Management Of Non-Idiopathic Clubfeet

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Abstract

Background: The management of idiopathic clubfeet with Ponseti method of casting has been well outlined in literature. However, management of non-idiopathic clubfeet, like the ones associated with arthrogryposis or myelomeningocele, has been challenging. Various treatment modalities including casting, soft tissue releases, bony procedures and talcetomy have been attempted, either in isolation or in combination, with variable results. The aim of the current study was to perform a comprehensive review of the literature related to the classification and management of non-idiopathic clubfeet. Recent literature has shown benefits of a trial of Ponseti method of casting for all non-idiopathic clubfeet. Most of these clubfeet, however, would require surgical treatment due to either insufficient correction or recurrence. We attempt to provide, based on the review of literature and our experience, a treatment algorithm to help with the management of complex, non-idiopathic clubfeet.

Keywords: Non-idiopathic clubfoot, CTEV, Ponseti, arthrogryposis, myelomeningocele.

Introduction

With the success of Ponseti casting technique, the management of most congenital talipes equinovarus deformity or idiopathic clubfeet, has been standardised. The management of non-idiopathic clubfeet, however, continues to be challenging. These non-idiopathic clubfeet are associated with many neuromuscular and congenital syndromes, the most common of which are arthrogryposis and myelomeningocele (Table 1). The diagnosis of the underlying condition associated with clubfeet is important as it can affect management and help predict prognosis. These feet have been considered more rigid with a higher rate of recurrence compared to idiopathic clubfeet. In the past, they have been considered to be resistant to non-operative treatment and have been treated with multiple surgical procedures including extensive soft-tissue releases and radical bony surgeries.

The purpose of this study is to perform a comprehensive review of the existing literature, review our experience and provide guidelines for management of non-idiopathic clubfeet. The article would first describe the distinct features of common conditions associated with non-idiopathic clubfeet and would discuss the need for special considerations, if any, in their orthopaedic management. This would be followed by general principles and treatment algorithm for management of non-idiopathic clubfeet.

ARTHROGRYPOSIS

Arthrogryposis is derived from the Greek words, joint (arthron) and hooked (grupon). It is a condition of non-progressive contractures of two or more joints often resulting from fetal akinesia. Arthrogryposis is not a specific diagnosis but a clinical finding resulting from fetal akinesia. Arthrogryposis is not a specific diagnosis but a clinical finding resulting from fetal akinesia. Arthrogryposis is not a specific diagnosis but a clinical finding resulting from fetal akinesia. Arthrogryposis is not a specific diagnosis but a clinical finding resulting from fetal akinesia.

It is a condition of non-progressive contractures of two or more joints often resulting from fetal akinesia. Arthrogryposis is not a specific diagnosis but a clinical finding characteristic of more than 300 disorders. Clubfeet in arthrogryposis are usually stiffer and the deformities are more severe than idiopathic clubfeet. The skin is thin and smooth, muscles are pale and thin and often replaced by fat and fibrous tissue, ligaments and capsular tissues are thickened and tight, and articular surfaces are abnormally shaped with absent skin creases around the joint. Deformities are usually bilateral but rarely symmetrical. Deformities are not progressive but untreated deformities become more rigid with growth. There is a resistance to correction and also a tendency to recur. Of the various types of arthrogryposis, clubfeet in amyoplasia have severe deformities, are stiffer, more rigid and more resistant to treatment than in distal arthrogryposis.

Concomitant contractures of the knee and/or hip pose a unique problem to clubfeet management in arthrogryposis. The knee could have a fixed deformity in extension (subluxated) or in flexion. During casting for clubfeet, a knee with an extension deformity can cause slippage of the cast and hence loss of correction. Simultaneous gradual correction of the knee and the foot in cast could be attempted. Failure of closed treatment of knee extension contracture may require surgical intervention. Simultaneous surgery of the knee (e.g. quadricepsplasty) and cast treatment of the clubfoot pose a challenge as well, as the knee would require early postoperative mobilization and this may compromise the correction of the foot. When knee has flexion contracture, soft tissue stretching/release or corrective extension osteotomy could be done after correction of clubfoot.

