Safe and Sound: Principles for Successful Complete Primary Repair of Bladder Exstrophy in the Boy

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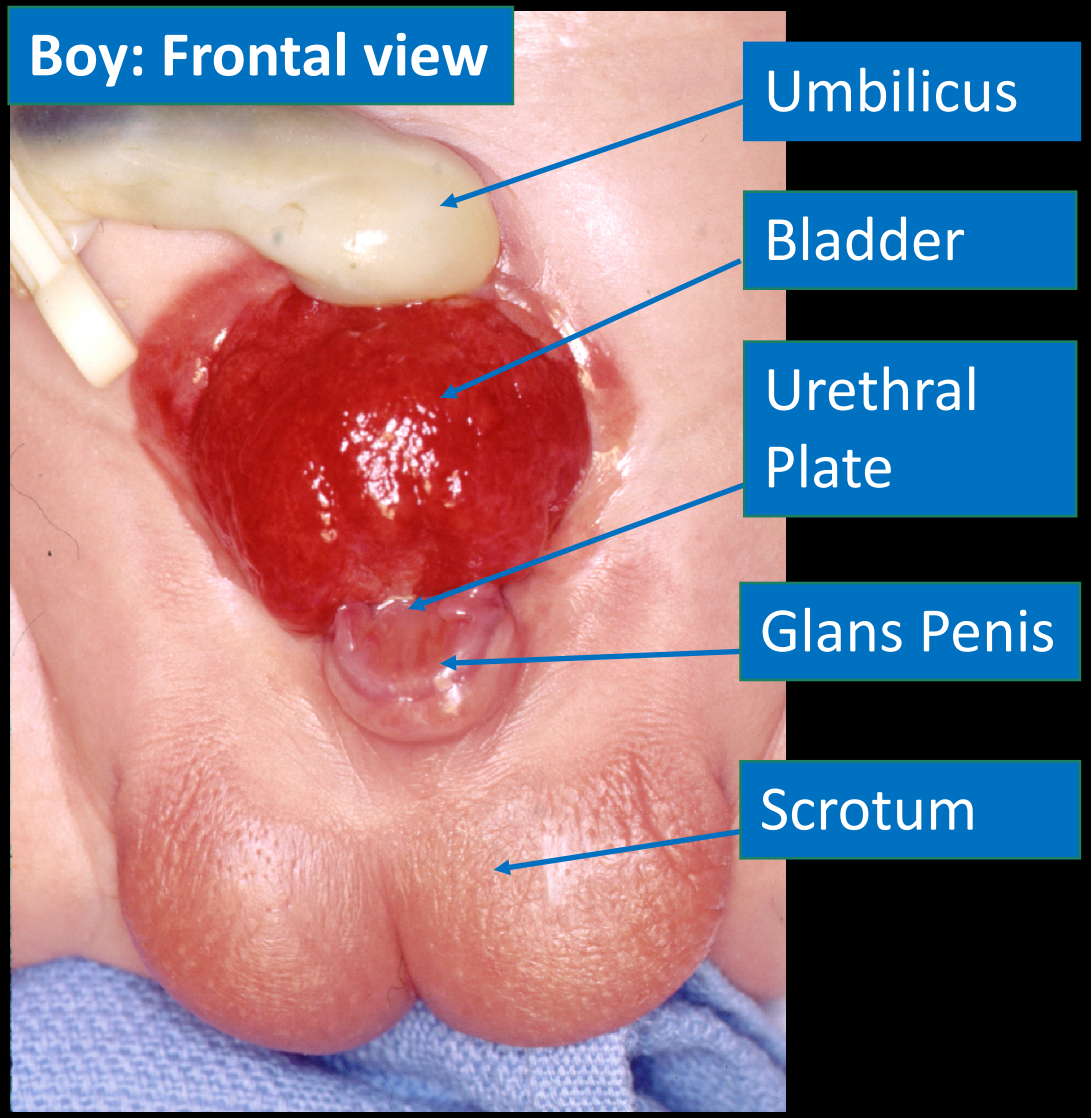
Introduction:

- Bladder Exstrophy is rare and complex
- Presents many challenges to optimal surgical reconstruction



Complete Primary Repair of Bladder Exstrophy (CPRE)

- Closure of bladder, reconstruction of the bladder neck and urethra
- Bilateral pelvic osteotomies
- Appropriate bladder outlet resistance
  - Normal bladder cycling
  - Optimal bladder growth and development

Objectives:

- Detail important elements of CPRE
- Special considerations
  - Increase safety
  - Limit tissue injury
  - Optimize functional and cosmetic outcome

Methods:

- CPRE in consecutive boys with Bladder Exstrophy (BE)
- Follow up per Multi-Institutional Bladder Exstrophy Consortium (MIBEC) IRB protocol

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Methods:

- CPRE at 2-3 months of age
  - Caregiver bonding and appreciation of BE
  - Decrease anesthesia risk
  - Tissue growth
    - Bladder with Valsalva
    - Genitalia growth

Methods:

- Skin hook(s) gentle retraction of tissue vs. forceps and repetitive grasping (i.e. marking of skin)
- Sharp dissection vs. electrocautery
- Intermittent topical dilute epinephrine or fine absorbable suture vs. epinephrine injection or monopolar electrocautery

Methods:

- Urethral tubularization and bladder closure
  - Incorporating serosa with little or no mucosa
  - Inverting mucosa to decrease fistula risk

Methods:

- Exploitation of fat planes to dissect skin from fascia and fascia from bladder (“fat is your friend”)
- Urethral plate dissection with initial ventral approach using manual traction to roll the corpora laterally
- Sharp dissection and bipolar electrocautery facilitates separation of spongiosum from cavernosum

Methods:

- Urethral width carried proximally to level of bladder neck to promote continence
- Glans color and perfusion pre- and post pubis approximation carefully assessed to decrease potential ischemic injury

Tip #1: “Fat is your friend”

(Dr. Michael E. Mitchell)

