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Current hypospadias management: Diagnosis, surgical management, and long-term patient-centred outcomes

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Abstract

In this article, we present the current approach to hypospadias, a review of the classification, preoperative evaluation, and factors that enable decision-making during surgery. We will then discuss patient-reported outcomes, evaluating the patients' and parents' perspectives regarding hypospadias repair.

Introduction

Hypospadias, a condition where the urethra opens on the underside of the penis with associated ventral penile curvature, is the second most common genital birth defect in boys, following cryptorchidism. With an incidence of one in 200 live male births, hypospadias correction is one of the common surgical procedures performed by pediatric urologists.^{1,2}

Classification

The initial diagnosis of hypospadias is typically made after birth during physical exam, where boys with hypospadias are found to have a ventral skin deficiency with a dorsal hood of foreskin and an abnormally located meatus with varying degrees of ventral penile curvature. The standard classification of hypospadias is based on location of the urethral meatus: distal, midshaft, or proximal (Fig. 1). This classification system is misleading, as some variants of distal hypospadias are associated with proximal spongiosal hypoplasia and penile curvature, which may require a more involved surgical reconstruction, while some apparent severe cases of proximal hypospadias present less of a surgical challenge when favourable anatomy is present.

Another form of distal hypospadias is termed megameatus intact prepuce variant, which is usually discovered when the foreskin becomes retractable or during a newborn circumcision. The meatus is widely open and extends proximally to the level of the corona. As universally recommended for hypospadias, circumcision should be deferred, although evidence suggests that surgical outcomes are comparable in circumcised penises with a distal hypospadias using the tubularized incised plate (TIP) repair.^{3,4}

In cases of proximal hypospadias associated with unilateral or bilateral non-palpable cryptorchidism, disorders of sexual differentiation are noted in 17–29% of patients.⁵⁻⁷ Therefore, initial evaluation should include serum electrolytes, 17-hydroxyprogesterone (17-OHP), karyotype, abdominal ultrasound to assess for Mullerian structures, and endocrine referral.

Surgical management

The surgical management of hypospadias aims to achieve:

1. A straight penis with a slit-shaped and adequate caliber meatus at the apex of the glans
2. A conical reconfigured glans
3. Either a circumcised appearance of the penis or if the parents opt for preputial reconstruction, a foreskin that is complete circumferentially and easily retractable
4. An acceptable cosmetic outcome judged objectively

The multiple surgical options available for hypospadias repair are a testament to the fact that no surgical procedure guarantees universal success by all surgeons. As is true for all reconstructive procedures, the best chance of a good outcome is with the first operation.⁸ All hypospadias surgery, even for an innocuous-appearing distal hypospadias, must, therefore, not be considered a minor procedure. The position of the meatus is not the sole determinant of the difficulty in reconstruction. Distal hypospadias can be associated with a more technically demanding repair due to small glans size, poor quality of the urethral plate, proximal spongiosal



Fig. 1. Increasing severity of hypospadias (left to right): distal (sub-coronal); midshaft (skin tethering); penoscrotal; and perineal.

hypoplasia, and possible ventral curvature (VC). Therefore, all hypospadias surgery should preferably be performed by a competent hypospadiologist, who ideally has an annual hypospadias surgical volume of 40–50 cases.⁹

Preoperative considerations

The physical examination of a child with hypospadias yields key information that allows operative planning.^{10,11}

1. Location of the meatus and the degree of proximal spongiosal hypoplasia
2. Presence and degree of VC
3. Quality (width and depth) of the urethral plate
4. Size of the glans and the depth of the navicular fossa
5. Degree of ventral skin deficiency
6. Scrotal abnormalities like penoscrotal transposition and bifid scrotum
7. Availability of the foreskin
8. Penile length

Role of preoperative androgen therapy

Preoperative androgen stimulation in the form of systemic testosterone, topical testosterone, and derivatives like dihydrotestosterone (DHT) and human chorionic gonadotropin (hCG) have been used to stimulate glans size preoperatively to allow better tubularization of the urethral plate and decrease the incidence of glans dehiscence.¹² Currently, there are no defined guidelines for recommending androgen use, nor a defined regimen.^{13,14} There is concern regarding the negative impacts of testosterone on wound-healing and increased bleeding during surgery.^{15,16} In addition, the impact of preoperative androgen use on decreasing the incidence of glans dehiscence has not been conclusively proven.¹⁷ A systematic review looking at preoperative therapy found significant heterogeneity in terms of the drug used, dosage, and evaluation of results or hypospadias outcomes.¹⁸ Despite a lack of high-quality evidence to support that testosterone supplementation improves outcomes, it remains a widely used practice reported by 78% of practicing urologists.¹⁹ Therapy must be stopped 1–2 months prior to surgery to avoid adverse effects during or after surgery.²⁰

Timing of surgery

Surgical intervention for hypospadias can be performed at any age, however, most authors recommend operative intervention at 6–18 months.⁹ The American Academy of Pediatrics suggests this time interval to limit psychological stress and subsequent behavioural problems seen in toddlers undergoing genital surgery.²¹ There are conflicting reports regarding whether increasing age at surgery leads to a higher complication rate.^{22,23}

