

Eisenmenger Syndrome: A Comprehensive Review

Bharti Sachdeva

Assistant Professor, Faculty of Nursing, SGT University, Gurugram, India

E-mail ID: [bharti.nursing\[at\]sgtuniversity.org](mailto:bharti.nursing@sgtuniversity.org)

Abstract: Eisenmenger syndrome (ES) is a medical complication, arising from large congenital anatomical shunts present in the heart. It is the most severe form of pulmonary hypertension characterized by elevated pulmonary vascular resistance and a reversed right-to-left shunting of blood in originally left-to-right shunting. Due to defects present at birth, pressure gradient initially results in a left-right shunt, which leads to severe pulmonary arterial hypertension (PAH) and elevated vascular resistance. Eventually, due to increased pulmonary vascular resistance, the left-to-right shunt becomes a right-to-left shunt, leading to significant hypoxemia and cyanosis. Although the syndrome is rare and a cardiovascular nurse may not get to see many patients with Eisenmenger syndrome, nurses need to have some basic knowledge about the syndrome, in case they need to care for such a patient in their clinical practice. This review aims to summarize the current knowledge on pathophysiology, clinical presentation and treatment options available for ES.

Keywords: eisenmenger syndrome, pulmonary arterial hypertension, congenital heart defect, left- to-right shunt

1. Introduction

Eisenmenger syndrome is defined as the complication of various congenital heart defects such as atrial septal defect, ventricular septal defect, atrioventricular septal defect, patent ductus arteriosus, truncus arteriosus and single ventricle anomalies [1]. The structural abnormalities of the pulmonary circulation are histologically similar to those seen in other forms of pulmonary hypertension. Eisenmenger syndrome is a multisystem disorder associated with numerous life-threatening complications, including hemoptysis, cerebrovascular accidents, brain abscesses, arrhythmias, and syncope. Exercise capacity is severely impaired in most patients with Eisenmenger syndrome. Although exercise limitation and exertional dyspnea may remain stable for years, poor exercise capacity identifies the patient at risk for hospitalization or death [2].

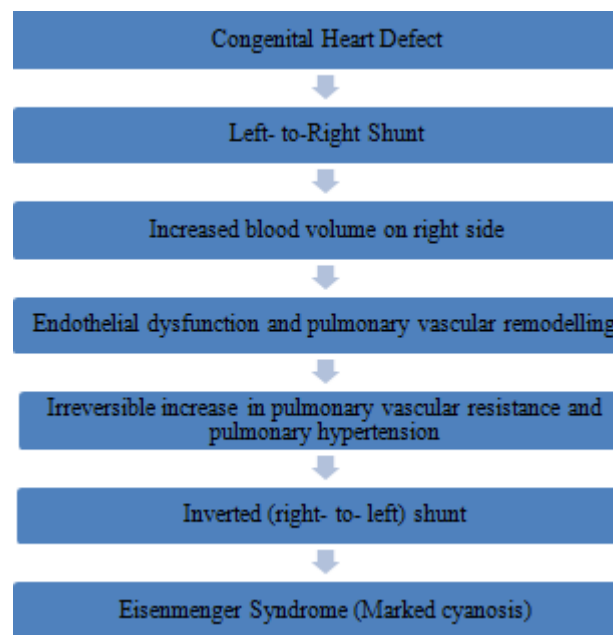
Epidemiology

No precise data on the incidence of Eisenmenger syndrome is available. It is estimated that around 5–10% of patients with congenital heart disease develop pulmonary arterial hypertension (PAH) while 1% to 4% develop Eisenmenger syndrome.[3] The probability of developing the Eisenmenger syndrome depends on the size and location of the defect. For example the incidence of ES is 3% with small or moderate-sized ventricular septal defects whereas it increases significantly to approximately 50% of infants with large unrepaired ventricular septal defects. [4] Almost all patients with unrepaired truncus arteriosus are at risk of developing ES. [5] The risk of mortality attributing to ES is approximately 10-12 folds high in patients with complex CHD. Similarly, 80% of the patients with patent ductus arteriosus or ventricular septal defects develop ES during infancy than do patients with atrial septal defects i.e. 90% during adulthood. [6]

Pathophysiology

Physiologically, the ES is preceded by the presence of a congenital defect in the heart allowing blood flow across the

pressure gradient i.e. from systemic to pulmonary circulation. Overtime, the increased blood volume and pressure in pulmonary circulation results in changes in pulmonary microvasculature leading to increased pulmonary vascular resistance and subsequently pulmonary hypertension. As soon as the pressure in the right side of the heart exceeds that in the left side, the blood starts mixing from pulmonary to systemic circulation (reversal of shunt) presenting marked hypoxemia and cyanosis. [7]



Clinical manifestations

- a) Clubbing of fingers/ toes
- b) Exercise intolerance
 - Shortness of breath
 - Fatigue
 - Chest pain
 - Central cyanosis
 - Syncope

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- c) Right sided heart failure
 - Peripheral oedema
- d) Compensatory erythrocytosis
 - Hyperviscosity
 - High haemoglobin and hematocrit levels
 - Increased risk of bleeding secondary to lower production of platelets and clotting factors
- e) Prominent parasternal ribs due to right sided hypertrophy
- f) Louder S₂
- g) Pansystolic murmur of tricuspid valve regurgitation

Management

Medical management

Primarily the medical management focuses on the symptomatic management (depending upon the clinical presentation of the patient), pulmonary vasodilation, and prevention and/ or treatment of heart failure.

Studies do not recommend calcium channel blockers for patients with ES since reduced systemic vascular resistance may lead to worsening of right to left shunting, syncope and sudden death. [9] Pulmonary vasodilation can be achieved by administering endothelin antagonists and phosphodiesterase (PDE) inhibitors. [10]

The use of aspirin is controversial to counter effect the hyperviscosity secondary to erythrocytosis because of high bleeding tendency. [9] Although, it may be prescribed in case of recurrent thromboembolic events after thorough assessment of risks and benefits. Patients with obvious signs of viscosity should be managed with isovolemic phlebotomy. In this procedure blood is slowly removed through a venipuncture (250 mL over 30 min.) and equal volume of normal saline is infused into the body. Prevention of pneumonia and endocarditis is advocated with yearly influenza and pneumococcal vaccination and prophylactic antibiotics respectively to delay deterioration and death.

Surgical management

Unfortunately, once the ES has developed; it is almost unlikely to reverse the underlying pathophysiology. Definitive treatment either includes repair of congenital heart defects with lung transplantation or heart-lung transplantation. [11]

2. Counseling

Patients are advised to avoid certain risk factors such as dehydration, chest infection, pregnancy, high altitude, strenuous exercise and smoking.

3. Conclusion

ES is a complex medical condition associated with significant morbidity and mortality. A multidisciplinary approach is advisable to improve the quality of life, prevent complications and delay death.

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