



## FONTAN OPERATION

### What is it?

The Fontan operation is an operation performed to help repair certain kinds of complex congenital heart disease. Unlike in the normal heart, which has two pumping chambers (medical term: ventricle), people with a Fontan operation only have one pumping chamber. (see The Normal Heart) There are several types of Fontan operation.

Heart problems can develop after a Fontan operation including:

1. Low exercise ability
2. Weakened heart muscle (medical term: heart failure)
3. Heart rhythm disturbances (medical term: arrhythmias)
4. Blood clot formation
5. Low oxygen levels (medical term: cyanosis)

### 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 Fontan operations to tolerate these changes depends on their specific type of Fontan operation, the strength of their heart muscle, their heart valve function, their oxygen level and if they have had arrhythmias or blood clots in the past.

Women with the Fontan operation are at increased risk of problems during pregnancy. For some women, this unfortunately means that pregnancy is very high risk and should be avoided. For other women, with careful monitoring before and during pregnancy, some problems can be avoided, but pregnancy is not risk-free.

Every pregnancy carries the risk for complications and this risk is increased because of your heart disease. Thus all women have to consider the safety of a pregnancy with their heart disease. Every woman is different and, as a consequence, so is the safety of pregnancy. Therefore, before trying to have a baby you must discuss your condition with a heart specialist who has expertise in caring for patients with heart disease in pregnancy.

### Issues for the mother

#### Which forms of birth control are safe?

For those women with the Fontan operation where pregnancy is very high risk and not advised, safe and effective birth control (medical term: contraceptives) is essential. Combined estrogen and progestin oral contraceptive pills (standard birth control pills) are not advised because they are associated with increased blood clot formation. Progesterone-only forms of contraception do not increase the risk of blood clots, but when used as pills ("mini-pills"), they are not reliable enough in preventing pregnancy. Some women at very high risk for pregnancy complications 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?**

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 echocardiogram, exercise test, or a magnetic resonance imaging scan (MRI scan) to better determine the risks of pregnancy.

Some women with the Fontan operation can have a successful pregnancy; however, there are a number of potential risks. The most common heart-related complications that occur during pregnancy are heart rhythm abnormalities (medical term: arrhythmias) or weakening of the heart muscle (medical term: heart failure).

Women with the Fontan operation who have good heart function and no history of heart rhythm problems have the best chance of a successful pregnancy. When the heart is weakened, when there are rhythm problems or when the oxygen levels are low, the risk is much higher and sometimes pregnancy is not advised. Other cardiac characteristics 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**

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

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

Women will be offered ultrasound screening of the baby's heart (medical term: fetal echocardiogram) at the end of the fifth month (20 weeks gestation) of pregnancy. The ultrasound will 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?**

Once pregnant, 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 likely 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 symptoms such as shortness of breath, chest pain, swelling of the legs, or heart palpitations.

If your symptoms are worrying 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.