The supination deformity and associated deformities of the upper limb in severe birth lesions of the brachial plexus

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We reviewed 42 consecutive children with a supination deformity of the forearm complicating severe birth lesions of the brachial plexus.

The overall incidence over the study period was 6.9% (48 of 696). It was absent in those in Narakas group I (27.6%) and occurred in 5.7% of group II (13 of 229), 9.6% of group III (11 of 114) and 23.4% of group IV (18 of 77).

Concurrent deformities at the shoulder, elbow, wrist and hand were always present because of muscular imbalance from poor recovery of C5 and C7, inconsistent recovery of C8 and T1 and good recovery of C6. Early surgical correction improved the function of the upper limb and hand, but there was a tendency to recurrence. Pronation osteotomy placed the hand in a functional position, and increased the arc of rotation of the forearm. The supination deformity recurred in 40% (17 of 42) of those treated by pronation osteotomy alone, probably because of remodelling of the growing bone.

Children should be followed up until skeletal maturity, and the parents counselled on the likelihood of multiple operations.

A supination deformity of the forearm (Fig. 1) in severe birth lesions of the brachial plexus arises from a combination of muscle imbalance, soft-tissue contracture and secondary skeletal deformity involving the shoulder, elbow, wrist and hand. It is part of a variety of deformities of the upper limb which produce a ‘begging’ posture of fixed flexion of the elbow, supination of the forearm, extension of the wrist and ulnar deviation. Many of these children also develop posterior subluxation of the shoulder and a thumb-in-palm deformity.

Each of these deformities must be addressed in order to restore function to the upper limb. We have attempted to identify those at risk, to explain the pattern of deformity by the neurological lesion and to evaluate the results of our strategy for improving the function of the upper limb and hand.

Patients and Methods

From a prospective database of birth lesions of the brachial plexus, we identified all the children seen and followed up at our centre between 1997 and 2004 who developed a supination deformity of the forearm. Of 696 children (6.9%) developed this deformity of whom 42 (6.0%) had adequate records and follow-up. Information concerning their nerve injury, treatment and the function of the upper limb was obtained from the database. Their complete medical records were reviewed for additional details. The distribution of the patients by gender, side of lesion and severity is shown in Table I.

We graded the severity of the lesions according to the classification of Narakas (group 1: C5-6 injury, group II: C5-7 injury, group III: C5-T1 injury without Horner’s syndrome, group IV: C5-T1 injury with Horner’s syndrome). During the study period there were 276 patients in group I, 229 in group II, 114 in group III and 77 in group IV. The severity of injury for each spinal nerve was assessed using a combination of methods including neurophysiological investigation which predicts the severity of the injury as shown in Table II, the intra-operative findings for each spinal nerve and the degree of neurological recovery (Table III). Function of the shoulder was assessed by a modified Mallet score which ranged from 5 (poor) to 15 (excellent). Elbow function was assessed by the range of active flexion and extension. Function of the hand was determined using a modification of Raimondi’s classification shown in Table IV.

Results

The nerve lesion. Neurophysiological investigation was carried out on 26 infants in whom
there had been poor recovery of shoulder or elbow function by three to six months of age. The plexus was explored in 27 infants and reconstructed by intraplexal grafting or extraplexal transfer in 19.

Table I. Distribution of patients with supination deformity by gender, affected limb and Narakas group

<table>
<thead>
<tr>
<th></th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>20</td>
</tr>
<tr>
<td>Female</td>
<td>22</td>
</tr>
<tr>
<td>Affected side</td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>20</td>
</tr>
<tr>
<td>Left</td>
<td>22</td>
</tr>
<tr>
<td>Narakas group</td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>0</td>
</tr>
<tr>
<td>II</td>
<td>13</td>
</tr>
<tr>
<td>III</td>
<td>11</td>
</tr>
<tr>
<td>IV</td>
<td>18</td>
</tr>
</tbody>
</table>

Fig. 1

Photograph showing an 11-year-old girl with Narakas group IV birth lesions of the brachial plexus and a supination deformity of the left forearm attempting to pronate both forearms simultaneously with the elbows flexed at 90°. There is concurrent ulnar deviation of the wrist, adduction deformity of the thumb and lack of intrinsic balance. The scar on the forearm is from the earlier harvesting of the superficial branch of the radial nerve for reconstruction of the brachial plexus.

