Hypospadias

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103.1 Introduction

Hypospadia is a common congenital malformation in boys that implies an abnormal opening of the meatus urethrae on the ventral side of the penis (Mouriquand and Mure 2004; Baskin and Ebbers 2006). Hypospadia arises due to a premature arrest of the male urethra development during gestational week 8–16. Depending on when the arrest arises, the malformation will have different severity with the meatus on the ventral side of the penis, in the scrotum or in the perineum in severe cases. The incidence is about 1:300 boys. The treatment is surgical, with different methods according to the severity. The long-term outcome is good overall, but there is a higher risk of being dissatisfied with the cosmesis and a higher risk for lower urinary tract problems. Males born with severe hypospadias can encounter more psychosexual problems.

103.2 Historical Overview

The word hypospadias is derived from the Greek words *hypos* meaning "under" and *spadon* "opening" or "fissure". Hypospadias was first documented by the Alexandrian surgeons Antyllus and Heliodorus and Galen during the first and second centuries (Hadidi 2017). References have been found in Greek, Roman, and Egyptian texts. Galen also described curvature and a treatment of division in order to gain fertility (Lambert and Snyder 2011). Historically, penile curvature and its effect on fertility have often been described. For a married couple in Malta, this was a reason for granting annulment. The king of France, Henry II (1547–1559), was married for 10 years, when his penile curvature was corrected and he then fathered ten children with his wife (Hadidi 2017).

Various surgical techniques have been presented over the centuries, ranging from stretching, tunnelling, partial amputation of the penis to the level of the urethral orifice to using flaps during modern times (Hadidi 2017; Lambert and Snyder 2011). The methods have successively been improved by better surgical instruments, introduction and refinement of anaesthesia and development of suture materials. Especially during the nineteenth and twentieth century, new techniques evolved. In 1838, Robert Liston reported a successful repair using a preputial flap. The introduction of ether anaesthesia in the 1840s

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revolutionized surgery, and more sophisticated procedures could be carried out with much less suffering for the patient. In 1874, Theophile Anger used two ventral penile skin flaps to tubularize the urethra and to cover the skin ventrally. The same year, Duplay described the three steps used for hypospadias repair, correction of chordee, skin flaps for creating a urethra and connection of urethra proximally to the neourethra. Ombredanne was the first to create a pedicled penile flap in 1911. During the mid-twentieth century, surgery was usually performed in two stages, relatively easy for many surgeons but with a risk of having hair-bearing skin in the urethra later. During the 1950–1960s, there was an increasing interest for single-stage procedures, popularized by Divine, with full-thickness preputial grafts for the urethra. In 1970, Hodgson introduced a pedicled flap. Byars technique, with two dorsal preputial flaps, facilitated skin coverage in 1955. In 1965, Nesbitt described the technique of treating the ventral curvature by removing dorsal part of the tunica albuginea. In 1994, Duckett and Baskin instead proposed plication dorsally, and now after anatomical studies by Baskin, instead this is performed exclusively in the midline when needed. With an increased incidence of milder hypospadias, other methods like the meatal advancement and glanuloplasty (MAGPI) were introduced by Duckett in 1981. Altogether, around 400 different surgical methods and modifications have been published (Hadidi and Azmy 2003).

103.3 Prevalence

The prevalence is generally estimated to be around 1:300 boys but varies a lot in different populations, probably due to both genetic and environmental differences, as well as different reporting to birth defect registries. The prevalence varies between 0.6 per 10,000 births in Malaysia and 464 per 10,000 births in Northern Europe (Denmark), with the majority revealing a prevalence between 5 and 50 per 10,000 births (Springer et al. 2016). There are also conflicting results whether or not its prevalence is increasing, for example, in Denmark and Sweden (Lund et al. 2009; Nordenvall et al. 2014; Bergman et al. 2015), and whether environmental factors can affect this (Bergman et al. 2015), since known risk factors cannot (Nordenvall et al. 2014).

103.4 Aetiopathogenesis

In an embryo with a normal male karyotype, 46, XY, the undifferentiated gonads will develop into testes during gestational week 8. Human chorionic gonadotropin from the placenta will stimulate hormone production in the Leydig cells. Testosterone is secreted from the testis and is converted by steroid 5-alpha reductase to dihydrotestosterone, which will cause growth and development of the genital tubercle through binding to the androgen receptor in the target genital organs. The meatus is primarily located in the perineum, but during male genital development, the urethra will gradually elongate when the phallus grows and the urethra forms by fusion of the urethral folds, until the opening will be located on the glans after 16 weeks of gestation (Fig. 103.1). The last event of male genital development is fusion of the prepuce. A premature arrest of urethral development will subsequently, depending on the timing of the arrest, cause varying degrees of hypospadias.

Fig. 103.1 Penile development during foetal life

fetal weeks   8   9   10   11   12   13   14   15   16
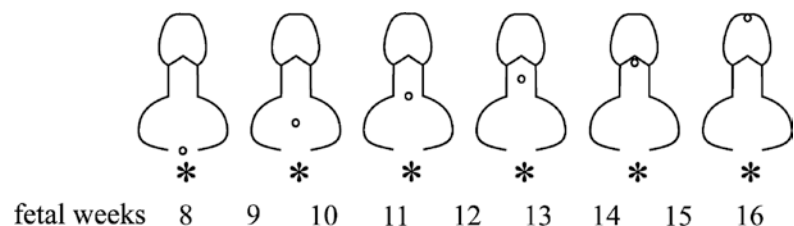
103.5 Pathophysiology

This malformation is a complex or multifactorial disorder meaning that both a genetic predisposition and environmental factors contribute to the developmental failure of the urethra (Van der Zanden et al. 2012). There are many families described with an autosomal dominant inheritance due to mutations in several different genes active during sex development. The concordance in dizygotic twins is 9% compared to monozygotic twins that are concordant in 27%, supporting the notion of a strong genetic factor. The risk for a brother of a hypospadiac boy is reported to be around 12–14% and the risk for a son is around 7–9%. If there are two first-degree relatives, the risk for the next sibling is as high as 26%. In about 10–15% of all cases, there is an additional relative with hypospadias; however, most cases are sporadic.

There are several gestational factors associated with hypospadias, and the most important is low birth weight, both adjusted for gestational age and twinning and is correlated to the severity of the phenotype (Fredell et al. 1998, 2002). This growth restriction is an early event, since it is also associated with low head circumference and low body length. Maternal risk factors are preeclampsia, diabetes mellitus, epilepsy and influenza during the first trimester. Hypospadias may also be associated with chromosomal aberrations and is part of many malformation syndromes, possibly due to lower birth weight in a majority of cases. In OMIM, there are over 300 hits for hypospadias associations to genes, chromosomal regions, syndromes and other malformations (https://www.ncbi.nlm.nih.gov/omim).

