

## Diagnosing and Managing Eisenmenger Syndrome in a Young Male

S. Aravinda Kumar<sup>1</sup>, S. Sujibala<sup>2</sup>

<sup>1</sup>Consultant Cardiologist, Kauvery Hospital, Heartcity, Trichy

<sup>2</sup>Duty Medical Officer, Kauvery Hospital, Heartcity, Trichy

\*Correspondence: aravindgowri@yahoo.co.in

### Abstract

Eisenmenger syndrome is a rare and complex condition that arises as a result of certain congenital heart defects. It is characterized by the reversal of blood flow within the heart, leading to severe pulmonary hypertension and subsequent heart failure. Managing this condition requires a multi-disciplinary approach, and early detection is crucial to improve the patient's quality of life. In this article, we discuss the case of a 20-year-old male who presented with symptoms of breathlessness on exertion and was diagnosed with Eisenmenger syndrome.

### Case Presentation

The patient, a 20-year-old male was diagnosed at the age of 8 months of life to have congenital heart disease for which appropriate treatment was not advised.

He was evaluated 10 years before current consultation. Echocardiogram had shown a large muscular ventricular septal defect (VSD) with inlet extension, resulting in a bidirectional shunt (predominantly left-to-right). It also showed severe pulmonary arterial hypertension (PAH) and good biventricular function.

Unfortunately, further details regarding cardiac catheterization were not available.

Since the age of 16years, he has been experiencing progressive breathlessness on exertion.

Recently, his symptoms worsened, prompting him to seek medical attention.

Physical examination revealed dusky appearance of extremities, indicative of significant disease progression.

His vital signs were within normal limits, but his oxygen saturation (SpO<sub>2</sub>) was alarmingly low, at 88% on room air. Electrocardiogram (ECG) findings showed Biventricular hypertrophy and Echocardiogram demonstrated a large inlet VSD measuring 10mm,

bidirectional shunt (predominantly right-to-left). Additionally, it revealed moderate left ventricular dysfunction, severe PAH, and mitral valve prolapse (MVP)

Based on the findings, the patient was diagnosed with Eisenmenger syndrome, characterized by the bidirectional shunting of blood due to the large VSD and severe PAH. This condition is often associated with poor prognosis and reduced life expectancy.

### ECG

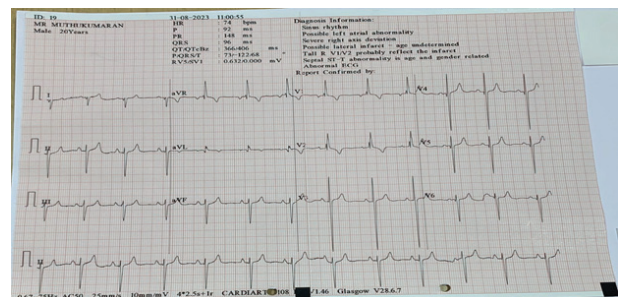


Fig (1). Finding showed biventricular hypertrophy suggestive of VSD with Severe PAH

### Discussion

Eisenmenger Syndrome is a rare and complex heart condition that often develops as a complication of another condition known as a congenital heart defect. This syndrome occurs when a defect in the heart's structure leads to high blood pressure in the lungs, resulting in irreversible damage to the blood vessels.

To better comprehend Eisenmenger Syndrome, it is essential to first understand congenital heart defects. These are abnormalities present at birth that impact the heart's structure and function. Some common examples include ventricular septal defects (VSDs), atrial septal defects (ASDs) and patent ductus arteriosus (PDA).

In a healthy heart, oxygen-rich blood is pumped from the left side of the heart into the main artery (aorta),

supplying it to the rest of the body. Simultaneously, oxygen-depleted blood returns to the right side of the heart. However, if an individual has a congenital heart defect, there may be an abnormal blood flow pattern, causing an imbalance in oxygenation levels.

If left untreated, these defects can lead to Eisenmenger Syndrome. As blood flows abnormally through the heart, the increased pressure in the pulmonary artery, which carries blood from the heart to the lungs, can cause damage to the blood vessels in the lungs over time. This ultimately results in the reversal of blood flow, with oxygen-poor blood mixing with oxygen-rich blood and being pumped back into the main arterial system.

Eisenmenger Syndrome has various symptoms, ranging from mild to severe. Fatigue, shortness of breath, and cyanosis (a bluish tint to the skin and lips) are common indicators of the condition. Diagnosing thorough examination of the patient's medical history, physical examination, and various diagnostic tests. Echocardiography, electrocardiography (ECG), and cardiac catheterization are required to evaluate the heart's structure, functioning, and blood flow patterns. Managing Eisenmenger Syndrome can be challenging, as there is currently no cure. However, a multidisciplinary approach involving cardiologists, pulmonologists, and other healthcare professionals can help alleviate symptoms and improve overall quality of life. Treatment aims to reduce complications and improve heart function, as well as optimize oxygen levels in the blood. Patients often undergo regular monitoring to evaluate the progression of the syndrome and minimize the risks associated with it. In severe cases, heart and lung transplantation may be considered.

### Conclusion

The case discussed highlights the challenges posed by Eisenmenger syndrome, a life-threatening complication of congenital heart disease. Its presentation in a young male emphasizes the importance of early detection and management to mitigate complications and improve the patient's quality of life.

In this case, the patient was advised to avoid physical exertion, medications to reduce pulmonary artery pressure.

The patient is also advised for heart-lung transplantation, depending on his overall health condition and availability of suitable donors.