ANOMALIES OF THE INFERIOR VENA CAVA: ABSENCE OF THE UPPER PART OF THE INFERIOR VENA CAVA WITH AZYGOS CONTINUATION

Emel Özmen, M. D.* and Ali Ertuğrul, M. D.**

Terminology

A variety of names have been used to describe the anomaly characterized by the absence of the upper part of the inferior vena cava with azygos continuation. Some of the terms to be found in the literature are: anomalous inferior vena cava with azygos continuation; absent inferior vena cava; anomalous inferior vena cava with azygos drainage; absence of the hepatic segment of the inferior vena cava; persistence of the supracardinal vein; persistence of the subcardinal vein; absence of the prerenal segment of the inferior vena cava; infrahepatic interruption of the inferior vena cava with azygos (hemiazygos) continuation; and continuation of the postcardinal vein. The very number and variety of terms used indicates that none adequately explains the exact nature of the anomaly.

This type of anomaly, which we shall refer to as the absence of the superior part of the inferior vena cava with continuation of the azygos vein, is a rare condition and only a few cases (41) have been reported in the literature. In 1958, during a study of macroscopic anatomy, Dr. Zeren and his colleagues [1] at Istanbul University, observed a case in which the upper part of the inferior vena cava was absent and a large azygos vein carried the blood from the lower part of the inferior vena cava to the right atrium.

Pleasants [2], in 1911, wrote a review on the subject of obstructions of the inferior vena cava. In nine of the cases he reviewed, the anomaly was considered to be of congenital origin and effected various parts of the inferior vena cava. In only one of the cases reported by Pleasants was there a complete absence of the upper por-

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From Ankara University Hacettepe Faculty of Medicine and Hacettepe Children's Hospital.

* Resident in pediatric cardiology.

** Assistant Professor in Pediatrics and Chief, Department of Pediatric Cardiology.

tion of the inferior vena cava with continuation of the azygos vein. This condition was verified by autopsy.

In 1951, Effler[3] reported a case in which, during an operation, a large azygos vein was ligated. The patient died, and at the autopsy it was discovered that the upper part of the inferior vena cava was missing and blood was carried by way of the azygos vein, which had been tied off during surgery.

In 1957, Abrams[4] described in detail types of normal and abnormal inferior vena cavas found in newborn infants.

In 1959, Kjellberg[5] encountered three cases of this type of anomaly among 668 patients with congenital heart disease.

An exact diagnosis of this anomaly in living persons became possible only after cardiac catheterization and angiocardiograms were developed as routine procedures. In 1952, Stackelberg[6] made the first diagnosis of absent inferior vena cava by angiocardiogram, and the following year Downing[7] diagnosed a similar condition by cardiac catheterization. As far as can be determined by a study of the literature, only 41 cases are on record as proven cases of this type of anomaly. Therefore, we feel it would be of value to report the case seen at our hospital.

Case Report

K.M., a 25 month old girl, was brought to Hacettepe Children's Hospital with complaints of cyanosis, fatigue, loss of appetite, dyspnea, and frequent upper respiratory tract infections. The patient was hospitalized for the purposes of performing a cardiac catheterization and obtaining an angiocardiogram.

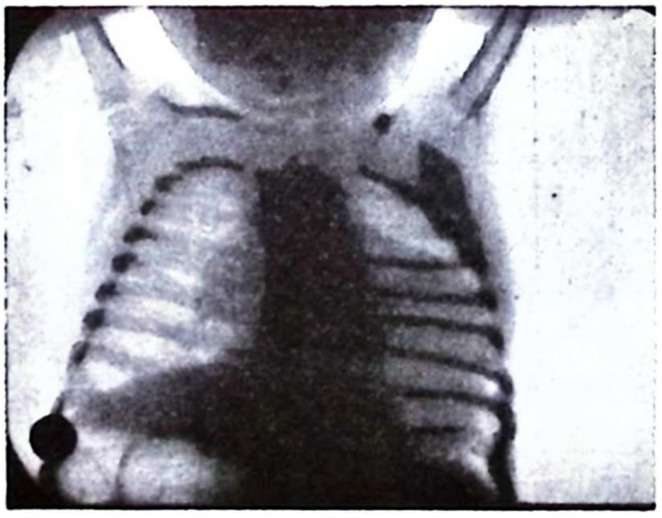

Figure 1. Antero-posterior view of the heart. The heart is enlarged 2 plus, the left borderline is distended and angular. Note the vascular shadow at the right hilar area where the superior vena cava enters the right atrium.

