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Surgery Illustrated – Surgical Atlas
Primary closure of bladder exstrophy

Timothy M. Phillips and John P. Gearhart
Paediatric Urology, Brady Urological Institute, Johns Hopkins Hospital, Baltimore, MD, USA

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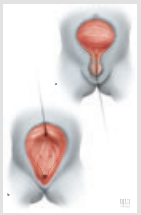

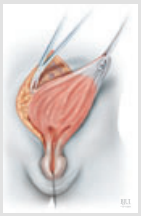
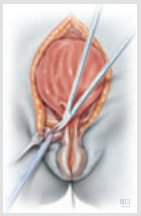
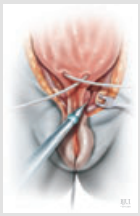

PLANNING AND PREPARATION

Before undertaking closure of primary exstrophy the surgeon must decide whether a formal osteotomy will be required, and if so, ensure that there is appropriate orthopaedic support for this. Also, the anaesthesia staff, nursing staff and parents need to be prepared to support a prolonged admission while the child is supine and in traction after surgery.

Supplies include:

- Interfragmentary pins, external fixator, and orthopaedic instruments (osteotomy, if required);
- Supplies for Buck's or Bryant's traction (as needed);
- 3- and 5-F paediatric feeding tubes (ureteric stents);
- 10 F Malecot drain (suprapubic tube);
- 2–0 silk ties (umbilical vessels);
- 3–0 and 5–0 polydioxanone sutures (bladder and urethral closure);
- 3–0 nylon, 4–0 chromic, 3–0 polyglactin (secure drains);
- No. 2 nylon sutures (intrapubic stitch);
- Paediatric Van Buren sounds.

INDICATIONS

Whilst other methods of treatment of the newborn with bladder exstrophy have been offered, we routinely use a modern staged reconstruction of bladder exstrophy. This includes: bladder and abdominal wall closure, and urethral closure onto the penis, in the newborn period, with bilateral innominate and vertical iliac osteotomy, if indicated (Stage 1); epispadias repair at 0.5–1 year old

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TABLE 1 The initial presentation and management of exstrophy of the bladder

Age	Problem	Possible solution
	Initial presentation	
0–72 h	Classic exstrophy with reasonable capacity and moderate symphyseal separation; long urethral groove; mild dorsal chordee	I: Midline closure of bladder, fascia, and symphysis to level of posterior urethra; no osteotomy. In *very* selected cases combined bladder closure and epispadias repair
0–72 h	Above findings with short urethra and severe dorsal chordee	II: Close as in I, adding lengthening of dorsal urethral groove by para-exstrophy skin (cautiously)
0–72 h	Above findings with very wide separation of symphysis or late presentation (>72 h, up to 1–3 years) for initial treatment	Osteotomy (combined anterior and vertical iliac) and closure as in I or II
0–2 weeks	Very small, non-distensible bladder patch	Prove by examination under anaesthesia, then non-operative expectant treatment awaiting internal or external diversion or delayed closure if bladder plate grows

(Stage 2); and bladder neck reconstruction along with antireflux procedure at age 4–5 years, when the child has achieved an adequate bladder capacity for bladder neck reconstruction and is motivated to participate in a postoperative voiding programme (Stage 3). Primary bladder, abdominal wall, and proximal urethral closure are outlined here (Stage 1). Other methods of primary bladder exstrophy closure, such as the complete primary repair of exstrophy or Kelly repair can be used, but the basic surgical principles of closure are similar.

PATIENT SELECTION

The size and the functional capacity of the detrusor muscle are important considerations for the eventual success of functional closure. Sometimes a good portion of previously unappreciated bladder can be discovered behind the fascia, under examination with anaesthesia. The exstrophied bladder that is estimated at the time of birth to have a capacity of ≥5 mL, and has elasticity and contractility, can be expected to develop to a useful size and capacity after successful bladder, posterior urethra and abdominal wall closure with early epispadias repair. Table 1 provides a guideline for managing bladder closure according to variations in initial presentation.

SPECIFIC PATIENT PREPARATION

At birth, although the bladder mucosa is usually smooth, pink and intact, it is also sensitive and easily denuded. In the delivery room the umbilical cord should be tied with 2–0 silk close to the abdominal wall, so that the umbilical clamp does not traumatise the delicate mucosa and cause excoriation of the bladder surface. The bladder can then be covered with a non-adherent film of plastic wrap (e.g. Saran wrap) or a hydrated gel dressing, to prevent sticking of the bladder mucosa to clothing or diapers. In addition, each time the diaper is changed the dressing should be removed, the bladder surface irrigated with sterile saline, and clean plastic wrap or dressing placed over the bladder surface area. The surgeon should ensure that any serious coexisting medical problems that would prevent bladder closure and subsequent immobilization are stabilized before taking the child to the operating room. Before and during the procedure, the patient is given broad-spectrum antibiotics. The procedure is performed under a combined general and tunnelled caudal epidural anaesthetic technique.

SPECIFIC PATIENT POSITIONING

For the entire procedure, including anterior osteotomy (if indicated) the patient is placed supine with the body circumferentially prepared, draped and exposed from the nipples distally. A small towel is placed beneath the sacrum and the bladder is covered with soft absorbent gauze during the time of osteotomy (if indicated).

SURGICAL STEPS

Figure 1

Children undergoing closure after 72 h of life, those with very wide separation of the symphysis (>4 cm), or those with a pelvis that is not malleable, will typically undergo a bilateral anterior transverse innominate and vertical iliac osteotomy, performed by orthopaedic colleagues. At our institution, this is done by the orthopaedic team before bladder closure. Interfragmentary pins are placed as shown.

After soft tissue closure and completion of the urological procedure, an external fixator is placed to buttress the pelvis in the corrected position. Modified Buck's traction is applied after surgery. Osteotomy together with posterior urethral, bladder and abdominal wall closure is a 5–7 h procedure in these infants.

