







# Congenital Clubfoot – Is the Ponseti Method the Definitive Solution?\*

## *Pé torto congênito – O método Ponseti é a solução definitiva?*

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### Abstract

#### Keywords

- ▶ foot deformities
- ▶ talipes/therapy
- ▶ congenital abnormalities
- ▶ treatment

### Resumo

#### Palavras-chave

- ▶ deformidades do pé
- ▶ pé torto/terapia
- ▶ anormalidades congênitas
- ▶ tratamento

Congenital clubfoot is one of the most common deformities at birth. The inadequacy or absence of treatment causes serious limitations for people with this condition. The initial treatment using the Ponseti method ensures functional results superior to other treatment modalities previously proposed. However, recurrences and neglected feet are still a challenge today. An understanding of the pathophysiology of the disease, as well as of the anatomy and local biomechanics and a thorough clinical and radiological evaluation of patients are essential to understanding the limits of the method and choosing the best treatment.

O pé torto congênito é uma das deformidades mais comuns ao nascimento. A inadequação ou ausência do tratamento provoca sérias limitações aos portadores desta condição. O tratamento inicial pelo método Ponseti garante resultados funcionais superiores a outras modalidades de tratamento propostas anteriormente, porém as recidivas e os pés negligenciados ainda são um desafio na atualidade. O entendimento da fisiopatologia da doença, da anatomia e biomecânica local e uma minuciosa avaliação clínica e radiológica dos pacientes são imprescindíveis para entendermos o limite do método e escolhermos o melhor tratamento.

### Introduction

Congenital clubfoot (CCF) is one of the most common musculoskeletal deformities at birth, affecting ~ 1 to 2 per 1,000 live births.<sup>1</sup> It is characterized by a three-dimensional deformity of the foot, in which four elements are present:

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cavus, forefoot adduction, hindfoot varus, and equine. The presence of the four deformities in the newborn without other comorbidities, not reducible to clinical examination, define the diagnosis of idiopathic CCF.

The method developed by Ignácio Ponseti is widely recognized as the gold standard for the treatment of this pathology, consisting of serial manipulations and application of plasters in weekly exchanges, in order to lengthen the contracted posteromedial structures and restore the anatomical relationships between the bones. Then, most of the time, a percutaneous tenotomy of the calcaneus tendon is required to correct the hindfoot equine. It continues in a period of correction maintenance with the use of a foot abduction orthosis to avoid recurrence of deformities.<sup>2</sup>

When applied early, performed by trained professionals, and observing the appropriate methodology, the method results in the correction of the deformity in most cases. It is important to highlight that the treatment goes beyond the plaster phase and requires care to ensure good results. The final success of the treatment is based not only on the knowledge and medical technique obtained, but on the effective orientation of parents and monitoring of adherence to the orthosis, which has been the main responsible for maintaining the correction obtained in the plaster phase.<sup>3</sup>

The protocol for the use of the abduction orthosis is of 23 hours a day until 3 months after the removal of the last plaster made at the time of tenotomy, and 14 hours a day until the child is 4 years old. This protocol is based on studies by Ponseti that showed that the risk of recurrence is higher in the first 5 years of life.<sup>4</sup> Ponseti reports that in the first 20 years of his practice, when there was still no awareness from parents about the use of the orthosis up until the child reached 3 or 4 years old, there was recurrence in half of the cases, on average, at ~2 and a half years old.<sup>4</sup>

Nonadherence to the use of the orthosis, either due to family or socioeconomic problems or to the lack of knowledge of the protocol itself, is the main factor associated with recurrence of CCF.<sup>5</sup> According to Ponseti, the use of the orthosis favors continuous stretching and prevents the formation of a new fibrosis in the ligaments and tendons around the medial aspect of the foot and ankle that lead to the return of the deformity. Nevertheless, the nonadherence to the orthosis does not explain all cases of recurrence that occur and, therefore, several studies have been published with the intention of identifying factors that may indicate a higher chance of recurrence of deformities.

Neurological, radiological,<sup>6,7</sup> electromyographic, and histological<sup>8</sup> studies found high rates of neural and myopathic alterations, including an increase in type I muscle fibers, fatty degeneration, and fibrosis.<sup>8</sup> Gelfer et al.<sup>9</sup> found that recurrence occurred in 16% of the children with idiopathic clubfoot, and showed that decreased activity of the right-hand muscles (short fibular, long fibular, and third fibular) is highly predictive of recurrence and has a greater

need for surgical intervention after 2 years of follow-up. Little et al.<sup>10</sup> suggested that the activity of the reverse muscles should be measured and added to the initial evaluation of the patient to propose a strategy to prevent recurrence. Factors such as age at presentation, initial Pirani score, and number of exchanges in the case were not significant indicators of recurrence or of need for additional surgical intervention.

