Scientific Foundation SPIROSKI, Skopje, Republic of Macedonia
South East European Journal of Cardiology. 2024 Jan 20; 5:1-5.
https://doi.org/10.3889/seejca.2024.6060
eISSN: 1857-9361
Category: *Case Report*
Section: *General Cardiology*

OPEN ACCESS

Successful Pregnancy with Uncorrected Tetralogy of Fallot: An Interdisciplinary Team Approach

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Edited by: Igor Spiroski
Citation: Millisani HI, Yogibuana V. Successful Pregnancy with Uncorrected Tetralogy of Fallot: An Interdisciplinary Team Approach. SEE J Cardiol. 2024 Jan 10; 5:1-5. https://doi.org/10.3889/seejca.2024.6060
Keyword: Uncorrected tetralogy of fallot; Pregnancy; Cyanotic congenital heart disease
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Received: 21-Aug-2023
Revised: 09-Dec-2023
Accepted: 11-Jan-2024
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Funding: This research did not receive any financial support
Competing Interests: The authors have declared that no competing interests exist
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Abstract

BACKGROUND: With an overall incidence of congenital cardiac disease, tetralogy of fallot (ToF) is the most clinical of cyanotic heart disease. The anomalies seen in patients with ToF are due to an inappropriate anterior and cephalad displacement of the interventricular septum's infundibular (outflow tract) component. Ventricular septal defect, right ventricular hypertrophy, overriding aorta, and pulmonary stenosis are the four defects that result from this condition. The physiology of healthy women will undergo several changes affecting body system, including the cardiovascular system. Pregnancy and delivery have a high risk for most ill women, including those with uncorrected ToF, due to extensive physiological adaption and alterations. For patients with ToF, it continues to be a significant contributor to maternal morbidity (62.5%), mortality (10%), and neonatal adverse outcomes.

CASE PRESENTATION: A 18-year-old female in her first pregnancy at 28 weeks was referred to Saiful Anwar Hospital with uncorrected ToF since she was 2 years old. She suggested repairing ToF at National Cardiovascular Center Harapan Kita Jakarta but her parents still refused. She married at 18 years old and she was pregnant unplanned before. Then, she was referred to RSSA for further management.

CONCLUSSION: Pregnancy has been related with higher risks of morbidity and mortality for women with congenital cardiac disease. ToF is the most prevalent cyanosis congenital heart disease. These are poorly tolerated and produce serious complications, including maternal and neonatal mortality. Prenatal counseling is crucial to identify risk factors for the patients and to reassure many patients who are at low risk.

Introduction

Tetralogy of fallot (ToF) is the most prevalence of cyanotic congenital heart disease (CHD) for 10% of all CHDs. The anomaly identified in ToF patients is caused by a single developmental defect, which is an anterior abnormality and cephalad displacement of the interventricular septum's infundibular. Most of patients require surgery, and just a few survive into adulthood if their ToF is not corrected. Ramage *et al*. previously showed 2114 newborns to mothers with adult CHD (ACHD) and suggested an association between neonatal and maternal adverse outcomes [1]. In comparison to women without ACHD, preterm births (37 weeks gestation) were 1.4 times more likely in women with ACHD. In addition, premature births that occurred at <32 weeks of gestation were more common in women with ACHD. A baby with a small gestational age (SGA) was also delivered by 12.8% of women with ACHD and 8.7% of women without ACHD [1], [2].

Case Report

A 18-year-old female in her first pregnancy in 28 weeks was referred to Saiful Anwar Hospital with uncorrected ToF. At 2 years of age, she had cyanosis and feeding difficulties. As a child, she also had failed to gain weight, recurrent respiratory tract infections, and dyspnea while playing with her friends. Her parents brought her to the pediatrician and diagnosed her with ToF at the age of 2 years. She suggested repairing ToF at National Cardiovascular Center Harapan Kita Jakarta but her parents still refused.

In 2021, she complain was shortness of breath while doing moderate activity and scoliosis. Her parents brought her to Baptis Hospital and she was referred to Saiful Anwar Malang Hospital. Then, she suggested echocardiography and diagnosed with ToF. She suggested referring to National Cardiovascular Center Harapan Kita Jakarta. In there, she underwent a stress echocardiography testing and showed ToF with the right aortic arch. The right heart catheterization showed ToF with the right aortic arch and dilatation

of the coronary artery. Then, she suggested repairing ToF and got Po. Propanolol 3×10 mg. Form the stress echocardiography testing showed tardokinesia at inferoseptal and anteroseptal segment, ventricular septal defect (VSD) subaortic with overriding aorta, with infundibular deviation anterocephal, severe pulmonary stenosis (PS), normal of left ventricle function, and right ventricle contractility.

