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Case Report

Successful Pregnancy in Uncorrected Tetralogy of Fallot with Right Aortic Arch: An Interdisciplinary Team Approach

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ABSTRACT

Background: Tetralogy of Fallot (ToF) is the most common cyanotic disorder, with a global prevalence of congenital heart disease nearly reaching 10%. The anomalies observed in individuals with ToF result from the anterior and cephalad displacement of the infundibular (outflow tract) component of the interventricular septum. ToF continues to be a significant contributor to maternal morbidity, mortality, and adverse outcomes in newborns with the condition.
Case presentation: An 18-year-old woman in her 28th week of pregnancy was referred to our tertiary referral center, Saiful Anwar Hospital, diagnosed with uncorrected Tetralogy of Fallot (TOF), which had been noticed since she was 2 years old. At that time, her parents refused treatment. She became pregnant unexpectedly and was then directed to RSSA for further management.
Conclusion: We present a successful case of pregnancy in a woman who has not undergone surgical repair for ToF. An interdisciplinary pregnancy heart team, including a cardiologist, obstetrician, anesthesiologist, midwives, and neonatologists at our tertiary hospital, developed a care plan based on an individualized assessment of the patient's comorbidities and ToF. The life expectancy of this pregnant woman and her baby is higher with a team approach. The choice of contraception and family planning should be part of the discussion and follow-up from the teenage years, allowing patients with ToF to control their fertility and make informed decisions about having children.

1. Introduction

Ten percent of all congenital heart diseases (CHD) are cyanotic, with Tetralogy of Fallot (ToF) being the most common cyanotic CHD. ToF remains the primary indirect cause of maternal mortality in pregnant women with CHD, especially those with uncorrected CHD from emerging countries.[1–3]

2. Case Report

An 18-year-old woman presented at 28 weeks of gestation and was referred to our center for further management. She was diagnosed with Tetralogy of Fallot (ToF) at the age of 2, and it remained uncorrected until now. At 16 years old, she experienced cyanosis and breathlessness. General examination revealed grade 4 cyanosis, clubbing, and 88% oxygen saturation by oximetry, along with a harsh systolic murmur at the upper left sternal border. Her X-ray showed a

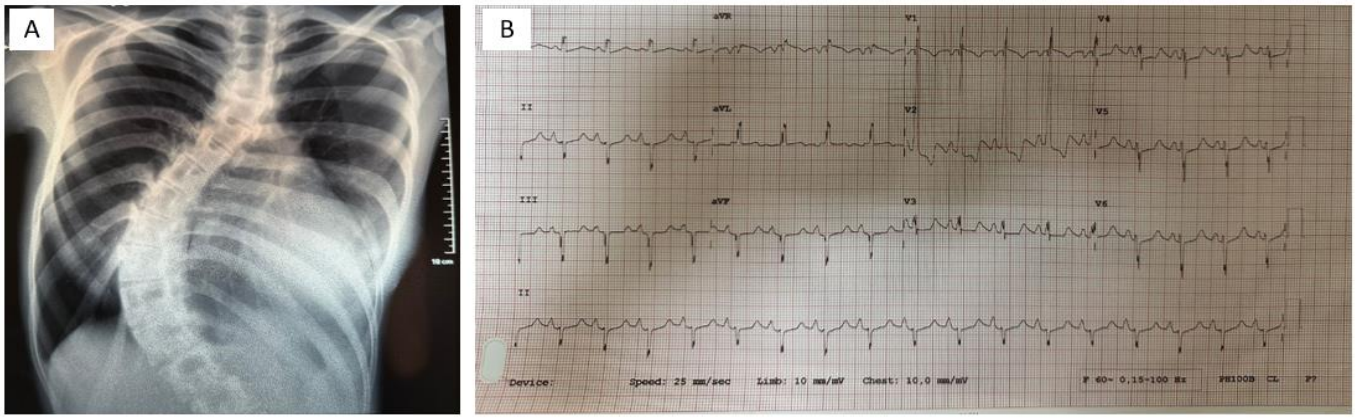

Figure 1 A. Chest X-ray anteroposterior projection showing cardiomegaly (boot shaped heart) and scoliosis; B. Electrocardiography

