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Primary Nerve Reconstruction in Brachial Plexus Birth Palsy: Current Concepts

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Abstract

This article discusses the role of primary nerve surgery for Brachial Plexus Birth Palsy (BPBP). Spontaneous recovery in brachial plexus birth palsy is known and in such cases the prognosis is good. However, the incidence of such recovery is 30–90%. In some infants however, the course of motor recovery is inadequate necessitating nerve repair. BPBP presents clinically as a lower motor neuron type of upper limb monoplegia at birth. Indications of primary nerve reconstruction or distal nerve transfer, the technique of exploration of brachial plexus, common anatomical variations and the methods for identification of intact roots are discussed in detail.

Keywords: Primary nerve reconstruction; Brachial Plexus Birth Palsy (BPBP); Nerve transfer; Co-contractions; Brachial plexus surgery.


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Introduction

This article discusses primary nerve surgery for Brachial Plexus Birth Palsy (BPBP). Spontaneous recovery in brachial plexus birth palsy is known and in such cases the prognosis is good. However, the incidence of such recovery varies between 30-90% [1]. In other infants however, the course of motor recovery is inadequate necessitating nerve repair. The diagnosis of BPBP is clinical in the form of lower motor neuron type of monoplegia in one upper limb at birth. Unlike cerebral palsy, there is no associated spasticity.

Indications of primary nerve reconstruction or distal nerve transfer, standard exploration of brachial plexus, common anatomical variations & intact root identification have been discussed in detail in this article.

Investigations in BPBP

Besides clinical findings, pre-operative investigations help in identifying the level, type & severity of the lesion, provides prognosis for recovery and plan surgical treatment strategy.

The following investigations are commonly performed in such cases.

- Plain radiographs of the chest and affected upper limb
- Electrodiagnostic study
- MRI Brachial Plexus (preferably 3 Tesla scanner)
- CT myelography (less common)

In BPBP, elevation of the diaphragm on the affected side on a plain X-ray of the chest indicates Phrenic nerve palsy. An X-ray of the affected limb helps to rule out pseudo-paralysis due to clavicle or humerus fractures and septic arthritis of the shoulder.

Electrodiagnostic study helps in diagnosing neuropraxia versus higher grade of axonal damage, severity of axonal damage and avulsion of roots. Preganglionic lesions are differentiated from postganglionic lesions by preserved sensory action potential and presence or loss of compound motor action potential (CMAP) in nerve conduction study. The complete absence of H-reflex has been correlated with poor prognosis in BPBP [2]. Awell documented electrodiagnostic study with clinical findings can give more accurate status of location, severity of the injury and indications of surgery [3].

The role of needle EMG in infants is controversial, however it is routinely performed in centres with an experienced electrophysiologist. Presence of fibrillation potential and positive sharp waves indicate denervation in muscle. The number and pattern of motor unit action potential denotes the extent of muscle reinnervation. It also helps in documenting co-contraction, a common phenomenon in BPBP. In delayed presentation, the time limit for nerve surgery may depend up on physiological status of the muscle. EMG may help in determining activity in denervated muscle.

3T MRI is a non-invasive modality but needs anaesthesia/sedation. It helps in localizing pseudo-meningocele, root avulsion (82% specificity) [4] and the extent of injury. The dorsal & ventral roots can be individually identified. It is particularly useful in case of root avulsion without pseudomeningocele formation.

Pseudomeningocele on CT myelography (CTM) indicates nerve root avulsion (pre-ganglionic injury to the brachial plexus). The diagnostic sensitivity for avulsion of C8 and T1 roots is 75% [4]. The procedure is performed by lumbar puncture and injecting iodinated contrast solution intrathecally to look for contrast between CSF and nerve roots by CT scan. In view of exposure to radiation and being an invasive procedure, it is less popular in cases of BPBP [5].

Classification of Brachial Plexus Birth Palsy (BPBP)

Dr Narakas from Switzerland gave the eponymous 'Narakas Classification' of Brachial Plexus Birth Palsy [6]. Table 1 shows the classification. Type I and II refer to upper plexus injury i.e. C5 C6 and C5 C6 C7, respectively; Triceps and wrist extensor function being the differentiating factor.

