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Lower Limb Deformity Management in Arthrogryposis - What to Correct and When

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Abstract
Arthrogryposis is a descriptive term involving non – progressive joint contractures of two or more joints at birth. It is associated with more than 300 diseases. Lower limb involvement is seen in about 95% of the cases with variable affection of the foot, knee and hip. Management depends on the severity, affection of number of joints and co-morbidities. Multidisciplinary management is crucial with realistic expectations. Prognosis should be discussed with parents prior to undertaking surgical intervention, especially the risk of recurrence as age advances. Deformities include soft tissue contractures, fibrotic hypoplastic muscles and in older children, deformed articular congruity. Foot is most commonly affected and Ponseti casting is the gold standard first line of treatment. Failed correction or late presentations are treated with soft tissue/bony surgery and/or fixator. Knee contractures can be flexion or extension with or without joint subluxation and patella involvement. Options for management are serial casting, soft tissue release, growth modulation, bony surgery, gradual correction with fixator or a combination of these based on the severity and age at presentation. Pterygium management is difficult due to proximity of neurovascular structures to the skin web and higher risk of recurrence. Hip contractures and dislocation when unilateral should be treated surgically. Treatment of bilateral affection is controversial and should be individualised.

Keywords: Arthrogryposis, Amyoplasia, Arthrogryposis multiplex congenita, Hip contracture, Knee contracture, Lower limb deformity, Atypical clubfoot.


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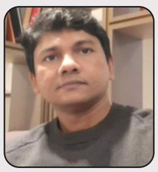
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Introduction

Arthrogryposis is a descriptive term and not a diagnosis. It was originally described by Otto in 1841 and etymology points to Greek origin meaning a 'curved joint'. It is a non-progressive condition with joint contractures involving two or more joints at birth. More than 300 diseases with arthrogrypotic features have been described. A specific diagnosis is achieved in only one half of the patients [1, 2].

Arthrogryposis multiplex congenita (AMC) is divided into 3 large groups: Group 1 has predominant limb involvement, e.g. classical arthrogryposis or amyoplasia. Group 2 involves limbs with other body parts, e.g. multiple pterygium syndrome, Escobar syndrome, Freeman–Sheldon syndrome. Group 3 shows central nervous system dysfunction with limb involvement, e.g. cerebro-oculo-facio-skeletal syndrome. Prognosis depends on the diagnostic group hence its identification is crucial [3]. Amyoplasia is the classic most common form where they present with rigid symmetrical contractures in extremity, hypoplasia of muscles with fibrofatty infiltration and normal intelligence. Lower limbs are involved in about 89 to 95% of cases [2-4]. Foot involvement is most common (90%) followed by knee (70%) and hip (40%) [5]. Limb deformities present with loss of flexion creases, fusiform appearance of limb and dimpling at affected joints (Fig. 1).

Single or multiple etiologic factors (genetic, muscular, neurologic) lead to loss of muscle mass with imbalance of muscle power at the joints. This provokes a collagenic

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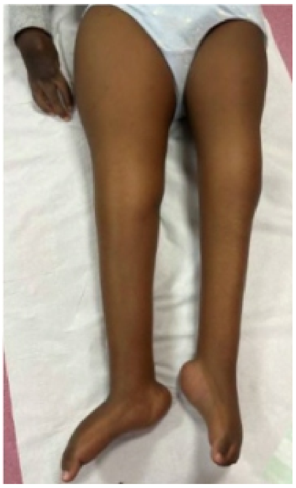

Figure 1: 5 year old with arthrogrypotic lower limb affection. Absence of flexion crease, fusiform appearance of lower limb. Knee extension contracture with bilateral foot deformity.

response by partial replacement of muscle volume with fibrous tissue and collagenous thickening of the joint capsules [6]. Joint movements are essential for articular cartilage nutrition, moulding of the joint shape and function of muscles, tendons and ligaments. The intrauterine hypomobility or akinesia contributes to fibrous tissue development around the joint. Ligaments are shortened, fibrotic and subcutaneous tissue is replaced by inelastic fibrotic tissue. All these factors contribute to joint contracture.