Table II. Neurophysiological investigation for spinal nerves in birth lesions of the brachial plexus

<table>
<thead>
<tr>
<th>Type</th>
<th>Nerve action potential</th>
<th>Electromyography</th>
<th>Lesion predicted</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Normal</td>
<td>No spontaneous activity, reduced number of normal motor units, increasing firing rates</td>
<td>Conduction block</td>
</tr>
<tr>
<td>B1</td>
<td>&gt; 50% amplitude of uninjured side</td>
<td>Relatively good motor-unit recruitment, mixture of normal and polyphasic units suggesting collateral re-innervation</td>
<td>Axonotmesis</td>
</tr>
<tr>
<td>B2</td>
<td>&lt; 50% amplitude of uninjured side</td>
<td>Few or absent normal motor units, collateral re-innervation</td>
<td>Neurotmesis</td>
</tr>
<tr>
<td>C</td>
<td>Absent (present in pre-ganglionic lesion)</td>
<td>Spontaneous activity, nascent units, poor recruitment</td>
<td>Complete rupture or avulsion</td>
</tr>
</tbody>
</table>

Table III. Grading of nerve recovery by spinal nerve modified from Birch et al

<table>
<thead>
<tr>
<th>Result</th>
<th>C5</th>
<th>C6</th>
<th>C7</th>
<th>C8 and T1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Good</td>
<td>Mallett* 13 or better</td>
<td>Flexion to reach mouth Biceps MRC† 4+</td>
<td>Full wrist extension</td>
<td>Digital flexion and extension MRC 4+, intrinsic balance, hands used appropriately in bimanual activity. Thumb abduction</td>
</tr>
<tr>
<td>Fair</td>
<td>Mallett 11 to 12</td>
<td>Flexion &gt; 90° MRC 3+</td>
<td>Wrist extension MRC 3</td>
<td>Digital flexion providing simple/weak grasp. Hand used to support or help the other side. Thumb adducted</td>
</tr>
<tr>
<td>Poor</td>
<td>Less than above</td>
<td>Less than above</td>
<td>Less than above</td>
<td>Less than above, hand with minimal function</td>
</tr>
</tbody>
</table>

* Modified Mallett score
† MRC, Medical Research Council grading of motor power

C5. Neurophysiological investigation predicted severe injury (grade B2 or grade C) in ten of the 26 nerves assessed. Of 27 nerves explored, 13 had been ruptured and two avulsed. Repair was performed on 13 nerves, of which
seven had a poor, four a fair and two a good result. Overall, the outcome for C5 was poor, with good elevation and external rotation of the shoulder being obtained in only four of the 42 children (9.5%). There was a fair recovery in 15 and a poor recovery in 27 children. The outcome was worst in group IV, in which 14 of 18 children (77.8%) had poor shoulder function.

C6. Neurophysiological investigation predicted severe injury in 11 of 26 nerves assessed. Of 27 nerves explored 14 had a rupture or avulsion, all of which were repaired. Of the 42 children, 39 (92.9%) regained fair or good elbow flexion, and all had active supination.

C7. Neurophysiological investigation predicted severe injury in 19 of 26 nerves, including all 11 in group IV. Ruptures and avulsions were found in 20 of 27 nerves and ten had been repaired. Irrespective of whether repair had been undertaken, none had a good outcome.

Of the 42 children, 14 had a fair outcome providing wrist or finger extension against gravity and in 28 the result was poor. All the children had weak or absent pronation of the forearm.

C8 and T1. Neurophysiological investigation was performed on 32 nerves (16 children) in groups III and IV, predicting severe injury in each case. In 27 children 54 nerves were explored. Ruptures and avulsions were found in 24 (12 children) of which only 11 were repaired. Good or fair recovery of wrist and finger flexion was seen in 26 of 29 children, but 14 lacked abduction of the thumb, which is predominantly a function of C8. Intrinsic balance was poor in two of the 11 children in group III and in 13 of the 18 in group IV.

The supination deformity. The supination deformity occurred with increasing frequency in each of the children in Narakas groups II, III, and IV. A total of 13 of the 229 patients in group II (5.7%) and 11 of the 114 patients in group III (9.6%) seen during the study period developed this deformity. Patients with complete lesions were at the greatest risk, with 23.4% (18 of 77) of those in group IV developing the deformity. These deformities did not occur in group I.

