Introduction
The incidence of brachial plexus birth palsies (BPBP) has been reported to be between 0.5 to 2 per 1000 live births,\textsuperscript{1,2} injury to the upper trunks (C5, C6) being the most common.

Causation
The mechanism of injury is usually traction. Affected infants are usually one half to one kilogram heavier than unaffected infants;\textsuperscript{2} thus factors leading to a larger baby or to cephalo-pelvic disproportion will increase the risk of brachial plexus birth palsy. It is commonly seen with maternal diabetes or in mothers with multiple previous pregnancies as birth weight tends to increase with successive pregnancies. Other common associations are shoulder dystocia and vacuum-assisted or forceps delivery, where there is lateral traction of the head away from the shoulder. However caesarean section is not protective for BPBP.\textsuperscript{3,4}

Initial assessment
Lack of movement of the affected arm usually leads to referral for orthopaedic opinion. Assessment aims to confirm the diagnosis, establish a baseline functional assessment, and exclude other diagnoses. This includes a full pregnancy and birth history looking for any risk factors including difficult labour and delivery involvement.

Differential diagnosis
Pseudo-paralysis of the arm may be caused by:

1. Fractures of the clavicle.
2. Fracture of the proximal humerus.
3. Infection, in or around the glenohumeral joint.
4. Rarely intra-uterine radial nerve compression.
The latter is caused by the position of the infant in-utero, and presents as a classical radial nerve neuropathy. It is treated expectantly, with full resolution expected.\(^5\)

**Clinical assessment**

Clinical examination usually reveals one of several distinct patterns of injury. Additionally, respiratory problems that may be indicative of C4 damage should be sought.

- Classical upper plexus (Erb-Duchenne) palsy. The primary deficit is in C5/6 \(+/-\) C7 affecting mainly the shoulder and elbow. The arm adopts the “waiter’s tip” position, with the shoulder adducted and internally rotated, the elbow extended, the forearm pronated, and the wrist and fingers flexed.
- Klumpkes’ palsy. This is rarely seen in birth palsy\(^6\) and involves the lower plexus (C8, T1).
- Total plexus. This is the most severe, and presents with complete atonia of the arm, with the infant often ignoring the arm.

Prognostically it is important to know whether the injury is pre or post-ganglionic.\(^6,7\) Pre-ganglionic lesions are avulsions from the cord that will not recover motor function spontaneously. Assessment of the function of nerves arising close to the ganglion can give an idea of the level and severity of the injury (Fig. 1).

The phrenic nerve is assessed by observing the pattern of breathing to see if it is symmetrical. Winging of the scapula suggests long thoracic nerve involvement, absence of the rhomboids, the dorsal scapular nerve and weak rotator cuff, damage to the suprascapular nerve. All of these suggest a more severe pre-ganglionic injury.

Further evidence of pre-ganglionic injury is the presence of a Horner’s syndrome. The pre-ganglionic sympathetic fibres that supply the eye arise from T1 and leave the spinal nerve, just as it exits the inter-vertebral foramen to join the sympathetic chain. Injury at this level gives the classic ptosis, meiosis, anhydrosis and enophthalmos of the ipsilateral eye.

**Treatment of neo-natal brachial plexus injuries**

Identifying those who would benefit from surgical intervention can be extremely difficult. At one end of the spectrum, for infants showing neurological recovery during the first month prognosis is universally good,\(^8\) and surgical intervention is unnecessary. At the opposite end of the spectrum, are infants with complete involvement of the plexus and a Horner’s syndrome. Their prognosis is universally poor without surgical exploration and reconstruction.\(^9\) However, for infants in the middle, because of knowledge as to the natural history without intervention, opinions are divided.

Gilbert and Tassin’s\(^10\) classic paper showed that shoulder function was better in infants who showed recovery of biceps and deltoid before 3 months of age. Peter Waters\(^8\) confirmed this, but also looked at infants who showed recovery in months 4, 5 and 6. Those who had surgery at 6

---

**Figure 1** Brachial Plexus (Reproduced with permission).
months, for failure of return of biceps function, did better than those who recovered biceps naturally at 5 months. This shows that surgery can improve the natural history but there is a still a question over the timing of surgery.

Howard Clarke's team in Toronto questioned the use of biceps function as a predictor of outcome. They showed that lack of elbow flexion at 3 months incorrectly predicted a poor outcome in 12%. When they looked at a composite score using elbow flexion and extension with wrist, finger and thumb extension, this gave a false positive rate of 5%. Those that fail this composite test score at 3 months are surgically explored. Those that pass are reviewed until 9 months when they undergo the "cookie" test. This is passed if the infant can place a cookie in their mouth without undue flexion of the neck. If the infant fails this test then surgical exploration is offered. For further information on this and the scoring system used, see Curtis et al.\(^{11}\)

**Technique**

Currently the standard surgical treatment for the nerve lesions is transection of the neuroma and grafting of the resultant gap, usually with sural nerve grafts, or, if a greater length of nerve is required, thoracic intercostal nerves.

