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Challenges in salvaging urinary continence following failed bladder exstrophy repair in a developing country

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Summary

Introduction
The management of urinary incontinence following failed bladder exstrophy repairs is not well defined, some surgeons recommend urinary diversion, others would pursue reconstructive surgery.

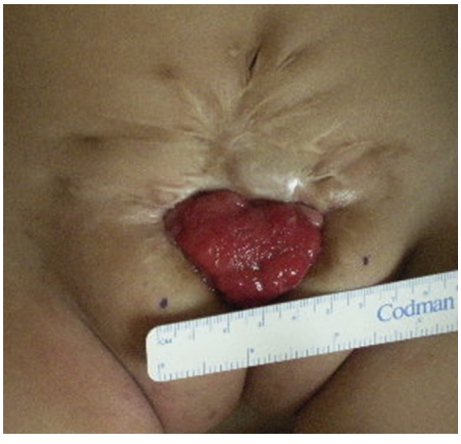
Objective
Herein we review our experience with various strategies for management of urinary incontinence and their outcome in 61 patients born with bladder exstrophy who failed their initial repairs. We also examine the impact of diverse factors in making the surgical decision for patients with limited resources in a developing country.

Material and methods
Between 1981 and 2014, 61 incontinent patients (age 3–18 years) born with bladder exstrophy were referred for secondary or tertiary repair. In nine children the bladder was exposed following wound dehiscence and 52 had had one or more attempted repairs resulting in contracted bladders. The primary treatment included: re-closure and bilateral iliac osteotomy in five patients (group 1); urinary diversion in Mainz II pouch in 15 patients (group 2); and 41 patients (group 3) underwent bladder augmentation and bladder neck reconstruction (31 patients) or bladder substitution and cystectomy (10 patients).

Results
In group 1, three out of five patients were voiding with dry intervals. One child was dry between catheterization, and one was incontinent and underwent Mainz II diversion. All 16 patients with Mainz II pouch were continent of urine and stool. Of the 41 patients in group 3, follow-up was available for 34 patients, of whom 31 (91%) were dry between catheterization. However, additional surgery was performed in 15/34 (44%). Malignant changes were noted in 2/26 cystectomy specimens.

Discussion
The quality of the bladder plate may deteriorate following failed bladder exstrophy surgeries. A few selected cases who had maintained a relatively healthy bladder plate were candidates for re-closure with osteotomy. Other surgical options included complex reconstruction with catheterizable channel, and internal urinary diversion. In this series the following factors were considered: a) pre-operative bladder biopsy to rule out pre-cancerous lesions, b) counseling the parents about possible additional surgeries after bladder augmentation or continent reservoir, and c) patients' geographic and socio-economic status.

Conclusions
Children born with bladder exstrophy are best treated at pediatric centers with expertise in their management. The exstrophied bladder should be protected and covered with non-adherent plastic wrap to prevent mucosal irritation and ulceration by the diaper. However, following failed surgeries the majority of incontinent patients can be salvaged to become continent/dry. It would appear that the Mainz II internal diversion offers a reasonable surgical option for selected patients, especially for females and those who have endured multiple surgical failures.



Summary figure Recurrent bladder exstrophy in 3 yr. old girl following 4 attempted repairs.

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Introduction

The early literature on the management of children born with bladder exstrophy (BE) indicated that the majority were treated with permanent urinary diversion by ureterosigmoidostomy [1] and subsequently ileal conduit. Bladder closure and multistage reconstruction were introduced by Jeffs in 1972 [2]. Continence using his approach was achieved in 70–80% of cases by other experienced surgeons [3,4]. In developing countries referral of newborns with bladder exstrophy is frequently delayed and surgical expertise is not always available, especially for patients with limited resources living in rural areas. After multiple surgical failures to achieve continence/dryness, many patients were social outcasts wearing diapers, and could not attend schools. The parents and their referring physicians believed that there was no remedy other than permanent urinary diversion by ileal conduit and abdominal stoma, which is socially unacceptable in some developing countries.

At present there are three approaches for management of children born with BE: multi-staged reconstruction, one stage complete primary repair of bladder and urethra, and primary urinary diversion. It has been reported that the chance of achieving continence after failed and multiple closure is markedly diminished [5].

