Contrast Transesophageal Echocardiography in the Diagnosis and Localization of Diffuse Pulmonary Telangiectasias*

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Pulmonary arteriovenous fistulas are rare, usually related to the Rendu-Osler-Weber syndrome, and are detected by chest CT scan or pulmonary angiography. In a 14-year-old boy without Rendu-Osler-Weber syndrome, but with clinical evidence of a right-to-left shunt, ancillary diagnostic studies were negative for pulmonary arteriovenous fistulas, and the final diagnosis was made by contrast transesophageal echocardiography. Saline echo-contrast medium injected peripherally was seen emerging from each pulmonary vein and filling the left chambers. These findings, in light of other negative test results, established the diagnosis of diffuse telangiectasias at the capillary level in both lungs. After 6 years of medical therapy, the patient remains cyanotic but functions well.

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Key words: arteriovenous malformation; contrast echocardiography; hereditary hemorrhagic telangiectasia; intrapulmonary shunts; pulmonary telangiectasias; Rendu-Osler-Weber syndrome; transesophageal echocardiography

Abbreviations: AVM = arteriovenous malformation; TEE = transesophageal echocardiography

Telangiectasias are vascular dysplasias consisting of dilated, convoluted venules connected directly to arterioles. They are found in many diseases and in some healthy persons. Typically, these 1- to 3-mm arteriovenous fistulas have a predilection for the skin, buccal mucosa, tongue, and conjunctiva.

Likewise, pulmonary arteriovenous malformations (AVMs) are direct, low-pressure, artery-to-vein connections of the lung, also lacking capillaries but much larger.1,2 Pulmonary AVMs are infrequent and are associated with Rendu-Osler-Weber syndrome in 47 to 60% of cases.1,3 Many clinical complications are described, including cyanosis, dyspnea, high-output heart failure, hemoptysis, cerebral abscesses, and stroke.1,3 Most commonly, pulmonary AVMs are large and multiple; appear in both lungs, preferentially in the lower lobes1–3; are visible on chest radiography and fluoroscopy, angiography, or high-resolution CT; and are amenable to surgical resection or embolization.1,2

In contrast, telangiectasias at the pulmonary parenchymal level are widely distributed, occur with unknown frequency, and are not detected by the methods usually used for the diagnosis of pulmonary AVMs, such as angiography and chest CT scan.1,2 Contrast echocardiography has been useful as a diagnostic tool in this situation.4

We report a case in which contrast transesophageal echocardiography (TEE) was used to detect the intrapulmonary shunt and to establish the anatomic diagnosis of diffuse, bilateral arteriovenous fistulas, consistent with diffuse pulmonary telangiectasias.

Case Report

A 14-year-old boy was referred to the Mayo Clinic for evaluation of cyanosis and dyspnea on exertion. Cyanosis was first noticed at age 4 years, when he was found to be polycythemic. After clinical evaluation, the patient was deemed healthy. During the remaining childhood years, he was well except for occasional periods of fingertip cyanosis, especially after exertion.

Two years before his referral, the patient experienced increased dyspnea and reduced exercise tolerance. His identical twin was healthy. The family history was negative for epistaxis and telangiectasias.

On physical examination, the patient was comfortable with the following signs: height, 175.4 cm; weight, 70.65 kg; BP, 120/90 mm Hg; and pulse, 80 beats/min. There was marked cyanosis of...
the fingertips, nose, and nail beds, without clubbing. Small telangiectasias were noted on the neck, chest, shoulders, and tip of the nose. There was mild gynecomastia, but there were no other stigmata of chronic liver disease. The heart and lungs sounded normal on auscultation. The rest of the findings from the physical examination were normal.

The results of laboratory evaluations revealed a hemoglobin level of 19.2 g/dL and a hematocrit of 55%. The results of liver tests were normal, as were those of a chemistry panel and hemoglobin electrophoresis.

Arterial blood gas levels were measured on room air and on 100% oxygen in both the supine and the standing positions. The shunt was calculated at 9.6% supine and 18.1% standing.

Pulmonary function tests demonstrated moderate exercise-induced bronchospasm that was reversible with bronchodilators.

