A Team Approach to Bladder Exstrophy
There is No “I” in Team

Pediatric Urology Nurse Specialists
September 26, 2019
Scottsdale, AZ

Lauren Cullen, CPNP and Joseph Borer, MD

Objectives

• Define elements of a multi-disciplinary team
• Appreciate challenges of complex patient care
• Identify opportunities for team development
• State how team approach can be applied to build a bladder exstrophy program

Disclosures

• There are no disclosures.

How far does care reach?

Local  Regional  National  International
Home Base

- Team Manager
- Coaches
- Players
  - Starting line up
  - Relief/support
- Dugout
  - Team huddles
  - Strategizing
  - Game film review
- Fans

Team Dynamics

- Communication is KEY
  - Speak up for safety: Any Role/ Any Time
  - Cross-check each other
  - Share ideas
- Leadership
  - Chain of command
- Commitment
  - Prioritize care and safety
  - Be cohesive in endeavors

Team Dynamics Cont.

- Learn/ identify strengths and weakness
- Pursue professional development
  - Contribution to team growth
- Team retreats/ evaluations
  - Annually or semi-annually
  - Reinforce positives and strategize solutions
- Communication planning
  - Identify channels for information sharing (ex. SBAR, IPASS)
Bladder Exstrophy Program
Mission Statement

• The **mission** of the BEP is to provide exceptional care for infants, children, adolescents and young adults with bladder exstrophy or other diagnoses within the exstrophy-epispadias complex.

Exstrophy-Epispiadias Complex (EEC)

• Epispadias
  • Exposure of dorsal urethra, exposing urethral plate

• Bladder Exstrophy (classic)
  • Eversion of bladder through abdominal wall defect
  • Exposure of bladder and urethral mucosa, and ureteral orifices

• Cloacal Exstrophy
  • Both urinary & intestinal tracts exposed through abdominal wall defect
  • Spinal, Renal, Limb, Omphalocele anomalies

Exstrophy-epispadias complex

<table>
<thead>
<tr>
<th>Condition</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epispadias</td>
<td>1:150,000</td>
</tr>
<tr>
<td>Bladder Exstrophy</td>
<td>1:40,000</td>
</tr>
<tr>
<td>Cloacal Exstrophy</td>
<td>1:400,000</td>
</tr>
</tbody>
</table>
Exstrophy-Epispadias Complex

- Alabama
- Alaska
- Arizona
- California
- Colorado
- Connecticut
- Florida
- Georgia
- Hawaii
- Idaho
- Indiana
- Iowa
- Kansas
- Kentucky
- Louisiana
- Maine
- Maryland
- Massachusetts
- Michigan
- Minnesota
- Mississippi
- Missouri
- Montana
- Nebraska
- Nevada
- New Hampshire
- New Jersey
- New Mexico
- New York
- North Carolina
- North Dakota
- Ohio
- Oklahoma
- Oregon
- Pennsylvania
- Rhode Island
- South Carolina
- South Dakota
- Tennessee
- Texas
- Utah
- Vermont
- Virginia
- Washington DC

Annual Diagnoses Types

Annual Patient Activity

National
Diagnosis of BE

- Prenatally
  - Absence of pelvic fluid filled bladder
  - Low lying umbilical cord
  - Low abdominal wall mass
- Postnatally
  - Classic BE anatomy at exam

Prenatal Dx Benefits

- Optimizes care
- Education and Preparation
- Continuity of care
Prenatal Dx of BE: Case Example

- Maternal Fetal Care Center (MFCC)
- Review Fetal Ultrasound/ MRI with Radiologist
- Parents meet with BEP team
  - Review diagnosis & Reinforce positives
  - Discuss Surgical options
  - Demonstrate care with simulation infant
  - Newborn care toolbox & BE Handbook
- Connect with Obstetrical team/ offer resources

Initial Surgical Options for BE

- Timing
  - Newborn
  - ~2 months*
- Technique
  - Complete Primary Repair of BE (CPRE)*
    - Including Iliac Osteotomies/ Spica cast
  - Modern Staged Repair of BE (MSRE)

Newborn Care Toolbox

1. Soft tie for umbilical cord
2. Transparent adhesive dressing for exposed bladder
3. Diapers to collect output

Family Journey

“At that point, we were in imagination land because finding credible and detailed information on rare conditions is challenging,” says Mom. “I just wanted to speak to someone who knew what they were talking about.”

