



# Diagnosis and Treatment of Idiopathic Congenital Clubfoot

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Idiopathic congenital clubfoot is the most common serious musculoskeletal birth defect in the United States and the world. The natural history of the deformity is to persist into adult life with a significant decrease in function and quality of life. The Ponseti method (serial casting, Achilles tenotomy, and bracing of the clubfoot) has become the most effective and accepted treatment of children born with clubfoot worldwide. The treatment is successful, particularly when the Ponseti-trained practitioner (often a pediatric orthopedic surgeon), the primary care clinician, and the family work together to facilitate success. An important factor in the ultimate success of the Ponseti method is parental understanding of the bracing phase. There is a very high rate of recurrent deformity when bracing is not done properly or is stopped prematurely. The importance of positive education and support for the parents to complete the entire treatment protocol cannot be overstated. The goal of treatment is a deformity-free, functional, comfortable foot. Ponseti clubfoot programs have been launched in most countries throughout the world, including many countries with limited resources. Ultimately, the goal is that every infant born with a clubfoot will have access to care with the Ponseti method. This clinical report is intended for medical practitioners who are involved in the care of pediatric patients with clubfoot. Understanding the standard of care will help these practitioners to care for patients and their families.

## abstract

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The guidance in this report does not indicate an exclusive course of treatment or serves as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

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## BACKGROUND

Congenital idiopathic clubfoot, also known as congenital talipes equinovarus, is the most common serious musculoskeletal birth defect that occurs in the United States and the world. Idiopathic clubfoot

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occurs in otherwise normal infants and should be distinguished from syndromic clubfoot and neurogenic clubfoot, which occur in infants as part of a syndrome or neurologic condition. Syndromic and neurogenic clubfoot are less common, are more difficult to treat, and have higher recurrence rates during and after treatment. Although syndromic and neurogenic clubfoot are important for the pediatrician to recognize, the focus of this report is on the more common idiopathic clubfoot.

The incidence of idiopathic clubfoot is approximately 1 to 2 per 1000 births, resulting in 150 000 to 200 000 infants with clubfoot born throughout the world every year. Approximately 80% of children with clubfoot are born in countries with limited resources,<sup>1-5</sup> and approximately 2100 infants born in the United States annually have clubfoot. There are varying rates of occurrence among different ethnic populations. Rates have been reported as high as 7 in 1000 in Hawaiian and Māori children, 2 in 1000 in Malawi children, 1 to 2 in 1000 in White children, and 0.5 to 1 in 1000 in Japanese and Filipino children. There is a 2:1 male predilection, and 30% to 50% are bilateral.<sup>6</sup>

Idiopathic clubfoot is typically an isolated congenital abnormality and is secondary to multifactorial environmental and strong genetic factors.<sup>1-3,7-11</sup> In support of a genetic cause, there is a concordance of 35% in affected identical twins.<sup>12</sup> If a father has a clubfoot or one child in a family is born with a clubfoot, the chance of a second child in that family being born with a clubfoot increases to 1/35.<sup>2</sup> Why males are affected with clubfoot about twice as often as females is unclear.<sup>2,10</sup> A polygenic threshold model in which environment and multiple genes

interact to produce the deformity has been proposed.<sup>10</sup> Clubfoot is not a deformity secondary to third trimester intrauterine crowding and should not be confused with the much more common benign positional foot and leg deformities that often resolve without treatment, including metatarsus adductus, calcaneovalgus, and positional cavovarus.<sup>2</sup> Early amniocentesis before 13 weeks' gestation and oligohydramnios at critical gestational periods are environmental factors that have been found to be associated with idiopathic clubfoot.<sup>13,14</sup>

### PRENATAL DIAGNOSIS

Clubfoot deformity may be discovered during prenatal ultrasonography. Clubfoot can be detected at 13 weeks' gestation using transvaginal ultrasonography<sup>15,16</sup> and at 16 weeks' gestation using transabdominal ultrasonography.<sup>17</sup> The visualization of a clubfoot deformity by prenatal ultrasonography requires a thorough examination to evaluate for additional findings indicating a syndromic or neurogenic clubfoot. Patients with syndromic or neurogenic clubfoot and, less often,

isolated clubfoot have higher rates of genetic abnormalities, which can be screened for with fetal genetic testing.<sup>18</sup> For example, fetal akinesia deformation sequence has associated anomalies that can be detected on ultrasonography, leading to more thorough genetic testing.<sup>19</sup> If syndromic clubfoot is noted, referral for fetal MRI has been suggested to reliably assess for associated findings.<sup>20</sup>

