

## Complex Repair of Abdominal Organs Exstrophy in an Adult Female-A Case report

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Short Title: Abdominal Organs Exstrophy case report

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**Citation:** Barel E, Cohen AA, Shay T, Yaacobi DS, Shachar T, et al. (2022) Complex Repair of Abdominal Organs Exstrophy in an Adult Female- A Case report. Annal Cas Rep Rev: 338.

**Received Date:** 19<sup>th</sup> August, 2022; **Accepted Date:** 26<sup>th</sup> August, 2022; **Published Date:** 31<sup>st</sup> August, 2022

### Abstract

A 42-year-old woman, employment immigrant, presented to our clinic with untreated bladder exstrophy with urinary bladder and uterovaginal prolapse and incontinence of urine. Surgical reconstruction was performed by a multidisciplinary team. Following the procedure, the patient was pleased with the aesthetic result, the resolution of her urinary incontinence freeing her from dependence on diapers, albeit the inconvenience of maintaining her urinary diversion stoma. Furthermore, she has noted all-in-all improved social as well as sexual function, although regarding the latter she has reported her difficulty in achieving orgasm. Thus, Surgical treatment of bladder exstrophy presents a complicated challenge, and even in the best of settings results often leave more to be desired.

**Keywords:** Absent clitoris, Bladder Exstrophy case report, Component separation, Pedicled flap, Rectus abdominis flap.

### Introduction

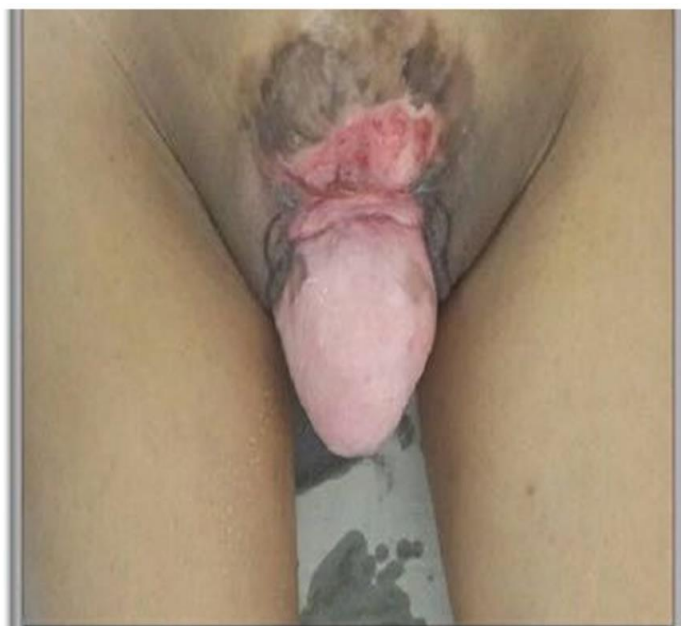
The term Bladder Exstrophy (BE) encompasses a class of infra umbilical anterior abdominal midline defect of varying expressions. BE affected structures may include the genitalia, urinary tract and bony pelvis [1,2]. Incidence of BE is estimated at 1:30,000 to 1:50,000 live births, BE is more frequent in male sex and Caucasian ancestry [3–5]. Recent literature has consistently depicted a spectrum of multi-system birth defects, encompassing under the title Exstrophy-Epispadias-Complex (EEC) a number of clinical entities, from isolated Epispadias, to the most common of these - ‘Classic Bladder Exstrophy’ (CBE) and the more morbid Cloacal Exstrophy (CE) [3]. A clinical definition arguably lacking in precision and uniformity, CBE is said to typically present with a lower abdominal wall defect exposing an open bladder and urethra, a wide diastasis of the pubic symphysis, and an epispadic urethral opening [4,6]. CE is similarly described, but for a bilobed bladder separated by a portion of cecum, and commonly presents with various other anomalies, such as malformations of the gastrointestinal, musculoskeletal, and central nervous

systems [7]. Embryologically the various forms of EEC are attributed to week 4 to 6 of gestation, when the cloacal membrane is prematurely invaded by mesoderm to build the abdominal wall for the genitourinary system. The timing of this developmental mishap would thus correlate with the phenotypic severity, i.e. If the membrane ruptures before 4 weeks of gestation, CE ensues, whereas if it ruptures after the urorectal septum has descended (at 6 weeks), isolated epispadias or CBE occurs [3].

