Current concepts in hypospadiology

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INTRODUCTION

Hypospadias can be defined as a 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 spongiosum, whose sides are represented by the two pillars of atretic spongiosum and whose base is the glans itself (Fig. 1).

In the middle of this triangle, from the tip to the base of the penis there are: a widely open glans, the urethral plate which extends from the ectopic urethral meatus up to the apex of the glans, the ectopic urethral meatus, and a segment of variable length of atretic urethra (not surrounded by any spongiosum) which starts where the corpus spongiosum divides [1].

Therefore it is possible to distinguish two main types of hypospadias: (i) one with a distal division of the corpus spongiosum with little or no chordee when the penis is erect (Fig. 2); and one with a proximal division of the corpus spongiosum with a marked degree of hypoplasia of the tissues forming the ventral radius, with a significant degree of chordee (Fig. 3). Besides these two main types there is also hypospadias for which several procedures have failed (hypospadias cripple; Fig. 4).

The causes of hypospadias are essentially unknown although several avenues have been explored to explain this anomaly: (i) Some endocrine disorders have been described in relation to hypospadias, mainly caused by an insufficient secretion of androgens, or insufficient response by the target tissues. However, in very few cases can these disorders be detected [2–4]; (ii) some genetic disorders [5] could explain why hypospadias can be found in several members of the same family; (iii) young and old mothers are more prone to carry a baby with hypospadias; babies of low birth-weight [6] and twins also have a higher risk of having hypospadias, possibly explained by a placental insufficiency [7]; (iv) the significant increase of hypospadias in the population over the last 20 years [8] raises the role of possible environmental factors [9], e.g. hormonal disruptors and pesticides, etc.; (v) abnormal or insufficient growth factors [10] could also be responsible for these penile anomalies and could explain the significant complication rate after surgery.

THE THREE MAIN STEPS OF HYPOSPADIAS SURGERY

Following the anatomical description, three surgical steps characterize hypospadias surgery [11]:

(i) The correction of chordee, which is essentially the result of the atresia of the ventral radius. The penile curvature is often a result of the tethering of the hypoplastic skin beyond the division of the corpus spongiosum onto the underlying structures, mainly the hypoplastic urethra. Degloving the penis represents the first step of this surgery and straightens the penis in 80% of cases. In 15% the persistent curvature is caused by an abnormal tethering of the urethral plate and the hypoplastic urethra onto the ventral aspect of the corpora. Freeing the urethral plate from the corpora is a valuable additional manoeuvre which straightens the penis in most cases (Fig. 5). Finally, in 5% of cases the penis remains curved although all the tissues forming the ventral radius have been freed. The residual chordee is caused by asymmetric corpora cavernosa and requires a dorsal corporoplasty (dorsal shortening of the albuginea of the corpora cavernosa; Fig. 6). In most cases the urethral plate can be preserved, although there are situations where the urethral strip is very poor and the two corpora twist around it; in these cases the urethral plate is usually sacrificed.

(ii) Once the penis is straight the missing urethra should be replaced. The technique chosen depends on the size and quality of the urethral plate. (a) If the urethral plate is wide and healthy, it can be tubularized following the Thiersch-Duplay technique (Fig. 7) [12,13]; (b) If it is too narrow to be tubularized, the Snodgrass urethrotomy [14] is one option (Fig. 8) or additional tissue can be laid on the urethral plate using a rectangle of pedicled preputial mucosa (onlay urethroplasty; Fig. 9 [15]), or a flap of ventral penile skin (Mathieu; Fig. 10 [16]); (c) If the segment of urethra to be replaced is short (< 2 cm) and if the distal urethra is not hypoplastic, complete mobilization of the whole penile urethra may be adequate to bridge the defect (Fig. 11). This technique [17], like the Thiersch-Duplay, has the advantage of avoiding the use of non-urethral tissue; (d) If the urethral plate is not preservable, a tube needs to be made to replace the missing urethra, using either a pedicled rectangle of preputial mucosa (Asopa-Duckett) [18] (Fig. 12) or buccal mucosa (Fig. 13) [19]; (e) Finally, in cripple hypospadias, no definite guidelines can be given, although the principles of repeat surgery are very similar to those of primary surgery. The procedures used are the same and sometimes need to be associated (composite urethroplasty) in complex situations.