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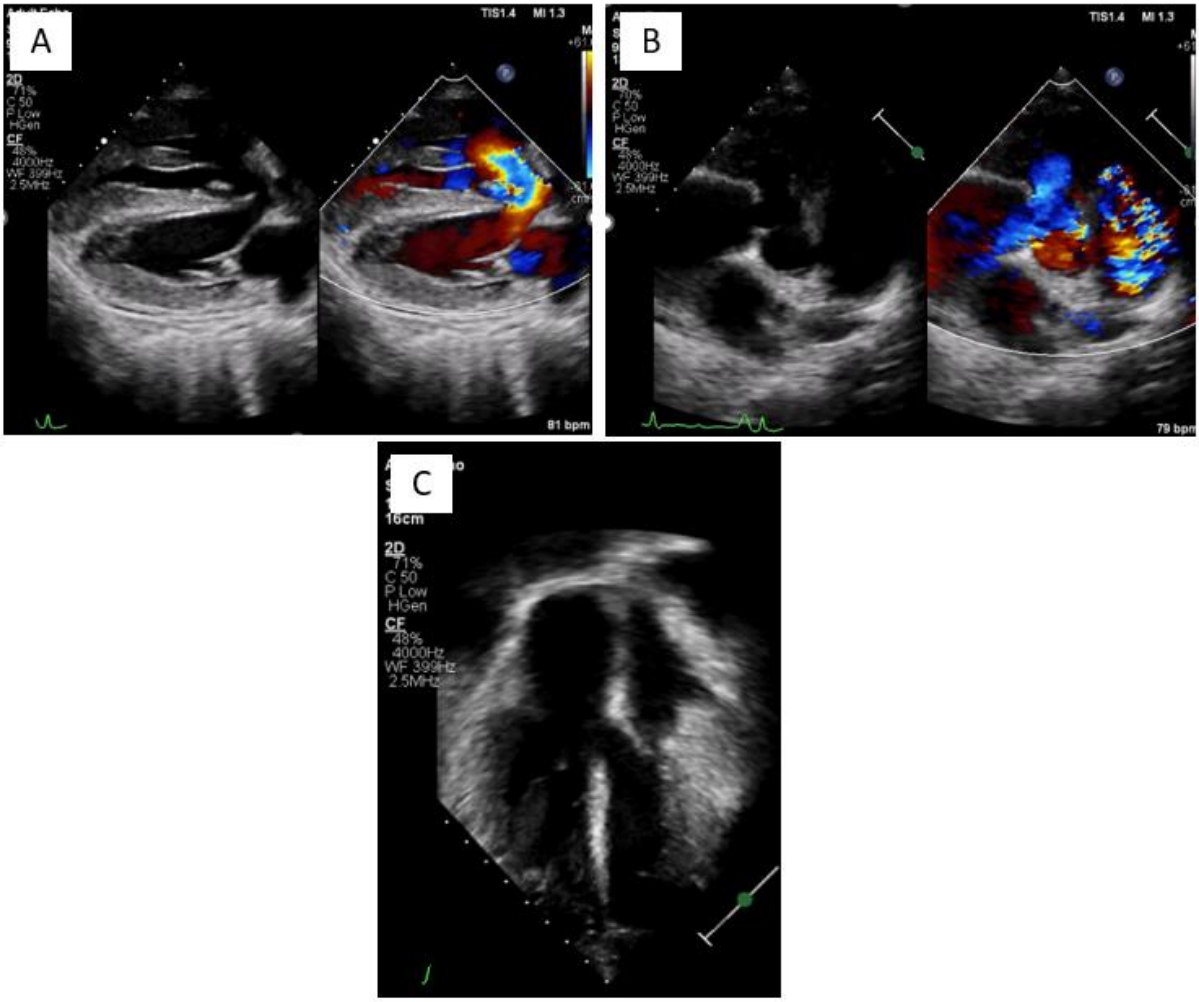

Figure 2. A. Echocardiography showing subaortic ventricular septal defect (VSD) with 50% overriding aorta; B. severe infundibular PS; C. RVH

boot-shaped heart and scoliosis (Figure 1A). Electrocardiography (ECG) revealed sinus rhythm, right axis deviation, a large amplitude of R wave at the anterior precordial lead, and right ventricular hypertrophy (RVH) (Figure 1B).

