Original Article

Single Stage Primary Closure of Bladder Exostrophy VS Two Stage Closure: Experience of 18 Patients

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ABSTRACT

Objective: To study the outcome and complications of single stage primary repair of Bladder Exostrophy and compare it with two stage repair.

Study Design: Descriptive study

Place and Duration of Study: The study was carried out at Paediatric Urology Dept, Children Hospital Complex & Institute of Child Health, Multan from 1st January 2008 to 30 December 2009.

Patients and Methods: We retrospectively reviewed the records of all patients operated for primary single stage and two stage repair of Bladder Exostrophy. Patients were divided into two groups based on the type of surgery performed. Group I consist of patients with single stage complete repair and group II consist of patients with two stage repair. Data of both groups was entered on a proforma and surgical outcome, complications and urinary continence was noted.

Results: There were total 18 patients 13 male, 3 female, Group I (3male 5female) and Group II (10 male). Age ranges from two days to one year in group one with mean age of four months and in group two age ranges from four days to one year with mean age of six months.

Average surgery time, bloodloss and anaesthesia recovery time was slightly higher in Group I but the complications like wound infection, wound dehience and reoperation rate was also more in Group I as compared to Group II. Long term outcome like VUR, continence interval was similar in both groups.

Conclusion: Single stage primary repair offers the advantage to correct the defect in one sitting and psychologically comfortable for the parents but it needs greater surgical experience and complications are higher in male patients. In our current setup single stage complete repair is suitable in female patients but in males two stage repair is more appropriate with satisfactory results.

Key Words: EEC (Exostrophy Epispadias Complex, VUR (Vesico Ureteral Reflux), Cantt-well Ransley Urethroplasty, Pelvic osteotomy, Urinary continence.

INTRODUCTION

Exostrophy Epispadias Complex (EEC) of genitourinary malformations can be as simple as glandular epispadias or an overwhelming multi system defect such as clocal exostrophy.

The first account of Bladder exostrophy is ascribed to Asyro Bablonia sources dating from 1st and 2nd millennia BC; however first clear description of Bladder Exostrophy is attributed to Schenck in 1595[1]. Its incidence has been estimated as between 1 in 10,000 to 1in 50,000 [2]and male ratio female is 5:1 to 6:1. The etiology of Bladder Exostrophy is not known but reports from two centers suggest possible hormonal cause[3] of the complex. A possible genetic basis for development of EEC is being explored.[4]

In Classic Bladder Exostrophy, most abnormalities are related to the defects of abdominal wall, bladder, genitalia, pelvic bones, rectum and anus. Abdominal wall shows triangular defect occupied by exostrophy bladder and posterior urethra, limited inferiorly by inter-symphysial band and laterally by divergent recti. There is diastasis of pubic symphysis along with external rotation of anterior and posterior pelvis. As a result there is wide separation of crural attachments, prominent dorsal chordee and shortened urethral groove. In females the vagina is short, orifice is stenotic, clitoris is bifid and labia, mons pubis, and clitoris are divergent.

Although a myriad of operations for exostrophy have been proposed and attempted, these operations can be broadly categorized into two approaches. First approach includes operations for urinary diversion and the second includes procedures to reconstruct the bladder either in multistage or in a single stage. In single stage procedure pelvic osteotomy is followed by primary bladder reconstruction, anterior urethroplasty with penile disassembly technique in male, pubic approximation and fixation, abdominal closure and skin coverage. In two stage repair pelvic osteotomy is followed by primary bladder and posterior urethra repair, pubic approximation and abdominoplasty in first stage . Bladder exostrophy is converted into complete epispadias in this stage. Anterior urethroplsty is prformed one year later after primary repair with modified Cantt-well Ransley urethroplasty.

The single stage approach offers the advantage to correct penile bladder and abdominal defect in one setting, but it requires greater expertise and complication rate is high, whilst the two stage operation offers the advantage of less complication and the results are equally satisfactory.

The purpose of this study is to compare the advantages and disadvantages of single stage complete repair versus two stage closure of primary bladder exostrophy.

PATIENTS AND METHODS

This study was carried out at Children Hospital Complex, Multan during two years period from 1st January 2008 to 30th December 2009. There were 18 children (5 females 13 male). Children included in this study ranges from 2 days to one year. Patients with associated exampholos, previous failed repair and small dysplastic bladder template unsuitable for primary repair were excluded from this study.

All patients presenting to out door or referred from other hospitals were considered. Common presenting symptoms were visible abdominal and genital defect with continuous leaking of urine from this defect.

Complete history was taken along with general physical examination performed. Blood tests Complete blood examination, Complete Urine Examination, serum urea, creatinine, Sodium and Potassium, x-ray chest x-ray KUB and Ultrasound abdomen performed in all patients. Bladder template was also assessed under anesthesia for suitability of closure regarding size, surface and elasticity. Bladder mucosa was immediately protected by thin moist plastic sheet which was continuously irrigated and changed frequently to prevent mucosal ulceration.