(iii) Once the urethroplasty is completed the ventral radius of the penis needs to be reconstructed. This includes: (a) Meatoplasty creating a slit-shaped meatus; (b) glanuloplasty to reconstruct the ventral aspect of the glans; (c) creating a mucosal collar around the glans [20]; (d) coverage of the neourethra using the lateral pillars of spongiosum (spongioplasty); (e) skin cover with a redistribution of the skin shaft bringing the excess dorsal skin to the ventrum; (f) and some prefer to reconstruct the foreskin, others favour circumcision.

COMMON PROCEDURES USED IN PRIMARY CASES OF HYPOSPADIAS

(i) The urethroplasty procedure does not need to chosen before straightening the penis, and requires a very clear assessment of the quality of the urethral plate. (a) The incision lines are always the same, following each side of the urethral plate from the tip of the glans down to the division of the corpus spongiosum (Fig. 14); (b) The two wings of the glans are dissected deeply and laterally until the corporeal surface is clearly identified; (c) On

FIG. 1. Anatomy of the ventral radius of the penis.

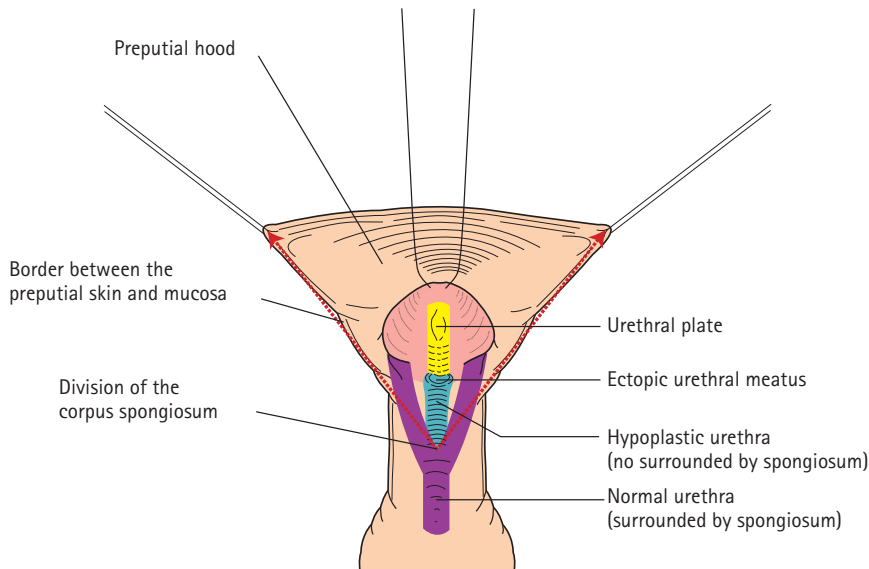FIG. 6. Dorsal plication of the corpora cavernosa.

FIG. 2. Hypospadias with a distal division of the corpus spongiosum.
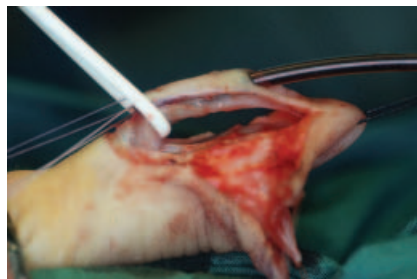

FIG. 3. Hypospadias with a proximal division of the corpus spongiosum.
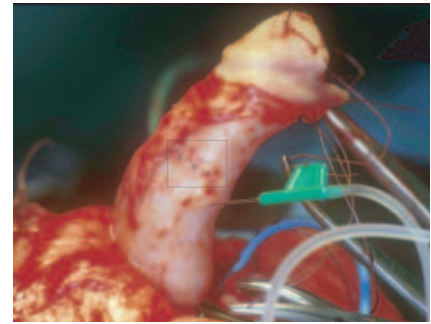

FIG. 4. Hypospadias cripple.

FIG. 5. The urethral plate is lifted off the ventral aspect of the corpora cavernosa.

each side, the incision line follows the border between the mucosal aspect and the cutaneous aspect of the preputial hood; (d) If the foreskin is not preserved the incision line continues on the dorsum following the same landmark; (e) The penis is fully degloved and a first erection test performed. As noted, the penile curvature is corrected in most cases but

in ≈20% it persists and for some the dissection of the urethral plate is by lifting it from the anterior aspect of the corpora. For others the urethral plate is kept attached to the ventral aspect of the corpora and a dorsal plication of the corpora performed using the Nesbitt principle.

(ii) If the urethral plate is wide and healthy, the Thiersch-Duplay (Fig. 7) procedure is used, where the urethral plate is tubularized around an 8 F catheter for children aged <3 years, using a 6/0 or 7/0 resorbable running suture.