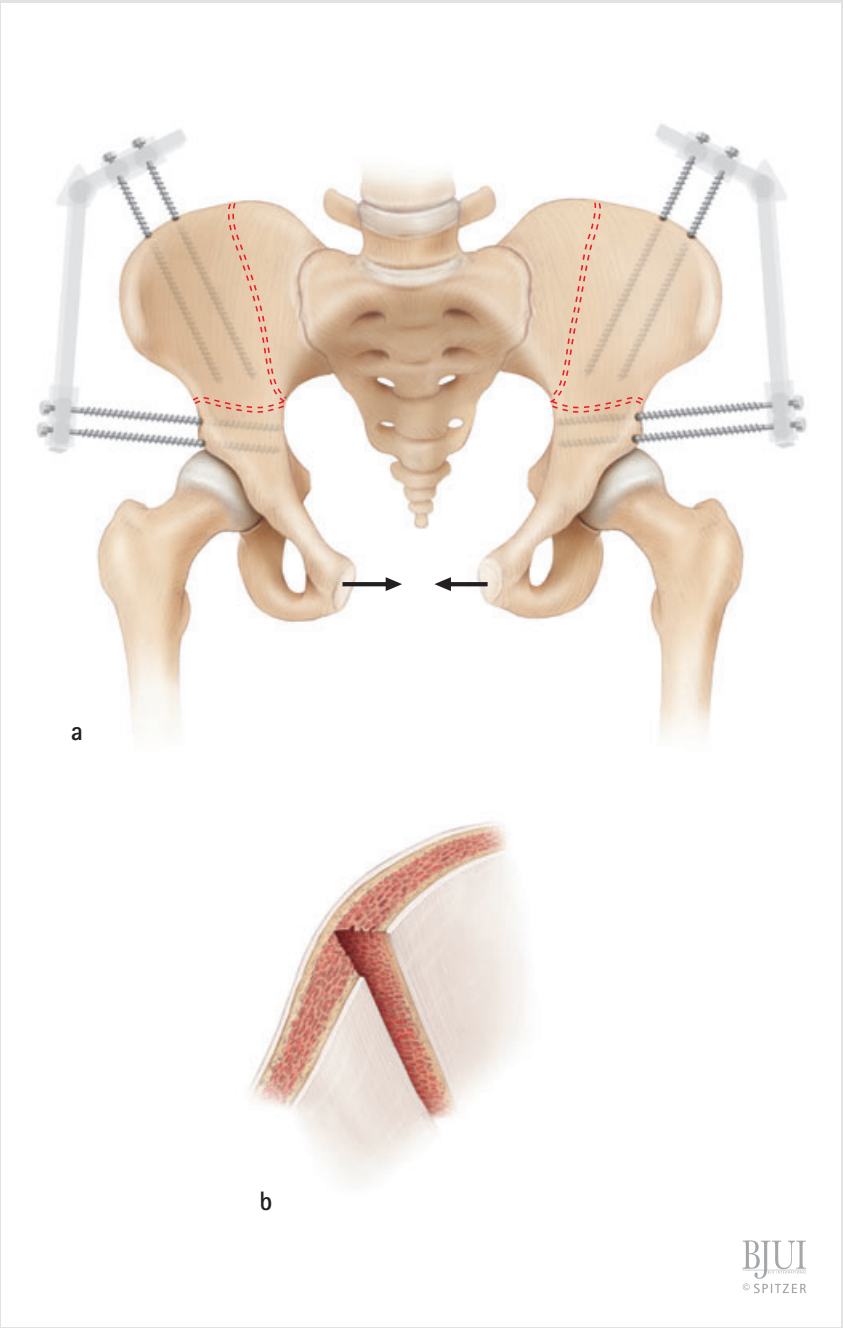


Figure 2

A strip of mucosa 2 cm wide, extending from the distal trigone to below the verumontanum in the male (a) and to the vaginal orifice in the female (b), is outlined and incised for prostatic and posterior urethral reconstruction in the male and adequate urethral closure in the female. The male urethral groove might be adequate, in which case no transverse incision of the urethral plate is needed for urethral lengthening. We tend not to incise the urethral plate unless the length of the urethral groove from the verumontanum to the urethral glans is so short that it interferes with eventual penile length and reduces dorsal angulation. If so, the urethral groove is lengthened using a modified Duckett para-exstrophy skin-flap technique (see below).

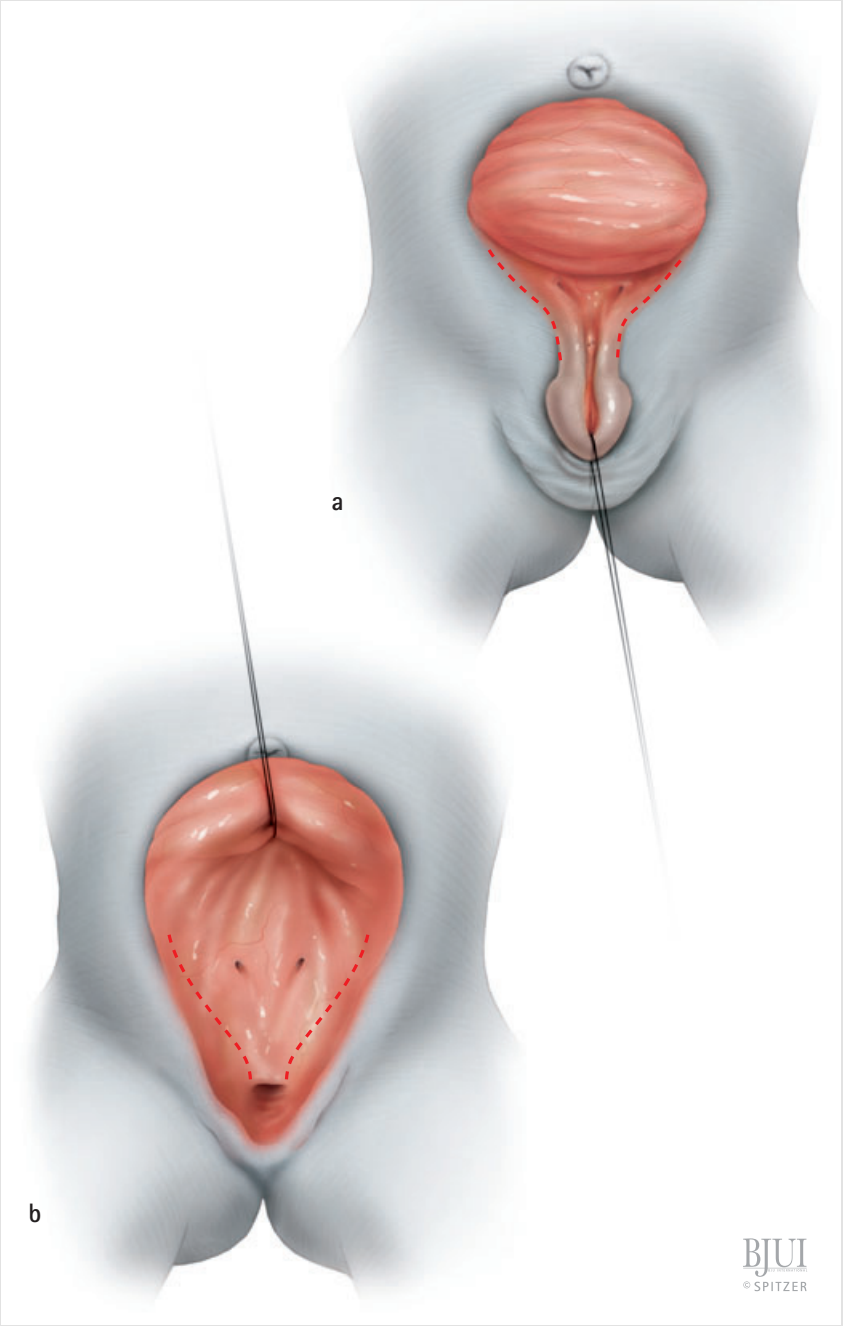


Figure 3

The bladder plate is then dissected off the anterior abdominal wall as shown, with an incision extending from just above the umbilicus, down the junction of the bladder and para-exstrophy skin, to the level of the urethral plate.

