



International Journal of Orthopaedics Sciences

E-ISSN: 2395-1958
P-ISSN: 2706-6630
IJOS 2020; 6(1): 1281-1285
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www.orthopaper.com
Received: 24-11-2019
Accepted: 26-12-2019

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Importance of operative intervention in the treatment of clubfoot associated with congenital constriction Band syndrome

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DOI: <https://doi.org/10.22271/ortho.2020.v6.i1q.1997>

Abstract

Background: Congenital talipes equino-varus is the most common congenital orthopaedic condition requiring intensive treatment. It has been associated with neuromuscular diseases and syndromes and specifically, CTEV associated with congenital constriction band syndrome forms the resistant variety of CTEV. This most often requires operative intervention in the form of postero-medial soft tissue release, JESS Fixator and Ilizarov fixators.

Material and Methods: 10 patients of less than 2 years with resistant CTEV associated with congenital constriction band syndrome, who required operative intervention were operated with JESS fixator at Govt. Medical College and Rajindra hospital, Patiala between august 2017 to august 2019. Following the correction, the assembly was held in the static position for the same period to allow for soft tissue maturation in the elongation position. Removal of the whole assembly was followed by well moulded above knee plaster cast in maximum correction for the same period followed by CTEV corrective shoes for 5 years to maintain correction and prevent recurrence.

Results: Patients were evaluated using functional rating system for clubfoot, hospital for joint diseases, orthopaedic institute as excellent, good, fair and poor. 7 feet (70%) were graded as excellent, 2 feet (20%) were graded as good, 1 feet (10%) as fair and 1 feet (10%) as poor. In this study, 1 feet (10%) had persistent oedema, 2 feet (20%) had flexion contractures of toes, 1 feet (10%) had tibial loosening of pin, 1 feet (10%) had rocker bottom foot and none of the feet had skin necrosis.

Conclusion: Congenital talipes equino-varus in a child of congenital constriction band syndrome forms the resistant variety of of CTEV, in which operative treatment is the mainstay of treatment. Joshi's external stabilisation system has proved to be an good treatment option in such cases.

Keywords: CTEV, Congenital constriction band syndrome, JESS

Introduction

Congenital talipes equinovarus (commonly referred to as clubfoot) is probably the most common (1 to 2 in 1000 live births) congenital orthopaedic condition requiring intensive treatment. It most likely represents congenital dysplasia of all musculo-skeletal tissues (musculotendinous, ligamentous, osteo-articular and neurovascular structures) distal to the knee. This conclusion is based on multiple investigators' observations of different abnormal anatomic findings and functional outcome of patients believed to have received optimal nonoperative or operative treatment. Clubfoot has been associated with neuromuscular diseases and syndromes. Therefore, an underlying neuromuscular or syndromic/dysmorphic etiology for all "idiopathic" clubfeet has always been suspected. Arthrogryposis diastrophic dysplasia, Streeter dysplasia (constriction band syndrome), Freeman Sheldon syndrome, Mobius syndrome and other conditions resulting from chromosomal deletions are just a few of the more recognisable systemic conditions with associated clubfeet. In contrast, idiopathic clubfoot is commonly due to a single musculoskeletal deformity in an otherwise normal infant. Many other theories on the aetiology of clubfoot have been proposed, including an arrest in embryonic development. A second hypothesis about the etiology of clubfoot proposes a retractive fibrotic response, not unlike Dupuytren's contracture, as a primary factor. Congenital constriction band syndrome, also known as congenital peripheral constriction rings originating

from soft tissues of the leg or as a Streeter band is occasionally observed in lower extremity^[1]. It is characterised with compression in the soft tissue, usually involving the deep fascia surrounding the leg at the time of birth. Lymphatic vessels and superficial vascular circulation are usually partially obstructed. At the distal side of the constriction, oedema and cyanosis could be seen. At the region of the constriction, fracture of tibia and fibula and foot deformities like clubfoot can be observed. Unlike congenital pseudoarthrosis, the fractures can be healed by means of releasing the constriction without need of an operation. According to Paterson's^[2], constriction ring can be classified into 4 subgroups:-

- 1) Simple constriction ring
- 2) Constriction ring with the deformity of distal part
- 3) Constricting with a fusion of distal parts
- 4) Complete intrauterine amputation.

