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Case Report

Successful Management of Neonatal Diaphragmatic Palsy with Home CPAP: A Case Report

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Abstract

Phrenic nerve palsy, a rare cause of respiratory distress in neonates, leads to diaphragmatic paralysis and significant respiratory compromise. While surgical interventions, such as diaphragmatic plication, are often considered, they carry inherent risks. This report highlights the use of home continuous positive airway pressure (CPAP) as a conservative treatment option. A late preterm infant diagnosed with right diaphragmatic palsy after birth was managed with home CPAP, avoiding surgery. The infant successfully transitioned to spontaneous breathing by 75 days of life, highlighting the effectiveness of non-surgical management. This case underscores the potential role of home CPAP as a viable alternative to surgical intervention in selected cases.

Keywords: *Diaphragmatic Paralysis; CPAP; Phrenic Nerve Palsy; Respiratory Distress*

Abbreviations

CXR: Chest X-ray; DOL: Day of Life; CPAP: Continuous Positive Airway Pressure; NICU: Neonatal Intensive Care Unit; BLS: Basic Life Support

Introduction

Phrenic nerve palsy is a rare but serious cause of respiratory distress in newborns, often arising from birth trauma or perinatal complications. This results in diaphragmatic paralysis and compromises normal respiratory function. Diaphragmatic paralysis can present unilaterally or bilaterally, with unilateral cases being more common. For affected neonates, this paralysis can lead to significant respiratory compromise, recurrent pulmonary infections, growth failure, and in severe cases, mortality Management of diaphragmatic paresis due to phrenic nerve palsy is controversial. Both supportive and surgical interventions, such as diaphragmatic plication, are considered in treatment. However, surgical intervention is not without risks, and evidence suggests that phrenic nerve palsy may spontaneously regress in some cases [1,2]. This report aims to highlight home CPAP as a viable conservative approach, reinforcing the importance of considering non-surgical options before pursuing surgical intervention.

Case Presentation

A late preterm female baby of 35 weeks and 3 days gestational age was born via spontaneous vaginal delivery to a primigravida mother with an Apgar score was 0, 2, 5 and 6 at the 1st, 3rd, 5th, and 10th minute respectively. The baby was severely depressed at birth which required extensive resuscitation in the form of bag and mask ventilation and chest compressions, subsequently requiring ventilatory support. On the 4th day of life (DOL), on routine Chest X-ray imaging (CXR), we noticed that the right dome of the diaphragm was significantly elevated by 2 rib spaces, thus suspecting that the respiratory failure is due to diaphragmatic palsy. The possibilities of Horner's syndrome and Erb's palsy were ruled out based on clinical examination. The baby was extubated and put on continuous positive airway pressure (CPAP). As

soon as we started tapering the CPAP, the baby started having apnea, respiratory distress, and desaturation. In light of suspected right diaphragmatic paralysis, ultrasound assessment revealed a marked reduction in right diaphragm excursion, indicative of diaphragmatic weakness. This observation accounts for the difficulties encountered during respiratory support weaning and suggests right diaphragmatic palsy as the probable etiology. Surgical intervention is indicated but the parents were not keen to opt for surgery. After consultation with the pediatric pulmonologist, the decision was made to discharge the baby on home CPAP. The infant continued receiving respiratory support via bubble CPAP until day 56 of life, during which multiple attempts were made to wean off CPAP. Despite these efforts, the baby was unable to be successfully weaned, and therefore, home CPAP was initiated at discharge. While the infant remained under monitoring in the NICU on home CPAP, the parents and home nurse received comprehensive training. This included recognition of critical warning signs, instruction in basic life support (BLS), proper use of the suction apparatus, administration of nasogastric tube feeds, and essential CPAP care and machine operation. The baby was discharged on home CPAP, set at 8/21% with a humidifier, SpO_2 99% with 21% FiO_2 room air 21%. After discharge, the baby was closely monitored through follow-up visits with both the pediatrician and pediatric pulmonologist. By day 75 of life, and following a consensus with the pediatric pulmonologist, the baby was gradually weaned off respiratory support and successfully transitioned to spontaneous breathing.

