Causes of Neonatal Brachial Plexus Palsy

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Abstract

The causes of brachial plexus palsy in neonates should be classified according to their most salient associated feature. The causes of brachial plexus palsy are obstetrical brachial plexus palsy, familial congenital brachial plexus palsy, maternal uterine malformation, congenital varicella syndrome, osteomyelitis involving the proximal head of the humerus or cervical vertebral bodies, exostosis of the first rib, tumors and hemangioma in the region of the brachial plexus, and intrauterine maladaptation. Kaiser Wilhelm syndrome, neonatal brachial plexus palsy due to placental insufficiency, is probably not a cause of brachial plexus palsy. Obstetrical brachial plexus palsy, the most common alleged cause of neonatal brachial plexus palsy, occurs when the forces generated during labor stretch the brachial plexus beyond its resistance. The probability of obstetrical brachial plexus palsy is directly proportional to the magnitude, acceleration, and cosine of the angle formed by the direction of the vector of the stretching force and the axis of the most vulnerable brachial plexus bundle, and inversely proportional to the resistance of the most vulnerable brachial plexus bundle and of the shoulder girdle muscles, joints, and bones. Since in most nonsurgical cases neither the contribution of each of these factors to the production of the obstetrical brachial plexus palsy nor the proportion of traction and propulsion contributing to the stretch force is known, we concur with prior reports that the term of obstetrical brachial plexus palsy should be substituted by the more inclusive term of birth-related brachial plexus palsy.

The causes of brachial plexus palsy in neonates can be classified based on the alleged time of occurrence, mechanism of injury, or according to their most salient feature. The classifications of the causes of brachial plexus palsy in neonates based on the alleged time of injury distinguish two major types: congenital and post-natal. Patients with congenital brachial plexus palsy can be further classified as having sustained the injury before the onset of labor (prepartum) or during labor (intrapartum).

The diagnosis of congenital brachial plexus palsy relies on observing arm weakness with a distribution consistent with a brachial plexus injury at birth. Observing the typical distribution of weakness associated with brachial plexus palsy is not always simple in neonates, because normal neonates have a relative paucity of arm movements shortly after delivery and the distribution of the weakness is similar in certain types of brachial plexus palsy and non-brachial plexus lesions. It is still more difficult to distinguish prepartum and intrapartum injuries among neonates with congenital brachial plexus palsy because: 1. the determination of the onset of labor is subjective; 2. a fetal deformity or the presence of an intrauterine CNS condition that produces arm weakness cannot be interpreted as a sine qua non indication of a prepartum origin of the brachial plexus injury; and 3. electromyographic studies, the only practical tool to time a brachial plexus injury, are only reliable when they yield very specific time-related findings in muscles innervated from the proximal region of the brachial plexus. A prepartum injury can be diagnosed when fibrillations are present during the first day of extrauterine life and the duration of labor is less than 24 hours, or when large motor unit potentials or nascent motor unit potentials are present during the first 10 days of life. An
Obstetrical Brachial Plexus Palsy

The incidence of obstetrical brachial plexus palsy is about 1 to 1.5 per 1000 live births.\textsuperscript{10-12} It has not changed despite the expected decrease that should have been brought about by the first 10 days of extrauterine life and appear in a second study performed days later.\textsuperscript{5,9} Hence, the classification of neonatal brachial plexus palsy based on the alleged time of occurrence is difficult and can seldom be accomplished.

The mechanism of action would seem a simple parameter to classify the causes of brachial plexus palsy in neonates, but it is not. The brachial plexus in neonates can be damaged by being stretched, compressed, infiltrated, or deprived of oxygen. Yet, stretch, the most common of these mechanisms, probably contributes to brachial plexus damage in patients with lesions primarily produced by any of the other mechanisms. In these cases, compression, infiltration, and oxygen deprivation by decreasing the resistance of the nerve bundles renders the plexus more vulnerable to stretch injury (Fig. 1).\textsuperscript{4}

The classification of causes of neonatal brachial plexus palsy according to their most salient associated feature seems, at first glance, the most imperfect one; however, it is probably the most appropriate. Delineating brachial plexus injuries by salient feature is widely used in the medical literature and allows the clinician the flexibility to catalog the causes of brachial plexus palsy in manageable groups without having to rely on rigid but uncertain parameters such as time of occurrence and mechanism of injury.

