

## Case Report

# MALE TERM NEONATE WITH BLADDER EXSTROPHY–EPISPADIAS COMPLEX

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### Abstract

Bladder exstrophy is a malformation of the bladder, in which the bladder and related structures are turned inside out. Bladder exstrophy affects one in approximately 50,000 livebirths. Male to female ratio is 2.3 : 1. Epispadias is commonly seen with exstrophy of the bladder. The cause and nature of the faulty development is not yet clear. The diagnosis is generally immediately apparent. Bladder exstrophy is a surgical correctable birth defect. In this case, bladder exstrophy–epispadias complex founded in male term neonate. On inspection was founded bladder everted through a midline lower abdominal wall defect, widening of the pubic symphysis and epispadias. Urology ultrasound revealed absent of the right kidney at right *fossa renalis* or in pelvic cavity, slight hydronephrosis of left kidney, no appearance of bladder in pelvic cavity. Patient underwent surgery during hospitalized in Sanglah Hospital - Denpasar. This patient was referred to the Congenital Anomaly Team in Dr. Soetomo Hospital - Surabaya because there was wound dehiscence during post operation care.[**MEDICINA 2009;40:60-4**].

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*Keywords: bladder exstrophy, epispadias, surgical correctable, dehiscence*

### Abstrak

*Bladder exstrophy* adalah suatu malformasi kandung kemih, dimana kandung kemih dan struktur yang terkait berada diluar tubuh. *Bladder exstrophy* dialami oleh 1 diantara 50.000 kelahiran hidup. Rasio laki-laki dibandingkan perempuan adalah 2,3 : 1. *Bladder exstrophy* sering disertai *Epispadias*. Kausa *bladder exstrophy* belum jelas. Biasanya diagnosis dapat segera ditegakkan. *Bladder exstrophy* merupakan suatu defek yang perlu dikoreksi dengan pembedahan. Pada kasus ini, *bladder exstrophy – epispadias complex* ditemukan pada seorang neonatus aterm, laki-laki. Pada inspeksi, ditemukan kandung kemih penderita keluar melalui defek dinding perut di garis tengah perut bagian bawah, symphysis pubis yang lebar, dan epispadias. Pada USG urologi, ginjal kanan tidak diketemukan, baik di fossa renalis kanan maupun kavum pelvis, hidronefrosis ringan ginjal kiri, dan tidak tampak adanya kandung kemih di kavum pelvis. Pasien dibedah di RS Sanglah – Denpasar. Namun karena penderita mengalami *wound dehiscence* saat perawatan post operasi, penderita dirujuk ke Team Anomali Kongenital RS dr. Soetomo – Surabaya. [**MEDICINA 2009;40:60-4**].

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*Kata kunci: bladder exstrophy, epispadias, pembedahan, dehiscence*

## INTRODUCTION

The word exstrophy is derived from the Greek word *ekstriphein*, which literally means to “turn inside out.” Bladder exstrophy is a malformation of the bladder, in which the bladder and related structures are turned inside out. The skin of the lower abdominal wall that normally covers the bladder also does not form properly and is separated, thus exposing the inside of the bladder to the external world.<sup>1,2</sup>

Epispadias is usually seen with exstrophy of the bladder. Epispadias occurred when the urethra has not formed completely, and it was be extremely short and split. Because of this, it open on the upper surface rather than on the end of the penis.<sup>2</sup>

Bladder exstrophy affects one in approximately 50,000 live births. Male to female ratio is 2.3 : 1. The risk of recurrence in a family is one in 100. Children born to parents with exstrophy have a one in 70 chance of having the defect.<sup>3</sup>

The condition of exstrophy occurs during the development of the embryo very early in the pregnancy, about 4 to 5 weeks after conception. The cause and nature of the faulty development is not exactly certain. It is 4 to 5 weeks after conception that the various organs and different types of muscles and tissues of the body begin to form from layers of cells that separate, divide and fold. One theory suggests that something goes wrong during this early folding and separation, causing the cloacal membrane fail to close, leaving the bladder outside of the abdominal wall. A second theory proposes that the layer of skin which forms over the bladder at this stage is thin and unable to hold in the bladder. It pulls apart, again leaving the bladder inside out.<sup>4</sup>