Multidisciplinary approach with specialised physiotherapist, orthotist, orthopaedic surgeon, neurologist, paediatrician and geneticist is essential in management of these conditions.

The aim is to improve quality of life and facilitate independent ambulation for daily living activities and recreational activities. Initial treatment goals are to improve and maintain range of movement (ROM) for better positioning and function. Therapy to improve muscle strength and splints to aid mobility form the cornerstone of rehabilitation. Following manipulation, resistant contractures and deformity should be considered for surgery based on co-morbidities and ambulatory potential. For any intervention, consider management of difficult venous access and intubation, increased risk of postoperative aspiration, gastrointestinal problems and concurrent deformities. In older children, upper limb deformity can restrict the use of walking aids following lower limb surgery.

Sequence of Correction

Simultaneous correction of all deformities is not recommended due to complexity of the procedures. However, two deformities can be corrected simultaneously depending on patterns of presentation and individual cases. Plan to deal with most distal deformity first.

Foot deformity + knee flexion contracture + hip dislocation: Foot can be corrected with Ponseti casting from 1 month of age, knee flexion deformity at 6-8 months of age and hip contractures and dislocation at 8-12 months of age.

Foot deformity + knee flexion contracture + hip adduction contracture: If hips have adduction contracture, Ponseti casting can be difficult causing problems in maintaining hygiene. In such cases, hips can be corrected first followed by foot and knee.

Foot deformity + knee hyperextension: Non operative management of foot and knee can be done simultaneously. If surgical correction of club foot is indicated, knee flexion is needed to maintain foot correction and hence knee surgery is performed before or at the same time as foot surgery [7].

Foot deformity + knee hyperextension + hip dislocation: Knee flexion is essential to maintain hip position after surgical reduction otherwise hamstring and quadriceps are under tension and tend to redislocate the femoral head. If sufficient knee flexion is not achieved by casting, knee surgery precedes hip surgery. Following which foot and hip can be managed simultaneously/sequentially.

Hip Deformity

The most common hip contractures are a combination of flexion, abduction, and external rotation known as the "Buddha position." For ambulation, isolated functional hip contracture <30 degree is needed [8]. However, with concomitant knee and foot deformities, compensation is affected, and lesser degree of contracture can affect ambulation. Congenital hip dislocations occur earlier in pregnancy (teratogenic) and are therefore stiffer and more proximally displaced [9]. The incidence varies from 14-42% [8, 10-13]. Non-surgical treatments (such as Pavlik harness) are generally unsuccessful [14].

Studies have shown that in cases of unilateral dislocations, the total arc of motion (flexion-extension) of the dislocated hip is only 12 to 20 degrees less than the contralateral side, both before and after relocation [15]. Yau et all reported good results with surgical interventions in a 20 year follow up of 19 patients (38 hips) with hip problems in AMC (dislocations, subluxations and hip contractures) [16]. Nema et al in a review article had 94 hips (59 children) from 7 studies concluded that

Figure 2: 2a: 6 months old child with right hip dislocation. **2b:** Four months post op image. Open reduction and femur derotation was done at 14 months. Adequate acetabular coverage was noted on arthrogram hence pelvic osteotomy was not undertaken. 2c: 6 years old now with subluxing right hip.

children less than 6 months of age had a good result with open reduction (medial/ anterior) while older children had a higher need of concomitant procedures (femoral and acetabular) [17] (Fig. 2). They also concluded children with AMC can be expected to ambulate with or without support in 90% of the cases after open reduction and concomitant knee and foot procedures.

Hence it is prudent to say that reduction of the unilateral teratological dislocated hip in a child with AMC would improve the chance of ambulation and longevity of the joint in later life. A higher incidence of avascular necrosis in AMC hips was attributed to a variety of reasons including difficult reduction, abnormal anatomy and revision surgery.