Surgical treatment aims to achieve urinary continence, closure of the bladder and abdominal wall, preserved renal function and functional as well as aesthetically acceptable external genitalia [7–10].

## Case Presentation

A 42-year-old woman presented to our clinic with untreated bladder exstrophy with bladder and uterovaginal prolapse and incontinence (fig. 1). An employment immigrant - she had no access to proper medical care in her native country, she had grave difficulty with social and sexual function, and had thus far been regularly wearing diapers. Computed tomography (CT) examination of her abdomen and pelvis revealed a malformed bony pelvis with the antermost pubic rami approximately 13cm apart.



**Figure 1:** Initial presentation-bladder exstrophy with uterovaginal prolapse and incontinence.

Our multidisciplinary team, comprised of Urology, Gynecology and plastic surgery specialists - devised and executed a surgical reconstruction: Cystectomy and urinary diversion via an ileal conduit (regarding the original deficit - she presented only a posterior bladder wall and no notable urethral remnants, as well as no distinct clitoris or parts thereof). The next part of her operation entailed fixation of the uterus to the pelvic floor musculature correcting for uterovaginal prolapse and colporrhaphy.

Given the significant remaining deficit in the anterior abdominal wall, our team performed a component separation - i.e. release of the external oblique aponeurosis to achieve medial translation of the rectus complex, with release of the rectus abdominis origin for mobilization of the posterior rectus sheath.

Notably, it was decided not to interfere with the bony skeleton due to the perceived risk of harm to her fully functional, stable and painless gait.

Following the aforementioned component separation, the anterior vulvar commissure was approximated and closed. Subsequently, an inferior pedicled (inferior epigastric artery based) vertical rectus abdominis myocutaneous (VRAM) flap with an overlying skin island corresponding to the remaining deficit was designed and raised, a mesh was applied to the donor site (fig. 2), as well as to the internal abdominal wall underlying the deficit, completing coverage

of the multilayered deficit with the VRAM flap (fig. 3). The patient has presently been undergoing follow up in our clinic, most recently (to the time of writing this text) 19 months after the aforementioned procedure. She is pleased with the aesthetic result, the resolution of incontinence freeing her from dependence on diapers, albeit the inconvenience of maintaining her urinary diversion stoma. Furthermore, she has noted all-in-all improved social as well as sexual function, although regarding the latter, she has reported difficulty in achieving orgasm.



**Figure 2:** The VRAM flap fixed via key sutures to the deficit, a second mesh applied to the iatrogenic abdominal wall deficit secondary to the VRAM flap harvest.



**Figure 3:** Final in-setting of the mesh and overlying flap at the primary (initial) deficit, overlying a drain.

## Discussion

Surgical treatment of bladder exstrophy presents a complicated challenge, and even in the best of settings results often leave more to be desired [11]. Albeit defining the desired end-point of medical & surgical intervention in compound congenital anomalies-is far from trivial, the goals of reconstruction as consistently stated in the literature are to (1) provide an adequate reservoir for urinary storage, (2) create functional and cosmetically acceptable external genitalia, (3) establish continence and (4) preserve renal function [8,9,12-14]. Notably, a variety of surgical techniques have been described towards these goals, where in patients with intricate deficits of tissue and domain - no 'gold standard' can yet be defined.

Accepted reported treatment strategies diverge to either a 'staged approach', in which the initial step involves closing the bladder plate and posterior urethra, with subsequent steps of epispadias repair and finally bladder neck reconstruction (BNR), or - complete primary repair of bladder exstrophy (CPRE), which involves closure of the bladder plate and epispadias repair, and may include ureteral reimplantation.

Whereas repair of epispadias is described in many instances in the literature pertaining to bladder exstrophy, there has been little mention of external female genitalia repair, but for the (qualitative) prevalence of a bifid clitoris. Our patient presented without any discernible clitoris or remnants thereof, thus the most anterior parts of the labia and would-be introitus were approximated as best was possible. Notably, at the time of writing this report it has been planned that she should undergo surgical reconstruction of the anterior commissure, which she is awaiting.

