

FONTAN CIRCULATION AND PREGNANCY: IS IT POSSIBLE

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ABSTRACT

More than 85% of congenital heart disease in children survive to adulthood according to recent advances in pediatric cardiology and heart surgery practice. Fifty percent of this population is made up of women, most of whom have reached child bearing age. In this group of patients, pregnancy represents a new challenge, whose natural history has been modified by surgery. In Fontan circulation, the ventricle unable to compensate for the major restriction in the bottleneck and non physiological suction have to pull blood through the lungs, although the ventricles provide the driving force for the circuit. Women with an adequate Fontan circulation can become pregnant without clinically significant long term sequelae. Pre-marital and pre-pregnancy consultation is important. A close collaboration between cardiologist, obstetrician and anaesthesiologist is mandatory in order to get the best outcome of pregnancy.

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INTRODUCTION

The Fontan procedure bypasses the normal systemic blood flow to the heart and is performed on people with a single ventricle.¹ In patient that unable to sustain biventricular anatomy and cannot be offered a 2 ventricle repair, this procedure is the goal surgery treatment in congenital heart disease.² Unlike in biventricular circulation, in Fontan Procedure, blood flow into the pulmonary arteries is passive with no intervening pump.^{3,4}

Recent advances in pediatric cardiology and heart surgery have allowed more than 85% of children with congenital heart disease to survive to adulthood. Fifty percent of this population is made up of women that have reached child bearing age. Pregnancy represents a new challenge for CHD patients especially in woman with history of Fontan procedure.⁵ Risk of maternal morbidity and mortality is increased significantly in pregnancy women with a fontan circulation. According to Guidelines of the European Society of Cardiology (ESC) in 2011, pregnancy woman with Fontan circulation were classified as having moderate to high risk of morbidity and maternal mortality (World Health Organization class III – IV).⁶

Due to high maternal and fetal risks, woman with Fontan circulation who reach adolescent are counseled against pregnancy. Complications can happened to both maternal and fetal. Maternal complications in Fontan circulation consist of arrhythmias, cyanosis, heart failure and impaired exercise capacity. On the other side, fetal complications consist of intrauterine growth retardation (IUGR), low birth weight and preterm birth. Even there are such complications, there is no report of maternal mortality in pregnancy women with history of a Fontan procedure in the current literature.⁷

FONTAN CIRCULATION AND PREGNANCY

There are two circulations in cardiovascular system consist of, pulmonary circulation and systemic circulation, serially connected and controlled by two pumps (right and left ventricle). Systemic venous return is connected to the pulmonary artery without passing the ventricle are found in Fontan circulation.⁸ In Fontan circulation, a neo portal system is made result in one network of small blood vessels into another vessels network without receiving energy from a pump or ventricle results in critical bottle neck. The pressure above and below

the bottleneck and bottleneck's resistance determine the output through this critical bottleneck. Those 3 features also become factor of physiologic impairment in Fontan circulation⁸. In Fontan circulation, the ventricle unable to compensate for the major restriction in the bottleneck and non physiological suction have to pull blood

through the lungs, although the ventricles provide the driving force for the circuit. Reducing of right ventricle role by the Fontan happened, the ventricle is not controlling the cardiac output nor can decrease the extent of congestion in the systemic veins.⁸

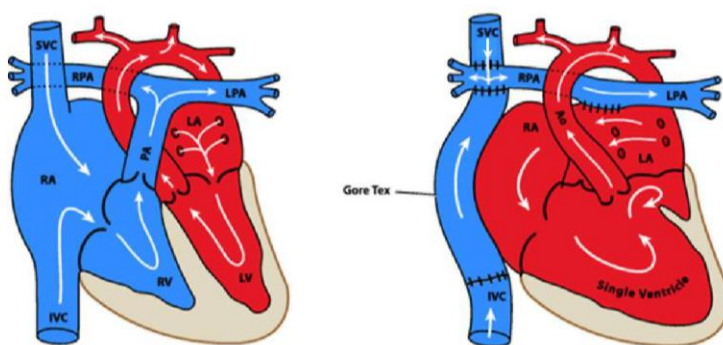


Figure 1. Schematic picture of normal heart (right) and fontan circulation (left)

It is impossible to create a Fontan circulation at birth. In this period, for over several weeks pulmonary vascular resistance continues to increase and the vessels- caval veins and pulmonary arteries are too small to preclude cavo pulmonary shunt.⁹ In the neonatal period, flow from the heart to the aorta must be unrestricted followed by well balanced limited flow to the lungs and limited return of blood from atrium to the ventricle. To develop the pulmonary vasculature, overload of volume in the heart is recommended.⁹ At the age of 4-12 months,

bidirectional cavo-pulmonary shunt can be done. At 1-5 years of age, the inferior caval vein is connected to the pulmonary artery to complete the Fontan circuit.⁹ It depends on growth of vascular structures and symptoms consist of cyanosis at rest and during exercise. There are different types of Fontan. The most common forms of the Fontan procedures are atriopulmonary connection and the cavopulmonary connection. In the atriopulmonary Fontan, the right atrial appendage is directly anastomosed to the main pulmonary artery, providing a pathway

for blood from the inferior and superior venae cavae to get the pulmonary circulation. The atrial septum is left intact, no shunting between the right and left atrium. In the very first Fontan, artificial valves were placed in the circuit. This was quickly abandoned due to high compliance, associated dilation of the right atrium, and calcific obstruction of the valves.¹⁰ Prolonged pleural effusion is common in post Fontan procedure, currently surgeons performed a fenestration hole diameter of 4-5 mm to connect the Fontan circuit and right atrium to reduce this risk.

