Bladder Extrophy and Epispadias

Treatment of bladder extrophy and epispadias

CHAPTER 6

Tom P.V.M. de Jong

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Abstract

Bladder extrophy and epispadias remain a challenging problem in pediatric urology. There are many therapeutic options ranging from different types of primary closure to the primary choice for diversion in extrophy patients. Further, there are several options for the treatment of male epispadias. In bladder extrophy, primary closure with meticulous reconstruction of the pelvic floor, combined with early clean intermittent catheterisation, seems to be the best first choice. In cases of female epispadias, a new technique will be described which provides a good chance to develop continence and voiding after one operation without the need for bladder neck reconstruction. The chapter tries to give a review of the contemporary treatment of extrophy and epispadias including some details of operative procedures.

Introduction

The exstrophy/epispadias complex represents, in most cases, dramatic congenital anomalies with lifelong consequences for both the child and the parents. It is rarely in mild epispadias only a cosmetic problem. The syndrome ranges from cloacal extrophy with involvement of both the urinary tract and the bowel, through classic bladder extrophy to epispadias.

Anomalies in the exstrophy/epispadias complex are rare. The prevalence of bladder extrophy is approximately 1 in 30,000 live births. The prevalence of epispadias in boys is approximately 1 in 40,000, in girls 1 in 200,000. The incidence of classic bladder extrophy in the Western world appears to be declining, possibly due to intrauterine detection with subsequent abortion of the fetus although specific numbers of prenatally detected cases cannot be found in the literature.

Many recent publications describe the results of surgical repair of extrophy and epispadias without giving details of treatment techniques - in this chapter a more practical approach will be adopted.
Anatomy of Bladder Exstrophy and Epispadias

In classic bladder exstrophy the bladder (fig. 1) and the urethra are open structures lying on the anterior abdominal wall. The amount of the bladder that is visible can range from a large bulging bladder plate to a hardly visible small patch. In males, the colliculus protrudes at the beginning of the urethral plate. The urethral plate runs to the glans penis. The glans is flattened and is connected to diverging corpora cavernosa fixed to the always separated halves of the pubic bones. In females, the urethral plate runs between the separate halves of the clitoris to the anteriorly positioned hymenal ring that is present in nearly all cases. The umbilical cord is implanted at the cranial margin of the bladder, thus caudal to the normal position.

Figure 1: Example of a new-born boy with bladder exstrophy. The child has a relatively large bladder plate, a good candidate for successful development of the bladder after primary closure. Notice the umbilical cord that has a typical, low implant adjacent to the bladder plate. The phallus is broad with a flattened glans. The arrow points at the left ureteric orifice.
In bladder extrophy the pubic bones are always widely separated, ranging from 2 to 8 cm. The anterior rim of the pelvic floor can be palpated between the pubic bones (fig. 2).

In male epispadias the urethral orifice is situated flush with the abdominal wall at the level of the symphysis or more distally at the anterior side of the penis, thus classified as glanular, penile and subsymphyseal. It can come together with symphyseal diastasis. In proximal epispadias, with or without symphyseal diastasis, incontinence based on congenital sphincteric insufficiency is common.

In female epispadias presents, there is no urethra, the bladder neck opens at the level of the hymenal ring. The hymen is intact in most cases. These girls are often misdiagnosed as intersex cases or can present at later age with continuous dribbling incontinence.
Finally, a wide range of extrophy variants exists. This can be a superior bladder fissure, an open bladder top with normal distal anatomy, a duplicate extrophy with a normal bladder and urethra in the abdomen, a bladder plate and a dorsal chordee at the surface of the abdominal wall, a covered extrophy with symphyseal diastasis in more or less normal anatomy of the lower urinary tract.\textsuperscript{1-7} All these cases need individually tailored therapy.

