# Anomalous Inferior Vena Cava with Azygos (Hemiazygos) Continuation

**Muhsin Saraçlar, M.D.***

Anomalous inferior vena cava with azygos (hemiazygos) continuation has been identified by various terms: persistence of the supracardinal vein, absence of the hepatic segment of the inferior vena cava, absence of the inferior vena cava, and absence of the upper part of the inferior vena cava with azygos (hemiazygos) continuation. Hemodynamically, the venous blood of the lower body empties into the superior vena cava by way of the azygos or hemiazygos vein instead of draining into the right atrium via the inferior vena cava. A case of anomalous inferior vena cava with hemiazygos continuation is reported because of its rareness.

*Case Report*

A three year old female (G. S.), who was the product of a normal pregnancy and cesarean section, contracted a severe pulmonary infection two months before she was examined at Hacettepe.

On physical examination, the patient weighed 12.800 gm, and measured 96 cm. Her pulse was 110 per minute and blood pressure 95/60 mm Hg. There was no clubbing of the fingers or cyanosis. Her lungs was clear. Her heart was hyperdynamic at the xiphoid and apex, and a systolic thrill was felt at the upper left sternal border. A loud systolic murmur was also present over the precordium, and was best heard at the third left intercostal space. There was a significant mid-diastolic rumble at the xiphoid and apex. The second sound in the pulmonary area was widely split. The liver was one cm below the right costal margin. The femoral and brachial pulses were normal in quality and intensity. There were polydactyly and pes cavus deformities.

From Hacettepe University Faculty of Medicine, Department of Pediatrics, Ankara.

* Attending Pediatrician and Pediatric Cardiologist.

Electrocardiogram showed a minus 75° left axis deviation, left atrial enlargement and biventricular hypertrophy (Figure 1). X-ray examinations revealed right and left ventricular enlargement, increased pulmonary vascularity and left atrial enlargement. Hemoglobin was 13 gm, and NPN 32 mg per 100 cc of blood.

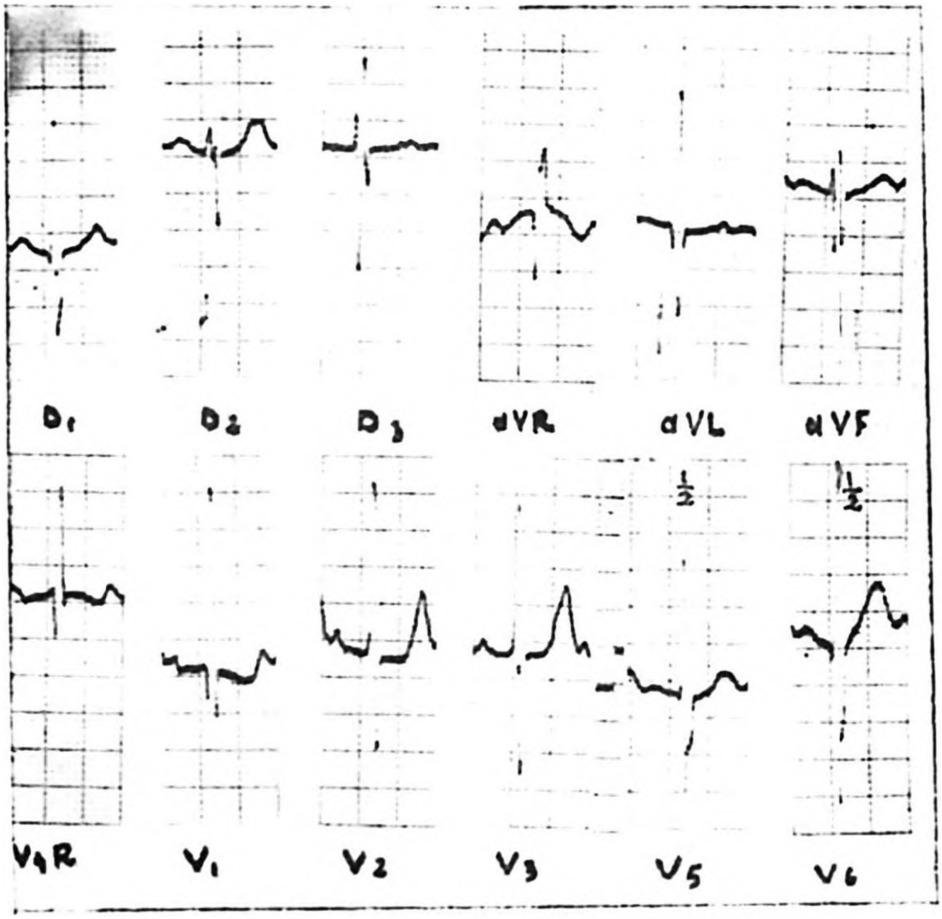



**Figure 1.** Electrocardiogram of the patient showing a minus 75 left axis deviation, left atrial enlargement, and biventricular hypertrophy.

