Current Management Strategies in Neonatal Brachial Plexus Palsy

Rajesh Meena[1] Ramesh S. Doddamani[1] Dattaraj P. Sawarkar[1] Deepak Agrawal[1]

[1]Department of Neurosurgery and Gamma Knife Center, All India Institute of Medical Sciences, CN Centre, New Delhi, India

Address for correspondence Deepak Agrawal, MCh, Department of Neurosurgery and Gamma Knife Center, All India Institute of Medical Sciences, Room No. 720, CN Centre, New Delhi 110029, India (e-mail: drdeepak@gmail.com).

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Abstract

Neonatal brachial plexus injuries are a common type of birth injury. Although spontaneous recovery occurs in most of the cases, there is a large subset of cases in which recovery does not happen and primary or secondary surgical interventions are required. With the advancement of microsurgical techniques, the results of nerve surgery are encouraging. Good results are possible with timely surgical intervention even in severe cases. In this review article, in addition to the anatomy, etiology, and pathophysiology of neonatal brachial plexus injuries, the importance of timely surgical intervention and various primary and secondary surgical procedures are discussed in detail.

Keywords
- ► Erb's palsy
- ► neonatal brachial plexus palsy
- ► nerve repair
- ► preganglionic and postganglionic injury
- ► primary reconstruction

Introduction

Erb's palsy or Erb–Duchenne palsy is a paralytic condition of the upper limb, resulting from traction injury to the upper trunk of the brachial plexus.[1-6] It is most commonly seen in the neonates and the most common cause is shoulder dystocia (delivery of the anterior shoulder of the baby is hampered by mother's pubic symphysis). Therefore, when additional traction is applied to the baby's head, the angle between neck and shoulder is widened forcefully, resulting in overstretching and injury to the ipsilateral brachial plexus. The incidence of neonatal brachial plexus palsy (NBPP) is between 0.42 and 2.9 per 1,000 live births.[2,3] Predisposing factors for NBPP are shown in ►**Table 1**.

Erb's palsy is rarely seen in adults, often associated with road traffic accidents when there is trauma to the head and the ipsilateral shoulder resulting in undue stretching of the upper brachial plexus. Other rare causes in adults include direct violence to the brachial plexus, gunshot injuries, traction to the arm, or violent attempts to reduce shoulder joint dislocation.[7,8]

Clinical Presentation

NBPP lesions can be clinically categorized into five types, depending on the specific nerve elements involved as shown in ►**Table 2** and ►**Fig. 1**. Resulting position due to brachial plexus injury (BPI) along with the involved muscles is shown in ►**Table 3**. Fractures of the clavicle may lead to pseudoparalysis, easily mistaken for BPI. Other conditions that may be mistaken for BPI include osteomyelitis of the humerus or clavicle and septic arthritis of the shoulder joint.[7,8]

Pathophysiology

During delivery, traction forces may cause the nerves to stretch, rupture, or avulse from the spinal cord (►**Fig. 2**).

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Table 1 Predisposing factors for neonatal brachial plexus palsy (NBPP)

Neonatal
• Large birth weight
• Breech presentation
• Congenital anomalies
Maternal
• Age > 35 years
• Cephalopelvic disproportion
• Obesity
• Gestational diabetes mellitus (macrosomia)
• Previous child with NBPP
Labor-related factors
• Shoulder dystocia
• Increased duration of second stage of labor (>60 minutes)
• Operative vaginal deliveries
• –Vacuum extraction
• –Direct compression of fetal neck during delivery by forceps

Table 2 Classification of neonatal brachial plexus palsy (NBPP) presentations

Common terminology	Root/nerve deficits	Typical posture
Erb's palsy	C5–C6	Shoulder Adduction, Shoulder internal rotation, Elbow extension and forearm pronation (Erb's posture)
Extended Erb's palsy	C5–C7(or C5–C8)	Erb's posture with wrist flexion ("waiters tip")
Pan plexus without Horner syndrome	C5–T1	Flail arm
Pan plexus with Horner syndrome	C5–T1 and sympathetic chain	Flail arm with Horner syndrome
Klumpke	C8–T1	Paralyzed hand

