Congenital Idiopathic Talipes Equinovarus
Benjamin D. Roye, Joshua Hyman and David P. Roye, Jr
Pediatrics in Review 2004;25;124
DOI: 10.1542/pir.25-4-124

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://pedsinreview.aappublications.org/content/25/4/124

Data Supplement at:
http://pedsinreview.aappublications.org/content/suppl/2005/01/26/25.4.124.DC1.html
Objectives  After completing this article, readers should be able to:

1. Describe congenital idiopathic talipes equinovarus (CTEV).
2. Differentiate CTEV from metatarsus adductus clinically.
3. Discuss how to rule out comorbid conditions such as torticollis and hip dysplasia when considering CTEV.
4. Describe the initial treatment of CTEV.
5. Discuss the genetic pattern of CTEV.

Historical Perspective
CTEV, commonly known as clubfoot, has been a recognized deformity since the time of the ancient Egyptians and was described independently by Hippocrates and the Aztecs. As implied by its name, the underlying deformity consists of a hind foot in equinus (plantarflexed) and varus (inverted). Additionally, there often is a cavus (abnormally high arch) and adductus component to the midfoot (Figs. 1 to 3). Essentially, the foot appears as though it was rotated inward and, in severe cases, as though it is on backwards. Historically, initial treatments were variations on the theme of manipulation (sometimes forceful and violent) and splinting. Surgical intervention began in the late 18th century with Lorenz’s Achilles tenotomy, but effective soft-tissue releases, osteotomies, and tendon releases did not evolve until the late 19th century with the advent of anesthesia and aseptic technique. Hiram Kite popularized gentle manipulation and serial casting in 1930, a technique championed and refined by Ignacio Ponseti in the 1950s. This remains the initial treatment of choice today. Although the basic concepts remain the same, surgical and nonsurgical techniques for the treatment of resistant CTEV continue to evolve, promising improved outcomes for children born with this condition. This article addresses primarily the more common idiopathic form of this condition, and unless otherwise stated, CTEV refers to idiopathic CTEV.

Epidemiology
The incidence of CTEV varies widely with race and geography. In Japan, the disease affects approximately 0.5:1,000 live births; in Caucasians, the incidence is 1.2:1,000 live births; and in natives of the South Pacific, the incidence is nearly 7:1,000 live births. All populations show a consistent 2:1 male predominance, with bilateral disease affecting approximately 50%.

Pathogenesis
Although the anatomic deformity of CTEV has been known for some time, its cause remains unclear, and innumerable theories have attempted to explain its origins. Prior to the 1980s, many physicians believed the theory introduced by Hippocrates that intrauterine compression created the fixed equinovarus deformity. This argument has been countered systematically and has fallen from favor. For example, CTEV has been documented in a 16-week-old fetus that had adequate room for its feet. Many other theories encompass environmental and genetic factors.

Several studies demonstrate abnormal muscle development in CTEV, including pre-
dominance and grouping of type 1 muscle fibers. Whether this is due to genetic factors, neurotrophic factors, or denervation is unclear. Several authors have examined the histologic and electron microscopic properties of soft tissues in the CTEV foot, finding predominance of type 1 muscle fibers in the lower legs of CTEV feet and a fiber type 2B deficiency with abnormal fiber grouping. Although there is no direct evidence of abnormal innervation, fiber grouping is known to be a consequence of denervation and reinnervation, which may play a role in the pathogenesis of CTEV. Other findings consistent with a neurogenic cause include changes in the sarcoplasmic reticulum and mitochondria and loss of myofibrils.

Recent evidence suggests that localized soft-tissue contracture may be involved. One researcher found that 100% of deltoid ligaments resected from virgin clubfeet stained positive for vimentin, a marker for myofibroblasts that are involved in wound and scar contracture. Only 8% of control specimens stained positive for vimentin.