Supination deformity of the forearm was first recorded at a median age of 5 years (1 to 13) and was treated initially by splinting and passive stretching exercises. When passive pronation was seen to be deteriorating, a pronation osteotomy with plate-and-screw fixation was carried out through a lateral approach at the junction of the middle and distal thirds of the radius, distal to the insertion of pronator teres. This was undertaken at a median age of 7 years (3 to 14), after which the patients were followed up for a median of 5 years (1 to 15). A mean correction of 88.5° (0° to 120°) was achieved. When extension of the wrist and fingers was also weak, these were corrected either earlier or concurrently with the pronation osteotomy. In three patients in whom dislocation of the head of the radius was present,
this was left alone. There were three complications; in two children the plate broke before the osteotomy had united and in one over-pronation developed after late recovery of active pronation.

The deformity recurred in 17 children (40.5%) after a median of 3 years (0.5 to 8). A repeat pronation osteotomy was performed in 14 children, while three declined further surgery.

Associated deformities of the upper limb

Shoulder. Posterior subluxation or dislocation of the shoulder with fixed medial rotation was present in 32 children. All had open relocation and lengthening of subscapularis as described by Khambampati et al10 at a median age of three years (4 months to 7 years). Of these, 20 later developed inadequate medial rotation because of weakness of subscapularis. They required a medial rotation osteotomy of the humerus (18) or transfer of pectoralis major (2) to allow the hand to reach the body.

Elbow. A fixed flexion deformity of the elbow of a median of 32.5° (10° to 90°) was present in 32 children. A fixed flexion deformity greater than 40° was seen in five children who were treated by serial plaster-of-Paris splinting, while the rest required only passive stretching exercises.

Wrist and hand. There was inadequate active extension of the wrist or fingers in 30 children. Of these, 19 also had weak abduction of the thumb resulting in a thumb-in-palm deformity, and 18 had ulnar deviation of the wrist. A flail hand was present in five (one in group III and four in group IV), and two in group IV had an insensate hand which they neglected. The other 23 had at least one strong wrist flexor and strong finger flexors. Active extension of the wrist and fingers and abduction of the thumb in these 23 children were restored by flexor-to-extensor tendon transfers. Ulnar deviation was due to imbalance between weak radial deviators (extensor carpi radialis longus and abductor pollicis longus) and strong ulnar deviators (extensor carpi ulnaris and flexor carpi ulnaris). This was corrected by transferring extensor carpi ulnaris to extensor carpi radialis longus or abductor pollicis longus in 15 children, and by transferring flexor carpi ulnaris for extension of the wrist or fingers in three. These tendon transfers were undertaken at a median age of 4.75 years (2.5 to 15). Further tendon transfers for recurrence of wrist or finger drop were required in 13 of 30 children.

Functional outcome. The final function of the upper limb was assessed at a median age of 12 years (8 to 26). The mean final modified Mallet score was 10.5 (6 to 15). The median final elbow flexion was 120° (90° to 125°) with a median extension deficit of 20° (0° to 70°). Elbow flexion was MRC grade 412 and above in 35 of 42 children. There

![Fig. 2](image-url)
was no clinically appreciable change in elbow function between the initial and final assessments. The final position of the forearm and the arc of rotation were greatly improved after pronation osteotomy (Fig. 2). The function of the hand improved between 1 and 3 grades after correction of the supination deformity and tendon transfers (Fig. 3), except in two children with recurrent supination deformity who declined further surgery, and in seven with flail or insensate hands.

Discussion

Secondary deformities of the upper limb are common in birth lesions of the brachial plexus and tend to increase with the severity of the lesion. 1,2,4,5 Khambampati et al 10 showed that 25% of patients developed subluxation or dislocation of the shoulder, particularly those in Narakas groups II and III. Sibinski et al 13 observed that the range of pronation and supination decreased with increasing severity of the lesion by Narakas group. Our study has shown that a supination deformity occurred in 6.9% of all 696 children with birth lesions, and was common in those with group-III (9.6%) and group-IV (23.4%) severity.

