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Uncomplicated Pregnancy with Residual Tetralogy of Fallot Post-Surgery

Ameliyat Sonrası Fallot Tetralojisi Rezidüleri ile Komplike Olmayan Gebelik

ABSTRACT

The most common cyanotic congenital heart disease (CHD) in adults is Tetralogy of Fallot (TOF), accounting for 10% of congenital heart anomalies and 75% of cyanotic CHD cases. It is a congenital heart disease that most often requires surgical correction within the first year of life. Corrective surgical treatment for TOF began in the 20th century, starting with a temporary shunt procedure and advancing to primary surgical repair. The current approach in infants often involves valve-sparing techniques. Following corrective surgery, more than 85% of patients with TOF can survive into adulthood. Without repair, however, patients with TOF rarely reach adulthood, and the prognosis for pregnant women with CHD is poor. Congenital heart disease is one of the leading causes of indirect maternal deaths. The literature indicates that pregnant patients with corrected TOF still face a higher risk compared to otherwise healthy women. According to the modified World Health Organization maternal cardiovascular risk classification, patients with repaired TOF have a mild risk of mortality and a moderate risk of morbidity, with the risk of maternal cardiac events ranging from 5.7% to 10.5%. Cardiac evaluations should be performed at least once during each trimester of pregnancy. In this case report, we discuss the pregnancy and successful, uncomplicated birth of a woman with TOF who underwent corrective surgery in childhood.

Keywords: Congenital heart disease, cyanosis, pregnancy, pulmonary stenosis, tetralogy of fallot

ÖZET

Erişkinlerde en sık siyanotik konjenital kalp hastalığı (KKH) fallot tetralojisidir (TOF). İnsanlardaki doğumsal kalp anomalilerinin %10'unu, siyanotik KKH vakalarının ise %75'ini TOF oluşturmaktadır. Hayatın ilk yılında en çok cerrahi düzeltme gerektiren doğumsal kalp hastalığıdır. Düzeltici cerrahi tedavi 20. yüzyıl itibariyle uygulanmaya başlanmıştır. Geçici şant işlemiyle başlayıp primer cerrahi onarım ile devam etmiştir. İnfantlarda güncel yaklaşım ise kapak koruyucu tekniklerdir. TOF için uygulanan düzeltici cerrahi ile hastaların %85'inden fazlası yetişkinliğe kadar hayatta kalabilmektedir. Onarım yapılmadığı takdirde TOF hastaları nadiren erişkin çağa ulaşır. Bu durumda hamile olan KKH'larında prognoz kötü seyretmekte ve dolaylı anne ölümlerinin önde gelen nedenleri arasında yerini almaktadır. Ancak onarıcı cerrahi geçiren kadınlarda gebelik sonuçları normal popülasyonla neredeyse aynıdır. Opere fallot, modifiye dünya sağlık örgütü (mDSÖ) maternal kardiyovasküler risk sınıflamasına göre; maternal risk açısından mortalitede hafif morbiditede orta dereceli riske sahiptir, maternal kardiyak olay riski %5,7-10,5 arasındadır. Gebelik boyunca her trimesterda en az bir kez kardiyak açıdan değerlendirilmelidir. Bizim vakamızda da çocukluk çağında düzeltici cerrahi geçirmiş olan TOF bir bireyin gebeliği ve komplikasyonsuz başarılı doğumu ele alınmaktadır.

Anahtar Kelimeler: Konjenital kalp hastalığı, siyanoz, gebelik, pulmoner darlık, fallot tetralojisi

CASE REPORT
OLGU SUNUMU

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Tetralogy of Fallot (TOF) is the most prevalent form of cyanotic congenital heart disease and represents a milestone in the successful surgical repair of congenital heart defects. Tetralogy of Fallot manifests through four primary components: ventricular septal defect (VSD), straddling of the aorta over the ventricular septum (with more than 50% of the aorta positioned above the right ventricle), right ventricular hypertrophy, and obstruction of the right ventricular outflow tract.[1] The clinical trajectory of TOF depends on the severity of right ventricular outflow tract stenosis, with patients experiencing severe obstruction facing central cyanosis and clubbing.[2] Untreated TOF is associated with increased maternal mortality during pregnancy due to hemodynamic changes, including increased plasma volume and cardiac output, as well

as decreased systemic vascular resistance, which elevate the risk of cardiovascular complications during gestation.[3] According to the modified World Health Organization maternal cardiovascular risk classification, women with repaired TOF face a mild risk of mortality and a moderate risk of morbidity during pregnancy. The likelihood of maternal cardiac events ranges from 5.7% to 10.5%. It is recommended that cardiac evaluations be conducted at least once per trimester throughout pregnancy.