Most often the prenatal diagnosis of clubfoot will occur at the routine ultrasonography appointment at 20 weeks' gestation, unless the pregnancy is being closely followed for other reasons. At this time, the parents may ask the pediatrician for information or treatment advice. No prenatal treatment is available for clubfoot; however, appropriate prenatal counseling is important. It is very helpful for the parents to have their pediatrician or obstetrician recommend a clinician who is experienced with the Ponseti technique. The parents may be comforted by accurate and reliable resources that are available on the Internet, but they may also find confusing and inaccurate information. Pediatric orthopedic surgeons are frequently willing to meet with parents in the prenatal period to



**FIGURE 1** Ponseti treatment (photo credit: Shafique Pirani).

review imaging studies, assess severity, and describe treatment options. It is best for patients to be treated by a practitioner with training and experience in the Ponseti method (Fig 1). In areas with no pediatric orthopedic surgeons and no orthopedic surgeons with experience in treating clubfoot, it is suggested that patients travel for care.

A prenatal diagnosis cannot always be made, even by high-quality ultrasonography, particularly if the deformity is mild or moderate. Conversely, a mild isolated foot deformity noted on prenatal examination may not be a true clubfoot at birth; it may be a positional resolving deformity, as the false-positive rate of idiopathic clubfoot on prenatal ultrasonography is estimated at ~19%.<sup>21,22</sup> Important to note is that the severity of clubfoot cannot be determined by ultrasonographic examination. The severity can only be determined by clinical examination of the foot.

### NEWBORN EVALUATION

The evaluation of a newborn with a clubfoot deformity involves a thorough general examination to determine overall health and development, exclude syndromes and neurologic conditions (eg, spina bifida, arthrogyriposis, limb formation anomalies), and provide a focused examination of the foot and limb. Examination of all joints for range of motion and stability, including the hips, is important, as is examination of lower extremities for equal length and symmetry. The severity of a newborn foot deformity is determined more by the foot's flexibility than by its appearance.<sup>23,24</sup> Newborn foot deformities that can be easily manipulated into an overcorrected position are considered positional rather than true clubfoot deformities. These resolve with

minimal or no treatment. Unless a limb deficiency such as fibular hemimelia, tibial hemimelia, or congenital short femur is noted, radiography is not necessary.<sup>25</sup>

Several syndromes are known to be associated with clubfoot, including classic arthrogyriposis, multiple pterygium syndrome, distal arthrogyriposis, amniotic band syndrome, and Freeman-Sheldon syndrome.<sup>26–28</sup> Geneticists can help to evaluate patients when there is suspicion of a syndrome. Neurologic causes include myelomeningocele, lipomeningocele, tethered cord syndrome, diastematomyelia, and sacral agenesis. A careful examination of the spine is needed to detect the sometimes subtle findings associated with a tethered cord, such as a sacral dimple or hair patch.

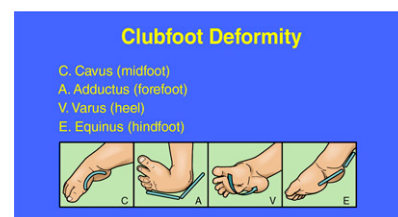
An infant with an isolated idiopathic clubfoot has a completely normal physical examination except for the involved foot and leg. The deformity has 4 key components that can be remembered by the acronym CAVE: The midfoot has a high arch (cavus), the forefoot is turned in (adductus), the heel is turned in (varus), and the hind foot is pointed down (equinus) (Fig 2). The deformity is quite stiff (not easily correctable with gentle manipulation), and the foot and calf may be slightly smaller than the opposite normal side. The untreated or relapsed clubfoot deformity results in the child walking on the lateral side or dorsum of the foot (Fig 3). Older children and adults with untreated clubfoot are unable to wear standard shoes and are limited in sports and employment.

### TREATMENT

Until the Ponseti technique became accepted as the superior method for the treatment of clubfoot, many treatments with initial good results became popular for a time, only to fall out of favor when the long-term

results showed a high rate of recurrent deformity, stiffness, and pain. The first recorded treatment of clubfoot is attributed to Hippocrates, who recommended “manipulation of the deformed foot and bandaging in the corrected position.”<sup>29</sup> Subsequently, other various nonoperative treatments involving casting, special shoes, braces, and even correction with a “wrench”<sup>30,31</sup> were proposed. Immediate results were sometimes satisfactory; however, in time, the deformity would almost always recur.