When present, it is always associated with one or more deformities of the upper limb. These include fixed medial rotation with posterior subluxation or dislocation at the shoulder, restriction of lateral rotation after relocation of the shoulder, fixed flexion deformity of the elbow, wrist and finger drop, ulnar deviation of the wrist and a thumb-in-palm deformity. They are caused by muscle imbalance at all joints in the upper limb resulting from the degree of neurological recovery which was fair or poor with C5 and C7 in all groups, inconsistent in C8 and T1 in groups III and IV; and good with C6 in all groups. We cannot explain why this pattern occurred, and further study into this is necessary. One possible explanation is that C5 to C7 are the most severely injured, but neurones preferentially regenerate towards biceps at the expense of the muscles of the shoulder and wrist.

The supination deformity arises from an imbalance between the strong supinators, and the weak or paralysed pronators. It becomes apparent only after three to four years when neurological recovery is mature and after the shoulder has been reduced. Before this, the supinators are still weak and the fixed medial rotation at the shoulder gives the forearm a pronated appearance. Improving the lateral rotation of the shoulder increases the active range of supination 10 and the strength of biceps as a supinator, 14 allowing the deformity to develop. Initially, it is passively correctable, but becomes progressively fixed because of soft-tissue contractures at the interosseous membrane. 1 In our series dislocation of the head of the radius from the unbalanced pull of the tendon of biceps was uncommon and occurred only in three patients.

The current literature 2,4,5 recommends re-routing of biceps with release of soft-tissue contractures 15 for deformities which are passively correctable, and pronation osteotomy of the radius 3,16,17 for fixed deformities. We have little experience with the biceps procedure. All the children in our series were treated by a pronation osteotomy, regardless of the passive range of movement. This not only put the hand into a more functional position, but surprisingly also increased the passive and active arcs of rotation. We cannot explain this finding, but consider that it may be due to a combination of release of the interosseous membrane while preparing the bone, and improvement in the length-tension relationship of the muscle fibres of both the supinators and the pronators.

Allende and Gilbert 5 reported a recurrence rate of 20% (9 of 44) following pronation osteotomy, which they attributed to the presence of residual passive rotation of the forearm at the time of osteotomy. In our series of 42 children,
17 (40.5%) had recurrence at a median of three years. Since the recurrence was gradual in all but three of the 17, we believe that in most cases it was due to persisting supinating forces principally brachioradialis, which remodelled the growing bone. Although a weak supinator in its anatomical position, after pronation osteotomy its supination vector is increased and it is put on stretch. It is attached to the distal position, after pronation osteotomy its supination vector is growing bone. Although a weak supinator in its anatomical belief that in most cases it was due to persisting supinating the recurrence was gradual in all but three of the 17, we

Deformities of the shoulder in these children arise from a combination of poor recovery of C5 and C7. Khambampati et al. give a detailed description of deformities of the shoulder arising from deficiency of C5 along with the technique and results of early correction. In patients who also have poor recovery of C7, although subscapularis is shortened and requires lengthening, it is weak and overpowered by the recovery of the C5-innervated external rotators. In our series, the loss of medial rotation after relocation of the shoulder was not due to retroversion of the head of the humerus as described by Khambampati et al. but because of a weak subscapularis. Flexion contractures of the elbow are due to early and strong recovery of biceps (C5, C6) and brachioradialis (C6) with weak or delayed recovery of triceps (C7 and C8). These may be corrected by serial casting. Wrist and finger drop and ulnar deviation occur because of weak extensors and radial deviators. The thumb-in-palm deformity results from weakness of the C8-innervated abductors of the thumb in the presence of strong flexors (C8 and T1). When possible, tendon transfers to correct these deformities should be done concurrently with correction of the supination deformity to minimise the number of surgical operations. Recurrent deformity after tendon transfers was common in our series, probably because re-innervated muscles were transferred and retracting of the transferred muscle in this age group was difficult.

A supination deformity occurs in patients with birth lesions of the brachial plexus of Narakas groups II to IV, particularly group IV, and is associated with deformities at all levels of the upper limb. Recovery of C5, C7 and to a lesser extent C8 is poor in these patients, and co-existing deformities at the shoulder, elbow, forearm, wrist and hand must be anticipated and addressed. Early correction of deformities and muscle imbalances allows early integration of the upper limb and hand into daily use and promotes early cortical development of useful patterns of function. Function of the hand increases considerably after the forearm has been placed in a functional position and the necessary tendon transfers performed to restore wrist and finger extension. The patient, parents and surgeon must be prepared for recurrent deformity as the child grows. Repeated surgery is often necessary, and patients should be followed up until skeletal maturity.

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References