Integrating the Spina Bifida Patient Into the General Dental Practice

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Neural tube defects, including spina bifida, affect one out of every 1,000 newborns each year. Due to advances in medical technology, the life expectancy of these patients is rising annually, and the dental community should be prepared to treat them. This article discusses how, with proper precautions and a little effort, patients with spina bifida can be treated in the general dental practice.

Introduction

The term neural tube defect (NTD) refers to any one of a group of malformations involving defects of the skull and/or spinal cord that occur during embryonic development. Neural tube defects affect one out of every 1,000 American newborns each year. Spina bifida (SB) is a type of NTD in which the neural tube and posterior vertebrae do not completely close during the first month in utero. Prior to the advent of antibiotics and the advances in surgery in the past several decades, the life expectancy for individuals with SB was brief, but currently, 85% to 90% of children born with the most severe form of SB survive into adulthood. In the US alone, there are an estimated 40,000 children and adults with SB. This article is intended to educate the dental health professional to the special needs of the SB patient, in order to integrate this population into the general dental practice.

Types of Spina Bifida

Among SB patients, 1% of the vertebral defects occur in the cervical region, 1% in the thoracic region, and 98% between the lumbar and sacral vertebrae. There are also differences in the severity of the condition (Figure 1). Spina bifida occulta is a bony defect in which one or more of the vertebrae fails to fully develop, but the skin is left intact. Detection of this type of defect requires radiographic examination.

Meningoceles are a more severe type of defect, where the meninges and cerebral spinal fluid protrude into the opening of the spinal canal, forming an external sac filled with cerebrospinal fluid. The sac is covered with skin, and nerve tissue is not involved.

The most severe form of SB, myelomeningocele (MSB), is the one to which people most commonly refer. This defect occurs when both the meninges and the spinal cord protrude into an external sac (Figures 1 and 2). The sac contains portions of the spinal cord, cord membranes, and spinal fluid without benefit of skin coverage. Neuromuscular function is affected below the level of the lesion, which is indicated by a decrease or absence of sensation (eg, pain, pressure, friction, temperature) in the lower body.

The cause of SB is unknown, although environmental factors (eg, infections, drugs, repeated x-ray exposure) and genetics have been implicated. Folic acid (0.4 mg per day) and multivitamins taken by the mother before and during pregnancy, as well as foods rich in foliates (eg, dark green leafy vegetables, orange juice, dried peas, beans, lentils) have been shown to reduce NTD. The Centers for Disease Control and Prevention (CDC) estimate that women taking the recommended dosage of folic acid...
during the childbearing years and early in pregnancy can prevent up to 3,000 serious birth defects annually.8

**Medical Complications**

Children born with MSB have a 90% chance of also having hydrocephalus, which is characterized by an accumulation of cerebrospinal fluid in the head that puts increased pressure on the brain. Causes may include developmental anomalies, infection, trauma, or brain tumors.9 Surgical placement of a shunt helps to relieve the pressure by redirecting the fluid buildup to the abdominal area.2,5,7 The shunt is inserted subcutaneously behind the ear, runs down the side of the neck, and ends in the abdomen, where the excess spinal fluid is reabsorbed. In many instances, the shunt needs to be revised or replaced as the child matures.5

There has been some disagreement about adverse effects associated with shunts. According to Jansen et al, there is a correlation between hydrocephalus patients with shunts and poor hand function.10 In contrast, a study by Muen et al reports that poor fine motor skills may be a result of cerebellum abnormalities, and the lack of strength in the small muscles of the hand may be due to cervical cord abnormalities.11

While most children with MSB are not mentally retarded, complications such as hydrocephalus or infections can lead to delays in learning.2 Dysfunctions, for example, short attention span, visual perception problems, sensory processing difficulties, decreased arm and hand function, poor eye-hand coordination, and memory deficits, are associated with MSB and are complicated by hydrocephalus.