Recurrence remains a challenge throughout childhood. Relapses occur more frequently and rapidly during the first 5 years of life, being extremely rare after the age of 7 years old.<sup>4</sup> In CCF adequately treated by the Ponseti method, recurrence rates range from 26 to 48%.<sup>11</sup> As this problem is not resolved spontaneously, early detection and immediate treatment of the affected feet are essential. The prevention of recurrence is one of the major challenges during the treatment of patients with idiopathic CCF initially treated by the method.

A recent systematic review reveals that few studies have a follow-up of patients beyond the age of 8 years old, and that this may reflect an underreporting of relapses in children older than this age.<sup>12</sup> In this same study, the authors found that the number of surgeries increases as follow-up increases.

## Relapse and the Ponseti Method

Knowledge of the pathogenesis of the clubfoot is essential for the prevention, recognition and understanding of recurrence. According to Ponseti et al.,<sup>13</sup> recurrences are caused by the same pathology that initiated the deformity and it is a mistake to believe that the recurrent deformity would be the same as the first, which has not completely corrected during treatment. The development of the deformity begins in the 2<sup>nd</sup> trimester of pregnancy and muscle and ligament changes continue to occur until the 3<sup>rd</sup> or 4<sup>th</sup> year of life.

The disease involves the entire posterior and medial region of the leg, ankle, and foot. There is a slight decrease in muscle size and an excess of collagen synthesis with fibrosis and retraction in the medial and posterior tarsal ligaments, in the deep fascia, in the Achilles tendon, and in the posterior tibial tendon. In 2012, Ippolito et al.<sup>14</sup> found a mean difference in volume and length between the three muscle compartments of the leg of the patients with diagnosis of unilateral CCF, with the muscles on the side of the foot slender and shorter than those on the normal side, showing involvement of the entire distal segment of the lower limb.

Histological studies showed fibrotic changes in tissues extracted from patients with CCF. Molecular studies revealed changes in the amount of type I, III, VI and profibrotic cytokines, such as transformative growth factor (TGF- $\beta$ ) and platelet-derived growth factor (PDGF). These studies have confirmed the presence of tissue with fibrosis, especially in the medial aspect, focusing on the medial malleolus, on the talus support, and on the navicular bone. This area presents a rigid nature with hyperproduction of extracellular matrix components.<sup>15</sup>

To fulfill the lack of prognosis, other possible methods to predict recurrence have been tested. Recently, a low eversion muscle activity has been considered as a significant recurrence parameter in CCF. The short, long, and third fibular eversion muscles of the foot have variable activity in patients with CCF. Gelfer et al.<sup>9</sup> revealed a rate of 16% in children with low eversion muscle activity. The form of evaluation followed the same pattern of the Pirani scale, in which 0 indicated normal muscle activity, 0.5 some activity, and 1 absence of muscle activity. In noncollaborative children, muscle strength was tested by manual stimuli in the lateral region of the foot and ankle.

Deformities that occur in an early treated foot may be residual or relapsed. Although it is not easily identifiable which were the deformities that returned to the old pattern versus those that were not corrected, the implication of this differentiation interferes little in the indication of treatment. However, late causes of recurrence require investigation of other associated diagnoses, such as neurological causes.

Recurrence severity considers the time, foot position, and stiffness of the deformity. Although the classifications for primary deformity are reproducible, relapses do not yet have a validated classification, perhaps due to the heterogeneity of presentation and different combinations of poor positioning of the bones of the foot or recurrences of some components in feet that have never been completely corrected. The most common order of recurrence is the return of the equine, followed by the varus. But this is not a rule. The equine may relapse on feet with hindfoot varus never previously corrected. With gait, mainly with equine and varus, the adduction and supination are installed, the latter also contributing to anterior tibial tendon hyperactivity. The muscle imbalance caused by weakness of the eversion muscles of the foot causes the anterior tibial tendon to present with greater activity. The lack of opposing strength to the action of the anterior tibial muscle, associated with its more medial and plantar insertion in the CCF, favors the development of dynamic supination during gait. The presence of tendon hyperactivity is an important factor to be evaluated in the dynamic inspection of the patient.