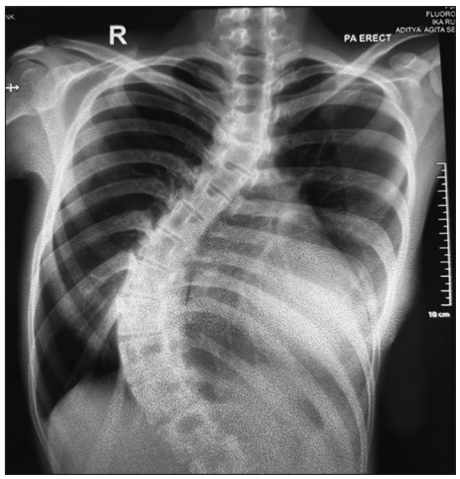


Figure 1: Chest X-ray showed cardiomegaly with scoliosis

She married at 18 years old and she was pregnant unplanned before. She routinely controls during pregnancy at an obstetrician and was referred to Saiful Anwar Hospital for delivery at 32 weeks. At the time of presentation to our department, she was shortness of breath with NYHA class II. On examination, we found blood pressure was 120/70 mmHg, heart rate was 80/min, and respiratory rate was 18/min. Saturation at upper extremities were 89% (dextra) and 92% (sinistra), lower extremities were 91% (dextra) and 90% (sinistra). On auscultation, we found that first and second heart sound was normal, continuous murmur grade 3/6 at the left lower sternal border, gallop (-). At obstetrical examination with fetal ultrasonography was small in gestation age and oligohydramnion. From laboratory finding, it was normal and chest X-ray that we found cardiomegaly with scoliosis (Figure 1). Electrocardiography sinus rhythm with p wave (P pulmonal) (Figure 2). Echocardiography showed a subaortic VSD bidirectional shunting, ±50% aortic override of septum with right of aortic arch, and PS valvar-sbuvalvar (peak gradient was 107mmHg). Left ventricular ejection fraction was 78% by teich (Figure 3).

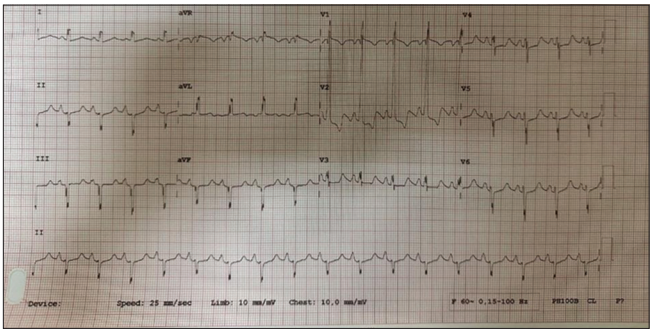


Figure 2: Electrocardiography

Pre-operative, she got Inj. Dexamethasone 2×6 mg for 2 days, neuroprotector with bolus $MgSO_4$ full dose 20% 4 g continue with drip $MgSO_4$ 40% 10 g for 24 h and. Inj. Clindamycin 2×300 mg. She was delivered by C-section with epidural anesthesia at 32 weeks with a multidiscipline team with risk score mWHO IV and Zahara score 3.2. Intraoperatively, there was no complication and her baby was a boy live birth (Weight: 1200 kg and Length 37 cm). After C-section, she suggested copper intrauterine device (IUD) insertion. After that, she and her baby were taken for 2 days to the intensive care unit. Clinical indications of cardiac disease were absent in her infant. On the 7th surgical day, the patient and her baby were both discharged.

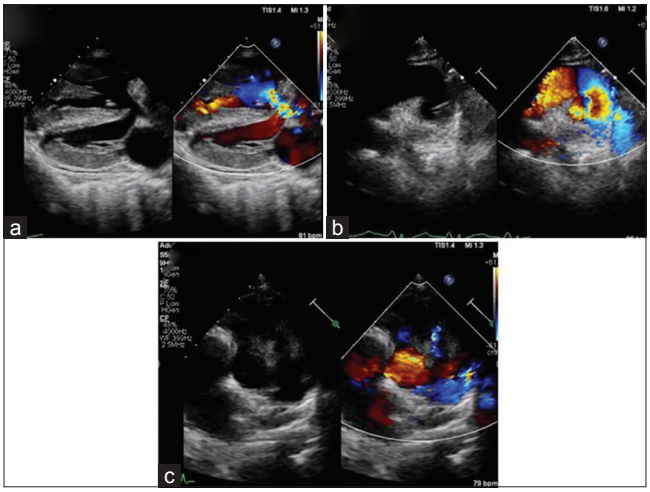


Figure 3: Echocardiography showed Sub-aortic ventricular septal defect (a), right of the aortic arch (b), and pulmonary stenosis (c)

