Platypnea Syndrome After Left Pneumonectomy*

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Contrast two-dimensional echocardiography (2DE) was used to demonstrate right-to-left shunting at the atrial level in a 49-year-old man with platypnea and orthostatic cyanosis which developed after a left pneumonectomy. This patient's systemic arterial saturation decreased with phlebotomy and increased with volume administration. This syndrome disappeared after repair of a previously unrecognized atrial septal defect. Right-to-left shunting in atrial septal defect is usually explained by a change in the relationship of right and left ventricular compliance with the right ventricle becoming less compliant (ie, stiffer) than the left. Pneumonectomy can affect atrial emptying either directly by mechanical means or indirectly by changing relationships in ventricular compliance. Contrast 2DE played a key role in initially establishing the etiology of cyanosis in this complicated case.

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Platypnea and orthostatic cyanosis is a rare syndrome seen after pneumonectomy in patients with unrecognized septal defects.1,2 As illustrated herein, its importance lies in the fact that it is completely reversible and need not be associated with pulmonary hypertension or progression of pulmonary disease.

Our report describes the clinical course of a patient with a right-to-left shunt which manifested itself after left pneumonectomy; it illustrates the usefulness of two-dimensional echocardiography (2DE) in the preoperative and postoperative evaluation of intracardiac right-to-left shunts.

The mechanisms of right-to-left shunting in two prior cases is not well understood. It has been suggested2 that right pneumonectomy played a role in the genesis of shunting. This patient, however, is the only reported case of right-to-left shunting after left pneumonectomy and illustrates that right-to-left shunting does not depend on right pneumonectomy.

CASE REPORT

A 49-year-old white man underwent left pneumonectomy at another hospital for squamous cell carcinoma on April 12, 1977. Two months after surgery, routine arterial blood gas (room air) determinations showed $\text{PO}_2$, 63 mm Hg; $\text{PCO}_2$, 27 mm Hg; and pH, 7.51. Five months later, he noted gradual onset of dyspnea exacerbated by sitting or standing. After admission to another hospital, arterial blood gas levels were as follows: $\text{PO}_2$, 36 mm Hg; $\text{PCO}_2$, 32 mm Hg; and pH, 7.49 (room air), and $\text{PO}_2$, 51 mm Hg; $\text{PCO}_2$, 35 mm Hg; and pH, 7.48 on 40 percent $O_2$ via Ventimask. A lung scan showed a possible segmental defect in the right lung and heparin...
therapy was begun. The arterial $P_{O_2}$ gradually deteriorated; because of the possibility of an intracardiac shunt, the patient was transferred to our hospital.

Physical examination showed a thin cyanotic white man in moderate respiratory distress. Blood pressure was 92/60 mm Hg, heart rate was 110 beats per minute, and respiratory rate, 24. The right lung was normal, but there was percussion dullness and absent breath sounds on the left. The PMI was in the anterior axillary line; $S_1$ was normal, and $S_2$ had an accentuated pulmonic component. No murmur was heard. Dyspnea and cyanosis were observed to improve in the supine position.

The ECG showed sinus tachycardia and right ventricular hypertrophy with strain. Chest x-ray film showed opacified left hemithorax with leftward shift of the mediastinum. Arterial blood gas levels on 40 percent $O_2$ via Ventimask were as follows: $P_{O_2}$, 29 mm Hg; $P_{CO_2}$, 19 mm Hg; and pH, 7.52. The 2DE (Fig 1, left panel) demonstrated enlarged right and left ventricles. A 10-ml bolus of normal saline solution injected in a peripheral vein produced a cloud of echoes appearing initially in the right and then left atrium, and subsequently in both ventricles indicating the presence of a right-to-left shunt at the atrial level (Fig 1, middle panel).

Cardiac catheterization confirmed the presence of right-to-left shunting in the absence of pulmonary hypertension. There was early appearance of green dye in the arterial system after right atrial injection. The mean right atrial pressure was 0 mm Hg, and the pulmonary artery pressure was 16/4 (mean 8). Arterial oxygen saturation of 93 percent (room air) fell to 86 percent after 150 ml phlebotomy and rose again to 93 percent after the injection of 45 ml of contrast medium for ventriculography. A similar increase in arterial oxygen saturation was noted with the infusion of large volumes of normal saline solution. Ultimately, he was given 10 to 15 L normal saline per day to maintain an arterial oxygen saturation of 98 percent on 50 percent $O_2$ via Ventimask. Because of his precarious clinical status, additional studies to elucidate physiologic mechanisms of right-to-left shunting were not carried out.

At surgery, a 4 sq cm secundum atrial septal defect (ASD) was found and repaired by primary closure without complication. Arterial blood gas levels (16th postoperative day) were as follows: $P_{O_2}$, 87 mm Hg; $P_{CO_2}$, 32 mm Hg; and pH, 7.46 on room air. Postoperatively, a 2DE with saline bolus injection demonstrated the appearance of echoes in the right atrium and right ventricle only (Fig 1, right panel). One year later, the patient continued to feel well with no recurrence of symptoms.

