Review Article

Congenital clubfoot

Sanjay Meena, Pankaj Sharma, Shreesh Kumar Gangary[1], Lalit Kumar Lohia[2]

Department of Orthopaedics, All India Institute of Medical Sciences, [1]Department of Orthopaedics, ESI Hospital, Okhla, New Delhi, [2]Department of Orthopaedics, Fortis Memorial research Institute, Gurgoan, Harayana, India

Address for correspondence:
Dr. Sanjay Meena,
Department of Orthopaedics, All India Institute of Medical Sciences, Ansari Nagar, New Delhi, India.
E-mail: sanjaymeena@hotmail.com

ABSTRACT

Clubfoot is the most common congenital deformity of lower limbs. Its etiology remains an enigma. The aim of the treatment is to obtain a plantigrate, painless and functional foot. Conservative treatment with Ponseti method of clubfoot is well-accepted and has been reported to result in good correction ranging from as low as 50% to as high as 90%. Surgical treatment is indicated only after failure of conservative methods, with limited release also known as "*a la carte*" release. This review looks at etiology, clinical assessment, scoring and provides an overview of operative and non-operative treatment options.

Keywords: *Congenital clubfoot, foot deformities, Ponseti method*

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Introduction

Clubfoot or Congenital talipes equino varus (CTEV) was introduced in the medical literature by Hippocrates around 300 B.C. The incidence of CTEV is 1–2 per thousand live births.[1] The involvement is bilateral in about 50% of cases and in unilateral cases the right side is affected slightly more common than left.[1] Idiopathic clubfoot is approximately twice as common in males as in females. Clubfoot is an obvious deformity easily recognized. It has four components: Equius, midfoot cavus, forefoot adduction and hindfoot varus. It is astonishing that in spite of the vast attention paid to it, there is still much uncertainty about its etiology and no consensus has been reached on methods of assessment and treatment.

The deformity in idiopathic club foot (CTEV) is both cosmetic and functional with associated hypoplasia of skin, muscles, tendons, bones, ligaments and neurovascular bundle on the medial side and the affected foot is smaller than the normal foot.[2]

Current treatment of clubfoot deformity consists of initial trials of manipulation and serial casting. 30–50% feet treated in this way eventually need surgical correction.[3] An average of 25% of the operated feet will have poor results and will need secondary surgical intervention.[3] The aim of the review is to provide an overview of clubfoot.

Etiology

Clubfoot may be associated with other congenital abnormalities but is usually an isolated finding which is idiopathic in nature. Multiple theories have been proposed to explain its etiology.

One is that any arrest during the development of the foot during intrauterine life would develop clubfoot.[4,5] Ignacio V. Ponseti supported the "arrest of development" theory.[6] Palmer supported multifactorial system of inheritance, possible with intrauterine factors having some effect.[7] Wynne Davis supported the polygenic theory and showed a rapid decrease in incidence of clubfoot from first to second to third degree relatives. About 2.9% of siblings in the first degree relatives had this deformity as compared to 1–2 per thousand in general population that is, 25 times more chances in siblings of an affected child.[8]

Insley reported the association of clubfoot with a deficiency of a part of the long arm of chromosome eighteen.[9] Sherman and Irani postulated primary defect in germ plasma as the cause of clubfoot. They dissected eleven extremities with talipes equino varus and fourteen normal feet. In their anatomic dissection they found no primary abnormalities of the vessels, nerves, muscles and tendon insertions. The constant abnormalities were found in the anterior part of the talus. The neck of the talus was found to be short. The anterior portion of the talus was always rotated

in the medial and plantar direction, so that the articular surface no longer faced directly forwards. Since the anlagen for the talus are fully formed at 6 weeks and tarsal joints are well-developed at 7 weeks, it is difficult to imagine an exogenous trauma at this stage, which could damage only the anterior part of the talus and that too often only in one foot.[10]

Environmental factors have also been implicated-. Some of these are: External pressure in utero (be it due to hydroamnios or oligoamnios), Infectious disease during pregnancy and maternal nutrition defects, vitamin deficiency, toxic agents like azaserine, d-tubocurarine, aminopterin etc., and maternal metabolic disorders.[11,12]

Clinical Assessment and Grading

Initially the assessment was purely subjective and based on the severity of the deformity and flexibility of foot.[13] Mac Even assessed the clubfeet by the degree of dorsiflexion possible, heel varus, forefoot adduction, calf atrophy and graded the result as excellent, good, fair and poor.[14] W. Davies developed the rating system and according to it every clubfoot was given 10 marks to begin with and marks were deducted from 10 according to the table.[15]

