Neonatal brachial plexus palsy: a permanent challenge

Paralisia do plexo braquial neonatal: um desafio permanente

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ABSTRACT
Neonatal brachial plexus palsy (NBPP) has an incidence of 1.5 cases per 1000 live births and it has not declined despite recent advances in obstetrics. Most patients will recover spontaneously, but some will remain severely handicapped. Rehabilitation is important in most cases and brachial plexus surgery can improve the functional outcome of selected patients. This review highlights the current management of infants with NBPP including conservative and operative approaches.

Keywords: obstetric paralysis, brachial plexus, birth injuries, peripheral nerve surgery, brachial plexus surgery.

RESUMO
A paralisia neonatal do plexo braquial (PNPB) tem uma incidência de 1,5 casos por 1000 nascidos vivos e não tem diminuído a despeito dos recentes avanços em obstetrícia. A maioria dos pacientes recupera-se espontaneamente, mas alguns permanecerão com sequelas graves. A reabilitação é importante na maioria dos casos e a cirurgia do plexo braquial pode melhorar o resultado funcional em pacientes selecionados. Esta revisão destaca o manejo atual de lactentes com PNPB, incluindo as terapêuticas conservadora e cirúrgica.

Palavras-chave: paralisia obstétrica, plexo braquial, traumatismos do nascimento, cirurgia de nervo periférico, cirurgia do plexo braquial.

Neonatal brachial plexus palsy (NBBP) is an ancient disease. There are references to this condition back to the the Old Testament, and Galen’s histories. The first scientific description was made by the Scottish obstetrician William Smellie, in 1768. The classical neurologic description of the upper brachial plexus lesion was done by Duchenne in 1872 and Erb in 1874. Augusta Klumpke, the first woman in France to be interne des hôpitaux, described the lower plexus lesion in 1885, including the ocular autonomic involvement. The most famous patient with this condition was Kaiser Wilhelm II, who ruled Germany during the First World War. Kennedy performed the first surgery in 1903, but it was not until the convincing results reported by Gilbert in the 80’s that surgical treatment became an option for these patients.

The incidence of NBBP varies from 0.5 to 3.0 cases per 1000 live births. Despite the advances in modern obstetrics, its incidence has not declined during the last decades. In Sweden, there has been actually an increase in its incidence for unknown reasons, but higher birth weight in the population has been probably contributed to this fact. There is no data of Brazilian incidence, but it is probably in the lower spectrum due to the high proportion of cesarean sections in our country. Nevertheless, we have seen over 400 cases in our hospital in the last 14 years. The recent efforts of Brazilian government to increase the proportion of vaginal birth may actually increase our incidence of NBBP.

RISK FACTORS AND PREVENTION

There are several well-known risk factors for NBPP; however, its occurrence remains an essentially unpredictable event. Most cases have no recognizable cause and just a minority of deliveries with identifiable risk factors will result in brachial plexus lesions.

The first point relates to the existence of congenital lesions. Although there are several convincing reports, these seem to account for a very small portion of cases. They have a different natural history, since limb atrophy is usually present since birth. Needle electromyography was once used to identify...
this situation based on the false assumption that an acute lesion would take about two weeks to generate abnormal muscle spontaneous activity, but there is experimental evidence that this time frame is considerably shorter in newborns.

The majority of NBPP is related to brachial plexus stretching during the delivery. The relative contributions of obstetric maneuvers and uterine propulsion have been fervently debated due to its legal implications. There are documented cases of NBPP without fetal head traction and the term "obstetric paralysis" has been condemned by several authors. The main risk factor for NBPP is shoulder dystocia which is reported in at least half of the cases. The fetal shoulder gets stuck under the pubic symphysis, opening the angle between the clavicle and cervical spine, and creating an upward tension gradient. This explains the higher incidence of upper brachial plexus lesions.

Birth weight is the most important fetal factor for NBPP, and it is clearly related to shoulder dystocia. A birth weight higher than 4.5 kg carries a ten-fold risk increase for brachial plexus lesions. Maternal diabetes mellitus is also related to this, but also seems to have some independent risk contribution. Other maternal risk factors include obesity, short stature, and previous shoulder dystocia.