Controversy exists in surgical treatment of bilateral hip dislocations in patients with arthrogryposis. Most authors report poor results [8, 10, 18] of surgical treatment while some suggested open reduction in children with less severe involvement [11-13]. Miao et all described eight patients with bilateral DDH who received open reduction surgery simultaneously, and seven of these patients did not require secondary revision surgery. There was no statistically significant difference in surgical age and revision surgery between the bilateral and unilateral hip groups [19]. Yau et al, in their series, had one patient with bilateral hip dislocation who did not receive surgical treatment and was confined to wheelchair and the other had open reduction at 8 months of age and was a community walker at final follow up [16]. Treatment should be individualized in bilateral dislocations and prognosis should be discussed with parents.

The treatment of hip subluxation/ dislocation in children with AMC is usually based on the age of the child and degree of involvement of the hip. The following algorithm is useful in managing these complex hip pathologies:

1. Hip joint contractures:

A. Isolated hip flexion or mild flexion/abduction contractures: Percutaneous release of the conjoint tendon of the sartorius and tensor fascia lata just inferior to the anterior superior iliac spine (ASIS) [9, 14, 20].

B. Multiplanar contractures: Proximal femoral reorientation osteotomy. This repositions the lower limb in line with the body, leaving the femoral head in its original position within the acetabulum and hence preserves the hip's total arc of motion making the limb more functional. Various authors have reported improved ambulatory capacity as compared to pretreatment [21].

2. Hip dislocation:

Early open reduction has been described in multiple studies with age ranging from 3-12 months with good results [19]. Both medial and anterior Smith Peterson approach has been described, with more leaning towards anterior approach. Depending upon the age, a higher need of concomitant procedures (femoral derotation, shortening and acetabular osteotomies) as expected has been reported. Up to 2/3rd of the patients need femoral osteotomies [14].

Multiple authors have described different timings and procedures for correction of hip dislocation [14]:

A. Early open hip reduction (medial/ anterior approach) at 3-6 months with no need for further procedures.

B. Beyond 10 months to 3 years: Open reduction using anterior approach along with femoral shortening derotation osteotomy and acetabular osteotomies

Complications included stiffness, redislocation, and avascular necrosis rate of upto 19%.

Knee Deformity

Knee contractures are difficult to treat, and severe contractures make ambulation difficult to impossible. The development and congruence of articular surface is affected [22]. The reported incidence is 38-85% [23]. Flexion contractures are common with only 50% becoming community ambulators compared to all children with extension contractures [24]. Knee can be hyperextended/subluxed or dislocated, patella subluxation/dislocation and patella alta can be associated. Flexion contractures have poor prognosis. For functional ambulation, knee flexion contracture should be of <20 degree, without hip and foot affection [8]. But these children often have concomitant affection of hip and foot making compensation of even mild contractures difficult. Risk of degenerative arthritis is higher in extensor type [25].

In flexion contracture, goal is to optimise extension for functional ambulation and in extension contracture, goal is to optimise flexion for sitting and rising from a chair.

Flexion contracture

Contractures particularly involve hamstrings and posterior capsule. Cruciates may be shortened, epiphysis is hypoplastic, it is not deformed at birth, but abnormal alignment and contractures lead to deformity over time. Radiographic abnormalities seen are patella elongation, flattening of femoral condyles, dome shaped proximal tibia epiphysis and joint incongruity [22]. Quadriceps strength affects ambulatory potential. Grade 3 or greater power is required for ambulation [8]. With flexion, the centre of gravity is shifted posteriorly which increases fatigue of the extensor mechanism.

Splinting and serial casting

Day and night splinting is essential following correction done with any method to maintain it. Knee ankle foot orthosis (KAFO) and ankle foot orthosis (AFO) help maintain the alignment, compensate for muscle weakness, support the extremity and improve ambulation. They are recommended until maturity following correction [24, 26, 27].

