Foot deformities in children with cerebral palsy

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Purpose of review
The present article describes common foot deformities in children with cerebral palsy and discusses treatment options for each of those deformities.

Recent findings
Bracing is a useful treatment to correct foot deformities during gait. Surgical correction of foot deformities is typically performed as a part of multilevel single-event gait improvement surgery that has increasingly become the standard of care for ambulatory children with cerebral palsy. Foot realignment may improve knee function during stance, probably because of change of lever arm.

Summary
Foot deformities are common among children with cerebral palsy. The three most common among them are equinus, planovalgus and equinovarus/equinocavovarus. Treatment consists of orthotics, physical therapy, spasticity reduction treatment and surgical correction. Guidelines for treatment are individualized and multifactorial. Important considerations include the child's level of function, the severity and flexibility of the deformity, the presence or absence of pain and skin irritation, and the changes in alignment observed over time.

Keywords
cavovarus, cerebral palsy, equinovarus, equinus, foot deformity, planovalgus

INTRODUCTION

Cerebral palsy (CP) is defined as an abnormality or affliction of the developing brain that results in a movement disorder. While the resulting encephalopathy is permanent and nonprogressive, the movement disorders that result may cause deformities of the musculoskeletal system to develop and these deformities may be progressive. The anatomy and location of the brain lesion determines the pattern of injury; a focal, unilateral lesion in the motor cortex, such as that which results from an infarct, may lead to unilateral involvement causing hemiplegic CP, while an injury in the periventricular region in preterm infants will affect the motor pathways to both lower limbs, more than the upper limbs, resulting in diplegic CP. A global anoxic event may cause injury to the very sensitive areas in the brainstem, leading to extrapyramidal movement disorders and total body involvement (quadriplegia). Most patients with CP have hypertonicity (increased muscle tone), the majority of which manifest as velocity-dependent increased tone, or spasticity. There may also be elements of dystonia, constant increase in tone not dependent on the velocity of movement. Other types of tone abnormality include athetosis, ataxia, hypotonia and mixed type, with components of several different tone abnormalities. All of these forms are characterized by loss of selective muscle control, weakness and muscle imbalance. The clinical expression is variable, and can range from very mild involvement in a fully ambulatory patient to complete absence of voluntary muscle control and inability to maintain antigravity posture, in a wheelchair-bound patient. The most frequently used classification for CP is the Gross Motor Function Classification System (GMFCS) [1]. It is a five-level scale, which classifies the severity of motor involvement of children with CP based on their functional abilities and their need for assistive mobility devices. Patients in GMFCS level 1 and 2 are independent walkers, patients in GMFCS level 3 need assistive device for ambulation, patients in GMFCS level 4 are mostly wheelchair bound but are able to stand for short periods with

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KEY POINTS

- The most common foot deformities in children with cerebral palsy (CP) are equinus, planovalgus and equinovarus or equinocavovarus.
- Management options for foot deformities in children with CP include stretching and strengthening, tone management (i.e. spasticity reduction), bracing and surgical realignment.
- The primary goal of bracing in the management of foot deformities in children with CP is to improve gait efficiency.
- The basic principles of surgical management of neuromuscular foot deformities are to realign the joints, correct the bony deformities and balance the muscles.

assistance and resist gravity, and patients in GMFCS level 5 are totally dependent and have no head and neck control.

Foot and ankle deformities are common among pediatric patients with CP at a reported rate of as high as 93% [2]. This is attributed to muscle imbalance, impaired motor control and abnormal tone. A spastic muscle group may overpower a normal or weak muscle group, creating abnormal foot position. Over time, this flexible, dynamic deformity can lead to a soft tissue contracture and, eventually, an osseous deformity. In general, foot deformities are characterized by the position of the ankle (equinus, calcaneus), the position of the hindfoot or heel (varus, valgus), the position of the midfoot (cavus, planus) and, occasionally, the position of the forefoot (adductus, abductus). The most common foot deformities among patients with CP are equinus, planovalgus and equinovarus. However, almost any combination of deformities is possible, such as equinovalgus, calcaneovalgus, equinocavovarus and others. The major principles of treating the most common three can typically be extrapolated to the other related deformities.

These foot deformities can vary from very mild and flexible to severe and rigid. As they become more severe and fixed, they can compromise standing and walking ability by affecting the stability of foot positioning in stance or the ability to clear the limb in swing. In the most severe cases, they can start to result in pain from uneven pressure and even cause skin irritation. The basic principles of treatment aim to relieve any symptoms of pain or skin irritation and optimize function.

