CHAPTER **42**

SPINA BIFIDA: IMPLICATIONS IN PODIATRIC MEDICINE AND SURGERY

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INTRODUCTION

Spina bifida (Latin "split spine") is a developmental defect caused by an incomplete closure of the embryonic neural tube during the first 3-4 weeks of gestation. The consequential vertebral defect may lead to motor and/or sensory loss below the lesion causing variable spasticity, paralysis, and muscle imbalance of the lower extremities. The worldwide reported incidence of spina bifida is 1-2 cases per 1,000 live births and 0.4-0.77 cases in the US. Spina bifida is more commonly observed in females (1-3) Hispanics exhibit the highest prevalence followed by Caucasians, African-Americans, and least in Asians (4). A thorough awareness of the different severities of spina bifida and appropriate treatment options can assist setting realistic goals for these patients.

TERMINOLOGY

Spinal dysraphism is a term that describes all forms of spina bifida, but three main subtypes are typically discussed. Spina bifida occulta, the mildest form, is an incomplete closure of the outer part of the vertebrae with no protruding neural tissue. The skin covering the lesion usually will present with a dimple, lipoma, hemangioma, birthmark, or hairy tuft. This abnormality is incidentally identified on 10-20% of radiographs of otherwise asymptomatic people.

Meningocele exhibits protruding meninges between the vertebral gaps due to the failure of the dura mater to fuse. Cystic lesions filled with cerebral spinal fluid (CSF) develop but neural tissue is not involved. Both spina bifida occulta and meningocele typically do not have associated long-term health problems as long as the neural tissue is not disturbed.

Myelomeningocele (MMC), the most common and most severe form of spina bifida, results in a hernial protrusion of the dura and arachnoid mater with incorporated meninges and spinal cord. The lesion is frequently observed in the lower thoracic and lumbosacral spinal regions. The epidermis is usually absent, exposing the neural tissue without protective covering. MMC is highly associated with long-term health problems due to the neural tissue involvement and therefore will be discussed in further detail.

SPECIAL CONSIDERATIONS

MMC is associated with abnormal development of the cranial neural tube, which results in several characteristic central nervous system (CNS) anomalies. Children commonly develop hydrocephalus, a condition where increased intracranial CSF pressures dilate the brain's ventricles potentially causing permanent cerebral damage. Therefore, 80-90% of MMC patients will require a ventriculo-peritoneal shunt, a device that has greatly prolonged survival since its introduction in the 1950s (4). MMC patients may also develop an Arnold Chiari II malformation involving the caudally displaced posterior lobe of the cerebellum through the foramen magnum, which can contribute to CSF flow impediment and excessive accumulation. This deformity may cause impairment of upper extremity function in addition to lower cranial nerves causing vocal cord paralysis, difficulty feeding, crying, and breathing (5).

Tethered cords also commonly develop where tissues adhere or scar to the spinal cord diminishing mobility within the spinal column. Progressively deteriorating neuromuscular symptoms are characteristically observed with the worsening of this condition (6). Therefore, a tethered cord should become a differential diagnosis with this clinical finding, and a neurosurgical consult may be warranted.

In addition to the multiple CNS concerns, 90% of MMC patients are born with a neurogenic bladder. Historically, chronic renal failure and consequential sepsis were the most common causes of mortality. Thus, clean, intermittent catheterization should always be practiced both at home and especially in the hospital (7). Furthermore, latex hypersensitivities have a reported incidence of 3.8-38% hypothetically due to repeat exposure from multiple surgeries at a young age. Patients seen whether in the clinical setting or the operating room should not be exposed to any form of latex (8-10).

The multiple medical comorbidities associated with spina bifida create a challenge for a physician's overall treatment plan. Frasier in 1929 described the first series of spina bifida patients treated with surgical resection of the MMC sac. Two-thirds survived until hospital discharge;

the 6-year survival rate was 23% (11). Due to the recognition of these comorbidities and through recent medical advances such as the ventriculo-peritoneal shunt, survival rate prognoses have greatly improved. Presently, aggressive resection of the MMC sac is recommended within the first 72 hours of birth to prevent long-term sequelae although in utero surgical intervention has yielded promising results (12-14).

CLASSIFICATION

Classification of the severity of MMC has been controversial in the literature over the years. The most commonly utilized classification is based on cataloging normal, spastic, and paralytic muscle function with a manual muscle test to predict the level of the spinal deformity (15). In 1964, Sharrard identified all lower extremity muscles and their specific spinal level innervations (16) (Figure 1). Despite the fact that variable patterns exist with each patient, Sharrard's postulation has proven to be generally accurate when compared with recent literature.

