ACQUIRED PES CAVUS IN CHARCOT-MARIE-TOOTH DISEASE

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ABSTRACT

Hereditary motor and sensory neuropathies, especially Charcot-Marie-Tooth disease, are frequently expressed with an acquired cavusvarus foot which is characterized by a fixed increase of the plantar arch and hindfoot inversion. Diagnosis of the underlying condition achieved through careful patient assessment and local evaluations is the keystone for decision-making about the adequate treatment. The cavus may present as an isolated deformity of the forefoot, hindfoot or it may be a combination of both locations. Related deformities, mainly the varus and toe clawing require appropriate evaluation; clinical characteristics such as severity of the deformity, impairment of the muscular power, flexibility and patient's age are important characteristics in the treatment decision. Conservative treatment of the cavusvarus foot with physiotherapy, insoles and shoe modifications are reserved to young patients and mild deformities. However, there is a tendency of the deformity to become more severe over time because of the progressive feature of the underlying neurological condition. So, the surgical treatment by using classical techniques is performed in early stages. Most importantly is the identification of the primary and main components of each deformity to properly correct them, if possible. Muscular transfers are used to treat the dynamic unbalance, retracted structures should be either divided or lengthened and localized osteotomies should be preferred over arthrodeses, which are reserved for stiff and severely deformed feet in adults.

Keywords – *Polyneuropathies; Charcot-Marie-Tooth Disease; Foot deformities*

INTRODUCTION

The pes cavus deformity is characterized by a fixed accentuation of the plantar arch[1-3]. It may be due to isolated changes localized in the forefoot or hindfoot, or as result of a combination of the two conditions[4-6]. It is a complex defect that can be associated with other deformities such as varus, calcaneus, equinus, adduction, and the claw toe deformity[3,7,8]. The most commonly used term throughout orthopedics is perhaps "cavovarus foot" which, classically, is characterized by plantar flexion of the first ray, forefoot pronation, and hindfoot varus[9].

Acquired pes cavus is often caused on the basis of a neurological disease, and hereditary sensorimotor neuropathies are the most common causes[1,10,11]. This makes the treatment of the foot in neuromuscular disease challenging, because the progressive nature of the muscle imbalance is likely to cause relapse, even after adequate surgical repair[11].

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ETIOLOGY

Generally, the first professional to assess the patient with acquired pes cavus is the orthopedist, for this disabling condition is caused by painful calluses, claw toes, and the deformation of footwear. However, the orthopedist should always consider that the etiology of the deformity most likely has a neurological basis, which should be investigated by a neurologist[12]. Although the most common condition causing pes cavus is a sensorimotor neuropathy, especially Charcot-Marie-Tooth disease, other very important conditions must be considered, such as tumors or birth defects of the spinal cord (diastematomyelia, syringomyelia, etc.)[13]. Clinical signs that should be alerting for these latter conditions are rapid progression, greatly asymmetric deformity, hyperreflexia, clonus, and changes in sphincter control[1,14]. However, even with adequate etiologic investigation, about 30% of cases are labeled as idiopathic[11,12]. Whatever the primary cause of pes cavus, the resulting muscle imbalance is the common denominator of them all.

Hereditary sensorimotor neuropathies

The current state of scientific knowledge regarding hereditary sensorimotor neuropathies, a heterogeneous group of diseases, reflects the evolution of many other areas of neuroscience[15]. This nonspecific name was tentatively proposed by Thomas et al.[16], and definitively adopted the following years by Dyck[17] who, from the anatomical and physiological knowledge of the time, developed a basic classification. A breakthrough occurred in cytogenetics and molecular biology that enabled the identification of several gene loci, such that new subtypes of the disease are still being described, resulting in a very complex classification, whose interest is not only academic, but with the practical importance of elucidating the progress of the disease and helping to conduct genetic counseling[15].

However, from the orthopedic point of view, the classical and simplified classification continues to prevail, which accepts the broad concept of sensorimotor neuropathies, mainly represented by Charcot-Marie-Tooth disease.

Charcot-Marie-Tooth (CMT) disease

Charcot-Marie-Tooth (CMT) disease is a progressive peripheral neurological condition that causes muscle atrophy and impaired proprioception[10], the diagnosis is most commonly associated with pes cavus and thus should be suspected in any patient with this deformity. It consists of more or less typical clinical manifestations, with varying degrees of severity, due to various complex genetic abnormalities that cause defects in the peripheral nerves[18].

