

COMPLETE PRIMARY REPAIR OF EXSTROPHY

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ABSTRACT

Purpose: The surgical correction of bladder exstrophy to achieve continence with voiding remains a challenging problem for the urologist. Since 1989 we have performed complete primary repair for exstrophy based on the concept that the primary defect of bladder and cloacal exstrophy is anterior herniation. Thus, the bladder and urethra must be treated as a single unit to move them posteriorly into the pelvis. We present this technique.

Materials and Methods: From 1989 to 1997, 18 patients with bladder exstrophy and 6 with cloacal exstrophy underwent complete primary repair of exstrophy. This procedure was done on day 1 of life in 18 patients. Mean followup is 44 months (range 4 months to 8 years).

Results: At a median followup of 48 months 4 boys and 4 girls have volitional voiding after complete primary repair of bladder exstrophy, 21 patients have continent intervals and 2 boys void with continent intervals after complete primary repair of cloacal exstrophy. No patient has had a loss of renal function in this series. Postoperative complications included urethrocuteaneous fistula formation in 2 cases. No patient had primary closure dehiscence.

Conclusions: The rate of urinary continence achieved with complete primary repair compares favorably to that of staged repair for exstrophy. Complete primary repair also minimizes the number of surgical procedures required to achieve urinary continence and potentiates bladder neck function that permits bladder cycling in year 1 of life. The complication rates of these techniques are significantly lower than those reported in previous series of primary closure of exstrophy.

KEY WORDS: bladder; abnormalities; surgical procedure, reconstructive

"The repair of congenital defects of the urethra and bladder constitutes one of the most difficult and for that reason perhaps one of the most interesting chapters in plastic surgery, and if the outcome be successful it may be regarded with intense satisfaction. During the past fifty years many surgeons have occupied themselves with the solution of this problem, and many have been the attempts to devise a method by the aid of which, even in the severe types of the deformity, a restoration of the normal bladder form and normal bladder function could be secured. Up to the present time, however, this ideal has not been reached."¹

F. Trendelenberg, 1906

Despite the efforts and ingenuity of various surgeons during the last 200 years, to our knowledge no ideal treatment exists for bladder exstrophy. Professional opinion regarding the surgical treatment of bladder exstrophy ranges from early urinary diversion to reconstruction of the exstrophic bladder in a staged or primary fashion.^{2–4} Initial efforts to repair the exstrophic bladder focused on primary reconstruction. The report of Young in 1942 of success using primary bladder reconstruction in a young girl suggested that reconstruction was feasible in exstrophy and epispadias.⁵ However, success in this endeavor was limited, leading to the abandonment of bladder reconstruction in favor of urinary diversion with ureterosigmoidostomy in the first half of this century. Early efforts at primary reconstruction were described as an all-in-one surgical tour de force at 1 planned stage, which included primary bladder closure, bladder neck reconstruction, abdominal wall closure and epispadias repair.⁴ A continence rate of less than 20% was reported for most of these techniques and complications secondary to infection or urinary tract obstruction were common.⁶

A sequential approach to bladder exstrophy repair was

subsequently developed by others and popularized by Jeffs, and so staged reconstruction has consequently been referred to as the Jeffs approach.^{7–9} Results of this type of exstrophy repair include improved rates of renal preservation and urinary continence compared to those of earlier 1-stage efforts. This approach to reconstruction of the exstrophic bladder is currently the most popular.¹⁰ However, recent efforts have focused on reconstruction of the bladder in a primary 1-stage fashion based on the principle that exstrophy represents anterior bladder herniation.¹¹ Therefore, repair demands aggressive posterior repositioning of the bladder, bladder neck and urethra. The bladder, bladder neck and urethra are considered 1 unit and no initial effort is made at bladder neck reconstruction. Mitchell and Bagli initially reported the penile disassembly (Mitchell) technique for epispadias, and demonstrated that the penis may be separated into its 3 components (2 corporeal bodies and the corpus spongiosum) and then reassembled.¹² This technique facilitates complete anatomical reconstruction of the bladder in the newborn period by optimizing surgical mobilization of the proximal urethra and bladder neck. Using this technique primary closure in the neonate optimizes the chance for early bladder cycling and potentiates bladder development. It may also obviate the need for multistage repair of bladder exstrophy, including further bladder neck reconstruction and penile reconstructive surgery. The goal of this complete primary repair approach is to combine the goals of staged reconstruction at a single operation, that is bladder closure, epispadias repair and the achievement of urinary continence.

MATERIALS AND METHODS

We retrospectively reviewed a consecutive series of patients with exstrophy who were admitted to our institution for primary repair. Between 1989 and 1997, 24 patients with cloacal (6) or bladder (18) exstrophy were hospitalized for

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primary exstrophy repair using an extension of the Mitchell complete penile disassembly technique in boys and analogous aggressive dissection in girls.

