46 Operative Brachial Plexus Surgery: Brachial Plexus Birth Injury – Neurodiagnostic Evaluation

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Abbreviations

BPBI	Brachial plexus birth injury
CMAP	Compound motor action potentials
EDX	Electrodiagnostic testing
EMG	Electromyography
NAPs	Nerve action potentials
SSEPs	Somatosensory evoked potentials

Introduction

The optimal timing of surgical treatment for BPBI constitutes a major dilemma and is the subject of considerable debate. Data from animal studies suggest that early nerve repair leads to better functional results, but currently, time is needed to clinically distinguish milder lesions, which do not require surgery from more severe lesions, which should be treated as soon as possible. Currently, neurological evaluation at 3 months is generally regarded as the main criterion for the fundamental decision to perform nerve surgery or not. Ideally, the aim of electrodiagnostic testing (EDX) should be to identify injuries that require surgery well before this 3 month time point. Depending on test characteristics, such a test would have two benefits: if the test is specific enough, surgery can be performed on those that require it at the earliest possible time, and a highly sensitive test would reduce uncertainty for parents of children with lesions that are expected to recover spontaneously.

Indeed, in adult patients with a brachial plexus traction injury, EDX is widely used and has been proven a useful tool for diagnosis and prognostication. There is controversy, however, whether the use of EDX is as useful in children with a BPBI. Some argue that EDX has no added value at all [1, 2], whereas others strongly recommend performing EDX on all patients with BPBI [3].

Similarly, the value of intraoperative neurophysiological testing is uncertain. A neurophysiological test that allows the identification of nerve elements or will show spontaneous recovery would greatly enhance surgical decision-making. Intraoperative neurophysiological monitoring is widely used, but its exact effect on the outcome of surgery remains uncertain.

A recent systematic review of the literature identified 16 observational studies with a total sample size of 747 children [4]. A wide variation was found in EDX techniques, outcome algorithms, and decision-making; pooling of data proved impossible. Risk of bias and quality of evidence were rated. Nevertheless, the most

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methodologically sound studies were claimed to support the use of EDX, at standardized time frames, as prognostic modality complementing clinical evaluation and neuroimaging.

To understand the difficulties in interpretation of EDX in children with BPBI, it is necessary to consider the specific pathophysiology of BPBI. As in any nerve, the traction injury in BPBI may result in neurapraxia, axonotmesis, neurotmesis, or root avulsion [5]. The main difference between adult traumatic lesions of the brachial plexus and BPBI is that the traction forces are low-velocity and with longer duration, which will probably lead to a different type of traction injury. In effect, during surgical exploration of BPBI, a true rupture as found in adults is encountered seldomly. The key finding of postganglionic injury in BPBI is the *neuroma-in-continuity* in which only impaired, disorganized axonal outgrowth has taken place [6, 7]. It may be more appropriate to classify these neurotmetic lesions between a grade 3 and grade 4 severity according to the Sunderland classification [8]. Some axonal outgrowth is usually encountered through the neuroma. This axonal continuity may result in electrodiagnostic continuity. The related clinical recovery is, however, variable and unpredictable. In this respect a gold standard of the diagnosis to compare neurophysiologic findings is unavailable, and there is a risk of circular reasoning [9]. Additionally, cross-fiber excitation inside the neuroma-in-continuity may further complicate interpretation [10]. Another striking feature in BPBI is axonal misrouting [11]. This may lead to unexpected electromyographic findings in different myotomes. A second difficulty for efficacy of EDX in BPBI is that the resulting nerve injury often consists of a mixture of postganglionic and preganglionic injury, in which the upper nerves (C5 and C6) are more likely to exhibit a postganglionic injury while the lower nerves (C7, C8, and T1) are more likely to be avulsed from the spinal cord, as has been shown in experimental settings and in imaging studies [12, 13]. The mixed character of the injury may complicate the interpretation of EDX findings. Due to these pathophysiologic mechanisms, the utility of electrodiagnostic continuity for prognostication is uncertain in children with BPBI.

The value and difficulties of different EDX modalities will be discussed for needle electromyography (EMG), nerve conduction studies, and intraoperative neurophysiology.