Right and left heart catheterizations were performed; a catheter inserted into the saphenous vein of the right leg passed upward along the posterior abdominal wall into the chest, but lay in a vessel behind the heart. When the catheter was pushed further, it entered the persistent left superior vena cava and passed downward into the coronary sinus and right atrium. Then the catheter entered the right ventricle, main pulmonary artery and left pulmonary artery. Biplane x-rays were taken to record the position of the catheter (Figure 2).

Oxygen saturation and pressure were measured in each chamber and vessel (Table 1). There was a marked rise of oxygen saturation in the right atrium with no further rise in the right ventricle or pulmonary arteries. Pressure in the pulmonary artery was 40/10 mm Hg; systemic arterial pressure was 88/43 mm Hg.

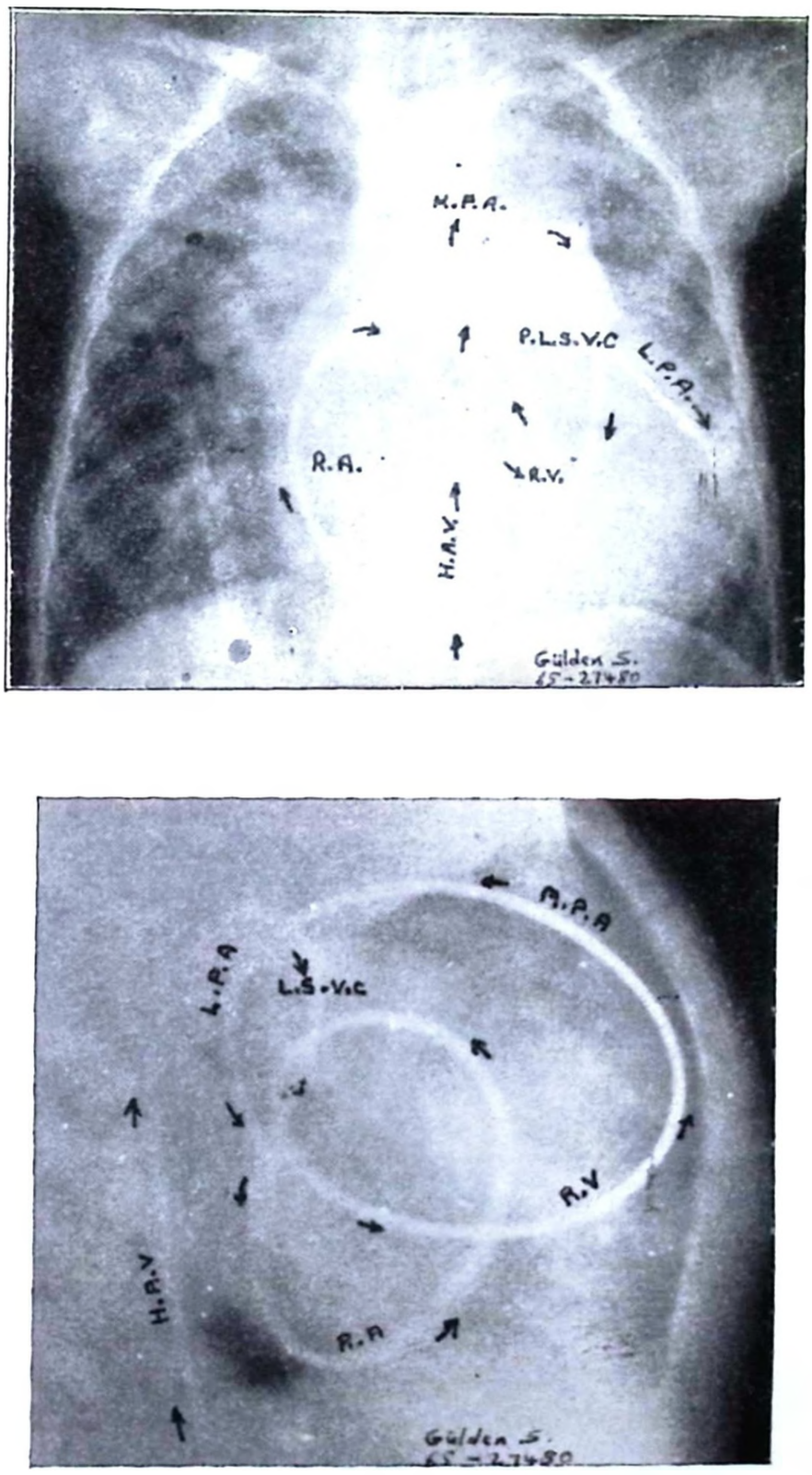



**Figure 2.** Posteroanterior and lateral roentgenograms showing the position of the catheter, which follows the hemiazygos vein, persistent left superior vena cava, coronary sinus, right atrium, right ventricle, main pulmonary artery and left pulmonary artery.

TABLE 1

