Pelvic osteotomies for exstrophy: a review of techniques and outcomes at Red Cross Children's Hospital

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Declaration

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Introduction

Bladder exstrophy is a rare congenital anomaly in which the open bladder is part of the anterior abdominal wall. Classic exstrophy comprises an open bladder, pelvic diastasis and complete epispadias. It is part of a spectrum of disorders involving the musculoskeletal system, the genito-urinary tracts and at times the intestinal tract. The severity ranges from simple balanic epispadias with closed bladder but widened symphysis, through classic bladder exstrophy to the severest form, cloacal exstrophy. The last has variable presentation, but includes in addition to the open bladder, omphalocoele. All share their common origin in the failure of ingrowth of mesoderm between the ectoderm of the abdomen and the cloaca during the fourth week with resultant absent midline closure of the inferior part of the anterior abdominal wall. In addition all have the potential for other associated congenital abnormalities and in particular neural tube defects and spinal deformities.[1,2]

Untreated, bladder exstrophy may lead to two lethal complications. Recurrent urinary tract infections result from continual exposure of the open bladder to the environment and results in renal failure. The infections and or exposure may also lead to endothelial metaplasia and malignant transformation[3,4]. The lesser complications are incontinence, cosmetically unacceptable genitalia, deficient anterior abdominal wall musculature, externally rotated lower limbs and inadequate development of bladder capacity.

Objectives of the surgical team are to obtain and maintain closure of the anterior abdominal wall as well as the urogenital tract. Secondary goals are to provide continence, to reconstitute cosmetically acceptable external genitalia and to correct gait deformities. The orthopaedic surgeon's role is to perform a pelvic osteotomy, which facilitates closure, especially of the bladder, by decreasing tension on the anterior abdominal wall. Approximation of the diastased symphysis pubis is attempted and may, or may not improve continence.

The purpose of this retrospective study is to review the outcome of these difficult pelvic osteotomies and primary closure in patients with exstrophy.

Incidence

Exstrophy is a rare condition. Marshall and Meuke quote Mowatt as first reporting it in 1747. The prevalence of classic bladder exstrophy is one 1 in 10 000 to 1 in 40 000 and that of the more severe cloacal exstrophy is 1 in 200 000. [4,5,6]. Boys are more commonly affected with a risk of 2.5 times that of girls. There are reports of identical twins with both twins affected, and with only one twin affected.[4] The risk to subsequent children born to parents of a child with exstrophy is 1:100, and the risk to children born to parents with exstrophy is 1:70. This equates to an increase in risk of 150-500 times that of the general population.

Embryology

The central feature of the exstrophy complex is a failure of mesenchymal cells to migrate to the midline in the fourth week of development. This causes a mesodermal defect between the ectoderm (skin) of the abdomen and the cloaca. The normal mesodermal structures – muscle and connective tissue and bone, fail to develop between the bladder and the abdominal skin. Subsequently the attenuated epidermis of the anterior abdominal wall and the endodermis of the bladder rupture exposing the posterior wall of the bladder.[8] There is no functional sphincter and urine drains continually from the exposed bladder.

The anomaly is characterised by this exposure and protrusion of the posterior wall of the bladder. The bladder trigone and ureteric orifices are exposed. The midline defect extends to the penis manifesting as an epispadias (opening of the urethra on the dorsum of the penis). In some cases the penis is split in two and the scrotum similarly bifid. The pelvis, a mesodermal structure, is separated (diastased), and anteriorly deficient.

Depending on the extent of midline mesodermal failure, a spectrum of disorders is created. The simplest is epispadias. Classic exstrophy follows in severity. Cloacal

exstrophy is a more rare and more severe variant. The bladder lies in two halves with the intestine prolapsed in between.

Interestingly, whereas most congenital anomalies occur as an arrest at an embryological stage (e.g. cleft lip and palate), exstrophy is not a normal embryonal stage.[7] There is also no reliably confirmed animal equivalent. In addition the complex is not confined to one organ system. Associated anomalies include in addition to defects of abdominal wall, bladder, genitalia and anus/rectum, those of renal anomalies, clubfeet, omphalocoeles, myelomeningocoeles, sacral agenesis[7,9]Presumably the causal insult acts simultaneously at multiple embryonic sites.

Pathoanatomy

Some controversy exists as to the causal events in the exstrophy complex. Marshall and Meuke[7] argue strongly in favour of a persistent overdeveloped cloacal membrane. This, they postulate, may either be an arrest in development, or as they favour, true overgrowth. As a result a "wedge effect" occurs, keeping midline structures diverged but intrinsically normal. They cite the normal musculature of recti and bladder to support this theory. In addition they mention the fact that the bones (mesodermal structures) are almost never underdeveloped or abnormal.

In contrast, Moore concluded that the soft tissue and bony abnormality is more likely due to a failure of normal mesodermal ingrowth.[8] In support of this is the fact that all anterior mesodermal structures are involved – recti, anterior bladder wall and pelvis. Failure to maintain soft tissue and bony repair is common in exstrophy. In another very different group of patients with wide diastasis – conjoin twins, but without midline mesodermal defects, Verrier and Hoffman found excellent medium term results with maintenance of bony anatomy. In this group of patients there is no