- Statement from Mom after prenatal diagnosis

https://thriving.childrenshospital.org/bladder-exstrophy-lifting-fog/
Initial Post-natal Care: Case Example

- DOL 1
  - Local team provides care

- Initial BCH office visit
  - Goal: within first week of life
  - Evaluate/ review anatomy
  - Review surgery plans and anticipated goals
  - Review/ demonstrate care to caregivers

Bathing
- Wash soap & water
- Dry para-exstrophy skin gently
- Avoid bladder plate

Apply dressing
- Transparent adhesive

Post-natal Care

Pre-op Planning: Case Example

- Pre-identify OR team for case
  - Outline staff intra-op plan
  - Identify/ resolve any team concerns

- Pre-op appointment
  - Answer family questions
  - Review simulation infant with parents

- Social work reviews local resources for planning
- Create/ distribute case PowerPoint to team
Initial Surgery (CPRE): Case Example

• Intra-op
  – Morning Huddles
    • Anesthesia, Surgeons (Urology + Orthopedics), Nursing, Cast tech
  – Universal time out
  – Nurse liaison updates family
• Initial Post-op (< 10 days)
  – ICU X 1 night
  – Inpatient care per standards

Follow up Care: Case Example

• Outpatient Post-op (> 10 days)
  – Nursing visits – twice weekly
  – Dressing care
  – Ensure urine output
• 4 week post-op
  – VCUG
  – Begin clamping trials
• Transition to home (6-8wks)
  – Follow-up per prospective grid
Long Term Follow up Care Coordination

- Ongoing evaluation of age appropriate development
- Social Work collaboration for local resource support
- Secure email/ Telehealth check-in
- Handoff with Primary Care team
- Compliance with long-term follow up

Family Journey

“The team made this incredibly scary time in our lives so much more manageable... Not only did you all give Julia exceptional care, but you also took care of the rest of my family and I will forever be grateful!”

-Mom

Family gracefully shared their story: https://thriving.childrenshospital.org/julias-journey-bladder-exstrophy-surgery/

Patient resources

- Development process:
  - Families questions/ concerns
  - Clinical staff input
  - Patient education team
    - Edit for diverse comprehension
    - Marketing: Facilitate production

Grant Award Funding

- Production of video demonstration tool
  - Care and management of unrepaired BE
- Co-developed with patient families
- Intended for dissemination to:
  - New parents
  - Care providers
- Current Status: Editing & Production

Funding provided by the Rosemary H. Grant Nursing Innovation Fund
BEP Psychosocial Care

- Social Work
  - Psychosocial evaluation and support
  - Develop 504 plans with families
  - Support transition to school
- Patient/ Family match
- Psychiatry/ Coping clinic
- Semi-Annual Support Group

Support Group Highlights

- 1992 – 1st ‘Support Group’ with Dr. Retik, (60 families present)
- 1994 – 1st Exstrophy picnic
- 1995 – 1st Panel presentation
- 2000 – 1st Prenatal cases in clinic & AFCC
- 2005 – Addition 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
- 2012 – 1st Formal urinary and reproductive health seminar for adolescents
- 2013 – Family-to-Family training
- 2014 – Presentation of the MIBEC by Dr. Borer
- 2015 – Night at Fenway Park for kids
- 2015 – Completion of comprehensive patient handbook
- 2016 – Addition of Program Coordinator
- 2017 – Addition of Nurse Practitioner
- 2018 – 1st Zoom live support group
- 2019 – A-BE-C Center of Excellence award

It takes a Village...

JB (MD) – Program Director
RG (RN) – Nursing Coordinator
DO (MD) – Urologist-in-Chief
AB (MD) – Chief Emeritus
VW (CPNP), MD (CPNP), LC (CPNP) – In-Out-Patient Nurse Practitioners
MJ, JN – Administrative Assistants
LZ (RN) – Maternal Fetal Care Center
YK (MD, PhD); EN (MD) – Orthopedic Surgery
CB (MD) – Anesthesia
JC (MD); CB (MD) – Radiology
AV – GW – Clinical Research Program
SL (PhD) – Assistant Professor, Surgery/ Path.
PS (LICSW); DP (LICSW) – Social Work
LM (PhD), LH (PhD) – Clinical Psychologist
CL (MD); BW (MD); NR (MD); BL (MD)...
National Resources

- Youth Rally
  - Mission: To provide an environment for adolescents living with conditions of the bowel and bladder that encourages self-confidence and independent living. www.youthrally.org

- MIBEC Family Picnic

National/International Resources

- Association-Bladder Exstrophy-Community
  - Pamela (Block) Artigas
  - Annual Conference
    - 2019 at CHLA
    - 2020 at Johns Hopkins

www.bladderexstrophy.com

Bladder Exstrophy Program

- Local, National, and International dissemination
  - Boston Children’s Hospital
    - Publications (50)
    - Presentations (25)
    - Videos (6)
  - Conferences presentations:
    - PUNS, SPU, AUA, ESPUNS, ESPU, SPUS

Teaching and Learning beyond Hunnewell...
...through collaboration

...tackling the complex and rare

- Successful initial surgery is paramount to optimal outcome in bladder extrophy (BE)
- Given BE rarity, gaining necessary surgical experience for proficiency is challenging
- “Personal Best…coaches.” Atul Gawande (The New Yorker, 10/3/11)

Multi-Institutional Bladder Exstrophy Consortium (MIBEC)
Institutional Team Managers

Joseph Borer
Boston
Douglas Canning
Philadelphia
Michael Mitchell
COACH
John Kryger
Milwaukee

Institutional Coaches

Boston
Not pictured: Alyssia Venna, Monique Joe, Jillian Nolan.