Obstetrical Brachial Plexus Palsy

Figure 1 The probability ($p$) of an obstetrical brachial plexus palsy (obpp) is directly proportional to the magnitude ($m$) and acceleration ($a$) of the stretching force, and the cosine formed by the vector of the stretching force and the axis of the most vulnerable brachial plexus nerve bundle ($\cos \theta @ \text{bp}$); and inversely proportional to the resistance ($r$) of the most vulnerable brachial plexus nerve bundle ($n$) and of the shoulder girdle muscles (mu), joints (j) and bones (b).

$p_{obpp} = m \cdot a \cdot \cos \theta @ \text{bp/r} (n+\text{mu+j+b})$

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The literature on predisposing factors for obstetrical brachial plexus palsy suffers from lack of precision, conflicting findings, and lack of correlation between the alleged predisposing factors and obstetrical brachial plexus palsy. Most articles equate neonatal brachial plexus palsy with obstetrical brachial plexus palsy, demonstrating a lack of precision in the literature. An example of conflicting findings relates to the duration of the second stage of labor. Some investigators found a precipitous second stage of labor to be a predisposing factor for brachial plexus palsy, whereas others have found a prolonged second stage of labor a risk factor.\textsuperscript{13-16} Most patients with obstetrical brachial plexus palsy do not have any risk factors accounting for their injury.\textsuperscript{10,11,13,14} Nonetheless, in order to understand the pathogenesis of obstetrical brachial plexus palsy, it is important to consider some of the predisposing and protective factors and their implications.

Predisposing factors for obstetrical brachial plexus palsy can parcel as maternal, labor-related, and fetal.\textsuperscript{15-17,19} The most important maternal factors are diabetes gravida and excessive weight gain during pregnancy. It is unclear whether diabetes gravida and increased maternal weight during pregnancy independently contribute to the production of obstetrical brachial plexus palsy when not associated with fetal macrosomia. The most common labor-related risk factors are shoulder dystocia, instrumented delivery, and a short second stage of labor; prolonged second stage of labor is probably a function of shoulder dystocia and of itself not a risk factor. Shoulder dystocia and instrumented delivery increase the stretch force brought upon the brachial plexus as a consequence of propulsive forces, traction forces, or both. The most common fetal risk factor is macrosomia. Other less frequent but pathogenetically revealing fetal risk factors include decreased fetal arm movements and the presence of a first cervical rib and clavicular fracture. Decreased arm movements may lead to atrophy of the shoulder girdle muscles and joints, thus rendering the brachial plexus more vulnerable to stretch forces. Decreased fetal arm movement may be due to uterine constraint or a central nervous system pathology associated with arm weakness.\textsuperscript{1,3,4} The presence of a cervical rib may, by modifying the anatomy of the region around the brachial plexus, alter the direction of the vector of the stretching force, increasing its alignment to the axis of the most vulnerable brachial plexus bundle (Fig. 1). Clavicular fracture, listed by some investigators as a predisposing factor, is more likely an associated lesion and the consequence of a shift in the direction of the vector of the stretching force toward a lower cosine value in relation to the brachial plexus nerve bundles.\textsuperscript{1,3,4,20-27}

Another point important to understanding the pathogenesis of obstetrical brachial plexus palsy is the relation between cesarean section and obstetrical brachial plexus palsy. Cesarean section has a significant protective effect, but by no means prevents obstetrical brachial plexus palsy.\textsuperscript{1,3,20,21} Although the importance of stretch, primarily traction, injury in the pathogenesis of brachial plexus palsy in neonates has been confirmed by detailed analysis of infants undergoing early brachial plexus surgical repair, obstetrical brachial plexus palsy is the consequence of an interplay of opposing physical forces (Fig. 1) and not...
merely an injury related to the magnitude, acceleration, and direction of the vector of the stretch force generated during delivery. Furthermore, the magnitude, acceleration, and direction of the vector of the stretch force is the product of the sum of the traction force generated by the obstetrician and the propulsive force generated by spontaneous or induced uterine contractions.25,26

