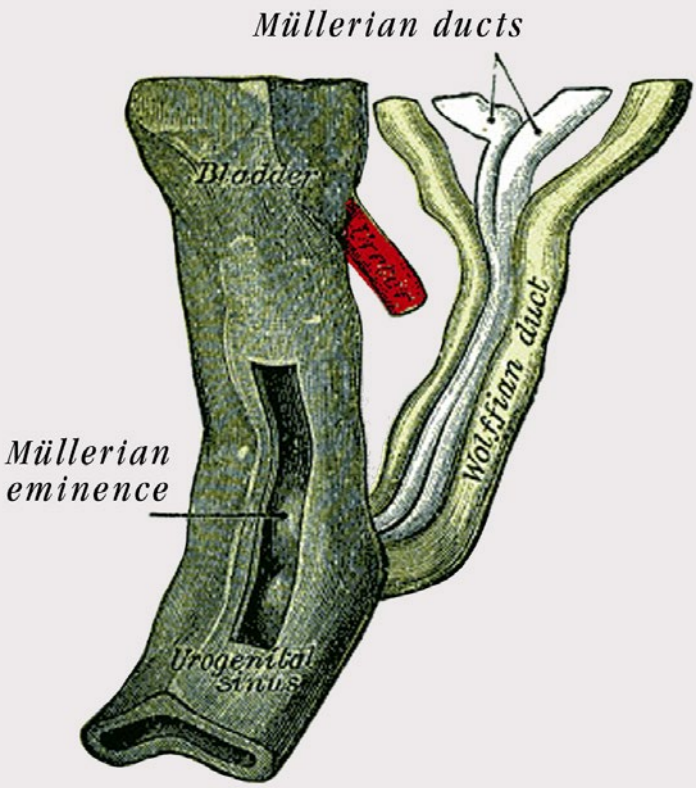
SWISS SOCIETY OF NEONATOLOGY

Exstrophy-epispadias complex in a female neonate



December 2008

Nava E, Hofmeister J, Haberlik A, Haecker FM,
Department of Neonatology (NE, HJ), Department of
Pediatric Surgery (HFM), University Children's
Hospital, Basel, Department of Pediatric Surgery (HA),
University of Graz, Medical School, Austria

© Swiss Society of Neonatology, Thomas M Berger, Webmaster

The exstrophy-epispadias complex is a rare congenital anomaly affecting about 1: 40'000 live births, with a male to female ratio of 2:1. It is thought to result from embryologic defects of unknown origin during the first trimester of pregnancy. Bladder exstrophy is caused by abnormal development of the lower abdominal wall and separation of the pubic bones (diastasis) leading to an everted bladder. Usually it is associated with an epispadius penis in affected males or a urethral opening located between a divided clitoris in female patients.

Surgical therapy may consist of a scheduled, staged repair procedure with 2 steps in female and 3 steps in male patients. The initial step includes primary closure of the bladder and abdominal wall, urethroplasty, reconstruction of the umbilicus and adaptation of the symphysis within the first 48-72 hours of life. In male patients, epispadia repair is recommended at the age of 6-8 months as a separate step. At the age of 2-3 years, bladder neck reconstruction, ureteral reimplantation and reconstruction of the external genitalia is performed in both male and female patients (1-4). As an alternative approach, some surgeons prefer to carry out a one stage procedure at the age of 6-8 weeks including all of the above mentioned measures and additional pelvic osteotomy if necessary (5, 6).

We report on a 3430 g full-term female neonate who presented with an exstrophy-epispadias complex which had been detected during prenatal routine ultrasound at 24 weeks of gestation. Prenatal chromosomal analysis revealed a normal 46, XX karyotype. The child was born by elective primary caesarean section. The Apgar scores were 6, 7 and 10 at 1, 5 and 10 minutes, respectively. On postnatal examination, the bladder was small with a diameter of 3 cm with an opened, flattened appearance and a thickened mucosa, covered with polyps. The vagina and the anus were anteriorly displaced. Additionally, a bifid clitoris, widely spread labia minora and an incomplete umbilicus were observed (Fig. 1). Abdominal, renal and spinal ultrasound revealed no pathological findings. The bladder was maintained in a moist condition by covering it with sterile NaCl 0.9% dressings and a plastic wrap.