Electromyography

One specific feature of needle EMG is that spontaneous muscle activity (denervation) is not as often found in BPBI as in adults. In view of the short length and smaller diameter of the axon, it is logical to expect signs of denervation earlier than in adults, and after restoration of some axonal continuity, denervation will disappear more quickly than in adults [14]. In fact, we found denervation activity starting after as few as 5 days, which contrasts with many textbooks indicating that denervation activity starts from 10 to 14 days after axonal discontinuity in adults. At the age of 1 month, two-thirds of children showed denervation in the deltoid muscle, which lasted through the age of 3 months in only 14% of children [15]. Other authors found that all denervation activity had disappeared after week 19 [16].

A traditional timing to decide whether surgical repair of BPBI is necessary was defined by Alain Gilbert as absent biceps muscle recovery at the age of 3 months [17]. EMG performed at that age usually shows a discrepancy: in the majority of patients, motor unit potentials are seen in clinically paralyzed muscles [18]. This can be explained in five ways: an overly pessimistic clinical examination; overestimation of EMG recruitment due to small muscle fibers; persistent fetal innervation; developmental apraxia; and misdirection, in which axons reach inappropriate muscles [14]. If the presence of motor unit potentials is interpreted as the start of recovery, and a nonsurgical treatment strategy is chosen, the eventual clinical recovery is often disappointing.

We prospectively studied a consecutive group of 48 infants with BPBI at the age of 1 week, 1 month, and 3 months and gathered clinical data

and EMG at these 3 time points [19]. As endpoints in this study, we defined a dichotomous outcome as either a severe lesion, i.e., the surgical findings of neurotmesis of root avulsion, or favorable neurological recovery after 2 years without nerve reconstruction. We could make a useful prediction of outcome already at the age of 1 month based on clinical recovery of elbow flexion and extension, aided with needle EMG of the biceps. This signifies that complete axonal discontinuity to the biceps muscle at the age of 1 month is a good predictor of eventual outcome. EMG of the biceps lost its predictive value at 3 months of age. In clinical practice this simple test enables early prognostication, which is of great value in early counseling the parents already at 1 month of age to express an optimistic or pessimistic expectation of outcome (Fig. 46.1). In our institution, neurological evaluation at 3–4 months remains the main indicator at which the fundamental decision to perform nerve surgery or not is taken.

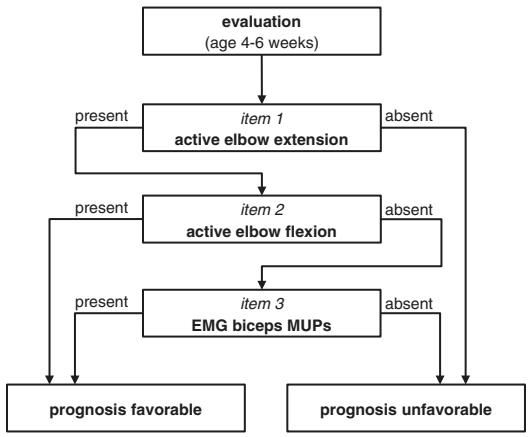

Fig. 46.1 Our proposed algorithm to evaluate the prognosis of a BPBI at the age of 4–6 weeks. An absent elbow extension against gravity reflects a lesion with involvement of the lower roots, so on clinical grounds it concerns a severe lesion with unfavorable prognosis. When both active elbow extension flexion and elbow extension demonstrate movement against gravity, the prognosis is favorable. When elbow flexion is not strong enough for movement gravity (with the child in supine position and the arm brought in 90 degrees abduction), an EMG is performed. The absence or presence of MUPs determines prognosis

Nerve Conduction Studies

One of the earlier papers that promoted the use of nerve action potentials (NAPs) dates from 1996 [20]. NAPs were recorded using surface (skin) electrodes from the median and the ulnar nerves in both forearms, with percutaneous stimulation at the wrist and recording at the elbow. A favorable nerve lesion was defined as a normal NAP amplitude (i.e., >50% of the uninjured side). This diagnostic algorithm was employed in a cohort of 73 children [3]. Based on a combination of NAP results and EMG findings, the authors predicted outcome correctly for C6 and C7 in 92% and 96% of the cases, respectively. The inability to record nerve action potentials for C5 led to a predictive value in a smaller proportion of cases (78%).