At birth, the diagnosis is generally immediately apparent. These are usually healthy babies with rare anomalies unrelated to the genitourinary and orthopedic defects. The exposed bladder is variable in size and generally has a soft, pliable membrane at birth. The kidneys are almost always normal. The umbilicus is low set and located just above the bladder. The bones of the pelvis (pubic rami) are widely separated. Boys will have a short penis with an upward bend (chordee). Oftentimes, the rectum is somewhat closer to the genitalia than usual.<sup>5</sup>

Bladder exstrophy is a surgical correctable birth defect. The complex surgical repair of the bladder exstrophy is performed with either the stage approach or the recently re-popularizes complete primary repair technique. The primary goals for reconstruction are: closure of the bladder and urethra; closure of the abdominal wall; preservation of the kidney and sexual function; improve appearance of genitalia; and urinary continence. Mortality with classic bladder exstrophy or epispadias is rare.<sup>6,7</sup>

Anatomic features of exstrophy include the presence of an open defect covered by a layer of urothelium on the anterior abdominal wall representing the bladder and urethra. At birth, the urothelium is usually normal in appearance. If left untreated after birth, the exposed urothelium will undergo squamous metaplasia in response to acute and chronic inflammation.<sup>4</sup>

The purpose of this paper is to report a case of bladder exstrophy – epispadias complex in a male term neonatal.

## THE CASE

BKR, a male baby delivered by SC in Sanglah Hospital Denpasar, at 18.45 p.m. May 31<sup>st</sup> 2006. He was born vigorously with Apgar score 7-9.

He is a second child in his family. Maternal ante natal care during gestation performed in Sanglah Hospital, and ultrasound determined that the pregnancy was normal. Mother realized that she was pregnant after her gestational age was 5 month and during the time she was continue taking contraception pil. During pregnancy, she never taken any other medication and never been sick (fever, urinary tract infection). She denied any symptoms of nausea, vomiting, vaginal discharge or bleeding, or any injury during pregnant. Diagnosis at delivery was: G2P1001, 41-42 weeks of gestational age, single/live, premature rupture of the membrane >6 hours. Cardiotocography: pathology.

On physical examination, activity of reflex tonus and cry was adequate. He had regular pulse rate, 140 beats per minute. The respiratory rate was 30 beats per minute. The axillary temperature was 36.7°C. His body weight and body length were 3200 gram and 50 cm. Head circumference 34 cm, chest circumference 33 cm.

Head normo-cephalic with flat fontanel. Strong and smooth hair. Eye examination showed no signs of anaemia, nor jaundice and palpebral oedema. Pupil reflex was positive in both eyes and isochor. Ear evaluation found perfect pinna and helix completely palpable. No enlargement of cervical gland found. Chest physical examination found that lung and heart were in normal state.

Abdominal revealed no distension, normal bowel sound and the liver and spleen were not palpable. Umbilical cord was fresh with slot beneath it. From the slot bladder appear and penis appear beneath of it in medial (**Figure 1**). Anal located very close to scrotum (<1 cm) (**Figure 2**). There was epispadias. Examination of lower extremities showed widening of pubic symphysis.

The result of blood laboratories investigation (on June 5<sup>th</sup> 2006) shown: leucocytes count was: 9.30 k/uL; haemoglobin was: 16.0 g/dL; hematocyte was: 47.5 %; platelets count was 328 k/uL. BUN was 13 mg/dL and SC was 0.6 mg/dL.