CATHETERIZATION DATA

| | O2 Saturation % | O2 Content cc/lt | Pressure mm/Hg |
|---|---|---|---|
| Main pulmonary artery | 91 | 110 | 41/10 |
| Right ventricle | 91 | 110 | 50/7 |
| Right atrium | 91 | 110 | a: 9 c: 6 v:6 |
| Coronary sinus | 72 | 90 | |
| Persistent left superior vena cava | 73 | 91 | |
| Left ventricle | 97 | 121 | 88/4 |
| Brachial artery | 97 | 121 | 88/43 |

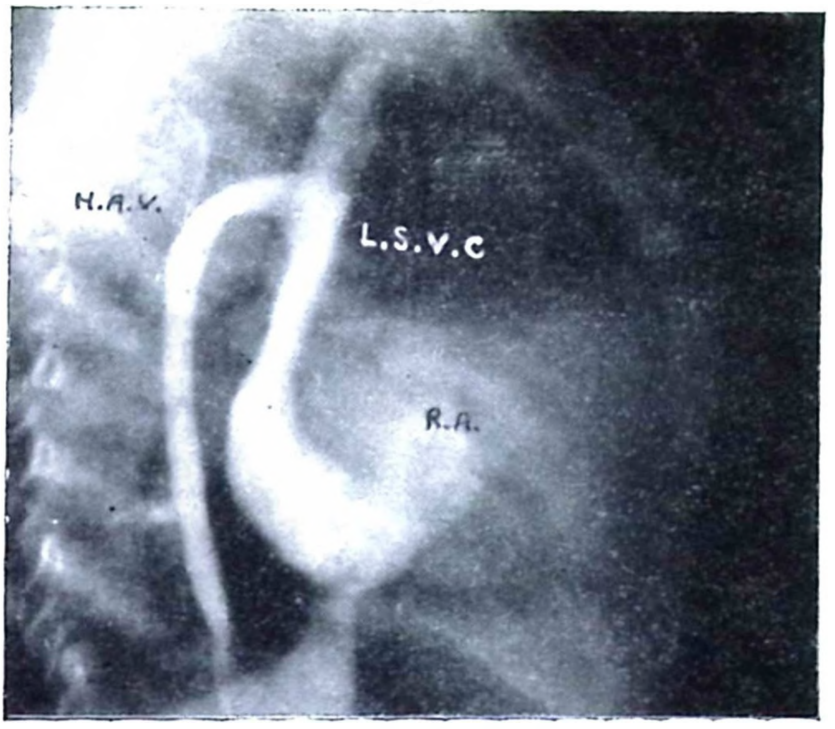
| | |
|---|---|
| Pulmonary flow | 16.3 lt/min |
| Systemic flow | 6 lt/min |
| Left to right shunt | 10.3 lt/min |
| Pulmonary resistance | 1 unit |

Radiopaque material was injected into the vein below the diaphragm to illustrate the anomoly. In Figure 3, the dye follows the hemiazygos vein, persistent left superior vena cava, coronary sinus and right atrium. A retrograde aortogram ruled out patent ductus arteriosus and coarctation of the aorta (Figure 4). The final diagnosisis was endocardial cushion defect and pulmonary hypertension associated with a venous anomaly.