Hennigan *et al.* conducted a study for resistant CTEV associated with constriction band syndrome in which results were graded as good, fair or poor, based on pain, residual deformity, need for special shoes, inserts or braces, presence or absence of calluses and limitation of activities. Children who had no functional limitations, no pain and little or no residual deformity, were considered to have a good result. Results were considered fair, if the child had occasional pain, mild to moderate residual deformity or limitations with strenuous activities or sports. Poor results were seen in patients who required amputation, with severe pain, with severe limitations in non-strenuous activities, or residual weakness, requiring a brace for a ambulation^[3]. Congenital constriction ring syndrome, also known as Streeter's bands or dysplasia or amniotic band syndrome is characterised by Syndactyly, hypoplasia, brachydactyly, symphalangism, symbrachydactyly, clubfoot, cleft lip, cleft palate, cranial defects. The prevalence of clubfeet in patients of congenital constriction bands ranges from 12 to 56%.^[4, 5, 6] Cowel and Hensinger in their series of 14 patients reported clubfeet among 25 patients with congenital constriction band syndrome^[7]

The location of constriction bands are divided into 4 zones:-

- Zone 1 - Bands occur between the greater trochanter and the knee
- Zone 2 - Bands occur between the knee and ankle
- Zone 3 - Bands occur between the ankle and metatarsophalangeal joints
- Zone 4 - Bands are limited to toes

Severity of bands is also considered. Grade 1 bands are subcutaneous but not to the level of fascia. Grade 2 bands to the level of fascia and do not compromise the circulation to the distal extremity. Grade 3 bands are to the level of fascia, such that lymphoedema or circulatory compromise necessitates surgical release. Grade 4 bands include all congenital amputations^[8].

Clubfoot associated with congenital constriction ring syndrome is mostly resistant to nonoperative treatment. Nowadays, most of the clubfoot patients are treated with serial casting with standard Ponsetti technique and results are promising. But the clubfoot associated with congenital ring syndrome, almost always requires operative intervention. This study has been undertaken to evaluate the role of operative intervention in all the patients of clubfoot associated with congenital constriction band syndrome.

Methods

10 patients in the age group of less than two years of either sex presenting with the clubfoot associated with congenital band syndrome attending the OPD /emergency of government medical College and Rajindra hospital, Patiala from August 2017 to August 2019.

Inclusion criteria

- a) Age less than two years
- b) Failed Conservative management
- c) Constriction ring with the deformity of distal part and without vascular compromise.

Exclusion criteria

- a) Age more than two years
- b) Constriction ring syndrome with intrauterine amputation or with vascular compromise.

Procedure

Photographic evaluation and radiological evaluation was done for all the patients of congenital talipes equinovarus associated with congenital constriction band syndrome (Zone 2). The patients for whom nonoperative treatment had failed, only those patients were included in the study. Preoperative clinical assessment of the patients was done along with the radiological assessment (Fig 1). The angles calculated are:-

- Talo- calcaneal angle (Antero- posterior)
- Talo- calcaneal angle (Lateral)
- Talocalcaneal index
- Tibio- calcaneal angle (Stress lateral)
- Tibio -metatarsal angle



Fig 1: Pre-operative condition of club foot

Patient is placed in supine position under general anesthesia. A pneumatic tourniquet was applied, but inflated only if required. All the patients were treated by Joshi's External Stabilisation System (JESS). The procedure was divided into 3 main steps:-

- 1) Insertion of K-Wires.
- 2) The creation of holds
- 3) The connections between the holds

Insertion of K- Wires: Wires are inserted at 3 sites. The upper one 3rd of the tibia, distal part of the metatarsals and the calcaneum. Two parallel k-wires are passed from lateral to medial side, a finger breadth distal to the tibial tuberosity. One transfixing k-wire is passed from the 5th to 1st metatarsal at the level of the neck. The transverse arch of the foot was not flattened in an attempt to impale all the metatarsals. 2nd k-wire was passed from the lateral side at the level of the metatarsals shaft. This wire engaged both the cortices of the lateral three metatarsals. The distance between the 2 wires was kept to 2 to 3 mm more than the distance between the 2 holes of the distractor. Similarly, another k-wire was

introduced from the medial aspect to engage both cortices of the 1st and 2nd metatarsals. Then a k-wire was passed from the tuber of calcaneum perpendicular to the long axis of the bone from medial to the lateral side. 2nd k-wire was passed, distal and parallel to the 1st wire using the distractor block as a guide with the 1st wire.

Creation of holds: After k-wire insertion, 3 holds were constructed in the following sequence after pre-stressing the k-wires.

