THE FAILED COMPLETE REPAIR OF BLADDER EXSTROPHY: INSIGHTS AND OUTCOMES

JOHN P. GEARHART AND ANDREW D. BAIRD

From the Division of Pediatric Urology, Department of Urology, The James Buchanan Brady Urological Institute, The Johns Hopkins Hospital and Johns Hopkins School of Medicine, Baltimore, Maryland

ABSTRACT

Purpose: We describe the complications of complete repair and their management.

Materials and Methods: A total of 19 patients were referred after failed complete repair. Total dehiscence occurred in 6 males, major bladder prolapse in 3, minor prolapse in 3, pubic separation in 1, impassable stricture in 1, and total hemiglans and corporal loss in 2. Overall, partial glans loss was seen in 7 patients, urethral loss in 5 and penile skin loss in 3. One female had complete dehiscence and 1 had major prolapse, both losing the urethrovaginal septum. One female had an impassable stricture.

Results: Six males with dehiscence underwent re-closure with osteotomy. Urethral replacement was performed with full thickness skin graft (FTSG) in 3 and with buccal mucosa in 3. Five patients underwent a modified Cantwell-Ransley (C-R) epispadias repair after placement of skin expanders, and 1 awaits repair. The 3 patients with major prolapse underwent re-closure with osteotomy. A urethral buccal graft was used in 1 patient, FTSG was used in 2 at a later operation and all 3 underwent C-R epispadias repair. Of the 3 patients with minor prolapse 2 underwent re-closure with osteotomy using urethral buccal graft or FTSG followed later with a C-R repair. The final patient with minor prolapse underwent re-closure with osteotomy and C-R repair after testosterone stimulation. One patient with pubic separation and urethral and skin loss underwent re-closure with osteotomy, C-R repair after skin expanders and later bladder neck repair. In 1 case a ureteral graft replaced a posterior urethral stricture. Of the 2 patients with hemiglans and corporal loss 1 underwent penile torsion repair and later hypospadias repair, while the other is being observed. Two females underwent re-closure with osteotomy and urethral replacement with tubularized bladder. The case of stricture was managed endoscopically.

Conclusions: Complications of complete repair are similar to those of other repairs but more serious if soft tissue loss occurs. Because of these increased risks, this procedure and its formidable complications are best managed by experienced exstrophy surgeons.

KEY WORDS: bladder exstrophy, epispadias, complications

The description of complete penile disassembly by Mitchell and Bagli in 1996 generated interest in combining this technique with bladder exstrophy closure in the newborn. The subsequent report of 18 patients was encouraging but followup was short and long-term outcomes were unknown. As more units began attempting this repair some early reports of complications appeared. Recently, concern was raised about loss of penile and corporal tissue associated with this method. Since complications can occur with any method of exstrophy closure, only time will establish the exact nature of such problems in those who undergo the complete repair and how they are to be treated most effectively. While all methods of exstrophy repair have merits, the quality and size of the bladder template, size of the phallus, depth of the urethral groove, width of pubic diastasis, and experience of the surgeon and associated staff still determine the outcome in all methods of repair. 

MATERIALS AND METHODS

Patients were identified from the institutional review board approved exstrophy/epispadias database at our institution, which is regularly updated and contains data on more than 830 patients seen since 1975 with classic exstrophy, cloacal exstrophy and epispadias. Sixteen males and 3 females have been referred to our unit after complete primary bladder exstrophy repair (CPRE) as newborns since 1996. In all patients closure was performed during the first week of life and in the first 48 hours of life in 9. None of the patients underwent pelvic osteotomy at the time of primary closure. Also, no external fixators or traction devices were used after reconstruction. Spica casts were used in 13 patients and “mummy wrap” was used in 6. Exstrophy closure was performed by a fellowship trained pediatric urologist in 13 patients, a nonfellowship trained pediatric urologist in 4, a general urologist in 1 and a general pediatric surgeon with an interest in pediatric urology in 1.

All cases had a pubic diastasis at the time of referral of at least 4 cm. Total dehiscence of the repair occurred in 6 males, major prolapse (bladder mucosa permanently seen protruding from a wide open bladder neck) in 3 (1 of whom had undergone 2 prior closures), minor prolapse (protrusion of bladder mucosa via bladder neck on straining) in 3, pubic separation in 1, impassable stricture in 1, and hemiglans and corporal loss alone in 2. Overall, partial loss of the glans was seen in 7 patients, urethra in 5 and penile skin in 3. Examples of boys with bladder prolapse/dehiscence and loss of hemiglans and urethral plate are shown in figures 1 and 2. One female had complete dehiscence and 1 had major prolapse, both losing tissue in the urethrovaginal septum. An impassable urethral stricture developed in 1 female.

