From dextropositio cordis to Scimitar syndrome

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We report on a female infant born to a 31-year-old G2/P1. At 23 weeks of gestation, the mother was transferred to a tertiary hospital for further assessment because of fetal thoracic anomalies. Ultrasonography showed dextroposition of the heart due to suspected left-sided congenital diaphragmatic hernia (CDH) and possible hypoplasia of the right lung. Since a cardiac defect could not be ruled out, pediatric cardiologists reassessed the patient at 27 and 29 weeks of gestation. Except for dextroposition, no cardiac anomalies were detected. The parents declined genetic analysis. Later in the course of pregnancy, right-sided pulmonary hypoplasia could no longer be confirmed.

The girl was born by Caesarean section due to breech presentation and failed tocolysis at 30 5/7 weeks of gestation. Apgar scores were 6, 6, and 7 at 1, 5 and 10 minutes, respectively. Arterial umbilical cord pH, birth weight, length, and head circumference were within normal ranges and percentiles.

Postnatal examination revealed respiratory distress with tachypnea of 80 breaths per minute, subcostal retractions and intermittent grunting. The heart sounds were shifted to the right. In addition, hypertelorism, down-slanting palpebral fissures, a high hairline, and a high-arched palate were noted. Because of respiratory distress, CPAP was provided from the third minute of life.
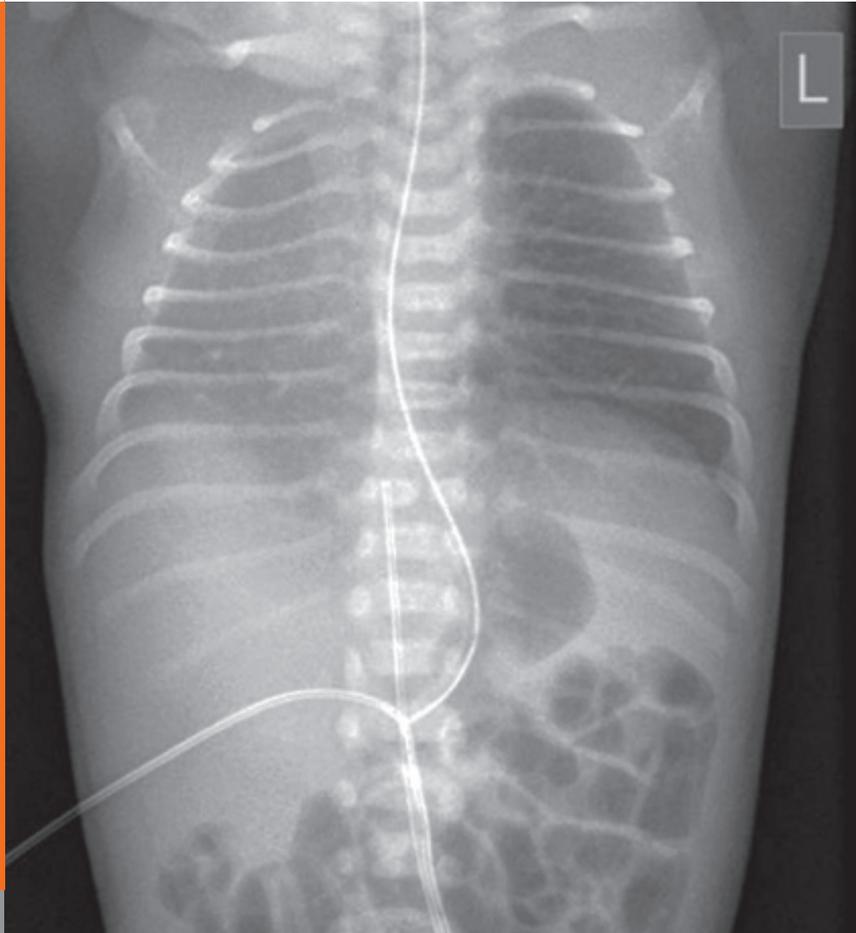
Fig. 1

Chest X-ray on DOL 1: dextroposition of the heart with a poorly outlined cardiac border.

