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Anomalous Inferior Vena Cava with Azygos (Hemiazygos) Continuation

A case report

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Anomalous inferior vena cava with azygos or hemiazygos continuation has sometimes been termed as absence of the inferior vena cava, absence of the hepatic segment of the inferior vena cava or infrahepatic interruption of inferior vena cava with azygos continuation.[1 2 3] Embryologically, a failure of the distal inferior vena cava to fuse normally with the hepatic portion results in an anomalous inferior vena cava with azygos or hemiazygos continuation. Hemodynamically, venous blood of the lower body drains via azygos or hemiazygos vein into the right or left superior vena cava.

In a previous report, we described a case of anomalous inferior vena cava with hemiazygos continuation.[4] Recently, we have encountered an additional case with different intracardiac anomalies. Since the anomaly is rare, this case is reported.

Case Report

S. K. (160729), 2-year-old female was admitted to Hacettepe Hospital for evaluation of congenital heart disease. She was the product of a normal pregnancy and delivery, and has been cyanotic since birth.

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On physical examination her blood pressure was 90/50 mm/Hg. on right arm, 100/70 mm/Hg. on right leg. She weighed 10 kgm., and measured 81 cm. There was no respiratory distress. Her nails and lips were cyanotic. Clubbing of the fingers was present. Heart was hyperdynamic at xiphoid. There was no thrill. A grade 3°/6 ejection type systolic murmur was present at the third left parasternal space. Sometimes a faint continuous murmur was heard at upper sternum. The second heart sound in the pulmonary area was permanently split. The intensity of the pulmonary component was normal. Other physical findings were within normal limits.

Hemoglobin was 14.07 gm./100 ml., hematocrit was 45 %. Heinz body and Howel Joly tests were negative. Electrocardiogram showed + 135° right axis QRS deviation, atrial inversion, and biventricular hypertrophy (Figure 1). Posteroanterior X ray films revealed cardiac enlargement (Figure 2). The aortic arcus was on the right side. Pulmonary vascularity was slightly increased. The stomach was on the right side, and liver was on the left.