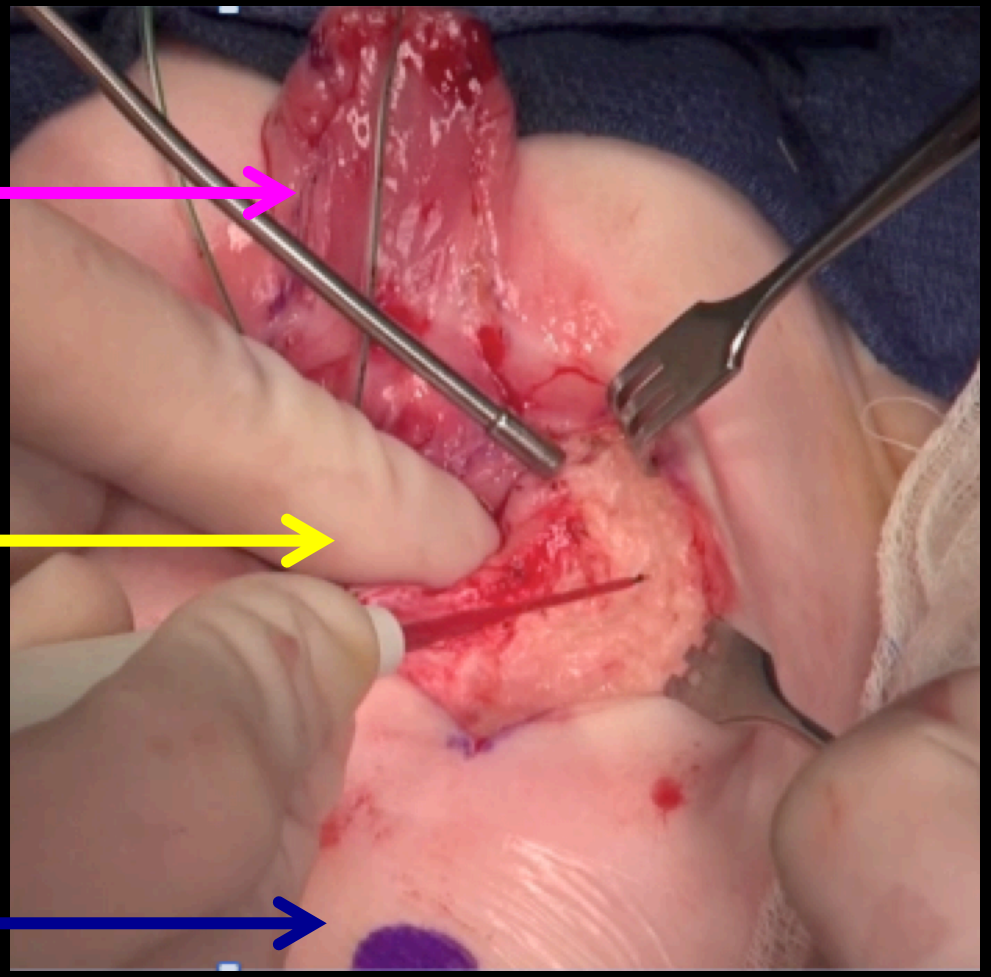
- Referring to initial dissection and development of the plane between skin and anterior rectus fascia
- Clarity in identifying this superficial tissue plane facilitates successful deeper dissection ➔ separation of bladder (detrusor) from rectus fascia and muscle

Tip #1: “Fat is your friend”

Penis

Subcutaneous fat

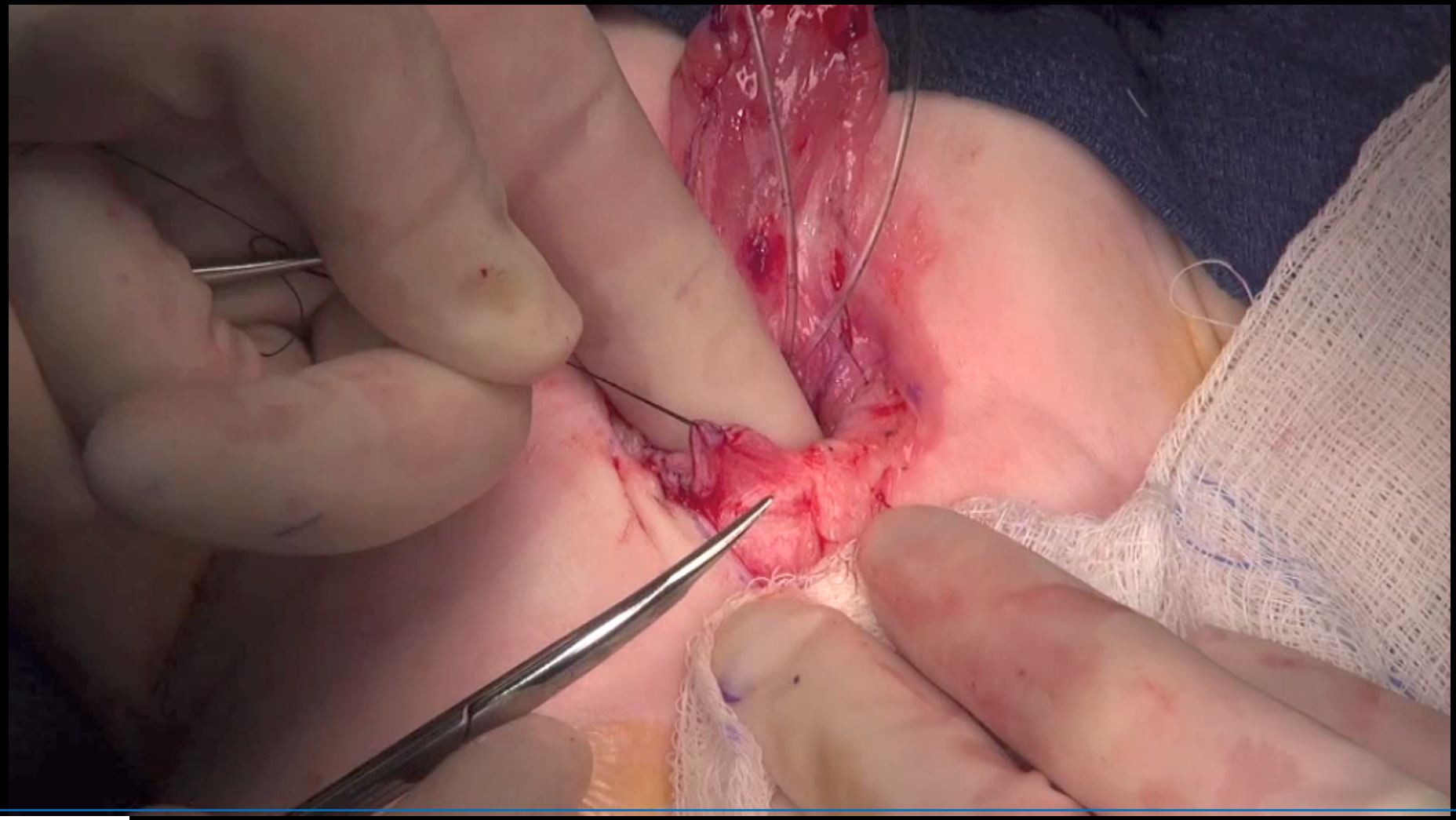
Neo-umbilicus site



Tip #2: Insert finger ➔ invert bladder

- Insertion of the surgeon's non-dominant index finger inverts the bladder and facilitates
  - Accurate identification of the plane between bladder & rectus, and
  - Safe, precise separation of these tissues