Hip joints in arthrogryposis are often subluxated or dislocated. These dislocations are often teratogenic and can rarely be treated.
Intravenous access may be difficult due to presence of tense skin and less subcutaneous tissue. Intra-operative positioning may be difficult due to multiple contractures. Perioperative respiratory issues and malignant hyperthermia should be considered in these children.

Diastrophic Dysplasia
Diastrophic dysplasia is an autosomal recessive disorder characterised by short stature, multiple joint contractures, cervical spine scoliosis, hitch hiker’s thumb, cauliflower ears and severe clubfoot deformity. Similar to arthrogryposis, clubfeet are usually rigid, difficult to treat and may be co-existent with hip and knee contractures. Surgical correction pose difficulty in these feet due to distorted anatomy. Recurrences after initial treatment are common and may require extensive releases and/or tallectomy.

Freeman-Sheldon Syndrome
This is a type of distal arthrogryposis characterised by ‘whistling mouth facies’, short stature, hand deformities in form of ulnar deviation and thumb in palm and rigid clubfeet.

Neurogenic Clubfoot
Clubfeet occur in 30-50% of spina bifida patients. It is most commonly seen in lumbar lesions at the level of L3-L4 and above. The level of lesion should be identified as it would affect the functional motor level and hence the ambulatory capacity. In a potential walker, the aim is to have a plantigrade foot that can weight bear; in a non-walker, the aim is to have a braceable foot to rest on the footpads of a wheelchair. Presence or absence of foot sensation should be assessed. The deformity in neurogenic feet can be

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attributed to varied causes including intrauterine malposition, muscle imbalance, spasticity and/or fibrotic contractures. These feet are rigid, difficult to treat and have a high risk of recurrence. There is an increased risk of pressure sores and skin breakdown due to altered or absent sensations. Physeal injury is not uncommon with manipulations. There is a high risk of fractures in these children undergoing manipulation which might not be diagnosed early due to absence of pain. There may be delayed healing of wound after surgery.

**Mobius Syndrome**

Mobius syndrome is a rare non progressive congenital neuromuscular disorder. It is characterised by palsy of the sixth/seventh cranial nerve and malformation of orofacial structures and limbs. Typically they have absent abduction of both eyes. Clubfoot is the most common deformity of the foot. Diagnosis should be suspected when multiple cranial nerve dysfunction exist with clubfeet.

**Streeter's Dysplasia**

This is a disorder of fetal development in which the amnion separates from the chorion and forms bands which encircle limbs. Part of the limb distal to the band may be amputated, anomalous or normal. Incidence of clubfeet with constriction bands is 12-56%. These children may or may not have a neurological deficit on the involved side. Neurologic deficit has been found more commonly with zone 2 bands (band between knee and ankle) and grade 3 severity i.e. (reaching up to the level of fascia compromising distal circulation). Clubfeet with constriction bands are rigid, respond poorly to conservative treatment and often require soft tissue release. Feet with neurological deficit have been more resistant to treatment by earlier authors and require more number of surgeries than ones without a deficit.

In case the constriction band causes circulatory compromise, emergent release should be done. All other bands may be released in a staged manner after correction of clubfoot. Paralytic feet may require tendon transfer depending on the involvement.

**Down's Syndrome**

Down's syndrome is the most frequent trisomy (chromosome 21). It is characterised by hypotonia and hyperlaxity, mental retardation, congenital heart disease, atlantoaxial instability, habitual dislocation of patella and hip and genu valgum. Though typical foot deformity due to hyperlaxity has been pes planus with metatarsus adductus, occasionally soft tissue contracture around the ankle, subtalar and mid-tarsal joints present as clubfoot. In spite of generalised ligament laxity, these feet are resistant to nonoperative treatment and often require surgical treatment. These feet, however have a tendency for overcorrection and calcaneovalgus deformity following treatment due to generalised hyperlaxity. Pre-operative evaluation should include cardiac evaluation and cervical spine x-rays to screen for cervical spine instability.

