

# Serial Operational Treatment of Arthrogryposis With Complete Dislocation of Hips and Knees

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**Introduction:** Arthrogryposis multiplex congenita (AMC) with incidence of 1 in 3000 of births is characterized by multiple rigid joint and soft-tissue contractures.

**Case Presentation:** We present a case of AMC with severe soft tissue and joints contractures, especially joints of lower limb and elbows, bilateral complete dislocation of hips (CDH) and knees (CDK) (type III), bilateral elbow joint dislocations and severe bilateral clubfeet, who had had some successful operations during five years on lower extremities.

**Discussion:** The main goal of our treatment was achieving the maximal function of lower limbs. Passive stretching range of motion on stiff joints especially of lower limbs must be started as soon as possible; serial castings, open reduction of CDH followed by derotational femoral osteotomy and shortening was helpful. Surgery performed at earlier age gives the best functional outcome.

**Keywords:** Arthrogryposis; Knee Dislocation; Hip Dislocation

## 1. Introduction

Arthrogryposis multiplex congenita (AMC) is a single unrelated nonprogressive disease with common phenotypic characteristic of multiple rigid joint contractures and defective muscles (1). More than 300 different conditions with arthrogryptic features have been described, including neuromuscular disease, skeletal dysplasias, chromosomal syndromes, etc. (2). The common end pathway of this disease is decreased fetal movements due to various factors such as neuropathic and myopathic processes, abnormality of connective tissue, and impaired fetal or intrauterine vascularity (3, 4). Various subtypes of AMC have been distinguished; the most common recognizable one is amyoplasia (4). It is a sporadic disorder characterized by multiple congenital contractures, symmetrical positioning of the limbs, and replacement of muscles by fibrous and fatty tissues (3, 5). The incidence rates of arthrogryposis and true amyoplasia have been 0.03% and 0.001% among newborn infants, respectively (6). Hip and knee dislocation are the most common joint contractures in these patients (4, 7). A well-planned treatment program should seek to accomplish the maximum possible functional improvement in as few operative procedures as possible, preferably finishing by the age of six (5). Our treatment started at the age of less than one year and finished at the fifth year. In this report, we pres-

ent a case of AMC with bilateral complete dislocation of knees (CDK type III) and hips (CDH). It is commonly believed that bilateral CDH associated with joint contractures versus CDK should not be reduced, while open reduction leads to poor results. This report represents our experience with surgical management of bilateral CDK and CDH in a child with AMC who could walk, run, sit and work after the following series of operations.

## 2. Case Presentation

A less than-one-year-old female was presented to Farahmandfar Hospital (Shiraz, Iran) with concern of AMC. She was the last child of a family of six individuals; parents were relatives with negative familial history of any congenital disease. The patient was from a normal vaginal delivery. She was regularly fed, mostly through breastfeeding. She had a history of developmental delay (e.g. the neck holding reflex was delayed because of the large size of the head; parachute and tonic-neck reflexes were also delayed). She was hypoactive due to her age. During physical examinations, the child was noted to have a large head (the head circumference was more than 95% for age) as well as a short stature and neck. Cardiovascular, respiratory and neurologic examinations were normal.

She had regular skin and nails. Cognition and language were normal. Shoulders had limitation of motion in all directions, deviation of elbows (cubitus valgus; elbows had flexion contractures of about 30 degrees) and both elbows and wrists had limitation of motion, but fingers did not. The patient had normal trunk and spines. Congenital bilateral complete dislocation of hips with coxa-valgum, hyperextension of both knees about 10 degrees, genu-valgum about 30 degrees and limitation of motion in flexions of both knees and hips were considered. She had severe clubfoot deformity. The patient was admitted for operations. A series of operations were performed on the patient's lower extremities, hip and knee. Two operations were performed on her; the primarily was on soft tissue for open reduction of femoral head (Somerville's operation), then internal fixation by Kirschner wires (pin) and casting (closed reduction of CDK with manipulation under anesthesia with serial casting) from toe to lower third of abdomen were performed. At the age of 1.5 year, two operations were performed on both knees for lengthening of quadriceps and releasing of quadriceps tendon and retinaculum (Curtis operation). At the age of three, the right hip was re-dislocated. The patient had hypoplastic medial condyle of both femurs. In addition, a close reduction was performed on both knees for increasing the flexion of both tibiofemoral joints. Shortening of femoral bone and derotational femoral osteotomy were performed. Inverted V-shaped lengthening of quadriceps muscle operation (King's operation for 2nd stage CDK treatment) was performed. The patient was observed every two months. Arthrogryposis rarely occurs and has varieties either in the disease criteria or the operational procedures. In this case, CDK and CDH made complications for operational procedures as these usually occur during arthrogryposis. Open reduction of both hips followed by derotational femoral osteotomy and shortening and serial casting plus manipulation under anesthesia for knee joint followed by quadriceps operation had very good results. At the age of 2.5, the patient was visited and examined. She could stand, walk and even run. A 30-degree flexion of knees was performed. Longitudinal growth of lower limbs was improved. Disproportions of head and neck to body were lessened. At the final follow-up, she walked without crutches or cane.

