

# Arthrogryptic Club Foot

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## Abstract

**Background:** Arthrogryposis Multiplex congenita is a rare disorder, characterized by multiple joint deformities i.e. multiple congenital contractures, with shapelessly cylindrical limbs and absent skin creases.

Club foot can be the only obvious deformity of this widespread disorder.

**Objective:** To assess the most frequent recurrent deformity after extensive soft-tissue release operations for arthrogryptic club foot and its appropriate treatment regarding combined tendon transfer and bony operations.

**Methods:** A retrospective study of 14 patients with arthrogryptic club foot (28 feet), had been operated on by multiple soft tissue and bony operations and followed in a period between January (1993) till February (1999).

Both clinical assessment including goniometer use to measure the recurrent deformity, and radiological assessment, was used in this study.

**Results:** In this series, 4 patients (7 feet) had recurrent deformity, 3 patients

(5 feet) out of these 4 had recurrent forefoot adduction, 2 of them had bilateral affection. The 4<sup>th</sup> patient had bilateral recurrent varus heel.

**Conclusion:** The main problem in arthrogryptic club foot is the liability for recurrence. The most frequent recurrent deformity was forefoot adduction. Combined tendon transfer and bony operations can correct the deformity once performed at the same session forefoot adduction. All required 2<sup>nd</sup> operation, in the 1<sup>st</sup> 3 patient combined Evan's (calcaneocuboid wedge-excision) and tibialis anterior transfer operations were performed. Talectomy was performed on one foot in the 4<sup>th</sup> patient, combined with capsulectomy of the calcaneocuboid joint, while triple arthrodesis done on the other foot.

**Key words:** arthrogryptic club foot, recurrent forefoot adduction, tibialis ant. Transfer.

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## Introduction

**A**rthrogryposis multiplex congenital, is a nonprogressive syndrome characterized by deformed and rigid joints, atrophy of muscles, Cylindrical limbs with diminished skin creases, contracture of joints Capsules, dislocation of joints, esp. of the hip or knee, intact sensation and mentality. It was originally described by Otto in 1841<sup>(1)</sup>, as a discrete clinical entity. In the recent years, Swinyard and Bleck<sup>(2)</sup> suggested multiple congenital contractures as a more appropriate name for the condition, and revealed more than 150 specific clinical entities associated with the disorder .

The condition is thought to be an intrauterine neuropathy or myopathy in 10% of cases, associated with oligohydramnios. Wynne-Davies, Williams, and O'Connor studied 132 patients, and concluded that arthrogryposis is a non-genetic disease of early pregnancy associated with unfavorable intrauterine factors, including viral agent.<sup>(3)</sup>

Brown, Robson, and Sharrard<sup>(4)</sup> identified two patterns of upper extremity involvement, each corresponding to a specific level of central nervous system involvement. Most deformities present at birth were paralytic in nature and thus required surgical procedures to balance muscle activity, associated contractures of capsular and periarticular tissues render the deformities extremely difficult to correct and quite prone to recur.

Palmer, McEwen, and Bowen, have devised a program of passive stretching exercises for each contracted joint to be carried out by the parents daily followed by serial splinting with thermoplastic splints. Most orthopaedic surgeons, however, have found that any improvement occurring after a physical therapy program is transient at best, and the deformity usually recurs all too promptly.

Drummond, Sellers and Cruess<sup>(5,6)</sup>, as well as Williams<sup>(7)</sup> concluded that, recurrence of deformity is the rule in arthrogryposis, because of the dense inelastic soft tissues about the joints not properly elongate with growth. They suggested combined osteotomies and transfer the range of motion to a more useful arc are beneficial, but only at or near skeletal maturity.

