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Educational Corner

Brachial Plexus Birth Palsy

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Background

The brachial plexus innervates upper limbs and is formed by the ventral roots of the 5th-8th cervical spinal nerves (C5-C8) and the 1st thoracic spinal nerve (T1). All peripheral nerves of the upper limb originate from different parts of the brachial plexus (1). Figure 1 illustrates the anatomy of this area.

The brachial plexus may be injured during childbirth, a condition known as brachial plexus birth palsy (BPBP). Its prevalence varies in different geographical regions, ranging from approximately 0.4 to 2 cases per 1000 live births (2-6). The perinatal risk factors include high birth weight (macrosomia), gestational diabetes, multiparity, difficult/ prolonged delivery, shoulder dystocia, and breech presentation. Following these conditions, especially shoulder dystocia, the main cause of injury is cervical traction and brachial plexus stretching. Given that the most common fetal presentation is left occiput anterior (LOA), the right side is more affected (7, 8).

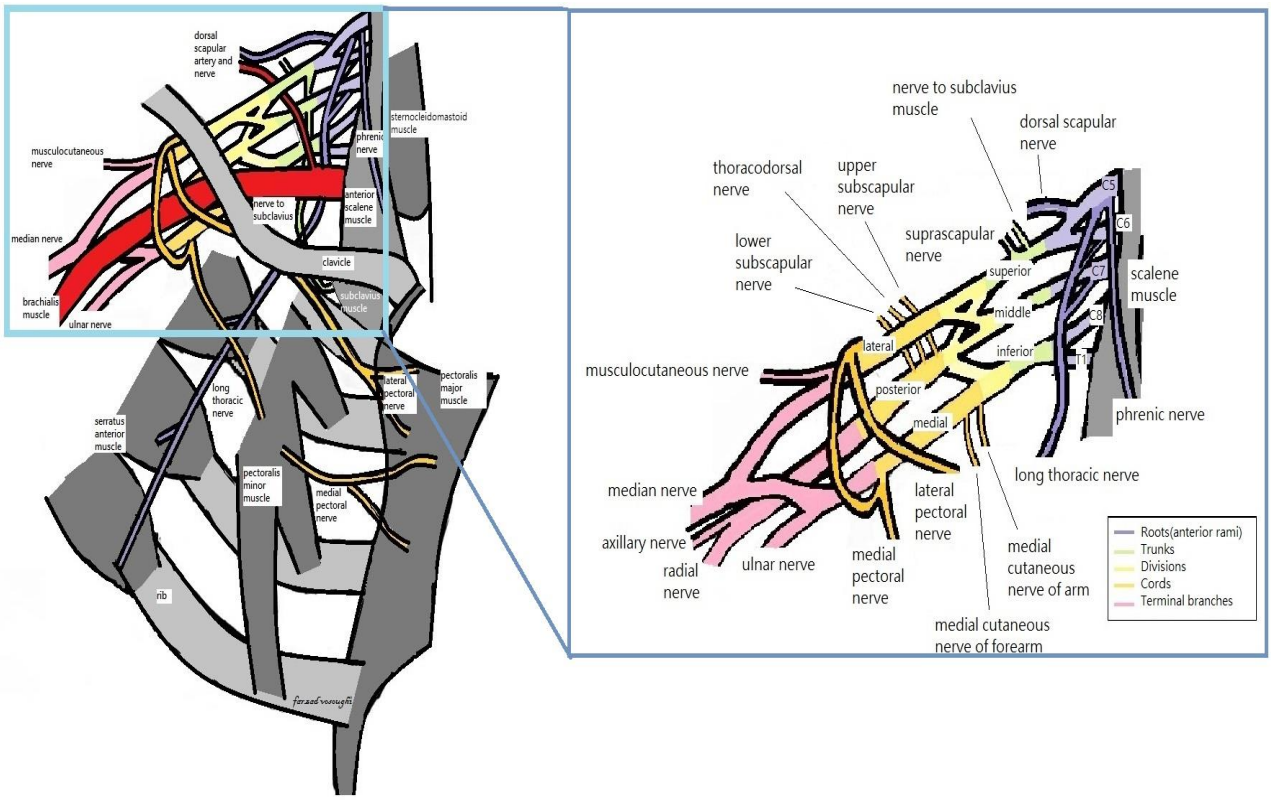

Figure 1. The brachial plexus originates posterior to the scalene and sternocleidomastoid muscle, passes posterior to the clavicle. Its terminal branches continue their way to the upper limb. The plexus is divided into roots, trunks, divisions, cords, and terminal branches. The cords are named based on their anatomic position relative to the brachial artery

Classification

The plexus injury may be preganglionic and in the form of avulsion, which is not repairable by surgery and has a weak possibility of spontaneous recovery. The other form, postganglionic injury, can appear as neuropraxia, axonotmesis, and neurotmesis (2) (Figure 2).