Environmental factors, like oestrogenic chemicals, have rendered much attention during the last decades, due to findings of increased incidences of hypospadia, as well as cryptorchidism and testicular cancer, in several countries, not readily explained by genetic factors.

103.6 Pathology

The abnormal positioning of the meatus is located along the ventral side of the penis, in the scrotum or in the perineum. Hypospadias has traditionally been described due to the position of the urethral meatus on the shaft of the penis and by the extent of ventral penile curvature and therefore has been classified as glandular, coronary, penile, penoscrotal, scrotal or perineal, with the mildest form being an isolated cleaved prepuce. In rare cases, the malformation is covered by a normal prepuce and discovered later when retraction of the foreskin is possible or when circumcision is performed.

The urethral plate distal to the hypospadiac meatus lies as a plate on the corpus cavernosum. In most penile or distal hypospadias, the corpus spongiosum and the urethral plate are well preserved, and the current surgical techniques are based on the preservation of the urethral plate that can be reconstructed to a "normal urethra" with all its layers (Fig. 103.2).

However, in the more proximal, penoscrotal or perineal hypospadias, there is often chordee due to different mechanisms. The chordee is a consequence of the abnormal proximal division of corpus spongiosum and the hypoplasia of the ventral tissues. Chordee is related to tethering of the ventral hypoplastic skin onto the underlying structures and a fusion of Buck's fascia, corpus spongiosum and tunica albuginea.

One can distinguish four different types of ventral curvature, including chordee associated with hypospadias. In the first type, the spongiosum is absent in the distal urethra and a fibrous layer prevents the penis from being straight. This type occurs in the perineal and penoscrotal hypospadias with a hypoplastic urethral plate.

In the second type, the urethra is completely developed with corpus spongiosum, but Buck's fascia and the dartos fascia are abnormal. This is common in the penile hypospadias, where the
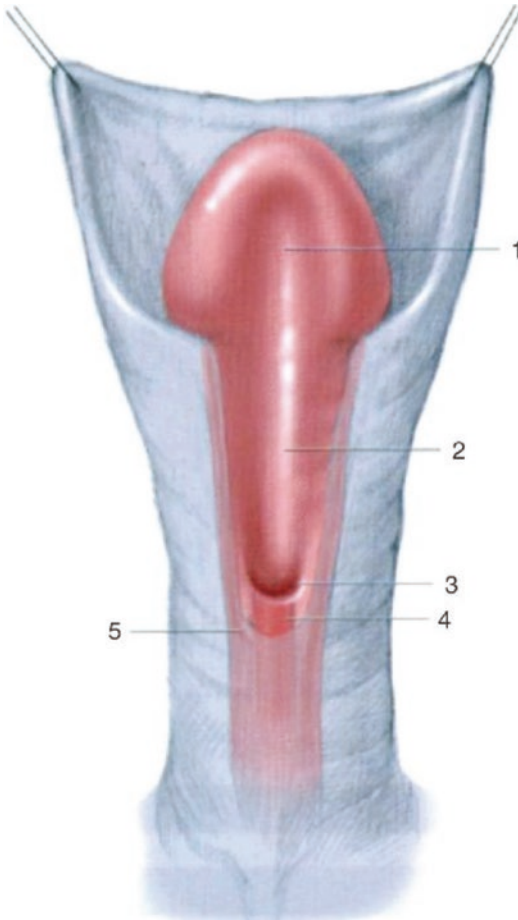
Fig. 103.2 Hypospadia from the tip to the base of the penis: the ventral aspect of the glans is wide opened (1). The urethral plate (2) extends from the apex of the glans down to the hypospadiac meatus (3). The point where the corpus spongiosum (4) deviates marks the proximal limit of the urethral malformation (5). This is also the point where the ventral raphe of the foreskin is separated

dissection of the urethral plate is often sufficient to get a good correction of the curvature.

The third type has been described as skin chordee and is caused by an abnormality of the superficial dartos fascia. Often a degloving and reconstruction of the penile skin are sufficient to correct the curvature.

In a few cases with ventral curvature, a transection of the urethral plate and resection of chordae may not suffice to straighten the penis, thus constituting the fourth type of ventral curvature, due to corpora cavernosal disproportion resulting in a persistent bending also after the penis has

been degloved. The only solution to achieve a straight penis is corporoplasty.

Consequently, the surgical techniques were based on the position of the meatus and the degree of curvature of the penis. The location of the urethral opening may sometimes be misleading. For example, a boy with a subcoronal meatus may have a distal urethra without a surrounding corpus spongiosum and that consists only of a thin epithelial tube extending from the point where the corpus spongiosum divides. Consequently, the preoperative assessment may mistake the location of the normal urethral end point and thus underestimate the severity of the hypospadia. Nowadays, the division of corpus spongiosum is regarded as marking the distal part of a normal urethra and the position of the urethral meatus. In order to help the surgeon make the correct classification of the hypospadias, it is very important to distinguish the division of the ventral raphe of the penile skin, which is the point where the corpus spongiosum deviates.

103.7 Diagnosis

The diagnosis is usually obvious from birth, due to the typical appearance. The most important neonatally is to identify if there is an associated meatal stenosis that in pronounced cases will have to be treated urgently. Later during childhood, the boy may be unable to void in a standing position due to the ventral orientation of the urine. Chordee that is diagnosed during erection can vary to a large extent and can be assessed by compressing the corpora cavernosa against the symphyseal bone or preferably during surgery with an artificial erection test. If untreated, chordee can unable normal intercourse. The fourth consideration is the aesthetic appearance of the penis, with different degrees of chordee and the cleaved prepuce that is mainly located on the dorsal tip of the penis described as a "hood". Fertility might be affected in uncorrected hypospadias, due to mechanical reasons or otherwise mainly due to the underlying cause of hypospadias, such as steroid 5-alpha reductase deficiency that sel-

dom is compatible with spontaneous fertility, due to the small prostate gland.

