Congenital Idiopathic Clubfoot: Prevention of Late Deformity and Disability by Conservative Treatment With the Ponseti Technique

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Clubfoot is a complex deformity in which the foot is completely turned inward at birth (Figure 1, 129). Clubfoot can occur in an otherwise normal child (idiopathic) or as a part of disorders such as myelomeningocele, or arthrogryposis. Idiopathic clubfoot occurs worldwide with an incidence varying from 0.39 to 8 per 1000 live births.1,2 It is the seventh most common congenital birth defect and the first for the musculoskeletal system.1 Boys are more commonly affected than girls (2:1 ratio), and up to 50% of the cases are bilateral, affecting both feet.3,4

Despite extensive clinical, epidemiological, and basic science research, the etiology and pathogenesis of clubfoot remains unknown.1-6 However, clubfoot clusters in families and affects family members across generations, suggesting that genetics may play a role in the causation of this deformity. A positive family history has been reported in up to one-third of patients, and twin studies demonstrate 32.5% concordance for monozygotic twins versus 2.9% for dizygotic twins.1 The occurrence rate is 17 times higher for first-degree relatives than in the general population. In addition, unaffected parents with an affected son have a 1 in 40 chance that another son will clubfoot. If the affected child is a daughter, the chance is 1 in 16 for a son, and 1 in 40 for a daughter. The chances are 1 in 4 if both a parent and a sibling are affected.1,3

Improvements in obstetric ultrasonography have led to an increase in the prenatal diagnosis of clubfoot. However, the false-positive rate is relatively high (40%), and more than 50% of patients are later found to have additional abnormalities.3,7 Prenatal diagnosis has led to an increase in parent consultations for counseling.

An understanding of clubfoot pathology, genetic risk, and methods of treatment and their long-term results is essential to better care for these patients.

PATHOANATOMY

Clubfoot is a deformity of the limb characterized by smaller calf muscles and adductus of the forefoot, cavus (increased longitudinal arch), varus of the heel (the heel is turned in), and equinus.
of the foot (the foot is in plantar flexion). Although the most severe deformity occurs in the hindfoot, all components of the deformity are interrelated. Clubfoot is present at birth (i.e., congenital), but it is not a malformation. A normally developing foot becomes a clubfoot during the second trimester of pregnancy.

Clubfoot rarely is detected by ultrasonography by the 14th to 16th week of gestation. The gene (or genes) responsible for clubfoot are active starting from the 12th to 14th week of fetal life until ages 4 to 6 years. After this age, relapses are very rare, although the calf atrophy will persist for the life of the patient (Figure 2, see page 130).

Pathologically, the ligaments of the posterior aspect of the ankle and of the medial and plantar aspects of the foot are shortened and thickened. The muscles and tendons of the gastrocnemius, tibialis posterior, and toe flexors are shortened and are smaller in size. There is an inverse correlation between muscle size and severity of the deformity. In addition, there is an increase in connective tissue rich in collagen that tends to spread into the Achilles tendon and the deep fascia.

Many have hypothesized that clubfoot is due to malignment of the bones and by deformation in their shape, specifically the talus. However, Pirani et al. demonstrated by magnetic resonance imaging (MRI) that the shape and relationship of the bones dramatically changes during treatment, resulting in a normal-looking foot. In addition, long-term radiographic studies demonstrate that the bones of the foot are very close to normal in many cases. These studies suggest that bony abnormalities may be secondary to the soft tissue abnormalities discussed above.
Differential Diagnosis

Congenital clubfoot must be differentiated from a postural equino-varus deformity, which is a deformation secondary to intrauterine molding. The later can be associated with some adductus of the forefoot, but it is flexible and it can be easily corrected. Importantly, there is no Achilles tendon contracture. Observation is indicated in such cases because they will resolve without treatment. However, in very premature babies (24 to 30 weeks gestational age), this type of deformity can represent an early stage of a true clubfoot. Therefore, these babies should be followed to make sure they do not develop any rigidity.

Clubfoot must be also differentiated from metatarsus adductus. Metatarsus adductus is a common neonatal foot problem in which the forefoot is adducted, and the lateral border of the foot is convex (Figure 2). This deformity differs from clubfoot in that the heel is not in equinus and varus. Importantly, the natural history of metatarsus adductus tends to be one of spontaneous improvement without treatment in the majority of cases.

To identify those patients who may need treatment, the flexibility of the foot is assessed. If the deformity is passively correctable, treatment is rarely needed. If the deformity is rigid, casting until the foot is fully correct and is indicated (usually 2 to 3 cast applications changed bi-weekly). The best timing for treatment is in the first 6 to 8 months of life, but improvement with casting can be seen until 2 years of age. Older patients with shoe problems or pain can be considered for surgical correction, although this is rarely indicated. Patients with mild to moderate deformity generally are asymptomatic at long-term follow-up.