Chest x-ray (Fig.1) confirmed dextroposition of the heart and revealed diffuse opacification of the right lung. Within 24 hours, both oxygen supplementation and CPAP could be discontinued. On the second day of life (DOL 2), echocardiography findings were felt to be compatible with scimitar syndrome (Fig. 2). Two days later, cardiac MRI (Fig. 3) confirmed the diagnosis of scimitar syndrome and the presence of right pulmonary hypoplasia with a hypoplastic right pulmonary artery, right-sided partial anomalous pulmonary venous return (PAPVR) to the right atrium. The arterial blood supply to the lower lobe of the right lung originated from the abdominal aorta, and there was a persistent left superior vena cava without vena innominata, an atrial septal defect (ASD), and a cor triatriatum sinistrum.

Over the following six weeks, the patient thrived satisfactorily but slowly developed congestive heart failure (CHF) with tachypnea and poor feeding. Treatment with hydrochlorothiazide and spironolactone was provided from DOL 44 to 55. On DOL 58, the patient´s overall condition rapidly deteriorated due to pulmonary hypercirculation as a result of PAPVR and pulmonary sequestration. Response to increased FiO2, diuretics, and digoxin was unsatisfactory (Fig. 4). On DOL 99, an episode of severe pulmonary arterial hypertension (PAH) resulted in circulatory arrest requiring cardiopulmonary resuscitation. Since her clinical status did not improve with conservative ma-
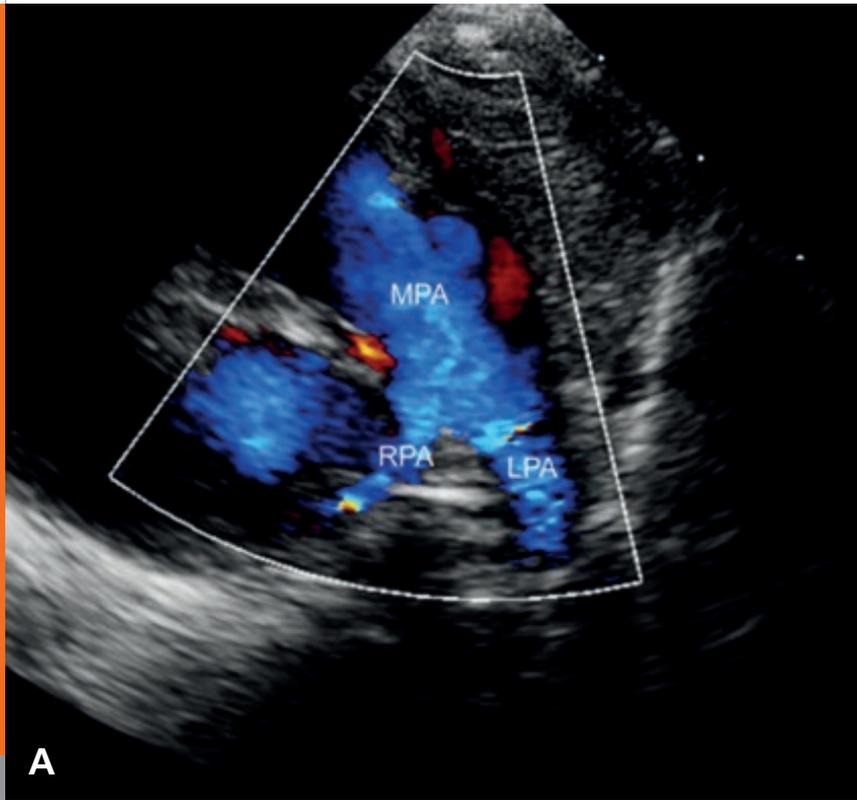
A