Additionally, the spinal accessory nerve, after it has given its branch to the cervical and upper thoracic parts of trapezius, can be grafted onto the suprascapular nerve to restore shoulder external rotation.

**Shoulder deformity**

When the upper trunk (C5/C6) is involved there is weakness of the shoulder external rotators and abductors (teres minor, infraspinatus and deltoid).

However, as they receive some innervation from the lower plexus the internal rotators (pectoralis major and subscapularis) usually retain their power and eventually dominate. As any muscle imbalance across a growing joint leads to joint deformity, untreated the internal rotation and adduction forces eventually progress to posterior subluxation and eventual dislocation of the shoulder joint. The shoulder pathology can be described anatomically or functionally.

The classic Mallet score (with or without the recent addition in some centres of hand to abdomen) (Fig. 2) gives a good functional description.

Assessment of the range of movement of the shoulder, demonstrating the main restriction of abduction and external rotation, is an anatomical description.

**Imaging**

The shoulder can be assessed by plain x-ray (with or without arthrography), CT, MRI or ultrasound. The aim is to assess the congruity of the joint and, if there is deformity, which side of the joint it lies.

Waters et al.\(^{12}\) used transverse CT slices of the shoulder to demonstrate the amount of subluxation or dislocation and then grade the deformity.

Waters classifies the deformity with reference to the normal side.\(^{13}\) (Fig. 3) assessing retroversion of the glenoid as the angle between a line drawn along the face of the glenoid and the long axis of the scapula. The retroversion angle is 90-\(\alpha\).

While in adults the glenoid is normally 7 degrees of retroverted, so that the glenoid forms an angle of 83 degrees with the sagittal plane,\(^{14}\) and Graichen\(^ {15}\) has shown that greater than 17 degrees of retroversion can lead to instability, there are few comparable studies in children or infants.

The long axis is also used to assess the percentage of humeral head translation, the amount of humeral head anterior to this line expressed as percentage of the whole diameter of the head.\(^{13}\) The normal should be about 50 percent.

Ezaki et al.\(^ {16}\) have applied the same principle but using posterior ultrasound. (Fig. 4) Due to difficulty in visualising the anterior glenoid rim, the longitudinal reference line is the posterior margin of the scapula. The advantages of this method of assessment are that no sedation is required, and use of ionising radiation is avoided. Pearl et al.\(^ {17}\) have described the arthrography to assess the deformity. This gives essentially the same information as CT but is invasive and is thus rarely used.

**Treatment options**

The aim of treatment is the restoration of abduction and external rotation. How this is achieved depends upon the degree of deformity of the glenohumeral joint and what functioning muscles are available around the shoulder for transfer.

The first step in management is non-operative. As the shoulder develops an internal rotation contracture, it may respond to physiotherapy and splinting, or even Botox injection.\(^ {19}\)

With progression of the contracture, the next step depends upon whether the contracture is developing due to absence of functioning external rotators, or if present, they are being overpowered by the relatively stronger internal rotators.

If the external rotators are absent, an attempt to reinnervate them with an accessory to suprascapular nerve transfer should be considered. Ideally this should be performed early, but good results have been obtained after the age of 10 months.\(^ {20}\) If they are overpowered by the internal rotators, then a muscle transfer to assist external rotation can be considered.

To aid decision making in the younger child, a trial of Botulinum toxin into the prime internal rotators (subscapularis and pectoralis major) to unmask any power of external rotation, may be useful. In the older child with a more established contracture this may not add a great deal to management.

**Soft tissue procedures**

In the milder, Waters I and II, deformities, soft tissue procedures to release tight structures and restore movement should be sufficient as the joint has not yet developed an established deformity.
Sever\textsuperscript{21} in 1918 described dividing subscapularis and pectoralis major to address the internal rotation contracture. L’Episcopo\textsuperscript{22} in 1934 attempted to restore external rotation by transferring latissimus dorsi and teres major to the posterolateral aspect of the humerus. Hoffer et al\textsuperscript{23} in 1978 modified this by moving the transfer to the rotator cuff to help stabilise the humeral head and improve abduction as well as rotation.

More recently Kozin et al\textsuperscript{24} used arthroscopy to visualise the joint and then to release the tight inferior and middle glenohumeral ligaments, and remove any structures blocking reduction.

The results of the soft tissue procedures are generally good and significant gains in abduction and external rotation can be achieved.\textsuperscript{25} However the early gains in abduction tend to tail off after about 6 years.\textsuperscript{26}

Waters, in case control studies, felt that soft tissue procedures are a reliably good option before joint deformity has developed, with average Mallet score increases of 9.5 to 15.6\textsuperscript{27} and 13 to 18.\textsuperscript{28}

In the presence of established joint deformity, soft tissues procedures alone are unlikely to be sufficient, though some authors have shown glenoid remodelling after tendon transfers without additional bony surgery. Waters showed a mean improvement in retroversion from 22 degrees (−3 to 45) to 16.5 (−8 to 40), and an improvement in humeral head subluxation from 30% to 37%, forty nine months after surgery (28). El-Gammal\textsuperscript{29} has shown remodelling, but suggests that the potential to remodel reduces after the age of 4 years.

