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Current concepts: Neonatal brachial plexus pals

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Abstract

Neonatal brachial plexus palsy continues to occur despite improvements in obstetric care. Recent research and publications have focused on early and accurate diagnosis and a more precise prediction of outcome based upon timing of muscle recovery. This information enables the physician to discuss realistic expectations for the infant with the family dependent upon the pattern of neonatal brachial plexus palsy. Reliable outcome measurements allow a better assessment of the natural history and the effect of various interventions. The precise surgical treatment algorithm to provide the best functional outcome is still evolving.

Introduction

Neonatal brachial plexus palsy may be decreasing in incidence; however, there are conflicting reports.¹ Regardless, neonatal brachial plexus palsy still has an incidence of 1-2 per 1,000 live births making this a frequent occurrence.^{2,3} The majority of infants with brachial plexus palsy spontaneously recover in the first 2 months of life and subsequently progress to near complete recovery of motion and strength.^{4,5} However, those infants who do not have substantial recovery by 3 months of age will have permanent limited range of motion, less strength, and a decrease in size and girth of the involved extremity. Currently, there continues to be debate about the timing and type of surgical intervention. The purpose of this article is to provide an update based on recent

literature regarding the anatomy, epidemiology, diagnosis, classification schemes, and treatment options for neonatal brachial plexus palsy.

Anatomy

The brachial plexus is formed by the ventral rami of the C5-T1 nerve roots and provides the basis for all sensibility and function of the upper extremity. This normal anatomic root pattern occurs in about 3/4s of the population.⁶ Prefixed cords receive an additional contribution from C4, whereas postfixed cords receive an additional contribution from T2. These have been documented to occur in 22 and 1 percent of the population, respectively.⁶

The brachial plexus is subdivided into roots originating from their respective spinal level, trunks where the roots combine, divisions where the trunks divide into anterior and posterior parts, cords that represent combinations of the divisions, and lastly, branches that proceed into peripheral nerves. In addition, various peripheral nerves branch off of various portions of the plexus.

The ventral rami of C5 and C6 combine to form the upper trunk, whereas the ventral rami of C8 and T1 combine to form the lower trunk. The middle trunk is a continuation of the ventral ramus of C7. Subsequently, each trunk will divide into anterior and posterior divisions. All three posterior divisions combine to form the posterior cord. The anterior divisions of the upper and middle trunk combine to form the lateral cord, whereas the anterior division of the lower trunk forms the medial cord.

The terminal branches continue to form the major nerves to the upper extremity. Specifically, the ulnar nerve arises from the medial cord, the radial and axillary nerves

arise from the posterior cord, the musculocutaneous nerve arises from the lateral cord, and the median nerve arises from a combination of the medial and lateral cords. In forming the median nerve, the lateral cord contribution is primarily afferent sensory fibers while the medial cord input is mainly efferent motor fibers.

Epidemiology & Etiology

Neonatal brachial plexus birth palsy occurs secondary to stretching of the trunks or avulsion of the roots. Risk factors include fetal macrosomia, instrumented delivery, prolonged labor, shoulder dystocia, multiparity, and gestational diabetes.^{7,8,9} Prior literature has suggested that breech delivery was a risk factor, however, a recent study by Sibinski and Synder found breech delivery was not associated with a higher incidence of nerve injuries.⁸ In addition, Sibinski and Synder found that a Caesarean incision reduced the risk of plexus palsy but did not eliminate it entirely.⁸ Fetal distress may be a contributing factor by contributing to relative hypotonia thus making the infant and plexus more susceptible to stretch during delivery.⁹

Recent literature has demonstrated that some infants have one or more risk factors while others have none.¹ Foad et al. demonstrated that 46% of children diagnosed with neonatal brachial plexus palsy had one or more known risk factors, whereas 54% had no known risk factors.¹ Additionally, they showed that shoulder dystocia had a 100 times greater risk, an exceptionally large baby (>4.5 kg) had a 14 times greater risk, and a forceps delivery had a 9 times greater risk.¹ Protective effects against the occurrence of neonatal brachial plexus palsy include having a twin or multiple birth mates and delivery by cesarean section.¹

The most common pattern of neonatal brachial plexus palsy (about 60%) involves the upper trunk (C5 and C6 nerve roots), and is known as an Erb's Palsy. Additionally, the C7 root may also be involved and this pattern is known as an extended Erb's Palsy (about 20-30%). Occasionally (approximately 15-20%), the entire plexus from C5 to T1 is injured and this pattern is known as a total or global brachial plexus palsy. An isolated lower trunk injury to C8 and T1 nerve roots is extremely rare and is known as a Klumpke palsy.¹⁰

