Long-term Bladder Exstrophy Voiding Potential and Sexual Function Outcomes

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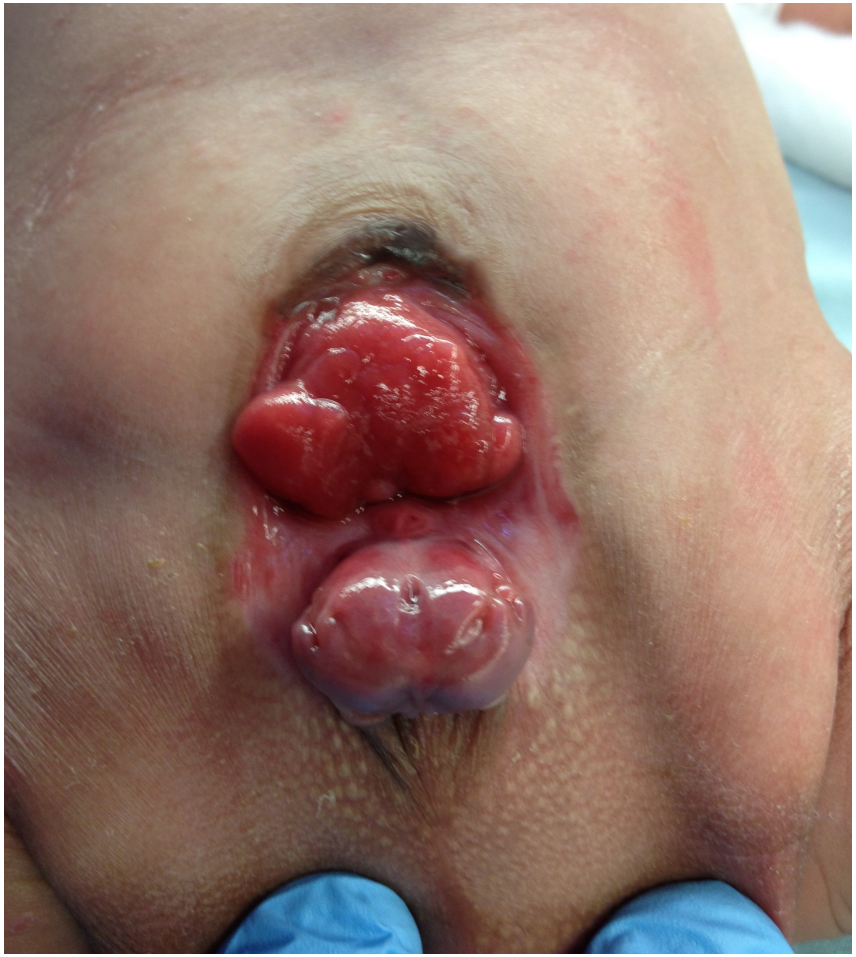
MIBEC
Multi Institutional Bladder Exstrophy Consortium

Incidence of Bladder Exstrophy

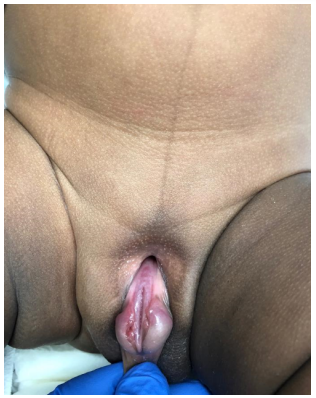
- Epispadias: 1/117,000 (M) and 1/484,000 (F)
- **Bladder Exstrophy: 1/50,000**
- Cloacal Bladder Exstrophy: 1/200,000



