The standard treatment of clubfoot has changed greatly in the past 10 years. Previously, extensive surgery was common in children born with this condition. The publication of long term evidence of good outcomes with more minimally invasive methods, such as the Ponseti technique, has led surgeons worldwide to change their approach. Ponseti treatment consists of sequential plasters and prolonged bracing, with minor surgical procedures.

This clinical review describes clubfoot and its current management. It is particularly aimed at general readers who are non-specialists but may be involved in the care of patients with this condition. The evidence underpinning this review is largely observational. Although the Ponseti method was first described over 30 years ago, it is only since the publication of long term outcomes of case series that it has been widely adopted.

What is clubfoot and who gets it?
Clubfoot, also known as congenital talipes equinovarus, is a developmental deformity of the foot. It is one of the most common birth deformities with an incidence of 1.2 per 1000 live births each year in the white population. Clubfoot is twice as common in boys and is bilateral in 50% of cases.

It is most often idiopathic but may be associated with other conditions in around 20% of cases. The most common associated conditions are spina bifida (4.4% of children with clubfoot), cerebral palsy (1.9%), and arthrogryposis (0.9%). Although it was previously thought to be associated with developmental dysplasia of the hip a recent prospective study did not support this.

Historical family studies suggest that there is a genetic component but not a recognisable pattern of inheritance. If one child has clubfoot, the risk of clubfoot in a subsequent child is increased 20-fold. The risk to the second of identical twins is one in three. If one parent has clubfoot the risk of having affected offspring is 3-4%, but if both parents are affected the risk is 30%.

How is it diagnosed?
Clubfoot is most commonly diagnosed postnatally during the routine baby check. The foot assumes the position shown in fig 1. The foot points downwards at the ankle (equinus) the heel is turned in (varus), the midfoot is deviated.

The foot is manipulated by gentle stretching and then left in a corrected position with the foot in a splint. The splint is worn for much of the day and removed for washing and for the child to play. The foot is manipulated several times a day and the splint is changed every few days.

Current best treatment is by casting and bracing according to the Ponseti method. Results are better with manipulative methods than with surgical release. Recurrences can occur and are normally caused by non-compliance with bracing.
Clubfoot is increasingly diagnosed on prenatal scans and these have a positive predictive value of around 85%.

**Why do I need to know about clubfoot?**

If diagnosis and referral to an orthopaedic surgeon do not occur prenatally or in the first few days after birth, the baby must be referred urgently when the deformity is first noticed. This is because the earlier Ponseti treatment is started (ideally around one to two weeks), the easier correction will be.

It is important for non-specialists to have an understanding of the standard treatment because success is largely related to the parents' compliance with the bracing protocol. This is prolonged and can be demanding. Support and encouragement from healthcare professionals can be helpful. It is important for parents to receive a clear message that the long term use of the brace is essential.

**What was the standard treatment previously?**

Although manipulation and casting were used in the past, this was not performed according to a formal protocol, and extensive surgery was often used. The aim of surgery was to correct the deformity by lengthening or dividing all structures that were tight. Few long term studies on the outcome of surgery are available, and most evidence comes from case series. One case series published in 2006 looked at 73 feet in 45 patients with a minimum follow-up of 25 years. This was a thorough review with the patients completing three independent quality of life questionnaires, including the Laaveg and Ponseti functional score, which is commonly used in clubfoot outcome studies.

On this measure, 34 feet (47%) had a poor outcome and most had more than one operation. In a case series looking at staged surgical release, 99 feet in 71 patients with severe clubfoot were studied. The average follow-up was 11.5 years and the relapse rate was 76%. Gait analysis has been used by some investigators to try to quantify the results of treatment. A case-control study using this technique compared the outcomes of surgical and non-surgical treatment. This showed that surgery led to a greater proportion of gait abnormalities.

