**Original article**

**Assessment of outcome of Quadricepsplasty in Knee contractures of patients with arthrogryposis**

*Moghadam MH¹, BirjandiNejad A², Ghoreishi SA³*

**Abstract**

Arthrogryposis multiplex congenita (AMC) refers to a group of conditions, unrelated to one another, which are similarly characterized by the contracture of multiple joints in the body. One of the common therapies in patients with AMC and knee extension contracture is quadricepsplasty. However, no comprehensive studies have been conducted on the results of the application of this method.

This prospective study included 14 patients (9 females and 5 males) with 23 hyperextension contractures of the knee. Before surgery, all patients had undergone non-surgical treatment (manipulation and serial casting) for at least 6 months, but none had responded to the treatment. Then patients underwent a Quadricepsplasty. Range of motion (ROM) of the knee and patients ambulation based on modified form of Functional Mobility Scale (FMS) before and after surgery were evaluated in a follow up 3 years.

The mean passive knee ROM before surgery was 7.17° (0° to 15°), which increased to 65.65° (35° to 105°) after the operation. According to t-test results, this increase was statistically significant (P<0.005).

The mean ambulation score (modified FMS) of the patients before surgery was 1.57, which increased to 3.71. According to t-test results, this increase was statistically significant (P<0.005).

Quadricepsplasty is the method of choice for the surgical treatment of knee extension contracture in AMC patients, who do not respond to non-surgical treatments. This method leads to a significant improvement in knee ROM. On the other hand, it preserves quadriceps strength at 4/5 level, and finally, along with other therapeutic interventions for other lower limb deformities, leads to ambulation improvement in these patients.

**Introduction**

Arthrogryposis multiplex congenita (AMC) refers to a group of conditions, unrelated to one another, which are similarly characterized by the contracture of multiple joints in the body ¹. AMC was first described by Otto in 1841 and was named by Stem ². The most common clinical presentation in AMC patients is the loss of joint movements, which can be caused by multiple disorders including neuropathy, muscular abnormality, and connective tissue disorders ³. There are approximately 150 conditions or syndromes, which are clinically presented by joint contractures ⁴.

AMC is a non-progressive disorder in which the patient suffers from joint contractures and muscle defects, while the organ sensations remain normal ⁵,⁶. Arthrogryposis is multifactorial in etiology and involves factors such as myopathic processes, connective tissue abnormalities, teratogenic factors, and genetic factors, which may all decrease fetal movements⁷.

Most cases of arthrogryposis are sporadic without

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any hereditary risks. This condition is inherited in an autosomal dominant pattern, particularly in case of distal arthrogryposis. The patterns of inheritance have been reported as recessive autosomal and x-linked recessive inheritance. The prevalence of arthrogryposis is approximately 1 per 3000 live births, and is classified into three major groups:

1- disorders with the involvement of main limbs [all four limbs (84%), only the lower limbs (11%), and only the upper limbs (5%)],

2- disorders with the involvement of limbs and other body organs, and

3- disorders with limbs involvement and central nervous system (CNS) dysfunction.

Multiple subtypes have been identified for arthrogryposis including distal arthrogryposis and amnioplasia, which is considered the most common subgroup.

The involvement of lower limbs in children with AMC is characterized by the hip flexion-abduction deformity or dislocation, knee involvement as flexion or extension contracture (flexion type is more common), and foot involvement as clubfoot or vertical talus.

Hip and knee functions are required for ambulation. Knee contracture in AMC is a debilitating deformity, and the patient should have at least 15-20 degrees of knee flexion for ambulation.

There are various treatments for modifying knee contractures including serial stretch casting (as the initial treatment) and surgical procedures involving quadricepsplasty, distal femoral extension, osteotomy, and the Ilizarov method of gradual correction.

One of the common therapies in patients with AMC and knee extension contracture is quadricepsplasty. However, no comprehensive studies have been conducted on the results of the application of this method. In this study, we aimed to evaluate the results of quadricepsplasty in patients with AMC and extension contracture of the knee to help provide treatment plans for these patients.

**Materials and methods**

This prospective study included 14 patients (9 females and 5 males) with 23 hyperextension contractures of the knee. The subjects underwent quadricepsplasty from 2007 to 2010 in the orthopedic ward of Imam Reza Hospital, Mashhad University of Medical Sciences, and were followed up prospectively for an average of three years.