Bladder Exstrophy

- Should be thought of as a malformation
- Bladder Exstrophy is a spectrum

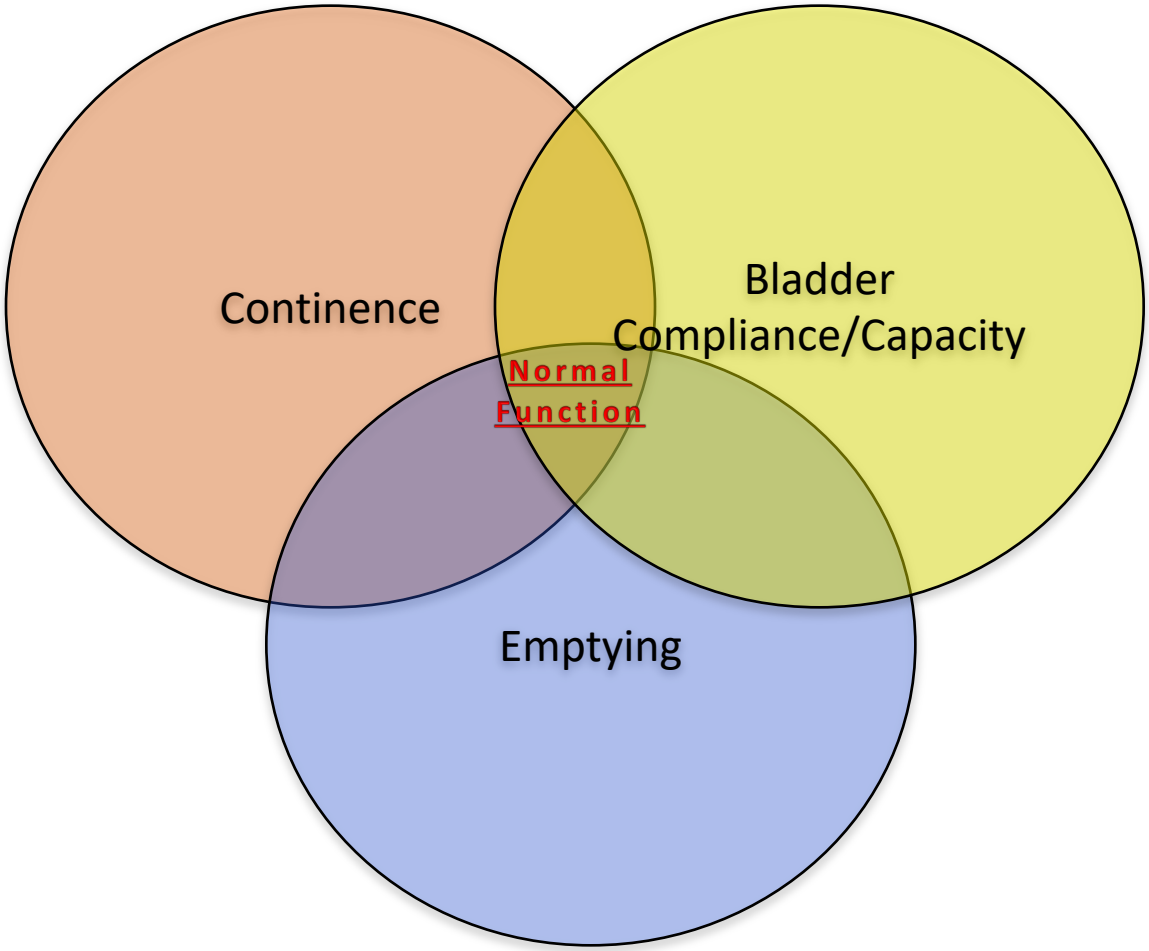
Epispadias → Bladder Exstrophy → Cloacal Exstrophy