Treatment of Clubfoot and Long-Term Results

We do not know the etiology of idiopathic clubfoot; therefore, we cannot influence the pathology in the ligaments, tendons, and muscles that seem to determine the deformity and its degree of resistance to correction. We currently can treat the results of clubfoot. The goal of treatment is to correct all components of the deformity so that the patient has a pain-free foot with good mobility, without calluses, and without the need to wear modified shoes and inserts. Most orthopedists agree that the initial treatment should be nonoperative; the preferred methods are manipulation and application of a plaster cast or physiotherapy started soon after birth.

The success of manipulation and serial casting depends on the severity and stiffness of the deformity, age of the patient, and the skill in performing appropriate manipulations and in applying the plaster casts. Accurate assessment of the severity of the deformity at birth is commonly believed to be important, and several classification systems have been developed. However, it is difficult to predict how a particular foot will respond to treatment. In some clubfeet, apparently tight ligaments seem to become easily stretchable with manipulation, and the alignment of the bones of the foot improves rapidly after application of a few casts. In other clubfeet, with the use of the same technique, correction is more difficult. Therefore, all clubfeet should be initially treated by a period of manipulation and casting regardless of the initial severity of the deformity.

During the past 40 years, the technique of manipulation and casting performed in most institutions followed the principles described by Kite. However, consistent reproducibility of Kite's reported success has not been observed, and the results have been very discouraging in most institutions, both in the United States and abroad. Reported results demonstrate incomplete or defective corrections and distressing outcomes in up to 95% of the cases. These outcomes included rocker-bottom deformity, metaphyseal fractures, flattening of the talus, lateral rotation of the ankle, and increased stiffness of the joints. To improve on these results, many other manipulative techniques have been proposed, but the majority of clubfeet in reported series ended up in surgical...
The only long-term follow-up study on the effects of extensive surgical releases for clubfoot (mean follow-up length 30 years) has been reported by Dobbs et al. The results demonstrate that 29 of the 45 patients (involving 73 individual feet) underwent more than one procedure, with an average of 3.3 (standard deviation: 1.5) per foot. Thirty of the 73 feet had a relapse, and 20 of these required a second extensive soft-tissue release operation. Seventeen feet required bony procedures for over-correction. Five feet had already developed a joint fusion because of painful arthritis. Clinically, there were no excellent results; 22 feet (30%) had a good result, 14 (19%) a fair result, and 37 (51%) a poor result.

Interestingly, the Medical Outcome Short-Form Health Survey (SF-36) physical scores (General Health Survey questionnaire) of these patients (score: 33.7) was very similar to that reported for several major medical conditions, such as Parkinson’s (score: 30.1), dialysis for kidney failure (score: 31.6), chronic heart failure (score: 32.8), or preoperatively to coronary artery bypass (score: 32.5). Importantly, these clubfoot patients were only between ages 25 to 32, and their physical impairment may continue to deteriorate over time. Although more studies are needed to fully understand the long-term implications of extensive soft-tissue releases in clubfoot, the Dobbs et al. report suggests that surgical treatment of clubfeet may lead to significant adult physical disability. Therefore, any treatment that prevents surgery would have a great effect on the overall outcome of patients with clubfoot.

**CHANGES IN THE NATIONAL STANDARDS OF CARE FOR CLUBFOOT**

The results of the treatment for clubfoot at the University of Iowa stand in contrast to those described above. We practice manipulation and serial casting.
as first described by Dr. Ignacio V. Ponseti in 1963. This method is based on a very specific manipulation and casting technique based on an understanding of the functional anatomy of the foot, and supported by a limited intervention (percutaneous Achilles tenotomy performed under local anesthesia in the office) and a foot-abduction brace to prevent relapses. Complete clubfoot correction can be achieved in more than 95% of the patients, in as soon as 16 days (Figure 3, see page 132). Patients do very well during childhood and adolescence, and long-term follow-up (average 34 years) has demonstrated satisfactory clinical and functional results in the majority of the patients. With the use of pain and functional limitation as the outcome criteria, 78% of the treated patients had an excellent or good result compared with 85% in a control group from the general population without clubfoot (no statistically significant difference).

The discrepancy in the results of manipulation and serial casting suggests that the attempts at correction by the Kite technique, or any of its modifications, have been inadequate. When the Ponseti method is properly performed, surgical release is indicated in less than 1% of patients, mainly those with a short, rigid foot, or those with a very severe deformity that does not respond to proper manipulations.

