Scimitar Syndrome

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This syndrome is characterized by anomalous drainage of the right pulmonary veins into the inferior vena cava below the diaphragm.[1] In this situation, some or the total of right pulmonary veins unite in a single vessel, which travels along the right atrial border and gives the appearance of a scimitar on posteroanterior chest roentgenograms. It is the rarest form of anomalous venous returns.

The anatomical description of this anomaly was first made by Chassinat[2] and Cooper[3] in 1836 on autopsy material. Grishman[4] in 1949 and Runström[5] in 1950 described the roengenographic features of the anomaly. Finally Halazs and his associates indicated the similarity in shape of the anomalous pulmonary vein with that of a scimitar,[6] Neill and associates gave the syndrome the specific name, "Scimitar Syndrome".[7] Dotter et al proved the live clinical diagnosis by using angiocardiography.[8] Some other anomalies of the heart, lung and diaphram are also associated with this vascular anomaly. Among children, it is more common in females than males.

Case Report

A. O. (68–48107) an 11-year-old Caucasian girl was brought to our clinic with the chief complaints of palpitation and dyspnea. These complaints had continued through childhood, but had became worse during the past year. Family history was not contributory.

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Physical examination revealed a fairly well-developed and well-nourished girl in no acute distress. There was no cyanosis, dyspnea or edema. Her weight was 24 kilograms, height 129 cm., temperature 36.5 °C., pulse 100 per minute and the blood pressure 120/60 mm. Hg. in the arm and 140/70 mm. Hg. in the leg. Femoral pulsations were of good quality. No thrills were felt, liver and spleen were not palpable. On auscultation, there was a grade three (out of six) low frequency systolic murmur, which was best heard over the pulmonic area. The pulmonary second sound was widely split, and did not change with the respiration (Figure 1).