Case Report

A 23-year-old primigravida female presented to our clinic for a cardiac assessment prior to undergoing a cesarean section. After a detailed examination, it was revealed that the patient had a history of corrective surgeries for TOF at an external center when she was 6, 9, and 14 years old.

In the first operation, at six years old, a conduit without a valve was placed in the right ventricular outflow tract (RVOT) between the infundibulum and the main pulmonary artery using an external patch. At nine years old, the RVOT grafts were obliterated, and a Modified Blalock-Taussig-Thomas Shunt was performed through a left posterolateral incision. In the final operation, at 14 years old, a redo RVOT procedure was performed, and the native pulmonary valve was dilated using bougies. After this operation, the cardiology council recommended a future procedure for complete recovery, which would include closure of the VSD once the pulmonary flow bed was suitable for the intervention. However, no additional operations were performed in the patient's history. The patient has no known additional diseases. She does not present any cardiac symptoms and is classified as New York Heart Association (NYHA) class I. On physical examination, lung sounds were normal, and no cyanosis or other abnormalities were observed. During auscultation, a 5/6 pansystolic murmur between the left 3rd and 4th intercostal spaces and a doubled second heart sound (S2) were noted.

The patient's blood pressure was 100/60 mmHg, her pulse rate was 100 beats per minute, and her oxygen saturation (SpO2) was 92%. A 12-lead electrocardiogram showed a sinus rhythm at 105 beats per minute with T-wave inversion in lead aVF. Echocardiography revealed an ejection fraction of 60%, right ventricular hypertrophy, mild aortic regurgitation, and straddling of the aorta (Figure 1). Additionally, a 2.0 cm defect in the subaortic part of the ventricular septum with a left-to-right shunt was observed (Figures 2 and 3). The pulmonary valves appeared fibrotic and thickened, with a maximal gradient of 74 mmHg and a mean gradient of 50 mmHg (Figure 4). Mild tricuspid regurgitation was present, with right ventricular systolic pressure calculated at 78 mmHg based on the tricuspid regurgitation jet (Figure 5).

ABBREVIATIONS

CHD	Congenital heart disease
NYHA	New York Heart Association
RVOT	Right ventricular outflow tract
SpO2	Oxygen saturation
TOF	Tetralogy of Fallot
VSD	Ventricular septal defect

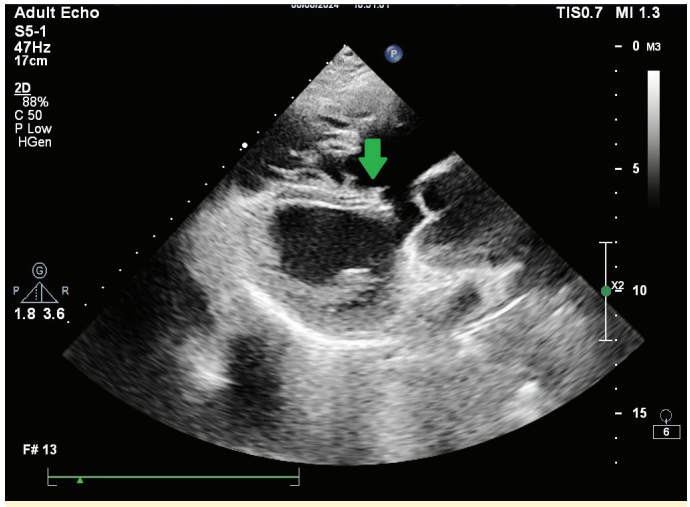

Figure 1. Echocardiography showing straddling of the aorta.

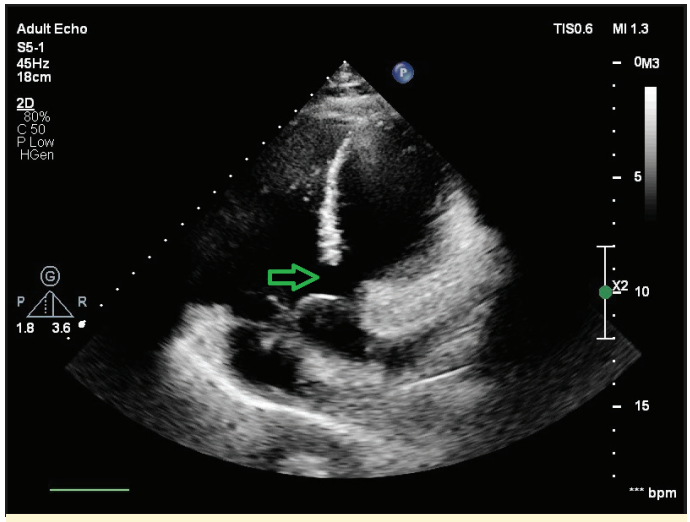

Figure 2. Echocardiography showing a 2.0 cm ventricular septal defect (VSD) located in the subaortic region.

