A REVIEW OF CURRENT MANAGEMENT

Congenital talipes equinovarus

Talipes equinovarus is one of the more common congenital abnormalities affecting the lower limb and can be challenging to manage. This review provides a comprehensive update on idiopathic congenital talipes equinovarus with emphasis on the initial treatment. Current management is moving away from operative towards a more conservative treatment using the Ponseti regime. The long-term results of surgical correction and the recent results of conservative treatment will be discussed.

Aetiology

The cause of congenital talipes equinovarus (CTEV), is unknown. Various theories have been proposed including vascular, viral genetic, anatomical, following a compartment syndrome, environmental factors and the effect of the position in utero. There is still a debate as to whether there is a neuromuscular basis for this disorder. Some studies have revealed ultrastructural and intracellular abnormalities in specimens of muscle in clubfoot, but others have found none.

One study observed that a deletion on Chromosome 2 (2q31-33) (related to the CASP10 gene) was associated with clubfoot. These genes encode proteins which are regulators of apoptosis, programmed cell death, which is particularly important during growth and development. Further studies are likely to reveal more about the molecular basis of the condition.

Most investigations of populations, families and twins suggest a genetic component, but the mode of inheritance does not follow a distinctive pattern. Studies on children with clubfoot support a single, major genetic factor, and observations on twins are useful in determining if the cause is principally genetic. Increased rates are found in monozygotic compared to dizygotic twins. A recent study from the Danish Twin Registry found that the risk of a second monozygotic twin having a club foot is one in three, suggesting that factors other than genetic are responsible.

Several studies have looked into the seasonal variation in the incidence of CTEV in different populations in an attempt to identify environmental factors which may be relevant. Robertson and Corbett demonstrated a seasonal variation and suggested that their data supported the view that CTEV may be caused by an intrauterine enterovirus infection. Barker and Macnicol also found a seasonal variation in the incidence, but another recent study failed to confirm this.

Epidemiology

Congenital talipes equinovarus occurs in 1.2 per 1000 live births in Europe and is twice as common in boys. First-degree relatives are at a significantly increased risk compared with the general population. A sibling of a patient has a 2% to 4% chance of having CTEV. If a child and another family member, or both parents, have clubfoot, the risk in another child increases by 10% to 20%. The more members of a family who are affected, the higher the risk of the condition occurring in other siblings, but the risk decreases significantly in second and third degree relatives.

In approximately 20% of cases, CTEV is associated with other congenital abnormalities. A comprehensive examination of the infant is necessary to detect physical signs which suggest that the condition is not idiopathic. An absence of toes may suggest tibial dysplasia and stiffness in the fingers may be associated with the distal form of arthrogryposis. Spina bifida is present in 4.4% of children with CTEV, cerebral palsy in 1.9%, arthrogryposis in 0.9% and other various neuromuscular defects in 7.7%. Other factors linked with the condition include amniocentesis, thyroid disorders, smallpox vaccination in the first
trimester, use of salicylate preparations by the mother in the first trimester and prenatal exposure to barbiturates.

A recent study in Australia found that four factors were significantly associated, namely maternal aboriginal race, male gender, maternal anaemia and maternal hyperemesis.14 A reduced risk of developmental dysplasia of the hip (DDH) in children with CTEV was also noted whereas most other studies have recorded an increased risk of DDH.

Pathoanatomy
The anatomical abnormalities in the limb such as malposition of the tarsal bones, atrophy of the calf muscle, and shortness of the foot, are well recognised. Although there are many descriptions of the morphological anomalies of the tibia and tarsal bones, there have been few concerning complicated abnormalities of the lower limb since the paper by Wynne-Davies in 1964.12 A recent study measured limb discrepancy in adolescence and early adulthood in patients with unilateral clubfoot. The girth of the thigh and calf was significantly smaller on the affected side, with the calf being more affected than the thigh. Radiological measurements of limb lengths also showed a significant difference between the two sides. The ankle height was decreased the most and the femoral length the least. The girth of the calf and the length of the tibia were found to be significantly less in patients undergoing operation than in those treated with casting. The discrepancy in limb length was 14.6 mm to 25 mm. There is both shortening of the limb and a generalised decrease in size, suggesting that CTEV may be part of a generalised disorder of development of the limb.

