

Diaphragmatic Paralysis Associated With Neonatal Brachial Plexus Palsy

Michyla Bowerson, BS*,
Virginia S. Nelson, MD[†], and
Lynda J.-S. Yang, MD*

Phrenic nerve palsy can occur in the context of neonatal brachial plexus palsy, yet neither outcomes nor definitive treatment guidelines have been established. Diaphragmatic paralysis alone in the newborn results in significant respiratory sequelae and failure to thrive. Reviewing the available literature revealed little information about the incidence of phrenic nerve palsy associated with neonatal brachial plexus palsy, or whether outcomes are associated with the severity of the brachial plexus palsy. Of patients with brachial plexus palsy evaluated during 2005-2009 (n = 166) at our institution, a minority (2.4%; n = 4) had clinically significant diaphragmatic palsy. Of these, a majority (75%; n = 3) manifested respiratory complications sufficient to warrant diaphragmatic plication. The severity of brachial plexus palsy failed to correlate with severity of respiratory consequences. None of the patients underwent nerve repair or reconstruction. We suggest that diaphragmatic paralysis should not be overlooked during a brachial plexus examination, and diaphragmatic paralysis in the very young may require aggressive intervention before the treatment of brachial plexus palsy. © 2010 by Elsevier Inc. All rights reserved.

Bowerson M, Nelson VS, Yang L J.-S. Diaphragmatic paralysis associated with neonatal brachial plexus palsy. *Pediatr Neurol* 2010;42:234-236.

From the *Department of Neurosurgery and [†]Department of Physical Medicine and Rehabilitation, University of Michigan Medical School, Ann Arbor, Michigan.

Introduction

In infants, diaphragmatic paralysis after phrenic nerve palsy is detrimental. Unlike respiration in adults, respiration in infants depends largely on their diaphragms. The baby's chest wall is more compliant, the mediastinum is more mobile, and the intercostal muscles are weaker [1,2]. Even unilateral phrenic nerve palsy in the newborn can result in significant respiratory sequelae such as repetitive respiratory infections with failure to thrive, often necessitating multiple intensive-care admissions, intubation, or tracheostomy.

Neonatal brachial plexus palsy may occur concurrently with phrenic nerve palsy. It is unknown whether the outcome of phrenic nerve palsy is associated with the severity of brachial plexus palsy. The incidence of this association has not been well-studied. Therefore, treatment guidelines remain uncertain with regard to the conservative versus surgical management of phrenic nerve palsy in the context of brachial plexus palsy. We present the University of Michigan experience, with a review of the literature.

Study Design

We performed a retrospective medical record review of 166 patients with neonatal brachial plexus palsy evaluated at the University of Michigan during the period 2005-2009. Four patients also manifested phrenic nerve palsies. The phrenic nerve palsies were evident on chest x-rays as elevations of the hemidiaphragm, and were confirmed and followed by ultrasound or fluoroscopy, indicating absent or paradoxical movements of the hemidiaphragm. Ultrasound or fluoroscopy was repeated after diaphragmatic plication. All patients were followed in the Brachial Plexus Program at the University of Michigan, and detailed brachial plexus examinations were performed at regular intervals. Approval for this study was granted by the University of Michigan Institutional Review Board.

Case Reports

The incidence of clinically significant phrenic nerve palsy in patients with brachial plexus palsy in our study was 2.4%. All hemidiaphragmatic and brachial plexus palsies occurred on the right side. Seventy-five percent (n = 3) of infants required diaphragm plication via

Communications should be addressed to:
Dr. Yang; Department of Neurosurgery, University of Michigan Health System; 1500 East Medical Center Drive, Room 3552, Taubman Center; Ann Arbor, MI 48109-5338.
E-mail: ljsyang@umich.edu
Received August 3, 2009; accepted November 2, 2009.

thoracoscopy, but no patients required surgical nerve repair or reconstruction (biceps muscle strength was used as a standard indicator for recovery from brachial plexus palsy). In patients undergoing thoracoscopic plication, supplemental oxygen and nasogastric feeds were discontinued by 1 month postoperatively, with subsequent steady weight gain. One patient presented with temporary respiratory distress, but did not require surgical intervention. Risk factors for a concurrent presentation of brachial plexus palsy and phrenic nerve palsy included breech presentation, forceps extraction, and shoulder dystocia. Increased birth weight was not evident in these patients. Details of patients' medical courses are presented below.