During the first half of the 20th century, Hiram Kite became an enthusiastic proponent of the nonoperative treatment of clubfoot.<sup>32,33</sup> Although his initial success was reported at 90%, the technique was lengthy, the results were not reproducible, and the deformity often recurred.<sup>34</sup> As a result of dissatisfaction with Kite's method of casting, surgical releases for resistant clubfoot, which dramatically corrected the deformity in the short term, became popular in the last quarter of the 20th century.<sup>1,35–43</sup> These extensive surgeries were performed at 4 to 18 months of age. The short- and medium-term results were good.<sup>40,41,43</sup> However, with growth, scarring from the surgery eventually led to decreased motion; the foot became painful, stiff, and arthritic; and activities and function became limited as the child reached adolescence and adulthood.<sup>44–48</sup>



**FIGURE 2** The 4 key features of a clubfoot deformity: cavus, adductus, varus, and equinus (CAVE).



**FIGURE 3** A 10-year-old boy with bilateral clubfoot deformity. As an infant, he underwent standard left foot surgical release, which later recurred and became as severe as the untreated right foot deformity.

In 1948, Ignacio Ponseti at the University of Iowa developed a method of treatment that used a new casting technique and minimal surgery.<sup>49</sup> Ponseti's methodology was clearly explained, easily understood, and reproducible. One of the distinctions of his method was the prolonged bracing phase, which lessened but did not eliminate recurrences. During the last quarter of the 20th century, while most centers had adopted extensive surgical releases for resistant clubfoot, Ponseti continued to perfect his method. The Ponseti method is based on a clear understanding of the pathoanatomy of clubfoot and how it affects foot mechanics.<sup>8,9</sup> Over the years, refinements of the Ponseti technique and improvements in bracing have decreased the problem of recurrence, and the method has now become the standard for the treatment of clubfoot.<sup>50,51</sup> Of note, although this report focuses on isolated idiopathic congenital clubfoot, syndromic and neurogenic clubfoot can also be treated with the Ponseti method, although the treatment is prolonged, with higher relapse rates.<sup>52</sup>

A second nonsurgical method of clubfoot treatment was developed in France and popularized and refined by Alain Dimeglio.<sup>53</sup> The technique, known as the French functional

method, also emphasizes gradual correction in infancy, with daily manipulations by an experienced physical therapist and elastic taping between therapy sessions. The method also includes a prolonged period of night splinting and limited surgery for recurrences. The time to correction with this method is several months; treatment is daily and much more labor intensive for both the family and the practitioner. Although the method has a few followers in Europe, unlike the Ponseti method, the long-term results of the technique are not known.

### THE PONSETI TREATMENT METHOD

Careful and complete use of the Ponseti method leads to optimal results. The method consists of 3 phases of treatment: manipulation and casting, tenotomy, and bracing.<sup>49,53-55</sup> The first phase involves manipulation and weekly serial above-knee casting performed by an individual trained in the technique. This phase typically lasts for 5 to 8 casts.<sup>56,57</sup> It is not necessary to initiate treatment in the newborn nursery. Treatment can start anytime in the neonatal period, ideally the first 1 to 3 weeks.<sup>58</sup>

In the second phase, after all of the elements of the deformity except equinus have been corrected, a percutaneous Achilles tenotomy is performed under local anesthesia by a surgeon in the clinic, which eliminates the perioperative risk of general anesthesia and concerns about anesthetic neurotoxicity in the infant.<sup>59</sup> Some surgeons prefer to perform the tenotomy in the operating room under general anesthesia, particularly in older children. A final cast is placed immediately after the tenotomy and worn for 3 weeks. Ninety percent of infants with clubfoot require a tenotomy.<sup>60</sup> A more extensive surgical release is not necessary.<sup>61</sup>

The third and most important phase is the bracing phase, which starts immediately after removal of the posttenotomy cast. The brace is a foot abduction orthosis that consists of 2 shoes or splints connected by a bar, which holds the feet shoulder width apart. During the first 3 months, the brace is worn 23 hours per day, allowing the brace to come off for dressing and bathing only. After the first 3 months, the brace is worn at nighttime and nap time only, with a goal of 12 to 14 hours of brace wear, until the child is 4 to 5 years old.