Table 3 Deficits resulting from brachial plexus injury

Weakness of movement	Muscles involved with cord segment	Resulting position
Abduction of shoulder	Deltoid (C5)	Adducted
External rotation of shoulder	Supra- and infraspinatus (C5)	Internally rotated
Flexion of elbow	Biceps, brachioradialis (C5, C6)	Extended
Supination of forearm	Supinators (C5, C6)	Pronated
Extension of wrist	Extensors of wrist (C6, C7)	Flexed
Extension of fingers	Extensors of fingers (C6, C7)	Flexed
Descent of diaphragm		Elevated

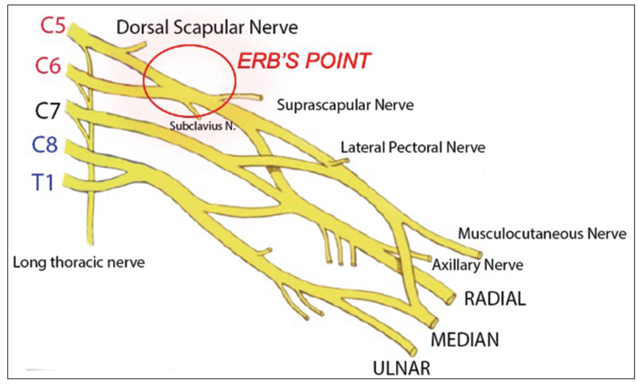

Fig. 1 Anatomical localization of Erb's palsy.

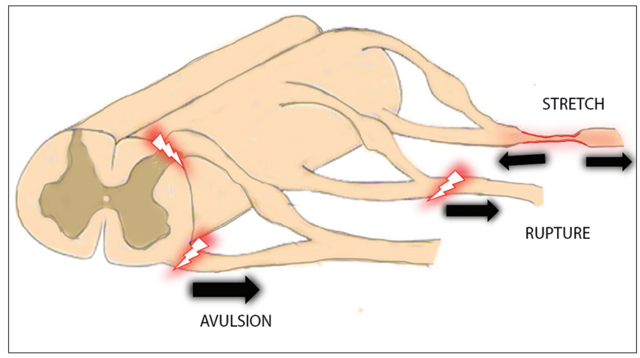

Fig. 2 Traction forces may cause the nerves to stretch, rupture, or avulse from the spinal cord.

Depending upon the anatomic disruption of axons and the surrounding endoneurium, perineurium, and epineurium, nerve injuries can be classified into following four types[9-11]

1. **Neuropraxic injury** It involves a disruption in the myelin sheath around the spared axon. It is the least severe form of injury, causing conduction block and allowing complete prompt recovery.
2. **Axonotmetic** It involves disruption of the myelin sheath as well as the axon, but preservation of the perineurium and epineurium. It is the intermediate form of nerve injury, allowing gradual recovery of the nerve function. The most important structure for the regeneration is the basal lamina tube surrounding the axons. In axonotmetic injury, usually the basal lamina tube is intact (thereby allowing axons to grow from the lesion site down into the basal lamina tube to the original end organ).
3. **Neurotmetic injury** It is the most severe type of peripheral nerve injury, in which there is complete rupture of the nerve involving the axon, myelin sheath, and connective tissues, including the perineurium and epineurium. In neurotmesis, there is rupture of the basal lamina tube and varying degree of perineurium and epineurium damage. Therefore, the outgrowth of the axons is blocked and most of them will not end up directly in an endoneurial tube. Rather they form an entangled mass of connective tissue and branching axons known as neuroma in continuity. Neuromas are classically seen in NBPP. However, some axons still manage to pass through the neuroma

and reach some tubes distal to the lesion site. Therefore, incomplete and aberrant axonal outgrowth will lead to end organ atrophy, distorted bony growth, joint incongruence, contracture formation, and possibly central apraxia. These changes become irreversible if there is no reinnervation.

4. R**oot avulsion** In root avulsion, there is complete discontinuity of the nerves between the spinal cord and the peripheral nervous system. Outgrowth of the axons, neuroma formation, or misrouting cannot take place.

Diagnosis

The mainstay of the diagnosis is the physical examination. The clinical assessment should include a thorough history (including gestational and birth history) and a careful and complete physical examination.[11-14] History and physical examination should be directed toward the following goals:

- Assessment of any associated nonneurologic injuries.
- Localization of the neurologic injury.
- Determining the severity of the injury.
- Delineating the risk factors for persistence.
- Monitoring for spontaneous recovery on sequential examination.