Diagnosis

Diagnosis of neonatal brachial plexus palsy is usually made shortly after birth by lack of shoulder, elbow, forearm, wrist, and/ or finger motion. The diagnosis is supported by assessing the presence or absence of neonatal reflexes that induce elbow flexion and wrist and digit extension. These maneuvers include the Moro reflex and the asymmetric tonic neck reflex.⁹ The Moro reflex is elicited by introducing a sudden extension of the neck, which subsequently causes the shoulders to abduct and the elbows and digits to extend including a spreading of the fingers. This reflex usually disappears by 6 months of age.¹¹ The asymmetric tonic neck reflex is elicited by turning the head to the side which subsequently results in extension of the arm and leg on the side to which the head is turned. Flexion of the upper and lower extremities is seen on the contralateral side, creating a position like a fencer. Additionally, the physician can assess for the presence or absence of Horner's syndrome, (ptosis, miosis, and anhidrosis), which indicates lower root injury and a poor prognosis.^{10,12}

Alternative diagnoses include fracture of the clavicle or humerus (pseudopalsy), cervical spine injury and cerebral anoxia. These entities are assessed by careful physical examination for crepitus, deformity, and lower extremity involvement. Since shoulder dystocia is a risk factor for both brachial plexus palsy and cerebral anoxia, we routinely ask for the APGAR scores and assess for signs of spasticity.

Additional radiologic studies have been utilized to determine the location and extent of nerve injury and as to whether the injuries are avulsions (preganglionic injuries) or extraforaminal ruptures (postganglionic injuries). Kawai et al compared evaluation of intraoperative findings with myelography, CT myelography, and magnetic resonance imaging. Myelography was found to have a true positive rate of 84%, a false positive rate of 4%, and a false negative rate of 12%. CT myelography increased the true positive rate to 94% and demonstrated that the presence of small diverticula to diagnose avulsions was only 60% accurate. The presence of large diverticula or frank meningoceles was 100% diagnostic. Magnetic resonance imaging had a comparable true positive rate to CT myelography.¹³

Electrodiagnostic studies including nerve conduction velocities and electromyography have also been utilized in an attempt to better evaluate the severity of neural injury. However, these studies have not been able to add additional information to the clinical picture. Heise and colleagues performed electromyography in 41 infants, between 3 and 12 months of age, with severe obstetric brachial plexopathy. Their study demonstrated that needle EMG fails to estimate or overestimates clinical recovery in the proximal muscles of the arm and shoulder.¹⁴

Once the diagnosis of neonatal brachial plexus palsy is made, it is imperative to determine the level and severity of neural injury. This determination will aid in predicting the potential for spontaneous recovery as well as the overall outcome of the child. Michelow and Clarke demonstrated that the rate and extent of spontaneous recovery of elbow flexion, shoulder abduction, and extension of the wrist, fingers, and thumb in the first 3 to 6 months of life will help predict outcome.¹² Gilbert and Tassin have shown that a lack of normal biceps function by 3 months of age yielded an abnormal outcome at 2 years of age.¹⁵ However, the Michelow and Clarke article demonstrated that return of biceps at 3 months still yielded a 12% rate of failure in detecting poor outcome.¹² This error was reduced to 5% by combining return of elbow flexion with return of wrist extension, digit extension, thumb extension, and shoulder abduction.¹²

Diagnosis of a neonatal brachial plexus palsy also requires evaluation of the glenohumeral joint for dysplasia, subluxation, or dislocation. Lack of passive external rotation of the glenohumeral joint is the hallmark of underlying joint deformity.¹⁶ A recent study by Dahlin and colleagues found a 7.3% incidence of posterior shoulder subluxation/ dislocation in infants less than one year of age with a diagnosis of brachial plexus birth palsy.¹⁷ Magnetic resonance imaging (MRI) has traditionally been utilized to assess for glenohumeral dysplasia following obstetrical brachial plexus palsy. However, a recent article by Vathana and colleagues assessed the intraobserver and interobserver reliability of ultrasound measures to assess the position of the humeral head with respect to the scapula.¹⁸ They concluded that amongst radiologists, pediatric orthopaedic surgeons, and orthopaedic residents and fellows there was a high intraobserver and interobserver reliability for these techniques with regard to both normal

shoulders and humeral heads posterior to the axis of the scapula.¹⁸ Ultrasound has the added benefit of being a dynamic evaluation and avoiding sedation or anesthesia, which is necessary for MRI.