Classification
One of the difficulties in assessing the results of treatment has been that many papers have not described the severity of the initial deformity. A structured evaluation of the foot is essential to quantify the severity of the deformity accurately and consistently before treatment, and to record progress. Radiographs of the infant foot are difficult to interpret and clinical examination remains the optimum means of assessment. Flynn et al16 investigated the systems of classification of clubfoot developed by both Pirani and by Dimeglio, with the aim of determining their reliability and reproducibility. They found that after the initial learning phrase, both systems had very good interobserver reliability and reproducibility, showing that either could be applied to clinical practice. We consider that the Pirani score is simpler to use. It comprises two parts: one gives a hindfoot contracture score assessing the emptiness of the heel pad, the heel crease and equinus, while the other defines a midfoot contracture score concerning curvature of the lateral border of the foot, the medial crease and the prominence of the head of the talus. Since the publication of Flynn’s study, the Pirani score has been modified and to our knowledge, has not been validated for reproducibility. It has, however, been shown to be of prognostic value in the initial conservative management since those with a higher score are more likely to require tenotomy.17

Antenatal diagnosis
Congenital talipes equinovarus can be diagnosed antenatally using ultrasound. There is a wide variation in the reported accuracy. A recent study found that the diagnosis of clubfoot by ultrasound had a positive predictive value of 83% with a false positive rate of 17%. All the inaccuracies concerned the diagnosis. No cases of a complex clubfoot (CTEV with other congenital abnormalities) were missed, but the authors stressed that sequential scans of the fetus with CTEV were essential as the complexity of the deformities changed in 25% of cases. A similar study by Mammen and Benson19 found the false-positive rate was higher for unilateral (29%) than for bilateral clubfoot (7%). They noted that associated anomalies were more frequent with bilateral (76%) than with unilateral clubfoot (55%). Bar Hava et al20 described a transient deformity in the early weeks of gestation resembling clubfoot which was attributed to late maturation or high flexibility of the foetal limb muscles at this stage. Scans at 20 to 24 weeks may be more reliable for the diagnosis than those taken earlier.

One study, which looked at the clinical features and treatment required in feet diagnosed antenatally, found that the degree of deformity was difficult to assess before birth.21 At birth, 26% were found to require no treatment, while 61% needed surgery. This has important implications for prenatal counselling. Antenatal diagnosis of any abnormality raises the issue of termination of the pregnancy. Given the results of the contemporary treatment of CTEV it is essential that families are adequately counselled before such a decision is made.

Imaging
Imaging has a limited role but enables clinicians to understand how their treatment works and to assess its results. Ultrasound and MRI can be used to visualise the non-osseous parts of the skeleton.

One group used 3-dimensional (3D) MRI to measure the total cartilaginous volume and the volume of the ossific nuclei in both the talus and calcaneum.22 They found that these measurements were less in clubfoot than in normal feet. In particular, the volume of the nucleus of the talus was found to be 20% smaller in affected feet. They also found that the ossific nucleus in both the talus and calcaneum of patients with clubfoot lies more anterior than in the normal foot.

A recent study used ultrasound to describe the morphological changes at the talonavicular joint and at the calcaneocuboid joint in two groups of children with clubfeet.23 One group was treated using the Ponseti method24 and the other by the Copenhagen regime.25 The same degree of anatomical correction at the talonavicular and calcaneocuboid joints was achieved in both groups, but was reached more quickly in the group treated by the Ponseti technique. The use of ultrasound confirmed that there does not appear to be malrotation of the talus in the ankle mortise. Dorsal displacement of the navicular has been noted to occur in 6%...
to 43% of clubfeet and is associated with a high rate of corrective surgery.²⁶ It could be identified at an early phrase of the treatment.

Various imaging modalities have been used following treatment to record joint motion. High resolution MRI was used to confirm the clinical finding of generally reduced movement in the bones of the hindfoot after surgical treatment of CTEV.²⁵ It was noted that the movement of the individual bones in the hindfoot was not only less in magnitude after surgery but also sometimes occurred in a completely different direction to the normal.

