CASE REPORT

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Successful management of Congenital Diaphragmatic Hernia (CDH) in an 8-day-old infant with moderate persistent pulmonary hypertension, moderate muscular Ventricular Septal Defect (VSD), and small Patent Ductus Arteriosus (PDA)

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ABSTRACT

Background: Congenital diaphragmatic hernia (CDH) is a rare, complex and severe abnormality in an infant. Globally, neonatal mortality as a consequence of congenital anomalies is increasing and is therefore outlined as an emerging priority to be addressed by the UN Sustainable Development Goals (SDGs) in the post-2015 child health agenda. Although numerous cases are discovered prenatally or in the early postnatal period, 5-25% of CDH are detected in delayed onset after birth. The aim of this article was to discuss the case of CDH with moderate persistent pulmonary hypertension, moderate muscular ventricular septal defect, and small patent ductus arteriosus.

Case Presentation: A-8 days old, 3,060 g, a full-term male infant was referred to our tertiary hospital with transient tachypnea of the newborn and a suspected case of congenital heart disease. The defect was repaired after stabilization of the cardiac output and gas exchange. During recovery after surgery, the patient was placed on high-frequency oscillation after surgery and was given potent inotropic support. The patient made an uneventful postoperative recovery.

Conclusion: The management of CDH patients should be multidisciplinary. More clinical studies were required to elaborate on a suitable management protocol for the CDH.

Keywords: *CDH, high-frequency oscillation, persistent pulmonary hypertension, SDGs*.
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INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a rare but complex and severe abnormality with a prevalence of 2-4 infants of 10,000 pregnancies.[1,2] Globally, mortality in children aged under 5 years has halved since 1990, mainly because of a sharp reduction in deaths from communicable diseases as a result of targeted child health strategies. Following this worldwide reduction, the relative contribution of congenital anomalies to neonatal mortality is increasing globally and is therefore outlined as an emerging priority to be addressed by the UN Sustainable Development Goals (SDGs) in the post-2015 child health agenda.

CDH is the defect in the diaphragm that allows the abdominal viscera to protrude into the thoracic cavity. The protrusion occurs more commonly in the left hemithorax (85-90% of cases), probably due to early closure of the right pleuroperitoneal opening. While many cases are discovered prenatally or during the immediate postnatal period, 5 to 25% of CDH can be diagnosed long after birth.[3,4] One of the CDH classifications is Bochdaleck hernias which results from the inadequate obliteration of the lumbar elements in the pleuroperitoneal area during the eighth and tenth weeks of intrauterine development.[5] Delay in the diagnosis of CDH may result in increased morbidity. However, there is no consensus on the absolute indications for surgery and the timing. The onset of complications carries the highest mortality and morbidity rates.[6,7] We present a case of such an example to increase the alertness of clinicians in diagnosing and managing late-presenting CDH.

CASE PRESENTATION

A-8 days old, 3,060 g, a full-term male infant was referred to our tertiary hospital with transient tachypnea of the newborn and suspected case of congenital heart disease. He was delivered at 38 weeks of gestational age by cesarean section due to premature rupture of the membrane. His mother has complete prenatal care. However, the ultrasonographic examination was not available.

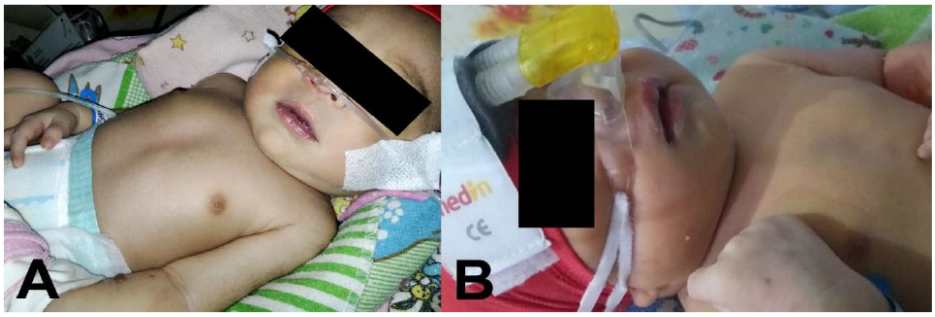

Figure 1. **A** and **B.** Scaphoid abdomen with subcostal and intercostal retractions.