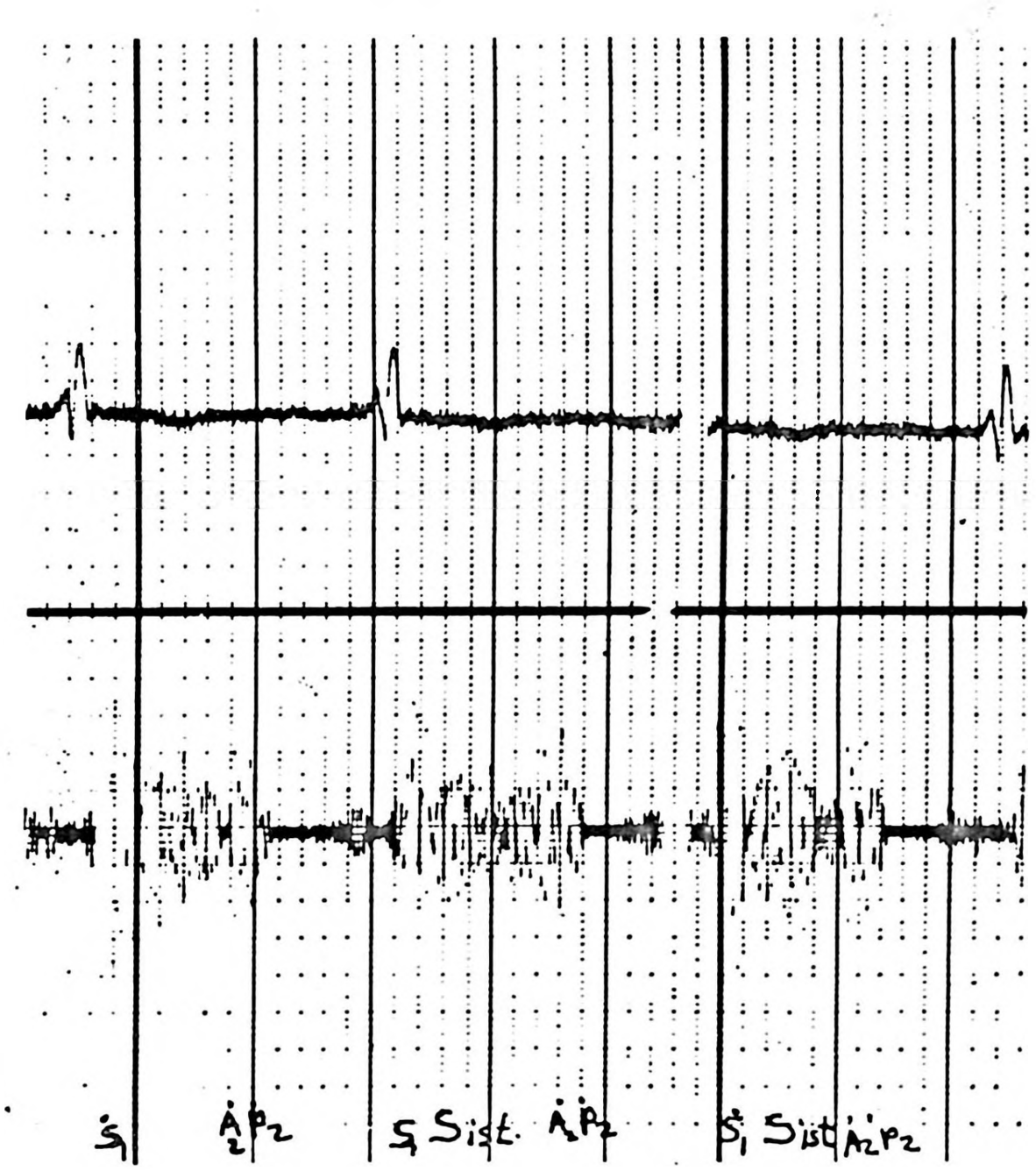

Figure 1. Phonocardiogram of the patient.

Laboratory Examinations: Her hemoglobin was 12.21 gm. per 100 ml.; hematocrit 40 per cent; white blood cell count 7.600 per cubic milimeter; nonprotein nitrogen 45 per cent. Urinalysis was normal. On roentgenographic studies, the heart was somewhat enlarged and the pulmonary vascular markings were increased. There was also a scimitar-like shadow along the right atrial silhouette on frontal chest roentgenogram (Figure 2). Electrocardiogram showed right axis deviation and right ventricular hypertrophy of diastolic overloading type (Figure 3).