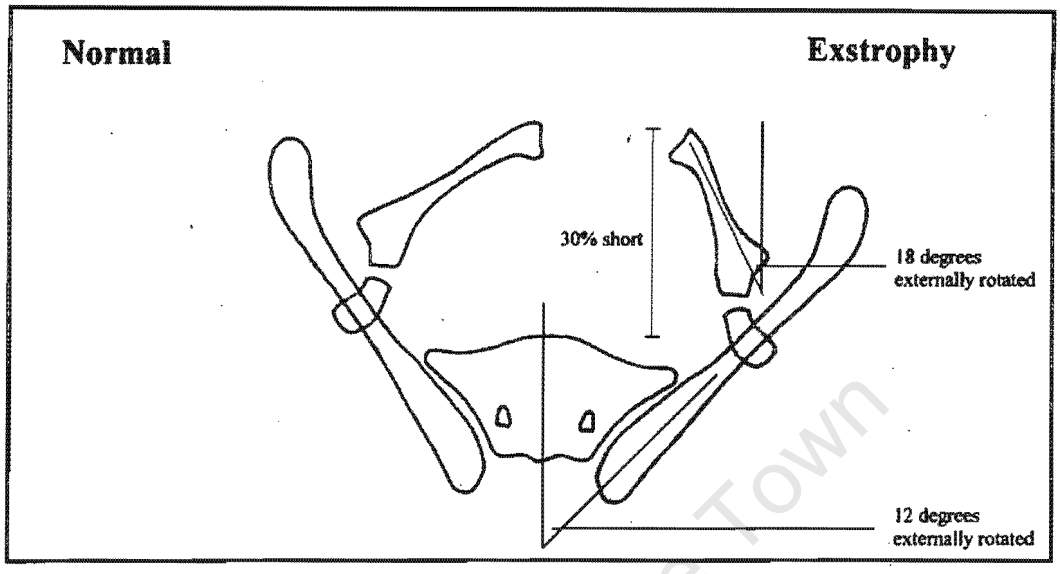
proposed mesodermal midline defect.[13] Lloyd Roberts[14] described shortening in two planes. Firstly, failure of the union of anterior midline structures (bladder, urethra, pelvic ring, anterior muscles and anterior abdominal wall). The second defect he considered to be saggital – the umbilicus drawn downwards, the urethra drawn upwards and shortened, the penis/vagina foreshortened and the anus drawn forwards in the perineum.

In addition to the soft tissue pathology, the bony abnormalities have been studied. Sponseller et al used computerised tomography (CT) scans of 24 patients with bladder exstrophy compared with aged-matched controls.[10] For descriptive purposes they divided the pelvis into an anterior (ischium and pubis), and a posterior (ilium) segment. They found that in classic exstrophy there was external rotation of the posterior segment of the pelvis and external rotation as well as shortening of the anterior segment. Additionally there was diastasis which increased with age. Their results are summarised in Table 1.

Table 1. Summary of abnormal findings in anatomy of the pelvis in exstrophy. Sponseller et al.[10]

	Classic Exstrophy	**Cloacal Exstrophy**
Posterior segment angulation	12° external rotation	25° external rotation
Anterior segment angulation	18° external rotation	24° external rotation
Acetabular version	13° retroverted	0° retroverted
Length of anterior segment	30% of normal	40% of normal
Interpubic distance (diastasis)	5.9 cm	8.0 cm

Figure 1: Composite view of pelvis comparing normal anatomy with exstrophy (redrawn from Sponseller et al.[10])



In a similar and complementary study, the bony anatomy of 6 children were studied by Yazici et al.[12] CT scans were used to compare the anatomy to that of normal controls. They too found the posterior and anterior segments to be externally rotated and the anterior segment shortened. They also found an apparent acetabular retroversion in the patients with exstrophy.

McKenna and Wedge utilised 3-dimensional computerised tomography (3D-CT) in 7 patients.[11] There were no normal controls. Plaster models were created using the 3D-CT's as templates. They found the apex of bony deformity to be at the level of the mid-ilium. They noted widened interpubic distance (diastasis), which increased with age.

Gargollo et al [15] used Magnetic Resonance Imaging (MRI) to evaluate the deformities in exstrophy. The used the iliac wing angle, obdurator internis angle and the ischial angle as markers. They measured the differences between exstrophy patients and normal controls. They also measured the differences in patients pre- and post-surgery. They found that surgery improved but never completely normalised

these values. They also found that the anatomy of those patients who became continent more closely approximated normal controls than those who remained incontinent.

Table 2. Pelvic anatomy pre-surgery, post-surgery and gender matched normal controls. Gargollo et al [15]

	Pre-surgery		Post surgery		Gender matched control
Iliac wing angle (degrees)	male	128.0	male	118.0	96.7
	female	123.5	female	105.7	
Ischial angle (degrees)	male	5.5	male		68.4
	female	12.8	female		
Obdurator internis angle (degrees)	male	22.9	male		55.8
	female	18.5	female		

Surgical management

The management of these complex deformities is difficult with a high attendant rate of morbidity and complications. They require a customised multidisciplinary approach, usually starting within the first few days of life, and often requiring procedures into adolescence and beyond. Despite wide variations within the details of current management, and recognising that considerable controversy exists, certain principles remain. The primary goals in management are to obtain secure closure of the anterior abdominal wall and the urogenital tract. Secondary goals are to obtain urinary continence and protect upper urinary tracts, to reconstruct functional and cosmetically external genitalia, and to restore normal gait.

Two main schools of thought exist in the management of the open bladder. The one option is urinary diversion, cystectomy and epispadias repair. This in effect converts the open bladder to an ureterosigmoidostomy. With access to better surgical and nursing care, this is largely being regarded as the inferior option. The attraction is that it is relatively simple, that it potentially saves on multiple procedures, and that the short term surgical morbidity is less. The downside lies in the fact that there remains a communication between gut and urinary tract. Magelli and Lattimer reported 87% of patients with ureterosigmoidostomy developing recurrent pyelonephritis. One third went on to ileal loop diversion, with its own attendant risk of carcinomatous endothelial change[16]. Other recognised complications include hyperchloraemic acidosis, urinary calculi and stricture formation. The alternative is reconstruction. This is achieved via initial primary bladder closure, bladder neck reconstruction and epispadias repair. Intuitively reconstruction is the more attractive option with the potential for continence, and the avoidance of both recurrent urinary tract infections and neoplasia. Continence, however has been variably successful with reported outcomes ranging from 45% to 90%[1,17,18]. Both approaches carry significant morbidity. Current trends recognise the increasing success with reconstruction and favour this approach with urinary diversion retained as a salvage procedure.