A condition common to all children with MSB is Arnold-Chiari malformation, a congenital structural defect where the brainstem is pushed into the cervical spine due to pressure from increased cerebrospinal fluid in the fourth ventricle. Common symptoms include headache, scoliosis, difficulty with balance and coordination, low muscle tone, voice alteration, decreased gag reflex, and respiratory difficulties.5,12 Other complications include neurogenic bladder, neurogenic bowel, and kyphosis (hunchback).2,7,13 Only 9% to 21% of those affected will need treatment for this condition,7,12 which involves the reduction of the pressure against the brainstem and spinal cord through surgical decompression, involving removal of small portions of bone.13

**Nutrition and Growth Complications**

A combination of poor self-esteem, dependence on family for care, and fears of maturity and independence put adolescent MSB patients at risk for eating disorders (eg, anorexia).14 Eating disorders may also be precipitated by caretakers who urge the patient to lose weight.6,14 In addition, 50% to 60% of children with MSB exhibit a disturbed growth pattern that may be a result of growth hormone deficiency.15

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Figures 1. A comparison of a normal spine to the types of spina bifida (differing in severity).
Dermatological Disorders

It has been reported that children with SB are 500 times more likely to have a reaction to latex than the general population. Other studies indicate 28% to 60% are affected to varying degrees. Symptoms may include urticaria (hives), conjunctivitis, and anaphylaxis. While some studies indicate that early and repeated exposure to latex from multiple surgeries can cause sensitivity, others indicate that the number of surgeries may not be the primary cause. A high association between latex sensitivity and foods such as avocados, bananas, and chestnuts has been reported, indicating that various foods are risk factors to SB patients. A study by Szépfalusi et al suggests the propensity for latex sensitization by the SB population may be caused by genetics.

Body positioning and assistive devices (eg, wheelchairs and leg braces) may create pressure and cause irritation to the skin. Due to the lack of sensation in the lower body, blisters and bedsores may go unnoticed by the patient. Skin infections also pose a potential danger to the spinal cord, particularly in the MSB patient, therefore, strict infection-control procedures are crucial. Bacteria do not have to travel far to infiltrate the nerves of the spinal column and cause irreparable harm. Efforts should be made to reduce the potential of irritation to the gibbus (ie, hump) when seating a SB patient in the dental chair (Figures 3 and 4). Pillows, a styrofoam ring (ie, donut), a mattress pad, or a body-size beanbag can be used to protect the gibbus and provide comfort for the patient. The patient should be allowed to shift his or her weight every 20 minutes to maintain circulation and prevent pressure sores.

Patient Management

All female patients of childbearing age can benefit from nutritional counseling regarding the use of multivitamins. Neural tube defects occur soon after conception, usually before a woman realizes she is pregnant, so the nutritional status of a woman at the time of conception is critical. As reported by Butterworth and Bendich, 60% of women who took multivitamins for three months prior to conception and three months after conception were less likely to have infants with NTDs than those women who did not take a supplement during the same time period.

Problems with obesity and eating disorders among SB patients make nutritional counseling a necessity. The dental hygienist can deliver such information in a non-threatening manner, which can serve as reinforcement for existing knowledge.

Table

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<tr>
<th>Common Items Containing Natural Rubber Latex</th>
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<tr>
<td>Latex gloves</td>
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<td>Medication stopper</td>
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<td>Window insulation</td>
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<td>Underwear elastic</td>
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<td>Stamps</td>
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Figure 2. Patient placed in an oxygen tent at birth to address respiratory problems. The presence of myelomeningocele is evident.

Figure 3. Back view of the patient.
Since patients with SB are considered at high risk for latex allergy, they should all be treated in a latex-free environment (Table). Powdered latex gloves have been reported to increase the latex aeroallergen level by as much as 115 times in areas where latex gloves are used, causing a reaction in sensitive patients. Nelson et al reported a case of anaphylactic shock by a dental patient, possibly caused by radiograph packets contaminated with powder from latex gloves.

Radiation Exposure
As diagnostic radiographs of the spine, neck, and head are important tools in long-term management, SB patients are exposed to high amounts of radiation over time. Gaskill and Marlin reported that excessive radiation exposure may be the reason patients with SB are at increased risk to develop bladder and rectal carcinomas. Definitive conclusions, however, regarding cumulative, long-term radiation exposure cannot be drawn at this time for the SB population. As additional SB patients grow into adulthood, more precise conclusions may be determined. Based on current knowledge, radiographs should be taken only if they will improve the ability of medical/dental professionals to care for the patient.

