ABSTRACT

The clubfoot is one of the most common congenital deformities affecting the lower limbs, it still presents controversial aspects regarding etiology and treatment. In spite of its relatively high frequency, the treatment is still challenging, since the long-term aim is achieving an everlasting flexible, plantigrade, pain-free and totally functional foot. The Ponseti method has gained attention and popularity because of its satisfactory results and surgery avoidance. Presently, surgical treatment is indicated only after failure of conservative methods, avoiding extensive soft-tissue release, but performing localized corrections of the deformities, a technique also know as “a la carte” release. The future perspective is based on the knowledge about long-term results and new understanding of the clubfoot etiology, especially in the genetic field, which may eventually be helpful for prognostic and treatment. Level of Evidence: Level II, systematic review.

Keywords Clubfoot. Foot deformities. Congenital abnormalities.

INTRODUCTION

The treatment of idiopathic congenital clubfoot (CC) has presented important evolution, since ample surgical releases were the rule a few years ago, in view of the unsatisfactory results of the manipulation techniques used at that time. Today, the Ponseti method is highly acclaimed due to the more satisfactory results and the reduction in the need for extensive surgical releases.

In the sector of Pediatric Orthopedics and Foot Disorders of Hospital das Clínicas da Faculdade de Medicina de Ribeirão Preto, the Ponseti method was introduced 10 years ago by the senior author (JBV), after discussion of treatment details with the creator of the method.

In comparison with Kite’s method there has been a complete change of the prognosis and results of the deformity, which were confirmed over the years not only in local experience, but also in other national and international centers.

It is the proposal of this review to present and discuss the main ideas about the disorder, based on literature and the authors’ experience, with the purpose of increasing the understanding and disclosure of modern concepts of etiology, anatomopathology and treatment of congenital clubfoot.

IDIOPATHIC CONGENITAL CLUBFOOT (CC)

CC is defined as a deformity characterized by complex poor alignment of the foot that involves soft and bony parts, with hindfoot equinus and varus deformity (talipes equinovarus) besides midfoot and forefoot cavus and adduction.1-5

With approximate incidence of one in every 1,000 live births, it predominates in males, in the proportion of 2:1, with bilateral involvement in 50% of cases.6-9 Population variations are found in relation to incidence, whereas in the Chinese, there are around 0.39 cases in every 1,000 live births, in Caucasians one to three cases per 1,000 live births, while in Hawaiians it occurs in about seven in every 1,000 live births.6

ETIOLOGY

With etiology still unknown, several theories were proposed to explain the origin of CC, considering intrinsic or extrinsic causes, including: intrauterine position of the fetus, mechanical compression or increase of intrauterine hydraulic pressure10,11; interruption in fetal development 12; viral infections 13; vascular deficiencies14,15; muscular alterations 16-20; neurological alterations21-27; defect in the development of bones structures 3,28,29 and genetic defects.7,30-39

The discovery of the existence of fibrotic tissue in the muscles, fasciae, ligaments and tendon sheaths of the postomedial region of the ankle and hindfoot18,20 corroborates the hypothesis of primary defect of soft parts and neuromuscular units that lead to bone alterations.16,18,21,22,24,26,40

The cytocontractile proteins and myofibroblasts identified in the postomedial contracted tissues of the hindfoot30,34 are structurally similar to those present in palmar fibromatosis and express high levels of type III collagen and certain growth fac-
tors, when compared to the non-contracted tissues.\textsuperscript{41,42} Shortening, fibrosis and retraction of the muscles and ligaments in CC are said to be genetically induced, resulting in abnormal retraction capacity that could possibly be related to primary congenital deformities and also to relapses that occur, even after adequate treatment. Genetic factors may be involved in the origin of CC as suggested by studies that evidenced an increase in family incidence and in identical twins.\textsuperscript{7,43} Several investigations identified genes with evidence of association with CC.\textsuperscript{33,36-38,44-46} On the other hand, external factors related to development were also considered\textsuperscript{6,31,35} and the deformity appears not to develop before the 12\textsuperscript{th} week of gestation, pursuant to a fetal ultrasonographic study.\textsuperscript{47} The consensus concerning the true genetic inheritance pattern has not yet been established, but a multifactorial polygenic inheritance pattern that can be influenced by external factors is suggested\textsuperscript{48,49}, with incomplete dominance and variable penetrance.\textsuperscript{35}

