Child Neurology: Brachial plexus birth injury
What every neurologist needs to know

ABSTRACT
While most often transient, brachial plexus birth injury can cause permanent neurologic injury. The major risk factors for brachial plexus birth injury are fetal macrosomia and shoulder dystocia. The degree of injury to the brachial plexus should be determined in the neonatal nursery, as those infants with the most severe injury—root avulsion—should be referred early for surgical evaluation so that microsurgical repair of the plexus can occur by 3 months of life. Microsurgical repair options include nerve grafts and nerve transfers. All children with brachial plexus birth injury require ongoing physical and occupational therapy and close follow-up to monitor progress. Neurology® 2011;77:695–697

CASE PART 1
A term male infant was delivered to a gravida 3 parity 3 mother after an uncomplicated pregnancy. Labor was uneventful; however, delivery was complicated by shoulder dystocia. An episiotomy was performed and the infant’s posterior shoulder (left) was grasped and delivered, followed by the anterior shoulder (right). The infant weighed 4,750 g, >97th percentile for age. In the delivery room he was noted to have a left upper extremity palsy, with an asymmetric Moro reflex.

Differential diagnosis. Brachial plexus injury is the most common etiology of a plegic arm in the neonatal period. Other considerations include a clavicular or humeral fracture, with pain limiting limb movement. Fractures can be diagnosed by feeling for “step-offs,” crepitus, or pain along the bone and obtaining plain films. Central causes, such as a focal cortical dysplasia selectively affecting the arm area of motor cortex, are rare. Poland syndrome, the absence or hypoplasia of the pectoralis muscles, can cause monomelic arm weakness; however, the structural abnormality is visibly apparent. A perinatal stroke typically does not cause hemiparesis in the neonatal period, but rather later in infancy.

CASE PART 2
On examination, there were no clavicular or humeral step-offs or crepitus, and a chest x-ray was normal. The parents were counseled that the brachial plexus injury would fully resolve. In pediatric follow-up at 2 months, however, the infant held the arm adducted and internally rotated at the shoulder. His forearm was pronated, his elbow extended, and his wrist and fingers were flexed in the “waiter’s tip” posture, consistent with injury affecting the C5-C7 root levels. There was no Horner syndrome. He was referred for neurologic and surgical evaluation.

Epidemiology. Brachial plexus birth injury occurs in 0.4 to 4 per 1,000 live births.1 It is most commonly associated with shoulder dystocia, an impaction of the infant’s anterior shoulder behind the maternal symphysis pubis. Lateral traction on the head, as part of the corrective maneuvers to deliver the infant, stretches the brachial plexus, leading to injury 4%–40% of the time.2

The strongest fetal risk factor for shoulder dystocia is macrosomia—birth weight greater than 4,000 g.2 Maternal risk factors for brachial plexus birth injury include diabetes or gestational diabetes, obesity, or a history of shoulder dystocia during a previous birth. A prolonged second stage of labor (pushing) and operative vaginal delivery also increase the risk.1–3 However, half of the cases have no identifiable risk factor.2

While the risk factors for shoulder dystocia are well recognized, they have poor predictive value.3,4 C-section decreases, but does not eliminate, the risk of brachial plexus injury, and introduces additional maternal morbidity.1,2

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Neurology and prognosis. The ventral rami of the C5 through T1 spinal nerves form the roots of the brachial plexus. Children with brachial plexus birth palsy have traditionally been classified clinically into 4 groups. The largest group (50% of cases) involves C5-C6 injury, classic Erb palsy, and generally has the best prognosis. The next group (25%) involves C5-C7 injury and has an intermediate prognosis. Children in these 2 groups hold the arm in adduction and internal rotation at the shoulder due to relative sparing of the shoulder adductor and internal rotation muscles. The imbalance of push-pull muscular forces across the glenohumeral joint at the shoulder causes the joint itself to develop abnormally, with increasing deformity as the child grows. Involvement of C7 is suggested by the presence of a wrist drop.

The third and fourth groups (together 25%) involve injury to the entire plexus. The arm is held in a neutral position with little to no movement. The fourth group is the most severely affected and can be distinguished by the presence of an ipsilateral Horner syndrome (miosis, ptosis, and anhidrosis) due to concurrent injury to the sympathetic chain as it exits the spinal cord. Isolated lower root injury (C8-T1), Klumpke palsy, is extremely rare.

Brachial plexus injuries can also be classified by the type of neuropathologic injury. The least severe is neuroapraxia, or stretch injury, causing conduction block, but no permanent structural damage to the nerve. Conduction block can last for hours to weeks, but ultimately fully recovers. Axonotmesis injury involves damage to axons, as well as supporting blood vessels and connective tissue, including perineurium and epineurium. If only the axons are disrupted, they regrow with full recovery. If the perineurium or epineurium are also disrupted, the likelihood of complete recovery decreases significantly. Neuromatosis injury indicates complete nerve rupture. Scar tissue forms between the proximal and distal ends of the nerve to become a neuroma. Recovery is limited because it is difficult for axons to regenerate through the neuroma. Root avulsion is the most severe injury, usually occurring at the nerve rootlets at or near the spinal cord. Avulsion injuries do not spontaneously recover so it is essential that these patients be identified for early intervention.