Classification Schemes and Outcome Measurements

A scoring system for surgical indications and subsequent outcome after nerve reconstruction has been proposed by Michelow et al and is termed the Toronto Test Score.¹² The scoring is based on recovery of shoulder abduction, elbow flexion, wrist extension, digit extension, and thumb extension. Each of these five functions is graded 0 to 2, where 0 is no function, 1 is partial function, and 2 is normal function. A combined score of less than 3.5 at 3 months of age or greater is an indication for microsurgery.¹²

The Hospital for Sick Children Active Movement Scale was developed to document upper extremity function during both treatment and recovery. Fifteen different upper extremity movements are tested first with gravity eliminated and then against gravity. A score of 0 to 7 is assigned based on the amount of motion that is able to be performed against gravity or with gravity eliminated. Assessing all fifteen movements provides information regarding assessment of the entire brachial plexus.¹⁹

The main outcome tool utilized to assess the shoulder after neonatal brachial plexus palsy is the modified Mallet system.¹⁵ This classification system has 5 categories to assess overall upper extremity limb function based on specific movements. These include global abduction, global external rotation, hand to neck, hand to mouth, and hand on spine. These categories are then graded on a scale from 0 to 5, with 0 being not testable and 5 being normal. Abzug and Kozin have recently proposed (submitted for

publication) the addition of a sixth category, hand to belly button. (Figure 1) This addition is graded the same as the others, but provides more relevant information regarding the child's ability to get to midline.²⁰

Bae et al. assessed the reliability of the Toronto Test Score, the Active Movement Scale and the modified Mallet system and determined that all three tests demonstrated positive intra- and interobserver reliability with aggregate scores. In addition, internal consistency (test-retest reliability) was excellent for the aggregate Toronto Test and the modified Mallet for all age groups tested.²¹

Nonsurgical Treatment

Once the child is diagnosed with a neonatal brachial plexus palsy without fracture, the initial treatment is passive range of motion of all joints. The newborn with birth palsy should have full passive motion. Limited passive motion is indicative of an underlying problem, such as joint subluxation or dislocation. Passive motion should be performed multiple times during the day and often requires the assistance and guidance of a therapist. The parents must be engaged in the therapy program to maintain supple joints. Particular attention should be paid to glenohumeral joint motion with scapulothoracic stabilization to prevent glenohumeral capsular tightness and subsequent deformity. (Figure 2) Additionally, tactile stimulation of the limb for sensory reeducation can be utilized.⁹

Surgical Treatment

Microsurgery

Microsurgical procedures for neonatal brachial plexus surgery continue to be a topic of debate with regard to indications and timing. Options include direct repair, neurolysis, nerve grafting, and nerve transfer. Direct repair is not possible since the stretching across the injured nerve results in an elongated area of damage and the formation of a large neuroma. Neurolysis has been shown by Clarke and others to have inferior outcomes when compared with resection and nerve grafting.²²⁻²⁶

Neuroma resection with nerve grafting is currently the gold standard treatment. Sural nerve grafts are harvested from the leg(s). Nerve grafting to the upper plexus have shown good return of shoulder function in 60-80% of patients with 80-100% having return of biceps function.²⁷⁻³⁰

Nerve transfers are gaining in popularity. Transfer of the spinal accessory nerve to the suprascapular nerve has been utilized as an adjunct to other microsurgical procedures for the treatment of neonatal brachial plexus palsy and is currently a viable option to restore shoulder motion. Suzuki et al. showed greater than two year follow-up in 12 patients who all had reinnervation of the supraspinatus and infraspinatus confirmed with electromyogram following spinal accessory nerve transfer to the suprascapular nerve in upper-type paralysis of the brachial plexus.³¹ Additional options for donor nerves to obtain shoulder motion include the radial nerve, intercostals nerves, the thoracodorsal nerve, the medial pectoral nerve, the long thoracic nerve, the phrenic nerve, the contralateral or ipsilateral C7 root, and the hypoglossal nerve.³²

Nerve transfers can also be performed to obtain elbow motion by directly transferring the nerve to the motor branches of the brachialis muscle and the biceps muscle, therefore increasing elbow flexion strength as well as the ability to supinate.³² Additional options include the thoracodorsal nerve, the hypoglossal nerve and the pectoral nerves, which can all be coapted to the musculocutaneous nerve.³² An assessment of available donors is mandatory and is determined by the type of brachial plexus palsy. Lesions that involve the upper trunk with or without middle trunk involvement allow for local nerve transfers including the ulnar or median nerves, which both have predominantly C8 and T1 root contributions to provide motor function. Global lesions mandate transfer of intercostals nerves since the local median and ulnar nerves are not available.³²

