

# Surgical Treatment of Hip Dislocation in Children With Arthrogyryposis Multiplex Congenita

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## Abstract

**Aim:** Arthrogyryposis multiplex congenital (AMC) is a rare disease with multiple joint contractures. In unilateral hip dislocation surgical treatment to reduce the hip is recommended to prevent pelvic obliquity and scoliosis. It is commonly believed that bilaterally dislocated hips should not be reduced because movement is satisfactory and open reduction leads to poor results. The purpose of this study is to report our clinical and radiological outcome of the surgical treatment of unilateral and bilateral hip dislocation in 5 patients (8 hips) having AMC.

**Methods:** During the period 2006 to 2010, we operated on 8 hips of 5 children with AMC (3 patients had bilateral hip dislocation and 2 patients had unilateral hip dislocation). Open reduction with capsular plication without any bony procedure was performed on 2 hips. Open reduction combined with Salter osteotomy of innominate bone was performed on 2 hips. Open reduction combined with Salter osteotomy of innominate bone and femoral shortening with varus derotational osteotomy was performed on 5 hips. In addition to that, open adductor tenotomy was done in all cases through a separate medial incision. The mean age at surgery was 16.6 months (range: 9-22 months) and the average follow up period was 2.3 years (range: 1-3 years).

**Results:** At the final follow up, two children were able to walk independently. One child had a unilateral dislocation and required a long leg brace and support for walking. One child had a unilateral dislocation and required a short leg brace for walking. One child was unable to walk because of severe contractures in all four limbs, and one child required revision surgery for a re-dislocation of the hip joint. The clinical results were good in 5 hips, fair in 1 hip, and poor in 2 hips. According to the Severin classification, all 8 hips were rated as good (5 hips in class I and 3 hips in class II). There was a low incidence of avascular necrosis where only one hip was in grade 1 according to Kalamchi classification, while 7 hips did not show any evidence of avascular necrosis.

**Conclusion:** We believe that open reduction combined with bony procedures should be performed for unilateral and bilateral dislocation of the hip in children with AMC, and it should be done at an earlier age to achieve the best functional outcome.

**Keywords:** Hip Dislocation, Arthrogyryposis, Pelvic Obliquity.

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## **Introduction**

Arthrogryposis multiplex congenital (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.<sup>1-3</sup> The incidence of AMC is rare, occurring in approximately 0.03% of newborn infants.<sup>1,3</sup>

Hip joint contracture, with or without dislocation, is reported in around 80% of patients with AMC.<sup>2,4</sup> Isolated contractures are usually treated conservatively by manipulation and splinting and only occasionally require operative intervention. Closed reduction of the dislocated hip in children with AMC has been relatively unsuccessful.<sup>2,5,6</sup>

In unilateral hip dislocation, open reduction is recommended to prevent pelvic obliquity, imbalance in sitting and scoliosis.<sup>1, 2, 4, 5</sup> For bilaterally dislocated hips, some authors believe that the hips should not be reduced since the pelvis remains level and movement is satisfactory<sup>5, 7, 8</sup> and open reduction has been associated with a poor outcome.<sup>6</sup> Others suggest that all dislocations should be reduced to restore normal hip mechanics and decrease the risk of future pain or stiffness.<sup>2, 9, 10</sup> We present the functional and radiological results in 5 children with AMC who were treated for both unilateral and bilateral dislocation of hip joint by open reduction combined with bony procedure in most cases performed at an early age since 2006.

## **Patients and Methods**

This is a retrospective study of AMC patients who underwent surgical treatment for dislocation of the hip (5 patients with 8 hips) at the department of Orthopedic Surgery in King Abdullah University Hospital from 2006 to 2010. Patients included in our study are those who had joint contracture at birth in at least 2 different parts of the body, generalized muscle wasting and absence of normal skin creases. Five children with 8 dislocated hips were treated. All of them had associated contractures in the upper and lower limbs (Table 1). Two patients had bilateral

dislocation of the knee, one patient had unilateral knee dislocation and flexion contracture of the other knee and one patient had flexion contracture of both knees. Two patients had bilateral talipes equinovarus and one patient had unilateral talipes equinovarus. Two patients had extension contractures of both elbows and one patient had flexion contracture of both elbows. Three patients had flexion contracture of both wrists and one patient had finger contracture and a clasped thumb (Table 1). Four patients were males and one was female. The mean age at surgery was 16.6 months (range: 9-22 months) and the average follow up period was 2.3 years (range: 1-3 years). In patients with hip and knee dislocation, the knee was operated on before the hip. This was done to facilitate positioning during hip surgery and the application of hip spica.