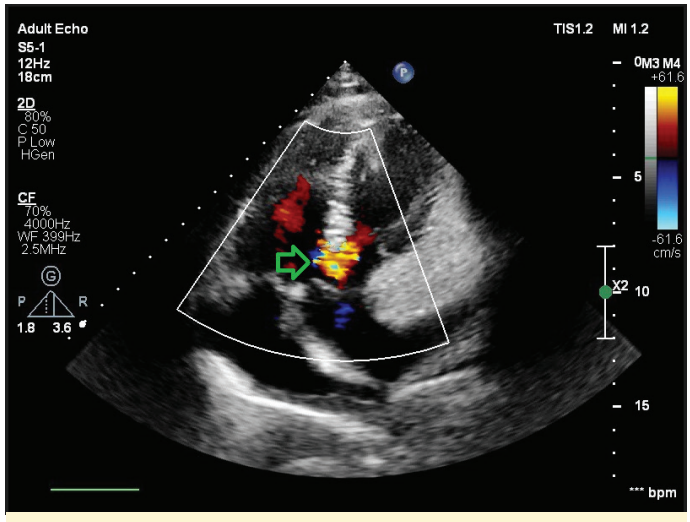

Figure 3. Doppler echocardiography demonstrating right-to-left shunts.

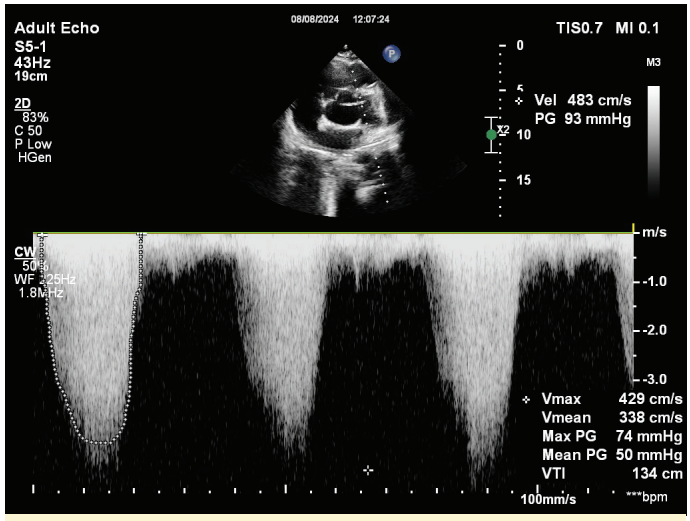

Figure 4. Echocardiography showing fibrotic and thickened pulmonary valves, with a maximal gradient of 74 mmHg and a mean gradient of 50 mmHg.

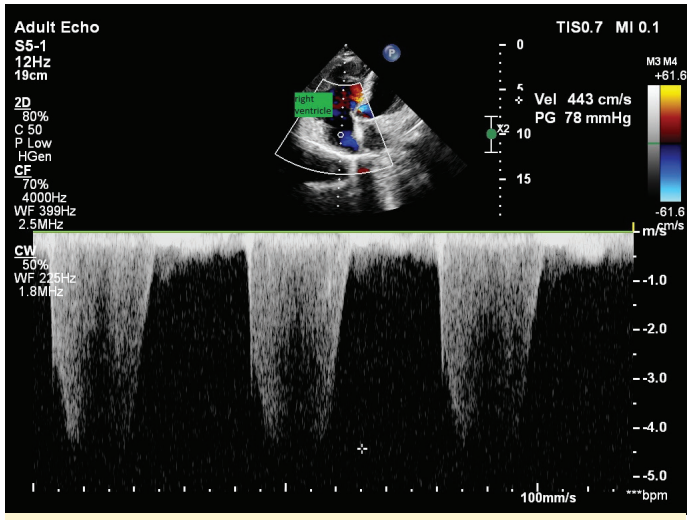

Figure 5. Echocardiography showing mild tricuspid regurgitation, with a calculated right ventricular pressure of 78 mmHg based on the tricuspid regurgitation jet.

Her blood test results were normal. Obstetric examination revealed one live cephalic fetus with a gestational age of 31 weeks. Based on these findings, a multidisciplinary team of cardiologists, cardiovascular surgeons, and obstetricians conducted a pre-partum assessment. Following the obstetric examination, it was determined that the fetus was mature and there were no contraindications for delivery. A cesarean section was recommended due to the time and effort required for vaginal delivery. Additionally, it was advised to monitor for severe bleeding and acute hypotension during the surgery, with the administration of antibiotics prior to the procedure. A 1,960 g healthy female baby was born two weeks later. During the surgery, 300 cc of blood loss was observed, and the patient was monitored in the intensive care unit post-delivery. The patient was discharged from the hospital after four days. She did not report any cardiac symptoms, and no additional pathology was identified. The patient was prescribed 100 mg of acetylsalicylic acid and advised to attend a follow-up visit one month later.