Serial casting can help in mild contractures, however response is variable [28] 2and recurrence rates are high with a risk of posterior subluxation of tibia due to abnormal shape of the proximal epiphysis [10]. This can be started within a few weeks of birth since the connective tissue is compliant and a large proportion of the bone is not mineralised [29].

Soft tissue releases

When conservative management fails, consider other deformities and ambulatory potential prior to planning intervention. Waiting until 2-3 years is suggested to assess ambulatory potential [1].

With poor ambulatory potential, acceptance of contracture aids in sitting. With good ambulatory potential - posterior soft tissue release along with posterior capsular release is indicated. This is demanding since muscle planes are absent and replaced by fibrous tissue. Use of tourniquet is inadvisable since identification of neurovascular bundle can be difficult [24]. All soft tissue often requires release including hamstrings, posterior capsule and posterior cruciate ligament (if taut) leaving only skin and neurovascular structures. Double incision posteromedial and posterolateral are used. The thick posterior capsule does not stretch [30] and recurrence is higher without capsular release [26]. Full extension might not be achieved even after release and serial casting is carried out after correction to improve this. Risks of surgical release include skin necrosis, iatrogenic neurovascular injury and contraction of scar tissue and recurrence. Knee releases improve function in immediate post operative period, however parents and children should be warned of decline in ambulatory ability as patients age and contractures recur [31]. Long term follow up shows revision surgery in 26-32% [23, 24, 26].

Guided growth

Anterior epiphysiodesis for distal femur has been advocated for contractures <20 degree. Correction is gradual over a number of years [26]. Palocaren T et al showed significant correction with anterior distal femoral 8 plates in <45 degree flexion contracture. Timing of intervention was recommended based on the size of femoral condyle and not bone age. In their series this was at 5-7 years since the smallest screw size for 8 plates is 16mm. Incisions were on either side of patella and plate was inserted 2-3 mm away from the medial and lateral edge of patella and removed after correction [32]. Correction occurs at the rate of 1.4 degrees per year [33]. A secondary distal femoral deformity is produced by this technique and recurrence is likely after removal of implants.

Bony procedures

Distal femur extension osteotomy with shortening can acutely correct large contractures (40 degree) especially with posterior release. It is also indicated in revision surgery. Shortening along with acute correction helps to reduce the stretch on neurovascular structures and is done with a trapezoidal wedge excision. These can be fixed with wires or a plate. Bone is often osteopenic and locking screws can be used for a stronger construct. Corrective osteotomy does not improve the range of movement (ROM) but changes the arc of ROM to a functionally desirable position [34]. The distal femur anatomy is altered but the abnormal joint articulation is not altered. These should be delayed beyond 12 years due to rapid remodelling and recurrence in younger children, however risks must be weighed with improved function and social benefits to these children. Recurrence is noted to be 1 degree per year in under 12 year olds [35]. Need for revision is reported in 35-38% of patients [35]. Yang et al showed that ambulatory gains were maintained at an average follow up of 4.9 years despite recurrence of flexion contracture following distal femoral extension osteotomy and/or gradual joint distraction with Illizarov circular fixator [36].

Feldman et al described a combination of posterior release, peroneal nerve decompression and proximal femoral shortening to improve the ROM and not just the arc of ROM. Results of this demonstrate good function [37].