Physical Examination: The patient weighed 12 Kg.; pulse rate was 124 beats per minute; respiration varied between 24 and 28 per minute. The temperature was normal at admission. There was moderate cyanosis and hippocratic fingers and toes. There was a grade 2/IV sistolic murmur which was transmitted to the rest of the precordium. The pulmonic second sound was single.

Roentgen ray examination of the heart in the antero-posterior position revealed a 2 plus enlargement. The left borderline was angular and distended (fig. 1). There was a vascular shadow at the junction of the upper part of the right atrium and the superior vena cava in the hilar area of the lung. This condition was also observed by Downing and others, and was thought to be the end of the azygos vein draining into the superior vena cava. The rest of the vascularity of the lungs consisted of tiny narrow and reticular veins, the «decreased vascularity of the lungs» type. The air shadow of the stomach could be seen at the right side.

In the right anterior oblique position, the esophagus was not displaced by the heart at any point. Above the right ventricle, about two to three cm. of lung could be seen (fig. 2).

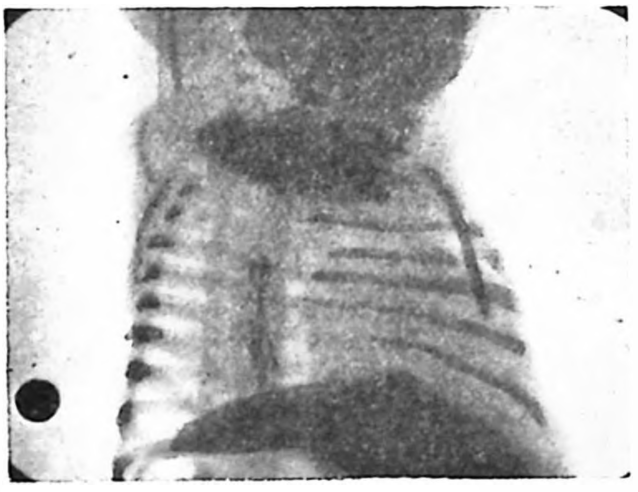

Figure 2. Right anterior oblique view. Two to three cm. of lung can be seen above the right ventricle.

In the left anterior oblique position, the heart shadow was beyond the vertebral column. The tip of this portion of the heart was slightly elevated, indicating a pseudo left ventricular hypertrophy (fig. 3).

An electrocardiogram revealed a heart rate of 105 beats per minute. There was sinus arrythmia and an electrical axis of -110 degrees. The heart was rotated clockwise to a semi-horizontal position. There was an incomplete right bundle branch block, and left atrial enlargement with right ventricular hypertrophy and a possible left ventricular hypertrophy (fig. 4).

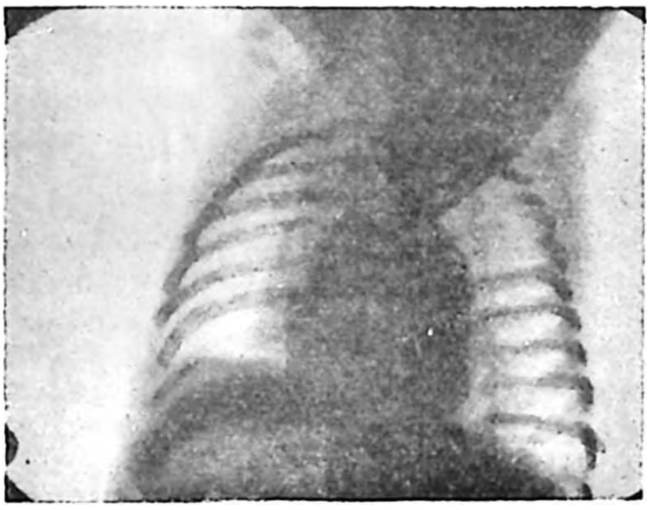

Figure 3. Left anterior oblique view. The heart shadow is beyond the vertebral column and the tip is elevated.