**Associated Anomalies and Diagnostics**

In classic bladder extrophy, in general, few associated anomalies occur. There can be vertebral anomalies in approximately 7\% and hip dislocations, rarely there are associated upper tract anomalies. The uterus can be partially or completely duplicated. Syndromes that include bladder extrophy/epispadias are not known, although inheritance of bladder extrophy is apparent in the relatively few cases that have proven fertility. Most patients have an anteriorly positioned anus. Inguinal hernias are common, in both girls and boys with bladder extrophy. In epispidias, the majority of children do not have other associated anomalies, apart from the diastasis of the pubic bones. The diastasis of the pubic bones, when untreated, seems to have little effect on the children other than external rotation of the hips with, sometimes, a waddling gait in patients with a wide separation of the pubic bones.\textsuperscript{8-11}

In cloacal extrophy many associated anomalies can occur, obviating the need of redundant diagnostics prior to the decision to start reconstructive operative treatment of the patient. Cloacal extrophy comes with spinal dysraphism, upper tract anomalies, cardiac anomalies and omphalocele in a high percentage.\textsuperscript{12-17}

Thus, in classic extrophy and epispidias, ultrasound of the urinary tract and a plain X-ray of the spine will provide sufficient information in most cases. In cloacal extrophy, additional investigations, including karyotyping and cardiac ultrasound are necessary.
Embryology of the Exstrophy/Epispadias Complex

Several explanations for the embryogenesis of exstrophy have been proposed. The bladder is separated from the gastrointestinal tract by the 6th week of gestational age. The ureters and part of the trigone, bladder neck and prostate (males) or urethra (females) are of wolffian origin. The most accepted theory for the development of bladder exstrophy is that the persistence of a large cloacal membrane prohibits development of the mesoderm leading to abdominal wall closure and phallic formation as described by Marshall ans Muecke. Several alternative theories exist, ranging from the low origin of the umbilical cord as the principal anomaly to the suggestion that the first anomaly lies in a lack of rotation in the pelvic ring primordia, thus suggesting that the exstrophy is secondary to malformation of the pelvic bones.14,18-20

Timing of Treatment and Treatment Choice

Generally, for reconstructive surgery of the lower urinary tract it is preferable to finish treatment before the age of 18 months. Psychologically, surgery of the genitalia before the onset of mental coupling of gender and genitalia is a great advantage. Nursing complications such as the unwillingness to void after removal of catheters can be avoided by operation before the age of 18 months.

In male epispadias, treatment of the phallus can be scheduled similarly as in hypospadias. Penile growth in the first year is relatively important. When the stretched penile length is insufficient, testosterone treatment by intramuscular injection or by ointment can be a great help. Operation will take place around the age of 10-12 months. When necessary, bladder neck reconstruction takes place at the age of 4-5 years.

In female epispadias, phallic size is unimportant, thus timing of the correction depends on the preference of the pediatric urologist. A small bladder capacity can be a relative contraindication to early surgery, a tendency of the bladder to prolapse can force early surgery.
Classic bladder exstrophy is considered to be a newborn emergency by most pediatric urologists, although groups exist that choose for a primarily delayed closure after weeks or months. Several factors support the advantage of bladder closure within the first 48-72h of life. In the first days after birth the symphysial diastasis can be closed primarily, without osteotomies, due to the circulating maternal hormones that maintain a flexible pelvic ring. Moreover, bacterial ingrowth of the bladder increases by time and tissue quality of the bladder changes rapidly because of edema and dryness. After primary bladder closure, in general, correction of the epispadias takes place at age 12-14 months, followed by bladder neck reconstruction at the age of 4-5 years. When necessary, bladder neck reconstruction is combined with bladder augmentation by ileocystoplasty. A catheterizable, continent stoma of the bladder is also possible. Both for epispadias and for exstrophy patients, nowadays, we try to achieve continence and satisfactory voiding in the majority of cases at primary closure, followed by temporary or permanent clean intermittent catheterization (CIC) if necessary.

The optimal treatment of classic bladder exstrophy, in Europe, was agreed on at the consensus meeting of the European Society for Pediatric Urology (ESPU) in 1992.

The following recommendations were made: (1) Early closure without osteotomies or delayed closure at 7-10 days with anterior (Salter type), osteotomies. (2) Epispadias repair at 12 months, when necessary after testosterone treatment. (3) Bladder neck reconstruction at the age of 4-5 years. Bladder capacity < 100 ml with ileocystoplasty, > 100 ml without cystoplasty. (4) When it is apparent that CIC is necessary, especially in girls, a continent catheterizable stoma of the bladder should be constructed. For those who do not follow these recommendations there are a number of options. Some opt for delayed closure with osteotomies after several months, some opt for primary deviation resulting in cystectomy with construction of a catheterizable continent urinary pouch constructed out of bowel.21-24 The disadvantages of these approaches are many. Long-term safety of bowel reservoirs constructed in early life has yet to be proven. For patients in countries with restricted possibilities for health care a rectal bladder with
sigmoid pull-through through the anal sphincter can provide a safe solu-
tion to obtain urinary continence without endangering the upper tracts
[personal communication with Prof. Dr. A. Pesamosca, Bucharest,
Romania, with 30 years of follow-up].