**Larsen's Syndrome**

Larsen's syndrome is a rare congenital
connective tissue disorder caused by mutation in gene encoding filamin B (FLNB). It has genetic heterogeneity with both autosomal dominant and autosomal recessive patterns. It is characterised by flat facies, cardiac defects and varied musculoskeletal issues that include joint hypermobility, vertebral anomalies, recalcitrant dislocations of multiple major joints and clubfeet. Early detection of the syndrome is important for management decisions and prognostic counselling. Management of cervical spine instability should be a priority in these children. When hips, knees and feet are involved, treatment of the knee should be a priority. Clubfeet in these children should be treated by conservative method soon after birth and surgical treatment should be postponed until knee dislocation is corrected. Due to hyperlaxity, care should be taken to prevent overcorrection of these feet as they can often develop calcaneovalgus deformity. Hip dislocation, if bilateral, should be managed by careful neglect due to poor results of surgical treatment.

Preoperative assessment should include thorough respiratory, cardiac and neurologic evaluation. Lateral cervical spine x-rays in flexion and extension should be mandatory prior to anaesthesia to evaluate cervical spine instability which is common in this syndrome.

**Principles of Treatment**

Conventional non-operative treatment has been considered ineffective in cases of non-idiopathic clubfeet. Serial manipulations and casting of these feet often resulted in incomplete correction and frequent recurrences. Previous literature thus focused on the operative treatment of these feet, with most feet requiring multiple procedures. Soft tissue releases, however, also resulted in recurrences and required brace wear till skeletal maturity. Due to high incidence of failure of soft tissue releases, talectomy has been considered to be the procedure of choice as a primary surgery as well as in cases of recurrence.

Ponseti popularised the method of serial casting to correct idiopathic clubfeet with excellent results. In non-idiopathic feet Ponseti method decreases the severity of deformity and hence decreases the need for extensive surgery. Trial of Ponseti method should be the first line of treatment for all clubfeet, irrespective of their etiology. In recent literature, short term results of Ponseti method for correction of non-idiopathic clubfeet have been encouraging (Table 2). Most of these feet were Dimaglio grade IV or Pirani grade 4 or above. More number of casts were required than idiopathic feet, however good initial correction was achieved in most non-idiopathic feet. Arthrogrypotic feet being more rigid than idiopathic, complete correction as in idiopathic foot might not be achieved. Usually, 40–45 degree of abduction and 5 degree dorsiflexion could be achieved. The recurrence rate has been higher, but the deformity tends to improve with re-casting in most cases. Moroney et al reported that after Ponseti casting, the Pirani score in feet which required surgery reduced from 5.1 to 3.7. Thus casting, even if not successful, reduced the morbidity of future surgery.

Severity in non-idiopathic clubfeet has not been classified in literature probably because of low incidence. Most often these feet are either not graded or are graded as per Dimaglio score or Pirani scores which have been described for idiopathic clubfeet. Since most non-idiopathic clubfeet are rigid and a trial of Ponseti method has been justified for most patients, we have classified non-idiopathic feet on basis of response to Ponseti casting to standardize management protocols (Table 3).

A treatment algorithm for non-idiopathic clubfeet has been outlined (Fig 1). After initial correction with Ponseti method, the main factor leading to relapse has been non-compliance of brace. The thick rigid capsule is incapable of stretching with growth and results in recurrence despite adequate bracing. Ill-fitting brace could contribute to non-compliance. In non-idiopathic clubfeet, it is important to maintain the correction obtained which might be less than that obtained in idiopathic feet. The brace may have to be modified to the degree of correction achieved i.e. 40 – 45 degrees of abduction and 5 degrees of dorsiflexion instead of 70 degrees and 20 degrees, respectively, as in idiopathic feet. The brace modification can improve compliance. As and when flexibility improves, more bend can be added to the brace as required. Often due to concomitant hip and knee contractures, Ankle Foot Orthosis (AFO) should be considered instead of Foot Abduction Brace (FAB) for better fit and compliance. Brace might have to be modified in case of a congenital constriction band with an amputation of the other leg – by using an AFO instead of an abduction bar. A brace in the form of an AFO should be generally worn till skeletal maturity.