On abdominal x-ray, there was widening of the pubic symphysis (**Figure 3**). Urology ultrasound reveals absent of the right kidney at right *fossa renalis* or in pelvic cavity (**Figure 4**). Left kidney was in normal size, normal cortex echo, normal cortex thickness, clear cortex sinus border, collecting system slightly widening without any ureter dilatation, no evidence of stone, a cyst or mass (conclusion : slight hydronephrosis of left kidney) (**Figure 5**). No appearance of bladder in pelvic cavity.

Based on history, physical examination, x-ray and urology ultrasound, we diagnosed as term neonatal with bladder exstrophy – epispadias complex. He was given breastfeeding on demand and prophylactic antibiotic cefotaxime I.V. The Bladder Surface irrigated with normal saline and covered with non adherent plastic wrap, then he was referred to urologist and orthopaedist. Patient underwent surgery during hospitalized on June 8<sup>th</sup> 2006. On January 31<sup>st</sup> 2007, patient referred to the Congenital Anomaly Team in Dr. Soetomo Hospital – Surabaya for revision of bladder closing surgery.

## DISCUSSION

Bladder exstrophy is a very rare congenital malformation in which the anterior wall of the bladder is absent, and the posterior wall is exposed externally. Male to female ratio is 2.3 : 1.<sup>3,8</sup> There are no data about incidence of bladder exstrophy in Indonesia, especially in Sanglah Hospital. In our case reported here, bladder exstrophy found in a male term neonate.

The extent of the exstrophy depends on how small or how large the opening is. One can think of exstrophy as a spectrum of conditions that ranges from the milder form, epispadias (only a urethral defect or opening), to the most severe form, cloacalexstrophy (defect of the urethral, bladder and bowel), with classic bladder exstrophy (defect of urethral and bladder) being in the middle.<sup>2</sup>

The diagnosis is usually made at delivery by the typical appearance of the lower abdomen. The typical manifestation of exstrophy-epispadias complex are: (1) Bladder everted through a midline lower abdominal wall defect; (2) Widening of the pubic symphysis; (3) Epispadias in males (dorsal cleft in the penis, exposing the urethral mucosa) and (4) Bivid clitoris in females, with a short "urethral strip" indistinguishable from bladder mucosa.<sup>5,9</sup>

In our case, it manifested as bladder that everted through a midline lower abdominal wall defect associated with widening of the pubic symphysis and epispadias. In addition, also found a single left kidney.

In case of bladder exstrophy–epispadias complex , there is no specific result found in laboratory test.<sup>3</sup> In our case, laboratory test revealed a normal limit of white blood cell, haemoglobin, platelet and blood urea nitrogen/serum creatinin.

Formerly, classic bladder exstrophy was thought only to show the characteristic widening of the pubic symphysis caused by malrotation of the innominate bones in relation to the sagittal plane of the body along both sacroiliac joints. In addition, there was an outward rotation or eversion of the pubic rami at their junction with the iliac bones.<sup>3,9</sup>

In our case, we also found a widening of the pubic symphysis caused by malrotation of the innominate bones in relation to the sagittal plane of the body along both sacroiliac joints.

Exstrophy can be determined by physical examination. Other diagnostic procedure may include renal ultrasound or renal scan. Renal ultrasound is used to determine the size and the shape of the kidney, and to detect a mass, kidney stone, cyst, or other obstruction or abnormalities.<sup>4</sup>

In our case, urology ultrasound concluded a left slight hydronephrosis, no appearance of the right kidney in fossa renalis and no appearance of the bladder in pelvic cavity.

