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Brachial Plexus Birth Palsy
An Overview of Early Treatment Considerations


Abstract
Since the description by Smellie in 1764, in a French midwifery text, that first suggested an obstetric origin for upper limb birth palsy, great strides have been made in both diagnosis and early and late treatment. This report presents an overview of selected aspects of this complex and extensive subject. Early treatment options are reviewed in the context of the present controversies regarding the natural history and the indications for and timing of microsurgical intervention in infants with brachial plexus birth injuries.

After the initial description of a brachial plexus birth palsy by Smellie in 1764 (Fig. 1),1 over a century passed before Duchenne confirmed that this complex insult likely resulted from birth trauma and was not “congenital” in origin. He described four cases of C5-C6 injury with the resultant shoulder paralysis. During the same period, both Erb and Klumpke further elucidated the anatomic details of different clinical presentations. During the early 20th Century, results following neurosurgical intervention were dismal. This lack of interest in early nerve repair remained unchanged for over half a century until 1984 when Gilbert demonstrated that using microsurgical technique coupled with intraoperative neurophysiologic monitoring could, in fact, dramatically change the outcome in properly selected infants.2 The role and timing of microsurgical repair and reconstruction remains the most debated issues among contemporary surgeons specializing in the care of these children.

Anatomy
The brachial plexus is composed of the ventral motor nerve roots from C5 to T1 (Fig. 2). Twenty-two percent of anatomic specimens receive contributions from C4 (prefixed) and 1% contributions from T2 (postfixed). The roots then combine to form trunks, divisions, cords, and branches. The C5 and C6 nerve roots form the upper trunk, C7 alone forms the middle trunk, and the C8 and T1 nerve roots form the lower trunk. Each trunk then has an anterior and posterior division. The posterior divisions of all three trunks combine to form the posterior cord. Each cord is named by its anatomic relationship to the axillary artery. The terminal branches of the posterior cord are the radial and axillary nerves. The anterior divisions of the upper and middle trunks form the lateral cord, whose terminal cord, whose terminal branches are the median and ulnar nerves.

It is important to note that the brachial plexus supplies every muscle of the upper extremity except the trapezius. Furthermore, the upper trunk provides primarily shoulder function, whereas the lower trunk provides primarily hand function.
The incidence of obstetric brachial plexus palsies (OBPP) is between 0.1% and 0.4% of live births. This statistic has remained relatively constant over time. The majority of these (40% to 50%) are C5-C6 lesions. C5-C6-C7 lesions comprise about 20% to 25% of OBPP. Panplexus or global lesions account for 25% to 50% of OBPP. Up to 10% of OBPP are bilateral, seen almost exclusively in breech presentations. Although rare in children born by C-section, the incidence of brachial plexus birth trauma in this group has been reported in the literature to be approximately 1%.

The existence of a true “Klumpke’s” isolated distal root palsy remains debated. Reports by experienced groups suggest that an isolated C8-T1 lesion is an exceedingly rare presentation. Lower root predominance may represent a complete plexus palsy with resolution of the upper plexus component. The experience of the senior investigator (JAIG) is the same.

Several risk factors for OBPP have been identified. These include: 1. large birth weight, 2. breech presentation, 3. shoulder dystocia, 4. prolonged second stage of labor, 5. vacuum or forceps delivery, 6. prior delivery of a child with OBPP, 7. multiparity, and 8. maternal diabetes. There has never been a proven association between race and gender and the risk of brachial plexus injury.

Clinical Examination
A newborn with an upper trunk lesion (Erb’s palsy) will hold the affected upper extremity internally rotated at the shoulder, pronated at the forearm, and flexed at the wrist. This classic posture is often referred to as a “waiter’s tip

Figure 1 Copy of Smellie’s initial description of a brachial plexus birth palsy, 1764.

is supplemented with neurophysiology and histopathology preoperative planning; intraoperatively, this information of CT myelogram is 93%.\textsuperscript{11} The MRI finding of a pseudo-tecting nerve root avulsions. The negative predictive value specificity, and a positive predictive value of 50% for de-

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replaced more invasive CT (computed tomography) my-
3) in the evaluation of a child with OBPP is useful and has
The utility of magnetic resonance imaging (MRI) (Fig.
Figure 3 Preoperative MRI axial image demonstrating neuroma of the left brachial plexus. Note the associated glenohumeral pathology with glenoid dysplasia and posterior subluxation of the humeral head.
hand.” The involved upper extremity will have decreased spontaneous movement and the Moro reflex will be absent. If the lower trunk is involved, the child may have an absent grasp reflex, the hand may be held in an intrinsic minus position, or the involved extremity may have a flaccid paralysis. Respiratory distress may also be seen when there is an associated injury to the phrenic nerve, although this is rare. A chest radiograph or ultrasound will confirm this clinical scenario. The child may also present with a Horner’s syndrome if there has been injury to the sympathetic chain. Anatomically the sympathetic chain lies in close proximity to the lowest roots of the brachial plexus, making it susceptible to injury. The clinical triad of Horner’s syndrome consists of miosis, ptosis, and anhydrosis. The upper eyelid ptosis is most readily detected.