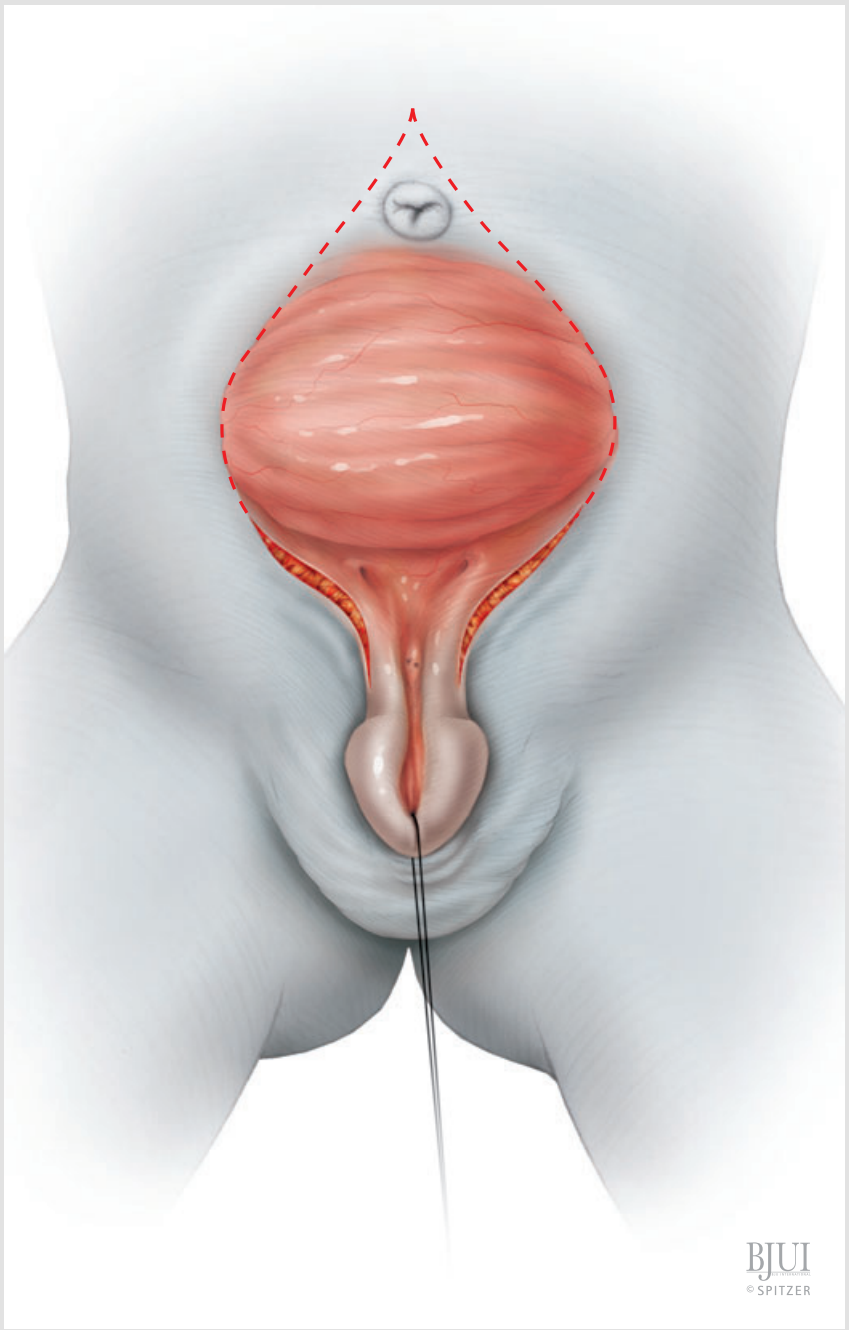


Figure 4

An appropriate plane is entered just above the umbilicus, and a plane is established between the rectus fascia and the bladder. The umbilical vessels are doubly ligated (3–0 polyglactin) and incised and allowed to fall into the pelvis. The peritoneum is taken off the dome of the bladder at this point so that the bladder can be placed deep into the pelvis at the time of closure. The plane is continued caudally down between the bladder and rectus fascia until the urogenital diaphragm fibres are encountered bilaterally.

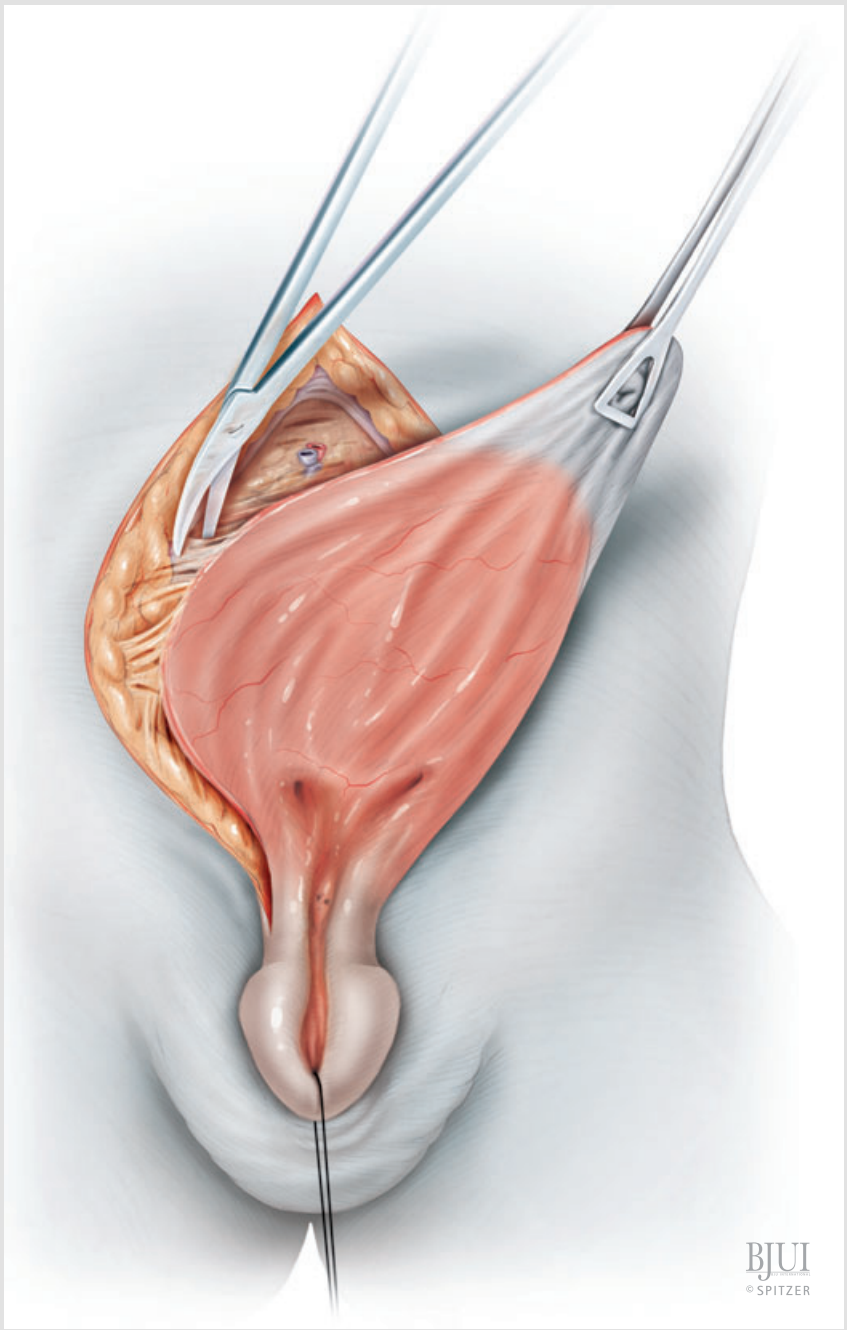


Figure 5

The pubis is encountered with continued caudal dissection. Gentle traction on the glans at this point shows the insertion of the corporal body on the lateral inferior aspect of the pubis. The penis is lengthened by exposing the corpora cavernosa bilaterally and freeing the corpora from their attachments to the suspensory ligaments on the anterior part of the inferior pubic rami.

