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Case Report

Pregnancy with Fontan circulation: A report of case series in Japan

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ABSTRACT

Owing to new surgical procedures and medications, more women who have undergone the Fontan procedure reach childbearing ages. We report five cases of pregnancy with Fontan circulation. Case 1 had subchorionic hematoma (SCH), fetal growth restriction (FGR), and preterm labor (PTL). She delivered a 1073 g infant via cesarean section at gestation week 28 because of hemorrhagic shock. Case 2 delivered 2142 g and 2232 g infants at gestation weeks 37 and 36, respectively. She had FGR, PTL, and postpartum hemorrhage (PPH). Case 3 had SCH, PTL, and heart failure. At 36 weeks, labor was induced and she delivered a 2546 g infant by vacuum extraction with epidural analgesia. Cases 4 and 5 resulted in miscarriage. All subjects experienced obstetrical complications. This report discusses pregnant women with Fontan circulation by focusing on affected Japanese women.

<Learning objective: More women who have undergone the Fontan procedure reach childbearing ages due to the new surgical procedures and medications. We report five cases and all of them experienced obstetrical complications. Anticoagulation might be one of the causes of postpartum hemorrhage. Labor epidural analgesia could contribute to minimized dynamic circulatory changes during labor. This report can be the literature regarding pregnant women with Fontan circulation in Japan.>

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Introduction

As a result of surgical procedures and medications, more women who have undergone the Fontan procedure reach childbearing ages. Pregnancy after the Fontan procedure carries one of the highest perinatal risks [1,2] and can result in disorders, including maternal heart failure, arrhythmia, hypertensive disorder of pregnancy, and postpartum hemorrhage. Fetal complications, such as intrauterine growth retardation, preterm labor, and low birth weight can also occur. Here, we report five cases of pregnancy with Fontan circulation in Japan (Table 1).

Case report

Case 1: A 33-year-old primiparous woman with a history of congenital pulmonary atresia and an intact vertical septum

presented to our hospital at 5 weeks of gestation. She had undergone the Fontan operation with an atriopulmonary connection (APC), an extracardiac total cavopulmonary connection (TCPC), and pacemaker implantation for a postoperative complete atrioventricular block with syncopal and vertiginous episodes. Her preconception brain natriuretic peptide (BNP) level was 38.8 pg/mL with a New York Heart Association (NYHA) class II functional status. Furosemide, spironolactone, hydrochlorothiazide, pilsicainide hydrochloride, and warfarin 4 mg per day were administered regularly. At conception, warfarin was replaced with unfractionated heparin (UFH) and acetylsalicylic acid, and pilsicainide hydrochloride was replaced with digoxin. The patient was admitted for massive genital bleeding due to subchorionic hemorrhage at 11 weeks gestation. The dose of UFH was 10,000 U per day and activated partial thromboplastin time (APTT) value was 47.5 s. When the bleeding began to decrease, heparin was replaced with danaparoid sodium. The dose of danaparoid was 1250 U per day. BNP levels were relatively elevated during the pregnancy, sometimes over 100 pg/mL, while ultrasound indicated that there was no considerable change in cardiac ejection fraction. At 28 weeks of gestation, the patient experienced hemorrhagic

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Table 1 Summary of six cases.

Case	Age of patient (years)	Cardiac complications	Gestational age at delivery (weeks)	Mode of delivery	Perinatal complications	Weight of neonate (g)	Medical complication	Maximum BNP during pregnancy (pg/mL)	EF at delivery (%)
1	33	Complete AVB Mild MR Pace maker	29	C-section	PTL, FGR, SGA, SCH, PPH	1073	Ascites, CHF	100.8	68
2-1	30	Mild TR Moderate TR	37	Vaginal delivery, Vacuum	FGR, SGA, PPH	2142	–	55.2	60
2-2	33	Mild TR Mild MR	36	Vaginal delivery	PTL, FGR, SGA, PPH	2232	–	46.1	58
3	28	Moderate AVVR Trivial AR	36	Vaginal delivery, Vacuum	HDP, PTL, SCH	2546	Heart failure	466	40
4	25	PSVT Moderate AVVR	14	Miscarriage	SCH, IUFD	–	Worse PSVT	135.5	
5	38	SSS Mild MR Pace maker	10	Miscarriage	IUFD	–	–	15.1	

BNP, brain natriuretic peptide; EF, ejection fraction; AVB, atrioventricular block; MR, mitral regurgitation; TR, tricuspid regurgitation; AVVR, atrioventricular regurgitation; AR, aortic regurgitation; PSVT, paroxysmal supraventricular tachycardia; SSS, sick sinus syndrome; HDP, hypertensive disorders; PTL, preterm labor; FGR, fetal growth restriction; SGA, small for gestational age; SCH, subchorionic hematoma; PPH, postpartum hemorrhage; IUFD, intrauterine fetal death, CHF, chronic heart failure; C-section, Cesarean section.

shock and imaging revealed the growth of a subchorionic hematoma. On that day, cesarean section was performed under general anesthesia. The volume of blood loss during the cesarean section was 1070 mL. The patient had a good postoperative course and resumed danaparoid sodium and warfarin on the third day following the procedure.

Case 2-1: A 31-year-old primiparous woman presented to our hospital at 6 weeks of gestation. She had undergone the Blalock–Taussig operation for a double inlet left ventricle at age 2 years, the Fontan procedure with an APC at age 12 years, and TCPC at age 27 years. Her BNP level was 29.9 pg/mL before conception and her NYHA functional status was class II. Furosemide, spironolactone, and warfarin 3.5 mg per day were administered regularly. Warfarin was replaced with UFH at conception and resumed at 15 weeks of gestation. She had a good perinatal course. She was admitted to our hospital at 36 weeks gestation for continuous intravenous heparin therapy to replace warfarin. During labor induction, which was started at 37 weeks, a central venous indwelling catheter was inserted to check the vital signs. She was delivered of her baby by vacuum. The volume of blood loss during labor was 1000 mL. Intrapartum central venous pressure remained below 15 mmHg. The patient had no episode of arrhythmia during the pregnancy and labor, and she had a good postpartum course.