The principle of bracing is that the corrected foot is maintained in an abducted and dorsiflexed position to prevent relapses. A well-designed brace is easily applied and removed, does not allow the foot to slip or escape, and has well-tolerated shoes (splints). In bilateral clubfoot, the shoes (splints) are externally rotated 60° to 70° on the bar. If the deformity is unilateral, the shoe (splint) on the affected side should be externally rotated 60° to 70° and the unaffected side 30° to 40°. The bar should be of the length between the child's shoulders and should be set with the ankles in 10° to 15° of dorsiflexion. Improvements in brace design using shoes or splints with soft linings that can be easily detached from the bar instead of stiff shoes that are rigidly fixed to the bar has made brace wear much easier for patients and families.

Adherence by the parents with the bracing phase of treatment has been shown to be the most important factor in the prevention of recurrent deformity and the ultimate success of the Ponseti method.<sup>57,62-69</sup> It is not easy to keep a child in a brace all night, every night, for 4 years; thus, the ability of the pediatrician and orthopedic surgeon to motivate and support the parents to complete the bracing program is as important as the initial correction of the



deformity. The pediatrician's knowledge of the standard Ponseti protocol and encouragement to the family to comply and complete the bracing phase of treatment are important to successful treatment. Recurrence of clubfoot deformity after treatment with the Ponseti method using the traditional foot abduction orthosis ranges from 8% to 56%.<sup>67,70-73</sup> Recurrences occur in only 6% of families who are compliant in completing the bracing phase but in up to 80% of families who are unable to complete the bracing phase.<sup>74</sup>

Adherence with bracing has many factors but is not believed to be related to family education, culture, or income level.<sup>75</sup> Adherence can be improved with culturally sensitive education, a positive communication approach, and greater family awareness of the importance of bracing.<sup>76</sup> Communication between the practitioners and family is very important to the family's understanding of the treatment process and adherence to bracing.<sup>68,77,78</sup> Both the pediatrician and the treating pediatric orthopedic surgeon should explore barriers to adherence and empower the families to succeed. Many treatment centers now use a clinic coordinator who instructs the family on proper brace wear, communicates through follow-up phone calls, assists with the financial challenges of bracing, and provides support throughout the phases of treatment.<sup>79</sup>

### TREATMENT OF RECURRENCE

Recurrence is defined as the reappearance of any of the components of the deformity.<sup>80</sup> The first sign of recurrence is often the development of equinus, or a tight heel cord. An intoeing gait develops with increased adduction of the forefoot, varus of the heel, and loss of heel strike. When the child has

developed an intoeing or adducted gait, the recurrence is quite advanced. Left untreated, the dynamic, flexible deformity gradually becomes more fixed. Recurrences are well treated by repeating the Ponseti method.<sup>51</sup> Despite repeating the Ponseti method, approximately 5% to 20% of appropriately treated patients with idiopathic clubfoot develop additional recurrences that manifest as a persistent intoeing gait with a flexible adducted and supinated forefoot. The deformity can be corrected with surgery limited to a tibialis anterior tendon transfer.<sup>49,81-84</sup> This is considered an anticipated part of the Ponseti method and should not be viewed by the family as a failure of treatment. Failure of the Ponseti method with residual fixed deformity that requires surgical release of the ankle or subtalar joint is rare.

### OUTCOME AFTER TREATMENT

Dr Ponseti first published his results in 1963. Of the 93 feet treated, 71% had a good result, 28% had a slight residual deformity, and only 1 had a poor result.<sup>49</sup> These results were excellent and unlike the results of surgical release, did not deteriorate with long-term follow-up. The long-term results of the Ponseti method, as practiced at the University of Iowa, were published in 1980<sup>53</sup> and then again in 1996.<sup>54</sup> After more than 30 years following treatment with the Ponseti method, adults who had been born with a clubfoot functioned nearly as well as a control group of adults born with normal feet. Children who undergo Ponseti treatment and are successful with bracing can be expected to participate in sports, wear normal shoes, and have excellent overall function.

