

[Case Report]

Two successful pregnancies with vaginal deliveries in a patient with uni-ventricle after a ventricular septation procedure: 5 year follow-up of pre- and post-partum in a rare surgery.

Yumi Shiina MD, PhD¹⁾, Koichiro Niwa MD, PhD¹⁾,
Michiko Yamanaka MD, PhD²⁾, Naoto Kawamatsu MD¹⁾,
Ayumi Suzuki⁴⁾, Makoto Nakazawa MD, PhD^{3,5)}

¹⁾Cardiovascular center, St.Luke's International Hospital, Tokyo Japan

²⁾Department of Obstetrics, St.Luke's International Hospital, Tokyo Japan

³⁾Department of Pediatrics, Tokyo Clinic, Tokyo Japan

⁴⁾Department of Radiology, Tokyo Clinic, Tokyo Japan

⁵⁾Pediatric and Lifelong Congenital Cardiology Institute, Southern Tohoku General Hospital, Fukushima, Japan

Abstract

A woman with uni-ventricular heart with double inlet left ventricle (DILV), who had undergone ventricular septation procedure, had two successful vaginal deliveries. She delivered a 2062g male infant (small for gestational age) at 37 weeks of gestation at 29 years old, and a 2604g female infant (appropriate for gestational age) at 38 weeks of gestation at 33 years old. She developed mild heart failure and took diuretics after the first delivery. During the second pregnancy, she sometimes complained of palpitations, but she had no severe cardiovascular complications. We think that pregnancy in woman with ventricular septation procedure could follow a favorable course if she has "appropriate paradoxical septal motion" as well as normal AV valve function, normal ventricular size, no bulbo-ventricular foramen outlet obstruction or no ventricular hypertrophy.

Key words : Uni-ventricle, ventricular septation procedure, pregnancy, congenital heart disease

Introduction

Ventricular septation procedure is a biventricular repair for uni-ventricular heart, and the procedure is amenable only for DILV type ¹⁾. It was started in 1956 and continued to be performed until 1990s, with an expectation of better postoperative hemodynamics compared with post-Fontan patients. But relatively high perioperative mortality had been reported compared to Fontan procedure and now this is extinct. Surgical mortality in most early series ranged from 36% to 47% and mortality in late series was from 25 to 27%, which was still worse than that of comparable patients undergoing Fontan palliation ²⁻⁵⁾.

Case report

She was diagnosed with uni-ventricular heart (double inlet left ventricle: DILV), with valvular pulmonary stenosis (PS) aged 4 months. At that time, her complex heart was considered to be inoperative and she had been observed conservatively. At the age of 8 years, she underwent ventricular septation procedure and pulmonary valvotomy on 1991. After the operation, she had been very well and had been in New York Heart Association functional class (NYHA FC) I throughout the course. Before the first pregnancy, she was a full time worker and in a stable condition without significant exercise intolerance. She had been on no medication.

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Corresponding author : Yumi Shiina, MD, PhD, Cardiovascular center, St.Luke's International Hospital, Tokyo Japan

9-1 Akashi Chuoku Tokyo 104-8560, E-mail: yushiina@luke.ac.jp

Catheterization data aged 8 years soon after ventricular septation procedure were the followings: left ventricular (LV) peak / LV end diastolic pressure (EDP) was 95/16 mmHg, right ventricular (RV) peak / RVEDP was 33/14 mmHg, cardiac index was 2.4 L/min/m², and the Qp/Qs ratio was 2.0. Her ventricular septation patch was large (approximately 60mm in diameter) and showed paradoxical motion before pregnancy (figure1). Left ventricular ejection fraction (LVEF) was around 55% and estimated cardiac output by echocardiogram was 4.5L/min (cardiac index 3.0 L/min/m²). There was also a patch leak or fenestration (approximately 7mm in diameter) with a residual shunt (left to right). There was mild right and left atrioventricular valve regurgitation. RV function was preserved and there is no significant pulmonary valve stenosis or regurgitation. There was no significant biventricular hypertrophy, bulbo-ventricular foramen outlet obstruction or pulmonary hypertension. Catheterization was not performed before the two pregnancies because she had been followed up in another clinic and she was referred to our hospital during the first trimester of the two pregnancies.

1. First pregnancy and delivery

She was referred to our hospital at 10 weeks of gestation by natural conception at 29 years old.

