Current Concepts Review

AMERICAN ORTHOPAEDIC FOOT & ANKLE SOCIETY

Foot & Ankle International®
2020, Vol. 41(7) 870–880

DOI: 10.1177/1071100720922220
journals.sagepub.com/home/fai

A Consensus Statement on the Surgical Treatment of Charcot-Marie-Tooth Disease

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Abstract

Background: Charcot-Marie-Tooth (CMT) disease is a hereditary motor-sensory neuropathy that is often associated with a cavovarus foot deformity. Limited evidence exists for the orthopedic management of these patients. Our goal was to develop consensus guidelines based upon the clinical experiences and practices of an expert group of foot and ankle surgeons.

Methods: Thirteen experienced, board-certified orthopedic foot and ankle surgeons and a neurologist specializing in CMT disease convened at a 1-day meeting. The group discussed clinical and surgical considerations based upon existing literature and individual experience. After extensive debate, conclusion statements were deemed "consensus" if 85% of the group were in agreement and "unanimous" if 100% were in support.

Conclusions: The group defined consensus terminology, agreed upon standardized templates for history and physical examination, and recommended a comprehensive approach to surgery. Early in the course of the disease, an orthopedic foot and ankle surgeon should be part of the care team. This consensus statement by a team of experienced orthopedic foot and ankle surgeons provides a comprehensive approach to the management of CMT cavovarus deformity.

Level of Evidence: Level V, expert opinion.

Keywords: Charcot-Marie-Tooth disease, CMT, cavovarus, cavovarus foot surgery

Charcot-Marie-Tooth (CMT) disease is the most commonly inherited neuropathy and affects 2.8 million people worldwide and 1 in 2500 in the United States.[22] The disease is named after the 3 physicians who first identified it in 1886, Jean-Martin Charcot and Pierre Marie in Paris, France, and Howard Henry Tooth in Cambridge, England. CMT disease is a motor-sensory neuropathy with multiple genotypes.[10] By comparison, the phenotypic expression of the disease is more uniform, with 2 main presentations. Most patients who need surgical care have a progressive cavovarus foot deformity, with muscle imbalance causing a nonplantigrade foot.[9] Surgery can be life-changing for these patients, allowing them to walk potentially brace-free with more endurance and less pain. Early realignment procedures may reduce progression of joint arthritis.[19,32] A minority of patients have diffuse paralysis below the knee. These patients are best treated initially with ground-reaction ankle-foot orthoses.

Due to the relatively rare and varied presentation of CMT, surgical treatment guidelines are lacking.[26] In an

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effort to address this deficit, the lead author, with the support of the Charcot-Marie-Tooth Association (CMTA), convened a 1-day meeting to address the challenges of CMT foot and ankle surgery. Our goal was to reach a consensus on surgical treatment, where possible. The core assembled group included 13 experienced orthopedic foot and ankle specialists, including 6 past presidents of the American Orthopaedic Foot & Ankle Society, and 1 neurologist specialized in CMT disease. Conclusion statements were formulated and voted on by the group. Although we considered all patients with lower extremity involvement from CMT, our focus was on cavovarus deformity.

Definitions/Terminology

We use *consensus* to describe a general agreement of 85% or more of the group. Differing opinions may be noted in the text. We reserve *unanimous* for a decision that was supported by every member of the group. These statements are highlighted in bold throughout the text.

The terminology used to describe a cavovarus foot is often confusing. The group recognized this problem and focused first on defining the terms to be used in our discussion.

Equinus

The term *equinus* is best reserved to describe the single-plane relationship between the talus and the tibia. While *equinus* is also often used to describe the position of the midfoot or forefoot, we found that terminology confusing. **Our consensus was that the term *equinus* should be reserved to describe a plantarflexion deformity of the talus relative to the tibia.** We recognized that in a cavovarus CMT deformity the talus is rarely plantar-flexed on the tibia, and a true equinus at the ankle is therefore uncommon.