In short, there is little consensus in the literature as to how much, if at all, the glenoid will remodel, and by what age procedures need to be done to maximise any remodelling potential.

The senior author’s preferred method of treatment for milder deformity involves initial clinical assessment of the tightness of the shoulder internal rotators to decide whether one or both of either pectoralis major and subscapularis are involved. This involves testing external rotation in adduction and then in 90 degrees of abduction, whilst stabilising the scapula. In abduction subscapularis should relax, so the range of external rotation will be greater if subscapularis alone is involved. If both muscles are affected then there will be no difference in the range of rotation between the two positions. If latissimus dorsi is functioning, and deltoid has at least MRC grade 3 power or above, then a subscapularis release off the anterior aspect of the scapula is undertaken.

This is followed by transfer of latissimus dorsi and teres major to the rotator cuff to help stabilise the humeral head during abduction, and thus assist the deltoid. Care must be taken not to insert it too high on the humerus to avoid an abduction contracture.
If necessary the pectoralis major tendon can be Z lengthened. A shoulder spica is then worn for 6 weeks in maximum shoulder external rotation and about 20 degrees of abduction to protect the transfer.

### Bony procedures

For the more severe deformities (Waters grade 3/4/5) and shoulders deemed unreconstructable, a humeral derotation osteotomy gives predictably good long term results by putting the severely internally rotated humerus into a more functional position. As it does not reconstruct the joint, with time further posterior subluxation and dislocation of the humeral head is possible, giving an unsightly lump in the back of the shoulder. Typical improvement in Mallet scores are from 9.5 to 15.1.\(^\text{18}\)

Nath et al have described a deformity of the scapula they feel is caused by muscular imbalance across the shoulder.\(^\text{19}\) They feel that the SHEAR (scapular hypoplasia, elevation and rotation) contributes to the classic internal rotation position, with lack of external rotation being due to impingement of the humeral head against the acromioclavicular triangle. The elevation of the scapula deforms the distal clavicle anteriorly and tilts the acromio-clavicular joint more anteriorly.

They have addressed this with the ‘triangular’ procedure, which aims to reposition the acromioclavicular triangle in a more neutral position by osteotomising the distal third of the clavicle and neck of the acromion. Their early clinical results at one year are encouraging, with an increase in Mallet scores of 13.6 to 18.6 and an average increase in active external rotation of 53 degrees.\(^\text{20}\)

Posterior opening wedge glenoid osteotomies have been used for the severe type 4 and 5 shoulders to correct the severe glenoid retroversion, but there is little published data on this.

<table>
<thead>
<tr>
<th>Classification</th>
<th>Radiographic Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>&lt; 5 degree difference in retroversion</td>
</tr>
<tr>
<td>Type II</td>
<td>&gt; 5 degree difference in retroversion</td>
</tr>
<tr>
<td>Type III</td>
<td>Posterior humeral head subluxation, &lt;35% anterior to scapular spine axis</td>
</tr>
<tr>
<td>Type IV</td>
<td>Presence of false glenoid</td>
</tr>
<tr>
<td>Type V</td>
<td>Flattening of humeral head, progressive/complete humeral head dislocation</td>
</tr>
<tr>
<td>Type VI</td>
<td>Infantile posterior dislocation</td>
</tr>
<tr>
<td>Type VII</td>
<td>Proximal humeral growth arrest</td>
</tr>
</tbody>
</table>

**Figure 3** Waters classification of shoulder deformity (Reproduced with permission).

If necessary the pectoralis major tendon can be Z lengthened. A shoulder spica is then worn for 6 weeks in maximum shoulder external rotation and about 20 degrees of abduction to protect the transfer.

Figure 4 Ezaki method of ultrasound assessment of shoulder deformity (Reproduced with permission).
For flail shoulders or those that become painful in the future, arthrodesis may be used to help improve upper limb function. These procedures are still in their infancy, and to date there is little published data on their effectiveness, with indications and outcomes still unclear.

Complications

There are some specific to this type of surgery. The conjoint tendon of latissimus dorsi and teres major is framed by the axillary nerve superiorly and radial nerve inferiorly and they are liable to damage when releasing the tendon off the humerus.

There may still be limitation of external rotation post-operatively due to joint deformity and glenohumeral incongruency. This is the one plane of movement that scapulothoracic movement cannot compensate for, and as such the "trumpet" posture may remain. This is where the patient, due to lack of external rotation, is unable to keep the elbow adducted by their side when trying to put their hand behind the head or to the mouth.

The release of subscapularis may be overly aggressive, leaving the patient unable to get the hand back fully to the midline, or worse, be stuck way out in external rotation. This may be seen in Z lengthening or complete release of subscapularis off the humerus.

If the tendon is transferred too high on the humerus, this can lead to an abduction deformity which can be difficult to treat even with aggressive physiotherapy. A similar deformity may also be seen when the patient first comes out of the post-operative spica, but this usually resolves with appropriate physiotherapy.

References