Forceps extractions are related to a higher risk for NBPP; however, it is not clear if this is due to fetal traction or just an associated factor present in a difficult delivery situation. Pelvic deliveries are related to severe and often bilateral lesions, which are probably caused by cervical hyperextension. Cesarean sections have a protective effect, but cannot avoid NBPP completely.

The indication of cesarean section for macrosomic babies would be a rational approach for prevention; however, fetal ultrasound is not very accurate for detection of large fetus. A cost-effective analysis indicated that it would take 3695 cesarean sections to prevent a single permanent NBPP in patients with estimated birth weight higher than 4.5 kg. The American College of Obstetricians and Gynecologists recommends cesarean section for estimated birth weight higher than 5 kg, which correspond to less than 4% of our cases.

**CLINICAL PICTURE**

The clinical presentation can be classified according to the anatomic structures compromised. NBPP is a closed supraclavicular lesion that affects sequentially the upper (C5-C6), middle (C7) and lower (C8-T1) brachial plexus trunks. The right side is affected in two thirds of the cases due to the most common fetal presentation. Bilateral cases are seen in up to 5%, but are usually asymmetric.

Isolated lesion of the upper trunk (C5-C6), also known as Erb’s palsy or Narakas grade I injury, occurs in about half of cases. The typical limb posture is called “waiter’s tip”, in which the arm is adducted and internally rotated, the elbow is extended, and the wrist is flexed (Figure 1). The Moro reflex is absent in the affected side, but the grasp reflex is normal. Motor deficit includes shoulder abduction, external rotation and elbow flexion. Biceps tendon reflex is lost, but pain sensitivity is usually preserved.

Upper and middle trunk (C5-C7) lesions, or Narakas grade II injury, accounts for one third of the cases. In addition to the motor deficits seen in Erb’s palsy, elbow and wrist extension are also compromised. Finger flexion is present, but usually weaker than the healthy side. All tendon reflexes are absent in the affected limb. Pain sensibility may be lost in the thumb or middle finger, and this is related to a poor prognosis.

Total plexus lesions (C5-T1) are seen in the remaining 17% of the cases. Some patients can still show minor finger movements and are classified as Narakas grade III injury. Narakas grade IV picture is of a complete flail arm, with abnormal sensibility, and sympathetic ocular involvement known as Claude-Bernard-Horner syndrome. Isolated lower plexus lesions, known as Klumpke’s palsy, are extremely rare. Most reported cases were probably total plexus lesions which recovered upper plexus function after a while. These patients develop a late posture of elbow flexion, wrist extension and supination known as “beggar’s hand”.

**Figure 1.** Patient with an upper brachial plexus lesion on the right side showing the classical “waiter’s tip” posture. The arm is adducted and internally rotated, the elbow is extended, and the wrist is flexed.

**Figure 2.** Patient with a total brachial plexus lesion on the right side showing a flail arm and Horner sign, which is characterized by miosis, partial ptosis, and enophtalmos. Hemifacial anhidrosis is usually not seen in this context.
ANCILLARY EXAMS

The diagnosis of NBPP is clinically obvious and no ancillary exam is necessary. Plain X rays can be helpful for detection of concurrent lesions, such as clavicular fracture or phrenic paralysis. Electrodiagnosis and image studies can be useful for prognostic and surgical planning providing data to characterize the root viability. To that extend the lesion can be divided in preganglionic or postganglionic injury according to the localization related to dorsal root ganglion (DRG). In the preganglionic injury the lesion is proximal to the DRG and is associated with root avulsion or intraforaminal root injury. Accordingly, the root cannot be used as donor to reconstruct the brachial plexus. The postganglionic injury is distal to the DRG and the repair can be performed by interposing nerve grafts from the viable root (or roots) to the distal plexus.