When the hypospadias is very severe, the child is born with an uncertain sex. Usually, a team consisting of paediatric endocrinologist, paediatric surgeon, psychiatrist and clinical geneticist should gather urgently after birth to perform a proper investigation and inform and involve the parents during the process of assigning the child a sex as accurately as possible, according to the up-to-date knowledge of sex development. The investigation includes karyotyping and hormone analysis, like 17-OH-progesterone, especially in order to diagnose a 46,XX girl with congenital adrenal hyperplasia that when left untreated can be life-threatening, even during the first week of life. In order to clarify the anatomical conditions of the malformation, ultrasonography of the abdomen, urethroscopy and laparoscopy with biopsies of the gonads may be needed. Mutation analysis is the nowadays standard and is the basis for an individualized treatment and is a more solid basis for gender assignment. Nowadays, several hundred genes have been identified to be involved during sex development, and whole genome sequencing can indicate a molecular cause in about one-third of disorder of sexual development (DSD) cases. The more common genes with mutational findings are the androgen receptor gene, the 5-alpha reductase gene, *SF1*, *HSD17B3* and *MAMLD1*, among others (Eggers et al. 2016).

103.8 Associated Malformations/ Differential Diagnosis

The most common associated malformation is cryptorchidism, which is found in up to 10% of the hypospadiac boys. Milder forms of hypospadias are often isolated as an anomaly, but in severe hypospadias, renal malformations, such as hydronephrosis, vesicoureteral reflux and renal agenesis, are found in a few percent of the cases.

Other minor penile anomalies, which can be found in combination with hypospadia, are torsion of the penis, concealed penis or webbed penis. In the more severe cases, a bifid scrotum is seen and sometimes partial or total penoscrotal transposition. Hypospadias is also part of many different syndromes (OMIM n.d.).

103.9 Management

The treatment is surgery and the goal is to restore the urethral meatus to its normal position on the glans, to correct the penile curvature if necessary and to reconstruct the glans and the foreskin, with or without circumcision. The goal is preferably achieved in as few surgical procedures as possible. In addition, one should strongly advise against performing a circumcision prior to surgery, since the prepuce may be essential for the urethroplasty or to cover the penile shaft. It is now generally agreed that surgery should be performed between 6 and 18 months of age, before the boy is aware of his body image and due to a better outcome. A lower risk for complications has been shown to correlate with younger age at surgery as well as the option to be operated on by more experienced surgeons (Lee et al. 2013). In experienced hands, the outcome in children and adults is comparable (Snodgrass et al. 2014a). Surgery before the age of 5 has been shown to not result in perioperative memories (Jones et al. 2009).

The mildest form of hypospadias variant is cleaved prepuce and that can easily be corrected at any age by a prepuceplasty, for the sake of psychological reasons if the parents or the child prefers that. Nowadays, there is a debate about whether or not to operate the glanular forms of hypospadias until the boy reaches an age when he can decide for himself. This means weighting the aspect of reconstruction of the mild malformation against any risk of urinary problems and a lower aesthetic outcome (Snodgrass and Bush 2018).

103.9.1 Preoperative Treatment with Androgens

Severe hypospadias combined with micropenis can be treated preoperatively with testosterone injections or dihydrotestosterone locally. This is the same treatment as that sometimes used diagnostically in disorder of sexual development (DSD) cases neonatally. In a systematic review, preoperative treatment with androgens in severe hypospadias seemed to increase the risk for more complications (Wright et al. 2013). Therefore, the use of preoperative use of androgens has diminished.

103.9.2 Preoperative Antibiotic Treatment

Antibiotics are often used, but there is no clear evidence or consensus of their use. A systematic review on the use of prophylactic antibiotics after stented distal hypospadias repair failed to show any clear advantage (Chua et al. 2019). An international survey showed that a clear majority use antibiotic prophylaxis for hypospadias surgery (84.9%) and, when stents are left postoperatively, as treatment or prophylaxis (84.2%) (Kim et al. 2018). In another survey, 78% used prophylactic antibiotics in hypospadias with a stent in place, and if not, 70% gave an extra dose when the stent was removed (Glaser et al. 2017). A change of the routine of continuous prophylaxis after surgery to two doses of antibiotics (preoperatively and at stent removal) did not increase postoperative infections but rather decreased the complication rate (Zeiai et al. 2016).

103.9.3 Surgical Techniques

The surgical technique differs depending on the position of the meatus and the eventual curvature of the penis. During the last century, a vast number of surgical techniques were described in order to repair hypospadias, probably reflecting different levels of success. In the end, each hypospadias surgeon should familiarize themselves with some techniques that are appropriate for different severity of hypospadias, since there is no technique that can correct all malformations.

As mentioned earlier, a two-stage procedure was the most common in penile or more proximal hypospadias repair. One-stage repairs were initially introduced in the 1960s and have even been used in proximal hypospadias with different kinds of vascularized flaps. During the last 15 years, the preservation of the urethral plate has been advocated and has led to many modifications of the tubularized urethral plate, according to Thiersch and Duplay. The urethral plate consists of the tissues distal to the hypospadia meatus, which are unique structures with a mucosal layer on an underlying corpus spongiosum, in essence constituting all the layers of a normal urethra. The preservation of the urethral plate has probably been the most important advancement in the reconstruction of penile and even proximal hypospadias and has changed our surgical techniques significantly. The urethral plate can primarily be tubularized without additional skin flaps after a midline relaxation incision of the urethral mucosa. The incorporation of the urethral plate (including spongioplasty) into the neourethra may reduce surgical complications. In addition, the urethral plate is rarely the cause of penile bending. The technique has been popularized by Snodgrass and is named the Snodgrass' procedure (tubularized incised plate, TIP; for details Snodgrass 2005). The most common surgical techniques used today are summarized in Figs. 103.3, 103.4, 103.5, 103.6, 103.7, 103.8, and 103.9.

