Special Needs: Realizing Potential

Addressing Sexuality in Spina Bifida

There is plenty to talk about when treating children and adolescents with spina bifida without ever talking about sex. Bladder and bowel functions alone can occupy most of the clinical visit. But it’s important for pediatricians to be vigilant in addressing the sexual development of patients with spina bifida and other disabilities.

Research suggests that fewer than 20% of adolescents with spinal cord injury seek information regarding their sexual or reproductive health, and just 16% of those who are sexually active use contraception.

Spina bifida affects all three areas of sexuality: psychosexual development, puberty, and sexual function. Issues of physical development and sexuality should be discussed regularly, starting during early childhood and continuing through the adolescent years. This can facilitate the transition of children with disabilities into adulthood, and assist parents in understanding their role in providing developmentally appropriate sexuality education.

In general, children with neurodevelopmental disabilities are 20 times more likely to experience early pubertal changes. Approximately 1 in 1,000 typically developing girls will experience idiopathic precocious puberty, while nearly 20% of females with spina bifida will enter puberty before the age of 8 years. About 90% of these will have hydrocephalus and shunts. The exact cause for the increased incidence isn’t known, but it’s likely due to malformation of the central nervous system and altered function of the hypothalamus and pituitary gland.

Gonadotropin-releasing hormone agonists are effective in the treatment of precocious puberty in most girls. Early menarche can increase the complexity of self-care and hygiene in patients with spina bifida. However, many parents worry too much about it. For girls still in disposable undergarments, menstrual flow can just go in the diaper. For most women with spina bifida, and disabilities in general, menstrual periods are no more of a problem than for typical women, and wearing diapers makes it easier to manage. I typically recommend a first pelvic examination at 18 years of age or earlier if the female patient is sexually active. When examinations are indicated, positioning may need to be modified to accommodate the needs of young women with orthopedic deformities.

Boys with spina bifida tend not to experience precocious puberty, but rather impairments to their sexual function due to spinal cord involvement. About 70% of males with spina bifida report erections and only 40% report ejaculation. In addition, spermatogenesis may be impaired and reproductive function may be compromised by lifelong urinary reflux into the reproductive tract. Infertility is common.

I often begin a discussion of sexuality and sexual behavior with individuals with spina bifida during the early teen years. I leave the door open for the adolescent to ask questions and avoid a prepared “sex talk.” I usually defer any evaluation of sexual function until they are 18 years of age, when they can legally make their own decisions and are mature enough to understand and use the information.

For males having difficulty with sexual intercourse or for those contemplating having children, a consult with a urologist often is necessary. Treatment options for erectile dysfunction, such as medications or implantable penile devices, are available. While fertility is generally preserved in females, genital sensation can be diminished or absent. Counseling can address alternative ways to achieve sexual satisfaction. Preconception counseling also is advisable to inform women with spina bifida of their higher risk of bearing children with neural tube defects, the protective effects of folate supplementation, and potential complications associated with pregnancy.

Both genders should be advised of the need for latex-free condoms to protect against sexually transmitted diseases and unplanned pregnancies.

People with spina bifida for the most part assimilate with the general population, but generally participate in fewer social activities and intimate relationships compared with typically developing peers. One survey revealed that of men with spina bifida older than 18 years of age, only 35% reported having attempted intercourse. Physical limitations also may impact psychosocial development by lengthening dependence on parents and caregivers for basic self-care activities. Dependence on others for intimate care also can make children and adolescents with disabilities more vulnerable to sexual abuse or victimization.

Monitor for early indications of abuse, and reduce the risk of abuse or exploitation by engaging in frequent discussions of sexuality. Unfortunately, I can’t remember a single patient with a disability who initiated a conversation about sex or sexual function. Boys and girls with spina bifida are often anxious or prudish, or don’t want to discuss issues related to sexuality in front of their parents. Conversely, most pediatricians acknowledge the need to have some private time with patients, but find it difficult to fit in time-consuming discussions.

Starting in the teen years, patients should have some time alone with their providers to address sensitive issues. Above all, patients with spina bifida and other disabilities need to know we are available, and that we can play an important role in their sexual development and ability to enjoy intimacy and sexuality.

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Botulinum Toxin Aids Drooling in Neurologic Disorders

By Jane Salodof MacNeil

Phoenix — Drooling by adults and children with neurologic disorders is an underrecognized problem that can be treated with botulinum toxin types A or B, Dr. Phillip C. Song said at a clinical dermatology conference sponsored by Medicis.

Especially for otherwise well-functioning patients with early Parkinson’s disease, drooling can be socially disabling and adversely affect quality of life, reported Dr. Song, an otorhinolaryngologist at the Massachusetts Eye and Ear Infirmary and Harvard Medical School, both in Boston.

About half to three-quarters of Parkinson’s patients complain about drooling, according to Dr. Song. Cerebral palsy, stroke, mental retardation, facial palsy, and amyotrophic lateral sclerosis (ALS) also put patients at risk, he said.

Yet neurologic patients often are not referred for treatment of drooling, according to Dr. Song.

“If you are a neurologist evaluating a Parkinson’s patient, you see it as a symptom, but not something to control,” he said, adding, “The problem is awareness.”

When patients are referred, he said, insurance companies may not cover botulinum toxin treatment despite clinical studies showing that injections into the parotid and/or submandibular glands can help to reduce saliva flow.

The treatment is not experimental, he emphasized in an interview after his talk. He said he has used botulinum toxin therapy for drooling in 15 patients, noting that it is available at other centers in the United States.

The primary problem for most patients is not hypersalivation, but insufficient elimination of saliva, according to Dr. Song.

“Treatment is attempted, the first step is usually to send the patient for speech and swallowing therapy, he said.

Other options include the medications glycopyrrolate and scopolamine, and radiation if the patient is terminally ill or elderly.

Surgical procedures—the rerouting or ligating of ducts, the excision of glands, or transtympanic neuroectomy—may also be tried. Comparing these alternatives with botulinum toxin injection, Dr. Song observed, “There are much more invasive and dangerous things we can do to people to control this problem.”

He said he uses 5-150 units of botulinum toxin type A (Botox) or 250-1,000 units of botulinum toxin type B (Myobloc) for drooling.

Typically, he starts by injecting the parotid glands, but will consider submandibular injection if parotid therapy fails. Treatments typically last 1-7 months, but he has had some patients come back more than a year later.

“Variability in patients is quite high,” he said.