**Table 1: The modified Functional Mobility Scale (FMS)**

<table>
<thead>
<tr>
<th>Ambulation method</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>The patient can walk without assistive devices on all surfaces</td>
<td>6</td>
</tr>
<tr>
<td>The patient can walk without assistive devices only on flat surfaces</td>
<td>5</td>
</tr>
<tr>
<td>The patient can walk with a walking stick</td>
<td>4</td>
</tr>
<tr>
<td>The patient can walk with a crutch</td>
<td>3</td>
</tr>
<tr>
<td>The patient can walk with a walker</td>
<td>2</td>
</tr>
<tr>
<td>The patient can only move by a wheelchair</td>
<td>1</td>
</tr>
</tbody>
</table>

All patients were diagnosed with the amnioplasia type of arthrogryposis along with hyperextension contracture of one or both knees (nine patients with bilateral involvement and others with unilateral involvement). The mean age of the subjects for surgery was 18 months (age range: 14-26 months). The deformities of the lower limbs in these patients included bilateral equinovarus foot deformity (10 patients), bilateral vertical talus (4 patients), unilateral hip dislocation (5 patients), bilateral hip dislocation (4 patients), and knee subluxation (6 patients).

Before surgery, all patients had undergone non-surgical treatment (manipulation and serial casting) for at least 6 months, but none had responded to the treatment.

Prior to surgery, all subjects were clinically examined. The active and passive range of knee motion (ROM) was calculated by an orthopedic ruler, and patient ambulation was evaluated based on the modified model of Functional Mobility Scale (FMS), which was completed by the physician (Table 1).

Before performing the surgery, seven patients were not able to walk with assistive devices. Five patients could walk with a walker, AFO (Ankle-Foot Orthosis), or KAFO (Knee-Ankle-Foot Orthosis); two patients were able to walk with a cane and without brace.

The surgical procedure was as follows: a longitudinal midline incision on the knee, extending proximally, along with medial arthrotomy, released the iliobibial...
band from tibia, and vastus lateralis from rectus femoris and quadriceps tendon; then, through medial arthrotomy, quadriceps and patellar tendons became fully mobile. Afterwards, all intra-articular fibrous tissues were debrided and eliminated, and a V-shaped incision was made on the quadriceps tendon using Y-V lengthening method, with the base toward the patella\(^\text{10}\). Finally, the knee was extended to maximum flexion and was repaired and sutured with a non-absorbable nylon suture of the tendon in the form of Y, while the knee was flexed at 90°.

After the operation, the knees were immobilized at 90° flexion using a long-leg cast for 3 weeks. After opening the cast, the joint movements of the knee were started, and the physiotherapist increased the ROM and muscle strength for one month; physiotherapy continued for at least 3 months by the parents at home.

The patients were re-examined 3 years (on average) after surgery (2 to 4 years). The knee ROM, the quadriceps muscle strength, and the ambulation (based on the modified FMS) were again reviewed and recorded, and all the data were analyzed using SPSS.

Table 2: Summary of the patients’ data

<table>
<thead>
<tr>
<th>Column</th>
<th>Sex</th>
<th>Age at surgery (month)</th>
<th>Knee side (L: left, R: right)</th>
<th>Passive flexion</th>
<th>Passive extension</th>
<th>ROM</th>
<th>FMS</th>
<th>Quadriceps muscle strength</th>
<th>Follow-up duration (year)</th>
</tr>
</thead>
</table>
Results
In this study, 14 patients (23 knees) with AMC and extension contracture of the knee were studied. The subjects were treated using the quadricepsplasty method, and the following results were obtained over an average of 3-year follow-ups (Table 2).
None of the patients had local skin problems, infections, neurovascular damages, or other surgical complications. In the clinical evaluation after quadricepsplasty, the strength of quadriceps muscle was 3/5 to 5/5 in all the patients (average of 4/5).
The mean passive flexion of the knee was 1.09° (-10° to 10°) before the surgery, which increased to 67.30° (35° to 105°) after the operation. The mean passive extension of the knee was 6.08° (-5° to 20°) before the surgery, which changed to -0.65° after the operation (-5° to 0°).
The mean passive knee ROM before surgery was 7.17° (0° to 15°), which increased to 65.65° (35° to 105°) after the operation. According to t-test results, this increase was statistically significant (P<0.005) (Table 3).
The mean age at surgery was 18 months (range: 10 to 26 months), and according to Pearson’s correlation test, there was no relationship between age at surgery, ROM change, and the patient ambulation. The mean ambulation score (modified FMS) of the patients before surgery was 1.57, which increased to 3.71. According to t-test results, this increase was statistically significant (P<0.005) (Table 3).