PATHOLOGICAL ANATOMY

Three-dimensional bone connectivity is altered in a complex manner and, according to Ponsset\textsuperscript{4}, the most severe deformities are located in the hindfoot, where the talus and the calcaneus are in accentuated equinus, the calcaneus is positioned medially and angulated in varus, and the navicular exhibits accentuated medial deviation. Moreover, the posterior ligaments of the ankle, such as those from the medial and plantar region, are shortened and thickened. The triceps surae, tibialis posterior and flexor muscles of the toes are shortened.

The talus presents morphological malformation, and the neck is angulated medially and plantarily, in comparison to a normal foot.\textsuperscript{12,17,18,20,26,60,62} Moreover, the talar neck is shortened and, in some cases, absent.\textsuperscript{26} The calcaneus is directed medially on the horizontal plane,\textsuperscript{5,50} in equinus, adducted and inverted, with the anterior tuberosity facing the lateral malleolus and under the talar head (varus).\textsuperscript{18,20,50,56} In more severe cases, there is medial angulation of the long axis of calcaneus.\textsuperscript{20,26} The navicular is in extreme inversion, medialized and displaced over the talar head.\textsuperscript{12,52,54} The cuboid is displaced medially in relation to the calcaneus.\textsuperscript{50,54} The tarsometatarsal joints and the metatarsal diaphyses are mediolateralized and cause adduction of the forefoot,\textsuperscript{20,53} which is pronated in relation to the hindfoot, since the first and the second metatarsals are angulated plantarily in relation to the fifth, which, in general, is aligned with the hindfoot. Such a relationship gives rise to cavus.\textsuperscript{53} There are complex and extensive anomalies in the posterior ligaments of the ankle and posteromedial ligaments of the hindfoot.\textsuperscript{12,20,50,55,57,58} Such as retractions that exercise deforming forces and resistance to corrections. The deltoid ligament and the plantar calcaneonavicular ligament (“spring ligament”) are shortened and thickened.\textsuperscript{20,55,58,59}

Moreover, in CC, the calf muscles are smaller\textsuperscript{1,12,28,50,56,57,60,61}, the foot size, as a whole, and that of the bones, individually, is also smaller.\textsuperscript{50} Thus, the triceps surae muscle is invariably contractured and shortened\textsuperscript{16-18,50,55-58}, although it is not clear whether the shortening is primary or secondary. Anomalous or accessory muscles are more frequent in idiopathic congenital clubfoot\textsuperscript{50,62-67} and there are also variations in the tendon insertions\textsuperscript{41,66}, including more medial insertion of the calcaneal (Achilles) tendon, which contributes to the varus angle.\textsuperscript{58} Vascular malformations were described in CC whose origin can be congenital, or otherwise adaptive to severe and prolonged deformity.\textsuperscript{14} The most frequent anomaly is absence or hypoplasia of the anterior tibial artery, which is present proximally in the leg, but ends abruptly in the ankle or in the calf, with deficient or absent anastomotic network.\textsuperscript{14,68-73} The absence of pedis pulse is more frequent in CC of greater severity and in older children.\textsuperscript{74} In rarer situations, there can be more accentuated vascular insufficiencies, which also compromise the perfusion provided by the posterior tibial artery.\textsuperscript{71,75-78} In these cases, the blood supply occurs separately through the fibular artery.\textsuperscript{73}

TREATMENT METHODS

Kite’s method

In 1932, Kite,\textsuperscript{79,80} in opposition to the methods then used, published a gentler manipulation method aimed at correcting each component of CC separately, and not simultaneously. Adduction correction consisted of foot abduction with fulcrum in the midfoot and support in the calcaneocuboid joint. Varus correction was performed with hindfoot eversion, with wedges or plaster cast changes. The manipulations successively forced abduction and pronation of the forefoot. After adduction and inversion correction, the forefoot and hindfoot equinus deformities were corrected with progressive dorsiflexion.