The surgical repair was performed according to the scheduled, modern staged repair procedure. Surgery included primary closure of the bladder and the abdominal wall as well as urethroplasty on day 3 of life. Bilateral ureteral stents (size Ch 4) were inserted (Fig. 2) and left in place until the 8th postoperative day, but no ureteral reimplantation was undertaken. Adaptation of the symphysis was achieved using a PDS (polydioxane) suture (Fig. 3). Pelvic osteotomy was not necessary. The bladder was drained for 4 weeks with a suprapubic catheter. The transurethral catheter (size Ch 8) was removed at the end of the operation to enable transurethral voiding.

In order to stabilize the symphysis, the modified Bryant's traction procedure (permanent 90° flexion of hips, knees and ankles held together and slight elevation of buttocks off the bed) was applied for three weeks (Fig. 4). At the age of three months, both hips were mature (Graf Ib). A VCUG at the age of 4 weeks revealed bilateral VUR III°. At the age of 6 weeks, the patient was finally discharged.

Renal ultrasound examinations performed at 6 weeks, 3 months and 6 months of age were normal without pyelectasis. Urodynamic studies at the age of 9 months demonstrated a bladder capacity appropriate for age. Clinical examination revealed an acceptable cosmetic appearance of the abdominal wall, but not yet fully corrected external genitalia (Fig. 5).

The second step of the scheduled, modern staged repair which includes bladder neck reconstruction, bilateral ureteral reimplantation and repair of the external genitalia, is planned to be undertaken at the age of 2-3 years. Until then the following treatments and examinations will be performed: long-term antibiotic prophylaxis and regular urinalysis, renal ultrasound examinations every three months, repeated VCUG after 12 month, DMSA scan to detect renal scars after 6 to 12 months, urodynamics (cystomanometry) after 6-9 months.

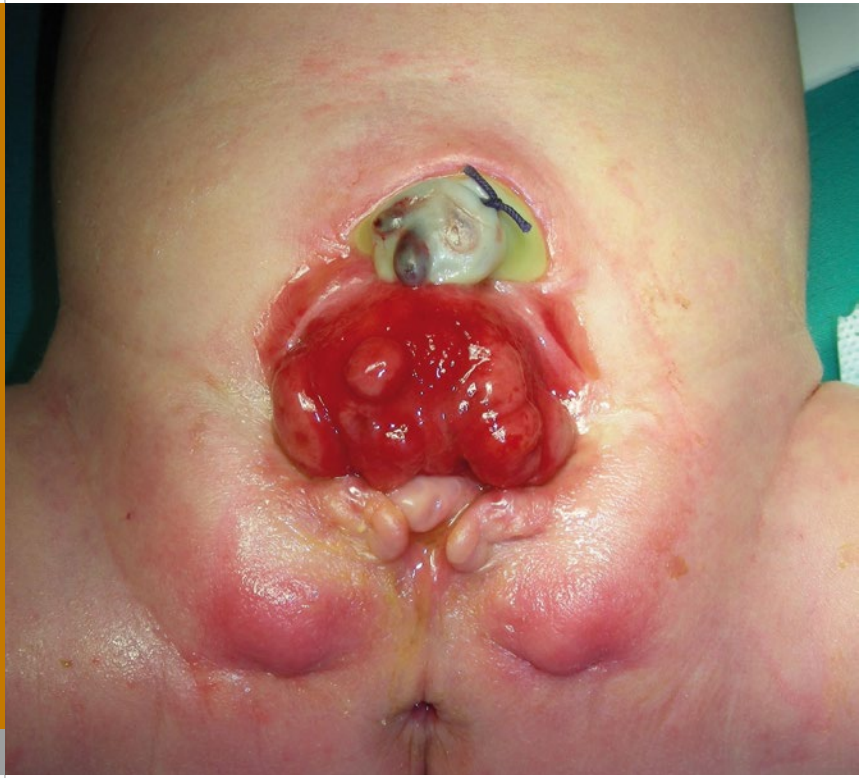


Fig. 1

Bladder exstrophy with anteriorly displaced vagina and anus, bifid clitoris and labia minora and incomplete umbilicus.

