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Two successful pregnancies in a patient with Fontan-repaired dextrocardiac univentricular heart

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Abstract

Univentricular dextrocardia is a rare complex congenital cyanotic heart disease. With the improvement of medical and surgical care, a greater number of women with complex congenital heart diseases have survived up to the childbearing age. Pregnancy with univentricular dextrocardia and Fontan circulation increases maternal and fetal mortality and morbidity. We report a case of successful management of two pregnancies in a woman with Fontan repaired dextrocardiac univentricular heart.

Key words: univentricular heart, cyanotic congenital heart disease, dextrocardia, Fontan circulation, pregnancy.

Introduction

In the univentricular heart, both atria empty into a single ventricular chamber with total mixing of systemic and pulmonary circulation. Congenital heart disease occurs in approximately 0.8% of live births, out of which 1-2% is contributed by univentricular heart disease.[1]

The Fontan procedure is used to repair the single ventricle by directing deoxygenated systemic venous flow to the pulmonary arterial circulation. Physiological changes that normally occur in pregnancy, i.e. increased cardiac output, increased heart rate, hypercoagulability state, increased intravascular volume and decreased systemic vascular resistance can adversely affect the cardiac stability of these patients leading to maternal and fetal complications.[2] We present a case of a woman with a dextrocardiac univentricular heart with Fontan conduit who had undergone two childbirths successfully with multi-disciplinary team management.

Case report

A 22-year-old second gravida woman, who had a history of dextrocardiac univentricular pathology that had been surgically corrected by the Fontan procedure, was admitted with premature rupture of membranes at the period of gestation (POG) of 34 weeks.

Her heart disease was detected at birth. The 2D echocardiogram revealed that the right ventricle had grossly dilated and hypertrophied, the left ventricle was rudimentary and both aorta and pulmonary artery arising from the right ventricle with 50% override. A large ventricular septal defect and pulmonary stenosis were diagnosed with a pulmonary valve pressure gradient of 60 mmHg. Right heart cardiac catheterization confirmed the diagnosis.

At the age of 4 years, she underwent bidirectional Glenn shunt (BDGS) and atrial septectomy due to worsening arterial oxygen saturation. When she was 10 years, completion of extracardiac Fontan (fenestrated) by using a 24 mm Gortex conduit was performed. She remained asymptomatic and had a good exercise tolerance throughout the adolescent period.

Pre-pregnancy counseling was done regarding the high-risk and adverse outcomes. When she was 19 years, she gave birth to her first child by emergency lower segment caesarian section. The indication for it was primi breech in labour at the POG of 35 weeks. A

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healthy baby boy was born with a birth weight of 1.580 kg. The post partem period was unremarkable. This uncomplicated pregnancy was managed by a multidisciplinary team throughout the pregnancy.

Following the first pregnancy, a Jadelle (levonorgestrel) subdermal implant was inserted for contraception but had to be removed due to abnormal uterine bleeding.

The second pregnancy occurred after 4 years due to contraception failure. It was an unplanned pregnancy, but she wanted to continue with it. She was cared for by a multidisciplinary team consisting of obstetricians, cardiologist, hematologist, anesthetist, and neonatologist.

An early pregnancy echocardiography done at 7 weeks of POG revealed well-functioning Fontan circuit and well-functioning bilateral bidirectional Glenn shunt with preserved ventricular function without atrioventricular valve regurgitation. Her symptoms, haemodynamical status, shunt circulation, and ventricular function were closely monitored. A detailed anomaly scan of foetus was done at the 20 weeks of POG and was normal.

The 2D echocardiography done at POG 29 weeks revealed a mild ventricular dysfunction (ejection fraction of 45-50%) with global hypokinesis. There was no valvular regurgitation. Fontan circulation and BDGS were well functioning. Her B-type Natriuretic Peptide (BNP) level was 23.4 pg/mL (normal < 100 pg/mL) at the 30 weeks of POG.

She remained asymptomatic throughout the pregnancy. Her oxygen saturation was maintained around 96-98% on room air. She was on warfarin for up to 30 weeks of POG, The doses were titrated to maintain the target INR at 2.0-2.5. Her hemoglobin level ranged around 9-11 g/dL with oral iron supplements.

She was admitted to the ward with signs of preterm labour at the POG of 34 weeks. There were no signs of foetal distress. Tocolytics, dexamethasone, and magnesium sulfate were given to improve foetal outcome. Warfarin was discontinued and subcutaneous enoxaparin 1mg/kg body weight, twice daily dose regimen was started. Emergency caesarian section was performed under graded epidural anaesthesia and a baby girl with a birth weight of 1150 g was delivered. The mother was closely monitored at the intensive care unit peripartum for two days. The intrapartum and postpartum period was uneventful. Her consent was obtained prior to surgery to do bilateral ligation and recession of fallopian tubes at the time of the caesarian section.

The baby was monitored at the neonatal intensive care unit for one month. She was free of any congenital cardiac anomalies.

