ISSN 1850-1044

CASE REPORT (English version)

Eisenmenger syndrome in a patient with atrial septal defects Study of a case

Fábio Azevedo Simão[1], Felipe Montes Pena[1], Fernanda Arêdo Carvalho[2], Carlos Augusto Cardozo de Faria[3]

Summary

The Eisenmenger syndrome (ES) represents the most advanced form of pulmonary arterial hypertension associated with congenital heart defects (CHD). Adults with CHD represent an expanding population requiring tertiary care in the long term. Around 8% of patients with CHD and 11% of those with shunts from left to right develop the framework for the ES. Efforts are directed at treatment for reducing pulmonary vascular resistance, left to right shunt, cyanosis, morbidity and mortality. We present a case of female, 41 years old, who had cyanosis at rest, lower limb edema and atrial septal defect type communication interatrial diagnosed in echocardiography who was tackled drug with calcium channel blockers.

Insuf Card 2010 (Vol 5)4:205-208

Keywords: Pulmonary hypertension - Eisenmenger - Congenital heart disease

Introduction

In 1897, Viktor Eisenmenger described a case of a patient with cyanosis and dyspnea since childhood, who died with massive hemoptysis at 32 years old. The autopsy showed a ventricular septal defect and severe pulmonary vascular disease[1]. In 1958, Paul Wood described the term Eisenmenger complex, consisting of "pulmonary hypertension in systemic levels with reversed or bidirectional shunt or a ventricular septal defect". Subsequently, the term Eisenmenger syndrome (ES) has been used to describe the pulmonary vascular disease and cyanosis resulting from the connection between the pulmonary and systemic circulation (as in atrial septal defects, ventricular septal defect, patent ductus arteriosus and aortopulmonary window)[2].

Therefore, Eisenmenger syndrome represents the most advanced form of pulmonary arterial hypertension (PAH) associated with congenital heart defects. Adults with congenital heart disease (CHD) represent an expanding population that requires long-term tertiary care. About 5% to 10% of them present PAH of variable severity that affects quality of life, morbidity and mortality[3].
We report the case of a 41-year-old female who presented with very high pulmonary artery pressure (PAP), lower extremity edema, cyanosis of the extremities and atrial septal defect, fulfilling the criteria for ES.

Case report

We present the case of a 41-year-old woman born in Rio de Janeiro, who was admitted to hospital with signs

[1] Specialization in Clinical Cardiology. Fluminense Federal University. University Hospital Antonio Pedro (HUAP). Niterói. Rio de Janeiro. Brazil.
[2] Pregrade in Medicine. Fluminense Federal University. Niterói. Rio de Janeiro. Brazil.
[3] PhD in Clinical Investigation. Fluminense Federal University. University Hospital Antonio Pedro (HUAP). Niterói. Rio de Janeiro. Brazil.

Correspondence: Dr. Felipe Montes Pena
Rua Mariz e Barros, número 71. Apartamento 601. Bairro Icaraí. Cep 24220-120. Niterói. Río de Janeiro. Brazil.
E-mail: fellipena@yahoo.com.br; fellipena@hotmail.com

Received: June 28, 2010
Accepted: September 25, 2010

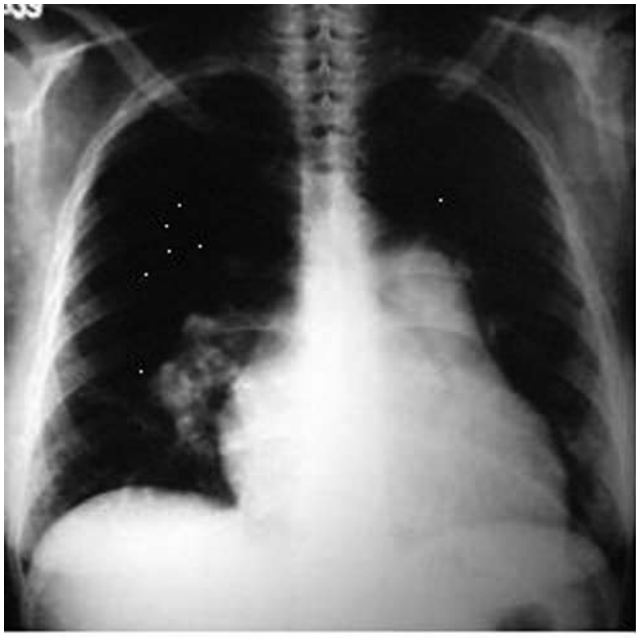

Figure 1. Pulmonary atrial protrusion at chest X-ray.