Fixator assisted deformity correction

Use of circular fixator with gradual correction has been described showing improvement with recurrence of lesser degree. Poorer results were noted with increasing severity of contractures [25]. For contractures >40 degree, posterior soft tissue releases and gradual arthrodiastasis using an Ilizarov external fixator has been effective in correcting and maintaining correction of knee flexion contracture. (Fig 3). Placement of the hinge at the knee centre of rotation (COR) is crucial. It is better to err inferior and anterior to avoid anterior impaction of tibia on femur in extension and posterior subluxation of tibia. Correction proceeds at 1 degree per day and is preceded by 5-10mm of joint distraction to prevent crushing of the articular cartilage [38]. Complications include

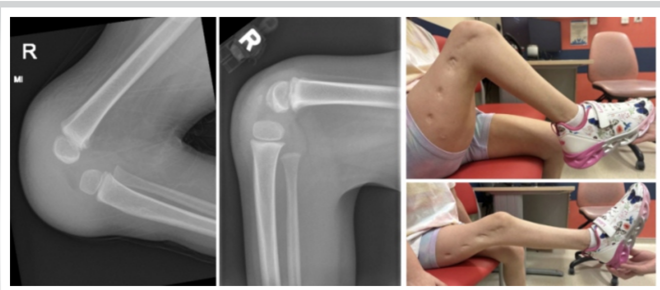

Figure 3: 3a,b: 3 year old with knee contracture – image in maximum flexion and extension. **3c,d:** 6 year post correction with soft tissue release and gradual distraction with frame in maximum flexion and extension

that of fixator treatment. Fractures are seen due to osteoporosis, manipulation of a rigid joint, secondary to shear forces and pre-existing stress fractures. Precautions include placing proximal femoral pins well centred in bone avoiding unicortical placement and thread diameter <25% that of femur isthmus. Posterior subluxation of tibia is seen and to counter the force causing it, the proximal tibia trans fixation wire is tensioned anteriorly.

Pterygium is a web of tissue with shortness of skin and other soft tissue. Popliteal pterygium often has a taut cord originating form ischial tuberosity and inserting into dorsal part of calcaneum causing a knee flexion contracture and foot equinovarus deformity. Serial casting can be started at birth including the foot. Early tendoachilles (TA) tenotomy improves the equinus contracture. If contracture is >20 degree, surgical correction is planned at 1 year of age prior to child gaining ambulatory potential. Surgical correction is difficult in pterygium due to proximity of neurovascular structures, maintenance of correction is also difficult, and recurrence is common. V-Y plasty of skin can be performed over the web. The ischiocalcaneal cord is excised along with TA lengthening. Soft tissue release is similar to flexion contracture with additional dorsal capsulotomy. Nerve and vessels should be identified and protected when performing the release. Post operatively knee is placed only in slight extension compared to pre-operative deformity and gradually stretched weekly to avoid an acute stretch on neurovascular structures [29]. Gradual stretching with a circular fixator has improved knee and ankle mobility but involves a greater number of procedures and higher complication rates [39].

Arthrodesis

Arthrodesis in extension closer to maturity is considered based on individual circumstances and demands of the patient. Ambulating short distances is easier, however sitting and wheelchair use is impaired [26].

Extension contracture

It is caused by fibrosis of the rectus part of quadriceps and tendon bound to suprapatellar part of femur. The anterior capsule, periarticular tissues and iliotibial band is tight with external rotation and valgus deformity. Entire extensor mechanism is displaced laterally and patella is displaced laterally and superiorly, hence development of trochlear groove is affected. Patella ossification is delayed beyond 3-4 years. Extension contractures are subclassified into simple recurvatum, anterior subluxation and anterior dislocation [40, 41].

Serial casting and manipulation

In anterior dislocations, collateral ligaments and hamstring are displaced anteriorly. These are started at birth by manipulation of knee with anteriorly directed force on distal femur and posteriorly directed force on proximal tibia followed by a dorsal splint. When started after a few days or weeks, initial longitudinal stretch is first applied to the knee to avoid nutcracker phenomenon of femoral condyles impinging on tibia. Concomitant hip flexion relieves stretch on the rectus making knee flexion easier. Too much pressure on the tibia can cause a plastic deformation [42]. This resolves with time but indicates a stiff knee needing surgery. It also carries a risk of proximal tibial physeal separation. Manipulation can fail to provide significant improvement in 15-58% especially when treatment is started at 1-2 years of age [7, 23, 24, 41, 43].