(iii) If the urethral plate is healthy but too narrow to be tubularized around an 8 F catheter, either the urethral plate is incised longitudinally on its midline from the ectopic meatus up the glans and subsequently tubularized around an 8 F catheter (Fig. 8) (the Snodgrass procedure, which leaves a dorsal raw area in the urethra which subsequently epithelialises); or non-urethral tissues can be laid on the urethral plate to create a conduit. These comprise: The Mathieu procedure (Fig. 10), where the incision line delimits a perimeatal-based skin flap that is folded over and sutured to the edges of the urethral plate using 6/0 or 7/0 resorbable running sutures; the Onlay procedure (Fig. 9) where a rectangle of preputial mucosa is pedicled down to the base of the penis and transferred to the ventrum of the penis to be laid on the urethral plate; a rectangle of buccal mucosa harvested from the inner aspect of the lower lip or the inner aspect of the cheek after identifying the Stenon duct. The buccal graft is laid on the urethral plate following the same principles as above (Fig. 13). If the segment of urethra to reconstruct is short (<2 cm) and the length of hypoplastic urethra proximal to the ectopic meatus is short, a full mobilization of the
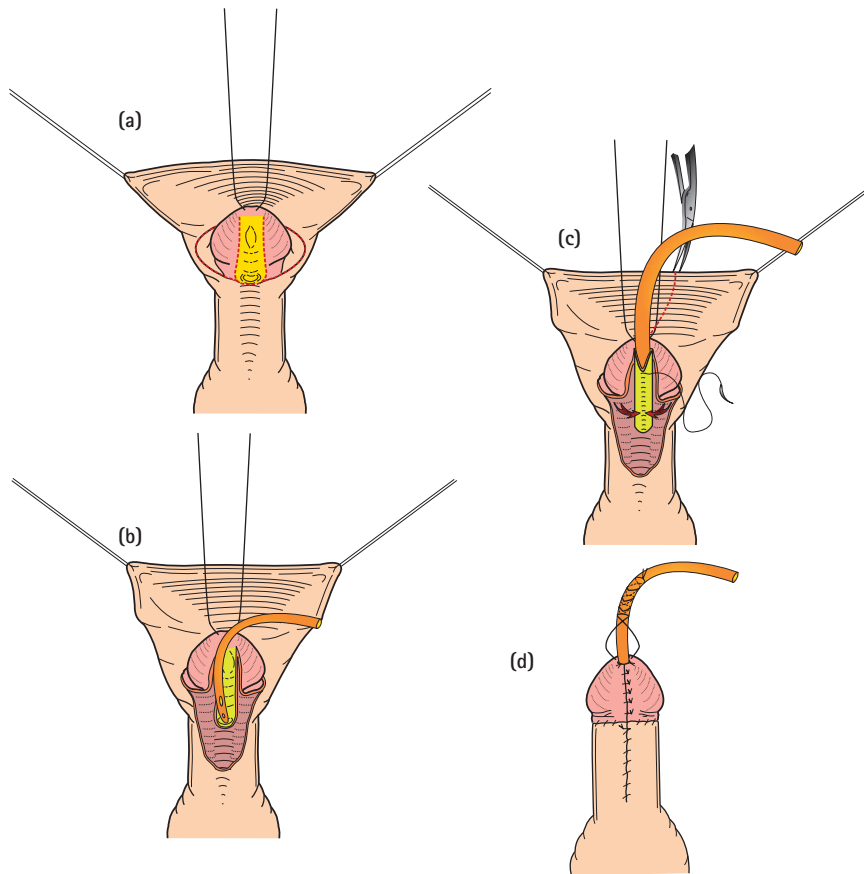
FIG. 7.
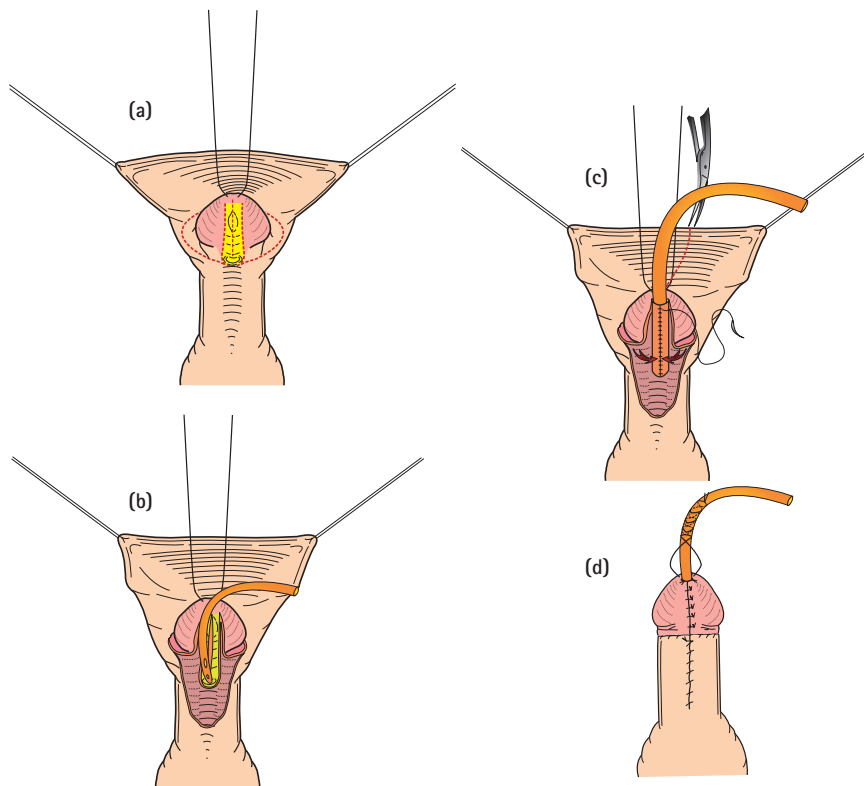Thiersch–Duplay procedure.