Cavus

Cavus is a multiplane deformity that describes a high-arched foot with increased plantar concavity, foreshortening of the midfoot, varying degrees of hindfoot varus and plantarflexion (from the subtalar joint), inversion of the midfoot through the cuneiforms (often referred to as supination), and plantarflexion of the medial metatarsals (most pronounced in the first).[2] There is significant variability in the contribution of each component to the cavus deformity. **We reached a consensus that the term *cavus* is acceptable but is best used in conjunction with the contribution of each component (ie, heel varus, first metatarsal plantarflexion, midfoot inversion). The use of the term *cavus* to describe the forefoot is confusing and not recommended.**

Hindfoot Varus

We had consensus that *hindfoot varus* is a midline deviation of the calcaneus relative to the long axis of the tibia. This can result from soft tissue contractures of the hindfoot joint capsules, with or without fixed arthritic changes of the joints, contractures of the posterior tibial or Achilles tendons, lateral ankle laxity, or changes in bone morphology. In most cases, the varus initially results from accommodation of the hindfoot to a valgus deformity of the forefoot (depression of the first ray) and the need for the foot to remain plantigrade when weightbearing.

Forefoot Valgus

We had consensus that *forefoot valgus* is eversion of the forefoot around the axis of the second metatarsal, commonly caused by plantarflexion of the first metatarsal in relation to the hindfoot.

Fixed vs Flexible Hindfoot Varus

The descriptors *fixed* or *rigid* are misnomers in all but the most severe patients with CMT. The varus is only truly fixed when there are severe arthritic changes or irreducible bony deformities in the hindfoot joints. The *fixed* designation should only be made intraoperatively after release of all of the potential deforming structures, including the subtalar and talonavicular joints, the posterior tibial tendon, spring ligament, gastrocnemius, or Achilles tendon. This is similar in many ways to a clubfoot correction. Only as each soft tissue structure is sequentially addressed can the surgeon know if the deformity is truly fixed.[21]

The group was unanimous in its recommendation that instead of *fixed* and *flexible* deformity, the terms *reducible, partially reducible*, and *irreducible* should be used when describing hindfoot deformity in patients with CMT. This language is not simply an issue of semantics but relates directly to how a surgeon should think about CMT reconstructive surgery. **We reached consensus that intraoperative evaluation was the best way to determine whether the deformity corrects without the need for fusion.**

Ankle Laxity vs Instability

There is a distinct difference between instability and laxity of the ankle. *Laxity* means that the ligaments are incompetent, as a result of an inversion injury, repetitive trauma to the ligaments, a hyperlaxity syndrome, or a connective tissue abnormality such as Marfan syndrome or Ehlers-Danlos syndrome. This laxity should be demonstrable on examination or stress radiographs of the ankle, either in the clinic or the operating room under anesthesia. **In contrast, we had**

consensus that most patients with CMT do not have true ankle ligamentous laxity but rather *instability* during gait from their nonplantigrade hindfoot, peroneal weakness, forefoot valgus, and altered proprioception. We reached consensus that the term *ankle laxity* should only be used when the lateral ligaments of the ankle are physically loose.

History and Examination

Patient History

Several components of the patient history warrant special attention.

History of cavovarus deformity. How long has the cavovarus been present, at what age did it begin, and is it progressing? The worse the cavovarus becomes, the harder it is to correct. Is the predominant problem pain, deformity, or both? It is important to try and distinguish neuropathic pain from the pain of walking on a deformed foot. Does the patient wear braces, and if so, what type? A better brace may preclude the need for surgery. If family members have CMT, what is the history of their deformity? What was the success or failure of their treatment?

A history of genetic testing should be noted. Although not required for surgical treatment, the CMT genotype may be helpful in better understanding the clinical presentation and prognosis. Patients with CMT1A, for example, typically have a more uniform and often less severe clinical presentation than those with other dominant forms of CMT.

Physical function. CMT is a progressive disease. Patients often experience a deterioration in function from increased pain, imbalance, and fatigue. A progressive cavovarus deformity is often at the root of all these problems. How does the patient's function compare to a year ago? Is there a difference in function throughout the day? Does your patient start the day strong, only to experience increasing limitations? Often, a discrepancy exists between what a patient reports and what the family and friends observe.[15] That realization can be important for a patient who is trying to decide on the benefit of surgery.

Expectations. CMT patients have often given up hope for improvement and have come to accept impairment as their new normal. They frequently have seen doctors who had little to offer, other than braces to help with ambulation. The hope of avoiding, or at least minimizing these braces is often the impetus for an orthopedic consultation. Many of these patients can be helped by an orthopedic foot and ankle specialist with expertise in CMT surgery. **We had unanimity that clear expectations from everyone involved is essential**. What is the chance that surgery will improve function and minimize the need for braces? How long is the recovery? When can the other foot be done? Will the deformity recur? These are just a few of the key issues that need to be discussed.