Most clinicians agree that there must be some genetic component to CTEV, based largely on the observation that nearly one of every four cases of CTEV has a positive family history. Recent evidence from the large Canadian Early and Mid-Trimester Amniocentesis Trial Group associates CTEV with early amniocentesis (11th to 12th gestational weeks) compared with mid-trimester amniocentesis (15th to 16th weeks). Other suggestions for which there is limited evidence include the presence of a talocalcaneal bar as well as chromosomal, viral, and vascular causes. Constricting annular bands also have been implicated. Although remaining elusive, the true cause of CTEV seems most consistent with multifactorial inheritance.

Neuromuscular disease also can lead to an equinovarus deformity. As with other pediatric conditions that have neurogenic causes, such as scoliosis, the neurogenic CTEV is more complex and difficult to treat than its idiopathic counterpart. Conditions known to be associated with an equinovarus deformity include arthrogryposis, spinal muscular atrophy, spina bifida, sacral agenesis, and other paralytic states.

**Clinical Aspects**

**Symptoms**

The infant who has CTEV has no symptoms other than the painless equinovarus deformity. The patient who has uncorrected CTEV walks on the dorsolateral aspect of the affected foot and develops a callus, hyperpigmentation, and a subcutaneous bursa. Surprisingly, some arti-

Figure 1. Clinical photograph of an 8-month-old boy who has bilateral CTEV.

Figure 2. Clinical photograph of a clubfoot from the medial aspect. Note the prominent crease in the midfoot.

Figure 3. Clinical photograph of a clubfoot with the child supine. The midfoot flexion crease is more notable on the left foot.
cles on the natural history of CTEV cite little pain in the affected foot.

**Signs**
The physical findings of CTEV have been well described. The affected foot is in equinus and varus. There is a crease of varying depth over the medial midfoot wherein the foot appears to be folded on itself (Figs. 2 and 3). The deformity may be supple, and the foot may be brought to a normal position with gentle manipulation. More frequently, there is some degree of rigidity that may be severe. The calf on the affected side is smaller than that on the normal side, a difference that persists even after correction of the deformity.

**Tests**
Radiographs are ordered routinely at diagnosis despite their limited utility because of the eccentrically located ossification centers in the mostly cartilaginous, hence radiolucent, tarsal bones. Radiographs of the foot should simulate weight bearing (Fig. 4). The angle between the long axis of the talus and calcaneus (Kite angle) is measured on the lateral and anteroposterior views. This can be challenging because the ossific nuclei in these bones are nearly circular in young children, and the direction of the long axis is not obvious (Fig. 5). In feet affected with CTEV, these lines are more parallel when compared with normal feet.

Although CTEV is a clinical diagnosis, radiographs can be used for confirmation and to help rule out associated abnormalities. However, no data support their value preoperatively as a prognostic indicator, and there is wide variability in reported norms. As noted in the literature, postoperative measurements of radiologic indices have an inconsistent relationship to functional outcome, and several studies have questioned the association between radiographic and functional outcomes.

The role of computed tomography (CT) or magnetic resonance imaging (MRI) in other than an investigative setting has yet to be determined.

Additional studies, including ultrasonography, gait analysis, pedobarographic analysis, and electrogoniometric analysis, may be obtained by the referring orthopedist, but the benefit of routine use of these modalities has not been established.

**Diagnosis**
CTEV traditionally is diagnosed clinically. Although today’s orthopedist has many diagnostic tools, including radiographs, ultrasonography, CT, and MRI, CTEV remains a clinical diagnosis made primarily in the office or at the cribside.

**Antenatal**
Over the past 15 years, routine use of prenatal ultrasonographic screening has led to the antenatal diagnosis of CTEV. As technology and experience improve, the antenatal diagnosis of CTEV will become more frequent. For mothers in whom the diagnosis is made, counseling and amniocentesis are recommended for genetic testing because of the high incidence (14%) of genetic anomalies in this population, including trisomy 18, Larsen syndrome, neural tube defects, and congenital heart defects.

**Postnatal**
CTEV can be diagnosed on physical examination in nearly every case. The equinus and varus posture of the
foot is immediately apparent on inspection (Figs. 1 to 3). Because of the high incidence of comorbid diseases, including torticollis and hip dysplasia, a complete and thorough physical examination must be performed. The examination should include a detailed neurologic evaluation as well as an assessment of all joints and the spine. Patients who have CTEV have smaller calves on the affected side compared with the normal side. The depth of skin creases on the medial side of the foot offers a clue to the severity of the deformity. Documenting the rigidity of all aspects of the deformity is important because this may have prognostic implications.