### 3. Discussion

Patients with distal arthrogryposis have fixed hand and foot contractures, but the major large joints of the arms and legs are spared (8). AMC may include many congenital and genetic symptoms, making it hard to be diagnosed as arthrogryposis multiple symptoms (9). AMC is a distinct entity which needs to be delineated from the other arthrogryposis types (~10 types so far) and other syndromes in which stiff joints are a part of the phenotype (~150 syndromes). In particular, distinction with distal forms of arthrogryposis can be challeng-

ing (10). Treatment should seek to obtain the maximum possible functional improvement in as few operative procedures as possible (5). Some contractures may seem to worsen with age, but no new joints become involved. At least 25% of affected patients are nonambulatory. An early program of passive stretching exercises for each contracted joint to be followed by serial splinting with custom thermoplastic splints is recommended (8). The reproducibility of walking patterns differs between patients with AMC and a healthy control group. This is probably due to the complex compensating strategies of gait, especially in the sagittal and frontal plane, which must be accepted as functionally important (11). An incidence of hip joint contracture of 80% with or without dislocation has been reported in patients with AMC (12). Most patients with dislocated hips have some active flexion or extension of the hip and a range of passive motion of 60 to 90 degrees. If flexion or extension is markedly limited, hip reduction may not be appropriate. If the child demonstrates little active hip movement and ambulation is clearly not likely, hip reduction will be unnecessary and positioning for sitting will be appropriate (13). Close reduction has been uniformly unsuccessful in children with arthrogryposis (14). Soft tissue releases may be considered; but if they are followed by hip flexion deformity, ambulatory function may be lost. Traditional recommendations say that bilateral teratological hip dislocations should not be reduced because reduction will not improve function (12). Canale et al. reported good results with early open reduction using a medial approach to the hip. This approach was used for unilateral and bilateral hip dislocations (8). DelBello and Watts found that if osteotomies are performed before skeletal maturity, the deformity would recur at a rate of one degree per month. Even with this tendency for recurrence, about 50% of corrections were maintained (15). Knee joint involvement has been reported in 70% of patients with arthrogryposis (4). The two most common deformities around the knee are flexion contracture and extension contracture. The initial treatment of flexion contractures is by serial splinting or casting in progressive degrees of extension. Ambulation is possible with residual knee flexion contracture of 15 to 20 degrees. If complete correction is not obtained by 6-12 months of age, posterior medial and lateral hamstring lengthening and knee capsulotomies will be indicated (8). The quadriceps is lengthened in a V-Y-plasty through the central tendon of the quadriceps. Alternatively, the femur may be shortened 2-3 cm at the midshaft and plated to reduce the need for quadriceps lengthening (13). The results of quadricepsplasty in arthrogryposis are rarely reported; flexion deformities predominate most reports and the need to treat extension deformities is unclear. Ideally, the goals of treatment would be to obtain a functional range of flexion (> 60 degrees) without sacrificing the quadriceps strength and to provide a knee with stable periarticular supporting structures. Often, these goals

are not realistic because the arthrogryptic quadriceps is congenitally weak and fibrotic (13). Contracture of the quadriceps mechanism can cause hyper-extension of the knee, which is treated initially by serial casting. If the deformity does not respond to conservative treatment by 6-12 months of age, surgical correction by quadriceps-plasty is recommended. In this study, despite the initial severity of involvement, she was able to walk independently at the end and could stand up on her own with 30 degrees of flexion correction. We believe that it must be a cause of the series of surgeries beside early intervention. In conclusion, our experience showed that the series of surgical steps mentioned above in patients with AMC with CDK and CDH were successful.

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### Authors' Contributions

Soheil Ashkani Esfahani, Shima Rafiee and Parisa Iloof helped in gathering the data as well as writing and editing the manuscript. Sedigheh Ebrahimi helped in follow up visits and editing the manuscript. Saeid Ashkani performed the operations and managed the complications.

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