Examination

The physical examination of a patient with CMT is challenging, even for the most experienced surgeon. A comprehensive examination[13] should consist of the following elements:

Inspection

- Perform a visual inspection of the patient sitting, standing, and walking barefoot.
- Observe the position of the hindfoot relative to the forefoot. Focus on the ability of the ankle to dorsiflex in swing phase and the hindfoot to evert at heel strike.
- Carefully observe the toe extensors for compensatory overactivity when the tibialis anterior is weak.
- Inspect the hands for interosseus muscle atrophy and clawing.
- Watch the patient walk in the braces they may have. It is key to determine if the feet are plantigrade in stance phase, in and out of braces.
- Examine lower extremity alignment, from the pelvis to the foot.
- Document callosities on the plantar aspect of the foot, especially at the base and head of the fifth metatarsal.
- Does the patient have scoliosis?
- Does the patient have a hyperlaxity syndrome?
- Are there toe deformities?

Sensory examination

- The sensation of the plantar foot can be evaluated with a 5.07 Semmes-Weinstein (10-g) monofilament, pin prick, or tuning fork.
- Toe proprioception and a Romberg test (when feasible) can help evaluate the contribution of sensory dysfunction to gait impairment.

Motor examination

- **We had consensus that the Medical Research Council (MRC) manual motor testing should be used to grade individual muscle strength from 0 to 5. However, grading can be highly subjective and may fail to detect mild muscle weakness.** Examination of each key muscle along with its antagonist should be performed to understand muscular imbalances.[20]
- **We had consensus that, if possible, the most accurate evaluation of CMT muscle strength includes several examinations at multiple time points. Accurate evaluation of muscle strength, as well as**

the subsequent choice of tendon transfers, is perhaps the most important component of a successful correction of the CMT cavovarus foot.

- The cavovarus position of the foot can confound an accurate evaluation of individual muscles.
- **We had consensus that the examiner may better isolate the tibialis anterior strength by passively holding the metatarsophalangeal (MTP) joints in slight plantarflexion while asking the patient to dorsiflex the ankle.**
- Weakness of the peroneus brevis (PB) can be masked by a strong extensor digitorum longus, which in the cavus foot provides some eversion power because of its lateral position on the dorsum of the foot. Grade eversion from a maximally inverted starting position.
- Peroneus longus (PL) strength often plays an important role in reconstruction. It is extremely important to assess PL strength separately from the toe flexors and gastrocsoleus. To isolate the PL, the examiner should place both thumbs under the first and fifth metatarsal heads and evaluate the plantarflexion power beneath the first.
- Assess hip, knee, and hand intrinsic strength.

Active and passive range-of-motion testing[28]

- The plantarflexed position of the first ray that is present in most patients with CMT may make it seem like there is a significant equinus contracture of the ankle.
- Distinguish an Achilles or gastrocnemius contracture from restriction due to mechanical impingement from the talus, which lies in a maximally dorsiflexed position within the tibial plafond. This can often be demonstrated on a lateral weightbearing ankle radiograph. If bony impingement exists at the ankle, an Achilles or gastrocnemius lengthening may not improve ankle dorsiflexion.
- An Achilles or gastrocnemius contracture, however, may contribute to the varus position of the heel, given the medial insertion of the tendon on the calcaneus. This subtle but important finding should be assessed during surgery. An Achilles lengthening may not increase ankle dorsiflexion but can decrease the varus pull on the calcaneus.

Hindfoot assessment

- Reduction of the varus hindfoot into a plantigrade position dictates surgical strategy. The precise etiology of the varus deformity often cannot be determined preoperatively. Inability to bring the hindfoot to neutral may be secondary to abnormal calcaneal morphology, irreducible varus laxity of the ankle, or soft tissue contractures.
- The Coleman block test is traditionally used to determine whether a varus hindfoot deformity is forefoot driven, ie "flexible." This is signified by hindfoot deformity correction out of varus with block placement under the lateral foot. If there is no correction, the literature suggests that a calcaneal osteotomy is needed because the hindfoot deformity is "fixed.". **The consensus was that the Coleman block test may not fully characterize the deformity and should not be used in isolation for surgical planning.**
- **We had consensus that it is preferable to examine a seated patient, in which the examiner manually attempts to counteract the medial deforming forces responsible for the varus hindfoot.** This can also be done with the patient prone and the knee bent.
- **We reached consensus that the need for a calcaneal osteotomy is best made intraoperatively, after the soft tissues have been addressed. Otherwise, there is a potential to overcorrect the heel varus. In some cases, soft tissue releases of the hindfoot, without calcaneal osteotomy, may be sufficient for correction.**

Evaluation of ankle ligamentous laxity[30]

- Ankle laxity in patients with CMT should be carefully evaluated in both the clinic and the operating room.
- **We had unanimity that ankle laxity should be evaluated by anterior drawer and varus stress tests under anesthesia, prior to the start of surgery.** Fluoroscopic examination can be very helpful in the assessment.