If the latter option, namely reconstruction, is taken as the preferred treatment, there are again two main schools of thought.

Gearhart and Jeffs et al from the John Hopkins Hospital, favour a staged approach to reconstruction[20,21]. In the first stage, usually in the newborn period, the bladder and abdominal wall is closed, and the urethra is closed onto the penis. Pelvic osteotomies are performed where necessary. Later the epispadias is repaired at around 1 year. Bladder neck reconstruction and an anti-reflux procedure is performed once the bladder capacity is sufficient and continence is feasible at around 4 years. In contrast Grady, Ansell and Mitchell [19,22,23] favour single stage anatomic reconstruction including bladder closure, pelvic osteotomy narrowing of the bladder outlet and an anti-reflux procedure. They stress the advantage of a single surgical event and a lower complication rate., but continence rates are lower.

Orthopaedic Management.

Addressing pelvic pathology has become an integral part of addressing soft tissue reconstruction. It allows midline approximation of soft tissue structures. This in turn aids healing by reducing wound tension. Bladder repair dehiscence is also reduced. It allows the levator ani and puborectalis muscles to add resistance to urinary outflow[14]. This in turn stimulates bladder development It may also improve gait by redirecting the acetabulae[25,26]. In the first 48 hours of life, and when the pubic diastasis is not too severe, Ansell[19] promoted the use of plastic deformation of the pliable pelvis. After the effect of relaxin has disappeared, or when simple manual pressure is insufficient, a formal osteotomy becomes necessary. A variety of differing procedures have been described, but all seek to aid midline closure and to provide soft tissue support.

Study

Aim

The aim of this study was to retrospectively review the surgical management of children with classic and cloacal exstrophy treated at Red Cross Children's Hospital.

Materials and Methods

Thirteen children with exstrophy were treated from 1990 to 2006. Seven had bladder exstrophy and six had the more severe cloacal exstrophy. In the cloacal group, one had a variant in which an intact bladder and urethra was present. One patient died from post-operative sepsis. One patient was lost to follow-up. The clinical notes, plain radiographs, and (when available) computerised tomography were assessed. Nine patients were available for clinical follow-up of >2 years.

There were six boys and seven girls. No genotypically male patients had gender reassignment. Associated congenital abnormalities were present in 10 patients. These included partial sacral agenesis, myelocystocoele, clubfoot and exomphalos.

Two patients had attempted closure with manual compression alone at 3 days, 4 days and 6 weeks respectively. In the remaining 11 patients, 12 osteotomies were performed. One patient, having been treated elsewhere, may have had a prior osteotomy. Eight osteotomies were done between 1 and 16 weeks of age (average 5.2 weeks). Four delayed osteotomies were done: one patient with cloacal exstrophy and a normal closure at 1 year 3 months, a patient with a failed closure at 10 weeks had a delayed closure at 4 years. Two patients had been treated elsewhere and had osteotomies at 5 and 16 years respectively. Of the osteotomies there were three posterior iliac osteotomies, three anterior pubic and seven mid-iliac-oblique osteotomies. Post-operatively the approximation was maintained in the initial cases

with polydioxanone sutures (PDS) accompanied by patient swaddling, and, more recently with external fixation.

Eleven patients were available for follow-up, between 6 months and 11 years of age (average 4.3 years). In these children five factors were evaluated:

- Facilitation of closure. This was a clinical subjective impression of the closure obtained post-osteotomy compared with the closure pre-osteotomy using lateral manual pressure alone.
- Post-operative wound dehiscence. This was taken to be any major wound breakdown requiring revision surgery and or prolonged dressings and antibiotics.
- Urinary continence. Without a consensus on the definition of continence, in this study we followed the criteria of Lottman et al[28]. Good is defined as complete continence with dry intervals of 3 hours, acceptable as dry intervals of 2 hours and poor as persistent incontinence or upper urinary tract deterioration.
- Restoration of pelvic anatomy. This was assed by measuring the interpubic difference (IPD, normal < 1 cm)) as a measure of diastasis. It was measured pre-operatively, immediately postoperatively, and at late follow-up. The percentage of pubic approximation was also calculated according to the formula proposed by Sponsellar[10]:
  $\frac{\text{pre-operative diastasis (mm)} - \text{post-operative diastasis (mm)}}{\text{pre operative diastasis (mm)} - 10\,\text{mm}} x100$. The denominator corrects for the 1cm symphyseal separation seen on X-ray as a result of the cartilaginous symphysis pubis.
- Functional orthopaedic outcome – gait, rotational profile and acetabular status assessed in 9 patients at late follow-up. The patients were observed for a Trendelenburg limp or a wide based gait. The rotational profile was assessed by noting the foot-progression angle during gait, and the thigh foot angle and internal and external rotation of the hip with the patient prone. On a standing anteroposterior radiograph of the pelvis, the acetabular index (or Sharp's angle in patients older than eight years), was assessed

Results

Facilitation of closure

A senior paediatric orthopaedic and urological surgeon was present at all the procedures. At surgery, all patients were noted to have easier closure following osteotomy compared with manual lateral pelvic pressure alone. Subjectively the easiest closure was that obtained mid-iliac oblique osteotomy.