It was first described by Friedrich Schultze in 1884. It received its names from the French neurologist and professor of pathological anatomy, Jean-Martin Charcot, and his student Pierre Marie, who described the disease in 1886, and Howard Henry Tooth, who independently published the description of the disease that same year in England. Charcot, Marie, and Tooth related the disease to an abnormal function of the peripheral nervous system cited in Beals and Nickisch[18] and Dyck et al.[19].

In the recent neurogenetic classification, the term "sensorimotor neuropathy" was finally replaced by Charcot-Marie-Tooth (CMT), which was classified into subtypes according to the clinical, cytogenetic, and neuro-myoelectric findings.

CMT disease type 1 is the most common expression (more than 50% of cases), and groups the demyelinating forms of the disease with autosomal dominant inheritance and low nerve conduction velocity (10 to 30 m/s) due to an abnormal myelin sheath[18]. It is subdivided into CMT-1A, which is a trisomy of chromosome 17 in the region containing the gene for peripheral myelin protein 22 (PMP-22), and represents most cases (60 to 90% of the demyelinating forms). The CMT-1B corresponds to the mutation of the gene for myelin protein zero (P0), and CMT type-1C are cases with unidentified defects. The CMT-1A and CMT-1B forms are the classical clinical presentations originally described by Charcot, Marie, and Tooth[15,20,21].

CMT disease type 2 (CMT-2) is the second most common form and accounts for 20% of cases, with axonal-type neuropathy. It is also autosomal dominant, but the nerve conduction velocity is almost normal, for there is no evidence of demyelination, and the amplitude of the pulse is diminished because of axonal involvement[18]. The CMT-3 disease is historically important (Dejerine-Sottas disease), more severe, and currently most of the cases have dominant inheritance. The rare recessive forms of the disease were classified as CMT-4. Type X CMT disease represents a set of changes linked to the X chromosome, so men are affected, but women rarely are[15,18,20].

CMT disease produces distal muscle imbalance, which in the lower limbs leads to bilateral foot deformity, with a slight asymmetry in relation to their strength and the deformity[18]. It is not the absolute weakness of a muscle that produces the changes, but the imbalance between muscle groups. Classically, it produces relative weakening of the muscles of the anterior and lateral compartment, but the preferential involvement is of the peroneus brevis and tibialis anterior muscles, because of the selective denervation that occurs in them. With progression, the disease also affects the posterior compartments, causing hypotrophy of all of the muscles of the legs and feet ("stork leg")[22-24].

Sensory changes occur later in the disease[22,23], are common and affect proprioceptive, vibration, pain, tactile, and thermal sensitivities[25]. In the upper limbs, the intrinsic muscles of the hand are the first to undergo symmetric hypotrophy, then the muscles of the forearm. The disease relatively spares the shoulder, torso, and facial muscles[22]. Muscle tendon reflexes are symmetrically diminished or abolished[20]. Symptoms start in the early decades of life and evolve slowly, progressing from distal to proximal. The intensity of involvement varies from case to case, as does prognosis, generally producing mild to moderate disability, but there may be significant limitations[20].

PATHOLOGICAL ANATOMY OF PES CAVUS

There is great anatomical variation in the set of deformities that produce cavus, and they can be didactically analyzed individually. The deformity can be mild, flexible, and painless, or severe, disabling, and rigid, with plantar calluses and lateral ankle instability. Thus, the pattern of muscle imbalance and degree of involvement of soft tissues should be interpreted in isolation[1]. Finally, there is no pattern of changes that is common to all cases of pes cavus and both the assessment and treatment must be individualized[9,11].

The cavus may be in the forefoot, hindfoot, or in both regions[2,4,6]. The radiographic evaluation of the foot in profile is useful in determining the main anatomical location of the deformity[6].

The genesis of most of the deformities is related to the imbalance of the intrinsic and extrinsic muscles of the foot and has very diverse patterns. It is believed that the weakness in CMT disease has an important component in the intrinsic muscles of the foot[23,26],

which is supported by the early onset of claw toes[27]; the process of fibrotic retraction of the intrinsic muscles would lead to accentuation of the longitudinal arch. However, there have been cases reported of cavus without claw toe deformity[28].