There are only small numbers of reports in fontan women who have gone through pregnancy. The interactions between pregnancy and fontan hemodynamics plays an important role. An increase in plasma volume found in normal pregnancy followed by cardiac output that increase up to mid-pregnancy as does circulating volume and heart rate.² At 6-12 weeks of gestation, plasma volume increases by 10-15%, progress rapidly until 30-34 weeks and remain stable thereafter until delivery. To accommodate these changes, right and left ventricular increasing their volume. Cardiac output first increasing at 5 weeks and then reaching the peak at 32 weeks gestation. In early pregnancy, 15-30% increase in the heart rate begins and persist throughout. Stroke

volume increases parallelly with plasma volume changes and reaches the peak at 36 weeks of gestation. Decrement in systemic vascular resistance followed by blood pressure alleviation, with a nadir at the end of the second trimester can be caused by hormonal factors and the development of the maternal-placental circulation. Pulmonary vascular resistance decreases, with no change in pulmonary artery pressure.¹¹ The key to successful pregnancy in woman with fontan circulation depends on the ability of the right atrium and functioning single ventricle to generate and tolerate the normal cardiovascular adjustment to pregnancy.²

Due to the risk for recurrence of CHD, all women of childbearing age with CHD especially with fontan circulation should receive pregnancy counseling by an expert. Counseling should be initially done at or soon after sexual maturity in the patient's teen years. For the woman with complex congenital heart disease, preconception planning is imperative. The woman and her partner should be fully aware of the potential risk of pregnancy for her and her offspring.¹⁴ Pregnancy woman with Fontan circulation were classified as having moderate to high risk of morbidity and maternal mortality (World Health Organization class III – IV) according to 2011 guidelines of the European

Society of Cardiology. There are risk score exist to evaluate maternal outcomes such as CARPREG score and ZAHARA score.

Cardiovascular complication during pregnancy included arrhythmia (such as supraventricular tachycardia), systemic venous tromboembolism (VTE), and heart Failure.¹⁷ Pregnancy intensifies the risk of VTE and increased during postpartum period. Pathophysiology mechanisms involve caval compression leads to venous stasis, endothelial injury during delivery, progressive increases in coagulation factors, a reduction in protein S and increasing resistance to activated protein C. Because of that, pregnant women with a Fontan circulation is recommended for prophylactic anticoagulation during pregnancy.¹¹ There is a high incidence of pregnancy complications such as misbirth, prematurity, and IUGR caused by limited capacity to respond to the increase in cardiac output required by pregnancy in Fontan circulation. One-third of pregnancies miscarriage complication happened in first-trimester. In pregnancy, mother may experienced symptoms of heart failure due to physiological changes influenced functional capacity, especially the right ventricle. During pregnancy, when gravid uterus compressed the inferior vena cava, maternal bradycardia and hypotension

can happened. Maternal bradycardia and hypotension can be worsen in the supine position.¹¹ Intrauterine fetal growth retardation was detected in this patient, therefore termination was done before the baby was aterm.

The risks and benefits of vaginal versus caesarean birth should be considered and planned carefully. Consideration of center facilities to deliver emergency cardiac, obstetric, anaesthetic, and neonatal care are important.¹² ESC guidelines 2011 prefer to vaginal delivery as a first choice. An early caesarean should be planned to avoid the development or worsening of heart failure, if ventricular function deteriorates.⁶ Some disadvantages of vaginal birth include the unpredictability of the progress of labour, the possibility of an emergency or immediate c-section being required, reduced likelihood of success if preterm.¹⁰ Provision anaesthesia for pregnant patients with a Fontan circulation follows the general principles such as PVR increasement prevention, ventricular function maintenance, and hypovolaemia prevention. Due to high risk of thromboembolic complication, fontan circulation in pregnancy woman will be fully anticoagulates.¹²

CONCLUSION

Women with an adequate Fontan circulation can become pregnant without clinically significant long term sequelae. Patient should have an adequate Fontan circulation, therefore could have uneventful pregnancy and delivery. Pre-marital and pre-conception counselling is mandatory, so that the patient and her husband know the risk fetal and maternal risk, and follow the cardiologist and obstetrician advice, included close monitoring at the third trimester.

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