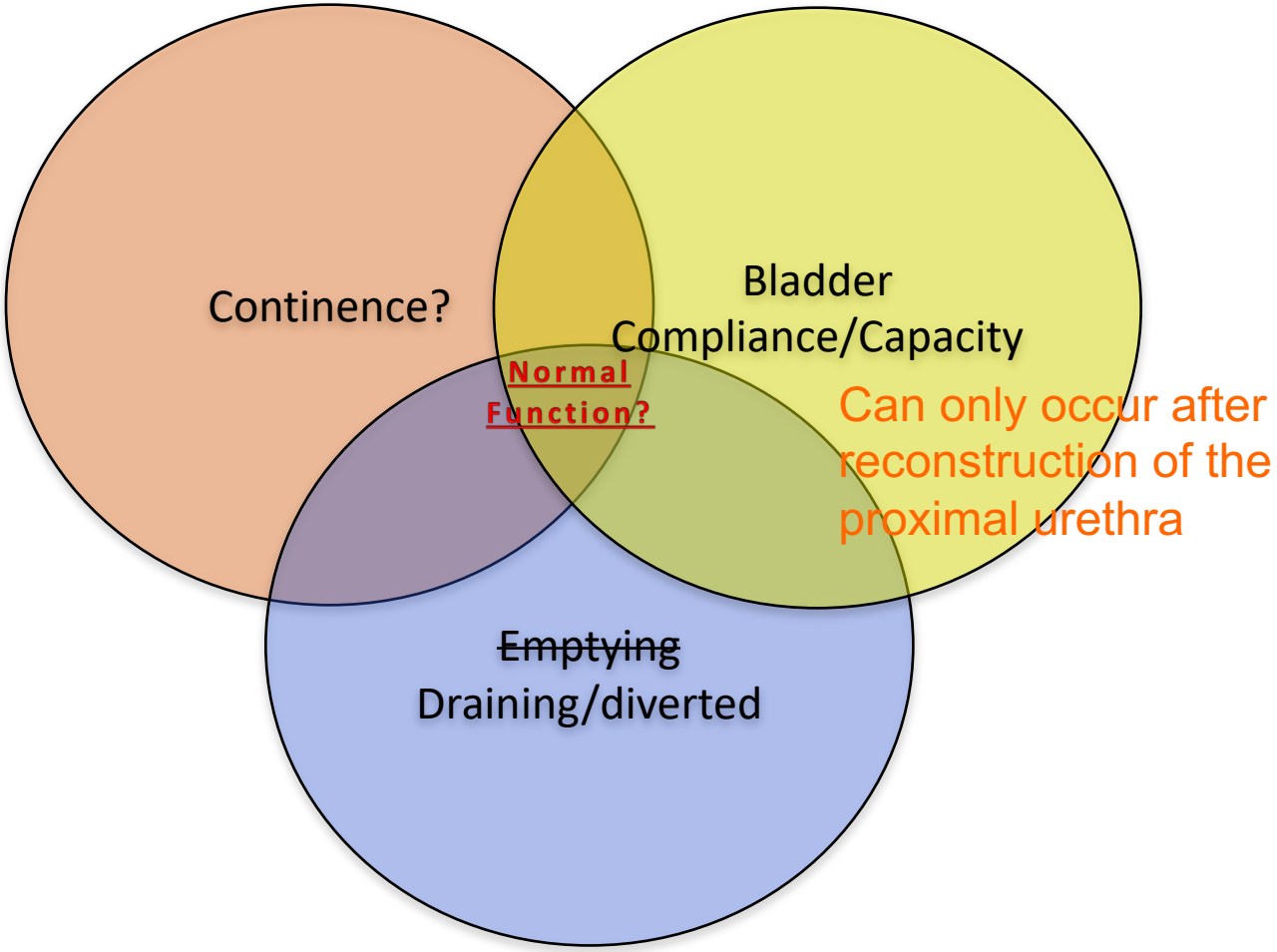
Can we help the Exstophy Bladder reach its potential?

- Goals of reconstruction:
  - Upper tract preservation
  - Anatomic reconstruction: creating functional and cosmetically acceptable external genitalia
  - Creating urinary continence

Development of Normal Bladder Function



Development of Bladder Function In Bladder Exstrophy



Bladder Exstrophy

- Surgical approaches to bladder exstrophy have changed over time
- Modern-Staged Approach Versus CPRE
- No standard surgery exists for bladder exstrophy

How are we doing?

- It depends on how we define success…
- Definition of continence has varied in literature:
  - Dry intervals of 2 hours and volitional voiding
  - Dry intervals of 3 hours or greater
  - Dry intervals with with assistance of CIC

Bladder Exstrophy

- Gearhart reported a single surgeon series of patients who underwent MSRE
  - Bladder closure as Neonate
  - Epispadias repair between 6 & 12 months (Males)
  - BNR 4-5 years
- 67 male and 41 female
- 3-hour dry intervals and urethral voiding
- 47/67 (70%) (M)
- 31/41 (75.6%) (F)