In male patients complete primary exstrophy repair was performed. After standard sterile preparation of the operative field 3.5F umbilical artery catheters are placed into each ureter and sutured in place with 5-zero chromic suture. To aid in dissection, transverse oriented, 4-zero polyglecaprone traction sutures are placed into each hemiglans of the penis. The bladder plate and urethral plate are stained with a marking pen to assist in differentiating the urothelium from adjacent epithelium. Dissection of the epithelium from the urothelium starts at the most superior aspect of the bladder plate and progresses inferior toward the bladder neck. We do not divide the urethral plate, since this may completely disrupt the main blood supply to the distal urothelium and corpus spongiosum, precluding the complete penile disassembly technique. Furthermore, division of the urethral plate is not necessary in this technique because the bladder and urethra are moved posterior in the pelvis as a unit.

Dissection continues as a circumcising incision along the ventral aspect of the penis between Buck's fascia and the overlying tissue, and should precede dissection of the urethral plate from the corporeal bodies. Since we most easily identify Buck's fascia ventrally, that is where we prefer to begin the circumcising incision. We routinely inject the surrounding tissues with 0.25% lidocaine and 1:200,000 units per ml. epinephrine to limit blood loss and assist in dissection. The urethral plate is dissected proximal to the bladder neck. It is important to avoid narrowing the urethral plate because it is tubularized later in the operation. To preserve the blood supply all spongiosal tissue remains with the urethral plate. Inclusion of the whole corpora spongiosa with the urethral plate gives this unit a triangular appearance. The neurovascular bundles are lateral to the lateral edge of the urethral plate within Buck's fascia in exstrophy.¹³ As in any penile surgery, careful dissection is required during separation of the penile shaft skin from the corporeal bodies laterally. The medial plane of dissection is the tunica albuginea of the corpora. This plane is followed proximally to the intersymphyseal ligament (anterior coalescence of the pelvic diaphragm).

This dissection allows the penis to be disassembled into 3 components (the right and left corporeal bodies with the respective hemiglans, the urethral plate and the corpora spongiosa). This dissection is easiest to initiate proximal and ventral at the level of the tunica albuginea on the corpora, which is a relatively avascular plane. Bleeding usually indicates injury to the corpora spongiosa or corpora cavernosa. After a plane is created between the urethral plate and corporeal bodies dissection is carried distal to divide the glans penis in the midline, which allows the 3 components to be separated, as previously described.¹² The independent blood supply of these 3 components allows this separation. The hemiglans exist on a separate blood supply based on the paired neurovascular bundles. The underlying corpora spongiosa must remain with the urethral plate. The blood supply to the urethral plate is based on this corporeal tissue, which is wedge shaped after it is dissected from the adjacent corpora cavernosa. This urethral corporeal component is later tubularized and placed ventral to the corporeal bodies.

Proximal dissection of the urethral plate from the corporeal bodies is critical to posterior placement of the bladder neck and proximal urethra. Incomplete posterior dissection of the bladder and urethral plate or inadequate division of the intersymphyseal ligament creates anterior tension along the urethral plate and prevents posterior movement of the bladder, bladder neck and urethra in the pelvis, which increases the likelihood of dehiscence and likely jeopardizes later urinary continence. Aggressive proximal dissection along each side of the urethra and deep incision of the intersymphyseal ligament posterior to the urethral plate allow the bladder to achieve a posterior position in the pelvis (fig. 1).

After the bladder and urethral plate are adequately dissected they are closed as a continuous unit (fig. 2, A). We routinely divert urine through a suprapubic tube brought out through the umbilicus. The edges of the bladder plate are reapproximated using a 3-layer closure with monofilament absorbable suture. The urethra is tubularized using a 2-layer running closure with monofilament suture. No special effort is made to narrow the bladder neck. However, there should be no step-off at the bladder neck when incision of the pelvic diaphragm is performed adequately. The ureteral catheters are brought out through the urethra.

