Sexuality in the Handicapped Patient

...or talking to parents about talking to their kids

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Disclosure

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Objectives

• Review the anatomically-based sexual function issues in this population

• Discuss methods to introduce the discussion of sexuality issues, based on developmental stage

• Identify treatment options appropriate to this population, including future fertility issues
Physiology of Normal Erection

- Initiated by parasympathetic nervous system overriding normal SNS tone
- Relaxation of trabecular smooth muscle in corpora cavernosa and vasculature
- Allows filling of sinusoids and expansion of the penis
- Combination of these two processes traps blood within the penis by preventing venous outflow, resulting in rigid erection
Cascade of Events Resulting in Penile Erection

Sexual stimulation
  Increased parasympathetic activity
Decreased peripheral resistance
  Increased blood flow in cavernous and helicine arteries
  Increased intracavernous pressure
  Relaxation of trabecular smooth muscle
Lacunar engorgement and penile erection

Ejaculation

- Ejaculatory process is under both central and peripheral nervous system control
  - Genital sensory input is coordinated with erotic imagery from cerebral cortex
  - 3 distinct physiological phases of the ejaculatory process: bladder neck contraction, emission, ejaculation
Spectrum of Ejaculatory Dysfunction

- Anejaculation
- Normal ejaculatory control
- Delayed ejaculation
- Retrograde ejaculation
- Premature ejaculation

Female Sexual Response Cycle

- Emotional and physical satisfaction
- Sexual arousal and responsive desire
- Sexual arousal
- Neutral but motivated
- Seeks/is receptive to
- Sexual stimuli
- Biological
- Processed
- Psychological

Basson R. J Sex Marital Ther. 2001;27:33-43. Figure 1.
Arousal and Vasocongestion: Vulval Structures

Women’s Sexual Response

- Women’s sexuality is different from men’s
- Definitions based on a male model of sex response
- Different models of sexual response have been described and are in clinical use
- New definitions in keeping with these models have been proposed and are being tested in clinical and research settings
Women’s Sexual Response

- Women have many reasons (motives) for having sex—sexual desire is an infrequent motive in their longer-term relationships.

- Women may typically sense desire once a sexual experience is underway.

- Psychological and biological factors influence women’s arousability.

- Women’s sexuality is highly contextual.

- Arousal is not primarily about the degree of vulval and vaginal vasocongestion.
Sex Therapy

- Identify desire issues
- Dissatisfaction
- Scheduling issues (work, children)
- Fear
- Tension about sex
- Fear of too much closeness
- Depression
- Drug effects

- Helps identify optimal conditions for sexual relations
- Methods for increasing arousal
- Methods to reduce anxiety
  - Relaxation
  - Yoga
  - Self-hypnosis
- Mind-power exercises
Overview of congenital GU conditions

Sexual and reproductive aspects
Undescended Testes

• Influence of undescended testes on fertility has been the subject of extensive literature but with incomplete conclusions

• Untreated unilateral undescended testis is compatible with normal fertility
  • disproportionate number of such men attending infertility clinics
Undescended Testes

• Man who has had an orchiopexy may find himself infertile 20+ years later

• By this stage the cohort of boys operated on is difficult to trace, and the incidence of infertility attributable to original maldescent cannot accurately be determined

• Up to 43% of undescended testes have abnormalities of epididymis and its testicular attachment

• Infertility may be due in part to obstruction
Fertility in females with exstrophy

• Problem in females is entirely anatomical

• Introitus narrow and almost always requires surgical enlargement
  • Thereafter, girls appear to have normal fertility
  • At least a 50% incidence of uterine prolapse
  • Few women with exstrophy are able to have more than 2 children before repair of the prolapse
Exstrophy in males

- Sexual and reproductive consequences of exstrophy in males well documented

- Erectile deformities may prevent vaginal penetration
  - once correction has been made, intercourse should be possible
  - shortness of the penis may prevent the delivery of semen to a point close enough to the cervix for insemination