Philadelphia

Milwaukee
Not pictured: Ed Bejtek, Melissa Lingongo

MIBEC Methods

• Pediatric urologic surgeons with special interest in Bladder Exstrophy care.
• The 3 sites alternating as host for scheduled surgeries, with observation, commentary and critique by visitors from the other 2 sites.

MIBEC Mission Statement

The mission of MIBEC is to perfect the surgical technique of Complete Primary Repair of Bladder Exstrophy (CPRE), and optimize care for patients within the Exstrophy-Epispiadas Complex.
MIBEC Methods

- Focus is CPRE
- CPRE at ~2 months of age
- Demographics, age @ CPRE, time of F/U
- Outcomes: Successful closure, complications
- Impact on CPRE technique and Sx volume

MIBEC

- Collate de-identified data between institutions regularly
- Travel monthly between institutions to operating room
  - Allows for shared experience
  - Mentorship / Coaching
- Semi-annual MIBEC conference
  - 2 days of 4 hour conference
    - Includes patient review, evaluate/analyze data, strategize on information dissemination and growth potentials

Complete Primary Repair of Bladder Exstrophy (CPRE)

CPRE 1996 - 2012
- previous at veru - taper into bladder
- improved continence

CPRE since Feb 2013
- current at BN - angle into bladder
- improved continence
**Operating Room Strategizing**

**CPRE in the Boy**

- Bladder dome
- Bladder neck

**Anatomy**
- Approximate width in mm
- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**CPRE: Bladder Dissection**

**CPRE: Reconstruction of Penis**

**Bladder dome**

**Bladder neck**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Bladder dome**

**Bladder neck**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Bladder dome**

**Bladder neck**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck

**Approximate width in mm**

- 20 mm
- 17-18 mm
- 15 mm
- 15 mm

- CEPHALAD
- MIDPOINT
- CAUDAL

- Bladder dome
- Bladder neck
Numbers Increased. So what?

- What are we learning?
- What are we doing right?
- What needs improvement?

MIBEC Consistencies Incorporated

- No bowel prep
- Same day admit
- Perioperative Antibiotic
- Spica-cast postop immobilization
- Specific technical aspects
  - Exquisite detail of dimensions (i.e. BN)
Benefits: Education/ Training

- Classical Halstead model
- Need now for modern strategy
- Complex surgery for a rare problem
- Residency down hours
- Trend of up minimally-invasive surgery

Benefits: Education/ Training cont.

- Intellectual and creative stimulation
- Another set of eyes - different perspective
- Healthy criticism (Harvard Business Rev.)
  - Expert opinion offered
  - Differing opinions discussed
- Critical reflection

Another set of eyes – you may see something that I do not!

Danger (sans Collaboration)

- Newton’s 1st Law of Motion
  - “Every object in a state of uniform motion tends to remain in that state of motion unless an external force is applied to it.” (Galileo’s concept of inertia, AKA law of...)
- A Urologist at rest stays at rest, unless...
- Nuance(s) that may enhance technique
Challenges to Collaboration

- Travel
- Expense
- Time
- Overstimulation
- Distractions in the OR

Antananarivo, Madagascar

NHK World Japan Somewhere Street
Antananarivo Madagascar

MIBEC – experience shared

- Presentations
  - National (20)
  - International (15)
- Publications
  - Manuscripts (5)
  - Chapters (3)
- Videos (18)
No endpoint, education in perpetuity…

“Chinese Pediatric Urology Training Program”
CHLA – Thursday, October 26, 2017
Special involvement of Evalynn Vasquez, MD and Roger De Filippo, MD

“It is the long history of humankind
(and animal kind, too) those who
learned to collaborate and
improvise most effectively have
prevailed.”

-Charles Darwin

With special thanks to all our teammates

- **Boston**
  - Joseph Borer, MD
  - Lauren Cullen, CPNP
  - Rosemary Grant, RN
  - Diane Price, LICSW
  - Paige Sable, LICSW
  - Alyssia Venna, MBS

- **Wisconsin**
  - Mike Mitchell, MD
  - John Kryger, MD
  - Travis Groth, MD
  - Elizabeth Roth, MD
  - Colleen Rosen, DNP

- **CHOP**
  - Doug Canning, MD
  - Aseem Shukla, MD
  - Dana Weiss, MD
  - Brynne Bonitatibus, CPNP
  - Jennifer Frazier, MPH

A special thank you to our patients and families as well for giving us
the honor and privilege everyday to care for them.
References

- Prophylactic Immobilization and Pain Management After Repair of Bladder Exstrophy. Roth E, Goetz J, Kryger J, Groth T.
- Clinical pathway for early discharge after complete primary repair of exstrophy and epispadias by using a spica cast. Sack BS, Kryger Jv, Mitchell ME, Dunkeel CT, Lyon R, Groth TTh.

References

- Bladder Exstrophy Program
- UpToDate, Inc.; Bladder Exstrophy care
- High Reliability Organization –AHRQ
  - https://psinet.ahrq.gov/primer/primer/31/High-Reliability
  - https://psjournal.org/opi/viewcontent.cgi?article=1287&context=journal
- Joint commission – Universal time out
  - https://www.jointcommission.org/assets/1/18/UP_Poster1.PDF

Thank You !!!