A variety of scales have been developed for the assessment of obstetrical BPI as shown in ►**Table 4**. These scales have shown to have moderate to near perfect interrater reliability for most movement tested and help in detecting changes on sequential examination. Few of them were subsequently incorporated into decision algorithms used for determining the surgical candidacy. The differentiation of preganglionic and postganglionic injuries is shown in ►**Table 5**.

Few bedside tests have also been developed for the assessment of motor function and decision-making, like the following:

- **Cookie test** When the infant is 9 months old, a cookie is placed in his or her hand and the elbow is then held against his or her side. The test is considered successful if the infant is able to get the cookie to his or her mouth without flexing the neck beyond 45 degrees.
- **Towel test** A towel is placed over the infant's eyes and the test is considered successful if he or she can take away the towel from the eyes with the involved limb versus the unaffected limb.

Table 4 Scales for obstetrical brachial plexus injury assessment

Mallet score
Active movement scale
Narakas grading system
Gilbert shoulder classification
Gilbert–Raimondi elbow classification
Raimondi hand and wrist classification
Medical Research Council scale
Gilbert and Tassin modified Medical Research Council scale

Table 5 Differentiation of preganglionic and postganglionic injuries

	Preganglionic BPI	Postganglionic BPI
Site	Avulsions proximal to dorsal root ganglion	Injury distal to sensory ganglion
Horner's syndrome	Present (disruption of sympathetic chain)	Absent
Winging of scapula	Present (loss of serratus anterior—long thoracic nerve and loss of rhomboids—dorsal scapular nerve)	Absent
Flail arm	Present	Absent
Sensory deficits	Present mostly with neuropathic pain	Present, limited to the involved segment and without neuropathic pain
Tinel sign	Absent	Present
Histamine test	Normal (C8–T1 sympathetic ganglion) Intact triple response	Abnormal histamine test (only redness and wheal but NO flare)
Radiological	Raised hemidiaphragm (phrenic nerve palsy) Cervical transverse process/first rib # MRI—(pseudomeningoceles rootlet abnormalities) A traumatic meningocele is a valuable sign of a preganglionic lesion, although it is not pathognomonic. Absence of roots is also an important sign in detecting a preganglionic lesion.	Edema and fibrosis of the brachial plexus can manifest as thickening of the plexus
Neurophysiology	NCV—preserved SNAP (but insensate)	NCV—if SNAP normal and patient insensate in ulnar nerve distribution, preganglionic injury to C8 and T1; if SNAP normal and patient insensate in median nerve distribution, preganglionic injury to C5 and C6
	EMG—may show loss of innervation to cervical paraspinals	EMG—shows maintained innervation to cervical paraspinals
Repair	Surgical repair/grafting	Neurotization
Prognosis	Little potential recovery of motor function (poor prognosis)	Capacity to regenerate (better prognosis)

Abbreviations: BPI, brachial plexus injury; EMG, electromyography; MRI, magnetic resonance imaging; NCV, nerve conduction velocity; SNAP, sensory nerve action potential.

In addition to motor testing and localization, other findings like presence of Horner syndrome (usually associated with a proximal injury to C8 and/or T1 and correspondingly predictive of a persistent deficit), asymmetrical chest expansion (may suggest injury to the phrenic nerve and hemidiaphragm paralysis), joint subluxation, or contractures should also be assessed clinically.[12,13,15]

Magnetic resonance Imaging (MRI) of cervical spine and brachial plexus may show evidence of nerve root avulsion such as pseudomeningocele or a pseudomeningocele with absent rootlets.[15,16]

The utility of electrodiagnostic tests is debated, especially in children as they are difficult to perform on infants. Also, there is often lack of concordance with clinical findings and variable interrater reliability. However, if electromyography denervation persists, there are little chances of spontaneous recovery. Absent fibrillations most likely points toward neuropraxia, while absent sensory conductions may be diagnostic of avulsion of nerve root from the spinal cord.[12-16]