Tips #1 & 2: friendly fat & index finger



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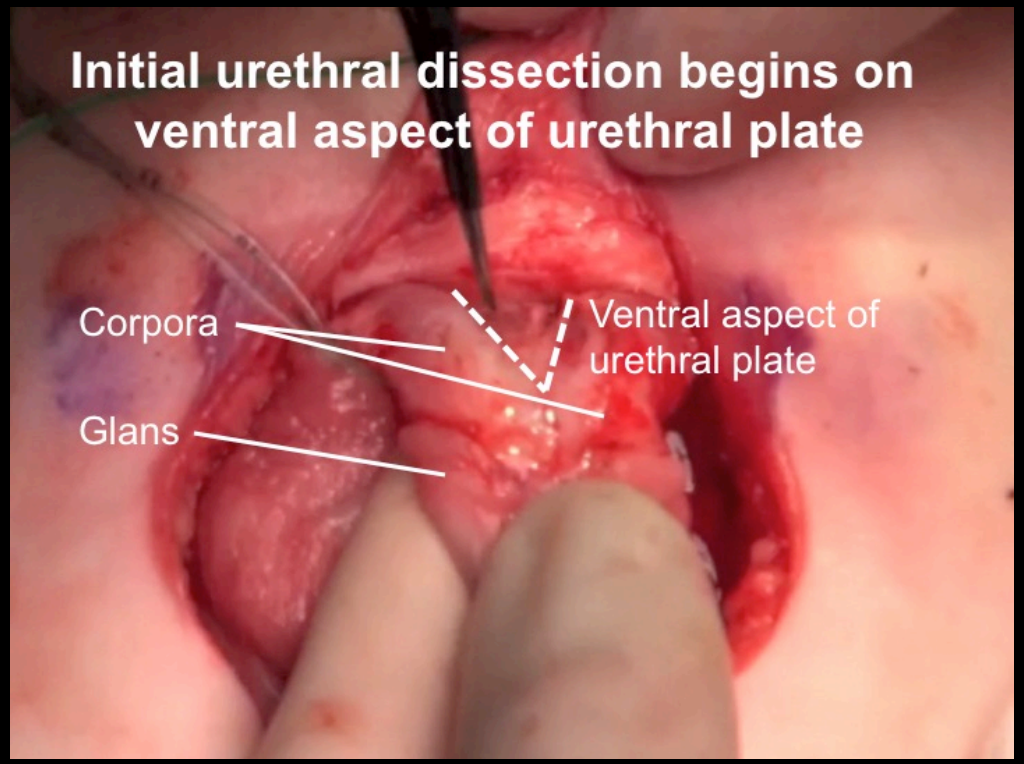
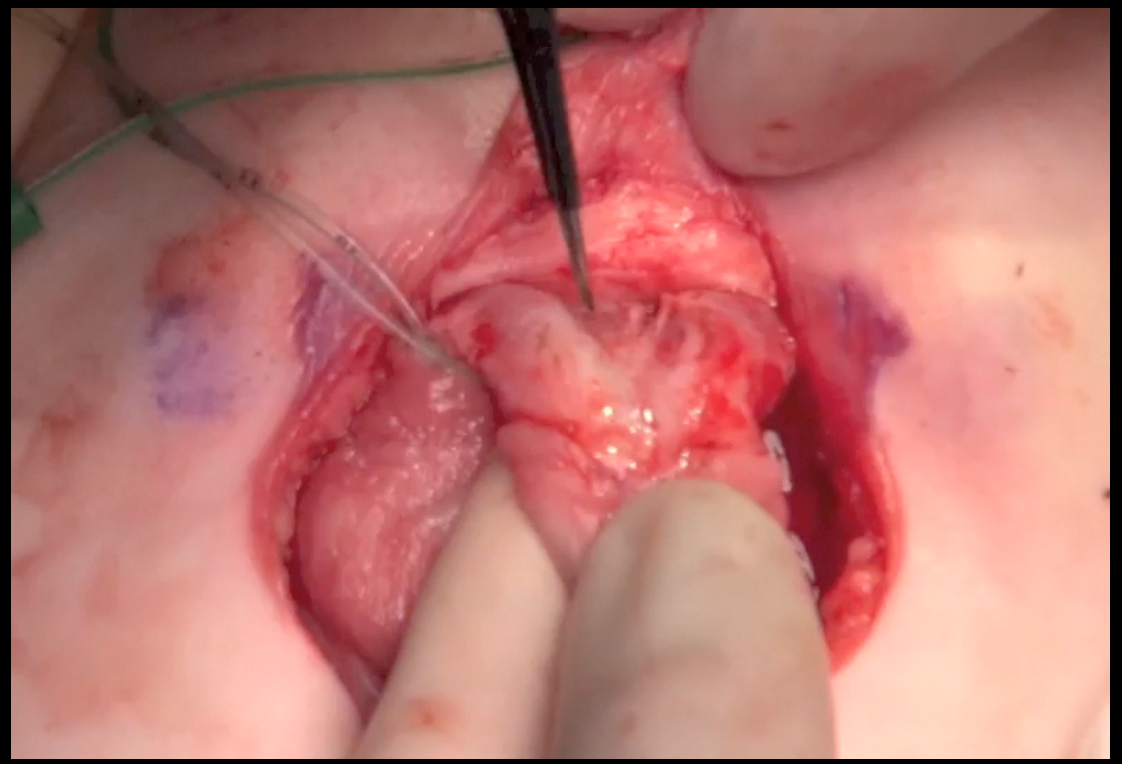
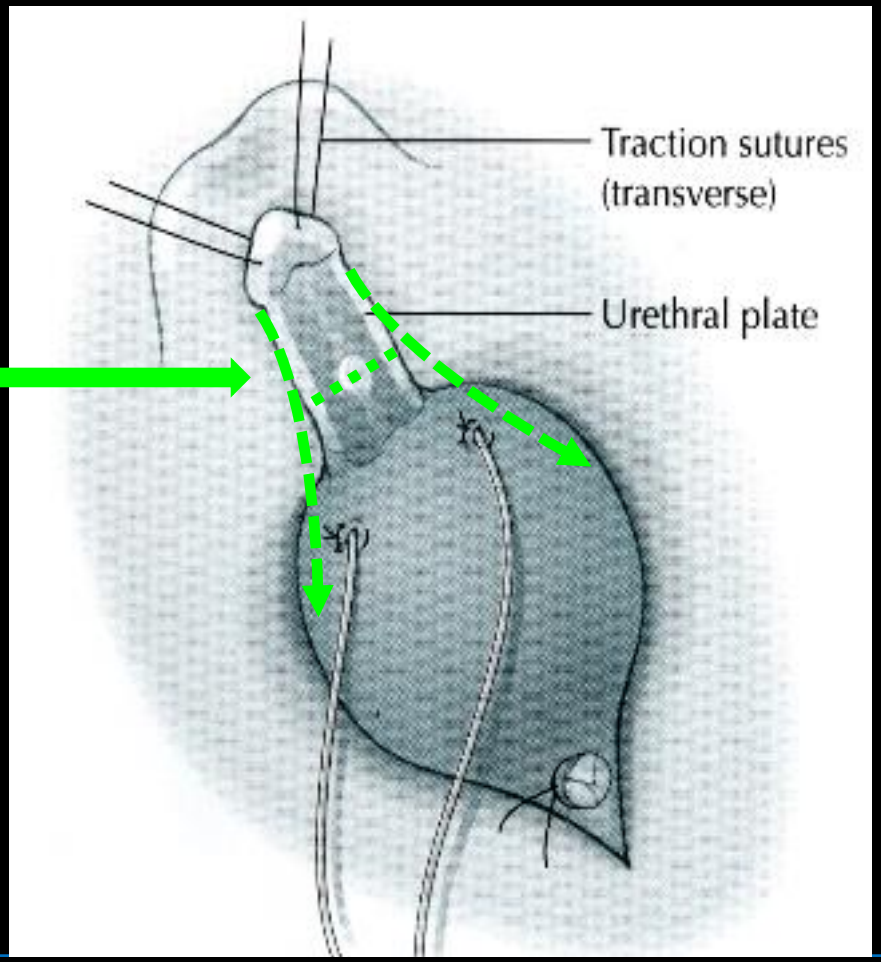
Tip #3: Urethral Plate Dissection
Ventral initially, Bipolar electrocautery