Patients were divided into two groups based on the type of surgery performed for closure of defect.

First group 8 patients (5 females 3males) were operated by single stage complete repair i.e bilateral posterior pelvic osteotomy, cystoplasty, abdomino-plasty, pubic approximation, total penile disassembly and anterior urethroplasty by modified Cantt-wel Ransley technique. Second group 10 patients (all male) two stage repair was done i.e bilateral posterior pelvic osteotomy, cystoplasty, abdominoplasty, pubic approximation and bladder neck repair early while total penile disassembly and anterior urethroplasty by modified Cantt-wel Ransley technique one year after primary repair was done.

After surgery patients were monitored in PICU for 24 hours and then moved to Pediatric Urology ward. Post operatively gallows were applied for 14 to 21 days in 15 patients and hipspica in 3 patients. Intravenous broad spectrum antibiotics were given for nine to ten days. All patients were discharged on low dose antibiotics and called for follow up in outdoor at 2 weeks, 6 weeks, 3 months and then after every 3 months for 2 years. During follow up wound was examined to see any dehiscence, uretheral fistula formation, motor and sensory system of lower limbs were also examined. Voiding history was taken and continence interval, bedwetting during day and night was noted. In every follow up Complete Urine examination and culture sensitivity, serum urea, Creatinine and Ultra-sonography of abdomen was done. Micturation cystouerthrography at six months & DTPA renal scan was done yearly to see bladder capacity, vesico ureteral reflux and any deteoriation in renal function. Data of both groups of patients was recorded on proforma and compared.

RESULTS

There were eight patients in group I (three males and five females) and ten patients in second group II (all male). Age ranges from two days to one year in group one with mean age of four months and in group two age ranges from four days to one year with mean age of six months.

Table No.1: Early Outcome of Surgery

	Group I	Group II
Surgery time	M 4.5 to 5 hours	M 2.5 to 3 hrs (stage I)
		M 1.5 hours (stage II)
Blood loss	M 100 to 150 ml	M 50 to 70 ml (stage I)
	F 80 to 100ml	M 15 to 20ml (stage II)
Anesthesia recovery	15 to 20 mints time	15 to 18 mints
Ventilator support needed	M 1	Nil

Table No.2: Comparison of Complications

	Group I	Group II
Infection (superficial)	3(37.5%) 1M(12.5%)2F(25%)	2(20%)
Wound dehiscence		
Abdomen (Complete)	1 M(12.5%)	None
Abdomen (normal)	1 F(12.5%)	1 M(10%)
Bladder (Complete)	1 M(12.5%)	None
Urethra (Complete)	1 M(12.5%)	None
Uretheral fistula	1 F(12.5%)	2 M(20%)
Osteotomy Complications		
Wound infection	1 F(12.5%)	None
Skin excoriation	2F (25%)	3M(30%)
Foot drop	None	1(10%)
Sepsis (shock)	1M (12.5%)	None
Re-operation	2 (25%) (1 M12.5%,1F12.5%)	None

Comparison of average surgery time, blood loss, recovery from anesthesia and needing ventilator for group I and group II are shown in the table 1.

All females were managed with single stage complete repair as female urethroplasty needed 30 to 40 minutes of surgery. In male approximately 1.5 hours is needed to perform urethroplasty and the surgical expertise required to complete this delicate operation is much greater in male patients as compared to female patients.

Comparison of complications between the two groups is shown in table 2.

Table No.3: Long Term Outcome

	Group I	Group II
Continence interval		
2.5 to 3 hours	5 (3F, 1M)	6 (M)
2 to 2.5 hours	1 (F)	2 (M)
1.5 to 2 hours	2 (1F)	2 (M)
Less than 1 hour	1 (1M)	None
Clean Intermittent Catherization (CIC)	3 (2F, 1M)	2 (M)

In, Group I, one male patient went into septic shock and was on ventilator for 8 days. He survived the sepsis but had complete dehiscence of repair and needed major reconstructive surgery later. Majority of patients with superficial infections healed with conservative treatment. In Group II, two patients developed peno-pubic area fistula after second stage repair. Both patients healed spontaneously with conservative treatment.

Bilateral posterior pelvic osteotomy was successfully done with no major complication. Pubic bones approximation was nearly complete. Skin excoriation occurred in 3 patients which healed spontaneously. One patient develop (male) in group II developed left foot drop after surgery which recovered partially with physiotherapy.

Comparison of results regarding continence and need for Clean Intermittent Catherization (CIC) one year after complete repair are shown in table 3.

In patients with single stage complete repair 2 females developed partial urethral stenosis and was later managed by CIC. All 8 patients developed Vesico-ureteral reflux of (5 of grade 3, 3 of grade 4) and managed conservatively. No incidence of recurrent Urinary tract infection was noted.

In 10 patients with two stage repair, all developed vesico-ureteral reflux (six of grade 3 and 4 with grade 4) & managed conservatively. No incidence of recurrent Urinary tract infection was noted

DISCUSSION

Primary closure is the most logical treatment of Bladder Exostrophy with restoration of abdominal wall defect and preservation of renal function. Preoperative assessment and post-operative care is the key for successful surgery.