Preoperative Imaging Studies

Numerous diagnostic tests are available for the evaluation of patients with CMT. **We had unanimity that all patients should have the following weightbearing radiographs: anteroposterior (AP), lateral and mortise views of the ankle, AP and lateral views of the foot, and a hindfoot alignment view.** A computed tomography (CT) scan and magnetic resonance imaging (MRI) may be indicated for select clinical presentations.[29] We had no consensus on routine CT scans or MRI imaging; the group believed that this was up to the clinical judgment of the surgeon. While emerging literature suggests that a weightbearing CT may be integral to the evaluation of complex foot and ankle deformity, there are insufficient data to suggest its use in patients with CMT.

We had unanimity that a gait study, electromyogram (EMG), nerve conduction study (NCS), and genetic testing are not routinely required in the *orthopedic* evaluation of patients with CMT.

Management of Cavovarus Foot

Brace vs Surgery for a Flail Foot

Approximately 20% of patients with CMT present to the orthopedic office with no motor function below the knee and no significant deformity. **We had consensus that the first line of treatment for these patients should be a ground-reaction ankle-foot orthosis (GRAFO).** Advances in bracing technology allow for a functional gait in these patients. An ankle fusion is rarely indicated as a first line of treatment, unless there is significant deformity at the ankle. Even when ankle arthritis is present with the flail foot, it is often painless because of the sensory neuropathy and is amenable to a GRAFO.

Surgical Treatment of Cavovarus

Timing of surgery. There is no evidence-based orthopedic studies to help determine optimal timing for surgery, and there is often contradictory advice from the patient's neurologist, physical therapist, and orthotist regarding the role of an operation. Many patients with CMT with cavovarus feet first face the issue of surgery during their teenage years.[6] It often takes until then for the deformity and weakness to progress to a point where there is a significant loss of function. They can no longer keep up with their classmates, have curtailed many activities, and may soon require braces. It is often the prospect of braces, in teens or adults, that leads to the first orthopedic consult.

Braces are reasonably the first line of treatment for these patients, as long as the foot is in a plantigrade position within the brace.[25] If it is not, surgery should be considered. Even if braces have to be worn after surgery, patients will invariably be more comfortable with their feet flat on the ground within the brace. Before going into a brace, all patients with CMT with a cavovarus foot or foot drop should probably first get an opinion from an orthopedic foot and ankle specialist to explore all potential options. **We had unanimity that an orthopedic foot and ankle surgeon should be part of the care team for patients with CMT early in the course of the disease.**

The longer a deformity is present, and the worse it becomes, the harder it is to correct with surgery. More severe contractures, skin ulceration, joint instability, and arthritis can occur over time. Early in its course, a cavovarus foot can usually be corrected with joint-sparing osteotomies, soft tissue releases, and tendon transfers to balance the foot.[15,16] Operative intervention at this stage may prevent increased deformity and the need for joint fusions in the future. Common sense therefore supports early surgical intervention. Surgery has an intrinsic risk, however, and the recovery can take many months. Another factor to consider is the progressive nature of CMT. No high-quality studies exist on the longevity of the surgical

Figure 1. Clinical weightbearing image of a 17-year-old patient with Charcot-Marie-Tooth disease, with uncorrected cavovarus deformity on the left and following surgical correction on the right (frontal view).

Figure 2. Clinical weightbearing image of the same patient (rear view).

correction. However, most patients would invariably prefer an operation to a brace if there is a high chance of success. That determination is based on shared decision making between the patient and the surgeon. **We had unanimity that early surgical intervention can minimize progression of the cavovarus deformity.**

Surgical strategies. Figures 1 and 2 show a teenage patient with uncorrected cavovarus deformity of the left and following surgical correction on the right.

Posterior Tibial Tendon

The posterior tibial tendon (PTT) is often the primary deforming force in patients with CMT. Its strength is usually preserved relative to its antagonist, the PB. When it is a deforming force, release of the PTT should be the first step in a CMT cavovarus reconstruction (see algorithm below). Commonly, the tendon is transferred through the interosseous membrane to the dorsum of the foot to supplement a weak tibialis anterior muscle.[7] It can also occasionally be transferred posterior to the tibia into the PB in the lateral compartment of the leg to supplement weak eversion strength.

