A 4-month-old infant girl was referred to hospital for evaluation of a heart murmur and failure to thrive. She had severe symptoms of congestive heart failure including poor weight gain, tachypnea, and respiratory retractions. Her body weight at the time of admission was 5.9 kg. A systolic murmur was clearly heard in the fourth right intercostal space. Echocardiography demonstrated right-sided pulmonary venous drainage into the junction of the inferior vena cava and the right atrium, and an atrial septal defect (6 mm in diameter). Chest roentgenography showed marked cardiac enlargement (cardiothoracic ratio: 65%) and pulmonary congestion. Electrocardiography revealed right axis deviation and right ventricular hypertrophy.

Cardiac catheterization revealed anomalous drainage of the right pulmonary veins into the inferior vena cava (Fig 1) and anomalous systemic arteries arising from the descending aorta and entering the lower lobe of the right lung (Fig 2). The left pulmonary veins were draining normally into the left atrium. Neither pulmonary venous obstruction nor stenosis was observed on angiography of the vessels on both sides. The pulmonary artery pressure was 85/30 mmHg (mean: 55), and the descending aortic pressure was 106/53 mmHg (mean: 76). Because large anomalous systemic arteries can cause congestive heart failure and pulmonary hypertension, coil occlusion of the collateral arteries was performed. Using a 4Fr right Judkins catheter, a 0.035-inch guide wire was advanced into the anomalous systemic arteries. Subsequently, the Judkins catheter was advanced into each collateral artery, and a coil was implanted one by one into each vessel. Selection of the coil basically depended on the diameter of each anomalous artery, and 3 arteries were finally completely occluded using 5-mm-diameter Gianturco coils and 3 with 3-mm-diameter Gianturco coils (Fig 3). Ten minutes later, the pulmonary artery pressure decreased to 57/14 mmHg (mean: 31). The symptoms of congestive heart failure resolved completely and good weight gain was observed. Chest roentgenography demonstrated marked improvement of pulmonary congestion (Fig 4). Surgical correction of the anomalous drainage of the right pulmonary veins was performed 2 months later without any complications. Resection of the lower lobe of the right lung or ligation of anomalous systemic arteries were not performed. Cardiac catheterization at 8 months after the surgery confirmed that reduction of pulmonary arterial pressure (mean, 16 mmHg) and persistent complete occlusion of the anomalous systemic arteries.

**Discussion**

Scimitar syndrome is a rare congenital heart anomaly consisting of abnormal pulmonary venous drainage into the right atrium, which is visible as a crescentic shadow of...
vascular density along the right border of the cardiac silhouette on chest radiography. The syndrome is also frequently characterized by varying degrees of hypoplasia of the right pulmonary artery and associated right lung bronchial anomalies. Dextrocardia is often present as a result of the hypoplasia of the right lung and the anomalous systemic arterial supply to it (the so-called 'pulmonary sequestration').

Although there are many reports of the clinical and pathologic findings of scimitar syndrome in children and adults, information concerning this disorder in infants is limited. Previous reports have demonstrated that patients in whom the diagnosis was made during the first year of life have more severe symptoms and a higher incidence of heart failure and pulmonary hypertension than do those patients in whom the diagnosis was made after 1 year of age. Several factors are probably responsible for the development of severe symptoms and pulmonary hypertension in infants and include (1) left-to-right shunting from the anomalous pulmonary venous return, (2) the presence of an abnormal systemic arterial supply to the lung (pulmonary sequestration) and associated cardiovascular defects, (3) constriction of the pulmonary vascular bed because of pulmonary hypoplasia with subsequent volume overload of the contralateral lung, (4) pulmonary venous obstruction, and (5) persistence of neonatal pulmonary hypertension. Gao et al emphasized the effect of the anomalous systemic arterial supply on the clinical symptoms and natural course. There are a limited number of studies of interventional treatments using embolization of the anomalous systemic artery in infants with scimitar syndrome. Although therapeutic embolization of the anomalous systemic arterial supply can be expected to decrease both the symptoms and the degree of pulmonary hypertension,
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the clinical efficacy of this procedure is unknown. Even if the systemic-to-pulmonary shunts are occluded, the anomalous drainage of the pulmonary veins and pulmonary hypoplasia play a major role in the development of congestive heart failure and pulmonary hypertension. In the present case, it was the lack of pulmonary hypoplasia that mostly influenced achieving an excellent result with coil occlusion. Perry et al reported the reopening of systemic-to-pulmonary collateral arteries necessitating repeat coil embolization? Although the cause of the reopening of the arteries in that case was uncertain, possibly angiogenesis de novo or the re-opening of previously existing channels, the efficacy of this procedure may be short-lived and corrective surgery should be performed within a few weeks or months. In patients with concomitant respiratory infection, this procedure should not be performed.

Despite these limitations, coil occlusion of systemic-to-pulmonary arteries in the infant patient with scimitar syndrome can dramatically improve symptoms of congestive heart failure, especially in patients with a extensive systemic-to-pulmonary collateral blood flow.

References