Bladder & Cloacal Exstrophy:
A 30 Year Journey of Innovation

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Objectives

Identify 3 systems involved in the Exstrophy Complex
Diagnosis
Define the procedure for management of the exstrophied bladder
State 2 components of psychosocial support for the Exstrophy Population

There are no disclosures
Exstrophy Complex

Exstrophy – Epispadias (EEC):
- Classic Bladder Exstrophy
- Epispadias

Cloacal Exstrophy (OEIS):
- Omphalocele
- Exstrophy
- Imperforate anus
- Spinal anomaly

Incidence - 1:10,000 - 1:50,000 live births
5:1 ratio of male - female births

Embryology
- Typically occurs between 9 and 12 weeks gestation
- Cloacal membrane ruptures prematurely AFTER separation of the GI and GU tracts

Presentation:
- Eversion of the bladder through an abdominal wall defect
- Exposure of the inner bladder mucosa
- Exposure of the dorsal urethra
- Lack of musculature in the anterior abdominal wall over the bladder
- Bladder is exposed and drains onto the abdomen

Exstrophy/Epispadias Complex (EEC)

Prenatal Diagnosis (Fetal US)

Courtesy of Carol Barneswold, MD
Bladder Exstrophy (Boy)

- Low-lying umbilicus
- Exposed (inside-out) bladder
- Urethra open on dorsum (top) of the penis

Bladder Exstrophy (Girl)

- Bladder open on abdominal wall
- Urethra open between bifid (split) clitoris in the girl

Cloacal Exstrophy

- Incidence: 1:200,000-400,000 live births
- Equal prevalence of males to females

Embryology:
- Occurs with spontaneous rupture of the cloacal membrane BEFORE division of the GU and GI tracts

Presentation:
- Bladder Exstrophy
- Omphalocele
- Imperforate anus
- Spinal cord abnormalities
- Split genitalia
**Cloacal Exstrophy (Boy)**
- Bladder halves divided by cecal plate
- Prolapsing ileum in midline
- Corporal body separation (boys)
- Absent anus

**Cloacal Exstrophy (Girl)**
- Bladder halves divided by cecal plate
- Prolapsing ileum in midline
- Clitoral body separation (girls)
- Absent anus

**Cloacal Exstrophy: OEIS Complex**
- Omphalocele
- Exstrophy
- Intestinal abnormalities
- Spinal deformity
Initial Postnatal Bladder Care

Secure umbilical stump with soft tie
Do not use clips
Apply Tegaderm over exposed bladder mucosa
Place diaper

Timing of Initial Surgery

Delayed Closure
Eliminate emergency
Limit parental anxiety / Promote bonding
Caregiver appreciation of Bladder extrophy and Cloacal Exstrophy
Infant development / Decrease anesthesia risk
Tissue growth: Bladder with Valsalva, Genitalia growth

Bladder Exstrophy Closure

Closure at 2-3 months
Complete Primary Repair of Bladder Exstrophy (CPRE)
Closure of bladder, reconstruction of the bladder neck, and urethra
Appropriate bladder outlet resistance
Normal bladder cycling
Optimal bladder growth and development
Bilateral pelvic osteotomies
Cloacal Exstrophy Staged Repair

Stage 1:
- Within first month of life
- Omphalocele closure
- Rescue the hindgut
- Separation of cecal plate (bladder halves) from intestinal tract
- Creation of end colostomy for stool diversion
- Approximation of bladder halves

Stage 2:
- 6-12 months of life
- Bladder closure

Goals For Bladder Care

Provide management techniques to protect exposed bladder mucosa surface and surrounding skin from:
- Diaper abrasion
- Fecal incontinence
- Exposure to noxious urine

Increase confidence level of caregivers and healthcare professionals

 Expedite appropriate care of surrounding skin complications

Intact dressing prior to removal.
**Cloacal Exstrophy (Boy) Post Stage 1 Repair**

