SEXUALITY ISSUES for the Person with Spina Bifida

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Forty years ago, we learned an important lesson in the development of a child with the Myelomeningocele form of Spina Bifida: 6 to 9 year old patients with myelomeningocele did not know whether they were boys or girls. A subsequent study demonstrated that parents were embarrassed to ask whether their infant or young child could become a competent reproductive human being! This lack of information is in stark contrast to what is actually known. Adult patients with myelomeningocele are able to reproduce. A patient's potential sexuality can be discussed in a straightforward manner. For parents of children initially treated elsewhere, their understanding of a particular concept should be clarified prior to any further discussion. If their understanding is correct, the issue will not require further discussion unless their knowledge is out of date. Therefore, we discuss with new families, regardless of age, the parents' knowledge of their fetus' or child's sexuality and reproductive potential.

Females with the various forms of Spina Bifida and normal reproductive organs will usually experience "orgasm" and are usually fertile and are capable of bearing children. Exceptions to this general statement are children with Exstrophy of the Cloaca, one of the rare forms of Spina Bifida Occulta, and the rare patient with additional malformations of the reproductive organs.

Males have a much more complex genitourinary system that depends on an intact autonomic nervous system. If a male with Spina Bifida has an anesthetic penis and desires such an important sensation, a branch of the ileo-femoral nerve to the dorsal penile nerve can restore sensation. A precondition for this operation is full sensation over the anterior and medial aspect of the upper thigh. That may also be insensitive in males with a high level of paralysis. Males with Spina Bifida can have erections that are due to urinary bladder base stimulation, erotic thoughts or noxious stimulation of the thigh, perineum, pubic area or penis. If a male with Spina Bifida does not have erotic erections that are sufficient for sexual intercourse by the time he has completed pubertal development, an artificial device can be inserted surgically to allow him to do so. Affected males can be fertile if their prostate, epididymus and at least one testicle are not malformed and have not been destroyed by infection. Ejaculation may be retrograde, forward or non-existent; ejaculation is not necessary for fertility. Some males with Spina Bifida as well as able bodied males will ooze prostatic fluid containing sperm during the sex act, successfully impregnating their partner without ejaculation. This phenomenon should be taught to ALL boys. Retrograde ejaculation can be reversed either by surgically adding an artificial sphincter proximal to the prostate or by closing the urinary bladder neck and adding a catheterizable stoma. For the 75 to 80% of males with Spina Bifida who are infertile, there is a 75 to 80% probability that needle aspiration of the testes can recover sperm or the DNA of -- precursor cells. These cells or DNA can be used to fertilize his partner's egg via intracytoplasmic sperm injection of his partner's egg outside her body and then reinserted into her uterus. The fourth and last portion of the male sex act is potency. This term is used to include the ability to satisfy a sex partner, not just satisfactorily performing sexual intercourse. There are many ways to achieve orgasm. Parents are unlikely to either understand or divulge such information, and able bodied peers cannot be relied upon! The male and female with any type of Spina Bifida need to learn from movies, television, a sexual counselor or sexually explicit literature how to perform these techniques to satisfy their sexual partners. There is available such literature specifically for the disabled.

The last aspect of reproduction discussed with both sexes is their ability to lower the recurrence of Spina Bifida in relatives, their parents and the affected child's children. A summary of the studies known to us about the recurrence rate of the most common form of Spina Bifida, (Myelomeningocele) in first-degree family members (mother's and dad's brothers and sisters of the affected child) suggest that the recurrence rate of the parents is 1 in 20, sisters 1:30 and brothers 1:170. For second-degree relatives (mother's sister's or brother's children) is approximately 1 in 220 (too few data are available to identify male and female risks independently). Others report a recurrence risk among mothers or an affected child's first degree relatives between 1 in 70 to 1 in 140. Recurrence rates in second and third degree relatives of 1 in 90 and 1 in 40 for sisters of the mother of an affected child and for the offspring of those sisters. One hundred sixty mothers who themselves had Spina Bifida reported to us only a 0.5 to 1 % chance of having a child with a neural tube defect. The percent of women in these studies with Lipomyelomeningocele, a form of Spina Bifida that has a much lower family recurrence than the Myelomeningocele form of Spina Bifida, was unknown.

The Medical Research Council of the United Kingdom first conclusively proved that when women with a prior child with a neural tube defect took 4 mg of folic acid daily beginning three months prior to conception, there was a 70% reduction in **recurrence** in subsequent offspring. Wald et al have recommended 5 mg daily. Because of the higher **recurrence** in first and second degree relatives, we recommend 4 mg daily beginning three months prior to a planned conception. We suggest the effect in the United States may be nearer to a 40-50% reduction for two reasons. First, Berry et al (1999) demonstrated a 79% reduction in **occurrence** in high incidence in north China but only 40% in south China when the women took 0.4 mg of folic acid pre-conceptually. The incidence of neural tube defects in the United States is closer to Southern than Northern China. Secondly, recent data from Canada and Mexico are the first to indicate that lower incidence communities on the North American continent can achieve a 45 – 55% reduction in occurrence with a regimen of 5 mg of folic acid per week and supplementation added to dietary fortification. Note which of these studies are these studies were of **occurrence** or **recurrence**. Also note that folic acid is not necessarily a deficiency in the mothers giving birth to a child with Spina Bifida since the preventive doses are far in excess of recommended daily doses and women with inherited or acquired folic acid deficiency do not have an increased incidence of offspring with Spina Bifida. Also note that only the Myelomeningocele form of Spina Bifida has been shown to be altered by large doses of folic acid.