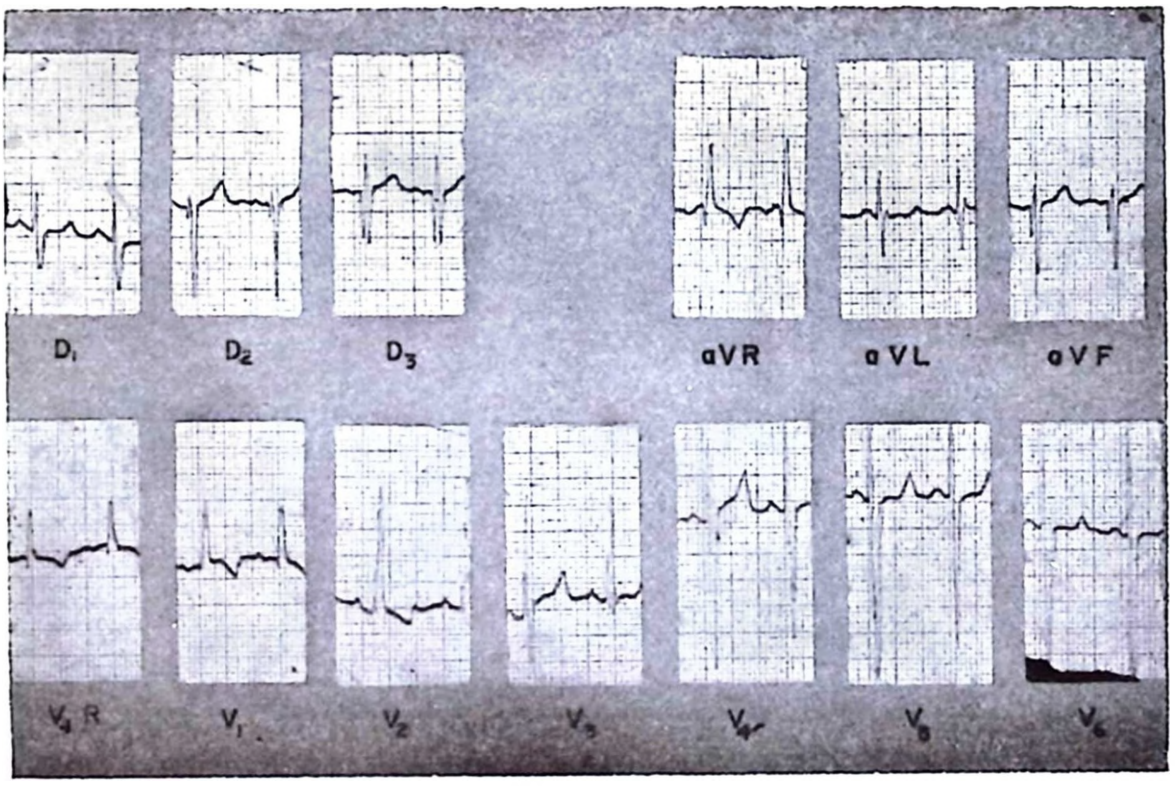

Figure 4.

Cardiac catheterization was performed with a French no. 7 catheter. Introduced via the left vena basilica, it did not penetrate further than the left axillary vein. The same size catheter was then introduced via the large saphenous vein. On each trial the catheter passed through the inferior vena cava, behind the heart shadow, to a point two to three centimeters superior to the right atrium, thence turned downward and entered the right atrium. Several at-

tempts were made via each of the pathways described, with no variation in the results. The angulation was at the right hilar shadow which can be seen in the antero-posterior roentgenogram.

From the right atrium the catheter entered either the hepatic vein, the coronary sinus or the left atrium. We were not able to pass the catheter into the right ventricle, although several attempts were made (figure 5, diagrams I, II, III, and IV). In order to verify

