Fig. 1. General appearance of the malformation in the frontal view. 1, Superior vena cava; 2, inferior vena cava; 3, left superior vena cava; 4, malaligned atrial septum and ostium primum defect.

restrictive and accounts for elevation of left atrial pressure. In accordance with Carpentier and colleagues,⁵ when the papillary muscles are basically normal, congenital mitral valve stenosis may be due to papillary muscle commissure fusion, excess valvular tissue, hypoplasia of the anulus, or supra-valvular ring. In this case, the supra-valvular ring was incomplete, and most of the stenosis was due to the overriding restrictive ostium primum.

REFERENCES

1. Van Mierop LHS. Pathology and pathogenesis of endocardial cushion defects: surgical implications. In: Davila JC, ed. Second Henry Ford Hospital International Symposium on Cardiac Surgery. New York: Appleton-Century-Crofts, 1977:201-7.
2. Corwin RD, Singh AK, Karlson KE. Double outlet right atrium: a rare endocardial cushion defect. *Am Heart J* 1983;106:1156-7.
3. Alivizatos P, Anderson RH, Macartney FJ, Zuber-buller JR, Stark J. Atrioventricular septal defect with balanced ventricles and malaligned atrial septum: double-outlet right atrium. *J THORAC CARDIOVASC SURG* 1985;89:295-7.
4. Nunez L, Gil-Aguado M, Sanz E, Perez-Martinez V. Surgical repair of a double outlet right atrium. *Ann Thorac Surg* 1984;37:164-6.
5. Carpentier A, Branchini B, Cour JC, et al. Congenital malformations in the mitral valve in children. *J THORAC CARDIOVASC SURG* 1976;72:854-66.

CONGENITAL TOTAL ABSENCE OF THE PERICARDIUM: CASE REPORT OF A 72-YEAR-OLD MAN AND REVIEW OF THE LITERATURE

Ayman J. Hammoudeh, MD, Michael E. Kelly, MD, FACC, and Haroutune Mekhjian, MD, FACS, FACC,
Paterson and South Orange, N.J.

Congenital absence of the pericardium (CAP) is rarely encountered during clinical practice. Most patients re-

main free of symptoms, and the diagnosis is made intra-operatively or at postmortem examination.¹ Some cases have been diagnosed by various imaging techniques, ranging from plain x-ray films to nuclear magnetic resonance. We report the case of a man with coronary artery disease (CAD) who underwent stress thallium scan. The scintigraphic images showed abnormal left ventricular (LV) orientation and levorotation. Intraoperatively, CAP was diagnosed.

Case report. This 72-year-old man with hypertension reported episodic chest pain, dyspnea, and palpitations for

From the Departments of Cardiology and Cardiothoracic Surgery, Saint Joseph's Hospital and Medical Center, Paterson, N.J., and Seton Hall University School of Graduate Medical Education, South Orange, N.J.

J THORAC CARDIOVASC SURG 1995;109:805-7

Copyright © 1995 by Mosby-Year Book, Inc.

0022-5223/95 \$3.00 + 0 12/8/59157

Fig. 1. Twelve-lead electrocardiogram from patient illustrating sinus bradycardia, right axis deviation, and clockwise rotation of QRS complexes in precordial leads.

about 20 years. Coronary angiography and cardiac catheterization in May 1993 revealed triple-vessel CAD and mild aortic stenosis. Because the major coronary lesions were mostly distal in location, medical management was undertaken. Chest pain recurred 2 months later and stress thallium scan was recommended. Pre-exercise examination revealed a blood pressure of 140/60 and a heart rate of 58 beats/min, and the cardiac apex was shifted to the left with an ejection systolic murmur at the left sternal border. The electrocardiogram (Fig. 1) revealed sinus bradycardia, right axis deviation, left posterior hemiblock, and QRS complex clockwise rotation. The patient exercised for 6 minutes (modified Bruce protocol) and achieved a highest exercise level of 1.7 mph at 5% grade for 3 minutes during which he had angina. The blood pressure dropped from 170/70 to 150/60 before he reached 85% of age-predicted target heart rate. No electrocardiographic changes were noted. Thallium-201 was injected 45 seconds before exercise termination and imaging was done immediately and 4 hours later in the anterior and 40- and 60-degree left anterior oblique views. The scintigraphic images showed cardiac levorotation and abnormal LV orientation with no increased lung uptake or LV dilation. Delayed tracer uptake was noted in the anterolateral wall.

On the basis of the poor effort tolerance, angina, and failure to sustain blood pressure the patient was referred for coronary artery bypass grafting. Intraoperatively, the heart was displaced to the left hemithorax with total absence of the pericardium. There was severe CAD with a left main coronary artery ostial lesion and a heavily calcified aortic valve with sclerotic leaflets and anulus. A large lingula and right lung accessory lobe were noted. Triple saphenous vein graft bypass and

aortic valve replacement with a Carpentier-Edwards porcine valve (Baxter Healthcare Corp., Santa Ana, Calif.) were done.