103.9.4 Correction of Chordee

The degloving of the penile skin sorts out the chordee related to the tethering of the ventral skin. If the angulation persists after freeing the skin, the dissection of the urethral plate from the ventral surface of the corpus cavernosum from the glans down to the normal urethra allows in most cases a straight penis. In a few cases, the bending still persists, and that is caused by a disproportion between the dorsal and the ventral
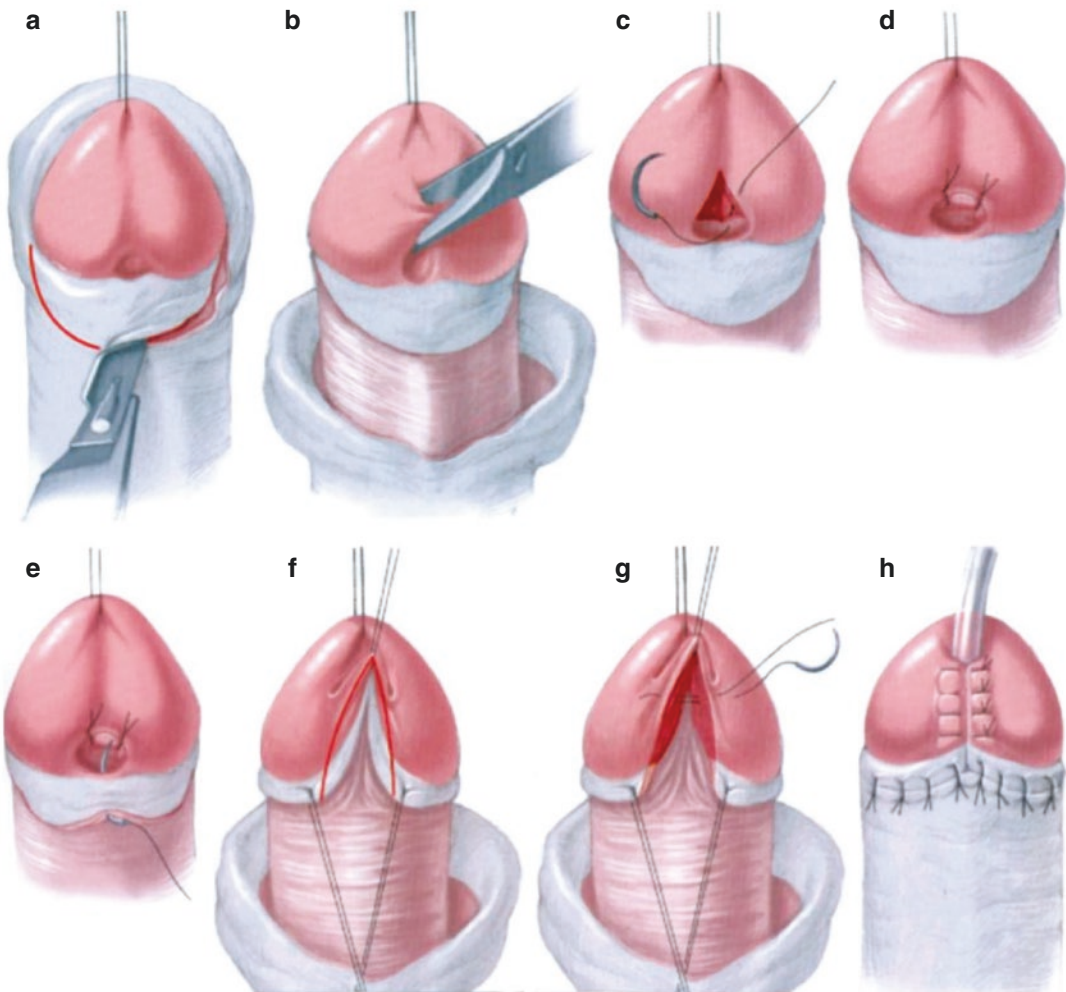
Fig. 103.3 (**a–h**) The meatal advancement and glanuloplasty (I) (MAGPI) procedure (Duckett and Snyder 1990). The incision line is drawn 5 mm behind the meatus (**a**). The glanular groove is incised (**b**). The dorsal meatus opens up and a diamond-shaped defect is created. This is closed transversely with 2–3 sutures (**c**, **d**). The ventral lip of the urethra is fixed with a holding stitch and brought forward (**e**). This allows the lateral wings of the glans to rotate to the ventrum (**f**, **g**). A sleeve approximation of the penile skin is done, excising all redundant tissue and leaving a circumcised appearance (**h**). No stent or catheters are required and the procedure can be done on outpatient basis. Secondary retraction of the meatus is quite common, and therefore, this procedure has become less popular

parts of the corpus cavernosum. This can be treated with multiple ventral incisions (Snodgrass and Prieto 2009; Snodgrass and Bush 2017), instead of making dorsal plication that was used earlier. In severe forms (scrotal or perineal), the urethra is dysplastic with associated chordee, such that the urethra plate needs to be resected to get a straight penis before urethroplasty.

103.9.5 Urethroplasty

Reconstruction of the missing urethra and the technique chosen depend on the size and quality of the urethral plate. In the very distal cases with a normal penile urethra, the MAGPI procedure still have a place (Duckett and Snyder 1990; Fig. 103.3).
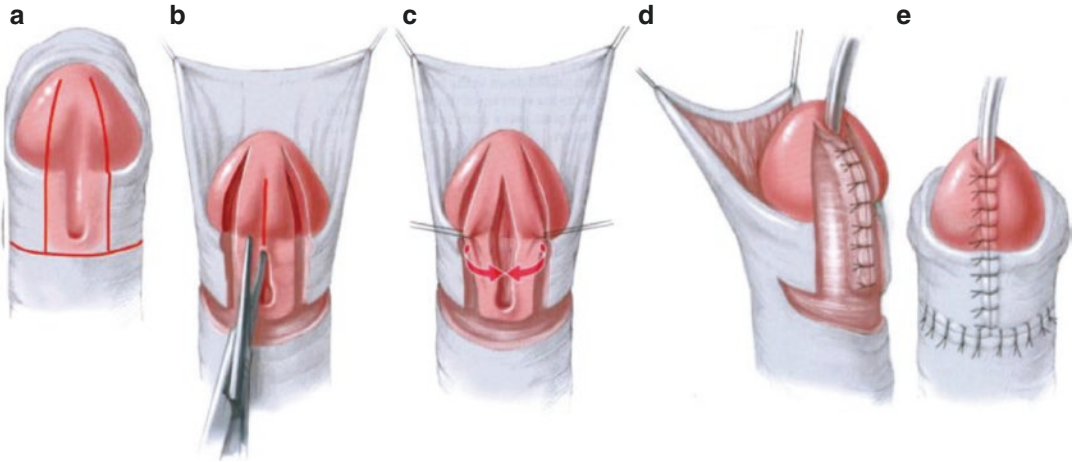
Fig. 103.4 (**a–e**) The TIP (Snodgrass) procedure (Snodgrass 2005). The Snodgrass' procedure is a development of the Thiersch–Duplay technique, saving even a narrow urethral plate for tubularization. The incisions are the same (**a**) and the distal urethral plate including corpus spongiosum is dissected (**a**). Deep glanular wings are created. The urethral plate is incised longitudinally in the midline from meatus up to the tip of the glans (**b**). The