The preoperative clinical and radiographic assessments were recorded. The mean fixed flexion contracture at the hip was 30°, mean flexion 60°, mean abduction 20°, mean adduction 20°, mean internal rotation 30°, and mean external rotation 35°. Open reduction through the anterior approach using bikini incision was performed. All the tight muscles, sartorius, iliopsoas and rectus femoris were incised. Any fibrous adhesions were dissected for thorough exposure of the joint capsule. The capsule of the hip joint was opened in a "T" shape. The acetabulum was cleared of any soft tissue. Transection of the transverse acetabular ligament was done. The ligamentum terese was removed. Capsular plication was performed. Open adductor tenotomy through a separate medial incision was performed in all cases. A complete reduction was confirmed by intra-operative radiography. Four patients (7 hips) required additional procedures, which included Salter innominate osteotomy in 4 patients (7 hips) and femoral shortening and varus de-rotational osteotomy in 3 patients (5 hips). Then the hip spica was applied with hip in slight flexion, slight internal rotation and 45° of abduction for 6 weeks. After that, it was changed to a petrie cast for another 6 weeks. Then the patients were allowed gradual ambulation and range of motion exercises without bracing or physiotherapy.

Follow up examination included assessment of the range of motion, walking and radiography. Motion in the joint was evaluated according to Gruel et al <sup>11</sup> criteria. Anteroposterior radiographs of each hip were graded according to the classification of Severin <sup>12</sup> and avascular necrosis by the classification of Kalamchi and MacEwen. <sup>13</sup>

**Results**

At the final follow up, the range of motion was good in 5 hips where flexion contracture was less than 5°, mean flexion was 100°, mean abduction 35°, mean adduction 10°, mean internal rotation 40°, and mean external rotation 40°. Fair in one hip where flexion contracture was 10°, flexion was up to 85°, abduction was 10°, adduction was 10°, internal rotation was 15° and external rotation 20°. Also, poor in 2 hips where flexion contracture was 20°, flexion was less than 80°, abduction and adduction were less than 10°, and internal and external rotation were less than 10° (table 2). Also, 5 hips were in class I and 3 hips were in class II according to the Severin classification.

Avascular necrosis was seen in one hip only which was in grade 1 according to Kalamchi (table2). Two of the five children were able to walk independently without crutches or support. One child had a unilateral dislocation and required a long leg brace and support for walking. One child had a unilateral dislocation and required a short leg brace for walking. One child who had a bilateral hip dislocation was unable to walk because he was having other severe joint contractures in the knees and feet as well as contractures in the elbows and wrist joints. In addition to that, he had recurrent attacks of a respiratory tract infection for which he required frequent hospitalization.