Chacko[16] (1976) described a preoperative grading of clubfoot depending upon the components of the deformity present; suppleness of the foot, amount of correction possible. Feet were graded from I to IV.[16] Douglas McKay[3,4] (1983) developed a rating system for routine follow-up and comparison of clubfoot, based on surgeons assessment and patients subjective complaints. His ten assessment criteria's were: Ankle motion, bimalleolar angle, strength of triceps surae, heel, forefoot, flexor hallucis longus, painful gait, subtalar pain, shoe wear and Sports. The system is based on the arbitrary assignment of 180 points according to deformity, loss of ankle mobility, loss of function and pain proportional to their deviation from normal.[17,18]

Classification systems given by Dimeglio and Pirani have stood the test of time and are most widely used [Figure 1 and Table 1]. Both systems give points according to the physical examination.[19,20]

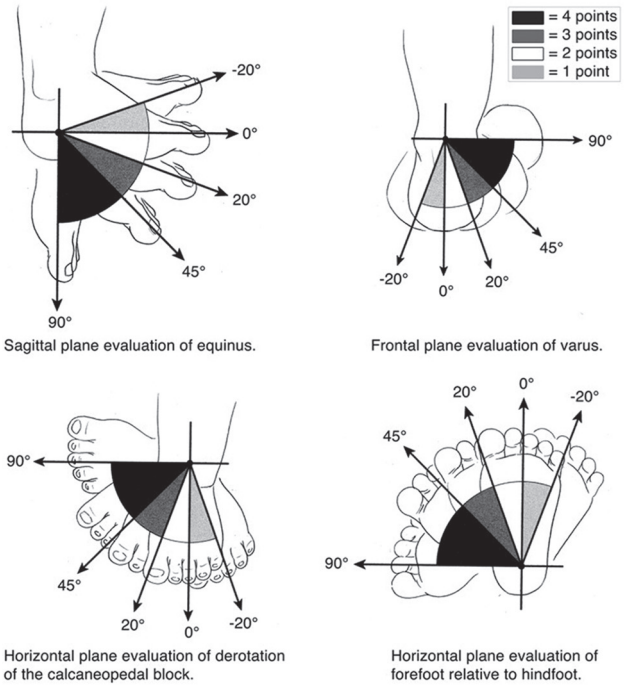
Radiological Assessment

Radiographs are indicated in talipes equinovarus to assess the degree of subluxation of the talocalcaneonavicular joint and the severity of the deformity before commencing treatment; to provide an accurate guide to progress during the course of closed non-operative treatment; to ascertain whether reduction of the talocalneaneonavicular dislocation and normal articular alignment have been achieved; to analyze the composite deformities preoperatively and to plan operative treatment accordingly; to determine intraoperatively whether concentric reduction of the talocalneanonavicular joint has been achieved; and to ascertain postoperatively whether normal articular alignment is being maintained.

Beatson and Pearson described the application of talocalcanial-index as a reliable tool for assessing correction at the end of the primary course of treatment, more in those cases in which correction was inadequate.[21] When the deformity in the hind foot is corrected the anterior end of the calcaneus swings laterally and upwards and resumes a more normal relationship with the talus. This increases the talo-calcaneal index. If this index moves into the normal range the deformity is corrected and will not recur. If it remains below this range, although in most instances the condition is improved, it is not adequately corrected and further treatment is advisable. If the talocalcanial index be less than 40 degrees even though clinically the foot appears corrected-further treatment should immediately be undertaken.

Classification

Classification Grade	Type	Frequency (%)	Score
I	Benign	20	(<5)
II	Moderate	33	(=5<10)
III	Severe	35	(=10<15)
IV	Very severe	12	(=15<20)



Assessment of Clubfoot by Severity Scale

Characteristics: Reproducibility	Points	Characteristics: Other parameters	Points
90°-45°	4	Posterior crease	1
45°-20°	3	Medial crease	1
20°-0°	2	Cavus	1
<0° to -20°	1	Poor muscle condition	1

Figure 1: Dimeglio classification of clubfoot. Each major component of clubfoot (equines, heel varus, medial rotation of calcaneopedal block, forefoot adduction) is graded from I to IV. Additional points are added for deep posterior and medial crease s, cavus and poor muscle condition

In the literature, numerous angle measurements for assessing talipes equinovarus have been described and the ranges of which vary among authors. The normal ranges according to Simons are[22] [Table 2].