If there has been partial correction with Ponseti method, surgery may be required. When no visible change is seen between two consecutive casts, casting should be stopped and surgery should be considered. The procedure of choice in such cases has been controversial. It would depend on the severity of the residual deformity, the age of child and ambulatory potential. Not all cases require extensive surgery. When only residual equinus and/or forefoot varus persists, a heel cord lengthening and anterior tibialis transfer can give good results (Fig 2). With more severe residual deformities, some authors prefer extensive soft tissue releases, reserving talectomy as a salvage procedure for the most severe of all deformities. Some favour talectomy primarily stating the higher failure of soft tissue releases. Extensive soft releases i.e. posteromedial and lateral have shown good results when performed by 1-2...
years of age to preserve articular congruity. If the child presents after 2 years of age or does not have ambulatory potential then a primary takedown should be preferred to avoid recurrence and repeat surgery. Often only a takedown is not enough to correct the deformity and should be accompanied by soft tissue release (Fig 3, 4). Cincinnati incision adequately provides the required extensive visualization for dissection medially, posteriorly and laterally.

In very severe rigid clubfeet, correction might not be achieved with Ponseti casting. Primary takedown with extensive soft tissue releases should be considered in these children for best results.

Soft tissue releases have shown good results when done at 7-8 months of age. When soft tissue releases are done, care should be taken to do as complete of a release as possible with excision of 1-2 cm of tendon to prevent recurrence. Soft tissue releases often correct the forefoot varus, though in cases of severe rigid equinus, the equinus is often not corrected even after release of tendo Achilles and posterior capsular releases. Complete excision of the talus provides enough laxity for correction of the equinus and varus deformity. Takedown however might not correct the forefoot deformity and additional procedure in the form of an osteotomy might be required for such severe deformities. Failure of soft tissue releases could be either due to inadequate initial correction because of tight skin and soft tissues on the medial side or due to a relapse after cast removal due to the wide medial gap created secondary to the severe deformity. However, soft tissue release is preferred in younger patients as later salvage in the form of takedown can be done in cases of failure. When presenting at a later age, the adaptive changes in joints preferably warrant a primary bony surgery to prevent failure and recurrence.

Takedown has shown satisfactory outcomes after short term as well as long term follow up. In younger children, however, less radical surgery may be preferred before takedown. If a child presents late, takedown could be performed primarily with the best results achieved when done at 1-5 years of age. Failure of takedown may be due to poor technique, due to either partial excision or incorrect placement of the talus under the calcaneus allowing for a later posterior drift and recurrence of equinus. A failure after poorly performed takedown, could be difficult to treat and may require a corrective osteotomy.

Triple arthrodesis is recommended if child presents after 10-12 years of age as earlier surgery may limit the growth of that foot. Non-idiopathic feet have a higher rate of recurrence and hence could require secondary and tertiary procedures. Recurrence within six months of previous surgery suggest incomplete correction and hence complete correction at the initial surgery should be attempted. In case of recurrence after extensive soft tissue releases, takedown should be considered instead of revision of soft tissue releases due to scar tissue. Ilizarov external fixator can also be used primarily or in case of secondary or tertiary recurrence.

Similar to idiopathic clubfeet, the primary aim of management of non-idiopathic clubfeet should be to achieve a painless, planigrade foot with minimal number of procedures as possible. When possible, this should be achieved before walking age to prevent adaptive changes in bone. The evaluation of result should be based on the final outcome as well as the number of procedures required to achieve it.

Summary
In recent literature, Ponseti method of casting has shown promising results in non-idiopathic feet albeit in short term. Although it may not avoid surgery in these patients in long term, it definitely reduces the severity of deformity and hence the magnitude of surgery required. It should be the first line of approach in all clubfeet, irrespective of their etiology. Later, depending on the amount of correction achieved, rigidity of the feet, walking potential and age of the patient, decision related to surgical treatment should be made. Attempt to ensure complete correction should be made in the first surgery itself to avoid recurrence. This often might require extensive soft tissue release with takedown with or without midfoot osteotomy to correct forefoot varus. Patients should be followed long-term to identify and treat recurrence and family should be strongly counselled about the importance of such follow-up visits.

References


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