Transthoracic echocardiography: A) parasternal short axis view with color Doppler: pulmonary bifurcation with a hypoplastic RPA; B) subcostal longitudinal view: the Scimitar vein (arrow) can be recognised entering the right atrium just above the IVC (IVC: inferior vena cava; LPA: left pulmonary artery; MPA: main pulmonary artery; RA: right atrium; RPA: right pulmonary artery).
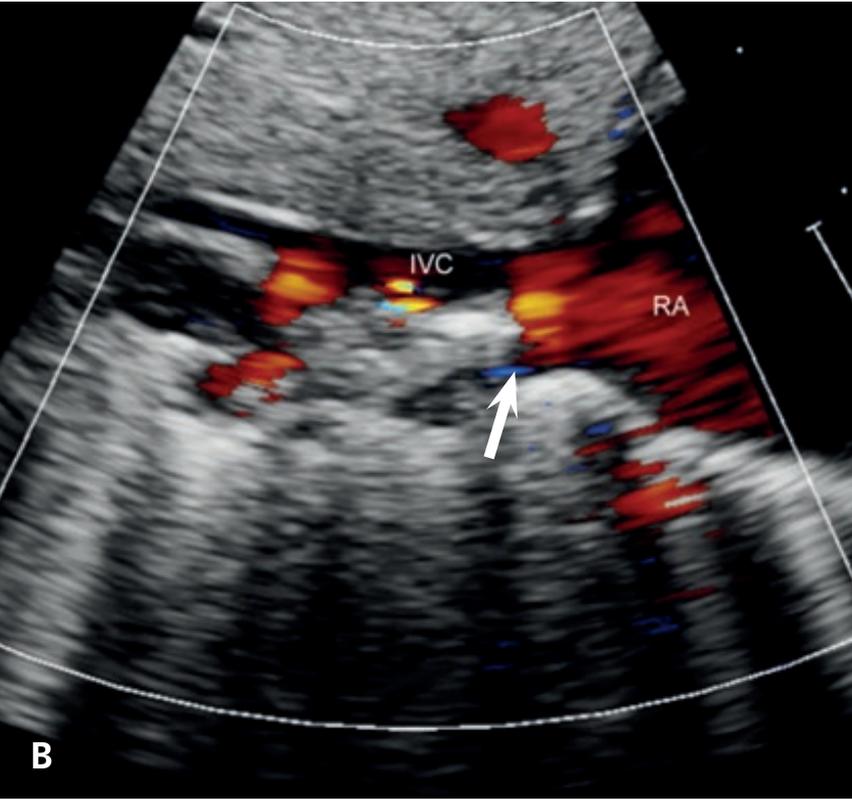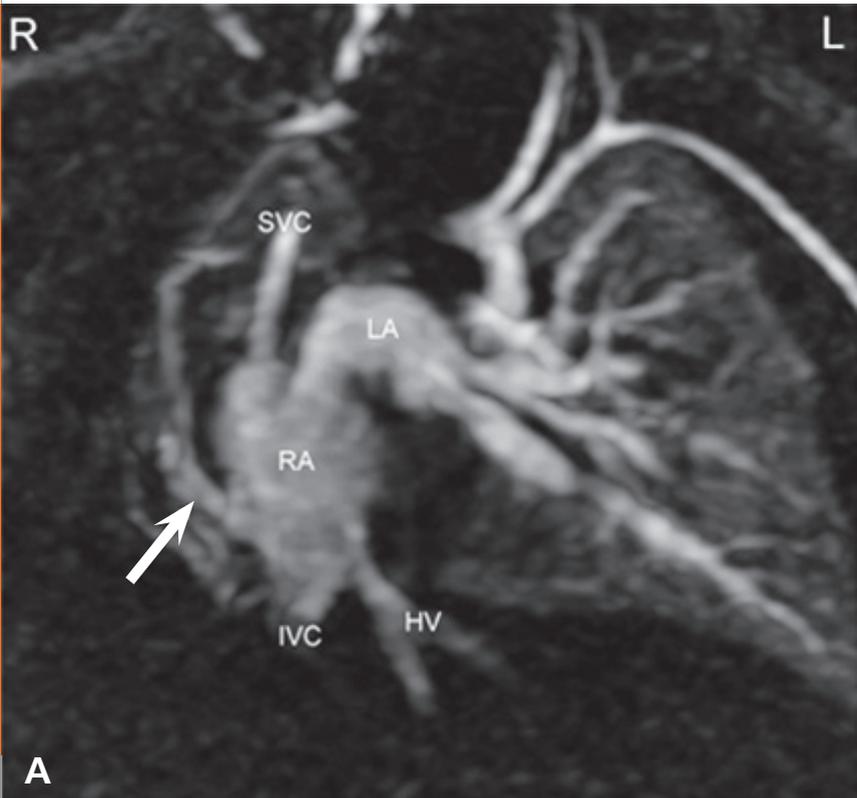
Fig. 3 **A**