Tendon transfers

An internal rotation contracture often results after a residual upper or extended-upper trunk lesion secondary to the pull of the normally functioning adductors and internal rotators overpowering the weakened external rotators. (Figure 3) A persistent internal rotation contracture will lead to glenohumeral deformity over time.⁹ Surgical options include musculotendinous lengthenings, tendon transfers, and/or joint reduction. In a study from 2005, Waters and Bae evaluated the effects of an extra-articular procedure, specifically latissimus dorsi and teres major tendon transfers to the rotator cuff with or without concomitant musculotendinous lengthenings, to assess shoulder function and glenohumeral remodeling. Their conclusion was that these extra-articular procedures improved shoulder function but no profound glenohumeral remodeling occurred.³³

Similar clinical and imaging findings were reported by Kozin and colleagues.¹⁶ Subsequently, Waters and Bae demonstrated that tendon transfers combined with musculotendinous lengthenings and open reduction of the glenohumeral joint for mild to moderate glenohumeral dysplasia secondary to neonatal brachial plexus palsy will improve global shoulder function, and demonstrate remodeling of the glenohumeral joint.³⁴ The important determinant of glenohumeral remodeling appears to be the formal open reduction of the glenohumeral joint. An alternative to open reduction is arthroscopic reduction. A recent paper by Pearl et al. demonstrated that arthroscopic release of the glenohumeral joint capsule and subscapularis tendon can result in improvement of external rotation and humeral head alignment within the glenoid.³⁵ Kozin et al. have also demonstrated that internal rotation contractures in association with glenohumeral dysplasia can be treated with arthroscopic release with or without tendon transfers.³⁶ This study demonstrated improvement in both joint alignment and clinical evaluations.

Osteotomies

The most common osteotomy performed for children with residual brachial plexus palsy is derotational humeral osteotomy. This procedure is traditionally performed in those patients with severe glenohumeral deformity and places the arm in a better functional position.³⁷⁻⁴¹ Ruhmann et al. recently reported that this procedure will also allow the child to flex the elbow to the mouth without striking the lower arm against the thorax.⁴² Waters and Bae reported that derotational humeral osteotomies improve shoulder function in patients with brachial plexus birth palsy who possess shoulder internal rotation contractures and/or advanced glenohumeral joint deformity.⁴³ The

surgical approach has traditionally been a deltopectoral approach with the osteotomy performed just superior to the deltoid insertion.⁴³ Abzug and Kozin recently presented (submitted for publication) a technique that utilized a medial approach to the humerus to perform the derotational humeral osteotomy. The results demonstrated significant improvements in activities associated with external rotation with a low complication rate.

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A new technique coined the “triangle tilt” operation was recently proposed by Nath et al. This procedure involves surgical leveling of the distal acromioclavicular triangle combined with tightening of the posterior glenohumeral capsule.⁴⁴ Their results demonstrated improvement in external rotation and Mallet scores.⁴⁴ However, long-term follow-up is necessary before wide acceptance of this procedure.

Outcomes

Outcomes regarding the aforementioned treatments are dependent upon the type of brachial plexus palsy present and whether the nerve lesions occurred at the root level or distal to the root level. Infants with full recovery by two to three months of age go on to have no long term sequelae. However, those infants that continue to have deficiencies at three months of age will have permanent limited range of motion, less strength, and a decrease in size and girth of the involved extremity.

Smith et al. reported long term follow-up of 28 infants who did not undergo microsurgery despite the absence of biceps recovery by three months of age. After utilizing validated outcome measures, it was confirmed that worse neurological injury lead to worse long term shoulder function.⁴⁵

Microsurgery involving nerve grafting and transfer has demonstrated early promising results and continues to be the mainstay of current treatment. However, no long term outcome studies exist to date. Future studies will need to be performed to demonstrate prolonged functional benefit in these children.

Conclusion

In conclusion, neonatal brachial plexus palsy continues to occur despite improvements in obstetrical care. It remains a challenging and complex entity for both the family and treating physician. Currently, our treatment algorithm involves examination and determination of the level of injury as soon as possible. Once the diagnosis is established, the family is instructed by occupational therapists to perform daily passive range of motion exercises on the involved extremity. Children with global palsies are scheduled for microsurgical intervention at approximately three months of age. Those children with upper trunk lesions are observed for return of biceps function. If the biceps has not returned by five to six months of age, microsurgery is performed. Future research evaluating long term outcomes will provide further insight into which surgical interventions yield the most successful clinical result.

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Figure Legends

Figure 1. Drawing of the newly modified Mallet System demonstrating the addition of the sixth category, hand to belly button.

Figure 2. Clinical photograph demonstrating that attention should be paid to glenohumeral joint stretching while stabilizing the scapulothoracic articulation to prevent glenohumeral capsular tightness and subsequent deformity.

Figure 3. Clinical photograph depicting an internal rotation contracture of the left shoulder secondary to the pull of the normally functioning adductors and internal rotators overpowering the weakened external rotators.