The Surgical Treatment of Bladder Exstrophy

Technical Aspects of Primary Closure

In bladder exstrophy, treatment starts immediately after birth. Irritation to
the bladder must be avoided, so plastic umbilical cord clamps are removed
or fixed cranial to the bladder. We treat the bladder after admission to the
hospital with an evaporator, normally used for upper airway problems,
located at a few centimeters from the bladder. Operation is preferably
schedued within the first 48 h after birth to avoid the need of primary
osteotomies and to preserve optimal quality of the bladder mucosa.

Because of the risk of developing latex allergy after multiple interven-
tions, we treat our patients preferably in a latex-free environment with
latex-free surgical gloves and catheters.

Surgery takes place under general anesthesia, preferably with a caudal
epidural catheter. We give amoxicillin/clavulic acid and gentamycin as
prophylactic antibiotic treatment. The operation starts with identification
of the ureteral orifices and the colliculus (in males). The umbilical cord is
fixed at the cranial margin of the bladder plate. The umbilicus is freed
from adjacent tissue, the umbilical arteries and vein are identified and
freed. Cosmetically we prefer to ‘transplant’ the umbilicus to the normal
position. This is done by excision of a segment of skin, approximately
1.5 cm in diameter, at the normal site of the umbilicus through the rectus
fascia into the preperitoneal space. After tunneling the plane between the
freed umbilical cord and the newly prepared site the umbilical remnant is
pulled into the new location. It is fixed to the skin with absorbable
sutures. Cosmetically this procedure is better than a man-made umbilicus
at the exit-site of a suprapubic catheter.
Next, the bladder is freed from the abdominal wall. It can be done in continuity with the urethral plate or disconnected with paraexstrophy flaps as proposed by Jeffs and Duckett, although many authors consider this procedure old fashioned. The advantage of leaving the urethral plate fixed at the bladder is a much easier primary procedure, the disadvantage is that certainly a secondary procedure to achieve continence will be needed. The theoretical possibility that the paraexstrophy flaps may prohibit easy intermittent catheterization has not been encountered in our patients, provided that they start CIC early after primary closure.

One advantage of the use of paraexstrophy flaps is the lengthening of the urethral plate, thus giving enough length to move the bladder neck into an intra-abdominal position, preferably with the proximal urethra cranial to the pelvic floor (fig. 3). The second advantage is that it provides the
opportunity to free the anterior pelvic floor from the rami of the pubic bones and to close the muscle around the proximal urethra. Thus, in many cases, continence can be achieved after the primary closure. In the past, this procedure was criticized because of upper tract deterioration caused by urethral obstruction. By starting CIC shortly after removal of the urethral catheter this problem can be avoided. After several weeks or months most patients will start to void spontaneously.

Freeing the bladder from the abdominal wall is a meticulous procedure. The bladder plate is circumcised into the preperitoneal fat providing optimal mobility. Care must be taken to leave the vascular pedicles intact. At the level of the prostate, bleeding from spongiosal tissue can be difficult to cope with. By cutting straight up to the level of the pelvic floor bleeding can be stopped by suturing the margins of the spongy tissue. In girls, there is a serious risk of producing a urethrovaginal fistula, the plane between the bladder and the vagina can be difficult to find as both organs are very thin-walled.

Paraexstrophy flaps are continuous with the urethral plate. After mobilizing the bladder upwards, they can be brought together in the midline, thus lengthening the urethral plate.

Bladder closure is done with interrupted 5.0 polyglycolic acid sutures, paraexstrophy flaps and bladder neck are modelled into a urethra with 6.0 sutures around an 8- or 10- Fr silicone catheter. The catheter is fixed to the bladder with a rapidly reabsorbable suture. When the bladder plate is large, ureteral splinting is not strictly needed. We perform a ureteral advancement into the midline, as described by Gil-Vernet, as a primary antireflux procedure at first closure of the exstrophy bladder. In our hands, this simple procedure prevents vesicoureteral reflux in approximately 60% of the cases. With a very small bladder plate, ureteral splinting with 4-Fr feeding tubes is carried out for 10-14 days.