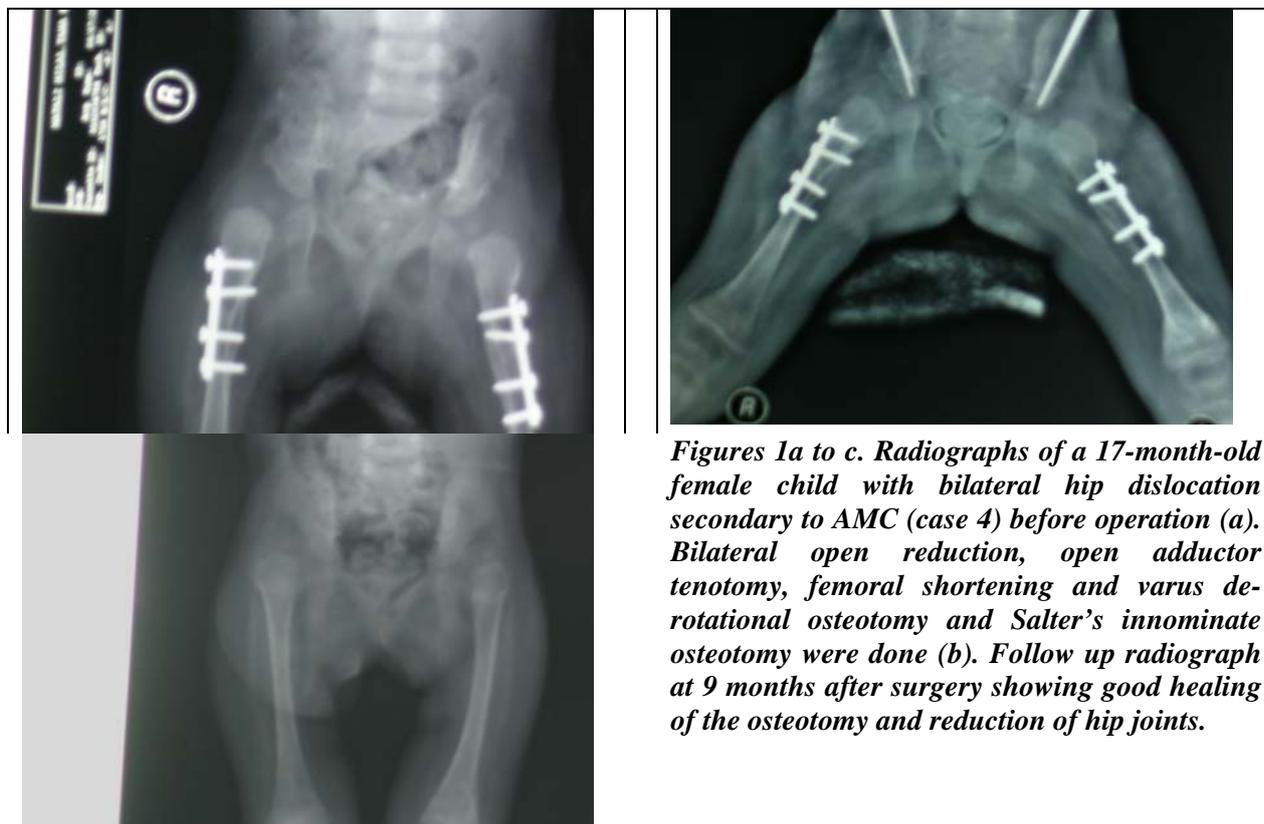
There was one complication in one hip where open reduction and open adductor tenotomy was performed without a bony procedure. Re-dislocation occurred after removal of casting. He was treated by revision open reduction combined with Salter innominate osteotomy and femoral shortening with derotational varus osteotomy. The patient did well after this revision and no re-dislocation occurred again.

**Table (1): Clinical details of the patients.**

<i>Case</i>	<i>Sex</i>	<i>Age at surgery (months)</i>	<i>Follow up (months)</i>	<i>Surgery performed</i>	<i>Other affected regions</i>
<i>1</i>	<i>M</i>	<i>Rt; 24 Lt; 28</i>	<i>Rt; 34 Lt; 30</i>	<i>Rt; Open reduction, open adductor tenotomy, Salter osteotomy Lt; same</i>	<i>Extension contracture of both knees, flexion contracture of both elbows.</i>
<i>2</i>	<i>M</i>	<i>Rt; 9 (primary surgery) Revision: 16</i>	<i>Rt; 22(after revision)</i>	<i>Primary; Open reduction, open adductor tenotomy. Revision; Open reduction, Salter osteotomy, femoral shortening with varus derotational osteotomy</i>	<i>Right knee flexion contracture, left knee extension contracture, bilateral equinovarus, l ulnar deviation of the both wrists.</i>
<i>3</i>	<i>M</i>	<i>Rt; 16 Lt; 24</i>	<i>Rt;32 Lt;21</i>	<i>Rt; Open reduction, open adductor tenotomy Salter osteotomy, femoral shortening with varus derotational osteotomy. Lt; same</i>	<i>Bilateral knee dislocation, , bilateral equinovarus, flexion contracture of both elbows, l flexion contracture of both wrists, contractures of both shoulders</i>
<i>4</i>	<i>M</i>	<i>Rt; 9</i>	<i>Rt; 22</i>	<i>Rt; Open reduction, open adductor tenotomy</i>	<i>Right equinovarus , flexion contracture of both elbows, flexion contracture of both wrists, bilateral clasped thumbs, scoliosis</i>
<i>5</i>	<i>F</i>	<i>Rt; 17 Lt; 18</i>	<i>Rt;13 Lt; 12</i>	<i>Rt; Open reduction, open adductor tenotomy Salter osteotomy, femoral shortening with varus derotational osteotomy. Lt; same</i>	<i>Extension contracture of both knees, flexion contracture of fingers of both hands.</i>