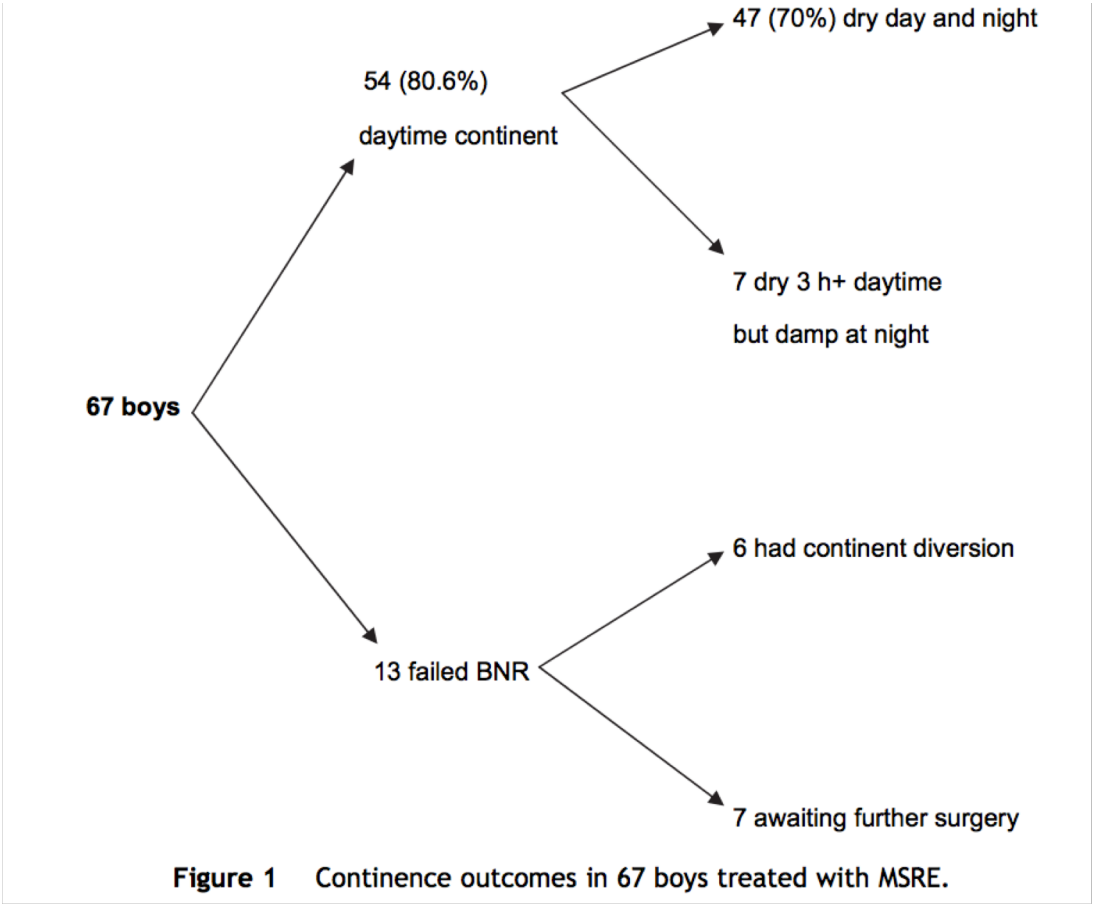


Figure 1 Continence outcomes in 67 boys treated with MSRE.

Purves J Ped Urol 2008

Gearhart et al, J Ped Urol 2007

Long-term Results

- How to define *long term*
- If it is defined as 20 years after repair…
- Repairs would have to have been done in the 1990s or earlier

Long-term Results

- Woodhouse and Redgrave identified 57 patients born between 1965 and 1974
- 13 of 57 children were initially continent and voiding normally at the end of first decade
- By end of the second decade only 3/13 could void (5.3%)
- All other patients needed either CIC with bladder augmentation or bladder closure or bladder neck surgery

Woodhouse and Redgrave BJU 1996

Long-term Concerns?

- If operations on the bladder neck produce continence by fixed obstruction
- Patients bladders would be at risk to deteriorate with time
- Bladders that were normal in childhood might not stay that way into adulthood

The Complete Primary Repair of Exstrophy

- Attempt to reconstruct to "normal anatomy"
- Anatomic Bladder neck dissection
- Proximal urethra placed deep in the pelvic diaphragm

	No. Bladder Exstrophy/Total No.		No. Cloacal Exstrophy/Total No.	
	Boys	Girls	Boys	Girls*
Urinary continence	4/12	4/6	2/4	0/1
Older than toilet training age	4/5	4/5	2/2	0/2
Dry intervals	12/12	6/6	2/4	1/1
Additional continence procedures	3†		1	1

* One girl died at age 2 months.