RESULTS

Six males with dehiscence underwent re-closure with combined anterior innominate and vertical iliac osteotomy along with an external fixation device and modified Buck’s traction. All patients remained re-closed after secondary repair. Full
thickness skin graft (FTSG) was used in 3 patients and buccal mucosal graft in 3. A modified Cantwell-Ransley (C-R) epispadias repair was done in 5 patients and 1 awaits repair. All 5 patients had placement of tissue expanders 6 weeks before epispadias repair. One urethrocutaneous fistula and 1 meatal stenosis responded to repair.

Three males with major prolapse underwent re-closure with bilateral innominate and vertical iliac osteotomy along with an external fixator and modified Buck’s traction. A buccal graft was used for urethral replacement at the time of re-closure in 1 case and a FTSG in 1. All 3 patients underwent modified C-R repair without complication. In 1 case re-closure and modified C-R repair were combined along with bilateral innominate and vertical iliac osteotomy after testosterone stimulation.

The patient with pubic separation and urethral and penile skin loss underwent re-closure and bilateral anterior innominate and vertical iliac osteotomy with an external fixator and modified Buck’s traction. Modified C-R repair was undertaken 6 weeks after placement of skin expanders without complications. This patient has undergone bladder neck repair and is currently wet at night and dry for 2 hours during the day. In 1 patient a urethral graft replaced a posterior urethral stricture. Of the 2 patients with hemiglans and corporal loss 1 underwent penile torsion repair and recent hypospadias repair, and the other is being observed. The results achieved in these 15 male patients are summarized in the table.

Two females underwent re-closure with bilateral innominate and vertical iliac osteotomy with an external fixator and modified Buck’s traction. In both cases loss of soft tissue in the common septum between the urethra and vagina was observed, which was believed to be due to exuberant soft tissue dissection during initial CPRE. Urethral replacement was accomplished at the time of re-closure with tubularized bladder mucosa to provide urethral continuity and allow soft tissue interposition between the urethra and vagina. A dense 1 cm stricture in 1 female was endoscopically treated with direct visual urethrotomy and intermittent catheterization.

**DISCUSSION**

Regardless of the type of repair chosen in the newborn period for exstrophy repair, certain long-standing principles remain, including radical dissection of the posterior urethra and bladder from surrounding pelvic structures especially the urogenital diaphragm; careful and precise suturing of the bladder, posterior urethra and last few cm of the penis if complete repair is attempted; firm closure of the abdominal fascia and pubis; and stable immobilization of the repair and protection from distracting forces of movement during heal-

<table>
<thead>
<tr>
<th>Presentation</th>
<th>No. Pts</th>
<th>Osteotomy (closure successful)</th>
<th>Urethral Replacement (No.)</th>
<th>Comments (No.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Males:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complete dehiscence</td>
<td>6</td>
<td>6 (6)</td>
<td>FTSG (3), buccal graft (3)</td>
<td>Fistula repaired (1), meatal stenosis dilated (1)</td>
</tr>
<tr>
<td>Major prolapse</td>
<td>3</td>
<td>3 (3)</td>
<td>FTSG (2), buccal graft (1)</td>
<td>Fistula spontaneous closure (1)</td>
</tr>
<tr>
<td>Minor prolapse</td>
<td>3</td>
<td>3 (3)</td>
<td>FTSG (1), buccal graft (1)</td>
<td></td>
</tr>
<tr>
<td>Pubic separation</td>
<td>1</td>
<td>1 (1)</td>
<td>Ureteral graft</td>
<td></td>
</tr>
<tr>
<td>Impassible stricture</td>
<td>1</td>
<td>1 (1)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Females:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complete dehiscence</td>
<td>1</td>
<td>1 (1)</td>
<td>Tubularized bladder</td>
<td>Managed endoscopically + clean intermittent catheterization</td>
</tr>
<tr>
<td>Major prolapse</td>
<td>1</td>
<td>1 (1)</td>
<td>Tubularized bladder</td>
<td></td>
</tr>
<tr>
<td>Impassible stricture</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
ing. The importance of a successful primary closure has been demonstrated by the fact that if the primary closure fails only 50% of those patients ever get an adequate bladder capacity and only half of those are ever dry. \(^7\) Bladder prolapse, dehiscence, bladder calculi and multiple closures must affect the long-term outcome of the bladder whatever the type of repair. While we remain unconvinced that penile disassembly is required to move the posterior vesicourethral unit into the pelvis, whichever method is chosen must be protected in the postoperative state so that proper healing can occur.