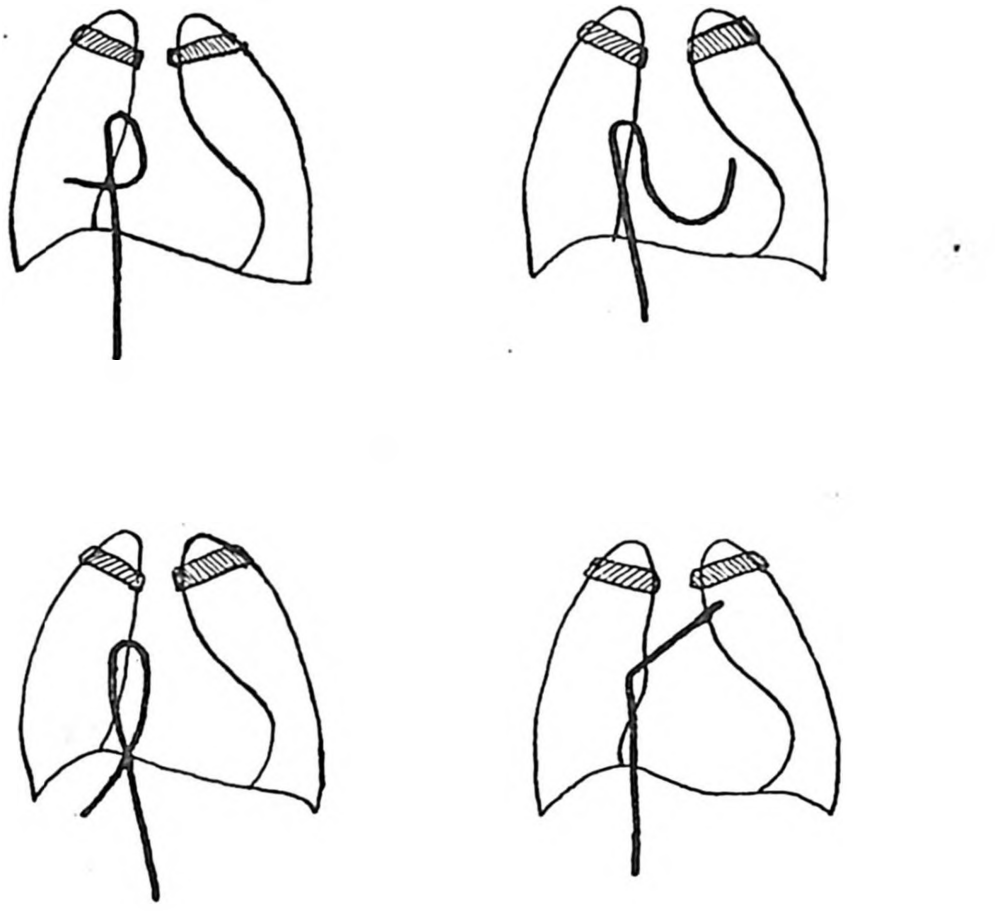

Figure 5. Diagrams I, II, III, IV. Diagram I shows catheter passed from inferior vena cava into azygos vein behind the heart shadow, then into the superior vena cava, right atrium and right pulmonary vein. Diagram III shows the hepatic vein drained into the right atrium directly.

the diagnosis, it was decided to obtain an angiocardiogram. One cc. of dye was injected into the azygos vein at the level of the right atrium, to test for any allergic reaction. Immediately after injection of the dye, the patient developed a severe reaction. Her respiration became irregular, shallow and apneic, and there were twitching and convulsive movements of the extremities. The angiocardiogram was postponed.