Discussion

A serious form of cyanotic CHD is known as the hemodynamic abnormality characteristics of ToF due to anatomic anomaly (Figure 4). Pregnancy with uncorrected ToF is a rare case and increases the risk for the adverse outcome of pregnancy and fetal abnormality. This condition is related to cyanosis get worsening, right to left shunt, and pulmonary hypertension. Uncorrected ToF with pregnancy was found in about 2%–14% of incidents with an increase in morbidity and mortality when surgical procedures. Uncorrected TOF patients may have worsening during pregnancy and delivery. These continue to be a significant contributor to maternal death (10%), morbidity (62.5%), and fetal adverse outcomes. A multidisciplinary pregnancy-heart team is required to support treatment and medication, consultation, and follow-up before and after delivery. Veldtman *et al*., [3] explained that unrepair ToF had premature delivery with infants who were smaller than patients with repaired ToF for their gestational ages (8.6%), spontaneous fetal loss with multiple factors

(24%), and cesarean delivery (21%). In our patient, she was an unrepair TOF with pregnancy and delivery at 32 weeks by cesarean with a small baby (weight: 1200 kg and length 37 cm) [1], [4], [5].

ToF with pregnancy was a serious problem and increased morbidity and mortality for the patient with uncorrected surgery. Pregnancy can result in increased cardiac stress and decreased cardiac function, which can increase maternal and perinatal morbidity. A previous study has found that patients with uncorrected ToF were more to experience cardiac and obstetric complications. The most common cause of cardiac and obstetric complications is uncorrected TOF, which increases the risk of premature birth, miscarriage, low birth weights, postpartum hemorrhage, paradoxical embolism, thrombosis events, congestive heart failure, infective endocarditis, and arrhythmias in both the mother and their baby [7].

The risk of pregnancy adverse outcome is determined by the underlying cardiac diagnostic, ventricular and valve function, functional class, the presence of cyanosis condition, pulmonary artery pressures, comorbidities, and other factors. Pregnancy in women with cardiac disease is at increased risk of developing adverse maternal cardiac events which can usually be estimated after a complete cardiovascular examination. The use of a modified WHO classification (mWHO) that incorporates the published risk factors into four risk steps corresponding to a negligible risk of cardiovascular complications (WHO I) to prohibitively high risk of maternal morbidity and mortality or (WHO IV). Risk assessment could be enhanced further by considering predictors identified in large populations with various conditions, such as the CARDiac disease in PREgnancy, ZAHARA, and registry of pregnancy and cardiac disease Studies [8], [9].

Before surgical repair, patients with ToF reached childbearing age, and successful pregnancy was uncommon. Complications occur at a rate of 7% due to the hemodynamic burden of pregnancy mixed with remaining cardiovascular lesions following repair. The most serious complication is progressive right ventricle dilation and ventricular failure. Arrhythmia, intrauterine growth restriction, and spontaneous fetal demise can all occur as a result of inadequate maternal cardiac output. Thromboembolism can increase aortic root dilatation, and endocarditis is some of the other problems [2], [10].

Patients with uncorrected ToF should be postponed pregnancy after their marriage to improve the survival of young adults with CHD and need discussions regarding family planning before marriage and contraception individually. In this situation, contraception should be used after marriage in a woman with CHD routinely. Several factors are to be considered that associated directly with cardiac conditions, including the risk of thrombosis and embolism event with estrogen-containing products, risk of infection and vagal response when insert of an intrauterine device during the procedure, and maternal risk in the event of contraception failure. All contraception has a significant failure rate, therefore not an optimal procedure for the woman in whom pregnancy is the best avoided [2], [10].

Women with cyanotic CHD-associated risk for the thromboembolic event should avoid estrogen-containing contraception because can increase the risk of arterial and venous thrombosis events. The risk of estrogen-associated thromboembolism is further increased by hypertension, smoking, diabetes mellitus, and obesity. Furthermore, hormonal contraception has interaction with several drugs, such as Bosentan an endothelin antagonist for the treatment of pulmonary hypertension. Progesteron-only contraception is useful option for women with CHD, such as the Levonorgestrel intrauterine system, progesterone implant, and depot medroxyprogesterone acetate injection. Furthermore, the IUD is nonhormonal and can use as another choice for long acting contraception, but this agent leads to vaginal bleeding.