Clinical Evaluation

The babies have usually suffered severe traction injuries and are likely to have associated skeletal and soft tissue injuries in Narakas type III and IV cases. It is not uncommon to notice a large cephalohematoma and clavicular humeral fractures. Chest and abdominal examination are essential to exclude diaphragmatic palsy. Presence of Horner's syndrome connotes a poor prognostic sign and an early need for surgery [7].

Motor examination to determine recovery and to measure functional outcome

Motor examination of the affected limb of the infant is the main clinical criterion. In Group I and II, this includes, shoulder abduction, shoulder external rotation, elbow flexion, elbow extension, wrist extension, prono-supination of the Radio Ulnar Joint. This is because the root values of muscles performing these functions are C5 C6 C7.

Motor examination can either be done by the Medical Research Council(MRC) scale 8 (Table II) or by the Toronto AMS system [9] (Table III). The AMS is popular in North America however in the rest of the world a majority of people use the MRC scale or the modified MRC for children (MMRC) [8].

Timing of nerve surgery

There is a lack of consensus with regard to the indications and timing of nerve surgery in BPBP. Periodic clinical assessment is the primary factor for determining treatment.

In upper plexus injury with good hand function, exploration of the brachial plexus is advised if there is no recovery of antigravity biceps function at three months of age. Infants with biceps recovery after 3 months of age had partial improvement [10]. The presence of Horner's syndrome and a flail limb at 3 months of age is an absolute indication for exploration of the brachial plexus, neuroma excision and nerve grafts or nerve transfers depending on the availability of roots [11]. Waters et al reported that amongst infants without antigravity biceps at three months of age, 85% of them subsequently recovered biceps function by 6 months of age. In infants with biceps recovery between 3 and 6 months, there was a reduction in shoulder function on follow-up beyond one year of age. This subset of children required secondary surgeries to improve shoulder function [12]. Al-Qattan proposed waiting until 4

Table 1: Narakas Classification

Type	Description
I	Erb's palsy involving C5, C6 Abduction/external rotation of shoulder & elbow flexion affected.
II	Extended Erb's palsy involving C5, C6 & C7
	Abduction/external rotation of shoulder, elbow flexion & extension affected Wrist extension is also affected.
III	Global palsy with no Horner syndrome involving C5, C6, C7, C8 & T1
IV	Global palsy with Horner syndrome involving C5, C6, C7, C8 & T1

Table 2: Modified MRC grading system (Kendall and McCreary, 1993)

Movement	Function of the Muscle	Grade		
No Movement	No contractions felt in the muscle	0	0	0
	Tendon becomes prominent or feeble contraction felt in the muscle, but no visible movement of the part	T	1	Trace
Supported in the horizontal position	**Movement in Horizontal Plane**			
	Moves through partial range of motion	1	2-	Poor-
	Moves through complete range of motion	2	2	Poor
	Antigravity Position	3	2+	
	Moves through partial range of motion			
Test Position	Gradual release from test position	4	3-	Fair-
	Holds test position (no added pressure)	5	3	Fair
	Holds test position against slight pressure	6	3+	Fair+
	Holds test position against slight to moderate pressure	7	4-	Good-
	Holds test position against moderate pressure	8	4	Good
	Holds test position against moderate to strong pressure	9	4+	Good+
	Holds test position against strong pressure	10	5	Normal

Table 3: Toronto Active Movement Scale

With gravity eliminated	
No contraction	0
Contraction without movement	1
Movement less than half of range of movement	2
Movement more than half of range of movement	3
Full movement	4
Against gravity	
Movement less than half of range of movement	5
Movement more than half of range of movement	6
Full movement	7