Treatment
Over the last ten years, the treatment of CTEV has changed. The spread of the technique described by Ponseti²⁴ has been remarkable, both in the developed and the developing world. The paper published in 1995 describing the 30-year follow-up of patients treated using this regimen is responsible for starting the change.²⁸ Little has been published concerning the long-term results of the traditional surgical approach.

Surgery
The surgical management of the correction of CTEV can be split broadly into two groups. An ‘a la carte’ approach as described by Bensahal et al.,²⁹ where structures are released only until full correction is obtained, and the ‘one-size fits all’ procedure as popularised by Turco³⁰ in 1979, where each foot undergoes the same operation regardless of the severity. Even those who advocate a surgical solution have agreed that surgery does not produce a normal foot.³⁰

One study compared the long-term results in a group of patients with idiopathic clubfeet who had a posterior release compared with a group of children who had a comprehensive procedure.³¹ The clinical and radiological outcomes were compared at an average of 21 years after surgery. The outcomes of the two groups were significantly different. Those with a comprehensive release had fewer subsequent operations, better muscle strength, less hindfoot varus, less subtalar stiffness, and a better radiological outcome. There was no difference in the outcome assessed using the Laaveg and Ponseti score.³² The majority of clinical and radiological findings did not correlate with pain or function at skeletal maturity.

Turco³⁰ conducted an extended study where he reviewed 149 cases treated surgically between the ages of six months and eight years, using a one-stage posteromedial release with internal fixation. The follow-up was for between two and 15 years, and 85% of the cases were found to have good or excellent results using the author’s own scoring system. The unsatisfactory results were, for the most part, a consequence of overcorrection. The lack of other long-term results of this regime has been a concern.

Recently Dobbs, Nunley and Schoenecker³³ published the long-term results of soft-tissue correction of CTEV,³³ reviewing operations on 73 feet in 45 patients after a minimum follow-up of 25 years. They represented 73% of those operated upon within the study period. Most had undergone a Turco style release and 87% had more than one operation, the second usually in adolescence. The Laaveg and Ponseti scores³² revealed 0% excellent, 33% good, 20% fair and 47% poor results. The SF36 results³⁴ were compared with published norms. They showed significantly reduced scores in physical functioning, role physical, general health, vitality, social functioning and physical components. The values obtained for physical components were similar to those found in individuals with pain in the cervical spine with radiculopathy, Parkinson’s disease, haemodialysis, chronic heart failure and those awaiting coronary artery bypass grafting. Moderate to severe degenerative changes were seen, mainly in the talonavicular and calcaneocuboid joints in 56% of the patients. There were fewer degenerative changes in the feet treated with posterior release alone.

Operation is required for recurrent or resistant deformities, particularly equinus at the talotibial joint. The use of per-operative arthrography has been described in an attempt to assess the structures responsible for the equinus deformity and the extent of the release.³⁵ In this study, sectioning of the tendon Achilles did not alter the position of the talus in the sagittal plane. Only division of the posterolateral fibrous knot, which includes the tissues deep to the peroneal tendons, allowed correction of the talus. In all cases, equinus was corrected without the need for opening the capsule of the ankle joint or sectioning of the posterior talofibular ligament. Capsulotomy produced only an artificial, temporary increase in dorsiflexion because of posterior gaping between the talus and the tibia.

Continuous passive movement (CPM) has been shown to be effective in the conservative treatment of idiopathic clubfoot. Its use after operation was investigated in a prospective, randomised clinical study to determine whether it could improve the results in resistant club feet which had required an extensive soft-tissue release compared to immobilisation in a cast.³⁶ The results indicated a significantly better outcome in the CPM group up to one year after surgery, but at 18 and 48 months the result was the same in each group. A recent paper investigating the compliance with a CPM protocol used post-operatively has shown that the duration of treatment was less than the recommended four hours per day in 79% of cases. The mean use was for 126 minutes (11 to 496) despite ‘intensive training and continuous support.’³⁷ The authors postulate that the results of using CPM after operation may be improved by ensuring better compliance.³⁷ Certainly the primary treatment using CPM requires a significant in-patient component to ensure compliance.