Tip #4: Elongation of urethra/ formation of bladder neck

veru



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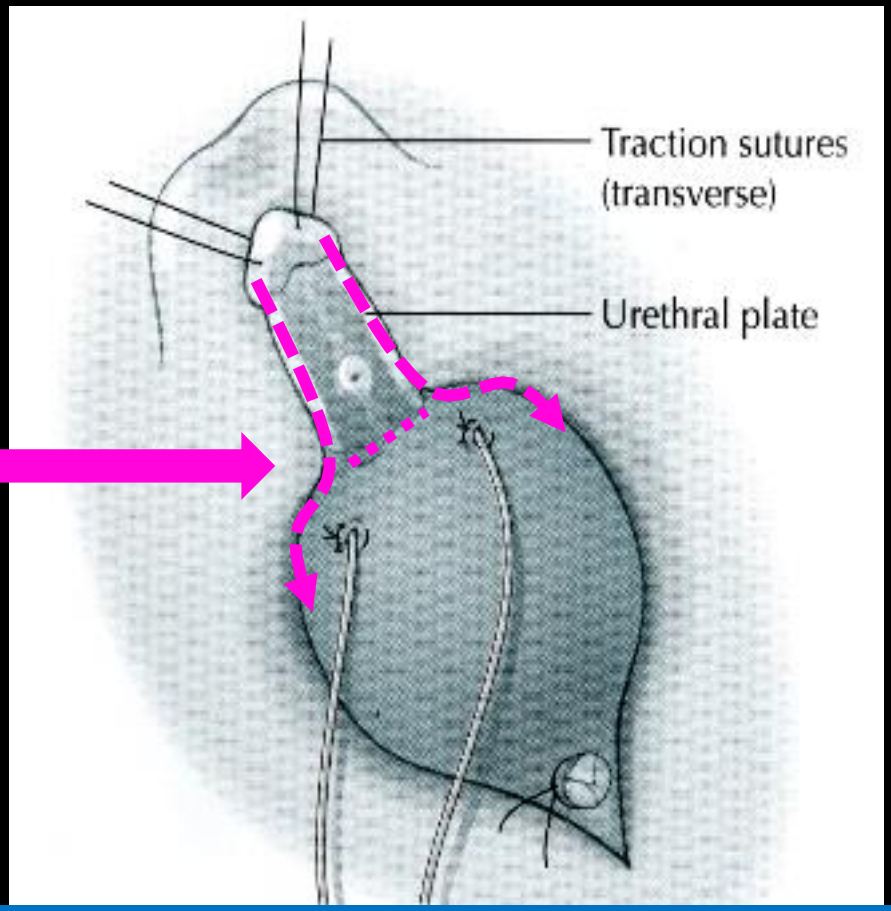

improved continence

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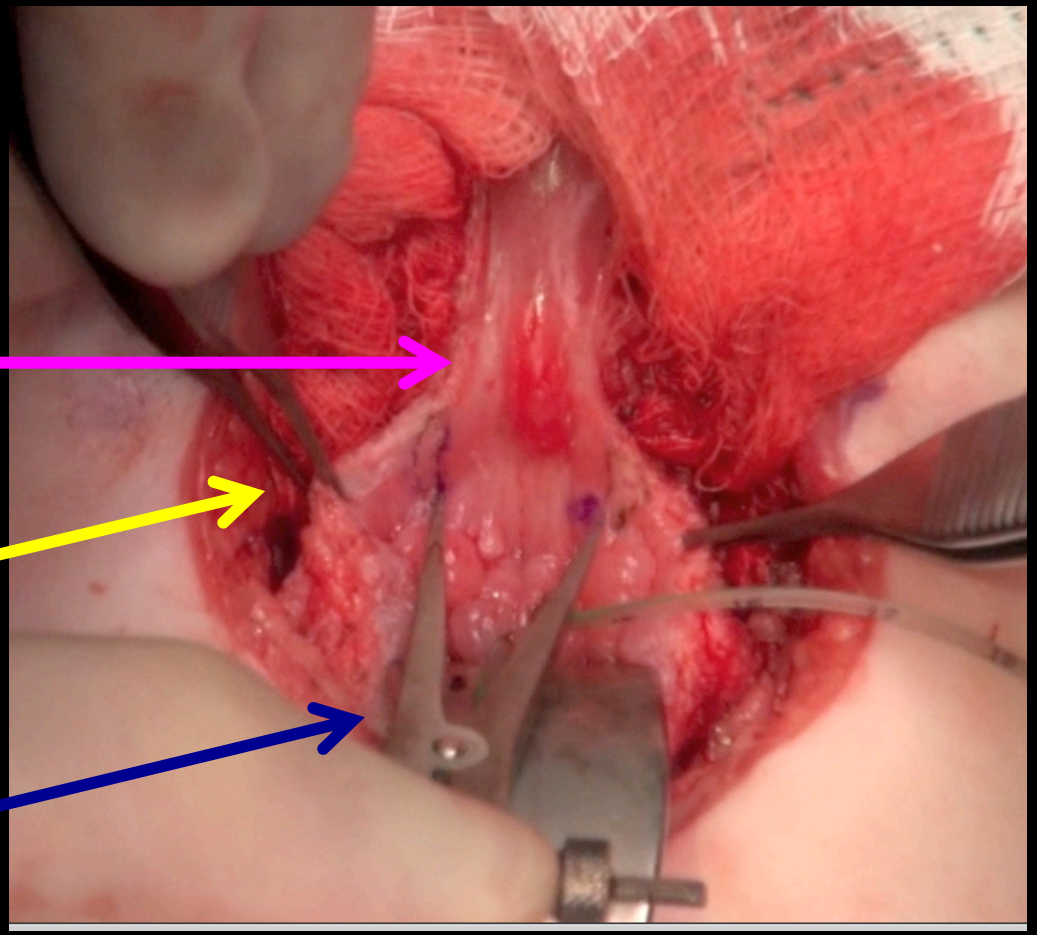
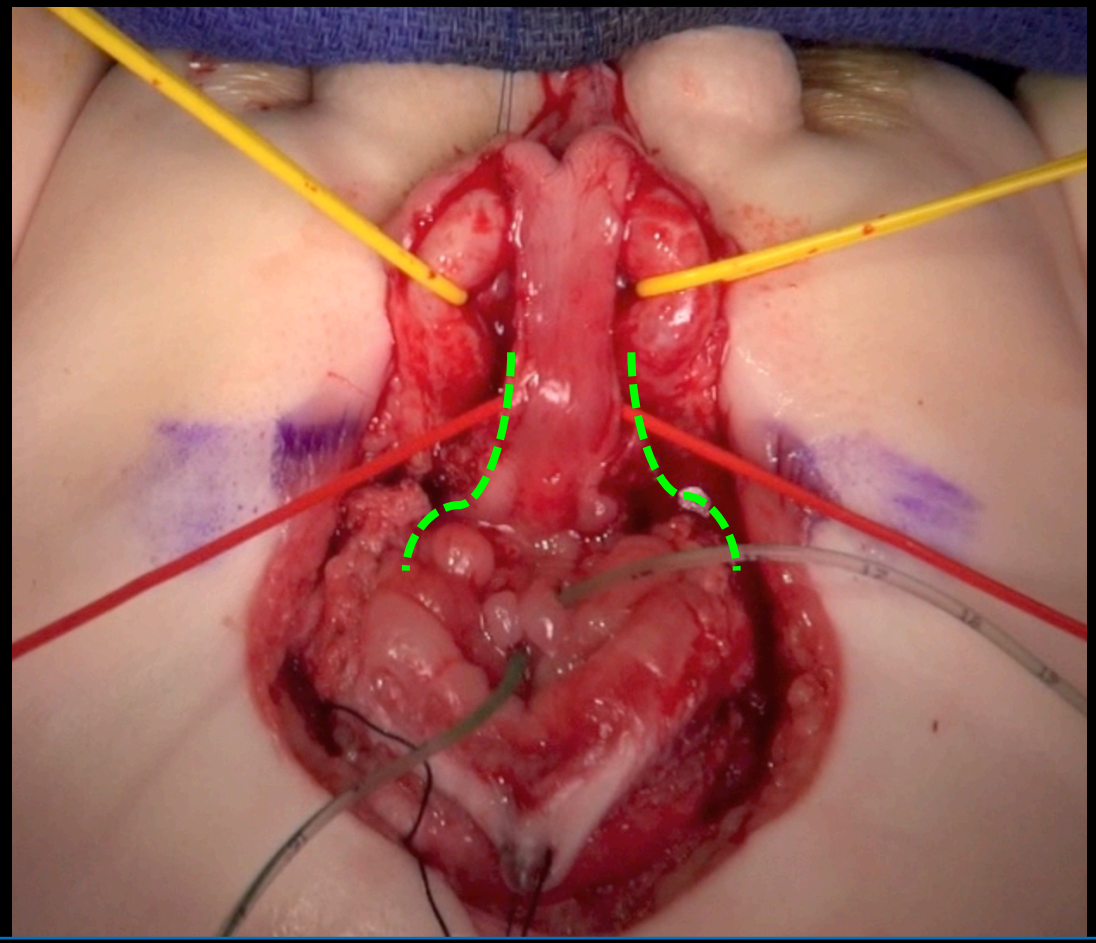
Tip #4: Elongation of urethra/ formation of bladder neck



