Birth brachial plexus palsy (BBPP) injury occurs in approximately 1 to 2/1,000 live births. This number has not changed significantly over the years. In most infants, clinical symptoms eventually resolve. As reported in the literature, between 50 and 95% of affected infants will recover.

In this chapter, we review risk factors and describe the anatomy involved. We also discuss the clinical evaluation and describe treatment approaches. Plain radiography and electrodiagnosis, including nerve conduction studies and electromyography, may be useful in clinical decision-making. Early treatment and immediate education of the family are essential. For a child with a flaccid, anesthetic arm, surgery is typically performed when he or she is 3 or 4 months old. If the child is capable of some arm movement, surgery is usually done when he or she is 6 to 9 months of age. Surgical intervention with nerve grafting has been available for more than 100 years. A multitude of muscle and tendon procedures also exist. In addition, more traditional orthopedic procedures, such as arthrodesis and derotational osteotomies, may be undertaken.

History

BBPPs have been described in the literature since the 1700s. In 1768, W.A. Smellie first published a description of BBPP. Then, in 1872, Duchenne first reported a unilateral BBPP in an article on electrical stimulation. Shortly thereafter, in 1874, Erb described the upper brachial plexus point, where there is a lower threshold for electrical stimulation of the plexus. Madame Klumpke described an adult with C8-T1 BBPP with Horner syndrome in 1885. In the 1890s, primary brachial plexus explorations with nerve grafts began and proceeded through the 1920s. The first publication describing the approach, in this case involving a C5-6 rupture and repair, was in 1903 by Kennedy in the *British Medical Journal*. Unfortunately, technical deficiencies at the time led to significant mortality secondary to the positioning of the great vessels close to the brachial plexus; therefore, the surgical procedures were, for the most part, stopped after the 1920s. The 1970s brought dramatic microsurgical advancements. Shortly thereafter, Dr. Alain Gilbert began performing these procedures routinely on infants with BBPP in Paris, and the procedure’s popularity spread from there.

The incidence of BBPP is approximately 1 to 2/1,000 live births. BBPP is frequently associated with shoulder dystocia at the time of delivery. The next most common associations are multiparity and large infants.

The most common description of an etiologic mechanism of action is a stretch, most commonly lateral. This is anatomically logical, as the site of the brachial plexus is just lateral to the sternocleidomastoid and just superior to the clavicle, leading to potential stretch of the brachial plexus in shoulder dystocia. Some intrauterine mechanisms also have been recently proposed, but they have not had wide support. Some infants clearly have anatomic variations or anomalies, including vascular, tendinous, or bony, which either cause BBPP or increase the susceptibility to it.

In any consideration of nerve injury, classification is useful. In the Seddon classification, the mildest type of injury is neurapraxia, in which there is no anatomic change and there is reversible loss of electrical conduction. This will completely resolve. The most severe type of nerve injury is neurotmesis, in which there is complete physical disruption of the nerve. This is called an avulsion when it

This chapter describes birth brachial plexus palsy (BBPP), including its incidence, the mechanism presumed to underlie the condition, associated risk factors, and level of neurologic involvement. Early and late treatment options will be presented, both therapeutic and surgical.
is preganglionic or proximal to the dorsal root ganglion and, therefore, immediately adjacent to the spinal cord. It is called a rupture when it is postganglionic. This distinction has pertinent surgical indications, but both require surgical repair. Axonotmesis is the third type of injury and it is the most difficult for evaluation because of variable severity. With this, the axon is disrupted, but there is preserved endoneurium.

The brachial plexus anatomy involved includes roots, trunks, divisions, cords, and the peripheral nerves. The roots are actually the anterior primary rami of C5, C6, C7, C8, and T1. There are upper, middle and lower trunks, as well as dorsal and ventral divisions and posterior, medial, and lateral cords (Figure 84-1). An infant with BBPP will have variability in the level of the roots involved. Erb’s palsy or C5-C6 palsy is the most common, occurring in approximately 75% of infants (Figure 84-2). This is anatomically logical, considering the position of the brachial plexus itself and the position of the baby during delivery. Klumpke’s palsy is a C8-T1 palsy with frequent concurrent Horner syndrome. There are many who question whether Klumpke’s palsy actually exists in BBPP, except as an anatomic anomaly, because of the lack of a plausible mechanism to involve C8 and T1 without involving the more superior roots during birth.