PRINCIPLES OF TREATMENT

Treatment of foot deformities can include the use of orthoses, tone management and surgical correction. Stretching is often considered a first line of treatment in cases of foot deformity, but evidence of its effectiveness in children with CP is limited [3]. A recent randomized controlled study by Maas *et al.* [4▪▪] suggested that a knee–ankle–foot orthosis used to prevent progressive loss of ankle motion was not well tolerated and did not prevent a progressive decrease in ankle motion over time.

Orthoses are intended to improve gait efficacy by applying mechanical constraint to the foot and ankle to correct malalignment, thereby optimizing gait. Orthoses, or braces, are named by the joint they cross; hence, the typical orthotic that spans the foot and ankle up to the calf, stopping short of the knee, is called an ankle–foot orthosis (AFO). Braces that span the knee are called knee–ankle–foot orthoses, and those that cross the hip are hip–knee–ankle–foot orthoses. In the management of children with CP, braces other than AFOs are rarely used as they add significant weight and tend to be cumbersome to don and doff. AFOs are most effective in the setting of dynamic deformities to correct sagittal plane abnormalities during gait. Most commonly, this includes dynamic equinus in swing, in which case AFOs improve foot clearance by keeping the ankle out of equinus, and excessive dorsiflexion in stance to help maintain upright posture. These goals are achieved by differences in brace design. A solid AFO (Fig. 1a) restricts ankle movement in stance and swing, and may be indicated for patients with dynamic equinus and significant weakness, in whom the desired effect is to restrict plantar flexion in swing and dorsiflexion in stance. A posterior leaf spring AFO is a one-piece AFO that is made of semirigid materials and allows some amount of plantar flexion at push off and dorsiflexion in the stance phase, but still restricts equinus in swing. Hinged AFOs (Fig. 1b) are most commonly prescribed with free dorsiflexion and plantar flexion stops at 90°. In diplegic patients, crouch gait might develop over time, and hinged AFOs (HAFOs) might no longer be appropriate, because of progressive dorsiflexion in stance. In this case, ground reaction AFOs (Fig. 1c), with a solid ankle and a rigid anterior shell that places an extension force on the front of the tibia just below the knee, can be used to minimize crouch [5].

Tone management can be achieved by focal or global methods. Global tone management is typically through the use of medications, most commonly baclofen administered orally or directly to the central nervous system by way of an intrathecal catheter. Focal tone management is

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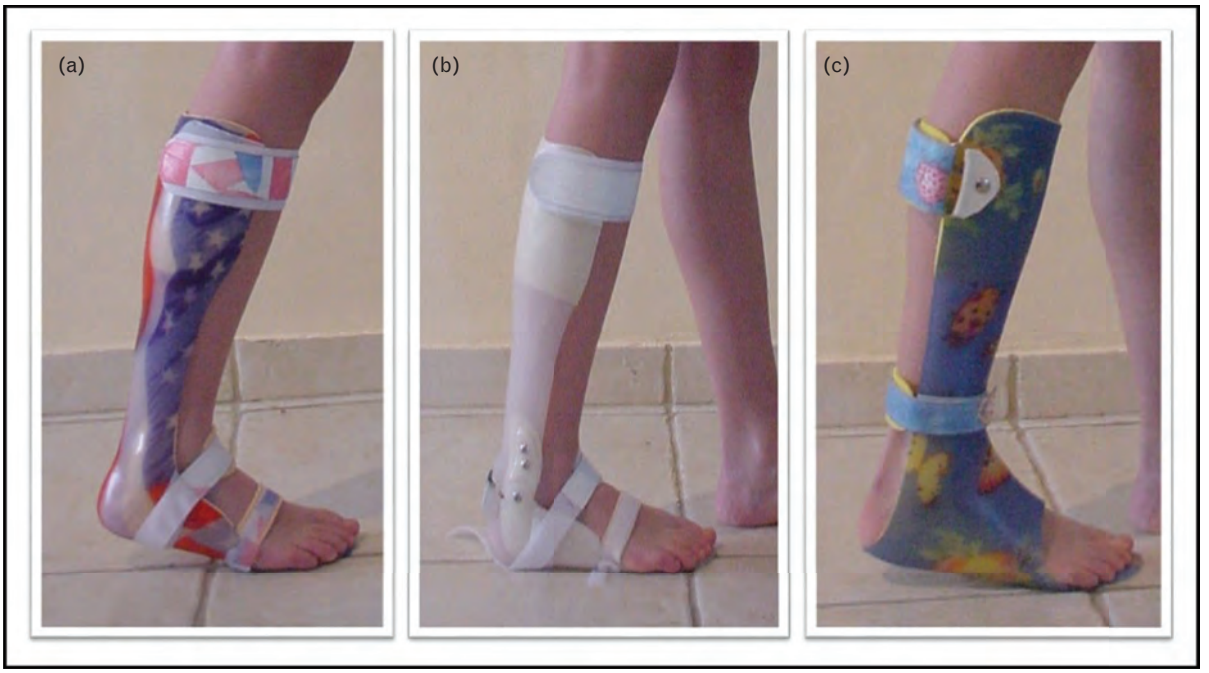


