

Pregnancy and Delivery in Functional Single Ventricle Patient; Successful Long-Term Outcome after Right Ventricle Exclusion and Fontan Operation

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Twenty-six-year-old Ebstein's anomaly patient, who had failed both biventricular and one-and-a-half repair, underwent right ventricle exclusion and Fontan operation. She completed pregnancy and delivery owing to the excellent long-term clinical course. Although the caesarean section was performed due to symptoms of heart failure on the gestational age of 32+6 weeks, preterm delivery was agreeable with neonatology support. In Korea, there has not yet been a case of pregnancy and delivery of functional single ventricle patient because most patients have been discouraged from getting pregnant. However, functional single ventricle patient can endure pregnancy and delivery, if valve function and ventricular contractility, status of Fontan pathway and absence of arrhythmia predict favorable outcome as presented in this case. Although the patient maintained her pregnancy without anticoagulation owing to laminar flow in the Fontan pathway and absence of thromboembolic event, anticoagulation should be considered, weighing the benefits and risks during the pregnancy. (**Korean Circ J 2016;46(1):111-114**)

KEY WORDS: Fontan procedure; Ebstein anomaly; Pregnancy; Delivery, obstetric; Heart failure.

Introduction

Many doctors discourage functional single ventricle (FSV) patients from becoming pregnant due to the concern that the cardiovascular burden during pregnancy and delivery may provoke peripartum mortality or persistent heart failure. While cases of successful pregnancy and delivery have been reported in the United States¹⁾ and Japan,²⁾ there have been no reported cases of

pregnancy in FSV patients in Korea so far. The patient studied in this case had already been reported for an immediate post-operative course of right ventricle (RV) exclusion and Fontan procedure.^{3,4)} However, her excellent long-term results, including pregnancy and delivery, should be notable with favorable prognostic factors and peripartum clinical courses.

Case

The patient was a 26-year-old female with Ebstein's anomaly disorder. When she was five years old, she underwent biventricular repair with Danielson's technique. However, after 11 years, she developed severe tricuspid regurgitation, resulting in RV failure. At that time, she underwent a tricuspid valve (TV) replacement and bilateral cavopulmonary bypass. Five years later, she was presented with severe dyspnea. Severe RV failure with paradoxical septal movement as well as atrial flutter were observed. Consequently, RV exclusion, right-side maze, and extracardiac conduit Fontan procedure were executed. The prosthetic TV was removed, and RV inlet was obliterated with a bovine pericardium patch with 5 mm

Received: December 5, 2014

Revision Received: March 11, 2015

Accepted: June 2, 2015

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• The authors have no financial conflicts of interest.

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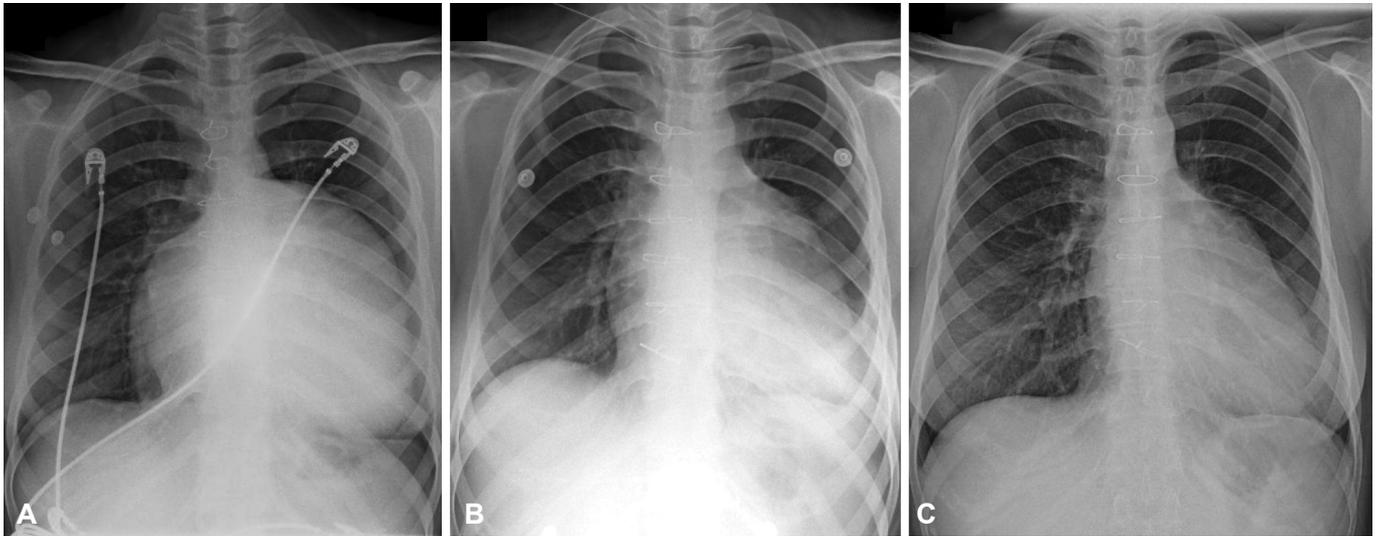


Fig. 1. Cardiomegaly has been improved on serial chest X-ray. (A) Before operation (B) Post-operative day 19 (C) 3 years after operation.

