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Images in Congenital Heart Disease

A case of large ventricular septal defect with right-sided diaphragmatic hernia

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A GIRL WAS BORN AFTER A GESTATION OF 37 weeks, displaying cyanosis and dyspnea. She was intubated immediately after birth, with radiographic findings suggestive of atelectasis of the left lung and likely congenital cardiac disease. Magnetic resonance imaging performed 14 days after birth accurately revealed a large perimembranous ventricular septal defect with a right sided diaphragmatic hernia. Both lobes of the liver, along with the right kidney, had herniated through the diaphragmatic opening into the right chest (Fig. 1). The inferior caval vein (A) coursed abnormally behind the liver before draining to the posterior aspect of the right atrium (Fig. 2). Echocardiographic studies, along

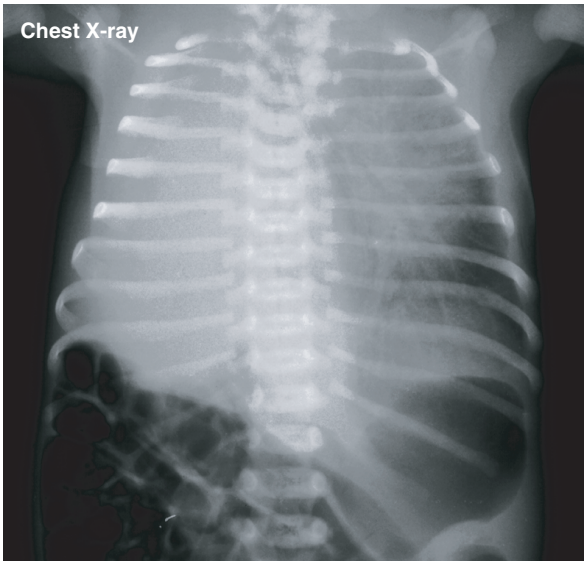

Figure 1.

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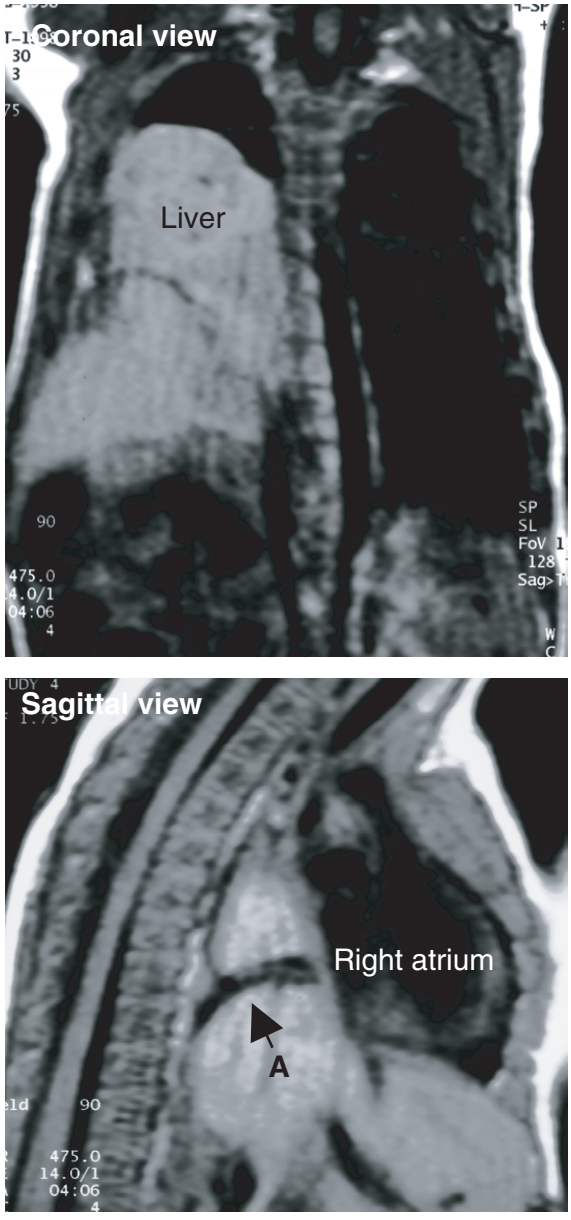

Figure 2.

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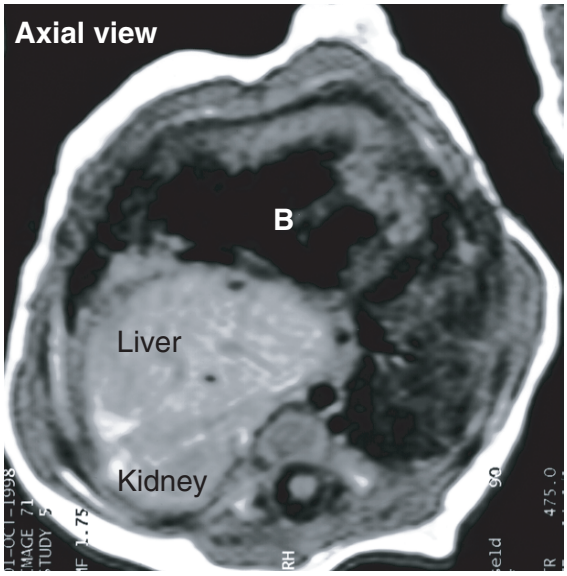

Figure 2. *continued.*

with the magnetic resonance imaging, showed that the ventricular septal defect (B) measured 18 mm in diameter (Fig. 2), and revealed severe hypoplasia of the right pulmonary artery, which was only 2 mm in diameter. A Doppler study demonstrated severe pulmonary hypertension. Despite our best attempts, it was not possible surgically to return the liver to the abdomen because of tight adhesions between the peritoneum and the epicardium. Continual high frequency artificial respiration was necessary to retain adequate oxygenation.

At the age of 1 year, she underwent closure of the ventricular septal defect, because of severe failure to thrive, and frequent pulmonary hypertensive crises. Unfortunately, she died of cardiac failure subsequent to the operative procedure.

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