FIGURE 1. (a) Solid AFO: ankle motion is eliminated. (b) Hinged AFO: can allow free dorsiflexion and plantar flexion, however, plantar flexion can be blocked by a plantar stop at neutral. (c) Ground reaction AFO: can be either solid or hinged with a dorsiflexion stop. The anterior rigid shell places extension force on the front of the tibia. AFO, ankle–foot orthosis.

accomplished by reducing tone in specific muscles. This can be accomplished by a neurosurgical procedure called rhizotomy, which entails selectively cutting the afferent nerve rootlets in the lumbar spine, or intramuscular injections of botulinum toxin. This neurotoxin blocks the release of acetylcholine at the neuromuscular junction and partially and temporarily denervates the spastic muscle. It was recently suggested [6] that Botox injections may result in loss of muscle volume and in architectural changes of the injected muscle, possibly leading to a decreased capacity for force generation and impaired function. A suitable rehabilitation program based on strength training might, however, prevent that effect [7]. Phenol is a proteolytic agent that can be used for neurolysis of motor nerves. Compared to Botox, phenol neurolysis is more painful, technically more demanding, and takes more time to perform [8]. Focal tone management with botulinum toxin, phenol or alcohol is the most common method to address foot and ankle deformities by tone reduction; however, it can only be used in the setting of dynamic deformities. Contractures and fixed deformities are contraindications to the use of focal tone reduction because, even if the tone is completely eliminated, the bone deformity or soft tissue contracture will still be present and will prevent the foot from achieving neutral alignment. In general, if the foot cannot be passively placed in the desired position, then injections to reduce tone will not be effective.

Surgical correction of foot deformities is often necessary as the deformity progresses. It is aimed at achieving a painless, stable, plantigrade and, if possible, supple foot. It includes soft tissue procedures (tendon lengthening, joint capsule and ligament releases or transfers) and skeletal procedures (osteotomies, arthrodeses). These are often performed in combination with other surgical procedures as a part of single-event multilevel surgery to optimize gait. This approach has increasingly become the standard of care for ambulatory children with CP [9,10▪▪,11].

Treatment and prognosis of foot deformities are based on the severity of the deformity, the age of the child at the time of intervention and the child's ambulatory status. Preserving energy and allowing an efficient, painless gait is the main goal of treatment of foot deformities in ambulatory children with CP (GMFCS levels 1–3). Among nonambulatory children (GMFCS levels 4 and 5), severe deformities are common and treatment is focused on enabling comfortable shoe and brace fit, addressing skin irritation resulting from these deformities and allowing therapeutic standing.