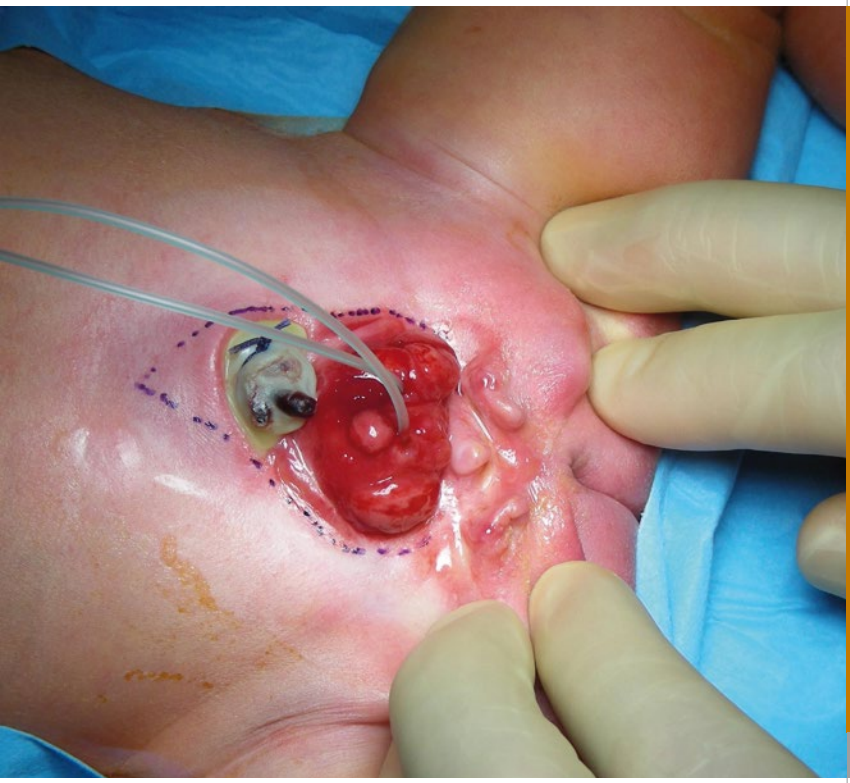


Fig. 2

Bilateral ureteral stents and incision markings.

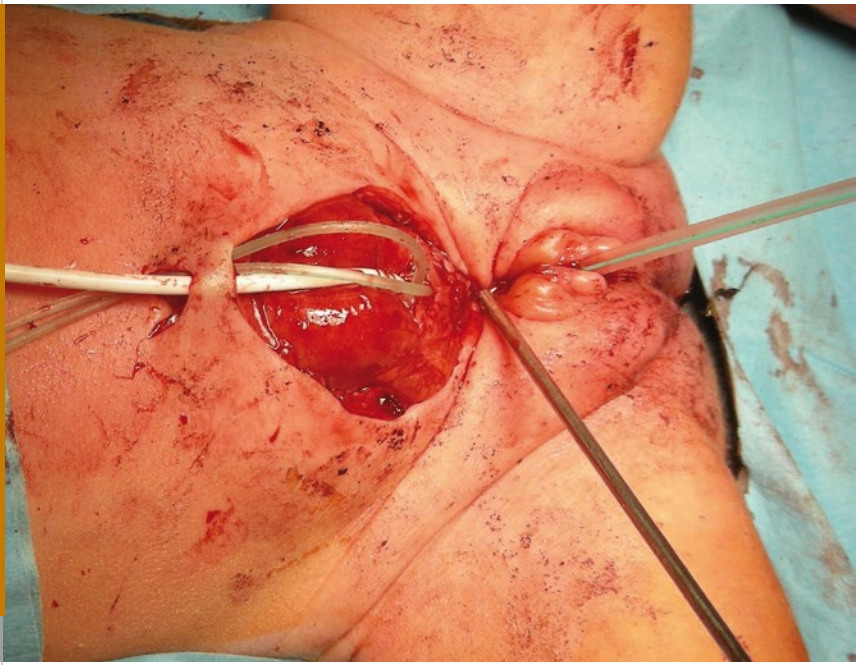


Fig. 3

Neoumbilicus, bilateral ureteral stents, suprapubic and transurethral catheters in place.