Transthoracic echocardiography showed RVH, a right-to-left shunt subaortic ventricular septal defect (VSD) with 50% overriding aorta, severe infundibular pulmonary stenosis (PS), and a right aortic arch (RAA). These features are suggestive of ToF with RAA (Figure 2). The patient was referred to the national heart center as the highest-level facility.

To confirm the diagnosis, she underwent cardiac CT, which revealed Tetralogy of Fallot (ToF) with a right aortic arch and coronary fistula. Based on the cardiac CT results, the patient was assessed for the coronary fistula using right heart catheterization (RHC) and stress echocardiography. Right heart catheterization showed ToF with the right aortic arch and dilatation of the coronary artery (Figure 3). Stress echocardiography revealed tardokinesia at the inferoseptal and anteroseptal segments, subaortic ventricular septal defect (VSD) with overriding aorta, infundibular deviation anterocephal, severe pulmonary stenosis (PS), good left ventricle function, and right ventricle contractility.

In our case, the patient was scheduled for ToF repair at the national heart center. While awaiting definitive treatment, she experienced an unintended pregnancy, and her obstetrician referred her to the cardiology department at our tertiary local hospital.

Upon examination at 28 weeks of pregnancy, her blood pressure was 120/70 mmHg, pulse rate was 80 per minute, and respiratory rate was 18 per minute. Oxygen saturation (SpO2) by pulse oximetry in both arms and lower extremities ranged from 89% to 91%. Echocardiography examination remained unchanged.

The patient was continuously monitored by a team consisting of an obstetrician, cardiologist, anesthesiologist, midwives, and neonatologists. The interdisciplinary team classified this patient with a high-risk score (mWHO IV and Zahara score 3.2) and recommended a cesarean section delivery at 32 weeks of pregnancy. The patient was admitted three days before delivery, and obstetric fetal ultrasonography showed small for gestational age (SGA) and oligohydramnion. Dexamethasone prophylaxis for the newborn was administered at 6 milligrams (mg) twice daily for up to two days, a 4-gram loading dose of magnesium sulfate followed by a 10-gram infusion drip for 24 hours as a neuroprotectant, and 300 mg Clindamycin injection twice daily. Blood analysis showed hemoglobin at 13.7 g/dL, white blood cells at 7370/μL, hematocrit at 40.1%, and platelets at 252,000/μL.

She had a cesarean section delivery with epidural anesthesia without complications. An IUD was inserted following the cesarean section. The newborn was a premature baby boy, weighing 1200 grams, measuring 37 cm in length, with a head circumference of 22 cm, an APGAR score of 6/8, breathing normally, and clear heart sounds. The patient and her baby were discharged for home treatment with recommendations for one week of cardiology and gynecological consultation.