Gross has recently described the technique of cancellotomy of the talus and cuboid, which rendered them hollow cartilaginous shells, followed by manipulation of the foot into the desired position of correction by crushing the treaded bones. He credits the original description of the procedure to Verebelyi in 1879; Ogston in 1902 and Kopits in 1974 reported its use in feet deformed by myelomeningocele. The procedure is performed by creating a window in the dorsal cortex of the cuboid and in the lateral cortex of the neck and the body of the talus. All cancellous bone is carefully curetted away and the deformity is corrected by manual manipulation. We have had no experience with this procedure.<sup>(17)</sup>

## Methods

Between January 1993 to February 1999, 14 patients with arthrogyrotic club foot, were seen at the out-patient clinic of the orthopaedic department of two general hospitals, i.e. Al-diwanaya, Hospital and Al-kindy Teaching hospitals. The patient included in this study fulfilling of the following criteria:

1. Availability of the patient for interview and reexamination.
2. A minimum follow-up period of 2 years.
3. All the feet in this series, were resistant club foot. They were not yielding to gentle attempts at passive correction and recognized by the atrophied legs, small inverted heel, prominent head of the talus on the dorsolateral surface of the foot, prominent cuboid and forefoot adduction. **(Figure- 1.1)**
4. All the patients in this series were subjected for a date to surgery usually at 6 months of age and more, depending on their age at the time for performance of initial surgery.
5. In our series, these cases were subjected for Turco's posteromedial release at 6 months of age,<sup>(8)</sup> plaster cast immobilization continued for a total of 3 to 4 months post-operatively, (fig. 1.2) followed by orthoses during periods of sleep. Once these children start walking outer-raised shoes were fitted for them. The final follow-up was at least one year lapse since the final correction.
6. Talectomy was performed as initial treatment together with posteromedial release, for serve cases (5 patients) in this series, and in one patient with recurrent varus heel and forefoot adduction.<sup>(9,10)</sup>
7. Every patient in this series, had antero-posterior and lateral x-ray of the foot with the foot in the plantigrade position, and after the foot subjected to operation, then post-operative x- ray taken also. **(Figure- 1.3 and 1.4).**
8. Examination of the back was performed as a routine in our cases, to exclude neurogenic club foot due to spinal dysraphism. Examination of the hip was done also to exclude cases of congenital dislocation of the hip (CDH) that may occur with bilateral club foot in female patient.

### Methods of Assessment

#### Clinical Assessment:

The objective clinical parameters include: the recognition of any significant visible recurrent deformity, i.e. forefoot adduction, varus heel, and equine deformity of the hind foot. The range of ankle planter flexion and dorsiflexion, subtalar inversion and

eversion had been observed. The power of extension and pronation of the foot had been recognized by tickling the sole of the foot especially in infants.

After walking, the gait had been recognized especially in-toeing.

The goniometer examination had been used in limited cases particularly in children beyond 2 years to assess the recurrent forefoot adduction and varus heel deformities. To calculate the angle of forefoot adduction, the goniometer is applied on the outer border of the foot with its axis opposite the styloid process of the 5<sup>th</sup> metatarsal while the body stood upright. The recorded angle is considered a deviation from the normal straight line tracing of the outer border of the foot. Normally this angle is up to 10 degrees. If more than 10 degrees indicate forefoot adduction.

To calculate a recurrent varus heel deformity, we apply the goniometer behind the foot with its axis opposite a line drawn from the heel-cord proximal to its deviation; the recorded angle of varus heel is considered as a deviation from the normal vertical line tracing of the heel cord. An angle of more than 5 degrees indicates recurrent varus heel deformity.

#### Radiological Assessment

In our series, 2 x-rays films were taken pre-operatively for every patient and in most especially those with recurrent deformity whether forefoot adduction or varus heel, 2 films were done post-operatively. An anteroposterior film with the foot plantigrade and the beam angled at 30 degrees to the vertical was performed for every patient our series. The talocalcaneal angle in this view is considered as the angle between the long axes of less than 20 degrees indicate recurrent varus heel deformity, especially if done post-operatively.<sup>(12)</sup>

A lateral x-ray with the foot in the neutral plantigrade position was done to measure the lateral talocalcaneal angle. Normally this angle ranges from 30-50 degrees.<sup>(12)</sup> an angle of less than 20 degrees indicates recurrent equine deformity of the hind foot.

To diagnose recurrent forefoot adduction, the talus-1<sup>st</sup> metatarsal angle was measured in an antero-posterior film with the foot plantigrade **(Figure 1.4)**. The normal value of this angle ranges from zero to (-20), if more than 10 degrees indicates significant forefoot adduction.

