HEART DISEASE & PREGNANCY

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MARFAN SYNDROME

What is it?

Marfan syndrome refers to a disorder that affects multiple parts of the body. The heart is commonly affected. Serious illness or death can occur if the aorta (large blood vessel which supplies the body) (see Normal Heart) becomes very large (medical term: aneurysm), or if the wall of the aorta tears (medical term: dissection).

How safe is it for me to become pregnant?

Pregnancy is associated with increased stress on the heart and the blood vessels. (see Cardiovascular Changes during Pregnancy) The risk of problems during pregnancy in women with Marfan sydrome largely relates to the size of the aorta, the large blood vessel that delivers blood to the body. However, even women with normal size of the aorta can have problems that develop related to pregnancy. Other medical conditions can have an impact on pregnancy outcomes. (see General Considerations)

If your aorta is significantly enlarged or is enlarging quickly, or if you have a past history of aortic tearing (medical term: aortic dissection), then pregnancy is very high risk for you and your unborn baby.

Pregnancy is generally not advised if you have a very large aorta, at least until you have surgery to address this problem. It is important that testing be done to look at the aorta before you become pregnant, as some important imaging tests are not safe in pregnancy. It is important to discuss your specific condition with a physician who knows about the care of women with heart disease in pregnancy. Some women with high-risk cardiac lesions may seek alternatives to pregnancy such as adoption or surrogate motherhood.

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

Issues for the mother

Which forms of birth control are safe?

Because of the high risk of pregnancy associated with a large aorta in women with Marfan syndrome, effective and safe birth control (medical term: contraception) is important. Oral contraceptive pills containing estrogen are best avoided in women with dilated aortas. Many of the progesterone-only forms of contraception are acceptable; however, progesterone-only pills can have high failure rates.

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 am pregnant?

In order to determine your risk during pregnancy, you should see your heart specialist before getting pregnant. You may be required to have additional heart tests such as an ultrasound of the heart (medical term: echocardiogram) or a magnetic resonance imaging scan (MRI scan) of your aorta to better determine the risks of pregnancy.

The most serious risk for women with Marfan syndrome is weakening and tearing of the wall of the aorta (medical term: dissection). The rate of dissection in pregnancy ranges between 1 and 10 per 100 pregnancies, depending on risk profile. You should discuss your risk profile with your doctor before you become pregnant.

Close follow up during pregnancy to monitor blood pressure and appearance of the aorta on echocardiography (generally every 6-8 weeks) is very important. Other cardiac characteristics can have an impact on pregnancy outcomes. (see General Considerations)

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

There is a 1 in 2 chance (50% risk) of having a baby with Marfan syndrome if one of the parents has the condition. Genetic testing can be performed during the first trimester of pregnancy to determine if the fetus is affected. Ultrasound examination of the fetal heart (medical term: fetal echocardiography) can be used to assess the size of the aorta in the fetus and is generally done halfway through pregnancy (20 weeks gestation).

There is a significant risk of early (medical term: preterm) delivery and having a small baby (medical term: low birth weight for gestational age).

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 specialists will determine the frequency of follow up 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 echocardiograms to help determine how your heart is adapting to the pregnancy.

A medication that controls blood pressure and is thought to protect the aorta is used throughout pregnancy (class of medication: beta-blocker).

It is important that you pay attention to symptoms during your pregnancy. Notify your doctor if you develop any worrying symptoms such as chest pain, back pain, dizziness 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.

Labour and delivery should be planned carefully with a team including a specialist in congenital heart disease, an anesthetist, and a high-risk obstetrician. Women with significantly enlarged aortas tend to be delivered by cesarean section.