Epispadias Repair and Primary Closure of Bladder Exstrophy

EPISPADIAS ONARIMI VE EKSTROFİ VEZİKA'DA PRİMER ONARIM

Adnan ABASIYANIK*, Burhan KÖSEOĞLU, Lütfi DAĞDÖNDEREN**, Ahmet Hamdi GÜNDOĞAN**, Mahmut KIŞ***, Alaeddin DİLSİZ***

* Assist.Prof.Dept. of Pediatric Surgery, Selçuk University, Faculty of Medicine
** Resident, Dept. of Pediatric Surgery, Selçuk University, Faculty of Medicine
*** Specialist, Dept. of Orthopaedics and Traumatology, SIA Ankara Hospital, ANKARA

SUMMARY

We treated five patients with the exstrophy-epispadias complex during 1991-1994. We performed a bilateral innominate iliac osteotomy in all our cases and primary bladder closure was established in the following two months, except for a 72 hour old baby whose bladder was reconstructed on the third day of admission. All of the patients underwent penile reconstruction following an average of 13 months of their previous operation. One of our patients later had a gastrocystoplasty and a uretero-neocystotostomy with the reconstruction of the bladder neck because of a grade four vesico-ureteral reflux. In two of the remaining patients, a uretero-neocystotostomy was performed because of grade 4-5 reflux.

We managed to obtain a satisfactory urinary continence in four of our patients. Our only patient with urinary incontinence at present is three years old and a future bladder neck reconstruction is planned.

Key Words: Bladder exstrophy, Primary closure

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ÖZET

1991-1994 yılları arasında, ekstrofi-epispadias kompleksi olan beş hasta tedavi edildi. Başvurusunun üçüncü günü mesane onarımı yapılmış olan 72 saatlik bir hasta dışında kalan tüm hastalara bilateral innominat osteotomi uygulandı ve osteotomiden yaklaşık iki ay sonra primer mesane onarımı uygulandı. Hastaların tümüne ilk ameliyatlarından ortalama 13 ay sonra penis düzeltme işlemi yapıldı. Hastalarımızdan birine daha sonra dördüncü dereceden veziko-üreteral reflü ve mesane kapasitesinin yetersiz olması nedeniyle üreteroneosistostomi, mesane boynu onarımı ve gastrosistoplasti uygulandı. Geriye kalan hastaların ikisine dördüncü ve beşinci dereceden vezikoüreteral reflü nedeniyle üreteroneosistostomi uygulandı.

Hastalarımızın dördünde yeterli derecede üriner kontinans sağladık. Şimdi üç yaşında olan üriner inkontinanslı bir hastaya ilerde mesane boynu onarımı düşünülmektedir.

Anahtar Kelimeler: Ekstrofi vezika, Primer onarım

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Extrophy of the bladder is a rare congenital anomaly with an incidence of 1/30.000 births. The treatment of the disease needs patience and meticulous work. Most of the time a satisfactory result could only be maintained through a series of staged operations. The first step is primary closure of the bladder with iliac osteotomy. This is followed by the reconstruction of the bladder neck and repairment of epispadias. The major aim of these series of operations is to maintain a continent child with properly draining urinary system.

PATIENTS AND METHODS

We treated five patients with bladder extrophy between January 1991-May 1994. Patients' ages ranged between two days and six years. All of the cases had accompanying pathologies; one had uniltaeral inguinal hernia and undescended testis, four had bilateral hernias, one of which additionally had duplex ureter on the left side.

In four cases, we performed bilateral innominate iliac osteotomy with external fixation. All inguinal pathologies were treated during this stage. Three months after this, primary bladder closure was performed using the paraextrophic flaps and pubic bones were approximated using a polydioxanone (PDS) band passing through foramina obturatum. Ureteral catheters were kept in place for 11-13 days, and a 10-12 French catheter was passed through the penopubic neourethra. Bladder mucosa was topically treated with 5% polyvinyl pyrolin-iodine 12 hours prior to operation. Parenteral ceftriaxone 75 mg/kg/day and amikacin sulphate 10 mg/kg/day were given for 15 days. We closed the bladder primarily in the 72 hours old baby without an osteotomy.

