



FONTAN OPERATION

Background

The Fontan operation is a palliative procedure performed in patients with a functionally or anatomically single ventricle that is not suitable for a biventricular repair.

The venous blood is diverted directly to the pulmonary arteries usually either via the right atrium (direct right atrium-pulmonary artery Fontan) or via a tunnel (either a lateral tunnel Fontan or extracardiac conduit). There is generally no subpulmonic ventricle to provide pulsatile blood to the pulmonary circulation. Pulmonary blood flow is thus dependent on changes in intrathoracic pressure and passive flow from the systemic veins to the pulmonary bed.

Patients with the Fontan operation are at risk of various complications including:

1. Arrhythmias – re-entrant atrial arrhythmias are particularly problematic
2. Heart failure
3. Pulmonary and systemic thromboembolism.
4. Hypoxia from intracardiac and intrapulmonary communications
5. Protein-losing enteropathy

Effects of pregnancy-related hemodynamic changes

Pregnancy is associated with hemodynamic changes including an increase in blood volume and cardiac output. (see Hemodynamic Changes in Pregnancy) The ability of a woman with a Fontan operation to tolerate these changes depends on ventricular systolic and diastolic function, the degree of atrioventricular regurgitation and presence and frequency of arrhythmias. Worsening of heart failure and arrhythmias are potential complications.

Patients with the Fontan operation are sensitive to changes in preload and afterload. An increase in volume can result in volume overload in women with systemic ventricular dysfunction. A decrease in preload can result in impaired flow through the Fontan circulation (systemic venous return to pulmonary arteries) subsequently resulting in reduced cardiac output. Increased intra-abdominal pressure during labor and peripheral vasodilatation, as a result of OR agents or anesthetics, can reduce preload at the time of labor leading to problems of low cardiac output.

Pregnancy is a hypercoagulable state. In women who have had a Fontan operation (particularly those with direct right atrial to pulmonary artery connections), there is already significant stasis of blood flow within the Fontan circulation, placing these women at an increased risk of thromboembolism.

Maternal cardiac complications

Women who have had a Fontan operation are at significant risk of complications including atrial arrhythmias (16%), deterioration in ventricular function, heart failure (4%), worsening atrioventricular valvular regurgitation and thromboembolism during pregnancy. (1,2,3).

Pregnancy risks are dependent on functional status, degree of cyanosis, ventricular function (systolic and diastolic), atrioventricular valvular regurgitation, and presence of atrial arrhythmias. Even in woman with well-functioning Fontan circulations (good functional status and ventricular function, and absence of cyanosis or atrial arrhythmias) the risk of an adverse cardiac complication during pregnancy remains high. Other cardiac characteristics can also have an impact on outcomes. (see General Considerations)

The late effects of pregnancy on the heart in women with Fontan physiology are not known.

Fetal complications

Fetal complications are common in women with the Fontan operation, occurring in just under half of pregnancies. This includes miscarriages, prematurity (28%) and small for gestational age birth weight (12%). (1) Perinatal mortality has been reported.

Management strategies

Preconception counseling/Contraceptive methods

Ideally, a comprehensive cardiovascular examination should be undertaken before embarking on pregnancy. This includes a careful history and physical examination, an electrocardiogram, an echocardiogram, and consideration of cardiopulmonary exercise testing and cardiac magnetic resonance imaging.

Women with significant ventricular dysfunction, poor functional status (NYHA III or IV) or recurrent atrial arrhythmia are at high risk of adverse cardiac events in pregnancy. Based on these variables, some women should be advised against pregnancy.

Some women who are fully informed and understand the maternal and fetal risk and complications may still choose to become pregnant. The importance of close monitoring with admission as needed to a hospital equipped with a multidisciplinary team experienced in the management of cardiac disease in pregnancy should be conveyed to the patient.

A discussion with caregivers about contraceptive methods is imperative. Contraceptive pills containing estrogen (combined contraceptive pills) are contraindicated due to their increased risk of thromboembolism. Progesterone-only formulations such as depot injections and subdermal implants (Implanon®) are a reasonable option. Progesterone-only pills are not optimal because of unacceptably low efficacy rates. The insertion of intra-uterine contraceptive devices can be associated with vasovagal reactions, which can reduce preload resulting in poor cardiac output. If this method of contraception is selected, insertion should only be done by a gynecologist with an understanding of the Fontan condition. Some women will consider sterilization due to the high-risk nature of a pregnancy. However, such a decision may have a major psychological impact and needs to be fully discussed with the appropriate caregivers. Moreover, the laparoscopic procedure carries risk in this population, as it requires insufflation of the abdomen with carbon dioxide, intermittent head down tilt and positive pressure ventilation, all of which reduce cardiac output and may be poorly tolerated. If sterilization is chosen, it is strongly recommend that it be performed at a centre with experience in the care of women with complex heart disease. Essure® is a new sterilization technique, involving the insertion of stents hysteroscopically into the Fallopian tubes using sedation and local anesthesia. Early studies suggest it may be safe and has a low failure rate (see Contraception).