Contrast-enhanced MR angiography: A) anterior view at the level of both atria: Scimitar vein originating from the right lung (arrow) is connected to the base of the right atrium; B) a systemic arterial collateral (arrow) originating from the abdominal descending aorta feeds into the base of the right lung (HV: hepatic vein; IVC: inferior vena cava; LA: left atrium; RA: right atrium; SVC: superior vena cava).
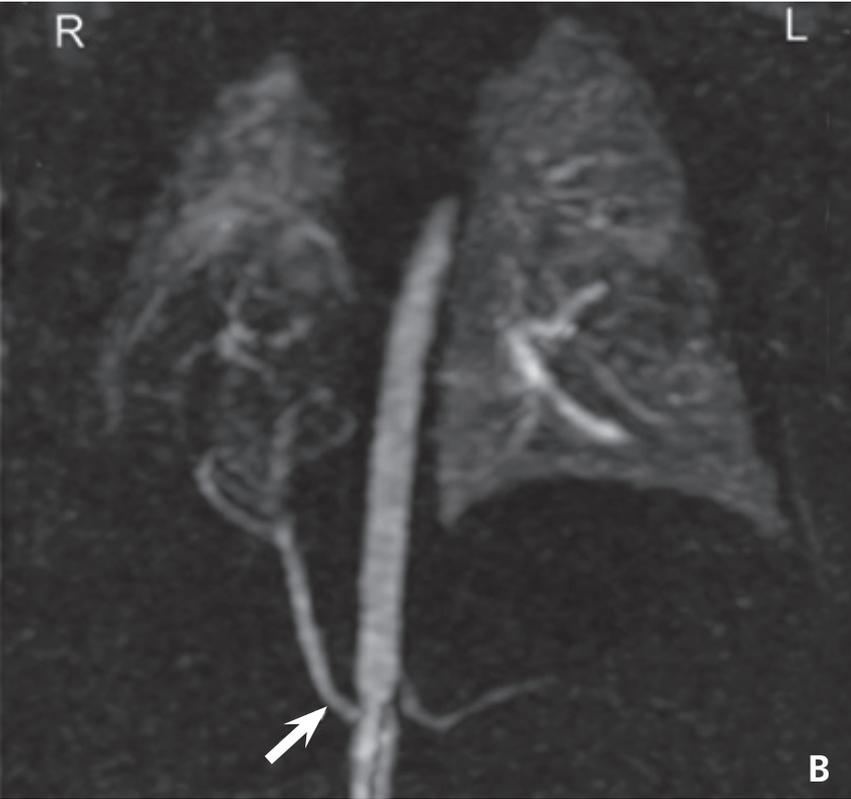
nagement, a decision was made to attempt complete surgical correction on DOL 105.