Discussion. Four decades after Columbus reported the first case of CAP in 1559,¹ Ellis, Leeds, and Himmelstein,² in 1959, described the roentgenologic sign of left-sided pericardial absence. CAP takes place in the eighth embryonic week when premature atrophy of the left common cardiac vein (Cuvier's duct) compromises the blood supply to the pleuropericardial membrane with subsequent formation of various degrees of CAP.¹ Complete or almost complete CAP is more common than partial pericardial defects.³ The loss of adequate cardiac support may explain the chest pain and palpitations. Shortness of breath and syncope are other potential symptoms.^{4,5} Although partial pericardial defects are potentially lethal because of myocardial herniation and incarceration in the young,⁶ total absence of the pericardium can remain asymptomatic and can be compatible with long life. To our knowledge, CAP was reported only in one patient (77 years old) older than ours.⁷

Patients with absent left pericardium have a leftward cardiac apex shift with systolic murmur. The electrocardiogram typically shows the same findings seen in our patient.⁸ On chest x-ray film, the heart is shifted to the left hemithorax with a large pulmonary trunk.⁹ The old diagnostic procedure of inflicting a left-sided pneumothorax to outline the pericardium¹⁰ is now obsolete because of the availability of echocardiography, computed tomographic scanning, and nuclear magnetic resonance imaging.^{9, 11, 12} CAP is associated with other cardiac and pulmonary anomalies in about 30% of the cases.

CAP should be suspected when myocardial perfusion scintigraphy shows abnormal cardiac position and LV orientation.

REFERENCES

1. Saito R, Hotta F. Congenital pericardial defect associated with cardiac incarceration: case report. *Am Heart J* 1980;100:866-70.
2. Ellis K, Leeds NE, Himmelstein A. Congenital deficiencies in parietal pericardium: a review of two new cases including successful diagnosis by plain roentgenography. *Am J Roentgenol* 1959;82:125-37.
3. Southworth H, Stevenson CS. Congenital defects of the pericardium. *Arch Intern Med* 1938;61:223-9.
4. Pernot C, Hoeffel JC, Frisch HR, Brauer B. Partial left pericardial defect with herniation of the left atrial appendage. *Thorax* 1972;27:246-50.
5. Robin E, Ganguli SN, Fowler MS. Strangulation of the left atrial appendage through a congenital partial pericardial defect. *Chest* 1975;67:354-5.
6. Altman CA, Ettedgui JA, Wozney P, Beerman LB. Noninvasive diagnostic features of partial absence of the pericardium. *Am J Cardiol* 1989;63:1536-7.
7. Glancy DL, Sanders CV, Porta A. Posterior chest wall pulsation in congenital complete absence of the left pericardium. *Chest* 1974;65:564-6.
8. Kansal S, Roitman D, Sheffield LT. Two-dimensional echocardiography of congenital absence of pericardium. *Am Heart J* 1985;109:912-5.
9. Schiavone WA, O'Donnell JK. Congenital absence of the left portion of parietal pericardium demonstrated by nuclear magnetic resonance imaging. *Am J Cardiol* 1985;55:1439-40.
10. Glover LB, Barcia A, Reeves TJ. Congenital absence of the pericardium: a review of the literature with demonstration of a previously unreported fluoroscopic finding. *Am J Roentgenol* 1969;106:542-9.
11. Nasser WK, Helman C, Tavel ME, Feigenbaum H, Fisch C. Congenital absence of the left pericardium: clinical, electrocardiographic, radiographic, hemodynamic and angiographic findings in six cases. *Circulation* 1970;41:469-78.
12. Burrows PE, Smallhorn JS, Trusler GA, Daneman A, Moes CAF, Rowe RD. Partial absence of the left parietal pericardium with herniation of the left atrial appendage: diagnosis by cross-sectional echocardiography and contrast-enhanced computed tomography. *Pediatr Cardiol* 1987;8:205-8.

INTENTIONAL DELAYED REPAIR OF ACUTE DISSECTION OF THE ASCENDING AORTA COMPLICATED BY STROKE

William Piccione, Jr., MD, Ian N. Hamilton, MD, and Hassan Najafi, MD, *Chicago, Ill.*

It is generally accepted that patients with acute dissection of the ascending aorta require urgent surgical reconstruction. However, the optimal treatment of patients with acute ascending aortic dissection complicated by stroke remains controversial.¹ The group at Stanford has suggested that stroke does not negate urgent surgical intervention, whereas others consider stroke a relative, if not absolute, contraindication to surgery.

We report the case of a 42-year-old man with phenotypic features of Marfan's syndrome who was taken to a suburban hospital with anterior chest pain and an acute left hemispheric cerebrovascular accident manifested by right hemiplegia and dysarthria. Magnetic resonance

scanning confirmed the presence of an ischemic left hemispheric infarct. Carotid noninvasive assessment demonstrated thrombosis of the right carotid artery along with an intimal flap in the left carotid artery. Computed tomography and magnetic resonance scanning confirmed the presence of an ascending aortic dissection involving the arch with extension into the brachiocephalic vessels (Fig. 1). The patient was transferred to our facility for emergency surgery.

On arrival in our institution the patient had a dense left hemispheric deficit with dysarthria, flaccid paralysis of the right leg, and severe weakness of the right arm. He was in hemodynamically stable condition with no right carotid pulse and mild aortic insufficiency by echocardiogram. The electrocardiogram showed no abnormalities.

Because of the patient's severe neurologic insult and hemodynamic stability, we elected to delay surgical intervention. He was treated with antihypertensive drugs and β -adrenergic blockade. He subsequently underwent physical therapy, with a marked recovery of motor function.

From the Department of Cardiovascular Thoracic Surgery,
Rush-Presbyterian-St. Luke's Medical Center, Chicago, Ill.

J THORAC CARDIOVASC SURG 1995;109:807-8

Copyright © 1995 by Mosby-Year Book, Inc.

0022-5223/95 \$3.00 + 0 12/8/61482