In cases of preserved ankle dorsiflexion, management of the PTT is not as straightforward. **The consensus of the group was that in such a case, the PTT does not require transfer, as long as it is not a deforming force.** In cases where the PTT is not transferred, the key is to make sure that there is adequate strength laterally to stabilize the foot.[31] This is usually accomplished by a transfer of a sufficiently strong PL into the PB. **We had consensus that lengthening of the PTT to minimize its pull should rarely be done.** While there is currently no scientific evidence to support this recommendation, lengthening of the PTT may preclude it from being used for transfer in the future should the deformity progress. One member of the group disagreed and noted that on occasion, a recession of the PTT at its musculotendinous junction can help balance slight residual overpull of the muscle.

When performing a PTT transfer, the following statements were the consensus of the group:

- The tendon is strongest when transferred directly into bone. An interference screw is a common technique.
- The tendon should be harvested as distal as possible. Distal portions of the tendon often extend past the navicular and insert into the medial cuneiform.[24] Adequate length will ensure that the tendon does not have to be overtightened when it is transferred. To dissect the PTT out to its cuneiform attachment, a sliver of bone can be elevated from the medial navicular so as not to thin-out the tendon too much.
- The tendon should be transferred through the interosseous membrane (IOM).[32,33] A large opening in the IOM will improve the tendon line of pull and prevent the muscle from binding down as the tendon moves from posterior to anterior. The opening can be made by gently spreading a large clamp, which is then used to pass the tendon from medial to lateral.
- The tendon should be transferred deep to the extensor retinaculum. One surgeon routinely transfers the tendon subcutaneously.
- The tendon should typically be transferred to the dorsum of the foot into the lateral or middle cuneiform.
- The PTT should not be split as this further reduces the motor grade of the muscle tendon unit.
- When tensioning the tendon at the end of the case, the ankle should be held in neutral to 5 degrees of dorsiflexion and the tendon fixed at its mid-excursion. The goal is to respect the Blix curve and preserve active function of the posterior tibial muscle. Overtightening of the transfer may only accomplish a tenodesis. Err on the side of slight overtightening if the Achilles is strong, as some stretching of the transfer will invariably occur over time.
- A modified Bridle procedure involving transfer of the PTT through the IOM and anastomosis to the tibialis anterior tendon is an option based on surgeon preference.[14]
- On rare occasions, the PTT can be transferred posterior to the tibia to supplement peroneal strength, if it is not needed to assist with ankle dorsiflexion.

Joint Capsules and Spring Ligament

We had consensus that longstanding cavovarus deformity leads to contractures of the subtalar and talonavicular joint capsules, with medial "over-coverage" of the navicular on the talar head. Once the PTT is detached in preparation for transfer, these joints are easily located. It is often impossible to obtain a plantigrade foot without wide release of these joint capsules, along with the spring ligament. As mentioned above, the approach mimics a clubfoot correction. The subtalar capsular incision should be extended far posteriorly, while protecting the neurovascular bundle and *flexor hallucis longus* (FHL) tendon. **We had consensus that only after these joints are widely released can the surgeon accurately assess the need for a calcaneal osteotomy.** A calcaneal osteotomy done prior to soft tissue releases may lead to overcorrection.

Calcaneal Osteotomy

Once the medial soft tissue structures are released, the position of the heel should be evaluated carefully. A lateralizing calcaneal osteotomy is indicated when the varus deformity of the heel remains irreducible. The goal of the osteotomy is to laterally translate the weightbearing aspect of the posterior calcaneus, normalize ankle joint contact pressures, and improve gait dynamics.[3] Lateralization of the calcaneus also transforms the gastrocnemius and soleus muscles from a deforming varus force to a correcting valgus force on the hindfoot.[5] **We had unanimity that an osteotomy is often, but not always, needed in CMT foot reconstruction.**

There are numerous techniques for a calcaneal osteotomy. In mild cases, simple lateralization of the posterior tuberosity of up to 1 cm can be performed.[18,23] For moderate to severe cases of CMT, this may be inadequate to address a multiplanar deformity.[17] A closing wedge osteotomy in conjunction with rotation of the tuberosity will provide correction in both axial and coronal planes.[1] A plantar fascia release at the level of the osteotomy facilitates mobility of the posterior tuberosity. The release can be done from lateral to medial through the lateral calcaneal osteotomy incision. More commonly, it is performed from medial to lateral, through a separate incision (see Plantar Fascia section below). **Although we did not have consensus for a single type of osteotomy, we had unanimity that correction of hindfoot varus is one of the critical components of a cavovarus reconstruction.** Superior translation of the tuberosity can be incorporated into the osteotomy to decrease a high calcaneal pitch angle. In select cases, a subtalar fusion may be needed if the osteotomy does not provide adequate correction. This approach is especially useful if there is no peroneus longus or brevis function.