Vanderwide *et al*., in a detailed study on measurements on X-ray of foot in normal infants and children noted that the mean values and normal values changed with age.[23] Moses *et al*., described predictive value of intra operative clubfoot radiographs on revision rates; 115 clubfoot in 73 children who underwent surgery were reviewed. A strategy using sequential release guided by intra-operative radiographs was employed in all cases. The intra-operative radiographic findings correlated with the need for early revision surgery, with residual radiographic evidence of cavus deformity associated with greatest risk of revision.[24]

Benjamin Joseph *et al*.,[25] (2001) stated that the lateral talo-calcaneal angles in normal feet were higher in dorsiflexion than in plantar flexion, whereas the converse was true in clubfeet. A lateral talo-calcaneal angle difference (between the stress dorsiflexion and plantar flexion angles) of 20 degrees suggests that there is a 93% probability that the hind foot deformity of clubfoot has been adequately corrected. A talo-calcaneal angle of 30 degrees or a talo-calcaneal index of 40 degrees does not ensure correction of clubfoot.[25] Magnetic resonance imaging (MRI)-scan have also been used to evaluate the deformity and to assess the correction. A MRI imaging protocol was devised to image the chondroosseus abnormalities of the virgin clubfoot deformities and illustrated in brief conventional radiography and tomography are not useful in early infant age due to unossified cartilaginous structures in limb while arthrography and MRI are not cost-effective and interpretation of these investigation are not uniform universally the changes that occur with Ponseti method of treatment.[26]

Table 1: Pirani scoring

Paramaters	Normal	Moderate	Severe
Midfoot			
Curved lateral border	0	0.5	1
Medial crease	0	0.5	1
Talar Head coverage	0	0.5	1
Hindfoot			
Posterior crease	0	0.5	1
Rigid equines	0	0.5	1
Empty heel	0	0.5	1

Table 2: Important radiological angle

	Normal range (in degrees)
Anteroposterior view	
Talocalcaneal	20-40
Talo-1st metatarsal	0-20
Calcaneal-2nd metatarsal	15-20
Lateral view	
Talocalcaneal	35-50
Tibiotalar in dorsiflexion	70-100
Tibiotalar in plantar flexion	120-180
Calcaneal 1st metatarsal	140-180
Talacalneal index	≥40

	Tuesday, October 28
CCHMC Updates – Dr. Hostetter (A8)	
PL1 – Town Hall Mtg (A8)	
Grand Rounds Noon Conference	
Ghory Lecture: Allergic Respiratory Diseases: Emerging Treatments and Challenges – Dr. Linda Cox (Sabin)	
Hospital Medicine: TBD – Drs. Statile, Unaka and Thomson (A8)	
	Purple QI (A8)
	No noon conference (A8)
Well – Rounded Resident (A8)	
Psych – TBD – Dr. Courtney Cinko (A8)	

Treatment

The aim of the treatment is obtain a plantigrate, painless and functional foot. Every conceivable form of treatment has been recommended by various authors, tried by many at different times with varying success rates.

Hippocrates (300 BC) said about clubfoot treatment that what could be done without force, could be done without harm. Bensahel *et al*., (1980) in France treated clubfoot by well-trained physiotherapist with daily manipulations of foot for three months, followed by taping of leg and foot to a splint. Physiotherapy softens the tissues making the foot more compliant. Ponseti argued that this style of repeated mobilization of displaced tarsal joint through physiotherapy will not reshape the foot as compare to what immobilization in proper position does. Moreover his method was lengthy and expensive and failed to correct more than one-fourth cases. Metaizeau[56] treated 37 clubfeet with a continuous passive movement (CPM) machine rather than by surgical release and founded that CPM treatment improved equinus and varus in all cases and thus can eliminate the need for surgery in mild clubfeet, and delay surgery in more severe cases.

Plaster Cast Treatment

The first recorded use of plaster casts is that by Guerin in 1836. Kite introduced the wedge plaster cast method.[27-29] He corrected the deformity by slow conservative manipulation without anesthesia and then applying a long leg corrected position. A wedge of plaster directed dorsolaterally was removed and the foot abducted to close the wedge. Once the adduction and inversion were corrected he removed a wedge from dorsal aspect of ankle and corrected the equinus. Later he realized that when a cast is wedged in abduction only forefoot is corrected and switched to change of plaster every time baby comes to correct both adduction and equinus. He continued the meticulous clubfoot cast application and molding correcting each component of the deformity separately instead of simultaneously. He was able to correct the cavus and to avoid foot pronation, but correcting the heel varus took many casts. He recommended getting all the correction by abducting the foot at the midtarsal joint with the thumb pressing on the lateral side of the foot near the calcaneocuboid joint. However, by abducting the forefoot

against pressure at the calcaneocuboid joint the abduction of the calcaneus is blocked thereby interfering with the correction of the heel varus. Therefore, it took many months and cast changes to slowly correct the heel varus and obtain a plantigrade foot.