Imaging

Chest X-ray

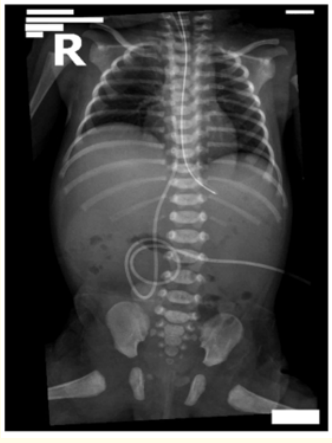

Figure 1: *Initial CXR on 1st DOL.*

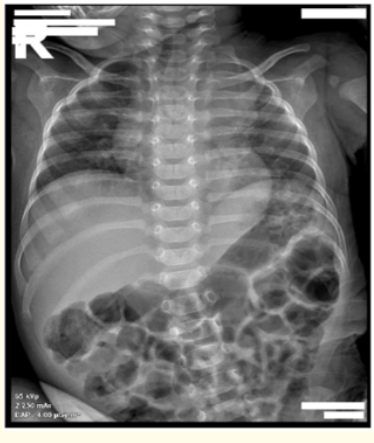

Figure 2: *Recovery after treatment.*

Discussion

The diaphragm is a dome-shaped muscle that separates the abdominal and thoracic cavities and its proper function is essential for adequate lung function. It is innervated by the phrenic nerve (C3-C5 cervical roots) on the left and right sides. Overstretching of the cervical nerves due to hyperextension or twisting and torsion of the head and neck during delivery can lead to diaphragmatic paralysis. Normally when the diaphragm contracts, it moves downwards, and a negative intrathoracic pressure is created which permits expansion of the lungs and air entry into the lungs for gas exchange [3]. Diaphragmatic paralysis prevents proper contraction of the diaphragm, disrupting normal negative pressure breathing and impairing effective lung ventilation, resulting in respiratory difficulties. In cases of unilateral diaphragmatic paralysis, the paralyzed side may exhibit paradoxical movement, rising during inspiration when the healthy side contracts. This paradoxical motion reduces the overall efficiency of breathing.

Based on the etiology, the weakness can be either temporary or permanent. In children, it can be a result of traumatic birth injury, complications of cardiothoracic surgery and neuromuscular disorders such as spinal muscular atrophy and congenital diaphragmatic muscle hypoplasia, although rare. Studies have found that other procedures performed previously have also led to diaphragmatic paralysis such as cannulation of internal jugular vein, subclavian vein, and insertion of chest tubes [4-9]. In this case, the baby was delivered via spontaneous vaginal delivery in the cephalic presentation which could have resulted in traction of the shoulder, however, unilateral paralysis of the diaphragm should be considered in cases of respiratory distress during the neonatal period, particularly in babies born after difficult delivery by breech presentation and/or forceps extraction [10,11].

The extent to which the phrenic nerve is damaged will determine the severity of the paralysis. The paralysis due to a mild degree of damage tends to recover within 3 months while in cases of severe damage, it could persist for up to 5 months and even warrant surgical treatment. The probability of diaphragmatic paralysis occurring in neonates who are in respiratory distress is increased if they have Erb's palsy however, in this case, Erb's palsy was ruled out through clinical examination and X-ray imaging [10]. One can reach a diagnosis of diaphragmatic paralysis through a combination of patient history, clinical examination findings, and imaging modalities. Although sensitive but not specific, CXR has been found to diagnose up to 90% of unilateral diaphragmatic palsy. When visualized using the frontal, inspiratory, upright, and lateral positions, the affected right hemidiaphragm should be at least two intercostal spaces higher than the left, and an affected left hemidiaphragm is one intercostal space higher than the right. If suspected based on CXR findings, it can be confirmed through fluoroscopic examination 'Snif test' [4,12,13]. Ultrasound scan is a preferred choice of investigation to confirm the diagnosis as it provides functional and dynamic images that can simplify diagnosis with the merit of being done at the bedside without exposure to ionizing radiation [14]. In infants receiving CPAP, the affected diaphragm may not be elevated or move paradoxically, hence ultrasound scan should be done while they are spontaneously breathing.

In our case, a CXR was done which revealed the elevated right dome of the diaphragm by 2 rib spaces leading to a suspicion of right diaphragmatic paralysis. Following this, a diaphragm excursion using an ultrasound scan further revealed significantly less movement of the right diaphragm indicating diaphragmatic weakness.

The management of diaphragmatic paralysis (DP) in children, particularly neonates and infants, remains largely unstandardized, with no established guidelines on indications, and timings for the interventions [2]. Standard strategies for diaphragm dysfunction generally include either conservative management such as prolonged mechanical ventilation with gradual weaning or surgical intervention via diaphragm plication or thoracotomy. Both CPAP and surgical plication are to stabilize the diaphragm and increase functional residual capacity [15]. The cases with mild degrees of paralysis have responded to supplemental oxygen via nasal cannula but more severely affected infants would require mechanical ventilation. However, in comparatively less severe cases, the use of CPAP reduces the paradoxical motion of the diaphragm thus allowing the infant to breathe spontaneously. One such case report found that an infant with bilateral diaphragmatic paralysis caused by birth injury was successfully treated with nasal CPAP, which was discontinued after 23rd DOL

upon the infant's improvement while another report revealed that nasal CPAP treated phrenic nerve palsy in an infant [16,17]. Surgical intervention too has previously shown rapid improvement in uncomplicated cases within a week of surgery.