## Results

Using our functional rating system, that depends on our clinical observation, goniometer measurement, and radiological assessment. Four patients (7 feet), out of 14 patients (28 feet), had recurrent deformities. The

most frequent recurrent deformity, was forefoot adduction in 3 patients (5 feet) out of these 4 patients (7 feet). Two patients out of these 3 had bilateral recurrent forefoot adduction deformity. The 4<sup>th</sup> patient in this series, had bilateral both varus heel and forefoot adduction recurrent deformities. All these patients had stiffness of both ankle and subtalar joints with a range of ankle planter flexion 20-35 degrees, ankle dorsiflexion 10-25 degrees and subtalar inversion and eversion range 5-10 degrees.(**Table -1**).

(**Table-1**)

**Recognition of the Recurrent Deformity, Range of Ankle and Subtalar Joints Motion Based on Clinical Assessment and Goniometer Measurement**

In child,

Normal range of ankle planter flexion = 35°-45°

Normal range of ankle dorsiflexion = 25°-30°

Normal range of subtalar inversion =20°-30°

Normal range of subtalar eversion =10°-15°

The results of roentgen graphic measurement of these patients showed the mean talus-1<sup>st</sup> metatarsal angle was 26 degrees (range20-30) indicating forefoot adduction deformity as compared to the normal(0- 20).

In the 4<sup>th</sup> patient, the mean antero-posterior talocalcaneal angle for his two feet, was 15 degrees (range 12-18), as opposed to the normal value of this angle (20-40), indicating recurrent varus heel deformity.( **Table- 2**)

(**Table-2**) show radiographic results, mean talocalcaneal angle and indices and talo-1<sup>st</sup> metatarsal angle.

The sex incidence among our cases, showed a predominantly affected males in the ratio of 10 males: 4 females. Among the 4 patients with recurrent deformities, 3 of them were males and one was female. In our series, all the patients presented with bilateral affection of the foot, i.e. 14 patients (28feet), indicating the multiple congenital contractures of arthrogryptic club foot. Out of these 14 patients, 4 patients (7 feet) had recurrent deformity after operative correction by Turco's release. three patients (5 feet) out of these 4, had recurrent forefoot adduction deformity. Two patients out of these 3, had bilateral recurrent forefoot adduction deformity. The 4<sup>th</sup> patient in this series, had bilateral recurrent varus heel and forefoot adduction deformities.( **Table- 3**).

(**Table-3**) show Liability of recurrence in arthrogryptic club foot and the main recurrent deformities.

In regard to the age of starting treatment, i.e. Turco's posteromedial release, we recognized the rate of recurrence tended to increase after the age of one year.

In our series, 3 cases of recurrence occurred after the age of one year and 1 case after the age of 4 years.

The initial surgical treatment in our series consisted of Turco's posteromedial release in all patients (28), and talectomy in 5 patients (7 feet). Turco's release (**Figure.1.5**) include resection of 1 cm. segment of both the gastrosoleus and the tibialis posterior tendons, generous lengthening of all other tendons posterior to the medial malleolus, posterior capsulectomy of the ankle and subtalar joints, and capsulectomy of the talocalcaneal, talonavicular, and calcaneocuboid joints. Talectomy (Astragalectomy) considered for severe arthrogryptic deformities. It relaxes the soft tissues enough to allow correction of equines and varus deformities of the hind foot and midfoot; however adduction of the forefoot is not corrected and needs additional procedure.

(**Figure. 1.5**) Turco's release.



The types of the initial surgical treatment affect the recurrence rate in our series, with the highest rate among those treated by Turco's release alone, and the lowest rate among those treated by talectomy and soft tissue release including Turco's release. In our series, 3 patients (5 feet) had recurrent deformities after Turco's release, including 2 feet in one patient with varus heel and forefoot adduction, and 3 feet with forefoot adduction alone. On the other hand, 2 feet recurred with forefoot adduction, after talectomy procedure.

The different modalities of treatment for recurrent deformities after initial surgical correction were illustrated in (**Table-4**) show For the 5 feet that recurred after Turco's release; the final surgery was as follows:

In 3 out of the 5 feet, forefoot adduction was the only deformity and treated by Evan's (dorsolateral wedge excision of the calcaneocuboid joint) combined with tibialis anterior transfer to the lateral cuneiform.