Surgical intervention

When there is no improvement with two serial casts or persistent subluxation/dislocation of the tibia on femur or failure to achieve 45 degree of flexion, surgical intervention is considered [43]. Soft tissue release in the form of quadricepsplasty can be undertaken. This can be done percutaneously [44] or mini open technique [42] or open technique with Z plasty or V-Y plasty [40]. Modified Curtis and Fisher quadricepsplasty through lateral incision show improved results with regards to wound healing, insufficient lengthening and instability. A tongue of rectus femoris is raised and vasti are mobilised without dividing the lateral retinaculae or collateral ligaments [32]. Open quadriceps plasty has shown satisfactory long term outcome in 62% of knees at 11.1 years with improved joint congruency, increase range of movement and relocation of patella in groove [45]. There is a risk of overlengthening the quadriceps tendon causing weakness of the already weak extensor mechanism [45].

Foot and Ankle Deformity

Foot and ankle contractures and deformities are common findings in arthrogryposis. Congenital talipes equinovarus (CTEV) being the most common presentation is seen in approximately 70% and congenital vertical talus (CVT) in 2–12% (Fig. 4). Less common deformities are the isolated equinus deformity, congenital metatarsus adductus, pes equinovalgus, or pes calcaneovalgus deformities (1%) [46].

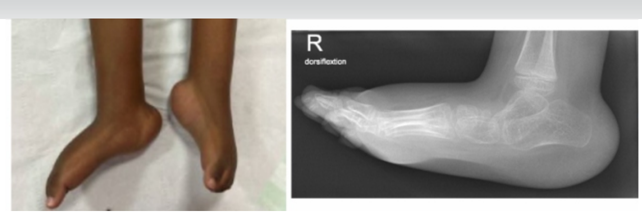

Figure 4: 4a: 5 year old with right CVT and left CTEV. **4b:** lateral radiograph showing right CVT

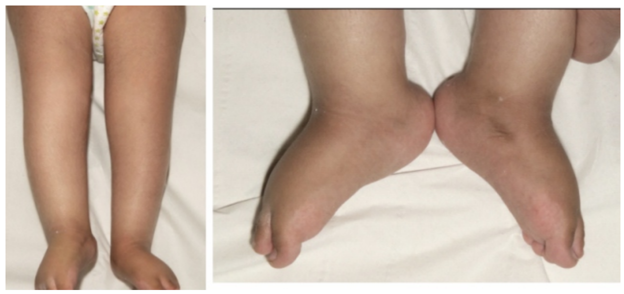

Figure 5: 18 month old with **5a:** bilateral knee extension contracture, **5b:** bilateral CTEV

Congenital talipes equinovarus (CTEV)

This is a more severe form compared to idiopathic CTEV and more resistant to treatment (Fig. 5).

Ponseti technique remains the gold standard as the first line of treatment. Morceuende et al [47] demonstrated good initial correction in patients following Ponseti serial casting and bracing in 15 out of 16 patients. They have stated that despite tenotomies, the foot achieved an average of 5 degrees of dorsiflexion. An average of 7 casts were required before tendo achilles tenotomy, with an older patient requiring 12 casts. Only one patient required posteromedial surgical release to achieve initial correction. The authors have suggested that recurrence can be up to 25 % and can be managed with repeat casting and tendo Achilles tenotomies in addition to tibialis anterior tendon transfers. A few patients might also require posteromedial surgical release. Mattar et al [48] achieved initial correction in all feet with Ponseti technique and they had satisfactory outcome in 10 out of 17 arthrogrypotic club feet (64.7 %).