A foot that rests in an equinovarus position but can be corrected passively to a neutral position is considered a positional clubfoot. These feet usually are treated with stretching or one to two serial casts.

One important aspect of the differential diagnosis is discerning between CTEV and metatarsus adductus (MTA). Children who have MTA have an adducted forefoot (pointed inward toward the midline) but a normal hindfoot (calcaneus and talus). With MTA, the ankle easily dorsiflexes past neutral, and subtalar joint motion (inversion/eversion) is normal. MTA lacks any component of the equinus or varus hindfoot deformity found in CTEV. Feet that have flexible MTA in which the forefoot can be corrected passively to or beyond the midline are treated with observation or stretching exercises. The natural history of MTA is benign, with 85% of children requiring no treatment. If the deformity is rigid or if it does not resolve in the first 6 months after birth, serial casting may become necessary.

Whenever a diagnosis of CTEV is being considered, secondary causes should be ruled out, usually with a thorough physical examination. Normal findings on the examination obviate further evaluation. However, if there are any other abnormal joints, any evidence of spinal dysraphism (such as sacral dimpling), or any abnormality on the neurologic examination, further evaluation, such as MRI, should be considered.

Classification
Orthopedists classify diseases to help determine the best treatment and prognosis. Unfortunately, no good classification scheme exists for CTEV that allows for accurate prognostication. Most schemes incorporate some combination of clinical (eg, flexibility) and radiologic aspects of the foot, but there is no standard scheme in the orthopedic literature.

Management
Nonoperative
Nearly all authors agree that initial treatment should be nonoperative, regardless of the severity of the CTEV. Historically, treatment consisted of forcible serial manipulations under anesthesia followed by casting. More recently, authors have championed the use of serial casting with minimal force (not requiring anesthesia) and have stressed the importance and potential for success of this nonoperative treatment (Fig. 6). Casting begins soon after birth to prevent the deformity from worsening and to take advantage of the neonate’s tremendous potential for remodeling. Casts are changed every 1 to 2 weeks until the foot has been corrected or improvement plateaus. Success can be defined as persistent restoration of the foot to a plantigrade position and avoidance of surgery. Kite claims successful treatment of more than 1,000 patients who had CTEV over 45 years with serial casting.

This “art” was lost to a large degree, as evidenced by studies reporting a success rate of not more than 50% and
usually closer to 15%. However, over the last few years, there has been renewed interest in manipulation and casting as a definitive treatment for CTEV. Reports from the University of Iowa have demonstrated a greater than 90% long-term (13- to 30-year follow-up) success for manipulation and casting, followed by an Achilles tendon tenotomy and bracing for several years. Although not strictly nonoperative (most children require an Achilles tenotomy, which can be performed percutaneously in the office, and many also require a tendon transfer later in life), this technique is less invasive than traditional surgical interventions, and other centers have published good short-term results. Critics of this technique point to the tremendous difficulty in making ambulatory children wear confining splints for 12 hours a day for several years. Without rigorous adherence to the splinting regimen, recurrence of the deformity is common.

Other nonoperative measures include bracing without casting, usually with some modification of the Denis Browne bar (Fig. 7). Some groups have reported successful treatment in nearly 65% of cases, although most believe that bracing alone does not constitute adequate treatment.

Although casting and splinting are the mainstays of nonoperative intervention for CTEV in North America, many Europeans employ physiotherapy and continuous passive motion without immobilization. Long-term results are not yet available, but early reports are encouraging. This aggressive physiotherapy approach also is being evaluated at centers in the United States with the addition of splinting (different from European centers) and sometimes continuous passive motion devices.

**Operative**

If conservative measures fail to correct the CTEV deformity, surgery is the remaining recourse. Unfortunately, there are no standard definitions of failure for nonoperative interventions. Most authors cite failure when the affected foot does not attain an anatomic or neutral position after 3 to 6 months of treatment or earlier if the rate of improvement from the treatment plateaus.