Tip #5: Proximal dissection of corpora cavernosa

- Scissor or bipolar electrocautery, in plane along Buck's fascia – preserves muscle/ nerves/ prostate
- Carrying this dissection along medial and lateral aspects of corpora cavernosa facilitates tension free approximation of the bladder neck & minimizes injury

Tip #5: Proximal dissection of corpora cavernosa



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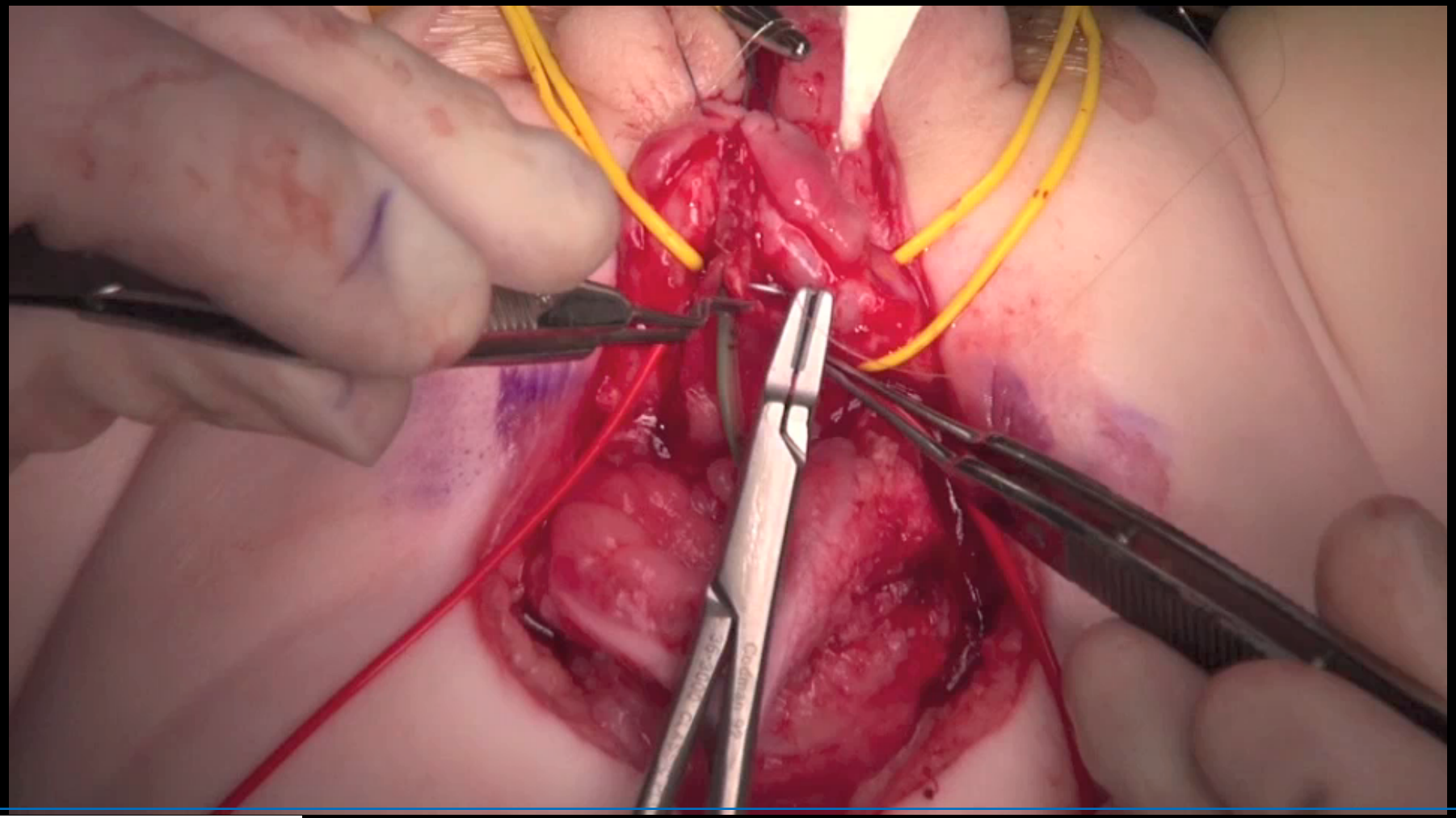
Tip #6: “up and in, down and out”

- Approximation of soft tissues: urethra, bladder neck and bladder
- Passing suture needle in an “up and in (first side), down and out (second side)”
- This trajectory of the needle in order to affect inversion of tissue edges & decrease fistula formation