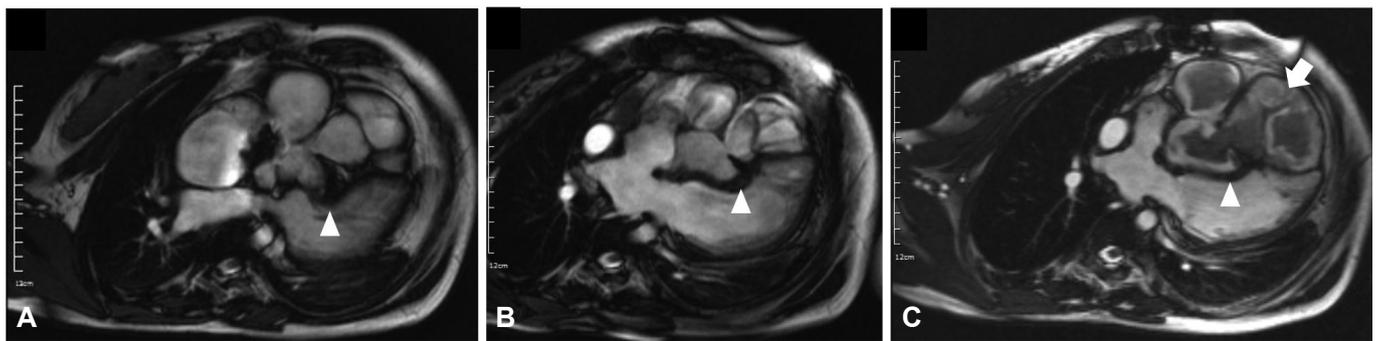


Fig. 2. Cardiac magnetic resonance imaging. (A) Before operation, right ventricle (RV) is enlarged, compressing left ventricle (LV). Interventricular septum (arrowhead) is deviated toward LV. (B) 1 month after operation, RV size is decreased. Interventricular septum (arrowhead) is still deviated toward LV. (C) 3 years after operation, RV is filled with thrombus (arrow) without inflow. LV function is improved with decreased paradoxical septal movement (arrowhead).

fenestrated window. At 19 days after operation, the patient was discharged without complications.⁴⁾ During the follow-up period, the patient maintained a stable cardiac function, with a B-type natriuretic peptide (BNP) value under 50 pg/mL, compared to the pre-operative value of 562 pg/mL. Moreover, the left ventricle (LV) ejection fraction (EF) on the echocardiography rose from 14.2% in the pre-operative state to 72% at the 3-year follow-up. Cardiomegaly on the chest X-ray was dramatically normalized (Fig. 1).

There were concerns that the inflow to the RV via TV fenestration might provoke the RV enlargement, compressing the LV again, or that thrombi in the RV might cause a paradoxical embolism. However, the patient neither had a thromboembolic event nor suffered from compromised LV function. Even though there had been RV inflow and enlarged RV during the early post-operative period, there had been no inflow into the RV since the patient's 3-year follow-up. LV was

not compressed, and paradoxical septal movement was decreased on the 3-year follow-up cardiac magnetic resonance imaging (MRI). The LV contractility improved as compared to the one-month post-operative MRI (Fig. 2).

Four years and six months after the last operation, the patient revealed her desire to become pregnant. Until then, she maintained a consistently normal sinus rhythm. At echocardiography, valve function was good except for mild degree mitral regurgitation. There were neither thrombi in the Fontan pathway, nor turbulent flow. Considering such favorable factors, we predicted that she could safely complete her pregnancy, and discontinued all her medications (enalapril, aspirin, digoxin, and spironolactone). She returned to the clinic in a pregnant state seven weeks later and was followed up along with her clinical symptoms, BNP level, and echocardiography.

Clinical visit including cardiac evaluation was scheduled on gestational age (GA) of 9+5 weeks and GA of 22+6weeks, while in the meantime, the routine prenatal screening test was executed in the local hospital. Although echocardiography showed slightly decreased EF of 50% on GA of 22+6weeks, valve regurgitation was not exacerbated. Diastolic function was evaluated with deceleration time (DT) of 157 ms that was not much different from the previous value.

BNP level was under 50 pg/mL. Her body weight increased to 58.4 kg that was 52 kg before pregnancy.