(Figure 1.6) Arthrogryptic Club Foot Corrected by Evan's Operation and Tibialis Ant. Transfer.



Evan's calcaneocuboid arthrodesis consists of modified medial soft-tissue release plus closing wedge resection of the calcaneocuboid joint to shorten the lateral column of the foot, the correction maintained by staples.

Out of the 5 feet, 2 feet in one patient recurred with varus heel and forefoot adduction deformities. The right foot had been subjected to talectomy and capsulectomy of the calcaneocuboid joint. Then the same patient brought to our clinic with recurrent left foot at the age of 8 years, triple arthrodesis was done at that time, consisting of wedge resection through the mid tarsal and the subtalar joints together with soft-tissue release, capsulectomy and elongation of the contracted side of the deformity.

Out the 5 patient (7 feet), treated by talectomy, 2 feet had recurrent forefoot adduction. These had been treated by Evan's and tibialis ant. transfer to the lateral cuneiform. (Table 4)

Table 4- The different modalities of treatment for recurrent deformities in arthrogryptic club foot

## Discussion

All the patients in our series were presented with bilateral affection of their feet. Thus, as suggested by Swinyard and Bleck multiple congenital contractures are a more appropriate name for arthrogryposis multiplex congenital.<sup>(1)</sup>

In our series, male children predominantly affected (ratio 10 males: 4 females).

Special emphasis had been placed by many authors on the roentgenographic criteria of club foot correction. In our series, two radiographic measurements were used, i.e. anteroposterior and lateral talocalcaneal angle. The anteroposterior angle was accurate indicator of recurrent varus heel deformity. The talo-1<sup>st</sup> metatarsal angle was the accurate indicator of recurrent forefoot adduction. This was supported by Turco (1971), and

Thompson<sup>(13)</sup>. Laaveg and Ponseti in their study on 70 patients (104 clubfoot), also augment this<sup>(14)</sup>.

The main problem is the disorder is the liability for recurrence. In our series, 4 patients (7 feet) out of 14 patients (28 feet), had recurrent deformity. The most frequent recurrent deformity was forefoot adduction.

Drummond, Sellers and Cruess<sup>(5,6)</sup> as well as Williams<sup>(7)</sup> have stated that recurrence of the deformity is the rule in arthrogryposis, because the dense inelastic soft tissue about the joints do not properly elongate with growth.

The possible causes of recurrent forefoot adduction, in our series, as predicted from the findings at the operative filed in the operations that were justified for its treatment. In 3 patients (5 feet) out of 4 patients (7 feet), who had recurrent forefoot adduction after Turco's release, a short thickened calcaneofibular ligament and bifurcated ligament was found in addition to the bony abnormality, i.e. inadequate mobilization of the calcaneocuboid joint. These patients respond well to Evan's operation. In addition to soft tissue release i. e. excision of the bifurcated ligament with its 2 part the calcaneocuboid and calcaneonavicular. These finding also had been recognized by Vizkelety in his article<sup>(15)</sup>. The 2<sup>nd</sup> possible cause of recurrent forefoot adduction was due to the hypertrophied tibialis anterior tendon acting as adynamic deformer.

In our series, tibialis anterior transfers done for 3 patients (5 feet), the age of the patients were beyond the walking age. During the swing phase of walking, the strong tibialis anterior muscle contracting unopposed by the weak peronei, will constantly invert the foot and lead to forefoot adduction.

In 1923, Dunn described lateral transfer of the anterior tibial tendon in the treatment of club foot and suggested that the transfer would be helpful in preventing or delaying the recurrence of the deformity.<sup>(14)</sup>

Peabody and Garceau believed that club foot is caused by prenatal muscle imbalance in which the pronators and the extensors of the foot are weak, that often the peroneals are permanently weak and that when this weakness can be demonstrated in recurrent deformities, lateral transfer of the tibialis anterior tendon is indicated. Weakness of the peroneals is suggested clinically when a varus deformity of the foot can be corrected passively but not actively and when roentgenograms show that the osseous deformity have been corrected.<sup>(16)</sup>

