Ebstein's Anomaly

1.0 WHAT IS IT?

Ebstein's anomaly is an abnormality of the tricuspid valve (right-sided heart valve). The valve is not located in its usual position (see The Normal Heart), and the abnormal valve often leaks. The right side of the heart, that pumps to the lungs, may also become enlarged and weak. This can lead to impaired ability of the heart to pump blood to the lungs to receive oxygen. Patients with mild disease may not have any heart problems but more serious disease can cause problems with abnormal heart rhythms (medical term: arrhythmias) or ability of the heart to pump strongly enough to meet the body's needs (medical term: heart failure). Rarely this can cause low oxygen levels (medical term: cyanosis).Surgery may be required for treatment of the abnormal tricuspid valve to improve symptoms.

2.0 HOW SAFE IS IT FOR ME TO BECOME PREGNANT?

Pregnancy requires your heart to work harder and pump more fluid (see Cardiovascular Changes during Pregnancy). The ability of women with Ebstein's anomaly to tolerate the changes of pregnancy 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 or a history of heart failure or arrhythmia prior to pregnancy

Planning of pregnancy reduces risk and use of reliable contraception should be discussed with a physician who has an understanding of your underlying heart condition. (see Birth Control) until you are ready to conceive. For patients 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.

3.0 ISSUES FOR THE MOTHER

Prior to Pregnancy: Ideally you should ideally see your heart specialist before getting pregnant to assess your personal risks. You may be advised to have additional heart tests, such as an echocardiogram, exercise test, or a magnetic resonance imaging scan (MRI scan). Your medications will be reviewed, and may be modified.

If you conceive without the benefit of a pre-pregnancy, please notify your doctor as soon as you know you are pregnant. Many tests can still be done and an urgent review of medications can be carried out. The Healthy Pregnancy Hub (www.healthypregnancyhub.ca) is an excellent resource, though medications should not be stopped without first reviewing with your doctor.

Issues During Pregnancy: Patients with Ebstein's anomaly generally have good pregnancy outcomes but do have a 4% risk of arrhythmia and a 3% risk of heart failure developing during pregnancy. This is often in patients with cyanosis, other cardiac issues or who experienced symptoms prior to conceiving. (see General Considerations). Your medical team will monitor you for symptoms and recommend periodic testing to look for early signs. Medications to control or modify symptoms (control heart rate/rhythm, improve heart function, prevent blood clots) can often be started before more serious problems develop.

4.0 ISSSUES FOR THE BABY

If a parent has congenital heart disease, the risk of a baby having a similar heart problem increases from 1% to about 5-10%. Ultrasound screening of the baby's heart (fetal echocardiogram) at 20 weeks of pregnancy can detect most major cardiac defects in the developing baby. Minor defects may not be detected until after birth. There is a 12% risk of poor fetal growth, which can affect pregnancy monitoring and delivery plans. Early delivery occurs in about 22% of pregnancies, which can increase other risks for newborns.

5.0 MEDICAL CARE DURING PREGNANCY AND DELIVERY

Your cardiologist will review your heart function tests and pregnancy risks to determine how often you should been seen by them during pregnancy. Any symptoms (irregular heart rate, shortness of breath etc) should be reported immediately to allow testing and treatment. They will talk with your pregnancy care provider to determine if your local hospital has the resources for care of your pregnancy. Patients at higher risk may need to deliver at specialized sites.

Your pregnancy care provider will monitor your blood pressure and the heart rate and growth of your baby. You may be seen more frequently and will have more ultrasounds to ensure concerns with the growth of the pregnancy is detected and monitored appropriately. There is increased risk of early labour, so you will be monitored for signs, to allow early treatment, if possible.

In labour, both baby and patient should be closely monitored. Vaginal delivery is generally recommended, due to a decreased risk of post-delivery complications, but there is an increased risk of needing Cesarean section, for a variety of reasons. Good pain control, generally with epidural is

recommended to make the labour process less stressful on the heart and to allow the head to descend into the pelvis prior to pushing. Some patients have to limit pushing for cardiac reasons and may be advised that vacuum or forceps delivery may be helpful. Cesarean section can safely be done, if needed for safety of mother or baby.

After delivery, postpartum care needs to include careful monitoring for heart failure and arrhythmia, which can increase at this time. Breastfeeding is safe and does not increase any risks. Your specialists will arrange for follow-up testing of heart function, with timing depending on your symptoms during pregnancy.

6.0 REFERENCES

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