The correct evaluation of the existing deformity is the best way to establish the treatment plan. The clinical evaluation depends on static and dynamic inspection, palpation, and special maneuvers. Recently, Pierz et al.<sup>16</sup> conducted a gait analysis study in a patient with recurrent idiopathic clubfoot and concluded that these patients presented changes in gait pattern explained by impaired ankle joint and compensation in the knee, hip, and pelvis. In this study, patients with unilateral clubfoot presented decreased dorsiflexion range of motion and greater associated gait impacts and compensations. Thus, the instrumented gait examination has proved to be an ally for the evaluation of dynamic changes during the gait of the child with foot deformity. However, its availability is low in most treatment centers, rendering it unusable at the moment.

Static evaluation with the patient initially standing is useful for detecting the presence of the varus (support of the calcaneus in the soil is displaced medially in relation to the midline of the leg) and of the equine (in case of not supporting the calcaneus or doing knee hyperextension). In dynamic gait inspection, the position of the varus support of the calcaneus is verified if it occurs in conjunction with the action of the anterior tibial with dynamic supination of the forefoot. Internal progression of the feet also occurs due to the presence of forefoot adduction and to the inversion of the subtalar, which is the most important factor.<sup>17</sup>

Podography or even the assessment of dirt or of calluses can give us important clues about the place of foot support in the gait. In the functional examination, plantar flexion and maximum dorsiflexion should be verified both passively and actively. Active dorsiflexion may be limited by a sural triceps contracture and may also reveal the tendency to inversion of the feet, evidencing the insertion of the anterior tibial in a more plantar region.

Although anamnesis and clinical examination combined with patient expectation are paramount in the decision-making process, radiographic evaluation is an important ally in this planning. A complete understanding of the alignment of the bones and joints of the medial and lateral columns as well as of the relationships between the hindfoot bones is essential to assess the severity of the foot. The three-dimensional relationship and understanding of residual or acquired bone deformities may be key to deciding on the best treatment method.

This investigation is primarily performed by radiographs and can be complemented with computed tomography (CT) and magnetic resonance imaging (MRI).<sup>18</sup> The radiographic series must include the anteroposterior incidences of the bearing, the axial, and the calcaneus, the Saltzman incidence, and the ankle anteroposterior incidence. Magnetic resonance imaging may demonstrate alterations related to the involvement of soft tissues and cartilaginous or fibrous joints between the bones. Although it is very useful for the evaluation of cases with late or repeated recurrences, MRI has the limitation of requiring sedation in younger children; therefore, its use is limited. Recent studies consider that the use of CT with 3D reconstruction can determine and classify the congenital clubfoot severity, as well as assess the progress of the intervention based on the relative position of the tarsus bones.<sup>19</sup> The three-dimensional relationship and the understanding of residual or acquired bone deformities may be key to deciding on the best treatment method.

### **The Choice of Treatment Method: The Ponseti Limit**

Correction by the Ponseti method is based on promoting the eversion of the subtalar joint and the abduction of the calcaneus under the talus; the block of the middle foot and forefoot accompany the correction, until the talus is covered by the navicular. Tissue elasticity allows

correction and congruence between bones and is facilitated by the ability of cartilage to reshape as it changes position.

The choice of treatment of recurrent clubfoot depends on several factors, such as the previous treatment method and the stiffness of the deformity. The Ponseti method can also be used for resistant and recurrent CCF.<sup>3,5,11,20</sup> The option for this method at a later stage goes beyond the severity of the deformity but should also consider individual and family characteristics. The limit of the method depends on anatomical changes such as the presence of degenerative joint changes, bone deformities, and tarsal coalitions, as well as tolerance and desire of the patient to regarding plastered treatment and the subsequent use of the foot abduction orthosis. In older children, complete correction is no longer possible due to intrinsic bone deformations, but the goal remains to attain functional plant position and absence of pain.

Eidelman et al.<sup>21</sup> nosologically divided the recurrences into three groups:

- Early relapse (from 6 months to 30 months old)
- Late recurrence (between 30 months and 8 years old)
- Relapse in adolescents ( $\geq$  9 years old)

This classification would make it easier to find the main reason for recurrence and, thus, define the best treatment method.

Early recurrences occur mainly due to failure to adhere to the orthosis. The first sign that should be observed during follow-up consultations is the loss of dorsiflexion. The foot ends up slipping through the orthosis and not settling in the correct way, which can cause wounds or areas of hyperpressure.

In the group of late relapses, children aged between 2.5 and 8 years old stopped using the orthosis either by medical recommendation or due to loss of follow-up. Many, however, have muscle factors that predispose the return of deformities. Recurrences in this group range from mild dynamic supination of the forefoot during walking to the multiple components of the deformity of the left foot (cavus, adduction, varus, and equine). The group classified as recurrence of adolescents usually has rigid deformity, and, with the dissemination of the method, recurrences in this age group are currently less common.

