
Approach to the Adult Hypospadias Patient

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Introduction

The aim of the surgeon in repairing a child's hypospadias is to create a straight penis, a neo-urethra of adequate caliber, a meatus at or near the tip of the glans penis, and normal voiding and good penile cosmesis with minimal complications. Modern techniques report an almost 90 % success rate but often have short-term follow-up. A successful repair in children is often judged by the cosmetic result and the quality of micturition. However, some urethroplasties deteriorate from childhood to adolescence. The high success rates often cited for various techniques are virtually impossible to report with certainty considering the fact that late stage failures are well documented and reported in the literature. A three-decade series from 1978 to 2009 by Prat et al. [1] found 4.6 % of 820 patients required further revision in adolescence. A recent population-based study on more than 5,000 patients found a 9 % secondary surgery rate for distal hypospadias repair ($n=3,553$) and 32.2 % ($n=423$) for proximal hypospadias repairs. Secondary surgery was

also more common in children presenting at an older age [2].

The degree of hypospadias and the type of repair utilized factored into the outcomes. In severe hypospadias, there is often proximal division of the corpus spongiosum and significant chordee. Achieving good long-term results for proximal hypospadias is far more challenging than for distal hypospadias. The definition of a successful outcome has changed over time. In the past, success was defined as a subterminal coronal meatus without chordee. Hinderer et al. [3] stated confidently that the long-term outcome of a hypospadias repair can be predicted only after 2 years of follow-up. Johansson and Avellan [4] reported favorably on the long-term results of Denis Brown, a technique that is currently obsolete. These reviews suggest that short-term patient satisfaction may extrapolate to comparable long-term outcome, although follow-up studies after sexual maturity has occurred are very limited and criteria for "success" have yet to be defined. As techniques have improved more literature has been published on psychosocial, micturition, ejaculation, and cosmetic outcomes. Some of these reports include functional and anatomical complications as well as perceived poor cosmesis with a desire for further surgical correction to look more normal. Bracka outlined the importance of the patient's own satisfaction in the cosmetic appearance of the penis. In a long-term survey, 40 % of his patients requested surgical revision [5]. The Cleveland Clinic experience [6] reported that the most

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Table 4.1 Adult and adolescent hypospadias repair 1980–2011 (18–39 year olds)

Presenting complaint	<i>N</i>
Chordee (11 fistula)	41
Urethral stricture + chordee or fistula	35
Lichen Sclerosus (LS)	11
Hairy urethra (stones or retention)	8
Spraying of urine/subterminal meatus	57
Aesthetic concerns ± spraying	59
Primary repair (6 distal, 4 mid-penile)	10
Total	221

Personal experience

common presenting complaints were voiding symptoms including dysuria, spraying, urgency, and urethrocutaneous fistula.

Hypospadias repair in adults can be divided into three groups: the first group are primary cases, the second group include patients who have had a previous repair during childhood and present with a complication, e.g., urethrocutaneous fistula, persistent curvature, urethral stricture, urethral diverticulum, and poor cosmesis and the third group are patients who have undergone several failed surgeries and previously referred to as “hypospadias cripples.” Failed multiple attempts at hypospadias repair often leaves the patient with a penis that is scarred, hypovascular, and shortened. In many patients, the type of repair as well as the number of surgeries that were done in childhood is not known. Frequently, the local tissues cannot be used to assist in the repair and extragenital sources must be utilized. Herein, we describe the issues surrounding the adult hypospadias patient and the management from our experience (Table 4.1) and the literature.

History

Perhaps no other surgical condition in pediatric urology has inspired more surgical innovations than hypospadias. There are more than 200 surgeries that have been described for hypospadias correction and a recent Medline search revealed more than 5,000 publications on the subject. There is nothing new in hypospadias surgery that has not been previously described. As with any

surgical procedure, the results we see today are directly attributable to the building process in medicine, capitalizing on other ideas, refining them and adding subtle improvements to end up with a surgical technique that is reliable and reproducible by other surgeons.

The significance of chordee was appreciated by Galen in the second century AD and forgotten until Mettauer [7] in 1842 recognized skin shortening as a cause of chordee. It was not rediscovered until 1967 by D.R. Smith [8] and 1970 by Lowell King [9] who reemphasized that the principal structure of the penile curvature is commonly proximal to the meatal orifice, a concept described by Mettauer more than 100 years prior when he advocated “a succession of subcutaneous incisions until the organ is liberated” [7].

