Obstetric Outcomes in Women with Unrepaired Tetralogy of Fallot: A Case Report

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Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart diseases and without corrective surgery, natural survival rate into the fourth decade was only about 3%. In pregnant patients with unrepaired TOF, pregnancy-related physiological changes is more difficult to tolerate compared to healthy pregnant woman. Discussed below a case of term pregnancy in a 20- year old woman with unrepaired TOF, was diagnosed at delivery.

Key Words: Obstetric outcomes, Pregnancy, Unrepaired tetralogy of Falllot.

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Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect and comprises 75% of cyanotic cardiac diseases.[1] Half of patients with uncorrected TOF will die before age 25 [2] due to ventricular arrythmia, cerebrovascular events, endocarditis and congestive heart failure or hypoxia.[3]

In this case report, we present a 20 year old woman with 5 consecutive first-trimester pregnancy losses who was diagnosed with TOF for the first time after caesarean section.

Case Report

Twenty-year old woman who had 5 consecutive first-trimester pregnancy losses was admitted to emergency room with diagnosis of early membrane rupture. Gestational age was 38 weeks due to last menstrual period. Her blood pressure was measured as 150/80 mmHg. The patient stated that she did not received antenatal follow-up. Obstetric ultrasound revealed a single viable fetus whose biometric measurements were accordant with 33 weeks' gestation. Amniotic fluid index was 60 mm and estimated fetal weight was 1800 gr. Cervical dilatation was 1 cm and effacement was 40%. Following initial evaluation, patient was admitted to delivery ward for further.

Routine blood tests revealed a hematocrit value of 48.7. Liver function tests were found in normal range and 200 mg/dl proteinuria was found in urine analysis. During fetal monitorization, late decelerations were observed and delivery was performed via caesarean section.

Intra- and early postoperative oxygen saturation was measured below 80% and central cyanosis was observed. Cardiological evaluation revealed second degree systolo-diastolic murmur and an increased cardiothoracic ratio and minimally prominent pulmonary conus in chest radiogram. Arterial blood gas measurements showed that p_{CO2}: 30.7 mmHg, p_{O2}: 46.5 mmHg and pH:7.4. Sinusoidal rhythm, right axial deviation and signs of right ventricular hypertrophy was found on electrocardiogram (Figure 1).

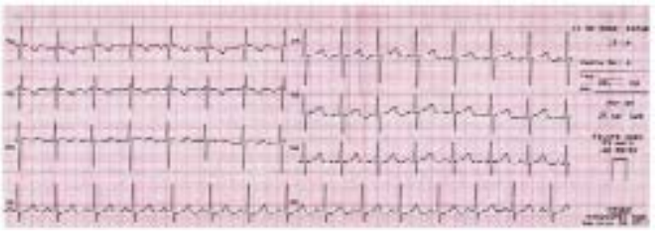

Figure 1: Patients electrocardiogram showing sinusoidal rhythm, right axial deviation and signs of right ventricular hypertrophy.

The patient was consulted to the cardiology department for further evaluation. Diagnosis of TOF was verified by echocardiography (Figure 2). A concomitant patent ductus arteriosus functioning as a compensatory arterio-venous communication by providing pulmonary blood was also demonstrated. In

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early postpartum period, supraventricular tachycardia was noted, and digoxin therapy controlled the heart rate. There is no other adverse maternal cardiovascular events.

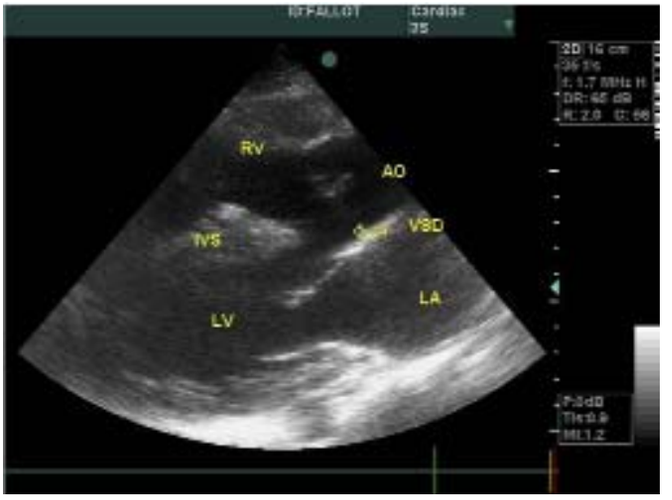

Figure 2: Echocardiographic findings of patient: Ventricular septal defect, overriding aorta, pulmonary artery atresia, and right ventricular hypertrophy.

The male infant weighed 1900 gr and had Apgar scores of 6 and 7 at 1 and 5 mins, respectively. Infant was admitted to neonatal intensive care unit and was given nasal continuous positive airway pressure for 3 days and supplemental oxygen for another.

No congenital cardiac anomaly was identified in infant.