Another classification is based on the location of the involved roots. The most common site of BPBP is C5-C6 roots, also known as Erb's Palsy. An extended Erb's palsy involves the C5-C6-C7 roots. The involvement is mostly postganglionic in these two types. The most severe type engages the whole plexus from C5 to T1 (pan-plexopathy). The rarest type, Klumpke's palsy, happens in the C8-T1 roots and is usually preganglionic.

Diagnosis

The diagnosis of the BPBP is based on clinical examination. Noticing the patient's prenatal and perinatal histories and awareness of the possible risk factors helps greatly in diagnosis. Decreased and asymmetric neonatal upper limb reflexes (e.g. grasp reflex and Moro reflex) and the existence of Horner syndrome hint to the diagnosis of BPBP (9-11).

However, we should consider the differential diagnoses of pseudoparalysis, including shoulder girdle and arm fractures, and shoulder infections (5). Other possible differential diagnoses are central nervous system disorders, neuromuscular disorders, and congenital anomalies. Plain radiography can be helpful in detecting the shoulder and arm fractures and is an imperative part of the initial diagnostic evaluation.

During the examination, the following findings are suggestive of preganglionic avulsion injury: paraspinal muscles dysfunction (injury of the dorsal branches of the nerve roots), rhomboid muscles dysfunction (injury of the dorsal scapular nerve), the scapular winging (injury of the long thoracic nerve), hemidiaphragm paralysis (injury of the Phrenic nerve), and Horner syndrome (sympathetic nervous system injury).

Aggressive imaging assessment (e.g. myelography), along with other modalities such as computed tomography (CT) scan and magnetic resonance imaging (MRI), and a combination of them can help the diagnosis. The presence of pseudomeningocele in the imaging workup is consistent with the preganglionic lesions (2, 3 ,6). The use of electrodiagnostic studies in BPBP is unreliable and inexpedient (2, 4).

Finally, based on clinical examinations, BPBP can be classified into four general categories:

1. Upper roots (C5-C6) palsy (classic Erb-Duchenne paralysis, Erb's palsy): it is the most common type and is associated with limitations in the abduction and external rotation of the shoulder, flexion of the elbow, and supination of the forearm. The wrist and finger movements are in the normal range. The chance of spontaneous recovery in this group is very high.
2. Upper roots along with C7 root palsy (Extended Erb's palsy): it is associated with limitations of wrist and fingers extension in addition to Erb-Duchenne paralysis parameters (waiter's tip position). It has a worse prognosis than the first group.
3. Palsy of the entire brachial plexus (Pan-brachial plexopathy): the upper extremity is completely flail without function.
4. Lower roots (C8-T1) palsy (Klumpke's paralysis): it is very rare and is characterized by the normal function of the shoulder and elbow, with restriction of wrist and hand movements.

The pan-brachial plexopathy and Klumpke's palsy may also be associated with Horner syndrome (miosis, ptosis, and anhidrosis).

Treatment

Many BPBP cases recover spontaneously within a few months, since the injury is usually incomplete (12, 13). Therefore, periodic physical examinations (at least once a month) using multiple scoring systems such as active movement scale (AMS) and modified Mallet scale are required to assess recovery.

Passive range of motion (ROM) exercises should be started immediately to prevent shoulder and elbow contracture. If the partial recovery is not achieved by 3 months of life, the complete transection of trunks and cords, root avulsion injuries, and whole plexus involvement (especially with Horner syndrome) are plausible (14-16).