The initial pattern of imbalance commonly found in CMT disease is the weakness of the peroneus brevis muscle, which does not balance the inverting power of the posterior tibial muscle, and muscle weakness of the tibialis anterior, with relative preservation of muscle strength of the peroneus longus and triceps surae[23,29-31]. There is relative preservation of the strength of the extensor hallucis longus muscle, which acts with the dorsiflexor of the ankle when the tibialis anterior muscle is weakened.

Forefoot

The cavus deformity of the forefoot consists of plantar flexion of the metatarsals, which is generally more pronounced in the medial column, especially in the first ray[3,4]. The critical point of this change is that it causes secondary hindfoot varus[32].

The muscle imbalance results from the action of the peroneus longus muscle not being counterbalanced by the tibialis anterior muscle, which is weakened[4,23,30]. Thus, the peroneus longus muscle flexes the first metatarsal plantarly, causes forefoot pronation, and thus produces cavus. However, the lateral column remains unchanged. When unloading weight, the foot as a whole is forced to supinate to accommodate the equinus deformity of the first ray, which leads to hindfoot varus[32,33].

In the early stages of deformity, although muscular forces maintain the deformity, the foot is flexible because the soft parts are still elastic and there are no associated bone or joint deformities. In this circumstance, the subtalar joint compensates for the pronation of the forefoot and varus does not develop. With disease progression, flexible structures such as the plantar fascia, joint capsules, and interosseous ligaments become contracted and the deformity is structured[3,32,34].

The evaluation of the cavus located in the forefoot on the radiograph is performed in profile with weight-bearing, where the longitudinal axis of the talus no longer continues with that of the first metatarsal. The interruption of the longitudinal axis occurs in the midfoot and equinus of the first metatarsal is then evident.

Adduction of the forefoot occurs when the posterior tibial muscle is active in the presence of peroneus brevis

muscle weakness[23]. Adduction builds more pressure on the lateral column of the foot and a stress fracture may be formed at the base of the fifth metatarsal, as well as a worsening of calluses[35-38].

Hindfoot

Although calcaneocavus deformity may occur in the CMT disease, classically, equinocavus deformity occurs, because the triceps surae and posterior tibial muscle hypotrophy later than the tibialis anterior muscle[11,22-24]. However, other authors reported that the hindfoot is dorsiflexed and not in plantar flexion, and therefore, the forefoot cavus is responsible for the appearance of the foot in equinus as a whole[39].

When the cavus is located in the hindfoot, it arises from the muscle imbalance due to the weakness of the triceps surae muscle and relative preservation of the intrinsic muscles[12]. Thus, there is excessive calcaneus inclination, with a trend toward verticalization[23] and soft tissue contracture. Clinically, there is worsening of the plantar arch, which is already enlarged, and its profile radiograph with weight-bearing is characterized by increased calcaneal-ground angle (angle of the grip of the heel or pitch) to greater than 30°[11].

In the pes cavus, the subtalar joint axis is more vertical and the head of the talus tends to be located on the anterior process of the calcaneus. Thus, there is less subtalar mobility and the transverse tarsal joints are also more rigid[6]. The navicular is positioned superiorly, not medially, to the cuboid, which restricts the mobility of the Chopart joint and blocks hindfoot inversion[3]. There is a decreased absorption of impact during gait by the restricted capacity for hindfoot eversion during the mid-stance phase[3,6,40], with lateral column overload of the foot[3], which can cause stress fractures[40].

Varus is compounded by the pull of the Achilles tendon and the contracture of the plantar fascia, which become more medialized[3,28]. The greater tendency to support using the lateral edge of the foot exacerbates symptoms[7]. The cavus associated with hindfoot varus, contracture of the plantar fascia and peroneus brevis muscle weakness causes overloading of the lateral ligaments of the ankle, which chronically produces varus instability of this joint, with future risk of tibiotarsal arthritis[3,23,41]. Moreover, it is believed that the cavovarus deformity may cause a pressure overload in the medial tibiotarsal joint, even when the ankle ligament complex is competent[42].

