A Morgagni hernia with an absent ductus venosus: An unusual case causing unusual consequences

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Abstract

A Morgagni hernia is a rare form of congenital diaphragmatic hernia (CDH), comprising only 3–5% of all CDH cases. Agenesis of the ductus venosus with direct umbilical vein blood flow to the heart is a relatively uncommon finding that is often fatal in utero. We present a case of a 2-month-old infant with Morgagni hernia and absence of the ductus venosus. These combined defects led to neovascularization of the liver, severe pulmonary hypertension and right heart failure. In this report, we describe a Morgagni hernia that’s presentation resembled that of a Bochdalek hernia likely because of concomitant absence of the ductus venosus causing severe pulmonary hypertension.

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Congenital diaphragmatic hernia (CDH) occurs in approximately 1 in 2500 births, accounting for about 8% of major congenital anomalies [1]. A Morgagni hernia is a rare form of CDH, comprising only 3–5% of all CDHs. Morgagni hernias are thought to be caused by a failure of the pars sternalis to fuse with the costal arches [2]. In infancy they are typically asymptomatic and usually present later with nonspecific symptoms [3]. The most common presentation is recurrent respiratory infection. Associated anomalies are relatively common and are usually cardiac in nature [3]. Surgical repair was previously accomplished via laparotomy or thoracotomy, but is now more frequently done through minimally invasive approaches leading to shorter hospital stays and fewer complications [4,5].

Pulmonary hypertension is commonly seen in infants with classic Bochdalek CDH, resulting in right to left shunting, hypoxemia and acute right sided heart failure in those most severely affected [6]. Agenesis of the ductus venosus associated with direct umbilical return to the heart is a relatively uncommon finding with a prevalence of 6/1000 fetal examinations [7]. These defects are associated with cardiomegaly, heart failure, hydrops fetalis and postnatal pulmonary hypertension [8–10].

Neovascularization occurs in ischemic states as a method of improving physiological function of organs with ischemic damage [11]. Neovascularization of the liver is most often seen in the literature in association with liver transplant and ischemia. Case studies involving transcapsular arterial collateralization of a liver transplant after hepatic artery occlusion have been seen adult patients [12,13] and failure of liver transplant in pediatric patients [14].

We present a case of a 2-month-old infant with Morgagni hernia and absence of the ductus venosus. These combined defects led to neovascularization of the liver, severe pulmonary hypertension and

![Fig. 1. Chest X-ray (AP) showing opacity obscuring the right medial hemidiaphragm consistent with Morgagni hernia.](image-url)
right heart failure. The authors are aware of no other presentation of Morgagni hernia associated with neovascularization and absence of the ductus venosus.

1. Case report

The patient was a 66 day old male infant who was born via Caesarian section as a result of maternal preeclampsia and breech presentation to a gravida 4 para 3 mother at 34 weeks and 2 days gestation. The pregnancy was complicated by maternal obesity and maternal insulin dependent diabetes. Prenatal imaging had revealed agenesis of the ductus venosus with umbilical vein draining into the right atrium, mild tricuspid and mitral valve regurgitation and dilation, cardiomegaly, shortened limbs, and liver calcifications at 29 weeks gestation. Apgar scores at birth were 7 and 8 at 1 min and 5 min of life. Birth weight was 3060 g. Several dysmorphic features were noted at birth, including low set ears, high arched palate, microphallus, unilateral palmar...
crease, multiple focal hemangiomas, cardiomyopathy with mildly depressed biventricular function, and hepatomegaly with fatty infiltrates. A TORCH infection work up and genetic SNP analysis were unremarkable. No other genetic analysis was completed. The patient was transferred to the NICU for management of tachypnea, pulmonary arterial hypertension, hypoxia, requiring CPAP and hypoglycemia. Follow up post-natal ultrasound confirmed prenatal diagnosis of agenesis of the ductus venosus with umbilical vein draining into the right atrium, mild tricuspid and mitral valve regurgitation and dilation, cardiomegaly, and liver calcifications.

The patient was able to be weaned down to 1.5 L of 100% FiO2 on nasal cannula by day 33 of life. The patient developed mild respiratory distress on DOL 66 with increasing oxygen requirements and positive blood cultures. The patient experienced persistent desaturation into the 70s, with rhonchi auscultated on the right side. Chest X-ray revealed an opacity obscuring the right medial hemidiaphragm contour, consistent with a Morgagni hernia containing the liver (Fig. 1). A CT of the chest was consistent with a Morgagni hernia that was occupied nearly completely by Liver (Fig. 2). Specifically, on echocardiogram, it was noted that the extension of the liver into the lower portion of the right hemithorax was causing mass effect on the adjacent heart and that there was significant pulmonary hypertension (Fig. 3).

Surgical correction was undertaken laparoscopically on day 69 of life after the pulmonary hypertension was medically optimized. The abdominal cavity was carefully examined. Significant neovascularization of the liver was noted (Fig. 4). There was a large diaphragmatic defect centrally located in the expected position based on the prior imaging. The umbilical vein connected directly to the right atrium and it was divided using bipolar electrocautery. The hernia sac was excised, avoiding entry into either the pleura or the pericardium. The suprahepatic diaphragm was mobilized away from the hepatic veins to allow for primary repair of the hernia without tension or chest wall deformity. There was no kinking of either the IVC or the hepatic veins. Blood loss was minimal. Fig. 5 shows a schematic of the defect before and after repair. Pre-operatively the patient was taking in 65% of his feeds PO. By post-operative day four, he was taking in 90% of his feeds PO. At 3 week follow-up, he was off oxygen, feeding and growing normally. Three months post-repair he was growing and developing normally without symptomatic respiratory or cardiac concerns.

2. Discussion

There have been three previous cases [7] reported in the literature of absent ductus venosus and anomalous drainage of the umbilical vein into the right atrium or coronary sinus in association with development of right sided congenital diaphragmatic hernia and dilation of the right atria and ventricle. One of these cases also involved desaturation and compression of the right atrium and ventricle with right to left shunting [7]. The authors are unaware of Morgagni hernia presenting with an absence of the venous ductus in any other published case study or review of the literature.

Extrahepatic umbilical venous drainage is associated with fetal malformations that involve significant agenesis of the fetal portal system, hepatic tumors and focal nodular hyperplasia [15].
hypothesize that the liver recruited increased arterial circulation and became hypervascular in order to compensate for the lack of portal flow resulting in the neovascularization. The ductus venosus allows a portion of the umbilical venous blood to bypass the liver and reach the main circulation more rapidly. It is able to change its diameter to adjust the distribution of blood into the heart. Absence of this structure and direct drainage of the umbilical vein into the heart causes complete liver bypass. This vascular configuration is associated with cardiac defects, high volume cardiac failure, cardiomegaly and pulmonary hypertension [9,16,17]. Thus, it likely that our patient's pulmonary hypertension was in part due to the neonatal effects of fetal hepatic bypass and an absent ductus venosus. Furthermore, the liver's lack of tethering by the falciform ligament and ductus venosus likely contributed to its ability to significantly herniate into the mediastinum. This allowed the liver to put pressure on the heart and contributed to the overall cardiac compromise. Thus, the combination of a Morgagni hernia and lack of a ductus venosus likely sets the stage for a hemodynamically significant congenital diaphragmatic hernia.

References


