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

• Men’s Health

Women’s Health

• Women’s Health

Sexual Health & Education

• 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%

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
    – 5/6 (83%) pregnancy unplanned “as they did not think they could get pregnant”
    – 1 IVF
  – Delivery – 4 vaginal; 2 C/S

"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

Courtney S. Steuer, a, * Christine L. Schafer a, Valerie P. Garcia a, Elisabeth H. Quint b, David E. Sandberg c, Claire Z. Kalpakjian c, Daniela A. Wittmann a

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

April 2018
Pregnancy & S

Duke Children'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
<table>
<thead>
<tr>
<th>Study</th>
<th>Year</th>
<th>Ever Sexually Active</th>
<th>Ever Sexually Active in Past Yr.</th>
<th>Ever Sexually Active in Past Mo.</th>
<th>Desired Sexual Contact</th>
<th>Desired Children</th>
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<td>Lassman, 2007</td>
<td></td>
<td>24%</td>
<td></td>
<td></td>
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<td>70%</td>
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<td>Verhoef, 2005</td>
<td></td>
<td>22%</td>
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<tr>
<td>Sandler, 1996</td>
<td></td>
<td>27%</td>
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<td>Game, 2006</td>
<td></td>
<td>35%</td>
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<tr>
<td>Cardenas, 2008</td>
<td></td>
<td>48/68%</td>
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<tr>
<td>Szymanski, SPU 2017</td>
<td></td>
<td>*91%</td>
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Additional statistics:
- Masturbation: 62%
- Vaginal intercourse: 70%
- Anal intercourse: 35%
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 – ¾ no ED; ¼ 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?

  – 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