At that time, she was 159cm tall and 50kg in body weight. During the second trimester, she was still fit and well without palpitations or

breathlessness. At 32 weeks pregnancy, she began to complain of fatigue. At 36 weeks gestation, her breathlessness worsened and auscultation of the heart revealed Gallop sounds. She had normal blood pressure, but proteinuria was +1 (total amount was less than 300mg/day) at that time. She had a mild peripheral edema but did not gain weight significantly. Serum brain natriuretic peptide (BNP) was elevated and her LVEF on echocardiography gradually decreased during the third trimester (figure2). This situation did not fulfill the criteria for preeclampsia, but we doubted whether or not this was a very early sign of preeclampsia. As for her fetus, cardiotocogram showed normal heart rate (HR), whereas amniotic fluid reduced and the fetal growth arrested. So, we planned to induce the labor within a couple of days. She delivered 2062g male infant (small for gestational age) with Apgar score 9/9 (1 minute / 5minutes) by forceps under the epidural anesthesia at 37 weeks of gestation. On the exact day, her blood pressure was less than 140/90 mmHg and the total amount of proteinuria was 1073mg, therefore this situation still did not fulfill the strict criteria for genuine preeclampsia. Blood loss was 1370ml. After delivery, isolated ventricular ectopics were sometimes seen. Next day, her chest X-ray showed mild pleural effusion. Her EF dropped to 35% and RV pressure, estimated by tricuspid regurgitation, increased. Particularly right-sided atrioventricular (AV) valve regurgitation worsened. LV diastolic parameters (lateral e' and E/e') also worsened.

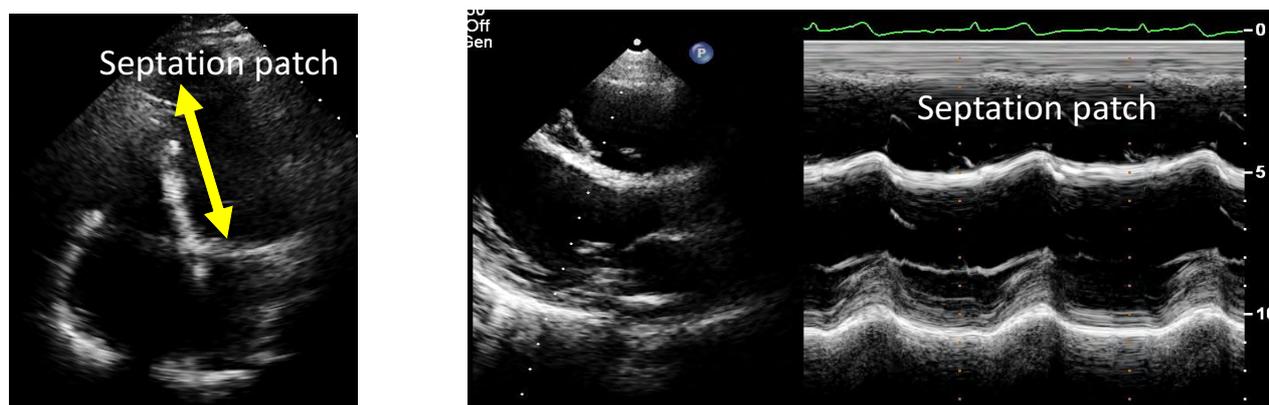
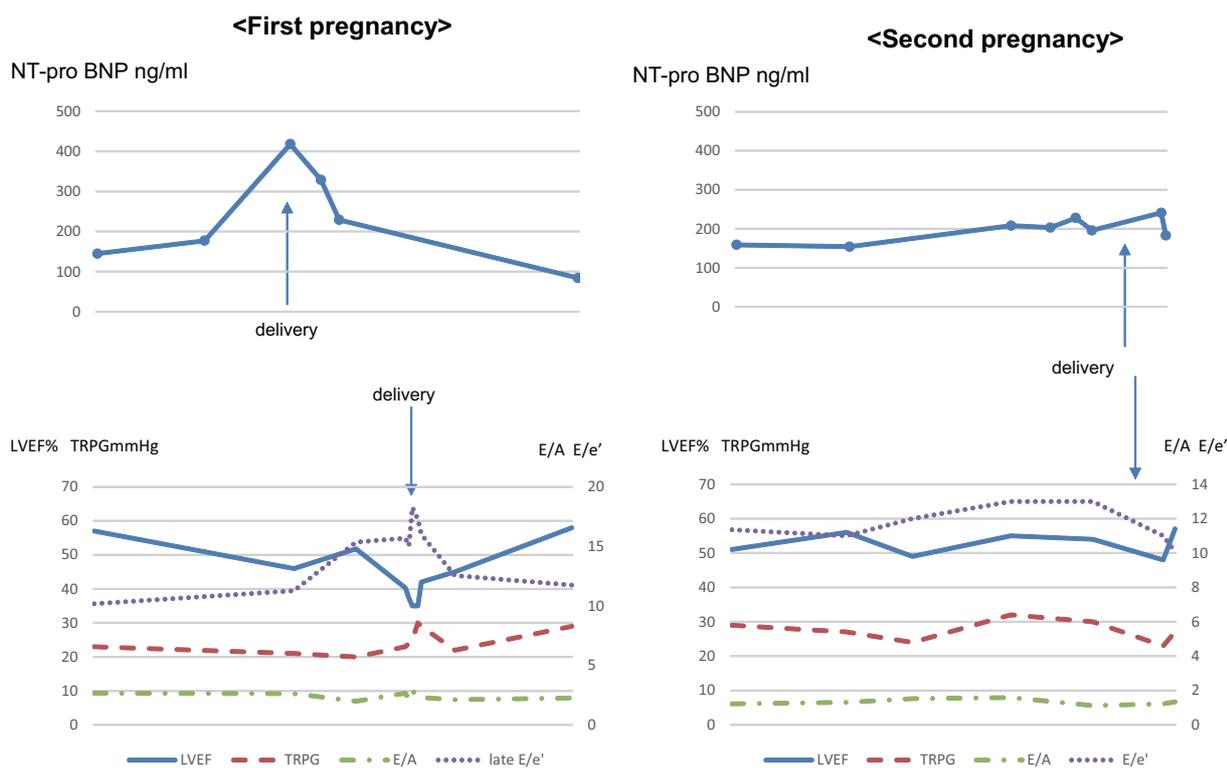