If a child presents with Klumpke’s palsy, there are three possibilities to consider. First, and by far most likely, is that the entire plexus was initially involved in the injury. C4, C5, C6, and, sometimes, C7 are held in the transverse process spinal nerve gutter by connective tissue, which is protective positioning for them. C8 and T1 do not have this protection and, therefore, are more at risk for a severe nerve injury, given equal amounts of force to all roots. Therefore, it is quite likely that an infant who presents for evaluation with C8-T1 brachial plexus palsy with a thorough history will have had a complete plexus involvement initially, with resolution of the better-protected fibers prior to evaluation. The dorsal roots are also at relatively less risk than the ventral, secondary to them being more cohesive and staying together into the dorsal root ganglion, as opposed to the ventral roots, which have the ganglion within the spinal cord and, therefore, are separated prior to leaving the cord. Because of this, sensory fibers are relatively better protected than motor fibers. The second possibility is spinal cord injury. The third possibility is an anatomic variation that changes the forces applied to the roots, such as a blood vessel, tendon, or anomalous rib that selectively affects C8-T1. There may be other combinations of plexus involvement, due to variation of types of nerve injuries that may have occurred and the various levels that may be injured. In the most severe case, the complete plexus is involved (Figure 84-3).
Evaluation

Primary questions to be asked when taking the medical history include the parity of the mother, the infant’s birth weight, and the presence of shoulder dystocia. Additionally, significant questions are the size of previous infants and their ages, presence or absence of gestational diabetes, and the clinical course since the time of birth, with any improvements or changes seen.

The physical examination includes sensory evaluation for any dermatomal or nerve root distribution deficits. A sensory exam in an infant may help with the motor exam by stimulating the child to move. A motor exam in an infant may be assisted by generating interest in reaching for toys or treats and by positioning the infant. Testing the primitive reflexes, particularly the Moro response, will help in showing the infant’s active proximal physical movement. Tendon reflexes will be absent or decreased in all infants with BBPP. Range of motion must be evaluated in the arm due to the common presence of contractures, which may be seen in shoulder adduction, internal rotation and extension, and finger flexion. Later, wrist flexion and elbow flexion contractures are frequently seen. The elbow flexion contracture is even found, ironically, in infants and children who do not have active elbow flexion and do have active elbow extension. This appears to be due to our natural frequent positioning in flexion. The pronated position is frequently overlooked. The size of the involved arm may give clues to the specific deficits and is frequently decreased, with both atrophy in the muscle and smaller bony structure. The shoulder joint is very frequently underdeveloped compared with the other side, as is quite common in infants with many motor problems involving the shoulder or hip. Occasionally, temperature may be asymmetric in the arms, particularly if there is Horner’s syndrome. Torticollis is commonly seen, with the neck rotated away from the involved arm the vast majority of the time.

Radiographic Evaluation

Plain radiographic films are an important part of early evaluation to rule out fracture of the clavicle or humerus. There are very rare instances of a bone tumor or osteomyelitis clinically mimicking BBPP; however, the history and time course differ. Computed tomography myelograms or magnetic resonance imaging (MRI) is routinely done for older patients for brachial plexus palsy. This has been evaluated and there are significant false-positives and false-negatives in doing these studies in infants with the current technology. As the infants must be sedated for these studies and studies have a low degree of reliability, these are currently not a recommended part of the evaluation. I believe it is likely that as the technology improves MRI will become a very useful study.

Electrodiagnosis

Electrodiagnostic evaluation can give crucial information about an infant with BBPP by revealing clinically problematic areas. This evaluation is a combination of a nerve conduction study (NCS) and electromyography. The NCS consists of a motor NCS, in which roots or nerves are stimulated for evaluation of subclinical electrical activity of muscle for indication of electrical continuity and potential for clinical recovery. This is useful because the electrical activity returns prior to having sufficient activity for clinical movement. Sensory NCS gives significant information if there is clinical sensory deficit. In this diagnosis, if sensation is intact in the arm, sensory NCS is not useful. If there is an area that is insensitive and the sensory NCS shows a normal response, then there is a preganglionic neurotmesis (avulsion). This is due to the continuity between the peripheral nerve and the dorsal root ganglion. Somatosensory evoked potentials (SEPs) are generally not performed as part of clinical evaluation both because of the overlap of innervation, making it less specific than ideal, as well as the fact that the child would need to be placed under general anesthesia for this study. Information on proximal
portions of the roots can instead be obtained from F waves and H reflexes during the NCS. The F wave is easily obtained and there is no significant difference in the comfort level for the patient. H reflexes require increased duration of stimulation and are, therefore, more uncomfortable and less well tolerated. They are more technically challenging to obtain as well.

Preparation is essential so that both the parents and the infant can tolerate the study with minimal discomfort. I gather all equipment needed first so that everything will be within reach, including toys for distraction. I tell the parents that the child will get a mild electrical shock, similar to that experienced after rubbing one's feet on carpet and touching someone, and that we will then be able to measure the electrical activity of the nerves and muscles. Some children describe this stimulus as a ticklish feeling, whereas others do find it uncomfortable. Next, I show them both a sensory level and motor level response on my wrist and then encourage the parents to feel a sensory level stimulation, as well.

I then initiate the study with the child as comfortable as possible, very commonly with the infant on the parent's lap. I perform the sensory nerve conduction study first, then the motor nerve conduction study. I then tell the parents that I will not be putting any more electrical input into the child but will use a wire electrode to look at the child's muscle electrical activity. I tell them that I will be putting the electrode through the skin into the muscle and that, very likely, the child will find this uncomfortable. If this is the case, however, and they fight the study, it gives the optimal electrical view of the functioning motor units. We discuss that there will be no after-effects of the study. I then perform the electromyography using a wire electrode in muscles that have motor deficit. Both NCS and electromyography are done in an individualized manner with only selective studies performed. Therefore, because approximately 75% of infants will have clinical involvement of the upper trunk, it is quite likely that the hand will be excellent and we will do no median or ulnar studies. Erb's point stimulation is commonly performed in the NCS of this group of children.

