Bladder Exstrophy v2.1: Prenatal

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

PHASE I

Inclusion Criteria
- Suspected diagnosis of bladder exstrophy

Exclusion Criteria
- Suspected diagnosis of cloacal exstrophy

Prenatal Consultations
- Urology
- Social Work

Delivery Recommendations
- Delivery at any location that is comfortable with bladder exstrophy care
- Provider to provider consultation available

Family Education
- Bladder Exstrophy: How to Care for your Child's Bladder Before Surgery PE3009
- Bladder Exstrophy PE174

Provider Education
- Outside Provider Goals of Care
- Nursing Guideline of Care 10149 (*for SCH only*)

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If child is stable, no need to transfer until mom can also be discharged

Go to Neonatal Phase

Bladder Exstrophy v2.1: Neonatal

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

PHASE II

!

Child can be cared for on the floor unless they meet standard NICU criteria

Inclusion Criteria
- Diagnosis of bladder exstrophy

Exclusion Criteria
- Age > 12 months
- Cloacal exstrophy

Cloacal Exstrophy
- Will need general surgery involvement for ostomy + bladder plate closure

Initial Management

Imaging
- Renal US
- Pelvic xray

Diet
- Regular

Antibiotics
- None

Bladder Exstrophy Care
- Tegaderm over bladder plate in hospital; Tegaderm or Press and Seal at home
- Silk tie on umbilical cord

Activity Restrictions
- None

Nursing Care
- Nursing Guideline of Care 10149 (*for SCH only*)

Consultations
- Urology
- Orthopedics (including Spica tiger brought to family)
- Social work
- Lactation

Standard Newborn Care

!

Do not give prophylactic antibiotics prior to bladder closure

Family Education
- Bladder Exstrophy: How to Care for your Child's Bladder Before Surgery PE3009
- Bladder Exstrophy PE174
- Latex Allergy PE001

Urology Folder: (Urology NP will give to family)
- Information about the Association for Bladder Exstrophy Community
- Medical Alert ID PE2417
- Urology Rounds PE3005
- Bladder Exstrophy: Care After Surgery PE2910
- Urinary Reflux PE181

Discharge Criteria
- Teaching complete
- Consults seen
- Follow-up scheduled
- Cleared by Pediatrics

Follow-Up Coordination
- PCP identified and appointment scheduled within 3-7 days of discharge
- Urology clinic visit in 2 months

Last Updated: January 2023
Next Expected Review: May 2023

Bladder Exstrophy v2.1: Operative

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

PHASE III

Timing of Surgery
Elective closure at 4-6 months of age with osteotomy and spica cast

Inclusion Criteria
- Diagnosis of bladder exstrophy

Exclusion Criteria
- Age > 12 months
- Cloacal exstrophy

Pre-operative Visits

2 months prior to surgery
- Urology clinic
  - Request iron deficiency anemia screen from pediatrician

2 weeks prior to surgery
- Urology clinic
- Orthopedics clinic
  - Order car seat
- PASS clinic
  - Type and Screen

Outpatient Pre-Operative Management
- Anemia screen, if positive treat and discuss delaying surgery
- Car seat ordered at orthopedic RN visit
- OR team e-mail sent – **Urology fellow after Monday AM conference**
  - Pre-surgical Overview PowerPoint
  - Full Body Prep instructions
  - Pick lists
  - Huddle time

Infection Control

2 weeks prior to surgery
- Bladder plate culture
  - **If positive**, treat with 7 days antibiotics to end on the day of surgery, then culture directive perioperative antibiotics
  - **If negative**, cefazolin only for perioperative antibiotics

Intra Operative Care

Anesthesiologist

Pain control:
- Epidural, TAP or iliac fascial blocks when anatomy permits
- Standing Tylenol post-operatively
- Standing Toradol post-operatively if >6 months and no renal anomalies

Fluids/Lines/Tubes:
- A-line and 2 large Ivs
- Type and cross
- Orogastric tube. Remove at the end of case.
- mIVF on a pump with separate replacement of deficit and blood loss
- Albumin 5% as needed
- Transfusion guide

