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Hypospadias is one of the most common urogenital anomalies, occurring in 3 in 1000 births. It is better defined as an arrest of the development of the genital tubercle (GT) during week 6 and week 16 of gestation, leading to hypoplasia of the tissues forming the ventral aspect (ventral radius) of the penis beyond the division of the corpus spongiosum. It is characterized by a ventral triangular defect whose summit is the division of the corpus spongiosium, whose sides are represented by the two pillars of atretic spongiosum, and whose base is the glans itself.

In the middle of this triangle, from the tip to the base of the penis, sit a widely open glans, the urethral plate (which extends from the ectopic urethral meatus up to the glans apex), the ectopic meatus, and a segment of variable length of atretic urethra (not surrounded by any spongiosum), which starts where the corpus spongiosum divides. Tissues sitting inside this triangle are dysplastic and do not grow at the same pace as the rest of the GT.

There are two main types of hypospadias:

- The hypospadias with a distal division of the corpus spongiosum with little or no ventral curvature when the penis is erected.
- The hypospadias with a proximal division of the corpus spongiosum, with a marked degree of hypoplasia of the tissues forming the ventral radius, associated with a significant degree of ventral curvature.

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### 64.1 Causes of Hypospadias

The causes of hypospadias remain essentially unknown. Several avenues have been explored to explain this congenital defect of the genital tubercle:

- Some endocrine disorders have been described in relation to hypospadias, mainly an insufficient secretion of androgens or insufficient response by the target tissues. These disorders can be demonstrated in very few cases, however.
- Some genetic disorders could explain why hypospadias may be found in several members of the same family.
- Young and old mothers are more prone to carry a baby with hypospadias. Low-birth-weight babies and twins also have a higher risk of presenting with a hypospadias. This risk could be explained by a placental insufficiency.
- The possible and controversial increase of hypospadias in the population over the past 20 years suggests a role for possible environmental factors such as oestrogenlike molecules, pesticides, and fertilizers.
- Abnormal or insufficient growth factors could also be responsible for these penile anomalies and could also explain the significant complication rate met in this surgery.

### 64.2 Surgical Techniques

Three surgical steps characterize hypospadias surgery: (1) Degloving of the penis and deep dissection of the glans wings; (2) Repair of the urethra (urethroplasty); (3) Reconstruction of the ventral radius of the penis.

Degloving of the penis and a deep dissection of the glans wings: In most cases, the penis is straightened and the common ventral glans tilt is corrected. In some midshaft and proximal hypospadias cases, the residual curvature

Hypospadias

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demonstrated by an erection test is due to asymmetrical corpora cavernosa and requires a dorsal corporeoplasty (dorsal shortening of the albuginea of the corpora cavernosa). In most cases, the urethral plate can be preserved, although there are situations in which the urethral strip is very poor and the two corpora writhe around it. In these cases, the urethral plate is usually sacrificed.

*Repair of the urethra (urethroplasty)*: Once the GT has been fully dissected, the length of urethra to be reconstructed can be precisely evaluated. The repair technique chosen depends on the size and quality of the urethral plate:

- If the urethral plate is wide and healthy, some will choose to tubularize it following the Thiersch-Duplay technique.
- If it is too narrow to be tubularized, the Snodgrass open urethrotomy is a popular option, or additional tissue can be laid on the urethral plate using a rectangle of pediculized preputial mucosa (onlay urethroplasty) or a flap of ventral penile skin (Mathieu procedure).
- If the segment of urethra to replace is short (<1.5 cm) and if the distal urethra is not hypoplastic, a complete mobilisation of the whole penile urethra may be adequate to bridge the defect. This technique (Koff) has the advantage of avoiding the use of nonurethral tissue.
- If the urethral plate cannot be preserved, a tube must be made to replace the missing urethra, using either a pediculized rectangle of preputial mucosa (Asopa-Duckett technique) or buccal mucosa.
- In major hypovirilization of the GT, the Koyanagi procedure, mobilising the tissues of the ventral and lateral aspects and dorsal prepuce with the blood supply, is a reliable option to reconstruct the missing urethra. Two-stage procedures (Cloutier-Bracka procedure) are an alternative for long urethroplasties using either preputial mucosa or buccal mucosa.

*Reconstruction of the ventral radius of the penis*: Once the urethra has been repaired, reconstructive steps are taken:

• Meatoplasty, trying to create a slit-shaped meatus.

