

# Budd-Chiari-like syndrome presenting with hydrothorax in a neonate with right diaphragmatic hernia

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## ABSTRACT

The diagnosis of congenital diaphragmatic hernia (CDH) is usually straightforward. However, cases with right-sided CDH can be challenging. We report a case of a neonate with right-sided diaphragmatic hernia presenting with hydrothorax.

**Keywords:** Budd-Chiari syndrome, diaphragmatic hernia, hydrothorax, neonate.

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## INTRODUCTION

Congenital diaphragmatic hernia (CDH) is an anomaly characterized by defective development of the diaphragm.<sup>[1]</sup> Ninety-two percent of CDHs are postero-lateral defects, and 80% of these seem to have a left-sided preponderance.<sup>[2]</sup> Symptoms of right-sided CDH are less severe and frequently inconspicuous at presentation. The association of right-sided CDH with hydrothorax is not an ordinary diagnosis, but it must be kept in mind. The mechanisms responsible for the cases presented with hydrothorax are obstruction of the superior vena cava and hepatic veins, respectively.

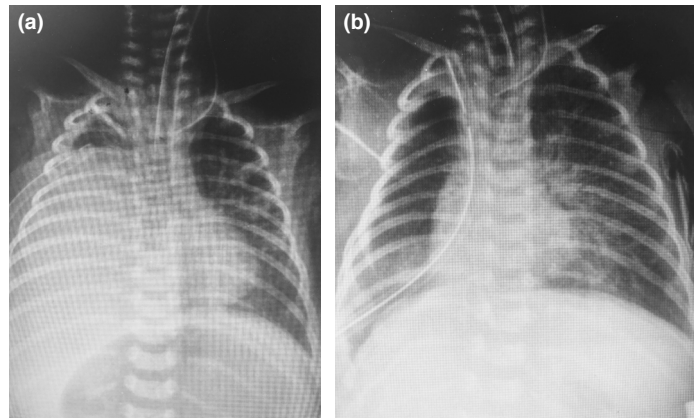
## CASE REPORT

A 1610 g male infant was born at 34 weeks of gestational age due to placental abruption. Prenatal ultrasound had revealed right-sided pulmonary hypoplasia. He required respiratory support with high-frequency oscillatory ventilation. A subsequent chest X-ray demonstrated opacification of the right hemithorax (Fig. 1a). Hydrothorax and atelectatic pulmonary parenchyma were seen on the right side with thorax ultrasound, and the liver was situated in an upper than normal position. A tube thoracostomy was carried out with the anticipation that draining the hydrothorax would relieve atelectasis in the lung. Pleural fluid rich in protein was drained via tube thoracostomy. An echocardiogram demonstrated tetralogy of Fallot. The right lobe of the liver was seen in the right hemithorax in computed tomography of the thorax. Following this finding, surgery was performed on the third day after birth. The diaphragmatic hernia was repaired and during the operation, the liver was found to be herniated into the right hemithorax and the three lobes of the right lung were seen (Fig. 2). After the operation, a chest X-ray demonstrated that the lungs had expanded bilaterally and there was no fluid accumulation in the right hemithorax (Fig. 1b). However, on the second day post-operation, abdominal ultrasound revealed fluid accumulation at the perihepatic area. Hepatic venous flow of the right hepatic veins was demonstrated to be partially obstructed with Doppler ultrasonographic evaluation. Budd-Chiari syndrome secondary to the displacement of the liver and liver vessels was suspected. However, this fluid accumulation did not last long and regressed spontaneously two days later. There were no operative findings of Budd-Chiari syndrome other than fluid accumulation and radiological findings. We were unable to extubate the baby due to complications associated with chronic lung disease and congenital heart disease. A tracheostomy tube was placed when he was 5 months old. He underwent surgery twice for tetralogy of Fallot. He is still being ventilated with a home ventilator and is followed up by our clinic.

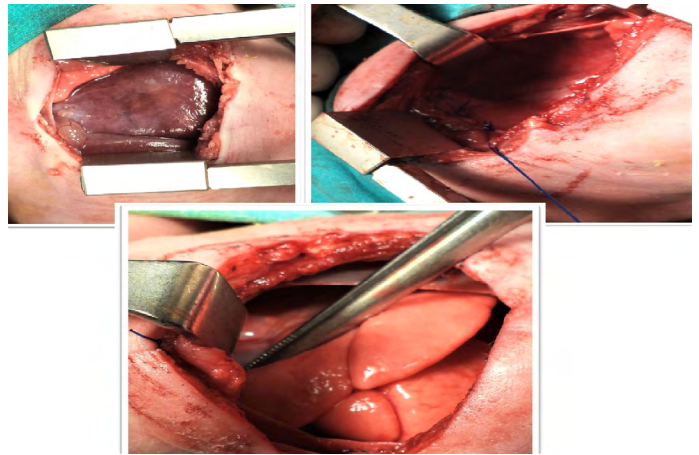
The parents of the patient were informed, and written informed consent was obtained.

## DISCUSSION

Our case described here presented with two complications—hydrothorax and ascites—both due to the right diaphragmatic hernia. Fernandez-Gonzalez et al.<sup>[3]</sup> reported a Budd-Chiari-like syndrome in two adults associated with rupture of the right diaphragm after abdominal trauma. They observed that venous outflow returned to normal after the liver was placed back into the in-



**Figure 1:** (a) Chest X-ray demonstrating opacification of the right hemithorax. (b) Chest X-ray demonstrating expanded lungs after the operation.



**Figure 2:** The liver in the right hemithorax and the three lobes of right lung was seen during operation.

traabdominal cavity. In our case, after the operation, there was no fluid accumulation in the right hemithorax anymore. Kaifi et al.<sup>[4]</sup> reported a 24-year-old woman who had a right-sided diaphragmatic hernia due to thoracoabdominal trauma. She developed irreversible Budd-Chiari syndrome as a result of right hepatic vein thrombosis and very late clinical presentation.<sup>[4]</sup> Gilsanz et al.<sup>[5]</sup> described five neonates with right-sided diaphragmatic hernia and hydrothorax in 1986 for the first time in the literature. Four of the infants with large right hydrothoraxes were found to have an incarcerated peritoneal sac with fluid. One of them had ascites. Distinctively, in our case, no peritoneal sac was seen during the operation, only the liver was found to be herniated into the right hemithorax. After the operation, fluid accumulation revealed by abdominal ultrasound lasted for a few days. The fluid was rich in protein content. These manifestations directed us to the diagnosis of a Budd-Chiari-like syndrome due to the right-sided CDH. Since Gilsanz's publication about this serious clinical entity, no case reports had been published regarding neonates.

In cases with hydrothorax without a clearly described etiology, the diagnosis of this entity should be kept in mind. Knowledge about the possibility of the co-occurrence of right-sided diaphragmatic hernia and hydrothorax is important for appropriate treatment.

## Statement

**Author Contributions:** Concept – AC, DA; Design – DA, AC; Supervision – GK, HA; Resource – AC, HFO; Data Collection and/or Processing – TG, KP; Analysis and/or Interpretation – AT; Literature Search – ST; Writing – DA, ST; Critical Reviews – AC, GK.

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