Anesthetic considerations for the pregnant patient with Fontan circulation
(Had Fontan Surgery, Now Pregnant)

Victor C. Baum, MD
Professor, Anesthesiology
&
Duncan G. de Souza, MD, FRCPC
Assistant Professor, Anesthesiology

The University of Virginia Health System
Charlottesville, Virginia

Goals
1. Understand the physiologic consequences of Fontan circulation
2. Understand the long term complications of patients with Fontan circulation
3. Understand how the Fontan operation has evolved in an attempt to reduce long term complications.
4. Understand how the physiologic changes of pregnancy affect Fontan circulation.
5. Understand the predictors we can use to determine how a patient will tolerate pregnancy, labor and delivery.
6. Understand the risks and benefits of the different anesthetic options for labor, vaginal delivery and Cesarean delivery.

Case
A 26 year old primigravida presented at 36 weeks gestation with spontaneous rupture of membranes. She was in not labor. The baby was in breech presentation. Emergency Cesarean section was scheduled.

She had congenital heart disease with a diagnosis of tricuspid and pulmonary atresia. She was palliated with a Blalock-Taussig shunt in the newborn period followed by an atrio-pulmonary Fontan at age 4. There was good functional status prior to pregnancy and the patient tolerated pregnancy well until 33 weeks when she developed atrial flutter with rapid ventricular rate. She spontaneously converted to sinus rhythm and since then remained free from arrhythmia. Her only medication was metoprolol 25 mg bid. The patient reported an increase in fatigue without dyspnea in the last few weeks of her pregnancy. She is still able to complete normal activities without limitation.

Cardiologist has suggested general anesthesia might be preferable in case it is necessary to cardiovert intraoperatively. What further information do you need? How will you proceed?
Questions for discussion

1. What is the Fontan operation? At what age is the operation performed?
2. What is the physiologic consequence of Fontan circulation? What is meant by trans-pulmonary gradient and what is its significance after the Fontan operation? Why is the atrium included in the Fontan pathway?
3. What are the three major long-term complications after atrio-pulmonary Fontan?
4. Why was the atrio-pulmonary Fontan abandoned?
5. What is the expected functional state of a patient with good Fontan physiology?
6. What are the cardiovascular changes of pregnancy? When do these changes occur? How do these changes affect a patient with Fontan physiology?
7. What do we know about pregnancy and outcomes in Fontan patients?
8. Can Fontan patients tolerate a trial of labor for vaginal delivery? How should analgesia be provided for labor? What are the benefits of epidural analgesia during stage I of labor? What problems might occur with bearing down and pushing during stage II of labor? What problems might occur immediately after delivery?
9. What are the pros and cons of the different anesthetic options for Cesarean delivery? Is invasive monitoring needed? What is the significance of the patient’s history of atrial flutter?
10. How will you manage post partum hemorrhage? How will you assess volume status? Is the choice of uterotonic drugs affected by the patient’s congenital heart disease?

Discussion

In 1971, Dr. Fontan published a series of three patients who underwent the pioneering operation that now bears his name. The patients all had single ventricle physiology with an intact left ventricle and an absent or rudimentary right ventricle secondary to tricuspid atresia. Conceptually brilliant, Fontan thought that the right atrium could be connected to the pulmonary artery to separate the systemic and pulmonary circulations. This would functionally create a circulation in series similar to that of patients with normal cardiac anatomy. The only difference is that the driving force for pulmonary blood flow would be the right atrium and central venous pressures. In the original operation by Fontan the right and left pulmonary arteries were divided. The superior vena cava was anastomosed to the right pulmonary artery and the right atrium anastomosed to the left pulmonary artery. (Figure 1) This is the classic Fontan. The modification by Kreutzer in 1973 kept the pulmonary arteries in continuity and connected the right atrial appendage to the main PA. This is known as the atrio-pulmonary Fontan. (Figure 2) The atrio-pulmonary Fontan, sometimes mistakenly called a classic Fontan, was done exclusively until the late 1980s when newer variations were conceived. From the beginning there were serious concerns about the viability of this unique circulation. Thus, eligibility criteria as listed below, were very strict.