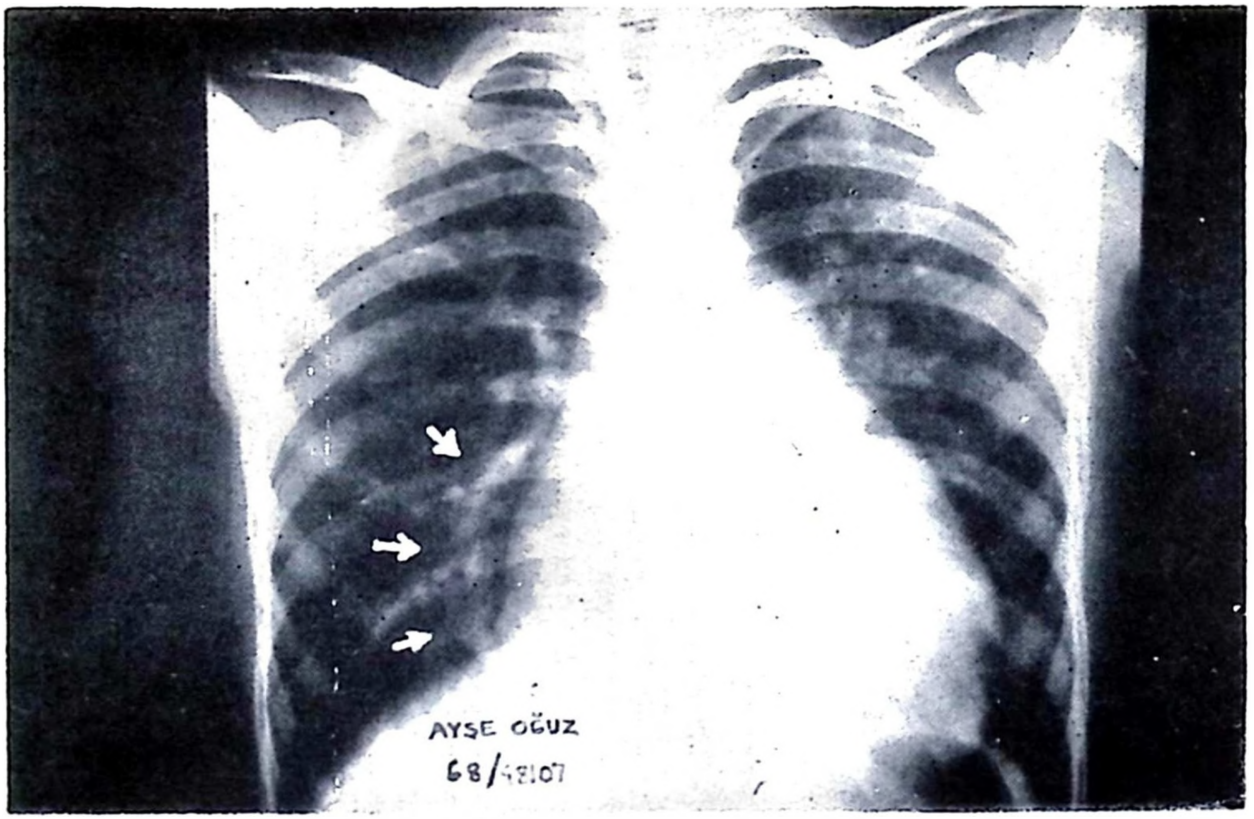

Figure 2. Posteroanterior chest roentgenogram (Arrows indicate the scimitar-shaped shadow).

Cardiac catheterization was done via right antecubital vein, using No. 7 F Cournand's catheter. Through the superior vena cava, the right atrium, right ventricle and pulmonary artery were reached. Blood samples were taken and pressures recorded throughout. During manipulation of the catheter, the left atrium was entered through an interatrial septal defect. As the catheter was introduced into the inferior vena cava an anomalous vessel was entered, which proceeded into the right lung field. This was proven to be a pulmonary vein by its blood oxygen saturation (Figure 4).

Selective angiocardiogram was done by injecting 30 ml. of 76 per cent Urografin into the right pulmonary artery and scimitar-shaped anomalous pulmonary vein, was demonstrated draining into the inferior vena cava