At the next clinic visit, on GA of 28+6weeks, she complained of headache, blurred vision and epigastric pain. However, admission was deferred because blood pressure was within the normal range. A dipstick test performed presented no albuminuria, and the patient's body weight was 60.7 kg, which was within the predicted range. On the next clinic visit, at GA of 31+6 weeks, she presented dyspnea (NYHA functional class III) and traces of albumin at dipstick test. Her body weight increased to 62.7 kg with generalized edema. With such findings, she was admitted for close monitoring. Peripheral capillary oxygen saturation (SpO₂) was 94% at room air, with nasal prong of oxygen flow of 2 L, SpO₂ increased up to 98%. During echocardiography, the mitral regurgitation increased from mild to moderate, while the LVEF was 51%. DT mildly rose to 165 ms.

At Holter monitoring, there was no arrhythmia. The patient's BNP level rose to 107 pg/mL on the third day at the hospital compared to 29 pg/mL on the day of admission. Although dyspnea was alleviated, elective caesarean section after antenatal corticosteroid therapy was planned because the overloaded volume during pregnancy could exacerbate congestive heart failure.

Prenatal cardiologic management included strict input/output balance. Intra-operatively, sufficient volume replacement was recommended to maintain adequate preload of Fontan circulation.

With an experienced neonatal care team available, the patient underwent an elective cesarean section at GA of 32+3 weeks under spinal anesthesia. Throughout the operation time, SpO₂ was maintained over 98%. Without the intra-operative event, the patient was transferred to the intensive care unit. Input/output balance was monitored closely and chest x-ray was followed up to determine the presence of pulmonary edema or cardiomegaly. Intermittent intravenous furosemide was used to maintain urine output and spironolactone medication was restarted post-operative day one. To avoid arrhythmia, electrolyte level was frequently checked, and potassium replacement was done. On post-operativeday one, the patient's BNP level dropped to 36 pg/mL, while her mitral regurgitation decreased to a mild degree, the LVEF rose to 50% with a deceleration time of 196 ms. Nasal prong oxygen supplement was discontinued because her SpO₂ was 96%

on room air. With diuretics, bodyweight decreased from 60.5 kg to 58.9 kg on the 6th day post-operatively. She was discharged on post-operative day 8 without any heart failure symptoms. Six-month follow-up echocardiography showed a LVEF of 60% and mitral regurgitation of negligible degree. However, diastolic function was compromised with DT of 238 ms. At that time, aspirin and enalapril were restarted.

The preterm neonate, weighing 1.73 kg, was admitted to the neonatal intensive care unit, and treated for neonatal respiratory distress syndrome and patent ductus arteriosus. With medical treatment including surfactant and indomethacin, the neonate was discharged without any complications.

Discussion

Pregnancy and delivery require a 30–80% increase in cardiac output.⁵⁾ FSV patients face the increased cardiac output demand with difficulty due to their compromised hearts. However, concerns about pregnancy in FSV patients appear to be too much in light of recently reported outcomes. For example, Canobbio et al.¹⁾ reported 71 pregnancies of 45 FSV patients, with no maternal mortality and a live birth rate of 73%.

Medications prescribed before pregnancy were aspirin, digoxin, enalapril and spironolactone. Enalapril is D and the other three drugs, including aspirin, are C in US Food and Drug Administration (FDA) pregnancy categories. Category C means that animal studies have shown adverse effect of the drug on the fetus and there are no adequate and well-controlled studies in humans. When it comes to the discontinuation of aspirin, considering increased thromboembolic risk of pregnancy and Fontan circulation, anticoagulation could have been used. The 2010 European Society of Cardiology guidelines and the American College of Cardiology/American Heart Association (ACC/AHA) 2008 guidelines recommend the use of anticoagulation in patients after Fontan operation, especially in the conditions where there is atrial thrombus, atrial arrhythmias or thromboembolic events.^{6,7)} ACC/AHA guideline adds atrial shunt as an indication for anticoagulation. In 2011, however, Monagle et al.⁸⁾ published results of multicenter, randomized controlled trial that showed no difference in the thrombosis rate of Fontan patients between the use of aspirin and heparin/warfarin. In this case, there was neither thrombus in Fontan pathway through the whole follow-up echocardiography nor the event of thromboembolism without anticoagulation. It was possible because the laminar flow on the Fontan pathway was well-maintained. However, according to the state and flow of Fontan pathway and existence of arrhythmia, low-molecular-weighted heparin can be used, which belongs to the

US FDA pregnancy category B, bearing bleeding complication risk.

It is suggested that maternal-fetal outcomes are affected by Fontan circulation status, functional capacity of a patient, systemic ventricular function, atrial arrhythmia, moderate-to-severe atrio-ventricular regurgitation and the degree of cyanosis.⁵⁾ Regarding these factors, pregnancy and delivery can be planned in Fontan circulation patients with favorable features.

Although the patient did not carry her child to full-term, preterm delivery was agreeable due to neonatology support. If meticulous cooperation exists between pediatric cardiologists, obstetricians, and neonatologists, as was the case, pregnancy in FSV patients should not be discouraged.

Acknowledgments

We deeply appreciate the effort of Chung Il Noh, MD, and Joong Shin Park, MD for the meticulous post-operative care and peripartum management of this patient as a pediatric cardiologist and an obstetrician.

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