Thoracic lesions have the worst prognosis and exhibit no active hip flexion and, subsequently, no distal motor function. Upper lumbar level lesions exhibit variable hip flexion and adduction (L1-L2) and quadriceps function (L3). Lower lumbar level lesions typically have active knee flexion against gravity as well as functioning tibialis anterior (L4) and extensor hallucis longus (L5). Sacral level lesions, having the best prognosis, display peroneal and intrinsic muscle weakness, active toe flexion, hip extension, and abduction.

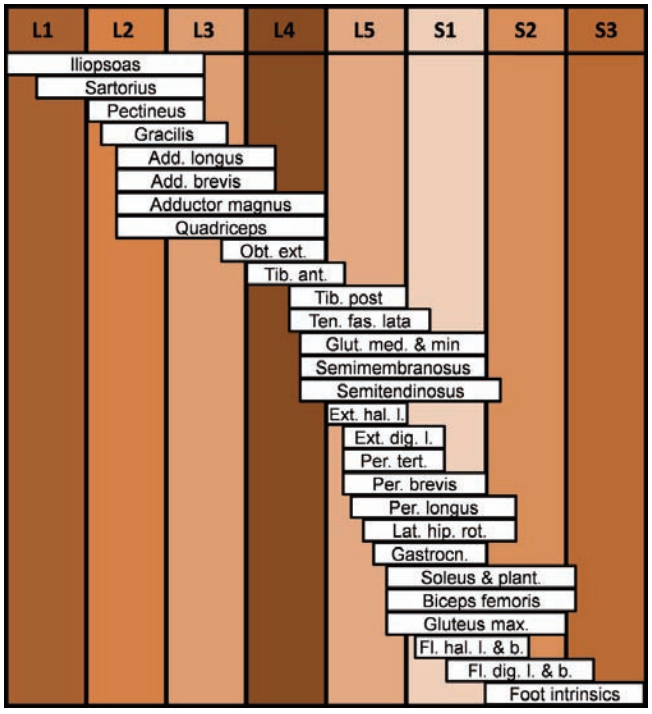

Figure 1 Neurosegmental innervation of lower limb muscles. (From Sharrard WJW. Posterior iliopsoas transplantation in the treatment of paralytic dislocation of the hip. J Bone Joint Surg Br 1964;46:426.)

Due to the fact that MMC began affecting the development of the CNS four weeks into gestation, more variations of clinical neuromuscular presentations are observed compared to other neuromuscular diseases such as cerebral palsy. Therefore, a combination of upper and lower motor neuron deficits may exist. Lindseth hypothesized that testing sensory loss at specific dermatomes within the first 18 months after birth can be used as a predictive tool in combination with a manual muscle test to more accurately determine the level of the spinal lesion (17).

CONSERVATIVE TREATMENT

After determining the suspected level, predictions can be made for long-term function and treatment goals. Generally, L2 level lesions or higher will be wheelchair bound. Two-thirds of patients with an L3-L5 lesion will be partially wheelchair-bound. Approximately 40% of MMC patients will be unable to ambulate, 30% will become functionally independent, and 30% will be employed. On average, maximal level of ambulation is achieved between ages 4-6 years and unlikely thereafter (18-20). Therefore, treatment goals for children at an early age should be based on expected function as an adult. If unable to effectively ambulate, then the goal may be directed at maintaining a stable posture in braces or in a wheelchair. Regardless, all treatment options and realistic outcomes should be discussed in detail with both the patient and the parent.

In order to achieve effective ambulation, both bracing and energy consumption should be minimized. An unsupportable torso with a collapsed posture and contracted hips and knees will require significant bracing and upper extremity function. However, 80% of patients with MMC have upper extremity impairment, which is required for bracing (21). The most important prerequisite for ambulation from a podiatric standpoint is having a plantargrade, supple, braceable foot. In addition, the spine should be balanced with the center of gravity over the pelvis. Sitting balance and posture can test for this and predict if a child may be able to ambulate. Although all lower extremity muscles play a key function with ambulation, functioning quadriceps and medial hamstrings are minimally necessary to support the torso (21).