Patient 1

The patient was a girl weighing 3.6 kg, born to a gravida 2, para 1 mother at term, aided by the use of forceps. The infant presented with right diaphragmatic paralysis associated with brachial plexus palsy (Narakas grade II) [3], resulting in moderate respiratory insufficiency. Her Apgar scores were 4 at 5 minutes and 9 at 10 minutes. She was hospitalized for 1 month in the intensive care unit, required continuous supplementary oxygen, and was discharged with oxygen via nasal cannula (1/16 L/minute). Because of her respiratory difficulties, the patient was unable to take food orally, and required placement of a nasogastric tube because of failure to thrive.

The patient presented at the Brachial Plexus Program at age 1 month. She manifested tachypnea at 66-100 breaths per minute, with difficulty feeding (weight, 3.94 kg; 22nd centile), and with antigravity biceps power. Although her right arm improved, repeated fluoroscopy demonstrated persistent paralysis of the right hemidiaphragm. Given her tenuous respiratory status and the requirement for nasogastric feedings, the patient underwent thoracoscopic plication of the right hemidiaphragm at age 2 months. Immediate improvement in respiratory effort was evident, with a normalized respiratory rate and less abdominal effort. She was weaned from supplementary oxygen by postoperative day 3, and from nasogastric feedings within 1 month. At age 11 months, she underwent a normal right upper-extremity examination with 5/5 biceps strength, indicating resolution of the brachial plexus palsy.

Patient 2

The second patient was a boy weighing 3.7 kg, born to a 33-year-old gravida 2, para 2 mother via spontaneous vaginal delivery at term, complicated by shoulder dystocia and diaphragmatic palsy associated with brachial plexus palsy (Narakas grade I) [3]. His Apgar scores were 5 at 5 minutes and 10 at 10 minutes. He required intensive care for 16 days with oxygen therapy via nasal cannula, and was discharged with an oxygen requirement of 1/4 to 1/2 L/minute. He initially did well, with adequate oral nutrition intake, stable oxygen requirement, and appropriate weight gain.

However, he required hospital readmission at age 27 days for labored breathing with worsening tachypnea (80-100 breaths per minute) and poor feeding, necessitating intubation and nasogastric feedings. The patient was diagnosed with pneumonia, and initial attempts to extubate resulted in a small pneumothorax, atelectasis, and intermittent desaturations. He was extubated on hospital day 6, but tachypnea persisted, with increasing respiratory labor. An ultrasound of the chest revealed no improvement in diaphragmatic function. Therefore, the patient underwent diaphragmatic plication at age 35 days. Supplementary oxygen and a nasogastric tube were discontinued by postoperative day 5. After plication, the patient manifested no additional respiratory difficulties, and his respiratory rate stabilized between 60-70 breaths per minute.

At age 5.5 months, the patient demonstrated a normal passive range of motion in his right arm, and his active range of motion continued to exhibit improvement. His biceps strength was 4+/5.

Patient 3

The patient was a boy weighing 2.87 kg, born at term to a 40-year-old gravida 2, para 1 mother via forceps-assisted vaginal delivery for fetal

tachycardia. The pregnancy was complicated by gestational diabetes requiring insulin, bipolar disorder, and maternal chorioamnionitis postpartum. His Apgar scores were 4 at 5 minutes and 8 at 10 minutes. He was diagnosed with right brachial plexus palsy (Narakas grade II) [3] and phrenic nerve palsy with right hemidiaphragm paralysis. He was hospitalized for sepsis, presumably resulting from the presence of maternal chorioamnionitis.

