

## Silent Pulmonary Artery Dissection in a Patient With Eisenmenger Syndrome Due to Ventricular Septal Defect: A Case Report

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### Abstract

A 62-year-old woman was admitted to our hospital because of fever in August 2002. She had been treated under a diagnosis of Eisenmenger syndrome with ventricular septal defect since 1988. On admission, echocardiography and color Doppler echocardiography revealed a markedly enlarged pulmonary artery with a mobile flap, and dissection of the pulmonary artery. The origin of the fever could not be identified, and the fever subsided spontaneously without specific treatment. She had no chest pain, but fever might have been a sign of dissection in this patient. Longstanding pulmonary hypertension may cause dissection, which may lead to sudden death or pulmonary hemorrhage often seen in patients with Eisenmenger syndrome. Our patient was a rare survivor without serious bleeding complication.

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### Key Words

■Echocardiography, transthoracic  
■Heart defects, echocardiography  
■Heart defects, congenital  
■Pulmonary artery

### INTRODUCTION

The prognosis for adult patients with Eisenmenger syndrome is characterized by complications such as congestive heart failure, brain abscess, and sudden death<sup>1)</sup>, and the long-term prognosis is poor. Sudden death generally considered to be due to ventricular arrhythmia is the most common cause of death in adult patients with Eisenmenger syndrome<sup>1-3)</sup>. However, a recent report<sup>4)</sup> emphasized that the cause of sudden death

is intrapulmonary hemorrhage following rupture of aneurysmal dilatation of the pulmonary artery (PA). We describe a case of Eisenmenger syndrome due to ventricular septal defect (VSD) with silent PA dissection identified by echocardiography.

### CASE REPORT

A 62-year-old woman was admitted to our hospital because of fever persisting for 2 months. Eisenmenger syndrome with VSD was identified by cardiac catheterization at another hospital in 1988

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when she was 48 years old. The PA pressure was 112/55 mmHg (mean: 79 mmHg), and left ventricular pressure was 118/2 mmHg. Since then, she had been receiving medication. She suffered breathlessness at rest and finger cyanosis in 1999 when she was 59 years old. She started home oxygen therapy in April 2000. She was admitted to our hospital in August 2002 because of low-grade fever persisting mainly in the afternoon for 2 months.

On admission, her blood pressure was 96/48 mmHg, pulse was irregular 87 beats/min, and body temperature was 36.8 °C. There was a systolic ejection murmur (Levine) and a high-pitched decrescendo diastolic murmur (Levine, pulmonary regurgitation) over a wide area. The liver was four fingerbreadths palpable below the right costal margin. Peripheral edema was absent. She had cyanotic fingers and clubbed toes.

Laboratory examination revealed erythrocytosis [red blood cells ( $614 \times 10^4/\mu\text{l}$ ), hemoglobin (18.1 g/dl), and hematocrit (60.9%)]; thrombocytopenia ( $12.7 \times 10^4/\mu\text{l}$ ); and increased total bilirubin (1.5 mg/dl), brain natriuretic peptide (122 pg/ml), and hypoxia ( $\text{PaO}_2$  47.2 mmHg). Inflammatory parameters showed no abnormality (white blood cells: 4,100/ $\mu\text{l}$ , C-reactive protein: 0.1 mg/dl).

Chest radiography revealed remarkable cardiomegaly (cardiothoracic ratio: 81%), and enlarged proximal PAs with clear lung fields (Fig. 1). Electrocardiography showed sinus rhythm, peaked P waves and right ventricular hypertrophy with right axis deviation (Fig. 2).

Transthoracic echocardiography revealed a VSD (perimembranous defect) with bi-directional shunting, and markedly enlarged PAs (diameter of the main PA: 8.5 cm, the right PA: 4.0 cm, and the left PA: 3.4 cm) with a mobile flap from just above the pulmonary valve (Fig. 3 - left). Color Doppler signal showed a flow from the true lumen to the false lumen, and dissection of the PA (Fig. 3 - right). The dissection started from 1 cm above the pulmonary valve, but the distal end was unclear. There was mild tricuspid regurgitation and severe pulmonary regurgitation. The degree of pulmonary regurgitation had not changed in 10 years. Doppler calculation of the right ventricular systolic pressure was 84 mmHg. The right atrium, right ventricle, and left atrium were enlarged. Left ventricular contraction was diffusely and moderately reduced (fractional shortening: 21%). A moderate amount



**Fig. 1** Chest radiograph showing severe cardiomegaly with cardiothoracic ratio of 81%, and bilateral prominent pulmonary arteries

