

Fig. 23.6 Intraoperative pictures showing steps of tubularized urethral plate urethroplasty along with excision of the vagina. (**a**) Proximal penile hypospadias. (**b**) Penile degloving and proximal mobilization of common urethra canal. (**c**) Opening of vagina. (**d**) Mobilization of vagina. (**e**) Excision of vagina. (**f**) Closure of cavity after excision of the vagina. (**g**) Tubularization of the urethral plate. (**h**) Scrotoplasty, preputioplasty, and skin closure

preputial grafts, with the current trend towards choosing a staged approach as shown in Table 23.1 [4, 22–30]. In a staged approach, the technique is standard one as described by Bracka in redo hypospadias surgery where he used buccal mucosal grafts [31]. After transection of the urethral plate, a preputial graft is harvested from the dorsal hood, quilted to the ventral aspect of corpora and wrapped around the receded proximal urethral opening. Distally, glans is incised in the midline and opened in an open book fashion, and the graft is sutured to the wide-open glans to allow a tension-free glansplasty later on. If corporotomies were needed for chordee correction, the graft would act as a scaffold to promote the healing of the incised tunica albuginea of the corpora [6]. Following the grafting, a bolster dressing is applied, which is left undisturbed for a week to promote graft uptake. The second stage of the repair is usually done after six months. The graft is tubularized over a urethral stent, with a tunica vaginalis flap being commonly used as a second layer cover.

3. *Refashioning of skin*

Efficient management of skin is another important concern in these children, which directly correlates with cosmetic results, and this may be in the form of scrotoplasty or a correction for associated penoscrotal transposition. Depending on the degree of undervirilization, scrotoplasty can consist of simple mobilization of surrounding skin or complex mobilization and rotation of scrotal flaps to the midline [19]. Though this procedure can be safely performed simultaneously with urethroplasty, whenever there are concerns about the vascularity of the skin flaps, it is advisable to perform scrotoplasty as a separate procedure later.

In some patients, an associated penoscrotal transposition leads to additional complexity of the