Infection Control:
- Antibiotics completed 30 minutes prior to incision

Extubation:
- Goal to extubate at end of case

Surgical tech
- Place urinary stents in a container to collect urine
- Monitor and call out urine output for anesthesia
- Open bladder exstrophy specific pelvic osteotomy kit

Nursing
- Bladder Exstrophy: Full body prep
- Radiopaque table
- Positioning at end of table
- **Fluoroscopy available for osteotomy**
- Page Orthopedic surgeon at end of case to place spica
- PACU nurse to petal spica

!
Fluid goal is euvolemia

Go to Post-Operative Phase

Bladder Exstrophy v2.1: Post-Operative

Approval & Citation | Summary of Version Changes | Explanation of Evidence Ratings

PHASE IV

NICU Admission
- At the discretion of the surgical and anesthesia teams
- Standard criteria for NICU discharge

Inclusion Criteria
- Diagnosis of bladder exstrophy

Exclusion Criteria
- Age > 12 months
- Cloacal exstrophy

Initial Management

Family Education
- Urology Rounds PE3005

Vital signs
- Standard vital signs per acute care guidelines
- Strict I/O

Activity
- Out of bed ad lib
- OK to hold baby

Fluids/Electrolytes/Nutrition
- D5 NS@maintenace IV+PO
- CBC, Chem7, Cr/BUN POD1
- Regular diet immediately after surgery

Medications
- Pain service consult in PACU
- Oxybutynin standing for bladder spasms

Wound Care
- Monitor incisions for early signs of infection
- GOC: Casts including Spica Casts 10207 (*for SCH only*)

Consults
- Lactation consultation if nursing or pumping
- Social work

!

STOP and RESOLVE if incisions are not clean, dry and intact

Infection Control
- Perioperative antibiotics for 24 hours after surgery
- No prophylaxis while bladder being continuously drained
- Urine culture 2 weeks prior to outpatient planned cast and tube removal
  - If positive, give culture driven antibiotics for 7 days (5 days prior to cystogram / VCUG, 2 days after)
- Antibiotic prophylaxis for vesicoureteral reflux if seen on outpatient VCUG at time of cast removal

Family Education
(once stable after surgery)
- Bladder Exstrophy PE174
- Bladder Exstrophy: Care After Surgery PE2910

Supplies and teaching (care coordination to initiate)
- Appropriate car seat
- Cather flush
- Petals / Mole skin
- Diapers

Follow-up Coordination
- Urology weekly visits x 6 weeks
- Spica cast removal at 4 weeks
  1. Orthopedics clinic visit for spica removal
     - Pelvic X-Ray to evaluate osteotomies
- 6 week visit for catheter removal
  1. Urology clinic visit for catheter removal
  2. Radiology:
     - Cystogram for bladder integrity; if (-), VCUG for VUR
     - Renal Ultrasound

Discharge Criteria
- Teaching complete
- Supplies ordered
- Car seat approved
- Follow-up appointments scheduled
- Medically cleared by Urology and Orthopedic surgery

Last Updated: January 2023
Next Expected Review: May 2023

Bladder Exstrophy Transfusion Guideline

Laboratory Monitoring

- Check blood gas every hour
- Check CBC and coags after osteotomies are complete or if concern for hemodynamic instability
- Consider TXA per formulary for high-risk patients (i.e., high blood loss expected, malnourished, history of prematurity)

Lab Results	**Intervention**
Hct < 21 or hemodynamic instability	PRBC 10-15 mL/kg
Platelets < 100k	Platelets 5 – 10 mL/kg
Fibrinogen < 150	Cryoprecipitate 2 - 5 mL/kg
TEG > 10 or INR > 1.5	*FFP 10-15 mL/kg*

Job Aid: Blood Transfusions – Transfusion and Dosing, 12056 (*for SCH only*)

Return to Operative

Bladder Exstrophy Outside Provider

Management of the bladder plate

- The goal is to keep the bladder plate clean and moist
- Tegaderm should be placed over the bladder plate overlapping with the surrounding skin
- Change the tegaderm prn dislodgement or soiling
- Clean the bladder plate with saline flush if soiled
- The child can be bathed but the bladder plate should not be scrubbed

Management of the umbilical stump

- Silk suture should be used for the umbillicus
- This is preferred over the umbi clamp to reduce irritation of the bladder mucosa

