

## Cardiac Tamponade in Eisenmenger Syndrome: A Diagnosing Complexity: Case Report

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### Abstract

### Case Report

Cardiac tamponade is a form of obstructive shock with clinical manifestations consistent with a low cardiac output and high central venous pressure. Features of a low cardiac output include low mean arterial pressure, cold peripheries and signs of poor end-organ perfusion. Characteristically, palpating the pulse reveals an apparent variation in pulse volume because of pulsus paradoxus. Jugular venous pressure is typically increased, with distended neck veins apparent. Sympathetic tone is increased in an attempt to compensate for the low cardiac output and manifests as tachycardia, diaphoresis, anxiety [1] and poor distal perfusion determining the need for emergent Pericardiocentesis. We report a case where an hemodynamically significant pericardial effusion with indication of pericardiocentesis had presented without classic signs of tamponade. It is an unusual and rare case of large pericardial effusion in the setting of Eisenmenger syndrome where only clinical signs as worsening hypoxia and cyanosis due to the right to left shunt was the warning sign of cardiac tamponade requiring pericardiocentesis.

**Keywords:** Cardiac, Eisenmenger, pulsus paradoxus.

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## CASE REPORT

We describe the case of a 20 year-old female known to have Down syndrome, uncorrected complete atrioventricular septal defect and consequently Eisenmenger Syndrome, who presented with dyspnea without orthopnea, hypoxia and cyanosis to the emergency department. She was found to be hemodynamically stable with a heart rate of 94 bpm and blood pressure of 127/78 mmHg with no pulsus paradoxus. She was notably cyanotic, and hypoxic with an oxygen saturation of 85%. Physical examination showed a muffled heart sounds with digital hippocratism.

Chest X-Ray revealed cardiomegaly (Figure 1), and the electrocardiogram showed sinus tachycardia associated to right ventricular hypertrophy without low voltage or electrical alternans. The Chest CT angiography didn't show pulmonary embolism or congestion.

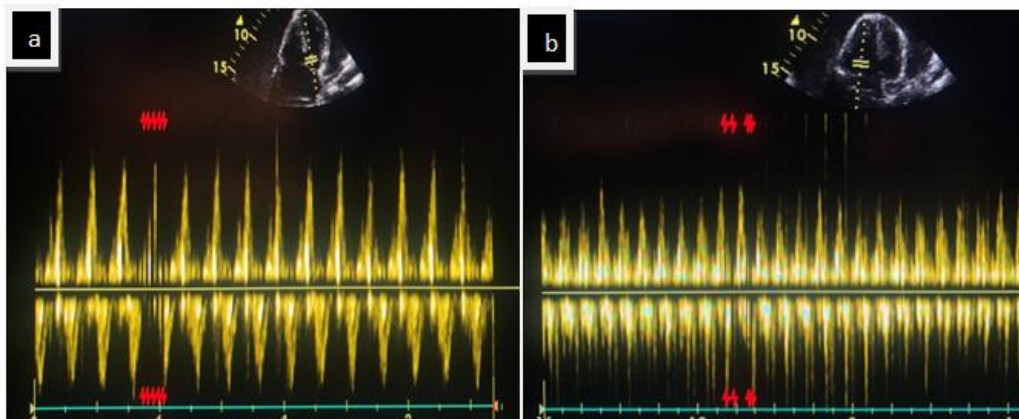


**Fig-1: Chest X-Ray showing cardiomegaly**

An urgent echocardiography confirmed the presence of large circumferential pericardial effusion with a maximum dimension of 30mm with no echocardiographic features of cardiac tamponade: no right atrial or right ventricular collapse, the respiratory variations in mitral, aortic and tricuspid inflow velocities didn't remain significant (Figure 2). A right

to left shunting was observed across the atrioventricular septal defect. The function of the right ventricle was correct with a wall thickness of 15 mm. Quantitation of

RV function (such as TAPSE, or fractional shortening), with severe pulmonary hypertension and sPAP (systolic pulmonary artery pressure) of 124 mmHg.



**Fig-2: Absence of significant mitral (a) and tricuspid (b) inflow variation during respiration**

The Challenge was to determine whether the symptomatology was due the large pericardial effusion or to Eisenmenger syndrome in the absence of the classic signs of tamponade which indicates an emergent management.