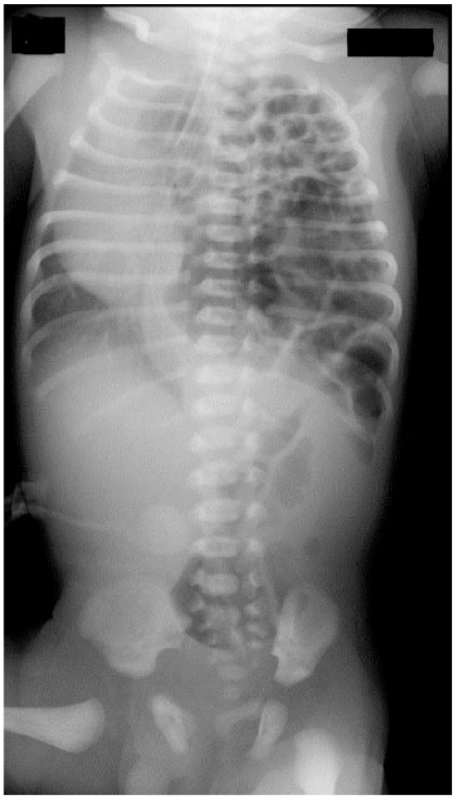

Figure 2. Babygram showed Congenital Pulmonary Airway Malformation (CPAM).

He suffered from respiratory distress with a respiratory rate of 80 breaths per minute, a heart rate of 160 beats per minute, and an oxygen saturation of 88% on room air. Subcostal and intercostal retractions, cyanotic extremities, and decreased breath sounds compared with bowel sounds on the left side of the chest were noted (Figure 1).

The babygram showed signs of congenital pulmonary airway malformation (Figure 2).

Echocardiography examination showed Moderate Persistent Pulmonary Hypertension in the Neonate (PPHN) with moderate Muscularis Ventricular Septal Defect (VSD) and small Patent Ductus Arteriosus (PDA). The patient was given Sildenafil 1.5 mg orally four times a day, dobutamine equal to 10 mcg/kg body weight/minute, and iloprost inhalation 2.5 mg every 3 hours a day. According to the examination, the patient was diagnosed with a Bochdaleq hernia.

Surgery

During exploration, we found the colon, stomach, and spleen herniated into the left hemithorax. The herniated organ was relocated into the abdomen, the defect was closed, and the chest tube (Water Sealed Drainage) was inserted. After surgery, the patient was supported with high-frequency oscillation (HFO) with setting pressure of 10 cmH2O, frequency 10 Hz, amplitude 60 and FiO2 50%, oxygen concentration and mean airway pressure were gradually decreased (Figure 3).

Babygram examination after surgery showed the mediastinum has shifted to the normal position, and the left lungs have been expanding fully to the superior lobe (Figure 4).

The breathing assistance was gradually removed according to the improvement of the patient's condition. The echocardiography evaluation showed PPHN was resolved, while the babygram evaluation showed better vascularization of the left lungs and normal bowel condition. He was discharged from the hospital uneventfully after 24 days.

DISCUSSION

Our patient present without prenatal ultrasonography result, where prenatal ultrasonography has a critical role in diagnosing CDH since 68% of CDH can be detected prenatally in either the first or second trimester using two-dimensional ultrasound (US). Meanwhile, MRI can help predict disease severity and prognosis.[8] In most cases, infants with CDH present with respiratory distress at birth or shortly afterward due to pulmonary hyperplasia, pulmonary hypertension, or both.[9] Late presentation of CDH was estimated to occur in less than 20% of all CDH cases, but 80% of delayed cases present with acute symptoms.[10] CDH is rarely found in the outpatient setting and often missdiagnosed.[11]

Other congenital abnormalities may coexist with CDH and should be suspected in CDH cases.[12] Since CDH is a complex and severe syndrome, insufficient knowledge of the pathophysiology of hypoxic respiratory failure, PPHN (Persistent Pulmonary Hypertension in the Neonate), and cardiac dysfunction and its inadequate management may cause sudden worsening.[2] Various pulmonary vasodilators have been investigated to overcome pulmonary hypertension in neonatal CDH. Inhaled nitric oxide (iNO) is recommended for confirmed supra systemic pulmonary arterial hypertension with preserved left-ventricular function.[13] Sildenafil may be considered as an alternative or adjunctive therapy.[14,15] Milrinone is indicated for cardiac dysfunction associated with pulmonary hypertension.[16] Prostaglandin E1 may be used to maintain or restore ductus arteriosus patency, thus reducing right ventricular afterload in patients with pulmonary hypertension and right-ventricular failure.[17]