The results of electrocardiography and abdominal Doppler ultrasonography were normal. A chest roentgenogram revealed a possible mild increase in pulmonary vascularity. The findings of a chest CT scan were normal. An outside pulmonary angiogram demonstrated a subtle increase of the transit of contrast medium from the arterial to the venous phase but no evidence of pulmonary AVMs.

The findings of transthoracic echocardiography were consistent with a normal heart but were limited because of hyperinflated lungs. During TEE, no heart defects were observed, but the injection of saline echo-contrast medium into a right dorsal hand vein demonstrated severe intrapulmonary shunting. Four or five cardiac cycles after initial opacification of the right atrium and right ventricle, the left atrium and, subsequently, the left ventricle became densely opacified (Fig 1). Each of the four pulmonary veins then was assessed individually. After each additional injection, the observed pulmonary vein showed dense echo contrast (Fig 2), confirming the intrapulmonary source of the right-to-left shunt. This observation is pathognomonic of diffuse pulmonary arteriovenous fistula.

In conjunction with the negative angiographic results, these findings were consistent with intrapulmonary right-to-left shunt secondary to diffuse telangiectasias. The diagnoses, therefore, were hyperreactive airway disease and diffuse intrapulmonary telangiectasia.

Six years later, the patient remains cyanotic during minimal exertion and sometimes at rest. He is polycythemic, requiring occasional phlebotomies to maintain an hematocrit at 55%. Furthermore, he uses oxygen at night and bronchodilators for wheezing and intermittent exacerbations of dyspnea.

**DISCUSSION**

Intrapulmonary shunting has been recognized in association with chronic liver disease (hepatopulmonary syndrome), hemorrhagic hereditary telangiectasia, and, very rarely, spontaneously occurring intrapulmonary telangiectasias.

In this instance of spontaneously occurring intrapulmonary telangiectasias, contrast TEE was used to diagnose diffuse pulmonary parenchymal arteriovenous fistulas when the results of other diagnostic technologies, including pulmonary angiography, which is usually considered the definitive procedure for the diagnosis of pulmonary AVMs, were inconclusive.

The rationale for the diagnostic use of saline echo-contrast medium is that the large bubbles (>50 μm) within the injectate cannot traverse the pulmonary capillaries. If the contrast solution is detected in the left chambers of the heart, it can be concluded that the bubbles bypassed the capillary bed through an arteriovenous shunt that is undetectable by pulmonary angiography. The delay of four or five cardiac cycles between the appearance of the bubbles on the right side and their subsequent appearance in the left atrium, as opposed to

**FIGURE 1.** Contrast echocardiography. Left: after injection of agitated saline solution into an antecubital vein, initially the right atrium (RA) and right ventricle (RV) are densely opacified. Right: after approximately four cardiac cycles, the contrast medium appears in the left atrium (LA) and subsequently completely opacifies it and the left ventricle (LV). This pattern of echo contrast is consistent with a large intrapulmonary shunt.
nearly immediate opacification of the left atrium, is the key to recognizing intrapulmonary shunts. On TEE, each vein can be visualized individually. Echo-contrast medium emerging from each of the four pulmonary veins confirms that fistulas are present throughout the pulmonary vascular bed.

This case illustrates the diagnostic usefulness of contrast TEE in the identification and localization of pulmonary arteriovenous fistulas. This technique can help to detect and localize pulmonary fistulas to a particular lobe or lobes of the lung and can obviate or indicate selective angiography.

For a patient in whom a right-to-left shunt has been detected by arterial blood gas analysis or clinical findings, the next step should be contrast echocardiography and TEE to localize the anatomic origin of the shunt so that a course of further documentation and management can be planned. In our patient, however, the fistulas are diffuse and not amenable to invasive intervention.

REFERENCES


Femoral Venoarterial Extracorporeal Membrane Oxygenation for Severe Reimplantation Response After Lung Transplantation*

Dirk Vlasselaers, MD; Geert M. Verleden, PhD; Bart Meyns, PhD; Dirk Van Baemdenck, MD, PhD, FCCP; Maurits Demedts, MD, PhD, FCCP; Antoon Lerut, PhD; and Peter Lauwers, MD