Nerve conduction studies and electromyography were commonly performed at three months of age as part of preoperative investigation. Technical issues and overly optimistic results led several surgeons to abandon this procedure. However, recent reports have shown that electrodiagnosis can be useful for prognostic estimation if performed earlier. Motor nerve conduction studies can estimate the percentage of motor axonal degeneration, which correlates with the functional outcome. Preservation of sensory potentials in a patient with severe paralysis is indicative of preganglionic lesion, which carries a grim prognosis. Biceps needle electromyography at one month of age has been used as part of Leiden’s University algorithm for surgical indication in these patients.

Detection of nerve root avulsions is the main indication for image studies. The classical finding is the pseudomeningocele, but the correlation of this marker with root avulsion is not perfect. Modern image studies can detect intraspinal nerve root continuity. The ideal method for evaluation is still a matter of controversy. Some prefer computed tomography myelogram due to a higher resolution. On the other hand, magnetic resonance image (MRI) is less invasive, allows multiplanar reconstructions, and can evaluate extraspinal lesions. Some authors report similar resolution between these methods. MRI is currently the method of choice in pediatric patients.

PROGNOSIS AND SURGICAL INDICATION

Data about NBPP prognosis is surprisingly confuse. The proportion of patients with complete recovery varies among different studies from 7% to 97%. Ancient publications had a grim perspective, which were followed by an overly optimistic view. Unfortunately, there is no perfect study to address this issue. The ideal design would be a population based on a prospective study, with patients enrolled soon after birth, followed for at least three years with no surgical intervention and with less than 10% of losses, and with a complete and reproducible final evaluation. Recent studies indicate a more balanced perspective: about 50% of the patients will be completely recovered, while about 15% will be severely handicapped. These would be the ideal candidates for surgical intervention. The remaining 35% of the patients will have a satisfactory outcome, but with some shoulder functional limitation. External rotation is usually the main problem, and these patients show excessive shoulder abduction (trumpet sign) while attempting to put hand to mouth.

Early surgery provides a larger time window for nerve regeneration and theoretically would have a better outcome. On the other hand, since the rate of spontaneous recovery is high, many children would be submitted to an unnecessary procedure. There is no agreement of which infants should be operated and when it should be done. The most popular criterion was introduced by Gilbert, based on the prognostic studies conducted earlier by Tassin: infants without biceps function at three months of age should be operated. This view has been endorsed by many other nerve surgeons, although some studies have criticized this approach due to a low specificity. Note that “biceps function” was originally related to biceps palpable contraction, but others use elbow flexion. Electric muscle activity detected by needle electromyography is not considered biceps function.

For patients with total plexus lesions, there is little controversy about the indication of early surgery. Some actually prefer to operate earlier than three months, while most wait up to this age due to anesthetic safety. For patients with C5-C6 or C5-C7 lesions, some surgeons prefer to wait a little longer, up to six months of age, which is probably a more cost-effective approach. Clinical evaluation should be not only based on elbow flexion, but also include shoulder abduction, elbow extension and wrist and finger extension. Surgery after twelve months of age is usually not very effective, although some late selective distal nerve transfers can still offer good results.

SURGERY

The supraclavicular approach usually provides adequate field for exploration and reconstruction of the brachial plexus structures. Combined infraclavicular approach through a deltoitoral incision is rarely necessary in cases of lower trunk lesions, but section of the clavicle is usually not performed. Intraoperative nerve stimulation is crucial for the identification of viable neural structures, but recording of nerve action potentials across the sites of lesions has not been proved to be advantageous in this particular situation.

The typical lesion found is the neuroma in continuity (Figure 3), in which there is an internal rupture of axons and disorganization of the supporting connective tissue, corresponding to lesions type 3 or 4 in the Sunderland classification. This leads to a local proliferation of axonal sprouts and fibroblasts,
but very few axons are able to effectively cross the lesion site. There are three possible surgical approaches in this situation: external neurolysis, nerve grafting, and nerve transfers. External neurolysis consists in removing scaring around the nerve. It has not been proved to be an effective isolated procedure, but it is a necessary step for other reconstructive strategies.

Nerve grafts are used to connect nerve stumps after the removal of the neuroma in continuity. The sural nerve is usually harvested for this purpose, but other nerves can be used as well. The grafts provide a path for nerve regeneration, but clinical results will take many months to appear, since the axonal sprouts will have to grow from the lesion site to the target muscle. After crossing the coaptation site, the axon grows at a rate of 1 to 5 mm/day.