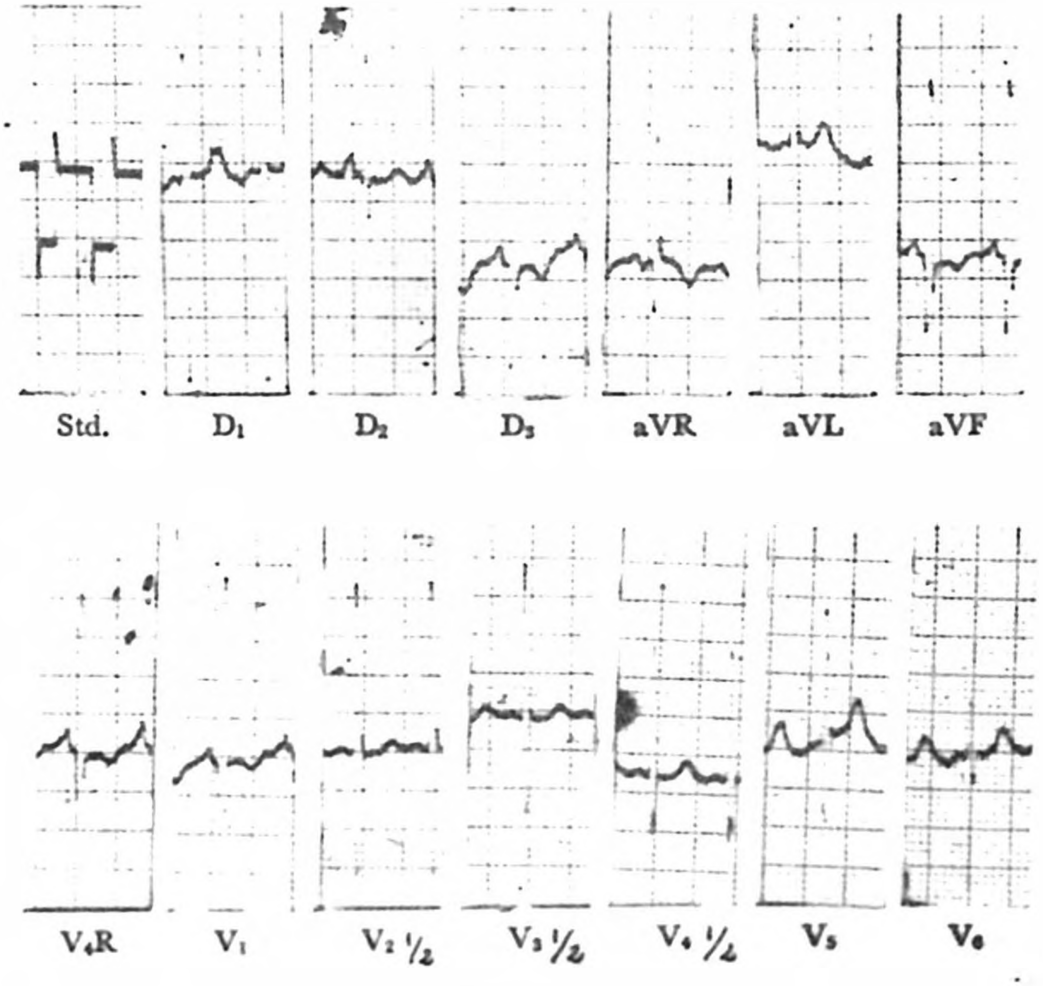


Figure 1
Electrocardiogram of the patient shows + 135° right axis QRS deviation, atrial inversion, and biventricular hypertrophy.

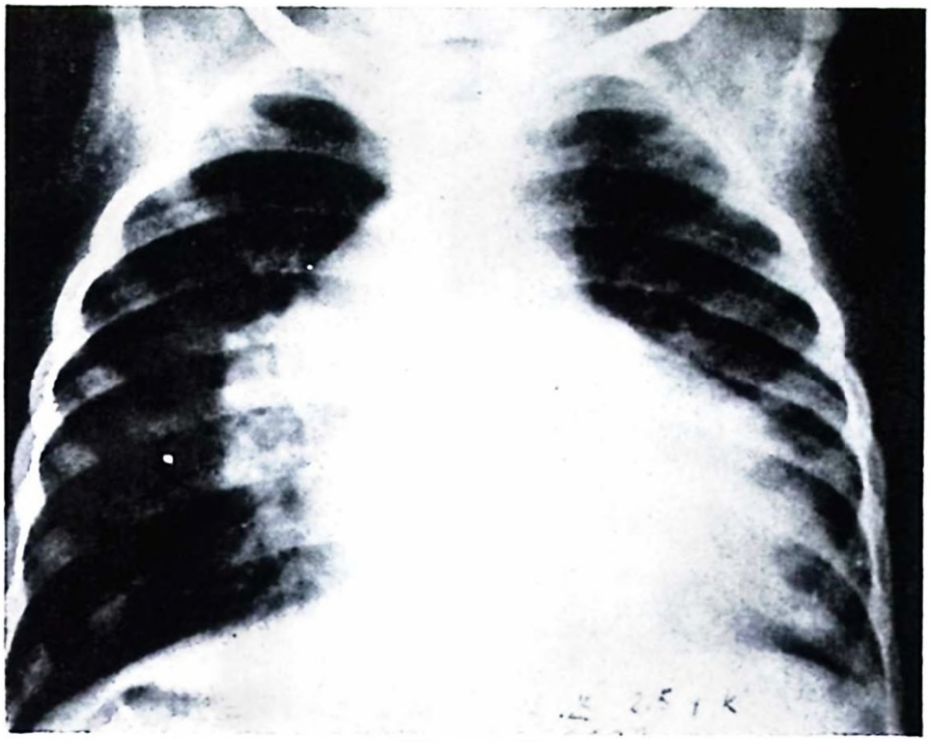


Figure 2

Posteroanterior telecardiogram of the case shows cardiac enlargement. The aortic arcus is on the right side. Pulmonary vascularity is slightly increased. The stomach is on the right side.