After determining that treatment has failed, the next question, which has vexed surgeons for as long as they have been operating on CTEV, is appropriate timing. There is no established “ideal” window for surgery, but there are some general guidelines. Attempts at surgical repair within the neonatal period have not proven successful. For children younger than 6 months of age, foot size makes the surgery technically more demanding and may affect outcomes detrimentally. Some surgeons prefer to wait 3 to 9 months; others wait for the foot length to reach a minimum of 8 cm. Another consideration is the safety of anesthesia in larger versus smaller infants. However, opinion is universal that the primary surgery should be performed in the first year of life if possible; after this time, much of the potential for growth and remodeling is lost.

Complications of surgery include wound healing problems, vascular injury, overcorrection, and undercorrection. Failure or recurrence (loss of correction) is a significant problem, and many clubfeet (7% to 47%, depending on technique and disease severity) require one or more revision surgeries. Clubfeet that fail surgical
treatment tend to be stiffer than conservatively treated feet, presumably due to scar formation.

**Summary**

The CTEV deformity remains a poorly understood condition in many respects. The mainstay of initial therapy, manipulation and plaster, has not changed conceptually in 2,500 years. The cause remains a mystery, and the morbid anatomy is just beginning to be understood with the application of modern imaging techniques. Evaluation continues to rely primarily on physical examination and plain radiography, although there is evidence that ultrasonography and MRI may assume a greater role. No method of classification or outcome assessment is widely accepted. Numerous described treatment strategies include early percutaneous Achilles tendon release with casting, casting and manipulation, surgery and dynamic treatment with physical therapy, and continuous passive motion devices.

**Suggested Reading**


PIR Quiz
Quiz also available online at www.pedsinreview.org.

7. The most likely cause of congenital idiopathic talipes equinovarus (CTEV) is:
   A. Genetic.
   B. Mechanical.
   C. Multifactorial.
   D. Neurogenic.
   E. Neuromuscular.

8. While instructing a group of medical students about examination of the newborn, you tell them that the diagnosis of CTEV is made best by:
   A. Computed tomography.
   B. Conventional radiography.
   C. Magnetic resonance imaging.
   D. Physical examination.
   E. Ultrasonography.

9. Among the following, the best method of distinguishing positional clubfoot from CTEV is to evaluate the:
   A. Depth of skin creases on the foot.
   B. Neurologic status of the infant.
   C. Presence of associated findings.
   D. Relative size of the calves of both legs.
   E. Rigidity of the deformities.

10. The nonoperative regimen used most commonly in North America for treatment of CTEV is:
    A. Bracing without casting.
    B. Casting and physiotherapy.
    C. Casting and splinting.
    D. Continuous passive motion.
    E. Continuous passive motion with splinting.

11. Most authorities agree that the optimal age for surgical intervention for CTEV is:
    A. 0 to 1 month.
    B. 1 to 3 months.
    C. 6 to 9 months.
    D. 9 to 12 months.
    E. 12 to 24 months.
## Congenital Idiopathic Talipes Equinovarus

Benjamin D. Roye, Joshua Hyman and David P. Roye, Jr

*Pediatrics in Review* 2004;25;124

DOI: 10.1542/pir.25-4-124

| Updated Information & Services | including high resolution figures, can be found at:  
| References | http://pedsinreview.aappublications.org/content/25/4/124  
| Subspecialty Collections | This article cites 11 articles, 0 of which you can access for free at:  
| Permissions & Licensing | http://pedsinreview.aappublications.org/content/25/4/124#BIBL  
| Reprints | This article, along with others on similar topics, appears in the following collection(s):  
| | **Fetus and Newborn Infant**  
| | http://pedsinreview.aappublications.org/cgi/collection/fetus_newborn_infant  
| | Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:  
| | /site/misc/Permissions.xhtml  
| | Information about ordering reprints can be found online:  
| | /site/misc/reprints.xhtml  

---

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN

---

Downloaded from http://pedsinreview.aappublications.org/ at Health Sciences Library State Univ Of New York on February 20, 2013