- Many boys with exstrophy do not ejaculate because of absence of circular prostate and bulbospongiosus muscle
  - Repeated operations to reconstruct bladder neck may cause damage to the vasa or seminal vesicles
  - Incidence rates of infertility in males with exstrophy unreliable
Exstrophy in males

- Males with exstrophy do not have an encircling prostate or a bulbospongiosus muscle → cannot achieve forceful ejaculation

- Most men report that some semen dribbles out with orgasm but others say that there is regular discharge of semen from the meatus unrelated to sexual activity

- Limited reports, but fertility better in boys who had had an early diversion rather than reconstruction
Spina bifida

- Congenital rural tube defects resulting from a lack of vertebral arches in the median line during the third or fourth weeks of gestation

- Incidence is 0.1 to 0.3% (4.6/10000 live births)
  - Female-to-male ratio of 2:1
  - Increased risk if one parent is affected by spina bifida
  - Risk factors multifactorial and include genetic, geographical area, maternal obesity, SES, epilepsy or diabetes, maternal exposure to drugs/alcohol/radiation
  - Life expectancy of these patients has been increased by improvements in medical and surgical management
  - 50 to 75% survived to adulthood
  - ...Resulting in typical adolescent emotional psychosocial and sexual issues
Spina bifida: Females

- Although sexual function is dependent on neurological level, females with spina bifida are thought to have normal fertility.
  - A difficult area of community policy is how to prevent unwanted pregnancies and sexual abuse in affected girls of low intelligence.

- Protection against conception of a baby with a neural tube defect is to give the mother folic acid supplements in the 3 months before conception and for first trimester.

- Common consequence of severe spina bifida is obesity.
  - Incidence of neural tube defect pregnancies is nearly double in women who are obese at the time of conception (BMI > 29).
Spina bifida: Females

- Few reports of pregnancies in females with spina bifida

- Good outcomes have been reported, especially with liberal use of cesarean section

- Several specific problems have been identified
  - Urinary tract infections are almost invariable
  - Bladder function often deteriorates
  - Small deformed pelvis makes accommodation of the fetus difficult leading to premature labor and increased need for cesarean section
Spina bifida: Males

- Men with spina bifida and intact sacral reflexes and 2/3 with absent reflexes but neurological level below L3 are likely to have normal sexual function.

- Levels up to T10 may have erections but many are not associated with sexual stimulus.

- Probable that men with low lesions have normal fertility.

- Men with higher lesions, fertility issues partially due to transport problem due to the impotence:
  - Impotent males with spina bifida were found to be azoospermic on semen analysis.
  - Sertoli cells only.
Spina bifida: Males

- Conventional management such as intracorporeal injection

- Sildenafil may be used with appropriate dose reduction
  - Other PDE5 medications

- Further study is needed in these patients to determine if current methods for treating azoospermia effective
Posterior urethral valves

- 40% of patients ejaculation is slow or absent
  - no proved reason for this phenomenon
  - marked dilatation of the prostatic urethra seems likely cause

- Ejaculation requires establishment of a bolus of semen that is forcefully expelled by the prostate
  - if the prostatic urethra is dilated, adequate pressure cannot be generated
  - Despite the open bladder neck, retrograde ejaculation rare

- Semen is grossly abnormal
  - pH is high, (range 8.4 to 9.2)
  - viscous
  - fails to liquefy
Uterine anomalies

- Didelphic, bicornuate, septate uterus → symptomless and menstruation is normal

  - Increased incidence of spontaneous abortion

  - If 1 side is larger than the other, it may be possible to establish an embryo in vitro and implant into the larger side
Klinefelter’s Disease

• Most common abnormality of sexual differentiation - 1:500 live births

• One of most common causes of primary hypogonadism

• Most common sex chromosome abnormality seen in infertile men

• Testicles are not absent, but their small size may lead to lack of recognition as testes
Klinefelter’s Disease

- **Presentation:** quoted typical triad is azoospermia, small testes, gynecomastia