Pelvic anatomy

The pre-operative interpubic distance (IPD) average was 5.1 cm (range 2.5 to 15cm) The immediate post-operative average was 2.4 cm (range 0.4 to 8cm) At long-term follow-up of an average of 5 years, the IPD was 4.7cm (range 3.5 to 7cm). The percentage of pubic approximation was 72% (range 36% to 140%). On antero-posterior radiographs and on 3D-CT of the pelvis, the best improvement of the pelvic anatomy was with the mid-iliac oblique osteotomy. (Figures 2,3 and 4)

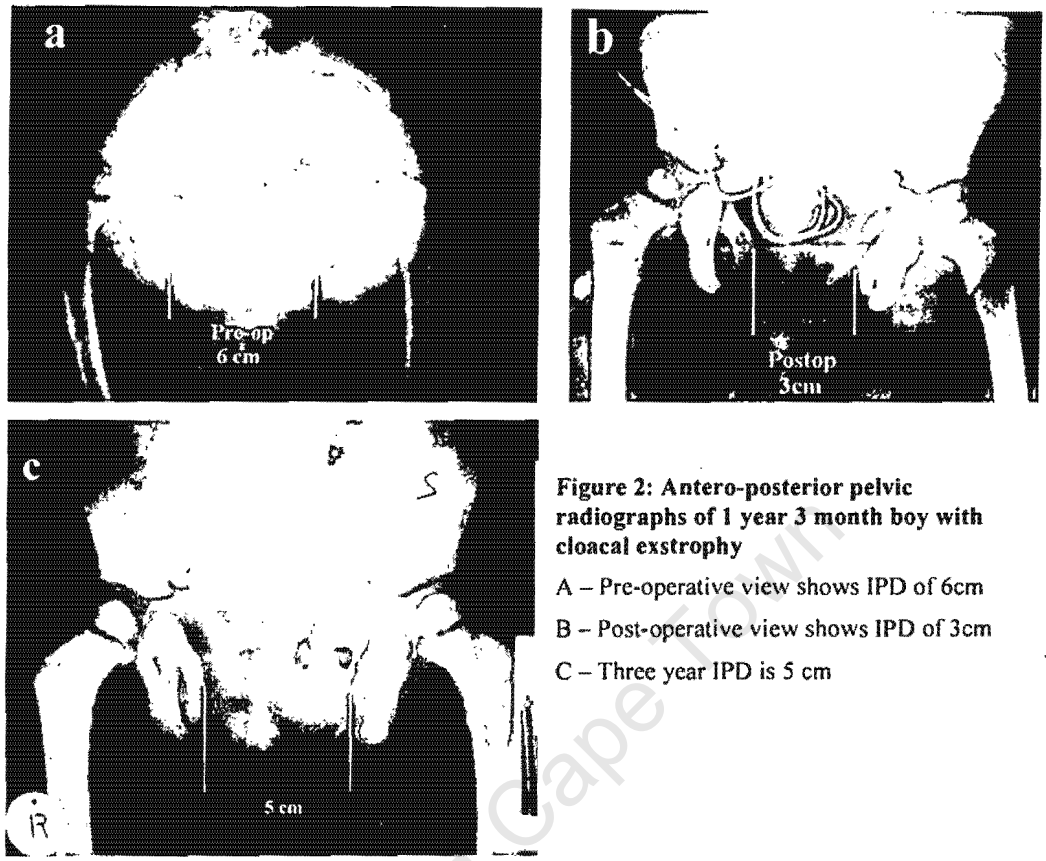


Figure 2: Antero-posterior pelvic radiographs of 1 year 3 month boy with cloacal exstrophy

A – Pre-operative view shows IPD of 6cm

B – Post-operative view shows IPD of 3cm

C – Three year IPD is 5 cm

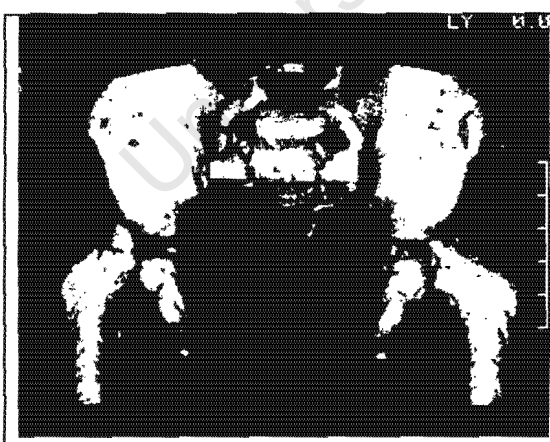


Figure 3: Pre-operative 3D-CT of patient in Figure 2

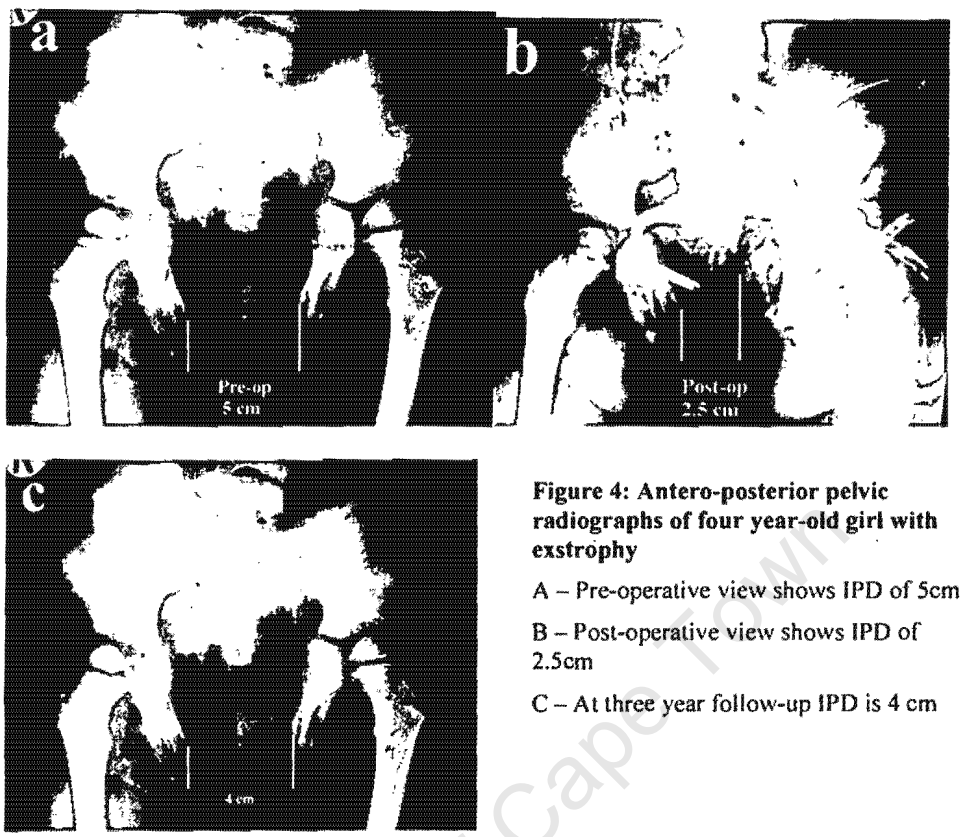


Figure 4: Antero-posterior pelvic radiographs of four year-old girl with exstrophy