**DISCUSSION**

The presence of cyanosis and right-to-left shunting in patients with ASD has traditionally been regarded as indicative of pulmonary hypertension and a relative contraindication to operative repair. However, there have been case reports of right-to-left shunting without pulmonary hypertension as a result of streaming of inferior vena caval blood into the left atrium across an ASD or as a result of a persistent left superior vena cava terminating in the left atrium.

This is the third reported case of an unrecognized ASD manifesting itself as a right-to-left shunt without pulmonary hypertension within the first seven months after pneumonectomy.

The pathophysiology of right-to-left shunting through an ASD in the postpneumonectomy patient is not well understood. Two previous patients reported had right pneumonectomy and the mechanical factor of a right hydrothorax placing increased extrinsic pressure on the right atrium was invoked as contributing to right-to-left shunting. Although mechanical factors cannot be excluded in our patient, our case indicates that right-to-left shunting is not limited to patients with right pneumonectomy.

Altered right heart compliance may also account for the effects of volume infusion as well as postural change on right-to-left shunting. Normal left and right ventricular diastolic compliance curves from animal studies are shown in Figure 2A. As Dexter observed, shunting across atrial septal defects occurs mainly in diastole and is determined by the difference between left and right ventricular compliance with shunting from the less compliant (stiffer) to the more compliant chamber. This would imply that in our patient, at low volumes, right ventricular compliance was less than left ventricular compliance, resulting in right-to-left shunting. As intravascular volume is increased, the curves would be expected to be closer together and the amount of shunting would decrease. At a point where the curves meet, one would predict no shunting across the defect. Thus, the postulated compliance curves in Figure 2B would explain the observed alteration in shunting with a volume load.

Mechanical alterations in right atrial configuration induced by pneumonectomy might favor right-to-left shunting. Streaming of venous blood across the ASD could account for significant right-to-left shunting without invoking altered ventricular compliance. However, this has hitherto only been observed in inferior, posterior ASDs rather than the secundum-type defect found here.

Two-dimensional echocardiography with saline solution injection was the key initial test used to confirm the diagnosis of ASD with right-to-left shunting in this patient. This technique provided a definitive, noninvasive demonstration of the presence and location of right-to-
left shunting and should be considered in the postpneumonectomy patient who develops unexplained cyanosis.

REFERENCES


Normal Left Ventricular Ejection Fraction in Systemic Arteriovenous Fistula*

Implications for the Use of Noninvasive Methods in Differential Diagnosis of Heart Failure

Robert Arnold Johnson, M.D., and Charles A. Boucher, M.D.

Heart failure is frequently a result of impaired systolic ventricular performance. We describe a patient with heart failure and a systemic arteriovenous fistula who had left ventricular enlargement and a normal left ventricular ejection fraction, demonstrated by gated cardiac blood pool scanning. The heart failure was relieved by surgical repair of the fistula, but the left ventricular ejection fraction did not change. These observations show that volume overload rather than impaired systolic function was the mechanism by which heart failure occurred in this patient with a systemic high output state and illustrate the value of gated cardiac blood scanning in noninvasively elucidating the mechanism of heart failure.

In this report, we describe a patient with severe biventricular failure resulting from a spontaneous systemic arteriovenous fistula. This report is one of a small number of reports that document that the left ventricular ejection fraction can be maintained despite volume-overload-induced heart failure, and demonstrates the importance of measurement of the left ventricular ejection fraction in distinguishing heart failure caused by intrinsic myocardial disease from heart failure caused by other mechanisms.

CASE REPORT

A 62-year-old man was admitted to the hospital because of six months of progressive lower extremity edema, orthopnea, and exertional dyspnea. Blood pressure was 17.3/9.3 kPa (130/70 mm Hg) and heart rate was 52 beats/min. Carotid pulses were brisk. The apex impulse was diffuse and laterally displaced, and a sternal impulse was present. A third heart sound was audible. A grade 3/6 mid-systolic murmur was noted at the upper left sternal border. Pitting edema extended to the thighs. A continuous murmur was audible in the right lower abdominal quadrant. The plain chest film showed cardiomegaly, bilateral pleural effusion, and interstitial pulmonary edema. The ECG showed nonspecific ST-segment and T-wave abnormalities.

Bedrest, digitalis, and increasing doses of furosemide produced only modest improvement. Abdominal aortography revealed a right common iliac artery atherosclerotic aneurysm that connected to the right common iliac vein by a fistula (Fig 1). A multigated cardiac blood pool scan demonstrated marked right and left ventricular enlargement (Fig 2); the left ventricular ejection fraction was 0.84. Because of the normal left ventricular ejection fraction, volume overload produced by the arteriovenous fistula was suspected to be the cause of heart failure.

A 9x9 cm saccular atherosclerotic aneurysm of the right common iliac artery was resected; closure of the arteriovenous fistula was performed, using a velour graft. Table 1 shows the hemodynamic findings immediately before and after surgery, measured by means of a balloon-tipped pulmo-

![Figure 1](image-url)

**Figure 1.** An aortogram showing a right iliac artery (R. Iliac A.) aneurysm. The inferior vena cava (IVC) opacifies early because it receives blood from a fistulous connection (not shown) between the aneurysm and the iliac vein.

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