In 1869, Professor C. Thiersch [10] reported that in 1857 and 1858 he tubularized the urethral plate in a child born with epispadias and credited the technique to August Brauser, his one-time assistant. Thiersch’s classic article illustrated the design of the flaps and the asymmetric lateral incisions so that the suture lines are unopposed. In 1874, Duplay [11] described the tubularization of the urethral plate distal to the hypospadiac meatus. He also stressed the importance of complete chordee release before urethroplasty.

In prior years, glandular and coronal hypospadias were often not repaired because the complications overshadowed the benefits of surgical correction. Duckett’s [12] meatal advancement and glanduloplasty technique (MAGPI) was designed to reduce the risks of formal urethroplasty in distal hypospadias. Zaontz [13] applied the Thiersch–Duplay principle to the repair of distal hypospadias. He reported excellent results with the glans approximation procedure (GAP), an operation indicated for patients with coronal meatus and a deep glandular groove. This procedure can be used in adults with equal success. Midline incision of the urethral plate was first reported by Reddy in 1975 [14]. He made the incision to excise the “fibrous tissue” in the midline that he believed to be the cause of chordee. He then combined the incision with the Thiersch–Duplay method and tabularized the urethral plate. Rich et al. [15] incised the plate to create a normal

slit-like meatus by hinging the distal urethral plate longitudinally in the midline, but Snodgrass [16] reported and popularized the tubularized incised plate urethroplasty (TIP) repair which is currently the most widely used technique.

Epidemiology

Hypospadias is the most common urogenital malformation second only to cryptorchidism and occurs in 1/250–300 [17, 18]. It has been previously postulated that the incidence of hypospadias in children is rising [19]; however, subsequent studies have not shown the same results [20]. Hypospadias has been associated with advanced maternal age [21, 22] and in vitro fertilization [23].

Given the dearth of longitudinal literature on hypospadiacs, little is known about the natural history of this population. While it is well documented that hypospadias patients—both previously repaired and never-repaired, frequently present with urinary complications in adolescence and adulthood [6, 24], the “hypospadias cripple” presents the greatest surgical challenge and these patients are subject to the highest complication rates [25]. The incidence of late complications is uncharacterized, with some studies reporting about 50 % of patients experiencing some complications. While about 1/3 of these complications occur in the

first 5 years, nearly 1/5 of the complications occur after 5 years [26]. Importantly, this study did not include patients who underwent staged reconstructions, and therefore, this cohort represents a group of patients with “mild to moderate” defects. Complications can come in the form of cosmetic issues, chordee, fistula/diverticulae, foreign material in the urethra (typically hair/stone), and recurrent stricture. Surgical therapy therefore must be tailored to the complication and a detailed discussion of all these problems is beyond the scope of this project.

Long-Term Outcomes: Psychosocial, Erections, Ejaculation, Micturition (Figs. 4.1, 4.2, 4.3, and 4.4)

Questions on the long-term adjustment of patients have now become de rigueur and a number of validated study questionnaires have been developed to follow these patients into adolescence and adulthood to assess the quality of repair and perhaps illustrate the importance of continued follow-up. Unfortunately, not all studies use validated questionnaires and therefore comparing results is problematic. As evidenced by the increasing literature on hypospadias failures presenting in adulthood, hypospadias in childhood has the potential to produce long lasting effects well into adulthood.



Fig. 4.1 116/221 adults complain of spraying from subterminal meatus and poor aesthetics

