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ORIGINAL RESEARCH

Short-term functional outcome in children with arthrogryposis multiplex congenita after multiple surgeries at an early age

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Purpose: The purpose of this study is to report our short-term functional outcome for 14 children with arthrogryposis multiplex congenita (AMC) who underwent multiple surgical procedures at an early age.

Methods: During the period 2002–2010, 14 children (11 males and three females) with AMC underwent multiple surgical procedures to treat deformities of the lower and upper limbs. About 81 procedures were performed, at a rate of 5.9 procedures per child. The mean age at the last follow-up was 5.9 years. The average follow-up period was 3.6 years (range, 1.5–6 years). The functional outcome assessment included walking ability and the activities of daily living for the upper limb function.

Results: At the last follow-up visit, six (43%) children (four males, two females) with a mean age of 8.3 years (range, 4–15) were independent walkers. Three children (males) with a mean age of 3.5 years (range, 2.5–5) were able to walk, but with support. One child (male), 3 years old, was a household ambulator. Three children (two males, one female) with a mean age of 4.2 years (range, 2.5–6) were nonfunctional ambulators. The last child (male) was nonambulatory at the age of 5 years. Activities of daily living were severely affected in the nonambulatory child. One child in the nonfunctional ambulators group had limitations in the activities of daily living; however, upper limb function was not affected in the remaining 12 children.

Conclusion: We believe that aggressive surgical treatment using multiple operations at an early age can improve the short-term functional and clinical outcomes of children with AMC.

Keywords: deformity, multiple surgeries, functional outcome

Introduction

Arthrogryposis multiplex congenita (AMC) is a syndrome complex characterized by several features, including multiple joint involvements with severe contractures, dislocations, lack of normal skin creases, and varying degree of fibrosis of the affected muscles.[1–3] The incidence of AMC is rare, occurring in approximately 0.03% of newborn infants.[1,3] The underlying cause of AMC is decreased fetal movements due to various factors, such as neuropathic and myopathic processes, abnormality of connective tissue, maternal disease, and impaired fetal or intrauterine vascularity.[4,5] Various subtypes have been distinguished; the most common one is amyoplasia, which represents one-third of all cases.[4,5]

Hip joint contracture, with or without dislocation, is reported in approximately 80% of patients with AMC.[2,6] Knee joint involvement has been reported in 70%, with flexion contractures being the most common, followed by extension contractures.[4,7] Foot deformities are frequently observed, with equinovarus adductus foot being the

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most common.[7,8] In the upper limbs, the shoulders are typically adducted and internally rotated, the elbows are often extended rather than flexed, and the wrists are flexed severely, with ulnar deviation.[9]

The goal of treatment is to obtain the maximum possible function through an early multidisciplinary approach.[3,10,11] According to Bamshad et al,[12] the primary long-term goals of treatment are increased joint mobility and muscle strength, as well as the development of adaptive use patterns that allow for walking and independence with activities of daily living. The ability to walk depends on the extent of joint range of motion, in particular in the hips and knees, as well as in the foot, with possibility of plantigrade foot position.[4] Muscle weakness in the lower extremities, primarily in the hip and knee extensor muscles, has also been reported to influence the walking ability.[13] The most important aim of the management of the upper limb deformities is to improve the ability to carry out daily activities, such as self-feeding and self-toileting. The second goal is to aid ambulation if there is a need for crutches or a wheelchair.[10]

The purpose of this study is to report our short-term functional outcome, including walking ability and upper limb function of 14 children with AMC who had multiple surgical procedures at an early age.

Patients and methods

This is a retrospective study of 14 AMC children who underwent multiple surgical procedures at an early age for deformities and dislocations of the joints in the upper and lower limbs at the Department of Orthopedic Surgery at King Abdullah University Hospital, from 2002 to 2010. In our study, we included children who had joint contracture at birth in at least two different parts of the body, generalized muscle wasting, and absence of normal skin creases. There were eleven males and three females, and the mean age at the last follow up was 5.9 years. Ten children had quadrimelic contractures, and four had bimelic involvement of the lower limbs. The hip joint was involved in 22 out of 28 hips, including 14 dislocations (six bilateral, one on the right side, and one on the left side). The remaining eight hips had contractures (six flexion contractures and two abductions with severe external rotation contracture). The knee joint was involved 22 times, including four dislocations, nine extension contractures, and nine flexion contractures. The feet were involved 16 times, including equinovarus deformity in 14 feet and calcaneovalgus deformity in the remaining two feet. In the upper limbs, four shoulders had adduction contractures, twelve elbows were involved by contractures (ten flexion and two extension contractures), and wrists and hands were involved 12 times. The spine was involved in two children (one had scoliosis and the other one had kyphoscoliosis). Table 1 shows the characteristics of the children. Eighty-one surgical procedures were performed in the lower and upper limbs (Table 2). The average number was 5.9 procedures per child. The average follow-up period was 3.6 years (range, 1.5–6 years).