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EQUINUS DEFORMITY

Ankle equinus is the most common foot deformity among children with CP [2,5]. It is defined as the inability to dorsiflex the foot beyond the plantigrade level, or neutral. It is the result of an overactivation of the triceps surae complex (soleus and gastrocnemius) compared with the ankle dorsiflexors. The midfoot and forefoot are in neutral alignment in equinus deformity. It is suggested that significant changes in kinetics and kinematics occur when the ankle is fixed in 10° of plantar flexion [12]. This deformity is initially dynamic, making it amenable to stretching and orthoses. This nonoperative management is often supplemented by treatment aimed at reducing spasticity that can be effectively achieved by a course of botulinum toxin injection to the spastic muscle [13,14]. Casting after this procedure is an option, and may include the knee in full extension in order to isolate the gastrocnemius muscle [15,16]. Surgical lengthening of the plantar flexors is considered when an equinus contracture has developed. Surgical lengthening can be performed along three zones of the gastrocnemius–soleus complex: in the muscle belly of the gastrocnemius, in the musculotendinous junction of the soleus and in the gastrocnemius or in the Achilles tendon. The more distal the procedure, the more correction that can be achieved but the greater the risk of causing weakness. Overlengthening might be the undesirable consequence of surgical correction of equinus deformity, especially after Achilles tendon lengthening. It may result in compromise of the patient's walking ability because of loss of ankle push-off power during late stance, and it should be performed judiciously and with caution. Firth *et al.* [17▪] suggested choosing the most conservative approach, that is, the most proximal procedure that can achieve proper correction. Children with diplegia are at particular risk for overlengthening, which can cause severe weakness and the development of crouch gait, and for postsurgical development of planovalgus deformity [18]. When they have contractures of the plantar flexors, it most commonly affects the gastrocnemius only, and not the soleus or Achilles. Children with hemiplegia are at greater risk for equinus of the gastrocnemius and soleus than diplegic patients. Young age at the time of surgery and a severe level of preoperative equinus are associated with a high recurrence rate [19,20▪]. Decision making as to the optimal procedure must take all of these factors into account, with the greatest emphasis on avoiding overlengthening, as the resultant weakness can be significant and oftentimes cause irreparable gait impairments.

PLANOVALGUS DEFORMITY

Planovalgus deformity is commonly seen among diplegic and quadriplegic patients with CP [21]. It is composed of hindfoot valgus, midfoot pronation and forefoot abduction and supination. The lateral column of the foot becomes functionally and structurally shorter than the medial column over time. The tarsal joints remain in 'unlocked' position during stance, a condition that can compromise foot stability. Planovalgus is often accompanied and exacerbated by equinus deformity of the hindfoot, with the tethered heel cord exacerbating the deforming force. External tibial torsion frequently develops over time, a factor that might further impair gait because of lever arm dysfunction [5].

This deformity is initially flexible and can be treated with foot orthoses. While orthotics attempt to improve stability in stance and may protect foot joints from excessive loads, the evidence of these effects on a planovalgus foot in children with CP is lacking [22▪]. The deformity often progresses because of increased body mass and foot load in ambulatory children or because of long-standing muscle imbalance in less ambulatory children. When surgical management is required, one needs to consider the three-dimensional, complex nature of the deformity. The goal of treatment is to establish a plantigrade, stable foot by restoring a correct relationship between the tarsal bones and restoring more normal foot progression angle during gait. An effort should be made to preserve the flexibility and mobility of foot joints in an ambulatory child, when possible, although keeping the midfoot stable is frequently an important goal as well. Lateral column lengthening, which was first described by Evans in 1975 [23], is an effective means of foot realignment in children with CP [24–26,27▪]. This method involves equalizing the lateral column length to that of the medial column by calcaneal opening wedge osteotomy. This procedure can be combined with peroneus brevis lengthening. Gastrocnemius lengthening is often required as well in order to treat gastrocnemius equinus (Fig. 2). The medial column should be assessed after lengthening of the lateral column; if there is residual supination, a plantar flexion osteotomy of the first ray or medial cuneiform can be added. In more severe cases, the medial column can be stabilized by talonavicular fusion [28–30]. The calcaneal–cuboid–cuneiform (triple-c) osteotomy, described by Rathjen and Mubarak [31], consists of a sliding calcaneal osteotomy, an opening wedge cuboid osteotomy and a pronation, plantar flexion closing wedge osteotomy of the medial cuneiform, and, accompanied by soft tissue release as needed,

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FIGURE 2. Preoperative (a and b) and postoperative (c and d) radiographs of a planovalgus foot in a child with diplegic cerebral palsy. GMFCS level 3, following surgical correction that consisted of lateral column lengthening osteotomies and medical sliding calcaneal osteotomies, performed as part of single-event multilevel surgery. GMFCS, Gross Motor Function Classification System.

can be also used to achieve proper foot alignment. Recurrence of the deformity after an isolated lateral column lengthening is common [25,26,27▪], especially among young patients and poor ambulators, and it can be successfully treated with additional medial column stabilization or with subtalar fusion [30]. Subtalar fusion combined with medial column stabilization provides good results in nonambulatory children who often present with severe and rigid planovalgus [26,27▪]. Triple arthrodesis (talocalcaneal, talonavicular and calcaneocuboid) is a reliable method of reconstruction of severe, rigid planovalgus deformities. It is indicated for nonambulators or limited ambulators, because of concern for development of degenerative arthrosis at the adjacent joints [32,33]. Interestingly, correcting a planovalgus deformity has a positive effect on knee function during stance, probably because of improvement of the lever arm function [34▪].