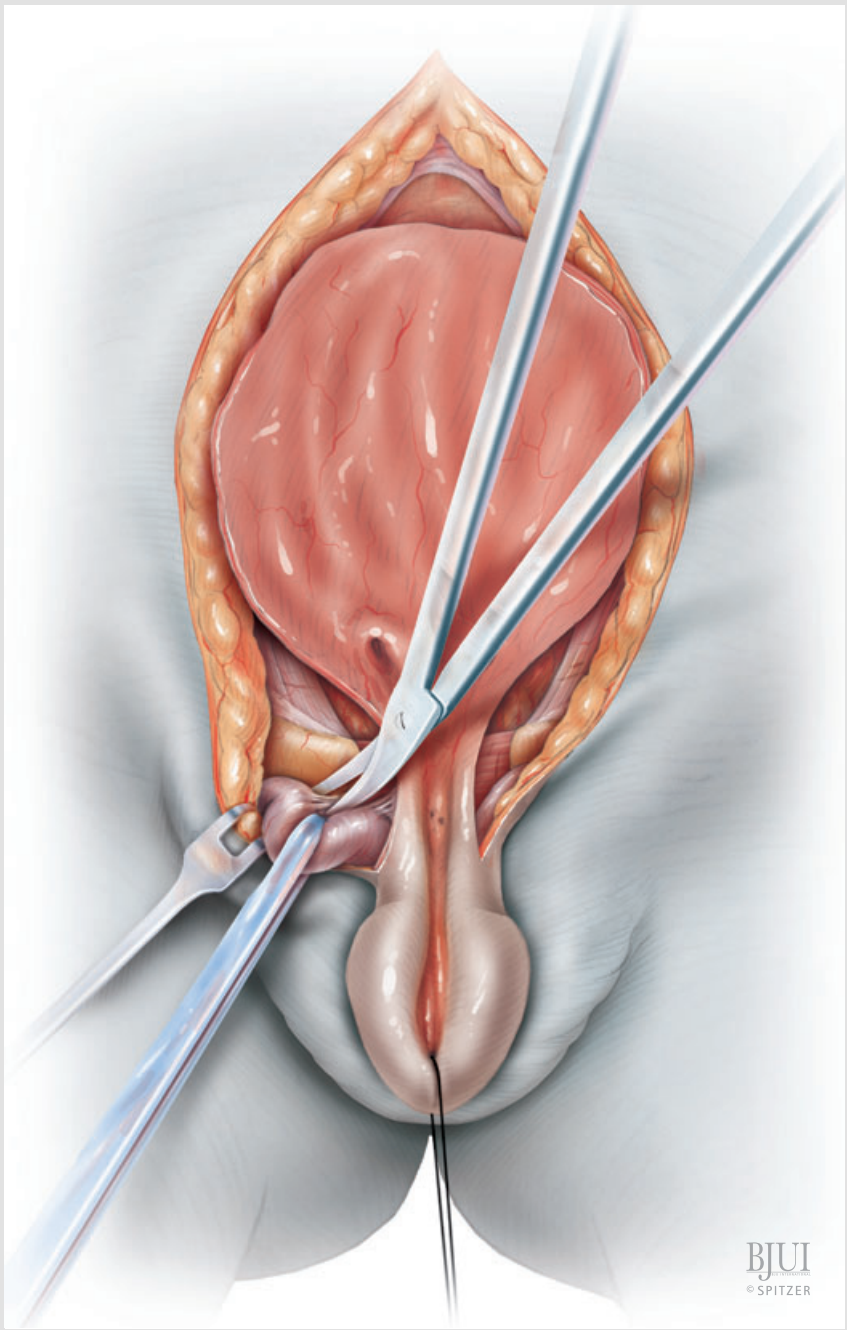


Figure 6

A double-pronged skin hook can be inserted into the bone at this time and pulled laterally to accentuate the urogenital diaphragm fibres, and to help the surgeon radically incise these fibres between the bladder neck, posterior urethra and pubic bone. The visualized urogenital diaphragm fibres are taken down sharply with electrocautery to the levator hiatus in their entirety. If this manoeuvre is inadequate the posterior urethra and bladder will not be placed deeply into the pelvis, and when the pubic bones are brought together, the posterior vesico-urethral unit will be brought anteriorly into an unsatisfactory position for later reconstruction. Ureteric catheters (3–5 F) are placed bilaterally, as well as a 10 F Malecot catheter being placed through the dome of the bladder for suprapubic drainage. The ureteric catheters will be passed through the anterior bladder wall at the time of closure. The ureteric catheters are secured directly to the ureteric orifice with 4–0 chromic sutures. The suprapubic tube is secured to the outside of the bladder with two sutures of 3–0 polyglactin, then at the skin with two sutures of 3–0 nylon. The ureteric stents are steri-stripped to the suprapubic tube at the skin. The corporal bodies are not brought together at this juncture, because later Cantwell-Ransley epispadias repair will require the urethral plate to be brought beneath the corporal bodies. If the urethral plate is left in continuity, it must be mobilized up to the level of the prostate to create as much additional urethral and penile length as possible. Further urethral lengthening can be done at the time of epispadias repair. The wide band of fibres and muscular tissue representing the urogenital diaphragm is detached subperiosteally from the pubis bilaterally. Reluctance to free the bladder neck and urethra from the inferior ramus of the pubis moves the neobladder opening cephalad should any separation of the pubis occur during healing, increasing the chance of bladder prolapse.