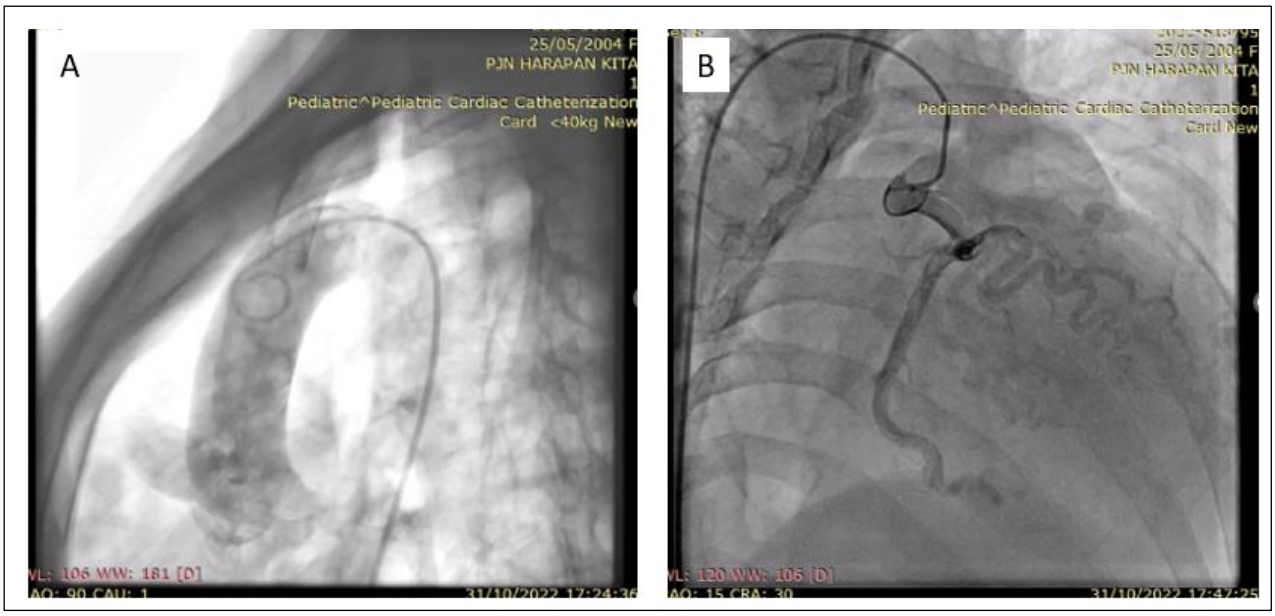

Figure 3. A. RHC showed right aortic arch; B. Dilatation of the coronary artery

2. Discussion

Unintended pregnancy is an unplanned or unwanted event occurring at any age in women, and adolescent girls and young women (15-24 years) account for 40% of such incidents worldwide. ToF, a serious form of cyanotic congenital heart disease (CHD), is characterized by hemodynamic abnormalities resulting from an anatomical anomaly and varying degrees of compromised cardiac function. Uncorrected ToF during pregnancy is an uncommon circumstance that increases the likelihood of unfavorable pregnancy outcomes and fetal growth abnormalities. Pregnant individuals with uncorrected ToF may experience worsening symptoms during pregnancy and delivery, significantly impacting maternal morbidity (62.5%), mortality (10%), and unfavorable fetal outcomes.

This case involves an 18-year-old woman with an unintended pregnancy and uncorrected ToF, presenting with complaints of shortness of breath during moderate activity since the third trimester of gestation.[4–6]

The application of a modified WHO classification (mWHO), which categorizes risk variables into four stages ranging from negligible risk of cardiovascular problems (WHO I) to prohibitively high risk of maternal death or severe morbidity (WHO IV), is one method of incorporating risk factors into published classifications. Utilizing predictors identified in large populations with various illnesses, such as the CARPREG (CARDiac disease in PREgnancy), ZAHARA, and ROPAC (Registry Of Pregnancy And Cardiac disease) Studies, could further enhance risk assessment. In our case, the pregnancy with uncorrected ToF, categorized as mWHO III and having a Zahara score of 3.2, posed a high risk.[7,8]