FIG. 8.
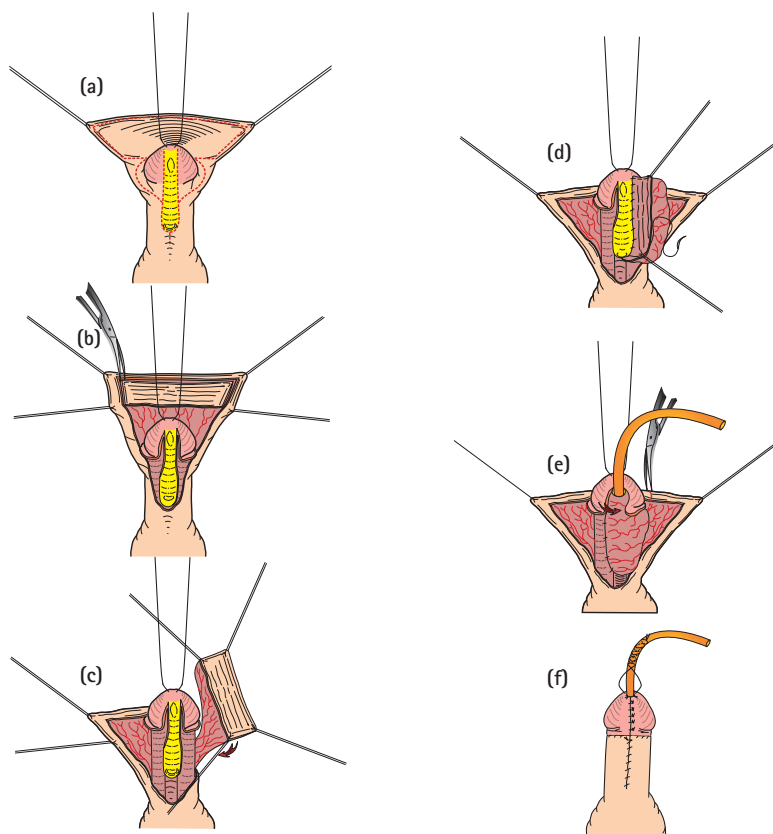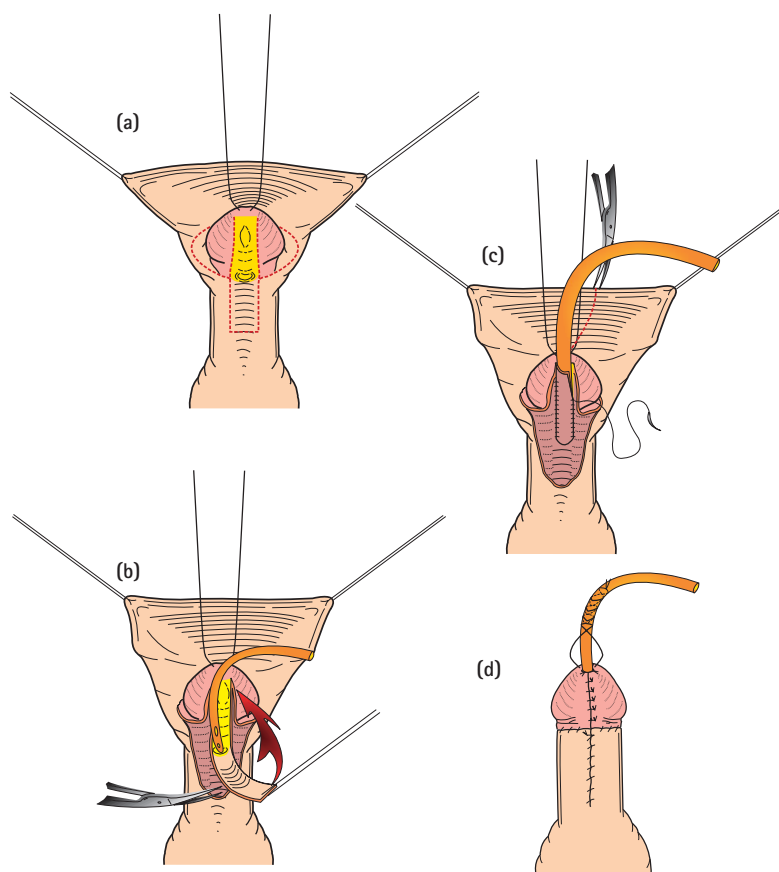Snodgrass procedure.