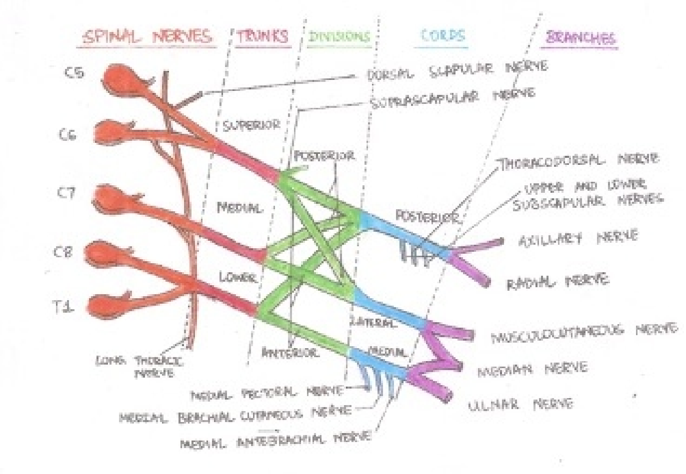

Figure 1: Pictorial presentation of brachial plexus

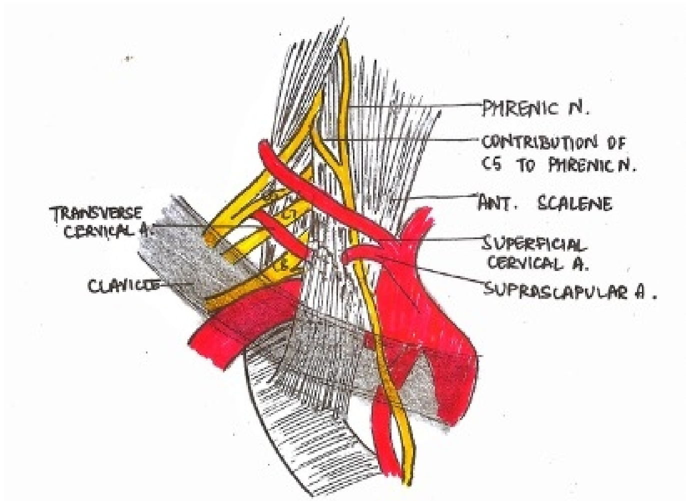

Figure 2: Brachial plexus in relation to muscles and clavicle.

months of age rather than 3 months before considering surgery. He also stated that if an infant develops biceps function at two months, probably child will never require nerve surgery [13]. According to Clarke and co-workers, waiting for nine months before embarking on nerve surgery is worthwhile if all other scores of movements except biceps are good and not otherwise. They suggested to perform cookie test at nine months and if child fails cookie test, it mandates nerve surgery [11].

Based on the literature, we follow an algorithm depending on the type and severity of injury. We consider electro-physiological study at one month and later at 3 months. In a case of total paralysis with or without Horner's syndrome, we recommend nerve surgery at 3 months. Timing in upper plexus injury is more complex and discussed in detail below.

Though nerve grafting can be done as late as two years in BPBP, the results are less encouraging than surgery performed at the right time.

Figure 3: Marking for surgical exposure.

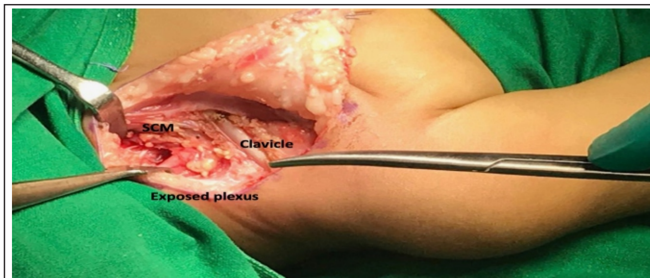

Figure 4: Exposed brachial plexus in relation to clavicle and sternocleidomastoid.

Management of Brachial Plexus Birth Palsy (BPBP)

Surgical anatomy & common variations (Fig. 1)

The brachial plexus receives contributions contiguously from the fifth cervical (C5) through the first thoracic (T1) ventral spinal nerve roots. Prefixed cords (22%) receive an additional contribution from the C4 nerve root, whereas the less common postfixed cords (1%) receive a contribution from T2 [14].

The proximal-to-distal orientation of the brachial plexus is nerve roots, trunks, divisions, cords, and terminal branches (Fig. 2). The divisions are located behind the clavicle [15].

Anatomic variations of brachial plexus have been studied by Emamhadi et al [16]. In a study of 32 cadavers, 18% of plexuses were prefixed in origin. The suprascapular nerve in 22% of plexuses was formed from posterior division of the trunk. Terminal branches variations were the highest wherein the ulnar nerve received a communicating branch from the lateral cord in 9% of cases. The median nerve was formed by 2 lateral roots from lateral cord and 1 medial root from the medial cord in 18% of cadavers.