An early neonatal closure without the need for an osteotomy is the ideal situation and this occurred within 48 hours in 9 of our patients (all closures were performed within the first week of life). No patient had an osteotomy at the time of primary closure. With very young premature cases, we have found the use of spica cast to be the standard method of immobilization through osteotomy. In a recent review by Meldrum et al the success rates in a large number of "mummy wrap" in 6, failures still occurred. In a recent review primary closure, although one with major prolapse had an osteotomy and by Meldrum et al the success rates in a large number of patients were nearly 75% in those with an osteotomy and modified Buck's traction, and 97% of second closures treated by Meldrum et al the success rates in a large number of patients were 93% in those with an osteotomy and modified Buck's traction at the time of secondary closure before referral. Unfortunately, with the use of a Spica cast in 13 cases and "mummy wrap" in 6, failures still occurred. In a recent review by Meldrum et al the success rates in a large number of primary closures were 95% in those with an osteotomy and modified Buck's traction, and 97% of second closures treated in the same fashion. \(^1\) Success rates were nearly 75% in those patients closed early without osteotomy but with modified Bryant's traction for 4 weeks. Also, primary closure failed in almost all patients without an osteotomy who were treated with only a Spica cast or "mummy wrap". \(^1\) While this was a select group of referred exstrophy failures, regardless of the method of repair chosen, proper immobilization with or without osteotomy depending on the pelvic diastasis and mobility is mandatory if proper healing is to occur. However, it is noteworthy that in reported successful series of CPRE the spica cast has been used as the standard method of postoperative immobilization when osteotomy has not been performed. \(^2\) \(^12\)

Special concerns other than dehiscence and prolapse must be appreciated with complete repair in the newborn. Loss of glans and/or corporal tissue is devastating to this particular group of patients with their inherent paucity of length and size. \(^13\) Two recent reports draw attention to these complications and include cases from other centers where a large exstrophy volume is seen. Hammouda reported ischemic changes in the glans in 5 of 21 patients who underwent penile disassembly along with exstrophy closure. \(^6\) Recently, Husmann and Gearhart reported on 9 patients (6 of whom are included in the present series) who suffered losses ranging from hemiglans and urethra in some to loss of both hemiglans, corpora and urethra in others. \(^7\) Speculation of the reason for these complications included technical misadventures, induction of venous congestion/arterial spasm or disruption of a congenitally abnormal blood supply. \(^5\)

Certainly some factors can come into play that may harm the small blood supply of the infant penis. The use of vasoconstrictive substances should be avoided at all costs as intractable vasoospasm could result. One of us (JPG) has not found penile vasocongestion to be a problem in any closure when an osteotomy has been performed, suggesting that perhaps osteotomy affords approximation of the pubis without undue pressure on the venous outflow of the corpora and avoids vascular compromise. It is interesting that in the previously cited article 2 patients sustained venous congestion with pubic reaproximation yet on release and resuturing of the pubis the congestion abated but the glanular blood supply remained at risk. \(^5\) Certain surgical handling of these tiny structures could cause endothelial damage with resultant thrombus and tissue loss. Lastly, successful reconstitution of the epispadiac penis relies on the consistency and preservation of its neuromuscular supply, and it is known that up to 20% of boys will have a congenital variant of the usual supply when 1 or more of the terminal arteries will meet the surface of the corpora at the junction of the corpus spongiosum and cavernosum, and form a variant accessory pudendal artery located medially on the anterior prostate surface. \(^6\) These terminal arteries are often difficult to reliably identify intraoperatively and, while usually anatomically consistent, could be located more medially and be damaged during dissection.

Loss of the distal urethral plate may result from arterial spasm or damage to the terminal arteries of the penis during dissection. The resultant acquired hypospadiac position of the urethral meatus will often require a further surgical procedure for the meatus to reach the tip of the glans. Loss of the entire urethral plate as seen in 5 patients may occur from overzealous dissection and damage to the bulbary artery as it exits into the corpus spongiosum from the common penile artery. We avoid this area in our modified C-R repair by staying on the bony aspect of the corpus, and moving cephalad towards the urogenital diaphragm but medial to the neurovascular bundle, thus remaining between the bundle and the arterial supply to the corpus spongiosum and urethra. The last remnants of the urogenital fibers are taken down by distracting the pubic bone outward, bringing any remnants of the diaphragm easily into view and allowing for their safe incision between the corpus and inferior pubic rami.