**What is the current preferred treatment?**

The current preferred treatment for clubfoot is the Ponseti method. This is a detailed method of manipulation and casting without major surgical releases, and it is the treatment of choice of most orthopaedic surgeons worldwide. A review of patients treated by Ponseti published in 1995 showed good long term results. In this study, 45 patients with 71 clubfeet were reviewed after an average of 30 years. Using pain and functional limitation as the outcome criteria, 35 of 45 patients (78%) had an excellent or good outcome compared with 82 of 97 (85%) of age matched patients without foot pathology. Although Ponseti originally published his method in 1963, it was only after this long term review that interest began to increase. Studies performed in other centres have confirmed his good results, although with shorter term follow-up. Acceptance by orthopaedic surgeons has been encouraged by

**Box 1 | Other common congenital foot deformities**

| Positional clubfoot: the foot assumes the same position as in congenital clubfoot but the deformity is correctable. This is probably a normal variant. |
| Metatarsus adductus: medial deviation of the forefoot on the hindfoot creates an adduction deformity. This may be correctable or fixed. The heel is in a neutral position (unlike clubfoot) and there is no equinus. This is more common than clubfoot, although its exact incidence is difficult to determine. Most cases will improve spontaneously, but more severe cases may require serial casting. |
| Positional calcaneovalgus: dorsiflexion of the whole foot to the extent that it may touch the tibia. This dramatic deformity is unlikely to be confused with clubfoot, but the appearance is concerning and it is important to rule out more serious conditions such as congenital vertical talus. Calcaneovalgus improves spontaneously with time. Both metatarsus adductus and calcaneovalgus are associated with developmental dysplasia of the hip and if diagnosed should lead to referral for assessment of the hips. |

**Box 2 | Grading of clubfoot: the Pirani score**

In this system, six clinical features of the deformity are graded 0, 0.5, or 1. The six scores are summed giving a total score of 0–6—the higher the score the worse the deformity.

The features scored are:
- Hindfoot: heel crease, equinus, and softness of the heel
- Forefoot: lateral border shape, medial border creases, and cover of head of talus

**Box 3 | Improving compliance with bracing**

- Parents need to understand why bracing is important and why it must continue for such a long time. Parents should be encouraged to accept responsibility for this phase of the treatment
- For older children, wearing the brace at night must be part of their normal routine. It should not be stopped for reasons such as illness
- Baby sleeping bags may help the boots stay on
- Socks with non-slip areas on the soles may also help the boots stay on
- The boots must fit well and be comfortable, and the bar width should be correct. If concerns exist, the parents should be directed back to the brace provider
QUESTIONS FOR FUTURE RESEARCH

• Which surgical techniques give the best results in feet that do not fully correct after Ponseti treatment?
• How effective is the “reverse Ponseti” technique for congenital vertical talus?
• What surgical technique is the best treatment for the “neglected” clubfoot in older children and adults?

parents who use the internet to seek out surgeons who use this technique.\(^\text{16}\) Parents prefer the more non-surgical approach and can become strong advocates for the technique. This method is particularly useful in developing countries where surgical services are limited. A descriptive study of its use in Malawi showed that it could be successfully carried out by suitably trained non-doctor personnel with similar results to those seen in other studies.\(^\text{17}\)

What is the Ponseti method?

Treatment starts as soon after birth as possible. Ponseti defined a precise sequence of manipulations of the clubfoot that lead to correction of the deformity. Exact details of the technique are available free on the internet.\(^\text{4}\) Ponseti stressed that the cavus should be corrected by raising the first metatarsal, which initially makes the deformity look “worse,” and that correction should occur around the head of the talus without the heel being touched. At weekly intervals the foot is manipulated into the maximum position of correction and then held in a plaster of Paris cast. Several studies have shown that this manipulation and casting can be carried out by doctors and other trained staff.\(^\text{17,18}\)

During the time in cast the immature collagen undergoes stress relaxation (stretches); this then allows greater correction at the next manipulation. After about six weeks of weekly cast changes the deformity of the midfoot and forefoot is generally corrected. The foot is often still in the equinus position at this stage (pointing down at the ankle), and in most cases this will not correct further with manipulation. Therefore, around 85% of children have an Achilles tenotomy carried out at this stage. This can be done under local anaesthetic in clinic or under general anaesthesia. The child then goes into a final cast for three weeks.