† Bladder neck reconstruction in 2 boys and augmentation cystoplasty in 1.

J Urol: October 1999

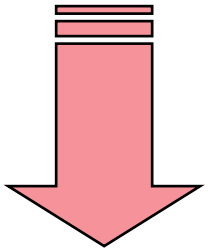
Why CPRE?

- The Bladder may have unique properties for development and healing in the first few months of life
- We know that PUV patients after early valve ablation do better then if they were diverted

Why CPRE?

Hypothesis:

“Normal” Filling and Emptying (Cycling) of the Bladder



Normal Bladder Development

Long-Term Followup of Complete Primary Repair of Exstrophy: The Seattle Experience

Margarett Shnorhavorian,* Richard W. Grady, Amy Andersen, Byron D. Joyner and Michael E. Mitchell

From the Children's Hospital and Regional Medical Center, Seattle, Washington

J Urol 2008

The Seattle Experience

- Since 1989, CPRE was performed
- Follow-up to previous report in 1999
- 39 children underwent CPRE
  - 22 boys and 17 girls
  - Median f/u entire series was 58 months
  - F/u for patients reported in 1999 was 106 months

The Seattle Experience

- Boys and girls over 4 years of age
  - 74% had daytime continence with voiding
  - n=23 patients
- 80% boys required bladder neck reconstruction
- 57% females required bladder neck reconstruction

The Seattle Experience

- Daytime continence was defined as being dry for 2-hour intervals with volitional voiding
- 4/23 children also required CIC through Mitrofanoff channels to assist with bladder emptying

BNR Incidence After CPRE

1994-2010 at Boston
N=32 (22 M, 11 F)
>3 yrs out from CPRE

Male

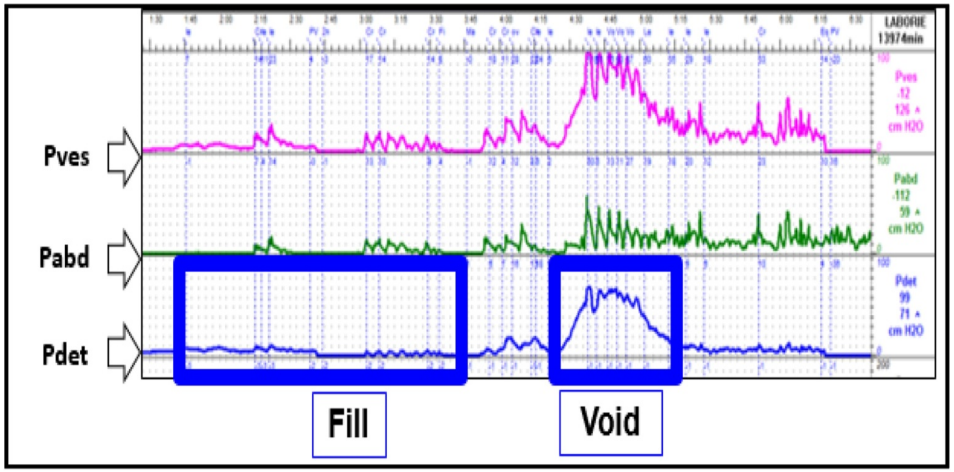
- 9/22 M BNR
- 9/22 M Potential BNR
- BNR needed (41-82%)

Female

- 3/11 F BNR
- 3/11 F Potential BNR
- BNR Needed (27-55%)

Exstrophy Bladder Function

- UDS performed on 13 consecutive pts with Bladder Exstrophy closed by CPRE
- EMG activity was normal
- Sacral reflexes were intact
- EUS silenced during detrusor contraction voiding