Fripp also advocated plaster treatment and found good results with it.[30] Reinmann used thermoplastic splints. Every time the patient visited the clinic the splint was heated and reapplied after over correcting it.[31]

Ignacio V. Ponseti (1963) described his method of cast application for conservative management of clubfoot.[32] According to Ponseti, all components must be corrected simultaneously but for equinus which takes place at ankle joint must be corrected last. Cavus is corrected first by supinating the forefoot with direct pressure under the first metatarsal. In majority of cases the cavus component is usually corrected by one cast. The hindfoot varus, fore foot adduction are then simultaneously corrected in the manipulation. The calcaneus cannot be everted unless it is abducted. Ponseti maintained the foot in cast for 5–7 days and the deformity can be gradually corrected with further manipulations in 5–6 casts. Later in 1980 in a long term follow-up of 10–25 year he published the functional results in which he found 89% satisfactory results by people treated with his plaster technique.[33] Today multiple studies from different centers around the world have shown the effectiveness of this method with low revision and complication rates.[34-37] However this method requires 2–4 years of intensive follow-up. Non compliance with the brace is a common problem and frequently lead to relapse.[38] To avoid this problem a new dynamic foot abduction orthosis is increasingly prefeered over Dennis brown foot abduction brace.[39] In developing countries steenbeek brace is popular, since it is cheap and has shown equally good results.[40] The patient characteristics at the time of presentation, such as the severity of the initial clubfeet deformity, previous treatment and the age at the initiation of treatment are important factors which predict the outcome after use of the Ponseti method for the treatment of idiopathic clubfeet.

Percutaneous achilles tenotomy lengthens the achilles tendon to help correct residual equinus. It reduces treatment duration, risk of recurrence, talarflattening ("nut-cracker" effect) or convex foot and the number of surgical releases required.[41,42]

Ponseti gave treatment of heel varus in complex clubfeet in which he says hind foot is abducted with counter pressure applied not only to the talar head but also to the lateral malleolus. The forefoot should not be abducted beyond its normal alignment. Once the heel varus is corrected, the flexed forefoot and the equinus are corrected simultaneously by forcefully dorsiflexing the metatarsals with both thumbs while applying a plaster cast reinforced by a posterior slab.[43]

Surgical Procedures

Surgery for clubfoot in today's scenario should only be considered when conservative management has failed. If surgery is necessary, most surgeons would do it only after 9–12 months. The effect of plaster cast and strapping become clear by this age. This also allows proper visualization of structures and prevents excessive scarring which is common with neonate surgery.[44] One of the common approach used is "*a la carte*" approach where only those structures are released which are required to obtain correction.[45] Surgical intervention should be delayed if the foot continues to repond to conservative management, however small improvement it may be Phelps a New York based orthopedist in 1890 described one stage medical plantar soft tissue release with tendon lengthening.[46]

Turco[1] (1971) performed a one stage posterior medial soft tissue release with internal fixation of talonavicular joint with a percutaneous Kirschner wire. He later in 1979 concluded that the best result of his operation was in age-group of 1–3 year.[47] Mckay advocated a more radial circumferential soft tissue release on medial, lateral, posterior and planar aspect of foot at an early age to obtain good results.[17,18] The Cincinnati incision affords excellent exposure both posterolaterally and posteromedially. However, skin closure at the conclusion of surgery at times is difficult.[48] Use of soft tissue expander before operation for clubfoot may help in primary skin closure after surgery for clubfoot in children would be difficult.[49] They are especially helpful before extensive surgery for clubfoot especially in revision procedures and for older children.

The 'Complete Subtalar Release consisted of a standard posteromedial release with additional release of the talonavicular joint, the calcaneofibular joint and the interosseus ligament. A greater degree of correction is obtained with the Complete Subtalar Release (both clinically and radiologically), superior alignment of the foot and knee is achieved, and the incidence of complication is no greater.[50] The Complete Subtalar Release produced significantly greater correction of the angles that were used to measure equinus and varus angulation and talonvicular subluxation.[51] With this procedure there is tendency to over correction unless the bones were precisely repositioned at the end of the procedure. The procedure is indicated when the foot is at least 8 cm long, the patient is at less than 4-years-old, and there is talonavicular subluxation or varus deformity or both, that has resisted non operative treatment. Contraindications include flat top talus and severly restricted planter flexion due to contracture of the ankle. Relative contra indications include rocker bottom deformity and marked pes planus.