FIG. 9.
Onlay procedure.
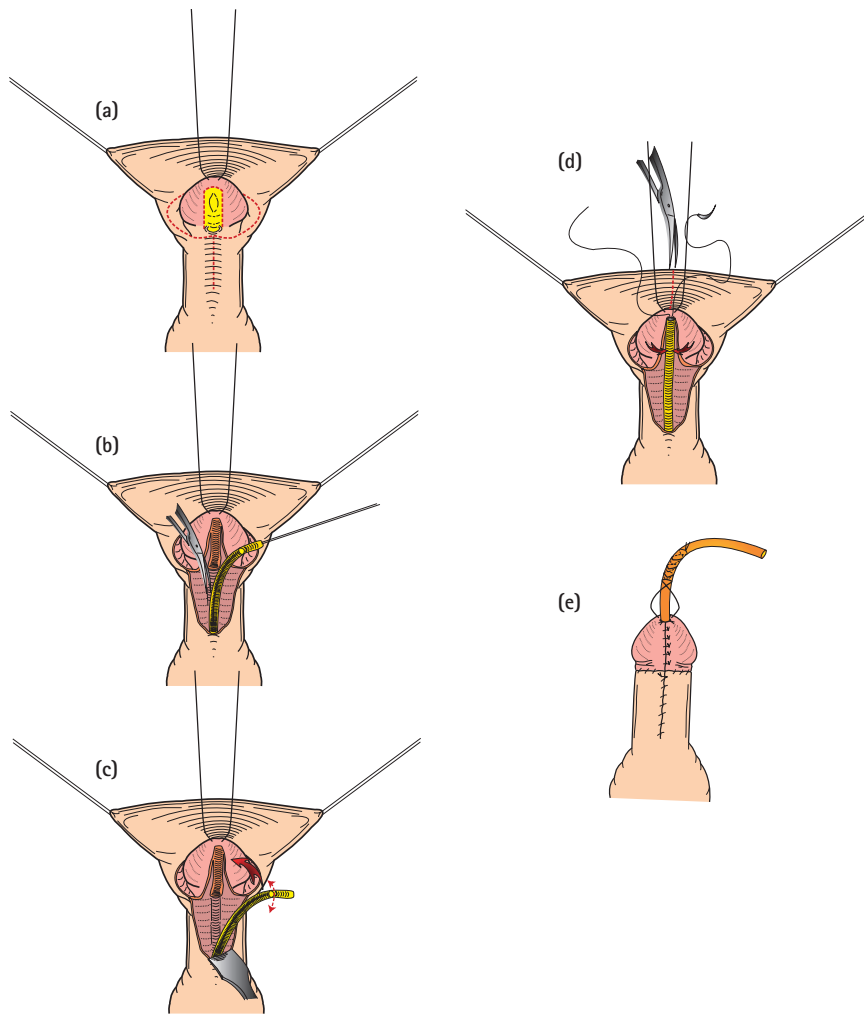

FIG. 10.
Mathieu procedure.