Van Bosse suggested that club foot deformity in arthrogryposis can present either as equinovarus or equinocavus foot deformity. The latter foot appears similar to atypical or complex club foot. These require the modified Ponseti casting technique. The index and forefingers of both hands are placed on the dorsum of the midfoot, just anterior to the ankle joint, the thumbs are positioned under the metatarsal heads and cavus is initially corrected with upward pressure on metatarsal head relative to midfoot followed by achillles tenotomy. In his experience, minimal interventional surgery and frequent serial casting maintains foot deformities while the child is growing, however these deformities recur despite the initial correction, equinocavus however have a lower tendency to relapse [49].

Many studies have reported that posterior and posteromedial surgical releases despite initial correction lead to unsatisfactory results in up to around 75% in the long term [10, 50-52]. Most of these patients would require further surgeries or casting [10, 51, 52].

Talectomies have also been used to treat arthrogrypotic clubfeet either as primary procedure or as salvage procedure. Shergui et al [53]performed a systematic review that included 232 feet treated with talectomy. Recurrent deformities and revision surgery were seen in 16.81% and 13.36% of cases respectively. The authors concluded that talectomy is a valid surgical option for severe arthrogrypotic foot deformities with favourable post-operative outcomes and low complication rate. Segal et al [54] reviewed their results of talectomies on arthrogrypotic clubfeet where they had good and fair results leading to their conclusion that primary talectomy in arthrogrypotic feet was more effective than posterior-medial releases or secondary (salvage) talectomy. They suggested though, that they had recurrent forefoot and cavus deformities after primary talectomies.

Eidelman and Katzman [55]published their preliminary results for the treatment of arthrogrypotic foot deformities that were managed with circular fixator with or without osteotomies. They concluded that the Taylor Spatial Frame is a reliable and accurate method of correction for complex foot deformities in children with arthrogryposis. Barbary et al [56] published their results of 66 clubfeet (23 arthrogrypotic clubfeet were included) treated with soft-tissue distraction using an Ilizarov frame with satisfactory results and plantigrade feet.

Congenital vertical talus

CVT is another deformity that can be seen in patients with arthrogryposis. Chalayon et al [57] published a minimally invasive technique for the management of congenital vertical talus with reverse Ponseti Technique that consisted of serial manipulation and casting followed by percutaneous tenotomy of the Achilles tendon and a small medial incision to reduce the talonavicular joint and ensure accurate reduction and pin fixation. This technique has resulted in excellent short-term deformity correction while preserving subtalar and ankle motion in patients with rigid congenital vertical talus associated with arthrogryposis.

Bouchard [58] published a stepwise algorithm for the patient of any age with congenital vertical talus. He suggested that casting should always be attempted prior to surgery, and surgery should be performed with à la carte approach for the irreducible deformities. He suggested medial subtalar joint release and release of any anomalous fibrous tissue between tibia and navicular for the subluxed talonavicular joints intraoperatively, followed by fractional lengthening of toe extensors if plantarflexion is less than 25° and lengthening of peroneals and lateral subtalar joint if adduction is less than 10°. For the more severe deformities where the navicular remains dislocated despite adequate castings, the author proposed dorsolateral surgical release when there is no significant dorsiflexion and posterior release of ankle and subtalar joint when equinus was severe. Naviculectomy is preserved for the older patients where the medial column is long to allow correction. Talectomy that has been used in the past for the initial management of CVT has been kept as a salvage procedure for the non-ambulatory patients.

Conclusion

Hip dislocation in arthrogryposis is a challenging problem. Non-surgical management is not recommended due to high failure rate. Long term studies suggest early intervention has the best results, with majority of the patients having a stable, well-developed hip and being satisfactory community ambulators. Treatment of bilateral hip dislocations is controversial but leans more towards surgical intervention. Knees can present with flexion or extension deformity. Early manipulation followed by various surgical procedures are based on the severity and age of the child. Recurrence is commonly seen but to a lesser extent. Foot deformities are the most common presentation including CTEV and CVT. Casting remains the gold standard followed by surgical intervention when there is no improvement. Salvage procedures are considered for recurrent and resistant deformities.

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