Tarsal Tunnel Release

The group discussed the need for a concomitant tarsal tunnel release at the time of the lateralizing calcaneal osteotomy. The release can theoretically prevent tibial nerve compression following the osteotomy.[20] **We had consensus that a tarsal tunnel release is not routinely needed.**

Peroneal Tendons

The peroneal tendons play a pivotal role in the restoration of muscle balance in CMT. The PB muscle is usually weaker than the PL. This produces eversion weakness and a deforming plantarflexion force of the first ray. As previously discussed in the examination section, it can be difficult to accurately assess PL strength. **We reached consensus, however, that if the PL is deemed to be functional—defined as a minimum 3/5 MRC strength—a PL to PB tendon transfer is recommended.** This requires that a stronger PTT is not left in place, as it will overpower the transfer.

The transfer can be accomplished by a Pulvertaft weave of the PL tendon into the PB tendon along the lateral border of the calcaneus, distal to the tip of the fibula, or in the distal leg, proximal to the superior peroneal retinaculum. **We had consensus in favor of a Pulvertaft weave over a side-to-side tenodesis**. To tension the graft, the hindfoot should be held in maximal eversion and the PL tied into the PB at the midpoint of its excursion. As with transfer of the PTT, the goal is to preserve active function of the PL muscle.

In the case of complete peroneal muscular paralysis, there is no best option. The alternatives include a Bridle-type procedure, a PTT transfer to the lateral cuneiform (or cuboid), a flexor hallucis longus (FHL) or flexor digitorum longus (FDL) transfer to the PB tendon, or a subtalar fusion.[4,12] **We had consensus that a tendon transfer of a strong PTT muscle to the lateral cuneiform, which lies lateral to the axis of the subtalar joint, is often sufficient to stabilize the subtalar joint, especially if the foot is plantigrade.** If the FHL or FDL is strong and functional, the group would consider a transfer of one of them into the PB insertion to further stabilize the hindfoot; however, there was no consensus on this. A subtalar fusion may be the only option if the heel cannot be brought into appropriate valgus. Several members of the group prefer a subtalar fusion in conjunction with a PTT transfer, to allow the PTT to have a single function as an ankle dorsiflexor. **We had consensus, however, that a subtalar fusion should be avoided if possible, especially in a young patient**.

Plantar Fascia

The plantar fascia (PF) is often contracted in patients with CMT. As the first metatarsal is plantarflexed and the hindfoot progresses into varus, the distance between the anterior and posterior foot narrows, leading to PF shortening. As intrinsic muscle wasting occurs and claw toes develop, the windlass mechanism further increases the contracture. **We had consensus that a complete release of the PF should usually be performed.**[11] The release can be done through a medial incision centered over the insertion of the fascia onto the posterior calcaneal tuberosity. The abductor can also be released with this approach. The release can alternatively be done through the lateral incision used for a calcaneal osteotomy. One member of the group only releases the plantar fascia in approximately 50% of cases, if needed after bony procedures and tendon transfers are complete.

The exact location and timing of the PF release is an intraoperative decision. In 2 specific scenarios, the PF has to be released to achieve adequate correction of the cavovarus foot. The first is when the posterior tuberosity requires significant lateral displacement after a calcaneal osteotomy. In this case, the PF should be divided directly below the osteotomy site from the lateral incision, as discussed above. The second is when a midfoot PF release is needed to allow elevation of a plantarflexed first metatarsal. This release can be accomplished with a small incision in the nonweightbearing aspect of the arch. In rare severe cases, the PF may need to be released in both locations, more proximally to facilitate lateralization of the calcaneal tuberosity, and in the midfoot to allow elevation of a severely plantarflexed first ray. **While an isolated PF release has been used in the past for the treatment of CMT, we had unanimity that a more comprehensive surgical approach is required.**

Achilles Tendon and Gastrocnemius

The group had an extensive discussion about the contribution of an Achilles or gastrocnemius contracture to a cavovarus deformity. **We reached consensus that in the majority of cases, neither an Achilles nor a gastrocnemius lengthening will significantly increase dorsiflexion of the ankle.** As discussed earlier, the talus may already be maximally dorsiflexed. This can be assessed preoperatively by a weightbearing lateral radiograph of the ankle and confirmed intraoperatively. One surgeon noted that although an Achilles contracture may not limit ankle dorsiflexion, it can contribute to varus deformity of the heel because of the medial insertion of the Achilles. A lengthening will facilitate deformity correction in these patients.