A – Pre-operative view shows IPD of 5cm

B – Post-operative view shows IPD of 2.5cm

C – At three year follow-up IPD is 4 cm

Dehiscence and continence

The early experience at Red Cross was disappointing. All six initial patients who had primary bladder closure had either wound breakdown or sepsis. Of the other four, one had a normal bladder which was not repaired, one died of post-operative sepsis and the other two were so severe no repair or reconstruction was attempted. Of the early surgical attempts at bladder reconstruction, all had surgical failure with incontinence. One later gained continence following a bladder augmentation and a Mitrofanoff procedure. The only continent child was the one with the intact bladder and urethra in whom no reconstruction was performed.

The results later in this series were more encouraging. Two patients had no dehiscence. The third resolved following dressings and antibiotics. Two gained

functional continence, the third at short-term follow-up too young to be assessed. Although numbers are small, subjectively the mid-iliac oblique osteotomy provided the best bony facilitation of soft tissue closure. Table I summarises the surgical complications.

Table II: Outcome of 13 patients with respect to primary healing and continence

Patient	Exstrophy Type	Osteotomy	Age at operation	Dehiscence	Continence
1	Bladder	Nil	6 weeks	No bladder repair	Ileal conduit
2	Bladder	Post. Iliac	16 weeks	Yes	Sigmoid conduit
3	Bladder	Nil	3 days	+	Mitrofanoff
4	Cloacal	Post. Iliac	1 week	- No bladder repair – too small	Cut. ureterostomy
5	Cloacal	Ant. Pubic	1 week	+	Ileal conduit
6	Bladder	Ant. Pubic	2 weeks	+	Ileal conduit
7	Cloacal	Ant. Pubic	2 weeks	+	Cut. ureterostomy
8	Cloacal	Post. Iliac	1 year 3 months	- Normal bladder	Continent
9	Cloacal	Mid Iliac Oblique Mid Iliac Oblique	10 weeks 4 years	+ - Bladder excised	Ileal conduit
10	Cloacal	Mid Iliac Oblique	9 weeks	-	Died post - operatively
11	Bladder	Mid Iliac Oblique	3 days	-	-
12	Bladder	Mid Iliac Oblique	5 years	-	Continent
13	Bladder	? Elsewhere Mid Iliac Oblique	? 16 years	+	Continent

Gait and rotational profile

No patient had a Trendelenburg limp or wide based gait. There was no radiographic evidence of acetabular dysplasia. Nine patients were available for follow-up of more than two years. The recorded rotational profile is shown in *Table II.* Patients with a follow-up of more than 5 years have a normal rotational profile. As a trend, patients with a longer follow-up have increasing diastasis. The amount of external rotation is unrelated to the diastasis. The three patients with a follow-up of two years had a foot progression angle outside the normal range of 20° (normal 25-30) and correspondingly had external rotation of the hip of more than the normal value expected for age. The one patient with delayed closure at four years had a pre-operative foot progression angle of 30°, and external rotation of the hip of 80°. This improved to 15° and 45° respectively after a mid-iliac oblique osteotomy. The two patients who had no pelvic procedures had a normal rotational profile at follow-up of eleven and seven years respectively.

Table III: Rotational profile of nine patients available for follow-up of > 2 years

Patient	Osteotomy	Age at operation	Duration of follow-up	Age at follow-up	Foot progression angle°	Internal rotation of hip°	External rotation of hip°	IPD (cm)
1	Nil	6 weeks	11yrs	11yrs	15	40	50	8
2	Post. Iliac	16 weeks	8yrs	8yrs	20	30	50	4
3	Nil	3 days	7yrs	7yrs	15	50	50	7
4	Post. Iliac	1 week	2yrs	2yrs	30	30	80	4.5
5	Ant. Pubic	1 week	5yrs	5yrs	15	30	50	6
6	Ant. Pubic	2 weeks	5yrs	5yrs	20	40	50	6
7	Ant. Pubic	2 weeks	2yrs	3yrs	30	10	80	3.5
8	Post. Iliac	1 year 3 months	2yrs	2yrs	25	20	70	5
9	Mid Iliac Oblique Mid Iliac Oblique	10 weeks 4 years	4yrs 2yrs	4yrs 6yrs	30 15	20 45	80 45	6.8 2.3

Discussion

The approach to surgical management of the exstrophy complex remains a multidisciplinary challenge. It also represents an evolutionary process. In grappling with this problem in the early 1960's the consensus lay with urinary diversion[7]. The complications of this – electrolyte abnormalities, upper urinary tract infection leading to eventual renal failure, and carcinomatous change led to a strive towards reconstruction. Aadelen and O'Phelan published early results of long term follow-up of patients who had undergone staged reconstruction, rather than diversion, starting in the late 1950's[1]. Sixty nine cases were evaluated at 5 years. Of their patients 45% and 20% had a satisfactory and fair urological result respectively. They did note that interpubic distance (as a marker of pelvic approximation) correlated with urologic outcome. Ansell[19] in 1979 reported on 28 cases of exstrophy seen. He found that in the first 48 hours of life, the pelvic bones are still pliable and may be brought together by simply squeezing with hand pressure. Osteotomies were, however still required in 19 of the 28 cases. Thereafter staged closure of the exstrophy was performed. In his series half of the patients achieved continence, but only 30% of the classic exstrophies were considered a surgical success. An average of >5 major procedures were performed per patient. He noted that the patients in whom closure is obtained do better than those who undergo diversion.