“up and in-down and out” suturing



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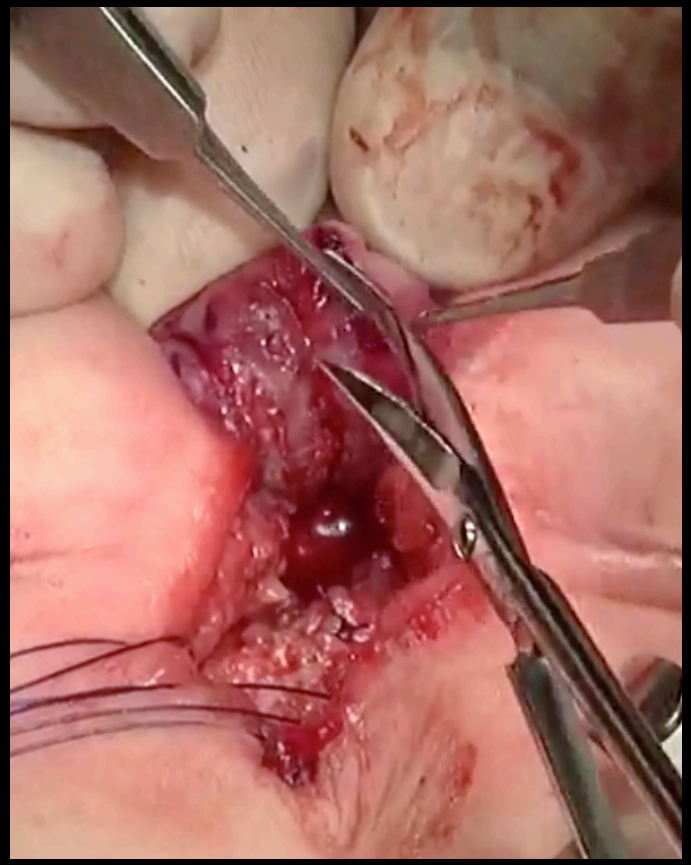
Tip #7: Approximation of pubis

- Careful assessment of color and perfusion, of glans and corpora cavernosa, both immediately prior to and after tying pubis approximating suture(s)
- Decreases risk of impaired perfusion and/ or ischemic tissue loss

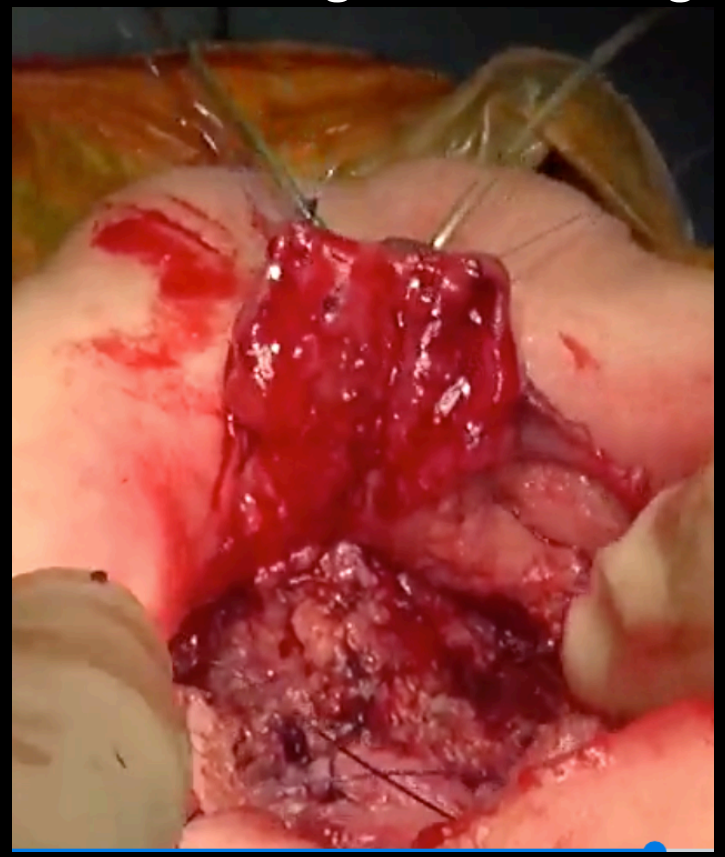
Tip #7: Approximation of pubis

1. pubis sutures tied– glans trimmed– no bleeding



2. decision to cut pubis approximating sutures

3. pubis sutures cut– immediate glans bleeding



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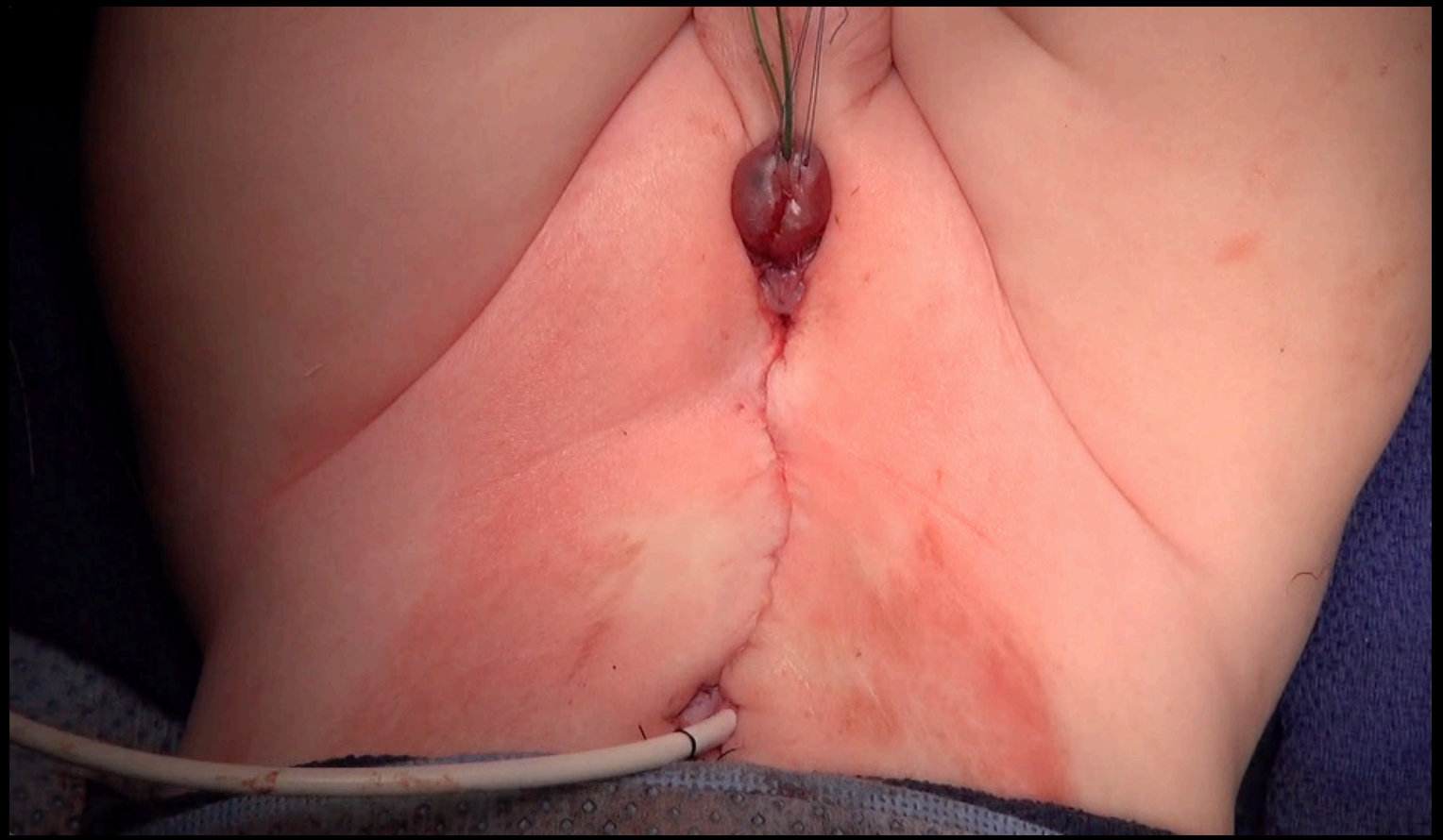
CPRE complete



