Syndromic Feet
Arthrogryposis and Myelomeningocele

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KEYWORDS

• Arthrogryposis multiplex congenita • Myelomeningocele • Spina bifida • Clubfoot • Calcaneus • Congenital vertical talus • Syndromic foot

KEY POINTS

- Myelomeningocele foot deformities can be congenital (clubfoot, congenital vertical tali), acquired (calcaneus, calcaneovalgus), or a combination (equinus).
- The type of myelomeningocele foot deformity seen is associated with the functional spinal level.
- Clubfeet of myelomeningocele or arthrogrypotic origin have a high recurrence rate, especially with surgical procedures, including soft tissue releases and talectomy.
- Calcaneus and calcaneovalgus foot deformities are best treated early with ongoing bracing to prevent regression and to decrease the number of late presenting deformities.
- Arthrogrypotic foot deformities are the clubfoot and its equinocavus variant, and the congenital vertical talus.

INTRODUCTION

Many of the congenital and developmental idiopathic foot deformities commonly seen also present in children with syndromic conditions. These foot deformities include talipes equino varus (clubfoot) and congenital vertical talus deformities as well as pes planus. The differences between the idiopathic deformities and their syndromic counterparts often related to how early in utero the deformity began to take shape, the subsequent non-pliability of the soft tissues, and the muscle imbalances that induced or maintained the deformity. The range of syndromes that include pediatric foot deformities is vast; this article focuses on only 2 conditions: myelomeningocele and arthrogryposis multiplex congenital. The foot deformities of these 2 conditions are often discussed together, especially clubfoot. Although there are similarities, such as the increased rigidity of the deformities in comparison to idiopathic feet, and imbalances or absence of muscle function, there are also important differences between these similarly appearing feet.

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MYELOMENINGOCELE

Although the incidence of myelomeningocele has decreased in recent years (1.9/10,000 births), it is still the cause for chronic disability in 70,000 to 100,000 persons in the United States,[1,2] with approximately 1500 pregnancies per year affected.[3] The associated foot deformities are particularly difficult to manage, because of the combination of motor paralysis/spasticity and sensory loss. Before the 1950s, the survival rate of afflicted children was low, and those that survived were severely mentally disabled, nonambulatory, and confined to a wheelchair.[4] With the implementation of urgent closure of the spinal defect and techniques to control the hydrocephalus, many if not most of these children will become at least therapeutically ambulatory; those with lower level spinal lesions, with proper care, will continue to be community ambulators in adulthood.[1,2,5–7]

Foot deformities in myelomeningocele can be either congenital or developmental, and the specific deformity that occurs depends to a great extent on the functional neurologic level of involvement.[2,6,8–11] Deformities can be secondary to functional muscles lacking innervated antagonists; denervated muscles reacting spastically rather than flaccidly, due to an intact reflex arc without otherwise intact spinal pathways; or acquired deformities resulting from the cumulative effects of weightbearing across an unbalanced joint. Although there is considerable overlap, the deformities largely stratify along levels of neurologic involvement.

- In the high-level lesions, thoracic or high lumbar (L1 or L2), a flail foot without deformity and an equinus foot are the most commonly reported.[6,8,9,11] Some of those with equinus, if aggressively treated, can progress to an iatrogenic calcaneus deformity.[6]
- The clubfoot is the most common deformity in infants with midlumbar lesions (L3 and L4)[2,6] and is the most common foot deformity in myelomeningocele overall, present in 30% to 50% of patients.[5,11–13]
- In the low-level lesions (L5 and sacral level), calcaneus deformities or neurologically normal-appearing feet without deformity are present.[6,9,11,14]
- The most common developmental deformity, usually due to years of weightbearing, is a valgus ankle or pes planovalgus. Most investigators note this as occurring in the lower level lesions,[6,15–17] but some have found this deformity to regularly develop in L3 level lesions as well.[18,19]
- The incidence of vertical tali is about 10%.[20] Sharrard[10] noted that most were congenital, but that some were also developmental, secondary to weightbearing, with innervation down to at least the L5 or S1 level.[11]

Treatment Considerations

The goal of treating foot deformities in myelomeningocele is very similar to treating all neuromuscular foot deformities: a plantargrade, supple foot that can be comfortably and safely braced, taking into account insensate skin.

- Procedures that stiffen the foot should be avoided, because they make it more difficult to avoid loading pressure points on the skin, which can lead to ulceration.[21–23]
- Cast correction is an important treatment option for most of the foot deformities,[24,25] but the casts must be well padded, pressure points must be avoided, and the casts must be changed relatively frequently to detect skin injury early.
- Most deformities, once corrected, will need constant bracing. Those feet without deformity should be braced to prevent acquired deformity. Other than those with