However, neither Kite’s method or other predecessor techniques of manipulation and plaster cast change techniques allowed complete correction of the deformities, and they resulted in feet with residual cavus, “rocker-bottom” feet, lateral torsion of the ankle, flattening and deformation of the upper side of the talar body, navicular subluxation, ligament and capsular rigidity, among other alterations.\textsuperscript{4} As other authors did not manage to reproduce the same rate of good results described by Kite\textsuperscript{51,82} they resumed surgical treatment when there was resistance to correction by the conservative technique. Codivilla’s classical medial release was performed often.\textsuperscript{53} The posteromedial release

In the 70s, based on the studies of Turco\textsuperscript{58,59}, the extensive release of soft parts became popular, with emphasis on posteromedial release. However, the complication of hypercorrection with hindfoot valgus was a common occurrence. Consequently, variations on the technique arose in the 80s and 90s.\textsuperscript{84-87} However, the long-term results continued unsatisfactory, with joint and ligament rigidity, ankylosis, weakness of the triceps and of the dorsiflexors, residual deformity due to hypocorrection or hypercorrection, navicular displacement, flattening and necrosis of the talus, skin necrosis, infections, scars with hypersensitivity, gait disorders, pain and late onset arthrosis.\textsuperscript{51,54,60,82-88,91} The current tendency is to avoid surgery with extensive joint releases\textsuperscript{4} and the use of surgery as a primary correction method is limited to the customized procedure, where just the structures required to achieve correction are released.\textsuperscript{92} (Figure 1)
Ponseti’s method

Ponseti, after in-depth studies on the pathological and functional anatomy of CC, starting in the 40s, developed and refined his own treatment method, in view of the poor results obtained with surgical and non-surgical treatments then in practice. He established details of manipulation and casting maneuvers, as well as follow-up after sectioning of the calcaneal tendon and strategies for prevention of relapses, based on the child’s age and on the parents’ cooperation. In addition, he identified and announced the most common mistakes.

Other initial treatment methods such as the use of the Denis Browne splint, physiotherapy, stretching and taping (French method) may have relative success, when adequately applied, but failure to achieve complete correction of the deformity is common.

The Ponseti method, composed basically of manipulations and serial plaster cast changes, percutaneous sectioning of the calcaneal tendon and use of abduction orthosis, has become the preferential method for the treatment of idiopathic CC in many countries, in the last ten years (Bor; Coplan, Herzenberg, 2009; Dobbs, Gurnett, 2009; Dobbs et al., 2004b; Herzenberg; Radler, Bor, 2002). With widespread acceptance, it was extended for use on older children, complex and resistant feet, including relapses after extensive surgical release, and also, in non-idiopathic cases, such as in myelomeningocele and distal arthrogryposis.

The grounds of the manipulation technique consist of correcting deformities by means of the plastic change of the contractured and shortened elements, which have a high elastic capacity in the younger child. Pirani, Zeznik and Hodges confirmed, with the use of magnetic nuclear resonance imaging that, with the Ponseti method, there is not just correction of foot bone connectivity, but also bone remodeling guided by mechanical stimuli, according to Wolff’s classical theory.

The treatment, according to Ponseti, should be started in the first days of life, with gentle manipulations, performed at intervals of five to seven days, followed by the application of a plaster cruropodalic cast, with the knee flexed ~90°. Cavus is the first deformity to be corrected with forefoot supination and plantar support on the head of the first metatarsal. Adduction and varus are corrected simultaneously in the next three or four casts, with counter-support on the lateral surface of the talor head and forefoot abduction, in supination. In achieving abduction of ~70°, varus should be corrected. Equinus correction should only be started after the correction of adduction and varus, with modeled plaster cast in the posterior part of the foot, with dorsal flexion. Ponseti gave the name “Kite’s error” to the support performed in the calcaneocuboid joint and the attempt to correct varus with pronation, as there is, respectively, blocking of the adducted calcaneus below the talor head (which prevents lateral rotation of the calcaneus and maintains the varus deformity) and cavus accentuation. Figure 1 illustrates a case treated by the Ponseti method.