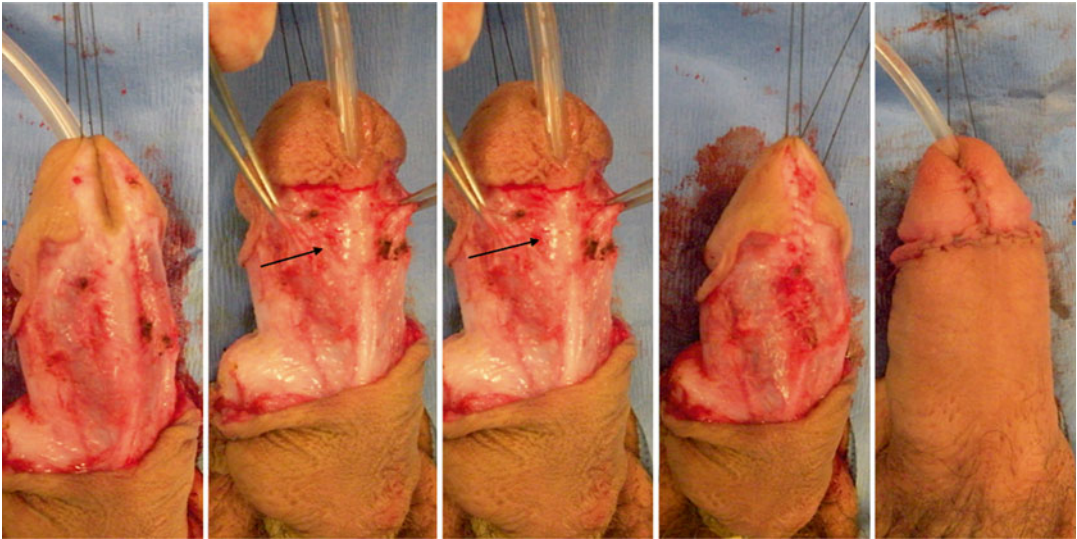


Fig. 4.2 Urinary spraying, subterminal wide meatus and spongiosum deficiency (*arrow*). GAP repair and spongiosal approximation to prevent diverticulum formation



Fig. 4.3 Distal urethroplasty and glandular sculpting for urine spraying and cosmesis

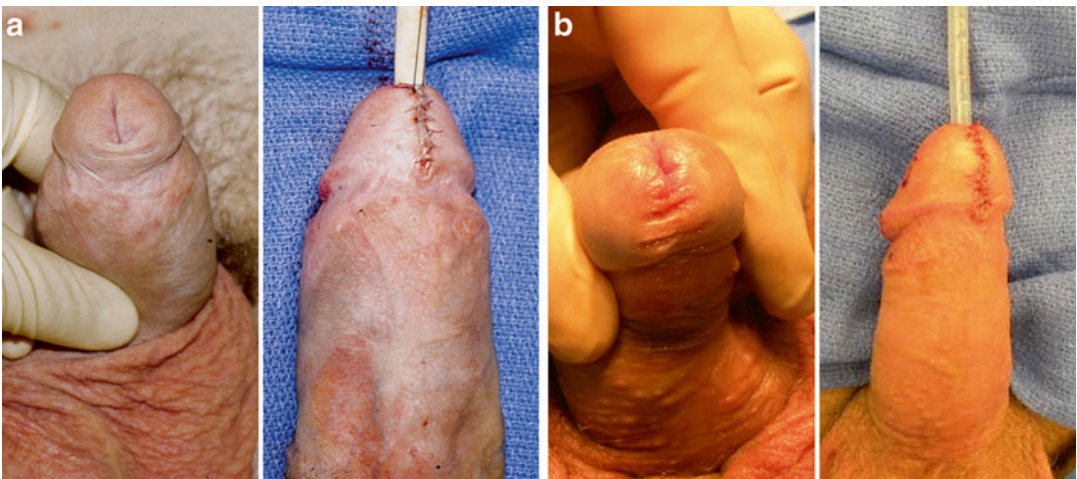


Fig. 4.4 (a) Urine spraying: GAP repair. (b) Suture tracts: glans sculpting

When reviewing studies on long-term patient-centered outcomes, it is important to consider the timeline in which the repairs were performed. Older techniques have historically worse outcomes and have fallen out of favor, influencing the reported outcomes. Adults and adolescents with subterminal or irregular meatus who had proximal hypospadias repaired during childhood often complain of spraying and angulation of their urinary stream. These patients also commonly note post-void dribbling of urine and the need to milk the ejaculate as the seminal fluid stagnates in the neourethra which lacks spongiosum tissue. In our experience, 116/221 (52.4 %) adolescents and adults with previous hypospadias repair complained of urine spraying and aesthetic concerns. They reported more sexual dissatisfaction as well as dissatisfaction with penile appearance. However, the majority had no difficulty with sexual intercourse except in patients with significant curvature.

