Transesophageal Echocardiographic Diagnosis of Pulmonary Arteriovenous Malformation

Role of Contrast and Pulsed Doppler Echocardiography


Contrast echocardiography produced by peripheral injection of agitated saline solution is widely used for detecting intracardiac and intrathoracic extracardiac shunts, like pulmonary arteriovenous malformation (PAVM). Currently, localization of PAVM requires pulmonary angiography even after detection by computed tomography of the chest. Pulsed Doppler along with contrast echocardiography of the pulmonary veins performed during transesophageal echocardiography may aid in the localization of PAVM and in its diagnosis.

(CHEST 1994; 105:1604-05)

IAS=interatrial septum; PAVM=pulmonary arteriovenous malformation; RIPV=right inferior pulmonary vein; TEE=transesophageal echocardiography

Pulmonary arteriovenous malformation (PAVM) is a rare congenital heart anomaly that may present with signs and symptoms of hypoxemia. The definitive diagnosis of this entity presently requires invasive diagnostic modalities, the gold standard of which is selective pulmonary arteriography. We present a case of PAVM diagnosed by contrast and pulsed Doppler transesophageal echocardiography (TEE) and confirmed by dynamic computed tomography (CT) of the chest.

Case Report

A 67-year-old white woman was referred to our hospital for evaluation of a transient ischemic attack, presenting as right hand weakness and slurred speech. Results of physical examination at the time of presentation were unremarkable. There was a 2/6 systolic ejection murmur auscultated at the lower left sternal border. Clinical evaluation at the outlying institution included an ECG, chest radiograph, and CT scan of the head that were reportedly normal. Additionally, two-dimensional echocardiography was performed to exclude an intracardiac thrombus and/or the potential source of a paradoxical embolism as the cause for her presenting symptoms. Contrast echocardiography showed a right-to-left shunt, although definite atrial septal defect, patent foramen ovale, or ventricular septal defect could not be demonstrated. The patient was referred to our institution for TEE to localize the cause of the patient's right-to-left shunt.

From the Likoff Cardiovascular Institute, Division of Cardiology, Department of Medicine, Hahnemann University, Philadelphia.

Reprint requests: Dr. Chandrasekaran, Cardiac Ultrasound Lab, Hahnemann University Hospital MS 313, Philadelphia 19102

The TEE was performed to better evaluate the interatrial septum (IAS), as well as to rule out a shunt across the IAS. Two-dimensional echocardiography showed normal left and right ventricular size and function, normal right atrium, and mildly enlarged left atrium. The valvular structures were normal. Subsequent to an agitated saline solution injection via the right brachial vein, contrast was sequentially visualized in the right atrium, right ventricle, pulmonary artery, and after three to five cardiac cycles, within the left atrium via the right inferior pulmonary vein (RIPV). Pulsed Doppler at this site demonstrated an abnormal spectral display in addition to increased peak and mean velocities, in comparison to the normal flow patterns observed within the remaining three pulmonary veins (Fig 1). This finding is suggestive of PAVM, localized to the right lower lobe vascular bundle. Thin-slice chest CT was performed and confirmed the diagnosis of a PAVM located proximally in the right hilum (Fig 2).

Discussion

The incidence of congenital heart disease in the United States is approximately 8 per 1,000 live births. Intracardiac shunts, including patent foramen ovale, atrial septal defect, and ventricular septal defect are not uncommonly found in the adult population. 

Pulmonary arteriovenous malformation is found in approximately 2 to 3/100,000 persons. Often it is detected as a “coin-lesion” on the chest radiograph in an otherwise asymptomatic individual. This rare extracardiac but intrathoracic shunt, however, is found within 6 to 15 percent of patients with hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber syndrome). Clinical presentation is primarily dependent on the degree of shunting. The symptomatic patient, usually with larger or more extensive vascular abnormalities, presents most often with signs and symptoms of oxygen desaturation: cyanosis, dyspnea, clubbing, or polycythemia. Routine diagnostic evaluation would include a chest radiograph, an arterial blood gas determination, and a CT of the chest if PAVM is suspected as the cause. However, more commonly when the diagnosis is unknown in the situation of systemic desaturation as in our patient, two-dimensional echocardiography with contrast is routinely used to document the presence of an intracardiac/intrathoracic shunt noninvasively.

Transesophageal echocardiography, by its ability to demonstrate the IAS, the insertion of the superior and inferior vena cavae into the right atrium, along with the insertions of the pulmonary veins into the left atrium has proved to be a reliable diagnostic tool in the evaluation of intracardiac shunts. Pulsed Doppler echocardiography of the pulmonary veins is used to assess the filling characteristics of the left ventricle and severity of mitral regurgitation. In our patient, TEE demonstrated intact IAS and interventricular septa, thereby excluding the presence of an intracardiac shunt. Furthermore, by its ability to visualize pulmonary veins, we were able to localize contrast entering the left atrium via the RIPV, thus indicating an extracardiac but intrathoracic shunt such as a PAVM. By demonstrating contrast entering the left atrium via the RIPV, the PAVM was localized to the lower lobe of the right lung. Pulsed Doppler examination supported the contrast echocardiography findings by demonstrating an increased velocity and abnormal spectral display consistent with an arterial flow pattern in the RIPV. This is in con-
Contrast to the normal flow patterns of the remaining three pulmonary veins (Fig 1). This confirmed the PAVM.

Transesophageal echocardiography with contrast and pulsed Doppler clarified the site of the right-to-left shunt detected by transthoracic echocardiography, but more importantly, localized the site as extracardiac and intrathoracic involving the lower lobe of the right lung. Since TEE is becoming so widely used in the evaluation of patients for shunts, performing contrast and pulsed Doppler echocardiography may aid in the diagnosis and localization of PAVM, obviating the necessity for pulmonary arteriography to localize a PAVM, especially in a patient with a small asymptomatic PAVM.

REFERENCES

2 Hodgson CH, Kaye RL. Pulmonary arteriovenous fistula and hereditary hemorrhagic telangiectasia: a review and report of


Pulmonary Artery and Noninvasive Hemodynamics During Lung Lavage in Primary Alveolar Proteinosis*

Jan R. Hemstad, M.D.; Bruce D. Spiess, M.D.; Thomas L. Marchioro, M.D.; F.C.C.P.; and Ganesh Baghu, M.D.

Noninvasive hemodynamic monitoring by bioimpedance has been compared to cardiac output measured by thermodilution. The technology of bioimpedance does allow for monitoring of static thoracic impedance, a

*From the Division of Cardiothoracic Anesthesiology (Dr. Spiess), Department of Anesthesiology (Dr. Hemstad); the Department of Surgery (Dr. Marchioro), and the Chest Clinic, Division of Pulmonary and Critical Care Medicine, Department of Medicine (Dr. Baghu), University of Washington, Seattle.

Reprint requests: Dr. Spiess, Department of Anesthesiology RN-10, University of Washington, Seattle 98195