Other Considerations

- Provider to provider consultation is available
- If child is stable:
  - No need to transfer until mom can also be discharged
  - Normal diet
  - No antibiotics
  - No activity restrictions

Bladder Exstrophy Approval & Citation

Approved by the CSW Bladder Exstrophy team for May 22, 2018 go-live

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Retrieval Website: http://www.seattlechildrens.org/pdf/bladder-exstrophy-pathway.pdf

Please cite as:
Seattle Children's Hospital, Merguerian P, Rowe C, Dearn J, Herrman A, McMannis N, Nelson V, Popalisky J, Sheets E, Turner A, Villavicencio C, 2018 May. Bladder Exstrophy Pathway. Available from: http://www.seattlechildrens.org/pdf/bladder-exstrophy-pathway.pdf

Evidence Ratings

This pathway was developed through local consensus based on published evidence and expert opinion as part of Clinical Standard Work at Seattle Children's. Pathway teams include representatives from Medical, Subspecialty, and/or Surgical Services, Nursing, Pharmacy, Clinical Effectiveness, and other services as appropriate.

When possible, we used the GRADE method of rating evidence quality. Evidence is first assessed as to whether it is from randomized trial or cohort studies. The rating is then adjusted in the following manner (from: Guyatt G et al. J Clin Epidemiol. 2011;4:383-94, Hultcrantz M et al. J Clin Epidemiol. 2017;87:4-13.):

Quality ratings are *downgraded* if studies:

- Have serious limitations
- Have inconsistent results
- If evidence does not directly address clinical questions
- If estimates are imprecise OR
- If it is felt that there is substantial publication bias

Quality ratings are *upgraded* if it is felt that:

- The effect size is large
- If studies are designed in a way that confounding would likely underreport the magnitude of the effect OR
- If a dose-response gradient is evident

Certainty of Evidence:

✪✪✪✪ High: The authors have a lot of confidence that the true effect is similar to the estimated effect
✪✪✪○ Moderate: The authors believe that the true effect is probably close to the estimated effect
✪✪○○ Low: The true effect might be markedly different from the estimated effect
✪○○○ Very low: The true effect is probably markedly different from the estimated effect
Guideline: Recommendation is from a published guideline that used methodology deemed acceptable by the team
Expert Opinion: Based on available evidence that does not meet GRADE criteria (for example, case-control studies).

Summary of Version Changes

- **Version 1.0 (05/22/2018):** Go live
- **Version 2.0 (2/21/2020):** Clarified Follow-up Coordination section on Post-Operative page
- **Version 2.1 (1/31/2023):** Removed Oxybutynin doing frequency (TID) under Post-Operative Phase IV. Removed TXA dosing and replaced with reference to formulary. Added Blood Transfusions job aid link to Transfusion Guideline information page

Medical Disclaimer

Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in treatment and drug therapy are required.

The authors have checked with sources believed to be reliable in their efforts to provide information that is complete and generally in accord with the standards accepted at the time of publication.

However, in view of the possibility of human error or changes in medical sciences, neither the authors nor Seattle Children's Healthcare System nor any other party who has been involved in the preparation or publication of this work warrants that the information contained herein is in every respect accurate or complete, and they are not responsible for any errors or omissions or for the results obtained from the use of such information.