urethral incision leaves a dorsal raw area in the urethra which will subsequently epithelialize (**c**). This allows the plate to be tubularized around a 6–8 Fr catheter (**d**). The dorsal foreskin is either resected (**e**) or saved and reconstructed. This technique may also be used even in proximal hypospadias with a good urethral plate, with ventral corporotomies when needed
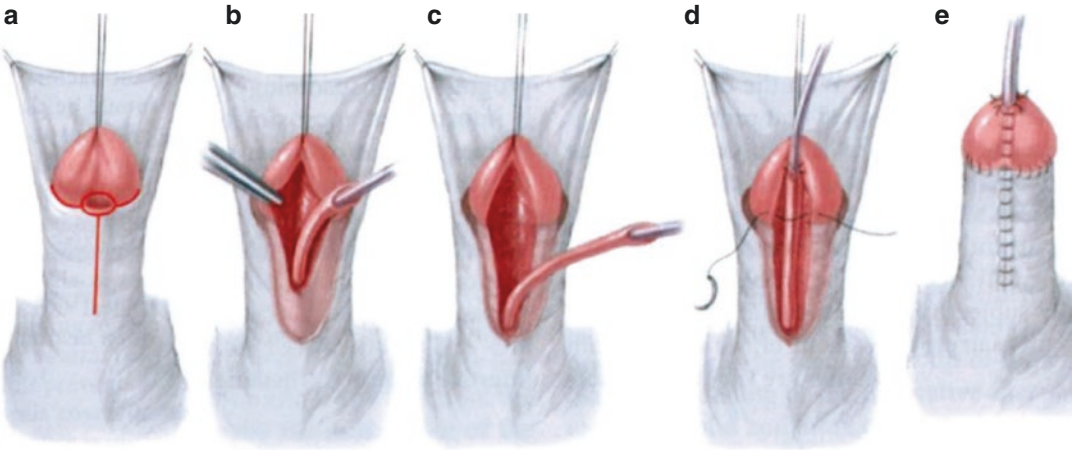

Fig. 103.5 (**a–e**) Koff's urethral mobilization technique (Koff 1981). This technique is based on the elasticity of the urethra. When the distance from the tip of the glans to the ectopic meatus is <2 cm it is possible to mobilize the penile and even the bulbous urethra to get a good "extra length", which is enough to get the normal urethra to the tip of the glans without causing ventral bending. After dissection of the urethra from the ectopic meatus and proximally enough to get the desired length (**b**, **c**), a deep

incision in the midline of the glandular urethra is performed (**c**). The dissected urethra is sutured to the tip of the glans, and the glans wings are sutured to cover the distal urethra (**d**, **e**). A 6–8 Fr catheter is often left in place but can be removed shortly after the operation because the distal urethra is "normal". Thus, the great advantage with this technique is that there are no urethral sutures and less risk for fistulas and strictures
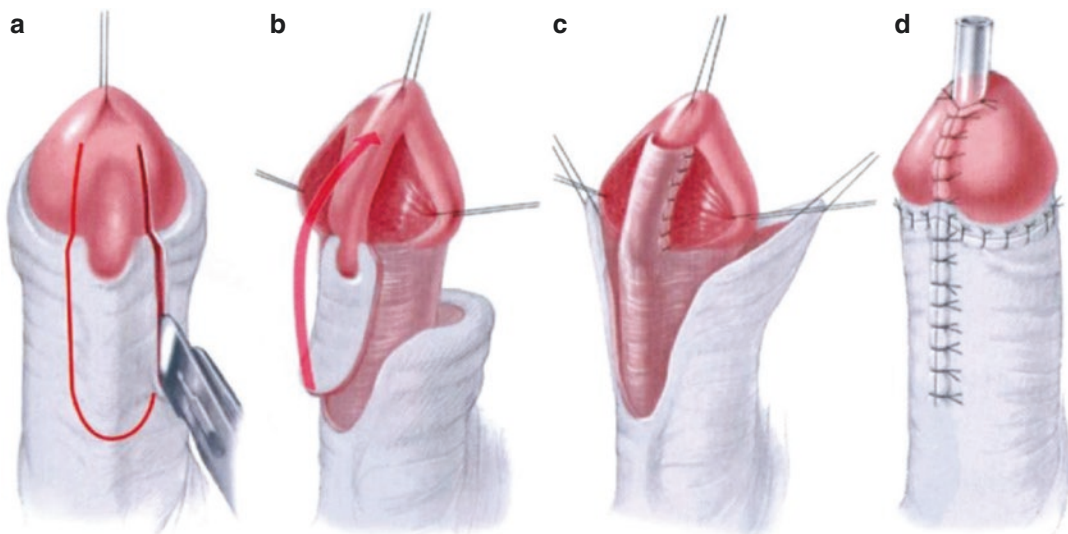
Fig. 103.6 (**a–d**) The Mathieu procedure (Mathieu 1932). The incisions are made on both sides along the distal urethra (**a**). A skin flap from the ventrum of the penile foreskin is prepared (**b**). This flap is based and attached to the distal urethra and is sutured to the dissected distal ure-thral plate. Deep glanular flaps are dissected (**c**) down to corpus cavernosum and enough to cover the distal urethra (**d**). There is also a possibility to reconstruct the foreskin if wanted. The Mathieu technique often gives a transverse meatus, which gives a less satisfactory cosmetic result

In most cases, the urethral plate is wide and healthy and can be tubularized, either directly according to the Thiersch–Duplay technique or the Snodgrass' technique with incision of the urethral plate that has replaced even the very distal reconstructions, and with better cosmetic results (Snodgrass 1994; Fig. 103.4). In some cases, when the urethral meatus is <2 cm from the tip of the glans, a complete mobilization of the penile urethra (Koff 1981) may be sufficient to get the urethral meatus to the tip of the penis (Fig. 103.5). If the distal plate is too narrow, there are optional techniques using a flap of the ventral penile skin, according to Mathieu (Mathieu 1932, Fig. 103.6), or an inlay flap in the incision of the urethral plate or using a pedicularized preputial flap as an onlay urethroplasty (Fig. 103.7).

In rare cases in perineal or penoscrotal hypospadias, the urethral plate may be hypoplastic and is not appropriate to use for the urethroplasty. The plate has to be divided in order to straighten the penis and to resect the chordee. A tube needs to be made to replace the missing urethra either using a pedicularized preputial flap in a one-stage procedure (Asopa–Duckett technique, Duckett 1980) or the Koyanagi et al. (1994) (Fig. 103.8).

The trend now is to use the two-stage technique, using either free prepuce or buccal mucosal graft, STAG (Bracka 1995; Altarac et al. 2012). When the urethra has been reconstructed, it is very important to cover the suture line with either a vascularized dartos flap or a flap from the Buck's fascia or surrounding tissue, thus preventing fistula formation. Even if a proper spongioplasty can be performed, covering of the suture line is necessary.

