

Complete Primary Repair of Exstrophy: Pitfalls and Practicalities

Douglas A. Canning, MD

Joseph G. Borer, MD

Dana A. Weiss, MD

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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:

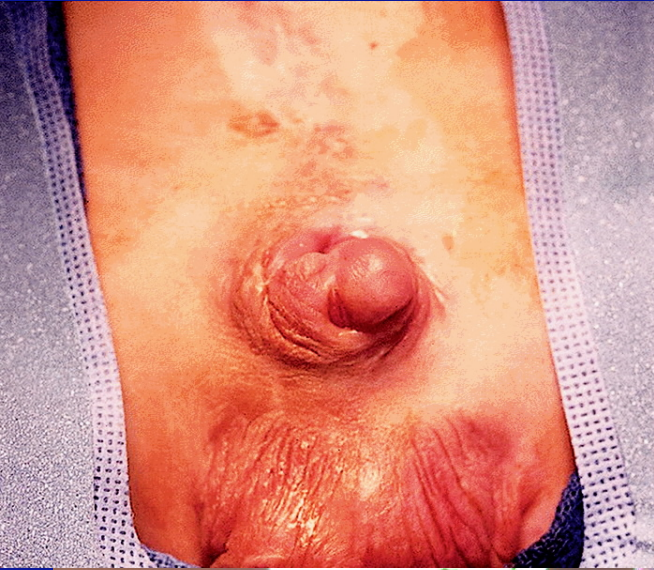
- What is your biggest concern during a CPRE?
 - A. Bladder dehiscence
 - B. Glans ischemia
 - C. Urethral fistula
 - D. Penopubic fistula
 - E. Lack of bladder growth over time due to poor outlet resistance

Topic 1: Penile ischemia

Early Experiences: Big Concerns

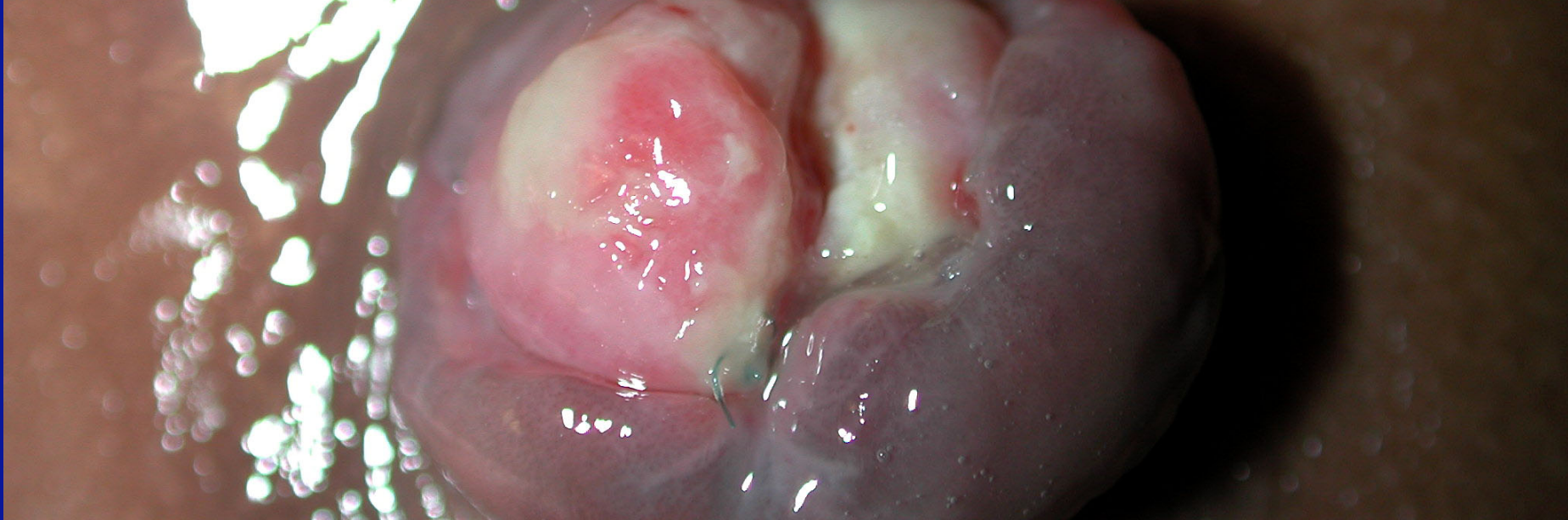
- Two dehiscences
 - both in boys without osteotomy
- Two glans/corporal injuries
 - one following separation
 - one in 6 month old with epispadias who had the symphysis split as part of the repair without osteotomy
- Take aways:
 - CPRE can be really dangerous
 - Osteotomy is critical to safe closure