The CDH surgery consists of abdominal viscera repositioning and diaphragm closure.[10] Rather than emergency surgical repair in the early neonatal phase, stabilization of the vital function is more crucial.[18,19] Unstable respiratory function increases the mortality rate during surgery, while stable vital functions may delay the surgery up to 7-10 days.[20] A consensus stated that an open surgical procedure is preferable for the minimally invasive (MIS) approach.[21] Minimal access surgery (MIS) is a popular open procedure (thoracotomy or laparotomy) that is preferable for a better cosmetic result.[22,23] However, CDH neonates who had MIS repair have a higher risk of recurrence.[24]

After surgery, the patient airway and breathing are supported by a mechanical ventilator setting High-Frequency Oscillation (HFO). CDH requires low-volume positive pressure ventilation or HFO and pulmonary vasodilatation. In most cases, due to different lung volumes and compliance, HFO is the optimal choice that allows for maintaining constant oxygen flow and continuous distending pressure with ventilation with the smallest volumes below dead space.[25]

Despite advances in surgical techniques and knowledge, the CDH mortality rate is more than 30%.[26] Prenatally diagnosed CDH is associated with higher morbidity and mortality due to larger defect size compared to the postnatally diagnosed CDH.[27,28] The most frequent post-surgical complications in subsequent months of life resulting from diaphragm dysfunction is gastro-oesophageal reflux (GER) (47%) which complicated to esophagitis, respiratory tract pathologies (bronchiolitis or pneumonia), and lung cirrhosis or complete lung dysfunction due to extreme pulmonary hypoplasia.[25]

The management of CDH patients should be multidisciplinary and consist of the pediatrician, pediatric surgeon, and anesthesiologist. Clinicians should consider the combination of the available evidence, expert consensus, practical diagnosis and treatment with HFO, proper surgical strategy, and adequate postoperative care to decrease mortality and morbidity in a patient with CDH. These case reports showed successful treatment of CDH with multidisciplinary management, but the limitation of case studies is that they cannot lead to conclusions regarding causality and may be atypical of the larger population. More clinical studies were required to elaborate on a suitable management protocol for the CDH.

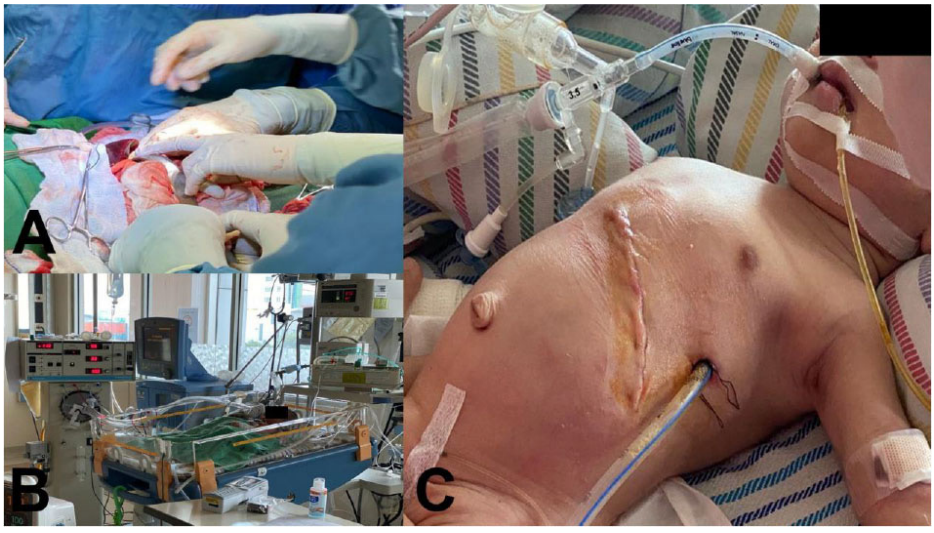

Figure 3. **A)** CDH operation procedure; **B)** HFO ventilation after surgery; and **C)** chest tube insertion after defect closure.