Transmission of congenital heart disease to offspring should be discussed with women with the Fontan operation. The risk of the fetus having structural cardiac defects varies between 5-10%, compared with the background risk of 1% for the general population

As certain anticoagulants and cardiac medications are contraindicated in pregnancy, medication use should be reviewed if a woman is contemplating pregnancy or is pregnant. The MOTHERISK website (<http://www.motherisk.org>) is an excellent resource.

Ante-partum care

If a woman with a Fontan operation becomes pregnant, coordinated care should be established early, involving a congenital heart disease specialist, high-risk obstetrician and an obstetrical anesthetist.

Close cardiovascular monitoring, with specific attention to volume status, is essential throughout pregnancy and the peripartum period. Serial echocardiograms are important to assess cardiac function.

These women are also vulnerable to thromboembolism. Thromboprophylaxis is essential at any time that the woman is relatively immobile. In women on Coumadin (warfarin), there is a risk of teratogenicity between 6-12 weeks of gestation. An appropriate anticoagulation plan should be devised with a hematologist/thrombosis expert when required.

Occurrence of a sustained atrial tachyarrhythmia can result in poor cardiac output and an increased risk of thrombus in the Fontan circulation. Prompt anticoagulation and conversion to sinus rhythm after exclusion of intracardiac thrombi is vital (see Arrhythmias).

Fetal echocardiography should be offered to the expectant mother to screen for congenital heart defects. A fetal echocardiogram is done at approximately 20 weeks gestation.

Labour and delivery

Labour and delivery should be planned carefully with a multidisciplinary team well in advance. It is important to communicate the delivery plan to the woman and to other physicians involved in her care. The best delivery plan is not useful if information is not readily available when needed.

Successful vaginal and cesarean deliveries have been reported. If vaginal delivery is chosen, good pain management is very important. During labour Valsalva maneuver should be avoided. To decrease maternal expulsive efforts during the second stage of labour, forceps or vacuum delivery is often utilized. To decrease potential harmful complications from difficult mid cavity-assisted delivery, uterine contractions are often utilized to facilitate the initial descent of the presenting part.

Various factors can result in reduced preload, poor cardiac output and hypotension, including blood loss, use of anesthetic agents, oxytocic drugs, and inferior vena caval compression. Ideally, women should lie on their side to avoid inferior vena cava compression in the supine position resulting in reduced venous return. Where oxytocic agents are necessary, bolus doses should be avoided.

The need for maternal monitoring is dictated by the functional status of the woman, her ventricular function, and propensity to arrhythmias. Maternal monitoring will often include telemetry and pulse oximetry. Some women may need invasive blood pressure monitoring. Central venous pressure monitoring is not indicated.

Compression stockings or thromboguards are very important around the time of delivery, along with early ambulation.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women with Fontan operations. However, some experts continue to administer antibiotics because they feel that the risks of adverse reactions to antibiotics are small and the risk of developing endocarditis has major health consequences.

For women with residual intracardiac shunts (i.e. fenestrations or leaks between the pulmonary Fontan circuit and the systemic circulation), air-particulate filters (bubble trap filters) are recommended for all intravenous lines at the time of labour and delivery.

Post-partum care

The hemodynamic changes of pregnancy may take up to six months to normalize. Women should be seen early after pregnancy (usually within 6-8 weeks). The frequency of additional follow up visits should be dictated by the clinical status of the women.

References:

1. Drenthen W, Pieper PG, Roos-Hesselink JW, et al. Outcome of pregnancy in women with congenital heart disease: a literature review. *J Am Coll Cardiol.* 2007;49:2303–2311.
2. Canobbio MM, Mair DD, van der Velde M, Koos BJ. Pregnancy outcomes after the Fontan repair. *J Am Coll Cardiol* 1996;28: 763-7.
3. Drenthen W, Pieper PG, Roos-Hesselink JW, van Lottum WA, Voors AA, Mulder BJ, van Dijk AP, Vliegen HW, Sollie KM, Moons P, Ebels T, van Veldhuisen DJ; ZAHARA Investigators. Pregnancy and delivery in women after Fontan palliation. *Heart.* 2006;92(9):1290-4.