- In reality, more common to observe a lack of development of secondary sexual characteristics
  - small [< 3.0cm] atropic testes
  - small phallus
  - diminished body hair
  - diminished muscle bulk
  - feminine, or truncal, rather than male, fat distribution that often includes gynecomastia
Klinefelter’s Disease

- Treatment options

- Issues due to genetic infertility and genetic hypogonadism

- Treatment with exogenous testosterone will prevent consequences seen with long-term hypogonadism

- No treatment to correct azoospermia
Medical genetics

- Development of specific tests looking for abnormalities of chromosome copy number and DNA sequence variants

- Many of these tests are now routinely available
  - often expensive → insurance issues
  - necessary analyses may take weeks if not months

- Appropriate use of these tests can be extremely useful

- They must be used in the context of a carefully structured approach to diagnosis to avoid unnecessary delay and expense in arriving at an explanation for the underlying condition
Medical genetics

• Why did it happen?
  • The need for an explanation of the cause of their child’s predicament is a strong driver
  • Underlying this need may be a feeling, often unvoiced, of guilt and of responsibility for their child’s symptoms

• What does it mean?
  • Uncertainty about what the future holds for the child and family is a source of great stress

• Will it happen again?
  • Where there is a potentially heritable cause of the phenotype exhibited, the issue of risk of recurrence
Exemplar case

Spina bifida
Spina bifida in males and females

• Research is limited on the effects of spina bifida on sexual response and function

• May be failure to develop normal sexuality
  • Impaired self-esteem
  • Dependence on caregivers
  • Lack of privacy
  • Urinary or fecal incontinence
  • Presumed asexuality, especially if wheelchair-bound

• Sex education often delayed

• Normal arousal patterns and sexual function can be seen
  • Frequency of sexual behavior dependent on level of the lesion
Spina bifida in males and females

- Patients seek a more active role from their healthcare provider, both in providing recommendations and initiating discussion

- Sex education specifically related to spina bifida problems is rarely discussed

- Incontinence in particular is an important obstacle for sexual activity

- It is important to raise these issues at an early age
  - Allows time first shift information
  - Respectful of developmental and chronological age
Spina bifida and males

• Precocious puberty may be an issue
  • Boys of spina bifida typically reach puberty in the range of 8-13 years
  • May demonstrate secondary sexual characteristics as early as nine

• Prevalence of erectile dysfunction is approximately 75%
  • Dependent on level of neurologic lesion
  • Intact sacral reflexes and urinary continence → potentially potent

• Variable ejaculation and orgasm; up to 75% experience ejaculation
  • Force may be decreased
  • Mosaic-type difficulties: neurological loss may not be accurately predicted by level of surviving motor function
  • Sensation of climax is variable in part due to absent or decreased penile sensation
Spina bifida in males

- Many young adults SB patients are sexually active
  - Health care professionals may be faced with issues regarding relationships and sexuality among young adults as spina bifida
  - Important to establish the information patients already have
- Small percentage of patients report they have never discussed sexuality issues with a provider
- Almost all spina bifida patients request further sex education
- Report to they would have discussed these issues if the provider had initiated the topic (Sawyer 1999)
Spina bifida in males

• Correlation between mobility and sexual activity exists for boys but not for girls

• Most studies focus on the relationship between sexual dysfunction and neurological loss

• Majority of male patients with lesions below L3 had more normal penile sensation, erections, ejaculation
  • In contrast only half of female patients with the lesion at or above L2 had vulvar sensation or climax
  • For both males and females, generalizations are difficult due to small number of patients in these studies
Spina bifida in males

• Sex education provided mostly by schools, and by parents

• Less than 1/5 had been informed about sexuality issues by a physician (Verhoef, 2005)

• Problems relating to spina bifida including fertility and heredity are rarely discussed

• STDs rarely discussed with male patients with spina bifida

• Many patients perceive obstacles in starting a relationship
  • Wheelchair
  • Incontinence
  • Many wish to have children
Treatment of sexual dysfunction