Marshall and Muecke reviewed the literature from 1906 to 1966.[29] In 329 bladder closures, urinary continence was achieved in only 16 cases (5%). Major surgical complications including dehiscence, urinary fistulas, incontinence, persistent reflux and pyelonephritis frequently led to urinary diversion. These sobering longer term results were echoed by Woodhouse and Redgrave[30]. They noted that despite promising initial results continence deteriorated in adolescence. Half of the patients with early success required augmentation in adulthood. Forty four percent underwent urinary diversion, a urological failure Our own early experience at Red Cross Children's Hospital reflected these views. In all cases of bladder repair, we encountered wound dehiscence or incontinence. The only success in terms of continence had an intact bladder and was not repaired. Seven out of an initial nine patients had urinary diversion. In the latter period of this study, however the results

have been more encouraging. One further patient has had major wound complications, while a further 2 have not. Two are continent and the last is too young to assess.

This improvement in outcome is also reported in the literature. Jeffs popularised the staged approach to closure.[5,31,32] The bladder is primarily closed and osteotomies of the pelvis performed to support this closure. The pubic symphysis is approximated after the bladder closure is completed. Bladder neck reconstruction is performed once the bladder capacity is sufficient, usually at the age of 3. It includes bilateral ureteral reimplantation, bladder neck plasty and bladder neck suspension. A functional obstruction to outflow is created in order to stimulate bladder growth. Lepor and Jeffs report 86% excellent continence, and only 3% dehiscence.[32] A number of factors have been identified in preventing dehiscence. Closure as soon as possible after birth to minimise changes in the exposed bladder mucosa. Closure of the pelvis, either with manual pressure in the first 72 hours of life, or by means of a pelvic osteotomy thereafter. Post-operative immobilisation of the pelvis to prevent shearing of the bladder neck. Lastly urinary diversion to protect the recently closed bladder and abdominal wall.[17,5]

In contrast there is an approach which favours complete primary repair.[4,19,22,23] The advantage is in reducing the number of surgical events. The tissues are not scarred and mobilisation of structures may be easier. Gearhart et al[18] usually a proponent of staged reconstruction acknowledges there is a place for combined repair. They advise its use in older boys with failed exstrophy closure and in delayed primary closure where the bladder plate was unsuitable for newborn closure. In a separate study, however Gearhart reviewed 7 patients in whom complete repair had been performed. All had major complications including dehiscence, bladder prolapse, loss of urethra, and loss of penile skin. He notes that the complications are similar to those seen with staged repair, but that the additional loss of penile skin and loss of urethral closure makes later reconstruction more difficult.

The contribution of the orthopaedic surgeon to this complex multidisciplinary challenge is in providing pelvic support usually with an osteotomy. This has a number of important contributions. It restores pelvic floor anatomy and reduces shear

forces on the anterior abdominal wall thereby aiding bladder closure. It reconstitutes the shape of the anterior abdominal wall, enhancing normal bladder development. It has not in this study been shown to alter eventual outcome of gait patterns, nor have we noted any evidence of acetabular dysplasia needing correction.

The bony pathology in exstrophy has been studied by Sponseller et al[10], Yazici et al[12], and McKenna et al[11]. All three groups used CT of children with exstrophy and compared these with normal controls. A number of abnormalities were demonstrated. There is external rotation of the ilium on the sacrum. The ischium and pubis are further rotated externally on the ilium. There is apparent retroversion of the acetabulae. The ischium and pubis are shortened, contributing to the diastasis. This diastasis which is present at birth, increases with age. All the above deformities are accentuated in cloacal exstrophy. The apex of the bony deformity was felt to be at the level of the mid-ilium.

A number of surgical approaches and techniques to address the bony abnormalities have been described. Ansell[19] in 1979 reported on 28 cases of exstrophy seen. He found that in the first 48 hours of life, the pelvic bones are still pliable and may be brought together by simply squeezing with hand pressure. Osteotomies were, however still required in 19 of the 28 cases. It is not described in this study how this is performed. The symphyseal closure is held with figure of eight polypropylene sutures. Post-operatively the patients are bandaged for 6 months.

The posterior saggital iliac osteotomy is widely used. Popularised by Aadalen and O'Phelan at al[1], it was first described by Schultz in 1958.[6] Through a bilateral posterior incision a vertical osteotomy is performed just lateral to the sacro-iliac joint. The symphysis is held with a malleable wire suture. Post-operatively the patient is immobilised in a double hip spica. Aadalen and O'Phelan at al reported on the first long-term follow-up in 125 cases (65 longer than 5 years). Their average interpubic distance was 1.5cm and they felt that a small interpubic distance correlated well with a good urological outcome. The disadvantage with this approach is the need to turn the patient.