Discussion

Advancements in surgical techniques and growing experience have enabled most young women who undergo surgical repair for TOF to survive into their reproductive years. Tetralogy of Fallot is associated with cardiac and obstetric complications during pregnancy, as well as increased maternal and fetal mortality, particularly in uncorrected patients. Even in women with repaired TOF, the risk of complications is higher than in healthy pregnancies, and morbidity is more common.[1]

The hemodynamic changes during pregnancy can lead to clinical deterioration by increasing the right-to-left shunt and exacerbating cyanosis. These volume shifts become more pronounced during delivery, and sudden blood loss can further intensify these effects.[2]

Adverse cardiovascular events may result from conditions such as right ventricular dysfunction, severe pulmonary hypertension, and significant pulmonary regurgitation accompanied by right ventricular dysfunction. Studies have shown that abnormal uteroplacental Doppler flow (UDF) correlates with parameters of right ventricular function, suggesting that maternal cardiac dysfunction may impair placental function or perfusion, thereby increasing the risk of obstetric and neonatal complications.[1] Reduced maternal cardiac output may lead to intrauterine growth retardation, which can occur even after corrective surgery. Adverse prognostic indicators in TOF include maternal hematocrit levels exceeding 60%, oxygen saturation below 80%, and a history of syncopal episodes. Pregnancy in these individuals poses a heightened risk of spontaneous abortion, preterm delivery, and low birth weight. Predictors of maternal morbidity may include functional status, ventricular dysfunction, significant arrhythmias, cyanosis, severe outflow tract obstruction, pulmonary hypertension, and the need for anticoagulation.[2] The use of anticoagulants during childbirth may increase the risk of maternal and fetal bleeding, contributing to complications such as spontaneous abortion, intrauterine growth restriction, prematurity, perinatal mortality, and congenital heart disease.[3] Conversely, residual shunting, right ventricular insufficiency, and pulmonary hypertension are risk factors for maternal and fetal morbidity in patients with repaired TOF. According to the literature, approximately 40% of women with TOF may develop heart failure during pregnancy, with maternal mortality rates reaching up to 10%. Additionally, fetal mortality rates as high as 36% have been reported in cases of uncorrected TOF.[4] The severity of these complications directly correlates with the degree of cyanosis in untreated TOF patients, who exhibit a higher incidence of fetal growth restriction, preterm birth, and lower birth weight compared to corrected cases.[3]

Surgical correction during pregnancy is not recommended due to the high rates of perinatal and maternal mortality. If surgery is necessary, it is usually considered safer after the 16th week of gestation.

Given the potential complications associated with pregnancy, women with TOF should receive thorough counseling before

conception. During preconception counseling, it is essential to evaluate risk factors, and ideally, any cardiac defects should be surgically corrected before pregnancy. Antenatal care should involve a collaborative approach between an obstetrician, cardiologist, perinatologist, and anesthetist. These patients require not only routine antenatal care but also more frequent examinations, restricted physical activity, iron supplementation, periodic arterial blood gas analysis, and close fetal monitoring. A planned delivery is crucial to optimize intrapartum management.[2] Excessive bleeding during delivery may exacerbate right-to-left shunting and increase cyanosis due to the elevated cardiac workload. The mode of delivery is a widely debated topic, with varying opinions in the literature. According to some studies, due to the high risk of complications, women with cardiac disease may be safer with a Cesarean section to avoid prolonged labor.[1,3] Conversely, other studies suggest that the majority of women with TOF should ideally have a vaginal delivery. Those without heart failure, ventricular arrhythmias, or obstetric indications should not routinely be scheduled for a Cesarean section.[2,4,5] Antibiotic prophylaxis is recommended before labor and Cesarean section to prevent infections. It is crucial to avoid hypotensive medications during anesthesia.[2,4] The second stage of labor should be shortened using instrumental delivery.[2,4]

To minimize thrombotic complications, postpartum thromboprophylaxis is given to all women with uncorrected lesions. However, its use remains controversial and is more commonly recommended for women with cyanosis and/or polycythemia due to their elevated risk of thromboembolic episodes.[3]

In this case, a successful Cesarean delivery was achieved for a patient at high risk of maternal mortality due to residual pulmonary stenosis. This outcome was facilitated through comprehensive evaluation, treatment, and follow-up by a multidisciplinary team comprising cardiology, obstetrics, cardiovascular surgery, and anesthesia. No complications were observed in either the mother or the baby during the follow-up period. A multidisciplinary approach is essential to reduce mortality and morbidity risks in pregnant women with TOF, especially in high-risk cases. This involves integrating evaluations of anatomy, pathophysiology, and clinical findings to guide personalized treatment and ensure vigilant monitoring.

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