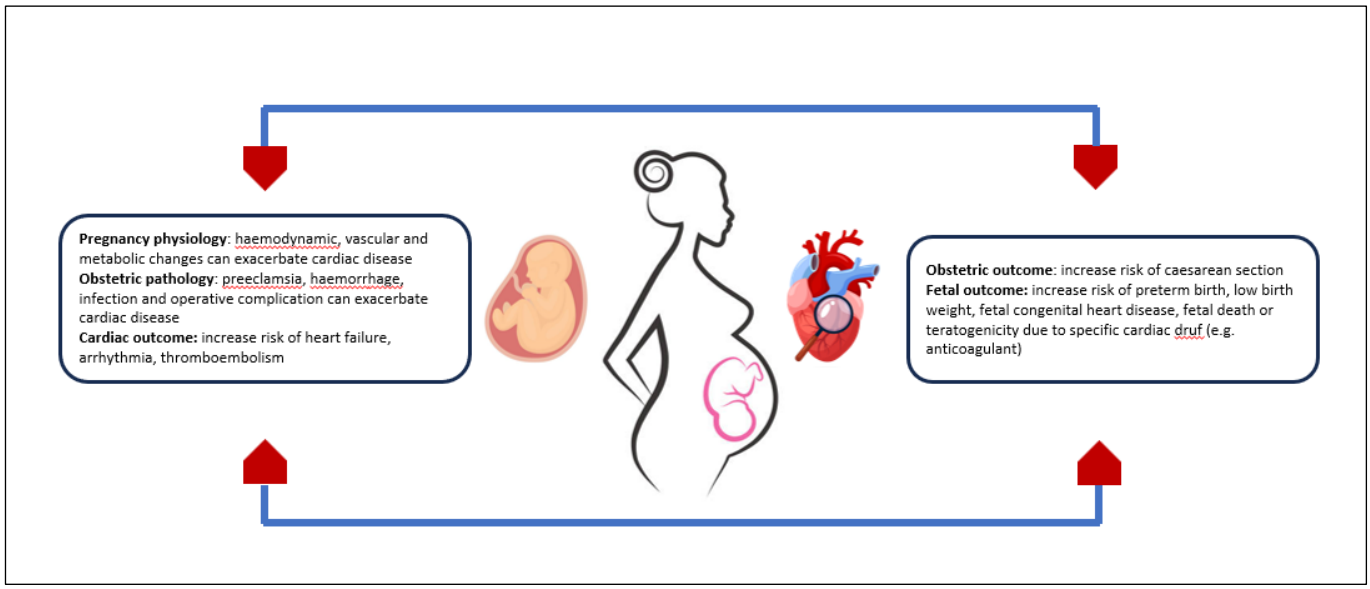

Figure 4. Hemodynamic during pregnancy.[6]