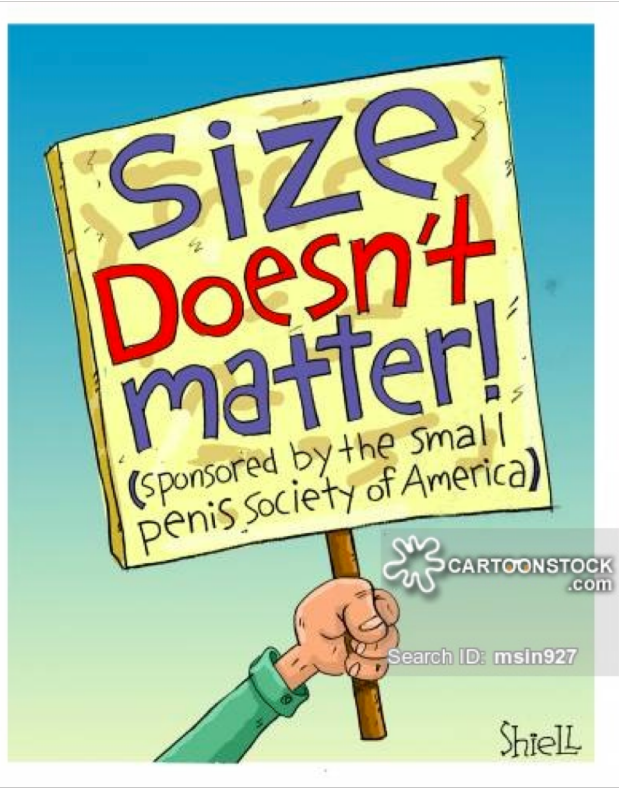
Borer et al., J Urol 2014

Bladder Potential

- Combining (Boston + Seattle) BNR for pts w/ CPRE
- 20% Males will not need BNR
- 45% Females will not need BNR

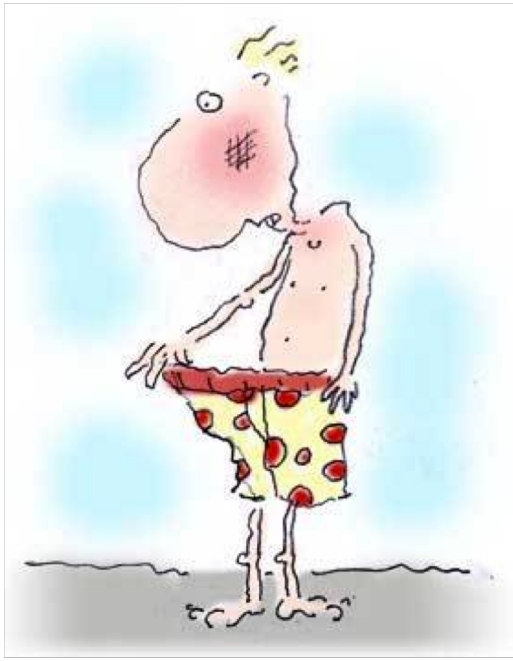
Sexual Function
Adolescent Anxiety (Gearhart)

- Male concerns at puberty:
  - Additional penile length
  - Residual dorsal chordee
  - Scars from prior surgery
  - Fertility and Sexual function
- Female concerns at puberty:
  - Sexual function, fertility, hair distribution
  - Appearance of genitalia, vaginal outlet
  - Vagina-short and wide like penis in males, cervix inserts into anterior vaginal wall not dome of vagina-prone to prolapse!