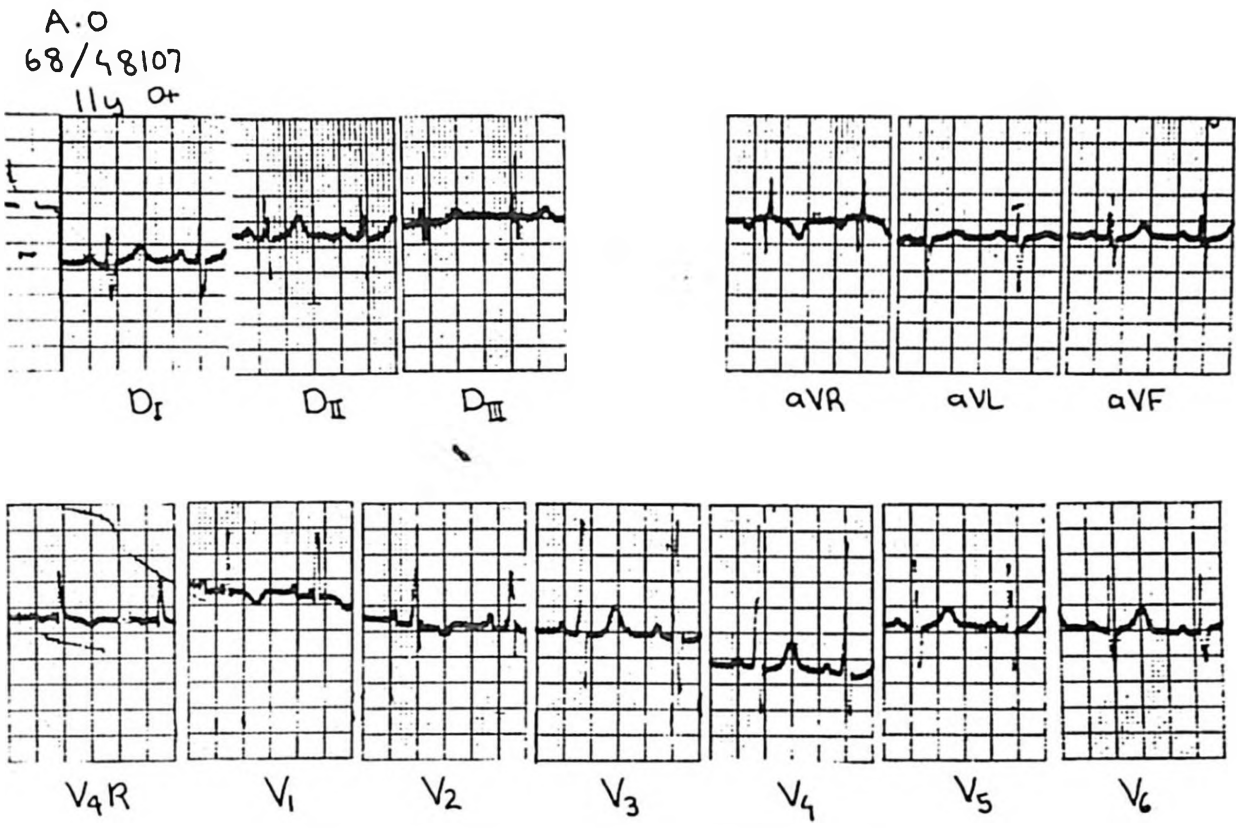

Figure 3. Electrocardiogram of the patient.

