Case Reports

Severe Hypoxia Related to Uncomplicated Atrial Septal Defect

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A 73-year-old man was admitted to our institution with severe hypoxia. Cardiac catheterization and transesophageal echocardiography revealed an atrial septal defect with an interatrial right–to–left shunt but with no pulmonary hypertension. Direct examination at surgery revealed an elongated thoracic aorta that caused the aortic annulus to reside at the level of the diaphragm and the heart to be positioned transversely in the mediastinum. Surgical closure of the atrial septal defect normalized oxygenation. We attribute the unusual occurrence of an atrial septal defect and a right–to–left shunt in the absence of pulmonary hypertension to the spacial and mechanical changes caused by the patient's elongated aorta and by the transverse position of the heart. ***(Texas Heart Institute Journal 1993;20:123-5)***

Atrial septal defect with cyanosis in the absence of pulmonary hypertension has been reported in association with pulmonic stenosis, tricuspid regurgitation, hypoplastic right ventricle, right ventricular infarction, and altered right ventricular compliance.[1,2]

Right–to–left shunting can be caused by conditions such as mechanical ventilation that elevate right-heart pressures (particularly after heart surgery)[3] or in conjunction with adult respiratory distress syndrome.[4] Mechanical factors may also cause right–to–left shunting. Eustachian and thebesian valves have been reported to divert blood from the inferior vena cava and the coronary sinus through an atrial septal defect.[5,6] A right–to–left shunt may also occur after a right pneumonectomy, during which procedure the angulation of the septum is changed, bringing the septum closer to the inferior vena cava.[7-9]

We describe our experience with a patient who presented with severe hypoxia and was found to have an atrial septal defect and an interatrial right–to–left shunt in the absence of pulmonary hypertension.

Case Report

A 73-year-old man was admitted to our institution because of shortness of breath, tightness in his chest, and cyanosis. He was placed on 100% inspiratory oxygen. His oxygen partial pressure was 43 mmHg, and his arterial oxygen saturation was 81%. His medical history included no cardiac or pulmonary disease. He had never smoked, but he had a long history of hypertension. His vital signs were normal, with a hemoglobin level of 17.6 g/dL and a hematocrit level of 50 mL/dL. The only abnormal feature shown by chest radiography was an elongated, tortuous thoracic aorta (Fig. 1). The patient's lung scan was normal, and a pulmonary arteriogram revealed normal pulmonary artery pressure and no evidence of pulmonary emboli.

Cardiac catheterization disclosed an atrial septal defect with an interatrial right–to–left shunt. Right- and left-heart chamber pressures were normal, as were the pulmonary artery pressures. The Qp was 2.76 L/min; the Qs, 3.53 L/min; and the Qp/Qs ratio was 0.783. Injection of microbubbles into the right atrium was monitored by transesophageal echocardiography and showed prompt passage of the gas into the left atrium. Repeat cardiac catheterization confirmed these findings.

The patient was taken to surgery. With the patient under cardiopulmonary bypass, hypothermia was induced to a level of 32 °C. The heart was exposed through a median sternotomy. Immediately apparent was a markedly elongated aorta with

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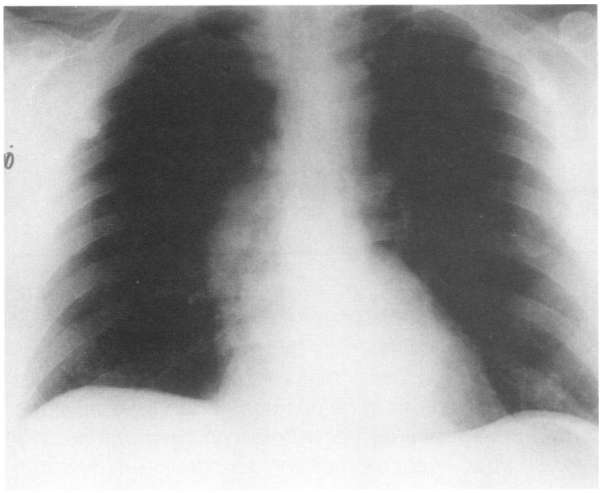