The patient was admitted to the intensive care unit at age 5 days because of hypoxia and physiologic jaundice. He required 1/4 L/minute oxygen via nasal cannula, and was discharged home with oxygen. At age 15 days, the patient was readmitted to the intensive care unit with fever, lethargy, tachycardia, tachypnea, and respiratory distress. A nasogastric tube was placed for feeding to avoid aspiration resulting from tachypnea. He remained tachypneic on 1 L/minute oxygen, with failure to thrive. At age 1 month, the patient underwent diaphragmatic plication. His respiratory rate slowly normalized with 1/4 L/minute oxygen, and his oral intake increased, soon leading to discontinuation of the nasogastric tube. The brachial plexus palsy continued to resolve spontaneously. At age 7 months, the patient's biceps strength was 4/5.

Patient 4

The patient was a boy, weighing 1.98 kg with right brachial plexus palsy and phrenic nerve palsy diagnosed by chest x-ray, born at 36 weeks of gestation to a gravida 1, para 1 16-year-old mother via spontaneous vaginal delivery. The pregnancy was complicated by intrauterine growth retardation and breech presentation, with possible head entrapment and birth asphyxia. His Apgar scores were 3 at 5 minutes and 4 at 10 minutes. The patient required positive pressure ventilation, and was subsequently intubated because of poor respiratory effort. He underwent selective cerebral hypothermia for 72 hours. He remained on continuous mechanical ventilation until age 8 days, and on oxygen until age 13 days. He was discharged after 17 days in the intensive care unit, and required no additional admissions.

The patient had continued respiratory difficulty requiring nebulized albuterol treatments three to four times per week, with one episode of pneumonia at age 5 months that resolved quickly with oral antibiotics. He was referred to the Brachial Plexus Program at age 9 months. By that time, his brachial plexopathy (initially Narakas grade III) [3] was resolving spontaneously. His biceps strength was 4/5.

Discussion

The incidence of perinatal diaphragmatic paralysis is approximately 1 in 15,000 to 1 in 30,000 live births, and mortality is estimated at 10-15% [4]. Of 166 patients in our clinic with neonatal brachial plexus palsy, the incidence of concurrent ipsilateral phrenic nerve palsy was 2.4% (n = 4), and the majority of these patients required diaphragmatic plication for continued respiratory distress. Al-Qattan et al. [5] reported an incidence of 4.2%, but respiratory outcomes were not addressed. Conversely, 66.7-71.4% of patients manifesting phrenic nerve palsies were reported to demonstrate some degree of brachial plexus palsy [4,6].

Risk factors for the simultaneous occurrence of both phrenic nerve and brachial plexus palsies include breech presentation, maternal diabetes, forceps or vacuum extraction, and shoulder dystocia. Other studies reported breech presentation as a risk for concurrent brachial plexus and phrenic nerve palsies [7,8]. None of the patients in our series were born via cesarean section. Cesarean sections may constitute a protective factor, decreasing the risk of birth trauma. Other potential risk factors include uterine

malformation [9], resulting in the intrauterine onset of brachial plexus and phrenic nerve palsy, and macrosomia [4]. We did not find increased birth weight to be a risk factor for the concurrent presentation of both conditions, although increased birth weight was reported to be a risk factor for neonatal brachial plexus palsy alone. The right hemidiaphragm was more commonly affected in our patients, in concordance with other reports (70-80%) [4,10].

A diagnosis of phrenic nerve palsy is suspected when a patient experiences respiratory distress at birth, and subsequent imaging studies reveal an elevated hemidiaphragm. Early fluoroscopy or ultrasound is recommended to assess diaphragmatic movement. Ultrasound is the preferred diagnostic modality, because it does not involve radiation [4,11].

Treatment guidelines for the management of patients manifesting continued respiratory distress resulting from diaphragmatic paralysis have yet to be established. Initial supportive management usually includes mechanical ventilation, oxygen, and nasogastric tube feedings to avoid failure to thrive and to ensure weight gain. A viable option is surgical management with diaphragmatic plication via thoracoscopy or thoracostomy. We suggest that indications for surgical intervention in phrenic nerve palsy with associated brachial plexus palsy include a continued need for respiratory or ventilatory support, and failure to thrive within the first few months after birth. The timing for diaphragmatic plication remains controversial, because the risks associated with surgical intervention are greater at a younger patient age, but efforts have been increasing to perform plication at the earliest sign of respiratory distress [4,12]. Stramrood et al. [4] reported that when plication was performed at around age 20 days, patients rapidly improved. In our study, 3 of 4 patients underwent diaphragmatic plication at age 1-2 months, and experienced significant, rapid respiratory improvement postoperatively. In addition, repeated admissions to the intensive care unit for respiratory distress necessitate surgical intervention.

