

Invited Review

## Surgical management of 46,XY differences in sex development

Kanishka Das<sup>1</sup>, Mainak Deb<sup>2</sup>, Attibele Mahadevaiah Shubha<sup>3</sup>, Akash Bihari Pati<sup>1</sup>

<sup>1</sup>Department of Pediatric Surgery, All India Institute of Medical Sciences, Bhubaneswar, Odisha, <sup>2</sup>Department of Pediatric Surgery, Rainbow Childrens' Hospital, Hyderabad, Telangana, <sup>3</sup>Department of Pediatric Surgery, St. John's Medical College, Bengaluru, Karnataka, India.



**\*Corresponding author:**

Kanishka Das,  
Department of Pediatric  
Surgery, All India Institute of  
Medical Sciences, Bhubaneswar,  
Odisha, India.

[kanishkadas@hotmail.com](mailto:kanishkadas@hotmail.com)

Received: 14 November 2025  
Accepted: 14 November 2025  
Epub Ahead of print: 02 January 2026  
Published: 23 February 2026

DOI  
[10.25259/JPED\\_81\\_2025](https://doi.org/10.25259/JPED_81_2025)

Quick Response Code:



### ABSTRACT

The surgical management of differences in sex development (DSD) has evolved over the decades with advances in diagnostic methods rendering clarity to the etiopathogenesis, surgical anatomy, natural history, and principles of treatment of various forms of 46,XY DSD. Cumulative insights into intermediate and long-term results of surgical cosmesis, functional outcomes, and gender dysphoria have tempered the surgical strategies with a global moratorium on elective interventions till the age of consent. Besides the pros and cons of early versus late surgery for 46,XY DSD, the narrative elaborates on the indications, surgical principles, timing of surgery, salient details of procedures, and their outcomes. Masculinizing procedures include gonadal surgery, penile reconstruction, surgery for urogenital sinus (UGS) and/or Müllerian remnants, and breast reduction. Likewise, feminizing procedures encompass clitorophallic reduction, surgery for UGS and/or vaginoplasty, gonadectomy, and breast augmentation. Also included are algorithms for surgical management in broad categories of 46,XY DSD and illustrative cases.

**Keywords:** 46,XY differences in sex development, Androgen insensitivity syndrome, Feminizing genitoplasty, Masculinizing genitoplasty, Persistent Müllerian duct syndrome, Surgical management

### INTRODUCTION

Surgical management options for children with differences in sex development (DSD) are continually evolving in the light of the changing attitudes and a greater awareness of the psychosocial and ethical issues that are involved in the treatment of these children. Historically, surgical management of the genitals (external genitalia and internal genital ducts) and gonads was aimed to benefit the child physically and psychosocially, yet perceived collateral harm in some has prompted a moratorium on all surgical procedures during childhood, excepting emergent ones. However, both in the developed and developing world, particularly the latter, parents continue to make decisions on surgical options for the young child with DSD to be able to rear the child in the prevalent society. With emerging statutory restrictions and consensus guidelines in this arena, it is important to offer a comprehensive multidisciplinary team (MDT) care through a shared decision-making (SDM) process.<sup>[1]</sup>

The diagnosis of 46,XY DSD is first established with a detailed clinical evaluation and systematic, directed algorithmic investigations.<sup>[2]</sup> A select panel of individualized investigations with combinations of genetic studies (including karyotype, fluorescent *in situ* hybridization, whole-

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, transform, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

©2025 Published by Scientific Scholar on behalf of Journal of Pediatric Endocrinology and Diabetes

exome sequencing, and androgen receptor [AR] studies), imaging (e.g., ultrasonography and pelvic magnetic resonance imaging), hormonal profile (including luteinizing hormone, follicle-stimulating hormone, and human chorionic gonadotropin-stimulated androgen levels), and additional relatively invasive studies (e.g., genitoscopy, laparoscopy, and gonadal biopsy) narrows the diagnosis to one of the three entities—central hypogonadism (hypothalamic–pituitary origin), a gonadal defect (androgen biosynthesis and conversion defects) or peripheral tissue unresponsiveness (androgen insensitivity). The varied anatomical and physiological issues that are addressed at surgery are related to the role of anti-Müllerian hormone (regression of Müllerian duct), testosterone (development of testis, epididymis, vas deferens, and seminal vesicle) dihydrotestosterone (development of prostate, conversion of urogenital sinus (UGS) to urethra, and development of penis and scrotum) in the prenatal development of internal and external genitalia.

There are compelling arguments for both early and delayed surgery in DSD.<sup>[3,4]</sup> However, the lack of registries, high-quality evidence, and longitudinal data on the outcomes limits the provision of educational material and informed decision-making [Table 1]. The surgical management is tailored to the assigned sex of rearing and endorsed after

detailed deliberations at the MDT.<sup>[5]</sup> Surgical interventions must suit the sex of rearing, generally male/female, or be held in abeyance till a decision is reached by a legally consenting individual. These may be emergent, semi-emergent, or elective. Emergent surgical procedures are mandatory and aim at creating unobstructed outlets for urinary and/or genital secretions or preventing permanent disability. Optional elective procedures address issues of gonadal and genital tract preservation, genital and breast cosmesis and/or desired appearance, voiding, sexuality, and procreation. A calendar of interventions is chalked out for the specific child with the minimum anesthetic exposure, and all interventions are supported by counseling sessions to foster psychosexual well-being.