The timing in electromyography for positive sharp waves and fibrillations has been reported as earlier in infants than in adults. The presence of motor units can be searched for in muscles that do not show clinical activity but do not have electrical activity. This may help guide possibilities expected for clinical recovery period.

**Early Treatment**

The treatment approach is presented to the family after the child has been examined. We first discuss the anatomy of the brachial plexus and pertinent surrounding structures. I present the related issue of the risk of repeat birth brachial plexus palsy due to size issues and recommend considering future scheduled cesarean section prior to going into labor. I stress the importance of a passive range of motion program that keeps the baby’s joints in excellent position as well as the importance of maintaining awareness of the baby’s involved arm. I recommend use of a wrist rattle on the involved arm as well as to have the family move the involved arm and hand up into a child’s field of vision and replicate movements the child does on the uninvolved arm. The parents are then given a prescription for occupational therapy (OT) or physical therapy (PT), depending on who is available in their area and their insurance coverage. The therapist then works on positioning the child immediately. The range of motion program is started at 2 weeks of age. Orthoses are frequently required, particularly for wrist extension early on and later for supination. Precautions are given to avoid overly strenuous shoulder abduction and external rotation or forearm supination to prevent shoulder dislocation and subluxation or dislocation of the radial head at the elbow. Seeing the therapist regularly and doing a home program several times a day to work on range of motion increase awareness of the arm and increase active use of the arm, which are crucial to gaining functional use of the arm.

**Surgical Decisions**

In most infants, BBPP spontaneously resolves, but findings reported in the literature vary dramatically. Those who report that up to 95% of their patients recovered counted any trace movement as recovery. Those who report that 50% or less recover demand perfect symmetry of the arm. There is some geographic variation in surgical timing. In France, an infant who does not have active elbow flexion will have nerve grafts at 3 months of age. In North America, if the child has an insensate, flaccid arm, recommendations are for surgery by 3 to 4 months of age. A child with an incomplete brachial plexus injury will be recommended for surgery at 6 to 9 months of age.

There are many reasons for attempting surgical intervention early. The primary one pertains to child development: a desire to have the child learning all bimanual activities with two hands from the beginning. The contractures also may progress over time, so that early intervention to minimize this is an aim. Quite frequently, growth is asymmetrical, with a decrease in arm length and atrophy. Lack of awareness may be a significant problem for these children, as they may grow up paying attention to one side and not having the other arm in the field of vision, leading to functional loss with daily activities. Shoulder asymmetry is extremely common, as is scapular winging. Pain is not usually an issue as it is in older patients. There are common reports of tenderness about the shoulder for approximately the first 2 weeks, particularly when changing clothes or bathing.
Surgery

Surgical intervention was begun on nerve roots on individuals with BBPP in the 1890s. The proximity of the great vessels made this technically quite difficult at the time. Advances in microsurgical techniques in the 1970s made this a relatively common procedure, and technical improvements continue to this day. Electrical stimulations are generally done on the nerves to determine electrical continuity. Nerve grafts are performed, most commonly using the sural nerve to join together proximal functioning, electrically connected fibers with distal nerve fibers. Neurolysis is regularly done to remove scar tissue that lacks functioning nerves. Nerve grafts are then done, ideally with direct end-to-end anastomosis. This is not generally possible, so donor nerves, such as the sural, are used. Combinations of these procedures are done in most cases. Nerve growth will follow in the months after surgical intervention. If no functioning nerve roots are found, C7 may be partially taken from the contralateral side and tunneled under the skin, with ulnar nerve or sural nerve graft. This approach, of course, leads to a longer time for nerve growth. Intercostal nerves are sometimes used, but because they are smaller, it is more difficult to have sufficient nerve fascicles for grafting.

The infants will then continue with therapies and a home exercise program while health practitioners monitor nerve and muscle recovery. Secondary procedures include the ulnar or median nerve transfer to musculocutaneous nerve.

If there are continued deficits, muscle transfers may be performed. The latissimus dorsi may be transferred for shoulder abduction. Muscle and tendon transfers may be done for wrist and finger flexion extension or thumb positioning. Another possibility is a muscle transfer of the free gracilis to the forearm with nerve grafts. A pronator transfer to change to supinator function is also a useful procedure. Advances continue to be made in this area with fine-tuning of muscle, tendon, and nerve transfers. Bony procedures about the shoulder are also commonly performed.

Summary

Birth brachial plexus injury occurs in 1 to 2/1,000 live births, though approximately 75 to 80% of these recover spontaneously. It is important for therapeutic intervention to be undertaken early with education for the parents, including consideration of cesarean section for any future deliveries. A home exercise program is vital. Many surgical options are available if a child does not show recovery by 6 months of age when they show some motor activity, or at 3 to 4 months of age when they have a flaccid, anesthetic arm.

Suggested Readings