With loss of the urethral plate and significant amounts of penile skin other sources of replacement tissues must be found. Of our patients 5 lost the entire urethral plate requiring replacement with either FTSG or buccal grafts. Two patients lost most of the penile skin and required new skin sources to cover the urethra and penile shaft. Six patients required replacement of tissue expanders along the penile shaft to allow the creation of new skin to cover the urethra and penile shaft. The tissue expander is inserted in a subcutaneous position beneath the penile shaft skin and incrementally injected percutaneously on a weekly basis with sterile saline in varying quantities depending on the amount of penile skin required. The neovascular pseudocapsule created by the expander allows a supple neovascular sheath to either cover or wrap the urethra with vascularized tissue. Most young infants can have an expander placed on either side of the penile shaft without difficulty. The expander is then removed without difficulty at the time of surgery. In 1 case the expander eroded and was removed. However, the remaining expander allowed enough skin for closure and repair. Eroded expanders can be replaced if needed but the erosion rate is higher than that of primary repair.

The use of FTSG and buccal mucosa to replace all or part of the urethra was required in 14 patients. As mentioned, 5 infants required replacement of the majority of the urethra. Also, in 11 patients either a FTSG or buccal graft was used to bridge the gap between the prostatic urethra and urethral plate in those in whom the plate was not entirely lost. Timing of urethral plate grafting depended on several clinical factors including the quality of urethral plate tissue at the time of referral, and the need for testosterone stimulation of the urethral plate and/or penile skin augmentation using tissue expanders. In the 6 patients with complete dehiscence a graft was essential to allow the bladder and posterior urethra to be placed deeply within the pelvis.

Intramuscular testosterone enanthate (2 mg/kg at 5 weeks and 2 weeks before surgery) is a useful adjunct in cases of reoperative epispadias repair and primary repair if there is a general paucity of penile size and penile skin, and topical 5% testosterone cream is applied to the urethral plate and urethral plate in those in whom the plate was not entirely lost. Timing of urethral plate grafting depended on several clinical factors including the quality of urethral plate tissue at the time of referral, and the need for testosterone stimulation of the urethral plate and/or penile skin augmentation using tissue expanders. In the 6 patients with complete dehiscence a graft was essential to allow the bladder and posterior urethra to be placed deeply within the pelvis.
loss of the urethrovaginal septum secondary to use of para-

exostrophy skin flaps. This type of soft tissue loss does not
appear to have been reported in cases of CPRE previously. In
our 2 patients with loss of the urethra prior similar recon-

structive experience was used along with a bladder tube to
make-up the difference with urethral loss. One female with
an impenetrable stricture required direct vision internal ure-
throtomy and intermittent catheterization until a stable ure-

thra was obtained.

In the re-closure of a failed exstrophy, whether it is a
modern staged closure, complete repair (CPRE) or other rec-
ognized techniques, the surgical principles are still the same.
The bladder and posterior urethra must be radically dis-
sected from surrounding structures and placed deep in the
pelvis. Helpful adjuncts include transverse bilateral innom-
inate and vertical iliac osteotomy along with a fixator and
modified Buck’s traction, proper postoperative sedation and
pain control to allow healing, and testosterone intramus-
cularly and topically on the urethral plate if needed. We have
used the combined repair (closure of bladder exstrophy along
with modified C-R) in a number of failed exstrophy cases with
excellent success.16, 17

CONCLUSIONS

Complete primary repair of exstrophy in the newborn is a
technically demanding procedure and not for the occasional

exstrophy surgeon. The degree of penile, urethral and glans
complications can confer a significant degree of difficulty to
the reconstructive efforts. While penile skin can be generated
by the use of tissue expanders and testosterone or even grafts
of nongenital skin, the loss of urethral tissue requires graft-
ing of buccal or FTSG with a reasonable chance of success.
The real difficulty lies in the loss of glanular and corporal
tissue for which good tissue substitutes do not exist. Techni-

cal advances in the laboratory with tissue engineering and

stem cell derivatives are likely to provide a vital source of
tissue substitutes and replacements for these particular pa-

tients.

The training and experience of the surgeon not only with
the exstrophy condition, but also with complete repair should
be considered when discussing this method of reconstruction
with parents. While two-thirds of the referred cases in our
series were treated by fellowship-trained pediatric urologists,
the remainder were performed by those who had not under-
gone fellowship training in advanced reconstructive pro-
ductures. While fellowship training is no guarantee of success, it
does allow exposure to advanced reconstructive techniques
required for a procedure of this magnitude in a newborn.
Finally, it should be remembered when evaluating the level
of complications in our series that these patients represent a
highly select subset who have had unfortunate and particu-
larly pronounced problems requiring challenging reconstruc-
tive surgery. Most patients undergoing CPRE do not suffer
such complications and contemporary series have reported
good functional results.