Kaiser Wilhelm Syndrome

The most controversial cause of brachial plexus palsy is placental insufficiency, or Kaiser Wilhelm syndrome. In 2005, Jain and colleagues28 reviewed the literature on the birth of Kaiser Wilhelm II and concluded that decreased oxygenated blood flow to the left brachial plexus during pregnancy, and not trauma during delivery, was the cause of Kaiser Wilhelm’s injury. They support their conclusion by medical, mathematical, and theoretical considerations. It has been documented that Kaiser Wilhelm’s mother sustained a severe fall during pregnancy and that he was small at birth. These findings were interpreted as an indication of placental insufficiency. The mathematical and theoretical data was derived from the creation of a computer model of a fetoplacental unit. Using this model, Jain and coworkers28 showed that a reduction in the placenta’s surface area by 50% produces significant hypoxia of the left brachial plexus and not of the right brachial plexus.

This syndrome is not universally accepted. The concept that placental insufficiency produces a brachial plexus injury by oxygen deprivation has not been supported by further reports. Most physician contemporaries to the birth of Kaiser Wilhelm II and he himself considered birth trauma as the most likely cause of the injury. Interestingly, Kaiser Wilhelm II blamed Dr James Clark, the English doctor and personal physician to Queen Victoria, for failing to timely call Professor Martin, a specialist, to help at the delivery. Some historians attach the Kaiser’s animosity toward the English and the beginning of World War I to this incident.28,30

Familial Congenital Brachial Plexus Palsy

Familial congenital brachial plexus palsy is characterized by the presence of arm deformity at the time of birth, weakness in the typical distribution seen in patients with brachial plexus palsy, and a positive family history.30,31 Mollica and associates30 reported one family with eight affected members. Zaki and colleagues31 reported two affected families; one of the families had 4 affected members and the other family had two. A history of conditions that predispose to focal neuropathy in later life, such as familial brachial neuropathy or neuritis with brachial plexus predilection, and familial recurrent polyneuropathy or tomaculous neuropathy were not reported in any of the patients or their families. Chromosomal studies were normal in the affected members of the three families. A history of maternal uterine malformation was not reported in any of the cases. Dermatoglyphic patterns between affected and non-affected hands were similar.

The suggested modes of inheritance of familial congenital brachial plexus palsy are autosomal dominant with incomplete penetrance, X-linked inheritance, and autosomal recessive. The alleged mechanism of injury in familial congenital brachial plexus palsy has not been elucidated. It seems reasonable to hypothesize that an inherited intrinsic nerve pathology affecting the brachial plexus, such as familial brachial neuropathy or neuritis with brachial plexus predilection, could decrease the resistance of the plexus, thus rendering it more vulnerable to the stretch forces generated by spontaneous uterine contraction or traction during delivery.

In addition to familial congenital brachial plexus palsy, a genetic predisposition to brachial plexus palsy was reported by Polovina and coworkers32 based on dermatoglyphic patterns.

Maternal Uterine Malformation

Maternal uterine malformation, such as bifid uterus, has been associated with congenital brachial plexus palsy.33-37 This condition is infrequent and must be diagnosed only when a proven and significant maternal uterine malformation is documented and the possibility of an intrapartum injury has been excluded by electromyography. Limb contractures, forearm atrophy, smaller size of the hand, and decreased humeral bone calcium density are not definite proof of prepartum injury. The possibility that the latter findings indicate predisposing factors for intrapartum stretch injury should be considered in all cases. Congenital amniotic band constriction involving the brachial plexus has not been reported; however, it has been shown to involve all the major distal branches of the brachial plexus.38

Congenital Varicella Syndrome

Congenital varicella syndrome may produce complete brachial plexus palsy. The neuroanatomical structures most significantly involved are the anterior horn motor neurons and the dorsal root ganglia neurons.39,40 Damage to these neurons at the spinal segments which contribute to the brachial plexus lead to malformation of the plexus. Weakness in these patients may result from the intrinsic brachial plexus malformation or from a prepartum or intrapartum injury superimposed on a weakened brachial plexus.

