

### What is Eisenmenger syndrome (ES)?

Eisenmenger syndrome was first described in 1897 by Dr. Victor Eisenmenger. It occurs as the result of a heart defect in which there is a defect or hole (shunt) between two chambers in your heart. With this defect, blood moves from the left side of the heart to the corresponding right chamber. This increases blood flow into the lungs. If the defect or hole is not closed, over time, this increased flow of blood to the lungs damages the lung arteries and tissues. As a result, pressure in the lung arteries increases. This is known as pulmonary arterial hypertension. This increased pressure eventually leads to shunt reversal. This means the blood now moves in the wrong direction from a right sided chamber (low oxygen) to a left heart chamber, causing low oxygen in the blood in your body. This leads to cyanosis and other manifestations of ES.

The incidence of ES has decreased over years from 8% in the 1950's to 4% in the 2000s. This is because septal defects are repaired more often and earlier. ES is more often diagnosed in young adulthood as the damage takes years to occur but may vary from one patient to another.

### Which congenital heart defects cause ES?

- Unrepaired ventricular septal defects (VSD) as small as 1 centimeter
- Unrepaired atrial septal defect (ASD) between 1.5 and 2.0 centimeters
- Atrioventricular (AV) canal defect or complete AV canal
- Patent ductus arteriosus (PDA).
- Other types of complex congenital heart disease, such as single ventricle or double inlet left ventricle.

### What are the signs and symptoms of Eisenmenger syndrome?

The symptoms depend on the organs that are affected. The most common visible signs are cyanosis (blueness) and clubbing (large rounded finger nails or toe nails).

Other signs and symptoms include:

- Chest pain
- Heart palpitations
- Coughing up blood (hemoptysis)
- Dizziness and fainting (syncope)
- Fever, poor appetite and sweating when there is blood stream infection
- Feeling tired
- Shortness of breath at rest or with walking
- Weight gain or leg swelling
- Abdominal pain
- Stroke like symptoms including visual disturbances and headache
- Gout

### How is Eisenmenger syndrome diagnosed?

The diagnosis of ES is based on the presence of three symptoms:

- A cardiac shunt or opening between the two chambers on opposite sides of the heart
- Pulmonary hypertension that cannot be reversed with medication
- Cyanosis or blueness

In addition to blood tests, EKG and chest x-ray, other tests are needed to confirm the diagnosis. These include an echocardiogram, cardiac catheterization and a 6-minute walk test.

### What are the possible long term medical issues?

- Your red blood cell count and hemoglobin is usually high. This occurs because cyanosis causes your body to produce more red blood cells to compensate.
- People with ES have increased tendencies to bleed and to clot. You should watch out for easy bruising, bleeding from the gum, nose bleeding, excessive bleeding during a woman's period, or coughing up blood. You should let your ACHD doctor know if you have any of these.
- Arrhythmias or irregular or fast heart beats are also frequent complications of ES. These are more commonly originating from the upper chambers of the heart.
- Ventricular dysfunction or heart failure can occur. Let your ACHD doctor know if you develop shortness of breath, swelling in your abdomen or legs, and fatigue.
- Rarely, years of cyanosis and pulmonary arterial hypertension can cause one of the coronary arteries to become compressed causing chest pain.
- The risk of lung, heart and skin infections is also increased. Brain infections can also occur.