Fig. 1 *Chest radiography reveals elongation and tortuosity of the ascending aorta.*

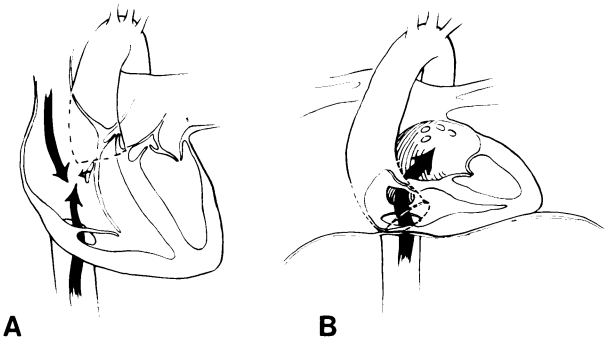

Fig. 3 *This illustration demonstrates the location of an atrial septal defect* ***A****) in a normally positioned heart and* ***B****) in the patient described. Elongation of the aorta has distorted the location of the atrial septum so that the atrial septal defect lies just above the inferior vena cava.*

the aortic annulus situated at the tip of the dome of the right diaphragm; the heart was positioned transversely in the mediastinum. Exploration of the right atrium revealed a 2-cm atrial septal defect that lay transversely within 2 cm above the inferior vena cava. Careful inspection of the atrium near the inferior vena cava revealed no evidence of a persistent eustachian valve or other anatomic abnormality.

The patient's postoperative course was without complication. On the 5th day after surgery, his room–air oxygen saturation was 96%.

Discussion

Our patient, admitted to the hospital with severe hypoxia, was found to have an atrial septal defect and a right–to–left interatrial shunt in the absence of pulmonary hypertension. We believe that a mechanical or spacial factor may have been responsible for the right–to–left shunt. The elongated thoracic aorta caused the aortic annulus to be situated at the tip of to the dome of the right diaphragm (Fig. 2). The position of the patient's heart was thus distorted so that the atrial septum, and therefore the atrial septal defect, lay just above the inferior vena cava (Fig. 3).

Although it is impossible to ascertain the pathophysiology of the right–to–left shunt (either in our patient or in others reported), we found that closing the defect resolved the cyanosis and hypoxia completely in our patient, which agrees with results in similar cases described.

We conclude that, in certain patients with an atrial septal defect, a right–to–left shunt can occur in the absence of either pulmonary hypertension or other obvious anatomic abnormalities. If the shunt is hemodynamically important, surgical correction is likely to resolve the accompanying hypoxia in similar patients.

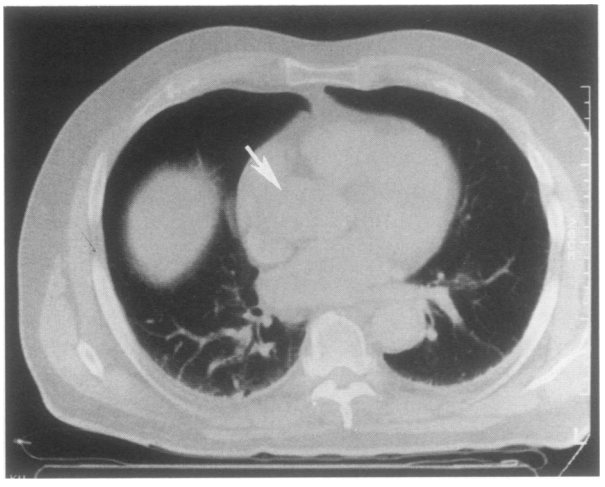

Fig. 2 *A computed tomographic scan of the chest shows the ascending aorta (above the aortic valve) at the same level as the tip of the dome of the right diaphragm.*

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