The medial surface of the pubic bones is freed from muscular and fibrous tissue from the anterior pelvic floor. Distally, the pelvic floor has strong attachments to the corpora cavernosa. Care must be taken to identify and free the corpora cavernosa, thus avoiding penile necrosis when closing the pelvic ring. When the symphysial gap is wide, the pelvic floor remnants cannot be closed around the neo-urethra before partial approximation of the pelvic ring.
Pelvic ring closure can be done with a No. 1 polyglycolic acid mattress suture through the pubic bones. The suture is passed through the rami of the pubic bones and tightened slightly to give the opportunity to close the pelvic floor, immediately afterwards the bones are completely approximated. When separation of the pubis is wide, external pressure at the hips is needed to bring the pubic bones into close contact. The abdominal wall is closed in layers. By closing the pelvic ring the rectus fascia can be easily closed in the midline, combined with a simple midline skin closure. Skin flaps are not needed in this procedure. Whether epispadias closure is performed at the primary closure depends on phallic size and preference of the pediatric urologist. Strict contraindications for primary epispadias closure do not exist. Operating time in the neonate can be a relative contraindication. Postoperatively, the patient is treated by Bryant’s traction for the pelvic ring closure: both legs are separately fixed upwards with a weight that leaves the buttocks just free of the bed. It offers the child relatively free movement of the legs, while securing the pelvic ring. The child can be taken out of this traction for breast feeding and cuddling in a plastic shell that fixes the legs at a right angle. In the past, we have tried external fixation with some success. Bone quality in the neonate prohibits secure fixation for a longer period of time.

In general, children are taken care of in a neonatal intensive care unit for 24 h. Broad-spectrum antibiotic prophylaxis is given for 24 h. Trimethoprim 2 mg/kg/day, once a day, is started on the 2nd day and continued for at least the first year of life. Ureteral stents are left for 5-15 days, depending on the size of the bladder plate. A transurethral catheter is left in situ for 3 weeks, after removal of the catheter CIC is started 5 times/day. All patients are kept on oxybutinin 0.5 mg/kg/day at 6-hour intervals for the first 6 weeks. The oxybutinin treatment obviates the need of extra care for bowel emptying. Sometimes, enema therapy is needed. During the first year, ultrasound examinations of the upper tract are carried out every 2 months after discharge from the hospital. Urinary cultures are performed every month.
Orthopedic Surgery in Exstrophy Patients

In delayed closure of the exstrophy bladder, osteotomies are mandatory to obtain closure of the pelvic ring. For continence, successful closure of the pelvic ring appears to be beneficial. Moreover, it prevents gait problems at a later age. Many techniques have been described. In our hands, Salter-type osteotomies have resulted in temporary femoral nerve palsy in the majority of cases. Still, this technique provides optimal reconstruction of the bony defect.\textsuperscript{8,10,11,19,28,29}

Surgical Treatment of Male Epispadias

Epispadias repair is subdivided into repair of primary, congenital epispadias or secondary epispadias after primary exstrophy closure. Preferably, surgery is timed around the age of 12 months, with or without treatment for phallic size with testosterone as a local ointment (2.5% testosterone in cream, twice daily for 3 weeks, to stop 3 weeks before operation, not to be used by the mother with bare hands) or systemic (2 injections of 25 mg of an androgen mix i.m, 60 and 30 days before operation). Many variations on dosage schedule exist. Preferably, before operation, information should be available on the quality of the external sphincter. This information can be obtained by observation of the infant when crying, does he lose urine with high abdominal pressure or not? Urodynamic study assisted by fluoroscopy can give an indication of the quality of the sphincter. Starting the procedure with a cystoscopy can also give additional information in most cases, although one should bear in mind that a caudal anesthetic block opens the sphincteric mechanism in boys under anesthesia.

The operation starts by placing a traction suture through the glans and opening of the skin alongside the urethral plate and around the urethral orifice. After degloving of the penis, the urethral plate is freed completely from the corpora and incised up to the top of the glans and finally rolled into a urethra with absorbable sutures. The tip of the glans is incised in the midline over a distance of a few millimeters and closed squarely to provide a better shape to the meatus (inverted MAGPI procedure, called
Cranial curvature of the penis is partly cured by excision of chordee-like tissue and finally by rotation of the corpora and fixation of the rotated corpora with or without opening and connecting the corporeal bodies in the midline. The corpora are connected anteriorly to the neourethra. Curvature is assessed by inducing an artificial erection using two needles and two syringes, one for each corpus because the connecting vessels existing in a normal penis are absent in epispadias. Finally, the glans is closed over the neourethra in two layers and the penile skin is repositioned.