### MASCULINIZING GENITOPLASTY IN 46,XY DSD

Patients assigned to or have opted for a male sex of rearing after the review by the MDT are planned for a masculinizing genitoplasty.<sup>[6]</sup> Hypospadias in DSD tends to be more severe and is associated with micropenis, bifid scrotum, and undescended testis. In addition to the paucity of tissue for reconstruction, hormonal deficiency and tissue responsiveness limit surgical success. The procedures aim at creating cosmetically acceptable and functional

**Table 1:** Early versus delayed surgery in 46,XY differences in sex development (DSD): Pros and cons.

	Pros	Cons
Early surgery	<ul style="list-style-type: none"> <li>• Emergency interventions avert life-threatening circumstances</li> <li>• Ambiguous genital anatomy – difficult childhood, poor social/parental support, parental rift/divorce</li> <li>• Gender identity (individualized, subjective) develops based on a defined genital/sexual anatomy, assigned sex of rearing, and social recognition during development</li> <li>• Genital surgery after careful investigation (anatomy, physiology, imprinting) reinforces the child’s sexual identity for developing the gender identity</li> <li>• Caters to self-initiated gender assignment</li> <li>• Recent studies showed a low sex reassignment rate and majority of 46,XY DSD were satisfied with their childhood surgery</li> </ul>	<ul style="list-style-type: none"> <li>• Decision making involves parents only, latent medical paternalism</li> <li>• Primary driving factors – parental anxiety, shame, desire for secrecy, stigma. Haste to “normalize” child’s appearance before being “fully informed” about options, risks, and benefits</li> <li>• Too early surgery – little tissue technically, disposes tissue that could be useful in later revision/reconstruction. Involves “irreversible” surgical interventions</li> <li>• Reports of unfavorable outcomes, complications, reoperation/revision rates, and patient dissatisfaction</li> <li>• Gonadectomy – under strong focus, loss of hormones, and fertility potential</li> <li>• Anesthetic harm in early life – multiple exposures!</li> </ul>
Delayed surgery	<ul style="list-style-type: none"> <li>• SDM is patient centered with partial/full disclosure (explicit acknowledgment, best available evidence discussed, considers patient’s/family’s values and preferences and provider’s guidance, decision support tools)</li> <li>• Respects patient autonomy and preserves options for future – body/physical, fluidity in gender choices/identity</li> </ul>	<ul style="list-style-type: none"> <li>• No solid research data on age when a child can make decisions on genital/gonadal surgery. Lack of evidence-base</li> <li>• Patient autonomy and respect are a social experiment with unknown consequences</li> <li>• Do we have a “cultural space” for a third gender/non-binary assignment?</li> <li>• Late surgery – too little tissue for the age!</li> </ul>

SDM: Shared decision-making, DSD: Differences in sex development.

external genitalia that allow penetrative intercourse.<sup>[7]</sup> The principles of surgical correction address varying degrees of discordance between chromosomal sex and external genital phenotype, gonadal position and anatomy, and the development of internal genital ducts—both mesonephric and paramesonephric [Table 2].

### Gonadal interventions

*Orchidopexy.* Most patients of 46,XY DSD undergoing masculinizing genitoplasty are likely to have testes, many of which are cryptorchid. These testes require early orchidopexy to best preserve potential for fertility. Inguinal testes may be brought down in a single stage into the labioscrotal folds with high success rates. Intra-abdominal testes are best managed laparoscopically; the low ones can be brought down into the scrotum in a single-stage operation. The high intrabdominal testis may be managed in a single-stage or two-stage procedure by the *Fowler–Stephen (FS) procedure*. This procedure involves division of the testicular vessels and dependence on collateral blood supply from two other sources – artery to the vas and cremasteric artery; hence, a single-staged procedure has a higher testicular atrophy rate compared to two-staged procedure where a gap of a few months is allowed

for the collaterals to develop. The *Shehata technique* is a two-stage procedure which employs traction lengthening of the testicular pedicle without vascular transection. The testicular atrophy rates of this procedure are comparable to those of the staged FS procedure.<sup>[8]</sup>

*Orchidectomy.* It is rare that malignant change occurs in a cryptorchid testis before the third decade of life; hence, orchidectomy is singularly uncommon in childhood. Dysgenetic gonads in 46,XY DSD are beyond the scope of this discussion. However, partial androgen insensitivity syndrome (PAIS) carries a 15–25% lifetime risk of malignant change and surveillance is easier in a scrotal testis.<sup>[9]</sup>

*Testicular prosthesis,* commonly made of silicone, may be chosen by an adolescent with unilateral scrotal testis in select cases—testicular regression, testicular atrophy after orchidopexy, or other surgery. This is ideally undertaken only after completion of pubertal scrotal growth to minimize prosthesis replacements.

### Penile reconstruction

The primary concerns in penile reconstruction are correction of ventral curvature/chordee (orthoplasty) and creation of a neourethral tube from the hypospadiac meatus to as close to the glans tip (urethroplasty).