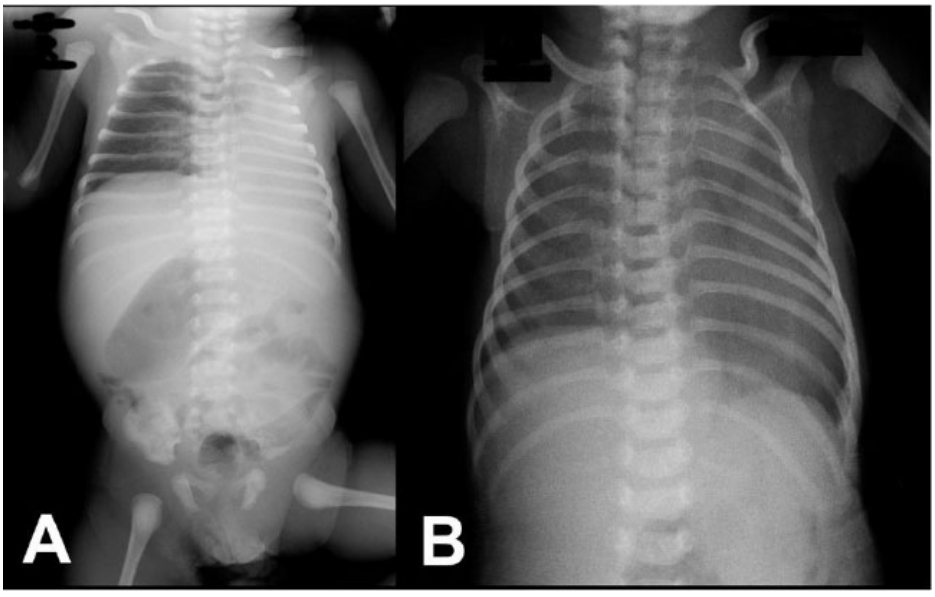

Figure 4. **A)** Babygram after defect closure, and: **B)** Babygram before discharged.

CONCLUSION

The management of CDH patients should be multidisciplinary, including the pediatrician, pediatric surgeon, and anesthesiologist. Clinicians should consider the combination of the available evidence, expert consensus, practical diagnosis and treatment with HFO, proper surgical strategy, and adequate postoperative care to decrease mortality and morbidity in a patient with CDH. More clinical studies were required to elaborate on a suitable management protocol for the CDH.

CONFLICT OF INTEREST

The authors affirm no conflict of interest in this study.

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AUTHOR CONTRIBUTION

All authors contributed to this research, starting from planning to the preparation of the manuscript publication.

REFERENCES

1. McGivern MR, Best KE, Rankin J, et al. Epidemiology of congenital diaphragmatic hernia in Europe: a register-based study. Arch Dis Child Fetal Neonatal Ed. 2015;100(2):F137-F144. doi:10.1136/archdischild-2014-306174.
2. Lakshminrusimha S, Vali P. Congenital diaphragmatic hernia: 25 years of shared knowledge; what about survival?. J Pediatr (Rio J). 2020;96(5):527-532. doi:10.1016/j.jped.2019.10.002.
3. Lava JR, Hettwer GA, Reginatto CJ, et al. Congenital diaphragmatic Bochdaleck hernia: case report. Int Arch Med. 2012;5(1):30. Published 2012 Oct 30. doi:10.1186/1755-7682-5-30.