- Erectile dysfunction contributes to
  - Lack of confidence
  - Poor body image
  - Difficulty maintaining intimate relationships
  - Parallel with non-spina bifida males

- Penile anatomy not altered by congenital issues

- Respond well to traditional treatments for erectile dysfunction
Fertility issues in males

- Testosterone is typically normal

- Infertility is an issue
  - Higher rates of neural tube defects compared with the general population

- Paternity percentage is greater with lower level lesions

- Higher-level lesions: issues may be secondary to anejaculation and erectile dysfunction

- Also the potential for Sertoli-cell only male infertility
  - Unclear etiology
Spina bifida and females

- Less likely to use hormonal contraception or do not use any method of birth control (Cardenas, 2010)

- Fertility is preserved → young women can achieve pregnancy and delivery

- 80% of women have some genital sensation, and one third have experienced climax

- Vulval sensation and climax are rarely described in patients with lesions at or above L2

- Female spina bifida patients are generally less affected than male patients (Visconti, 2012)
Spina bifida and females

- Higher incidence of precocious puberty and premature activation of hypothalamic-pituitary-gonadal axis higher in SB girls than healthy counterparts
  - Timing of puberty may be earlier
    - 10.9-11.4 years (Trollmann 1998)
  - Unknown issues relative to rhythm and intensity of menstruation in this population

- Preconception counseling strongly recommended, due to higher risk of having an infant with a neural tube defect
Spina bifida and females

- No special protocol to follow during pregnancy care of women with spina bifida
- Increased risk for UTI, urinary stasis, glycosuria
- Promotion of vaginal delivery
- C-section delivery higher rate of surgical complication
  - Small pelvis, hip bone deformities, kyphoscoliosis
Sex education
Statistics

- In the context of sexual behaviors, adolescence encompasses ages 10 to 18

- Most recent survey of adolescent sexual behavior
  - 50% of youth in grades 9 to 12 are sexually active (CDC, 2008)
  - 7.1% initiated intercourse before age 13
  - 14.9% had intercourse with four or more partners
  - 38.5% had not used a condom during their last sexual intercourse
  - Currently there is an estimate of 9.1 million cases of sexually transmitted infections among people aged 15 to 24 (CDC, 2008)
Gap in sex education

• Parents are in a unique position to engage in dialogues about sexuality related issues
  • Before initiation of sexual activity

• Parent-child discussions can be continuous
  • Build upon previous discussions as the child ages
  • Respectful of developmental level
  • Time sensitive to immediate questions rather than predetermined, such as school curricula

• Parents can focus discussions based on their value and belief system about sexual behavior that outside sources
Gap in sex education

• Need to reach youth early with sexual risk prevention messages

• Need ways to overcome communication barriers

• Need to enhance responsiveness of parents to promote safe sex behaviors

• Parents should be encouraged to be responsive and practice having discussions about sex

• Increase effectiveness when children are young: beneficial to address before the complex and emotionally charged issues of teen years...
Responsiveness

• A critical role and key determinants in effectiveness of parent-child discussions about sex
  
  • Key role in route to seeing teen sexual health risk
  
  • Increases attentiveness in teens when mothers are responsive
  
  • Lowered level of adolescent sexual risk in the context of parental responsiveness
  
  • Protective effect of the discussion conditional on responsiveness
    • Communication with partners about condom use
Repetition

- Repetition of sexual communication is important
  - More robust predictor of teen perceptions of the quality of the relationship with their parents

- Right message at the right time

- Repeated communication increases feelings of comfort

- Fosters a connected parent-teen relationship

- Conveys parental interest

- Implications for promoting sexual health and adolescence
...what to tell parents
Discussion

• Teenagers perceive the gap between topics covered by their parents and the topics about which they are concerned (Nat’l Campaign to Prevent Teen Pregnancy 2002)