103.9.6   Glanuloplasty

The reconstruction of the glans is very important as it determines the final cosmetic outcome of the surgery. The tubularization of the urethra allows a slit-like urethral opening and necessitates a dissection of glandular wings to cover the glanular part of the urethra to get the best cosmetic result.

103.9.7   Foreskin Reconstruction

In distal hypospadias, there is always a possibility to preserve the foreskin. Depending on the
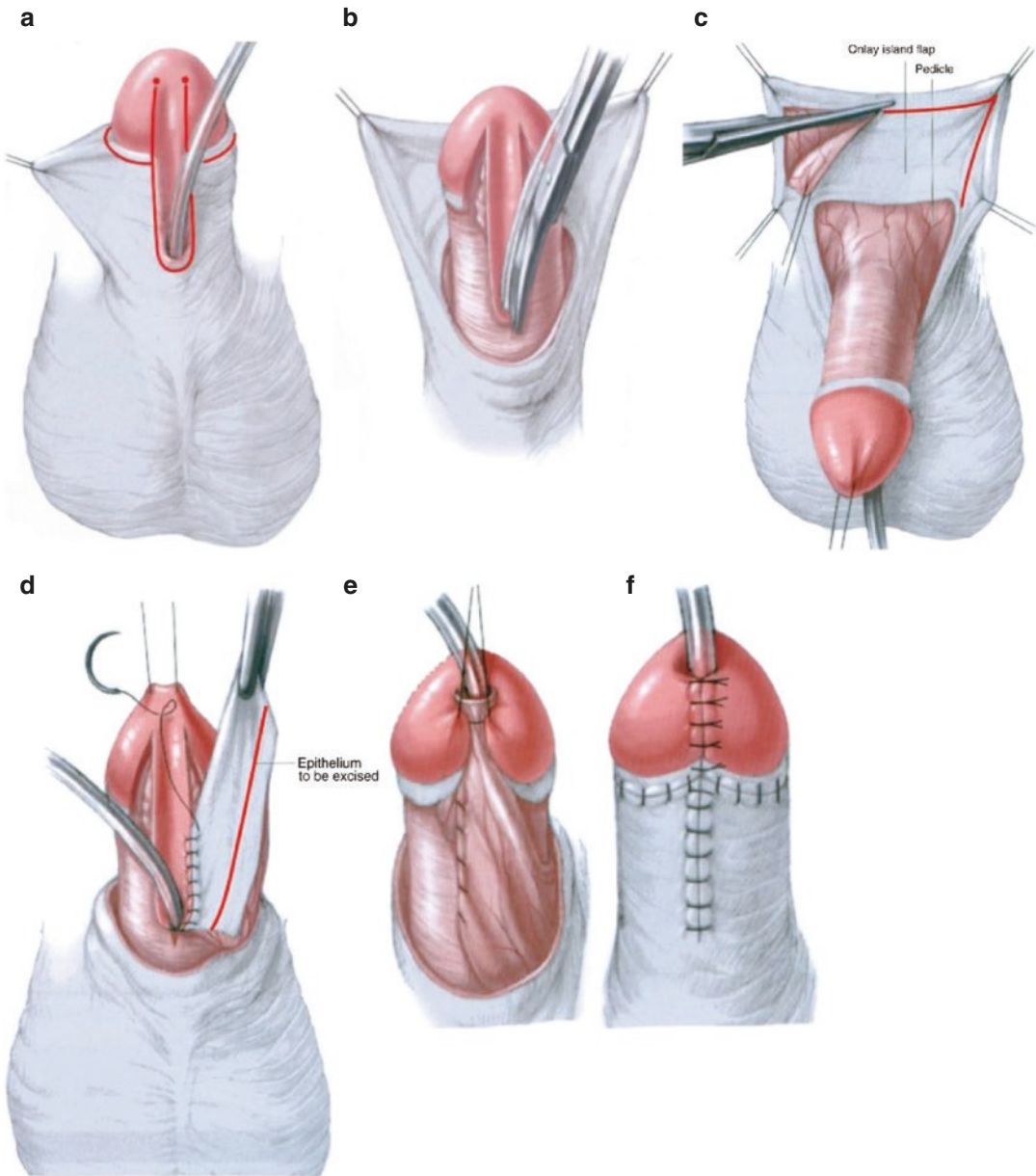
Fig. 103.7 (**a–f**) Onlay plasty with the transverse island flap technique (Elder et al. 1987). In cases with a narrow urethral plate that do not allow tubularization even with a TIP, the plate may be preserved. Providing the penis is straight, an onlay procedure may be the method of choice. The penis is degloved (**b**) and a rectangle flap of the inner preputial mucosa is pediculized (**c**) and transferred to the ventrum of the penis to be laid on the urethral plate using interrupted or a running 6–0 or 7–0 suture over an 8 Fr catheter (**d**). The suture line is covered with a dartos flap (**e**), and deep glanular wings are dissected and sutured, and the foreskin is reconstructed (**f**). In the severe hypospadias (scrotal or perineal), the urethral plate is often dysplastic and associated with severe chordee causing

