Case Report

A Rare Presentation of Bilateral Bochdaleck Congenital Diaphragmatic Hernia, a Case Study

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Abstract

Introduction
Congential diaphragmatic hernias (CDH) are a rare process that have a prevalence of 1–4 cases per 10,000 live births, and of these, bilateral Bochdalek congenital diaphragmatic hernias are even rarer entities that occur in about 1% of cases. This finding is rarely documented in literature. This paper offers a detailed methodical timeline documentation of the events that occurred after delivery and includes how to approach the diagnosis and management of such a rare and volatile condition.

Presentation
The neonate was born at 35 weeks and 6 days gestation. Unfortunately, despite intensive cardiopulmonary resuscitation efforts, the neonate expired within less than one hour after delivery. Diagnosis of a bilateral diaphragmatic Bochdalek hernia was confirmed during the autopsy report and was evidenced by chest x-rays and clinical course. The autopsy revealed pulmonary aplasia/hypoplasia with weight being 12.8% of normal lung weight. Hepatomegaly, splenomegaly, nephromegaly and abdominal contents were in the bilateral pleural cavities.

Conclusion
This case report can be used to guide health care practitioners who come across neonates presenting with possible signs and symptoms of a rare but severe case of bilateral CDH. Management with enhanced prenatal care and hospital resources is needed to improve the chances of successful resuscitation in a neonate that presents with bilateral CDH.

Keywords
neonate; diaphragmatic hernias; congenital diaphragmatic hernias; congenital abnormalities; internal hernia; Bochdaleck hernia; diagnosis

Background
A congenital diaphragmatic hernia (CDH) is defined as an anatomical defect in the diaphragm, which allows for the protrusion of abdominal contents and potential cardiopulmonary complications in a neonate.[1] Bochdalek hernias specifically occur posteriorly due to inadequate obliteration of lumbar elements in the pleuroperitoneal area during weeks eight to ten of intrauterine development.[2] The prevalence of CDH is approximately 1–4 cases per 10,000 live births.[3,4] Approximately 95% of CDHs are classified as Bochdalek with mortality rates ranging from 40% to 80%.[5,6] Classification is based on location of the diaphragmatic hernia. Other types of CDHs include anterior defects or Morgagni hernias and, less frequently, central hernias.[7]

An international cohort study conducted between 1995 and 2015 involving 80 patients with bilateral CDH reported a high mortality rate of 74%.[8] All survivors were treated with primary or patch repair along with 19 needing extra-corpeal membrane oxygenation (ECMO). The management of infants with bilateral CDH remains a difficult problem with significant mortality. In addition, bilateral CDH is associated more frequently with other major anomalies

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such as neural tube and cardiac defects, when compared to unilateral CDH, with a prevalence as high as 56%.[9]

Case Presentation

We present a 26-year-old African American female, gravida two para one, with a history of adequate prenatal care. This included prenatal ultrasound imaging, which was reported as normal. The presumption was that the liver was confused for lung tissue. Neonatology was only consulted on this case upon time of delivery, and the report was received from the obstetrician-gynecologist. The only pregnancy complication was pregnancy-induced hypertension (PIH), which was treated with methyldopa. Labor was preterm at 35 weeks and 6 days gestation along with delivery complications of nuchal cord and meconium-stained amniotic fluid. Based on the above perinatal history, the neonatal resuscitation team was in attendance at the delivery. Below we provide a timeline of events:

11:48 PM Neonatology was called to the delivery at approximately five minutes of life. Upon arrival, the infant was already intubated and ventilation attempts were being performed with very little to no chest movement.

11:54 PM The infant was bradycardic with a heart rate of 60–80 beats per minute. Chest compressions were initiated. Due to poor chest excursion, the infant was re-intubated, with vocal cords easily visualized. Regardless of re-intubation, chest excursions were not able to be obtained. Bilateral trans-illumination of the chest was performed at this time with no evidence of pneumothorax. The decision was made to change the ventilation mechanism from the Neopuff to an Ambu bag. However, this change was not effective.

11:58 PM First dose of epinephrine was administered via the endotracheal (ET) tube. Meconium aspiration was attempted via a meconium aspirator in the case of a mechanical obstruction due to a meconium plug. However, this maneuver to potentially unplug the airway of a meconium plug was unsuccessful.

12:03 AM An umbilical venous catheter (UVC) line was inserted at 8 cm.

12:04 AM A second dose of epinephrine was given via the UVC line.