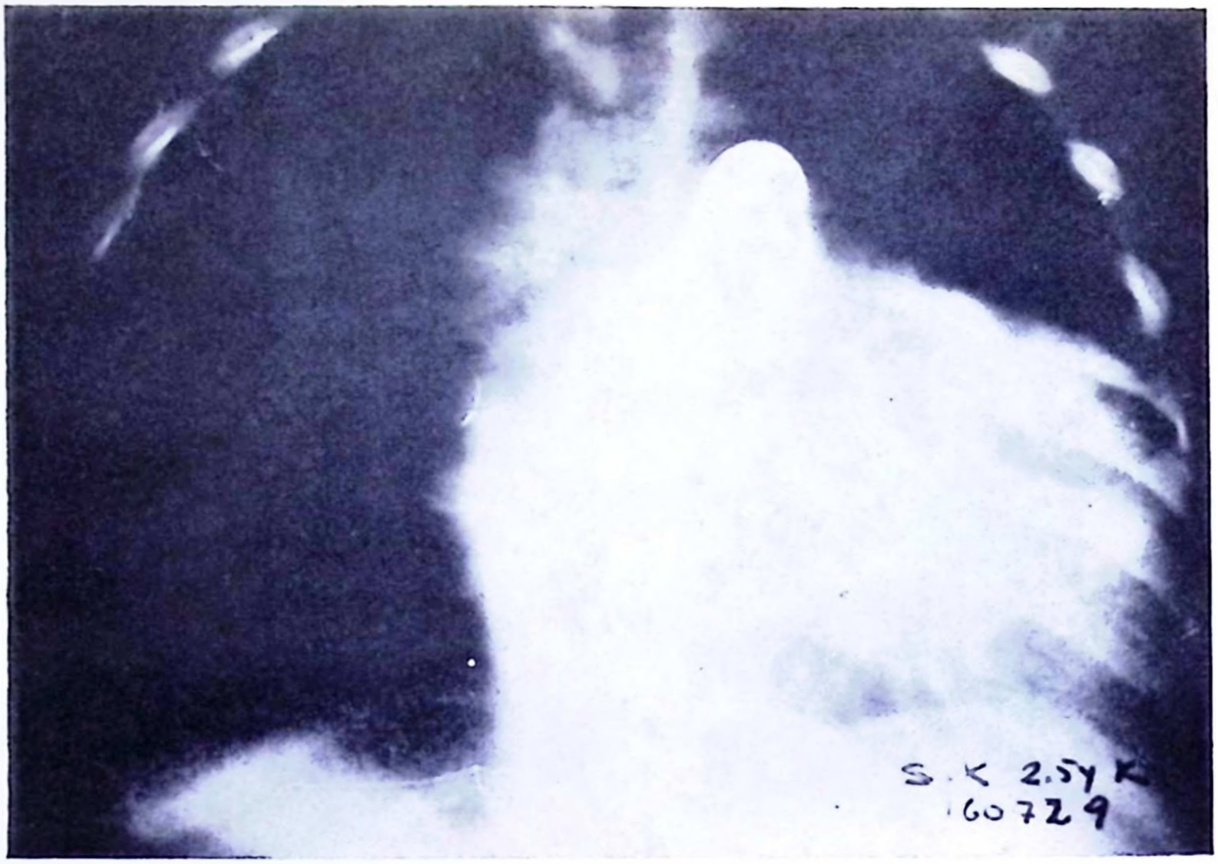
In order to obtain definite diagnosis, the patient was catheterized. The catheter was put into the right saphenous vein. During manipulation, a curve of the catheter could not be made when the tip was in the right atrial silhouette. Anomalous inferior vena cava was suspected. The catheter was pulled back into the abdomen, and radiopaque material was injected to show anomaly. Then the catheter entered the right atrium and anomalous right pulmonary vein via hemiazygos vein and superior vena cava. Blood samples for oxygen saturations were taken and pressures were recorded from the cardiac chambers entered (Table I).

TABLE I

CATHETERIZATION DATA

	O_2 Saturation* %	Pressures mm/Hg.
Pulmonary vein	96	–
Right atrium	96	m: 6
Superior vena cava	50	–
Femoral artery	70	95/60

* Analysis of the oxygen saturations reveales a rise at the right atrial level. There is arterial oxygen unsaturation.