The functional outcome assessment included walking ability and activities of daily living for upper limb function. Walking ability was assessed using Hoffer's classification,[10,13] as follows: independent ambulator walks without any aid; community ambulator is able to walk with aids in the community and does not need a wheelchair; household ambulator is able to walk with aids in the household and use a wheelchair in the community; nonfunctional ambulator uses a wheelchair and is capable of transfer; nonambulator always uses a wheelchair and is not capable of transfer.

Results

At the last follow-up visit, six (43%) children (four males, two females) with a mean age 8.3 years (range, 4–15) were independent walkers. They had excellent range of motion, with residual mild flexion contracture (less than 15°) in four hips. Knee motion in these patients was good, with mild limitation of motion. One patient still had mild bilateral flexion contracture of both knees. All of these patients had plantigrade feet, but one patient had significant recurrent adductus deformity. Although some of these patients still have contractures in the upper limbs, all of them can perform their daily activities without limitation. One patient (male) has severe kyphoscoliosis that requires surgery, but the family has refused.

Three children (males) with a mean age of 3.5 years (range, 2.5–5) were able to walk, but with support. They had decreased range of motion in the hips and knees. One still had mild flexion contracture of the knees, with external rotation contracture of the hips. All of these patients had plantigrade feet. The upper limbs were not involved in these patients.

One child (male) was a household ambulator at the age of 3 years. He was able to walk with bilateral, long-leg orthosis, but he sometimes needs more support and uses a wheelchair for community ambulation. This patient has bilateral hip dislocation. Open reduction was performed on one hip at the age of 8 months, but redislocation occurred, and he required revision surgery at 12 months. The other hip was not operated on. He had limitation of motion of both hips, with bilateral residual flexion contracture of 30°. His knees were

Table 1 Clinical profile of children with arthrogryposis multiplex congenita

Order of patient	Sex	Age at last follow-up	Duration of follow-up	Hip	Knee	Foot	Shoulder	Elbow	Hand and wrist	Spine
1	M	6	4	Bilateral dislocation	Bilateral extension			Bilateral flexion contracture		
2	F	4	2.5	Bilateral dislocation	Bilateral extension					
3	M	6	4	Bilateral dislocation	Bilateral dislocation	Bilateral equinovarus	Bilateral adduction	Bilateral flexion contracture	Bilateral extension contracture	
4	M	4	3	Rt: dislocation	Rt: flexion contracture, Lt: extension contracture	Bilateral equinovarus			Bilateral flexion contracture with ulnar deviation	
5	M	4	3	Rt: dislocation		Rt: equinovarus		Bilateral flexion contracture	Bilateral flexion contracture, bilateral clasped thumbs	Scoliosis
6	M	5	4	Bilateral flexion contracture		Bilateral equinovarus				
7	M	2.5	2		Bilateral dislocation			Bilateral flexion contracture		
8	M	3	2.5	Bilateral abduction and external rotation contracture	Bilateral flexion contracture	Bilateral equinovarus				
9	F	2.5	1.5	Bilateral flexion contracture	Bilateral flexion contracture					
10	M	3	2	Bilateral dislocation		Rt: equinovarus				
11	M	6	5		Bilateral flexion contracture	Bilateral calcaneovalgus				
12	F	13	6	Bilateral dislocation	Bilateral flexion contracture			Bilateral flexion contracture	Bilateral flexion contracture	
13	M	8	5	Bilateral dislocation	Bilateral extension	Bilateral equinovarus	Bilateral adduction contracture		Bilateral flexion contracture	
14	M	15	6	Bilateral flexion contracture	Bilateral extension	Bilateral equinovarus		Bilateral flexion contracture	Bilateral flexion contracture	Kyphoscoliosis

Table 2 Surgical procedures performed

Type of surgery	Number	Range of age (months)	Mean of age (months)
Hip			
Open reduction (primary)	13	8–28	18.5
Open reduction (revision)	2	12–16	14
Pelvic osteotomy	11	16–28	20.8
Derotation varus osteotomy of femur with shortening	5	16–24	18.2
Open adductor tenotomy	8	9–28	18.1
Knee			
Posterior soft tissue release (primary)	9	8–24	17.9
Posterior soft tissue release (revision)	4	24–32	28.5
Distal femoral extension osteotomy	4	26–42	33.5
Quadriceps plasty	9	4–28	19.6
Foot			
Complete subtalar release (primary)	14	8–32	20.6
Complete subtalar release (revision)	2	26–66	36
Upper limb			
Posterior elbow release and triceps plasty	1		28
Carpal osteotomy	1		36

not affected. He had plantigrade feet after surgical correction of equinovarus deformity in the right foot. His upper limbs were not affected.