Predicting the respiratory outcomes of diaphragmatic paralysis based on the occurrence or severity of brachial plexus palsy has not been investigated. The phrenic nerve originates at the level of C₄ in the cervical spine, with contributions from the C₃, C₄, and C₅ nerve roots. Concurrent phrenic nerve injury and brachial plexus palsy (at C₅, C₆, possibly C₇, C₈, and T₁) indicate a wider proximal zone of injury, which was thought to indicate poorer outcomes. Our study illustrates that the severity of respiratory signs resulting from phrenic nerve palsy does not correlate with the severity of the brachial plexus palsy, because the brachial plexus palsies in all four of our patients resolved spontane-

ously. Recovery of arm function has little value in framing a prognosis in the context of phrenic nerve palsy. The patient with the most severe brachial plexus palsy (from C₅ to T₁) was the only patient who did not require diaphragmatic plication. Similarly, Al-Qattan et al. [5] reported that the presence of phrenic nerve palsy in newborn babies with brachial plexus palsy did not imply worse spontaneous motor recovery of the limb.

Diaphragmatic paralysis resulting from phrenic nerve palsy in the context of neonatal brachial plexus palsy can lead to dire consequences. Our data indicate that spontaneous recovery from brachial plexus palsy does not prognosticate a better respiratory outcome from phrenic nerve palsy. Early evaluations of patients with brachial plexus palsy should include an investigation of the phrenic nerve. The indications and timing for diaphragmatic plication are not established. However, accumulating data suggest that earlier interventions improve outcomes. Further investigations will be necessary to develop more definitive guidelines for the treatment of this condition.

References

- [1] Meyers BF, Kozower BD. ACS Surgery: Principles and Practices, 6th ed., Chapter 4.6. Philadelphia: B.C. Decker Inc., 2008, <http://www.acssurgery.com/acs/Chapters/ch0406.htm>.
- [2] Simansky DA, Paley M, Refaely Y, Yellin A. Diaphragm plication following phrenic nerve injury: A comparison of paediatric and adult patients. *Thorax* 2002;57:613-6.
- [3] Zafeiriou DI, Psychogiou K. Obstetrical brachial plexus palsy. *Pediatr Neurol* 2008;38:235-42.
- [4] Stramrood CA, Blok CA, van der Zee DC, Gerards LJ. Neonatal phrenic nerve injury due to traumatic delivery. *J Perinat Med* 2009;37:293-6.
- [5] Al-Qattan MM, Clarke HM, Curtis CG. The prognostic value of concurrent phrenic nerve palsy in newborn children with Erb's palsy. *J Hand Surg [Br]* 1998;23:225.
- [6] Hughes CA, Harley EH, Milmo G, Bala R, Martorella A. Birth trauma in the head and neck. *Arch Otolaryngol Head Neck Surg* 1999;125:193-9.
- [7] Al-Qattan MM. Obstetric brachial plexus palsy associated with breech delivery. *Ann Plast Surg* 2003;51:257-65.
- [8] Blaauw G, Slooff ACJ, Muhlig RS. Results of surgery after breech delivery. In: Gilbert A, editor. *Brachial plexus injuries*. Florence, KY: Taylor & Francis, 2001:217-24.
- [9] Dunn DW, Engle WA. Brachial plexus palsy: Intrauterine onset. *Pediatr Neurol* 1985;1:367-9.
- [10] Rennie JM. Respiratory problems of infants with neurological disease. In: Greenough A, Milner AD, editors. *Neonatal respiratory disorders*. London: Hodder Arnold, 2003:254.
- [11] Lemmer J, Stiller B, Heise G, et al. Postoperative phrenic nerve palsy: Early clinical implications and management. *Intensive Care Med* 2006;32:1227-33.
- [12] Tsugawa C, Kimura K, Nishijima E, Muraji T, Yamaguchi M. Diaphragmatic eventration in infants and children: Is conservative treatment justified? *J Pediatr Surg* 1997;32:1643-4.