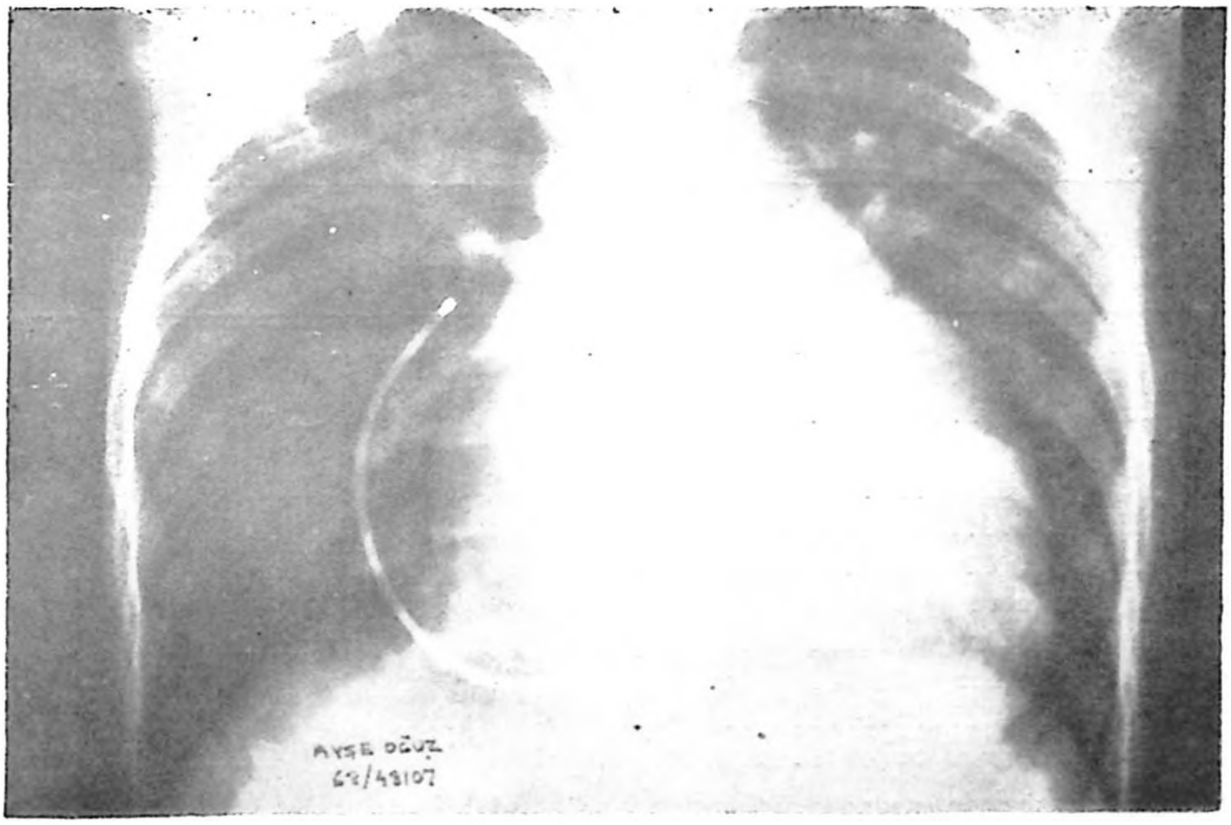

Figure 4. Posteroanterior spot film, showing the catheter in the anomalous pulmonary vein draining into the inferior vena cava.

below the diaphram (Figure 5). As shown in Table I, there was an increase in the blood oxygen saturations in the inferior vena cava and right atrium. Pressures in the right ventricle and pulmonary artery were

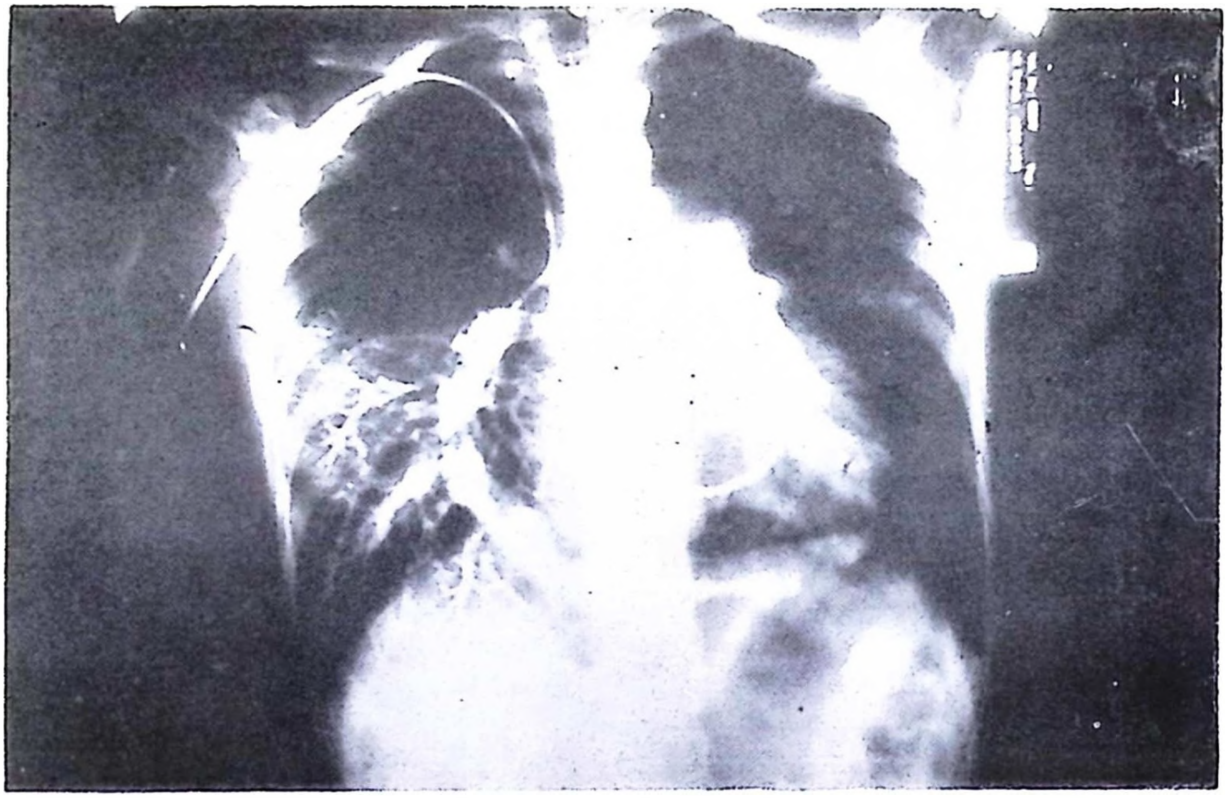

Figure 5. Posteroanterior angiocardiogram taken at venous phase.