Three children (two males, one female) with a mean age 4.2 years (range, 2.5–6) are nonfunctional ambulators and are unable to walk, even with support. They require a wheelchair for ambulation but are capable of transfer. One of them (male) had unilateral hip dislocation, for which open reduction was performed. He does not have knee involvement. He had complete subtalar release for equinovarus deformity of the right foot. He had bilateral flexion contracture of the elbows and bilateral flexion contracture of the wrist joint, with bilateral clasped thumbs. In addition, he has progressive scoliosis.

Two children had bilateral flexion contracture of the knee joint, for which posterior soft tissue releases were performed. The female child has good range of motion after surgery, without recurrence of the deformity. She had limitations in upper limb function. The male child required revision surgery and distal femoral extension osteotomy, due to the recurrence of the flexion deformity, and he ended up with significant residual flexion contracture of both knees. In addition, he had calcaneovalgus deformity of both feet, for which he had surgical correction. The upper limbs were not involved.

The last child (male) was nonambulatory at the age of 5 years. He was dependent on a wheelchair and was not capable of transferring alone. He had severe involvement of all joints in the upper and lower limbs at birth (Figure 1). He had open reduction combined with pelvic and femoral osteotomies for bilateral dislocation of the hip joint and bilateral complete subtalar release for equinovarus deformity of both feet. In addition, he had bilateral dislocation of the knee joint. Involvement of the upper limbs include adduction contracture of both shoulders, extension contracture of both elbows, and flexion contracture of both wrists, resulting in severe limitation of upper limb function. In addition, this patient had recurrent attacks of chest infections, which required multiple hospital admissions.

A summary of the functional outcome is shown in Table 3.

Discussion

Hip joint contracture, with or without dislocation, is reported in approximately 80% of patients with AMC.[2,6] In our study, we had a similar prevalence, in that the hip joint was involved in twelve children (86%): eight children (57%) had dislocation (six had bilateral and two had unilateral dislocation) and four children (29%) had contractures without dislocation (three had bilateral flexion contractures and one had bilateral abduction with external rotation contracture). Hip contractures were treated by aggressive physical therapy. At the last follow-up, one child was an independent walker, two children were community walkers, and one

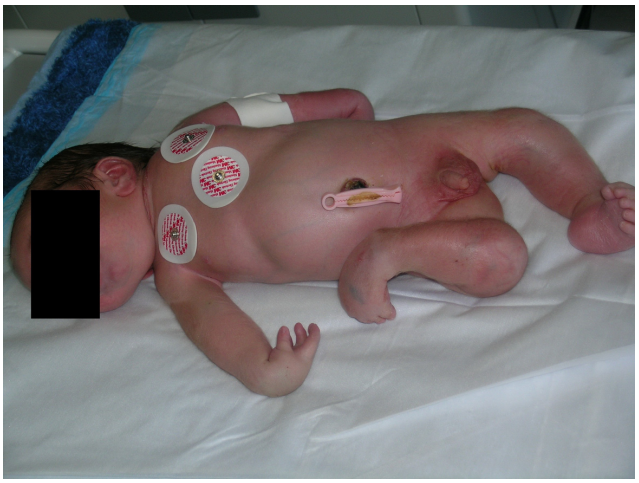

Figure 1 Clinical picture of the child who was nonambulatory at last follow-up, showing severe involvement of the upper and lower limbs at birth.

Table 3 Functional outcomes for children with arthrogryposis multiplex congenita

Functional status	Number of patients	Mean age (years)	Activities of daily living
Independent walker	6	8.3	Not affected
Walks with support	3	3.5	Not affected
Household walker	1	3	Not affected
Nonfunctional ambulator	3	4.2	Not affected (two patients) Limitation in one patient
Nonambulator	1	5	Severe limitation

child was nonfunctional ambulatory. It is generally agreed that unilateral dislocation in AMC should be reduced to prevent pelvic obliquity, sitting imbalance, and secondary scoliosis.[6,14–16] The management of bilateral dislocation, however, is controversial.[2] The high rate of complications, such as redislocation, stiffness, and avascular necrosis, has led to the recommendation that bilateral dislocation should be left unreduced.[1,2,6] Closed reduction to reduce hip dislocation in our patients failed; this result also has been reported previously.[2,15] Lloyd-Roberts and Leltim[15] operated on six patients with bilateral hip dislocation, and all of them ended up with stiff joints. Staheli et al[16] operated on three patients with bilateral dislocation in AMC. He performed a combination of open reduction, femoral varus-derotational osteotomy, and/or pelvic osteotomy, and showed good results. Akazawa et al[2] operated on five patients with bilateral dislocation in AMC and performed complete circumferential capsulotomies, but the incidence of avascular necrosis was high (70%). Huurman and Jacobsen[6] reported that better functional results could be obtained with subtrochanteric extension osteotomy. Asif et al[1] operated on four patients with bilateral dislocation in AMC. He performed open reduction in two patients (four hips); the other two patients (four hips) required additional bony procedure in the femur in all four hips and Salter innominate osteotomy in two hips. He reported good clinical and radiological results.