We decided to perform a pericardiocentesis as the dyspnea got worse with an oxygen blood saturation of 78% with spectacular improvement of the clinical state.

## DISCUSSION

Eisenmenger syndrome is a complication due to chronic systemic to pulmonary shunting, leading to an increase in pulmonary blood flow responsible of endothelial dysfunction, vascular remodeling and elevated pulmonary vascular resistance. Besides, RA dilatation and RV hypertrophy are the result of a chronic exposure of the pulmonary arterial bed to systemic pressure. Eventually, the existing shunt becomes bidirectional or inverted. This manifests as hypoxia, platypneic and cyanosis [2].

Progression to right-sided heart failure is relatively slow and may be attributed to a more resistant right ventricle. However, once it develops, it is a strong predictor of death [3]. Pericardial effusion is common in Eisenmenger syndrome, and although it is a manifestation of heart failure, this has not been shown to be of prognostic significance [4].

Cardiac tamponade occurs when overwhelming intrapericardial pressure from either a rapidly expanding or sizeable pericardial effusion leads to collapse of the cardiac chambers and impairment in diastolic filling. The right side of the heart is usually affected before the left, but a loculated collection around the left heart may cause left-sided effects. Echocardiography findings consistent with pericardial tamponade include collapse of the cardiac chambers,

inferior vena cava dilatation, increased respiratory variation in the intracardiac blood flow measured with Doppler and excessive leftward shift of the interventricular septum during spontaneous inspiration. Collapse of cardiac chambers occurs at the time of lowest pressure during the cardiac cycle (right atrial systolic and RV diastolic collapse). With impaired blood flow into the right atrium the IVC progressively dilates and loses its usual variation in diameter with respiration. The associated clinical and echocardiographic signs result from the exaggerated shifting of the interventricular septum towards the left ventricle as the compressed RV fails to accommodate the increased venous return during inspiration. This leads to a reduction in left ventricular size and stroke volume [1].

To date, the characteristics of the clinical and echocardiographic parameters observed in the association of tamponade and Eisenmenger syndrome have not been illustrated in literature. This case shows the difficulty in recognizing the perturbation in hemodynamics caused by the presence of a pericardial effusion in the setting of complex congenital cardiac diseases, even when aided by echocardiography. In the case of our patient two mechanisms contributed to this: First, the absence of an intact septum eliminated the effects of ventricular interdependence on stroke volume. Instead, the unimpeded flow through the AVSD allowed for more right to left shunting and resulted in worsening of the patient's hypoxemia. It has already been described that five patients with atrial septal defect and proved cardiac tamponade had neither paradoxical pulse nor the inspiratory reduction of left ventricular internal dimension associated with this sign. It is assumed that equilibration of flow across the atrial septal defect prevented paradoxical pulse. Patients with a large atrial septal defect and tamponade do not manifest a paradoxical pulse [5].

Another case of Eisenmenger syndrome complicating a 31 year old patient with atrioventricular septal defect, described that only hypoxemia was the alarming sign of tamponade, and explained that was a result of increased right-to-left shunting due to non-compartmentalized diastolic restriction [6]. In addition, as with primary pulmonary hypertension, markedly elevated pulmonary pressures, non-compliance of the pulmonary vasculature and right-sided chambers, and right ventricular hypertrophy in Eisenmenger's syndrome, protects against diastolic chamber collapse in the face of a significant pericardial effusion [7]. As a result, right chamber collapse will likely be a late finding as seen in our patient's case (Figure 3).



**Fig-3: AVSD and large pericardial effusion with no collapse of the right atrium**

## CONCLUSION

This case explains how the consequences of pulmonary hypertension in right chambers in the setting of Eisenmenger syndrome can protect against the collapse and hemodynamic compromise of tamponade.

Besides, the assessment of a hemodynamically significant pericardial effusion in the presence of complex congenital heart disease and to determine if the symptomatology of the patient is due to tamponade or the chronic evolution of Eisenmenger syndrome seems

inconclusive. Moreover, septal defects modify the effects of ventricular interdependence on stroke volume, resulting in the absence of a pulsus paradoxus and its associated traditional echocardiographic findings.

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