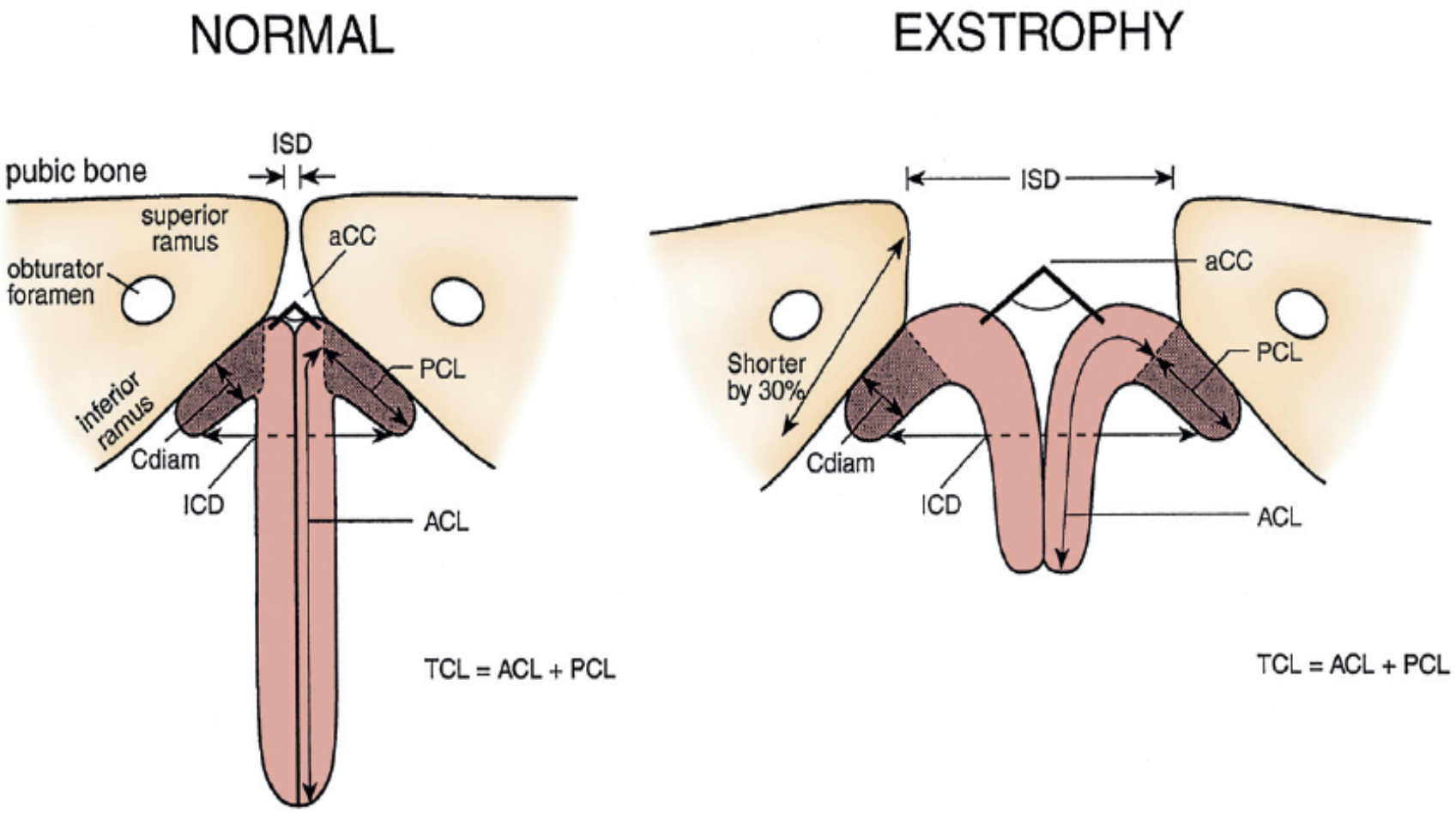


Exstrophy Long-term: Penile Concerns

- With CPRE Versus MSRE
  - Does Cantwell Ransley affect outcome of straight penis?
  - Does Salle flap allows pendulous appearance
  - Most happy with erections and sex
  - All!!! --Want more length
- Additional length and correction of dorsal chordee; Silver, Gearhart (J Urol 1998)
  - Dermal grafts (allografts of dermis, Alloderm)



Corporal Differences in Exstrophy



Exstrophy Long-term Concerns: Fertility

- Stein et al. (J Urol 1994)
  - 23/28 ejaculate, 5 none
  - Sperm counts low to none, low volumes, none fertile
- Ben-Chaim et al. (J Urol 1996)
  - Sperm counts low, most with azospermia, none fertile
  - Females all fertile
- Hopkins Exstrophy Database: 1280 patients-26 IVF's no birth defects so far

Exstrophy Long-term Concerns: Sexual Function

- Ben-Chaim et al. (J Urol 1996)
  - All good erections for sex; 6 felt penis too small
  - 78% satisfied with orgasms
  - Females: all normal libido, all normal menses, all satisfied with orgasms
- Mathews et al. (BJU Int 2002): 83 females
  - All became pregnant who wanted to
  - Normal libido, sexual activity began 20.2 yrs
  - 85% normal orgasms, 15% unsure, 6 dyspareunia