Management

Although most of the infants with NBPP show spontaneous complete recovery, ~20 to 30% have incomplete spontaneous recovery.[17,18] Early recognition of persistent NBPP gives these infants the best chance of restoration of function. With early recognition and timely surgical intervention, the majority of the cases showed good functional outcomes in most of the retrospective studies. Advancements in microsurgical techniques, development of new techniques like nerve transfers, and understanding the secondary sequelae of NBPP have also widened the scope of safe and reliable outcomes in the majority of the patients with persistent NBPP. However, there is a limited time period during which the primary surgical intervention can provide benefit.[1,4-6] The decision to perform surgery is a balancing act—on one end is the need to allow sufficient time for demonstration of spontaneous recovery, while on the other end are data suggesting that outcomes are improved with early surgical intervention. Therefore, the timing of surgery is controversial.[1-6,11,12] Based on various recommendations, an algorithm has been shown in ►**Fig. 3**).

Salient features are as follows:

- Most of the authors agree that 80 to 96% of the patients with NBPP recover completely in the first year of life.[1-6]
- Most authors agree that infants with global/pan plexus palsies or flail and Horner syndrome should undergo surgical intervention at the earliest, preferably within 2 or 3 months after birth.[1-6,11-15]
- About 70 to 80% of the patients with NBPP have Erb's palsy or extended Erb's palsy. There is no consensus on the timing of surgical intervention in these cases. However, intervention after 9 months yielded poorer results, especially with regard to the hand function. Therefore, majority favored operative intervention, if there is no recovery of shoulder external rotation and/or elbow flexion/extension by 4 or 5 months of age.[1-6]
- Surgical interventions can be divided into categories: **Primary reconstruction** is the initial surgical management and may include nerve surgery/reconstruction (e.g., direct repair, neurolysis, nerve grafting, nerve transfers) and/or soft tissue procedures (e.g., functioning free muscle transfer, tendon transfers). **Secondary reconstruction** is considered when the desired functional outcome can be improved or refined further with surgical intervention or when there has been no further improvement/recovery after primary surgical intervention. This includes soft-tissue reconstruction (e.g., tendon/muscle transfer, FFMT, capsulotomies) and osseous procedures (e.g., arthrodesis, osteotomy).
- The rate of nerve regeneration is estimated to be around 1 mm/day or 1 in/month. Therefore, clinical results may not be seen for 1 to 2 years after the surgical intervention.[3-5]
- The shorter the distance to the target muscle, the lesser the time for reinnervation. While waiting for reinnervation to occur, regular physiotherapy should be done to prevent contractures.
- Grafting is not possible for **preganglionic injuries** as they are discontinuous from the spinal cord. **Postganglionic injuries** can be grafted as they remain in continuity with the spinal cord and the proximal stumps can be used as a viable nerve source for reconstruction of distal targets.
- Potential donor nerves include viable postganglionic roots of C5 or C6, ipsilateral C7, **intraplexal sources** (fascicle of the ulnar or median nerves, triceps nerve branches, medial pectoral nerve) and extraplexal nerves (spinal accessory nerve, intercostal nerves, phrenic nerve, and contralateral C7 nerve).[7-10]
- Surgical options for NBPP include exploration alone, nerve grafting, internal neurolysis, and/or neuroma excision, and more recently nerve transfers. Nerve grafting involves removal of the injured nerve segment and a conduit (usually an autogenous sural nerve graft) is placed between the proximal stump and the distal target organ. Nerve transfers involve sacrificing a nerve (to a muscle with redundant function) or a fascicle (with intraneural redundancy) and connecting it to a denervated target. Examples include median or ulnar fascicle to the biceps branch of musculocutaneous nerve. Nerve transfer can be performed for preganglionic injury or to accelerate recovery in postganglionic injuries by shortening the distance between the site of nerve repair and the motor end plate[11].
    - Most cases of upper brachial plexus injuries (C5–6) can be managed by supraclavicular exposure alone. However, some cases may require infraclavicular exposure or extension depending on the severity of NBPP on surgical exploration.
    - After exposure, electrophysiological studies are required to delineate the extent of injury and plan definitive surgical procedure.
    - Neuromas, if found, can be explored surgically using a microscope and microneurolysis can be attempted. However, most of the cases will require excision of neuromas followed by direct repair if feasible or sural nerve grafting.