Discussion

Uncorrected TOF is related with 4% maternal and 30% fetal mortality due to hypoxia[3] and incidence of congenital heart disease in newborn is between 1.1%[4] and 14%.[5] Pregnancy related changes such as decreased peripheral vascular resistance, increased blood volume and venous blood return will worsen right-to-left shunt.[6] This leads to further cyanosis, a compensatory rise in hematocrit, and a corresponding decrease in arterial oxygen saturation and risk of cardiac failure will be increased in these cases. A poor prognosis exists for patients whose shunting is of such a degree as to result in a hematocrit 60% or more or an arterial oxygen saturation of less than 80%.[6] In TOF cases pulmonary artery abnormalities, particularly in the presence of pulmonary regurgitation, may adversely affect augmentation of maternal cardiac output and this may result in depressed placental blood flow and, subsequently, intrauterine growth retardation.[7]

In a study of 144 pregnancies of which 74 mothers had congenital heart disease, found that 24 out of these had cyanosis and 9 had previously known TOF (2 uncorrected). Among women with cyanosis, rate of spontaneous abortion was 19.4% and perinatal mortality was 13%.[8] Patients with unrepaired TOF had smaller infants than the patients with repaired TOF.

From the cardiovascular standpoint, vaginal delivery is preferred for most TOF patients. But in our case, delivery with caesarean section itself was first step leading us to diagnosis because continous intraoperative monitorization of oxygen saturation was possible. A monitorization would not be performed if she had delivered via the vaginal route because lack of clinical evidence pointing out a cardiac disease and diagnosis would be overlooked.

It is well known that patients with TOF who do not undergo operation in childhood have short survival, which depends predominantly on the degree of pulmonary artery stenosis and early development of collateral circulation to the lungs. Long-term persistence of natural aortopulmonary anastomosis with systemic collateral circulation to the lungs and remodeling of the heart, with better hemodynamic balance as well as the presence of pulmonary atresia probably enhanced the survival of our patient.

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Düzeltme Operasyonu Uygulanmamış Fallot Tetralojili Gebelerde Obstetrik Sonuçlar: Olgu Sunumu

Fallot tetralojisi erişkin yaşta en sık görülen siyanotik konjenital kalp hastalığıdır. Klasik olarak pulmoner stenoz, ventriküler septal defekt, sağ ventrikül hipertrofisi, aorta dekstropozisyonunu kapsayan dört ana bulgusu vardır. Düzeltme operasyonu uygulanmamış olgularda ortalama yaşam süresi 12 yıldır ve olguların sadece %3'ü yaşamın 40'lı yıllarına kadar yaşayabilir. Gebelikte gerek hormonal faktörler, gerekse kan volümü ve kardiyak outputdaki artış gibi hemodinamik değişikliklerin etkisiyle taşiaritmilerin insidansında artış gözlenir. Sağlıklı gebelerde bu taşiaritmi atakları çoğunlukla antiaritmik tedaviye gereksinim göstermezken Fallot tetralojisi gibi altta yatan kardiyak hastalığı olan gebelerde ciddi hemodinamik bozukluklar görülebilir. Bu çalışmada herhangi bir düzeltme operasyonu uygulanmamış olan ve doğum anında tanı alan bir 20 yaşında term gebelik olgusu sunulmaktadır.

Anahtar Kelimeler: Obstetrik sonuçlar, Gebelik, Düzeltilmemiş fallot tetralojisi

References

1. Campbell M. Natural history of cyanotic malformations and comparison of all common cardiac malformations. Br Heart J 1972;34:3-8.
2. Ikeda M., Hirosawa K. Tetralogy of Fallot. Circulation 1968; 38: (suppl. V):21.
3. Warnes CA, Elkayam U. Congenital Heart Disease and

Pregnancy. In: Elkayam U, Gleicher N, editors, Cardiac Problems in Pregnancy. 3rd edn. New York: Wiley- Liss Press, 1998, pp 39-54.

4. Whittemore R, Hobbins JC, Engle MA. Pregnancy and its outcome in the woman with and without surgical treatment of congenital disease.Am J Cardiol 1982;50:641-51.
5. Meyer EC, Tulsky AS, Sigmann P, Silber EN. Pregnancy in the presence of tetralogy of fallot: observations on two patients.Am J Cardiol 1964;14:874-9.
6. Davies GAL, Herbert WNP. Congenital Heart Disease in Pregnancy. J Obstet Gynaecol Can 2007;29(5):409-14.
7. Veldtman GR, Connolly HM, Grogan M, Ammash NM, Warnes CA. Outcomes of pregnancy in women with tetralogy of fallot. J Am Coll Cardiol 2004;44;174-180.
8. Shime J, Mocarski EJ, Hastings D, Webb GD, McLaughlin PR. Congenital Heart Disease in Pregnancy: short- and long-term implications: published erratum appears in Am J Obstet Gynecol 1987;156:313-22.