Some fibres from the C7 root contributed to the musculocutaneous nerve in 25% of cadavers.

Identification of landmarks for important nerves in the brachial plexus

Standard exposure of the brachial plexus in the angle between the clavicle and sternocleidomastoid is shown in Fig. 3 and 4. The spinal accessory nerve can be traced on the deep anterior surface of the trapezius muscle. After identifying omohyoid muscle, Erb's point is to be identified first followed by C7 root, C5-C6 below and superiorly and C8 T1 on the medial end near scalenus medius [17].

Phrenic nerve is located on the anterior aspect of scalenus anterior muscle. Suprascapular nerve is dissected on the outer border of Erb's point mainly at the posterior division of the upper trunk.

For infraclavicular exposure, the incision proceeds distally between the middle and lateral thirds of the clavicle to the coracoid and further distally to the anterior axillary fold. This incision can be connected with the supraclavicular fold on the medial aspect. For better exposure of the retroclavicular plexus, a clavicle osteotomy can be performed although this is seldom necessary in practice. It also carries the risk of non-union/malunion as one of the senior authors has observed [18].

Intraoperative identification of a nerve root [19]

The continuity of nerve roots has to be traced at intra-foraminal level under microscope or loupe magnification. The usefulness of a root for neural reconstruction is judged based on pre-op clinical findings, electrodiagnostic study and intra-operative neuroma formation with its extent, location and state of fibrosis around the scaleni muscles. The nerve root may be considered appropriate if found intact at the intra-foraminal level, good quality at the juxta foraminal level and less fibrosis around epineurium of root. Inter-fascicular continuity while dissecting the neuroma helps in differentiating axonotmesis and neurotmesis. Electrical stimulation of the root proximal to the neuroma may show contractions in muscle distally. An example of a good quality, usable nerve root is shown in Fig. 5.

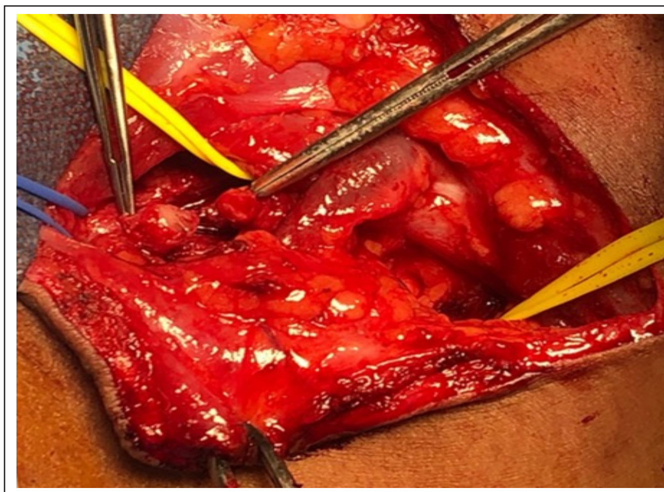

Figure 5: Good quality nerve root.

Consensus on Narakas type 1 & 2 treatment strategy

The current debate centres around two issues

1. Whether spontaneous recovery occurs in a significant proportion of cases.
2. How long does one wait?

As mentioned earlier, various studies estimate the rate of spontaneous recovery between 30-90% [1]. For the 2nd point, we do not think one clear answer is possible, however a review of literature will be useful to help us make our own conclusions.

Decision Making

There are two schools of thought with regard to the ideal timing of nerve surgery in BPBP. Gilbert and Tassin [20] published their findings that for upper plexus injury achieving 'anti-gravity elbow flexion' by 3 months of age was a sign that primary nerve surgery was not required. Clarke and co-workers from Toronto developed the AMS score, validated it and used it for decision making with a different strategy of waiting up to 9 months of age prior to considering nerve surgery. They also developed the 'cookie test' which essentially entails the infant eating a cookie/biscuit with the affected limb. This task demonstrates the ability for antigravity elbow flexion in addition to a successful hand-to-mouth reflex.

Consensus Building

Multiple groups have attempted to answer the question of spontaneous recovery and timing of nerve surgery [21]. To encapsulate the findings, the following general statements can be made.