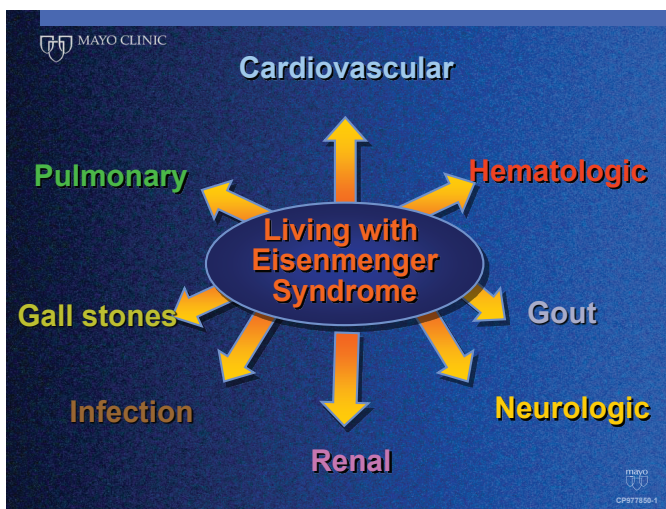


Illustration courtesy of Naser M. Ammash, MD, Mayo Clinic, Rochester, MN

- Strokes can occur in about 7-14 percent of Eisenmenger patients.
- Gout, an inflammation of the big toe or other joint, can occur. This is due to large levels of uric acid in your body.
- Abnormal kidney function occurs in more than 1/3 of patients with ES. To help prevent this, it is important to stay well hydrated, to avoid NSAIDS and any drugs that are toxic to the kidney.
- The development of gall stones is also increased. This causes abdominal pain after eating.
- Musculoskeletal complications such as curvature of the spine (scoliosis) and joint and long bone pain are possible.

*With recent advances in management and treatment, many people with Eisenmenger syndrome live longer and lead more active lives.*

### What are the options for treating Eisenmenger syndrome?

Treatment for ES is three-fold:

- Management of complications
- Medical therapy
- Heart/Lung transplant versus surgical repair of the heart defect and lung transplant

Management of complications includes:

- Followup with an ACHD specialist regularly
- Avoid adding salt to your diet
- Not smoking
- Avoid drinking alcohol
- Avoid high altitude
- Avoid hot tubs and saunas
- Do not take aspirin or non-steroidal medications such as Aleve and Ibuprofen
- Avoid strenuous exercise and competitive sports
- Prevent infections by having excellent oral health (mouth, gums, teeth)
- Take antibiotics to prevent subacute bacterial endocarditis when indicated
- O2 supplementation if needed or if it makes you feel better
- Have non-cardiac surgery in ACHD centers
- Take the Pneumovax and Flu vaccines
- Air travel should be okay in pressurized cabin
- Avoid pregnancy and seek counseling

Three groups of drugs are generally considered in treating the pulmonary hypertension. Your ACHD doctor will decide which is best for you. These are:

- A prostacyclin analogue (like Inhaled Iloprost)
- An endothelial receptor antagonist (like Bosentan)
- A vasodilator (like Sildenafil)

Heart/lung transplant is an option for patients whose hearts and lungs continue to fail after getting the best treatment

available. It must be done in a center that does heart/lung transplants and has an experienced ACHD team to care for the patient before, during and after the procedure.

The prognosis for patients with Eisenmenger syndrome is improving with better diagnosis and care.

### Can I have a baby?

Pregnancy is a very high risk for women with Eisenmenger syndrome. The maternal death rate is up to 50 percent. The risk of miscarriage is around 30 percent. Women are cautioned to avoid getting pregnant. If pregnancy occurs, close monitoring and bed rest will be necessary and this likely will require hospitalization.

### What about birth control?

You should talk to your doctor about contraception. Acceptable birth control can include use of the Mirena Coil IUD, and the option of endoscopic sterilization (Essure). Estrogen contained birth control pills should be avoided. Birth control pills that primarily contain progesterone are generally okay to use.

### What things can I do to improve my quality of life?

If you have Eisenmenger syndrome, you should:

- Be seen on a regular basis by an ACHD specialist
- Stay hydrated
- Avoid hot tubs, saunas and other activities that cause dehydration
- Make sure iron deficiency anemia is treated appropriately
- Avoid inappropriate phlebotomies
- Eat a low salt diet
- Don't smoke
- Avoid high altitudes
- No ASA/NSAID
- Exercise is okay—avoid strenuous/competitive sports.
- Talk to your doctor before beginning any exercise program
- Avoid infections—take care of skin, finger, toes and face
- Take antibiotics before dental procedures
- Have non-cardiac and cardiac surgery at centers with expertise in Eisenmenger syndrome
- Use supplemental oxygen if it makes you feel better (no proof it helps)
- Get your flu and pneumovax vaccines
- Travel by air is okay as long as you stay hydrated.

### Living longer and better

With recent advances in management and treatment, many people with Eisenmenger syndrome live longer and lead more active lives. If you have this condition, it is essential that you are treated by an ACHD doctor and a team with expertise in managing your condition and associated symptoms. Care from doctors who understand the challenges you face and can provide the high level treatment you need is crucial. Educating yourself is key to understanding this complex and challenging condition. Education and the right care will help assure you have the longest and best quality of life.