- Tibial hold
- Metatarsal hold
- Calcaneal hold

Connection between the holds:-

Finally, the 3 holds were connected in the following manner.

- Tibio- Metatarsal connection
- Calcaneo metatarsal connection
- Tibio-calcaneal connection

After the final construct is made, the ends of wires were cut and bent. (Fig 2)

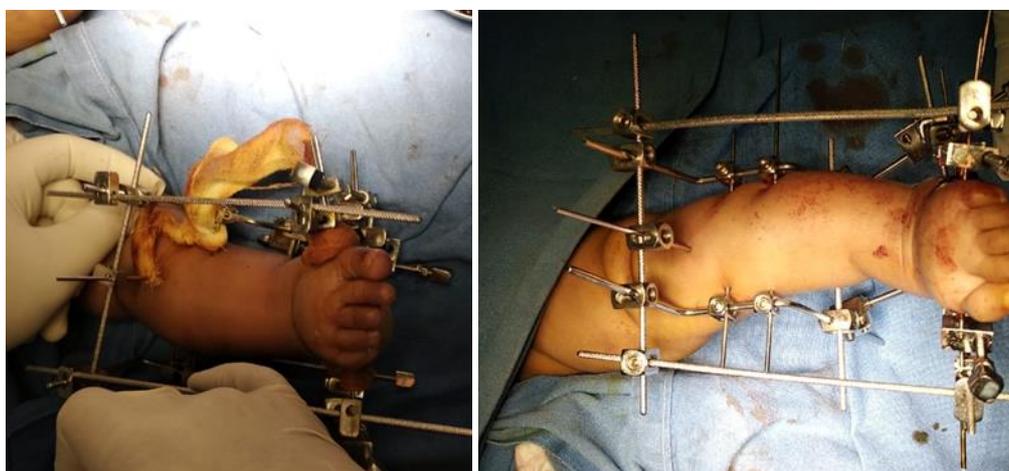


Fig 2: Post operative condition of the foot

Post-operative management:- The pin sites were covered with a dry dressing and frame was encased with a thin layer of soft foam or a cardboard wrap, to prevent injury from protruding wires. On the 3rd postoperative day, differential fractional calcaneometatarsal distraction and tibio-calcaneal distraction on the medial side were started at twice the rate than that on the lateral side (medial – 0.5 mm every 12 h; lateral – 0.25 mm every 12 h). The above explained distraction was very clearly demonstrated to the patient's attendant and supervised for 2 days. 7 days following the surgery, the patient was fit enough to be discharged and was advised for a regular follow-up at weekly intervals for 6 weeks to look for a progressive correction of the deformity, persistent edema, rule out pin tract infections, and tighten the loosened link joints. Afterwards they were required to come monthly for 3 months and then biannually.

The dressings of the pin tract were changed regularly and all link joints were tightened every week or whenever required. Following the correction, the assembly was held in the static position for the same period to allow soft tissue maturation in the elongation position. Single-stage removal of the whole assembly was done under mild sedation and a well-molded above knee plaster cast was applied in maximum correction for the same period it took for correction. Full correction of

forefoot adduction, varus, and equinus was achieved usually at the end of 6 weeks. For all patients, CTEV corrective shoes were advised for 5 years to maintain correction and prevent recurrence. Postoperatively, patients were evaluated using Functional rating system for clubfoot, Hospital for Joint Diseases, Orthopaedic Institute as excellent, good, fair and poor. Another method for evaluation is ICFSG score, where results were classified as excellent (0–5), good (5–15), fair (16–30), and poor (>30) at follow-up intervals of 3 and 6 months. The parents care and compliance played an important role in the success of this procedure. This is a 60- point international clubfoot study group score and is based on foot morphology (12 points), radiologic (12 points) and functional assessment (36 points) testing the eight main leg muscles. It is however only moderately adapted to idiopathic clubfoot.^[9]

Results

The age of the patients was less than 2 years, with an average of 1.1 years. Out of 10 feet, 6 (60%) were males and 4 (40%) were females. There were 6 feet (60%), which were unilateral and 4 feet (40%) were bilateral cases. All that 10 feet (100%) were resistant cases and none of them responded to serial casting by Ponsetti method. In this study, all the 10 feet treated by Jess, none of the foot had skin necrosis, 1 foot

(10%) had persistent oedema, 2 feet (20%) had flexion contractures of toes, and 1 foot (10%) had loosening of the tibial pin, which was changed in general anesthesia and assembly was reconnected. 1 foot (10%) had rocker bottom foot, prior to application of JESS Fixator, which was treated accordingly.