- Glanuloplasty, by stitching together the two glans wings over the reconstructed urethra to reconstruct the ventral aspect of the glans.
- Creation of a preputial collar around the glans.
- Coverage of the reconstructed urethra (spongioplasty), using the lateral pillars of spongiosum or the pedicle of the onlay flap.
- Skin cover with a redistribution of the skin shaft, bringing the excess dorsal skin to the ventrum.
- Reconstruction of the foreskin, or circumcision.

The patient's age at surgery for primary hypospadias repair is usually between 6 and 24 months.

Pre-operative hormonal stimulation of the penis using beta human chorionic gonadotrophin ( $\beta$ HCG), testosterone, or dihydrotestosterone is sometimes indicated in case of a small penis (dorsal radius <25 mm long or glans width <15 mm in the first year of life) or in case of redo surgery. It remains unclear how safe these treatments are on a long-term basis. Some publications have pointed out their detrimental action on the healing process.

General anaesthetic is the rule, often associated with caudal or penile anaesthesia. Magnification is commonly used in this surgery. Coagulation is often not needed in this surgery when the tourniquet is used followed by a slightly compressive dressing. Other surgeons prefer bipolar coagulation or adrenaline injection prior to incision.

Urine drainage via a suprapubic catheter, a transurethral bladder catheter, or dripping urethral stent varies from one surgeon to another; some do not drain at all.

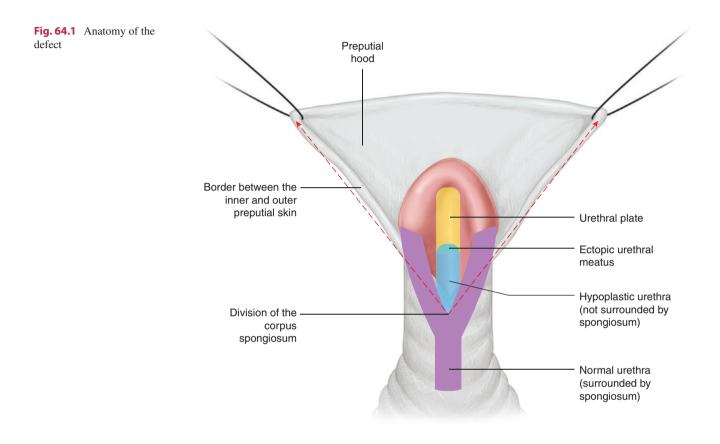
Antibiotic protocols also vary greatly from one centre to another, and their efficacy needs to be proven. A dressing is essential after this surgery. We prefer the so-called daisy dressing because it is very comfortable for the patient and contains postoperative bleeding, but some others prefer OPSITE dressing, Silastic foam dressing, or Tegaderm dressing.

Postoperative pain control is essential, using morphine instillations, anti-inflammatory medications, an anticholinergic (oxybutynin chloride), and diazepam to reduce bladder spasms caused by the bladder catheter.

### 64.3 Specific Surgical Procedures

From the tip to the base of the penis, the ventral aspect of the glans is widely opened. The urethral plate extends from the apex of the glans down to the ectopic meatus (Fig. 64.1). Behind the ectopic meatus sits a segment of hypoplastic ure-

thra not surrounded by any spongiosum. The division of the corpus spongiosum marks the proximal limit of the malformation. It defines a triangular defect whose summit is the division of the corpus spongiosum, whose base is the glanular cap and whose sides are represented by the two lateral pillars of atretic spongiosum.

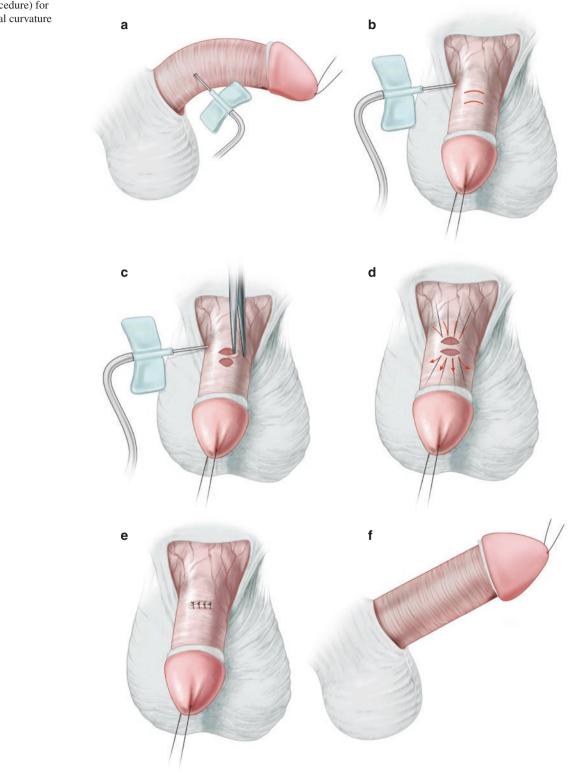


### 64.3.1 Correction of Ventral Curvature: TAP Procedure

Most penises are fully straightened after skin shaft degloving and extended dissection of the glans wings. In hypospadias with a proximal division of the corpora cavernosa, a