FIG. 11.
Koff procedure.

penile urethra can be used following Koff's technique (Fig. 11).

(iv) The urethral plate is too poor to be kept a full tube urethroplasty is needed using either a pedicled rectangle of preputial mucosa (Fig. 12) or a rectangle of buccal mucosa. The main disadvantage of these techniques is that a proximal circular urethral anastomosis is used, which exposes the repair to the risk of urethral stenosis.

(v) Once the penis is straightened and the urethroplasty completed the ventral radius of the penis is reconstructed following the steps mentioned above.

OTHER PROCEDURES

There are many other procedures described to repair the missing urethra, which clearly shows that no one is totally satisfactory.

(i) The MAGPI [21] procedure was very fashionable in the 1980s to repair distal hypospadias; in fact it is not a meatal advancement but a flattening of the glans, which gives the illusion that the meatus reaches the glans apex. Secondary retraction of the meatus are quite common and therefore this procedure has become less popular.

(ii) Various flip-flap procedure have been described using the Mathieu principle. The Devine-Horton [22] procedure was quite fashionable in the 1980s until it was noted that the urethral plate should be preserved in most cases.
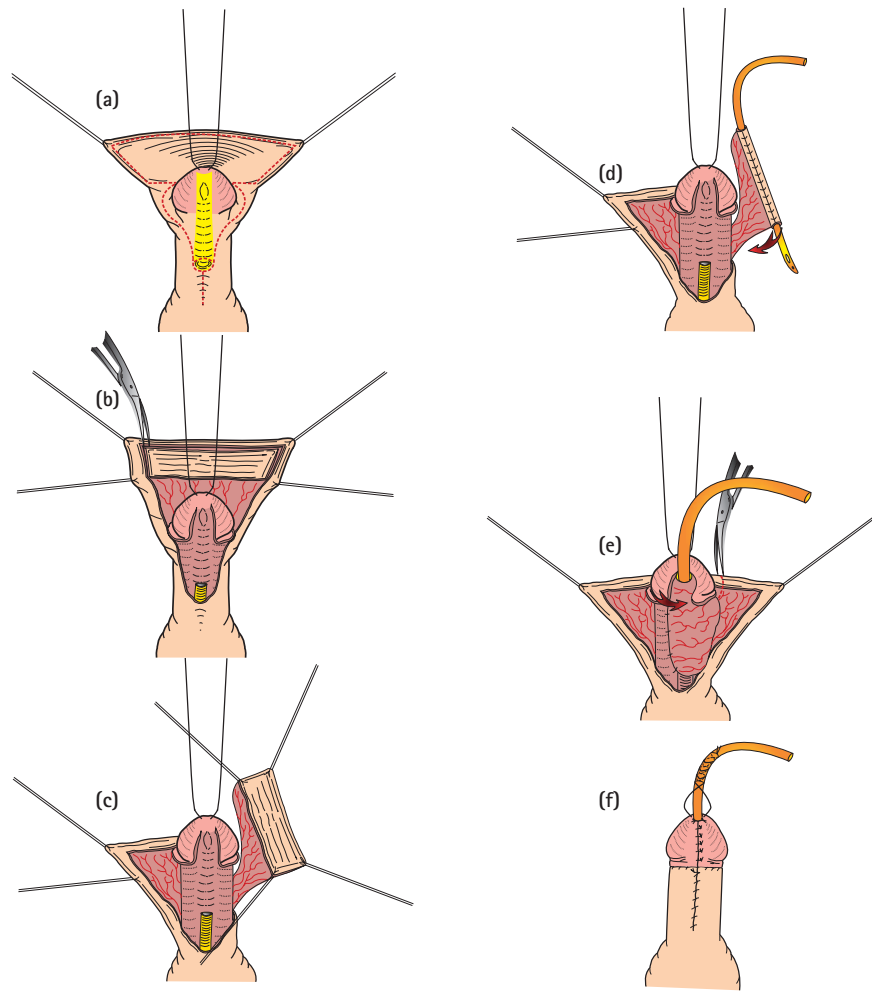
(iii) The bladder graft urethroplasty (Fig. 15) [23] seemed to be a logical option in the late 1980s as it is the same cell lining as the urethra. Unfortunately complications were common and with the use of buccal mucosa, this technique has become obsolete.