REFERENCES

1. Mitchell, M. E. and Bagli, D.: Complete penile disassembly for
2. Grady, R. W. and Mitchell, M. E.: Complete repair of bladder
3. Hammouda, H.: Results of complete penile disassembly for ep-

4. Gearhart, J. P.: Complete repair of bladder exstrophy in the
newborn: complications and management. J Urol, 165: 2431,
2001
and/or corpora following primary repair of bladder exstrophy
using the complete penile disassembly technique. J Urol, 172:
1696, 2004
6. Gearhart, J. P.: Failed bladder exstrophy closure: evaluation and
7. Gearhart, J. P., Ben-Chaim, J., Scortino, C., Sponseller, P. D.
and Jeffs, R. D.: Multiple reoperative bladder exstrophy clo-

ures: what affects the potential of the bladder? Urology, 47:
240, 1996
8. Baka-Jakubiak, M.: Combined bladder neck and urethral and
penile reconstruction in boys with exstrophy-epispadias com-

plex. BJU Int, 86: 513, 2000
reconstruction der blassen exstrophy. In: Verh Ber Otsch Ges
383, 1984
10. Stein, R., Fisch, M., Black, P. and Hohenfellner, R.: Strategies
for reconstruction after unsuccessful or unsatisfactory pri-
mary treatment of patients with bladder exstrophy or incon-
11. Grady, R. W., Ben-Chaim, K. K., Scortino, C., Sponseller, P.
D.: Methods of pelvic and extremity immobilization following bladder exstro-

phy closure: complications and impact on success. Urology, 62:
1109, 2003
repair of exstrophy: further experience with neonates and
13. Silver, R. I., Yang, A., Ben-Chaim, J., Jeffs, R. D. and Gearhart,
J. P.: Penile length in adulthood after exstrophy reconstruc-
14. Gearhart, J. P., Forshner, D. C., Jeffs, R. D., Ben-Chaim, J. and
Sponseller, P. D. Combined vertical iliac and horizontal pelvic
osteotomy for primary and secondary repair of bladder exstro-
pelvic osteotomy in the modern era of bladder exstrophy re-
construction, Unpublished data

bined bladder exstrophy closure and epispadias repair in the
reconstruction of bladder exstrophy. J Urol, 160: 1182,
1998
17. Baird, A. D., Mathews, R. I. and Gearhart, J. P.: The use of
combined bladder and epispadias repair in males with classic
bladder exstrophy: outcomes, complications and consequences.
J Urol, 174: 000, 2005

EDITORIAL COMMENTS

The authors provide a descriptive account of 19 patients referred
with complications following CPRE. Of the 19 cases 6 were previ-
ously discussed (reference 5 in article). Some patients underwent
initial closure beyond 72 hours of life without iliac osteotomy. This
practice is contrary to conventional wisdom and placed these indi-
viduals at an increased risk for total dehiscence and bladder pro-
lapse. These specific complications occurred in 12 males and 2 fe-
nales in the series. This fact is concerning and has more to do with
surgeon experience and familiarity with indications for osteotomy as
an adjunct to initial management and less to do with preference for
CPRE or the first stage of the staged approach.

It is clear that injection of vasoactive agents increases the risk of
permanent tissue loss, which is true for any exstrophy or epispadias
repair, and should be avoided. The authors are to be commended for
their discussion of adjunctive measures for tissue reconstruction in
this complex subgroup of cases. The complications discussed may
occur with either CPRE or a staged approach. This report highlights
the need for this technically demanding surgery to be performed by
a surgeon with sufficient experience and proficiency in all aspects of
bladder exstrophy reconstruction. This tenet applies regardless of
technique or training.

Joseph G. Borer
Department of Urology
Children's Hospital Boston
Boston, Massachusetts

The authors are to be congratulated for continuing their long-
standing commitment to increasing our understanding of the care of
children with exstrophy. The data presented here emphasize the
importance of familiarity with technique regardless of the approach chosen to repair exstrophy. The next steps to better understanding should include hypotheses why these complications occur (as the authors have done) and increased rigor to baseline reporting criteria for clinical research studies in these areas to increase our ability to analyze and compare results. Such changes will allow us to more critically evaluate factors that impact the success or failure of our efforts to correct exstrophy.

Richard Grady
Division of Pediatric Urology
Children’s Hospital and Regional Medical Center
University of Washington School of Medicine
Seattle, Washington


DISCUSSION

Dr. Alberto Lais. I assume your cases were referrals from previously pitfalls of closure. You stated that you performed an osteotomy. How many of these patients had had a previous osteotomy?

Dr. John P. Gearhart. None of those patients had had a previous osteotomy.