4. Ghabisha S, Ahmed F, Al-Wageeh S, et al. Delayed presentation of congenital diaphragmatic hernia: a case report. Pan Afr Med J. 2021;40:242. doi:10.11604/pamj.2021.40.242.32113.
5. Chiu PP, Langer JC. Surgical conditions of the diaphragm: posterior diaphragmatic hernias in infants. Thorac Surg Clin. 2009;19(4):451-461. doi:10.1016/j.thorsurg.2009.08.009.
6. Antoniou SA, Pointner R, Granderath FA, Köckerling F. The Use of Biological Meshes in Diaphragmatic Defects - An Evidence-Based Review of the Literature. Front Surg. 2015;2:56. doi:10.3389/fsurg.2015.00056.
7. Testini M, Girardi A, Isernia RM, et al. Emergency surgery due to diaphragmatic hernia: case series and review [published correction appears in World J Emerg Surg. 2019 Oct 17;14:48]. World J Emerg Surg. 2017;12:23. doi:10.1186/s13017-017-0134-5.
8. Ruano R, Lazar DA, Cass DL, et al. Fetal lung volume and quantification of liver herniation by magnetic resonance imaging in isolated congenital diaphragmatic hernia. Ultrasound Obstet Gynecol. 2014;43(6):662-669. doi:10.1002/uog.13223.
9. Hon KL, Fung RCM, Leung AKC. Delayed Presentation of Congenital Diaphragmatic Hernia with Acute Respiratory Distress: Challenges in Diagnosis and Management. Case Rep Pediatr. 2020;2020:6109487. doi:10.1155/2020/6109487.
10. Spinelli C, Ghionzoli M, Costanzo S, Pistolesi F, Massart F. Acute respiratory distress for late-presenting congenital diaphragmatic hernia. Turk J Pediatr. 2008;50(5):480-484.
11. Rouse C, Schmidt L, Brock L, Fagiana A. Congenital Diaphragmatic Hernia Presenting in a 7-Day-Old Infant. Case Rep Emerg Med. 2017;2017:9175710. doi:10.1155/2017/9175710
12. Chandrasekharan PK, Rawat M, Madappa R, Rothstein DH, Lakshminrusimha S. Congenital Diaphragmatic hernia - a review. Matern Health Neonatol Perinatol. 2017;3:6. Published 2017 Mar 11. doi:10.1186/s40748-017-0045-1.
13. Canadian Congenital Diaphragmatic Hernia Collaborative, Puligandla PS, Skarsgard ED, et al. Diagnosis and management of congenital diaphragmatic hernia: a clinical practice guideline. CMAJ. 2018;190(4):E103-E112. doi:10.1503/cmaj.170206.
14. Noori S, Friedlich P, Wong P, Garingo A, Seri I. Cardiovascular effects of sildenafil in neonates and infants with congenital diaphragmatic hernia and pulmonary hypertension. Neonatology. 2007;91(2):92-100. doi:10.1159/000097125.
15. Uslu S, Kumtepe S, Bulbul A, Comert S, Bolat F, Nuhoglu A. A comparison of magnesium sulphate and sildenafil in the treatment of the newborns with persistent pulmonary hypertension: a randomized controlled trial. J Trop Pediatr. 2011;57(4):245-250. doi:10.1093/tropej/fmq091.
16. Patel N. Use of milrinone to treat cardiac dysfunction in infants with pulmonary hypertension secondary to congenital diaphragmatic hernia: a review of six patients. Neonatology. 2012;102(2):130-136. doi:10.1159/000339108.
17. Lawrence KM, Berger K, Herkert L, et al. Use of prostaglandin E1 to treat pulmonary hypertension in congenital diaphragmatic hernia. J Pediatr Surg. 2019;54(1):55-59. doi:10.1016/j.jpedsurg.2018.10.039.
18. Chatziioannidis I, Mouravas V, Babatseva E. When is the appropriate timing of surgical repair for congenital diaphragmatic hernia?. Hippokratia. 2014;18(4):381.
19. Gaiduchevici AE, Cîrstoveanu CG, Socea B, et al. Neonatal intensive care unit on-site surgery for congenital diaphragmatic hernia. Exp Ther Med. 2022;23(6):436. doi:10.3892/etm.2022.11363.
20. Moga M, Anastasiu C, Arvătescu C, Mironescu A. Congenital diaphragmatic hernia - A case report. Archives of the Balkan Medical Union. 2013;48(4):431-434. doi:10.18535/jmscr/v6i9.21.
21. Kirby E, Keijzer R. Congenital diaphragmatic hernia: current management strategies from antenatal diagnosis to long-term follow-up. Pediatr Surg Int. 2020;36(4):415-429. doi:10.1007/s00383-020-04625-z.
22. Vijfhuize S, Deden AC, Costerus SA, Sloots CE, Wijnen RM. Minimal access surgery for repair of congenital diaphragmatic hernia: is it advantageous?--An open review. Eur J Pediatr Surg. 2012;22(5):364-373. doi:10.1055/s-0032-1329532.
23. Zani A, Zani-Ruttenstock E, Pierro A. Advances in the surgical approach to congenital diaphragmatic hernia. Semin Fetal Neonatal Med. 2014;19(6):364-369. doi:10.1016/j.siny.2014.09.002.
24. Putnam LR, Tsao K, Lally KP, et al. Minimally Invasive vs Open Congenital Diaphragmatic Hernia Repair: Is There a Superior Approach?. J Am Coll Surg. 2017;224(4):416-422. doi:10.1016/j.jamcollsurg.2016.12.050.
25. Kociszewska-Najman B, Rawicz M, Pietrzak B, et al. Successful complex management of a congenital diaphragmatic hernia--a case report. Ginekol Pol. 2011;82(10):788-791.
26. Kim DJ, Chung JH. Late-presenting congenital diaphragmatic hernia in children: the experience of single institution in Korea. Yonsei Med J. 2013;54(5):1143-1148. doi:10.3349/ymj.2013.54.5.1143.
27. Colvin J, Bower C, Dickinson JE, Sokol J. Outcomes of congenital diaphragmatic hernia: a population-based study in Western Australia [published correction appears in Pediatrics. 2006 May;117(5):1870]. Pediatrics. 2005;116(3):e356-e363. doi:10.1542/peds.2004-2845.
28. Burgos CM, Frenckner B, Luco M, et al. Prenatally versus postnatally diagnosed congenital diaphragmatic hernia - Side, stage, and outcome. J Pediatr Surg. 2019;54(4):651-655. doi:10.1016/j.jpedsurg.2018.04.008.