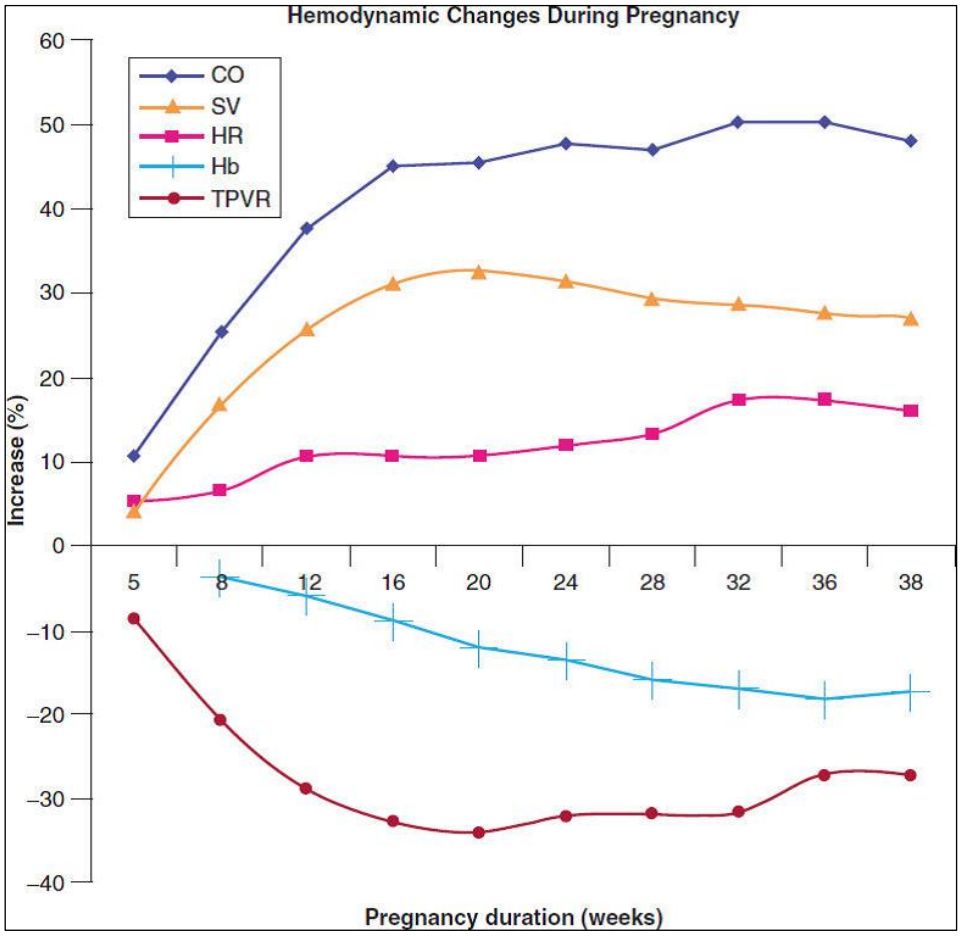

Figure 5. Hemodynamic changes during pregnancy.[9]

For the vast majority of women with CHD, vaginal delivery is the recommended method. This preference is based on the differing hemodynamic stresses experienced during vaginal delivery and cesarean section, which can lead to serious cardiovascular complications, including increased blood loss in the case of cesarean section. Premature births and small for gestational age (SGA) birthweights are the most common neonatal complications in pregnancies affected by CHD. The delivery date did not significantly impact the occurrence of obstetric issues associated with increased risks for newborns, such as hypertensive disorders of pregnancy (HDP), fetal growth restriction (FGR), and gestational diabetes. Full-term cohort patients were more likely to be categorized as mWHO class I, while those in the early-term cohort had a higher likelihood of being mWHO class II or above. Anesthetic treatment goals for patients with uncorrected ToF aim to maintain systemic vascular resistance (SVR) and prevent a decrease in peripheral vascular resistance, as these changes could potentially worsen the existing right-to-left shunt. Following interdisciplinary team discussion, she underwent elective cesarean section at 32 weeks of gestation due to the indication of SGA and oligohydramnios conditions.[10–12]

Patients with uncorrected ToF should delay pregnancy until after marriage to enhance the survival of young adults with CHD. Discussions about family planning and contraception should be initiated individually before marriage. Progestin-only contraception is a useful option for women with CHD. Additionally, the copper intrauterine device (IUD) is a nonhormonal alternative for long-acting contraception, although it may lead to vaginal bleeding. In our patient, based on interdisciplinary team recommendations, the use of a copper IUD (Copper T) was suggested after cesarean section and has been discussed with her family.[11,13]

3. Conclusion

We present a successful case of pregnancy in a woman with unrepaired ToF. The interdisciplinary pregnancy heart team, consisting of a cardiologist, obstetrician, anesthesiologist, midwives, and neonatologists, played a crucial role in managing this case. The life expectancy of both the pregnant woman and her baby is higher with a collaborative team approach.

4. Declaration

4.1 Ethics Approval and Consent to participate
Patient has provided written informed consent prior to involvement in the study.

4.2. Consent for publication
Not applicable.

4.3 Availibility of data and materials
Data used in our study were presented in the main text.

4.4 Competing interests
Not applicable.

4.5 Funding Source
Not applicable.

4.6 Authors contributions
Idea/concept: HIM. Design: VY. Control/supervision: VYSP. Data collection/processing: HIM Analysis/interpretation: HIM, VY. Literature review: HIM. Writing the article: VY. Critical review: VY. All authors have critically reviewed and approved the final draft and are possible for the content and similarity index of the manuscript.

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