

The Societies for Pediatric Urology



PEDIATRIC UROLOGY FALL CONGRESS

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Fertility & Spina Bifida

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John S. Wiener, M.D.

Professor of Urologic Surgery & Pediatrics



Disclosures



- **Financial**

- Grants

- **Centers for Disease Control & Prevention**
 - **Sexual Medicine Society of North America**



GUIDELINES FOR THE CARE OF PEOPLE WITH **SPINA BIFIDA**



Men's Health

Workgroup Members: Hadley Wood, MD, FACS (Chair); Dominic Frimberger, MD;
John S. Wiener, MD

Introduction

Until recently, adult sexual function in men and women with Spina Bifida had not been widely

- **Men's Health**
- **Women's Health**
- **Sexual Health & Education**

1. Evaluate and characterize penile and genital sensation.
2. Evaluate and characterize erectile function.
3. Evaluate and characterize orgasmic and ejaculatory function.
4. Maximize fertility potential of men with Spina Bifida, if desired.
5. Ensure sexual education and safe practices (Sexual Health and Education Guidelines).
6. Determine the sexual activity and interest in men with Spina Bifida.

Tertiary

1. Describe known therapies for decreased genital sensation, erectile/orgasmic/ejaculatory dysfunction, and infertility.
2. Assess the impact of fertility and sexual function on the quality of life in men with

SB = Success Story



- **Prior to 1960:**
 - **Survival rate in infancy = 10-12%**
- **1989-94:**
 - **Survival rate in first year = 91%**

Pruitt LJ, *Pediatrics*, 2012

Wong L & Paulozza L, *Paediatr. Perinat Epidemiol.*, 2001

SB = Success Story



- **CDC estimates:**
 - **166,000 Americans living with SB**
 - **Majority are adults**
- **Therefore, sexual health & fertility in adults living w/ SB are garnering more attention**

Female Fertility



Female Fertility



- **Not usually affected by spina bifida**
 - Cardenas (Seattle) 2008 - ~ 20% married/been pregnant
- **Orthopedic issues could affect positioning**
- **Neuropathy can affect:**
 - Clitoral/vaginal/genital sensation
 - Orgasmic function
 - Uterine support by pelvic floor

Female Fertility



- **Biggest problem may be knowledge gap**

- **25 women with SB interviewed**

- **22 (88%) had desire for pregnancy**
 - **6 (24%) had at least one pregnancy**

"He told me it would be extremely selfish of me to even consider [having kids]": The importance of reproductive health to women with spina-bifida and the lack of support from their providers

- **5/6 (83%) pregnancy unplanned "as they did not think they could get pregnant"**

Courtney S. Streun^{a,*}, Christine L. Schafer^a, Valerie P. Garcia^a, Elisabeth H. Quint^b, David E. Sandberg^c, Claire Z. Karpakjian^a, Daniela A. Wittmann^a

- **1 IVF**

- **Delivery – 4 vaginal; 2 C/S**

June 2019

Pregnancy & SB



Urologic Congenitalism

Complications and Outcomes of Pregnancy and Cesarean Delivery in Women With Neuropathic Bladder and Lower Urinary Tract Reconstruction



Joshua D. Roth, Jessica T. Casey, Benjamin M. Whittam, Konrad M. Szymanski, Martin Kaefer, Richard C. Rink, Frank P. Schubert, Mark P. Cain, and Rosalia Misseri

Pregnancy & S



Pregnancy & SB



- **Pregnancy outcomes in Indiana**
 - 18 pregnancies in 11 women
 - 15 live newborns via C/S – 53% term
 - 13/15 – new/worsening hydronephrosis
 - 6 had nephrostomy
 - 8/15 had difficulty catheterizing
 - 66% of those per urethra
 - 44% of those per channel

Pregnancy & SB



- **Pregnancy outcomes in Indiana**
 - **Urologist present for all 15 C/S – 10 emergent**
 - **Complications in 40%**
 - 5 cystotomies – all emergent
 - 1 bowel deserosalization
 - 1 vaginal laceration
 - 3 developed urinary fistulae

Pregnancy & SB



- **“A scheduled C/S at the earliest reasonable time should be strongly considered with involvement by urologists and OBs.”**
- **“a high rate of intraop & postop complications”**

Pregnancy & SB



- **Unanswered question**
 - **Should women with SB deliver per vagina?**
 - Both in those w/ and w/o history of reconstruction
 - Will pelvic floor/continence be compromised?
 - What if routine delivery becomes emergent?