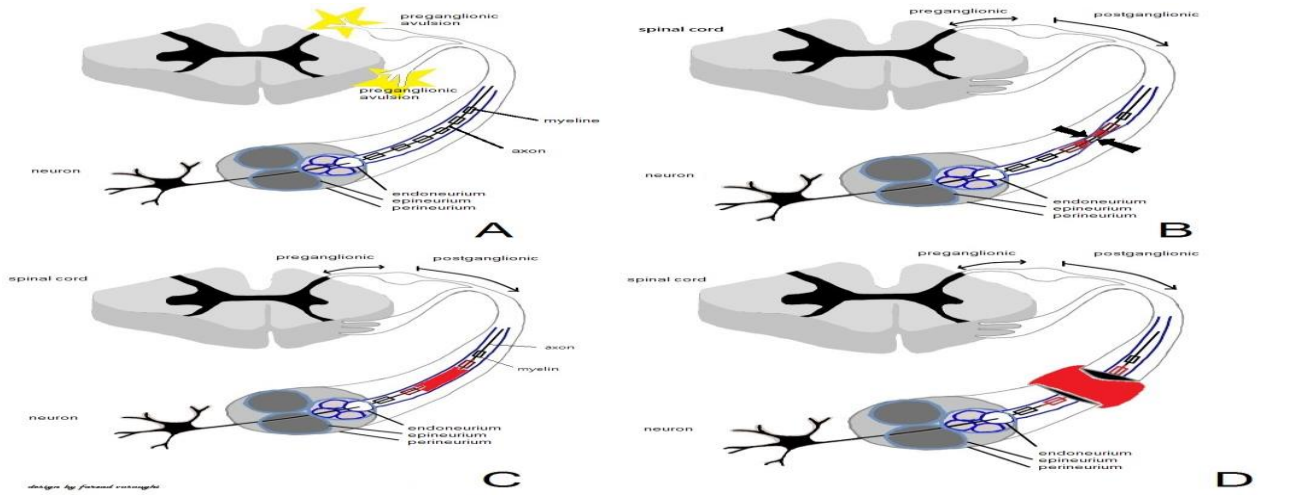

Figure 2. An injury to the spinal roots can be preganglionic (before the spinal ganglion) (A) or postganglionic. B) The neural injury can be caused by compression of the nerve without disruption of its anatomy (neuropraxis). C) The injury may disrupt the anatomy of some axons without damaging the endoneurium (axonothmesis). D) The most severe neural injury causes complete disruption of the neural axons, endoneurium, perineurium, and epineurium (neuronothmesis).

The use and timing of surgery are controversial. Many surgeons accept the failure of biceps recovery to act as an antigravity flexion three months after birth as an indication for microsurgical procedures (2-6, 17 ,18). The reason for assessment of the biceps function relates to the fact that it is the only muscle that is exclusively innervated by the C5-C6 roots.

In the case of partial recovery within 3 months of life, longer follow-up is advocated. Complete failure of biceps function recovery by the 6th month or the child's inability to place a cookie in the mouth without excessive neck flexion by the 9th month (positive cookie test), can be a sign of inappropriate recovery and requires surgical intervention (19) (Figure 3).

The surgical procedures include:

1. Neurolysis: it has no acceptable effects on its own and is performed along with other surgical procedures.
2. Neuroma resection and nerve repair: direct repair is not recommended because of failure to achieve tension-free coaptation, and the use of a nerve graft, mainly the Sural nerve, is mandatory.
3. Nerve transfer (neurotization): this method is widely used nowadays especially in neurotmesis and avulsion injuries. The commonly used nerve transfers are as follows (2, 4, 18-21) (Table 1).

A) Transfer of a branch of the spinal accessory nerve to the suprascapular nerve to improve external rotation and abduction of the shoulder.

B) Transfer of a branch from the radial nerve innervating the long head of the triceps to the motor branch of the axillary nerve to improve shoulder abduction.

C) Transfer of an intercostal nerve to the biceps motor branch of the musculocutaneous nerve to improve elbow flexion.

D) Transfer of a fascicle of the ulnar nerve and/or a fascicle of the median nerve to biceps and brachialis motor branch of musculocutaneous nerve to improve elbow flexion and forearm supination (Oberlin's procedure). This transfer can be applied in upper plexus palsy with intact lower roots.

E) In the case of paucity of donor nerve especially in the whole plexus palsy, some surgeons use the phrenic nerve as a source for transfer.

Secondary Musculoskeletal Problems

If the shoulder, elbow, and hand fail to recover properly, the permanent contractures and subsequent dysfunction can reduce the quality of life (QOL).

Shoulder

Restriction in external rotation and abduction of the shoulder following subscapularis muscle contracture is the most common functional impairment of chronic BPBP injury. This condition can lead to glenoid changes such as retroversion and subluxation of the humeral head. MRI can be helpful to assess glenohumeral deformities. In the early stages of this situation, in which the morphological changes are not predominant and the joint is reduced but subluxable, physical therapy should be performed to improve the ROM. If the subluxation is reducible, the mainstay of treatment is the injection of botulinum toxin in the subscapularis muscle and daily passive external rotation exercises by occupational therapist and parents.

The follow-up visits should be performed frequently to assess the shoulder ROM and plan the necessary interventions at the proper time. In more advanced cases, surgical procedures are performed. Soft tissue surgeries include anterior shoulder release, subscapularis muscle release, and tendon transfer to enhance abduction and external rotation of the shoulder using latissimus dorsi and trapezius muscle.