of progressive dyspnea on minimal effort, which began six months ago with a significant worsening in the last three months functional class, pleuritic chest pain with non-productive cough and severe edema of the lower extremities. Medical history recounts drug treatment hypertension, heart murmur in childhood and smoking. Cardiovascular auscultation physical examination showed a hypophonetic S2 with sisto-diastolic murmur in the lung area. He was undergoing tachypnea, cyanosis of the fingertips at rest, and increased positive sign Dressler on palpation of the chest.

Complementary examinations were requested, such as biochemistry, rheumatic activity evidence, thyroid, hemoglobin electrophoresis, within normal parameters. It was requested a chest radiography, showing a significant protrusion in the area of the pulmonary artery (Figure 1), electrocardiogram identified first degree atrioventricular block with right bundle branch block (Figure 2), and transthoracic echocardiography (TTE) showed an increase in the atria, significant tricuspid regurgitation, enlarged right ventricle, PAP of 123.7 mm Hg and ventricular ejection fraction (LVEF) 47% (Figure 3). Subsequently, we performed transesophageal echocardiography (TEE) that demonstrated the presence of an atrial septal defect (ASD) with left to right shunt (Figure 4). At thorax computed tomography the main finding was a protrusion of the pulmonary artery (Figure 5). A room air arterial blood gases revealed the following data: pH=7.43, pCO_2=23.4 mm Hg, pO_2=53.9 mm Hg and O_2 saturation=89.8%. Based on the clinical case proposed we diagnosed ES with high PAP, cyanosis of the limbs at rest and the presence of ASD.

Initially we opted for drug treatment, suggesting the use of diltiazem in progressive doses of 720 mg/day,