In our series, we operated on 13 hips of eight patients (14 hips) with AMC (six had bilateral dislocation and two had unilateral dislocation). All patients had open reduction and capsular plication through anterior approach using bikini incision and open adductor tenotomy through a separate medial incision. This procedure was combined with Salter innominate osteotomy in six patients (eleven hips). Five hips (three patients) required additional femoral shortening and varus derotational osteotomy to improve reduction, decrease the pressure on the femoral head, and provide further stability.

Two children with unilateral hip dislocation who underwent open reduction with capsular plication had redislocation after cast removal. Orthotic treatment to reduce the hip failed. Revision open reduction combined with Salter innominate osteotomy was performed on one child. This procedure was combined with femoral shortening and varus derotational osteotomy for the other one. They did well and no further complications occurred. At the last follow up, five children (four had bilateral and one had unilateral dislocation) were independent walkers, and they had good range of motion in the hip joint. One child who had bilateral dislocation was household ambulatory. One child with unilateral dislocation was nonfunctional ambulatory, and the last child with bilateral dislocation was nonambulatory. The complications and the outcome of surgery were almost similar for both unilateral and bilateral dislocation.

Knee joint involvement has been reported in 70% of patients with AMC, with flexion contractures being the most common, followed by extension contractures.[4,7] In our series, eleven children (79%) had knee involvement, and extension and flexion contractures were equal (nine joints each). Two children had bilateral knee dislocation, four had bilateral extension contracture, four had bilateral flexion contracture, and one child had flexion contracture of one knee and extension contracture of the other one. Conservative treatment, which included serial casting and physical therapy, was effective in two children only (four knees) who had bilateral extension contractures. The remaining nine children underwent surgical release of the contractures early, because of the failure of conservative treatment. Flexion contractures were treated by posterior capsular release and hamstring tenotomy in five children (nine knees). Two of those children (four knees) had recurrence of the deformity, which was treated first by revision of the procedure, and then by distal femoral extension osteotomy. Extension contractures were treated by quadriceps plasty in five children (nine knees). At the last follow-up visit, six children were independent walkers; they had good range of motion in the knees. Two children were community ambulators, two were nonfunctional ambulators, and the last child was nonambulatory.

The feet were involved in nine children (64%). Rigid equinovarus was the most common deformity, which was present in 14 feet of eight children (six bilateral and two unilateral deformities). Bilateral calcaneovalgus deformity was present in one child. Most authors reported that more radical approaches, such as talectomies, are required either as a primary procedure or after failure of less radical treatment.[17–19] In our series, complete subtalar release, using a Cincinnati

incision, was used as a primary procedure to treat rigid deformity in 14 feet and as a revision for two feet that had recurrence of the deformity. At the last follow-up, seven children had plantigrade feet (two of them were independent walkers, two were community ambulators, two were nonfunctional ambulators, and the last one was a household ambulator). The remaining two children (four feet) had recurrent equinovarus deformity that need to be corrected; one was an independent walker and the other one was nonambulatory.

The upper limbs were involved in eight children (57%). Bilateral adduction contractures of the shoulders were present in two children, six children had bilateral contractures of the elbows (five had flexion and one had extension contractures), and six children had bilateral flexion contractures of the wrist joint. Conservative treatment was used to control the deformities. Only two surgical procedures were performed on the upper limbs: posterior release of the elbow with triceps lengthening in one child, and carpal osteotomy to correct flexion contracture in another one. At the last visit, all the children in the study were able to use their upper limbs for activities of daily living without significant limitations, except for two: one was nonambulatory and the other was the nonfunctional ambulator.

Spine deformity was present in two children (14%), which was lower than what has been reported by other studies.[20,21] One was an independent walker with severe kyphoscoliosis in the upper thoracic spine, and the other one was the nonfunctional ambulator, who had scoliosis.

In our short-term follow-up for children with AMC who had multiple surgical procedures at an early age, 43% were independent walkers. Activities of daily living were not affected in the majority of the children.

We believe that aggressive surgical treatment using multiple operations at an early age can improve the short-term functional and clinical outcomes for children with AMC.

Disclosure

No financial support was given from any organization and the authors report no conflicts of interest in this work.

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