On removal of this final cast the foot position is reviewed. If correction is complete the child then goes into “boots and bar.” This is an orthotic device that holds the feet in an abducted, externally rotated, and dorsiflexed position about a shoulder width apart (fig 2). In a unilateral case the affected side is externally rotated 60-70° and the unaffected side is rotated 30-40°. The child wears this device all the time for three months and then at night time and during naps until 4 years of age. If treatment is successful the child will be left with a supple well corrected foot. It should look similar to the unaffected foot but may be slightly smaller (around one shoe size). The calf may also be smaller than on the unaffected side.

Can clubfoot recur?

Yes, recurrence occurs in around 15% of patients and can be at any stage in the treatment process. It occurs most commonly during the time in the boots and bar device, as a result of poor compliance.\(^\text{17}\) At initial signs of recurrence it is important to ensure compliance with the boots and bar. Simple measures and advice may increase compliance and lead to correction of recurrence (box 3). If these measures are not successful, recurrences can be treated with a further period of manipulation and casting. If the recurrent deformity is dynamic supination of the forefoot this can be corrected by a tibialis anterior tendon transfer. This is described by Ponseti and is the only other surgical procedure that is a standard part of the Ponseti method.\(^\text{4}\) It is carried out far less often than Achilles tenotomy (around 15% of cases).

Are outcomes better with current treatment?

No randomised trials have compared the Ponseti method and surgical management of clubfoot. Long term studies of the Ponseti treatment suggest that the results are much better than with surgical treatment, however.\(^\text{11,12,15,20}\) In the 30 year review of Ponseti’s patients, 35 of 45 (78%) had an excellent or good outcome using the Laaveg-Ponseti technique. The technique is carried out far less often than Achilles tenotomy.
seti functional score. This compares with 24 of 73 feet (33%) in the previously discussed case series, which had a similar follow-up of surgically treated patients. It is unusual to find studies where case series from the same authors can be compared, but in one such paper, with long term follow up, 20 of 47 feet (43%) treated surgically had an excellent or good result compared with 38 of 49 (78%) treated by the Ponseti method.

Can the Ponseti method be used for non-idiopathic clubfoot?

Yes, although syndromic clubfoot is more difficult to treat, may need more plasters, and may not correct fully with Ponseti treatment. However, this treatment will improve the position and make subsequent surgery easier; it also is practical, Ponseti treatment can also be used for late presenting clubfoot.

What other forms of treatment are available?

Other specific methods of manipulation have been suggested. One of these is the French method of manipulations (also known as functional or physiotherapy method). This requires daily manipulation of the foot and taping. A non-randomised study in one unit found it to be as effective as the Ponseti method. When given the choice, however, parents were twice as likely to choose the Ponseti method as the French method, probably because of the need for daily attendances with the French method.

Ilizarov frames are external fixators that can be used for gradual correction of deformity. They can be used to stretch soft tissues or to alter the foot shape through osteotomies. They have been used for recurrences after Ponseti treatment and residual deformity in older patients. Not all studies show good outcomes however.

Surgery still has a part to play in the management of clubfoot. The Ponseti method does not fully correct the defect in a proportion of patients, and these children will need surgery. Patients with syndromic clubfoot are also more likely to need surgery. During surgery, postoperative splints are removed or lengthened to allow the foot position to be corrected. Older patients with residual deformity of the foot may need osteotomies in addition to soft tissue procedures.