12:06 AM There was an attempt to dilute a potential meconium plug with normal saline, which was unsuccessful. Bilateral trans-illumination of the chest was performed again at this time and showed no evidence of a pneumothorax.

12:08 AM The patient was re-intubated again with a larger-sized ET tube (4.0). However, this intubation did not yield positive chest movement or improveme the patient's oxygenation status.

12:13 AM The neonate was extubated.

12:14 AM Bagging was initiated via face mask with successful chest expansion, but no improvement of oxygen saturations was achieved.

12:17 AM Bilateral trans-illumination was again performed with no evidence of a pneumothorax. Reintubation was attempted using a size 3.0 ET tube, which was advanced into the right mainstem bronchus in order to illicit any form of ventilation response, but it was unsuccessful. The tube was also pulled just below the vocal cords in an unsuccessful attempt to ventilate. The ET tube was then pulled back and placed into the infant's esophagus. This different placement of the tube seemed to produce better chest expansion.

12:19 AM The infant was extubated again with bagging reinitiated. Bilateral trans-illumination was used to verify the absence of a pneumothorax.

12:22 AM A third dose of epinephrine and a fourth re-intubation was performed with a 3.5-sized ET tube. However, this action was also unsuccessful.

12:25 AM The neonate was extubated and bagging was again restarted.

12:26 AM A chest x-ray was obtained to document the ET placement and revealed no signs of a pneumothorax. The chest x-ray showed no aeration of the lungs and a gaseous mass located

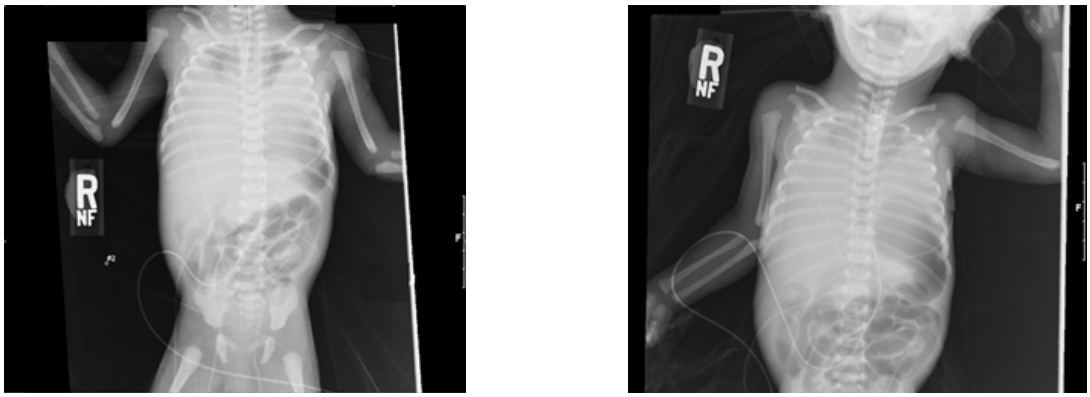

Figure 1. Anterior-Posterior Babygram indicating ET placement. Report included no signs of pneumothorax, no aeration of the lungs, and a gaseous mass located in the left lower hemithorax.

in the left lower hemithorax. (**Figures 1A and 1B**) Eventually, the neonate expired despite resuscitative efforts. Time of death was 12:26 AM.

Discussion

The clinical profile of respiratory failure in our case report showed the inability to ventilate due to lung underdevelopment. The only cue could have been the physical examination findings of an excavated abdomen, displaced heart sounds and decreased respiratory lung sounds with the findings of bowel sounds in the chest. However, these findings are very hard to elicit during resuscitation.[10] At the time, we did not obtain any prenatal history or prenatal sonograms. Bilateral CDH could potentially be diagnosed prenatally via color and power Doppler, demonstrating hepatic vessel within the cardiothoracic space.[11] Diagnosis in the prenatal period leads to a better prognosis, in addition to the delivery occurring in a center well equipped to make necessary interventions, thereby minimizing neonatal complications and optimizing the chances of survival. However, in this severe case of lung aplasia/hypoplasia (autopsy results of combined weight of 4.7 grams, being 12.8% of normal weight) it would likely not have made a difference. During resuscitation, the possibility of tracheal obstruction with meconium or tracheal agenesis with a broncho-esophageal fistula was considered due to the lack of chest expansion despite proper ET tube placement. Later, it was determined that the chest excursion elicited with mask bag ventilation and an endotracheal tube in the esophagus was a direct result of the stomach being inflated and deflated in the left hemithorax. In this case, the bilateral CDH allowed for the migration of abdominal contents into the thoracic cavity, compressing the lungs bilaterally and causing severe pulmonary hypoplasia untenable for extra-uterine life.