*Orthoplasty* involves a stepwise surgical progression depending on the severity. Complete degloving of the penile skin may suffice in mild or skin-level tethering. Minor residual chordee thereafter is corrected with dorsal tunical plication, the obvious disadvantage being foreshortening of the already short phallus. More severe chordee requires further techniques such as urethral plate mobilization with the underlying corpora spongiosal delta, urethral plate division, and corporal lengthening procedures (ventral superficial or deep corporotomy incisions) to achieve a complete straightening.<sup>[7,10]</sup>

*Urethroplasty* in proximal and severe hypospadias that are common in DSD are conventionally staged but can be done in a single stage along with orthoplasty. The choice between the two depends on the surgical anatomy, tissue adequacy, vascularity, androgen responsiveness, and surgical expertise. Cases with mild chordee where the urethral plate can be preserved are suitable for single-stage urethroplasty using a variety of techniques dictated by the surgeon's preference. Commonly employed options are the tubularized incised urethral plate urethroplasty (*Snodgrass*) or a preputial flap onlay repair (*Duckett, Asopa*). The *Koyanagi technique* can also be used to achieve chordee correction and urethroplasty in a single sitting if the preputial anatomy is favorable.

**Table 2:** Aims and indications for genitoplasty in 46,XY differences in sex development.

A. Aims of surgical interventions
<ul style="list-style-type: none"> <li>• Prevent urinary/genital tract outflow obstruction</li> <li>• Restoring near normal external genital appearance</li> <li>• Functional anatomy to allow penetrative intercourse</li> <li>• Preservation of fertility</li> <li>• Gonadal concordance with sex of rearing</li> <li>• Mammoplasty – reduction/augmentation</li> <li>• Foster psychosocial well-being and social acceptance</li> </ul>
B. Indications for masculinizing surgery
<ul style="list-style-type: none"> <li>• Phallus – small/buried phallus</li> <li>• Ventral penile curvature – chordee</li> <li>• Hypospadiac external urethral meatus</li> <li>• Undescended testes</li> <li>• Scrotal anomalies – labioscrotal folds, bifid scrotum, penoscrotal transposition</li> <li>• Müllerian duct and urogenital sinus remnants</li> <li>• Mammoplasty – reduction</li> </ul>
C. Indications for feminizing surgery
<ul style="list-style-type: none"> <li>• Phallus – large</li> <li>• Urogenital sinus, hypospadiac urethral meatus</li> <li>• Testicular gonad</li> <li>• Labial anomalies – posterior labial fusion, hypoplastic labia</li> <li>• Vaginal anomalies – absent/short vagina</li> <li>• Mammoplasty – augmentation</li> </ul>

Composite repairs using a combination of urethral plate tubularization (*Duplay*) along with tubularized preputial flaps (*Duckett*) have been used. In proximal hypospadias with severe chordee, orthoplasty is achieved with tissue cover for the ventral raw area using hairless skin, inner preputial tissue (vascularized or free graft), or free labial/buccal mucosa graft (*Bracka*) in the first stage and a delayed urethroplasty as a second stage. The two-stage procedure is the favored approach in most patients and is associated with fewer complications and better outcomes.<sup>[7,10,11]</sup>

*Phalloplasty*, or reconstruction of a penis, is a rare surgical undertaking and indicated in select cases of 46,XY DSD—aphallia (congenital absence of the penis), micropenis (isolated - gonadotropin failure, associated with hypospadias or gonadal abnormalities in primary testicular failure), severe undervirilization syndrome (e.g., PAIS), and some children with bladder or cloacal exstrophy. It is a multistage operation using various free/pedicled flaps to reconstruct the penis and the urethra; neurotization allows for sensation in the neophallus.<sup>[12]</sup>

### 3. UGS remnant or vaginal pouch and Müllerian remnant/prostatic utricle

Some children with 46,XY DSD and proximal hypospadias, especially due to 5-alpha reductase type 2 (5ARD2) deficiency, have UGS remnants/pseudovaginal pouches distal to the verumontanum with varying length and caliber. They are largely asymptomatic;<sup>[13]</sup> small ones are left alone while larger accessible ones are excised or incorporated into the proximal repair during urethroplasty.<sup>[14]</sup>

Müllerian remnants and prostatic utricles open at the verumontanum in the rare persistent Müllerian duct syndrome (PMDS). The prostatic utricle is usually asymptomatic. Most symptomatic ones are managed by cystoscopic widening of the mouth to facilitate drainage. A large remnant is excised to eliminate recurrent urinary tract infections (UTIs) and epididymo-orchitis and reduce the risk of endometrial malignancy. The anomalous vas deferens may be entirely or partly embedded in the wall, particularly at its caudal end, and is prone to injury or devascularization during excision. Where complete excision is not safely feasible, it is longitudinally divided with cautious removal of mucosa; this division also facilitates bilateral orchidopexy with ease. An objective, practical guide to the approach, longitudinal splitting or division, or near-total excision has been proposed based on three anatomical parameters: gonadal status, relation to the vas, and degree of Müllerian development.<sup>[15]</sup> Excision can be achieved by laparoscopy aided by cystoscopic guidance or other open

routes (perineal, transperitoneal, and transvesical anterior sagittal with/without splitting the anterior rectal wall) with risk of seminal vesicle injury.<sup>[16,17]</sup> Except for the large ones, these are addressed after the urethroplasty is complete. A low resistance, good caliber neourethra minimizes complications arising from these remnants and many remain asymptomatic. Symptoms are due to retention of urine, UTIs, or mechanical obstructions to the flow of urine and rarely due to mucosal secretions.