EQUINOVARUS AND EQUINOCAVOVARUS DEFORMITY

Equinovarus in children with CP typically involves hindfoot varus, midfoot adduction and supination with medial peritalar subluxation of the navicular on the talar head. While it is considered as being the result of tibialis posterior spasticity, tibialis anterior overactivity may also play an important role in the formation of this deformity in children with CP [35]. Cavovarus is defined as plantar flexion of the forefoot with associated hindfoot varus. Similarly to planovalgus deformity, and unlike what is seen with neuromuscular disorders of a paralytic nature, cavovarus in children with CP may be associated with equinus. The foot is typically stiff, with poor shock absorption and a diminished weight-bearing area, all of which might impair stability and increase the risk of stress injuries [36]. Equinovarus is observed mostly in hemiplegic patients, although it may

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FIGURE 3. Preoperative (a and b) and postoperative (c and d) radiographs of an equinovarus foot in a child with hemiplegia, GMFCS level 2, following surgical correction that included a gastrosoleus fractional, lengthening plantar fascia releases, tibialis posterior, cuboid closing wedge osteotomy, lateral sliding calcaneal osteotomy and a split tibialis anterior tendon transfer to the cuboid. GMFCS, Gross Motor Function Classification System.

occur in young diplegic patients as well. Orthoses are often used for mild, flexible deformities. Surgical intervention will be required if the deformity progresses.

The basic principles of managing any foot deformity with a neurologic cause are to realign the joints, correct the bony deformities and balance the muscles. In the case of equinovarus or equinocavovarus deformities, in which the muscle imbalance is most severe, following these principles becomes most critical. Muscle lengthenings, soft tissue releases and tendon transfers are effective in the setting of flexible deformities. For rigid deformities, osteotomies are added in order to achieve adequate correction.

Tendon transfers are a main component of correcting equinovarus deformities in children with CP. These are performed almost exclusively as split tendon transfers, either split tibialis anterior or split tibialis posterior, leaving half the tendon at its insertion and moving the other half to the opposite side of the foot. Decision making to indicate the best procedure is controversial and may include physical exam elements and gait analysis with dynamic electromyography and pedobarography (plantar pressures). Some authors have recommended using the timing of the deformity for decision making. They suggest that when the deformity is present during the stance phase, the overactive tibialis posterior should be transferred to the peroneus brevis, while tibialis anterior transfer is indicated in the case of dynamic supination during swing [37]. Transfer of the entire tibialis posterior is not recommended for young, diplegic patients who are poor ambulators because of the risk of poor outcomes and late valgus

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deformity [38]. Using gait analysis, the phase of gait in which the muscle is firing can be identified. When a muscle is firing out-of-phase, split transfer of that tendon is most reliable. In both split anterior tibial transfer and split posterior tibial transfer, the distal part of the tendon is split longitudinally, and half of the tendon is detached from its medial insertion and is reattached to the lateral side of the foot [39,40].

Osteotomies are required in more severe equinovarus and equinocavovarus deformities. These should be planned on the basis of the location of the deformities, paying attention to basic principles of deformity correction, such as identifying the apex of the deformity or the center of rotation of angulation. Correction of cavus, adductus and rigid supination is commonly accomplished with osteotomies of the midfoot (cuboid or medial cuneiform), first metatarsal or occasionally arthrodesis of the first tarsometatarsal joint or talonavicular joint (Fig. 3). A fixed hindfoot deformity should be corrected by a calcaneal osteotomy, which can be in the manner of a lateral calcaneal slide, laterally based closing wedge osteotomy or a combination of the two. A triple arthrodesis may be required to correct malalignment in the most severe and most rigid cases of equinocavovarus.

CONCLUSION

Foot deformities are common among children with CP, because of impaired muscle balance, control and tone. The most common segmental malalignments in this population are equinus, planovalgus and equinovarus, and can vary from very mild and flexible to severe and rigid. Management of flexible deformities is possible through orthoses, stretching and spasticity reduction treatment. The natural history of foot deformities is unpredictable, but they tend to progress over time, and become fixed. In such cases, surgical intervention is necessary, and includes a combination of soft tissue procedures, osteotomies and, at times, arthrodeses. Treatment and prognosis of foot deformities differs on the basis of the severity of the malalignment, the age of the child and their functional ability.

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Conflicts of interest

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