Other components

The plantar fascia acts physiologically as a spring mechanism to absorb shock, increase the plantar arch, flex the metatarsal plantarly, and invert the calcaneus. In pes cavus this structure is contractured[7] and, as it is anatomically thicker and stronger in its medial portion, its retraction not only maintains the accentuation of the plantar arch, but also contributes to adduction of the forefoot and varisation of the calcaneus[6]. It is also a source of medial plantar pain (plantar fasciitis)[3].

The deformities in the toes vary by type, severity, and flexibility. The claw toes and the hyperextension of the metatarsophalangeal joints (MTP) restrict the ability to suport load during the release of the foot and its propulsive power. They arise from the intrinsic muscle weakness and relative preservation of the strength of the extensor digitorum longus, which overstretch the MTP joints, and the flexor digitorum longus, which flex the interphalangeal joints[23,31].

In cases with more pronounced plantar flexion of the metatarsals, the plantar pad of the forefoot migrates distally in relation to the metatarsal heads, which are more exposed to the load[1,9,43,44]. In addition, distal migration of the plantar pad reduces the support of the toes. The hyperextension of the MTP, with callus under the metatarsal head, and the interphalangeal flexion, with dorsal callus, are a significant source of pain and disability[45].

The extensor hallucis longus muscles and toes, which act as auxiliary ankle dorsiflexors, are not counterbalanced by the weakened intrinsic muscles and exacerbate the clawing of the hallux and other toes[23].

DIAGNOSIS

The typical patient is young, complains of progressive deformation of the feet with calluses, difficulty in wearing shoes and painful deformities in the toes[27], usually with similar cases in the family. When it appears to be acquired pes cavus, examination by a neurologist will be necessary[11], but the orthopedist must perform the preliminary assessment of sensitivity, atrophy, and tendon reflexes. The upper limbs and balance as well as the progression of changes should not be overlooked, because these aspects are important in deciding treatment.

The orthopedic examination is conventional, but with attention to identifying the main complaints, establishing the degree of deformity and the associated changes. A good evaluation of calluses is essential because they are

the causes of pain and reflect the existing mechanical disturbance. The presence of calluses suggests that the deformity is no longer flexible[44]. The gait examination should analyze the contact of the foot with the ground and verify whether there is a tendency to "drop foot" in the swing phase, as well as hyperextension of the toes resulting from the action of the long extensor trying to compensate for the weakness of dorsiflexion.

The Coleman test is performed to estimate the influence of the forefoot on the hindfoot varisation and the flexibility of the deformity[32]. The test can be accomplished in three steps; however, the first and second are the most important. The test consists of relieving the load of the first metatarsal head by using only the heel and lateral edge of the foot for support on a board. A clear improvement of the varus means that the deformity is reducible and secondary to excessive inclination of the first ray[32]. When all the forefoot is in equinus, supporting only the calcaneus on the board will cause the varus to disappear (if the deformity is flexible).

Lateral ankle instability is a common finding due to the hindfoot varisation associated with failure of stabilizing forces (peroneus brevis), which favors the occurrence of sprains[3,23,40,41]. Anterior impingement of the tibiotarsus should also be assessed.

The radiographic investigation should include axial views of the calcaneus, and front and profile views of the feet with weight-bearing. In profile, the angle between the longitudinal axis of the first metatarsal and the talus (Meary angle greater than 5°) indicates that there is cavus in the forefoot. In the cavus of the hindfoot, the calcaneus is positioned more vertically, with an increased calcaneal-ground angle (greater than 30°) and an increased angle between the longitudinal axis of the calcaneus and the first metatarsal (Hibbs angle greater than 90°). The height of the navicular is increased. Since radiography of the cavovarus foot in profile generates an oblique view of the ankle, there is an artifactual image where the fibula appears posteriorized and talar domes have a flattened appearance[3].

The diagnostic confirmation of the underlying disease with thorough neurological, electroneuromyographic, and cytogenetic examination, and other imaging tests is usually performed by a neurologist.

CONSERVATIVE TREATMENT

Conservative treatment is restricted to early cases with slow disease progression, often with a foot that is still flexible, mildly painful, fairly plantigrade, with good muscle strength in younger patients. It consists of the use of insoles, orthotics, shoe modifications, and physiotherapy[18]. This aims to maintain foot flexibility with the exercises emphasizing gains in global mobility and the stretching of retracted structures[30]. In general, conservative treatment is performed on a temporary basis[44].