(iv) Two-stage procedures have been popularized again by Bracka [24], who resurrected the Cloutier [25] procedure. In a first step the penis is straightened after complete removal of the urethral plate. The ventral radius of the penis is grafted by preputial mucosa or buccal mucosa and then tubularized a few weeks later. Although these techniques are not commonly used they certainly have solid indications in severe cases or repeat surgery.

SURGICAL TIPS

The age at surgery for primary hypospadias repair is usually 6–24 months. Hormonal stimulation of the penis using βhCG or testosterone or dihydrotestosterone is sometimes indicated in patients with a small penis, or for repeat surgery; it is unclear how safe these treatments are in the long-term. General anaesthesia is the rule, often

FIG. 12.
Asopa-Duckett procedure.

RESULTS

Assessing hypospadias surgery requires extensive experience and honesty, as there are no reliable objective methods to evaluate functional and cosmetic results. Very few publications report long-term outcomes of these various procedures. There are also large differences between what the patient (or family) thinks and the surgeon's evaluation. Urinary flow studies are often abnormal even after a successful hypospadias repair, as quite often the child changes his bladder behaviour after repair. However, the aim should be a straight penis with no redundant skin, with a slit-shaped meatus, with regular scars and an aesthetically reconstructed ventral aspect of the glans. The child should be able to pass water while standing, with no spraying, no straining, no pain, in a single stream from the apex of the glans. The penis should be straight when erected.

MAIN COMPLICATIONS

A poor cosmetic result is the most common complication, often related to irregular and asymmetric scars with skin 'blobs' and an excess of ventral skin (Fig. 17). The mucosal collar is an important addition to improve penile cosmesis.

Urethral fistula is the second most common problem met in this surgery (Fig. 18), often lateral at the coronal level. The temptation to simply close the fistula is dangerous, as recurrent fistulae are quite common at this level. Often it is more advisable to repeat the whole urethroplasty. Urethral fistula can also be associated with a urethral stricture, which should be treated concomitantly. The causes of fistulae remains unknown although it is likely that local infection, local ischaemia, an inadequate procedure or the poor

associated with caudal or penile anaesthesia. Magnification is commonly used in this surgery but coagulation is often not needed when the tourniquet is used, followed by a slightly compressive dressing. Other surgeons prefer bipolar coagulation or adrenaline injection before incision. Antibiotics protocols are extremely variable among centres, and again the efficacy needs to be confirmed. The use of urine drainage via a suprapubic catheter, a transurethral bladder catheter or dripping urethral stent varies among surgeons; some use no drainage. The dressing is essential after surgery and also varies. The 'daisy dressing' (Fig. 16) is our favourite as it is very comfortable for the patient and contains postoperative bleeding. Others prefer Opsite, silastic foam or Tagaderm dressings. Pain control is essential after surgery using morphine instillations, anti-inflammatory medications, anticholinergics and diazepam to reduce bladder spasms.

FIG. 13. (a) Buccal graft urethroplasty: the inner aspect of the lower lip. (b) Onlay buccal graft urethroplasty (hypospadias cripple).

FIG. 14. The incision lines follow each edge of the urethral plate from the apex of the glans down to the division of the corpus spongiosum. The lines following the border between the preputial skin and the preputial mucosa cross where the corpus spongiosum divides.

FIG. 15. (a) Bladder graft urethroplasty: anterior aspect of the bladder after detrusorotomy. (b) Bladder graft urethroplasty (tube).
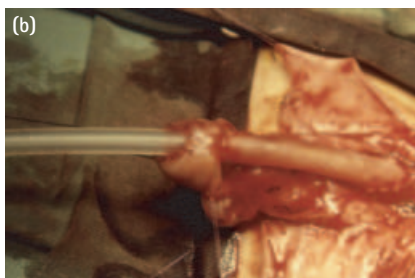

FIG. 16. Daisy dressing and double nappies.

