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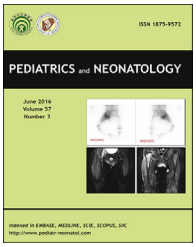

CASE REPORT

Scimitar Syndrome and H-type Tracheo-esophageal Fistula in a Newborn Infant

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Scimitar syndrome is a rare congenital anomaly characterized by partial anomalous pulmonary venous drainage of the right lung to the inferior vena cava (IVC) creating a tubular opacity paralleling the right cardiac border on chest radiography which resembles a curved Turkish sword or scimitar. Associated pulmonary and vascular anomalies have been reported in cases of Scimitar syndrome, most commonly hypoplasia of right lung, dextroposition of the heart, hypoplasia of the right pulmonary artery, and aberrant arterial supply from the descending aorta to the affected lobe of the right lung. To the best of our knowledge, this is the first case of Scimitar syndrome with an H-type tracheoesophageal fistula that has ever been reported.

1. Case Report

A male term newborn infant was born via vaginal delivery to a 22-year-old female. At birth, the infant was vigorous with spontaneous respirations and normal Apgar Scores. Besides a two-vessel umbilical cord, the infant appeared to be normal and was roomed in with the mother. At a few hours of life, a choking episode with discoloration was noted during the first bottle feed. Repeat examination revealed more prominent heart sounds on the right side of the chest. A chest x-ray revealed a cardiac silhouette that occupied right hemithorax (Figure 1). The infant was transferred to our institution for further evaluation.

Upon arrival, the infant appeared well and was breathing comfortably on room air. An echocardiogram

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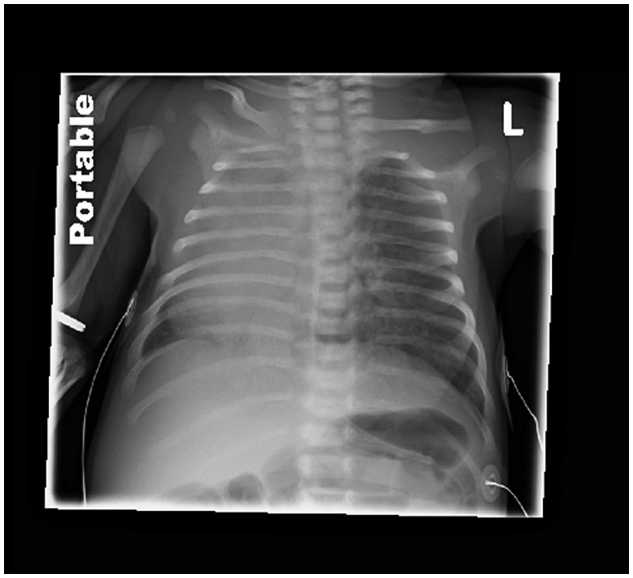

Figure 1 Chest radiography demonstrated the dextroposition of the cardiac silhouette occupies almost entire right chest.

demonstrated situs solitus, dextrocardia, concordant atrioventricular and ventricular great arterial connections, with normal drainage of the left-side pulmonary veins to the left atrium, but the connection of the right-sided pulmonary veins was not defined. In addition, there was evidence of suprasystemic pulmonary hypertension with bidirectional shunting through a 2-mm patent ductus arteriosus (PDA). The infant was started on oxygen via nasal cannula for pulmonary hypertension. He continued having persistent tachypnea despite normal oxygen saturation. A repeat echocardiogram demonstrated partial anomalous connection of the right pulmonary veins to the inferior vena cava with mild obstruction of the Scimitar vein at the diaphragm and persistent pulmonary hypertension (Figure 2E and F). He was started on furosemide and was weaned to room air successfully. Magnetic resonance angiography (MRA) confirmed the diagnosis of Scimitar syndrome: anomalous pulmonary venous drainage of all right-sided pulmonary veins to a vertical, scimitar-like vein that crossed the diaphragm and emptied into the IVC; there was normal left-sided pulmonary venous drainage, hypoplasia of the right lung, and a very small (2 mm) systemic arterial collateral arising from the abdominal aorta supplying the right lower lobe (Figure 2A—D). Repeat echocardiography demonstrated improvement of his pulmonary artery hypertension and spontaneous closure of ductus arteriosus. A decision was made to continue medical management with furosemide, maintain close follow-up and perform surgical repair of the partial anomalous pulmonary venous return at a later date.

Initially, a nasogastric tube was placed without difficulty and he tolerated the tube feeds. As his tachypnea resolved and he was able to maintain his saturation in room air, oral feeding was reintroduced and the episodes of choking and gasping for air during bottle feeding were noted. At that point his feeding difficulty was felt to be unrelated to his cardiac defect. Further evaluation by speech therapy noted the abrupt choking and aspiration after taking only a small volume of formula. A swallowing barium esophagogram was then obtained and demonstrated an H-type tracheoesophageal fistula (Figure 3).

Bronchoscopy confirmed the presence of the fistula in the membranous portion of the cervical trachea. Ligation and division of the H-type tracheoesophageal fistula was performed through a right-side incision. He recovered well and was kept NPO (*nil per os*) for 4 days. A repeat barium esophagogram demonstrated no esophageal leakage. He

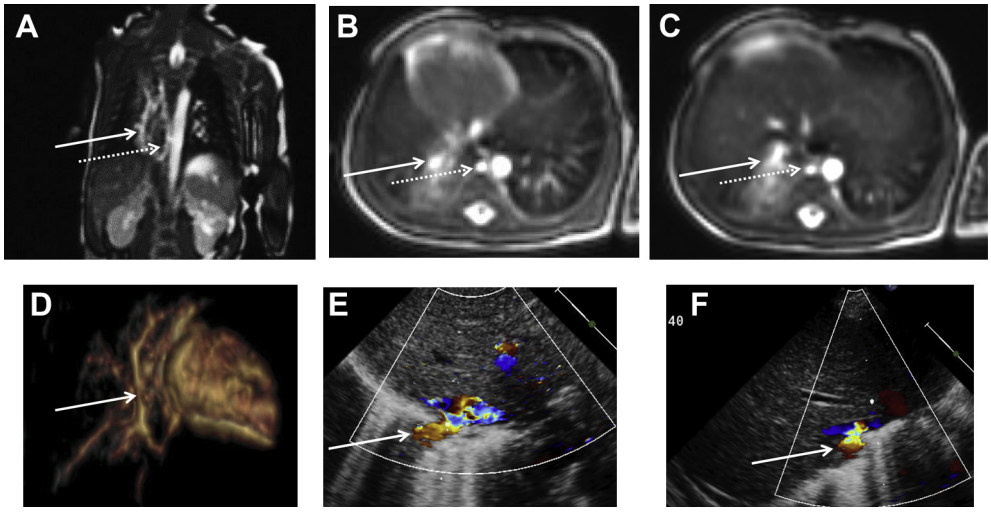

Figure 2 (A—C) Steady-state free precession magnetic resonance images. (A) Coronal view demonstrates the scimitar vein (white arrow) descending to the level of the diaphragm, and the azygous vein (dotted arrow). (B,C) Axial images reveal the scimitar vein crossing the diaphragm and inserting into the infradiaphragmatic, intrahepatic portion of the inferior vena cava (IVC); the azygous vein is also seen. (D) Three-dimensional reconstruction of gadolinium-enhanced magnetic resonance angiography (MRA) from a right posterior view displays the Scimitar vein. (E,F) Modified axial color Doppler echocardiographic views demonstrate similar findings to MRI views in B and C.