The successful results of the Ponseti method have been duplicated at many other treatment centers in the United States, with initial correction rates reported to be 90% to 100%.<sup>63-66,85-87</sup> A survey of the

Pediatric Orthopedic Society of North America revealed that 97% of members use the Ponseti method as their preferred treatment of idiopathic clubfoot.<sup>5</sup> As a result, the rate of radical surgical releases performed for idiopathic clubfoot in the United States has decreased from 70% in 1996 to 5% to 10% in 2006. Parent satisfaction at 2 years is more than 90% in numerous studies.<sup>88</sup> Through computer gait analysis of children treated by the Ponseti method compared with nonaffected children, researchers have shown good clinical results with high function.<sup>89</sup> Children treated by the Ponseti method had greater ankle power than children treated surgically; however, children who were treated for clubfoot had a small decrease in ankle power compared with normal feet.<sup>90,91</sup> Residual intoeing was sometimes noted.

### CHALLENGES AND COMPLICATIONS

Ponseti casting is safe but can have complications even when performed by experienced practitioners.<sup>87</sup> The complications include skin irritation and breakdown from the casting. Complications from the tenotomy are rare but can include bleeding, infection, pseudoaneurysm, and damage to the neurovascular bundle.<sup>87</sup> Minimal delays are seen in gross motor milestone achievement in children treated with the Ponseti method.<sup>92</sup>

The most prevalent challenge to successful treatment is adherence with the entire treatment protocol. Patients and families can experience distress with bracing, hardship related to cost of the braces, and the distance necessary to travel for treatment. It cannot be overstated that nonadherence with brace wear will result in a recurrence 100% of the time in the first year, 80% of the time in the second year, 60% of the time in the third year, and 30% of the time in the fourth year.

## CLUBFOOT IN COUNTRIES WITH LIMITED RESOURCES

Although seldom witnessed in the United States and other industrialized countries, children born with clubfoot in countries with limited resources reach adulthood unable to wear shoes and walk on the tops of their feet with their toes pointed backward. Cultural bias and prejudices often relegate these children to a life of poverty, lack of education, and difficulty finding a marriage partner. To these individuals, clubfoot correction is life changing. Eighty percent of the nearly 200 000 infants born with clubfoot worldwide every year are from countries with limited resources.<sup>4</sup>

The Ponseti method is particularly well suited for use in resource-limited countries because the casting and brace supervision can be performed by physician extenders instead of physicians. Results of treatment by physical therapists trained in the Ponseti method are as good, if not better, than the results obtained by physicians.<sup>56,57</sup> In countries with limited resources, effective and economical braces can be made using local systems. Ponseti programs have been started in many developing countries, including Uganda, Nepal, India, and Ecuador, with the ultimate goal of providing access to treatment with the Ponseti method for every child in the world born with a clubfoot.<sup>4,93-97</sup>

However, many challenges remain, including sustainable funding, access to care, long travel distances, training of staff, performance of a safe tenotomy, and access to affordable, quality braces.<sup>98</sup> A care delivery value chain recommends the following 6 steps for optimal program success: diagnosing early, developing high-volume Ponseti

centers, training nonphysician health workers, engaging families in care, addressing barriers to access, and providing follow-up in the patient's community.<sup>99</sup>

## CONCLUSIONS AND FUTURE DIRECTIONS

The treatment of idiopathic congenital clubfoot with the Ponseti method is now accepted as superior to other methods in the United States and the world. The Ponseti method is effective but needs to be followed closely by trained practitioners and surgeons. The importance of parents adhering to the entire bracing program cannot be overstated. A positive communication style between the parents and the pediatrician can improve brace adherence and treatment success. Ponseti treatment programs are now available in many countries with limited resources. In the future, the goal of every child in the world born with a clubfoot having access to care with the Ponseti method will become a reality.

## WEB RESOURCES

### Global Help

Global Help is an organization dedicated to making medical and health publications available and accessible at no cost. This site contains links to copies of *Clubfoot: Ponseti Management*, an authoritative and easy-to-follow guide for health care professionals. The publication is available in 30 languages. Available at [http://global-help.org/products/clubfoot\\_ponseti\\_management](http://global-help.org/products/clubfoot_ponseti_management).

### Ponseti International

Ponseti International is an organization based at the University of Iowa, where Dr Ponseti developed his method. The mission

is to provide global leadership in building high-quality, locally directed, and sustainable capacity to deliver the Ponseti clubfoot care pathway at the country level. This site also has a downloadable version of Dr Ponseti's book *Congenital Clubfoot: Fundamentals of Treatment*. Available at <http://www.ponseti.info>.

## Global Clubfoot Initiative

Global Clubfoot Initiative is a nongovernmental organization that supports and provides resources and links for those who are providing clubfoot treatment in low- and middle-income countries. Available at <http://globalclubfoot.org>.

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