Approximately 18 months after bladder closure, we constructed a neourethra using the penopubic flaps, following dorsal chordeectomy and penile lengthening procedure, in patients whose ages ranged between 13 months-7 years (Figure 1). A 10-12 French catheter was

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Yazışma Adresi: Dr.Adnan ABASIYANIK
Aziziye Cad. Köşem İşhanı
No:2/102
42030 KONYA

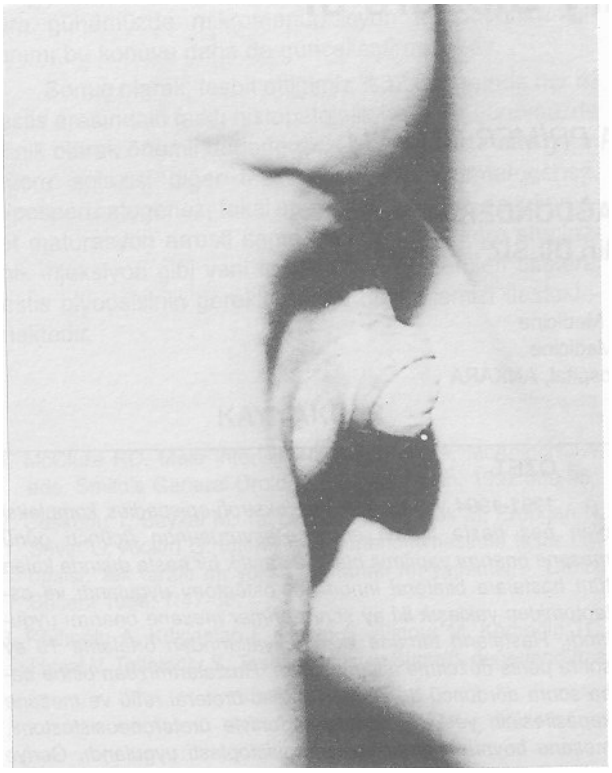

Figure 1.

used as a stent in the neourethra between 12-20 days. Under general anesthesia, we measured bladder capacity and obtained cyctograms in each case prior and a year after this procedure.

RESULTS

We observed wound infection in one of our patients following osteotomy which healed after drainage and antibiotherapy. One case developed a vesicocutaneus fistula on the 20th postoperative day after catheters were removed, which healed spontaneously. We did not see any major complications as bladder wall dehiscense (Table 1). One case developed an urethral fistula on the 12th postoperative day following removal of the stent, which also healed spontaneously after two weeks (Table 2).

Bladder capacity of the patients prior to epispadias repair was calculated to be between 21-43 mL. In four cases the capacity ranged between 54-65 mL approximately a year after epispadias repair. Cystograms revealed a unilateral grade four vesico-ureteral reflux on the left side in one case and grade five on the right and grade three reflux on the left side in another case. We performed uretero-neocystostomies in these patients and have satisfactory cystograms two months postoperatively, but one developed ileus on the seventh postoperative month. Two other patients both have unilateral refluxes, grade one on the right and grade two on the left side respectively (Table 2).

Table 1. Documents and results of patients

	Age/Sex	Inguinal Pathology	Osteotomy/Complications	Primary Bladder Closure/Complications	Bladder Capacity (Before Epispadias Repair)
B.Ö.	13 m/M	Bilateral inguinal hernia	Yes/No	Yes/No	21 ml
S.K.	1 y/M	Left inguinal hernia+Undescended testis	Yes/No	Yes/No	43 ml
O.K.	6 y/M	No	Yes/No	Yes/No	25 ml
M.A.	10 m/M	Bilateral inguinal hernia	Yes/Wound enfection	Yes/Vesicocutaneous fistula (closed spontaneously)	30 ml
R.U.	2 d/M	Bilateral inguinal hernia	No/No	Yes/No	36 ml

m: month, y: year, d: day M: male

Table 2. Documents and results of patients after epispadias repair

	Age/Sex	Epispadias repair complications	Upper urinary tract pathologies	Ureteroneocystostomy/ Complications	Bladder capacity	Continence
B.Ö.	22 m/M	Yes/No	Grade 1 right VUR*	No/No	65 ml	Continet (dry 3 hr)
S.K.	3 y/M	Yes/Bladder stone	Grade 4 left VUR and left double ureter	Yes/No	62 ml	Continent (dry 3 hr)
O.K.	7 y/M	Yes/Urethrocutaneous fistula (closed spontaneously)	Bilateral Grade 4 VUR	Yes+Gastrocystoplasty and bladder neck reconstruction/vesicocutaneous fistula (closed spontaneously)+Hematuria and dysuria syndrome	82 ml	Intermediate (90 min)
M.A.	2.5 y/M	Yes/No	Grade 5 right VUR	Yes/Ileus	55 ml	Continent (dry 3 hr)
R.U.	13 m/M	Yes/No	Grade 2 left VUR	No/No	54 ml	Wet (dry 25 min)