Table 2: t-test results with regard to knee ROM and patient ambulation

Table 3: t-test results with regard to knee ROM and patient ambulation

<table>
<thead>
<tr>
<th>Variables</th>
<th>The mean before the surgery</th>
<th>The mean after the surgery</th>
<th>SD before the surgery</th>
<th>SD after the surgery</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Knee ROM</td>
<td>7.17</td>
<td>66.65</td>
<td>4.47</td>
<td>16.94</td>
<td>0.000</td>
</tr>
<tr>
<td>Ambulation (FMS)</td>
<td>1.57</td>
<td>3.71</td>
<td>0.64</td>
<td>1.20</td>
<td>0.000</td>
</tr>
</tbody>
</table>

Discussion
In AMC patients with extension contracture of the knee (similar to other joints), non-surgical procedures such as passive stretching and serial casting or splint are initially performed 11,12.
Palmer and colleagues in their study showed that passive motion improves the joint ROM in these patients 13.
Ryoppy and So¨dergard in their study reported the positive results of non-surgical treatment 14. Similarly, Murrary et al. showed the significant results of knee non-surgical treatment in AMC patients15. In the study of Drumand et al., it was found that manipulation treatment does not improve the knee ROM in AMC patients 14. The failure of manipulation treatment in the improvement of knee ROM is an indication for performing surgery 15.
Patrica and colleagues in their long-term review found that non-surgical treatment (manipulation and casting) causes damage to articular cartilage, which leads to early degenerative changes in these patients 17.
Some authors consider 6 to 3 months of age as the best time for performing surgery. One of the factors leading to the failure of non-surgical treatment for knee extension contracture in AMC patients is the age older than 1 to 2 years, during which the results are poor 17.
Some other researchers suggested that the optimal time for surgery is when the non-surgical treatment fails and the patients are 6 to 12 months of age 1. However, in the current study, age at surgery ranged from 10 to 26 months (mean: 17 months, SD: 3.03), and no association was found between the results of surgery and age at surgery in this age range.
Another indication for early surgery in these patients is severe extension contracture, i.e., the knee cannot be flexed more than 35° 18.
Most researchers believe that quadricepsplasty is the best method to improve joint ROM and preserve the strength of quadriceps muscle 14,15,18. In this study, the surgical method was used, which both improved the joint Rom and preserved the strength of quadriceps muscle at 4/5.
Patrica and colleagues in a study, which evaluated the long-term results of quadricepsplasty in knee contracture extension found similar results, and reported that surgical intervention improved joint ROM and preserved the strength of the quadriceps; it also improved the patients’ gait 17. The results of the study by Murary et al. confirmed our findings 15. In this study, most patients had deformities in the lower limbs and all the patients were treated for these deformities in 3-year follow-ups. We observed a significant increase in the ambulation improvement, based on the modified FMS after the 3-year follow-ups, which shows the effectiveness of orthopedic treatment in the improvement of the patient’s function and ambulation.
The ambulation did not significantly improve in 2...
patients. This is due to the involvement of the hip and even the ankle, in addition to knee function, in the ambulation. In these two patients, due to lack of hip function improvement during therapy, despite the improved knee function, no progress was observed in the ambulation.

One of the strengths of this study is that the number of our patients was higher than other previous studies. On the other hand, one of the study limitations is the short-term 3-year follow-ups for the evaluation of the results.

**Conclusion**

Quadricepsplasty is the method of choice for the surgical treatment of knee extension contracture in AMC patients, who do not respond to non-surgical treatments. This method leads to a significant improvement in knee ROM. On the other hand, it preserves quadriceps strength at 4/5 level, and finally, along with other therapeutic interventions for other lower limb deformities, leads to ambulation improvement in these patients.

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**References**