Nerve transfers were originally developed for nerve repair when a viable proximal nerve stump was not available, such as in cases of root avulsions. The donor nerve may be part of the brachial plexus itself (intraplexual transfer) or a nearby nerve outside the plexus (extraplexual transfer). Examples of intraplexual transfers include the use of the medial pectoral nerve, transfer from a triceps motor branch to the axillary nerve, or from a fascicle of the ulnar nerve to the biceps motor branch (Oberlin procedure). Examples of extraplexual transfers include from the accessory nerve to the suprascapular nerve or from the intercostal nerves to the musculocutaneous nerve. The phrenic nerve is also a possible donor in adults, but is not used in infants. Nerve transfers provide a more distal source of motor axons with a single coaptation site. This means that recovery is usually faster and that late procedures can still be effective.

**LONG TERM COMPLICATIONS**

Internal rotation contractures and posterior humeral subluxation are by far the most common long term complication in NBPP. It is related to muscular imbalance due to poor active external rotation. It leads to a progressive shoulder deformity according to the Waters classification, ranging from mild glenoid retroversion (Waters grade II) to a complete posterior luxation with false glenoid and proximal humeral deformity (Waters grade VII). Early referral to an orthopedic specialist is crucial.

**REHABILITATION**

Limb immobilization has been associated with shoulder deformities and is not recommended, except if bone fractures are also present. Some advocate that immobilization may be useful for pain treatment during the first week, but it is difficult to evaluate pain in these patients. It seems that NBPP is not painful, at least in older patients. This picture is very different from that seen in adults with brachial plexus lesions, who usually show severe neuropathic pain, especially after root avulsions.

Physical therapy and occupational hand therapy are important, but it is essential to involve the parents in the rehabilitation program. Passive range-of-motion exercises are critical to avoid muscle contractures and should be done several times on a daily basis. It is a good idea to include it in other routine activity such as changing dippers. As soon as the child shows intentional voluntary control, it is important to stimulate the affected limb to avoid developmental apraxia. Encouraging bimanual activities is an interesting strategy for that. Wrist splinting can help to enhance hand function in cases of wrist drop, as long as it does not prevent limb use during daytime. Aberrant reinnervation can result in biceps-triceps cocontraction, which can be treated with botulinum toxin. This can also be used to prevent muscle contracture of the shoulder internal rotators.

**Figure 3.** Surgical view after supraclavicular approach to the right brachial plexus in a child with paralysis related to upper trunk. A large neuroma in continuity (N) of the upper trunk was identified after external neurolysis. C5: fifth root; C6: sixth root; D: distal; L: lateral; PN: phrenic nerve; SN: suprascapular nerve.

**Figure 4.** Surgical view of intercostals nerves transfer to musculocutaneous nerve in the right thoracic region. All brachial plexus roots were avulsioned during the supraclavicular approach. 3th R: third rib; 4th R: fourth rib; 5th R: fifth rib; D: distal; IN: intercostal nerves; IS: intercostal space; MN: musculocutaneous nerve.
surgeon is important to avoid glenoid dysplasia and possibly shoulder pain. Other orthopedic deformities can also be seen, such as scapular winging, elbow flexion contracture, radial head luxation, fixed pronation or supination posture, and claw hand deformity. Growth imbalance between the upper limbs is common in severe cases of NBPP.

Little attention has been devoted to sensory disturbances, since the prognosis of sensory deficits is usually good. However, some children can develop a self-mutilating biting behavior. This is more common after brachial plexus surgery and is probably related to some kind of uncomfortable paresthesia. This is only temporary and it is crucial to prevent the child from eating off their own fingers and assure the parents that it will pass after a few months.

**CONCLUSION**

NBPP is a common situation and there is no perspective of adequate prevention in the near future. Most affected newborns will recover spontaneously, but some might be severely handicapped without appropriate care. Early referral to specialized centers with multidisciplinary approach should be provided to all patients that do not recover after a couple of weeks.

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