m: month, y:year
*VUR: Vesicoureteral reflux

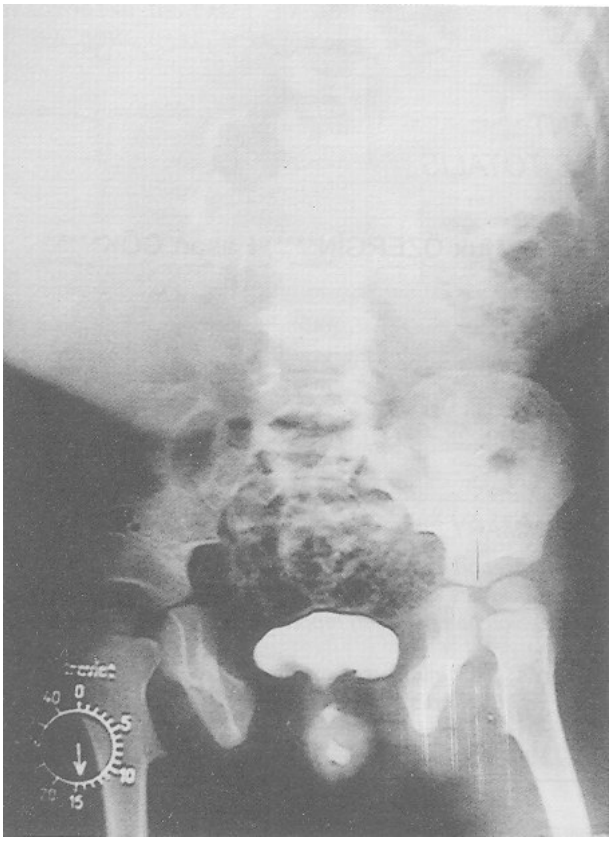

Figure 2.

Three of the cases can stay dry for three hours and the remaining one who is 2.5 years old can stay dry for half an hour. This patient has 54 mL of bladder capacity and we are planning to perform a bladder neck reconstruction in the future.

The remaining one case is an eight year old boy who had 25 mL of bladder capacity with bilateral grade 4 reflux. We performed a gastrocystoplasty+bilateral ureteroneocystostomy (cephalotrigonal reimplantation), and a Young-Dees-Leadbetter bladder neck reconstruction (Figure 2). On the 18th postoperative day following removal of the catheter, he developed a vesico-cutaneus fistula which healed spontaneously after 12 days. He had 82 mL of bladder capacity two months postoperatively. He can stay dry for a period of 80 to 90 minutes and can micturate on his own will.

One of our patients underwent ESWL for a bladder stone which was formed two years after bladder closure.

DISCUSSION

Numerous studies that have been published for the past 20 years, have revealed that the best treatment for this congenital anomaly is staged operations (1-3). Mainstay of our approach to patients with this anomaly is primary closure of the bladder, provided that they have sufficient bladder capacity. It has been postulated that primary closure in the first 72 hours of life is an important factor which may lead to better results (4). However, Husmann et al (5) state that there is no major difference in the incidence of bladder wall dehiscense following primary closure, performed either within 72 hours or one month. We also believe that primary closure should be performed under one month of age.

We used innominate iliac osteotomy since it seems to provide a better approximation for the pubic rami, and we repaired the inguinal pathologies during same stage (6). It is stated that bladder wall dehiscense is seen mostly due to wound infections after osteotomies or hernia repairs (5). In order to avoid such complications we performed our bladder closure operations at a second stage.

In order to increase bladder capacity (7), we repaired the epispadias complex earlier (13-22 months) in two of patients. We observed a 50% to 200% increase in bladder capacity in these patients with unilateral grade one and grade two reflux respectively. It is also important to mention that in one patient who was eigth years of age, bladder capacity failed to increase following epispadias repair with bilateral grade four reflux, and we had to do an augmentation cystoplasty and cephalotrigonal reimplantation of the ureters (8). It is obvious that bladder neck reconstruction has to be accomplished before bladder undergoes structural refractory changes.

The distance between pubic bones is important when it comes to repair the penile anomaly with good cosmetic result. Therefore osteotomy has to be done with great care to obtain a penile body with sufficient length (9).

As for conclusion, we managed to keep three of our patients dry for a period of three hours. A proper osteotomy, successful primary closure of the bladder with a urethral lenght greater than two cm and 60 cm H2O urethral closing pressure is mandatory to achieve a continent urinary system (10,11).

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