ToF is known to be related to abnormalities of coronary arteries. The most common anomaly

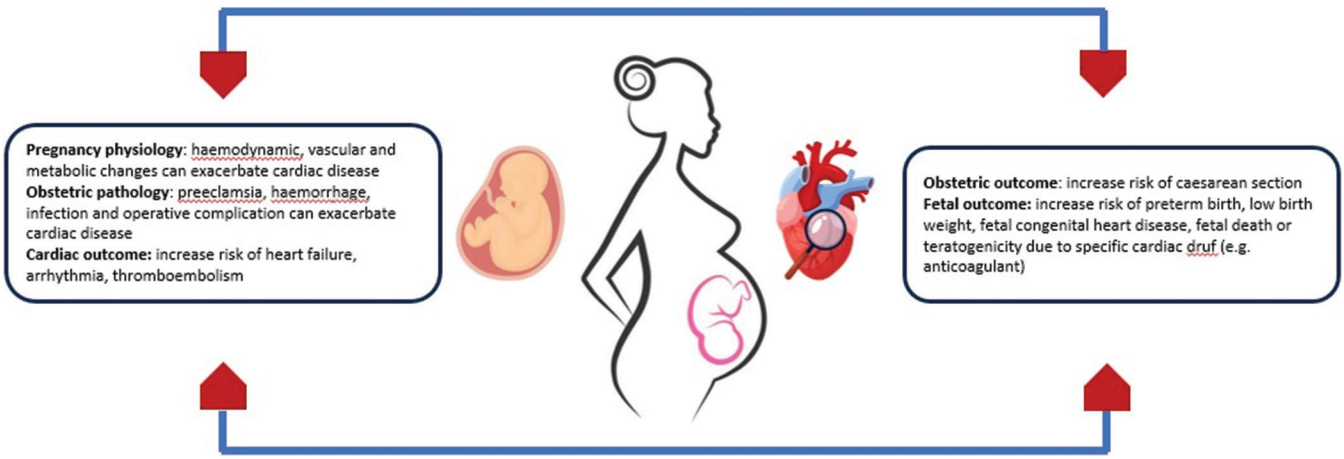


Figure 4: Hemodynamic during pregnancy [6]

of coronary arteries was found in an anterior descending artery from the right coronary artery or from the right coronary sinus. In our patient, she was diagnosed with ToF from the echocardiography and right heart catheterization showed ToF with dilatation of coronary artery and right aortic arch condition [5], [11].

Premature of birth is related to increase neonatal mortality and morbidity when compared with delivery at or beyond 39 comparable at a rate of 21%. Premature births are the most frequent neonatal complication in CHD-affected pregnancies, followed by SGA birthweight. Increased rates of newborn newborns admitted to intensive care units and SGA babies were the main causes of neonatal problems in the early-term group, followed by an increase in the prevalence of respiratory distress syndrome. The anesthetic treatment goals for the patient with an uncorrected ToF are to maintain SVR and prevent a decrease in peripheral vascular resistance because these alterations could potentially worsen the pre-existing right-to-left shunt [7], [12], [13].

A multidisciplinary approach involving obstetricians, cardiologists, anesthetists, intensivists, midwives, and neonatologists is essential. The circulation should be examined carefully, paying special attention to right ventricular dysfunction, pulmonary regurgitation, and right ventricular outflow tract obstruction. Early hospitalization for rest and treatment of related complications, as well as a delivery plan, should be decided upon. Preterm delivery should be taken into consideration if right ventricular failure occurs [14].

Forceps or ventouse extraction can be used during vaginal birth in combination with epidural anesthesia to shorten the second stage. General anesthesia might be necessary if the localized method fails or if a life-threatening emergency arises. Fontan techniques that have been modified have improved the long-term survival of patients with single ventricle physiology. The most frequent obstetric issues among pregnant women are preterm rupture of the membranes and premature labor. Fetal death and postpartum hemorrhage are also possible outcomes. At least 26% of pregnancies are complicated by arrhythmias, most frequently supraventricular, with atrial-pulmonary connections being the most common; the older kind of Fontan surgery [14], [15].

Conclusion

A case of pregnancy in a patient with uncorrected ToF has been discussed in this case. In patients with congenital heart abnormalities, pregnancy has a major impact on the hemodynamics of the heart and it leads to increase fatal outcomes. Due to the prevalence of pregnancy in patients with significant uncorrected CHD and the lack of evidence on the subject, preconception education and consultation in this population should be privileged for risk stratification to make decisions regarding management without optimal either obstetrical or medical management.

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