A recent debatable approach proposes leaving a male vagina or perineal utricle *in situ* in boys with perineal hypospadias to minimize genital tissue removal in children without a definite gender identity to facilitate future gender reassignment surgery.<sup>[18]</sup>

### Management of breast

Gynecomastia is a prominent clinical feature in nearly 80% of adolescent boys and men with PAIS with *AR* gene variant. This is a cause of psychological distress and hence requires treatment. Medical management includes administration of selective estrogen receptor modulators (tamoxifen, raloxifene, and clomiphene citrate), aromatase inhibitors (testolactone, anastrozole, and letrozole), and androgens. However, their efficacy in PAIS is comparatively modest than in other types of pathological gynecomastia and many require surgical intervention eventually. Bilateral mastectomy by circumareolar or sub-mammary approach yields good results.<sup>[19]</sup>

### FEMINIZING GENITOPLASTY IN 46,XY DSD

Select children with 46,XY DSD (e.g., typical female genitalia as in complete androgen insensitivity syndrome (CAIS) or atypical genitalia more akin to females, e.g., PAIS with microphallus reared as females or those with a female gender identity) are candidates for feminizing genitoplasty.<sup>[20]</sup> The timeline for surgery remains controversial. Surgery during infancy relies on better quality of the genital tissues and vascularization with postnatal maternal estrogens. Early surgery aligns with the sex of rearing and the phenotype and allays parental anxiety about the external genital appearance. However, vaginal dilation is not optimal in childhood, and evolving stenosis requires revisional surgery at puberty. Testicular gonadectomy is scheduled after completion of puberty, after the age of consent and before malignant transformation sets in.

Feminizing genitoplasty involves reconstruction of the genitals to appear and function in a typical female pattern. Important goals include achieving a feminine external genital

appearance, preserving genital sensitivity, and providing a vaginal structure for satisfactory coitus.<sup>[21]</sup>

### Clitorophallic reduction

Clitorophallic reduction with dorsal neurovascular bundle preservation has replaced complete removal. The reduced phallic structure is anchored at the pubis; its investment retains erotic sensitivity and improves sexual function. In addition, the redundant phallic skin after reduction is used to reconstruct the labia minora that may be missing. Careful vascular preservation prevents necrosis or atrophy of the clitoris. Likewise, large labioscrotal folds may be reduced by a Y-V technique.<sup>[22]</sup>

A more conservative surgical approach of corporeal sparing dismembered clitoroplasty (*Pippi Salle*) preserves the enlarged corpora. The hemi-corpora after separation from one another are rotated laterally and inferiorly and buried in the subdartos plane within the labia majora. The retained corpora in the labia majora have a certain mobility, permitting painless erections and scope for reversibility using the retained erectile tissue for phallic reconstruction in case of gender dysphoria in the future. Apparently, psychologically, it eludes the feeling of castration sensed by some with the excision of the erectile tissue. Anecdotal unpleasant labial engorgement during clitoral erection may be addressed by removing the hemi-corpora through labial incision.<sup>[23]</sup>

### Surgery for UGS and vaginoplasty

A common urethra and vaginal channel (UGS) in some forms can be associated with post-void dribbling (vaginal voiding), recurrent UTI, and difficulty with coitus. Its length at cystogenitoscopy dictates the choice and complexity of the surgical procedure. These include minor procedures such as cut-back incision (for two separate urethral and vaginal introital orifices, but covered by skin due to mild posterior labial fusion or cranially extended fourchette) and Y-V perineal flap (for vaginal introital stenosis) and partial urogenital mobilization (urethra and vagina mobilized as one vascular unit distal to the pubo-urethral ligaments to preserve urinary continence). More extensive procedures that are used to exteriorize both orifices in a high confluence in congenital adrenal hyperplasia, like Passerini–Glazel procedure and partial or total urogenital mobilization, are rarely employed here.

Most patients with CAIS have an adequate vaginal length that is amenable to dilatation and does not need augmentation. In PAIS, an adequate vagina for satisfactory coitus may be

created by dilatation of an existing vaginal pouch by the patient (vaginal dilators or molds) or surgically (Vecchiotti procedure with acrylic olive) or by creating a neovagina with variety of material (McIndoe–Reed vaginoplasty [split skin], Davydov procedure [peritoneal flap], or intestinal [ileum or sigmoid vaginoplasty]), each with its outcomes and specific complications.

### Gonadectomy

At present, gonadal tumor risk prediction and timing of gonadectomy are based on the molecular diagnosis. In CAIS, both testes are retained to achieve spontaneous puberty, optimal breast and bone development due to high levels of androgens being converted to estrogen by aromatase. The risk of testicular malignancy in CAIS is low before adulthood. In PAIS, the risk of early malignant transformation is higher, warranting gonadal placement in an accessible location for careful surveillance till puberty and early gonadectomy thereafter. In PAIS and some CAIS, gonads are inguinal; the intrabdominal gonads in CAIS are managed with a laparoscopic approach.<sup>[24,25]</sup>