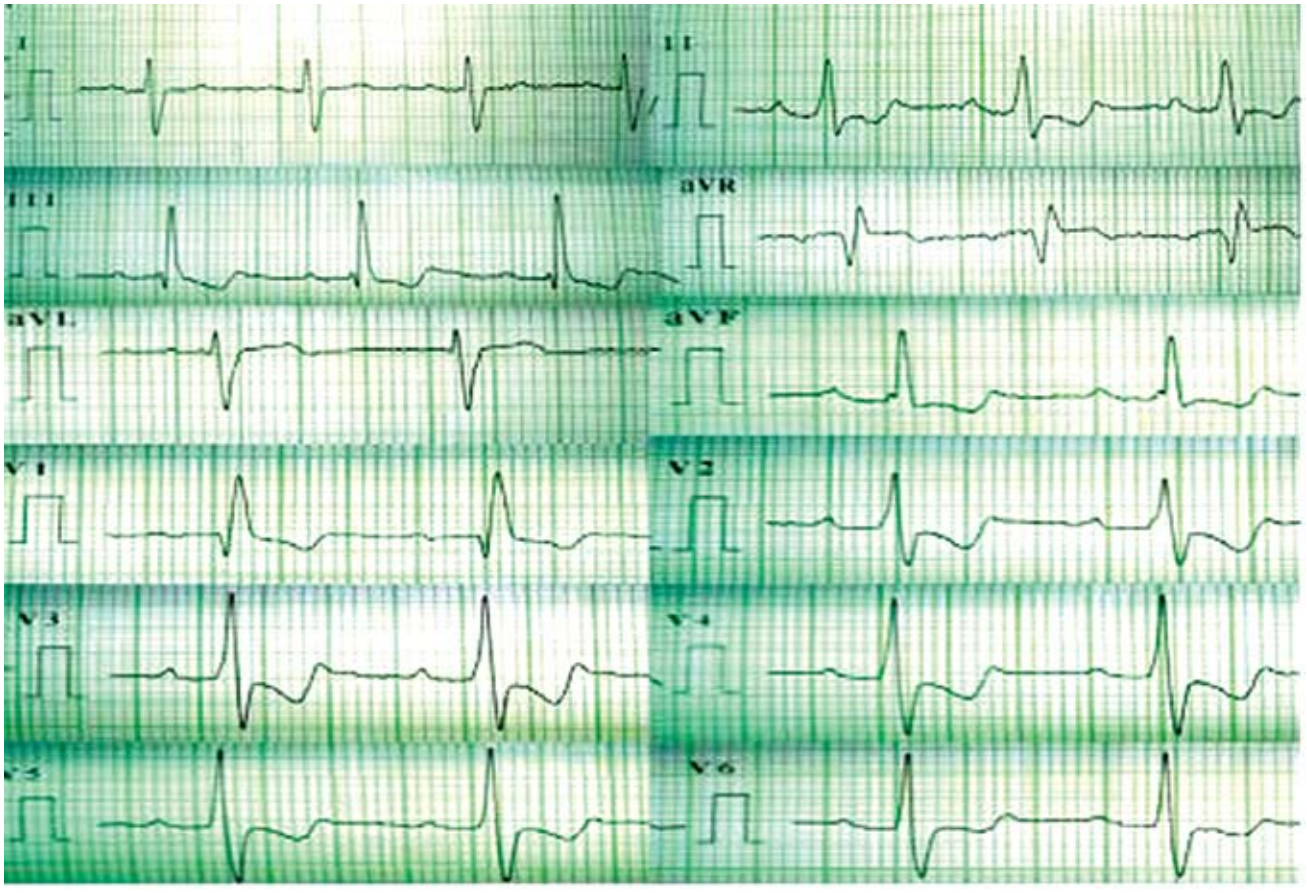

Figure 2. Electrocardiographic changes according to described pathology.