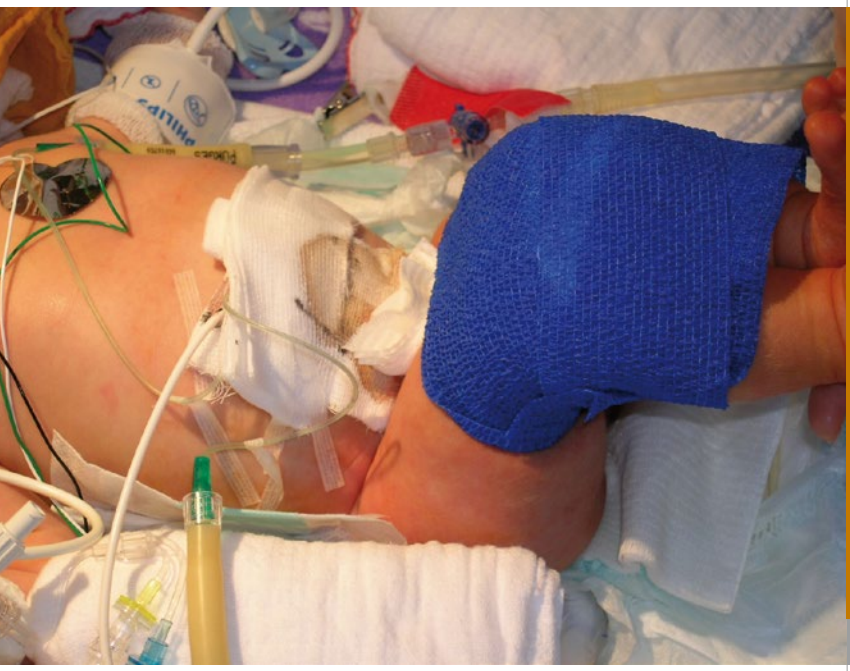


Fig. 4

Modified Bryant's traction to stabilize the symphysis.

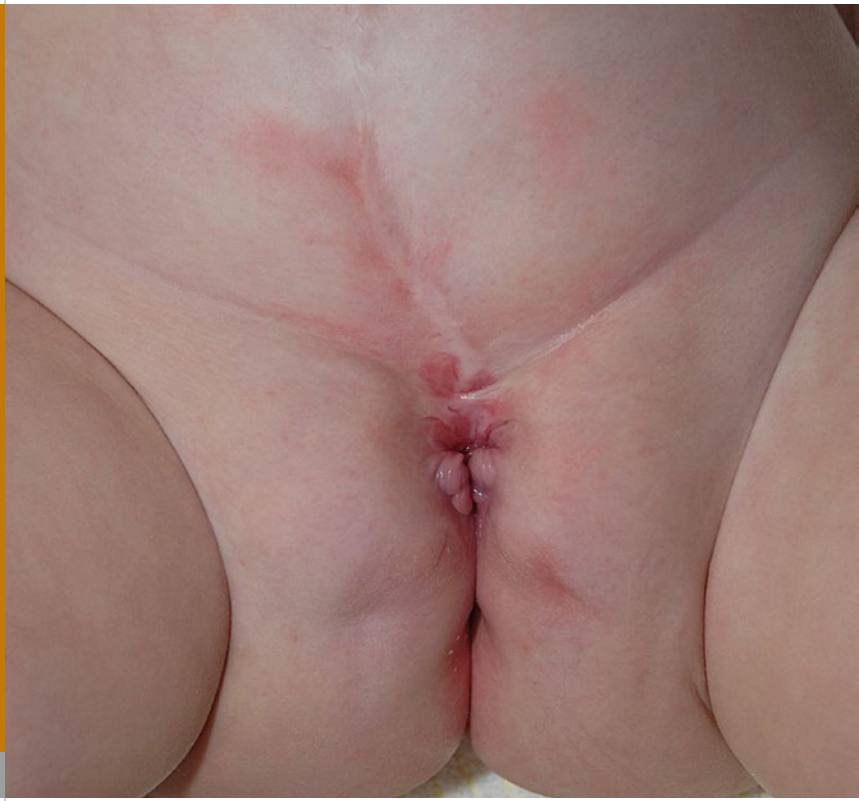


Fig. 5

Cosmetic appearance at 9 months of age.

