



Bladder Exstrophy in the School Age Child

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The Goal and the Challenge

- Bladder and abdominal wall closure
- Preserve upper tracts
- Achieve urinary continence
- Functional and normal appearing genitalia
- Sexual function/fertility





Exstrophy Challenge

I challenge anyone to show me an adult male with bladder exstrophy who:

- Voids normally and is dry day and night
- Ejaculates
- Has normal sexual function

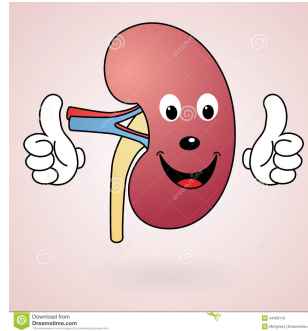
Most recently in Curitiba, BZ 2005

The Continuum

- Infancy
 - Achieve a secure closure of the bladder, abdominal wall and epispadias repair
- Toddler and school age
 - Attempts at toilet training, frustration.
 - Concerns begin about genital appearance
- Teenage/Adult
 - Genital appearance
 - Sexual intercourse and impotence/infertility

Focus on the Three's

- 1. Upper tracts
 - New hydro?
 - GFR stable?
 - Scar?
- 2. Infection
 - Episodes of pyelo?
 - UTI's?
- 3. Contenance
 - Volitionally voiding?
 - CIC?
 - Constantly wet?
 - Dry when supine?



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

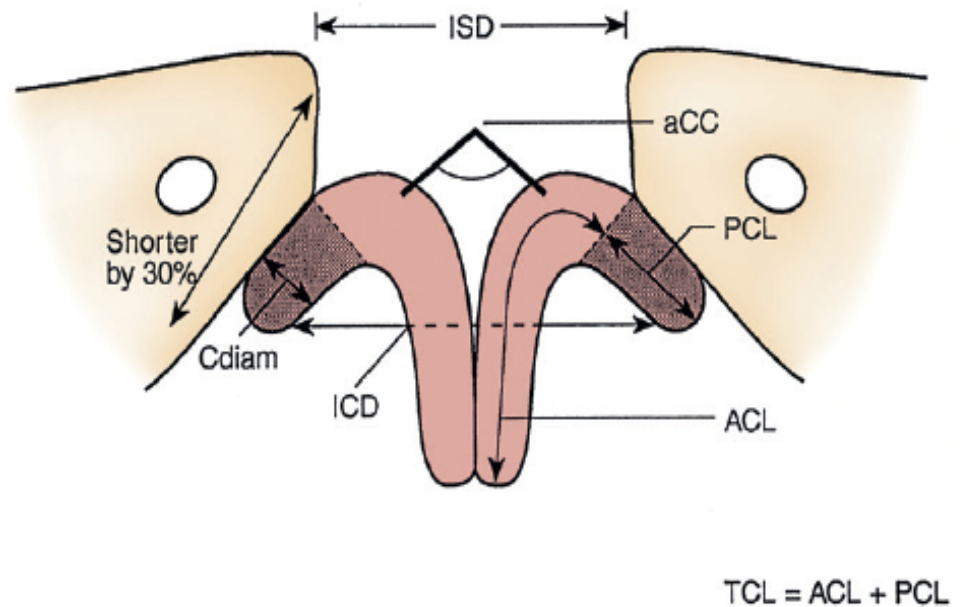
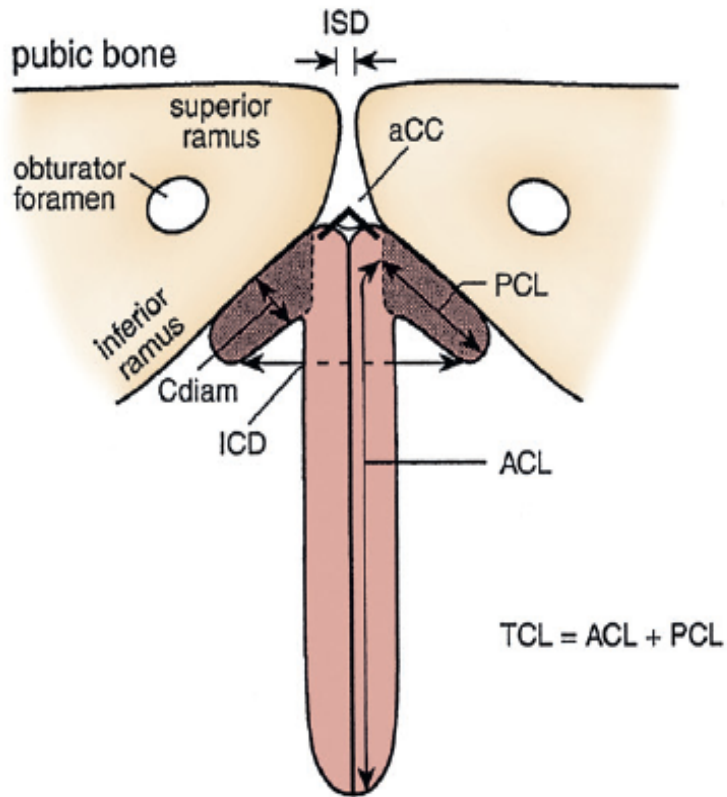
- With CPRE Versus MSRE
 - Does Cantwell Ransley affect outcome of straight penis?
 - Does Salle flap allow pendulous appearance?
 - Most happy with erections and sex
 - All!!! Want more length
- Additional length and correction of dorsal chordee (Silver, Gearhart, J Urol 1998)
 - Dermal grafts (allografts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



Corporal Differences in Exstrophy

NORMAL

EXSTROPHY



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%)

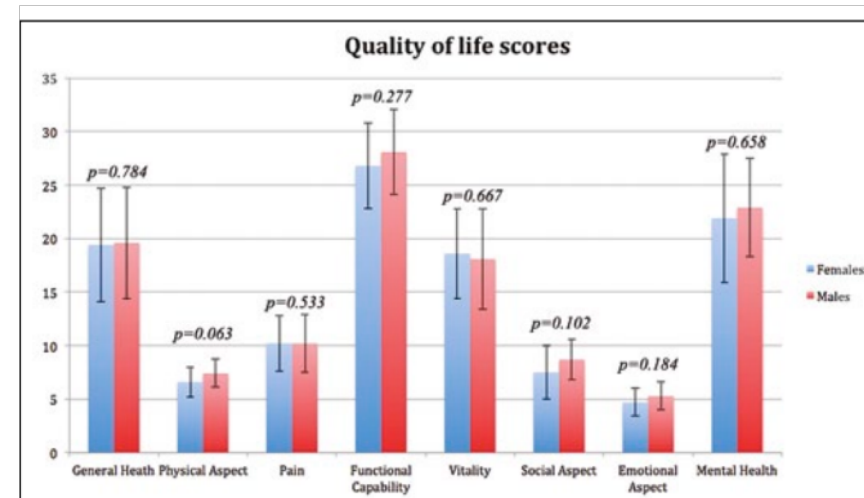


Figure 1. Values of the domains assessed by SF-36 questionnaire.

Da Cruz, Mattos, et al, 2016

Exstrophy: Long-term Fertility Concerns

- 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 Sexual Function Concerns

- Ben-Chaim et al. (J Urol 1996)
 - Males: 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 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 had 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 or 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