Surgical repair included ASD closure with fenestration, excision of the left atrial membrane, and resection of the large lung sequester. Significant bleeding occurred during redirection of the scimitar vein to the left atrium. Due to suprasystemic PAH weaning from cardiopulmonary bypass failed and extracorporal membrane oxygenation (ECMO) was initiated. Cranial neuro-imaging revealed severe generalized cerebral edema and multiple widespread hypoxic lesions. On DOL 108, due to the patient's poor prognosis, ECMO was discontinued and the patient died.
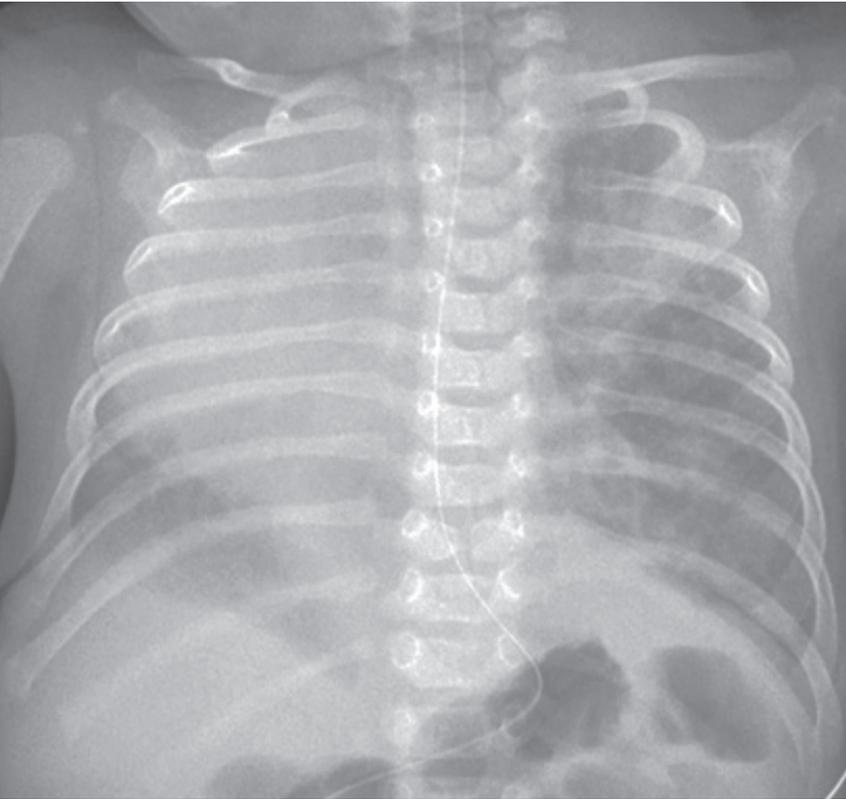
Chest x-ray on DOL 97:dextroposition of the heart.

Cooper and Chassinat first described scimitar syndrome in 1836 based on postmortem examinations. The first to diagnose this condition in a sick patient were most likely Dotter et al. in 1949 using cardiac catheterization and angiography (1). In 1956, Halasz et al. used the term "scimitar" (i.e., sword of oriental origin with a curved blade) for the first time (Fig. 5). On chest x-rays, they found a scimitar-shaped opacity caused by a pulmonary vein with abnormal drainage into or close to the right atrium (Fig. 6). Neill et al. were the first to use the term "scimitar vein" in the context of "scimitar syndrome" in 1960 (1, 3).

An indispensable finding in scimitar syndrome is partial or total abnormal pulmonary venous return from the right lung to the inferior vena cava (IVC), the inferior cavo-atrial junction or the right atrium. In two thirds of all cases, the scimitar vein (usually a singular vessel) drains the complete right lung, in one third of all cases, only its lower lobe with the upper pulmonary veins connecting normally (1, 4, 5). Only very few cases with left-sided scimitar veins have been described (11, 12). Further characteristics in scimitar syndrome are abnormalities of lung lobation and bronchial branching leading to a widely varying degree of lung hypoplasia, possible pulmonary sequestration, dextroposition of the heart secondary to lung hypoplasia, hypoplasia of the right pulmonary artery and anomalous systemic arterial blood supply from the descending aorta to the right lung (5). Associated congenital cardiovascular

anomalies include ASD (predominantly of secundum type), ventricular septal defect (VSD), coarctation of the aorta, and abnormalities of the aortic arch (5). Numerous nonvascular findings include right-sided diaphragmatic defects, accessory diaphragm, Bochdalek hernia, small hemithorax, scoliosis, and horseshoe lung (parenchymal continuity between right and left lung) (1-5).