TABLE I

CATHETERIZATION DATA

	O_2 Saturation (per cent)	Pressures (mm. Hg)
Superior vena cava		
High	74	
Low	70	
Right atrium		
High	80	
Mid	84	
Low	86	
Inferior vena cava		
Below diaphragm	86	
Below hepatic vein	73	
Right ventricle	85	50/ 0
Pulmonary artery	88	34/15
Left atrium	92	
Pulmonary vein	94	
Brachial artery	88	85/60

somewhat elevated, but there was no significant systolic pressure gradient between these two chambers. The final diagnosis was partial anomalous pulmonary venous drainage into the inferior vena cava below the diaphram (Scimitar syndrome), associated with an atrial septal defect of secundum type.

Discussion

Scimitar syndrome is a rare congenital cardiovascular anomaly. Mathay and his associates found only three cases among 55 different type of anomalous pulmonary venous returns, studied between the years 1958 and 1966.[9] At the pediatric cardiology department of Hacettepe Children's Hospital, we have diagnosed 44 cases of anomalous pulmonary venous return by cardiac catheterization and/or angiocardiography between 1958 and 1970. Three of these cases were total anomalous pulmonary venous return and the rest were partial, draining into the different chambers of the right heart. This is the first case of scimitar syndrome that we have encountered and diagnosed clinically. To our knowledge, this is going to be the first reported case in Turkey.

Malformations of the pulmonary venous system often produce characteristic roentgenographic features, which by themselves, are virtually diagnostic of the condition. Thus, total anomalous pulmonary venous drainage with connection to the left innominate vein gives rise to the "figure of eight" or "snowman" configuration of the cardiac silhoutte on frontal roentgenograms of the thorax. Total anomalous pulmonary venous return with connection below the diaphram yields the "ground glass" or "reticular" appearance in the peripheral lung fields on plain roentgenograms. Anomalous drainage of all or some of the right pulmonary veins to the inferior vena cava produces another characteristic roentgenographic appearance. In this malformation, the right pulmonary veins unite in a single vessel, which produces a scimitar-shaped paracardiac shadow in the right lower lung field and drains into the inferior vena cava or hepatic vein below the diaphram. This shadow is usually misdiagnosed as a recurrent pulmonary infection.

This malformation of the pulmonary venous system is frequently associated with hypoplasia of the right lung, which may cause the displacement of the heart to the right. In this situation, the characteristic shadow may be obscured by the heart, however an overpenetrated film or tomography can help in making the diagnosis. Hypoplasia of the right lung with the displacement of the heart may be present in variable degrees. In many cases no hypoplasia of the right lung is discernible on the

frontal roentgenogram of the thorax.[10] Our case did not show any evidence of lung hypoplasia on plain chest roentgenogram (Figure 2).

The clinical symptoms depend upon the associated lesions of the heart or lung. Therefore some of the patients may be asymptomatic and no treatment is needed. In symptomatic cases surgical treatment may be necessary.[11] Most commonly associated cardiac lesions are: ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot and atrial septal defect. Our case was associated with an atrial septal defect of secundum type.

Summary

A case of Scimitar Syndrome in a child is presented and the features of the syndrome were discussed. The main pathology of the syndrome is the anomalous drainage of all or some of the right pulmonary veins into the total the inferior vena cava below the diaphram. These veins form a characteristic scimitar-shaped shadow along the right atrial silhouette on frontal roentgenograms. In the pediatric age group, it is more common in females. The prognosis of the individuals with this malformation is related to the severity of the pulmonary hypoplasia and associated cardiac malformations.

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