Between 4 to 15 years of age          Mean PA pressure < 15 mmHg
Normal sinus rhythm                 PVR < 4 Woods
Normal systemic venous return       PA diameter at least 75% of aortic diameter
Right atrium of normal volume       Normal ventricular function
No atrio-ventricular valve regurgitation
Blood can only flow between any two points when a pressure gradients exists between those same two points. The pressure that creates pulmonary blood flow in Fontan circulation is known as the trans-pulmonary gradient. It is simply the pressure in the Fontan circuit (SVC, IVC, right atrium) minus the pressure in the left atrium. The difference between the two represents the pulmonary vascular resistance (PVR) that must be overcome. These scenarios illustrate the concept.
Scenario 1: Fontan pressure 15, LA pressure 10, trans-pulmonary gradient 5
Scenario 2: Fontan pressure 20, LA pressure 15, trans-pulmonary gradient 5
Scenario 2: Fontan pressure 20, LA pressure 10, trans-pulmonary gradient 10

Scenario 1: Normal Fontan and LA pressures and normal PVR.
Scenario 2: Elevated Fontan and LA pressures. There is ventricular dysfunction or valvular disease but PVR remains normal
Scenario 3: Elevated Fontan pressures but normal LA pressures. The elevated Fontan pressures are caused by raised PVR.

This fundamental relationship underlies the success or failure of Fontan circulation. Purely passive central venous pressure was thought inadequate to provide pulmonary blood flow and therefore the right atrium was incorporated in the original Fontan operation. It was hoped that central venous pressure with the assistance of atrial contraction would successfully provide pulmonary blood flow.

It soon became apparent that Fontan circulation could lead to three major complications: atrial arrhythmia, thrombosis and protein losing enteropathy (PLE). The right atrium over time became dilated and lost most of its contractile function. Atrial dilation from any cause is a known precipitant of arrhythmia. Atrial arrhythmia with a rapid ventricular response leads to acute decompensation. Given that the right atrium is significantly dilated, attempts to restore sinus rhythm were usually not successful. Controlling ventricular response was the only therapy. However, the persisting atrial arrhythmia led to long term deterioration in function of the systemic ventricle which is very poorly tolerated in Fontan circulation. It must be noted here that independent of atrial arrhythmia, in some patients with Fontan circulation the systemic ventricle would fail through a combination of ventricular dysfunction, valvular disease or both. There are few therapeutic options for decreased ventricular function in the setting of Fontan circulation. The dilated right atrium with stagnant blood flow provided a ready source for thrombosis. With the right atrium directly connected to the pulmonary circulation, thrombosis easily led to pulmonary embolism with potential significant elevations in Fontan pressures. The etiology of thrombosis is primarily stasis but by poorly understood mechanisms, Fontan patients are believed to exist in a mildly hypercoagulable state. Appropriate thrombo-prophylaxis is controversial. Most patients are maintained on aspirin with warfarin reserved for those with proven thrombosis. The last major complication is PLE, which remains as confusing as it is serious. The purely mechanical explanation is that deteriorating Fontan circulation with elevated central venous pressures leads to portal hypertension and thus, a protein losing enteropathy. This explanation is insufficient. The correlation between central venous pressures and PLE is poor. Some patients with PLE have relatively normal central venous pressures while other patients with very elevated central venous pressures do not have PLE. Unless PLE is due to an anatomic obstruction or stenosis in the Fontan pathway that can be corrected, it responds poorly to all medical therapy and portends a very poor prognosis. Some combination of atrial arrhythmia, thrombosis, PLE or low cardiac output state has been termed the “failing Fontan”.