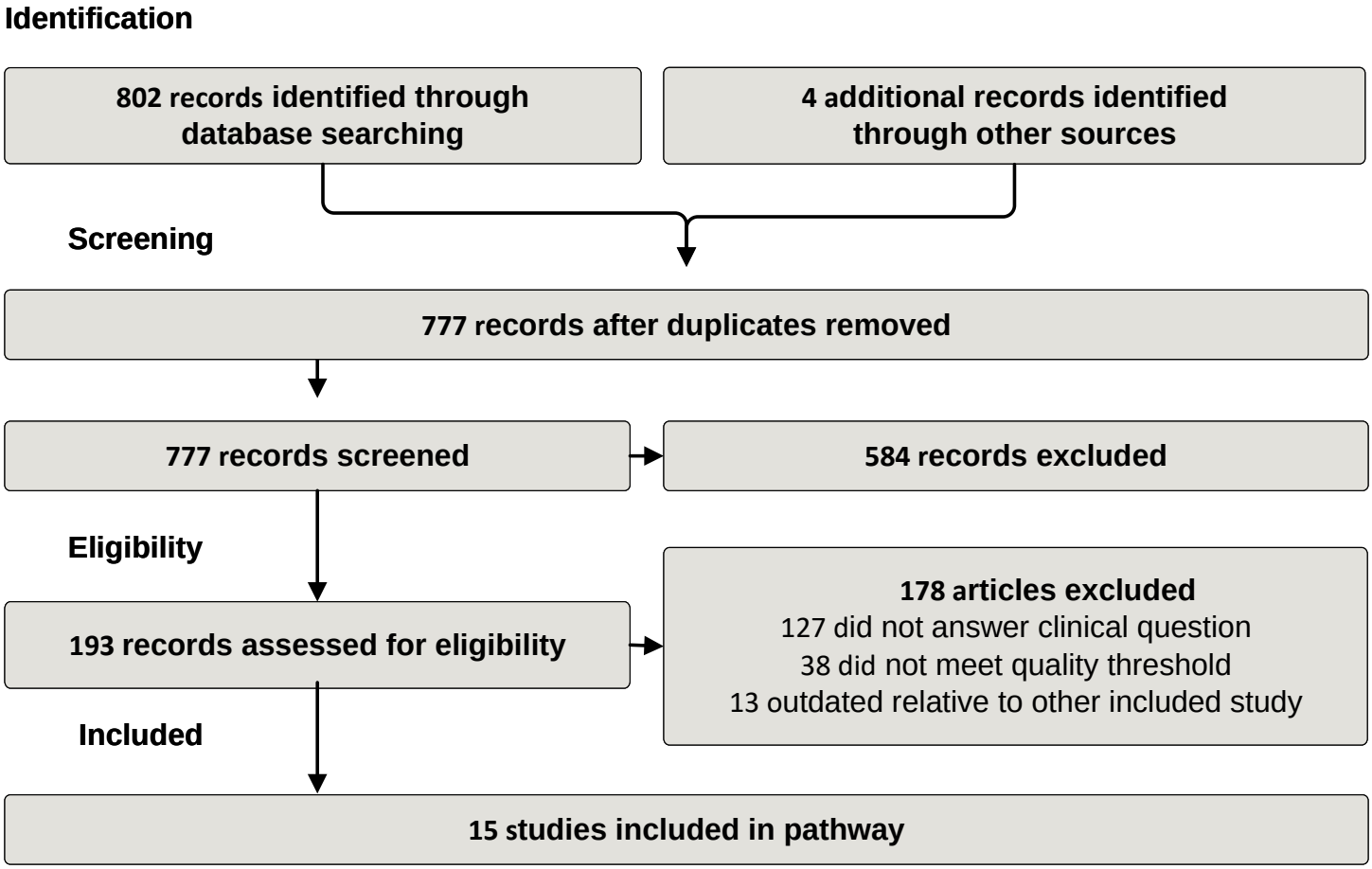
Readers should confirm the information contained herein with other sources and are encouraged to consult with their health care provider before making any health care decision.

Bibliography

Search Methods, *Bladder Exstrophy* Clinical Standard Work

Literature searches were conducted in two phases and executed by a medical librarian, Jackie Morton. The initial search in June 2017, targeted synthesized literature on bladder exstrophy, pelvic fixation methods, the use of anticholinergics, transfusion criteria, as well as the use of antibiotics and pain control and surgical recovery. It was executed in Ovid Medline, Cochrane Database of Systematic Reviews, Embase, National Guideline Clearinghouse and TRIP. The second search, in September 2017, was conducted in Medline and Embase to retrieve primary studies, focusing on pelvic fixation methods, transfusion criteria and the use of anticholinergics or any additional studies on bladder exstrophy. All searches were limited to items published in English, from Jan 2007 to date. The team added 3 citations not retrieved with the search strategy scope and limits. Results were exported to RefWorks for system de-duplication, then to Excel for the screening process.

Jackie Morton, MLS
April 30, 2017



Flow diagram adapted from Moher D et al. BMJ 2009;339:bmj.b2535

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