The use of sensory NAPs has been advocated as a diagnostic method for detecting root avulsions. The presence of a normal sensory NAP or an amplitude greater than 50% of normal compared to the uninvolved site together with EMG abnormalities in the corresponding muscles is accepted as a main criterion for avulsion. One patient series that promotes the use of NAPs examined 13 infants and claims that electromyography can be of great value to identify patients with a poor prognosis [21]. In this study, however, all children with an upper-type lesion recovered well, and all children with a total lesion had a poor outcome, suggesting that in this patient series, clinical outcome was determined by the extent of the lesion and the added value of NAP measurements to clinical evaluation was limited. Another study in 54 patients claimed an overall accuracy of the detection of preganglionic lesions of 74% for EDX (a combination of sensory NAPs and EMG) for all nerve roots [22]. The presence of a preganglionic injury implies an indication for surgery, as spontaneous recovery does not occur in these avulsion injuries. The actual diagnosis was made during surgery. The overall sensitivity of detecting preganglionic lesions by EDX was low (31%), but the specificity was high (90%). The authors compared EDX to imaging, where they found an overall sensitivity of detect-

ing preganglionic lesions of 66%, and an overall specificity was 70%. The low sensitivity to identify preganglionic lesions limits NAP use as a sole diagnostic entity, although the combination of NAP and needle EMG is of use to detect root avulsions.

Intraoperative Neurophysiology

Direct stimulation of nerves during surgery and evaluation of elicited muscle contractions and strength is widely performed, but no clinical studies have evaluated the effectiveness of this qualitative evaluation. Intraoperative NAP recording was introduced by Kline in 1969 [23]. It was employed in BPBI, specifically to decide whether a neuroma in continuity should be resected or that simple neurolysis was sufficient [24]. When an amplitude decrease of 50% or more across the neuroma was present, neuroma excision and grafting was performed. This threshold has never been validated, which was the reason for us to perform a study in 95 infants [25]. We measured NAPs and elicited compound motor action potentials (CMAP) during surgery and classified the severity of the nerve lesion irrespective of the EDX findings. Although axonotmesis, neurotmesis, and avulsion could be distinguished on group level, we were unable to identify valid cutoff points for the individual patient to facilitate the decision of whether to resect a neuroma-in-continuity and graft or leave the nerve-in-continuity and perform neurolysis.

Intraoperative EDX was also advocated by Chin et al. They used CMAP recordings after direct intraoperative stimulation, but instead of comparing the amplitude resulting from stimulation proximal and distal to the neuroma, they looked at the morphology of the CMAP to help in the decision-making process [26]. Roughly two-thirds of their surgically treated patients (22 of 32) underwent neurolysis only, which resulted in good recovery of abduction and elbow flexion but poor recovery of external rotation. In our opinion, external rotation is the hallmark function of recovery in BPBI patients, and therefore their algorithm was unable to predict poor shoulder recovery, which makes it less useful.

Another modality that has been used intraoperatively is the somatosensory evoked potential (SSEP) to detect root avulsions. The combination of an intact sensory nerve action potential with an absent cortical SSEP is indicative of a dorsal root avulsion lesion. However, data on the clinical utility of this technique is very limited [26].

SSEPs cannot be used to evaluate integrity of motor pathways. In adults, motor evoked potentials have been used to identify root avulsion injuries [27], but to our knowledge, this technique has not been used in BPBI patients.

Conclusion

Although EDX is often performed for the evaluation of lesion severity, the added value to a proper serial neurological evaluation is uncertain. Needle EMG at 3 months, the timing when a decision has to be taken to perform nerve reconstruction or not, is often too optimistic. Nerve conduction studies to detect root avulsions have a high specificity but low sensitivity. Moreover, as root avulsions primarily occur in the lower roots, the added value to neurological evaluation is questionable. Intraoperative neurophysiology was, in our hands, insufficient to aid in intraoperative decision-making. The main indication for EDX in our view is needle EMG at 4–6 weeks of age which expresses an optimistic or pessimistic prognosis for outcome, which is helpful for counseling parents at an early stage and enables timely referral to a specialized center.

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