The autopsy reported bilateral diaphragmatic hernias with both pleural cavities filled with large portions of the liver, spleen and the majority of the stomach in the left pleural cavity. (**Figures 2A and 2B**) During the autopsy, it was found that the neonate's lungs weighed 12.8% of normal weight, implying significant pulmonary compression. The diagnosis of a bilateral diaphragmatic hernia was only made during the autopsy.

It is noted that the increased degree of compression from the abdominal organs against the lungs caused a direct loss of pulmonary vasculature and bronchioles, thus the presentation of severe pulmonary hypoplasia.[7] Subsequent to the loss of pulmonary vasculature, a neonate may develop persistent pulmonary hypertension (PPHN), which can further result in cardiac dysfunction, with the neonate maintaining fetal circulation to the lungs via blood flow through the foramen ovale and ductus arteriosus.[12] Right ventricular hypertrophy and left ventricular hypoplasia, which are often times found to be more severe in PPHN associated with CHD compared to other causes, can also occur as a result of this right-to-left shunt. The aforementioned changes to physiology ultimately present with a severe case of cyanosis and decreased pulmonary compliance.[13] Postnatal management of CDH consists of stabilization of the neonate's oxygenation,

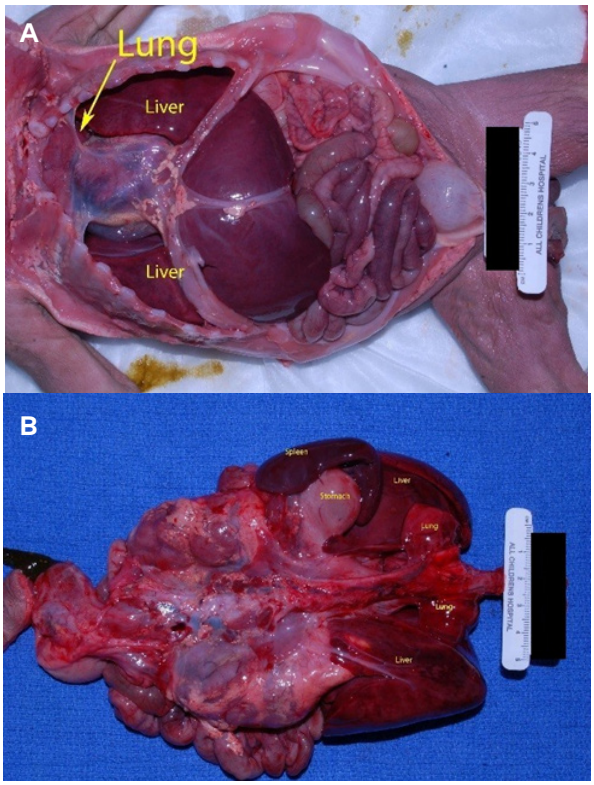

Figure 2. A) Autopsy results indicating underdevelopment of bilateral lungs, with bilateral diaphragmatic hernias and both pleural cavities filled with large portions of the liver, spleen and stomach. B) Contents within pleural cavities, labeled spleen, stomach, liver and lungs. The neonate had severe pulmonary aplasia/hypoplasia with the combined weight of the lungs at 4.7 grams (normal combined weight being 36.7 grams), hepatosplenomegaly with liver weight of 138.5 grams (normal weight being 82 grams), splenomegaly with spleen weight of 8.3 grams (normal weight being 6.5 grams), nephromegaly, combined weight of lungs at 36 grams (normal combined weight being 18.1 grams), cerebral edema, diffused lymphadenopathy, unicornuate uterus and blind-ending proximal fallopian tube.

blood pressure and acid base status, as acidosis and hypoxia can worsen pulmonary hypertension. Blood pressure should be adequately maintained so as not to worsen the right-to-left shunt and subsequent hypoxia. Definitive management must involve reduction of the abdominal viscera and surgical repair of the diaphragmatic defect.

Conclusion

This case report can be used to guide health care practitioners who come across neonates presenting with possible signs and symptoms of the rare but severe case of bilateral CDH. Despite our cardiopulmonary resuscitative efforts, the neonate expired. Management with enhanced prenatal care and hospital resources is needed to improve the chances of successful resuscitation in a neonate that presents with bilateral CDH.

Conflicts of Interest

The authors declare they have no conflicts of interest.

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