## *Discussion*

Anomalous inferior vena cava with azygos (hemiazygos) continuation is a rare malformation. Formerly reported cases were mostly from dissecting laboratories. Recently the malformation is being diagnosed more frequently because of increased application of cardiac catheterization and angiography. In 1961, Anderson[1] published 15 cases of his own and collected 26 cases from the literature which had detailed clinical information. To our knowledge, cases reported since 1961 are as follows: Ertuğrul[2] 1, Hood[3] 4, Dupuis[4] 5, LePere[5] 1, Peterson[6] 1, Milledge[7] 5, Bussat[8] 1, and Derra[4] 5. Only 14 of these 77 cases had hemiazygos continuation as in our patient.

Although anomalous inferior vena cava with azygos (hemiazygos) continuation may occur as an isolated anomaly[3-6], it is usually associated with other congenital cardiovascular anomalies.[1,7] It has been stated that



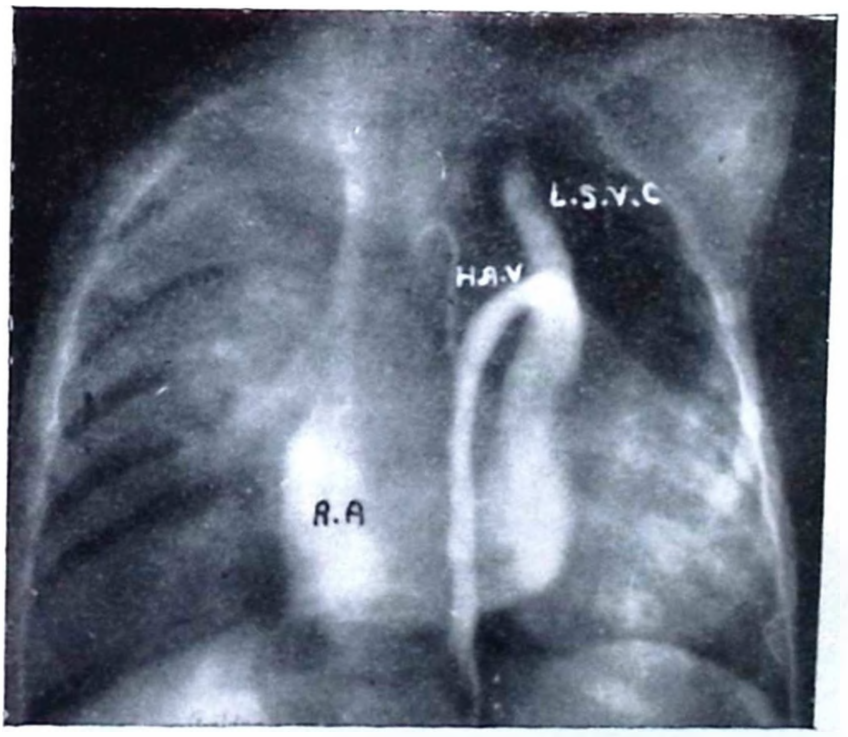
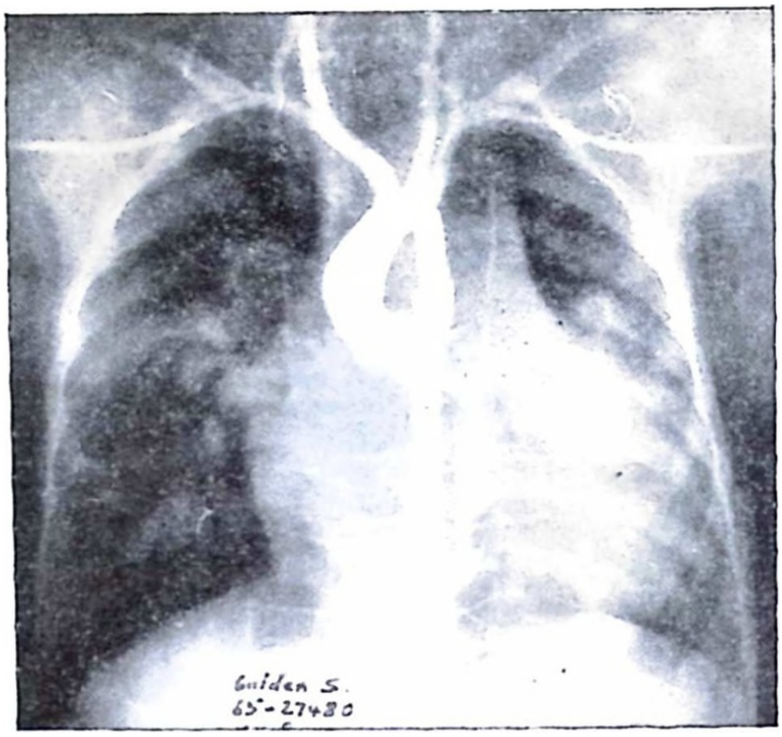




**Figure 3.** Posteroanterior and lateral angiocardiograms. Radiopaque material follows the hemiazygos vein, persistent left superior vena cava, coronary sinus and right atrium.