Herein we report on our experience with 61 children born with classic BE who were referred after attempted surgical repairs. They were treated by one stage salvage surgery to achieve urinary continence/dryness over the last three decades. The aim of this study is to demonstrate that successful repair is achievable in developing countries despite multiple surgical failures.

Material and methods

Between 1981 and 2014, 61 children born with BE were referred following failed surgeries to achieve urinary continence, all were incontinent of urine. In most cases the size and quality of the bladder plate, medical records, and prior operative details were unavailable or inadequate, and we often had to rely on medical history provided by the parents.

There were 45 boys and 16 girls aged 3–18 years, all had had several operations ranging between two and 14 surgeries. Nine children had wound dehiscence and their bladders re-exstrophied and three underwent urinary diversion using an ileal conduit, their parents requested urinary undiversion. Forty-one children had had one or more attempted closures and surgeries to achieve urinary continence, resulting in various degrees of bladder contracture and/or fibrosis, associated with bladder prolapse in some cases. However, in five children (group 1) the bladder plate appeared to be relatively healthy and was judged to be suitable for reconstruction (Table 1). Only four out of 49 had had prior iliac osteotomy and one patient had had bilateral pubic osteotomy elsewhere.

Initial evaluation early in the series included radiologic study of the upper urinary tract by intravenous pyelography and subsequently ultrasonography. Blood chemistry, complete hemogram, and examination under anesthesia included cystoscopy and multiple bladder biopsies in all cases with bladder dehiscence and when indicated in others, such as if the bladder wall appeared polypoid or keratinized. When the pathologic specimens showed extensive cystitis glandularis and squamous metaplasia, cystectomy and bladder substitution were performed (10 patients). Early in the series in three out of 49 patients the symphyseal gap appeared to be too wide, and posterior vertical iliac osteotomy was performed at the same sitting as the bladder augmentation and substitution. As our experience increased, we were able to close abdominal wall defects without osteotomy. However, when functional closure was contemplated in five cases, we believed that additional osteotomy would decrease the tension on the repeat bladder and abdominal wall repairs. As our experience increased, we were able to close abdominal wall defects without osteotomy in groups 1 and 2.

Table 1 Surgical repair of 61 incontinent bladder exstrophy patients.

Group1: Reclosure and iliac osteotomy	5[a]
Group 2: Mainz II pouch	16[a]
Group 3: a Bladder augmentation	31
b Bladder substitution	10

[a] One patient from group 1 was incontinent and underwent Mainz II diversion.

Group 1 included five children (Table 1) who underwent one stage posterior iliac osteotomy, bladder and Young-Dees bladder neck reconstruction [6,7], with bilateral cross-trigonal ureteral re-implantation in four patients, and single ureteral re-implantation with transuretero-ureterostomy for dilated ureters in one patient.

Fifteen patients (group 2) for whom intermittent catheterization was either impractical or unacceptable to the parents, and one boy who failed one stage repair and was incontinent of urine, a total of 16 patients underwent modified Mainz II ureterosigmoidostomy [8]. Some of these cases had been reported previously [9]. The ureters were re-implanted in sub-serosal tunnels using the surgical technique reported by Abol-Enein and Ghoneim [10] instead of submucosal tunnels.

In patients with small capacity bladder (<50 mL), various segments of bowel were utilized for bladder augmentation and replacement (group 3 a,b), with the most common being the ileo-cecal-appendiceal segment. At surgery the ureters were re-implanted into the bladder remnant and a urinary reservoir was created from the cecum and ascending colon based on the right ileo-colic vessels. The ceco-appendiceal junction was assessed intraoperatively for competence [11]. A cystotomy tube was placed in the cecum and the tip of the appendix was excised, the cecum was filled with normal saline and infused via gravity at 60 cm above the bladder level to evaluate the competence of the ceco-appendiceal sphincter. When leakage from the tip of the appendix was noted, the cecum was plicated over the base of the appendix to re-inforce the sphincteric mechanism [12]. This bowel segment was used for bladder augmentation (20 patients) and for bladder substitution (10 patients).