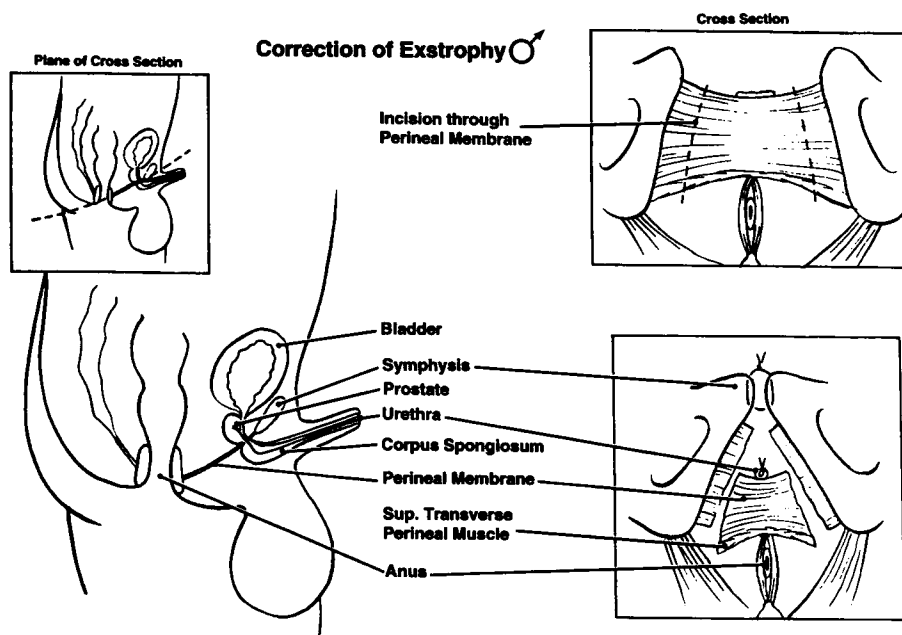


FIG. 1. Pelvic view of male exstrophy repair. *Sup.*, superior

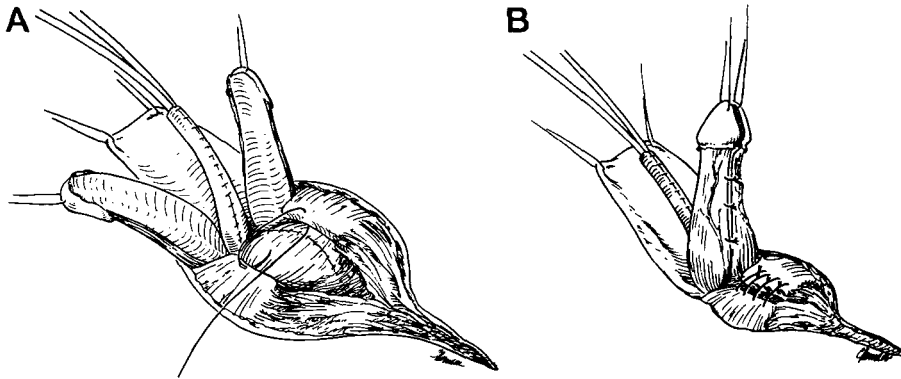


FIG. 2. A, closure of urethral plate and bladder as continuous unit. B, placement of urethra ventral to corporeal bodies via positioning of bladder, bladder neck and urethra posteriorly in pelvis.

We do not perform ureteral reimplantation at primary repair because the exstrophic bladder is still immature. However, vesicoureteral reflux is assumed to exist in these patients until proved otherwise. The corporeal bodies tend to rotate medial, which assists in correcting dorsal chordee and may be readily appreciated by observing how the horizontally placed glans traction sutures lie vertically. Usually the lateral aspect of the urethral plate is dissected off of the corpora toward the midline. The corpora are reapproximated with fine interrupted suture along the dorsal aspect. The tubularized urethra is then brought up to each hemiglans ventrally to create an orthotopic meatus (fig. 2, B). The glans is reconfigured using deep interrupted polydioxanone mattress sutures followed by horizontal 7-zero monofilament mattress sutures to reapproximate the glans epithelium. The neourethra is matured with 7-zero braided polyglactin suture, similar to our standard hypospadias repair. If necessary, glans tissue reduction is performed to create a conical appearing glans. We routinely note excess tissue at the base of the glans dorsally, which should be trimmed. Occasionally the urethra lacks sufficient length to reach the glans. In this situation the urethra may be matured along the ventral aspect of the penis, creating hypospadias that may be corrected later. We believe that this represents an inherent lack of length in the urethral plate and hypospadias results from aggressive posterior mobilization of the bladder and urethra. Redundant shaft skin is left in place ventrally in these patients to assist in later penile reconstructive procedures.

To reapproximate the pubic symphysis we use No. 0 or 1

polydioxanone interrupted sutures. Knots are placed anterior to help prevent suture erosion into the bladder neck. The rectus fascia is reapproximated using a running 2-zero polydioxanone suture. Penile shaft skin is reconfigured using primary dorsal closure or reversed Byars flaps if needed to provide dorsal skin coverage. Skin covering the abdominal wall is reapproximated using a 2-layer running closure of absorbable monofilament suture.