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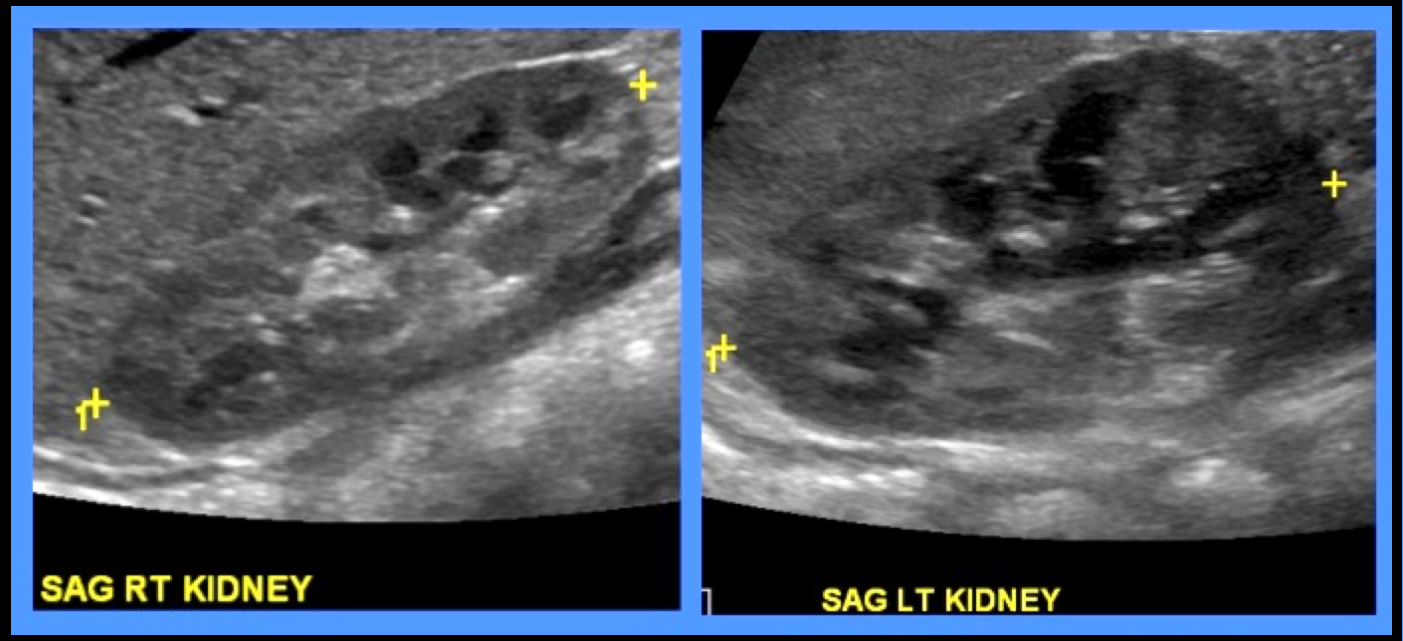
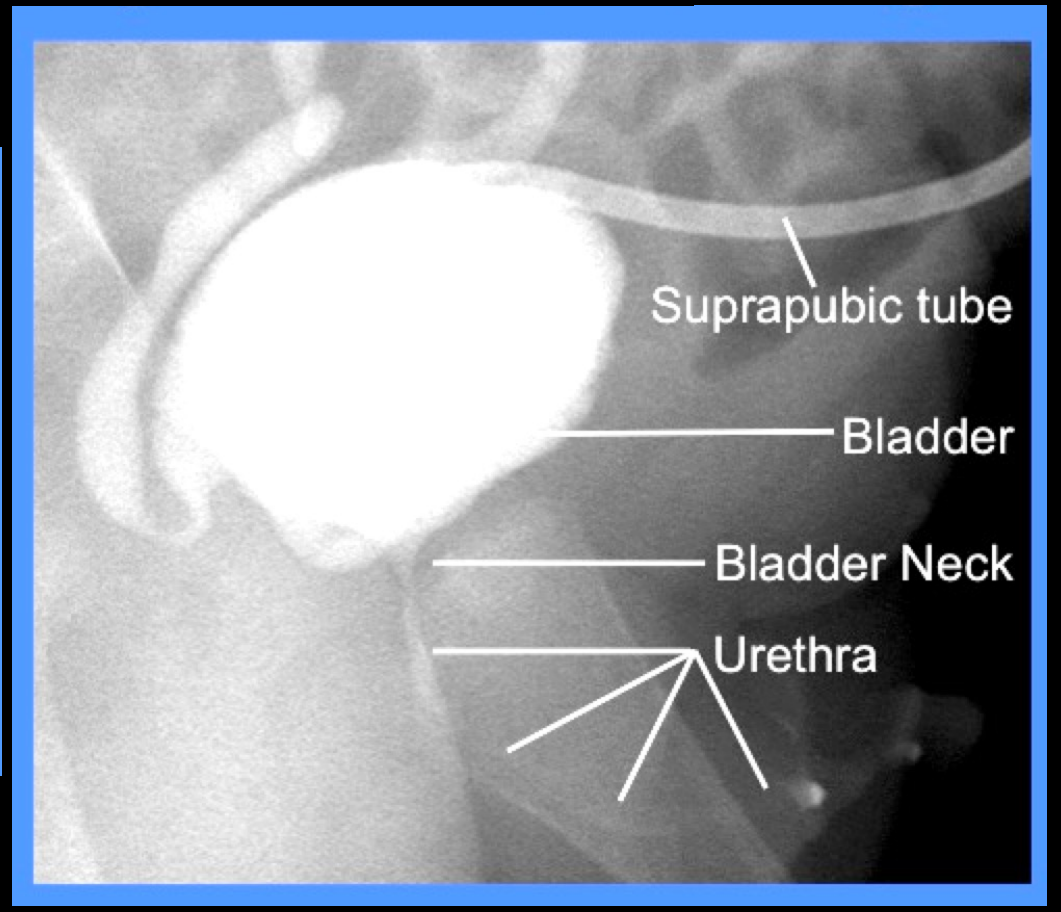
Spica cast

Ultrasound / VCUG 4 wk post-CPRE





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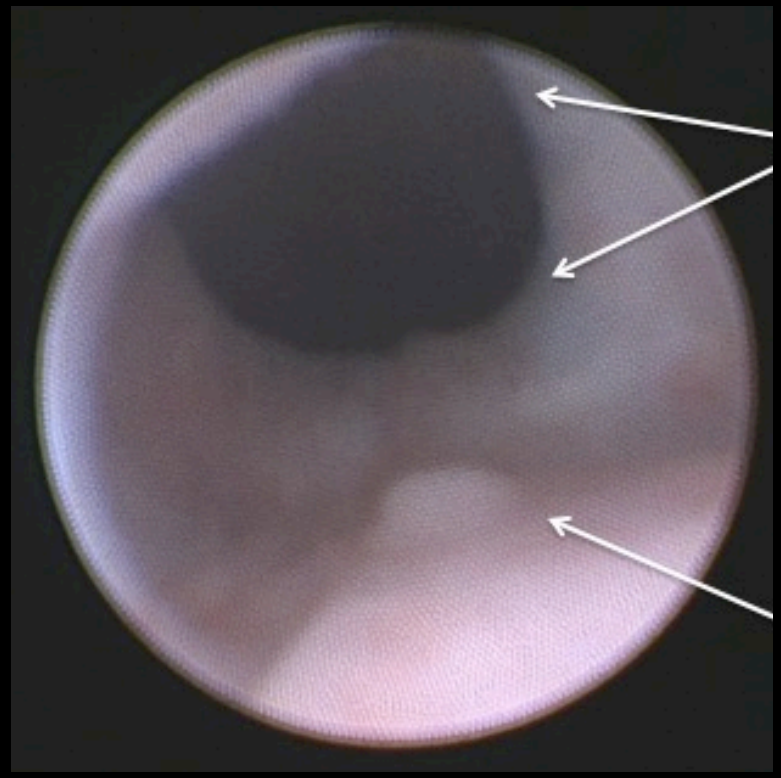
Void and cystoscopy 4 months post-CPRE

