Complete Primary Repair of Exstrophy: Pitfalls and Practicalities

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Session Structure

- **Topic 1 Discussion:**
  - Goals of Complete Primary Repair of Exstrophy
  - Pitfalls of CPRE

- **Case discussion:**
  - Case 1: near miss – glans ischemia
  - Case 2: Protocol change to address the near miss

- **Topic 2 Discussion:**
  - Utility/Futility of ureteral reimplant during CPRE

- **Case Discussion:**
  - Case 1: Cephalo-trigonal reimplants at time of surgery
  - Case 2: Delayed cephalo-trigonal reimplants
Hendren Project Webinar Goals

• Audience participation!
  • Makes session more dynamic, interesting, and valuable
  • Participate in the Question polls--all answers are anonymous!

• Please use the COMMENTS & QUESTIONS box (located on the right side of the screen) to ask as many questions and to offer as many opinions as possible!
Background: The Delayed Complete Repair

- CPRE described in 1998 by Mitchell
  - Closure of bladder
  - Reconstruction of bladder neck
  - Urethroplasty
  - Epispadias repair

- Goal:
  - Create appropriate bladder outlet resistance
  - Allow for normal bladder cycling from early age
  - Allow for optimal bladder growth and development
Background – Concerns with CPRE

• Immediate concerns:
  • Dehiscence
  • Penile Ischemia → Glans/Corporal Injury
  • Balance between too tight and too loose (bladder neck)

• Later concerns:
  • High volume reflux
  • Persistent incontinence
Question 1:

• How do you close a primary bladder exstrophy?
  A. CPRE
  B. Modern staged repair
  C. Kelly procedure
  D. Primary diversion (Mainz II)
Question 2:

- What is your biggest concern during a CPRE?
  - A. Bladder dehiscence
  - B. Glans ischemia
  - C. Urethral fistula
  - D. Penopubic fistula
  - E. Lack of bladder growth over time due to poor outlet resistance
Topic 1: Penile ischemia
Early Experiences: Big Concerns

• Two dehiscences
  • both in boys without osteotomy

• Two glans/corporal injuries
  • one following separation
  • one in 6 month old with epispadias who had the symphysis split as part of the repair without osteotomy

• Take aways:
  • CPRE can be really dangerous
  • Osteotomy is critical to safe closure
Glans/corporal injury

Newborn with epispadias:
• Primary closure with epispadias repair
• No osteotomy
• Bladder neck reconstruction
• Complete disassembly with anatomical closure

What do you do to prevent glans and corporal ischemia?

A. Don’t close pubic symphysis  
B. Posterior osteotomy  
C. Anterior osteotomy  
D. Leave a gap in pubis if there is sign of ischemia  
E. Sprinkle papaverine on penis during closure
“Be No” Event

• How to avoid the “Be No” Event
  • Keep Glans intact
  • Tempered closure of pubis
  • Incise glans to watch for bleeding
  • BE AWARE of the potential, and open the pubis if the glans appears dusky
A Cautionary Tale

PUBLIC CLOSURE DURING A COMPLETE PRIMARY REPAIR OF BLADDER EXSTROPHY

CHILDREN’S HOSPITAL OF PHILADELPHIA
Topic 2: CPRE with Bilateral Ureteral Reimplant
The concurrent re-implantation

- **Cephalotrigonal technique**
  - Described by Canning and Jeffs in 1993
- **Done at time of CPRE**
  - Described by Braga and Pippi Salle in 2010
Concurrent CPRE-BUR outcomes

Table 2. Outcome comparison between patients who underwent CPRE with and without ureteral reimplantation

<table>
<thead>
<tr>
<th>Variables</th>
<th>No. CPRE-BUR (%)</th>
<th>No. CPRE (%)</th>
<th>p Value</th>
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<tbody>
<tr>
<td>Postop hydronephrosis grade:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>2 (13)</td>
<td>3 (13)</td>
<td>0.05</td>
</tr>
<tr>
<td>II</td>
<td>—</td>
<td>3 (13)</td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>—</td>
<td>3 (13)</td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>—</td>
<td>1 (4)</td>
<td></td>
</tr>
<tr>
<td>Postop febrile UTIs</td>
<td>1 (7)*</td>
<td>11 (48)</td>
<td>0.01</td>
</tr>
<tr>
<td>Postop VUR</td>
<td>0 (0)</td>
<td>17 (74)</td>
<td>0.004</td>
</tr>
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* Febrile UTI after voiding cystogram.
Question 4:

Do you do a ureteral reimplantation at the time of CPRE?

A. Yes
B. No
C. Depends on bladder size
D. Depends on bladder polyps
## The re-implant question

<table>
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<tr>
<th>PRO</th>
<th>Con</th>
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<tr>
<td>• Allows bladder to cycle efficiently</td>
<td>• Small bladder plate may not allow for long tunnel</td>
</tr>
<tr>
<td>• Avoids progressive ureteral dilation during cycling</td>
<td>• Persistent reflux may occur</td>
</tr>
<tr>
<td>• May prevent UTI’s</td>
<td>• Adds time to already long procedure</td>
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BUR at CPRE – case for and against in Boy and Girl
Thank you for your participation!