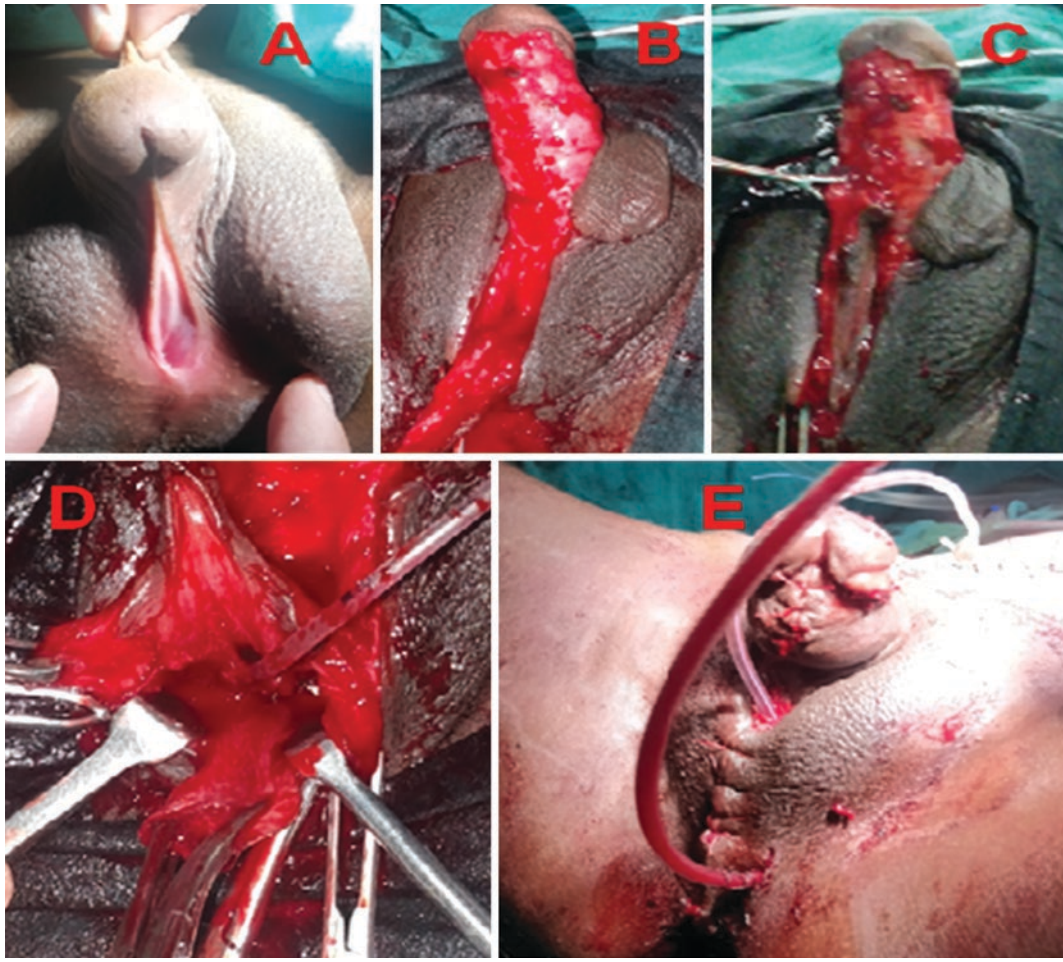

Fig. 23.7 Intraoperative pictures showing steps of two-stage repair including chordee correction, tubularization preserved urethral plate, and scrotoplasty. (**a**) Perineoscrotal hypospadias. (**b**) Transection and mobilization of the urethral plate with spongiosum to correct the chordee. (**c**) The mobilized urethral plate is preserved for later tubularization of the urethral plate. (**d**) Mobilization of proximal urethra and vagina. (**e**) Scrotoplasty and neo-urethral plate formation by Byars flap

repair, correction of which is usually achieved during the 1st or 2nd stage of the hypospadias repair; However, as mentioned above, in situations where the surgeon has doubts about vascular supply to the skin flaps due to extensive dissection during the hypospadias repair, it can be deferred to a later date and done as a stand-alone procedure [6].

4. A further important consideration in these children with DSD is that most of them will need additional procedures for the management of gonads and Mullerian structures.
   (a) *Management of gonads/testes*—Many of these children with hypospadias and DSD, especially those with partial gonadal dysgenesis, PAIS, or disorders of testosterone biosynthesis, will have associated undescended testis (UDT either unilateral or bilateral), which require to be managed along with the hypospadias. Orchidopexy in these patients can be done either before or during hypospadias repair, with the approach (open or laparoscopic) being decided by the position of the gonads [32]. In children with ovotesticular DSD, conservative gonadal surgery, with preservation of gonadal tissue that is concordant with the gender identity, is possible when

Table 23.1 Summary of the series of patients with hypospadias and DSD who underwent masculinizing genitoplasty

Series	No of patients, *n*	Types of DSD	Types of Hypospadias	Pre-op hormone treatment/mode of supplementation	Median age at first surgery, years	Procedure done	Auxiliary surgeries	Mean number of procedures *n*	No of complications, *n* (%)	Complications	Follow-up, years
Farkas [22] (1993)	16	46 XY=16	n.a	16/ 16, i.m injections	n.a	Primary =2 Staged=14	Orchidopexy =11 Testicular biopsy=5	n.a	6(37.5)	Meatal stenosis =2 UCF=3 Dehiscence=1	n.a
Chertin [23] (2005)	39	46 XY=25 46 XX=10 SC=4	Penoscrotal=30 Perineal=9	7/39, i.m injections	1.8	Primary=39	Gonadal biopsy=39 Gonadectomy=2 Transurethral incision of the utricle opening =1 Open removal of the utricle=1.	n.a	14(35.9)	Rec chordee=4 UCF=5 Dehiscence=3 Recurrent infection due to persistent vaginal pouch =2	6
Fekete [24] (2006)	25	46 XY=19 46 SC=6	n.a	n.a	n.a	n.a	n.a	n.a	5(20)	UCF=4 Penile deformity=1	n.a
Sharma [25] (2008)	356	46 XY=298 46 XX=7 SC=51	Penoscrotal= 242 (68%) scrotal =61 (17%), perineal =53 (15%)	351/356, local application after first stage	23.6 months	Primary=5 Staged=351	n.a	n.a	83(23.3)	UCF= 56 Stricture=12 Baggy urethra=8 Recurrent infection due to persistent vaginal pouch =5 Diverticula=3 Hair growth in urethra=5	n.a