Williams had described osteotomies and transfer the range of motion to a more useful arc are beneficial but at or near skeletal maturity or the deformity will promptly recur with growth.<sup>(7)</sup>

The 4<sup>th</sup> patient in our series presented with bilateral recurrent varus heel and forefoot adduction deformities. Talectomy was performed for the right foot, in addition to capsulectomy of the calcaneocuboid joint. Also talectomy was done as initial surgery for server deformities in 4 patients (7 feet). We obtained permanent hind foot correction in 5 of 7 primary talectomies. A common cause of recurrence in retention and subsequent growth of a fragment of the talus. Adequate surgical exposure and meticulous removal of the entire talus are important, along with capsulectomy and soft tissue correction.

Drummond and Cruess obtained permanent and lasting hind foot correction in 5 of 11 primary talectomies, and 1 other foot was improved by revision talectomy.<sup>(5,6)</sup>

Green, Fixsen, and Lloyd-Roberts obtained satisfactory results in 24 (71%) of 34 feet treated by talectomy for equinovarus deformity.

Recurrence of the deformity was attributable to the either a retained fragment of talus or inadequate tendo calcaneus release.<sup>(9)</sup>

Likewise Hsu, Jeffray, and Leong have reported satisfactory results in 9 of 15 feet treated by talectomy, 7 of which developed spontaneous tibiocalcaneal fusion.<sup>(10)</sup>

Spontaneous fusion of the calcaneus to the tibia is a frequent occurrence following talectomy, and recurrent deformity in this instance can be corrected by osteotomy of the fusion mass.

In our series, the 4<sup>th</sup> child brought again to our orthopaedic department, with recurrent varus heel and forefoot adduction deformities of his left foot, which was neglected by the family till the age of 8 years. We decided to perform triple arthrodesis for his left foot. The procedure include a dorsolateral wedge resection of bone from the mid tarsal area, the distal cut is made through the cuboid and the proximal part of the cuneiforms and the proximal cut is made through the distal part of the calcaneus and the neck of the talus. The correction is maintained by staples and plaster cast for 3 months until solid bony union occurs. Again, capsulectomy and soft tissue correction are required with elongation of the short or the contracted side of the deformity.

Drummond and Cruess have resorted to triple arthrodesis in younger children, even as young as 6 years of age, with a satisfactory plantigrade, although shortened, foot.<sup>(5,6)</sup>

As a rule, triple arthrodesis should be deferred until skeletal maturity for tow reasons: (1) to prevent foot shortening from surgically inhibited growth, and (2) to prevent recurrence of the deformity in the growing foot.

## Conclusion

Arthrogryposis multiplex congenita is a rare disorder as noticed in our series. This disorder characterized by multiple joint deformities i.e. multiple congenital contractures; coincide with all our cases, presented with bilateral affection of the foot.

Clubfoot can be the only obvious deformity of this widespread disorder, evident in our selected cases for the study. Of the 14 patients (28 feet), 4 patients (7 feet), have had recurrent deformities after initial surgical correction by Turco's posteromedial release. The main problem in this disorder is the liability for recurrence due to the dense, inelastic soft tissue about the joint. The most frequent recurrent deformity, in our series, was forefoot adduction (5 out of 7 feet).

Combined Evan's calcaneocuboid arthrodesis and lateral transfer of the tibialis anterior tendon can correct the deformity once performed at the same cession.

Talectomy, performed as primary procedure on 5 patients (7 feet), achieved permanent and lasting hind foot correction in 5 of 7 feet.

Lastly independent walking is almost always possible in arthrogryposis.