Cystoscopy 4 months post-CPRE



February 2013 – September 2017

Institution	Classic BE	Epispadias	Cloacal/Variant	Total Cases
Site 1	20	8	2	30
Site 2	23	5	4	32
Site 3	9	6	2	17
Total	***52***	***19***	***8***	***79***

Results:

- From February 2013 – November 2017
  - Successful CPRE in 29 consecutive boys
  - No bladder dehiscence
  - Penopubic skin loss in 1 boy
  - Hypospadias result in 6 boys

Results: 3 year post CPRE

- Several boys
  - Toilet training success
  - Voiding with stream observed
  - Normal urodynamics
- Urethrocutaneous fistula in 5 boys
  - 2 with large posterior urethra/ penopubic fistula
    - 1 repaired, 1 awaiting repair
- Pyelonephritis in 2 boys
- No complications in spica cast immobilization

Normal voiding 3.5 years post-CPRE

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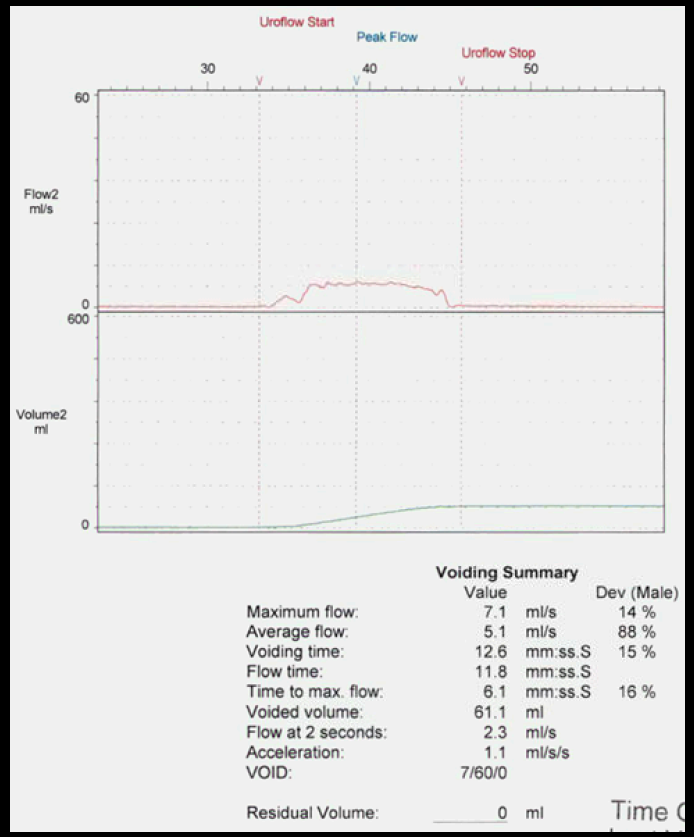
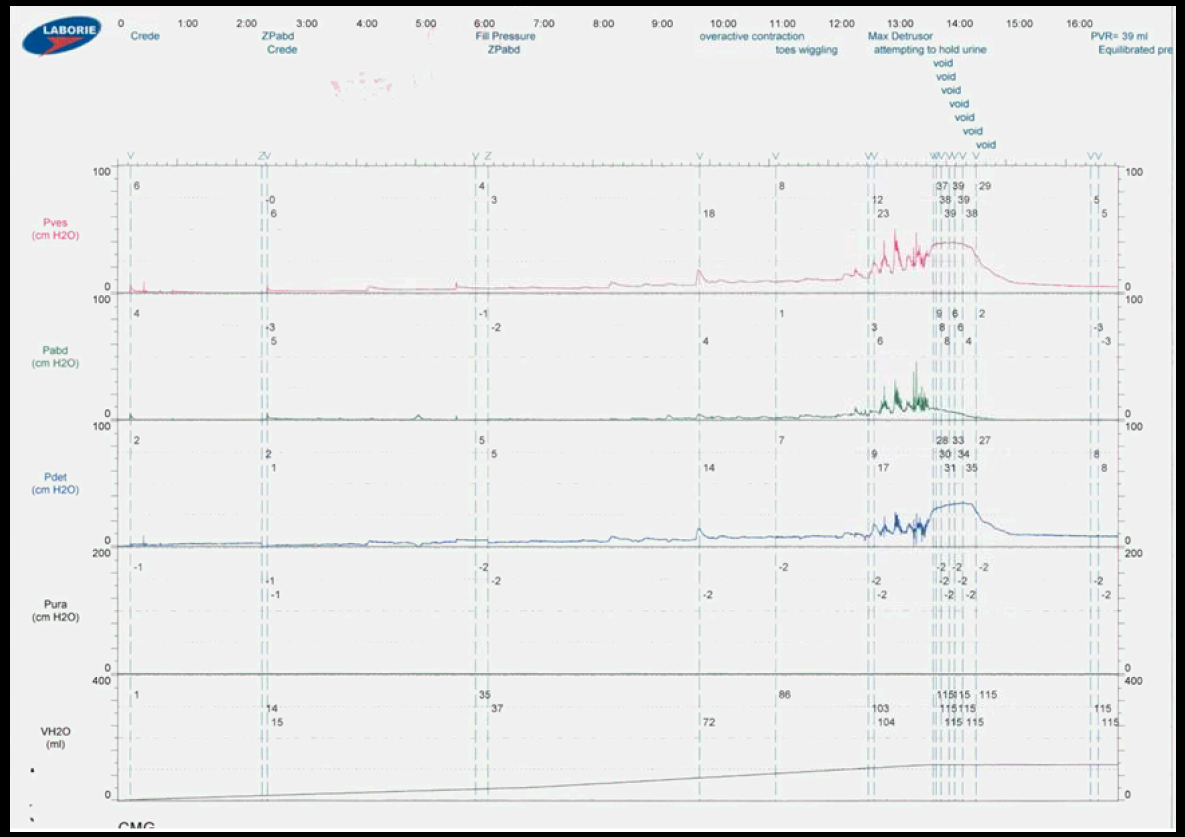
Uroflowmetry and Cystometrogram 3.5 years post-CPRE





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Conclusions:

- Principles developed promote:
  - Safety
  - Limit tissue injury
  - Optimize functional outcomes
  - Optimize cosmetic outcomes
  - Refinement of CPRE technique

Tip #8: Collaborate

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

Charles Darwin

Thank You!