FIG. 17. A poor cosmetic result.
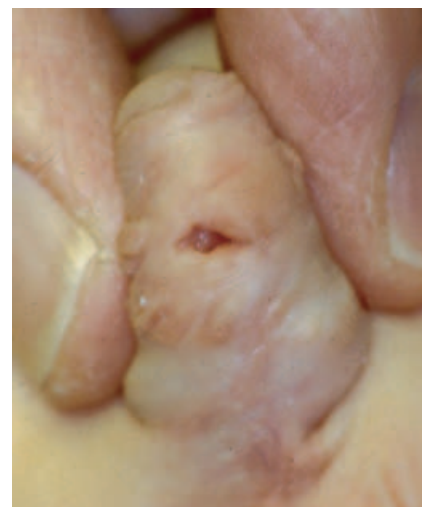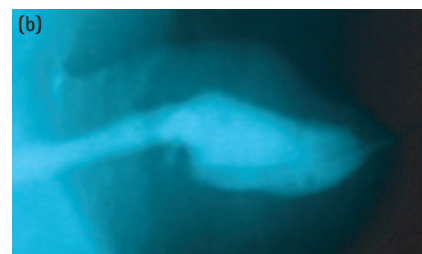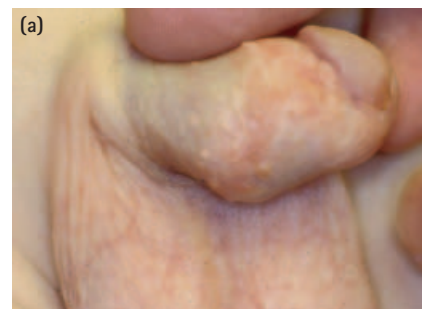

FIG. 18. Urethral fistula.

FIG. 19. (a) Urethrocele with a distal stenosis of the urethra. (b) X-ray of the urethra of the same patient.

healing abilities of the patient might be involved.

Urethral strictures are less common as onlay urethroplasties are more popular and avoid circular anastomosis, which certainly increases this risk. Urethral dilatations are very poorly accepted in children. Internal urethrotomies have a poor record and often a segmental urethroplasty is needed using a patch onlay with buccal mucosa. In other cases a full repeat urethroplasty is preferable. Meatal stenosis remains a common complication and requires either dilatation or better, a meatotomy or meatoplasty.

A urethrocele (Fig. 19) is caused by the progressive distension of the reconstructed urethra, possibly resulting from a distal urethral stenosis or the difference of compliance of the tissues used for urethroplasty compared to the native urethra. Excision of the redundant tissue is the usual treatment, with a good backing of the neourethra with the surrounding tissues to avoid further urethral distension.

Balanitis xerotica obliterans is an annoying complication as it tends to recur; it causes recurrent meatal stenosis and sometimes pain on voiding when it develops inside the reconstructed urethra. Its cause is unknown.

Urethral prolapse (bladder mucosa), urethral stones (hairy skin), sticking meatus (bladder mucosa), meatal retraction (MAGPI) are also reported but are probably less common with modern techniques.

Results with some of the most common techniques include: (a) The Mathieu procedure gives excellent results, with 1% meatal stenosis and ≈5% fistula rate [26]. One common criticism of the Mathieu procedure is its half-moon shaped meatus. (b) The Koff urethral mobilization has a very low fistula rate but meatal stenosis is common, especially if the distal urethra left in place is hypoplastic. In our experience the meatal stenosis rate is ≈20% [27]. (c) The Duplay-Snodgrass procedure has a much higher complication rate in proximal hypospadias (about a third of patients required complementary surgery) [28]. (d) The Onlay procedure has a complication rate of 15–50%; Elbackry [29] reported a 7% breakdown, 23% fistulae, 9%

strictures and 3% diverticulae. (e) Buccal graft urethroplasties often require subsequent surgical revisions. One group reported a 57% complication rate with five (of 30) with meatal stenosis, seven strictures, two fistulae and one breakdown over a 5-year follow-up [30]. (f) The Asopa/Duckett tube has a poor long-term record, as does bladder mucosa graft urethroplasty; 66% of Ransley's patients required 1–9 additional procedures to treat complications [31]. The most common complications were meatal stenosis and/or urethral prolapse. (g) Two-stage procedures produce a good cosmetic result but significant urethral complications, e.g. stenosis in 7% [24]. (h) Composite repair using the prepuce has a reoperation rate of almost half [32].

CONCLUSIONS

Hypospadias surgery remains a difficult challenge, as several factors contributing to success remain unknown. One of the most intriguing is the variation in the 'healing ability' among patients. With the development of tissue engineering it is hoped that urethral substitution using the patient's urethral tissue might be a future avenue to resolve the current difficulties. A long-term follow-up of these patients appears to be crucial for assessing and validating the various techniques currently available. The problem is how to follow these patients. Clinical examination of the penis is highly subjective; assessing the urinary stream is difficult, as urine flow studies are very often abnormal after urethral reconstruction. Finally, the experience and honesty of the paediatric urologist remain the two most important factors in hypospadiology. Collaboration with paediatric endocrinologists is also important to increase the chances of surgical success. Treatment before and after surgery may be helpful for improving the patient's 'healing ability'.

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