For flexible deformities, the insole must have a soft coat, be made in a mold, so that it fits the foot and provides load relief in the regions of anomalous pressure, usually the heads of the metatarsals, and should have retrocapital support. A study of the distribution of loads with a baropodometer can help with this step[46]. In flexible feet with equinus deformity of the first ray, the lateral elevation in the forefoot may be beneficial in accommodating the metarsal and preventing varisation of the hindfoot[11]. The shoes should be common but comfortable and with a high toe box to accommodate the foot and insole. Semirigid orthotics for hindfoot alignment, including lateral stabilizer bars, may be prescribed in severe cases and those with lateral ankle instability[47], but have garnered little patient acceptance because of their discomfort and aesthetic compromise.

It has been suggested that conservative measures may slow the progression of the deformity[30] or even cause its reversal[28]. There have also been suggestions that surgical treatment could be delayed because surgery is not always necessary and many patients could be treated conservatively[9]. This is especially true in mild cases. However, many authors believe that the conservative treatment of pes cavus, over time, is insufficient and that delaying surgical correction would only worsen the deformity[6], because muscle imbalances can lead to bone deformations in the immature skeleton[9,23]. According to these authors, the ideal would be to act early in the deformity, while it is flexible, through releases, tendon transfers, and localized osteotomies[6,23,30,48,49], procedures that are simpler and less aggressive[50].

SURGICAL TREATMENT

The goal of surgery is to obtain a plantigrade, painless, and stable foot with flexibility and muscle strength, if possible. It is important to remember that in the pes cavus, one deformity may be secondary to another and, if the foot is flexible, correction of the primary deformity will lead to correction of the secondary. The prime example is secondary hindfoot varus, for the correction of the equinus deformity of

the first ray would correct the varus in flexible feet[32]. If the hindfoot is rigid and persists in variation in the Coleman test, the Dwyer-type calcaneal osteotomy[7,28] should be associated.

Flexible pes cavus and without established bone deformity should be treated with soft tissue procedures, such as the medial plantar fasciotomy with or without tendon transfer[6,34,49], the most common being the transfer of the peroneus longus to the brevis and the Jones procedure. Tendon transfers are especially indicated when there is muscle imbalance with progressive characteristics, and are important to prevent relapse[44].

Feet with specific osteoarticular deformities that are still flexible, besides undergoing soft tissue procedures, should also undergo osteotomies, such as elevation of the distal first metatarsal, and midfoot (Cole, Japas) or calcaneus osteotomy (Dwyer, with or without cranial displacement of the posterior fragment). As there are usually associated deformities, the procedures are combined for the correction of each primary component of the deformity[51].

Surgical options that preserve the joints are always preferable, but more severe and rigid cases must be submitted to triple arthrodesis, which is also applied as a salvaging procedure in previous surgical failures[6,22,43,50,52-54].

Cases with equinus deformity secondary to retraction of the triceps surae muscle require special assessment of gait and quadriceps strength, as lengthening the already weakened tendon and the limb with quadriceps weakness will worsen gait. This situation should be handled by accepting the hindfoot equinus deformity and making the foot plantigrade by triple arthrodesis using the classic Lambrinudi technique[55]. This procedure, a legacy of the techniques used in the sequelae of poliomyelitis, has the advantage of restoring a plantigrade foot while maintaining the equinus, or, in other words, without further weakening the triceps surae. In addition, the equinus that arises in knee extension and disappears with its flexion can be properly treated with the selective lengthening of the gastrocnemius[31,47]. The lengthening of the Achilles tendon in the calcaneocavus foot is iatrogenic, since it worsens the calcaneal gait[33].

Mild and flexible claw toe deformities often resolve spontaneously with surgical correction of midfoot cavus[28]. The need to correct these mild deformities can be researched semiologically before surgery by passively lifting the forefoot and seeing if they disappear; however, the claw toe deformity may not be completely corrected or it may be a primary deformity and be completely rigid. When it is reversible, the indication is the transfer of the long flexor tendon of the toes to the toe extensors, according to the Girdlestone-Taylor technique[56,57]. If there is fixed hyperextension of the metatarsophalangeal joint, the indication is to lengthen the extensor tendons and perform wide capsulotomy of this joint[1,45]. If the dorsal subluxation of the phalanx persists after capsulotomy and tendon release, Weil osteotomy for metatarsal shortening is indicated[9,58,59]. The fixed flexion deformity of the interphalangeal joint requires arthrodesis or resection arthroplasty (DuVries)[1,45].

