Hydrops Fetalis due to Direct Hepatic Compression of the Right Atrium in a Newborn with Right Congenital Diaphragmatic Hernia

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Abstract

Right sided Congenital Diaphragmatic Hernia (CDH) has been previously associated with venous or lymphatic obstruction of vessels by the herniated liver. We report an unusual case of direct cardiac atrial compression by the liver causing limitation of preload and resultant fetal non-immune hydrops fetalis.

Keywords: CDH; Congenital diaphragmatic hernia; Non-immune hydrops

Case Report

A 28 year old, G_3P_2 mother with an unremarkable pregnancy presented to a rural hospital with premature rupture of membranes at 36 weeks gestation. Breech presentation was noted on ultrasound, so referral to an urban tertiary care hospital for expectant delivery by C-section was arranged. Upon arrival, a repeat ultrasound demonstrated hydrops fetalis with a right sided diaphragmatic hernia and fetal liver in the thorax.

A male infant was born with an estimated weight of 4.0 kg and aside from generalized edema there were no obvious features to suggest a syndromic form of CDH. His apgar scores were 2/1/3/3 at 1, 5, 10 and 15 minutes respectively. The venous cord gas showed a pH of 7.27, pCO_2 54 mm Hg, pO_2 20 mm Hg, HCO_3 20 mmol/L. His caseroom resuscitation included intubation, paralysis, normal saline bolus, one minute of chest compressions and a single dose of epinephrine. A chest radiograph confirmed the antenatal ultrasound findings (Figure 1). Following these events, the first umbilical arterial gas on conventional mechanical ventilation revealed significant compromise with a pH of 6.83, pCO_2 111 mm Hg, HCO_3 9 mmol/L, lactate 6.4 mmol/L and a pre-ductal oxygen saturation of 34% despite receiving 100% FiO_2.

Upon transfer to the NICU he was briefly placed on the High Frequency Jet ventilator (HFJV) but experienced severe bradycardia during transition so was quickly changed to high frequency oscillatory ventilation (HFOV). During his brief period on the HFJV he again received chest compressions, and had inhaled nitric oxide, morphine and epinephrine infusions initiated. Due to persistent hypotension, metabolic acidosis and poor perfusion he received another four 10 mL/kg boluses of NS.

Pediatric Cardiology performed an echocardiogram at 3 hours of age due to volume resistant hypotension. This revealed severe pulmonary hypertension, a hypoplastic right pulmonary artery, displacement of the heart by the herniated bowel and liver, a dysplastic aortic valve with mild to moderate aortic regurgitation as well as a large PDA with a small amount of pericardial fluid seen. What was

most unusual however was that the herniated liver was noted to be compressing the right and left atria potentially causing limitation of preload (Figure 2). Although both atria were affected the compressive effect was deemed to be greater on the right than the left.

Over the next five hours, he experienced transient improvement with his highest pre-ductal saturation being 94%. He then developed a further deterioration with respect to oxygenation and systemic systolic blood pressures consistently in the low 30s. After escalating doses of epinephrine failed to address this issue and with worsening oxygenation, his parents chose to withdraw support. He died at 13 hours of age. The family declined an autopsy and genetic testing was unavailable.
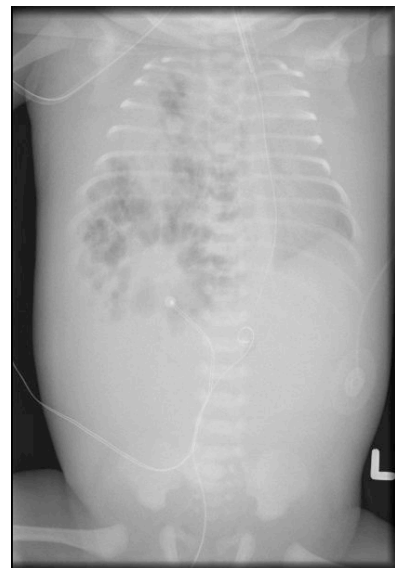
Figure 1: Radiograph demonstrating significant subcutaneous edema and left mediastinal shift by the right CDH.
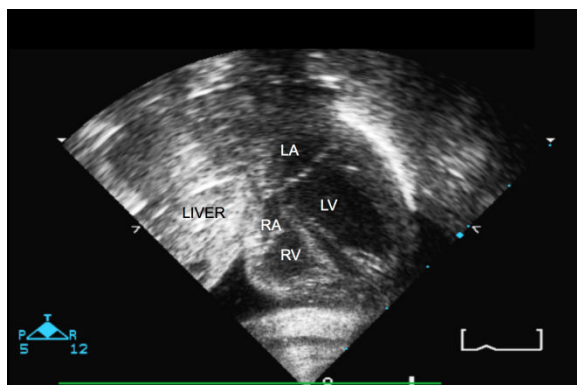
Figure 2: Significant compression of the right atrium by the herniated liver.

Discussion

The incidence of Congenital diaphragmatic hernia is between 1 in 2500 to 4000 live born infants [1,2]. The majority of patients have left sided defects with 14-25% of patients having a defect on the right and less than 1% having bilateral involvement [3,4]. In a recent modern cohort, a survival rate of 55% was reported for those with right sided defects [5]. Some reports suggest a lower survival rate for patients with right sided defects and the need for ECMO may play a role, which has been estimated to be 40% in right sided defects versus 15% in those on the left [4]. Additionally, prenatal diagnosis of right CDH is less common possibly due to liver echogenicity masking the defect [6,7]. Inability to plan for such deliveries likely contributes to both significant morbidity and increased mortality as well.

Superior vena cava syndrome may result from the presence of the liver in the fetal thorax obstructing IVC or other venous inflows [8,9]. This disturbance in flow may compromise preload culminating in biventricular compromised output. Alternatively, intrathoracic liver may be associated with impaired lymphatic flow leading to hydrothorax and lethal pulmonary hypoplasia [10].

Hydrops fetalis in late gestation fetuses with CDH appears to be a rare event. In 2002, Sydorak et al. described their experience with 175 patients referred to a single centre for CDH [11]. Of these patients, only 10 had isolated CDH with hydrops and of these 6 were right sided. Only five of these patients survived and of these, all received fetal intervention. In this series, the development of hydrops was speculated to be secondary to fetal venous compression by the herniated viscera. The liver may have been responsible for this compression as in the series described above, 9 of the 10 patients had liver in the fetal thorax.

What makes this case unique is the visualization of direct compression of the right and left atria by the liver. Although left heart hypoplasia has been described by others this was found in infants with liver herniation in the presence of a left CDH [12]. To the best of our knowledge this is the first reported case of direct hepatic atrial compression causing non-immune hydrops fetalis. We postulate that the development of hydrops in this case was attributable to limitation of venous return to the right atrium. The recalcitrant hypotension may have been impacted by limitation of pulmonary venous return as well. This would have been doubly affected by decreased output from the right ventricle and direct compression of the left atrium. This infant proved resistant to both fluid and inotropic support as evidence of this restricted systemic output. Although the largest study of CDH and hydrops to date did not find a benefit to delivery in an ECMO centre 11 we propose that if hepatic compression of the atria prior to the development of hydrops is diagnosed antenatally, consideration should be given to delivery in an ECMO centre. Furthermore the receiving team should be prepared to initiate ECMO expeditiously after delivery as a means to early surgical intervention. This combined approach could confer a survival benefit provided the gestational age was advanced enough to consider early delivery.

In conclusion, hepatic atrial compression may limit venous return of both systemic and pulmonary blood flows and should be considered as an etiology for hydrops in this population. Identification of this condition antenatally may help in planning for postnatal management.

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