Premature Opening of the Pulmonary Valve in Endomyocardial Fibrosis

Mitsunori Okamoto, M.D.,* Hideyo Amioka, M.D.,
Masaki Hashimoto, M.D., Hiroyuki Shimamoto, M.D.,
Eiichiro Sakura, M.D., Yuji Yokote, M.D., Togo Yamagata, M.D.,
Yukiko Tsuchioka, M.D., Hideo Matsuura, M.D.,
and Goro Kajiyama, M.D.

SUMMARY

Two-dimensional and Doppler echocardiographic findings in a 67-year-old man with endomyocardial fibrosis (EMF) are described. The two-dimensional echocardiogram showed typical features of EMF, right ventricular endomyocardial calcification, a thickened right ventricular wall, obliteration of the apex of the right ventricle and marked dilatation of the right atrium. In addition, premature opening of the pulmonary valve was observed during late diastole. The Doppler echocardiogram revealed forward flow from the right ventricle to the pulmonary artery, indicating the conduit state of the right ventricle. These findings were supported by cardiac catheterization and autopsy. Thus, two-dimensional and Doppler echocardiography are useful not only in making the diagnosis, but also in understanding the hemodynamic condition in EMF.

Additional Indexing Words:
Endomyocardial fibrosis  Doppler echocardiography  Two-dimensional echocardiography  Pulmonary valve  Restrictive cardiomyopathy

ENDOMYOCARDIAL fibrosis (EMF) is characterized by a patchy or uniform fibrosis of the endocardium, involving mainly the apical and inflow regions of either or both ventricles of the heart.1,2) EMF is accompanied by endocardial rigidity, atrioventricular valve incompetence secondary to papillary muscle involvement and progressive reduction of the size of the ventricular cavities. This condition is usually seen in tropical countries and is rare in Japan. There have been many investigations of myocardial involvement in EMF by M-mode and two-dimensional echocardiography. However, Doppler echocardiographic features have not been characterized.

From the First Department of Internal Medicine, Hiroshima University School of Medicine, Hiroshima City, Japan.

* Present status: Clinical Laboratory, Hiroshima University.

Address for reprint: Mitsunori Okamoto, M.D., First Department of Internal Medicine, Hiroshima University School of Medicine, 1-2-3 Kasumi, Minami-ku, Hiroshima City 734, Japan.

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We present a patient with EMF, in whom premature opening of the pulmonary valve with diastolic forward flow toward the pulmonary artery was observed by pulsed Doppler and two-dimensional echocardiography.

**CASE REPORT**

A 67-year-old man with a 20-year history of exertional dyspnea was admitted to our hospital. He had undergone gastrectomy because of gastric

![Fig. 1. Four-chamber view echocardiograms in end-diastole (left) and end-systole (right) showing right ventricular endocardial calcification and hypertrophy (white arrow).](image)

RV = right ventricle; RA = right atrium; LV = left ventricle; ED = end-diastole; ES = end-systole.

![Fig. 2. Two-dimensional and M-mode echocardiograms showing opening of the pulmonary valve during late diastole (white and black arrows).](image)

PA = pulmonary artery.
carcinoma 2 years previously. Physical examination revealed jugular venous
dilatation, a systolic murmur at the lower left sternal border and several
metastatic skin tumors. Eosinophilia was not found on the blood smear.
The electrocardiogram showed right axis deviation and right ventricular over-
load. The two-dimensional echocardiogram demonstrated right ventricular
endomyocardial calcification, a thickened right ventricular wall, obliteration
of the apex of the right ventricle and marked dilatation of the right atrium
(Fig. 1). The interventricular septal motion was reversed. There were no
abnormalities of the endocardium and myocardial wall of the left ventricle.
Mural thrombi were not found in either ventricle. The most conspicuous
finding was the pulmonary valve opening in mid to late diastole (Fig. 2).
The pulsed Doppler echocardiogram revealed forward flow from the right
ventricle toward the pulmonary artery during late diastole (Fig. 3). Tri-
cuspid regurgitant signals were also observed in the right atrium, but there
were no mitral regurgitant signals in the left atrium. Right heart catheteriza-
tion was performed and intracardiac pressure was measured by a catheter-
tipped manometer and a fluid filled system (Table I). The right ventricular
pressure curve exhibited a dip and plateau pattern and the diastolic pressure