• Conversation often poor quality
  • May be defensive avoidance impersonal unsupportive and rule oriented
  • Studies have shown that parents are dominant prevent turn taking by the adolescents
  • Adolescents may demonstrate contempt, dishonesty and avoidance
Discussion

• Large proportion of adolescents were not communicating with parents about key topics before sexual debut (Beckett, 2009)

• Communication about various topics is almost always earlier with daughters than with sons

• Communication with daughters occurs earlier relative to their sexual activity, leaving parents less time to communicate preemptively with sons
Discussion

• 3 categories
  • talk before the act: topic was 1\textsuperscript{st} discussed prior to when sexual act first occurred
  • talk after the act: act 1\textsuperscript{st} occurred prior to when topic was first discussed
  • ambiguous: topic first discussed in same time period when act 1\textsuperscript{st} occurred
Discussion

- **Presexual stage**
- Topics typically addressed deal primarily with sex in relationships
  - how to choose friends
  - Homosexuality
  - why not to have sex
  - how to decide on sex
  - female physical development (e.g., pregnancy, menstruation)
  - how boys’ bodies change
  - pregnancy consequences
  - how to make healthy decisions
Discussion

- **Precoital phase**
- Parents and adolescents begin to communicate
  - STD prevention
  - birth control (eg, condoms/STDs, birth control efficacy)
  - continue talking about sex in relationships
    - how to ask for a date
    - recognizing love
  - topics related to male physical development (eg, masturbation)
  - making healthy decisions
    - not pressuring for sex
    - why people like sex
    - refusing sex
Discussion

- Genital touching is an important sexual milestone
- It is the act that precedes sexual debut
- May present a critical period during which youth may especially benefit from communication about sexuality, including conversations about how to practice abstinence or safe sex
Discussion

• **As adolescents are initiating intercourse**
  • They and their parents communicate about additional topics
    • STDs and pregnancy prevention (eg, recognizing STD symptoms, how to use condoms, choosing birth control)
    • How sex feels (eg, sex in relationships)
    • what to do if a partner refuses a condom (eg, making healthy decisions)
Discussion

• Parents can play an important role in the sexual socialization of their children by educating and talking to youth about sexuality and by reinforcing safer HIV-related and pregnancy prevention behaviors (Miller, 1998)

• Timing and content of this communication in relation to an adolescent’s sexual behavior may be critical

• Information regarding the timing of parent–child discussions about sexuality and youth sexual behavior can inform pediatricians and others as they counsel parents to talk with their children about sexuality
Discussion

• Previous studies
  • discussions with mothers that occurred before first intercourse were associated with more condom use (ie, with more protected intercourse), when compared with no discussion

• Clawson and Reese-Weber (2003) found that mother–adolescent communications before an adolescent’s first intercourse predicted older age of first intercourse, fewer lifetime partners
  • also predicted greater likelihood of a pregnancy

• On-time father–adolescent communication (before 1st intercourse) predicted older age of first intercourse
Discussion

• Talking about sex is not an all-or nothing event

• Repetition of sexual discussions—talking about topics more than once—associated with adolescents’ feeling closer to the parent and having a sense of open communication

• Content of parent–adolescent sexual discussions can cover a range of topics
  • Biological and developmental issues (eg, puberty)
  • Values
  • Healthy relationships
  • Pregnancy and STD prevention
Discussion

- Parents and adolescents discuss a range of sexual topics in relation to adolescent sexual experiences

- Strong grouping when topics were discussed according to the sexual experiences of the adolescents

  - When parents believe that their children have not yet initiated intercourse, parent–child communication focuses on parent values regarding teen sex

  - Once parents suspect that their adolescents have initiated intercourse, parents focus on more concrete matters such as
    - birth control
    - STDs
Discussion

• Parents have significant potential to reduce adolescent sexual risk behaviors and promote healthy sexual development

• Studies have found that parents communicating with adolescents about sexuality results in
  • delayed intercourse
  • contraception use
  • fewer partners

