where there is herniation of the heart, cineangiography has been used.

In our case, although cineangiography was done to confirm the diagnosis, the possibility of pericardial defect was first suspected by the cardiac blood pool isotope imaging. Filling of the protuberance with isotope on first pass confirmed it to be part of the right atrium, but increase in its size in later phases was not clearly explained, although the possibility of the right ventricle forming part of it was considered. Multiple gated isotope imaging of cardiac blood pool was not performed in this case. Retrospectively, it is considered that if multiple gated imaging was performed, it could have provided the definitive diagnosis by showing the motion of the wall of herniated cardiac chambers, thus possibly obviating the need for cineangiography for diagnosis.

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Partial Anomalous Pulmonary Venous Connection to the Azygous Vein with Intact Atrial Septum*

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A five-year-old girl with partial anomalous pulmonary venous connection to the azygous vein with intact atrial septum is reported. The clinical and roentgenographic features suggested the correct diagnosis. Surgical cor-

rection of this previously unreported defect was accomplished by creating an atrial septal defect and constructing a baffle to direct the blood flow from the azygous vein to the left atrium.

Defects of the atrial septum generally accompany partial anomalous pulmonary venous connection (PAPVC).1,2 Although many cases with intact atrial septum have been reported, we could find no report of an intact atrial septum accompanying PAPVC to the azygous vein. We report such a case here to point out the characteristic clinical and hemodynamic findings and discuss the surgical approach used for correction.

CASE REPORT

An asymptomatic five-year-old girl with a heart murmur since infancy was referred for cardiac evaluation. Examination revealed a prominent left parasternal lift. S1 was normal and S2 was widely split but varied appropriately with respirations. A grade 3/6 medium pitched systolic ejection murmur was heard at the upper left sternal border and an early to mid-diastolic rumble was heard at the lower left sternal border. The remainder of the examination was normal.

An electrocardiogram revealed right axis deviation (+ 130°) and right ventricular hypertrophy. The chest roentgenogram showed cardiac enlargement, increased pulmonary vascularity and prominence of the azygous vein (Fig 1). Right ventricular enlargement and paradox intraventricular septal motion were noted echocardiographically.

Unsuccessful attempts to cross the interatrial septum were made during diagnostic cardiac catheterization from the right femoral vein. The venous catheter passed easily from the superior vena cava to the azygous vein, where the O2 saturation was 90%. A large step-up in O2 saturation was noted between the high superior vena cava and the right atrium. The calculated Qp/Qs was 2.4 to 1. Right ventricular and pulmonary arterial systolic pressures were elevated at 38-40 mm Hg. Mean right atrial and right pulmonary artery wedge pressures were equal at 4 mm Hg. Mean left pulmonary artery wedge pressure was 8 mm Hg. Selective right pulmonary artery angiography showed that all the pulmonary veins from the right lung connected anomalously to the azygous vein (Fig 2). Angiographic visualization via the left pulmonary artery showed normal pulmonary venous connection to the left atrium and no evidence of intracardiac left to right shunting.

The absence of an interatrial septal defect was confirmed during surgical repair. A 2 cm2 defect was created in the cephalad and posterior portion of the interatrial septum. Pericardium and a flap of right atrial wall were used to divert the flow from the enlarged azygous vein to the atrial septal defect. A second pericardial patch was used to enlarge the superior vena cava-right atrial junction. The postoperative course was completely uncomplicated. No dysrhythmias or clinical signs of superior vena caval obstruction developed. The postoperative chest roentgenogram showed a decrease in cardiac size and normal pulmonary vascularity.

DISCUSSION

To the best of our knowledge, PAPVC to the azygous vein with an intact atrial septum has not been reported previously. In our patient, the diagnosis was suspected from the physical findings and the noninvasive labora-
The approach cross sectional cineangiogram (dye injection into anomalous pulmonary veins, i.e., PAPVC) has been diagnostically useful.

When the atrial septum is intact, a number of hemodynamic and anatomic factors will influence the degree of pulmonary blood flow through the anomalous route. These include the number of anomalously draining veins, the relative vascular resistances of the anomalously draining veins and normally connected lung segments, and the pressure and compliance of the respective atria. Normally, the right lung receives about 54 percent of the total pulmonary blood flow. With PAPVC involving the entire right lung and an intact atrial septum, this proportion may approach 66 percent. This is due to the greater compliance of the right atrium as compared to the left, and the increased pulmonary pressure difference across the anomalously draining lung.

The surgical correction was similar to the procedure used in patients with PAPVC to the superior vena cava. A patch was constructed and sewn along the wall of the superior vena cava to include the ostium of the azygous vein. The patch was carried into the right atrium and sewn about the surgically created atrial defect. The anomalously connected veins drained through a tunnel whose posterior wall was made up of the superior vena cava and posterior right atrium and whose anterior wall was constructed from an atrial flap and pericardium. To prevent superior vena cava obstruction, a pericardial gusset was placed at the caval-right atrial junction. The repair resulted in a small amount of systemic venous blood from the azygous vein draining to the left atrium, but this minor hemodynamic abnormality should be well tolerated.

Isolated PAPVC has long been considered a relatively benign condition. It is now recognized, however, that PAPVC with an intact atrial septum may result in progressive pulmonary vascular obstructive disease at an early age. It is recommended, therefore, that these lesions be surgically corrected when the Qp/Qs is 1.7 or greater in the absence of a significantly elevated pulmonary vascular resistance.

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