Arthrodesis

While joint-sparing options are the preferred method of deformity correction, especially in adolescents and young adults, certain clinical scenarios require a subtalar fusion or triple arthrodesis.[16] **We reached consensus that the indications for a subtalar or triple arthrodesis include painful arthritic joints (often in older patients with a long history of disease) and irreducible joints that cannot be restored to a plantigrade position through soft tissue releases and osteotomies.**[34] Arthrodesis may also be considered when there is insufficient muscle function to power a joint or when abnormal bone morphology may preclude other correction. In severe or long-standing CMT disease, degeneration of the talonavicular (TN) and calcaneocuboid (CC) joints may necessitate a triple arthrodesis in addition to osteotomies and tendon transfers to balance the foot.[27] **We reached consensus that in isolation, an arthrodesis may not be sufficient to correct deformity and preserve function. Soft tissue balancing with tendon transfers is often still required.** A supramalleolar osteotomy may have a role in rare cases for the correction of severe deformity. The group had limited experience with this technique.

Midfoot

There are multiple different osteotomies and fusion techniques for CMT midfoot deformities. **We had unanimity that CMT midfoot deformity is always 3-dimensional. We reached consensus to describe the deformity based on its apices in the sagittal and transverse planes.** Preoperative planning is particularly important if a midfoot osteotomy is required. A nonweightbearing CT scan with 3-dimensional reconstruction can be very helpful in elucidating the deformity. A weightbearing CT may not be beneficial, as it can exaggerate the midfoot deformity by driving the hindfoot into varus because of a plantarflexed first metatarsal.

Midfoot osteotomies are not commonly required, except in the most severe cases. As a general rule, if a dorsiflexion metatarsal osteotomy of more than the medial 2 metatarsals is needed, a midfoot osteotomy may be the best way to correct the deformity.[8] The osteotomy is typically performed through the cuneiforms, naviculocuneiform joints, or tarsometatarsal (TMT) joints. Motion of the transverse tarsal joints is thereby preserved. We had no consensus on which osteotomy is best. For overload at the base of the fifth metatarsal, an osteotomy of the cuboid may be helpful to elevate the lateral border of the foot.

A truncated wedge is removed at the midfoot osteotomy site to adequately decompress the tissues and allow for whatever dorsiflexion, abduction, or rotation is needed to correct the deformity. An additional closing wedge osteotomy of the first metatarsal may occasionally be required once the midfoot is fixed.

Forefoot

The valgus deformity of the forefoot in CMT results from an imbalance between a strong PL muscle and weak tibialis anterior, which draws the medial border of the foot into a plantarflexed position. Progressive contracture of the PF further contributes to the metatarsal imbalance. The first metatarsal is always involved, the second occasionally, and the third rarely.

The goal of the forefoot component of CMT surgery is to elevate and realign the weightbearing plane of the metatarsal heads. All soft tissue releases should be completed before the forefoot is addressed. A dorsal closing wedge osteotomy of the first metatarsal base is commonly performed. An additional metatarsal osteotomy may be considered if there is residual plantarflexion deformity with plantar prominence of the second metatarsal head.

It is difficult to close more than 7 to 8 mm of the dorsal metatarsal cortex without a fracture of the stabilizing plantar cortex. A closing wedge of the dorsal medial cuneiform can be added if there is residual deformity, or an open physis precludes a metatarsal osteotomy. A plantar opening wedge osteotomy of the cuneiform is another option, filled with the surgeon's choice of graft material. **We had consensus that a dorsal closing wedge osteotomy of the proximal first metatarsal is needed in almost all cases of CMT reconstruction.** One member of the group stated that once the PL and PF are divided, an osteotomy may not be needed, as the plantarflexed metatarsal will correct with weightbearing.