The Fontan operation has continued to evolve. After the first fifteen to twenty years experience, it became clear that incorporation of the right atrium provided no assistance to pulmonary blood flow and in fact was directly related to the major Fontan complications. Its presence as a dilated chamber led directly to thrombosis. The creation of the atrio-pulmonary anastomosis involved extensive atrial
suture lines that provided a focus for arrhythmia. Clearly, the atrium could not be included in the Fontan circuit. In the late 1980s to early 1990s the atrio-pulmonary Fontan was abandoned for a total cavo-pulmonary connection. The cavo-pulmonary connection exists in two forms. (Figure 3). The lateral tunnel form creates a channel within the right atrium. A dilated atrium cannot occur but the surgery still involves atrial suture lines. The extra-cardiac connection takes the desire to avoid atrial suture lines even further by creating an “extra-cardiac” channel. Both of these versions are a clear improvement from the original atrio-pulmonary connection. The choice of lateral tunnel or extra-cardiac Fontan is usually institutional preference reflecting the fact that neither has yet shown a clear advantage over the other.

**Figure 3** Modern Fontan operation

Despite the known long term problems with Fontan circulation, it has allowed patients to survive well into adulthood and consider starting families of their own. This is a truly remarkable achievement. When a patient with Fontan circulation of any type considers pregnancy, the best predictor of a successful pregnancy is her pre-pregnant functional state. This would seem to portend success because in the absence of the three major complications, most Fontan patients report few if any restrictions of functional activity. This is deceptive however, because even well functioning Fontan patients approach pregnancy with significant limitations of cardiac reserve. The decreased cardiac reserve is often not appreciated by the patients themselves because they have lived with it for so long. They function well with their normal daily activities but rarely if ever, attempt anything strenuous. Exercise testing has
shown significant reductions in maximal oxygen consumption and cardiac output when compared to normal controls. The cardiovascular changes of pregnancy begin early in gestation and plateau by late second trimester. The changes then remain stable until the onset of labor. During labor there are further increased cardiac demands. (Figure 4) Fontan patients are at increased risk of thrombosis due to their own hypercoagulable state and that superimposed by pregnancy.

**Figure 4** Cardiac output during pregnancy, labor, and the puerperium. Values during pregnancy are measured at the end of the first, second, and third trimesters. Values during labor are measured between contractions. For each measurement, the relative contributions of heart rate (HR) and stroke volume (SV) to the change in cardiac output are illustrated.


The key question is how the patient with Fontan circulation will cope with the increased cardiac work. Those with evidence of a failing Fontan circulation will clearly further deteriorate during pregnancy. Within the larger group of patients with per-existing good functional status, there are those that will cope well and those that will not. Our ability to predict which ones will deteriorate is limited. Those with an atrio-pulmonary Fontan are probably at higher risk of developing arrhythmia as the atrium dilates to accommodate the increased blood volume of pregnancy. Pregnancy itself is the “stress test” and we only know in retrospect which patients will pass and which will fail. The literature is also colored by selection bias – only some are allowed to take the test. Women with moderate to severe physiologic limitations are likely counseled to avoid pregnancy or obtain early termination of pregnancy, limiting most pregnancies to those with expected better results.

Our knowledge of pregnancy in Fontan patients suffers from the lack of a comprehensive database to track outcomes. Most reporting is of the anecdotal or case report variety. The questions to be answered are the following:
Is there reduced fertility?
What is the occurrence of congenital heart disease in the offspring?
What is the rate of spontaneous abortion or miscarriage?
What is the rate of prematurity?
How can we predict if a Fontan patient can successfully carry a pregnancy to term without a deterioration functional status?
Is there a significantly increased of thrombosis? Should thrombo-prophylaxis be started specifically for pregnancy?
What is the ideal timing and mode of delivery?