The Ponseti method is being widely publicized, both in developed countries and in those under development, on account of the good correction rates achieved, which are close to 90%, while in Kite’s technique, around 50% of cases require surgical intervention and around 40% present residual deformity. Another important factor is that the treatment time with Kite’s technique is approximately twenty-two months, while with the Ponseti method, the time is from two to four months.

Herzenberg, Radler and Bor reported that the Ponseti method was able to significantly reduce the need for posteromedial surgical release (3% against 94% by the traditional technique of that time).

However, most cases treated by the Ponseti method present residual equinus and require percutaneous sectioning of the calcaneal tendon, originally performed with an ophthalmic scalpel blade, through a small incision in the skin. Tenotomy is indicated when the hindfoot does not attain 15° of dorsiflexion, after achieving varus and adduction correction, and is necessary in 70 to 90% of patients. Attempts at forced correction of equinus with plaster produces the classical “rocker-bottom” deformity. Furthermore, relapses after treatment can be frequent and are

Figure 1. Child with bilateral CC resistant to treatment by the Ponseti method. A, B and C – clinical aspect at 12 months of age, after treatment attempt (Ponseti), with residual cavus, varus and adduction. D, E and F – clinical aspect at three years of age, after posteromedial release on the right (deformity in calcaneus, residual varus and adduction) and plantar fasciotomy on the left. (Authors’ material).

Figure 2. Child with bilateral CC treated by the Ponseti method. A and B – clinical aspect at 2 months of age, prior to treatment. C and D – clinical aspect at two years of age, after correction of deformities using the Ponseti method and percutaneous tenotomy of the Achilles tendon. The feet are plantigrade, flexible and without deformities or scars. (Authors’ material).
part of the actual natural history of the disease. They are caused by the same pathological factors that initiated the deformity. The use of abduction orthosis and daily manipulation at home should be encouraged, as they can act preventively. Relapses can be treated fast with two or three plaster cast changes, but cases with a strong tendency for dynamic supination are candidates for transfer of the tibialis anterior tendon to the third cuneiform bone. Surgery can prevent future relapses and correct the talocalcaneal angle. The occurrence of varus and/or residual adduction can also be treated with localized surgical corrections, such as osteotomies on the midfoot (cuboid subtraction wedge or combined wedges) or in the calcaneus (Dwyer), which avoid surgical joint releases.

Calcaneal tendon sectioning
Percutaneous sectioning of the calcaneal tendon was performed widely for a long time, although without any descriptions of the complications and risk of the procedure. However, those in the orthopedic field express misgivings about the possibility of injury to the adjacent structures and concern regarding the quality of tendon healing.