The elevation osteotomy of the distal first metatarsal[60] is rarely done as a single procedure, but is usually combined with the medial plantar fasciotomy and even with the calcaneal osteotomy[1]. A well-accepted technique consists of the dorsal access on the proximal region of the bone, inserting a screw perpendicular to the longitudinal axis of the first metatarsal, dorsally-based wedge resection, leaving the plantar cortex intact to undergo a greenstick fracture and subsequent closure of the wedge. The correction is maintained by transosseous cerclage distally anchored in the bone and proximally anchored in the screw[1]. Eventually, the second metatarsal also needs to be corrected. In the case of an immature skeleton, the growth plate of the first metatarsal, which sits at the base of the bone, should not be injured.

Dwyer's calcaneal osteotomy technique[7] is a classic procedure used in the presence of rigid varisation of the hindfoot. Access to the bone is performed through a straight lateral oblique incision in the heel just below the peroneus longus tendon, going from the extreme anterior and superior region of the posterior calcaneal tuberosity and ending at the inferior junction of the posterior tuberosity with the body of the calcaneus. The wedge is removed laterally; the superior cut must be made immediately below the peroneus longus tendon. The medial cortex is preserved and serves as a fulcrum in the closure of the osteotomy. The fixation may be performed with staples, screws, or Kirschner wires[1].

In cases of hindfoot cavus with an elevated calcaneal-ground angle, osteotomy with cranial displacement of the lower fragment of the calcaneus[61,62] is indicated, with or without Dwyer osteotomy (modified Samilson procedure)[1]. However, this procedure weakens the triceps surae, which may decrease the strength of propulsion.

The osteotomies in the midfoot region are indicated when the cavus is a consequence of equinus deformity of the whole forefoot and not just the medial rays. The Cole osteotomy[30] consists of dorsally removing a wedge in the midfoot, but has the undesirable effect of causing shortening[4]. The Japas osteotomy[4] consists of a V-cut in the midfoot with correction of the equinus in the forefoot. The Akron proximal crescentic osteotomy has the power of multidirectional correction[63]. Although they have the advantage of acting at the apex of the deformity, the cuts of the osteotomies cross the joints of the midfoot and cause arthrodeses, which may not be desirable. There are other osteotomies in this region, although with a more distal location[27,64], which means a decreased ability for correction, and can cause S deformities. Giannini et al.[65,66] reported good results with the combination of plantar fasciotomy, naviculocuneiform joint arthrodesis, and osteotomy of the cuboid in flexible feet.

Arthrodesis is reserved for more severe or rigid cases, or cases with significant and symptomatic osteoarthritis after skeletal maturity[1,6,9,11,22,43,50,52-54]. It has the advantage of associating stabilization with correction of the deformity and the disadvantage of generating overload and arthrosis in the adjacent joints[52-54]. However, when there has been much loss of sensitivity, particularly deep sensitivity, it can result in Charcot type joint degeneration in the long-term[67,68].

Finally, for very deformed feet that have usually already undergone surgery, with poor soft tissue and skin conditions, correction is possible by following the principles of the Ilizarov technique[69,70].

The recurrence of the deformity has been observed even after triple arthrodesis and is attributed to residual muscle imbalance and the progressive nature of the disease[44,52,53,71]. The use of well-adapted orthotics and insoles in the postoperative period is valid and aims to maintain correction and prevent the onset of neuropathic tegument injury[44].

A recent study reported low rates of degenerative changes and reoperations in the long-term in patients with CMT disease and flexible cavovarus feet undergoing plantar fasciotomy, transfer of the peroneus longus to the brevis and elevation osteotomy of the first metatarsal. Recurrence of varus and occurrence of a small residual cavus were common, but most of the feet had good functional indexes[71]. However, few studies related to the postoperative results in surgery for cavovarus foot in CMT disease have been published and, considering the small number of patients and the heterogeneity of the disease, no absolute evidence exists to establish a gold standard treatment[18]. The current trend in the treatment of cavovarus foot in CMT disease is early intervention in the foot and ankle in order to alleviate the muscle imbalance to decrease the loss of function and long-term morbidity[1,21].

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