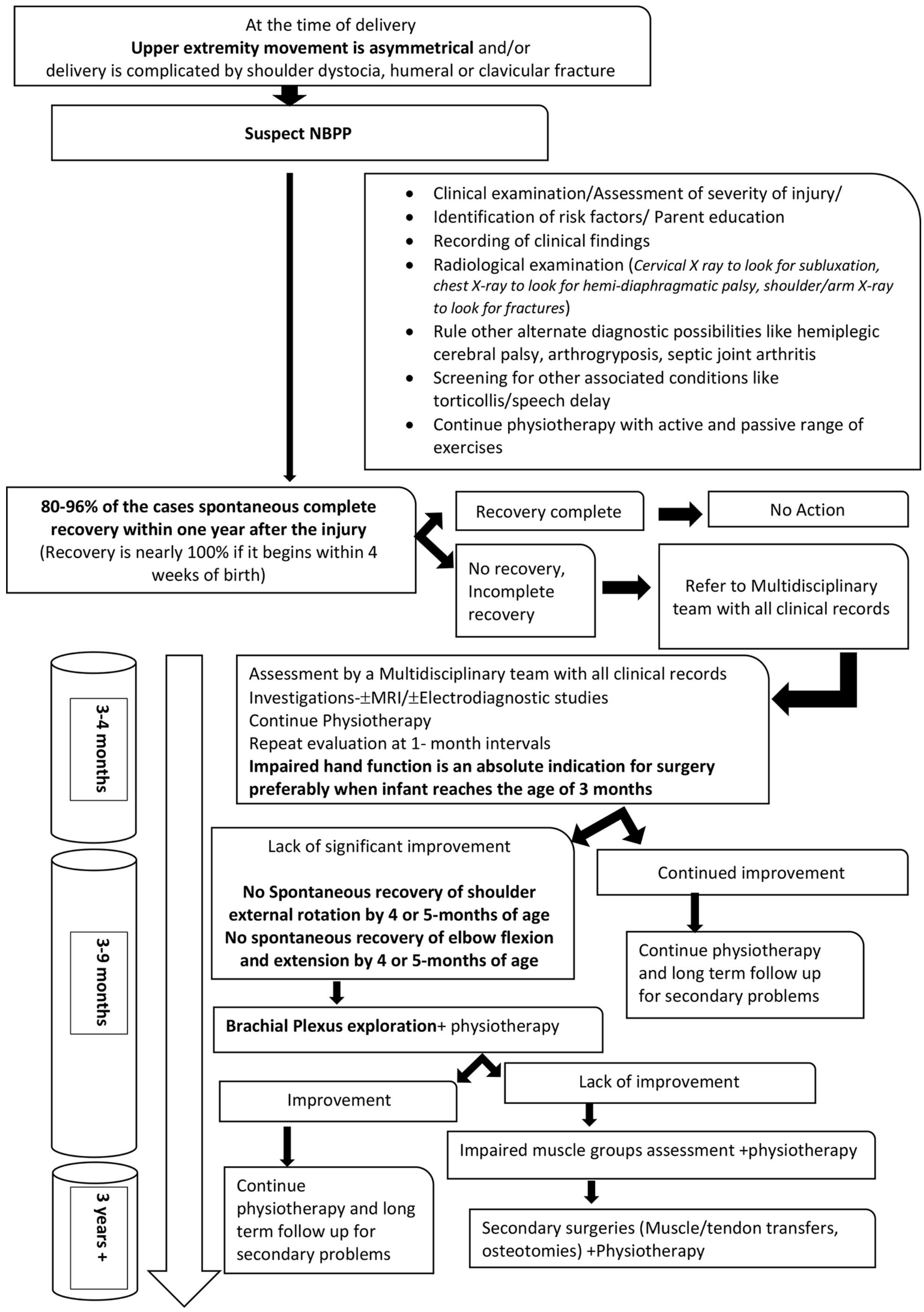

Fig. 3 Graphic on algorithm.

It is important to make a distinction between an avulsed nerve roots (disconnected from the spinal cord) and viable nerve roots, as the latter can be used proximal sites of nerve graft anastomosis.[6-11]

Two viable spinal nerves If two viable nerve roots are available (typically C5 and C6), these can be used to restore shoulder function and elbow flexion.