Series	No of patients, *n*	Types of DSD	Types of Hypospadias	Pre-op hormone treatment/mode of supplementation	Median age at first surgery, years	Procedure done	Auxiliary surgeries	Mean number of procedures *n*	No of complications, *n* (%)	Complications	Follow-up, years
Sircili [26] (2010)	59	46 XY=44 46 XX=7 45XY/45 X0=8	n.a	n.a	6	Staged=59	n.a	2	43(72.8)	UCF=30 Stricture=13	14.1± 9.2
Sharma [27] (2012)	6	46 XX=6	n.a	no	14.2	Staged=6	Mullerian structures removal=6 Bilateral mastectomy=6	4.9	1(16.7)	Rec UTI due to persistent vaginal pouch =1	9.2
Palmer [28] (2012)	17	46 XY=8 SC=7 Others=2	Proximal shaft=2 Penoscrotal=3 Perineal=12	n.a	2.37	Primary=7 Staged =10	Orchidopexy= 8 (47.1) Gonad biopsy= 4 (23.5) Gonadectomy= 6 (35.3)	2.06	5(29.4)	n.a	2.07
Saltzman [29] (2018)	30	46 XY=26 46 XX=2 Others=2	Midshaft=2 Penoscrotal=9 Perineal=19	7(24.1)	9 months	Primary=6 Staged =24	Orchidopexy= 18	n.a	24(80)	UCF=8 Stricture=1 Dehiscence=8 Skin issue=4 Graft issue=1 Diverticulum=1	35.1 months
Ochi [4] (2019)	58	46 XY=51 46 XX=1 SC =4 47XY +21=2	Perineal=26 Scrotal=16 Penoscrotal=15 Midshaft =1	n.a	n.a	Primary=6 Staged =52	Orchidopexy= 17(29.3)	n.a	8 (13.8)	Stenosis=3 Diverticulum=2 UCF=2 Rec curvature=1	5.16 ± 0.56

Key: *n.a.* not available, *DSD* Disorders of sex development, *SC* Sex chromosomal, *UCF* urethrocutaneous fistula

guided by intraoperative frozen section analysis of gonadal tissues to define the margins between ovarian and testicular components [33]. Rarely, when there is a high risk of malignancy (dysgenetic gonads), gonadectomy is recommended, (Fig 23.8b) which can be combined with concurrent testicular prosthesis insertion in young adults [19, 34].

(b) *Mullerian structures*—In DSD patients, rudimentary Mullerian structures are not an unusual occurrence. Among these, a commonly encountered structure during hypospadias repairs is the prostatic utricle, a cystic structure that communicates with the urethra at the level of the verumontanum. Apart from causing some difficulty in catheterization during or after hypospadias repair in these patients, most utricles are asymptomatic and do not affect urethroplasty. Rarely, it can become symptomatic due to the stone formation or vasal efflux leading to recurrent episodes of epididymorchitis, which necessitates its removal either laparoscopically or through an open sagittal posterior approach [35–37]. Other Mullerian duct structures like vagina and hypoplastic uterus with fallopian tubes require excision (Figs. 23.8a and 23.6d). Resection of these structures may be combined with the hypospadias surgery or can be done separately.

23.7.2 Complications

In general, complications following HP repairs can be divided into anatomical (urethral healing failures (fistula, dehiscence), urine flow impairments (stenosis, diverticulum), persistent penile curvature) and functional (poor cosmetic results, sexual disorders) complications. The anatomical complications like urethrocutaneous fistulas, urethral strictures, meatal stenosis, and glans dehiscence usually present in the immediate postoperative period and will require surgical intervention, which may vary from a simple meatal dilatation to complete redo repair.

Being a more complex surgery than distal repairs, complication rates for proximal hypospadias repair are significant, and rates in excess of 50% have been historically reported [21, 38–40]. Unfortunately, very few studies have focused on the outcomes following HP repairs in patients with a definitive diagnosis of DSD [4, 22–29]. As shown in Table 23.1, among the few studies that have studied outcomes in this cohort of patients with DSD, the complication rate ranges between 13 and 72%. Regarding the question of whether there is a difference in outcomes in children with and without DSD diagnosis, Palmer et al. showed that though boys with a specific DSD diagnosis have significantly more atypical anatomy and are more likely to require procedures for the management of the gonads, they do not have an increased risk of complications or number of surgeries [28]. However, this observation has been refuted by Saltzman et al., who in a more recent study observed that proximal hypospadias repair on patients with DSD is associated with higher reoperation rates in the first two years than a standard proximal hypospadias repair (80 vs 45.9%), with staged repairs being a particular risk factor for reoperation [29]. A similar observation has been made by Lucas-Herald et al. also who, in their study on outcomes in partial androgen insensitivity syndrome, observed that young men with an AR mutation were more likely to have multiple procedures [41]. Ultimately, the bottom line is proximal hypospadias repair with or without DSD is still a challenging and humbling experience, even for the most experienced surgeons. This aspect should be truthfully informed to all the stakeholders involved [6]. More importantly, since many of these patients will need to undergo multiple genital surgeries to ensure good anatomical and functional results, including enabling optimal sexual functioning, all clinicians involved in managing this challenging condition should emphasize the need for long-term follow-up in these patients and caregivers.