The differential diagnosis of OBPP can include infections, such as septic arthritis of the shoulder and acute osteomyelitis. Fracture of the humerus or clavicle will also present with a pseudoparalysis of the shoulder. Although rare, congenital malformations of the plexus can also occur. Spinal cord injury, cerebral palsy, or other central nervous system (CNS) lesions can also be initially present with hypotonia of the involved upper extremity.

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The utility of magnetic resonance imaging (MRI) (Fig. 3) in the evaluation of a child with OBPP is useful and has replaced more invasive CT (computed tomography) myelography. In a young child, this test requires sedation or formal anesthesia, which is not without risk in this patient population. While myelographic or MRI evidence of pseudomeningoceles is associated with nerve root avulsions, the testing characteristics of these modalities in infants remain suboptimal. CT myelogram has only 69% sensitivity, 89% specificity, and a positive predictive value of 50% for detecting nerve root avulsions. The negative predictive value of CT myelogram is 93%.\textsuperscript{11} The MRI finding of a pseudomeningocele has a low sensitivity (approximately 50%), but a high specificity (approaching 100%) in the diagnosis of nerve root avulsion.\textsuperscript{12} MRI information is valuable for preoperative planning; intraoperatively, this information is supplemented with neurophysiology and histopathology data for decision making.

Electomyelography and nerve conduction studies can have prognostic value in infants but must be performed according to a strict protocol, as described by Birch\textsuperscript{11} and others. Reliance on serial physical examinations of these children is of primary importance.

**Classification**

Perhaps, for us, the most useful classification of brachial plexus birth lesions is a clinico-anatomic one and will form the basis of the remainder of this discussion. C5-C6 lesions involve paralysis of primarily upper trunk innervated muscles. Clinically, these infants present with weakness of shoulder elevation and external rotation, absent biceps, and the classic shoulder internal rotation and elbow extension posture. C5-C6-C7 lesions also show weakness of both triceps and wrist extension, and sometimes thumb weakness. Global palsies present at birth with a generally flail, insensate limb, and often upper eyelid ptosis.

We present here a focused review of the contemporary early treatment options for these infants. Delineation of the precise natural history of OBPP remains elusive because of the heterogeneous cohorts of patients, methodological flaws, and inability to ensure long-term follow-up in these children in early retrospective series. These limitations explain the intense controversies that continue to surround the critical questions regarding the specific indications and optimal timing of “early” microsurgical intervention in these children.

**Natural History Following Nonoperative Treatment of Brachial Plexus Birth Injuries**

A critical analysis of recent published data\textsuperscript{4,5,7,11,14-19} suggests that, indeed, the majority of infants will demonstrate spontaneous ongoing motor recovery with observation alone. A study from the British pediatric surveillance unit found that 53% of children with OBPP spontaneously recovered to normal or nearly normal levels, while an additional 39% regained “good” function of the upper limb.\textsuperscript{19} It is well accepted that classic Erb’s palsy has the best prognosis, with up to 90% reported rates of spontaneous recovery.\textsuperscript{20} C7 involvement, however, is associated with an 80% risk of poor recovery in upper plexus palsies. Lower plexus involvement has the poorest prognosis, with some studies showing that less than 10% will recover any useful function of the hand.\textsuperscript{4} Due to its poor prognosis, many centers use pan-plexus lesions as an indication for early (i.e., 3 months of age) surgical intervention.