Postoperatively patients are maintained in Bryant's traction for 2 to 3 days and then fitted with an exstrophy splint to maintain hip adduction. We have also used spica cast immobilization for 3 weeks. Urethral catheters left in place and brought out through the urethra are removed 7 to 10 days after the operation. We do not use a urethral catheter.

Principles of closure in female patients are similar to those in male patients. Several points are important. The clitoris is separated, which precludes the need to disassemble it. The perineal incision must be extended around the vagina and deep into the pelvis in a Y-V advancement. The vagina and urethral plate are considered a single unit and never separated. Analogous to male repair, the pelvic diaphragm is incised deeply lateral to the vagina (fig. 3). Closure of the bladder, urethra and clitoris is analogous to bladder, bladder neck, urethral and penile repair in male patients.

RESULTS

From 1989 to 1997, 12 male and 6 female patients with bladder exstrophy, and 4 male and 2 female patients with cloacal

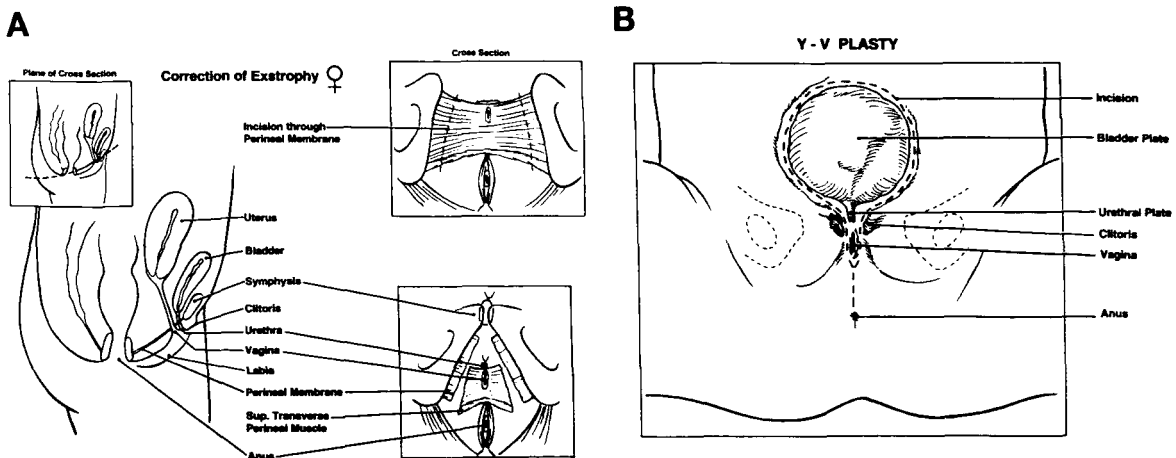


FIG. 3. Female exstrophy repair. A, pelvic view. Sup., superior. B, Y-V plasty

exstrophy underwent complete exstrophy repair as a primary procedure. This procedure was done on and after day 1 of life in 18 and 6 patients, respectively. One female and 2 male patients required medical stabilization before primary repair. In another patient closure was delayed because the condition was misdiagnosed as epispadias. Two patients underwent delayed repair secondary to nonmedical reasons. Notably the first 2 male patients in this series underwent the initial exstrophy operation as a portion of planned staged primary repair (table 1). A modification of the complete primary repair technique was used in these 2 cases but it represented an early version of the current procedure.

Median age of gestation at birth was 39 weeks. Apgar scores at birth ranged from 6/9 to 9/9. Two patients required intubation shortly after birth, of whom 1 was born at 25 weeks of gestation. The other patient was born at 35 weeks of gestation with significant concomitant congenital defects, including a tracheo-esophageal fistula, imperforate anus, diaphragmatic hernia and myelomeningocele. This latter patient subsequently died at age 2 months due to complications of pulmonary dysplasia.

Initial assessment preoperatively revealed a small bladder plate in 1 case, while in the remainder size was considered satisfactory. Operative time ranged from 2.4 to 5.1 hours (median 4.1 overall, and 4.7 and 3.2 in male and female patients, respectively). Estimated blood loss associated with this operation ranged from 10 to 100 cc (median 45). Osteotomies were performed in 1 patient in whom bladder exstrophy was repaired at age 9 months and in 3 in whom cloacal exstrophy was closed after 72 hours of life.