Exstrophy Long-term Results: Social Concerns

- Feitz et al. (J Urol 1994): All ileal loop
  - All positive attitude toward life
  - Males concerned about penile length, all good erections
  - Females: all orgasms, all had children that wanted to become pregnant
- Social follow-up; Ben-Chaim (J Urol 1996)
  - 50% of males & all females describe relationships as serious and long-lasting
  - 15/20 higher degrees
- Schaeffer et al. (J Urol, 2012): 49 Bladder exstrophy or epispadias completed CHQ-CF87
  - Mean age of 14.3 years
  - Good scores relative to reference populations from two previous studies with questionnaire

Long-term Psychiatric Follow-up

- Montagnino et al. (J Urol 1988)
  - Sophisticated psy instruments
  - Some difficulty adjusting in school
  - No major depression or anxiety
  - Continent by age 5 and 6 did better
- Reiner et al. (J Child Psy 2008)
  - 60 exstrophy patients-teens and above
  - Overall very normal teens
  - Not many psychiatric problems
    - Mainly self image
    - Need early intervention
- New data: Hankinson et al. (J Ped Urol 2013)
  - EEC children have greater likelihood of experiencing emotional and behavioral problems
  - Older children have worse, internalizing symptoms and adaptive functioning

Need more data

- Bladder exstrophy is a rare disease
- Surgeries have changed over the years making it difficult to predict true outcomes along with surgical variations per institution and per surgeon
- No standardized approach and follow-up

Conclusion

- Bladders in patients with bladder exstrophy have the potential to void normally
  - Storage of urine
  - Continence mechanism that's controllable
- Patients with bladder exstrophy that are doing well i.e. continent and voiding volitionally are still at risk for possible bladder deterioration as they become older AND need to be followed closely

THANKS

Adolescent Anxiety

Table 1. *Number of surgeries and bladder augment*

	Number of surgeries		Bladder augment	
	Less than 5	More than 5	Yes	No
Female	9 (64.3%)	5 (35.7%)	3 (21.42%)	11 (78.58%)
Male	13 (46.4%)	15 (53.6%)	9 (31.03%)	20 (68.97%)

Table 2. *Satisfactory cosmesis and Incontinence in the patients treated*

	Satisfactory cosmesis		Incontinence	
	Yes	No	Yes	No
Female	10 (71.42%)	4 (28.58%)	7 (53.84%)	6 (46.16%)
Male	15 (53.57%)	13 (46.42%)	13 (44.82%)	16 (55.18%)

Table 3. *Male sexual profile*

	Satisfactory penile length		Ejaculation	
	Yes	No	Yes	No
Sexually active (n=20)	3 (15.00%)	17 (85.00%)	19 (95.00%)	1 (5.00%)
Not sexually active (n=9)	0	9 (100.00%)	3 (33.33%)	6 (66.66%)

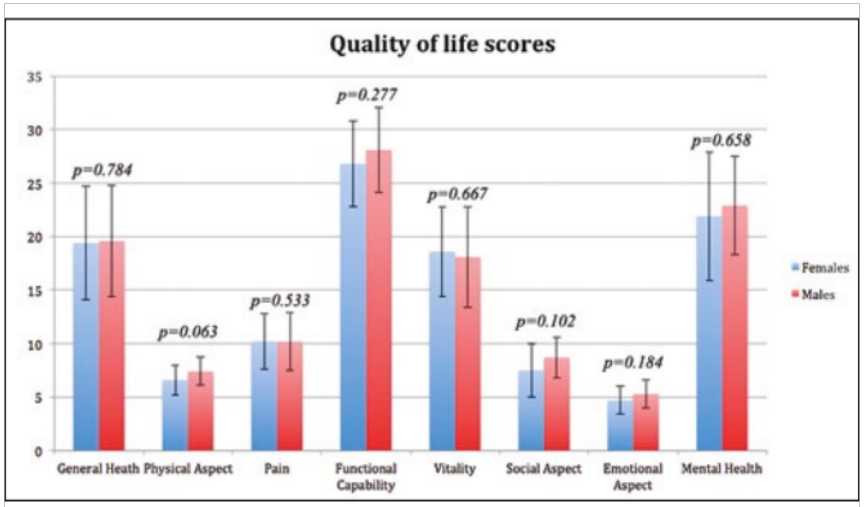


Figure 1. *Values of the domains assessed by SF-36 questionnaire.*

Da Cruz, Mattos, et al, 2016