Gilbert and Tassin’s\textsuperscript{2} landmark series demonstrated that the absence of biceps anti-gravity function by 3 months of age portends a poor prognosis with regard to shoulder function. A single-institution longitudinal study\textsuperscript{15} of 91 infants treated nonoperatively with only physical and occupational therapy over a 7-year period highlighted the variable subcohorts comprising this complex patient population and their associated unique clinical courses with observation alone.
Specifically, 63 children with an upper or upper-middle plexus injury (Group 2) recovered good-to-excellent shoulder and hand function at a minimum of two-year follow-up. Additionally, serial evaluations documented British Medical Research Council M4 or better motor strength in the biceps and deltoids (Table 1). In contrast, 12 infants with global palsies (Group 1) demonstrated only M0 to M1 in these critical muscles at 6 months of age. Sixteen patients with a similar clinical presentation to Group 2 (Group 3) demonstrated minimal spontaneous recovery of biceps and deltoid function by 6 months of age. These observations suggest that children with rapidly progressive motor recovery during the first 3 to 4 months of life are manifesting resolution of a neuropraxic injury, while infants with upper and upper-middle plexus injuries, who fail to show motor improvement between 3 to 6 months of age (i.e., Group 3), have likely sustained a neurotemetic or dense axonotemetic injury, and should be managed in a similar fashion to infants with global palsies. Furthermore, all children with minimal spontaneous recovery of deltoid and biceps function by 6 months of age were found to have only poor-to-fair shoulder function and fair-to-satisfactory hand function on the Gilbert Shoulder and Gilbert-Raimondi Hand scales, respectively (Tables 2 and 3). It is thus crucial to perform thorough serial examinations in order to identify this subcohort of children who should be offered early microsurgical intervention.

Similarly, in a prospective study of 66 patients designed to compare the natural history of nonoperative management of OBPP with that of microsurgical repair in patients who demonstrated no biceps function at six months of age, Waters\textsuperscript{14} found that children without biceps recovery after 3 months were all Narakas groups 3 and 4 (i.e., more severe neurological involvement with persistently weak digital extensors or flail arm). The later the return of biceps function the poorer the ultimate shoulder function. The scores for every aspect of the Mallet shoulder function (Fig. 4) decreased with delayed biceps return. In this series, no child with biceps recovery after 3 months had full spontaneous recovery of shoulder function.

In a review of 28 patients with absent biceps function at 3 months, Smith and colleagues reported that no patient recovered normal shoulder function, and only 12 patients regained good shoulder function.\textsuperscript{6} Specifically, shoulder function decreased with increasing delay in biceps recovery. All patients with C5 and C6 lesions regained biceps function by 6 months. Conversely, no patient with Horner's syndrome at birth regained biceps function by 6 months.

These recent data\textsuperscript{5,6,11,14-23} allow for several conclusions that impact the current algorithm of treatment in these children:

1. The majority of upper brachial plexus birth injuries are transient;
2. Global palsies have a poor prognosis with nonoperative treatment;
3. Failure to recover anti-gravity biceps function by 3 to 6 months of age is a poor prognostic sign; and
4. Infants with C5-C6 or C5-C6-C7 injuries may sometimes continue to demonstrate spontaneous improvement between 3 and 6 months of age; thus, precluding the need for early surgery.

Therefore, the true challenge to the treating physician is the correct identification of the subcohort of infants who will either plateau or fail to show any neurological improvement. This highlights the crucial need for careful serial, complete neurological examinations.

**Early Microsurgical Treatment Options**

It is generally accepted that the case of an infant presenting with a global C5-T1 injury (i.e., flail, insensate limb and upper eyelid ptosis) and no significant recovery within the first 8 to 10 weeks of age is an absolute indication for surgery by 4 to 5 months of age.\textsuperscript{24-27}

For C5-C6 and C5-C6-C7 lesions, there is no Level I evidence indicating that microsurgical intervention at less than 6 months of age yields superior results to those obtained following surgery at 8 to 9 months. Some very experienced groups advocate extending this “window” of observation to allow an additional small percentage (albeit not previously quantified) of these patients to manifest additional motor recovery and thus avert surgery. For example, Michelow and associates\textsuperscript{7} observed that if at 3 months elbow flexion

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**Table 1** Medical Research Council Muscle Grading System

<table>
<thead>
<tr>
<th>Observation</th>
<th>Grade</th>
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<tbody>
<tr>
<td>No contraction</td>
<td>M0</td>
</tr>
<tr>
<td>Trace contraction</td>
<td>M1</td>
</tr>
<tr>
<td>Active movement-gravity eliminated</td>
<td>M2</td>
</tr>
<tr>
<td>Active movement against gravity</td>
<td>M3</td>
</tr>
<tr>
<td>Active movement against gravity and resistance</td>
<td>M4</td>
</tr>
<tr>
<td>Normal strength</td>
<td>M5</td>
</tr>
</tbody>
</table>