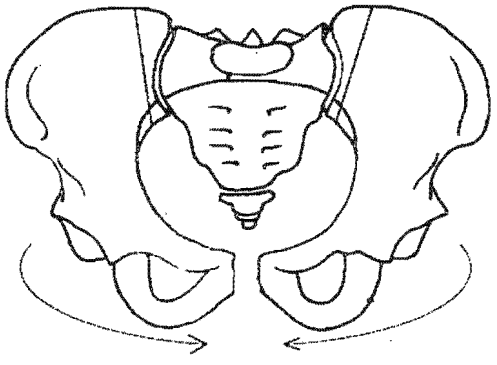


Figure 5: Schematic of posterior saggital iliac osteotomy

Frey and Cohen described an osteotomy of the superior ramus of the pubis. The medial aspects of both pubic rami are tilted towards the midline and approximated. The cartilaginous ischiopubic junction acts as an articulation and the rectus sheaths are sutured together with resorbable sutures. The major advantage is in avoiding the need to turn the patient and re-drape. Additionally they felt that the mobility gained with a posterior osteotomy was not always sufficient. Postoperatively a spica cast was used

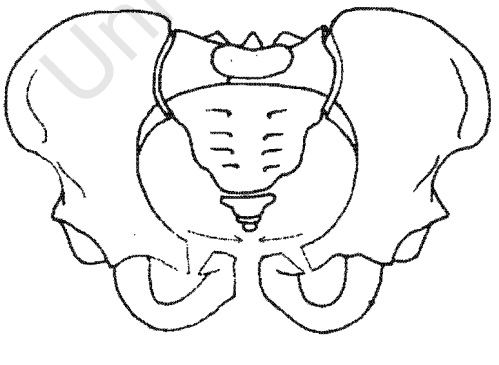


Figure 6: Schematic of anterior superior ramus osteotomy

In 1991 Sponseller, Gearhart and Jeffs reported on the use of a transverse iliac osteotomy. It utilises a similar surgical approach as used to perform a Salter osteotomy. A horizontal iliac osteotomy was made from the sciatic notch posteriorly, exiting anteriorly at a point midway between the anterior superior and the anterior inferior iliac spines. This is more cephalad than for a Salter osteotomy. The ischiopubic segments were internally rotated 30° to 45°, and the held with a small external fixator through both iliac blade and iliac body. In the older patient internal fixation to the symphysis was added They had reservations about the mobility of the pubis with the anterior procedure and occasional delayed or mal-union of the ilium with the posterior procedure. They also found it useful not to have to turn the patient and re-drape during the procedure. No mention is made of how to fix this osteotomy in very small infants. None of the patients the current study had this osteotomy.

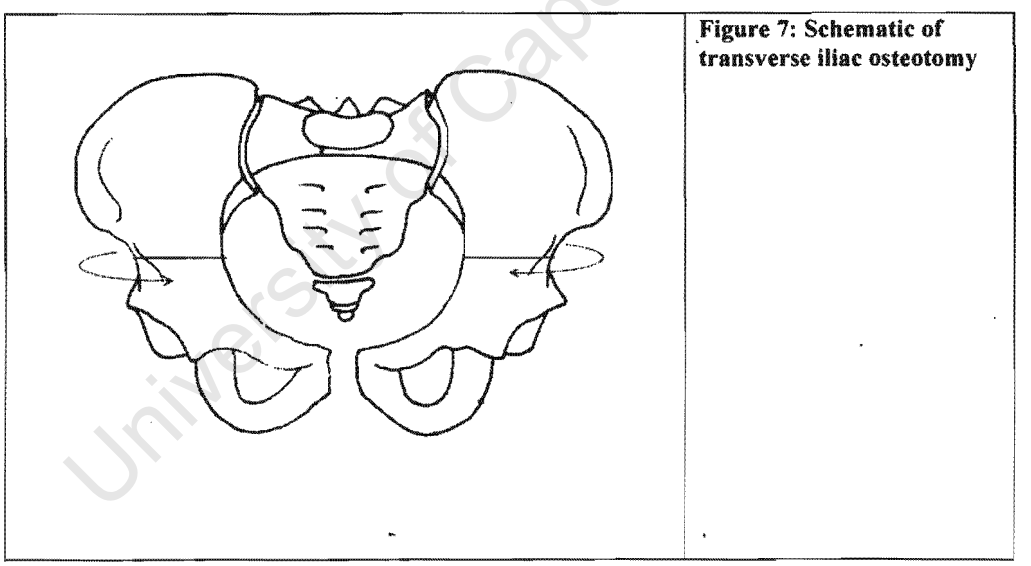


Figure 7: Schematic of transverse iliac osteotomy

McKenna and Wedge et al[11] developed a model based on 3D-CT of a three year old patient with classic exstrophy. On this model they performed a number of osteotomies including posterior, transverse, superior pubic and their own, the mid-iliac oblique. All but the anterior osteotomy corrected the diastasis. Their osteotomy addressed the apex of deformity. This is an oblique osteotomy made from the sciatic notch to a point on the iliac crest at the junction between the anterior third and the posterior two-thirds. The osteotomy is through the thinnest point of the ilium and is parallel to the arc of pelvic rotation. No mention is made of the method of fixation. Compared to the posterior and anterior osteotomy this obtains the most optimal transverse and anteroposterior diameter. They unfortunately describe no long-term follow-up. In our study this osteotomy provided the best radiographic improvement in pelvic anatomy.

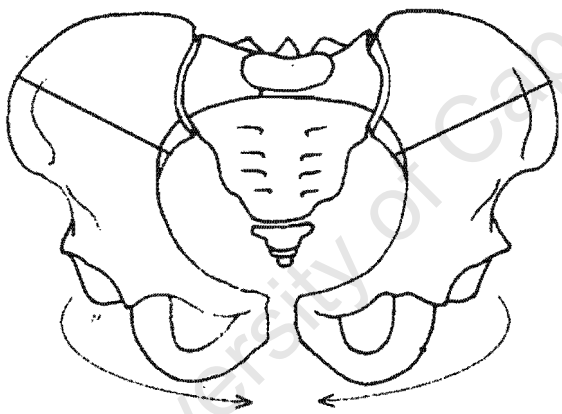


Figure : Schematic of mid-iliac oblique osteotomy

Facilitation of closure and pelvic anatomy

In this study, there was a progression from no osteotomy (2), to posterior iliac (3), to anterior pubic (3), to mid-iliac oblique (6). No transverse iliac osteotomy was done. This reflected both a progression in the literature of understanding of the management of exstrophy and local surgical expertise. All those who underwent osteotomy were assessed intra-operatively as having facilitated closure. The mid-

