Two Cases of Congenital Heart Diseases those Experienced Intracardiac Repair after Termination of Index Pregnancy

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Abstract

A 24 year patient with double-chambered right ventricle (DCRV) got pregnant and echocardiography revealed the pressure of high pressure chamber was equal to the left ventricle and she was told the risk for the index pregnancy and had intracardiac repair after termination of the pregnancy. Another 23 year patient with Ross procedure was told there was a severe pulmonary stenosis of the composite graft and was recommended re conduit replacement after termination. These two patients had not enough cardiac examination and did not understand well their own cardiac performance. For patients with congenital heart diseases, both education of their cardiac status and preconceptional counseling is necessary when they reach childbearing age depending on the patients’ age and development.

Keywords: Pregnancy; Echocardiography; Pulmonary stenosis; Congenital heart diseases

Introduction

Recently many patients with congenital heart disease, even severe or complex congenital heart disease, could become adulthood and childbearing age. And they intend to be pregnant, but some of them don’t know the risk of pregnancy. It is truly important to inform the risk of pregnancy and delivery to them. It is wise to avoid pregnancy in women with severe congenital heart disease because pregnancy will result in cardiac failure or arrhythmia and possibly maternal and fetal death. Also it will induce cardiac dysfunction and that will be lasting long even long-term after delivery. All women with congenital heart disease should know the risk of pregnancy and delivery before conception. We present two cases with severe congenital heart diseases became pregnant and resulted in artificial abortion due to very high risk of continuation of pregnancy. This report is focusing on importance of preconceptional counseling and education to the patients with congenital heart diseases.

Case 1

The patient was diagnosed as congenital heart disease at birth. And she had denied propose of intra-cardiac repair (ICR) at birth and at 20 year she had come Japan to study abroad and at 24 year she was 19 weeks of gestation, when she was referred to our hospital because of large Ventricular Septal Defect (VSD), pulmonary stenosis (PS) and whether she could continue pregnancy. She was diagnosed as double-chambered right ventricle (DCRV) on echocardiography, abnormal muscle bundles were admitted in the right ventricle (RV), perimembranous VSD was 20 mm in diameter and was open to high pressure chamber, pulmonary valvar stenosis was observed and the pressure of high pressure chamber was equal to the left ventricle (LV) [1].

Resting systemic pulse oximetry (SpO2) in the sitting position was 98%. After multidisciplinary team review, she was terminated at 21 weeks of gestation because volume load by pregnancy may cause right heart failure. Cardiac catheter revealed that the pressure of high pressure chamber was as high as 90% of systemic pressure, pressure gradient between main pulmonary artery (PA) to the RV inflow was 60 mm Hg, pulmonary to systemic flow ratio was 3.11. Patch closure of VSD, resection of abnormal muscle bundle, pulmonary commissurotomy was performed [2]. After two years, on echocardiography there was mild pulmonary stenosis and the pressure of RV is 30 mm Hg, she got pregnant again and she is in the 25 weeks of gestation with favorable cardiac condition and will continue the pregnancy.

Case 2

A systolic heart murmur was audible at 3 months old and the patient was diagnosed as aortic stenosis at 2 year and cardiac catheterization at 4 year revealed aortic valvar stenosis with pressure gradient of 35 mm Hg. At 6 year, she lost her consciousness when running, however, cardiac catheterization showed no exacerbation. She experienced loss of consciousness again and felt chest pain at 7 year and at 8 year, Ross procedure was performed at our hospital, RV outlet was replaced with 22 mm trileaflet polytetrafluorethylene valved couduit [3,4]. At 10 year, there was no aortic valve (native pulmonary valve) stenosis/regurgitation and coronary angiography showed no stenosis or kinking problems. At different hospital her cardiac function was to be followed, however she had repeatedly declined the follow up cardiac catheterization from 12 to 22 year. We had planned cardiac catheterization when she was 23 year because she was going to marry. However she got pregnant before it and at 19 weeks of gestation, she entered our hospital. On echocardiography, pressure gradient between the RV to PA conduit was 82 mm Hg with poor mobility of pulmonary valve. Left ventricular systolic function was good without aortic valve regurgitation, nor enlargement of the aorta. After multidisciplinary team review, she was terminated on 21 weeks of gestation because volume load by pregnancy may cause right heart failure.

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failure due to severe PS. Cardioangiography revealed valvar supravalvar stenosis in the composite graft and pressure gradient between RV inflow and main PA was 50 mm Hg. Pressure of the RV was 80% of the LV. Conduit replacement was performed with 26 mm trileaflet polytetrafluorethylene valved conduit [4]. The Post-operative course was favorable and the estimated RV pressure was 28 mm Hg. And she has regular cardiac check-up afterwards.

Discussion

Case 1 was a severe DCRV case and the pressure of high pressure chamber was 90% of the LV and the pressure gradient of main PA to the RV was estimated as 60 mm Hg. The case 2 had a severe PS, the pressure gradient between the RV and PA conduit was 82 mmHg. Case 1 had an indication of operation, because the high pressure chamber was as high as systemic chamber and the resection of the abnormal bundles, patch closure of the VSD are not the high risk operation [5,6]. Case 2 also had indication of conduit replacement because severe PS with the pressure gradient between main PA and the RV inflow >50 mm Hg [7]. Both patients had the wish for bearing baby, however continuation of pregnancy may have led to heart failure in both cases [8]. Perhaps surprisingly, signs and symptoms of right-heart failure, arrhythmia did not occur in the study subjects, although they might appear if the pregnancies was allowed to continue.

During pregnancy those with congenital heart diseases might experience heart failure, thromboembolism, arrhythmia, cyanosis, even though they were asymptomatic and in NYHA class 1 before conception [8]. One reason is circulation blood volume gradually increases by approximately 50% up to around 30 weeks of gestation and then reaches a plateau [9]. Another reason is, during pregnancy, there is an increase in all circulating coagulant factors, including fibrinogen and a hypercoagulative state occurs [10]. Preconceptional counseling by experts of congenital heart disease and pregnancy is especially important to assure safety for both the mother and the fetus and to avoid artificial abortion like these two cases [11]. Further the most important issue in these two cases is the patients had not the regular cardiac examination and had not known their own cardiac problems [12,13]. To be educated on their cardiac status is important to those who have congenital heart diseases, especially for girls at adolescence before they reach child bearing age [14]. When they reach child bearing age, it is important to gradually inform the risk for pregnancy in their current cardiac performance. This information includes physiological changes of pregnancy, effect of the drug to the fetus, expected course of pregnancy, or necessity of ICR before conception [15]. This information must be conveyed gradually but certainly depending on the patients’ mental and physiological developments [16,17].

In summary, as pregnancy is accompanied with increased volume overload, increased coagulopathy, some patients with congenital heart diseases becomes symptomatic during the second or third trimester or postpartum course. Especially for girls with congenital heart diseases, both education of their cardiac status and preconceptional counseling is necessary when they reach childbearing age depending on the patients’ age and development.

References