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**USEFUL RESOURCES FOR NON-SPECIALISTS AND PATIENTS**

**Resources for healthcare professionals**

Ponseti International Association (www.ponseti.info)—Website of the unit where Dr Ponseti practised. The website promotes the Ponseti method and provides education for healthcare professionals

Global Help (www.global-help.org)—Clubfoot: Ponseti management. This document contains detailed information on the Ponseti method and practicalities of treating patients with this technique

**Resources for parents**

Steps charity (www.steps-charity.org.uk)—A UK charity website that has information on clubfoot as well as other lower limb conditions

Clubfoot.co.uk (www.clubfoot.co.uk)—Website set up by the parents of a child with clubfoot that contains good basic information and describes their experience

Ponseti International Association (www.ponseti.info/parents)—Website specifically for parents that is intended to promote the Ponseti method. It has information on the method itself and doctors who offer it

**TIPS FOR NON-SPECIALISTS**

- Although patients should be seen as soon after birth as is practical, Ponseti treatment can also be used for late presenting clubfoot
- Recurrence is associated with non-compliance with bracing. Parents must be encouraged to continue using the boots and bar
- Ponseti treatment is successful in around 85% of patients. Parents can be reassured that a good outcome is likely

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**CLINICAL REVIEW**
A toddler with pallor and recurrent infection

1. This child’s initial symptoms were non-specific, although their long duration suggested an underlying disorder such as leukaemia. Recurrent infections suggested immunodeficiency, but as the history unfolded, lethargy and pallor indicated anaemia. The combination of anaemia and jaundice raised the possibility of haemolysis, perhaps, given the infective symptoms, virally induced. Joint pain suggested a septic or juvenile arthritis, but the concurrent anaemia made leukaemia or other malignancy more likely, especially because several joints were affected. The finding of hepatosplenomegaly with adenopathy made leukaemia highly likely, but neuroblastoma would be a plausible alternative.

2. Children with acute leukaemia are at risk on presentation of overwhelming sepsis, haemorrhage, leucostasis, airway obstruction as a result of a mediastinal mass, acute heart failure secondary to profound anaemia, and renal failure precipitated by tumour lysis syndrome.

3. Examination of the bone marrow is the investigation of choice. Although many of the diagnostic tests for leukaemia can also be performed on peripheral blood samples, definitive diagnosis is still based on examination of a bone marrow aspirate.

4. Acute leukaemia is most likely to cause jaundice through cholestasis, both intrahepatic (owing to infiltration of the hepatic parenchyma by leukaemic cells) and extrahepatic (owing to compression of the biliary tree by enlarged abdominal lymph nodes).

5. Five year survival for all types of leukaemia in children is now over 80%, although prognosis varies considerably among subtypes.

PICTURE QUIZ

Gradually worsening shortness of breath

1. The chest radiograph shows hyperinflation with bullous emphysematous changes in the lower zones bilaterally.

2. Findings are consistent with deficiency of the enzyme inhibitor $\alpha_1$ antitrypsin. This protease inhibitor is synthesised by the liver, and protects lung tissue against proteolytic damage from the enzyme neutrophil elastase. Deficiency is associated with early onset emphysema that characteristically affects the lung bases. Basal emphysema is not always obvious on plain radiography, and the condition is often diagnosed as regular chronic obstructive pulmonary disease (COPD).

3. Airflow obstruction should be confirmed by spirometry. Serum $\alpha_1$ antitrypsin concentrations: concentrations less than 11.0 $\mu$mol/l (normal range 20-60) increase the risk for developing emphysema. Genotyping is encouraged, along with family testing after diagnosis. High resolution computed tomography of the chest should also be performed.

4. Smoking cessation is the most important factor and has the greatest effect on survival. Treatment is similar to that for standard COPD and should follow the guidelines for this disease: short and long acting inhaled $\beta_2$ agonists, anticholinergics, inhaled corticosteroids, and pulmonary rehabilitation. Disease specific treatment includes enzyme replacement, which is used commonly in the United States but rarely outside of trials in the United Kingdom. Observational trials have suggested benefit for this treatment, but no controlled trial has confirmed this. All patients with COPD, but especially those with early onset disease, should be investigated for $\alpha_1$ antitrypsin deficiency.