Female



Men's Health



- **Cryptorchidism**
- **Hypogonadism**
- **Penile Sensation**
- **Erectile Function**
- **Ejaculatory Function**
- **Fertility**

Male Sex



	Ever	ed al ct	Desired Children
Lassman, 2007	2		70%
Verhoef, 2005		%	
Sandler, 1996	2	%	
Game, 2006			
Cardenas, 2008	48		
Szymanski, SPU 2017	7		
D	*91%	l intercourse	



Cryptorchidism in SB



- **Kropp & Voeller (Toledo) 1981 – 23%**
- **Hutson (Melbourne) 1988**
 - 15% boys - 6-19 years
 - International Myelodysplasia Project (unpublished) -**23%**
- **Ferrera (Rome) 1998 – 15%**

Cryptorchidism in SB



- **Patel (Philly) 2008 – biopsy data in 6**
 - 2 – no germ cells
 - 3 - severely reduced # germ cells & delayed maturation

Hypogonadism in SB



- **Sandler, Worley (UNC/Duke) 1996**
 - 10/15 small testes (<12 cc)
 - 7 were soft
- **Hultling (Stockholm) 2000**
 - 2 – 15 cc
 - 1 – 12 cc
 - 6 – ≤ 8 cc

Hypogonadism in SB



- **Decter (Hershey PA) 1996**
 - Testosterone level – normal in 40/44
 - Low T level – $\frac{3}{4}$ no ED; $\frac{1}{4}$ fathered child

Hypogonadism in SB



- **Reilly** (Erie PA) – **AUA 1992** – nine men
 - Small soft testes
 - Small SV in 5 with TRUS
 - Low T in 4/9; Elevated FSH in 5/9
 - EEJ in 1 – azoospermia
 - Quoted as all had azoospermia and testis biopsies with Sertoli only

Hypogonadism in SB



- **Hulting (Stockholm) 2000**
 - 2/9 provided semen & 7 EEJ
 - Motile spermatozoa noted in only 5/9
 - Most abnormal morphology
 - Best had only 10K motile sperm
 - Testis biopsy in 7/9
 - Normal spermatogenesis in 3/9 but low #
 - Reduced spermatogenesis in 1
 - Sertoli cell only in 3/9

Hypogonadism in SB



- **What can we learn from SCI world?**

- Celigoj FA, Ibrahim E,... Brackett NL. Semen quality in men who sustained a SCI during prepubertal period. J. Urol. 188:521, 2012
 - Miami Project to Cure Paralysis – Male Fertility
 - 533 men – 7 injured prior to age 12y
 - All (3) injured before age 10y – azoospermia
 - Two injured at 10 and 11.6y – oligospermia
 - Two injured at 11.9 y – normospermia



– NORMAL NEURAL INPUT AT EARLY AGE MAY BE REQUIRED FOR NORMAL SPERMATOGENESIS

Penile Sensation in SB



- **Penile sensation – S2-4**
 - **SB lesion level-dependent**
- **Diminished penile sensation assoc w/ ED**

Penile Sensation in SB



- **Sandler/Worley (NC) 1996 – 20% normal**
- **Gatti (Parma) 2009 –**
 - 7% normal in L2 SB and above
 - 53% normal in L3-5 and sacral SB
- **Verhoef (Utrecht) 2005 – 27-32% “less genital sensitivity than desired”**

Erectile Function in SB



- **Most report inability to maintain erections**
 - **Likely related to decreased sensation**
- **Shiomi (Nara, Japan) 2006 – 26 men**
 - 85% had psychogenic erections by AV stimulation
 - 54% reported rigidity with tactile stimulation
- **Roth 2017 SPU – online survey - 122 men**
 - 41% - “sufficient firmness for intercourse”

Ejaculatory Dysfunction



- **Semen emission may be altered**
 - 73-88% report ejaculation
 - Higher than normal erections or orgasms
 - Most report dripping and not w/ orgasm
 - Szymanski 2017 – only 17% forceful
- **Orgasmic dysfunction**
 - Limited data – 20-66% report orgasms

Fertility in Men with SB



Paternity in SB



- **Laurence (Wales) 1975**
 - 9/11 married men – 23 offspring
 - None with MMC

Paternity in SB



- **Cardenas (Seattle) 2008**
 - 4/27 (15%) w/o hydrocephalus; 0 w/ HC (mean: 25y)
- **Decter (Hershey) 1997**
 - 7/10 with L5/sacral SB – all amb w/o HC
 - 1/39 w/ higher lesion attempted – success
- **Hultling (Stockholm) 2000**
 - 2/46 men in clinic had fathered children

Fertility in SB



- **SUMMARY**

- Lesion level (and VP shunt) matter
- Spermatogenesis is impaired in many
 - Altered testicular innervation?
 - Altered ejaculation?
 - Congenital testicular maldevelopment?
- Future – Role of EEJ & ART?

Final Thought



- **Discuss increased risk of SB in offspring**
 - One estimate 1:23 – no difference M vs. F
 - Prior to folate supplementation
 - CDC recommends **folate** 4000 µg daily for 1-3 months prior to conception & through 1st trimester