In 1989, Bracka published a long-term follow-up of 213 patients with a history hypospadias in childhood of whom 196 had undergone surgery [27]. This landmark study followed patients with questionnaires and physical examinations for those who had a “meatal ventralizing or terminalizing repair” of hypospadias. Patients were assessed for location of meatus, urinary dysfunction, appearance, penile size, sexual behavior and performance adequacy of follow-up and guidance. The impetus for review was based on older reports that patients did not need long-term follow-up after hypospadias repair. The number of procedures required for repair varied from 3.6 to 7.2 with higher numbers in those with “short urethra” and proximal hypospadias. On review, many of the patients had a sub-glandular meatus, which Bracka theorized was due to retraction during growth. Not surprisingly, these patients also reported more spraying on the questionnaire. 38 % of patients reported feeling deformed and 72 % felt appearance was just as important as function. When asked about understanding of their condition, a surprising 60 % had never heard of hypospadias and the Bracka postulated that ignorance of their condition caused further social anxiety. In fact, on further review 44 % of patients requested

further surgery for dissatisfaction with stream, appearance, residual curvature, or stricture. In 181 patients who had reached sexual maturity, 1/3 felt inadequate about the size of the penis and in those who underwent proximal hypospadias repair penile length was shorter in flaccid, stretched and erect position compared to the more distal repairs. While 77 % reported having had satisfactory sexual intercourse, ejaculatory complaints predominated with 33 % of patients reporting dribbling ejaculation and 4 % dry ejaculate.

Mureau et al. [28] reported outcomes in 116 patients who underwent hypospadias repair between 1960 and 1992 at two different hospitals. The patients were compared to a group of 88 controls who had undergone inguinal hernia repair. A semistructured interview was performed in boys with an average age of 14.4 years vs. 13.9 years for controls. Patients with hypospadias were more likely to have anxiety and inhibition in seeking sexual contact and 25 % were dissatisfied with penile appearance. The severity of hypospadias or number of surgical procedures did not seem to differ between the groups. However, 19 % of patients in whom surgery was performed after 6 years of age were less satisfied with the surgical result and had a worse genital view than the younger children. Older boys at the time of study (13–18 years) were less satisfied with appearance than their younger counterparts (9–12 years), 31.6 % vs. 13.2 %, respectively. Overall, 39 % patients expressed desire for further correction. The importance of early hypospadias surgery was reiterated by Jones et al. [29] who demonstrated that using validated questionnaire, Hypospadias Objective Scoring Evaluation (HOSE) 80 % of patients had excellent surgical outcome. They concluded that when surgery was completed before age 5 years, boys had no preoperative memories. An association was found between no recollection of surgery and satisfaction with body appearance. The HOSE scheme comprises five domains including meatal location, meatal shape, urinary stream, curvature at erection and fistula.

In a recent study, Chertin et al. [30] reported objective and subjective sexual outcomes in 119 adult patients (older than 18 years) who had their

hypospadias repair performed in childhood between 1978 and 1993. In addition to the Index of Erectile Function (IIEF), they used an invalidated questionnaire of patient perception of psychological well-being and penile appearance. Patients were divided into three groups based on the location of the original meatus: group 1 included 45 patients with glandular hypospadias, group 2 consisted of 56 with distal hypospadias and group 3 included 18 patients with proximal hypospadias. Multiple techniques were used at the time of initial repair. Almost all patients who had glandular and distal hypospadias were satisfied with penile appearance whereas only 11 % in the proximal group were pleased. Mild erectile dysfunction was reported in 50 and 72.2 % in those with distal and proximal hypospadias, respectively; however, 16.7 % of the proximal hypospadias group reported moderate erectile dysfunction. Premature ejaculation was common, reported to be present in 88 %. Additionally, more patients with proximal hypospadias reported decreased sexual quality of life compared with the distal and glandular hypospadias groups.

Long-term outcomes in patients with a history of proximal hypospadias are scarce. Lam et al. [31] performed two-stage hypospadias repair in 44 boys with severe hypospadias and chordee. The average age at response was 15.6 years. The surgeons reported all patients had a satisfactory physical examination. However, ten patients reported minor urine spraying, ten had to milk the urethra to completely eliminate, seven complained of minor post-void dribbling and five others complained of pain or a weak stream. Of the 20 patients who reported ejaculation, 9 had to milk the urethra. Aulagne et al. [32] also reported outcomes in severe hypospadias in 27 patients (age 20–35) all of whom had associated chordee. Various surgical techniques were used. In addition to non-validated questionnaires, the authors used the Hypospadias Objective Scoring evaluation (HOSE) questionnaire. Additional repairs not named included scrotal and penile skin flaps. Micturition was evaluated, and only 15 % of patients had no urinary symptoms. Almost half the patients experienced dribbling. None of the

patients reported problems with erection while 11.1 % reported retrograde ejaculation. Cosmesis was noted to be slightly abnormal by 51.9 % patients while 22.2 % felt it was considerably different. Five patients reported slight to moderate curvature.

