Surgical management of hip dislocation in children with arthrogryposis multiplex congenita

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Arthrogryposis multiplex congenita (AMC) is a rare disease with multiple joint contractures. It is widely believed that bilaterally dislocated hips should not be reduced since movement is satisfactory and open reduction has had poor results. Since 1977 we have performed a new method of open reduction using an extensive anterolateral approach on ten hips in five children with AMC. The mean age at surgery was 31.5 months (17 to 64) and the mean follow-up was 11.8 years (3.8 to 19.5).

At the final follow-up all children walked without crutches or canes. Two managed independently, one required a long leg brace and two had short leg braces because of knee and/or foot problems. The clinical results were good in eight hips and fair in two and on the Severin classification seven hips were rated as good (group I or group II).

We recommend the extensive anterolateral approach for unilateral or bilateral dislocation of the hip in children with arthrogryposis or developmental dislocation of the hip.

J Bone Joint Surg [Br] 1998;80-B:636-40.

Received 29 July 1997; Accepted after revision 1 December 1997

Arthrogryposis multiplex congenita (AMC) is an uncommon congenital disease in which there is multiple involvement of joints with severe contractures, dislocations, limited muscle strength, and a lack of normal skin creases. An incidence of hip contractures of 80%, with or without dislocation, has been reported in patients with AMC.¹

Isolated contractures are usually treated conservatively by manipulation and splinting and only occasionally require operative intervention, but closed reduction of the dislocated hip in children with AMC has been relatively unsuccessful.^{2,3}

In unilateral dislocation, open reduction is recommended through an anterior approach to prevent pelvic obliquity and scoliosis.^{1,2} Treatment by the medial approach is seldom used since it gives limited access to the hip. By contrast, in bilateral dislocation, some authors believe that the hips should not be reduced since the pelvis remains level and movement is satisfactory,^{2,4,5} while others suggest that all dislocations should be reduced to restore normal hip mechanics and decrease the risk of future pain or stiffness.^{6,7}

Since 1977 we have performed a new method of open reduction using an extensive anterolateral approach in children with AMC and with developmental dislocation of the hip.⁸ As the exact aetiology and pattern of AMC are unknown, it is difficult to compare accurately the results of various treatments. We now present the functional and radiological results of our procedure in five children with AMC although our series is too small to make any statistically significant conclusions.

Patients and Methods

From 1954 to 1995, we have examined 48 arthrogryptic children at the Department of Orthopaedic Surgery in Okayama University Medical School. We included in our study only patients who fulfilled the following criteria: 1) joint contracture at birth in at least two different areas of the body; 2) evidence of a non-progressive neurological disorder; 3) diffuse muscle wasting with fusiform joint degeneration; 4) generalised reduction in muscle bulk, shortening of muscles with decreased power and skin dimpling over joint contracture; 5) the presence of webbing; and 6) the absence of normal skin creases. Of these, there were 11 patients with 21 hip dislocations; ten had bilateral and one unilateral dislocation. After 1977 we attempted actively to reduce the dislocated hips in five of these patients. All had associated contractures, in four in the upper and lower limbs and in one only in the lower limb (Table I).

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0301-620X/98/48216 \$2.00

Table I. Details of the five patients with arthrogryposis multiplex congenita

Case	Sex	Age at first visit	Preoperative treatment*	Age at surgery (mth)	Follow-up period (yr)	Additional surgery	Other affected joints	Brace
1	F	10 days	PH	R:36 L:28	R:18.8 L:19.5	None	Dislocations of both knees Bilateral equinovarus	Long leg brace
2	F	4 yr	None	R:57 L:64	R:12.1 L:11.5	R:Femoral shortening osteotomy L:Femoral shortening osteotomy	Contractures of both shoulders Extension contracture of both knees Bilateral vertical talus	Short leg brace
3	M	8 mth	PH,OHT	R:30 L:19	R:12.6 L:13.3	R:Salter's osteotomy Femoral derotational osteotomy L:Femoral varus derotational osteotomy	Flexion contractures of all fingers Bilateral polysyndactyly Dislocations of both knees	None
4	M	14 days	PH	R:17 L:23	R:11.6 L:10.9	None	Extension contracture of both elbows Dislocations of both knees Bilateral planovalgus	None
5	F	2 mth	PH	R:24 L:17	R:3.8 L:4.3	None	Bilateral club hands Contractures of both shoulders Extension contractures of both knees Bilateral equinovarus	Short leg brace