When examining the brachial plexus in a neonate, the emphasis should be on looking for signs of injury to proximal nerve structures as these are highly suggestive of avulsion. Given the proximity of the sympathetic chain to the spinal cord, the presence of Horner’s almost always implies a root avulsion injury. Additional signs of avulsion include winging of the scapula, indicating long thoracic nerve injury, and asymmetry in chest wall excursion, indicating phrenic nerve injury. In cases of complete plexus palsy, a chest x-ray should be performed to rule out hemidiaphragm paralysis.

Diagnostics. The diagnosis of brachial plexus birth injury and the assessment of severity are both made clinically based on history and examination findings.

Some groups support the routine use of EMG/NCS or MRI for diagnosis early in the patient’s course to confirm the presence of avulsion-type injuries; however, as the decision to intervene surgically is exclusively based on whether there is adequate recovery on physical examination over time, these studies typically do not aid clinical decision-making.

Therapeutics. In the first few days of life, the patient’s arm can be temporarily immobilized via swaddling if there is pain from an accompanying fracture. Caregivers should be instructed in appropriate positioning to avoid contractures, pressure ulcers, and unnecessary traction. If the patient tolerates it, gentle range of motion exercises may be started either immediately or at latest by 7 to 10 days of life. Physical therapy should be continued until the child’s brachial plexus injury recovers. For cases that result in permanent functional deficit, therapy should be tailored to the patient’s age and developmental stage.

Ideally, infants with brachial plexus injuries should be referred to a multidisciplinary specialty clinic for treatment. Teams at these clinics include pediatric neurologists, orthopedic surgeons, neurosurgeons, physical and occupational therapists, and social workers. If this is not possible, the infant should be followed closely by a neurologist to monitor the pace and extent of neurologic recovery. If antigravity biceps function does not return before 6 weeks of age, a referral to surgery is appropriate, as a subset of these infants will require microsurgical reconstruction of the plexus. In cases of suspected avulsion or rupture injuries where spontaneous recovery is impossible or unlikely, it is generally agreed the infant should undergo microsurgical reconstruction by age 3 months for avulsions and by 6 months for nerve ruptures. Early surgery minimizes motor end-plate loss and maximizes recovery time.

In less severe injuries, the indications for, and timing of, surgical interventions remain controversial. Most groups agree that lack of antigravity biceps function by 3 to 6 months is an indication for surgical intervention, while others continue to observe and operate as late as 9 or 10 months of age.

Surgical intervention for brachial plexus palsy includes early microsurgical repair of the brachial plexus using nerve grafts or nerve transfers. In both
cases, the neuronal scar tissue (neuroma) is resected. For rupture injuries, a donor nerve, most often the sural nerve, is inserted into the area of discontinuity. Nerve transfers, in contrast, redirect an uninvolved healthy nerve, such as the spinal accessory nerve (CN XI), to the distal site of nerve injury and rely on neuroplasticity for adoption of functional control by the transferred nerve.1

Outcomes. Most children with brachial plexus birth palsy recover well. A recent prospective study demonstrated full recovery in 50% of patients by 3 months of age, and 82% by 18 months. However, roughly one in 5 affected infants have some degree of permanent nerve damage.12

While patients with permanent injury have lower functional scores than their peers, these children have equivalent rates of individual and team sports participation as their peers.13 Most children with persistent injury can manage their activities of daily living, albeit with varying degrees of difficulty.11

CASE PART 3 The infant regained gravity-assisted biceps function at 3 months of age and antigravity biceps function at 5 months. Now 18 months old, he is able to use his left hand and arm, though still with weakness and range of motion limitations. He continues with intensive physical and occupational therapy and his ultimate outcome is not yet determined.

DISCUSSION Brachial plexus birth injuries are usually transient, but can result in permanent functional deficits. Signs of nerve root avulsion, indicating severe injury that will not recover spontaneously, include a total plexopathy (complete arm paralysis), Horner syndrome, or phrenic nerve involvement. These infants should be referred for microsurgical evaluation immediately so that reconstruction of the plexus, if indicated, can be performed by 3 to 6 months. All infants with brachial plexus nerve injuries need close follow-up to monitor progress, and early and ongoing physical and occupational therapy to maintain range of motion, prevent glenohumeral joint deformity, and maximize function.

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Dr. Pham: drafting/revising the manuscript, study concept or design, analysis or interpretation of data. Dr. Kratz: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, acquisition of data. Dr. Jelin: drafting/revising the manuscript. Dr. Gelfand: drafting/revising the manuscript.

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REFERENCES