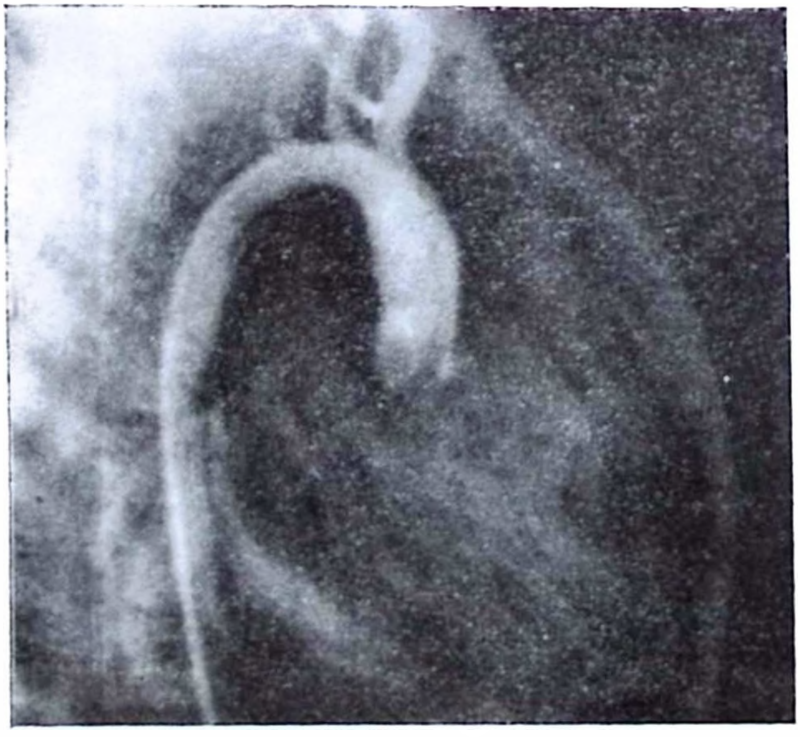



**Figure 4.** Retrograde aortograms ruling out patent ductus arteriosus and coarctation of the aorta.

the incidence of this condition is 0.6 per cent in patients with congenital heart disease. Abnormal position of the heart is also a common association.

Embriologically this malformation is interpreted by Anderson[1] as an interruption or failure of fusion of the inferior vena cava combined with persistence of either the right lumbar azygos vein or left lumbar hemiazygos vein.

There are no signs of this malformation on physical examination. Electrocardiogram shows only the changes of the associated congenital heart disease; however, on x-ray a widened azygos or hemiazygos vein may point out the malformation.[3 6 10] Heart catheterization via a leg vein is the easiest way to discover the anomaly. During fluoroscopy the tip of the catheter cannot be manipulated as usual when it is on the right atrial silhouette, because it is in the azygos or hemiazygos vein beyond the heart.

When the catheter is pushed further, it enters the superior vena cava and right atrium. The anomaly may not be noted in cases where catheterization is done via an arm vein. However, anomalous inferior vena cava can be suspected if the catheter cannot enter the inferior vena cava from the right atrium.

Anomalous inferior vena cava with azygos (hemiazygos) continuation *per se* is harmless, however, it may be of major importance in a patient with portal hypertension in whom a porto-caval anastomosis is suspected.[10] In 1951, Effler[11] reported the first case of surgical complication of this malformation. The patient died following ligation and partial excision of the anomalous vein. It is important to be aware of the anomaly in surgical correction of a cardiac malformation, especially when the heart-lung machine is used, because it may require special techniques.

## *Summary*

A case of anomalous inferior vena cava with azygos (hemiazygos) continuation is presented and the literature reviewed. Only 19 of 77 previously reported cases of anomalous inferior vena cava had hemiazygos continuation similar to that of our patient.

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