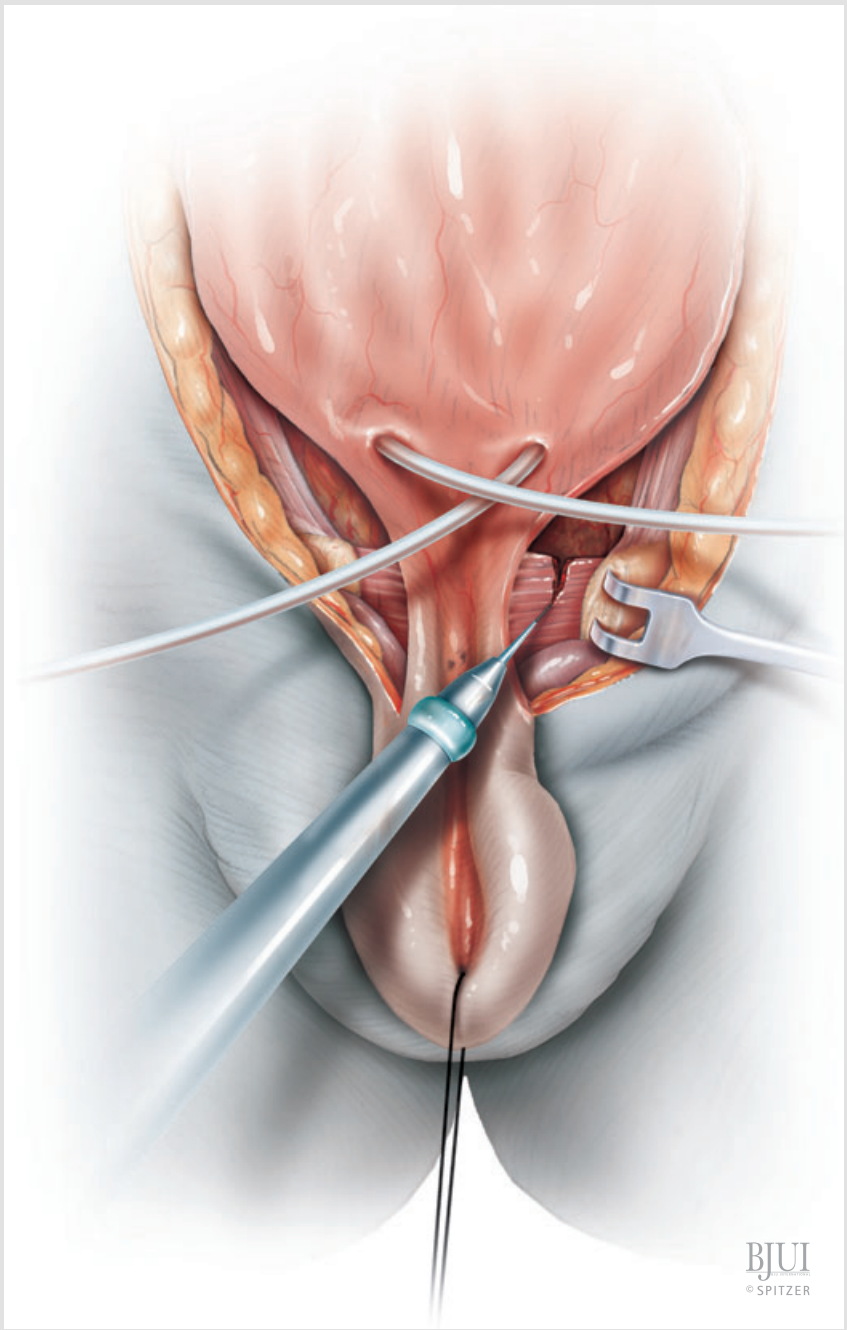
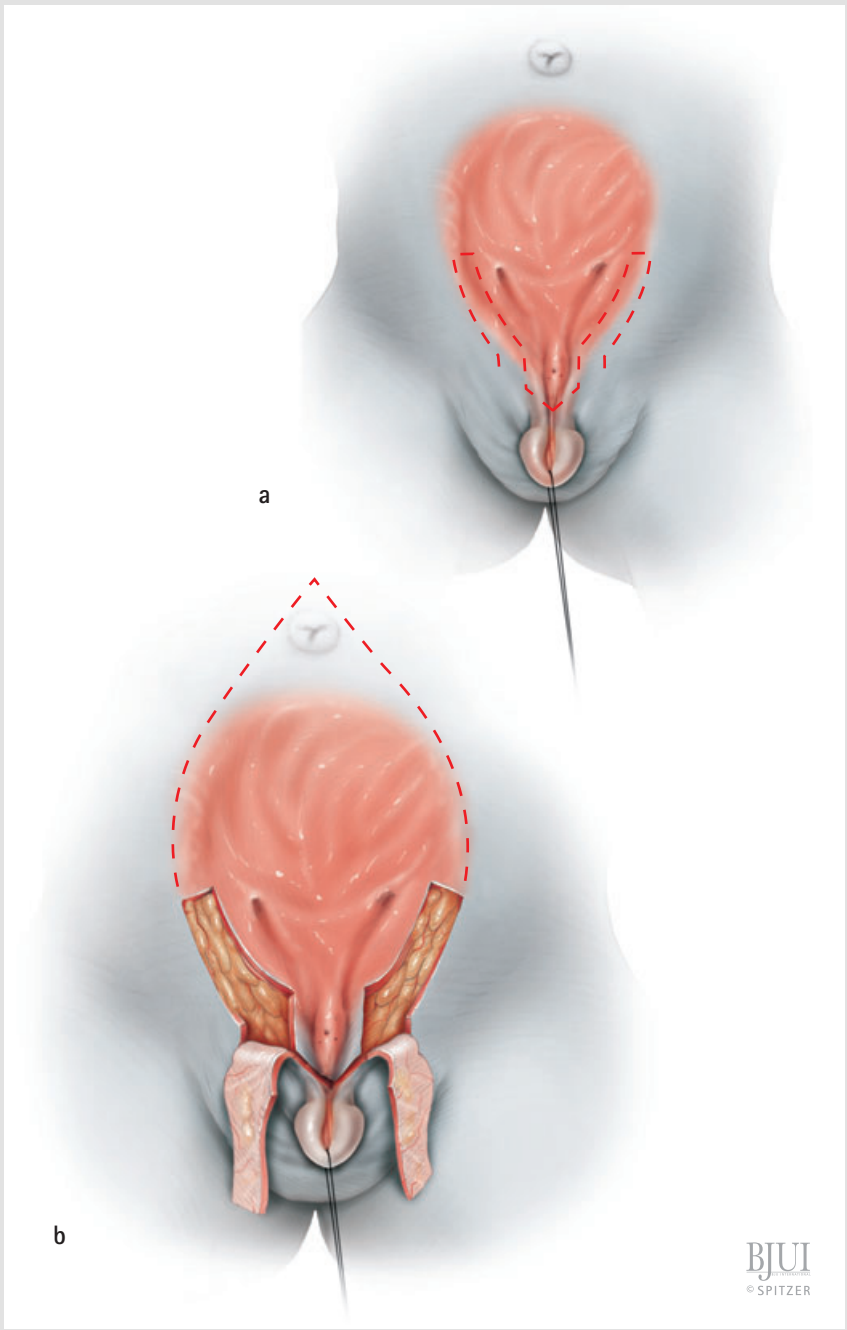