**Fig. 64.2** (**a–f**) Tunica albuginea plication (TAP procedure) for correction of ventral curvature

complementary corporeoplasty may be needed to complete the straightening, either by tunica albuginea plication (TAP procedure), plicating the dorsal albuginea of the corpora cavernosa on the dorsal midline (Fig. 64.2), or by using the Nesbit procedure. Some suggest grafting the ventral aspect of the corpora to extend its length.



### 64.3.2 Thiersch-Duplay Procedure

If the urethral plate seems to be wide and healthy, it can be tubularized following the Thiersch-Duplay technique. Its best indication is in the glanular hypospadias with intact prepuce (megameatus), where the urethral plate is thick, deep, and healthy. The incision lines follow each side of the urethral plate from the tip of the glans down to the division of the corpus spongiosum (Fig. 64.3). The two wings of the glans are dissected deeply and laterally until the corpora are clearly identified. The urethral plate is tubularized around an 8F catheter for children under 3 years of age, using a 6/0 or 7/0 absorbable running suture. The neourethra is then covered by two wings of the glans in one or two layers.

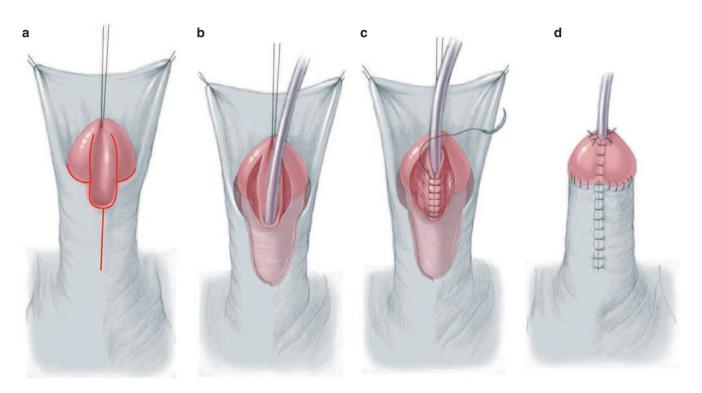


Fig. 64.3 (a-d) The Thiersch-Duplay technique for hypospadias repair

# 64.3.3 Snodgrass Procedure

In the Snodgrass procedure (Fig. 64.4), the urethral plate is incised longitudinally on its midline from the ectopic meatus up to the glans and subsequently tubularized around an 8F

catheter. This procedure leaves a dorsal raw area in the urethra, which is subsequently epithelized. Some suggest grafting the raw area with either foreskin or buccal mucosa; the technique is then called the inlay urethroplasty or "Snodgraft" procedure.

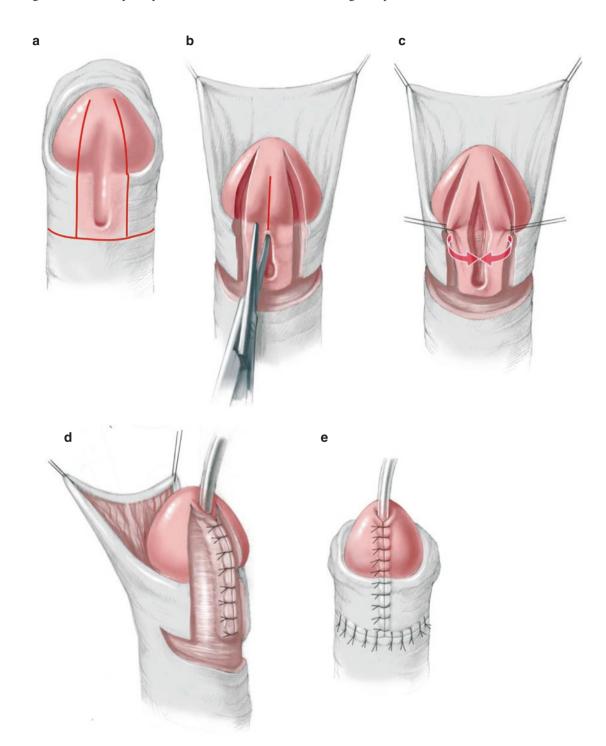


Fig. 64.4 (a–e) Snodgross procedure for hypospadias repair

# 64.3.4 Onlay Procedure

In the onlay procedure (Fig. 64.5), a rectangle of preputial mucosa is pediculized down to the base of the penis and