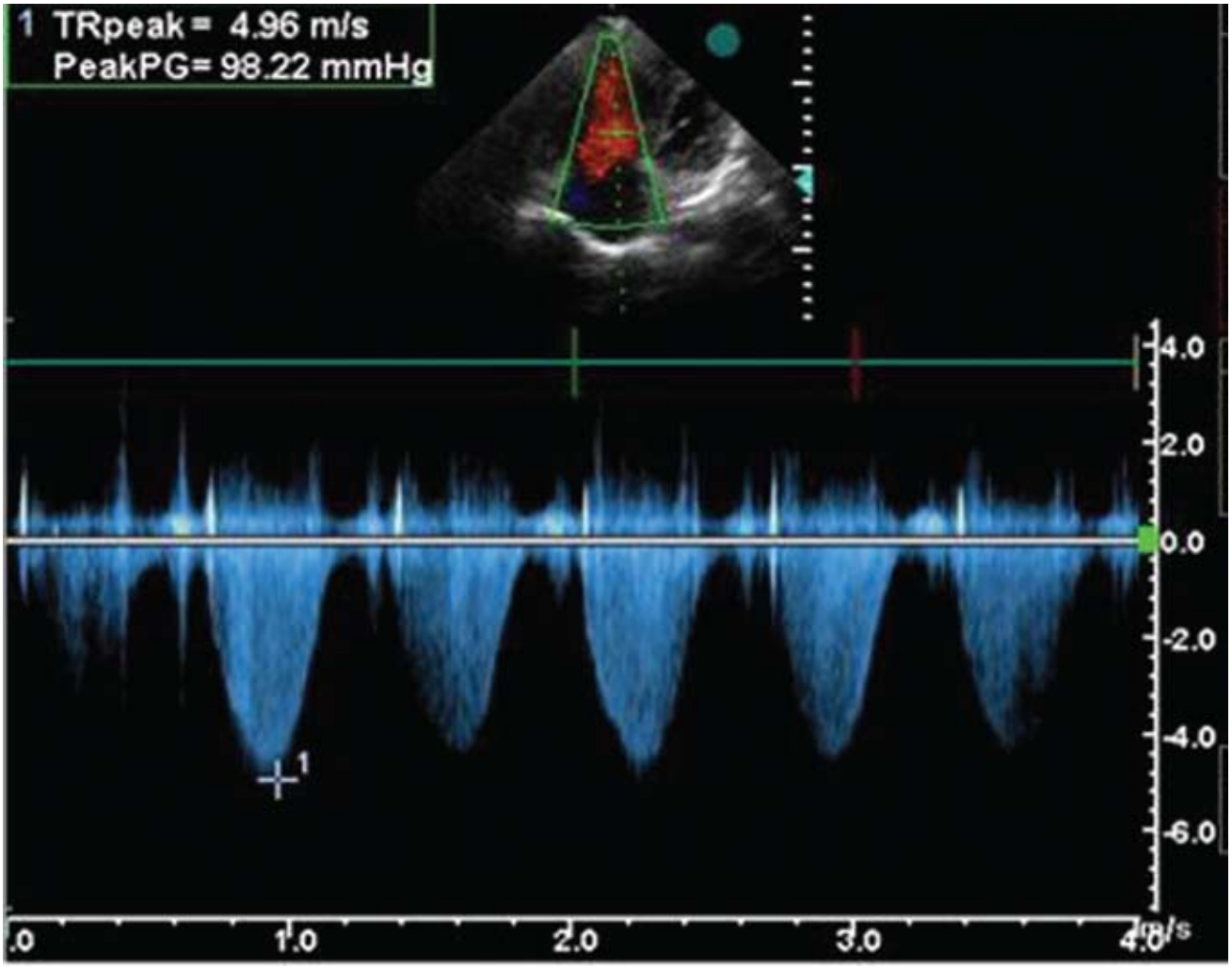

Figure 3. Pulmonary hypertension echocardiographic demonstration.

according to Venetia Consensus[4], furosemide 20 mg/day and spironolactone 25 mg/day. After 15 days of use in increasing doses of calcium antagonist, diltiazem, there was a 20% drop in the PAP (98 mm Hg) measured by TTE by the same previous operator. Currently, the patient is stable as an outpatient.

Discussion

The incidence of CHD in the general population is 1%. About 8% of patients with CHD and 11% of those with left to right shunt develops a framework for ES[5]. Increased pulmonary vascular resistance can occur between 5% and 10% of patients with untreated atrial septal defect, predominantly in women. The pathogenesis of PAH in some patients is unknown. Generally, we consider the presence of an ES when the ASD is large and not restrictive, in presence of cyanosis at rest[6]. In the description of this case we observed, according to literature, a patient with PAH, ASD and cyanosis at rest.

Physical examination of these patients is varied and can reveal central cyanosis that may be affected due to increase vascular resistance, when subjected to higher temperatures, exercise, fever, high altitude or systemic infection. The signs of PAH on physical examination present the palpable pulmonary valve closure and hyperphonetic second heart sound components. Blood pressure is usually palpable or decreased. There may be a diastolic murmur of pulmonary regurgitation (Graham-Steel)[7].

For differential diagnosis with additional tests in those patients who present at chest radiography a prominent pulmonary artery and ventricular septal defect (VSD), the cardiothoracic ratio is decreased, while the ASD carriers have cardiomegaly with dilated right ventricular secondary to increased pressure load[8]. TTE can identify valvular or cardiac defects. Associated Doppler allows identification of shunts. TEE is useful in those patients in whom there is difficulty in identifying the pulmonary artery pressure or septal defects[9]. Magnetic resonance imaging (MRI) can identify intracardiac defects and patent ductus arteriosus, especially in those with previous cardiac surgery. MRI can detect shunts from left to right or bidirectional, but have no obligation to quantify the shunt. Cardiac catheterization is very useful to detect, locate and quantify the shunt and to determine the severity of pulmonary vascular disease, however it has fallen into

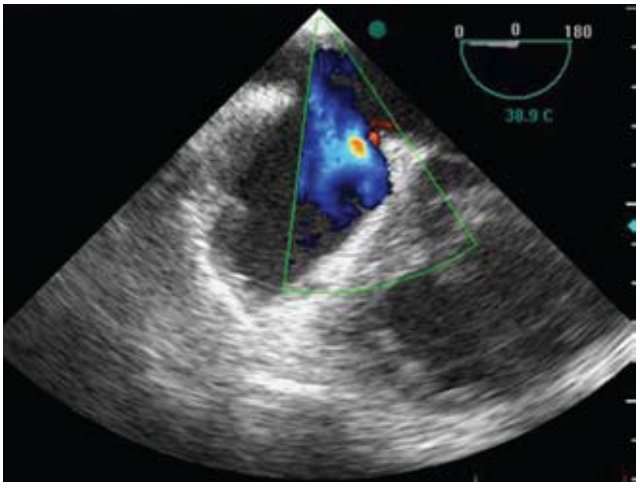

Figure 4. Transthoracic Doppler echocardiography showing interatrial communication presence.