1. Biceps function is used because it is the only muscle with C5-C6 innervation responsible for a single movement namely elbow flexion. This provides a unique insight into the continuity of the C5, C6 nerve roots.
2. Many children with upper plexus injury who do not have an antigravity Biceps at 3 months will develop it later, some as late as 1 year.
3. Those who do not develop antigravity Biceps by 6 months will have a poorer overall outcome, especially in shoulder function.
4. Biceps function therefore, is not the end goal, but a useful indicator of upper plexus recovery.

The current algorithm therefore is of constant monitoring of the child using electrophysiological studies and CMAP of the muscles in comparison to the opposite side and monitoring and documenting co-contractions which can hamper clinical goals. For example, a strong Biceps-Triceps co-contraction can hamper elbow flexion despite adequate CMAP and innervation of both muscles. In this situation, Botulinum Toxin injection to the triceps will allow biceps function to become more evident during clinical examination. If, however there is neither a strongly felt biceps nor any co-contraction, the decision to explore the plexus should not be delayed beyond 6 months of age.

Intra-plexus Repair versus Distal transfers

The current literature shows an overwhelming preference for intra-plexus repair from root stumps to Trunks/Divisions/Cords [22]. However, following Oberlin's publication of the Ulnar to Biceps distal transfer, some authors have used distal nerve transfers in BPBP [23]. A review article on distal transfers is available that provides further details [24, 25]. In cases where the roots are avulsed, there is no option but to perform distal transfers. However, in South Asia (and perhaps many other countries outside western Europe and North America) there is the added parental pressure to not excise a neuroma with partial/inadequate function, prompting the surgeon to opt for distal transfers.

Distal nerve transfers to biceps (with ulnar nerve fascicle as a donor nerve) and suprascapular nerve (with spinal accessory nerve as a donor nerve) have produced encouraging results, irrespective of the type of injury (Needs reference). The International Federation of Societies for Surgery of the Hand (IFSSH) document on the subject states that there is no general consensus to determine the choice of nerve surgery [26].

Consensus on management of Narakas Type 3 & 4 cases

The severity of injury is the key determinant of prognosis and the need for surgical intervention. Narakas et.al. [6] have classified these injuries in four groups. Group 3 cases have total paralysis with a flail extremity and is reported in 9-26% of obstetrical palsies [27]. Group 4 cases have Horner's syndrome in addition to total paralysis and represent approximately 2% of brachial plexus palsy cases [28]. These two groups carry a worse prognosis when compared to Group 1 and 2 especially where intra-plexus donors are unavailable due to avulsion. Therefore, early nerve reconstruction surgery is recommended in all children with a Group III/IV injury.

Surgical Reconstruction

Unlike post-traumatic adult brachial plexus injuries, restoration of hand function is of paramount importance in Group III/IV injuries. The next priority is restoring elbow flexion, followed by shoulder stabilization and shoulder adduction.

During exploration, an attempt is made to identify one or more viable roots. If present, nerve grafts are placed between the available root/roots and distal plexus. If all 5 roots are available, which is rare, they can simply be reconnected to their distal parts with nerve grafts. Where three roots are available, each one of them is directed to the lateral, posterior and medial cords respectively. In the presence of two viable roots, the better root is targeted towards the lower trunk and the other root to the posterior or lateral cord. When the posterior cord is chosen as the second recipient, the musculocutaneous nerve is innervated with three intercostals nerves. A single available root is always directed to lower trunk or medial cord. Additional extra-plexal nerve transfers such as XI nerve to Suprascapular Nerve and Intercostal Nerves to Musculocutaneous nerve are performed to improve shoulder and elbow function (Fig. 6).

In the absence of an intact root, extraplexal nerve transfers are performed with spinal accessory nerve, intercostal nerves and contralateral C7 as donor nerves. Spinal accessory nerve is preferred for suprascapular nerve. Good results have been reported when three intercostal nerves

