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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.

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

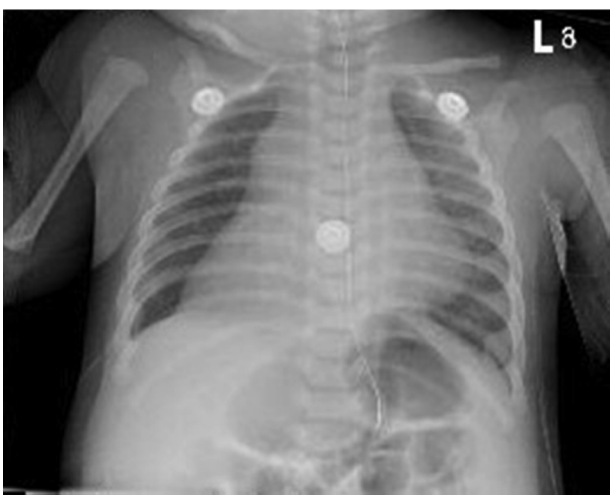

Fig. 1. Chest X-ray (AP) showing opacity obscuring the right medial hemidiaphragm consistent with Morgagni hernia.

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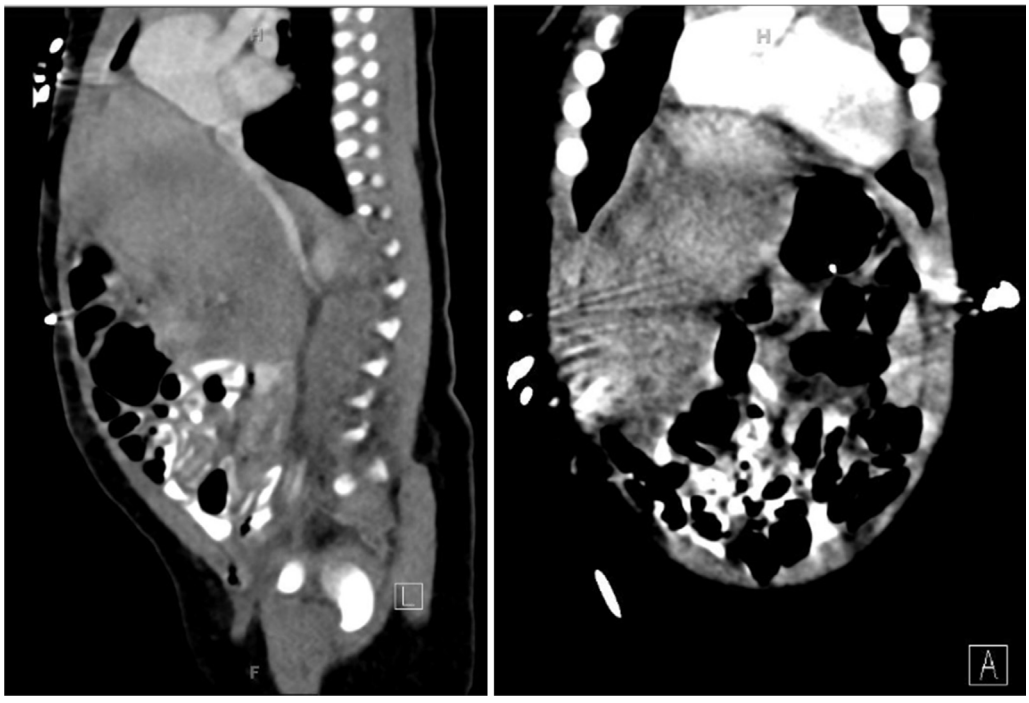

Fig. 2. Sagittal and coronal CT scans showing the Morgagni hernia.

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

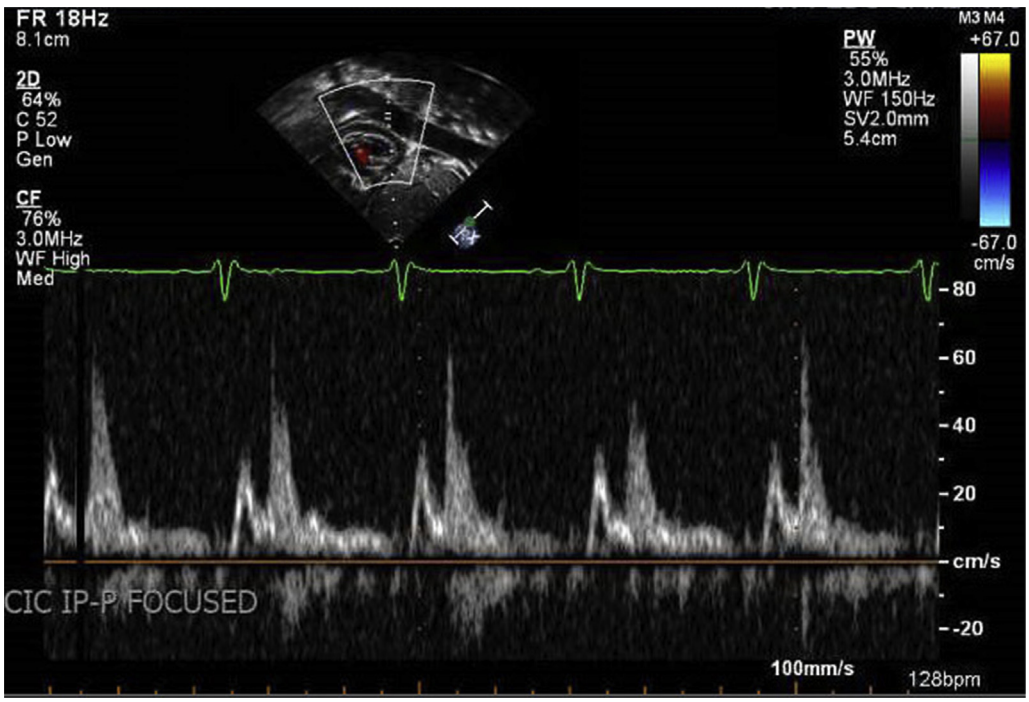

Fig. 3. Echocardiogram showing mass effect of liver on the heart.