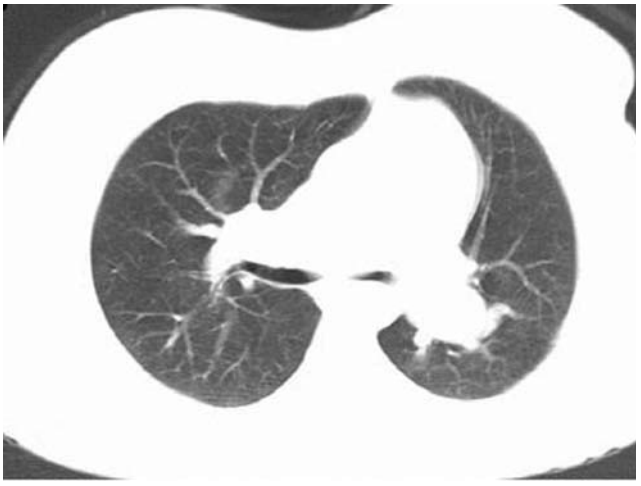

Figure 5. Pulmonary artery significant dilation at computed tomography.

disuse as advances in echocardiography have helped to achieve these measurements[10].

Efforts are directed to a treatment to reduce pulmonary vascular resistance, left to right shunt, cyanosis, morbidity and mortality. These measures have been disappointing. Calcium channel blockers lower systemic atrial pressure and decrease shunt, and may lead to syncope and sudden death. Their use is controversial, existing results of good development and at the same time, adverse outcomes in others. Oxygen therapy is not routinely recommended but is useful in patients with profound hypoxemia, dyspnea at rest or limited activity[11,12].

The long-term prognosis of patients with ES is better than other conditions associated with PAH, such as primary pulmonary hypertension. These patients have a survival of 80% at 10 years, 77% at 15 years and 42% at 25 years. The prognosis is not influenced by the location of the intracardiac defect. Variables associated with worse long-term prognosis are: syncope, right cavity inflation pressure and severe hypoxemia[13]. The patient described has a rare disease with atypical presentation, considering that age is not typical of the disease or the response to treatment with diltiazem at high doses, maintaining a good outcome so far.

References

1. Eisenmenger V. Die angeborenen Defects des Kammerscheidewand des Herzen. Z Klin Med 1897;32(Suppl):1-28.
2. Wood P. The Eisenmenger syndrome or pulmonary hypertension with reversed central shunt. Br Med J 1958;2:701-709.
3. Kidd L, Driscoll DJ, Gersony WM, Hayes CJ, Keane JF, O'Fallon WM, Pieroni DR, Wolfe RR, Weidman WH. Second natural history study of congenital heart defects: results of treatment of patients with ventricular septal defects. Circulation 1993;87:I38-I51.
4. Galiè N, Rubin LJ, eds. Pulmonary arterial hypertension. Epidemiology, Pathobiology, assessment, and therapy. J Am Coll Cardiol 2004;43(Suppl S):1S-90S.
5. Young D, Mark H. Fate of the patient with the Eisenmenger syndrome. Am J Cardiol 1971;28:658-669.
6. Webb G, Gatzoulis MA. Atrial septal defects in the adult: recent progress and overview. Circulation 2006;114:1645-1653.
7. Vongpatanasin W, Brickner ME, Hillis LD, Lange RA. The Eisenmenger syndrome in adults. Ann Intern Med 1998;128:745-755.
8. Rees RS, Jerrerson KE. The Eisenmenger syndrome. Clin Radiol 1967;18:366-371.
9. Chen WJ, Chen JJ, Lin SC, Hwang JJ, Lien WP. Detection of cardiovascular shunts by transesophageal echocardiography in patients with pulmonary hypertension of unexplained cause. Chest 1995;107:8-13.
10. Boehrer JD, Lange RA, Willard JE, Grayburn PA, Hillis LD. Advantages and limitations of methods to detect, localize, and quantitate intracardiac right-to-left and bidirectional shunting. Am Heart J 1993;125:215-220.
11. Wong CK, Yeung DW, Lau CP, Cheng CH, Leung WH. Improvement of exercise capacity after nifedipine in patients with Eisenmenger syndrome complicating ventricular septal defect. Clin Cardiol 1991;14:957-961.
12. Wimmer M, Schlemmer M. Long-term hemodynamic effects of nifedipine on congenital heart disease with Eisenmenger's mechanism in children. Cardiovasc Drugs Ther 1992;6:183-186.
13. Saha A, Balakrishnan KG, Jaiswal PK, Venkitachalam CG, Tharakan J, Titus T, et al. Prognosis for patients with Eisenmenger syndrome of various aetiology. Int J Cardiol 1994;45:199-207.