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**Figure 4** Mallet Shoulder Classification.
Table 2  Modified Gilbert Shoulder Evaluation Scale

<table>
<thead>
<tr>
<th>Grade</th>
<th>Criteria</th>
</tr>
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<tbody>
<tr>
<td>0 (None)</td>
<td>Completely paralyzed shoulder or fixed deformity</td>
</tr>
<tr>
<td>1 (Poor)</td>
<td>Abduction = 45°  No active external rotation</td>
</tr>
<tr>
<td>2 (Fair)</td>
<td>Abduction &lt; 90° Bi active external rotation</td>
</tr>
<tr>
<td>3 (Satisfactory)</td>
<td>Abduction = 90° Active external rotation &lt; 30°</td>
</tr>
<tr>
<td>4 (Good)</td>
<td>Abduction &lt; 120° Active external rotation 10° - 30°</td>
</tr>
<tr>
<td>5 (Excellent)</td>
<td>Abduction &gt; 120° Active external rotation 30° - 60°</td>
</tr>
<tr>
<td>6 (Very Good)</td>
<td>Abduction &gt; 150° Active external rotation &gt; 60°</td>
</tr>
</tbody>
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Table 3  Gilbert/Raimondi Classification of Impairment of the Hand in Patients with Obstetric Palsy

<table>
<thead>
<tr>
<th>Grade (Function)</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 (None)</td>
<td>Complete paralysis or slight finger flexion of no use, useless thumb—no pinch, some or no sensation.</td>
</tr>
<tr>
<td>1 (Poor)</td>
<td>Limited active flexion of fingers; no extension of wrist or fingers; possibility of thumb lateral pinch.</td>
</tr>
<tr>
<td>2 (Fair)</td>
<td>Active extension of wrist with passive flexion of fingers (tenodesis)—Passive lateral pinch of thumb (pronation).</td>
</tr>
<tr>
<td>3 (Satisfactory)</td>
<td>Active complete flexion of wrists and fingers—mobile thumb with partial abduction—opposition intrinsic balance—no active supination; good possibilities for palliative surgery.</td>
</tr>
<tr>
<td>4 (Good)</td>
<td>Active complete flexion of wrist and fingers; active wrist extension—weak or absent finger extensor; good thumb opposition with active ulnar intrinsics; partial prosupination.</td>
</tr>
<tr>
<td>5 (Excellent)</td>
<td>Hand IV with finger extension and almost complete prosupination.</td>
</tr>
</tbody>
</table>

was used to predict recovery at 12 months, the proportion of patients whose recovery was incorrectly predicted was 12.8%, implying that 1 in 8 patients would have had surgery unnecessarily or would not have had surgery when required. When elbow flexion was combined with elbow, wrist, thumb, and finger extension, recovery was incorrectly predicted in only 5.2% of cases. Similarly, Clarke and Curtis observed that 10% to 15% of infants without biceps contraction at 3 months would still spontaneously recover by 9 months. The senior investigator’s (JAIG) observations support this as well.

Persistent deficits in active wrist extension at 4 to 5 months with recovery of hand function is highly suggestive of a poor outcome, especially regarding the shoulder without surgical intervention.

The clinical approach for children older than 8 to 12 months of age with persistent proximal motor weakness and fixed or progressive shoulder deformity has received little attention. Often, delayed orthopaedic procedures (i.e., contracture release, tendon transfers, humeral osteotomy) are recommended. Birch and coworkers reported promising results following late grafting into the suprascapular nerve. Subsequent series suggest that late nerve reconstruction in properly selected infants with persistent severe shoulder sequelae and upper plexus injuries may be beneficial. Grossman and colleagues reported an improvement of two grades or more on the modified Gilbert shoulder scale in 22 infants who underwent combined plexus and shoulder reconstruction at a mean age of 16 months. Grossman and associates have also shown that following neurolysis alone (n = 3) or neurolysis with bypass grafting (n = 8) at a mean age of 13 months, the median increase by the modified Gilbert shoulder scale was 2 grades and that 6 of 11 cases improved by at least 3 grades. Van Ouwerkerk and coworkers reported promising results in 54 children following isolated spinal accessory to suprascapular nerve transfers, performed as a primary or secondary procedure at a mean of 21 months of age in children without return of active shoulder external rotation. A recent review of our experience suggests that direct spinal accessory to suprascapular nerve transfer provided similar clinical and functional outcomes when performed prior to or after 9 months of age. While these studies are retrospective and without a control group in which established musculoskeletal procedures (contracture release, tendon transfer, humeral osteotomy) are performed, the results achieved following late nerve reconstruction appear to warrant this approach in experienced centers.