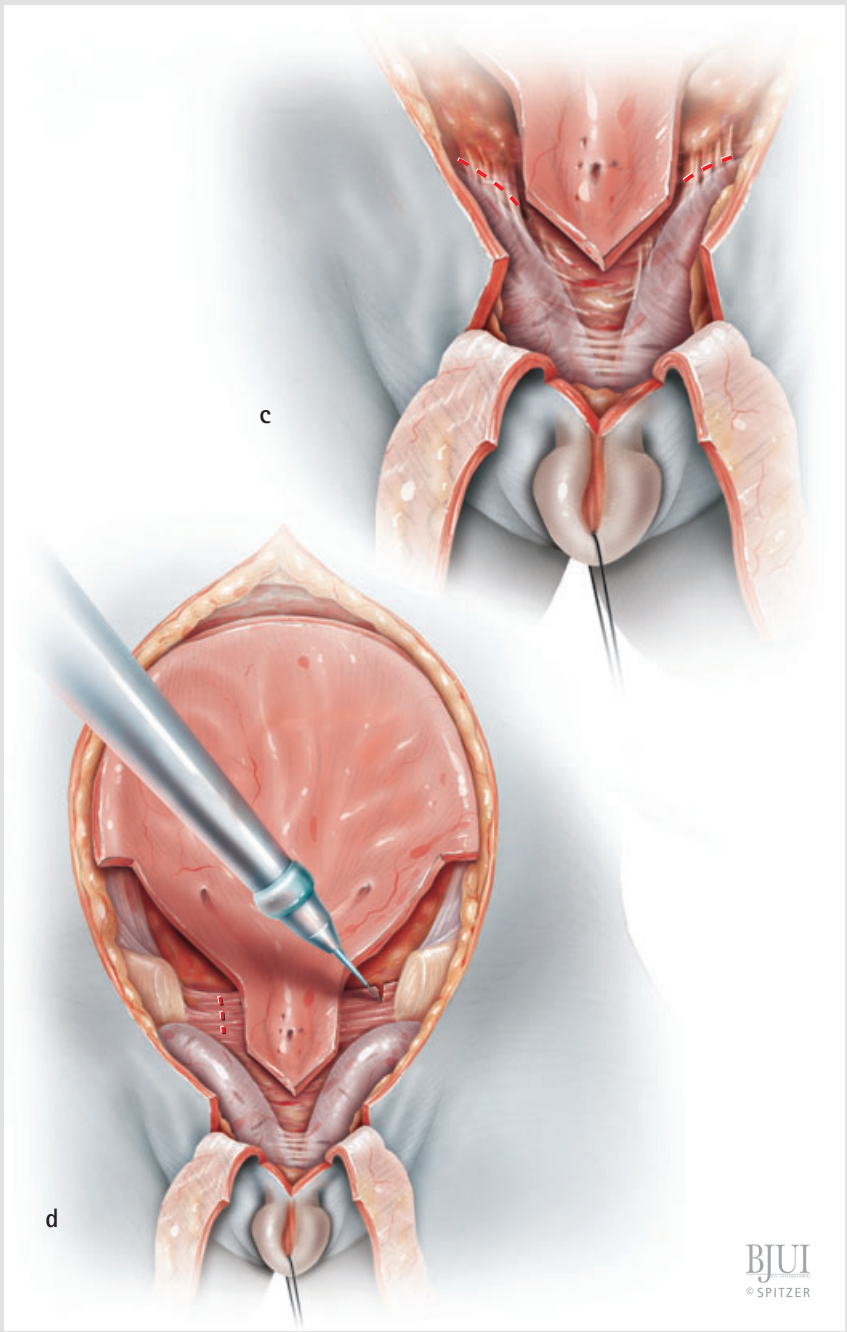
Figure 7

If the decision is made at this point that the urethral groove is to be transected, caudal glabrous para-exstrophy skin at the inferolateral borders of the bladder template are outlined and incised (Fig. 7a). A V-incision is made transversely through the urethral plate 1 cm distal to the verumontanum, and full-thickness para-exstrophy skin flaps are mobilized (Fig. 7b).

The corporeal bodies are further freed from the pubic bone by incising the suspensory ligaments (Fig. 7c). Again, complete and radical transection of the urogenital fibres is a critical step to allow placement of the posterior vesico-urethral unit deep into the pelvis (Fig. 7d).

Dissection is carried out cephalad and medial behind the prostate and lower aspect of the bladder (Fig. 7e). The para-exstrophy skin flaps are sutured to the lateral aspect of the prostatic urethra (5–0 polydioxanone). They extend to the inferior border of the bladder to effectively widen the plate used for tubularization (Fig. 7f).





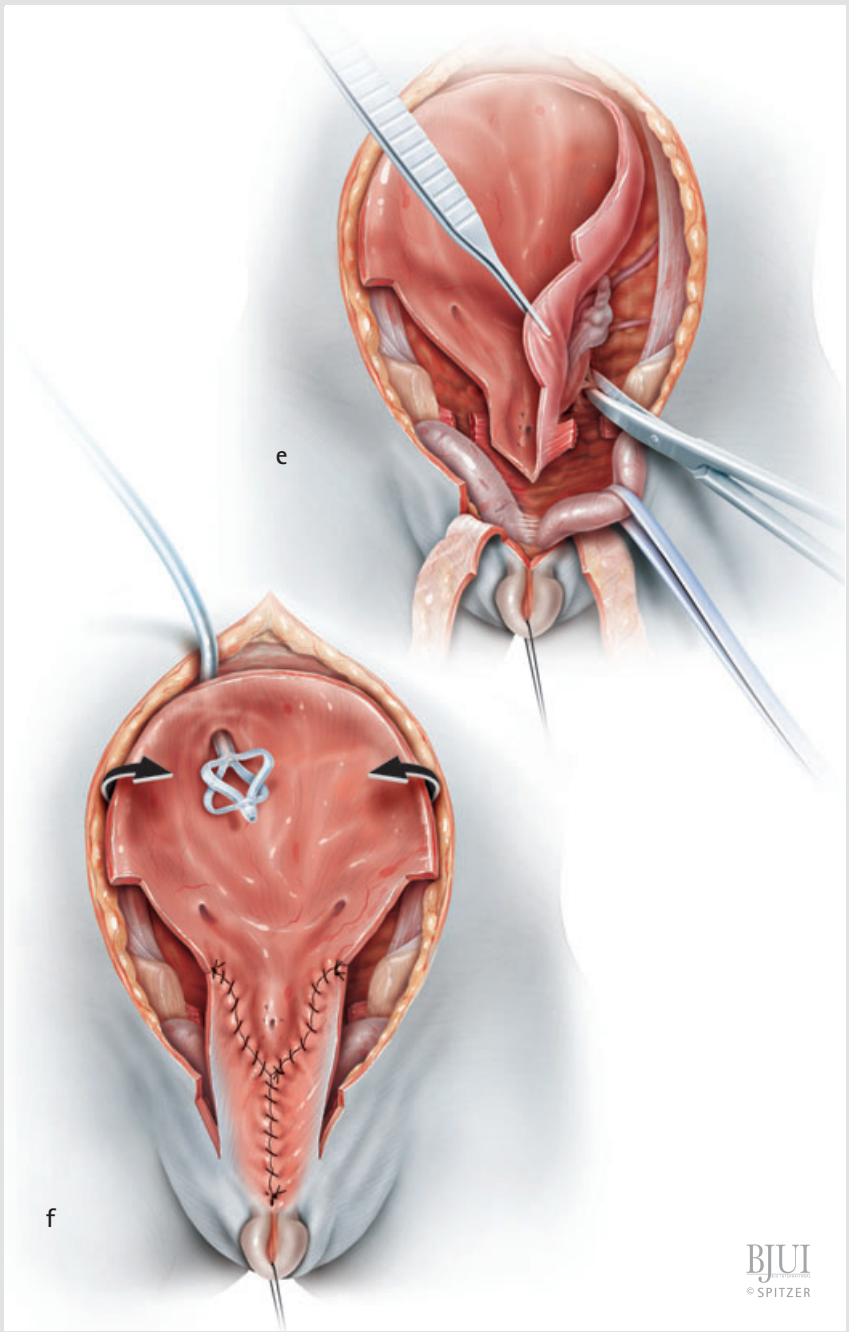


Figure 8

The mucosa and muscle of the bladder are closed in single layer with interrupted 3–0 polydioxanone figure-of-eight sutures. Starting at the vesico-urethral junction, the urethra is then closed well onto the penis in the midline anteriorly with full thickness, interrupted 5–0 polydioxanone. The urethra should accommodate a 10–12 F sound comfortably. The size of the opening should allow enough resistance to aid in bladder adaptation and to prevent prolapse, but not enough outlet resistance to cause upper tract changes. The posterior urethra and bladder neck are buttressed with a second layer of local tissue if possible. The urethra is not stented, to avoid necrosis with accumulation of secretions in the neourethra.

