

Eisenmenger Syndrome

In the late 1800's, a physician by the name of Victor Eisenmenger was the first to describe the medical condition of a patient with congenital heart disease and pulmonary hypertension.

Today, the term Eisenmenger syndrome is used to describe this very same condition.

Certain congenital heart defects that are not surgically repaired can cause Eisenmenger syndrome. These defects include:

- (ASD) Atrial Septal Defects
- (VSD) Ventricular Septal Defects
- (PDA) Patent Ductus Arteriosus
- (AV Canal) Atrioventricular Canal Defect
- (DILV) Double Inlet Left Ventricle
- (DORV) Double Outlet Right Ventricle
- (HLHS) Hypoplastic Left Heart Syndrome
- (D-TGA) D- Transposition of the Great Arteries
- (L-TGA) L - Transposition of the Great Arteries

Normal Heart Circulation

The ventricles are the pumping chambers of the heart. The right ventricle pumps blood to the lungs and the left ventricle pumps blood to the body.

In a structurally normal heart there is no mixing of blood between the two ventricles. See figure 1.

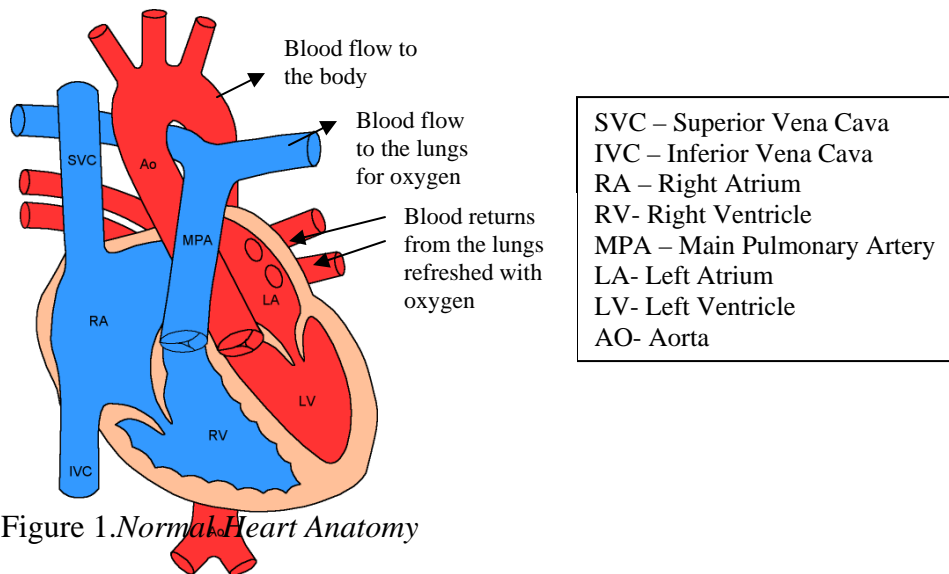


Figure 1. *Normal Heart Anatomy*

Abnormal Heart Circulation

An example of one of the most common congenital heart defects that can lead to Eisenmenger syndrome is known as a ventricular septal defect. See figure 2.

A ventricular septal defect (VSD) means that there is a hole in the heart between the right and left ventricles.

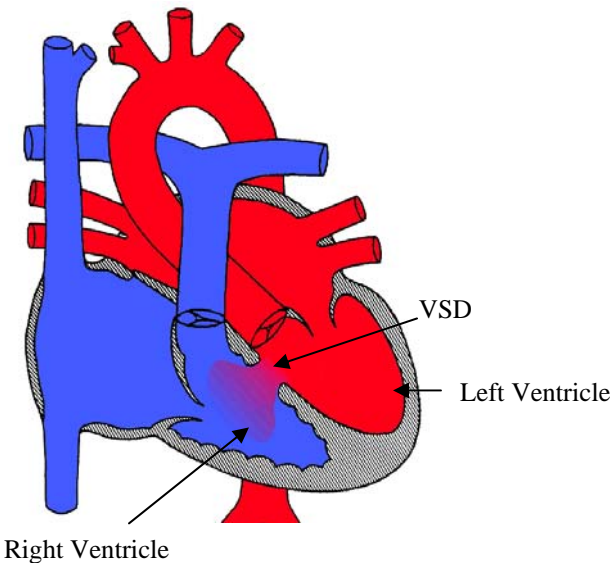


Figure 2. *Ventricular Septal Defect*

Because the blood pressure in the left side of the heart is much higher than the blood pressure in the right side, the hole (defect) permits blood to flow (shunt) from the left side of the heart into the right side of the heart. This results in high pressure and increased amount of blood sent to the blood vessels of the lung.