Case Report
Patient History
A seven-year-old white male patient presented for a routine oral prophylaxis. His medical history included myelomeningocele affecting the spine at T8, with kyphosis, hydrocephalus, and Arnold-Chiari malformation. The patient was born by cesarean section and had accompanying respiratory complications. Spina bifida correction surgery was performed at the age of one week and again at two weeks. A ventriculoperitoneal shunt was placed to reduce the pressure caused by the hydrocephalus. The shunt was examined annually by a neurosurgeon and had not been replaced or presented complications. Although both hips are dislocated, the parents and physician have decided not to correct this condition since the patient will never be ambulatory. Decompression surgery was performed at the age of two for the Arnold-Chiari malformation. Although the patient exhibits a neurogenic bladder and bowel, the child’s mother reported a history of only two urinary tract infections. The patient appeared to have difficulties with visual perception despite corrective lenses. When reading, he tilted his head to one side to see properly. The patient took 2 mL per day of senna, an over-the-counter peristaltic agent to prevent constipation.

The patient used a specially designed wheelchair with support straps and steadied himself with his left hand on a stable surface when not strapped in the wheelchair or otherwise secured. To avoid pressure sores on the skin, the back of the chair had a cushion with an opening to allow for the gibbus. The child’s mother reported that ulcerations occurred on the gibbus approximately three to four times per year.

The patient brushed his teeth once or twice daily with a soft-bristled toothbrush utilizing fluoridated toothpaste. No other oral hygiene products were employed. His mother reported that she occasionally brushed his teeth as a follow-up. The patient did not report any oral complications (e.g., dry mouth, aphthous ulcers, sensitivity).

Treatment
One hour before the scheduled procedure, the patient took 12 ccs of 250 mg/5 mL oral suspension amoxicillin trihydrate. A beanbag pillow was placed on the dental chair for the comfort of the patient (Figure 5), and he was transferred from his wheelchair by his mother.

A resident pediatric dentist performed an oral examination, and a dental assistant recorded the findings. The oral examination revealed a Class I amalgam restoration on T. A carious lesion was discovered on J. The patient exhibited a Class I occlusal relationship on the right side and a Class II occlusal relationship on the left side. The mother reported that she had a similar occlusal relationship. There was a lack of interproximal spacing in the primary dentition. Due to the slow growth rate of the patient, there was a possibility of crowding of the permanent dentition. The maxillary left buccal mucosa appeared slightly
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## Discussion

Annual tests to detect latex sensitivity have had negative results. Respiratory infections have been minimal, occuring only two or three times over the patient’s lifetime. There have been no complications related to the shunt; nonetheless, future replacement of the shunt may be indicated as growth occurs. No treatment has been necessary for the Arnold-Chiari malformation. Since the patient appears to be small for his age, the parents plan to have him tested for growth hormone deficiency, which is common in 50% to 60% of persons with SB.15

## Conclusion

The life expectancy of patients with NTDs is rising annually. As there are an estimated 40,000 children and adults in the US with SB, the dental community needs to make provisions to treat this population. With some minor modifications, there should be little difficulty treating the SB patient in any general dental practice. Latex-avoidance precautions, wheelchair access, limited radiation exposure, and accommodations for comfort in the dental chair are further examples of ways to integrate SB patients with safety and comfort. Placing a beanbag pillow or an egg crate mattress pad will provide additional comfort for patients with back problems as well as the SB patient. Placing a hand on the shoulder of the SB patient as the dental chair is reclining will give the patient a feeling of stability. An alternative is to leave the patient in the wheelchair during the procedure. Verbal as well as written self-care instructions are also extremely beneficial, due to the patient’s visual and perceptual difficulties. These simple procedures can easily be adapted within the dental practice and can provide comfortable accommodations for physically challenged patients.

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## References