Followup in this group of patients ranges from 4 months to 8 years (median 48 months). Approximately half of the patients in this series have reached toilet training age. Serum creatinine levels available in 5 patients are within normal limits when corrected for age. Kidney development and growth have been followed by serial ultrasonography. Growth results on serial ultrasound have served as a surrogate for serum studies when serum studies were not available. Of the 18 patients with bladder exstrophy renal ultrasound was normal in 12 and 6 had mild hydronephrosis in 1 or each kidney at the most recent followup. It has been less than 2 years after the operation in all patients with hydronephrosis. Mild hydronephrosis was also noted in 4 other patients but it resolved on serial examination. In 1 patient ultrasound revealed evidence of severe hydronephrosis and subsequently a ureteral stricture was treated. In 3 patients with cloacal exstrophy ultrasound showed normal kidneys and 3 have mild hydronephrosis (table 2). Based on serial ultrasound and serum studies 98% of the renal units have no evidence of deterioration.

Vesicoureteral reflux documented in 12 of the 18 patients with bladder exstrophy is unilateral in 2 and bilateral in 10. In 2 patients voiding cystourethrography showed no reflux after primary exstrophy repair only. All patients with reflux are maintained on suppressive antibiotic therapy. However, despite suppressive antibiotic therapy febrile urinary tract infections have necessitated ureteroneocystostomy in 9 patients. In 6 patients who have not yet been evaluated for vesicoureteral reflux the condition is presumed to exist and they are maintained on suppressive antibiotic therapy. In the cloacal exstrophy group unilateral and bilateral reflux has been documented in 1 patient each, while 2 had no reflux

TABLE 1. *Timing of primary exstrophy repair*

Closure	No. Male/No. Female Pts.	
	Bladder Exstrophy	Cloacal Exstrophy
Newborn	10/5	2/1
Delayed	2/1	2/1
Totals	12/6	4/2

TABLE 2. *Radiographic studies*

	No. Bladder Exstrophy	No. Cloacal Exstrophy
Hydronephrosis on renal ultrasound:		
None	12	3
Mild	6	3
Severe	0	0
Reflux on voiding cystourethrography:		
None	2	2
Unilat.	2	1
Bilat.	10	1

after primary closure only based on voiding cystourethrography findings (table 2).

Urinary continence was defined as volitional voiding with dry intervals of 2 hours or more, and 4 boys and 4 girls who underwent primary closure of bladder exstrophy meet this criterion. Six patients with bladder exstrophy who underwent primary repair have not yet achieved toilet training. Of 2 boys with significant stress urinary incontinence 1 underwent delayed primary closure at age 9 months and 1 had an extremely small bladder plate at primary closure as a newborn. All 18 patients with bladder exstrophy in this series have documented dry intervals after primary repair. Two patients who underwent bladder neck reconstruction to improve urinary continence were the initial patients in this series in whom exstrophy closure was done as a planned staged procedure. One of these patients who underwent concomitant bladder augmentation and Mitrofanoff stoma construction now empties by intermittent catheterization. To date no patient with bladder exstrophy who underwent complete primary repair after these 2 boys has required a bladder neck procedure or bladder augmentation. Overall urinary continence in this group approaches 80% after toilet training, including 8 of 10 patients (4 girls and 4 boys) with bladder exstrophy who have reached an appropriate age for toilet training.

Of the patients with cloacal exstrophy 2 boys have achieved urinary continence, including 1 who had inclusion of the hindgut in the bladder at primary closure. Thus, effectively he underwent augmentation cystoplasty. One girl with dry intervals between voids is not continent, and 1 boy is not continent and does not have dry intervals (table 3). One girl with cloacal exstrophy required bladder neck reconstruction to achieve dry intervals.

Other procedures included inguinal herniorrhaphy in 4 patients performed as a separate operation. Also, we performed minor epispadias revision in each boy who underwent ureteral reimplantation as a concomitant procedure. Epispadias revision was done in these cases to correct dorsal chordee and/or improve the cosmetic appearance of the penis. Complications of primary exstrophy repair included urethrocuteaneous fistula in 2 cases, of which 1 did not respond to urinary diversion by catheter drainage and required operative closure. No patient had primary closure dehiscence. Two patients have recurrent episodes of cystitis and 13 have had episodes of pyelonephritis. One patient underwent nephrostomy tube placement to evaluate persistent hydronephrosis after ureteral reimplantation with subsequent ureteral

TABLE 3. *Urinary continence results*

	No. Bladder Exstrophy/Total No.		No. Cloacal Exstrophy/Total No.	
	Boys	Girls	Boys	Girls*
Urinary continence	4/12	4/6	2/4	0/1
Older than toilet training age	4/5	4/5	2/2	0/2
Dry intervals	12/12	6/6	2/4	1/1
Additional continence procedures	3†		1	1

* One girl died at age 2 months.