When, in primary epispadias, sphincteric activity is thought to be ineffective, we have adopted a policy of trying to find the anterior side of the pelvic floor, free it from the pubic bone and wrap it around the urethra or taper it anteriorly to the urethra. This can be done even with a closed pubic symphysis. Success has been variable and the numbers are too small to draw conclusions.

When incontinence is apparent at a later age, physical therapy can be tried in mild cases, followed by injectional therapy with collagen, silicon particles, Teflon or other materials. Severe incontinence will end with formal bladder neck reconstruction, with or without clean intermittent catheterization and bladder augmentation.

The Surgical Treatment of Female Epispadias

Female epispadias is a rare entity, series in the literature of more than a few cases do not exist. Straightforward operation techniques other than remodelling a urethra, combined with bladder neck reconstruction, have not been published. We have devised a technique that gives hopeful results with 3 out of 4 patients continent, 3 out of 4 voiding and 1 on CIC. One patient is too young to assess continence. The operation starts with an incision alongside the urethral plate that runs between the halves of the clitoris, the incision is lengthened around the urethral orifice. The urethral plate is completely mobilized and tubularized. The bladder neck and trigone are freed completely from the vaginal wall and pelvic floor, permitting mobilization of the bladder outlet and the urethra into the
abdominal cavity. Long intravenous infusion canulas are passed from the inguinal region along the inner side of the pubic symphysis into the wound, two on each side. Nonabsorbable sutures are placed at the bladder neck and passed upwards through the canulas. When the sutures are tightened and knotted at the external abdominis fascia, bladder neck and urethra slide into a physiologic position. The pelvic floor is identified and closed in the midline between bladder neck and vagina. Finally, the vulva is reconstructed by joining the clitoral halves and approximation of the labia majora anteriorly.\textsuperscript{33,34}

The Treatment of Cloacal Exstrophy

Cloacal exstrophy is a heavy burden for the children, their parents and doctors. The bladder consists of two halves joined in the midline by the remnants of the extrophied colon and the protruding ileum. Primary urologic therapy is not very much different from classic exstrophy although a multidisciplinary approach combined with general pediatric and orthopedic surgeons is mandatory in most cases. Problems arise with related congenital anomalies that can be impressive. One should consider very thoroughly all the ins and outs for the baby and parents before starting therapy. The list of operations and complications is a very long one. Therefore, in our hospital, we think it is justified to advise the parents not to have the baby treated resulting in early death. The few cases that we have treated, according to the parents’ wishes, have done relatively well although complications such as a short bowel syndrome, urinary tract infections, and, not to forget, major psychologic complications have been impressive.\textsuperscript{13,16} Worldwide, this policy of nontreatment is not possible in most countries, forcing the medical teams to try and offer optimal therapy. In general, lifelong medical guidance is mandatory for these children.
General Care of the Patients

The extrophy/epispadias complex patients are subject to a lot of medical technology resulting, in the best cases, in a relatively good situation of continency and voiding with the possibility of a relatively normal sexual life, in the worst cases with a continent catheterizable stoma and poorly functioning sexual organs. The hospitalization alone, even with a good end result, can produce psychologic distress. When surgery leads to a situation with a frustrated body image psychologic complications can be impressive, necessitating of thorough follow-up and treatment by a specialized team that takes the child through puberty into adult life.

CIC in these children produces significantly more aversion to the treatment then in spina bifida patients because of the normal sensibilty of the urethra.

Sexual life can be normal in female patients with normal fertility. However, prolapse of the uterus as a consequence of the absence of normal uterine suspension can occur very early, even in teenagers. Male patients mostly have a broad and relatively short penis with normal function and orgasm during intercourse. Ejaculation and fertility are hampered.

Psychosexual development in the exstrophy/epispadias patients is in need of professional support by psychologists, urologists and gynecologists with special knowledge in the field of gender identity and congenital malformations of the urogenital tract.11,35-49
Literature