iliac oblique gave the best impression of aiding soft tissue approximation. The post-operative radiographs also showed that the best anatomic improvement was with the mid-iliac osteotomy. In this regard, osteotomies fulfilled their function. When the long-term results were assessed, the initial improvement in interpubic distance later deteriorated. The average pre-operative diastasis was 5cm, which improved to 2,4 cm post-op. At 5-year follow-up, however this had deteriorated to 4.6 cm. This gradual deterioration in the diastasis has been noted before by a number of authors.[34,36,37,38]. Kantor et al[38] in a 13 year follow-up noted a mean pubic diastasis of 6.4 cm (range 4-11.5 cm). There were no clinical problems associated with the diastasis. Schmidt et al39 noted that most patients eventually developed diastasis equivalent to the width of their sacrum. They did not have any associated functional defect. Sutherland et al found that the diastasis increased with age and in patients who had not had an osteotomy

The reasons for the inevitable increase in pubic diastasis with age are threefold. Firstly, as shown by Sponseller et al[10] the mesodermal deficiency, especially shortening, in exstrophy affects mostly the anterior segment. This growth deficiency will increase with age. Ongoing growth will therefore worsen the defect. In contrast in a study in patients without a mesodermal defect by Verrier et al[13] this progression did not occur. These patients in whom osteotomies were performed as part of separation of ischiopagus twins, had marked pre-operative diastasis. In patients who have pelvic lengthenings performed utilising distraction callotasis this recurrence of diastasis still occurs.[40]

Secondly, the type of osteotomy performed may not have addressed both the posterior segment pathology (external rotation) and anterior segment pathology (external rotation and shortening). On plain radiographs and on 3D-CT, the best reconstitution of the pelvic anatomy was with the mid-iliac oblique osteotomy, and this is our current preference

Lastly, the post-operative fixation of the osteotomy may not have been sufficient. Initially in this series, the symphysis was held with PDS sutures and the patients swaddled. Later, an external fixator was applied in the larger children or, in neonates plaster of Paris immobilisation used. Of the three possibilities, it seems most likely that the anterior mesodermal deficiency plays the biggest role. Post-operative immobilisation is the one factor we are able to address and the current preference is

to utilise external fixation where possible.

Table IV: Methods of fixation used by Various authors		
Authors	Fixation	Immobilisation
Aadelen and O'Phelan[1]	Symphyseal wiring	Spica for 4 months
Scherz, Kaplan et al[24]	Fascia lata	Bryant's Traction
Allen et al[25]	Polyglyconate suture	Nil
Frey and Cohen[41]	Polyglyconate suture to rectus	
Schmidt et al[39]	Non-absorbable suture	Nil
Sponseller[34,42]	External fixation Anterior internal fixation in children >8	External fixation
Kandemir et al[40]	External fixation	External fixation

Wound integrity and continence

The incidence of primary healing versus dehiscence and continence versus incontinence vary considerably in the literature. Aadalen and O'Phelan et al[1] in one of the earliest long term studies had 95% primary healing. Of 69 patients followed up for longer than 5 years, 45% were continent. Continence correlated well in this series with an IPD of less than 2 cm. Sponseller, Gearhart and Jeffs et al[4], however, found no correlation between continence and pubic diastasis. Lepor and Jeffs report 86% excellent continence, and only 3% dehiscence.[32] In contrast, Woodhouse and Redgrave[30] found that with longer follow-up, the initially promising results deteriorated with 44% of patients undergoing urinary diversion. While the examination of all urological factors related to satisfactory results is beyond the scope of this study, most authors agree that pelvic osteotomy plays an important role in successful urologic outcome.

Gait, rotational profile and acetabulum

Both Green et al[2] and Gokcora and Yazar et al[43] have noted an association with exstrophy and acetabular dysplasia. In this study we found no significant acetabular

dysplasia, concurring in this with Kantor et al[38] and Sutherland et al[37] who also found no evidence of dysplasia. In addition, none of our patients had a waddling gait.

In untreated patients with exstrophy, there is increased acetabular retroversion of 13 degrees as a result of the additive effect of both anterior segment (30°) and posterior segment (12°) external rotation. As a result all untreated patients have increased external rotation of the hip on examination and an externally oriented foot progression angle when walking. This is especially so under the age of 2 years. [10] The pelvic osteotomy corrects this externally rotated gait as illustrated by patient 9 in Table III. This patient had delayed closure at 4 years with improvement of the pre-operative foot-progression angle of 30° to15° and external rotation of the hip from 80° to 45°.

With the increasing diastasis with increasing age previously mentioned, a similar increase in external rotation of the hip would be expected. This is not the case. Cracchiolo and Hall[44] and Schmidt et al[39] found increased external rotation of the hip, but no functional or cosmetic impairment. Sutherland et al[37] failed to find a waddling gait. In their study the patients tended to have more external than internal rotation. Hip external rotation was not related to the degree of diastasis. The majority of patients grew to develop a normal gait. While the foot progression was increased by 10°, this was still within normal limits as described by Staheli et al.[45] Kantor et al,[38] in a long-term study (average 13 years) found marked pubic diastasis, but no associated clinical problems associated with it. He performed CT scans on 7 patients and found remodelling of the femora and acetabula. This accounts for the invariable return to near normal hip function and gait. The present study confirms these findings. The amount of diastasis did not correlate with external rotation of the hip Older patients had the most diastasis, but those with a 5 year or longer follow-up had normal hip function and gait.

Conclusion

The purpose of pelvic osteotomy is to facilitate soft tissue closure. It may have a role in improving outcomes in continence. The success of both soft tissue healing and continence continues to improve with evolving knowledge and surgical skill. In this study the best reconstitution of pelvic anatomy was achieved with mid-iliac oblique osteotomy. Fixation of the pelvis with rectus fascia and symphyseal suture should be augmented with external fixation when feasible.

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