• Limited, as research in this area almost exclusively focuses on parents of teens aged 13 and older

• Can be hindered because most parents do not feel comfortable or competent talking about sexual issues

• Conversation may be limited to developmental changes and negative consequences
Closeness of parent-teen relationship

- Implications for adolescent sexual health

- Positive connection with their parents
  - Less likely to initiate intercourse at a young age
  - Less likely to engage in frequent intercourse
  - More likely to use contraception
  - Fosters high quality and repeated parent-teen sexual communication
  - Reduces teen sexual risk-taking if teens feel their parents are open, skilled, and comfortable with discussions of sex-related topics (Whitaker, 1999)
Discussion

• American Academy of Pediatrics recommends that pediatricians and other clinicians encourage parents to educate their adolescents about sexuality beginning early in life

• Many adolescents report little or no communication about sexuality with their parents

• Several findings suggest that the higher the quality of the parent-teen relationship is a particularly important protective factor for girls
  • Especially true of paternal influence
Role of the health care professional
Influences on adolescent sexual behavior outcomes

• Academic ability or achievement

• Social and behavioral competence

• Emotional competence

• Subjective norms regarding sex

• Perceived norms of others

• Religiosity
Counseling and Minors

- Ethical/legal requirements
  - Must be in best interest of patient
  - If able to understand, minors should be presented with choices
  - Sexual maturity of patient

- Encourage information specific to the needs of adolescent females and adolescent males individually
  - Avoid assuming their needs are identical
Counseling

- Define risk to fertility, for STDs, for pregnancy, when possible

- Discuss with parents early in treatment planning, to allow for most potential options and time for discussion
  - Spina bifida Association of America (sbaa.org)

- If pre-pubertal: Vital to consider maturity of patient, as some wish to discuss without parents present

- Females: issue with post-pubertal minors

- Support of the family may be important also
Counseling: Barriers

- Lack of time
- Lack of motivation
- Lack of confidence/hope
  - Extent of disease
  - Limited knowledge of emotional impact of condition
  - Limited knowledge of impact on attitudes about having children
- Perceived issues relative to ethnicity/cultural domains
- Modesty of patient
- Modesty of provider
- Lack of learning opportunities or opportunities for questions
- Emotional issues
  - Diagnosis
  - Prognosis
  - Treatment course
  - Fear regarding unknown cost of any eventual therapies
Tips for counseling

• Because of the potential for cognitive difficulties in these populations, information should be relayed in a manner that is sensitive to this
  • Hydrocephalus especially may be an issue

• Information should be broken down into units and encourage the use of visual aids

• Information should be written down

• Individual should be asked questions to ensure comprehension

• Provide opportunities to maximize ability to learn and process this information

• These strategies are appropriate for parents and healthcare providers alike
Counseling

- Issues of sexuality in teenagers and young adults with spina bifida or other congenital GU issues is often overlooked
  - Or avoided

- In particular, issues such as self-esteem and self-image are important to address

- Resources for transition planning
  - New York State Institute for health transition training for youth with developmental disabilities (healthytransitionsny.org)
  - Adolescent health transition project (depts.Washington.edu/healthtr)
  - Health Care Transition Initiative (hctransitions.ichp.ufl.edu)
Conclusions

- Discussion of specific sexual problems can generate the ability to take greater responsibility for their sexuality
  - Decreased risk of sexual abuse
  - Decreased risk of STDs
  - Decrease unwanted pregnancy

- Assessing sexual risk behavior involves looking at the spectrum of possible sexual behavior
Conclusions

- Sex education should discuss major topics
  - Self-esteem
  - Body image
  - Public versus private body parts
  - Changes of puberty – Menstruation, nocturnal emission
  - Physical mechanics of sex
  - STDs
  - Appropriate and inappropriate expressions of sexuality
  - Safe sex
  - Birth control

- Issues with transition of care for this population from pediatric management to adult
  - Education on sex and sexuality should be part of the supportive care of all young adults
Final Thoughts

Questions?

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