Case 2-2: This is the same patient of Case 2-1. After 2 years had passed, she had a good perinatal course and BNP levels ranged between 10 and 50 pg/mL. She was admitted at 36 weeks to start continuous intravenous heparin instead of warfarin. Labor started at 36 weeks and 4 days of gestation. A central venous catheter was placed. She was delivered an infant by vacuum extraction. The volume of blood loss during labor was 1000 mL. Intrapartum central venous pressure remained below 20 mmHg. She had a good postpartum course.

Case 3: A 28-year-old primiparous woman was admitted to our hospital at 5 weeks of gestation. She had undergone an aortopulmonary shunt operation at age 2 years, a bidirectional Glenn shunt operation due to a double inlet right ventricle at age 9 years, and the Fontan procedure with TCPC at age 14 years. Her preconception BNP level was 18.3 pg/mL, with a NYHA class II functional status. She had been administered aspirin regularly and it was substituted with apixaban from 15 weeks of gestation. She was admitted for face and leg edema, 6 kg weight gain for 2 weeks, and proteinuria at 35 weeks. Her blood pressure was 145/90 mmHg. Chest radiography revealed

the enlarged cardiothoracic ratio of 50% and pleural effusion and that her BNP level had risen to 466 pg/mL. We started furosemide, spironolactone, and continuous intravenous heparin administration to replace apixaban. At 36 weeks, labor was induced. Epidural analgesia catheter was inserted 6 h after stopping heparin. She was placed supine with left uterine displacement. Fentanyl 25 µg was injected intrathecally. The first stage of labor was 7 h and 30 min, and the second stage of labor was 2 h and 54 min. Avoiding the labor effort, we used vacuum extraction to shorten the time of second stage. The blood loss during labor was 300 mL. The patient had no episode of arrhythmia during the pregnancy and labor. Her heart failure had improved, and her BNP level was 28.4 pg/mL at 2 weeks postpartum.

Case 4: A 25-year-old primiparous woman was admitted to our hospital at 5 weeks of gestation. She had undergone the Blalock–Taussig operation for a double inlet left ventricle at 1 month, the Fontan procedure at age 6 years. Paroxysmal supraventricular tachycardia (PSVT) appeared at the age of 20 years. She was taking antiarrhythmic therapy with propafenone hydrochloride, verapamil hydrochloride, and perindopril erbumine which was stopped at conception. She also took atenolol. PSVT began to occur frequently after conception. Rivaroxaban was replaced with UFH. She had miscarriage at 14 weeks of gestation.

Case 5: A 25-year-old primiparous woman was admitted to our hospital at 5 weeks of gestation. She had undergone the Fontan operation and pacemaker implantation for a postoperative sick sinus syndrome. Warfarin 2.5 mg per day was replaced with UFH. She had miscarriage at 10 weeks of gestation.

Discussion

Our report shows high perinatal risks in pregnant women with Fontan circulation. Recently, there were some studies reporting successful pregnancies with Fontan palliation [3,4].

We experienced postpartum hemorrhage (PPH) in Cases 1, 2-1, and 2-2. Blood loss of ≥ 500 mL for vaginal delivery and 1000 mL for cesarean section is classified as PPH. A previous study reported that women with a Fontan circulation were most prone to PPH [5]. Regarding anticoagulation, the advice is to stop taking low molecular weight heparin (LMWH) at first stage of labor and omit administration for at least 12 h (preventive) or 24 h (therapeutic) until delivery. On the other hand, another study reported pregnant women with therapeutic LMWH had a greater risk of PPH despite

stopping 24 h prior to delivery [6]. In our facility, warfarin is usually replaced with UFH at conception and resumed at 15 weeks of gestation. UFH is started at 10,000 U per day and adjusted the dose to keep the APTT level ranged from 50 to 80 s. After 36 weeks of gestation, warfarin is replaced with UFH again. Heparin is stopped 6 h before starting induction of labor. Although there is a possibility that anticoagulation is one of the causes for PPH, anticoagulation is important therapy for thromboembolic disease and valve thrombosis which are causes of maternal mortality. It remains a great issue whether the anticoagulant therapy is necessary or not for people with Fontan circulation.

Labor epidural analgesia could contribute to minimized dynamic circulatory changes during labor in patients with congenital heart disease. In case 3, the patient could avoid effort in the second stage of labor and stabilize circulation dynamics. There are several studies on the successful use of labor epidural analgesia for congenital heart disease [7,8]. The diminished venous return was prevented by avoiding the labor effort. Adequate pain relief with minor hemodynamic changes prevented tachycardia and hypertension. In case 2-1 and 2-2, epidural anesthesia had been planned but not been used because labor progressed rapidly. Further research is required to assess the effect of labor analgesia.

Some drugs, for example, diuretics, antiarrhythmic drugs, and anticoagulants are administered only when it is judged that the therapeutic benefits outweigh the risks during pregnancy. In all cases, the drugs administered before pregnancy were essential for the maternal treatment during pregnancy. That is why we continued the therapy to keep maternal cardiac condition stable.

There are few reports of pregnancy with Fontan circulation. It is important to recognize the individual risks associated with pregnancy in women with Fontan circulation. This report can be

the literature regarding pregnant women with Fontan circulation by focusing on affected Japanese women.

Conflict of interest

The authors declare that there is no conflict of interest.

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