The prevalence of scimitar syndrome is estimated to be 1-3 per 100'000 live births; its true incidence may be higher due to asymptomatic patients (3). There is a female predominance of about 2:1. To date only two cases of familial occurrence have been described (1, 13, 14). The exact pathogenesis of scimitar syndrome is unclear, but abnormal development of the lung bud in early embryogenesis is presumed (15).

Clinically, two different forms are distinguished: a neonatal/infantile form with severe symptoms, poor prognosis and high mortality rate (16-64%) (1-4, 6, 7, 14), and a childhood or adult form with an oligo- or asymptomatic course.

The neonatal/infantile scimitar syndrome is usually diagnosed within the first 2 months of life (1, 3) when patients present with CHF. Factors contributing to early onset and severity of CHF include severe and usually right-sided obstruction of pulmonary venous connections, additional cardiac malformations, and aberrant

Fig. 5 A

A) Scimitar sword; B) Scimitar-shaped opacity caused by a pulmonary vein draining abnormally into or close to the right atrium (from reference 5).
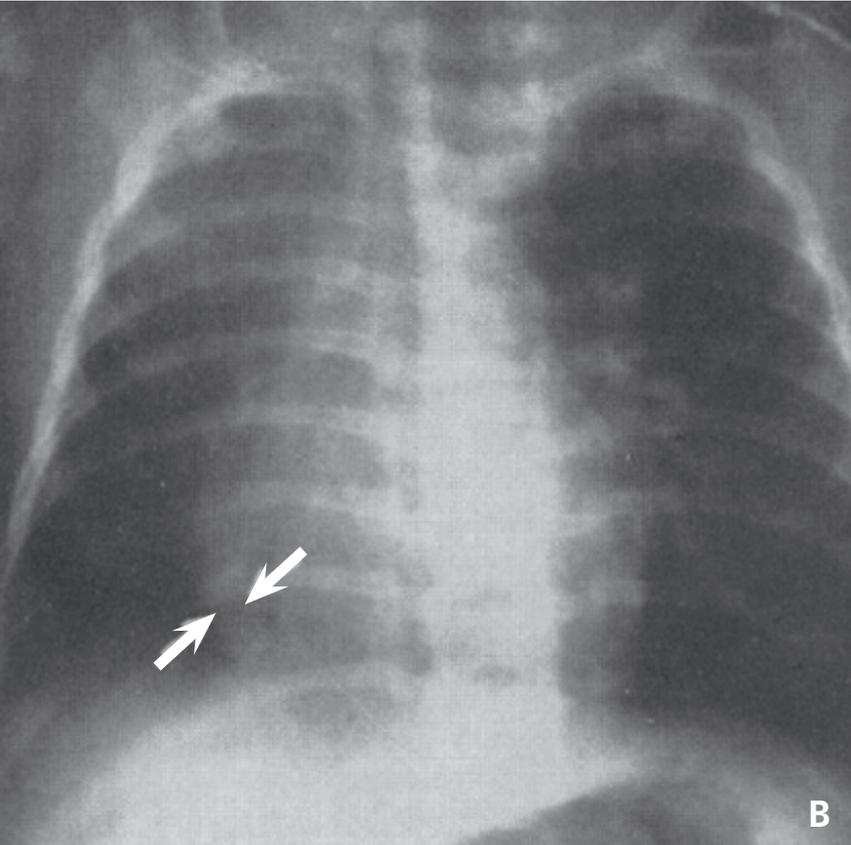
B

arterial supply of the lung sequester leading to significant left-to-right shunt with consecutive right heart failure due to volume overload (1-4, 8).