A discussion of etiology, genetics and prevention is also important to help parents learn about causation and to decrease any remaining guilt associated with the diagnosis of a fetus or infant with one of these congenital anomalies. Guilt-ridden parents have greater difficulty in helping their child develop an intact personality. Frequently, the parents' imagined causes for their child's neural tube defect occurred long after the closure of the neural tube at 27 days gestation; the parents can be firmly reassured their guilt is misplaced.

Sexuality Training for and Learning by the Child without a Disability

An important, but frequently overlooked, aspect of transition and a necessary precursor for independent toilet training is the child's ability to undress and dress. These two skills are learned gradually during late infancy and early childhood. The removal of clothing also encourages autonomy. However, unless undressing occurs in the proper social circumstances, it leads to condemnation rather than approval. Parents teach their young children not to expose their perineum – frequently called their "private parts" - in public, particularly in the presence of the opposite sex.

During this developmental period, infants and children fondle their genitalia and parents tell them that this should not be done; at least, the child soon learns not to do so in the presence of others. Masturbation is a normal aspect of development. The social disapproval of masturbation should be explained in terms appropriate for the child's age but should not include threats of bodily harm. The young child of three to four years of age may also ask questions about where babies come from: their interest is usually brief and superficial. Simple explanations will both correctly inform the child and end their questioning. They observe that mother develops a large abdomen, comes home from the hospital or comes out of a home birthing room with a great reduction in her abdominal girth and a baby. Stories such as a stork delivering the baby are not appropriate.

Children age 3 to 5 years of age also notice differences between boys and girls, men and women. Some will satisfy this natural curiosity by comparing their "private parts" to other children of both sexes. Children with mental disabilities only are no different in their curiosity or methods of exploration. Children born with the various forms of Spina Bifida most recently have anesthetic genitalia that are sequestered from their exploration by excreta collecting devices such as diapers. In addition, they have their genitalia exposed to both adult males and females indiscriminately during toileting that resulting in gender identification confusion.

Sexuality Training for the Infant or Early Toddler Hood with Spina Bifida

When normal early childhood genital exploration amongst peers is postponed to later childhood, there can be severe social consequences. We have knowledge of 5 late childhood and early teenage boys with Spina Bifida convicted of and incarcerated for sexual molestation after engaging in curiosity driven comparison of their genitalia with younger children, despite my testimony. A sixth was successfully exonerated after an embarrassing social criminal investigation. Another female with Spina Bifida was reportedly accused of indecent exposure for demonstrating her clean intermittent catheterization technique to some boys and girls in their school.

Playing nude in front of a mirror is effective in teaching very young children the difference between boys and girls. Infants and young children enjoy learning about themselves from a mirror and exploring their body parts. We recommend that caretakers of the child with Spina Bifida consider directing the child's attention to the genitalia seen in the mirror, explaining and showing the difference between girls and boys.

Sexuality Training for the Preschool and Early School Years for the Child with Spina Bifida

The suggestions in the sexuality training section for infants and early toddler with Spina Bifida also apply to children in their preschool and early school years. At these ages, parents and caregivers can more easily explain sex differences and appropriate behaviors with language that the child can understand. Children at these ages are more likely to ask questions about sex and childbirth. For younger children, the explanations should be accurate but superficial. Explanations for older children should also inform the children about their future sexual potential, particularly the recurrence risk for their birth defect in their own children. The aspects of privacy and protection of their genitals should also be covered to avoid the problems mentioned above.

Sexuality for the Later Childhood and Preadolescent Child with Spina Bifida

Several independent activities should be acquired by children in the Myelomeningocele group prior to 9 years of age. The most notable of these skills are dressing, undressing (and when that is appropriate), personal toileting, hygiene for cleanliness with absence of bad odor, teeth, hair and nails; personal choices when shopping; being on time for activities outside the home; knowing their own telephone number; using a telephone directory; understanding time as measured by a clock; monitoring their spending and remembering special dates such as birthdays. Data for more intimate situations such as first date of sexual intercourse, on the other hand, are not well known.

They should also be taught to report to their like-sexed parent any touching of their genitals by others than their like-sexed caretakers. Girls with Spina Bifida are at risk for seduction or rape if they lack understanding about sexuality.

Children born with Spina Bifida, like their same age peers without disabilities, develop an interest in their ability to develop intimate relationships and procreate. Their lack of mobility and socially appropriate stool and urine hygiene potentially interferes with achieving these goals. We, therefore, recommend that teachers and medical staff bring these issues surrounding normal development and sexuality to the attention of parents of children with disabilities. When the child shows evidence of entering into puberty, we recommend discussing with them all the aspects of sexuality pertinent to any teenager. Open communication between parent and child is a psychosocial independent variable that predicts better adjustment at this age. These discussions should include the medical reasons for not participating in indiscriminate sexual intercourse to avoid pregnancy for the girl, responsibility for a child born out of wedlock for the boy and venereal diseases for both. For girls we also discuss the complications of pregnancy, the increased frequency of urinary tract problems and the possibility of developing herniated vertebral discs and need for cesarean section birth. In addition, we discuss recurrence risk of their disability in the affected child's children, prevention, prenatal diagnosis, and prognosis for various types and degrees of neural tube defects, particularly a defect seen more commonly in families with Spina Bifida, absence of the brain, Anencephaly. Recurrence risks should include the different levels of paralysis associated with Spina Bifida and the associated risk for normal or abnormal brain function (less with very minor low level Myelomeningocele and essentially normal potential for fetuses with Lipomyelomeningocele and most of the other rare forms of Spina Bifida Occulta.