Many bracing options exist and are based on functional level. The ankle-foot orthosis (AFO) improves swing phase and prevents dropfoot due to a weak tibialis anterior. They

also improve the stance phase and prevent a crouching gait due to weak plantarflexion. The knee-foot-ankle orthosis (KFAO) can be used with weak quadriceps function requiring knee stabilization. The hip-knee-ankle-foot orthosis (HKAFO) is used with lower lumbar level lesions and severe internal torsion of the legs causing weak quadriceps function and weak stride placement. The reciprocating gait orthosis assists with alternating hip flexion and extension for upper lumbar level lesions causing hip contractures and weak flexion (21-25).

SURGICAL PRINCIPLES

Surgical intervention is commonly performed for severe deformities at 12-15 months, roughly the time of ambulation. Most often, the goal for correction is not to restore muscle function but to create a plantargrade, supple, braceable foot (26). Therefore, tenotomies are typically preferred over tendon transfers due to their inherent weakness and spasticity. Joint-sparing osteotomies are also preferred over arthrodesis due to the fact that most patients are insensate and are at an increased risk of developing neuropathic ulcerations (27, 28). Length of casting for manipulation should be kept to a minimum due to ulceration risk and also because pathological fractures can develop due to neuropathy and osteopenia (29). Unfortunately, reoccurrence rates are reported to be high due to shifting muscle imbalance, and multiple surgical procedures are not uncommon (30).

PRESENTATIONS

Equinus deformities are observed with higher lumbar and thoracic lesions. These are usually acquired due to gravity or the infant lying in a prone position with chronic plantarflexion. An Achilles lengthening may be performed if flaccid, but a tenotomy or even tendon resection may be preferred for spasticity especially if concerned with scarred adhesions and reoccurrence (31). Severe cases may require flexor tenotomies, a posterior capsulotomy, or a talectomy (32-34).

Talipes equinovarus deformities present in 30% of patients at birth (35). They differ from idiopathic clubfoot due to the increased rigidity. Some studies show promising results with serial casting followed by Achilles tenotomy and foot abduction casting (36). However, the reoccurrence rates remain high, and many will require a posteromedial release, radical complete circumferential subtalar release, or a talectomy (37-39).

Ankle and hindfoot valgus deformities are observed with lower lumbar lesions. The gastroc-soleus muscle function is diminished or absent. If ambulatory, loading of the medial foot can potentiate skin breakdown over the talar head (40, 41). The valgus deformity may originate from the subtalar or ankle joint or an osseous deformity of the tibia. After determination of the level of the deformity, the appropriate procedure can be selected, which may include a calcaneal osteotomy, hemiepiphyseodesis, or a distal tibial osteotomy (42, 43).

Cavovarus deformities are observed with sacral level lesions. Although joint sparing osteotomies and soft tissue releases are preferred over an arthrodesis, Olney and Menelaus reported satisfactory results for a triple arthrodesis after a 10-year follow-up (44). Regardless, skin breakdown is common and should be monitored closely.

Calcaneus deformities present in 35% of patients typically with L5-S1 lesions. The calcaneovalgus deformity is most common. These patients are at high risk for heel ulcerations. Early bracing attempts often fail due to the high reoccurrence. An anterolateral release may be performed only if the gastroc-soleus muscles are not spastic in order to prevent newly aquired equinus (45). The transfer of the tibialis anterior to the posterior calcaneus has been reported in the literature with varying satisfactory results (46-49).

Vertical talus deformities are observed in 10% of patients with MMC. The congenital form is typically more rigid while the developmental form is suppler. Immediate bracing attempts are performed at birth until ambulatory in braces around 12-18 months. This rocker-bottom deformity has a poor prognosis for conservative treatment and many require at least a posteromedial-lateral release or talectomy (50).

Other orthopedic deformities should be considered that may contribute to the lower extremity deformity. A full examination should include assessment of motor and sensory function and range of motion of the spine, hip, knee, ankle, and foot. A multi-disciplinary team approach including orthopedic surgeons as well as neurosurgeons, urologists, physiatrists, and orthotists most often is necessary to formulate a common goal and prevent complications.

CONCLUSION

The most common form of spina bifida, myelomeningocele, can present with different patterns and severities. Manual muscle tests in addition to sensory dermatomal tests should be performed on bilateral extremities to assess the level of the lesion. A predicted functional level of the patient can therefore be evaluated to determine a treatment plan. Whether conservative or surgical, a plantargrade, supple, braceable foot typically yields the best results. Regardless, treatment of the spina bifida patient remains a challenge, and realistic long-term goals should be thoroughly discussed with both the patient and the parent.

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