Fig. 3. Blood flow signals in the right ventricular outflow tract (upper
panel) and just above the pulmonary valve in the pulmonary artery (lower
panel). Forward flow is seen during late diastole in both the right ventricle
and pulmonary artery (black arrows). SV=sample volume.
Table I. Right Heart Catheterization Data

<table>
<thead>
<tr>
<th>Syst/Diast (mean) mmHg</th>
<th>l/min/m²</th>
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<tbody>
<tr>
<td>Pulmonary artery</td>
<td>15/10 (12)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>22/5-12 (12)</td>
</tr>
<tr>
<td>Right atrium</td>
<td></td>
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</tbody>
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Fig. 4. Pullback pressure tracing from the pulmonary artery to the right atrium through the right ventricle.

Fig. 5. Right ventriculography showing obliteration of the apex, narrowing of the right ventricular cavity and severe tricuspid regurgitation.

was almost the same in the right atrium, the right ventricle and the pulmonary artery (Fig. 4). Typical features of EMF, including obliteration of the right ventricular apex, right ventricular endocardial calcification, marked dilatation
of the right atrium and severe tricuspid regurgitation, were noted on right ventriculography (Fig. 5). The forward flow from the right ventricle toward the pulmonary artery and opening of the pulmonary valve during late diastole were also confirmed by right ventriculography. This patient died of cachexia resulting from metastases of his gastric carcinoma to the liver, bone, lymphonodi and skin.

At autopsy fibrous thickening of the right ventricular endocardium and narrowing of the right ventricular cavity were found (Fig. 6). There were no mural thrombi in the cardiac cavities. Histologic examination showed a prominent layer of hyaline fibrous tissue in the thickened endocardium. Thus, the pathologic findings supported the clinical diagnosis of EMF.

**DISCUSSION**

EMF was first described in Uganda by Davies. Other cases have been reported from tropical and nontropical countries. Its recognition has been mainly established by the characteristic angiographic obliteration of the ventricular apex. Echocardiography has been reported to be helpful in making the diagnosis of EMF. On M-mode echocardiography, paradoxical interventricular septal motion, left atrial enlargement, increased right ventricular dimension, and an abnormal increase in posterior wall thickness have been described. However, two-dimensional echocardiography is a more sensitive and specific method than M-mode echocardiography. Some authors
have described obliteration of the apex of the involved ventricle, a grossly dilated atrium with normal sized ventricle, endocardial calcification, thickening of the posterior left ventricular wall and thickening of the atrioventricular valv on two-dimensional echocardiograms.3-7) In the present case, two-dimensional echocardiogram demonstrated the right ventricular involvement. In addition, premature opening of the pulmonary valve was evident on M-mode and two-dimensional echocardiography. This abnormal pulmonary valve motion was accompanied by forward flow toward the pulmonary artery, which was detected by Doppler echocardiography. In EMF, such pulmonary valve motion and blood flow abnormalities have never been characterized by two-dimensional echocardiography nor by pulsed Doppler echocardiography. However, a similar pulmonary valve motion seen on the echocardiogram has been described in patients with constrictive pericarditis, right ventricular infarction, Uhl's anomaly, rupture of an aneurysm of the sinus of Valsalva and after cardiac surgery.8-12) In these diseases, the right ventricular diastolic pressure is elevated because of impaired diastolic filling due to a calcified pericardium in constrictive pericarditis, severe right ventricular failure in right ventricular infarction and Uhl's anomaly, and shunt flow to the right ventricle in rupture of an aneurysm of the sinus of Valsalva. Accordingly, the right ventricular pressure may sometimes exceed the pulmonary artery pressure in end-diastole, so that the reverse diastolic gradient may cause premature opening of the pulmonary valve. Some authors have described a reverse diastolic gradient in patients with EMF.13) In the present case, end-diastolic pressure was almost the same in the right atrium, the right ventricle and the pulmonary artery. In addition, the diastolic forward flow toward the pulmonary artery was observed by Doppler echocardiography and right ventriculography. These abnormal hemodynamic conditions may have produced the premature opening of the pulmonary valve in the present case. The premature opening of the pulmonary valve with forward flow toward the pulmonary artery means the conduit state of the right ventricle in EMF. Thus, two-dimensional and Doppler echocardiography are useful not only in making the diagnosis, but also in understanding the hemodynamic condition in EMF.

REFERENCES
PREMATURE PULMONARY VALVE OPENING IN EMF

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