In all relapses, the principles of Ponseti may be sufficient to restore foot correction.<sup>11</sup> The goal of returning to the plaster is to recover the angle of 60 to 70° of abduction of the foot that was achieved in the initial treatment and to obtain the maximum dorsiflexion. Again, it is imperative to institute a vigorous orthosis program to maintain the correction achieved. In patients with late recurrences > 2.5 years old who present a dynamic supination component of the forefoot, adjuvant treatment through transfer of the anterior tibial tendon to the third cuneiform should be evaluated. It is worth remembering that this procedure will only have the expected success if the deformities of the foot are better corrected.<sup>22</sup>

Most authors recommend that all patients with recurrences should return to treatment by the Ponseti method, since even if we do not get a full correction of deformities, we can improve the position and avoid very extensive surgeries.<sup>3,5,11,20,23</sup> In these cases, other methods should be considered: open release, osteotomies, or gradual distraction with external fixator.<sup>24</sup> In nonreducible deformities, bone procedures are often necessary to treat structural abnormalities.

In a study with a cohort of 33 patients with residual deformity and 55 patients with recurrent clubfoot, half of the patients underwent treatment by the Ponseti method; one quarter of them also required adaptation with the foot abduction orthosis protocol, and a quarter required surgery.<sup>20</sup> The Ponseti method was effective when recurrences or residual deformities were present before 5 years of age.

The eversion of the subtalar is the primary objective to be achieved, and this condition must necessarily be evaluated in the case of recurrences. The absence of adequate positioning of the navicular in relation to the talus in the radiographic evaluation, without proper correction of the Kite angle, indicates that the calcaneus was also not abducted under the talus, so the method is indicated to perform this correction.

Treatment of progressive loss of dorsiflexion can be challenging. Repeated percutaneous tenotomies can make tissue increasingly rigid and fibrotic. Open procedures of tendinous stretching, capsulotomy, and even anterior hemiepifyosis of the distal tibia<sup>25</sup> have been described and used.

In some patients, it is no longer possible to obtain the rotation of the subtalar, either due to the stiffness of this joint in older children or to the union between the talus and the calcaneus. In them, the incongruity between the bones does not allow peritalar release. Also, often, the alignment of the forefoot and middle foot is corrected, and the deformity is concentrated in the hindfoot. In such cases, many procedures can be used to improve alignment. The understanding of clinical and radiological deformities, through CT with triplanar reconstruction, allows an individualized evaluation, being essential for decision-making.<sup>18,19</sup>

Soft tissue release procedures can be associated with bone procedures to obtain a plantigrade and functional foot. Fasciotomy, associated with the releases of the plantar capsules of the medial spine joints, as well as the stretching of the achilles tendon and flexor of the fingers and allux, are the most common soft tissue release procedures. Among the bone procedures, the correction of the heel varus can be performed through the osteotomy of lateral wedge resection, as well as by insertion of a medial wedge (usually removal of the cuboid) to correct the adduction. This prevents shortening and improves the arm and lever of the sural triceps. The adduction can be improved with a combined medial opening wedge osteotomy (medial cuneiform) lateral closing wedge.<sup>26</sup> The cavus can be treated by osteotomy of dorsal wedge removal from the middle foot, while residual supination can be corrected by elevating



osteotomy of the first metatarsal, associated with stretching of the anterior tibial tendon or through its transfer to the lateral wedge.

In more severe cases, other procedures can be performed: naviclectomy combined with lateral calcaneus-cuboid shortening, as described by Mubarak et al.,<sup>27</sup> or talectomy associated with shortening and fusion of the lateral spine.<sup>28</sup> Modelling triple arthrodesis is reserved for patients > 12 years old with complaints of severe pain and deformities. The use of circular external fixators with or without additional osteotomies is also a method that can be used, allowing for a correction in multiple planes and gradual deformity.<sup>24</sup>

## Final Considerations

The initial treatment of congenital clubfoot has a very well-defined protocol. However, despite the good results of the Ponseti method, the prevention and treatment of relapses remains a challenge. Knowledge of pathogenesis, strict regular follow-up, and regular use of abduction orthotics are crucial for treatment success. Identification of factors with worse prognosis and early recognition of relapses make the treatment more effective.

In relapses, especially in the presence of rigid deformities, the association of open surgical procedures may be indicated. For the surgeon, it is essential to know the entire surgical arsenal, which should include soft tissue release procedures, stretching and tendon transfers, osteotomies, addition or resection of wedges, and modeling bone fusions. Thus, from the identification of the deformity and stiffness of the patient, they will be able to choose the best technique to achieve an aligned and stable foot.

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### Conflict of Interests

The authors have no conflict of interests to declare.

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