transferred to the ventrum of the penis to be layed on the urethral plate using interrupted 6/0 or 7/0 or a running suture.

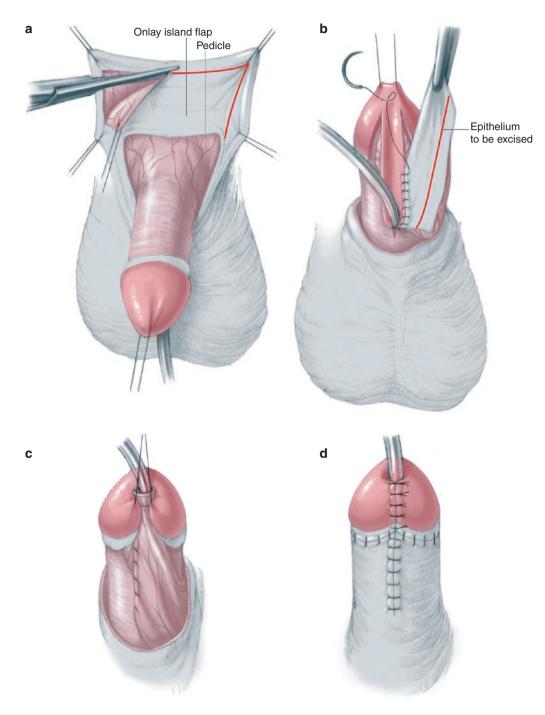


Fig. 64.5 (a–d) Onlay procedure for hypospadias repair

## 64.3.5 Mathieu Procedure

In the Mathieu procedure (Fig. 64.6), two parallel incisions are made on either side of the urethral plate up to the tip of the glans and deep down to the corpora cavernosa. The

incision line delimits a perimeatal-based skin flap that is folded over and sutured to the edges of the urethral plate. The lateral wings of the glans are generously dissected from the corpora cavernosa and approximated together, producing a conical shape of the glans.

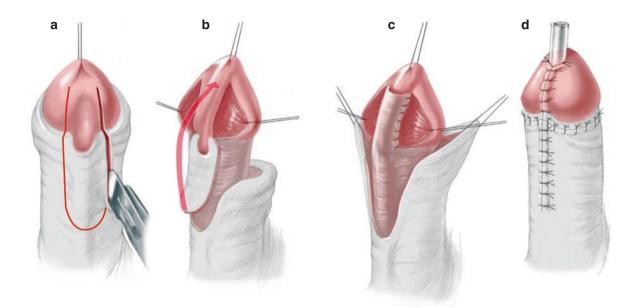


Fig. 64.6 (a–d) Mathieu procedure for hypospadias repair

### 64.3.6 Koff's Urethral Mobilisation

When the segment of the urethra to reconstruct is short (<1.5 cm) and when the distal urethra is healthy, a full mobilisation of the penile urethra can be performed following Koff's technique (Fig. 64.7). In these cases, the penile urethra is detached down to the base of the penis, and then it is moved upward to bring the meatus to the tip of the glans. The gain of length may be up to 15 mm. It is important to reattach the freed urethra to the corporeal surface using interrupted 7/0 resorbable stitches to avoid a ventral tilt of the glans and an iatrogenic curvature.

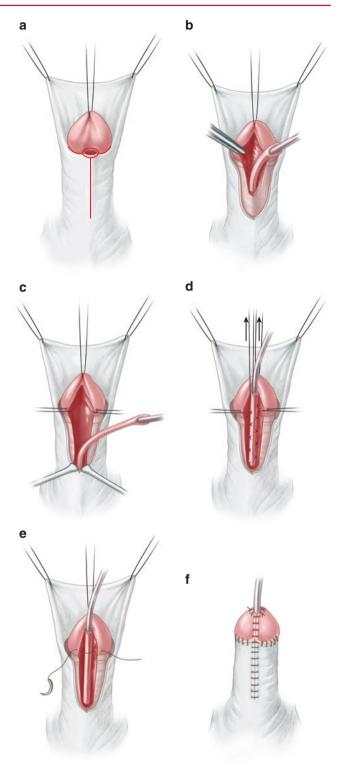
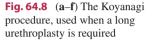