Figure 1. Echocardiography in uni-ventricle with ventricular septation procedure before pregnancy



Abbreviations: LVEF: left ventricular ejection, TRPG: tricuspid regurgitation peak gradient, E/A : LV inflow E wave/A wave, E/e': LV inflow E wave / tissue Doppler lateral e'

Figure 2. BNP and echo parameters during and after pregnancy

Paradoxical septal motion was mildly improved, probably because biventricular end-diastolic pressures were elevated (figure3). Clinically she had heart failure after delivery and started diuretics (furosemide 10mg daily and spironolactone 25mg daily). After taking diuretics, she had enough urine volume and mild pleural effusion disappeared. Her EF and diastolic function gradually recovered and became normal before discharge. She was discharged 8 days after the delivery. Diuretics intake was discontinued 2 weeks after discharge. She raised her baby with breast feeding in the daytime and her mother supported bottle feeding at night.

2. Second pregnancy and delivery

Two years after the initial pregnancy, she became pregnant again by natural conception at the age of 33. Her cardiac function was in NYHA FC II. Before the second pregnancy, she sometimes felt fatigue and palpitations, whereas there was no fatal arrhythmia on a 24 hour tape.

In the second trimester (22 weeks pregnancy), she sometimes complained of palpitations. On a 24 hour tape, both isolated supraventricular and ventricular ectopics were identified, but they were not sustained arrhythmias. We recommended her to take a beta blocker for her palpitations, but she declined to take it because of possible adverse effect on the fetus.

At 36 weeks, she began to complain of mild fatigue, whereas she did not get edema or gain weight. She had normal blood pressure, but proteinuria was +1 (total amount was less than 300mg/day) again. The fetus showed normal HR on cardiotocogram and normal amniotic fluid. There was no significant worsen of LV systolic function, and paradoxical septal motion was mildly improved. We scheduled to induce the labor at 38 weeks of gestation. She delivered 2604g female infant (appropriate for gestation age) with Apgar score 8/9 (1minute/5minutes) by vacuum extraction under the epidural anesthesia at 38 weeks. The blood loss was 615ml. Since her anemia had not improved after delivery and the hemoglobin level decreased to 7.6g/dl, so she

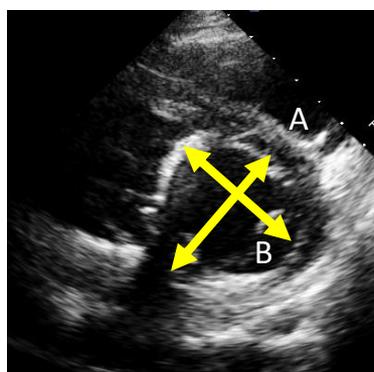
received blood transfusion. After delivery, the patient maintained normal blood pressure and heart rate. LV diastolic function slightly worsened, but recovered 2 days after delivery. No sign of heart failure or arrhythmias were seen, whereas a low-dose furosemide intake was started as a prophylaxis of heart failure. She was discharged 6 days after delivery. She continued to take diuretics for two weeks and fit and has been well so far.