**Table (2): Clinical and radiological outcome of the patients.**

<u>Case</u>	<u>Sex</u>	<u>Side</u>	<u>Gruel classification</u>	<u>Severin classification</u>	<u>Avascular necrosis (Kalamchi)</u>
1	M	Rt	Good	I	None
		Lt	Good	I	None
2	M	Rt	Good	II	None
3	M	Rt	Poor	II	I
		Lt	Poor	II	None
4	M	Rt	Fair	I	None
5	F	Rt	Good	I	None
		Lt	Good	I	None



**Figures 1a to c. Radiographs of a 17-month-old female child with bilateral hip dislocation secondary to AMC (case 4) before operation (a). Bilateral open reduction, open adductor tenotomy, femoral shortening and varus derotational osteotomy and Salter's innominate osteotomy were done (b). Follow up radiograph at 9 months after surgery showing good healing of the osteotomy and reduction of hip joints.**

**Discussion**

The dislocated hip in a patient with AMC is usually defined as a teratological dislocation that has occurred early in fetal development. Closed reduction invariably fails.<sup>2, 6</sup> It is generally agreed that unilateral dislocation in AMC should be reduced to prevent pelvic obliquity and secondary scoliosis. The management of bilateral dislocation, however, is controversial.<sup>2</sup> The high rate rate of complications such as re-dislocation, stiffness and avascular necrosis has led to the recommendation that bilateral dislocation should be left unreduced.<sup>1, 2, 4</sup>

Lloyd et al.<sup>6</sup> operated on 6 patients with bilateral hip dislocation and all of them ended up with stiff joints. Staheli et al.<sup>9</sup> operated on 3 patients with bilateral dislocation in AMC. He did a combination of open reduction, femoral varus derotational osteotomy and/or pelvic osteotomy and showed good results. Akazawa et al.<sup>2</sup> operated on 5 patients with bilateral dislocation in AMC in which complete circumferential capsulotomy was done, but the incidence of avascular necrosis was high (70%). Huurman and Jacobsen<sup>4</sup> reported that better functional results may be obtained using subtrochanteric extension osteotomy. Asif et al<sup>1</sup> operated on 4 patients with bilateral dislocation in AMC.

He did open reduction only in 2 patients (4 hips) and the other 2 patients (4 hips) required additional bony procedure in the femur in 4 hips and Salter innominate osteotomy in 2 hips. He reported good clinical and radiological results.

In our series, we operated on 8 hips of 5 patients with AMC (3 had bilateral dislocation and 2 had unilateral dislocation). All patients underwent open reduction and capsular plication through anterior approach using bikini incision and open adductor tenotomy through a separate medial incision. This was combined with Salter innominate osteotomy in 4 patients (7 hips). Five hips (3 patients) required additional femoral shortening and varus derotational osteotomy to improve reduction, decrease the pressure on the femoral head and provide further stability.

There was one complication in our series. In one patient with unilateral hip dislocation, open reduction with capsular plication only was done. The patient developed re-dislocation after cast removal. Orthotic treatment to reduce the hip failed. Revision open reduction combined with Salter innominate osteotomy and femoral shortening with varus derotational osteotomy was done. The patient did well and no further complications occurred.

At the final follow up, 5 hips had a good range of motion, one hip had fair and 2 hips had poor range of motion. Two of 5 patients were able to walk independently without crutches or support. One patient had unilateral dislocation and required a long leg brace and support for walking. One patient had unilateral dislocation and required a short leg brace for walking. One patient that had bilateral dislocation was non ambulant, and he was having severe contractures with a marked limitation of motion of the upper and lower limbs. Also, this patient had recurrent attacks of respiratory tract infections which required frequent hospitalization.

In our series, we have a low incidence of avascular necrosis since most of our cases (7 hips) did not show any evidence of avascular necrosis and only one hip was in grade I according to Kalamchi classification. We believe

that femoral shortening with varus derotational osteotomy has an important role in this result because it decreased the pressure on the femoral head and enhanced the stability of reduction. This may have decreased the risk for avascular necrosis and led to an improvement in the functional outcome.