* PH, Pavlik harness; OHT, overhead traction

In four patients, we first tried closed reduction without success. Open reduction was performed on only one child as the primary treatment because of her age (four years) at the first visit. On all five children (ten hips), we then used open reduction by an extensive anterolateral approach. The mean age at operation was 31.5 months (17 to 64) and the mean follow-up 11.8 years (3.8 to 19.5). Two patients required additional surgery. One had a Salter innominate osteotomy and a femoral varus derotational osteotomy on one hip, and a femoral derotational osteotomy on the opposite side. The other had concomitant shortening osteotomies on both hips since the dislocations were high.

Operative technique. A transverse incision is made from the medial border of the sartorius to the greater trochanter 3 cm distal to the anterior superior iliac spine. The fascia lata and the tensor fasciae latae are transversely dissected at the level of the incision. Gluteus medius and minimus are temporarily detached from their insertions. The rectus femoris, with its reflected head, is exposed and the edges of the head clearly defined by blunt dissection. The straight head of rectus femoris is then detached from the anterior inferior iliac spine to obtain a better view. The psoas tendon is also detached from the lesser trochanter and later transferred to the anterolateral surface of the proximal femur. Any fibrous adhesions are dissected for thorough exposure of the joint capsule. Tendons of the short external rotators, such as piriformis and gemellus superior, are transected at their insertions because of shortening. At this point, the ascending branch of the medial femoral circumflex artery is protected. The joint capsule is incised circumferentially near the acetabular rim. Transection of the transverse acetabular ligament alleviates inversion of the labrum. The ligamentum capitis femoris is hypertrophied and is removed along with the fibrofatty tissue in the acetabulum. The redundant parts of the capsule are trimmed away. We

found capsulorrhaphy to be unnecessary. While the hip is held in slight flexion, full internal rotation, and 30° of abduction, a complete reduction is confirmed by radiography. The transected and detached muscles are reattached except for the short external rotators. A hip spica cast is then applied in the above position for eight weeks.

Bracing or physiotherapy was not required. Follow-up examination included assessment of the range of movement and walking and radiography. Motion in the joint was evaluated on the criteria of Gruel et al.⁹ Anteroposterior radiographs of each hip were graded according to the classification of Severin¹⁰ and avascular necrosis by the classification of Kalamchi and MacEwen.¹¹

Results

At the final follow-up all children walked without crutches or canes. Two walked independently. One required a long leg brace and two needed short leg braces because of knee and/or foot problems. The range of movement was good in eight hips and fair in two (Table II).

Table II. Functional and clinical results in the five patients

Case	Sex	Side	Gruel ⁹ classification	Severin ¹⁰ classification	Avascular necrosis (Kalamchi and MacEwen ¹¹)
1	F	R L	Good Good	IIa IIa	III None
2	F	R L	Fair Fair	III IIa	II II
3	M	R L	Good Good	IVb III	II II
4	M	R L	Good Good	Ia Ia	None None
5	F	R L	Good Good	IIb IIb	II II

Fig. 1a

Fig. 1b

Fig. 1c

Fig. 1d

Case 2. Figure 1a – General appearance of a four-year-old girl with bilateral AMC. Figures 1b to 1d – Radiographs before operation (b), at one year two months after operation on the right hip and seven months on the left (c) and showing that the result was Severin group III in the right hip and group IIa in the left at 16.8 years of age (d).

Two hips were in Severin class Ia, three in class IIa, two in class IIb, two in class III, and one was in class IVb. Hence, seven hips had good results (class I or class II) (Fig. 1).