ventral bending of the penis. The plate is then too poor to be kept, and a full-tube urethroplasty needs to be performed using either a pediculized flap from the inner preputial layer (Duckett 1980) or a buccal mucosal graft. The main disadvantage of these techniques is the risk of proximal urethral anastomosis stricture and dilatation of the distal neourethra. There is a high complication rate associated with the tubularized flap-techniques, up to 50% needs a redo surgery (Pippi Salle et al. 2016). Therefore, these techniques have become less popular and instead the two-stage procedures using either preputial flaps or buccal mucosal graft as replacement of the urethral bed in a two-stage procedure, STAG (Bracka 1995; Altarac et al. 2012)
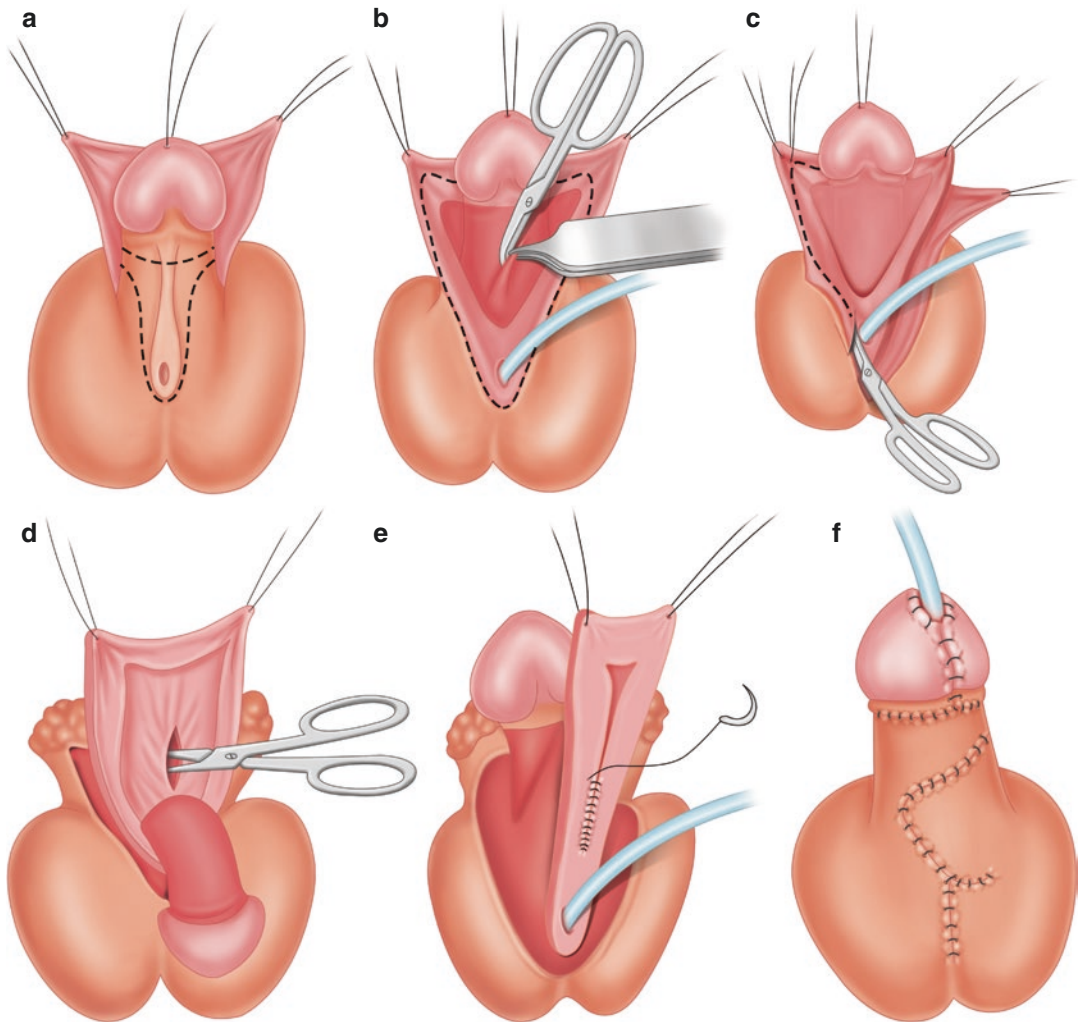
Fig. 103.8 (**a**–**f**) Koyanagi procedure (Koyanagi et al. 1983, 1994). The Koyanagi technique was reported already in 1983 and is used mainly for the reconstruction of proximal hypospadia in experienced hands and later included modifications (Hayashi et al. 2001). The combined technique is illustrated in figures (**a**–**f**). (**a**) Skin flaps for the urethroplasty are outlined along a coronal circumference with a transection of the urethral plate just below the glans. A racket-shaped incision is done for the mobilization of the urethral plate and the adjacent tissues with the inner aspect of the preputial hood. (**b**) The urethra is dissected free and the chordee is released. An artificial erection is done to test the release. A U-shaped skin incision is made surrounding the meatus to extend the skin along the marked line. (**c**) This second incision is extended laterally and dorsally onto the dorsal prepuce ≈8 mm parallel to the first incision. (**d**) A buttonhole is made through the pedicle of dartos. (**e**) The urethra is reconstructed with the dorsal wall of the urethra and then the ventral wall. (**f**) The meatus is created by splitting the glans and the divided dorsal Byar's flaps are sutured to cover the ventral skin defect

cultural or religious traditions in each country, either a circumcision is performed or the foreskin can be preserved and reconstructed (Fig. 103.9). This is a simple procedure that in most cases of distal hypospadias even saves operative time. The complication rate is not higher, and there are

indications that there are less complications compared to a circumcision (Snodgrass et al. 2013; Zeiai et al. 2016). The skin incision is performed between the inner and outer layer of the foreskin. After the urethral and the glanular reconstruction have been performed, the inner layer is sutured
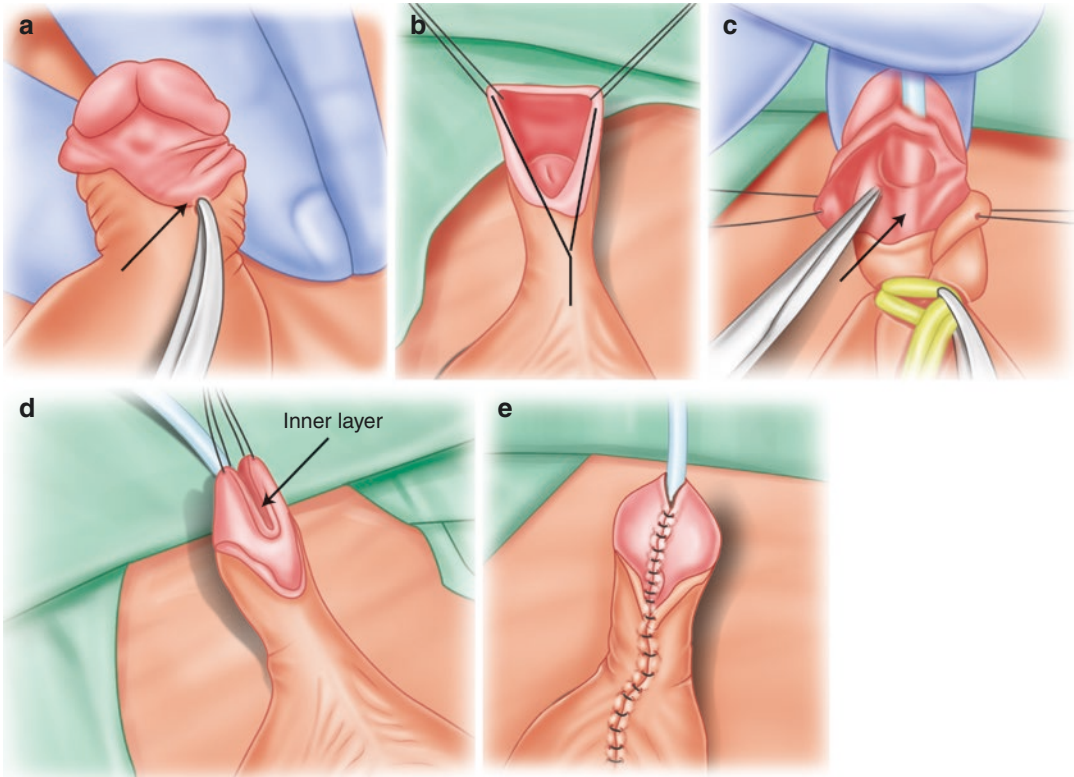
Fig. 103.9 (**a**–**e**) Prepuceplasty (Snodgrass 2005). Preservation of the foreskin in hypospadia reconstruction. (**a**) Point of the deviation of the ventral raphe (arrow). (**b**) Outline of the incision. (**c**) End point of the normal ure-
thra. (**d**) Reconstruction of the inner and outer preputial layer with a subcutaneous layer in between. (**e**) A well-preserved foreskin

