



EBSTEIN ANOMALY

What is it?

Ebstein anomaly is an abnormality of the tricuspid valve (a right-sided heart valve). The valve is not located in its usual position (see The Normal Heart), and the abnormal valve often leaks. In some cases, parts of the right side of the heart may also become weak, either enlarged or sometimes too small.

A common related cardiac abnormality is a hole between the filling chambers of the heart (medical term: atrial septal defect). Ebstein anomaly can be associated with other conditions such as narrowing of the pulmonary valve.

Ebstein anomaly can vary in severity. Some adults with a mild form of the disease, will not have any heart problems. Others with more serious forms of the disease will have problems with abnormal heart rhythms (medical term: arrhythmias) or heart failure. Less commonly, adults can develop low oxygen levels (medical term: cyanosis). Some women may require heart surgery because they need to have the abnormal tricuspid valve, repaired or replaced.

How safe is it for me to become pregnant?

Pregnancy does require your heart to work harder and pump more fluid (see Cardiovascular Changes during Pregnancy). The ability of women with Ebstein anomaly to tolerate these changes depends on the function of the heart valves and the size and strength of the heart muscle. Pregnancy is much riskier if you have low oxygen levels. If oxygen levels are really low, there is an increased risk of miscarriage.

Every pregnancy carries some risk for complications and this risk may be increased by heart disease. All women have to consider the safety of a pregnancy with their heart disease. Every person's heart condition is different and therefore the safety of pregnancy differs too. Before trying to have a baby you should discuss your situation with a heart specialist who knows about caring for women with heart disease in pregnancy.

Issues for the mother

Which forms of birth control are safe?

For most women with Ebstein anomaly, the choice of birth control measures (medical term: contraceptives) is usually not limited by their heart condition. However, in women with abnormal heart rhythms (medical term: arrhythmias), holes in the heart, or low oxygen levels, estrogen-containing birth control pills and patches can be a problem because of the risk of blood clots. Contraception should be discussed with a physician who has an understanding of your underlying heart condition. (see Birth Control)

In order to determine your risk during pregnancy, you should see your heart specialist before getting pregnant. You may benefit from additional heart tests such as an echocardiogram, exercise test, or a magnetic resonance imaging scan (MRI scan) to better determine the risks of pregnancy.

Many women with Ebstein anomaly will have a successful pregnancy. Potential heart complications include heart rhythm abnormalities (medical term: arrhythmias) and weakening of the heart muscle (medical term: heart failure). If you have low oxygen levels, you will be at higher risk for complications during pregnancy, including miscarriages. If you had heart failure or rhythm problems before pregnancy, your risk for complications during pregnancy is higher. Other cardiac issues can have an impact on pregnancy outcomes (see General Considerations). It is very important to see a congenital heart specialist before pregnancy to discuss your risk of pregnancy.

Some medications are not safe in pregnancy. Do not stop medications without first checking with your doctor, but do check your medications out before pregnancy so you will have a plan. If you did not do that, then do so as soon as you know you are pregnant. The MOTHERISK website is an excellent resource. (<http://www.motherisk.org>)

Issues for the baby

In women with Ebstein anomaly there is an increased risk of early delivery (medical term: preterm) and of having a small baby (medical term: low birth weight or small for gestational age). Babies born too early are at increased risk for health problems after birth.

In the general population, the risk of having a baby with congenital heart disease is about 1%. If a parent has Ebstein anomaly, the risk increases to 5-10%.

Women will be offered ultrasound screening of the baby's heart (fetal echocardiogram) at the end of the fifth month (20 weeks gestation) of pregnancy. The ultrasound can detect most major cardiac defects in the developing baby. Minor defects may not be detected until after birth.

Medical care during pregnancy and delivery

Where should I be followed?

Depending on the severity of the Ebstein anomaly, you may need to be followed at a center that specializes in high-risk pregnancy. Your specialists will determine how often you need to be seen through your pregnancy.

What can I do and expect during pregnancy?

Your heart specialist will arrange for check up visits during your pregnancy. In addition to your clinic visits, your doctors will arrange ultrasounds of your heart (medical term: echocardiograms) to help determine how your heart is dealing with pregnancy.

It is important that you pay attention to symptoms during your pregnancy. Notify your doctor if you develop any worrying symptoms such as shortness of breath, chest pain, swelling in your legs, or heart palpitations.

If you are concerned about symptoms 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.

Labour and delivery should be planned carefully with a team including a specialist in congenital heart disease, a cardiac anesthetist, and a high-risk obstetrician. A vaginal delivery is usually recommended. Good pain management is important.