Ponseti regime
The Ponseti regime²⁴ involves serial casting of the lower limb using a strictly defined technique. The casts may be changed every five or seven days.³⁸ Once the foot is cor-
rected, an abduction foot orthosis must be worn full time for 12 weeks, and then at night and at nap time, up to the age of four years. Percutaneous tenotomy of the tendon Achilles and transfer of the tibialis anterior tendon are integral parts of the protocol. The tenotomy of the tendon Achilles is usually performed once the talonavicular joint is reduced, but when the hindfoot remains in equinus and shows no sign of correcting, a position which has been termed ‘hindfoot stall’. Transfer of the tibialis anterior tendon is indicated for recurrent deformity which develops in children over two and a half years of age. Any fixed deformity should be treated by cast beforehand. The key paper on the Ponseti regimen was that published by Cooper and Dietz in 1995. They reviewed a group of 45 adults, with 71 clubfeet, who had been managed with the Ponseti method, 30 years after treatment. The results were compared with normal controls. The investigators used a questionnaire, the findings on a structured examination, radiographs, electromyography and measurements using a pedobarography, to study both groups. Using the Laaveg and Ponseti score, the results in the normal controls and in those with treated clubfeet were the same, with 62% excellent. Radiographs showed that the feet were not completely corrected, but functioned well despite this. A further study from Iowa described the short-term results of a more recent series of 256 feet. Correction was obtained in 98% of the patients with between one and seven casts. Percutaneous tenotomy of the tendon Achilles was performed in 86% of the cases. The mean angle of dorsiflexion of the ankle after tenotomy was 20° (0° to 35°). Minor complications from the cast were encountered in 8% of patients and 2.5% required extensive corrective surgery. The rate of relapse after initial successful treatment was 10%.

Tenotomy of the tendon Achilles is an integral step in the Ponseti technique. After percutaneous section just above the calcaneal insertion, the tendon will unite within three weeks, as has been shown using ultrasound. There is no information concerning the strength of the healed tendon but late rupture after the procedure has not been reported.

One study found a 2% incidence of serious bleeding following tenotomy and it was suggested that a careful surgical technique would reduce the rate of this serious complication.

The need for tenotomy of the tendon Achilles is not universal. Scher et al. found that the more severe the foot deformity, the more likely that a tenotomy would be required.

The Ponseti method is successful in children who present after the neonatal period. Bor, Herzenberg and Frick reviewed a number of children who were first seen after the age of three months in whom conservative treatment had failed. After a period of 24 months, only one of 36 feet (2.8%) required extensive surgery.

Traditionally, the application of casts for CTEV has been performed by either clinicians or physiotherapists. A recent study has confirmed that a trained physiotherapist could achieve good results. Clubfoot is responsible for much disability in developing countries where access to surgical services is limited. The Ponseti regimen has been used in Malawi to good effect. The incidence of CTEV in Malawi is twice that of Western Europe. It was shown that the technique could be learned and practised effectively by orthopaedic clinical officers who are not doctors. They work in the remote and rural parts of the country, which enables many families to receive treatment which would otherwise have been inaccessible and unaffordable.

Recently Alvarez et al. used Botulinum toxin-A injected into the gastrocsoleus complex of children with CTEV as an alternative to Achilles tenotomy after a course Ponseti casting. They found that this procedure produced satisfactory results with less skin scarring and deep tissue fibrosis than a percutaneous tenotomy. However, a recent prospective randomised double-blind study of 20 newborns with CTEV found no significant difference between injections of a placebo or Botulinum toxin-A in conjunction with serial manipulation and casting as regards the speed of correction, the need for tenotomy of the tendon Achilles and the risk of relapse.