with continuous 6–0 or 7–0 sutures to the tip of the foreskin, leaving an opening that allows the foreskin to be retracted without difficulty. The inner layer should be covered with a subcutaneous layer to promote healing. Thereafter, the outer foreskin is sutured with intracutaneous 6–0 sutures. Hypospadiac cases with an abundant dorsal foreskin (a big dorsal "hood") may have to be reconstructed in several stages.

103.9.8 Postoperative Care

The milder cases are treated as outpatients. In most cases, even in distal hypospadias, a urethral stent is left in place. We prefer the Cook double diaper catheter that is sutured to the glans. The

catheter is kept for 1 week and used as a dripping urethral stent with a double diaper. The dressing is variable from one centre to another and also the time of catheter drainage. One commonly used dressing is a Tegaderm dressing that will be removed together with the stent.

103.9.9 Complications

Early and more common complications are bleeding, infections and urinary retention that can be treated appropriately in the postoperative period. Later complications are described below and treated accordingly. Urethrocutaneous fistulas, which have to be reoperated, are reported in several percent in different degrees depending on

the severity of the malformation. Meatus stenosis and urethra stricture are examined postoperatively by checking the urinary flow. The most common treatment is meatoplasty and urethral dilatation, respectively, but sometimes internal urethrotomy is required. Usually these complications arise early, around 80% during the first postoperative year, and the risk is correlated to the glans width (Snodgrass et al. 2014b; Bush et al. 2015). Diverticulum of the reconstructed distal urethra can arise, especially after two-stage procedures and if the meatus is relatively stenotic. Residual curvature is examined at puberty since it will need further surgery. Balanitis xerotica obliterans (BXO) can arise after surgery and render residual strictures. Rupture of the reconstructed foreskin can happen and will also need a reoperation. The genital appearance is important and should be recognized as it is of significant importance to the patient. Sometimes, the initial result is not optimal and will require smaller skin corrections. Also, the patient should be informed about a large potential for better aesthetic outcome depending on the underlying condition and due to growth at puberty.

103.9.10   Postoperative Follow-Up

The first postoperative follow-up is due after 1–3–6 months, followed by medical evaluations at 5, 10 and 15 years of age. These patients are important to follow until puberty because of the risk of recurrent curvature and seldom a risk of urethral stricture that has to be controlled for with flowmetry and residual urine after the age of 5 years. The final postoperative result can only be judged at puberty and the boy should be motivated to come for a final examination. At the final visit, thorough information should be given, preferably in writing, and a decision has to be made whether or not to refer for future visits to an andrologist or urologist.

103.9.11   Postoperative Long-Term Consequences for Life

Generally, one can expect a normal life in all aspects, such as education, income and marriage/partnership (Skarin Nordenvall et al. 2017; Örtqvist et al. 2017a, 2017b). Urologically, it is more common to experience a weak stream, urinary dribbling or spraying (Rynja et al. 2011; Keays et al. 2016). Other problems that can arise, and more so in severe hypospadias, concern the genital appearance, such as a shorter penis or erectile dysfunction. However, most still declare a satisfactory sex life (Ruppen-Greeff et al. 2013; Even et al. 2015; Örtqvist et al. 2017a). There is an interest for more psychological support, especially in males born with severe hypospadias, and also peer support (Even et al. 2015; Örtqvist et al. 2017a). Studies evaluating coping strategies have detected a tendency for avoidance (Örtqvist 2017a; Rynja et al. 2019). In an effort to explore the consequences of not being operated for hypospadias through a Facebook group, the result was that these men had experienced lower sexual health, more problems with curvature and more difficulties with having intercourse (Schlomer et al. 2014). In addition, they were unhappy about the meatus and it was more common to sit and void.

Fertility in hypospadias has not been studied as much as would be expected. Many different factors like coping, partnership, dissatisfaction with genital appearance, erectile dysfunction and postoperative urological complications can affect this outcome. In two studies, hormone levels (FSH and LH) were examined, as well as semen quality (Asklund et al. 2010; Kumar et al. 2016). These parameters were normal, except in some cases of proximal hypospadias and especially in association with cryptorchidism. In a follow-up study, males with proximal hypospadias (mean age 34 yrs.) had children less often (19%) than controls (50%) (Örtqvist et al. 2017b). The age for the first child,

however, did not differ. In national register studies (Finland and Sweden), 24% had at least one child compared to 29% in the general population (Asklund et al. 2010) and 21% of all hypospadias males (in proximal form only 10%) compared to 27% (Skarin Nordenvall et al. 2020), respectively.

103.10 Conclusion

Important advances in the aetiology and the management of hypospadia have been made over recent decades. Increased knowledge of the embryology and the genetic background can help us to better select the optimal treatment for boys born with hypospadia.

A distal meatus may be a penoscrotal hypospadia. It is very important that the surgeon recognizes the diagnostic signs for the different types of hypospadias, in order to provide the correct information and treatment. We have to accept that there is no minor hypospadias.

The preferred methods for reconstruction have undergone changes from two-stage surgery, even in simple cases, to one-stage procedures in almost all cases. Now, there is a larger acceptance of performing two-stage procedures again, not only in the surgery of the hypospadias cripples. It is very important that each surgeon familiarizes themselves with different techniques and judges each case individually. It is also very important to follow the patients long term, at least until they are in puberty, as there are those who will require further surgical reconstruction or medical attention.

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