Graph of post operative complications is graph 1

7 feet (70%) were graded excellent, 2 feet (20%) were good, 1 foot (10%) was graded fair, 1 foot (10%) was graded poor, as graded by the Hospital for Joint Diseases Orthopaedic Institute, Functional rating system for clubfoot. Radiological assessment was done using a talo-calcaneal index and it was compared with other case series showing good radiological correction. As in table 1.

Table 1: Average calculated preoperative and post-operative talo-calcaneal parameters

Pre operative			Post-operative		
Talo-calcaneal angle AP View	Lateral view	Talo-Calcaneal Index	Talo-calcaneal angle AP View	Lateral view	Talo-Calcaneal Index
13 degree	18 degree	29 degree	23 degree	30 degree	53 degree

Table 2: Comparison of results from various studies

Series	Pre-operative (in degrees)			Post-operative (in degrees)		
	AP	Lateral	Index	AP	Lateral	Index
Graham and dent ^[10]	-	-	-	25	20	45
Lau <i>et al.</i> ^[11]	-	-	-	16	22	38
Stromqvist <i>et al.</i> ^[12]	-	-	-	15	24	39
Ryppy and sairanen ^[13]	-	-	-	26	30	56
Present study	13	18	29	23	30	53

Discussion

Various forms of treatment are available for congenital talipes equinovarus, including serial casting with ponsetti method. Functional method, which is implemented during the 1st months of life, and is based on daily physiotherapy, decoaptation, of the navicular from the medial malleolus, correction of forefoot adduction and calcaneal varus, CPB derotation and then talus re-integration with equinus correction associated to evertor muscle stimulation. Percutaneous Achilles tendon tenotomy, that helps to correct the residual equinus. It reduces treatment duration, risk of recurrence, Talar flattening (nut cracker effect) or convex foot and the number of surgical releases required ^[14]. Postero-medial soft tissue release used to do the lengthening of the tendons and sectioning the aponeurosis, which hinder reduction soft tissue release corrects Tibio -Tarsal & Subtalar equinus, CPB adduction and medio-tarsal adduction. This is not a light surgery and should not be performed before one year of age and should respect the sub-talar structures. ^[15] Beginning with the posterior release it moves on to the medial part of the foot. The Talonavicular joint is temporarily pinned in the maximum reduction to avoid navicular dorsal subluxation. Correction is achieved in 75 to 80% of cases with the 20 to 40% recurrence, which may require heavy revision surgery ^[16]. Joshi's external stabilisation system (JESS) is another form of treatment based on controlled differential distraction and is a simple, versatile light fixator system with the tremendous potential. ^[17] It includes a bloodless, semi- invasive procedure that avoids fibrous tissue formation, further shortening (unlike bony procedures), post-operative complications and scarring. As all the cases of congenital Talipes equino- varus associated with congenital constriction band syndrome almost always needs operative intervention and as all the cases in this study were less than 2 years, so we operated all the patients with joshi's external stabilisation system (JESS). Another Form of operative treatment available is Ilizarov fixators. But JESS fixators are lighter in weight, shorter, cheaper, and have an easier application than Ilizarov's fixators. The results of our study employing JESS proved to be better than the outcome of the study of Ilizarov's fixator conducted by Fernando where only 58.3% of cases showed excellent results ^[18] and the study

conducted by Bradish and Noor where only 47% of cases were successful ^[19].

Conclusion:

Congenital talipes equinovarus in a child of congenital constriction band syndrome forms the resistant variety of ctev in which operative treatment forms the mainstay of treatment. Joshi's external stabilisation system (JESS) has proved to be an good treatment option in such cases which is simple, versatile and light fixator system with the tremendous potential. JESS ensures proper control of all the components of correction by causing acute physiological lengthening and histiogenesis of soft tissues thereby reducing the pressure on the growing epiphysis. Differential distraction technique gives good result in children, but the results are excellent in younger children, and in those who have not undergone any previous operative procedure. All cases of CETV are not amenable to this technique, but all cases of CETV associated with the congenital constriction band syndrome should be operated and Jess has proved to be a novel technique for all those cases, although this technique has many advantages. One should not forget that injudicious and unsupervised distraction may lead to catastrophic results in the small developing foot. Long-term studies are required to accurately assess the functional outcome of treatment of clubfoot associated with the congenital constriction ring syndrome by JESS.

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