Complications were reported recently after percutaneous tenotomy of the Achilles tendon. These include excessive bleeding, attributed to lesion of the posterior fibular artery or saphenous vein and formation of pseudoaneurysm. Clinical studies with late follow-up showed that tenotomy does not cause late onset effects such as tendon weakening and ruptures. Previous studies on other types of disease showed that there is complete repair of the calcaneal tendon after its total sectioning at the myotendinous junction, or more distally. The tendon appears to undergo complete repair within six weeks after the sectioning. Repair of the calcaneal tendon after percutaneous tenotomy in the CC was studied more recently verifying fast healing, with recovery of the mechanical transmission of movements in all the cases three weeks after the sectioning. At six months after sectioning, the repair tissue was similar to the normal tendon and formation of pseudoaneurysm. Several instruments were used for percutaneous sectioning of the calcaneal tendon. Ponseti initially advocated the use of the ophthalmic scalpel blade. As it is long and pointed, the use of a shorter ophthalmic blade with a rounded end was suggested. Common scalpel blades such as no. 11 and 15 are widely used. The 1.6mm caliber needle has been employed more recently. The surgical technique of tenotomy is more important than the instrument used. The entry point should be on the level of the medial edge of the calcaneal tendon, about 1.0 cm above the insertion, in order to avoid the posterior tibial neurovascular bundle. The section should be assessed by palpation, with the extremity in accentuated plantar flexion, both medially and laterally, besides shortening and hyperextension of the hallux. The calcaneal tendon is under more tension than usual, is long, fibrotic, and palpable up to the proximal half of the calf and produces accentuated equinus, with deep posterior fold in the ankle. The forefoot, besides adduction, is in accentuated plantar flexion, both medially and laterally. The lateral malleolus is more protuberant. The talus appears smaller and its head is not easily palpable, as in habitual CC, and it is frequently confused with the anterior calcaneal tuberosity. Attempts at correction by the habitual technique do not work as support occurs in the anterior calcaneal tuberosity, with hyperabduction in the Lisfranc joint and deterioration of plantar flexion of the metatarsals. There are a large number of plaster cast changes and the casts slip easily. For this reason, many orthopedists opt for surgical treatment. In these cases, Ponseti himself recommended a modified manipulation technique. Correction initially requires correct identification of the talar head, which is smaller than usual, in front of the malleoli, with dynamic perception of the navicular and of the anterior calcaneal tuberosity. At the time of the counter-support, for manipulation and casting, it is necessary to make sure the procedure is performed in the talar head, and not in the prominent anterior calcaneal tuberosity. The abduction should reach around 40º, after two or three plaster cast changes. Afterwards cavus correction is started with support under the head of the first and fifth metatarsals, for correction of forefoot plantar flexion, with an assistant providing counter-support at

**CLASSIFICATION OF CC**
CC has variable expression and there are classifications that consider only clinical aspects, while others also take into account the radiographic aspects. However, no classification system has prevailed until now, but the main classifications are those of Dimeglio and of Pirani.
satisfactory results. In older children modifications were suggest-
be posterior release of the tibiotarsus and subtalar joint.

If residual equinus remains after tenotomy, there can 
allow greater accommodation and remodeling of the soft and

manipulations and plaster cast changes are performed every two weeks, 

abduction of ~40° is prescribed for the patient.

However, the upper age limit, both for treatment using the 
Ponseti method, and for the performance of percutaneous te-
notomy, is not well established.

The use of the Ponseti method was also extended to relapsed feet, 
including after surgical releases, with reports of good outcomes101.

TERATOLOGICAL, SYNDROMIC AND NEUROLOGICAL CC

The Ponseti method has been used for the treatment of CC in ar-
throgryposis with satisfactory short-term results104,105,132, although 
with modifications such as performing percutaneous sectioning 
of the calcaneal tendon as a first corrective measure104 or even 
accepting abduction gain of 40-50°132, including in the abduction 
orthosis. Apparently, flexibility of the foot improves with the 
evolution of treatment and a lower degree of surgical correction 
is necessary, yet relapses are common.132

The method has also been applied in cases of myelomeningo-
cele102,103, but it is worth remembering that in diseases with foot 
sensitivity alterations, treatment with plaster cast may be danger-
ous and provoke severe skin lesions, stress fractures and plastic 
deforation, especially after acute corrections, such as calcaneal 
tendon sectioning.103

In general, when compared to idiopathic CC, the Ponseti method 
in the treatment of syndromic CC results in a higher average 
number of plaster cast changes and greater frequency of failures, 
relapses and the need for additional surgical procedures. How-
ever, the correction produced is satisfactory in most cases.133

The use of the abduction orthosis should also be encouraged in 
spite of the tendency for reduced family participation and higher 
incidence of skin lesions. Abduction should be the same as that 
achieved at the end of the treatment with plaster cast.103

FINAL CONSIDERATIONS

To guarantee that patients with CC are adequately treated in 
a progressively less invasive manner, with functional, flexible, 
painless feet, without deformities or callosities and that do not 
need special footwear, it will be necessary not only to know the 
disease pathogenesis and other technicalities, but also the late 
functional results of the various types of treatment. Most related 
surveys are still of short and medium terms and compare the 
Ponseti method with other surgical or non-surgical methods. 
Thus in the future, new evidence will help to clear up the current 
uncertainties and controversies related to the treatment of CC.

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