Discussion

Univentricular heart is a form of congenital cyanotic heart disease characterized by both atria emptying through a common atrioventricular valve or through separate valves into a single ventricular chamber, with total mixing of systemic and pulmonary venous return. The prominent ventricular chamber may have left, right, or indeterminate ventricular anatomic characteristics. The other ventricle may be small, rudimentary, or absent altogether. The aorta and pulmonary artery both usually arise from a prominent single ventricle although one of the great vessels may originate from the rudimentary ventricle. The aorta may position posterior, anterior, or side by side to the pulmonary artery. Pulmonary stenosis or atresia is commonly associated with univentricular hearts.[1,3]

The Fontan procedure is a palliative staged operation for all children who cannot undergo two ventricular repair. The BDGS surgery is usually performed at the age between 2-6 months followed by Fontan correction at 2-3 years of age when the child is ambulatory.[1] Staged surgical palliation showed better outcomes compared to the originally described Fontan procedure. Our patient had undergone both procedures with a significant delay at the age of 6 years.

In BDGS, the superior vena cava is divided and detached from the right atrium, and the pulmonary artery is divided and detached from the right ventricle. Then the superior vena cava is anastomosed end-to-side to the pulmonary artery,[1] which reduces the volume load on the ventricle and reduce the progression of ventricular dysfunction.

The originally designed Fontan procedure has been modified for better results. Our patient had undergone the modified Fontan procedure, which uses an extracardiac conduit between the inferior vena cava and right pulmonary artery instead of the originally described intra-atrial routing of blood. The inferior vena cava was anastomosed directly to the pulmonary artery through a homograft or Gore-Tex tube running outside the heart (external conduit Fontan). This procedure is known as cavo-pulmonary isolation, and it allows the desaturated blood from both vena cavas to direct into the pulmonary arteries, and oxygenated blood returns to left atrium followed by left ventricle and ejected into the systemic circulation.[1] The Fontan operation eliminates the volume overload of the functioning systemic ventricle at the expense of limited ability to increase cardiac output.

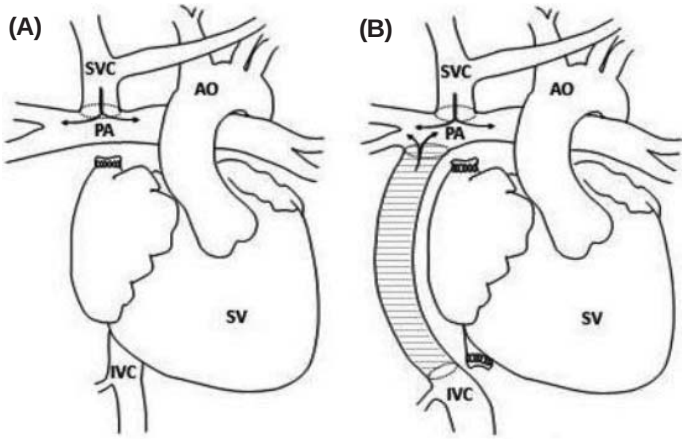

Figure 1. Demonstration of BDGS procedure (A) and the modified Fontan procedure (B).

According to the modified World Health Organization classification of maternal cardiovascular risk, unrepaired cyanotic heart diseases or uncomplicated Fontan circulation is categorized into mWHO risk category 111 (maternal cardiac event rate 19-27%), and Fontan circulation with any complication categorized into category 1V (maternal cardiac event rate 40-100%). In asymptomatic women with Fontan circulation and with good ventricular function without significant atrioventricular valve regurgitation, pregnancy can be considered with extreme care. A multidisciplinary approach is essential with close monitoring for possible complications such as systemic venous congestion, increased atrioventricular valve regurgitation, atrial and ventricular arrhythmias, thromboembolism, and if the Fontan is fenestrated for paradoxical emboli.[5] In addition, they are at increased risk of maternal complications such as maternal heart failure, pregnancy-induced hypertension, pulmonary oedema, infective endocarditis, haemorrhage, thrombosis, and maternal death.[3] It is recommended that Fontan patients should have at least monthly monitoring during the pregnancy and the first week after the delivery. They should be managed at a tertiary care center by a multidisciplinary team, that consists of an obstetrician, cardiologist, haematologist anesthetist, and neonatalogist.[4,5]

The risk of premature delivery, fetal mortality, and small for gestational-age offspring is significantly high. Higher frequency of preterm premature rupture of membranes has been reported among these mothers.[4] Our patient also had a similar presentation.

Iron deficiency anaemia is often caused by increased haematinic requirement with compensatory erythrocytosis for low oxygen saturation. Thus, iron level and haemoglobin should be checked regularly and replaced as required. Therapeutic anticoagulation should be considered as per the increased risk of thromboembolic events. Atrial arrhythmia should be recognized and treated. All women should have deep vein thrombosis prophylaxis following caesarian section.[4]

Equally good outcomes have been reported following vaginal delivery as well as caesarian section. Vaginal delivery is recommended in pregnant women with functionally normal univentricular hearts unless an obstetric indication for caesarian section.[3] During 48-72 hours after delivery, there is an increased risk of cardiac failure and pulmonary oedema due to autotransfusion phenomenon and therefore a need for close monitoring during this period. Repeat echocardiography between 6-8 weeks after the delivery is preferred to identify any delayed complications.[4]

Conclusion

A successful pregnancy is achievable in a woman with an univentricular heart and uncomplicated Fontan circulation with careful assessment, monitoring, and interventions by a multidisciplinary team even though there is an increased risk of maternal and fetal complications during the pregnancy and peripartum period.

Author declarations

Consent

Informed consent for publication was obtained from the patient.

Conflicts of interests

None.

Criteria for authorship

All the authors contributed equally to the areas of conception and design of the work, drafting, and revising of the manuscript. All authors have read and approved the final manuscript.

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