The first reported case of a successful pregnancy in a patient with Fontan circulation was published in 1989. Since then we have only further case reports and two retrospective studies. There have been two series of pregnancy outcomes in women with Fontan physiology, both relatively small and suffering from the limitations mentioned above. Drenthen et al. identified 38 women of child bearing age from two Fontan registries. There were ten pregnancies from six women, resulting in four live births, five miscarriages and one ectopic pregnancy. The live birth pregnancies were complicated by functional deterioration, atrial arrhythmias, prematurity and intrauterine growth retardation. The paper ended with the controversial statement that “pregnancy is not advisable”. Ten years earlier, however, Canobbio came to a different conclusion. In the largest review to date; they described 33 pregnancies from 21 women. The outcomes were 15 live births, 13 miscarriages and 5 elective abortions. Functional status remained good throughout the pregnancies in all but one case. There was no significant risk of prematurity and no increased risk of congenital heart disease in the infants. The authors concluded that “the tendency to routinely discourage pregnancy may need to be reconsidered.” With time, more women will presumably have had the modern Fontan repair (lateral tunnel or extra-cardiac), with hopefully better physiologic results.

A review of the literature suggests that Fontan patients with good pre-existing function can successfully complete a pregnancy but have increased risk of spontaneous abortion, prematurity and deterioration of NYHA class. Pre-pregnancy thrombo-prophylaxis, if any, should be continued but starting therapy specifically because of pregnancy is not warranted. The timing and mode of delivery, in absence of cardiac complications, is guided by obstetric indications. Patients with known major complications should probably not become pregnant but there are too few cases to state definitively. It should be noted the above mentioned studies have small numbers and are published by cardiologists with obviously no mention of the anesthetic concerns surrounding labor and delivery. The anesthetic literature is entirely comprised of case reports data accompanied by some review articles. We believe the Fontan patient should be seen early in pregnancy and followed in a high risk obstetric clinic. Consultation with an anesthesiologist knowledgeable in adult congenital heart disease is required if possible. The assessment consists of a history and physical in the usual manner with specific attention to functional state and the presence of any of the known major complications of Fontan circulation. The type of Fontan operation must be noted. If not done recently, an echocardiogram should be ordered and once again, performed by cardiologist familiar with adult congenital heart disease. Follow up appointments and testing is determined by how the pregnancy is being tolerated.

By the time of parturition, the stresses of the hemodynamic alterations of pregnancy have usually been successfully faced. The patient now has passed the “stress test” of pregnancy. There is no evidence that operative delivery is inherently safer than vaginal delivery, thus the timing and mode of delivery are...
dictated by obstetric indications. A review of the limited case reports in the anesthesia literature shows
that epidural analgesia is well tolerated and indeed recommended for the first stage of labor. Effective
analgesia will help to smooth out large fluctuations in pulmonary blood flow secondary to the
tachypnea associated with contractions. Although the decreases in pulmonary blood flow caused by
bearing down and pushing in the second stage of labor might decrease cardiac output, perioperative
complications are low and peripartum cardiac decompensation is rare. The caesarian section rate seems
to be higher than the general population. Neuraxial anesthesia for caesarian section, in addition to its
usual benefits, preserves spontaneous ventilation, which is desirable in Fontan patients. The gradual
onset of epidural anesthesia is preferable to the rapid sympathectomy and resulting fall in preload from
spinal anesthesia. No increased risk from general anesthesia has been identified. There is no absolute
indication for additional physiologic monitoring. Certainly in patients with any hemodynamic
compromise a peripartum arterial catheter is reasonable. Oxytocin will decrease systemic vascular
resistance and increase heart rate and pulmonary vascular resistance, and methylergonovine
(Methergine®) will increase systemic vascular resistance, thus close observation is required during this
immediate postpartum period. Prevention of uterine atony is very important because post partum
hemorrhage is poorly tolerated. Patients with Fontan physiology are preload dependent with no
pulmonary ventricle to maintain pulmonary blood flow in the presence of hypovolemia. The
physiologic reserve is compromised, making the maintenance of intravascular volume the key to
managing post partum hemorrhage.
References