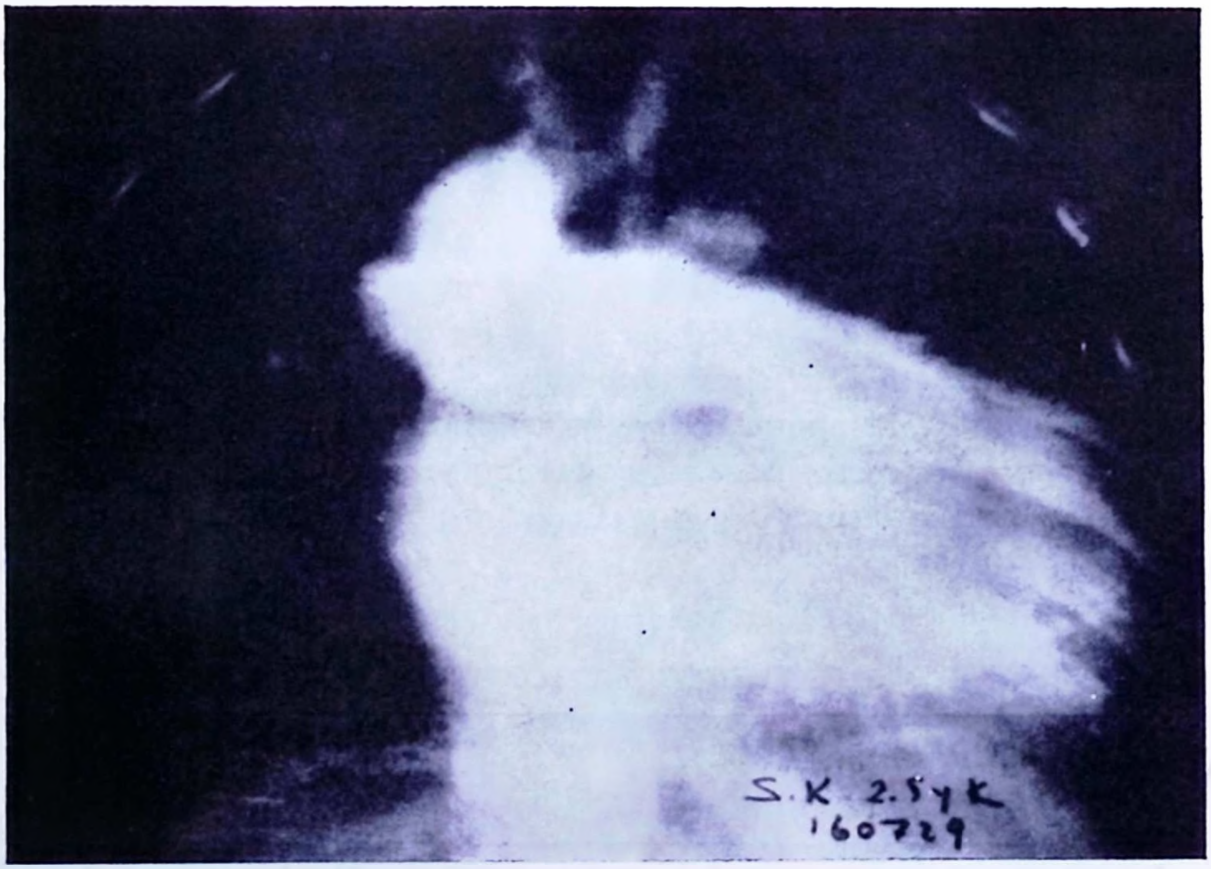


Figure 3 a ve b

Angiocardiograms. Radiopaque material was injected into the vein, below diaphragm. Dye follows hemiazygos vein, persistent left superior vena cava, right atrium, and right ventricle. Both great vessels filled from a common trunk. Descending aorta is on the right side.

a– Early anteroposterior film. b– Late anteroposterior film.