The complications and the outcome of surgery were almost similar for both unilateral and bilateral dislocation. Although our series is small and there is no non-surgical control group to draw statistical conclusion, we believe that both unilateral and bilateral hip dislocation in AMC patients should be treated at an early age with an extensive surgical approach which includes open reduction, open adductor tenotomy combined with femoral shortening and pelvic osteotomy to achieve the best clinical and radiological outcome.

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## العلاج الجراحي لخلع الورك في الأطفال الذين يعانون من اعوجاج المفاصل الخلقية المتعدد

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### الملخص

**الهدف:** اعوجاج المفاصل لمتعدد الخلقية هو مرض نادر، وهو يتميز بوجود تقوس وتشنج في معظم مفاصل الجسم في حالات خلخ الورك في جانب واحد من المستحسن العلاج الجراحي للحد من انحراف الحوض واعوجاج العمود الفقري. ويعتقد عموماً أنه لا ينبغي التدخل الجراحي في حالات خلخ الوركين ثنائياً، وذلك لأن الحركة عادة تكون مقبولة بينما العلاج الجراحي قد يؤدي إلى نتائج سيئة. والغرض من هذه الدراسة هو تقرير النتائج السريرية والإشعاعية لدينا من العلاج الجراحي لخلخ الورك الأحادية والثنائية في 5 مرضى اردنيين (8 مفاصل وركيه).

**الأساليب:** خلال الفترة 2006 حتي 2010 ، تم اجراء عمليات جراحية 8 مفاصل وركيه ل 5 اطفال يعانون من اعوجاج المفاصل المتعدد الخلقية ( 3 من المرضى الذين لديهم خلخ في مفصلي الورك معاً و 2 من المرضى يعانون من جانب واحد في خلخ الورك). تم إجراء عملية ارجاع المفصل عن طريق فتح المحفظة المفصالية ومن ثم ترميم المحفظة دون أي إجراء عظمي في مفصلي اثنين. وأجريت عملية فتح للمفصل جنباً إلى جنب مع قص العظم الورك في مفصلي اثنين. كما تم عملية فتح المفصل جنباً إلى جنب مع قص عظم الورك وتقصير عظم الفخذ في 5 مفاصل وركيه. وبالإضافة إلى ذلك ، تم فتح الوتر المقربة في جميع الحالات من خلال شق وسطي منفصل. وكان متوسط العمر عند اجراء الجراحة 16,6 شهراً (9 - 22 شهراً) ومتوسط فترة المتابعة كان 2.3 سنة (1-3 سنوات).

**النتائج:** عند نهاية الدراسة كان اثنان من الأطفال قادرين على المشي بشكل مستقل ، وطفل واحد (قد خلخ من جانب واحد) احتاج لجبيرة طويلة الساق والمساعدة في المشي. طفل واحد (قد خلخ من جانب واحد) احتاج لجبيرة قصيرة الساق للمشي.

وطفل واحد كان غير قادر على المشي بسبب شدة التقلصات في أطرافه الأربعة. طفل واحد تطلب إعادة اجراء الجراحة له نتيجة لعودة الخلخ من مفصل الورك. وكانت النتائج السريرية جيدة في 5 مفاصل وركيه ومتوسطة في مفصل واحد وسيئة في مفصلي اثنين. اما النتائج الاشعاعية على تصنيف (severin) فقط صنفت جميع المفاصل ال 8 جيدة (5 مفاصل في الدرجة الأولى و 3 مفاصل في الدرجة الثانية). كان هناك معدل منخفض للنخر الوعائي في مفصل واحد حيث كان في الدرجة (1) وفقاً لتصنيف Kalamchi ، في حين أن 7 من المفاصل لم تظهر أي دليل على نخر وعائي.

**الخلاصة:** نعتقد أن العلاج الجراحي عن طريق فتح المفصل مع قص العظم ( سواء كان عظم الفخذ او عظم الورك او كليهما معا ) يجب استخدامه في علاج حالات خلخ المفصل الورك ( الاحادي الجانب او الثنائي ) للاطفال الذين يعانون من مرض اعوجاج المفاصل المتعدد الخلقية، ويجب استخدامه في عمر مبكر للحصول على افضل النتائج .

**الكلمات الدالة:** خلخ الورك، اعوجاج المفاصل، انحراف الحوض.