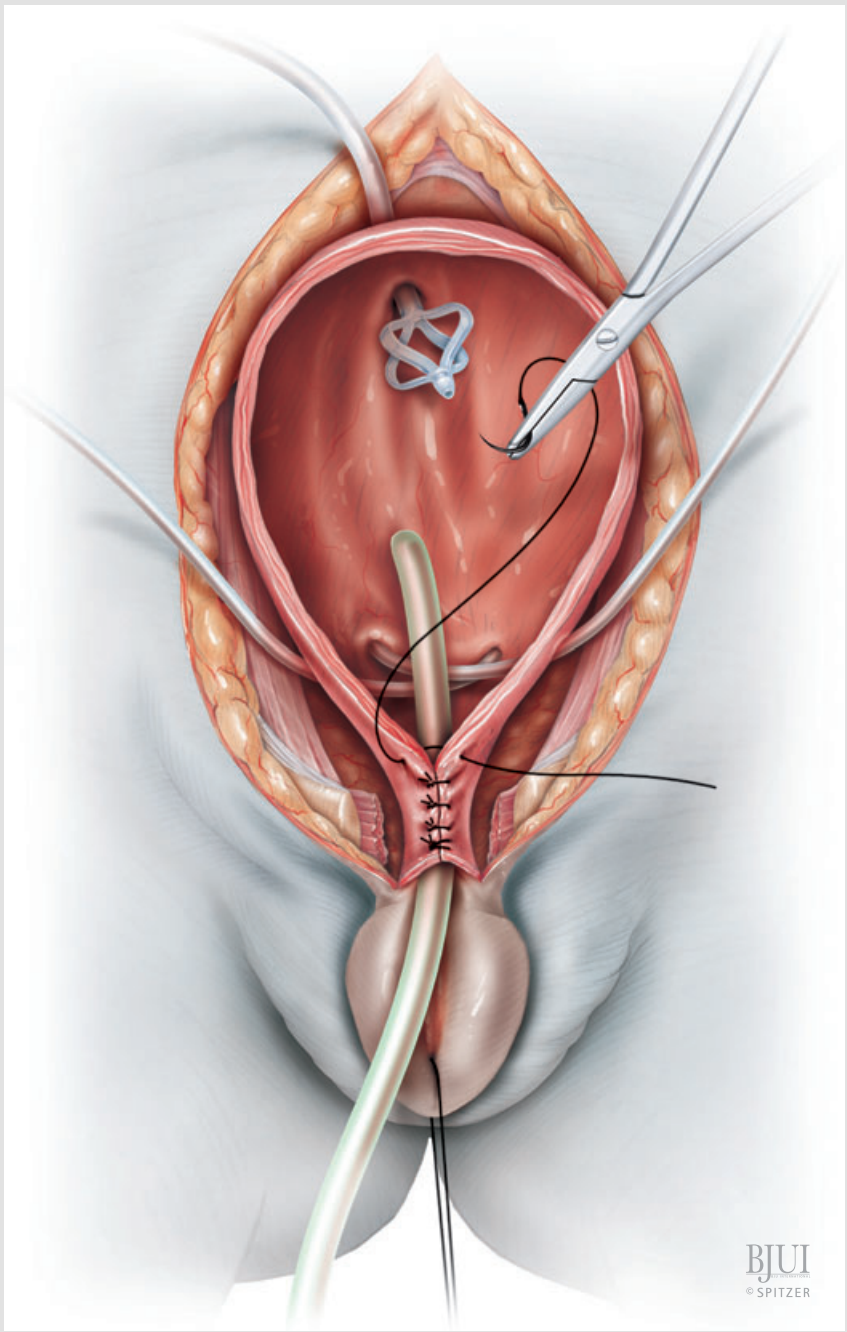
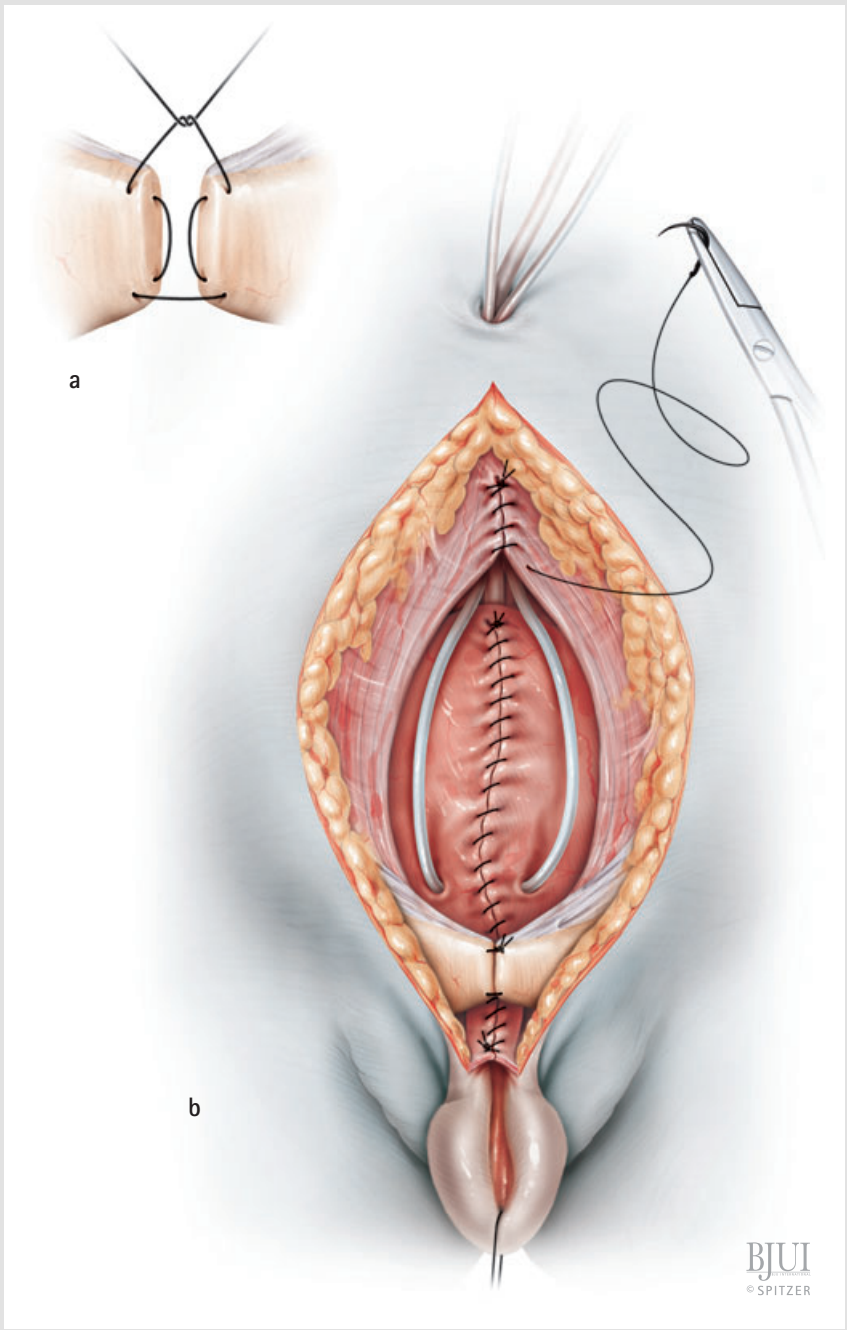


Figure 9

When the bladder and urethra have been closed and the drainage tubes placed, pressure over the greater trochanters bilaterally allows the pubic bones to be approximated in the midline. Horizontal mattress sutures are placed in the pubis and tied with a knot away from the neourethra (Fig. 9a). Often, we are able to use another stitch of no. 2 nylon at the most caudal insertion of the rectus fascia onto the pubic bone. This manoeuvre adds to the security of the pubic closure. The neourethra can then be matured to the surrounding lower abdominal and penile skin with interrupted 5–0 polyglactin. A V-shaped flap of abdominal skin at a point corresponding to the normal position of the umbilicus is tacked down to the abdominal fascia with 5–0 polyglactin, and the drainage tubes exit this orifice (Fig. 9b).