In our care plan, we implemented home CPAP at discharge to avoid surgical intervention, as the neonate was deemed unfit for surgery and required continued respiratory support. This conservative approach with home CPAP proved effective, with the infant being successfully weaned off all respiratory support by 2.5 months of age This outcome demonstrates that non-surgical management can be effective in some cases of diaphragmatic paralysis, potentially avoiding the risks associated with surgical intervention. Our approach of home CPAP is a non-invasive intervention that can be tried to give the diaphragm time to improve and the surgery can either be delayed or avoided altogether.

Conclusion

In conclusion, the management of diaphragmatic paralysis in neonates and infants remains an unstandardized area of care. While traditional approaches have included prolonged mechanical ventilation or surgical intervention, our case demonstrates the potential effectiveness of a conservative, non-invasive approach using home CPAP [18]. With this case, we propose that home CPAP should be considered as a potential management option in particularly in cases where surgery is not immediately feasible or desirable. This can either facilitate eventual surgical intervention or potentially avoid it altogether.

Conflict of Interests

The authors declare that they have no competing interests.

Financial Disclosure

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Bibliography

1. Murty VSS., *et al*. "Phrenic nerve palsy: a rare cause of respiratory distress in newborn". *Journal of Pediatric Neurosciences* 7.3 (2012): 225-227.

2. Kadraoui Romaissa., *et al*. "Phrenic nerve palsy: a rare cause of respiratory distress in newborn". *International Journal of Advanced Research* 11.9 (2023): 577-581.

3. O'Toole SM and J Kramer. "Unilateral diaphragmatic paralysis". StatPearls (2023).

4. Kokatnur L., *et al*. "Diaphragm disorders". National Center for Biotechnology Information (2024).

5. Rizeq Yazan K., *et al*. "Diaphragmatic paralysis after phrenic nerve injury in newborns". *Journal of Pediatric Surgery* 55.2 (2020): 240-244.

6. Van Smith C., *et al*. "Minimally invasive diaphragm plication in an infant". *The Annals of Thoracic Surgery* 65.3 (1998): 842-844.

7. Pleasure JR and VL Shashikumar. "Phrenic nerve damage in the tiny infant during vein cannulation for parenteral nutrition". *American Journal of Perinatology* 7.2 (1990): 136-138.

8. D'Netto MA., *et al*. "Unilateral diaphragmatic palsy in association with a subclavian vein thrombus in a very-low-birth-weight infant". *American Journal of Perinatology* 18.8 (2001): 459-464.

9. Arya H., *et al*. "Neonatal diaphragmatic paralysis caused by chest drains". *Archives of Disease in Childhood* 66.4 (1991): 441-442.

10. Anagnostakis D., *et al*. "Diaphragmatic paralysis in the newborn". *Archives of Disease in Childhood* 48.12 (1973): 977-979.

11. Volpe JJ. "Injuries of extracranial, cranial, intracranial, spinal cord, and peripheral nervous system structures". Neurology of the Newborn, 4th edition, W.B. Saunders (2001): 813.

12. Alexander C. "Diaphragm movements and the diagnosis of diaphragmatic paralysis". *Clinical Radiology* 17.1 (1966): 79-83.

13. Greene W., *et al*. "Paralysis of the diaphragm". *American Journal of Diseases of Children* 129.12 (1975): 1402-1405.

14. Nason Laura K., *et al*. "Imaging of the diaphragm: anatomy and function". *Radiographics* 32.2 (2012): E51-E70.

15. Haller JA Jr., *et al*. "Management of diaphragmatic paralysis in infants with special emphasis on selection of patients for operative plication". *Journal of Pediatric Surgery* 14.6 (1979): 779-785.

16. Zajkowski EJ and RE Kravath. "Bilateral diaphragmatic paralysis in the newborn infant: treatment with nasal continuous positive airway pressure". *Chest* 75.3 (1979): 392-394.

17. Bucci G., *et al*. "Phrenic nerve palsy treated by continuous positive pressure breathing by nasal cannula". *Archives of Disease in Childhood* 49.3 (1974): 230-232.

18. Helvacioglu Caglar., *et al*. "Concomitant brachial plexus and phrenic nerve palsy due to birth trauma: a case report". *Medicine Science: International Medical Journal* 10.3 (2021): 1054-1056.

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