Bladder exstrophy is a rare, but severe, life-altering congenital deformity of the genitourinary tract. The primary goals in the surgical management are closure of the bladder, preservation of renal function and reconstruction of the external genitalia. To reach a satisfactory quality of life, long-term interdisciplinary support is mandatory.

The outcome of exstrophy-epispadias complex after staged reconstruction is generally good, even though some patients may have long-term medical problems. Recurrent urinary tract infections are often caused by VUR or nephrolithiasis and require antibiotic or surgical treatment and can lead to kidney dysfunction (1, 7).

Urine incontinence is reported in about 30% of these patients (3). In these patients, further operations such as augmentation cystoplasty or placement of a catheterizable stoma are needed. A recent study from the John Hopkins Group reports that 70% of patients are continent day and night and are voiding per urethra, but only 10% achieve social continence (dry for more than 3 hours during the day) (7).

Reports about sexual function are controversial in the literature. Some authors report an almost normal sexual function (1) after genital reconstruction, but psychological assistance is recommended to overcome

anxiety about genital appearance and sexual performance (7-9). Female genitalia reconstruction shows acceptable cosmetic results in the majority of cases, whereas male dissatisfaction with penile length due to the anatomical shortness and broadness of the corpora clearly exists. Fertility in females is preserved, although there is an increased risk for uterine prolapse and incontinence after vaginal delivery. Male patients have reduced fertility, especially in the case of recurrent urinary tract infections (2, 6).

Finally, the risk of bladder cancer is elevated due to prolonged bladder exposure.

For related case reports, see **COTM 02/2006**: Exstrophy of the cloaca sequence and **COTM 03/2008**: OEIS Complex: developmental insights into a severe congenital abnormality of body wall development.

REFERENCES

1. Gearhart JP. Exstrophy, Epispadias, and Other Bladder Anomalies. In: Campbell's Urology, 8th ed. Edited by Walsh PC, Retik AB, Vaughan ED, Wein AJ. Philadelphia: WB Saunders Co, 2002. p. 2136-2196

2. Gearhart JP. The bladder exstrophy-epispadias-cloacal-exstrophy complex. In: Pediatric Urology. Edited by Gearhart JP, Rink RC, Mouriquand PDE. Philadelphia: WB Saunders Co, 2001. p. 511-546

3. Baird AD, Nelson CP, Gearhart JP. Modern staged repair of bladder exstrophy: A contemporary series. J Pediatr Urol 2007;3:311-315

4. Connor JP, Hensle TW, Lattimer JK, Burbige KA. Long-term follow-up of 207 patients with bladder-exstrophy: an evolution in treatment. J Urol 1989;142:793-796

5. Schrott KM. Komplette einaktige Aufbauplastik der Blasen-exstrophie. Plastisch-rekonstruktive Chirurgie in der Urologie 1999;430-438

6. Silver RI. Surgical management and postoperative care of children with bladder exstrophy. UpToDate® 2007;1-9

7. Chan DY, Jeffs RD, Gearhart JP. Determinants of continence in the bladder exstrophy population: predictors of success?. Urology 2001;57:774-777

8. Montagnino B , Czyzewski DI, Runyan RD, Berkman S, Roth DR, Gonzales ET Jr. Longterm adjustment issues in patients with exstrophy. J Urol 1998;160:1471-1474

9. Ebert A, Scheuering S, Schott G, Roesch WH. Psychosocial and psychosexual development in childhood and adolescence within the exstrophy-epispadias complex. J Urol 2005;174:1094-1098.

10. Reiner WG. Psychological and psychiatric aspects of genitourinary conditions. In: Pediatric Urology. Edited by Gearhart JP, Rink RC, Mouriquand PDE. Philadelphia: WB Saunders Co, 2001. p. 696-704

concept & design by mesch.ch

SUPPORTED BY

NYCOMED

CONTACT

Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch