Case Report

Two-Dimensional Contrast Echocardiography in Pulmonary Arteriovenous Fistula

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SUMMARY

Definitive clinical diagnosis of congenital pulmonary arteriovenous fistula is extremely difficult. In order to evaluate the diagnostic value of echocardiography, 2 cases with suspected pulmonary arteriovenous fistula were studied.

In the first case, there was a solitary pulmonary arteriovenous fistula, while in the second multiple minute pulmonary arteriovenous fistulas were illustrated. The solitary lesion was demonstrated by two-dimensional and peripheral vein contrast echocardiography. However, in the second case direct visualization of the lesion was not possible but peripheral vein contrast echocardiography showed abnormal filling of the left atrium with echo contrast material.

Additional Indexing Words:
Two-dimensional echocardiography Contrast echocardiography Pulmonary arteriovenous fistula

Congenital pulmonary arteriovenous fistula is a rare anomaly.1,2) Definitive clinical diagnosis of this condition is extremely difficult. Cyanosis, digital clubbing and dyspnea without a significant cardiac murmur suggest this condition. For precise anatomic diagnosis, angiocardio-

graphy is necessary. However, as a noninvasive diagnostic method echocardio-

graphy seems to be useful in pulmonary arteriovenous fistula.

In order to evaluate the diagnostic value of two-dimensional and peripheral vein contrast echocardiography, 2 cases with suspected pulmonary arteriovenous fistula were studied.

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Case Report

Case 1: A 10 year old boy was admitted for evaluation of central cyanosis and digital clubbing. There was no history of hematemesis, hemoptysis, seizures, or congestive heart failure, although some decrease in exercise tolerance over the last 6 months had been noted. Physical examination revealed cyanosis with digital clubbing of the fingers and toes. The heart sounds were normal. There was no significant heart murmur. The hemoglobin was 15.85 gm/dl and Hct 50%. The electrocardiogram was within normal limits. There was a rounded opacity close to the right heart border seen on telecardiogram (Fig. 1). On two-dimensional echocardiography, the left atrium and ventricle were slightly enlarged. No intracardiac pathology was detected. In apical four-chamber view an echo free lesion was seen beyond the left atrium (Fig. 2). This lesion was also detected in the parasternal short-axis view in which the right atrium, right ventricular outflow tract, pulmonary artery, aorta and left atrium could be visualized (Fig. 3). To confirm this impression peripheral vein contrast echocardiography was performed. In this procedure 10 cc of 3% saline solution was injected rapidly into an antecubital vein 6 times. In one injection the transducer was located in the apical four-chamber view (Fig. 2), and in the others the parasternal short-axis view (Fig. 3). In the apical four-chamber view the contrast material was first seen in the right atrium, then in the right ventricle, vascular lesion and left atrium. In the parasternal short-axis study the contrast appeared first in the right atrium, then the right ventricle, pulmonary artery, vascular lesion and left atrium. These studies confirmed the previous impression of a pulmonary arteriovenous fistula draining into the left atrium. For further diagnostic confirmation, cardiac catheterization and angiocardiography were performed. Injection of radio-opaque
Fig. 2. Two-dimensional echocardiography (Case 1). In apical four-chamber view an echo-free lesion was seen beyond the left atrium (LA) which was thought to be an arteriovenous fistula (above picture). Following injection of contrast material into the peripheral vein, right atrium (RA), right ventricle (RV), fistula lesion, LA and LV (left ventricle) filling were seen (below picture). Black arrows indicate the fistula lesion.

Fig. 3. Two-dimensional echocardiography (Case 1). In parasternal short-axis view the echo-free lesion of the arteriovenous fistula was seen beyond the LA. Following peripheral vein contrast injection RV, pulmonary artery (PA), fistula lesion, and LA were filled with echo-contrast material. Black arrows indicate the arteriovenous fistula.

material into the right pulmonary artery demonstrated immediate filling of the fistula, left atrium and left ventricle (Fig. 4).

Surgical treatment of the lesion with right lower lobectomy was carried out. The postoperative course was uneventful.

Case 2: An 8 year old male was admitted to the hospital for evaluation of cyanosis and clubbing. He also complained of limitation of exercise capacity. Physical examination revealed a child with cyanosis and clubbing of the fingers. No significant murmurs were heard. Heart sounds were normal. ECG was within normal limits. X-ray examination of the chest showed reticular appearance of the lung fields.

Two-dimensional echocardiography did not show any significant lesion
Fig. 4. Cineangiogram of right pulmonary artery of Case 1 illustrating the arteriovenous fistula (arrow).

Fig. 5. Cineangiogram of right pulmonary artery of Case 2 showing multiple fistula lesions (arrow).

as described in the previous case. Peripheral vein contrast echocardiography in the parasternal short-axis view revealed left atrial filling with echo contrast material, following pulmonary artery filling. In the parasternal long-axis view left atrial contrast filling was seen following the right heart opacification with contrast. These findings were evaluated as indirect evidence of communication between the pulmonary artery and vein.

At cardiac catheterization and angiocardiography arterial oxygen saturation was 69%. Pulmonary artery cineangiograms revealed multiple pulmonary arteriovenous fistulas (Fig. 5). The patient was thought to be inoperable.

**DISCUSSION**

Pulmonary arteriovenous fistulas vary in number, size, distribution and configuration. At one extreme, there may be multiple minute arterio-
venous fistulas distributed throughout most or all lobes of both lungs. At
the other extreme, there is a solitary large arteriovenous aneurysm usually
situated in the lower lobes of the entire lung, more often on the right than on
the left. The 2 cases presented are examples of these extremes.

In the first case central cyanosis, digital clubbing and rounded opacity
on telecardiogram were suggestive of pulmonary arteriovenous fistula. It
was diagnosed by 1) two-dimensional and contrast echocardiography and 2)
angiocardiography. Then, the diagnosis was confirmed by surgery. As a
noninvasive method echocardiography was very useful to illustrate the lesion.
It could be seen directly by two-dimensional echocardiography. In addi-
tion, contrast material filling of the lesion following pulmonary artery filling
could be illustrated (Fig. 3). On review of the video recordings consecutive
filling of the related cardiac structures namely, the right atrium, right ven-
tricle, pulmonary artery, fistula lesion and left atrium were seen in the para-
sternal short-axis view.

Although some authors have demonstrated left atrial filling follow-
ing right ventricular outflow tract opacification with contrast material by
M-mode contrast echocardiography, the superiority of two-dimensional
echocardiography to M-mode is well known.

The peripheral vein two-dimensional contrast echocardiographic meth-
od can be diagnostic in solitary large pulmonary arteriovenous fistulas as it
was in our first patient. However, in cases of multiple minute fistulas as seen
in our second case, only filling of the left atrium with echo-contrast material
can be illustrated.

Normally, contrast substance in the pulmonary artery would be removed
by passage through the pulmonary capillary bed. As a result of this,
contrast material does not appear in the left heart. Appearance of contrast
material in the left atrium may suggest a pulmonary arteriovenous fistula,
as it did in both of our cases. However this must be differentiated from an
atrial septal defect. Although by using the M-mode technique such differen-
tiation is not possible, the filling of the left atrium from the right atrium in
an atrial septal defect is directly seen on two-dimensional peripheral vein
contrast echocardiography. This can more clearly be visualized in the
apical four-chamber projection. We could not see any passage of contrast
material through the atrial septal defect in either of our cases.

In conclusion, two-dimensional and contrast echocardiography may be
diagnostic in cases of a solitary large pulmonary arteriovenous fistula. How-
ever, in those with multiple minute lesions, definite diagnosis cannot be made
using only two-dimensional echocardiography, although contrast echo may
be suggestive.
In order to prepare patients with pulmonary arteriovenous fistula for surgery, pulmonary artery angiocardiology is also recommended. By angiocardiology multiple lesions can be detected. Even in cases with solitary lesions, angiocardiology may provide more information about the lesion.

To our knowledge this is the first report in the medical literature evaluating the diagnostic role of two-dimensional echocardiography in cases of pulmonary arteriovenous fistula.

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