



EISENMENGER SYNDROME

What is it?

Unlike in the normal heart, (see The Normal Heart) women with Eisenmenger syndrome have high blood pressure in the lungs (medical term: pulmonary hypertension) that develops as a result of extra blood passing through a hole in the heart. The most common types of holes in heart are known as atrial septal defect and ventricular septal defect. Eisenmenger syndrome can also develop in women with an abnormal blood vessel connection known as a patent ductus arteriosus.

Because of the high blood pressure in the lungs, some of the oxygen-poor blood ('blue' blood) returning back to the heart flows directly to the body, instead of being sent through the lungs for loading with oxygen. As a result, women with Eisenmenger syndrome have low oxygen levels and appear 'blue' (medical term: cyanotic). Women can develop heart failure and abnormal rhythm disorders (medical term: arrhythmias). Low oxygen levels also results in many health issues including bleeding problems, blood clotting problems, kidney troubles and infections.

How safe is it for me to become pregnant?

There is a very high risk of complications for you and your unborn baby. Pregnancy is not advised if you have Eisenmenger Syndrome.

Because of the high-risk nature of pregnancy for women with Eisenmenger syndrome, it is important to discuss your specific condition with a physician who has expertise in the care of women with congenital heart disease in pregnancy. Some women with high-risk cardiac lesions, such as Eisenmenger syndrome, will seek alternatives to pregnancy including adoption.

Issues for the mother

Which forms of birth control are safe?

Because of the high risk of pregnancy associated with your heart condition, effective birth control (medical term: contraception) is essential. The combined progesterone/estrogen oral contraceptive pill is best avoided because it is associated with increased blood clot formation. Many of the progesterone-only forms of contraception are acceptable; however, progesterone-only pills can have high failure rates. Some women may choose more permanent forms of contraception. Contraception should be discussed with a physician who has an understanding of your underlying heart condition. (See Birth Control)

What are my risks if I become pregnant?

The most serious risk for women with Eisenmenger syndrome is that many women (some studies suggest up to 50% of pregnant women) will not survive the pregnancy.

Other problems that can develop during pregnancy include weakening of the heart (medical term: heart failure), abnormal heart rhythms (medical term: arrhythmias) and the development of blood clots (medical term: deep venous thrombosis, pulmonary embolism, and thromboembolism causing stroke). Even if you remain well, there is a significant chance that you will go into early labour.

There is a high risk of miscarriage in women with Eisenmenger syndrome.

Some medications are not safe in pregnancy. Medication use should be reviewed with your doctor if you are contemplating pregnancy or are pregnant. Do not stop medications without first checking with your doctor. The MOTHERISK website is an excellent resource. (<http://www.motherisk.org>)

Issues for the baby

There is a significant risk of early (medical term: preterm) delivery and having a small baby (medical term: low birth weight). Prematurely born babies are at greater risk for having serious health problems.

If you have a congenital heart defect, the risk of your child having a congenital heart defects varies between 3% and 50%, compared with the background risk of 1% for the general population. This risk will depend on the underlying cardiac lesion of the mother.

Fetal echocardiography is offered to mothers to screen for congenital heart defects in the unborn baby. A fetal echocardiogram is done at approximately 5 months (20 weeks gestation) into the pregnancy.

Medical care during pregnancy and delivery

Where should I be followed?

If you choose to become pregnant or continue your pregnancy, you should be followed at a center that specializes in high-risk pregnancy. Your specialist will determine the frequency of follow-up through your pregnancy.

What can I do and expect during pregnancy?

If you become pregnant, your heart specialist will arrange for frequent check up visits during your pregnancy. In addition to your clinic visits, your doctors will arrange echocardiograms to help determine how the heart is adapting to the pregnancy. You may also be seen by a pulmonary hypertension specialist.

You need to be attentive to symptoms related to your heart. Notify your doctor if you develop any concerning symptoms such as shortness of breath, swelling of the legs or stomach, chest pain, coughing up blood, fast heart beats (palpitations) or fainting. If you develop complications you may be admitted to hospital for closer care and monitoring.

If your symptoms are concerning and you cannot get in touch with your doctor, go to your nearest emergency department. It is helpful to keep a letter from your doctor explaining your condition so that other health care professionals can better help you in an emergency situation.

Many women will be treated with special medications to lower the blood pressure in their lungs.

Some women will need to be admitted to the hospital for monitoring during pregnancy.

Some women will need to have treatment for weakening of the heart muscle (medical term: heart failure).

Labour and delivery must be planned carefully with a team including a heart specialist, a pulmonary hypertension specialist, a high-risk obstetrician and an anesthetist. The risks of pregnancy for a woman with pulmonary hypertension does persist for sometime after delivery, and therefore special care is needed for several weeks after delivery.