† Bladder neck reconstruction in 2 boys and augmentation cystoplasty in 1.

nent inflammatory cell infiltrate composed of plasma cells, lymphocytes and histiocytes, and occasionally neutrophils and eosinophils^{30,31} (fig. 1, C). At some sites, such as the lung, mesentery and retroperitoneum, the lesion may have an aggressive clinical course with multiple recurrence and even distant metastasis. The etiology of inflammatory myofibroblastic tumor remains unclear. The recent demonstration of lesion production and elevated serum concentrations of interleukins 1 and 6 associated with systemic manifestations in a child with a pulmonary inflammatory myofibroblastic lesion suggests that altered regulation of cytokine expression may have a role in pathogenesis.³² Others recently described heterogeneous clonal chromosomal aberrations or aneuploidy (hyperdiploidy) in some cases at some sites, excluding the bladder, and suggested that there may be a neoplastic basis for at least some cases of inflammatory myofibroblastic tumor.^{12,33-36} In the bladder this entity has been associated with urinary tract infection,^{1,4,14,15} or previous surgery or trauma^{4,15} but in most patients and especially in children no etiological factor has been determined.³

The most common forms of presentation of inflammatory myofibroblastic tumor in the bladder are hematuria and dysuria.^{3,4} Urinary tract infection is absent in most cases.⁴ In our series all 3 children presented with dysuria and urine culture was negative. Two children had gross hematuria, which was severe in 1. The diagnosis of a bladder mass was initially made by ultrasonography in all children. CT did not add significant information concerning the bladder mass but it helped to confirm that the lesion was confined to the bladder. Inflammatory myofibroblastic bladder tumors are usually located in the fundus, or lateral or posterior walls and in only 1 case has a tumor been described arising from the trigone.^{3,4}

Since all bladder inflammatory myofibroblastic tumors reported to date have been benign, the goal of treatment is tumor resection with bladder preservation. Therefore, although some have suggested open surgery as the initial approach, we think that the initial diagnostic surgical technique should be transurethral resection of the bladder mass.^{3,14} All of our patients were initially treated with endoscopy and they subsequently required open surgery. Partial cystectomy was done in 2 children due to intensive bleeding in 1 and suspicion of a malignant tumor in 1. The remaining child underwent open tumor debulking and unilateral ureterovesical obstruction to avoid a maiming procedure, as suggested by Freud et al.¹⁵ At the last followup all 3 children with an inflammatory myofibroblastic tumor were clinically asymptomatic, 2 were tumor-free on ultrasonography and 1 who underwent tumor debulking has had a stable residual mass for more than 3 years.

Differentiating these tumors from a malignant lesion may be difficult but it has important consequences for therapy. Inflammatory myofibroblastic masses have been misdiagnosed as rhabdomyosarcoma, leiomyosarcoma and lymphoma.^{13,37,38} Inflammatory myofibroblastic tumor and sarcoma may be similar in regard to clinical aspects, since each is a hemorrhagic, polypoid or myxoid lesion, and histologically tumor cells may resemble smooth muscle cells and infiltrate beyond the bladder wall.³⁷ The histological features that differentiate inflammatory myofibroblastic bladder tumor from sarcoma may include minimal nuclear atypia, normal mitotic figures not exceeding 5 per 10 high power fields, intense chronic or mixed inflammatory cell infiltrates, delicate granulation tissue-like vascularity and rare tumor necrosis.³⁸ Immunohistochemical staining may demonstrate variable expression of the cytostructural proteins vimentin, desmin and actin in myofibroblastic tumor and bladder sarcoma. Immunohistochemical studies must be interpreted with caution and within the context of other histopathological features. Only the second patient with a myofibroblastic tumor in our series was evaluated using immunohistochem-

ical studies, which revealed that the tumor contained spindle cells that were vimentin positive but actin negative and desmin negative. However, other adjunctive diagnostic modalities may provide more useful differential information. Some cases of embryonal rhabdomyosarcoma have displayed chromosome 2 trisomy³⁹ or loss of heterozygosity for chromosome 11p.⁴⁰ Flow cytometry of the myofibroblastic tumors has revealed a diploid cell population,³¹ and so aneuploidy may be an ominous sign.

Based on these observations we agree with others^{37,41} that the term pseudotumor has little scientific meaning and it is inappropriate for describing these inflammatory tumors. Although these lesions are not malignant, they have the potential for severe local destruction if not managed aggressively. We suggest that they should be termed inflammatory myofibroblastic tumors due to the usually intense inflammatory infiltration, and spindle cell myofibroblastic and fibroblastic proliferation with granulation tissue-like vascularity but no significant cytological atypia and no abnormal mitosis or extensive necrosis.

The etiology of inflammatory eosinophilic tumor also remains unclear. Allergic reaction with peripheral blood eosinophilia or previous trauma has been associated with some cases.⁴²⁻⁴⁷ The lesion usually presents with dysuria, frequency, hematuria and abdominal pain.^{9,27} Two of our children presented with all of these features. Microscopic hematuria was noted in only 1 child but urine culture was positive for bacteria in each. As in those with an inflammatory myofibroblastic tumor, ultrasonography was the imaging study of choice that initially detected the bladder lesions.