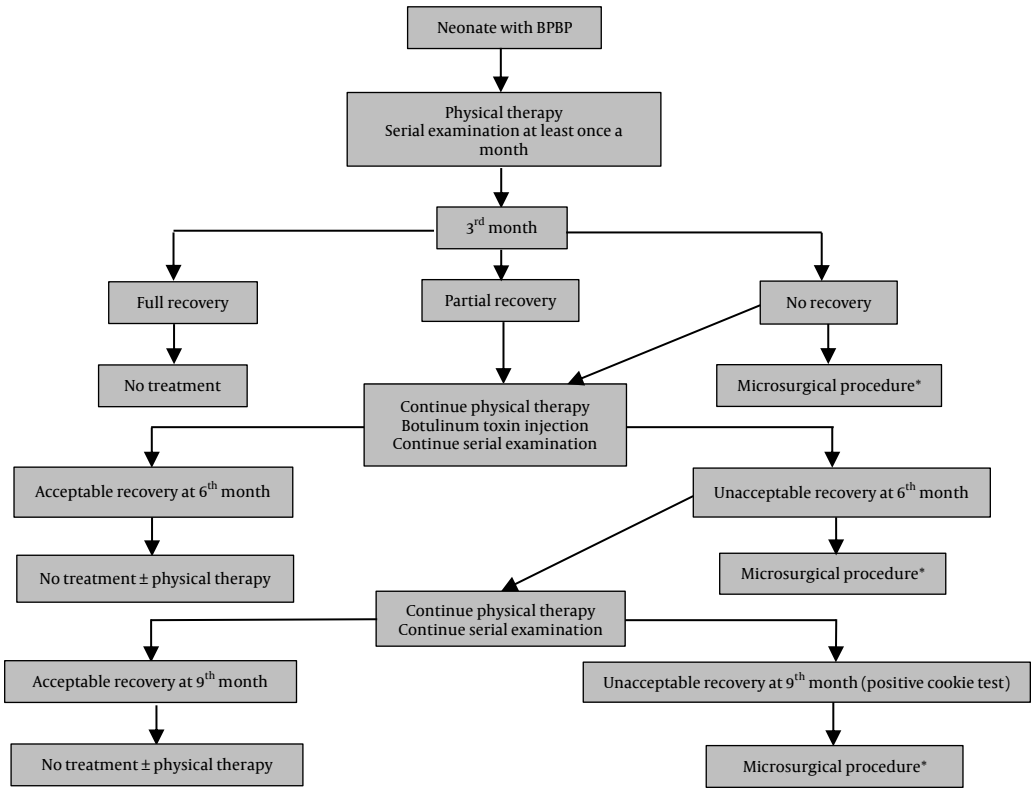

Figure 3. Suggested approach to a patient with brachial plexus birth palsy (BPBP).
*The timing of microsurgical procedure intervention is controversial and depends on the surgeon's preference.

Table 1. Common nerve transfers in brachial plexus birth palsy (BPBP)	
Type of palsy	**Suggested nerve transfer**
Erb-Duchenne palsy	Intercostals to the biceps/brachialis motor branch or Oberlin's procedure Spinal accessory nerve to suprascapular nerve Radial nerve (branch of long head of triceps) to the axillary motor branch
Extended Erb's palsy	Oberlin's procedure Spinal accessory nerve to suprascapular nerve Intercostals to the axillary motor branch Medial pectoral or intercostals to the radial nerve
Pan-brachial plexopathy	Intercostals or phrenic nerve to musculocutaneous nerve Spinal accessory nerve to suprascapular nerve Intercostals to radial nerve
Klumpke's palsy	Nerve to brachialis to ulnar/median nerve Nerve to brachialis/supinator to posterior interosseous nerve

Bony procedures include glenoid osteotomy to correct retroversion and derotation osteotomy of the humerus (2, 3, 5, 20, 21).

Elbow and Forearm

If elbow flexion is not achieved, tendon transfer (latissimus dorsi, pectoralis major, and triceps), Steindler flexorplasty, or free gracilis flap can be used (2, 3).

The forearm supination contracture is due to functional dominance of forearm supinators over pronators. In the early stages, there is no radial head anterior subluxation and it is passively correctable. Therefore, biceps rerouting (Zancolli procedure), which makes biceps muscle a forearm pronator, is one of the therapeutic options. Radius osteotomy, alone or along with ulnar osteotomy, can be considered in severe contractures or when there is profound palsy with no functional muscle available, to place hand in a more functional and/or cosmetic position.

Wrist and Hand

To balance residual wrist and finger movement limitations, tendon transfer, free gracilis transfer, and arthrodesis can be helpful (2, 3).

In the end, although obstetric care is improved nowadays, BPBP is still a challenging problem. Despite recent advances in surgical treatment and achieving good results, it is not possible for many patients to attain complete function. This should be addressed during parental consultation before initiating treatment.

Conflict of Interest

The authors declare no conflict of interest in this study.

Acknowledgments

None.

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