POSTOPERATIVE CARE

The modern staged reconstruction of bladder exstrophy converts a patient with exstrophy into one with incontinent proximal shaft epispadias. In males, the second stage epispadias repair is at 0.5–1 year old, with bladder neck reconstruction and ureteric reimplantation at age 4–5 years, when the child has achieved an adequate bladder capacity (>100 mL) and is motivated to participate in a postoperative voiding programme.

Ensure all catheters are well secured and a sterile surgical dressing of choice is used.

Adequate postoperative pain control is essential in maintaining pelvic immobilization. Epidural analgesia is maintained with a continuous infusion of 1 mg/mL lidocaine at 0.8 mg/kg/h. Epidural infusion rates are proactively adjusted to maintain serum lidocaine levels at 5 mg/L. It is desirable to maintain epidural catheters for ≥3 weeks. If analgesia remains inadequate, catheters are removed and i.v. or oral opioids are administered. In addition to epidural analgesia, infants receive enteral acetaminophen 10–15 mg/kg every 4–6 h around the clock and i.v. or enteral diazepam 0.1 mg/kg, every 4–6 h, as needed. Diazepam also assists with controlling bladder spasms. All children who receive benzodiazepines and/or opioids for >2 weeks are considered drug-dependent and are, at the conclusion of their hospital stay, weaned slowly (10–20% of their total daily dose per day) to prevent symptoms of withdrawal.

All children are given oral oxybutynin (0.1–0.2 mg/kg) three times daily until all catheters are removed. Also, appropriate continuous prophylactic antibiotics should be continued, as all children with bladder exstrophy have VUR.

PATIENT INSTRUCTION (ACTIVATION/ IMMOBILIZATION)

All patients who receive pelvic osteotomy are immobilized after surgery, using modified Buck's traction and external fixation. The patient is placed in minimal skin traction in a nearly horizontal position for ≈4 weeks for infants and 6–8 weeks for older children, depending on patient age and amount of healing. Pelvic radiographs are taken to ensure adequate bony healing and callus formation before the external fixator is removed and the child allowed to bear weight and mobilize. If the pubic diastasis is not completely corrected initially, it can be gradually corrected at the bedside in succeeding days by gradual approximation of the inferior fixator bars. These fixation devices are removed at the bedside or clinic using a mild sedative at 6 weeks after surgery in children who are ≥2 years old. Children aged <2 years might have the devices removed as early as 4 weeks after surgery in a similar manner.

Newborns undergoing closure without osteotomy in the first 48–72 h of life are immobilized in modified Bryant's traction while supine, where the hips have 90° of flexion. When modified Bryant's traction is used, it is maintained for 4 weeks. Spica casts and 'mummy wraps' are all inadequate for proper immobilization and are associated with an increased failure rate.

CATHETER/STENT HANDLING

The bladder is drained by a suprapubic non-latex Malecot catheter for 4 weeks. Before removing the suprapubic catheter the bladder outlet is calibrated with a urethral catheter or sound, to ensure free drainage. Also, residual urine is estimated by clamping the suprapubic tube, and specimens for culture are obtained before the patient leaves the hospital and before tube removal. Should bladder outlet resistance be such that urine is retained within the bladder, then reflux and ureteric dilation could develop, with a subsequent risk of urinary infection. In these cases, it might be necessary to dilate/incise the urethra or begin intermittent catheterization. Ureteric stents provide drainage during the first 10–14 days after closure, as swelling caused by the pressure of closure of a small bladder can obstruct the ureters and give rise to obstruction and transient hypotension. If there are no problems with the stents during healing, we leave them in for up to 3–4 weeks. A complete ultrasonography examination is obtained to ascertain the status of the renal pelves and ureters after catheter removal.

ADJUVANT THERAPY

While there is no specific adjuvant therapy in neonatal exstrophy closure, all older children undergo a structured voiding-improvement course before bladder neck reconstruction, to achieve appropriate postoperative voiding dynamics. They are taught to relax the pelvic floor and use the pelvic floor muscles properly. This is accomplished by a team of senior nursing staff using one-to-one coaching and modern biofeedback equipment.

FROM SURGEON TO SURGEON

The difficult case: Small bladders that are of questionable size for closure are a special challenge. For example, a small, fibrotic bladder patch that is stretched between the edges of the small triangular fascial defect without elasticity or contractility cannot be selected for the usual closure procedure. Examination with the patient under anaesthesia might sometimes be required to assess the bladder adequately, particularly if considerable oedema, excoriation, and polyp formation have developed between birth and the time of assessment. Decisions on the suitability of bladder closure, or the need for waiting (delayed primary closure) should be made only by surgeons with extensive experience of the exstrophy condition. Some conditions preclude primary closure, including penoscrotal duplication, ectopic bowel within the extruded bladder (a relative contraindication), a hypoplastic bladder, and significant bilateral hydronephrosis. Ideally, waiting for the bladder template to grow for 4–6 months in the child with a small bladder is not as risky as submitting a small bladder template to closure in an inappropriate setting, resulting in dehiscence and allowing the fate of the bladder to be sealed at that point. If the bladder does not grow to sufficient size after 4–6 months, other options include excision of the bladder and a nonrefluxing colon conduit or ureterosigmoidostomy. Another alternative involves urinary diversion with a colon conduit and placing the small bladder inside, to be used later for the posterior urethra in an Arap-type procedure. Lastly, if the bladder is small and the presentation is for late primary closure, bladder augmentation, ureteric reimplantation, and an outlet procedure, in addition to a continent urinary stoma, can be considered.

THINGS TO MAKE LIFE EASIER

We have found that having a paediatric orthopaedic surgeon with an interest in pelvic osteotomy and the orthopaedic care of children with bladder exstrophy is essential to

successful closure. Along those lines, the tunnelled epidural catheter, as described previously, has proved crucial in ensuring long-term postoperative pain control and immobilization. Performing osteotomy on a child without a clear plan for immobilization is doomed to failure.

Also, in an effort to decrease the incidence of vesicocutaneous fistulae and intrapubic suture erosion after surgery, we have recently been placing a ≈2 × 4-cm patch of human acellular dermis (HAD, Alloderm, LifeCell Corporation, Branchburg, NJ, USA) between the posterior urethral/bladder neck and the pubic closure to avoid these potential complications. The HAD is secured to the pelvic floor and bladder with interrupted 4–0 polyglactin before pubic bone apposition. The pubic bone is then closed over the HAD adjunct and urethral/bladder neck closures. The long-term results of this technique are pending.

The single most important aspect when encountering intraoperative problems is having previous experience with closure of a wide variety of exstrophy variants.

Correspondence: Timothy M. Phillips, Paediatric Urology, Brady Urological Institute, Johns Hopkins Hospital, 600 N. Wolfe Street, Baltimore, MD 21287, USA.
e-mail: tphill24@jhmi.edu