Mondaini et al. [33] performed a cross-sectional analysis of men with hypospadias and compared them to 500 controls. Men with hypospadias were less likely to initiate sexual contact, especially if they had more operations. Only 16.6 % had sexual intercourse compared to 42 % of controls; however, all reported satisfaction with the experience. More patients reported poor genital appearance (26.1 % vs. 2 %) compared to controls as well.

In a comprehensive review of the literature, Rynja et al. [34] performed a meta-analysis and reviewed general results, micturition, uroflowmetry, cosmesis, sexuality, and relationships. They included 20 studies with 1,069 patients and reported outcomes compared to controls if included in the original study. The authors also evaluated patients with severe hypospadias. Mean age at follow-up was 27 years (14.0–34.7) and the patients had an average of 2.7 operations. There were 742 subjects in the control group. Patients with hypospadias were more likely to complain of lower urinary tract symptoms including spraying and dribbling (>50 % of the time). Uroflowmetry revealed lower Q_{max} compared to controls (5–39 mL/s vs. 11.9–64.6 mL/s) but mean Q max did not differ.

Patients with severe hypospadias were significantly less satisfied with penile appearance compared to other hypospadias patients and controls, although overall 33.5 and 35.4 % of hypospadias patients and controls, respectively, felt their penis was abnormal compared to their peers.

It is important to note that surgeon perception may be different than patient perception regarding successful outcome. Mureau et al. [35] found overall patients were less satisfied compared to the surgeon with overall genital perception including glandular size and shape, penile thickness, flaccid penile size and appearance of scrotum and testes. After physical exam 11 % of boys

went onto further surgery due to fistula, curvature, and location of meatus, findings that did not prompt them to seek out surgery before the office visit. This study highlights the importance of patient-reported outcomes in long-term studies related to penile surgery.

Conclusions

Body image development occurs in stages and puberty stands out as a particularly sensitive time as the teenager undergoes major changes in his physical appearance. Adolescents become more self-aware and develop the capacity for self-reflection and sexual experience. They depend heavily on what others think and respond with complex emotional reactions. There is significant pressure to conform to normality in the present image-conscious society. Hypospadias patients may have more negative genital appraisal and anticipated ridicule by a partner [28]. A significant number of the adults born with proximal hypospadias encounter micturitional, ejaculatory, and psych-sexual difficulties.

The majority of adults born with distal hypospadias corrected by a modern well-executed “terminalizing” technique are satisfied with their genital appearance. However, long-term outcomes for the modern procedures of the twenty-first century remain to be elucidated in the future.

For primary hypospadias repair, a near-perfect functional and aesthetic result represents a successful outcome and is indeed achievable for many using modern techniques. However, for the minority who require multiple surgeries, which compromised the quality of the genital tissues, the outcome can be severely disabling and the patient is required to accept a lower standard for success. For example, a patient who has endured multiple surgical failures is happy to void through a subterminal or even a coronal meatus, but such an outcome would be unacceptable in a primary repair of a “virgin” hypospadias. Progress in hypospadias surgery will require long-term patient-reported outcome studies to better determine how the patients fare in adolescence and adult life.

Summary

The aim of the surgeon in repairing a child’s hypospadias is to create a straight penis, a neourethra of adequate caliber, a meatus at or near the tip of the glans penis, normal voiding, and good penile cosmesis with minimal complications.

Hypospadias repair in adults can be divided into three groups:

- Primary cases
- Patients who have had a previous repair during childhood and present with a delayed complication in adulthood
- Several prior failed repairs (“hypospadias cripples”)

Rates of secondary surgery for hypospadias are not entirely characterized, but studies suggest:

- 9 % secondary surgery rate for distal hypospadias repair
- 32 % for proximal hypospadias repairs

Problems encountered in postpubertal life include:

1. Urinary—spray, deviated stream, weak stream, dribbling
2. Sexual—erectile dysfunction, ejaculatory dysfunction
3. Infertility
4. Cosmesis—scarring, persistent chordee
5. Psychosocial—sexual inhibition, dissatisfaction with appearance, overall decreased QoL

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