If this shunting continues for many years without closure of the hole or shunt, the over circulation of blood and increased pressure results in abnormal changes in the lung blood vessels. The blood vessels will become constricted and this constriction will limit the flow of blood through these pulmonary vessels.

These changes in the pulmonary blood vessels will cause the blood pressure within the vessels to rise. This is known as pulmonary hypertension. Unlike the body's blood pressure, blood pressure within the lungs should be very low- about 1/5th the bodies blood pressure.

Eventually the blood pressure in the pulmonary circulation will equal and may even be higher than the body's blood pressure. When this occurs, the direction of blood flow through the hole will reverse. This is known as Eisenmenger syndrome. See figure 3.

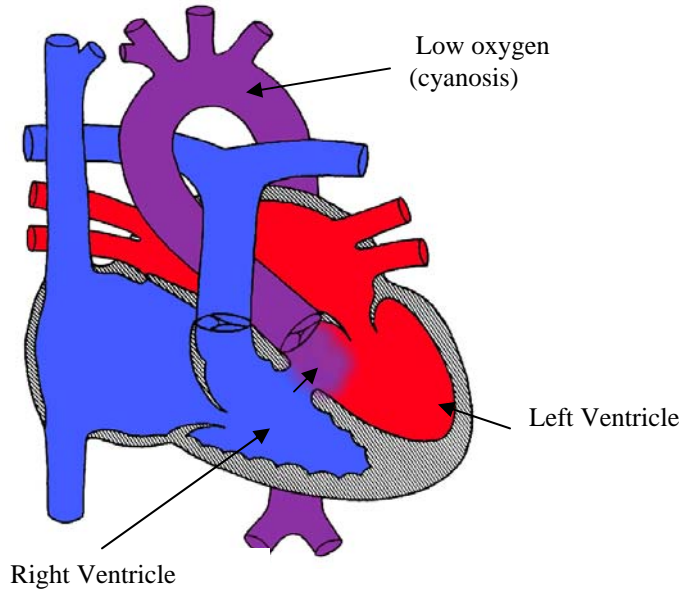


Figure 3. *Ventricular Septal Defect. Right to Left Shunt. Eisenmenger syndrome*

This reversal of blood circulation means some of the un-oxygenated blood from the right side of the heart will bypass the lung circulation and cross (shunt) through the hole to the left side of the heart and out to the body.

Although some of the un-oxygenated blood does travel to the lung to be refreshed with oxygen the majority of blood shunts across the hole to the left side of the heart and eventually out to the body. This is called a right to left shunt. Figure 3.

People who develop Eisenmenger syndrome may have no symptoms for many years, especially during their childhood years. However many people with Eisenmenger syndrome develop blue appearing skin, particularly the lips and nail beds. This is due to the low oxygen levels caused by the shunting of blood from the right side of the heart out to the body. This lack of oxygen can also cause shortness of breath (difficulty breathing)

Symptoms of Eisenmenger syndrome

- Shortness of breath – Difficulty breathing with physical activity.
- Shortness of breath at rest
- Fatigue
- Chest pain or pressure
- Palpitations (heart racing)
- Dizziness or syncope(passing out)
- Cyanosis (blue color) skin, lips, fingers, toes and nail beds due to low oxygen levels in blood
- Impaired exercise tolerance – inability to perform simple or moderate activities without feeling shortness of breath

Treatment/Long Term Outlook

People with Eisenmengers can have a relatively healthy life for many years. Today, we have several medical therapies available that have been shown to improve quality of breathing for patients living with Eisenmenger syndrome. The goal of treatment with Eisenmenger syndrome is to decrease the pulmonary blood pressure and improve heart function.

Medications: Patients living with Eisenmenger syndrome have several treatment options to improve symptoms of the disease and their quality of life. Available therapies include the following:

- Intravenous Flolan
- Intravenous Remodulin
- Subcutaneous Remodulin
- Endothelin Receptor Antagonists: Tracleer (Bosentan)/ Ambrisentan(Letaris)
- Phosphodiesterase type 5 inhibitor: Viagra (Sildenafil)
- Inhaled Iloprost (Ventavis)
- Clinical research of new medications
- Lung Transplant
- Combined Heart and Lung Transplant

Oxygen

Oxygen may be worn with activity or during sleep to improve symptoms of shortness of breath.

Surgical Care: There is no surgical care that is available to correct the cardiac defect once Eisenmenger syndrome has developed.

Transplant: Combined Heart-Lung Transplant or Lung transplant with repair of congenital heart defect are the only possible surgery options available. Transplant is reserved for patients who are doing poorly despite medical treatment.