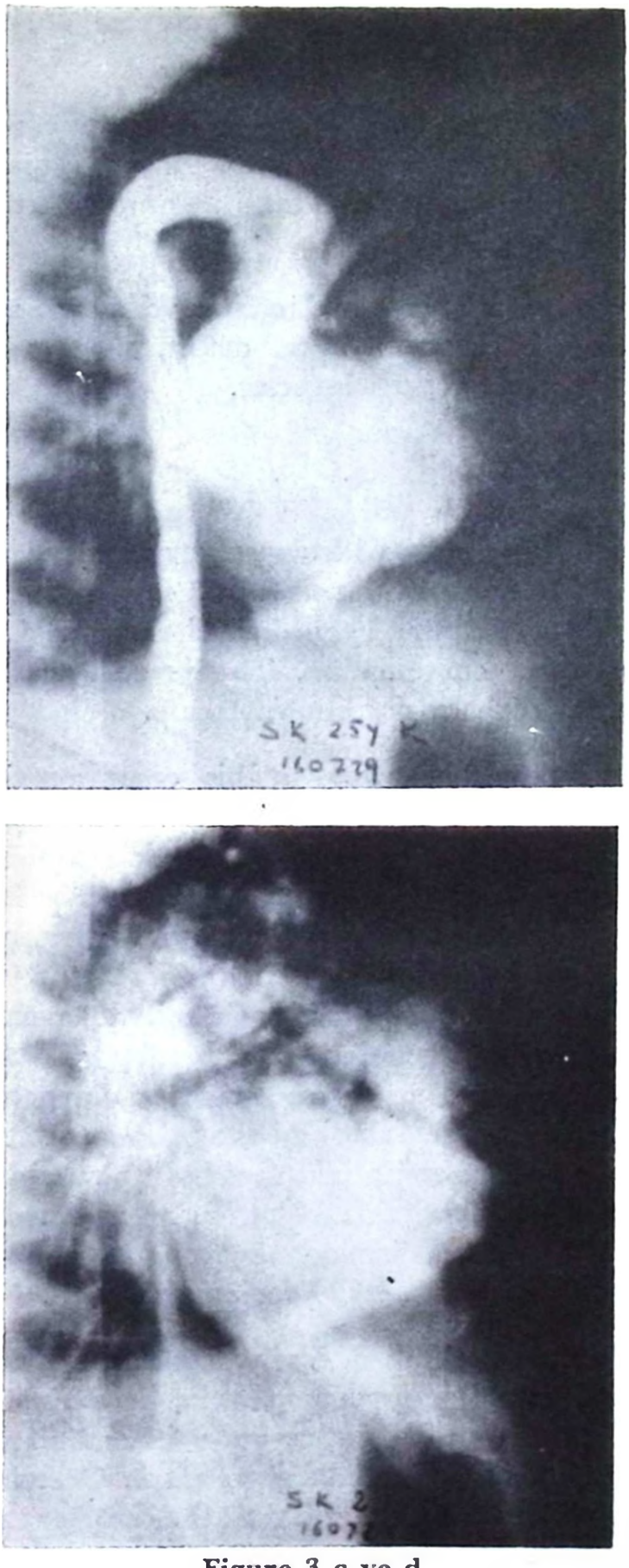


Figure 3 c ve d

Angiocardiograms

c– Early lateral film. d– Late lateral film.

Analysis of the oxygen saturations revealed a rise at the right atrial level. There was arterial unsaturation.

On angiocardiograms, radiopaque material followed hemiazygos vein, persistent left superior vena cava, right atrium and ventricle (Figure 3). Both great vessels filled from a common trunk. Descending aorta was on the right side.

Final diagnosis was anomalous inferior vena cava with hemiazygos continuation, levocardia, atrial septal defect, single ventricle, partial anomalous venous return, situs inversus.

Discussion

Anomalous inferior vena with azygos or hemiazygos continuation is a rare anomaly. In our previous report 77 cases of anomalous inferior vena cava (AIVC) were found in medical literature.[4] Only 14 of these cases had hemiazygos continuation. The present case also showed hemiazygos continuation as did our first case.

There are no physical findings in this condition since the venous anomaly is functionally insignificant. Therefore, it could not be diagnosed clinically. In the case presented, systemic venous anomaly could not be suspected upon physical examination. It has been mentioned that a radiologic finding in the plain posteroanterior chest X-rays, consisting of a slight bulging shadow in the right supracardiac border at the region of the superior vena cava might sometimes be noted in cases with azygos continuation.[2 5 6 9] Berdon and Baker[7] reported two cases of A.I.V.C. with azygos continuation diagnosed by preangiographic plain films. They emphasized that the association of a right mediastinal mass and of right paravertebral pleural deviation in a patient with a normal left aortic arch strongly suggested this anomally. We have not been able to diagnose the venous condition from plain X-rays, since our case had hemiazygos continuation.

The diagnosis is established during cardiac catheterization. The catheter, inserted into the saphenous vein, is pushed until cardiac silhouette. It enters azygos or hemiazygos vein and seems, as if, it was in a vessel instead of being in the right atrium. Occasionally, a curve is made in the superior vena cava and then the catheter can be advanced into the right atrium. In our case the diagnosis of venous anomaly was established during the manipulation of the catheter in the right atrial silhouette.

Venous angiography below the diaphragm is diagnostic. We confirmed the diagnosis by means of venous angiography.

The anomaly is often associated with complex cyanotic cardiovascular defects frequently encountered are cor biloculare, atrioventricular canal, anomalies of pulmonary venous return, origin of both great vessels from the right ventricle, pulmonary stenosis or atresia and combination of the above.[3] Our case showed complex intracardiac lesions.

Hepatic veins open directly into the right atrium since the venous interruption is below them. However, in some cases with levocardia and situs inversus, they open into the left atrium. During atrial systole, regurgitation of radiopaque material into the hepatic veins from right atrium was seen on the angiograms of our case.

Polysplenia or asplenia may be associated with this anomaly.[7 8] Asplenia was not present in our case.

In regard to the position of the viscera and the heart, Anderson et all classified 40 cases they reviewed. None of the cases showed situs inversus with levoversion and hemiazygos continuation as in our case.

Because of the fact that the venous anomaly, by itself, is harmless, surgical correction is not necessary.

Summary

A case of anomalous inferior vena cava with hemiazygos continuation is reported, and the diagnostic methods of this anomaly are discussed.

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