(Table- 1)  
*Recognition of the recurrent deformity, range of ankle and subtalar joints motion based on clinical assessment and goniometer measurement*

No. of patients	Visible recurrent deformity	Range of ankle dorsiflexion	Range of ankle planter flexion	Type of gait	Range of motion of subtalar joint
3 (5 feet)	Forefoot adduction	10°-25°	20°-35°	In-toeing	5°-10°
1 (2 feet)	Varus heel and forefoot adduction	5°-10°	10°-15°	In-toeing	5°-10°

(Table-2)  
*Radiographic Results, Mean Talocalcaneal Angle and Indices and Talo-1<sup>st</sup> Metatarsal Angle*

Number of cases	Lateral talocalcaneal angle	Anteroposterior Talocalcaneal angle	Talocalcaneal index	Talo-1 <sup>st</sup> metatarsal angle
3 (5 feet)	Mean range 31° 30-35°	Mean range 22° 20-26°	Mean 53°	mean range 26° 20-30°
1 (2 feet)	18° 16-20°	15° 12-18°	33°	20° 16-24°

(Table-3) -*Liability of Recurrence in Arthrogryptic Club Foot and The Main Recurrent Deformities*

No. of patients	No. of arthrogryptic club foot	Main recurrent deformity After surgical correction
10	21 feet	No deformity
3	5 feet	Forefoot adduction
1	2 feet	Varus heel and forefoot adduction
Total 14	28 feet	

(Table- 4)  
The different modalities of treatment for recurrent deformities in arthrogryptic club foot

Initial Treatment	Main recurrent deformity	No. of feet	Final surgical treatment	no. of feet
			Talectomy and capsulectomy of calcaneocuboid joint	1
Turco's Release	Varus heel and forefoot Adduction	2	Triple arthrodesis	1
	Forefoot adduction	3	Evan`s and Tib.Ant.transfere operation	3
Talectomy	Forefoot adduction	2	Evan`s and Tib. Ant. transfer Operation.	2

**References**

- Otto, A.G., Genetic aspects of arthrogryposis multiplex congenital, Clin. Orthop. 1841;194:44.
- Swinyard, C.A., and Bleck, E.E., The etiology of arthrogryposis (multiple congenital contractures), Clin. Orthop. 1985;194:15.
- Wynne-Davies, R.W., Williams, P.F., and O'Connor, J.B.F, The 1960s epidemic of arthrogryposis multiplex congenital, J. Bone Joint Surg. 1981; 63-B: 76.
- Brown, L.M., Robson, M.J., and Sharrard, W.J.W, The pathophysiology of arthrogryposis multiplex congenital neurological, J. Bone Joint Surg. 1980; 62-B: 291.
- Drummond, D.S., Siller, T.N., and Cruess, R.L, The management of the foot and ankle in arthrogryposis multiplex congenital, J. Bone Joint Surg. 1978; 60-B: 96.
- Drummond, D.S., Siller, T.N., and Cruess R.L, the management of arthrogryposis multiplex congenita. In American Academy of Orthopaedic Surgeons: Instructional course lectures, 1974; vol. 23, St. Louis.
- Williams, P.F. The management of arthrogryposis Orthop. Clin. North Am. 1978; 6: 967.
- Guidera, K.J., and Drennan, J.C, Foot and ankle deformities in arthrogryposis multiplex congenita, Clin. Orthop. 1985;194:93.
- Green, A.D.L., Fixsen, J.A. and Lloyd-Roberts, G.C. Talectomy for arthrogryposis multiplex congenita, J. Bone Joint Surg. 1984; 66-B: 694.
- Hsu, L.C.S., Jaffray, D., and Leong, J.C.Y. Talectomy for clubfoot in arthrogryposis, J. Bone Joint Surg. 1984; 66-B: 694.
- Weal M.F. clubfoot, a cord Retainer. J of pediatric orthopaedic 1989; 9: 575-578.
- Sharrard W.J.W. Pediatric orthopedic and fractures. 1979;P. 279.
- Thompson G.H. and Richardson A.B, surgical Management of Resistant Talipes equinovarus.JBJS 1982; 64-A: 652.
- Laaveg S.J. and ponseti I.V. long-term result of congenital clubfoot, JBJS 1980; 62-A: 23.
- Vizkelety T. and Szepesi k; Reoperation in treatment of clubfoot, J. of pediatric orthopedic 1989; 9: 144-147.
- Hofmann A.A. and Coleman S.S. Osteotomy of the 1<sup>st</sup> cuneiform as treatment of residual adduction of the forefoot in clubfoot. JBJS 1984; 66-A: 985
- Gross, R.H. the role of the Verebelyi-Ogston procedure in the management of the arthrogryposis foot, Clin. Orthop. 1985;194: 99.

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