### Management of breast

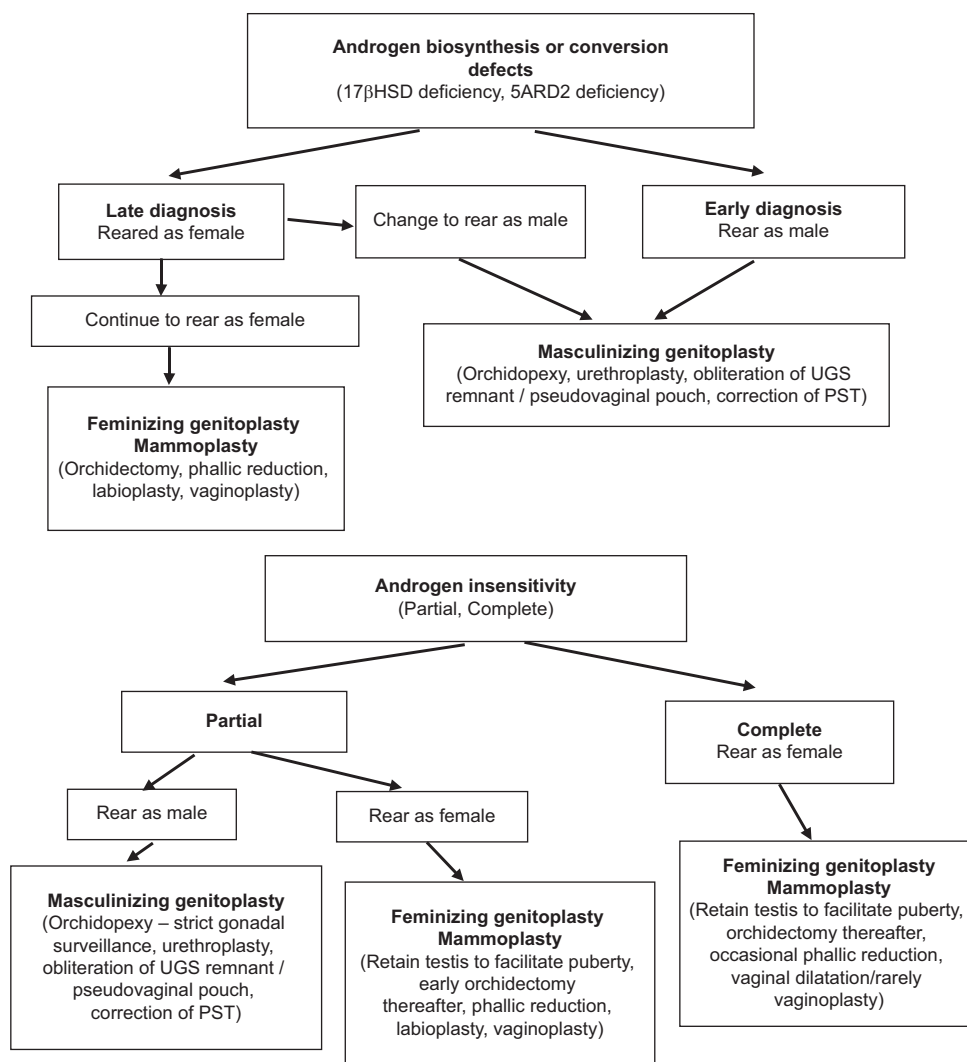
Children with 17 $\beta$ -hydroxysteroid dehydrogenase (17 $\beta$ HSD) deficiency and occasionally in PAIS may require breast augmentation either by medical (hormonal supplementation) or surgical (insertion of silicon prosthesis) means.

The management is individualized according to the diagnosis and sex of rearing. Figure 1 illustrates the summary of surgical treatment for specific categories of 46,XY DSD—testosterone biosynthesis or conversion defects and androgen insensitivity syndromes. Illustrative cases are depicted and detailed in Figures 2-4.

## COMPLICATIONS AND OUTCOME OF SURGICAL MANAGEMENT IN 46,XY DSD

There has been a gradual shift from early surgery based on the optimal gender policy to SDM, full disclosure, and delayed surgery. However, published literature on long-term outcomes is sparse, particularly where prenatal androgenization (as in 5ARD2 deficiency, 17 $\beta$ HSD deficiency and PAIS) renders them prone to gender dysphoria.<sup>[26]</sup>

Complications in genital surgery vary (20–60%) and are often seen beyond 2 years too. Besides surgical expertise, patient factors such as genetic variants (*AR* variant), additional comorbidities, complex urogenital malformations, suboptimal anatomy, and severe under-masculinization play a pivotal



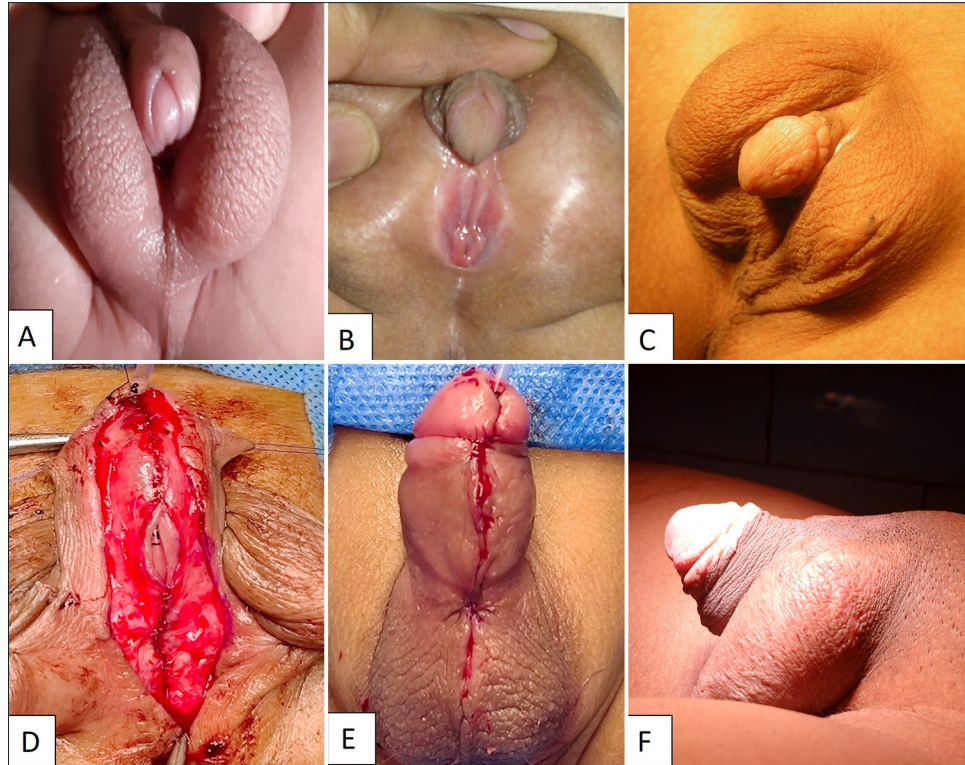
**Figure 1:** Summary of surgical management for specific categories of 46,XY DSD. 17βHSD :17-beta-hydroxysteroid dehydrogenase, 5ARD2: 5-alpha-reductase type 2, PST: Penoscrotal transposition, UGS: Urogenital sinus, DSD: Differences in sex development.