Humeral or Vertebral Osteomyelitis

Neonatal brachial plexus palsy may be associated with humeral and vertebral group-B streptococcal osteomyelitis. The mechanism of injury is probably related to ischemia of the plexus. The clinical presentation is characterized by sudden onset of arm weakness several days after birth. Neonates with brachial plexus palsy associated with humeral and vertebral group-B streptococcal osteomyelitis may be afebrile but are usually very sensitive to motion of the arm.
or palpation of the affected region. Bone scan is the study of choice to detect early osteomyelitis.41,42

Exostosis of the First Rib
Exostosis of the first rib may be seen in neonates with brachial plexus palsy. The brachial plexus is involved at the level of the divisions. A bony mass under the clavicle should alert to the possibility of exostosis of the first rib. The diagnosis is established by conventional radiology. It is not known whether the injury in these patients was due to compression or stretch of the brachial plexus or if it had occurred before or during labor. The association of exostosis of the first rib and brachial plexus palsy has only been reported once.43

Tumors
Tumors originating from or involving the brachial plexus in neonates are rare. They include neurofibromas, rhabdoid tumors, and myofibroma. Neurofibromas and myofibromas produce slowly progressive arm monoparesis that may be associated with shoulder paresthesias. Rhabdoid tumors produce rapidly progressive arm weakness and a supraclavicular mass. Brachial plexus injury due to tumors results from compression, infiltration, or both.44,45

Hemangioma
Hemangioma in the shoulder girdle region is a rare cause of brachial plexus palsy.46,47 The mechanism of brachial plexus involvement could be compression or secondary to oxygen deprivation. The latter phenomenon is produced by the hemangioma shunting arterial blood from the brachial plexus. Hemangiomas may occur as part of a systemic disease or as an isolated condition.46 In one report, early intravenous corticosteroids resolved the arm weakness in a patient with cavernous hemangioma and brachial plexus palsy.47

Intrauterine Maladaptation
Brachial plexus palsy due to intrauterine maladaptation refers to: 1. a suspected or proven prepartum brachial plexus injury of unknown cause, or 2. an apparent intrapartum brachial plexus injury without the use of significant traction forces at the time of delivery. The alleged mechanism of injury of the brachial plexus in intrauterine maladaptation is the propulsive force generated by spontaneous or provoked uterine contraction prior to or during labor.

Supporters of intrauterine maladaptation as a cause of congenital brachial plexus palsy point to the poor correlation between predisposing factors and obstetrical brachial plexus palsy, involvement of the posterior shoulder, the occurrence of other congenital peripheral nerve injuries not related to traction (such as congenital facial and peroneal nerve palsy), and the absence of applied traction during vaginal delivery or cesarean section extraction in a significant number of neonates classified as having obstetrical brachial plexus palsy. Opponents of intrauterine maladaptation as a cause of congenital brachial plexus palsy point to lack of documentation or minimization of the reported traction force used.48-52

A possible explanation for the production of brachial plexus palsy despite an apparently normal delivery could be that the propulsive force generated by spontaneous or provoked uterine contraction prior to or during labor can be enough to disrupt the continuity of the brachial plexus nerve bundles if their resistance or the resistance of the shoulder girdle structures is decreased (Fig. 1).

Conclusion
The causes of brachial plexus palsy should be classified according to their most salient associated features. The causes of brachial plexus palsy are obstetrical brachial plexus palsy, familial congenital brachial plexus palsy, maternal uterine malformation, congenital varicella syndrome, osteomyelitis involving the proximal head of the humerus or cervical vertebral bodies, exostosis of the first rib, tumors and hemangioma in the region of the brachial plexus, and intrauterine maladaptation. Kaiser Wilhelm syndrome, neonatal brachial plexus palsy due to placental insufficiency, is unlikely to be a cause of neonatal brachial plexus palsy. Intrauterine maladaptation may be related to decrease resistance of the brachial plexus nerve bundles or of the shoulder girdle structures. Because in most cases labeled obstetrical brachial plexus palsy the contribution of the different factors involved in the production of brachial plexus injury is unknown, we concur with prior reports that the term birth-related brachial plexus palsy should be used to refer to these injuries in the future and that the term obstetrical brachial plexus palsy, implying traction as the sole contributor to brachial plexus injury, should be abandoned.22,53

Disclosure Statement
The author does not have a financial or proprietary interest in the subject matter or materials discussed, including, but not limited to, employment, consultancies, stock ownership, honoraria, and paid expert testimony.

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