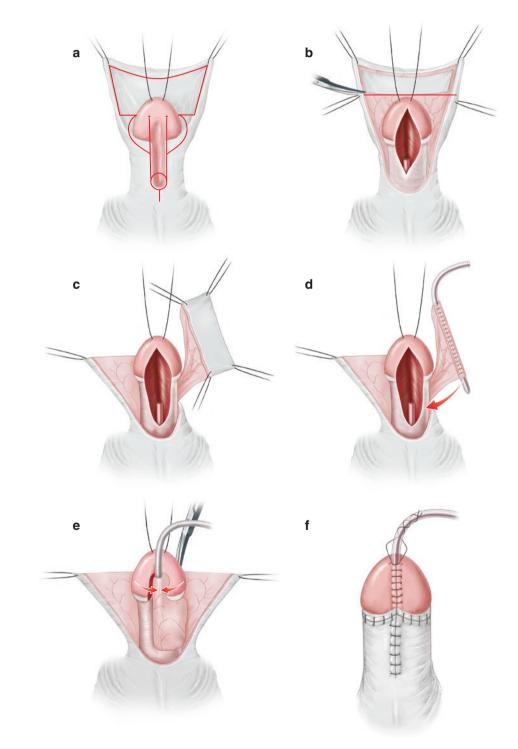
Fig. 64.7 (a–f) Koff's uretheral mobilisation technique for reconstruction of a short segment

### 64.3.7 Koyanagi Procedure

In the Koyanagi procedure, the initial incision lines follow a coronal circumference with a transection of the urethral plate just below the glans (Fig. 64.8). Then a second, racket-shaped incision allows complete mobilisation of the urethral plate and the adjacent tissues with the inner aspect of the preputial hood. A long, wide strip of well-vascularized tissue is freed with its blood supply down to the base of the penis.

The whole graft is transferred to the ventral aspect of the penis after the midline dorsal division of the inner aspect of the preputial hood. The skin flaps are joined together to reconstitute the basement of the future urethral plate, which is then duplay-ed from the ectopic meatus up to the glans. In the Hayashi modification, dorsal preputial skin is left intact to improve the blood supply of the distal graft. In the original Koyanagi procedure, the dorsal preputial skin is divided on the midline as shown in Fig. 64.8.





repair

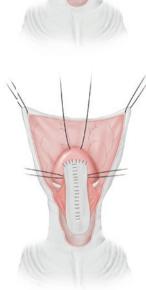
### 64.3.8 Bracka-Cloutier Procedure

Fig. 64.9 (a–f) The two-stage Bracka-Cloutier

Repair of some proximal hypospadias or some redo hypospadias may benefit from a two-stage procedure: The first stage aims at bringing fresh tissue (the internal aspect of the preputial hood or buccal mucosa) onto the ventrum of the

genital tubercle (Fig. 64.9). These free grafts need to find adequate blood supply to survive. The graft is stitched flat on the ventrum of the corpora from the ectopic meatus to the glans wings. It will be subsequently duplay-ed 3 months later once the graft take is confirmed.

b procedure using a free graft for hypospadias а С d







f

#### 64.4 Results and Conclusions

Hypospadias surgery remains a difficult challenge, as several factors of success remain unknown. One of the most intriguing aspects is the variation in the healing abilities of patients. With the development of tissue engineering, it is hoped that urethral substitution using the patient's own urethral tissue might be a future avenue to resolve current difficulties. Long-term follow-up of these patients appears to be crucial to assess and validate the various techniques currently available. The problem is how to follow these patients. Clinical examination of the penis is highly subjective, and assessment of the urine stream is difficult, as urine flow studies are very often abnormal after urethral reconstruction even if the caliber of the neourethra is correct. At the end of the day, the experience and honesty of the paediatric urologist remain the two most important factors for progress in hypospadiology.

Parents should be clearly informed that a significant number of hypospadias repairs will require further surgical attention during the patient's life. Because minor hypospadias are much less common than believed, this surgery should always be performed by experienced paediatric urologists. Collaboration with paediatric endocrinologists is also important, to increase the chances of surgical success. Preoperative and postoperative treatment may be helpful to improve each patient's healing abilities.

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