role.<sup>[27,28]</sup> The DSD life study focused on a twin assessment of physician review (clinical and physical examination) and patient-reported outcomes on both genital appearance and function and correlated the findings with diagnosis, procedures, and examination.<sup>[29]</sup> In general, patients operated for hypospadias and undescended testis were satisfied with early surgery; but procedures that patients consented to themselves (e.g., mammoplasty) had a better impact on their lives.

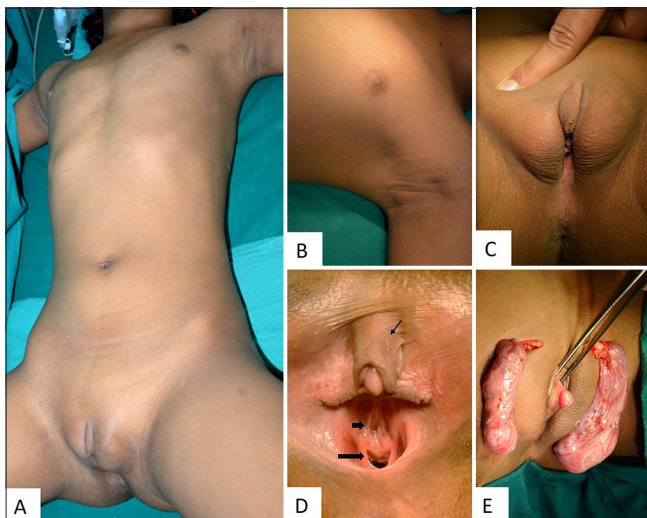
Self-reported satisfaction is usually lower than the observer's evaluation for both cosmesis and function.<sup>[30]</sup> The satisfaction levels with genital appearance were rated higher by physicians while those with genital function were rated higher by the patients. Two studies (in 2011 and 2022) separated by a decade showed a similar percentage of satisfaction

with appearance (37–38%) and function (20–29%), the satisfaction in non-operated cases being much lower.<sup>[30,31]</sup>

Hypospadias surgery for proximal hypospadias was the most common procedure and was performed early. Of these 60% had a revision, largely for stenosis and fistulae. Penile length was shorter than peers (shortest with PAIS) and correlated with satisfaction with appearance. Staged procedures seemed to have a better potential to preserve the intrinsically short penile length during reconstructive procedures. Glans sensitivity was normal in less than 50%, probably due to lack or loss of foreskin, dorsal plication, and other surgical maneuvers and could explain sexual dissatisfaction. Penile erections and orgasms were fairly satisfactory in the majority; 25% had erectile dysfunction, delayed orgasm, or premature ejaculation. Besides these, men with PAIS reported higher



**Figure 2:** 5-alpha reductase (5ARD2) deficiency: (A-C) The severe chordee is corrected and the vaginal pouch excised in the first stage of the repair, while (D and E) urethroplasty and correction of penoscrotal transposition are tackled in the second stage to achieve (F) a near normal circumcised phallic appearance.



**Figure 3:** Partial androgen insensitivity syndrome (PAIS): A 13-year-old child, reared as female and presented at puberty with atypical genitalia and recent change in voice. Note the feminine habitus (A and B), poor breast development (B), and sparse axillopubic hair (B and C). Both gonads were palpable in labioscrotal folds (C) beside a short phallic structure (thin arrow, D). There were two orifices: the external meatus (short arrow, D) and vaginal introitus (long arrow, D). The management included bilateral orchidectomy (E), vaginal dilatation, and hormonal supplementation.

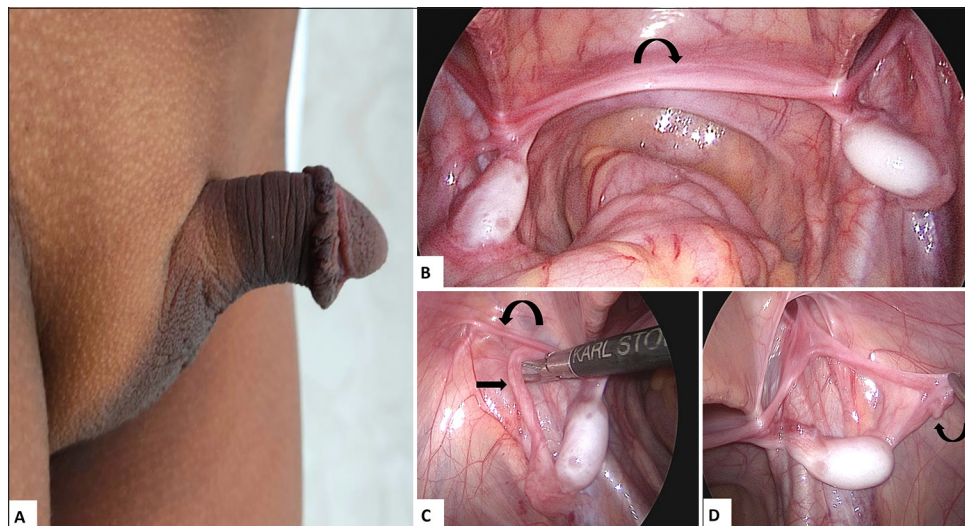
dissatisfaction with penile shape and urogenital infections.<sup>[30]</sup> Most patients undergoing phalloplasty can achieve standing micturition and penetrative intercourse and are satisfied with the cosmetic and functional outcomes.<sup>[32]</sup>

In 46,XY DSD, partially androgenized females, feminization surgery yielded mixed results with problems in clitoral arousal (47%) and dyspareunia (57%); several had partners of female (9%) or both sexes (18%) too. Those with CAIS had significant problems with desire (82%), arousal (64%), and dyspareunia (70%) too.<sup>[33]</sup>

The participants were mostly dissatisfied after gonadectomy, even if they were largely performed to ameliorate the risks of neoplasia. Outcomes with regard to sexuality were varied and inconsistent; a satisfactory sex life seemed to depend on other factors besides genital appearance and function, e.g., partner support, coping skills, and hormonal balance. Thus, long-term satisfaction with esthetic and functional outcomes depended on both initial anatomy and the surgeries performed and not necessarily on the specific diagnosis.

## CONCLUSION

The tenets of surgical management in 46,XY DSD include multidisciplinary care, establishing a molecular diagnosis,



**Figure 4:** Persistent Müllerian duct syndrome (PMDS): A 6-year-old child, reared as male, presented with bilateral impalpable undescended testis, symmetrical hypoplastic rugose scrotum and a normal penis (A). Laparoscopy, (B, C and D) revealed bilateral low intraabdominal testis with a symmetrical Müllerian remnant. Note (B) the slender uterus (arrow), (C) left round ligament (curved arrow) and fallopian tube (straight arrow) and (D) right fimbriae (curved arrow). A midline division of the uterus with endometrial denudation, fallopian tube excision and bilateral orchidopexy was done.

charting individualized surgical calendar for a given patient, integrating research data from patient registries, and understanding long-term outcomes to moderate specific medical and surgical interventions. Areas that need continuing study include changing epidemiology, morbidity relating to specific etiologies, optimal options for genitoplasty, and quality of life in terms of genital cosmesis, sexuality, and fertility.

**Ethical approval:** Institutional Review Board approval is not required.

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship:** Nil.

**Conflicts of interest:** There are no conflicts of interest.

**Use of artificial intelligence (AI)-assisted technology for manuscript preparation:** The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

## REFERENCES

- Gardner M, Sandberg DE. Navigating surgical decision making in disorders of sex development (DSD). *Front Pediatr* 2018;6:339.
- Das K. Approach to diagnosis in children with differences of sex development. In: Ratan SK, editor. *Children with differences in sex development*. Singapore: Springer; 2024:185-8.
- Hemesath TP, De Paula LC, Carvalho CG, Leite JC, Guaragna-Filho G, Costa EC. Controversies on timing of sex assignment and surgery in individuals with disorders of sex development: A perspective. *Front Pediatr* 2019;6:419.
- Weidler EM, Grimsby G, Garvey EM, Zwayne N, Chawla R, Hernandez J, et al. Evolving indications for surgical intervention in patients with differences/disorders of sex development: Implications of deferred reconstruction. *Semin Pediatr Surg* 2020;29:150929.
- Sarin YK, Singh D, Babu R, Das K, Rao S, DSD Guidelines Committee of Indian Association of Pediatric Surgeons. Indian association of pediatric surgeons guidelines on the management of differences in sex development. *J Indian Assoc Pediatr Surg* 2022;27:376-80.
- Wisniewski AB, Batista RL, Costa EM, Finlayson C, Sircili MH, Dénes FT, et al. Management of 46,XY differences/disorders of sex development (DSD) throughout life. *Endocr Rev* 2019;40:1547-72.
- Mouriquand PD, Gorduza DB, Gay CL, Meyer-Bahlburg HF, Baker L, Baskin LS, et al. Surgery in disorders of sex development (DSD) with a gender issue: If (why), when, and how? *J Pediatr Urol* 2016;12:139-49.
- Bidault-Jourdainne V, Botto N, Peycelon M, Carricaburu E, Lopez P, Bonnard A, et al. Staged laparoscopic orchidopexy of intra-abdominal testis: Spermatic vessels division versus traction? A multicentric comparative study. *J Pediatr Urol* 2024;20:498.e1-8.
- Ovidiu B, Marcu DR, Mischianu DL, Poiana C, Diaconu CC, Bungau SG, et al. The challenges of androgen insensitivity syndrome. *Arch Med Sci* 2022;18:881-9.
- Chertin B, Koulikov D, Hadas-Halpern I, Farkas A.

- Masculinizing genitoplasty in intersex patients. *J Urol* 2005;174:1683-6.
11. Palmer BW, Reiner W, Kropp BP. Proximal hypospadias repair outcomes in patients with a specific disorder of sexual development diagnosis. *Adv Urol* 2012;2012:708301.
  12. Alba B, Nolan IT, Weinstein B, O'Neill E, Fritsch A, Jacobs KM, *et al.* Gender-affirming phalloplasty: A comprehensive review. *J Clin Med* 2024;13:5972.
  13. Karakurt G, Keskin H, Doğan HS, Tekgül S. Excision of prostatic utricle: When? How? *J Urol Surg* 2025;12:115-7.
  14. Bose S, Das K, George B, Raman V, Shubha AM, Mahadevappa K, *et al.* 46 XY disorder of sex development (DSD) due to 5 alpha (SRD5A2) deficiency - Experience from a multidisciplinary pediatric gender clinic. *J Pediatr Urol* 2022;18:492.e1-8.
  15. Abd El-Monsif MS, Wishahy AM, Arafa N, Elbarbary MM, Eltagy G, Marei MM. Excision versus division of Müllerian duct remnants in male disorders of sexual development and differentiation: A prospective study to generate anatomical assessment criteria. *Pediatr Surg Int* 2025;41:238.
  16. Krstić ZD, Smoljanić Z, Mićović Z, Vukadinović V, Sretenović A, Varinac D. Surgical treatment of the Müllerian duct remnants. *J Pediatr Surg* 2001;36:870-6.
  17. Ark JT, Moses KA. Operative considerations for late-presenting persistent Müllerian duct syndrome. *Urol Ann* 2016;8:363-5.
  18. Wolffenbittel KP, Holmdahl G. Perineal hypospadias repair with preservation of a coincidental vagina or perineal utricle in boys with disorders of sex development. *J Pediatr Urol* 2019;15:626.e1-5.
  19. Patjamontri S, Lucas-Herald AK, Bryce J, Van Den Akker E, Cools M, Globa E, *et al.* Gynecomastia and its management in boys with partial androgen insensitivity syndrome. *J Clin Endocrinol Metab* 2025;110:e2018-25.
  20. Hughes IA, Houk C, Ahmed SF, Lee PA, LWPES Consensus Group, ESPE Consensus Group. Consensus statement on management of intersex disorders. *Arch Dis Child* 2006;91:554-63.
  21. Lee P, Schober J, Nordenström A, Hoebeke P, Houk C, Looijenga L, *et al.* Review of recent outcome data of disorders of sex development (DSD): Emphasis on surgical and sexual outcomes. *J Pediatr Urol* 2012;8:611-5.
  22. Kaefer M, Rink RC. Treatment of the enlarged clitoris. *Front Pediatr* 2017;5:125.
  23. Pippi Salle JL, Braga LP, Macedo N, Rosito N, Bagli D. Corporeal sparing dismembered clitoroplasty: An alternative technique for feminizing genitoplasty. *J Urol* 2007;178:1796-800; discussion 1801.
  24. Döhnert U, Wunsch L, Hiort O. Gonadectomy in complete androgen insensitivity syndrome: Why and when? *Sex Dev* 2017;11:171-4.
  25. Lucas-Herald AK, Bryce J, Kyriakou A, Ljubicic ML, Arlt W, Audi L, *et al.* Gonadectomy in conditions affecting sex development: A registry-based cohort study. *Eur J Endocrinol* 2021;184:791-801.
  26. Babu R, Shah U. Gender identity disorder (GID) in adolescents and adults with differences of sex development (DSD): A systematic review and meta-analysis. *J Pediatr Urol* 2021;17:39-47.
  27. Long CJ, Van Batavia J, Wisniewski AB, Aston CE, Baskin L, Cheng EY, *et al.* Post-operative complications following masculinizing genitoplasty in moderate to severe genital atypia: Results from a multicenter, observational prospective cohort study. *J Pediatr Urol* 2021;17:379-86.
  28. Wang, AM, Tsang V, Mankowski P, Demsey D, Kavanagh A, Genoway K. Outcomes following gender affirming phalloplasty: A systematic review and meta-analysis. *Sex Med Rev* 2022;10:499-512.
  29. Scougall K, Bryce J, Baronio F, Boal RL, Castera JR, Castro S, *et al.* Predictors of surgical complications in boys with hypospadias: Data from an international registry. *World J Pediatr Surg* 2023;6:e000599.
  30. Rapp M, Duranteau L, Van De Grift TC, Schober J, Hirschberg AL, Krege S, *et al.* Self- and proxy-reported outcomes after surgery in people with disorders/differences of sex development (DSD) in Europe (dsd-LIFE). *J Pediatr Urol* 2021;17:353-65.
  31. Van De Grift TC, Rapp M, Holmdahl G, Duranteau L, Nordenskjöld A, dsd-LIFE Group. Masculinizing surgery in disorders/differences of sex development: Clinician- and participant-evaluated appearance and function. *BJU Int* 2022;129:394-405.
  32. Rynja SP, De Jong TP, Bosch JL, De Kort LM. Functional, cosmetic and psychosexual results in adult men who underwent hypospadias correction in childhood. *J Pediatr Urol* 2011;7:504-15.
  33. Köhler B, Kleinemeier E, Lux A, Hiort O, Grüters A, Thyen U, *et al.* Satisfaction with genital surgery and sexual life of adults with XY disorders of sex development: Results from the German clinical evaluation study. *J Clin Endocrinol Metab* 2012;97:577-88.

**How to cite this article:** Das K, Deb M, Shubha AM, Pati AB. Surgical management of 46,XY differences in sex development. *J Pediatr Endocrinol Diabetes*. 2025;5:130-8. doi: 10.25259/JPED\_81\_2025