Avascular necrosis was seen in seven hips. Six were in group II and one in group III.

There were no postoperative redislocations or infections.

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.²

It is generally agreed that unilateral dislocation in the arthrogryptic child should be reduced to prevent pelvic obliquity and secondary scoliosis. The management of bilateral dislocations, however, is controversial. The high rate of complications such as redislocation, stiffness, and avascular necrosis has led to the recommendation that bilateral dislocations should be left unreduced.

Staheli et al⁶ reported good results using open reduction by the medial approach principally because minimal operative dissection was required. The anterior approach has been recommended for bilateral dislocations because the femoral head usually rides high on the ilium. Drummond et al² found that stiffness and persistent flexion deformity usually followed open reduction of an arthrogryptic dislocation.

We attempted to remove any obstacles to a successful reduction. We performed a complete circumferential capsulotomy followed by release of joint contractures so that our patients did not require physiotherapy. It is thought that complete capsulotomy may cause iatrogenic avascular necrosis of the femoral head giving an unsatisfactory result. It is important to preserve the essential blood supply to the femoral head, mainly the lateral epiphyseal artery (a branch of the medial femoral circumferential artery). We believe that capsulotomy near the acetabular rim does not affect the blood supply to the femoral head, because the branch comes from behind the greater trochanter and passes through the posterior capsule at the base of the femoral neck. We therefore reflect the gluteus medius and minimus and incise the capsule under direct vision.

The incidence of avascular necrosis in our series was 70% which is considered high but the deformity was small since six of the seven hips were in group II, one was in group III, and there was none in group IV. We believe that two reasons for the high incidence of avascular necrosis are the mean age at surgery, which was 31.5 months (17 to 64), and the fact that undue force had been used in the pre-operative conservative treatment. In cases 1, 4 and 5, treatment with a Pavlik harness, combined with long leg casts because of knee and/or foot problems, had been attempted; the hips were then forced into a position of 90° of flexion and abduction. At the time of operation, a

deformed femoral head was found in all hips, particularly in case 2, in which it was pear-shaped. After open reduction the femoral head gradually remodelled regaining its normal round shape.

Huurman and Jacobsen¹ report that better functional results may be obtained using subtrochanteric extension osteotomies. We agree that femoral shortening can be helpful in the reduction of the high femoral head in AMC or in congenital dislocation of the hip. The high dislocated femoral head is compressed against the iliac wall and the posterior capsule is adherent to the ilium. Itadera et al¹² reported that the contracted posterior capsule and short external rotators pulled the femoral neck posteriorly, resulting in residual subluxation. We believe that a femoral osteotomy is a simple procedure but may be less satisfactory and may weaken the muscle power of the affected leg. Our results show that extensive complete release did not result in a stiff hip. We believe that a less radical release may be the cause of hip stiffness.

Initially, we attempt conservative reduction before walking age, but there is little chance that this will be successful in AMC. After one year, we perform open reduction as early as possible using the extensive anterolateral approach, since in our experience surgical intervention should optimally be performed under the age of three years.⁸

According to Huurman and Jacobsen¹ it may be necessary to use a brace at night for several years to attain maximum remodelling. Furthermore, Gibson and Urs⁵ stated that the prognosis was relatively poor for children with affected hips: 25% are confined to a wheelchair, 25% are dependent on braces for walking, and only 50% can walk independently. Children with AMC often have concomitant knee and/or ankle problems and some require braces or wheelchairs for mobility. Our patients, all of whom had hip and knee and/or ankle problems, were able to walk independently after our procedure without crutches or wheelchairs.

Although our series is small, we believe that our extensive anterolateral approach can successfully treat unilateral or bilateral hip dislocation in arthrogryptic children. We also believe that the method is useful for the treatment of developmental dislocation of the hip.

We wish to thank John E. Herzenberg, MD, FRCS C, Associate Professor of Orthopaedic Surgery and Paediatrics of the University of Maryland School of Medicine, for help in the preparation of this manuscript.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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