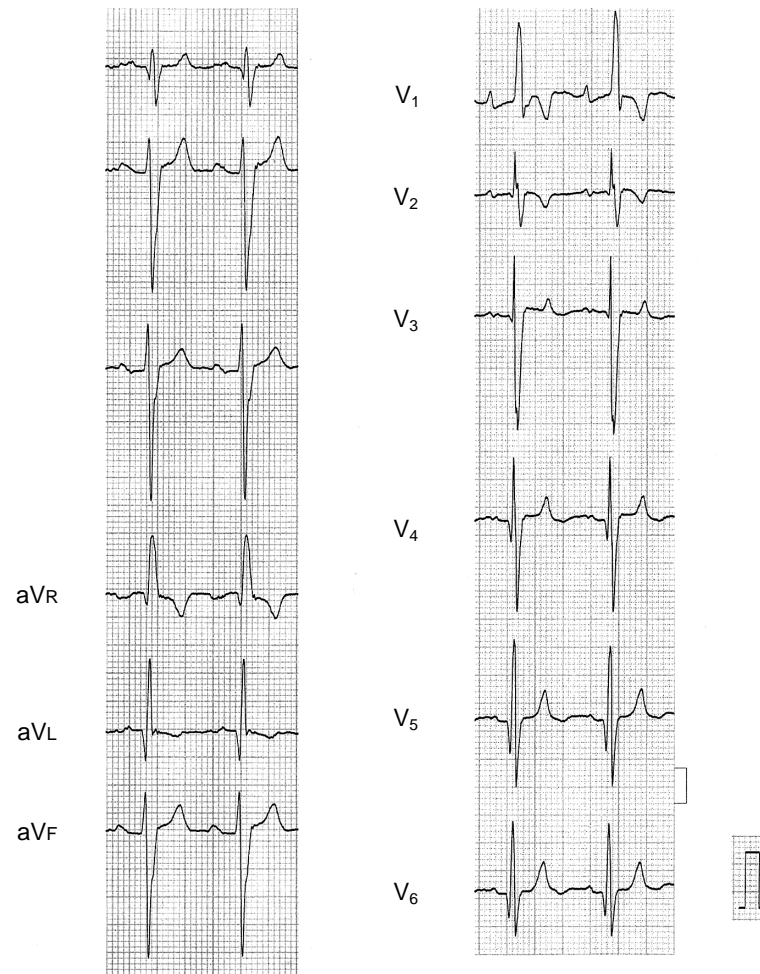
of pericardial effusion was present.

Chest computed tomography revealed markedly enlarged PAs (diameter of main PA: 10.0 cm, right PA: 5.0 cm, and left PA: 4.0 cm) without calcification, and a moderate amount of pericardial effusion (Fig. 4).

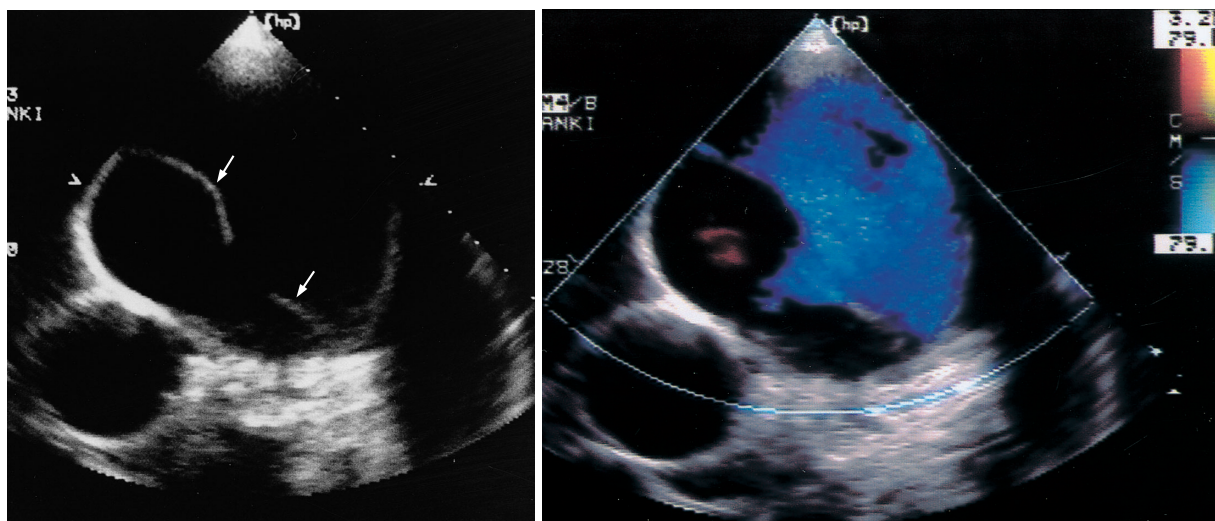
To identify the cause of fever, many examinations to detect infection, malignancy and collagen disease were carefully performed, but all results, including blood culture, were negative. Laboratory data did not reveal the presence of inflammation. After admission, her low-grade fever mainly occurred in the afternoon (37 °C). The origin of the fever was not identified, but it subsided spontaneously without any specific treatment and breathlessness improved after 10 days. However, echocardiography revealed markedly enlarged PAs with dissection. PA dilatation had progressed compared with 2 years previously, when PA dissection had not been observed.