Discussions

There is only one report searched with the index words 'pregnancy' and 'ventricular septation procedure'. The first and only one successful pregnancy after ventricular septation procedure was reported in 1990 ⁶⁾. The report concluded that pregnancy in the woman with a septation procedure can follow a favorable course if she is acyanotic without functional or cardiovascular compromise. Ohuchi et al reported that the exercise capacity of patients after ventricular septation procedure was better than that after Fontan procedure when the patient had no significant AV valve regurgitation ⁷⁾. When patients were appropriately selected, ventricular septation procedure still may be an attractive surgery for uni-ventricular heart of DILV type, especially in relation to pregnancy and delivery. Poor clinical outcome of ventricular septation procedure is

associated with abnormal AV valve function, small ventricular size, bulbo-ventricular foramen outlet obstruction and ventricular hypertrophy ⁸⁻⁹⁾. In our case, the patient was well-selected for septation procedure and good current cardiovascular status, therefore she underwent two successful pregnancies.

As for paradoxical septal motion, we should carefully interpret it in this challenging population ¹⁰⁾. In end-diastole, the non-contractile interventricular septum shifted toward the LV side, obviously the result of pressure difference between the two ventricles. The diastolic leftward movement of the septum is crucial for "earning" of diastolic volume of the RV. In other words, this paradoxical motion of the septum obviously reduces pump function of the LV, but augments pump function of the RV. Thus, the paradoxical septal movement is an essential factor for systolic function of the RV and also to equalize stroke volume of the two ventricles (10). During pregnancy, blood volume is gradually increased and subsequently biventricular volumes are also increasing. As a result, the paradoxical motion is improving (figure3), particularly in the third trimester. In our case, her LV filling pressure seemed to be higher than right-side pressure even in end-diastole. This "non-paradoxical" motion should be advantageous to the pump function of the LV



Deformation ratio = A/B

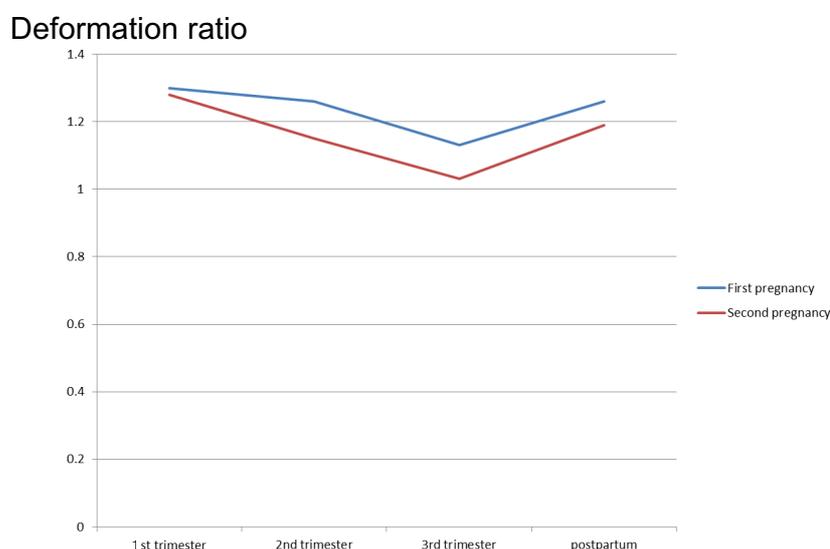


Figure 3. LV deformation in end-diastole

generally, but may not be advantageous for the right heart hemodynamics resulting in elevation of filling pressure of the RV.

We think that pregnancy in woman with ventricular septation procedure can follow a favorable course if she has “appropriate paradoxical septal motion” as well as normal AV valve function, normal ventricular size, no bulbo-ventricular foramen outlet obstruction or no ventricular hypertrophy.

Conclusions

A woman with uni-ventricular heart with double inlet left ventricle (DILV), who had undergone ventricular septation procedure, had two successful vaginal deliveries. We think that pregnancy in woman with ventricular septation procedure can follow a favorable course if she has “appropriate paradoxical septal motion” as well as normal AV valve function, normal ventricular size, no bulbo-ventricular foramen outlet obstruction or no ventricular hypertrophy.

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Details of ethics approval: We got the informed consent from this patients.

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