- Colostomy
- Umbilical Stump
- Bladder
- Scrotum

**Special Considerations for Care**

- Tegaderm adherence difficulty
- Neighboring Colostomy or stoma
- Para-exstrophy skin rash / breakdown
- Para-exstrophy skin infection
  - Fungal or bacterial

**Duoderm**

- Colostomy
- Duoderm Horseshoe
- Bladder
Skin Care Treatment

- General skin care measures
  - Frequent diaper changing
  - Thorough cleansing/bathing
- Aquaphor™
- Criticaid™ clear/paste
- Nystatin cream/ointment
- Nystatin powder
- Mycolog™ Topical Cream

Rash Example

Fungal Rash Example
Neighboring Stoma

Address exstrophied bladder first
  Directs urine away from stoma
Adhere stoma appliance
  Or cover with absorbent gauze
Fasten diaper

Communication: Secure Image Messaging

Electronic images transmitted
  Securely
Local and long distance triage and support
Timely expert assessment
  Normal vs. Abnormal
  Reassurance vs. Recommendation(s)
It takes a village ...

Our multidisciplinary team consists of physicians, nurse practitioners, nurses, social workers, researchers, child life specialists, and volunteers.

Psychosocial support for BE/CE

Psychosocial support is a key component of comprehensive care across the continuum

- Inpatient support through child life and social work
- Outpatient support through nursing and social work in clinic
- Outreach to caregivers at school
- Utilization of medical coping program
- Coordination of outpatient therapy
- Participation in support group activities
Case study- MG

28 yo with prenatal diagnosis male BE  
Presentation at birth CE male 
Multiple surgeries:  
  Bowel diversion  
  Gonadectomy  
  Bladder closure with augmentation  
  Spinal cord detethering  
  Vaginoplasty with buccal mucosa

Case study- MG (cont.)

Psychosocial challenges:  
  46 XY karyotype raised as female  
  Chronic bowel and bladder incontinence  
  Compromised family dynamic  
  School disruption  
  Social isolation  
  Clarity of birth gender at age 18

Case study- RG

Newborn prenatal diagnosis male CE  
Presentation at birth: diagnosis confirmed CE  
To be raised as a male  
1st stage CE surgery complete
Case study- RG (cont.)

Physical challenges:
- Wound ostomy and skin care issues
- Bladder closure with osteotomy and spica casting
- Continence procedures to be determined
- Genitoplasty

Psychosocial challenges
- Emotion burden on family due to multiple hospitalizations
- Financial burden on family due to extended stay & multiple return visits

Exstrophy complex innovations over thirty years

- Sophisticated prenatal evaluation and referral- AFCC
- Transition to delayed closure for CE and BE
- Decreased length of stay in ICU and floor
  - ICU- from 4 nights to 1 night
  - Floor- from 6 weeks to 2 weeks
- Transition from traction to spica casting
- Telemedicine
- Peer mentor/support group
- Multi institution collaboration (MIBEC)

Support Group Timeline

- 1992- Initial ‘Support Group.’ First speaker, Dr. Retik, with 60 families in attendance.
- 1994- 1st Exstrophy picnic
- 1995- 1st panel presentation with adult patients
- 2000- 1st prenatal cases seen in clinic and AFCC
- 2005- Introduction of Social Work
- 2007- Live web cast- including closure and family experience
- 2008 – 1st ‘Teen Chat’
- 2010- 1st Co-ed ‘Teen picnic’
- 2012- Introduction to technology in patient care
- 2012- 1st formal urinary and reproductive health seminar for adolescents
- 2013- Family-to-Family training
- 2014- Presentation of the Bladder Exstrophy Collaboration by Dr. Borer
- 2015- Night at Fenway park for boys
- 2015- Completion of comprehensive patient handbook
- 2016- Addition of Program Coordinator
- 2017 Addition of Nurse Practitioner
Institutional Collaborators

MIBEC
Multi Institutional Bladder Exstrophy Consortium

With sincere appreciation to our patients and families who inspire us every day...

With sincere thanks...

Joseph Borer, MD
Lauren Cullen, CPNP
Brittany Szabo, BHS
References