Bladder exstrophy is a surgical correctable birth defect. Reconstruction of exstrophy-epispadias complex remains one of the greatest challenges facing the pediatric urologist. Many modification in surgical procedures have improved the outcome for these patients, but the optimal approach remains uncertain. Longitudinal prospective assessment of both surgical approaches is critical to optimize functional and cosmetic outcomes. The

complex surgical repair of bladder exstrophy is performed by either the staged approach or the recently re-popularized complete primary repair technique (CPRE).<sup>5,6</sup>

The stage approach consists of three surgical components: (1) The bladder, posterior urethra and abdominal wall are closed during the newborn period; (2) The urethral closure (epispadias repair) is done at 6 months to 1 year of age; (3) The bladder neck reconstruction for urinary continence and bilateral ureteral re-implantation (for vesicoureteral reflux) are performed at 4 to 5 years of age, when the child has adequate bladder capacity, and is willing and able to participate in voiding program after the surgery. Using a stage approach, urinary continence rate as high as 88%. Preservation of kidney function with multistaged reconstruction represents a significant improvement over previous efforts at bladder closure. Evidence of renal damage at a rate ranging from 13% to 20%.<sup>7,9,10</sup>

Complete primary repair technique combines the goals of staged reconstruction into a single operation, so complete surgical repair is done sooner and normal bladder function can begin earlier. This allows for bladder growth and development. Early reconstruction including restoration of urinary continence would allow for mechanical cycling of the bladder that does not consistently occur with multistaged reconstruction of bladder exstrophy. Primary closure of the exstrophied bladder in the newborn period may offer the best opportunity for normal bladder development and may optimize the potential for urinary continence. Early primary bladder reconstruction would decrease the number of surgical procedures necessary to repair bladder exstrophy and minimize the emotional trauma that often accompanies conditions associated with numerous hospitalizations and surgical procedures.<sup>6,11</sup>



Proponents of urinary diversion believe that the exstrophic bladder is intrinsically defective. The use of the native bladder will thus likely require later bladder augmentation with intestinal segments to achieve a functional bladder storage capacity. Ureterosigmoidostomy became the treatment of choice for patients with bladder exstrophy as a primary and secondary therapy.<sup>12-14</sup> Issue of sexual function, fertility, and self esteem become more important long - term issues for children born with bladder exstrophy.<sup>9,15</sup>

In our case, Patient referred both to urologist and orthopaedist. Surgery for this patient performed in Sanglah Hospital Denpasar, June 8<sup>th</sup> 2006. First session of surgery, operation conducted by orthopaedist in an osteotomy surgery for right and left innominate, associated with placement of bilateral spica. Subsequently, continues with second session of surgery, conducted by urologist, consisted of closing bladder and bladder neck reconstruction.

After primary closure, combine closure or complete repair, failures can manifest as complete bladder dehiscence, bladder prolapse or neourethral stricture and obstruction. It is prudent to consider referral of these complex management situations to a center where special expertise and experience in dealing with the exstrophy condition exists.<sup>1</sup> In dr. Soetomo Hospital Surabaya, the overall primary closure successful rate is 70%.<sup>16</sup>

In our case, complication during post operation care was wound dehiscence on June 16<sup>th</sup> 2006. On January 31<sup>st</sup> 2007, patient referred to the Congenital Anomaly Team in Dr. Soetomo Hospital - Surabaya for revision of bladder closing surgery.

## **SUMMARY**

We has been reported a case of a male term neonate with bladder exstrophy–epispadias complex. The diagnosis is established from it manifested as bladder that everted through a midline lower abdominal wall defect associated with widening of the pubic symphysis and epispadias. Patient referred both to urologist and orthopaedist. Patient underwent surgery by orthopaedist in an osteotomy surgery for right and left innominata and by urology surgeon, consisted of closing bladder and bladder neck reconstruction during hospitalized. Complication during post operation care was wound dehiscence. Due to evidence of complicated post operative wound dehiscence, the patient then referred to Dr. Soetomo Hospital Surabaya. The fate of the case after 6 months was still in doubt.

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**Figure 1.** Bladder exstrophy - epispadias complex



**Figure 2.** Anal located very close to scrotum



**Figure 3.** Widening of the pubic symphysis caused by malrotation of the innominate bones in relation to the sagittal plane of the body along both sacroiliac joints



**Figure 4.** No appearance of the right kidney either in *fossa renalis* or in pelvic cavity



**Figure 5.** Slight hydronephrosis of left kidney