Operative principles

The operative steps of hypospadias surgery involve penile degloving, correction of VC (orthoplasty), reconstruction of the urethra (urethroplasty), providing a vascularized coverage for the urethroplasty, reconstruction of the glans (glansplasty), and finally a cosmetic skin coverage to create a circumcised penile appearance. The key surgical principles to achieve optimal surgical outcomes include the use of magnification, fine instruments and sutures, minimal and atraumatic tissue-handling, careful hemostasis, and good surgical assistance.^{9,11}

Most surgeons perform stented repairs, although distal hypospadias repairs can be done without the use of stents.²⁴ There is no uniform consensus regarding dressings²⁵ and use of prophylactic antibiotics.^{26,27} Surgeon preference often dictates these variables. Most hypospadias surgery is performed as an outpatient and caudal or dorsal penile nerve blocks are used for postoperative analgesia. There is ongoing controversy whether caudal blocks lead to a higher incidence of urethral fistulas, although the evidence is of poor quality and needs confirmation.²⁸

Correction of ventral penile curvature

Correction of VC is a key component of hypospadias surgery to achieve a straight penis, while preserving penile length and erectile function. VC is assessed preoperatively, but the surgical decision regarding the method of VC correction is made after penile degloving.^{11,20}

The steps of VC correction involve:

1. Penile degloving to the penopubic and penoscrotal junction with release of all penoscrotal tethering bands
2. Release of the glans wings and hypoplastic spongiosum from the corporal bodies
3. Dorsal plication using the Baskin modification of the Nesbitt procedure²⁹
4. Proximal dissection of the spongiosum and distal division of the urethral plate³⁰
5. Multiple fairy cuts to incise through the fibrotic spongiosal tissue without entering the corporal bodies³¹
6. Corporotomy to incise through the tunica of the corpora cavernosum from the 3–9 o'clock position, elevating the tunica to create an ellipsoid defect (lengthening the ventral penile length) and coverage using dermal grafts, small intestinal submucosa (SIS), or a vascularized tunica vaginalis flap from the testes³²

VC below 15° degrees usually does not require any correction. VC between 15–30° can be managed with dorsal plication, while curvature greater than 30° may require more extensive maneuvers like urethral plate transection. The move towards ventral reconstruction with higher degrees of curvature is guided by the concern that dorsal plication shortens penile length and is associated with higher rates of recurrent curvature compared to ventral correction in boys with a >30° curvature.³³

Surgical options for urethroplasty

There are multiple surgical options and several proposed algorithms to guide urethroplasty decision-making.³⁴ Current practices appear to suggest that procedure selection is often guided by surgeon experience. Ongoing efforts to prospectively track surgical outcomes will hopefully lead to more standardized, evidence-based hypospadias decision-making. Surgical procedures can be divided into one- or two-stage procedures and into procedures that involve urethral plate (UP) tubularization, UP augmentation, and UP replacement.

UP tubularization

This urethroplasty technique comprises several procedures that involve tubularization of the UP distal to the hypospadiac meatus. The Thiersch Duplay technique³⁵ and the glanular approximation procedure (GAP) procedure³⁶ involve simple tubularization of the UP after lateral incisions circumscribing the meatus. The TIP procedure proposed by Snodgrass in 1994 involves a midline incision of the UP to widen it and achieve a better caliber urethra, which is then tubularized.³⁷ The superior cosmetic results and the low incidence of complications has led to wide consensus that the TIP urethroplasty is the most commonly used technique for distal

hypospadias.³⁸ An alternative technique, which discards the distal UP and relies on urethral mobilization to bring the meatus to an acceptable location, involves dissection of the urethra proximally to allow an advancement of the meatus to the glans.³⁹ This procedure obviates potential complications, such as urethrocutaneous fistula, but achieves limited advancement even after extensive proximal mobilization.^{39,40}

UP augmentation

Snodgrass et al, who devised the TIP procedure, have shown that UP width does not impact results of tubularization using the TIP technique.⁴¹ However, alternative UP augmentation techniques have been used selectively in cases with poor UP quality. An inner preputial free graft, referred to as a "Snodgraft"⁴² procedure places a preputial inlay graft on the incised surface of the UP with the premise that a graft covering a deep UP incision would decrease scarring. The Mathieu procedure flips a rectangular piece of foreskin distally, which is sutured to the edges of the UP on either side as an onlay flap to the UP.⁴³ A transverse island flap, described by Asopa and Duckett, is derived from the dorsal preputial hood and can also be translocated ventrally to provide an onlay flap to augment the UP.^{44,45} Several studies with blinded raters have found superior cosmetic outcomes for TIP compared to Mathieu or onlays.^{46,47}

UP replacement

In the presence of significant VC, a proximal hypospadias, and a poor UP, urethral transection may be required, thereby, precluding the use of UP tubularization. There are several management options for urethroplasty in this situation. Most surgeons prefer to perform a two-stage urethroplasty with an initial preputial graft placement or a Byars flap followed by a second stage tubularization after six months. Alternative options include the Koyanagi repair and its modifications or a composite repair involving tubularization proximally and distally with a transverse preputial flap onlay in the middle.

Outcomes

Distal hypospadias

The outcomes of distal hypospadias repair are favourable, with a low incidence of redo surgery, but complications are encountered in 5–10 % cases. The TIP procedure has been well studied, with several authors showing consistently favourable results.⁴⁸ A systematic review of outcomes of the TIP urethroplasty and the Mathieu procedure for distal hypospadias showed a lower fistula rate in the TIP group (3.8% vs. 5.3%) and a lower stenosis rate in the Mathieu group

(0.7% vs. 3.1%).⁴⁹ The use of dartos coverage and stents was associated with lower complication rates in this review.

Proximal hypospadias

Complication rates for proximal hypospadias with severe curvature show a high and variable complication rate of 15–56%.^{50,51} A systematic review comparing TIP to onlay preputial flaps for proximal hypospadias without significant VC showed no significant difference in urethroplasty complication rates.⁵² The Koyanagi repair has shown favourable results for proximal hypospadias, with a 17% complication rate in a series of 151 proximal hypospadias children.⁵³ Two-stage repairs for proximal hypospadias repair in boys with >30° VC provides a reliable and reproducible option for success, although the evidence for this approach is weak. The use of preoperative androgen stimulation, tunica vaginalis flap coverage of the repair, and extended glans wings dissection are other factors presumed to decrease complication rates.

Complications

Postoperative complications can usually be identified early on in the first few months after surgery in most cases, but long-term followup is mandatory because delayed presentation with a urethral fistula and recurrent curvature of the penis following puberty spurt have been documented.⁹ Postoperative assessments may include observed voiding and post-void residue assessments or formal uroflowmetry.

The common complications following hypospadias repair include:

1. Urethrocutaneous fistula
2. Meatal stenosis
3. Urethral stenosis
4. Glans dehiscence
5. Urethral diverticulum or urethrocele, which can lead to infections and post-void dribbling
6. Cosmetic issues: Excess residual skin, skin tags, inclusion cysts, skin bridges, suture tracts
7. Hair-bearing urethra
8. Recurrent or persistent penile curvature
9. Spraying or misdirected urinary stream and/or irritative symptoms
10. Erectile dysfunction
11. Balanitis xerotica obliterans of the urethra leading to strictures

Management of complications

The management of hypospadias repair complications are performed after a period of healing over 4–6 months, with the exception of urethral or meatal stenosis, which require more emergent attention. Urethral fistula closures involve

excision and closure of the fistula with adequate dartos flap coverage after excluding distal urethral stenosis. Coronal or more distal fistulas may also require a redo glansplasty. Symptomatic meatal stenosis will often require a dilatation or a meatotomy. Glans dehiscence can be managed with reoperative glansplasty. When a redo urethroplasty is required, the degree of postoperative scarring and the possibility of balanitis xerotica obliterans (BXO) may dictate reoperative management. A redo TIP procedure can be a viable option in the presence of a non-scarred urethral plate or primary glans dehiscence.⁵⁴ In the absence of dorsal preputial tissue, a buccal graft harvested from the lip or cheek can be used to perform a staged redo procedure for more scarred or proximal repairs.

Functional outcomes after hypospadias repair

The majority of hypospadias outcome papers focus on surgical complications and there are few publications assessing long-term functional outcomes. A recent systematic review of the long-term functional outcomes following hypospadias repair demonstrated that patient reported urinary symptoms, such as obstructive voiding, spraying, and deviated stream, more often than controls.⁵⁵ Obstructive flow based on evaluation of Qmax lower than the 95th percentile was found in 13.5% of patients compared to 2.9% of controls. Fortunately, low flow rates have been found to improve in a majority of patients by puberty.⁵⁶ A recent review of 93 adults with hypospadias presenting to a reconstructive urologist highlight the long-term issues that can occur following childhood repairs, including lower urinary tract symptoms (55%) and significant rate of urethral stricture (47%), although there is a selection bias in this study.⁵⁷ Long-term sexual issues, such as erectile dysfunction,⁵⁸ ejaculatory difficulties,⁵⁹ lower self-esteem, teasing, and negative genital perception, have been reported in patients with hypospadias.^{60,61} Several studies have found overall sexual function to be equivalent or slightly lower compared to controls.^{53,62}

There is very limited information on the long-term outcomes of uncorrected hypospadias. A study of 56 adults with primarily distal hypospadias showed that 5% expressed dissatisfaction with the appearance of their genitals and 32% were unaware they had an anomaly. Over a third of these patients had voiding issues and 5% sat to void. This is in contrast to a study of adult men with hypospadias evaluated during transurethral surgery, who reported that they could void in a standing position and have sexual intercourse without problems.⁶³

Conclusion

Hypospadias is a common genital condition in boys with varying severity and long-term functional implications.

Modern surgical techniques have significantly reduced complication rates, but standardization of reporting and well-designed prospective studies will further aid surgical decision-making. There is an increasing body of evidence suggesting that long-term followup and patient-reported outcome measures should form the key to assess surgical results.

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This paper has been peer-reviewed.

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