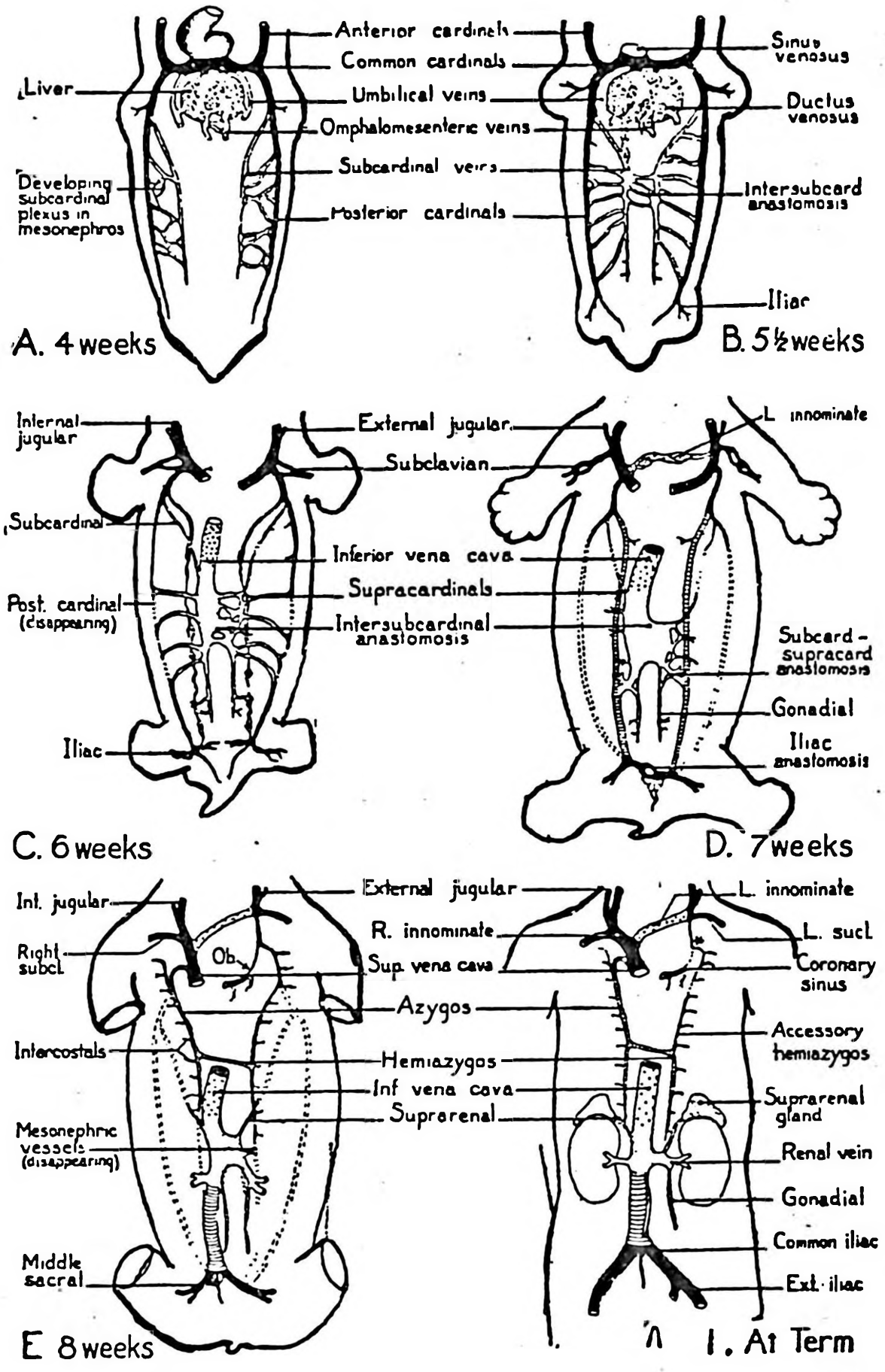

Figure 6. Embryologic development of the inferior vena cava (B.M. Potter Human Embryology, modified).

Embryological Development of the Inferior Vena Cava

The development of the inferior vena cava in the fetus is complicated and subject to considerable variation. Embryologists and anatomists have described this embryologic pattern and its variations, and many complex studies have been made. Figure 6 is a simple explanation of the normal fetal development of the inferior vena cava.

The veins of the caval system are called cardinal veins in the embryo. The inferior vena cava is formed by the fusion and alternation of components of several different primitive venous pathways, and can thus be viewed as consisting of distinct segments. The partial fusion of the two afferent vessels of the omphalomesenteric vein plexi contributes to the formation of the portal vein segment. The prerenal, renal, and postrenal segments of the inferior vena cava develop after numerous anastamoses between the posterior cardinal, subcardinal and supracardinal veins. The azygos - hemiazygos segments are developed from the anterior part of the supracardinal veins. If the intermediate vein, which connects the supracardinal to the subcardinal veins, remains, then the blood will flow into the azygos and hemiazygos veins from the lower part of the inferior vena cava instead of from the lumbar vein. This is the type of anomaly we saw in our patient.

Discussion and Comment

As pointed out by embryologists[8], the development of the inferior vena cava is so complex that a great many variations occur. Since most of the anomalies of this part of the venous system are important, it is essential to recognize them early and attempt to correct them. Further it is important to be able to recognize the associated vascular anomalies and cardiac malformations. Patients with the type of anomaly considered in this report often have congenital cyanotic heart defects. Corbiloculare is common, with endocardial cushioning defect next in frequency. Abnormal positions of the heart, and partial inversion of the abdominal viscera are also common. There have been cases in which older persons were discovered at autopsy to have an anomalous inferior vena cava with azygos continuation, apparently isolated and benign.

The results of physical examinations vary according to the presence or absence of other additional congenital heart anomalies. An electrocardiogram will usually indicate associated cardiac de-

fects. Roentgen ray examination of the heart may reveal an enlarged azygos vein at the junction of the superior vena cava and the right atrium.

Cardiac catheterization by way of the arm veins may be normal except for the impossibility of advancing the catheter into the inferior vena cava. If cardiac catheterization is carried out via the saphenous vein, the right atrium will be entered only from the superior vena cava.

In our patient the hepatic vein drained into the right atrium directly. Since the catheter did not go further into the coronary sinus, this seemed to indicate the absence of the left superior vena cava entering the coronary sinus. There was a normal superior vena cava. The condition was interpreted as a probable tricuspid atresia. The presence of congenital heart malformation in addition to the absence of the upper part of the inferior vena cava with azygos continuation is not uncommon.

Although the malformation is rare, we feel it should be taken into consideration with a patient who is being catheterized via the saphenous vein or the inferior vena cava.

Summary

The authors discuss anomalies of the inferior vena cava and present the case history of a 25 month old girl with absence of the upper part of the inferior vena cava with azygos continuation.

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