## DISCUSSION

The natural history of 47 patients with Eisenmenger syndrome due to VSD aged 23 to 69 years (mean: 39.5 years) showed 10 of the 14 deceased patients died suddenly during the follow-up period of 5 to 18 years (mean: 7.2 years)<sup>1</sup>. Autopsy of nine patients found the cause of sudden



**Fig. 2** Electrocardiogram showing first-degree atrioventricular block, right atrial enlargement, and right ventricular hypertrophy



**Fig. 3** Echocardiogram, short-axis view, showing a dilated pulmonary artery and intimal flap in the lumen (left, arrows) and color Doppler echocardiogram showing flow from the true lumen to a false lumen (right)



**Fig. 4** Chest computed tomogram showing a huge dilated main pulmonary artery with a diameter of 10.0 cm, right pulmonary artery of 5.0 cm, and left pulmonary artery of 4.0 cm

death was massive intrapulmonary hemorrhage in two patients and rupture of the aneurysmal pulmonary trunk in one. No patient suffered tachyarrhythmic sudden death. Therefore, PA damage is an important cause of death in patients with Eisenmenger syndrome.

PA dissection is a very unusual event. Review of 52 patients with PA dissection found that dissection occurred in 39(75%) patients with pulmonary hypertension of various causes<sup>5)</sup>. Among these 39 patients, 28 had secondary pulmonary hypertension and eight had primary pulmonary hypertension. Among the 28 patients with secondary pulmonary hypertension, 23 had congenital cardiovascular anomaly, eight had ductus arteriosus, and two had VSD. The clinical symptoms of PA dissection were non-specific: chest or parasternal pain in 16

patients, cyanosis in 11, dyspnea in nine, and shock in four. In 38 of the 52 patients, rupture caused hemorrhage in the surrounding organs and tissues, including the pericardium(30 patients), lungs (four), mediastinum(three) and pleural cavity (one). PA dissection led to sudden death in all 38 patients. In the majority of reported cases(45 of 52 patients, 86.5%) the diagnosis was made at autopsy.

PA dissection should be suspected if a patient with pulmonary hypertension complains of severe chest pain and dyspnea. However, since PA dissection often results in sudden death, the diagnosis has rarely been made in living patients. Only seven cases of PA dissection have been identified in life, using echocardiography(two patients)<sup>6,7)</sup>, pulmonary arteriography(one)<sup>8)</sup>, magnetic resonance imaging(two)<sup>9,10)</sup>, or computed tomography (two)<sup>11)</sup>. Anatomically, the dissection was located in the first 2 or 3 cm of the pulmonary artery mainstem in most patients.

Longstanding pulmonary hypertension may cause PA dissection, which is associated with the sudden death or pulmonary hemorrhage often seen in patients with Eisenmenger syndrome. Our patient was thought to be a rare survivor without serious bleeding complications. The PA asymptomatic dissection was identified by echocardiography. In our case, no chest pain was observed, but fever of unknown origin might be a sign of PA dissection.

Only three cases of PA dissection have been successfully treated with surgical repair of the PA<sup>5,10,11)</sup>. Our patient and her family rejected invasive treatment, so we selected conservative therapy.

## 要 約

### 無症候性に肺動脈解離を認めた心室中隔欠損症による Eisenmenger 症候群の 1 例

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症例は62歳、女性。1988年に他院で心臓カテーテル検査を受け、Eisenmenger症候群を伴う心室中隔欠損症の診断を受けていた。2002年8月、発熱精査のため入院となり、心エコー図検査により肺動脈径の著明な拡大と肺動脈解離の所見を認めた。発熱の原因は特定できなかったが、入院後に自然軽快した。胸痛の訴えはなかったが、発熱が解離の症状であった可能性が考えられた。長期間にわたる肺高血圧は肺動脈の解離の原因となり、Eisenmenger症候群の症例において肺出血や突然死をもたらすとされている。62歳まで重篤な出血に至らず、無症候性に肺動脈の解離が生存中にみつかったまれな症例と思われた。

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