Toes

Weakness of the tibialis anterior can lead to recruitment of the extensor hallucis longus (EHL) and extensor digitorum longus (EDL) muscles to achieve ankle dorsiflexion. Over time, the overpull of the long extensors, in addition to loss of intrinsic motor function, leads to claw toe deformities—characterized by extension contractures of the MTP joints and flexion contractures of the proximal interphalangeal (PIP) joints. Patients have symptoms from the pressure of shoes over the fixed flexion at the PIP joints and metatarsalgia from subluxation or dislocation of the MTP joints. Several different surgical options are available for lesser toe clawing. For the great toe, fusion of the interphalangeal (IP) joint, with transfer of the EHL into the first metatarsal, is a well-accepted procedure. Patients with CMT rarely need surgical correction for a passively correctable toe deformity. Flexor tenotomies of the long toe flexors may be needed if flexion deformities occur as the foot is brought up into a plantigrade position during surgery.

In the correction of claw toes, the long extensors, both EHL and EDL, may be transferred to the metatarsals or cuneiforms, both to decrease their deforming force on the toes and to augment dorsiflexion strength of the ankle. A modified Hibbs procedure transfers the EHL into the medial or middle cuneiform and the grouped EDL tendons into the lateral cuneiform, depending on the balance needed for the foot. We had no consensus, however, on the optimal treatment of CMT toe deformities.

Consensus Statement on the Sequence of Surgery for CMT Cavovarus

We reached consensus on the sequence of surgical correction of the CMT cavovarus foot. Given the complexity of the deformity, it is often not possible to know preoperatively which procedures will be required. This should be discussed in detail with the patient and included in the operative consent.

1. Evaluation of ankle and hindfoot range of motion, ankle laxity, and hindfoot reducibility once the patient is anesthetized
2. Medial soft tissue releases, including the talonavicular joint, subtalar joint, spring ligament, and abductor hallucis longus
3. Harvest of posterior tibial tendon, if needed for transfer
    a. Transfer the PTT through the interosseous membrane to the dorsum of the foot. The tendon should not be fixed until the end of the case, so appropriate tension can be determined.
4. Lateralizing calcaneal osteotomy, with or without a closing wedge and rotation of the tuberosity, for persistent hindfoot varus. It is important to determine the need for an osteotomy only after all soft tissues have been released. Otherwise, although rare, overcorrection of the cavovarus is possible.
    a. Subtalar fusion if the varus deformity remains irreducible after the osteotomy or the joint is arthritic
    b. Complete plantar fascia release through the lateral calcaneal incision or a medial incision to facilitate translation of the tuberosity
5. Peroneus longus tendon release for transfer to the PB
    a. Pulvertaft weave distal to the tip of the fibula or proximal to the retinaculum in the distal leg
6. Modified Broström procedure, or other lateral ligament reconstruction, if needed for ankle laxity
7. Midfoot osteotomy, if needed, to correct lateral column overload, severe midfoot deformity, or plantarflexion deformity of more than the first and second metatarsals
8. First and possibly second metatarsal dorsal closing wedge osteotomy. A release of the plantar fascia in the midfoot may be needed to allow correction of a severe metatarsal plantarflexion deformity.
    a. Concurrent closing wedge osteotomy of the medial cuneiform, if needed, to correct residual plantarflexion of the first ray or if the physis of the first metatarsal remains open. An opening wedge osteotomy of the cuneiform with a bone graft is a less common option.
9. Correction of claw toes (may be performed in a second surgery)
    a. Extensor tendon transfers to the metatarsals or cuneiforms
    b. Arthrodesis of the great toe interphalangeal joint
10. Gastrocnemius or Achilles lengthening if there is an equinus contracture or the Achilles is a deforming varus force
11. Tension and fixation of the PTT and any extensor tendon transfers

Postoperatively, patients are kept nonweightbearing for 6 weeks, then transitioned into a weightbearing cast boot and physical therapy.

Conclusion

Patients with CMT present with a wide range of foot and ankle deformities that often worsen as the disease progresses. These are complex deformities that require highly specialized care. Early surgical intervention should reduce deformity progression and help preserve a plantigrade foot. We believe that a multidisciplinary approach involving neurology, physical therapy, and orthopedic surgery is in the patient's best interest. An orthotist should be involved as needed. Shared decision making with the patient, family, and multidisciplinary team will provide the highest quality of care. Our expert group has reached a consensus on the salient issues that face an orthopedic reconstruction of the CMT cavovarus foot. We believe our work is valuable, especially given the paucity of evidence-based guidelines on this topic. We do not have all the answers, but through our collective experience, we believe that these guidelines will help patients with CMT receive the best possible treatment.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article. ICMJE forms for all authors are available online.

Funding

The author(s) disclosed receipt of the following financial support for the research, authorship, and/or publication of this article: Charcot-Marie-Tooth Association.

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