Efforts to reconstruct the genitourinary system have been tried for over hundred years. Primary closure cannot be achieved when there is hard fibrotic bladder with a small capacity. Trendelenburg (1906)[5] for the first time attempted to reconstruct an exostrophic bladder to achieve urinary continence but his patients did not gain satisfactory continence. Such discouraging results let to abandonment of the functional reconstruction and cystectomy with uretero-sigmoidostomy became the treatment of choice. Young[6] in 1942, Ansell [7] in 1971 and Montagnani[8] in 1982 reported several patients with single stage functional bladder closure. Because of lower rate of urinary continence 0% to 45% and high incidence of renal damage 90% due to bladder outlet obstruction, reconstructive surgical efforts were directed toward staged bladder reconstruction, an approach pioneered and advocated by Dr. Robert Jeffs.[9]

To gain satisfactory results from surgery certain points must be considered. Early operative treatment and in newborns with a wide pubic diastasis anterior or posterior pelvic osteotomy are major factors in maintaining a large bladder volume. Schmidt[10] reported experience of bilateral posterior pelvic osteotomy series of patients in 1993 that osteotomies assist closure and enhance anterior pelvic support which may improve later urinary continence. Purves [11](2007) performed combined vertical iliac and horizontal in nominate osteotomy in staged bladder reconstruction and reported satisfactory results. Similar findings were noted by Salman Riaz[12] at Agha Khan University hospital in 2005. Our experience of osteotomy (posterior pelvic in 17, anterior iliac in one)was also satisfactory. It helped in closure of abdomen and contributed towards achievement of continence. There were no significant complications except one patient who developed left foot-drop which late rimproved.

The single stage anatomic approach offers the advantage to correct the penile, bladder, and bladder neck abnormalities in one setting.[13] In a review of records of patients evaluated for the genital complications following complete repair of bladder exostrophy from 1996 to 2003 at myo clinic, Minnesota by Husmann, [14] 9 patients were noted to have serious genital injuries including complete loss of glans and corporal bodies and penile urethra. Baired[15] after reviewing records of 38 patients at Brandy Urological institute with previous failed primary closure or delayed closure reported very high complication rates, lot of patients needed additional surgery and continence rate at best was reported as 50%. In our experience of 8 patients with single stage repair, two females had superficial wound infection one male developed sepsis and complete dehiscence of wound. These findings dampen our enthusiasm for this procedure. These new techniques currently the Mitchell[16] or CPRE technique have a lower complication rate and urinary continence

can be achieved for many of these patients without the need for further bladder neck reconstruction.

After unsuccessful primary treatment in patients with EEC, the options for a surgical solution to preserve the upper urinary tract, to achieve complete continence and to reconstruct the genitalia are limited. Stein[17] from Germany (1999) reviewed 128 patients after unsuccessful or unsatisfactory primary closure and concluded that first operative intervation in patients with bladder exostrophy determines their fate. Similar results were reported by Osterling[18] at John Hopkins (1987) after review of 144 patients. In our experience in patient with complete dehience of wound, secondary repair was difficult, continence interval short and bladder was small in capacity which later needed augmentation cystoplasty. But our experience with 5 females & 2 male patients was excellent and both healed successfully with minimal complications. In females single stage repair has shown good results. There are no serious early complications and long term outcome like urinary continence rate is also satisfactory.

Since 1970 the staged reconstruction of bladder exostrophy has yielded consistent surgical success. Baker[19] in 1998 at John Hopkins reported early abdominal closure in two stages with bilateral pelvic osteotomy, bladder, abdomen and posterior urethra at first stage and after 1 year anterior urethra closure by modified Canttwell Ransley technique in patients suitable for primary closure. This approach usually results in a continent, voiding patient with pleasing external genitalia and preserved renal function. Our experience of 10 male patients with two stage repair was satisfactory with no serious complications and good long term outcome. Two stage repair with pelvic osteotomy has shown world wide good results with minimal complications, satisfactory cosmetic appearance after closure and urinary continence rates are good.

Baka[20] in 2000 reported a series with two stage repair continence rate of 75% in patients with classic bladder exostrophy. Similarly Baired [21]in 2006 after reviewing the record of 131 patients reported that patients with a good bladder template who develop sufficient bladder capacity after successful primary closure can achieve acceptable continence without bladder augmentation and intermittent catherisation.

Single stage closure saves the patient from second surgery and anesthesia and psychologically comfortable for the parents, but it needs greater surgical expertise with prolong anesthesia time. We recommend that such surgery should only be done in specialized institutions with pediatric and neonatal intensive care units. Although our experience with male patients for single stage repair is limited (2 Patients) but complications were high as compared to females. Two stage repair require two separate surgeries with anesthesia and prolong hospital stay but the results were good even in male patients. We therefore recommend that single stage complete repair of Bladder Exostrophy may be done with satisfactory results in females but for primary closure in males two stage repair is more appropriate.

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