To our knowledge the optimal treatment of an inflammatory eosinophilic tumor has not been established. Various treatment modalities have been proposed, such as antihistamine drugs, steroids, surgical intervention in some cases^{27,42,48} and even no treatment, as suggested by Sutphin and Middleton.¹⁹ In our series we treated each patient surgically with transurethral resection of the lesion and necrotic material drainage. In 1 case endoscopic resection resolved the mass, as confirmed by followup ultrasonography. In the other case there was resolution after transurethral resection and open drainage of the tumor, which extended into the true pelvis. Neither child with an inflammatory eosinophilic tumor required open surgical resection within the bladder.

Inflammatory eosinophilic tumors may also be difficult to distinguish from malignant tumors, such as sarcoma,^{27,42,48} and from inflammatory reactive lesions, such as interstitial cystitis, bacterial, viral and tuberculous cystitis, and Langerhans' cell histiocytosis.^{9,19,27} Histologically the early lesion of inflammatory eosinophilic tumor is characterized by an intense inflammatory infiltrate containing eosinophils and lymphocytes with edema and congestion (fig. 2, C). The eosinophilic infiltrate is more prominent in cases of muscle necrosis. More chronic lesions tend to have fewer or even absent eosinophils, scarring of the lamina propria and superficial muscular interstitium, and proliferative or squamous metaplasia of the overlying epithelium. Although eosinophils may be present in the inflammatory myofibroblastic tumors, they are not prominent. Spindle cell myofibroblastic and fibroblastic proliferation confirms the diagnosis of the myofibroblastic rather than the eosinophilic subtype. When histological examination reveals multinucleate giant cells, despite the presence of eosinophils, the possibility of a granulomatous process may be considered, as in children with chronic granulomatous disease.⁴⁹

Eosinophilic bladder lesions may present as diffuse eosinophilic inflammation of the bladder wall without a mass, or as a nodular or sessile lesion.^{9,20,29} In our opinion the term eosinophilic cystitis should be reserved for cases of infiltrative and global cystitis without a mass. Cases with a mass should be referred to as inflammatory eosinophilic tumor.

stenting for a ureteral stricture, which has been successfully managed with ureteral balloon dilation.

DISCUSSION

The goals of bladder exstrophy reconstruction include preservation of kidney function, creation of urinary continence, decreased episodes of urinary tract infection, and creation of functionally and cosmetically acceptable external genitalia. Goals underlying the numerous operations for exstrophy repair have remained constant since the initial operations were proposed and attempted in the 1800s. Then and now operations for exstrophy include those designed to remove the exstrophic bladder and replace it with a form of urinary diversion, and reconstructive procedures designed to reconstruct the bladder in multiple stages or in 1 stage.

Reconstruction of the native exstrophic bladder requires autologous tissue that may grow and develop normally after surgical repair. It is generally believed that the exstrophic bladder is primarily abnormal. However, the muscarinic receptors of the exstrophic bladder are normal compared to those in control bladders.¹⁴ Furthermore, Toguri et al showed that some bladders may achieve normal detrusor contractions and the urethral sphincter may function in a normal and coordinated fashion after it is closed.¹⁵ The observation that some patients who have undergone primary bladder reconstruction have achieved urinary continence and volitional voiding without catheterization or sacrificing renal function also demonstrates that these bladders have the potential to achieve normal function.^{3, 4, 16}

Initial successes in exstrophy repair were marked by the observation that successful exstrophy closure was intimately associated with anatomical reconfiguration of the pelvic anatomy. Trendelenberg noted this in 1906, indicating that exstrophy repair failed because he did not reapproximate the pubic symphysis and pelvic diaphragm.¹ He advocated molding the infant pelvis with time to cause desired changes in the bony structures of the pelvis by orthopedic measures. Schultz rediscovered the benefit of iliac osteotomy in the plastic repair of exstrophy and in 1958 he described it in a case report of successful 1-stage exstrophy closure.¹⁷ Others, such as Ansell,¹⁶ also recognized the importance of true anatomical reconfiguration of the pelvic anatomy in the repair of exstrophy.

In the past the disappointing results of planned 1-stage reconstruction of the exstrophic bladder earned the operation a poor reputation that remains with it today. Series of the 1970s indicated 20 to 30% rates of renal deterioration after 1-stage repair.^{6, 18} King and Wendel noted a 90% incidence of kidney damage in their series.¹⁹ Continence rates were also suboptimal in these studies with results of 0 to 50% (average 20%) noted by most investigators.^{6, 18, 19} Because of these results and the increasing popularity of multiple stage reconstruction, many surgeons avoided further attempts at 1-stage reconstruction of bladder exstrophy.