Regarding the skeletal component, BE is said to typically present with shortened pubic rami as well as a wider angle of external rotation of the innominate bones (sagittal axis), thus attempting to close the bony pelvis may be advocated for a few reasons: (1) approximating the margins of the abdominal / pelvic deficits so as to reduce closure tension; (2) hypothetically may reduce uterine prolapse; (3) hypothetically, re-approximating the pelvic floor musculature may improve future continence in the presence of a patent pelvic urethra. Notably, the skeletal anomaly associated with exstrophy, though potentially producing an externally rotated foot progression angle potentially invoking a wide based gait potentially invoking early hip arthrosis, is not in itself regarded an indication for surgery. In the case of our patient, whom had no pain nor other difficulty in ambulation, given the increasingly painful and prolonged recovery (including mandated prolonged immobilization) that pelvic osteotomy ensues, and the tensionless closure devised by our surgical team, it was decided not to pursue skeletal intervention.

Several complications had been consistently described in the literature including but not limited to: (1) recurrent urinary retract infections [0-65%] & pyelonephritis [0-54%]; (2) bladder dehiscence [0-4%]; (3) hydronephrosis [0-33%] & renal function impairment [0-22%]; (4) bladder

calculi [0-24%]; (5) Bladder obstruction [0-13%] and (6) Urethral stricture [0-21%].

Of particular interest regarding bladder exstrophy patients is the absent standard definition of continence. With regard to a prevalent definition that is used in bladder exstrophy literature, continence is defined as maintaining dry periods of 3 hours or greater than 3 hours during the day with no stress incontinence. Notably, the aforementioned is not consistent with the definition of continence as established by the International Children's Continence Society. Reported urinary continence rates after repair of bladder exstrophy vary, ranging from 12% to 83%, albeit lacking uniform definition to continence and inconsistently reported correlation with severity of bladder neck malformation and need for initial repair and revision thereof. Arguably, given continence is pinnacled among the treatment goals, it is paramount to set well defined, achievable goals and adequate patient expectations when deciding and designing a treatment plan.

Regarding sexual function, our patient has presented a problem to which we could not find has been addressed in the present literature- a difficulty achieving orgasm following reconstructive surgery wherein no discernible clitoris was observed, perhaps an awake mapping of our patient's (or rather, future similar cases) erogenous zones prior to surgery may aid in identifying aberrant significant sensate tissues.

In summary, bladder exstrophy as presented here, is a multidimensional problem which necessitated an intricate multi-disciplinary repair, which was performed in this instance successfully - achieving significant objective restoration of near normal anatomy and function as well as significant patient satisfaction.

### Disclosure:

Regarding consent, our patient had given her formal consent for our team to present her case to the medical community at large, expressing a hope that her misfortune and the treatment thereof may serve to educate, and perhaps benefit others with similar conditions in the future. Ethical approval is not required at our institution to publish an anonymous case report.

### Statements

**Funding:** The authors did not receive support from any organization for the submitted work.

**Conflict of interest/ Competing interest:** The Authors declares that there is no conflict of interest.

**Ethics Approval:** Ethic approval was waived by our local ethics committee in view of the anonymous nature of the study and all the procedures being performed were part of medical routine care.

**Consent to participate:** Verbal informed consent was obtained prior to the interview.

**Consent for publication:** The participant has consented to the submission of the case report to the journal. The authors affirm that human research participant provided verbal

informed consent for publication of the images in Figures 1, 2 and 3.

**Availability of data or materials:** Data will be available upon reasonable request.

**Author Contribution:** EB - Project administration, methodology, investigation, original draft preparation, review and editing. AAC - Project administration, methodology, investigation, original draft preparation, review and editing. TS- Investigation, methodology, original draft preparation. DSY- Investigation, methodology, original draft preparation. TS- Investigation, original draft preparation. HK - Review and editing. JB- Review and editing. DDA- Project administration, review and editing.

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