- C5 can be grafted (sural nerve cable graft) to the supra-scapular nerve and posterior division of the upper trunk (to axillary nerve) and C6 can be grafted to the anterior division of the upper trunk (to musculocutaneous nerve). This will restore motor function as well as help in restoration of sensibility.
- Alternatively, a hybrid of nerve grafting to shoulder and nerve transfer for elbow flexion (an ulnar fascicle transfer to the biceps motor branch ± median nerve to the brachialis nerve branch) can be done.
- One viable spinal nerve For upper trunk injuries with one viable nerve root, the viable spinal nerve can be grafted to the SSN and posterior division of the upper trunk, and the distal nerve transfer(s) described previously can be performed for elbow flexion.

- No viable spinal nerve Distal nerve transfers to restore shoulder external rotation, abduction and elbow flexion are the only option. For shoulder stability, abduction, and external rotation, a common strategy is to perform two nerve transfers—an SAN to spinal accessory nerve (SAN) transfer (via anterior approach or posterior approach) and a branch of the radial nerve to triceps to anterior division of the axillary nerve transfer. Elbow flexion can be restored by either a single or double nerve transfer as previously described.

Pan-plexus injuries It is utmost important to explore the brachial plexus thoroughly for the possibility of viable nerve donors as it can provide an additional source for reconstruction in pan-brachial plexus injuries. Surgical options are limited and directed toward restoration of shoulder stability and elbow flexion. Source of nerves for reanimation of the extremity is all extraplexal.[7-10]

- For restoration of shoulder function, the SAN to SSN transfer (for limited external rotation, stability, and abduction).
- For restoration of elbow flexion, extraplexal donor nerves to the musculocutaneous nerve (or biceps motor branch).

Postoperative Management

The infant's upper body is placed in a prefabricated cast, to limit movements of the neck and the affected arm, for a week or two. Gentle range of motion can be allowed after 2 to 3 weeks. Regular physiotherapy and follow-up in a rehabilitation center for a minimum period of 5 years is recommended for assessment of recovery and determination of potential secondary reconstructions to improve function.[11]

Complications

In a meta-analysis by Coroneos et al on primary nerve repair by obstetrical brachial plexus palsy, that included 512 patients, major adverse effects were reported in 1.5% and minor adverse effects were reported in 5% of the cases.[3,4] Major adverse events were persistent hemidiaphragm paralysis, disconnection of transferred nerves, and laryngeal edema/reintubation. Minor events were transient hemidiaphragm paralysis, wound infection, and co-contraction. No deaths were reported.[3,4]

Secondary reconstruction includes muscle or tendon transfers, tendon releases, arthroplasties, and osteotomies.

Secondary Procedures for Shoulder Restoration[4,7,11,17,18]

Classical Sever–L'Episcopo procedure This procedure involves transferring the latissimus dorsi (LD) and teres major tendon to the external aspect of humerus to augment external rotation of the shoulder. There are high chances of axillary nerve damage, so the procedure is abandoned.

Hoffer procedure This is a variation of the Sever–L'Episcopo technique, with better results both functionally and cosmetically. It involves transfer of the LD and teres major to the rotator cuff posteriorly through a transaxillary route.

Rotational osteotomy of the proximal humerus is done to improve external rotation.

Secondary Procedures for Elbow Restoration

Steindler flexorplasty This is the most commonly used secondary procedure. Flexor-pronator muscles arising from medial epicondyle are detached along with a bony cuff and then transposed to a more proximal point on the humerus bone and fixed with a screw. Various authors have reported a good postoperative outcome in elbow flexion in majority of the cases.

Transfer of free functioning gracilis muscle from thigh This procedure is indicated in patients in whom Steindler flexorplasty is contraindicated or for those who present very late for surgical intervention. It is a complex procedure with a nerve and vascular anastomosis, and the rate of success is low.

Secondary Procedures for Hand and Wrist Restoration

Both primary and secondary procedures for restoration of hand and wrist function are challenging in obstetrical brachial plexus palsy as results are palliative in most of the cases. It includes tendon transfers to restore absent wrist extension, muscle transfers (e.g., free functional gracilis muscle transfer to forearm extensors), joint release surgeries, and a variety of tenodesis.

Conclusion

NBPP should be evaluated properly and timely surgical intervention and regular follow-up is the key for the successful management.

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Conflict of Interest
None declared.

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