For diagnosis of scimitar syndrome, echocardiography, cardiovascular magnetic resonance tomography, and angiography are used (6, 10, 16). Treatment of neonatal/infantile scimitar syndrome is initially conservative but requires surgical intervention, as most symptomatic patients respond poorly to drug therapies only. However, surgical repair of scimitar syndrome at a young age is challenging and associated with high mortality and morbidity rates (1, 3, 4, 9).

REFERENCES

1. Freedom RM, Yoo S, Mikailian H, Williams WG. The natural and modified history of congenital heart disease. Blackwell Publishing 2004, ISBN: 1-4051-0360-4

2. Wang CC, Wu ET, Chen SJ, et al. Scimitar syndrome: incidence, treatment, and prognosis. Eur J Pediatr 2008;167:155-160

3. Gudjonsson U, Brown JW. Scimitar syndrome. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2006:56-62

4. Goa YA, Burrows PE, Benson LN, Rabinovitch M, Freedom RM. Scimitar syndrome in infancy. J Am Coll Cardiol 1993;22:873-882

5. Midyat L, Demir E, Askin M, et al. Eponym Scimitar syndrome. Eur J Pediatr 2010;169:1171-1177

6. Sarquella-Brugada G, Prada F, Mortera C. Double venous drainage in Scimitar syndrome. Ideal anatomy for percutaneous complete cure. Cardiol Young 2011;4:1-4

7. Argueta-Morales R, Meador LC, Nykanen DG, DeCampli WM. Infantile form of Scimitar syndrome with contralateral pulmonary vein stenosis. Pediatr Cardiol 2010;31:550-552

8. Bhide A, Murphy D, Thilaganathan B, Carvalho JS. Prenatal findings and differential diagnosis of Scimitar syndrome and pulmonary sequestration. Ultrasound Obstet Gynecol 2010;35:398-404

9. Karthekeyan RB, Saldanha R, Sahadevan MR, Rao SKG, Vakamudi M, Rajagopal BK. Scimitar syndrome: experience with 6 patients. Asian Cardiovasc Thorac Ann 2009;17:266-271

10. Khan MA, Torres AJ, Printz BF, Prakash A. Usefulness of magnetic resonance angiography for diagnosis of Scimitar syndrome in early infancy. Am J Cardiol 2005;96:1313-1316

11. D'Cruz IA, Arcilla RA. Anomalous venous drainage of the left lung into the inferior vena cava: a case report. Am Heart J 1964;67:539-544

12. Rutledge JM, Hiatt PW, Wesley Vick G 3rd, Grifka RG. A sword for the left hand: an unusual case of left-sided Scimitar syndrome. Pediatr Cardiol 2001;22:350-352

13. Ruggieri M, Abbate M, Parano E, DiStefano A, Guarnera S, Pavone L. Scimitar vein anomaly with multiple cardiac malformations, craniofacial and central nervous system abnormalities in a brother and a sister: familial Scimitar anomaly or a new syndrome. Am J Med Genet 2003;116:170-175

14. Dupuis C, Charaf LAC, Breviere GM, Abou P, Remy-Jardin M, Helmius G. The adult form of Scimitar syndrome. Am J Cardiol 1992;70:502-507

15. Schramel FMNH, Westermann CJJ, Knaepen PJ, Van Den Bosch JMM. The Scimitar syndrome: clinical spectrum and surgical treatment. Eur Respir J 1995;8:196-201

16. Berrocal T, Madrid C, Novo S, Gutierrez J, Arjonilla A, Gomez-Leon N. Congenital anomalies of the tracheobronchial tree, lung and mediastinum: embryology, radiology and pathology. Radiographics 2004;24:e17

17. Halasz NA, Halloran KH, Liebow AA. Bronchial and arterial anomalies with drainage of the right lung into the inferior vena cava. Circulation 1956;14:826-846

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