Glans/corporal injury



Newborn with epispadias:

- Primary closure with epispadias repair
- No osteotomy
- Bladder neck reconstruction
- Complete disassembly with anatomical closure



Question 3:

- 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

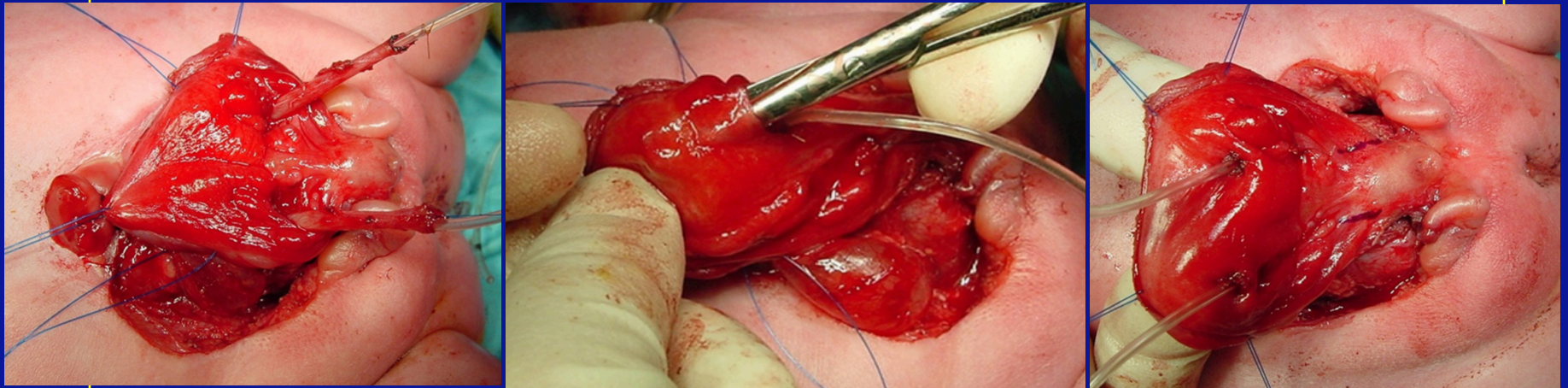
PUBIC CLOSURE DURING A COMPLETE
PRIMARY REPAIR OF BLADDER EXSTROPHY

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

Variables	No. CPRE-BUR (%)	No. CPRE (%)	p Value
Postop hydronephrosis grade:			0.05
I	2 (13)	3 (13)	
II	—	3 (13)	
III	—	3 (13)	
IV	—	1 (4)	
Postop febrile UTIs	1 (7)*	11 (48)	0.01
Postop VUR	0 (0)	17 (74)	0.004

* 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

PRO

- Allows bladder to cycle efficiently
- Avoids progressive ureteral dilation during cycling
- May prevent UTI's

Con

- Small bladder plate may not allow for long tunnel
- Persistent reflux may occur
- Adds time to already long procedure

BUR at CPRE – case for and
against in Boy and Girl

Thank you for your
participation!