Transfer of pectoralis major in arthrogryposis to restore elbow flexion

DETERIORATING RESULTS IN THE LONG TERM

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We present the long-term results of pectoralis major transfer to restore elbow flexion in seven patients (ten procedures). The early results in all the patients were encouraging but with longer follow-up a gradual and progressive flexion deformity was observed with a decrease in the arc of flexion in eight elbows, reaching ≥90˚ in all cases.

The results of pectoralis major transfer deteriorate with time due to the development of a recalcitrant flexion deformity of the elbow. With bilateral involvement we now recommend that the procedure be undertaken on one side only to allow the hand to reach the mouth for feeding, while the opposite side remains in extension for perineal toilet.

The upper limb is commonly involved in arthrogryposis multiplex congenita either in isolation or in combination with the lower limbs. Two patterns of involvement have been described. The most common (type I) presents with adduction and internal rotation at the shoulder, extension at the elbow, pronation of the forearm and a flexion deformity of the wrist, suggesting involvement of the C5/6 neural segments. The less common (type II) shows adduction and internal rotation at the shoulder, a flexion deformity of the elbow and flexion and ulnar deviation of the wrist, suggesting partial involvement of C5, complete involvement of C6 and partial involvement of C7. These deformities can be disabling. Feeding and drinking are particularly difficult, although some of these children develop very effective trick movements. Function of the upper limb is frequently improved by special orthotics. Surgical intervention is only indicated when such measures fail and the basic activities of daily life, feeding, toilet hygiene and picking-up objects require assistance. The deformities at all levels should be considered as a whole and not in isolation when planning surgery.

Inability to flex the elbow in arthrogryposis has a major effect on function and surgical correction is often indicated. Many tendon transfer procedures have been described to restore flexion. In 1985 Atkins, Bell, and Sharrard described a modified pectoralis major transfer with satisfactory results in the medium-term. However, longer follow-up has identified deteriorating function in the majority of patients leading to an unsatisfactory outcome.

Patients and Methods

Clinical examination and a review of the medical records of seven patients (ten procedures) who had undergone a modified pectoralis major transfer were carried out. The surgical procedure and post-operative rehabilitation was similar to that reported previously. Three patients underwent simultaneous tricepsplasty to improve the arc of flexion. Four patients (cases 3, 5, 6 and 7) had a tricepsplasty two to three months prior to pectoralis major transfer.

The age at operation ranged from 2.5 years to 14 years. Three patients had staged bilateral procedures. The follow-up ranged from seven to 19 years (Table I).

Two patients (cases 5 and 6) were subjects in a previous study by the senior author (MJB).

Results

The early results in all the patients were encouraging. All except one (case 7) retained useful power (Medical Research Council grade 3-4) in the transferred pectoralis major muscle and maintained the arc of flexion obtained following tricepsplasty. Both the parents and patients were satisfied with the initial results.

However, with time, a gradual and progressive increase in flexion deformity and decrease in the arc of flexion were observed in eight of the ten elbows. The deformity was 90˚ or more in all cases. One child (case 6) developed a flexion deformity of 100˚ and required an exten-
sion osteotomy of the distal humerus to improve function and appearance. Case 5 underwent a bilateral staged transfer of pectoralis major for extension contractures of both the elbows at the age of 2.5 and 5 years (Fig. 1). He had good function in the right elbow following operation but developed progressive flexion contractures in both elbows and at the age of 15 had the deformities shown in Figure 2.

Case 6 was first seen at the age of 1.5 years. She underwent left-sided tricepsplasty followed by pectoralis major transfer at the age of 2.5 years. Similar procedures were carried out on the right side at the age of three. After operation, the function and appearance were very good but after four years she began to develop a flexion deformity of both elbows. Although she was able to reach her mouth, she could not reach her perineum and needed help with toi-

<table>
<thead>
<tr>
<th>Case</th>
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<th>Age at operation (yrs)</th>
<th>Follow-up (yrs)</th>
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<td>14</td>
<td>10</td>
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<tr>
<td>4</td>
<td>R</td>
<td>M</td>
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<td>7</td>
<td>Fixed flexion deformity</td>
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<td>5</td>
<td>Bilateral</td>
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<tr>
<td>7</td>
<td>L</td>
<td>M</td>
<td>8</td>
<td>9</td>
<td>Transferred tendon is weak</td>
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</table>

Discussion

Inability to flex the elbow, commonly bilateral, is a source of significant disability in arthrogrypotic children, particularly in relation to feeding and personal hygiene. Non-operative treatment should be tried before embarking on surgery to improve function. There are various operations available of which pectoralis major transfer is one. It has the advantage of retaining the function of the triceps muscle, which is essential for the use of crutches and a wheelchair. The power of the triceps muscle was grade 3 or more on the MRC scale in all our patients. Various techniques of transferring the pectoralis major have been described. We continued to use the technique reported from the Sheffield Children’s Hospital,\(^8\) which showed satisfactory medium-term results. However, with longer follow-up it became obvious that the functional results were deteriorating due to a gradual increase in the flexion deformities in the operated elbows and a decrease in the effective arc of flexion. No deterioration in the power of the muscle transfer was noted. The cause of this deformity is unclear but appears to be related to the muscle transfer, as a similar deformity was not observed in the elbows which had not undergone operation. The only patient who retained good
long-term function (case 3) had surgery at the age of 14 years, whereas all other patients except case 7 were younger than five years of age at the time of operation. Function deteriorates dramatically following the development of a flexion deformity in both elbows. With one elbow flexed, patients can still bring the hand to the mouth to feed themselves but with both elbows fixed in flexion perineal care becomes difficult if not impossible (case 6). Surgical correction of the flexion deformity may become necessary in such cases. Williams also reported poor long-term results following triceps transfer due to increasing flexion deformity at the elbow. The cause for this deformity was initially thought to be the triceps transfer but because nearly all the mobile or flexed elbows, whether subjected to surgery or not, suffered a similar fate, it was concluded that the transfer was not the cause, which remained unknown.

In our experience the early encouraging results of transfer of pectoralis major to treat an extension contracture of the elbow in arthrogryposis deteriorate with time due to the development of a recalcitrant flexion deformity of the elbow. We now recommend this procedure on one side only in cases of bilateral involvement. If the operation is carried out on one side only it is possible to get the hand to the mouth for feeding while the unoperated side in extension can be used for perineal toilet. Parents should also be advised that there is a risk of introducing a new deformity by muscle transfer in the growing child and that the object of transfer is to balance the muscle forces around the paralysed joint.

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.

References

Fig. 2
Flexion deformity in both elbows in case 5 at 15 years of age.

Fig. 3
Flexion deformities in both elbows in case 6 prior to right supracondylar extension osteotomy to restore extension.