

Bladder Exstrophy v1.0: Prenatal

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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)

!

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

Go to Neonatal
Phase

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PHASE II

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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

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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 lvs
- 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

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Fluid goal is euvolemia

Go to Post-Operative Phase

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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@maintenance IV+PO
- CBC, Chem7, Cr/BUN POD1
- Regular diet immediately after surgery

Medications

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

Wound Care

- Monitor incisions for early signs of infection
- GOC: Casts including Spica Casts (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](#)

Follow-up Coordination

- Urology weekly until spica removed
- 6 week visit for catheter and spica removal
 1. Orthopedics clinic visit for spica removal
 2. Radiology:
 - Pelvic X-Ray to evaluate osteotomies
 - Cystogram for bladder integrity; if (-), VCUG for VUR
 - Renal Ultrasound
 3. Urology clinic visit for SPT removal and initiation of prophylactic antibiotics if evidence of VUR on VCUG

Supplies and teaching (care coordination to initiate)

- Appropriate car seat
- Cather flush
- Petals / Mole skin
- Diapers

Discharge Criteria

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

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 for high risk patients (ie high blood loss expected, malnourished, history of prematurity)
- **50 mg/kg** (maximum 2000 mg) intravenous over **15** min followed by 5 mg/kg/hour continuous I.V. infusion until skin closure

Lab Results

Hct < 21 or hemodynamic instability

Platelets < 100k

Fibrinogen < 150

TEG > 10 or INR > 1.5

Intervention

PRBC 10-15 mL/kg

Platelets 5 – 10 mL/kg

Cryoprecipitate 2 - 5 mL/kg

FFP 10-15 mL/kg

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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 umbilicus
- 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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Medical Staff Services, Owner
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Retrieval Website: <http://www.seattlechildrens.org/pdf/bladder-exstrophy-pathway.pdf>

Please cite as:

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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.):

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

Guideline – Recommendation is from a published guideline that used methodology deemed acceptable by the team.

Expert Opinion – Our expert opinion is based on available evidence that does not meet GRADE criteria (for example, case-control studies).

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Summary of Version Changes

- **Version 1.0 (05/22/2018):** Go live

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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.

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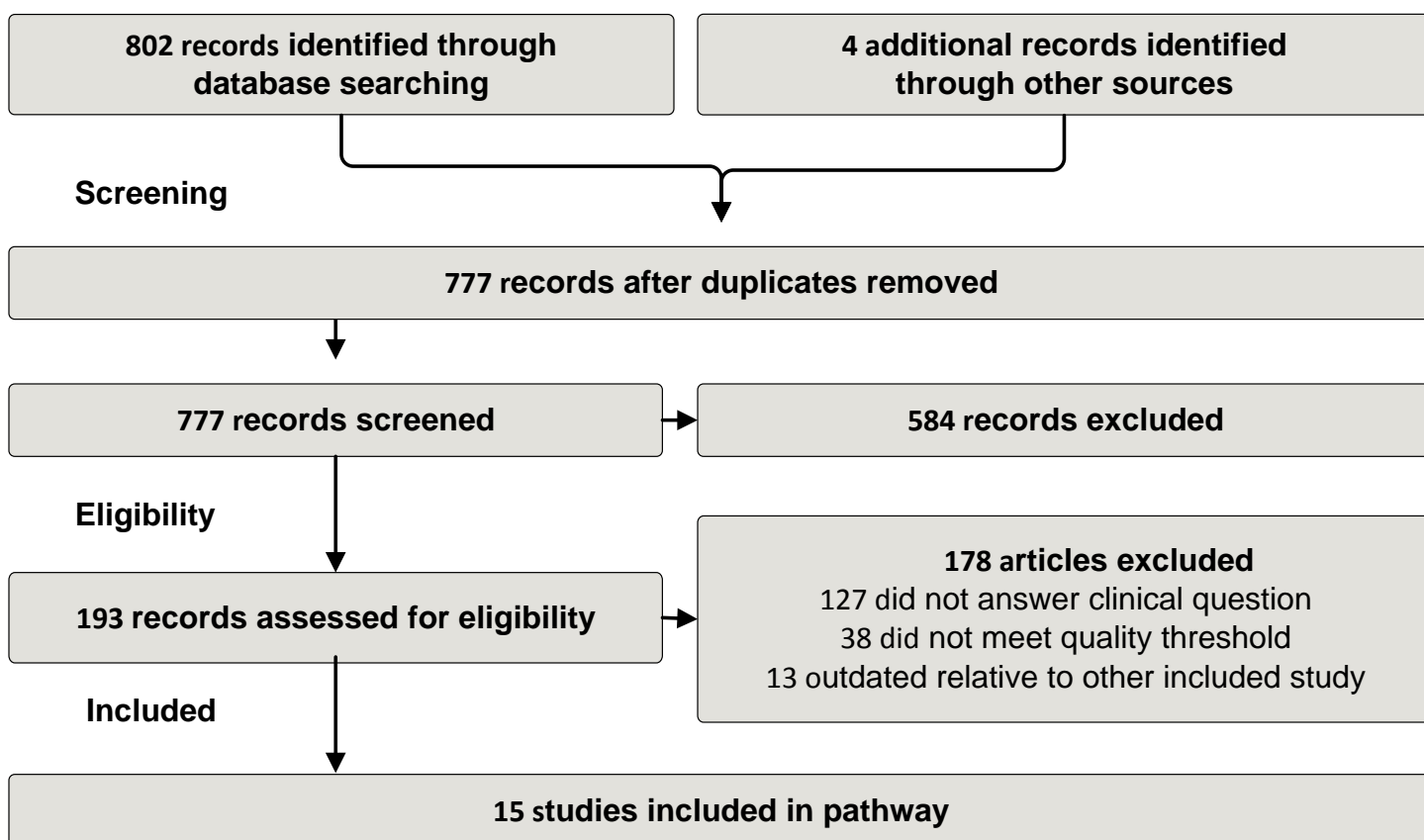
Bibliography

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

Identification



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

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