World Initiatives for Treatment of Clubfoot by the Ponseti Technique

As a noninvasive treatment for congenital idiopathic clubfoot, the Ponseti method has been proven to be a superior method of correcting clubfoot deformity and avoiding major surgical interventions. The results of this method have encouraged national efforts to make this method the gold standard in the treatment of congenital idiopathic clubfoot. The Ponseti International Foundation for the Advancement of Clubfoot Treatment has initiated a program for the diffusion of the method worldwide. Under the sponsorship of The Bone and Joint Decade 2000-2010, an initiative aimed at improving quality of life for people with musculoskeletal disorders, this project is designed to ensure that every child born with clubfoot worldwide is identified at birth, and then provided with effective corrective treatment by the Ponseti method. The objective is the establishment of comprehensive programs to foster partnership among government, education, and community-based institutions to implement the Ponseti method at a national level in each country.

Experience with the Ponseti method in the US and worldwide has been most encouraging, and the program has been started successfully in more than 40 countries. In industrialized countries, through visits to our center, courses on the Ponseti method, and national and international meetings, many orthopedists have had the opportunity to be trained in the method. Many have become experts on the subject and have developed courses in their own countries.

However, the major driving force behind changes in the standards of care for children with clubfoot has come from the parents and their use of the Internet (Sidebar). Given that the results of surgical treatment can be so discouraging, parents have been looking for alternative treatments. Traditionally, parents would have relied on health care professionals for advice and treatment. However, the development of the Internet and the explosion of health care information have allowed them to obtain disease-specific information in a timely manner.

From information available through the Internet, parents became aware of the Ponseti method, as well as of the controversy that existed over whether extensive surgical treatment for the correction of the deformity is needed. Clubfoot is a very "visual" deformity — ie, it is very easy for the parents to see and assess the results of any treatment. The fact that the Ponseti method allows full correction of the deformity in a very short period of time and without the need for extensive surgery is of critical importance. Parents have created and actively used the Internet support groups on clubfoot to...
grams for the treatment of clubfoot has been initiated (eg, England, Sweden, Norway, Portugal, Brazil, and Chile),
with training of health professionals, designation of treatment centers and patient referral patterns, and developing registries and multicenter studies. The success in these countries has been excellent, with early results demonstrating more than 95% of children successfully treated without the need for extensive corrective surgery (private communication, 2005).

The major challenge for the implementation of clubfoot programs is in developing countries. This is of utmost importance because 80% of the patients born with clubfoot are in developing countries, where healthcare resources are very limited. As a result, many cases are neglected. If left untreated, clubfoot persists as a rigid, unsightly deformity. In the most extreme cases, the toes point backward during ambulation (Figure 4). Special footwear is required to accommodate the deformity, and every step is unnatural and excruciating. Children have difficulty playing with their peers and going to school. They are frequently subject to ridicule that can be associated with long-term psychological problems. Adults with untreated clubfoot may experience pain and disability, and many have a great deal of difficulty finding jobs that can accommodate their limitations. They are frequently relegated to welfare or even homeless lives. Globally, neglected clubfoot is the most common cause of physical disability among musculoskeletal birth defects.

Paradoxically, treatment of neglected cases is very complicated and expensive and has a significant rate of complications. The type of procedures needed to obtain a plantigrade foot range from extensive soft-tissue releases, to complex corrections using different types of external fixators, to corrective osteotomies and triple arthrodesis. Furthermore, in many developing countries, there are a limited
number of orthopedic surgeons. For example, in 2000, Malawi, with a population approaching 12 million people, had just three orthopedic surgeons and 12 physiotherapists. As a result, most of their efforts were directed toward coping with large numbers of trauma cases and infections. Therefore, a treatment method that is safe, quick, effective, economical, and easy to teach can have an enormous effect on the lives of many thousands of children born with clubfoot.

In 1999, Norgrove Penny and Shafique Pirani developed the Uganda Clubfoot Project. Using a public health approach, a collaboration among the study group, the Disability Section of the Ministry of Health, and the Department of Orthopaedic Surgery at Makerere University was developed. A national strategic plan was implemented with a community and public awareness program that used posters. Training of healthcare professionals was undertaken, including orthopedic and primary medical officers who are not physicians. An inexpensive, easy-to-make foot abduction brace also was developed. In a pilot study from November 1999 to October 2002, 155 patients were treated. They observed 97% corrections without the need for extensive surgical releases. Because of social, economic, and cultural issues, 25% of children did not complete the corrective phase of treatment.

This study is very important because it has demonstrated that this method of treatment is suitable for developing countries. Because of its success, it has been exported to many other African countries, where very good results have been observed. The experience in other countries (eg, Brazil, Chile, Argentina, Nepal) has demonstrated that neglected clubfoot can be corrected with the Ponseti method up to early adolescence, providing a suitable treatment for most clubfoot cases worldwide (Figure 4).

**SUMMARY**

The Ponseti method has become the gold standard for the treatment of clubfoot. It is very safe, efficient, economical, and easy to teach, and it radically decreases the need for extensive corrective surgeries. Awareness of the excellent results provided by the Ponseti method is essential for counseling and providing treatment advice to the families of children born with this deformity.

**REFERENCES**


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