Using a staged approach to reconstruct the exstrophic bladder urinary continence rates as high as 88% have been reported.^{7, 20, 21} However, others have been able to achieve continence rates of only 10 to 30% with this approach.^{22, 23} Preservation of kidney function by multistage reconstruction represents a significant improvement over previous efforts at bladder closure. Various series provide evidence of renal damage at a rate of 13 to 20% in this patient population based on radiographic evidence of hydronephrosis or persistently elevated serum creatinine.²¹⁻²³

Complete repair of the exstrophic bladder as a primary procedure offers many theoretical and practical advantages. Experimental studies in animal models suggest that mechanical force is necessary for bladder growth and regeneration.^{24, 25} Early reconstruction, including restoration of urinary continence, allows mechanical cycling of the bladder,

which does not consistently occur after multistage bladder exstrophy reconstruction. Interestingly the ability of the bladder to regain normal function decreases significantly if outflow obstruction persists beyond year 1 of life in patients with posterior urethral valves.²⁶ In turn this implies that the bladder progresses through developmental milestones in the first few months of life, which may be irreversibly lost if missed. Precedence for this form of organ development is noted in the brain with the acquisition of language and visual perception.²⁷ Primary complete repair of the exstrophic bladder in the newborn period may provide the best opportunity for normal bladder development, which would optimize the potential for urinary continence.

Our results of complete primary repair in the newborn period compare favorably to successful results of exstrophy closure using a staged reconstructive technique. Urinary continence rates approached 80% in our patients who have attempted toilet training. Dry intervals were noted in all patients with bladder exstrophy and 3 of the 4 with cloacal exstrophy. To date only 2 boys early in the series in whom initial closure was performed in anticipation of a staged approach have required later bladder neck reconstruction to treat urinary incontinence. To date no patient who underwent complete primary repair has required a bladder neck procedure to achieve continence.

We make no attempt to reconstruct the bladder neck at primary repair. We believe that proper posterior positioning of the bladder and urethra as a unit allows more anatomical reconstruction of the pelvic floor, which has contributed to the urinary continence rate in this population without the need for formal bladder neck reconstruction. This posterior positioning may only be achieved by effective dissection of the bladder and urethral plate from the adjacent tissue lateral to these structures. Posterior movement of the bladder neck and urethra also facilitates reapproximation of the pubic symphyses, which in turn helps to prevent anterior migration of the urethra and bladder neck, creating a more anatomically normal muscular pelvic diaphragm.

Complication rates also compare favorably to those reported in series of staged reconstructive procedures of the exstrophic bladder, and they are markedly improved from earlier reported series of 1-stage exstrophy repair. We did not observe deteriorating renal function in this series. Significant complications included episodes of pyelonephritis in 9 patients with vesicoureteral reflux, which necessitated ureteral reimplantation. Notably 4 patients in this series did not have reflux after undergoing complete primary repair only. Early postoperative complications included urethrocutaneous fistula in 2 cases. However, dehiscence was not noted in any patient in our series. We believe that posterior positioning of the bladder and urethra decreases the chance of this complication because it relieves anterior tension on the incision that is present if the urethra and bladder neck are not moved posterior. By using aggressive dissection the Mitchell technique decreases anterior tension on the urethra because it is dissected free from its attachments to the corporeal bodies, which are inferior and lateral to the urethra, and pushed anterior by the intersymphyseal ligament.

CONCLUSIONS

Complete primary repair of exstrophy as described decreases the number of procedures necessary to achieve urinary continence. It may also facilitate bladder development, obviating the need for bladder augmentation. Complications in our series were minimal compared to those of previous efforts at primary closure. Long-term data are necessary to compare the results of complete primary exstrophy repair and staged reconstruction. At this point 85% of our patients who are old enough to achieve toilet training have spontaneous volitional voiding with dry intervals of 2 hours or more.

No patient who has undergone complete primary repair as an initial procedure for bladder exstrophy has required bladder augmentation, urinary diversion or intermittent catheterization for bladder emptying.

The previous successes of others with 1-stage primary reconstruction of bladder exstrophy demonstrate that it may be done.^{1,4,16} Our efforts build on the recognition of a principal factor that successful treatment of bladder exstrophy involves the reconfiguration of pelvic anatomy in an anatomically normal fashion. The results of our series suggest that the application of these principles by extending the Mitchell complete penile disassembly technique to complete primary exstrophy repair may achieve reproducibly satisfactory results in cases of bladder and cloacal exstrophy.

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