Microsurgical Options

With advancement in microsurgical techniques by Narakas, Millessi, Gilbert and Tassin, Kawabata and colleagues, and others, interest in the microsurgical reconstruction of brachial plexus injuries grew. An in-depth discussion of current microsurgical techniques and options used for brachial plexus reconstruction is beyond the scope of this paper, but the spectrum includes neurolysis, neuroma resection with nerve grafting, neurolysis and bypass grafting, and nerve transfers (Fig. 5). Direct repair is rarely performed due to the extensive nature of these injuries and the inability to achieve a tension-free repair without interposition grafting.

While Laurent and associates advocate performance of neurolysis alone in the presence of a neuroma-in-continuity, which demonstrates greater than 50% maintenance of action potential upon intraoperative stimulation, many surgeons have abandoned this approach. Capek and colleagues reported better long-term results with neuroma resection and grafting, as compared to neurolysis alone in the setting of conducting and nonconducting neuromas, despite an initial
down-grading following resection. To avoid initial down-grading, some experts advocate neurolysis with bypass grafting over neuroma resection and graft reconstruction. The intraoperative decision-making process is complex and requires consideration of multiple preoperative and intraoperative findings. Interestingly, despite the long duration and complexities of these procedures, the complication rate is extremely low. In an analysis of a single-surgeon experience of 100 consecutive cases over a 30-month period, there were no major complications (i.e., phrenic nerve injury, pneumothorax, chylothorax, pulmonary edema, or wound infection), and only eight minor complications (intraoperative wheezing, n = 5; prolonged hospitalization for poor oral intake, n = 2; and bronchiolitis, n = 1).

Glenohumeral deformity occurs early and is progressive in children who do not demonstrate spontaneous recovery, its occurrence is secondary to the muscle imbalance that develops about the shoulder. At the time of primary plexus exploration, the glenohumeral joint must be assessed. Our indications to perform a concomitant glenohumeral reconstruction include inability to obtain full passive external rotation of the shoulder when the upper limb is positioned adducted at the patient’s side and the elbow flexed, or if there is joint instability. Reconstruclive options for the associated internal rotation contracture include capsuloplasty, open reduction and posterior capsulorrhaphy, pectoralis major tenotomy, and subscapularis slide. Further, there is data to support consideration of botulinum toxin type A injections into the internal rotation muscle groups about the shoulder (i.e., pectoralis major and the latissimus dorsi-teres major complex) at the time of the primary plexus reconstruction (based on the experience of AP and VAIG). Use of botulinum toxin in this setting has been shown to reduce the incidence and severity of recurrent internal rotation contracture of the shoulder following a subscapularis slide. Finally, it must be emphasized that in the reconstruction of global injuries, priority must be given to reinnervation of the hand as opposed to adult cases where emphasis is placed on recovery of shoulder stability and elbow flexion. In infants, the short distance of nerve regeneration needed to reach the hand allows for this approach. Furthermore, unless some hand function and sensibility is recovered, the extremity will be essentially ignored even with excellent active shoulder and elbow motion.

Multidisciplinary Approach
A multidisciplinary approach is crucial to optimizing the outcome of these children. Our model consists of a team comprised of specialists in child neurology, neurophysiology, and occupational therapy who serially evaluate these infants. The collective combined expertise of these specialists is used in collaboration with the treating orthopaedic and peripheral nerve and hand surgeons for decision making in the immediate perioperative period and during long-term follow-up. Experienced neuroradiologists, pathologists, and pediatric anaesthesiologists as well as pediatric hospitalists round out the team.

Summary
Infants with brachial plexus birth injuries represent a complex, heterogeneous group of patients. Current controversies surrounding optimal timing and early microsurgical intervention will continue even as large patient cohorts and long-term follow-up designed to answer these questions are available. There is no “cookie-cutter” approach available to treat these unique patients; intraoperative decision making is based on the extent of the injury, preoperative and intraoperative findings, available reconstructive options, and surgeon experience. The learning curve is significant with regard to both the nonoperative and operative management of these children. Serial and thorough clinical examinations of the infant are paramount to identifying the small segment of patients who will definitively benefit from early surgical intervention. A multidisciplinary approach incorporating various health professionals is crucial to optimizing functional outcomes in these children.

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None of the authors have a financial or proprietary interest in the subject matter or materials discussed, including, but not limited to, employment, consultancies, stock ownership, honoraria, and paid expert testimony.

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