In the child whose tarsal and metatarsal bones have become deformed and resist correction, a combination of soft tissue release and various bony procedures are considered.[52-53] In older children between five to eight years of age, a combination of soft tissue release and Lichtblau procedure is recommended.[54] In those older than nine years of age, the lateral column of the foot is shortened and stabilized by calcaneocuboid resection and fusion.[55] A combination of soft tissue release with a medial opening wedge osteotomy of calcaneum and insertion of a bony wedge is also described.[56]

In general, bony procedures are rarely if ever, indicated in the infant and young child as these will disturb the normal growth and development of the foot. In a skeletally mature foot (more than ten years old), osteotomy of the os calcis, tarsal reconstruction and triple arthrodesis are required as salvage procedures.[57] Metatarsal osteotomy at their bases will correct the varus footfoot, Dwyers osteotomy of the calcaneus corrects hindfoot varus and medial rotation osteotomy of the tibia may be indicated to correct severe lateral rotational malalignment of the tibia and fibula.[58,59] Occasionally, a talectomy is performed.

External fixators

Even with surgical management, some foot are either resistant to correction or relapse. This may require repeated surgery in about 20% of cases. Repeated surgery may cause small, stiff and painful foot. External fixators are very useful in such cases. This method added a new dimension to the ability to solve 3 dimensional problems previously more difficult or less safe with conventional technique.

Grill and Frankie used Ilizarov fixator for treating relapsed or neglected clubfoot. They achieved plantigrade foot with satisfactory clinical and radiological results in all 10 feet treated.[60] Gupta and Bither used Ilizarov fixator in treating 15 patients (16 feet), achieving plantigrade foot in all except one with good or excellent appearance at follows up.[61] They supported the use of iilizarov as the benefits offered outweigh the risk of associated complications, making this potentially cumbersome appartus a useful tool in the armantarium to treat relapsed clubfeet following soft tissue release. However, use of this apparatus should be reserved for children above 3 years of age.

Dr. Joshi designed his J.E.S.S (Joshi's External Stabilizing System), an external fixator system for correction of deformities in clubfoot.[62] It was based on the principle of Differential Fraction Distraction laid down by Ilizarov. He used this fixator system successfully in all presentation of clubfoot in children aged 3 months to adulthood. Subsequently other authors have shown good result with jess fixator in deformities remain uncorrected by plaster-of-paris casts and manipulation as well as recurrent clubfoot [Figure 2].[63,64] We also had a good experience with this fixator in resistant and neglected clubfoot and give as good result as complete subtalar release.[51]

Conclusion

Clubfoot is one of the frequent conditions encountered in the practice of orthopaedic surgeon. Conservative management with ponseti technique has shown excellent result and decreased the requirement of surgery drastically. Surgery should be done as "*a la carte approach*". Grading system is simplistic and does not adequately assess the severity. Ilizarov fixator has a definitive role in relapsed, recurrent clubfoot. Jess fixator may safely be used in small childrens (less than 3 yrs) with equally good result as Ilizarov.

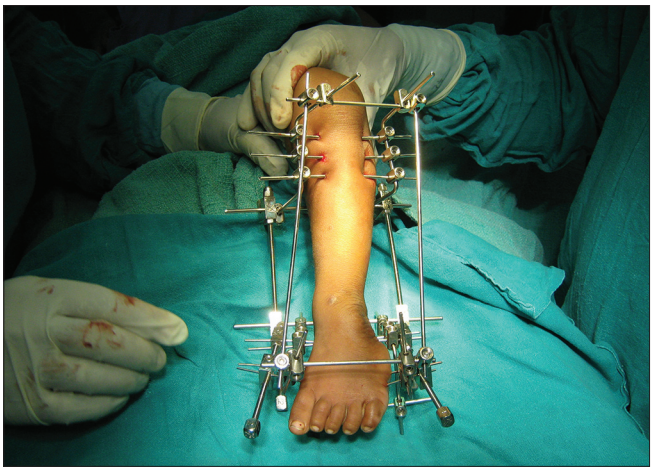


Figure 2: Resistant clubfoot managed with Ilizarov fixator

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