concentration, which had been high prior to treatment, increased further, probably due to liberation of the antigen resulting from the destruction of the cysts. Only after three months of treatment did the level of IgE concentration begin to fall (Table 1). The rise of IgE was not accompanied by allergic reactions.  

The titer of indirect hemagglutination for hydatidosis, high prior to treatment, fell continuously during the administration of mebendazole. The results of indirect immunofluorescence were less evident: its titer fell during the first months of treatment but later rose again, perhaps due to further liberation of antigen (Table 1). Both tests had remained positive for a long period (even years) notwithstanding successful surgery.  

We are not certain whether the echinococcosis in this patient, though clinically cured, has been eradicated permanently, or whether he will need another course of mebendazole, but the case shows mebendazole to be an effective treatment for patients with disseminated echinococcosis, whose longterm prognosis had been hopeless.

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Partial Right-Sided Congenital Pericardial Defect With Herniation of Right Atrium and Right Ventricle*

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In an unusual case of right-sided pericardial defect with herniation of the right atrium and right ventricle, cardiac blood pool isotope imaging is introduced as a new, noninvasive diagnostic procedure. Surgery which consisted of excision of right-sided pericardium relieved the chest pain which was the only symptom the patient had prior to surgery.

Congenital pericardial deficiency, first described in 1559 by Realdus Columbus,1 had been considered a rare anomaly with the diagnosis seldom being made prior to death. In 1959, Ellis et al2 found 97 cases on reviewing the literature and added two of their own in whom clinical diagnosis was made for the first time. Since the review of Ellis et al,2 the number of cases reported in the literature has steadily increased. According to Chang and Leigh,3 the foramen type of defect is found in about 75 percent of the cases, and in 67 percent, the defect is left-sided. The right-sided

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defect is rare. Of 87 cases reported between 1937 and 1967, Glover et al. found that 77 cases were left-sided and only 5 had a right-sided defect. Since 1967, at least two more cases of right-sided defect have been reported in the literature. In 1965, Chang and Amory reported the first case of right-sided pericardial defect with herniation of the right atrial appendage. To the authors' knowledge, however, there is no reported case with herniation of both right atrium and right ventricle. The purpose of this paper is to report such a case and to introduce a new technique for the diagnosis of this condition.

CASE REPORT

A 39-year-old white man was in excellent health until one month prior to hospitalization when he suffered from a head cold accompanied by right-sided chest pain. The chest pain was intermittent, dull, pressure-like, increasing on deep inspiration and after meals, was localized, and was not related to exertion. Symptoms of cold had disappeared about one week after the onset, although chest pain persisted. There was no family history of congenital heart disease.

Physical examination results were essentially within normal limits, with pulse rate of 60 beats per minute, respiration 20 per minute, and blood pressure, 122/80 mm Hg. There was no evidence of clubbing, cyanosis, or edema. The lungs were clear, and heart size and sounds were normal. No adventitious sounds were heard. Peripheral pulses were normal.

ECG was normal. A chest roentgenogram revealed an abnormal contour of the right cardiac border. A spot film showed a prominence over the right cardiac border just above the diaphragm, and the possibility of pericardial cyst was raised. A cardiac blood pool isotope imaging was performed after intravenous injection of 127mCoI of human serum albumin labeled with radioactive technetium (99mTc). Both the nuclide angiogram and the equilibrium images demonstrated a contour abnormality which filled with isotope simultaneously with the right atrium (Fig 1). Transmission images previously obtained with 57cobalt showed this protuberance to be inseparable from the expected position of the right atrium. The possibility of this to be part of the right atrium and the right ventricle was considered. Cardiac catheterization and angiography were performed. The right and left heart pressures were normal. A cineangiogram with injection of contrast material into the right atrium filmed in a shallow left anterior oblique projection demonstrated that the lateral aspect of the right atrium formed the upper portion of herniation, while the inferior portion was formed by the basal portion of the right ventricle (Fig 2).

Thoracotomy performed four weeks later revealed a foramen-type defect in the right lateral pericardium through which parts of the right atrium and the right ventricle were herniated. The shape of both the atrium and the ventricle appeared distorted, giving the appearance of a longstanding herniation. An attempt to suture the defect failed because of its large size and resultant increased compression of the heart. Therefore, the pericardium on the right side was completely excised, freeing the herniated heart. Postoperatively, the patient's course was entirely benign. The chest pain was relieved after the surgery.

DISCUSSION

Congenital partial pericardial defects are generally regarded as asymptomatic. In some cases, vague chest pain occasionally resembling angina pectoris has been described. Ellis et al. reported two cases where no other cause of chest pain was found. Herin et al. described a case of chest pain relieved by enlargement of the defect through which the left atrial appendage had become incarcerated. Three cases of herniation of heart resulting in sudden death have been described.

Our patient had intermittent right-sided chest pressure which increased on deep inspiration. Since no other cause was found, and the chest pressure was relieved after surgery, it is presumed that the cause of chest pain was cardiac herniation. It seems likely that worsening of the chest pain during deep inspiration was related to an increase in the size of herniation secondary to increased venous return to the right side of the heart during deep inspiration.

DIAGNOSTIC CONSIDERATIONS

The radiologic findings in most cases provide a clue to the diagnosis, and cardiac herniation through a foramen-type defect may be seen as an abnormal configuration of the heart on the chest roentgenogram. Confirmation of the diagnosis has been made in the past by diagnostic artificial pneumothorax. In cases

Figure 1. Cardiac pool study demonstrating abnormal contour of right cardiac margin filling simultaneously with right atrium and right ventricle. SVC indicates superior vena cava; PA, pulmonary artery.

Figure 2. Cineangiogram showing right atrium (RA) and right ventricle (RV) herniated through pericardial defect.
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 correction 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). 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. $S_2$ was normal and $S_1$ 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 paradoxic interventricular 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 O$_2$ saturation was 90%. A large step-up in O$_2$ 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 cm$^2$ 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-