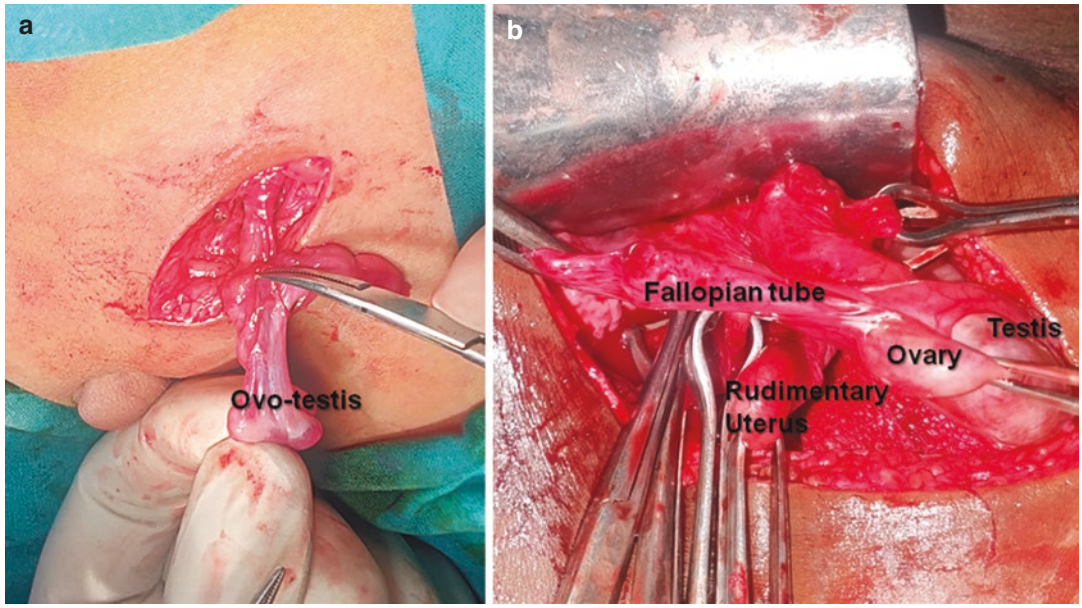

Fig. 23.8 Intra-operative picture showing Mullerian duct strictures. (**a**) Ovotestis in hernia sac of undescended testis. (**b**) Testis, Ovary, Fallopian tube, and rudimentary uterus

23.8 Long-term Follow-up and Outcomes

As mentioned earlier, an important aspect of hypospadias repair that has been stressed time and again has been the need for long-term follow-up to assess long-term functional outcomes. This is all the more important in this unique subgroup as, along with the anatomical results, the aspect of gender identity also comes into play as these children get older. Further, many patients, specifically those with 46 XY DSD, will need hormone supplementation in monthly intramuscular testosterone to induce puberty and maintain adequate hormone levels through adulthood [42, 43].

In recent years, validated instruments like the Pediatric Penile Perception Score (PPPS), the Penile Perception Score, Hypospadias Objective Scoring Evaluation (HOSE) have been developed to report patient perceptions of anatomical and functional outcomes hypospadias repair [6]. There have also been studies that have reported on the patient-reported urinary and sexual function outcomes in adults who had undergone masculinizing genitoplasty for DSD in childhood [42–47]. In the majority of these series, although specific complaints about reduced penile length, difficulties in voiding and sexual activity were frequently reported, patients were overall satisfied with the long-term results of masculinizing genitoplasty. In one of the earliest studies by Miller et al., men who underwent staged hypospadias repair for severe ambiguous genitalia with perineoscrotal hypospadias had continued difficulty with both ejaculation and micturition, but no major psychiatric or psychological disturbance [47]. Some of the common problems reported by adults, who had undergone masculinizing genitoplasty in childhood are dissatisfaction due to reduced penile length, poor or premature ejaculation, erectile dysfunction that sometimes translate into even fear of sexual contact. In general, good results may be expected if the initial phallus size is adequate (with or without testosterone supplementation); however, the results tend to be poor in cases with micropenis

and minimal virilization [24, 27]. Interestingly, despite the problems, men with DSD are more likely to be satisfied than women with regard to their surgical outcomes and sexual function [19, 26, 45]. Presently, prospective collection of patient-centric data in this group of proximal hypospadias with DSD is an active area of research. This data is vital to improve care in this difficult condition further [6].

23.9 Conclusion

To summarize, patients with hypospadias and a diagnosis of DSD are a unique subpopulation of patients with proximal hypospadias where management includes both surgical and psychosexual care for promoting positive adaptation. Because of the predominantly proximal type of hypospadias seen in these patients, two-stage repair with a preoperative course of testosterone gives better anatomical and functional results in most of these patients [27]. An individualized approach with a focus on integrated communication between the multidisciplinary medical team and the patient's family, which continues beyond the initial post-surgical period to adulthood, is crucial for successful outcomes.

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