Non-compliance with the use of the foot-abduction orthosis is the primary risk factor for recurrence of the deformity. The level of parental education is also important. Recurrence is not dependent on the initial severity of the deformity, the age of the initial treatment, the number of casts required for correction or whether the patient had previous non-operative treatment of the deformity. One study reported the success of two different casting regimes. One group comprised feet treated with a modification of the Kite technique and the other feet treated by the Ponseti method. There was no significant difference between the two groups in the initial severity of the deformity but there was a difference in the mean follow-up of the patients, with 29 months (16 to 45) for the Ponseti group and 54 months (44 to 68) for the Kite patients. At follow-up, 57% of the feet in the Kite group needed surgery and 44% had a residual deformity. In the Ponseti group, only 6% of the feet had residual deformity and were operated upon.

Bensahel/Dimeglio regime

The emphasis on non-operative management has stimulated interest in this French method. It requires daily manipulations of the newborn’s clubfeet by a skilled physiotherapist and temporary immobilisation with elastic and non-elastic adhesive taping. A CPM machine is used for further mobilisation during the hours of sleep. Most of the improvement occurs during the first three months of treatment. If successful, the programme continues and is performed daily by the parents until the child is walking. The results of this method have been investigated in a retrospective study. A review of 142 feet with greater than moderate severity treated by the French method was undertaken after 35 months. Poor results were found in 20.4% of the
cases. Surgery was not required in 42% of the feet, 9% had a tenotomy of the tendon Achilles, 29% needed a posterior release and 20% a comprehensive posteromedial release. The Dimeglio classification scale was prognostic in determining the outcome of the treatment, with moderate feet having the best results and very severe feet the worst outcome.

The Ilizarov method
Recurrence of the clubfoot deformity remains a complex problem. The Ilizarov technique of gradual distraction and correction of deformities using an external frame has been described as an alternative treatment in cases of recurrent or resistant feet. Long-term results have been lacking, but a recent study showed unexpectedly fair or poor results in 86% of feet. Surgery was required for recurrent deformity in half of these. This is the first study that has demonstrated that the Ilizarov method for treatment of resistant clubfoot deformities may not be as successful as had been hoped. We were unable to find published results for use of the Ilizarov method as a primary treatment.

Surgical vs conservative treatment
Few studies have directly compared feet which had undergone operation with those treated with more conservative measures. One analysed the findings on gait analysis in children aged two who had undergone the French physical therapy programme with a matched group of patients who had undergone posteromedial release. Only feet with severe deformity (Dimeglio scores 10 to 17) were included in the study. Of those undergoing an operation, 29% of the feet were in calcaneus and 8% were in still in equinus. Intoeing was more common in the surgical group (45.1%) compared with the physical therapy group (30.8%). Few children in any group had completely normal parameters of gait, but the highest incidence of normal gait was seen in the physical therapy group with 32.7% described as normal compared to 33.7% in the surgical group.

Another study used CT at skeletal maturity to compare patients treated by different protocols. One group was treated with manipulation and serial casting, followed by posteromedial release for the resisting feet. The second group included patients who were treated by a modified Ponseti regime in which open z-lengthening of the tendon Achilles was carried out in preference to percutaneous tenotomy. The Ponseti group had better correction of cavus, supination and adduction and several radiological parameters were also significantly better. It is important to note that 85% of feet had an abnormal subtalar joint regardless of the method of treatment. Residual subluxation of the navicular was seen in 75% of those treated by surgery and in 92% of the Ponseti group. External torsion of the ankle mortise was higher than normal in both groups.

Outcomes
In order to study the outcomes in many paediatric disorders, it is important to use valid instruments to measure the results of treatment. There has been growing emphasis on the use of patient-based assessment tools. The clinical results of 46 patients with clubfoot were assessed using both traditional radiological and patient generic and disease-specific measures 16 years after surgery. The findings from this study agreed with others which have shown that radiographs are not good indicators of outcome. We recommend that both disease-specific and generic outcome measures should be used in assessing the results of the treatment of CTEV.

Conclusions
There is still much to learn about idiopathic CTEV. The over-riding principle of management is to achieve and maintain a painfree, plantigrade and pliable foot. The few long-term results of surgical correction are disappointing. The initial results of treatment with the Ponseti regimen used across the world are encouraging. Longer term follow-up will be required to see whether the technique lives up to its expectations. The management of resistant and recurrent deformities continues to remain a challenge.

References