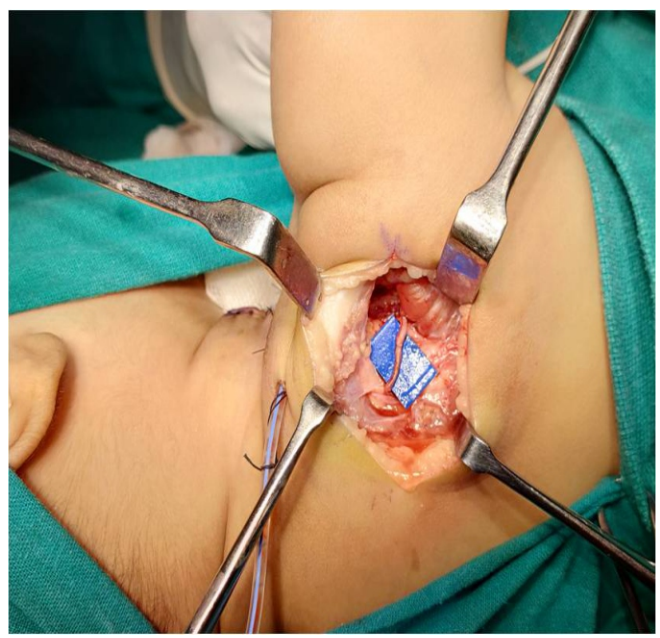

Figure 6: Nerve transfer between XIth cranial nerve to Suprascapular nerve.

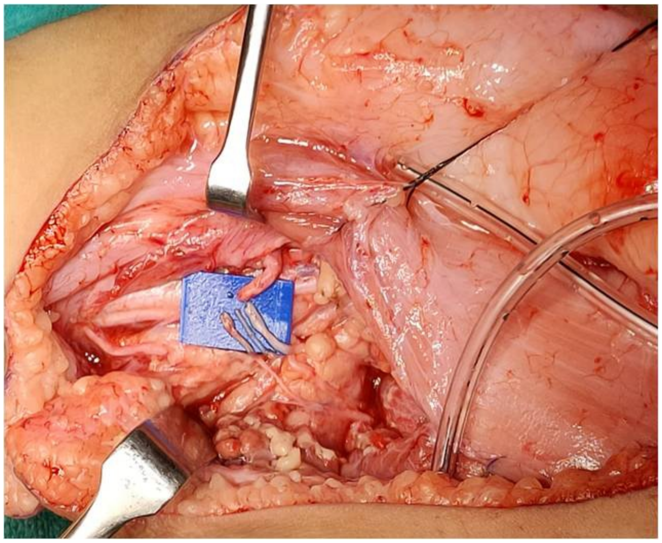

Figure 7: Nerve transfer between ICNs to Musculo-cutaneous nerve (MCN).

Figure 8: Narakas Type 4 pan brachial plexus birth palsy with Horner syndrome.

Figure 9: M3 grade elbow flexion following ICNs to MCN.

are transferred to the musculocutaneous nerve [29] (Fig. 7) with 82-100% of patients obtaining M3 elbow flexion (Fig. 8 and 9). Intercostal nerve transfer can be undertaken safely in infants if the ipsilateral phrenic nerve is functioning normally. However, intercostal nerves harvesting risks alterations in chest growth and breast development [10].

Transfer of the contralateral C7 via a vascularized ulnar nerve graft has been described for total plexus palsy with 4 or more avulsions [29]. This transfer necessitates the use of a vascularized ulnar nerve graft which precludes possibility of intrinsic hand function from spontaneous recovery. However, good results are seen with the use of cable grafts which allows the ulnar nerve to be innervated as well.

The recovery of shoulder function after nerve reconstruction is far superior to the natural history in Narakas 3 or complete lesions. Early nerve reconstruction followed by tendon transfers can provide 70% better elbow and hand function at 8-years follow up [30].

Summary

- Up to 50% of children with BPBP show natural recovery. Recovery of anti-gravity biceps function at 3 months is an important positive prognostic sign for spontaneous recovery.
- Completely flail upper limb with or without Horner syndrome needs early intervention.
- Nerve reconstruction or nerve transfer depend upon availability of intact roots, consent from parents to deal with neuroma and a targeted strategy to restore maximal hand function.
- Children with presence of weak or slow recovering biceps at 3 to 6 months should be assessed for co-contractions through clinical examination and electrophysiological & imaging studies; a final decision is taken by 6 months in most cases.
- We recommend that every infant with BPBP needs to be examined and treated by an experienced clinical team. Even cases with severe grade injuries can be expected to achieve reasonably good function following nerve reconstruction.

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