

## Tetralogy of Fallot May Escape Attention Even in Pregnancy: a Late Presenting Case of a Multiparous Woman

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### Abstract

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease. A 47-year-old woman presented to our clinic with cyanosis and dyspnea on exertion. She had previously had three successful and healthy pregnancies. From the results of laboratory and imaging techniques a diagnosis of tetralogy of Fallot was made and the patient was referred to the cardiovascular surgery for the repairing operation. Here, we report a case of tetralogy of Fallot in a multiparous woman. (*JAEM 2014; 13: 36-8*)

**Key words:** Tetralogy of Fallot, pregnancy, congenital heart disease

### Introduction

Tetralogy of Fallot is the most common cyanotic heart defect after infancy and is characterized by a large ventricular septal defect, aorta overriding the interventricular septum, right ventricular outflow obstruction, and right ventricular hypertrophy. Survival into late adulthood of patients with unoperated TOF is rare. Also, TOF in pregnancy is very rarely seen in pregnancy for several reasons, because most patients undergo surgical repair early in life. Patients with repaired surgery tolerate pregnancy well. Unrepaired TOF pregnancy presents serious risks, including maternal and fetal mortality (1).

### Case Presentation

A 47-year-old woman presented to our clinic with cyanosis and dyspnea on exertion. She described a history of progressive shortness of breath on exertion in the previous two years. Three months earlier the patient's condition deteriorated and she described dyspnea on mild exertion. She had no previous history of diabetes mellitus, hypertension, usage of any medication and smoking. She had three successful and healthy pregnancies. Children were of normal weights. She had no significant complaint after or during pregnancies. Previous deliveries were vaginal in the home environment monitored by an obstetrician. On physical examination, the patient appeared to be in moderate respiratory distress. Her functional capacity was New York Heart Association (NYHA) class III. She was cy-

anic. Her heart rate was 66 beats per minute and regular; her blood pressure was 100/60 mmHg. The respiratory rate was 35 breaths per minute, with an oxygen saturation of 84% while she was breathing ambient air. A cardiovascular examination revealed a right ventricular impulse and systolic thrill at the left sternal border. Respiratory examination was normal. There was no peripheral edema.

The hemoglobin level was 20.7 g/dL and hematocrit was 59.8%. Biochemical investigation was within normal limits. An initial electrocardiogram (ECG) showed normal sinus rhythm with 66 beats per minute and an incomplete right bundle branch block with biventricular enlargement signs and right axis deviation. Echocardiography was performed and revealed a ventricular septal defect of 1.5 cm, which extended from the membranous septum, 50% aortic override, an obstruction with an right ventricular outflow tract gradient of 80 mmHg, RV hypertrophy (13 mm) and dilation (50 mm) with normal systolic function at end-diastolic phase (Figure 1). RV systolic motion was measured as 15.4 cm/sec in the right ventricular wall. The right atrium was dilated (49 mm) and a moderate to severe tricuspid regurgitation was present. The left atrium was dilated (43 mm) and the left ventricle was of normal dimensions. An estimated LV ejection fraction measured with modified Simpson method was 60%. Cardiac magnetic resonance imaging (MRI) was consistent with a diagnosis of uncorrected tetralogy of Fallot (Figure 2).

Cardiac catheterisation was performed and demonstrated a normal LV function and coronary arterial system. The RV systolic pres-



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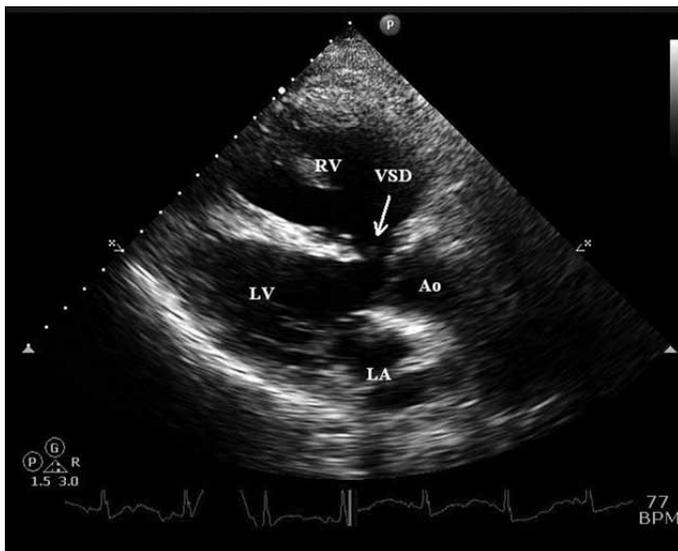
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sure was approximately 96 mmHg. In room air, the arterial saturation was 84% with a pulmonary arterial (PA) saturation of 64%. The peak gradient across the pulmonary valve/RVOT was 80 mm Hg. The patient was consulted to have cardiovascular surgery and was referred to the complete TOF repairing operation. Total correction of TOF was performed. Subsequently she was DOIng well and was discharged from hospital on her 5<sup>th</sup> postoperation day.

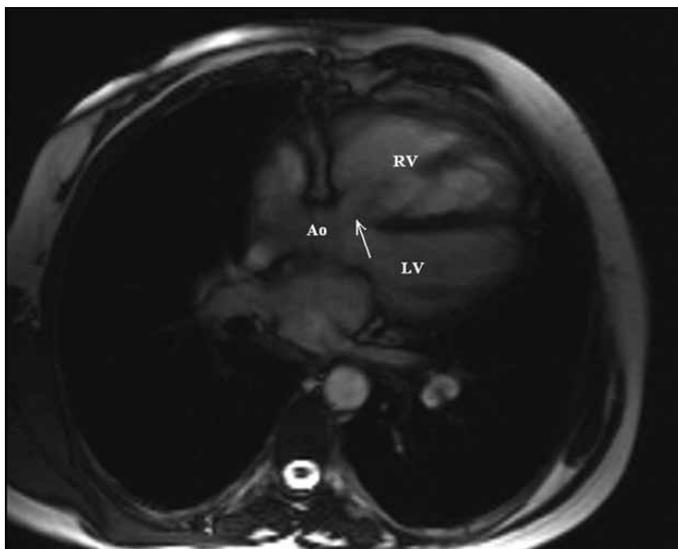
## Discussion

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect after infancy and approximately 35% of patients with untreated TOF die during the first year of life, and 75% die by age ten (1).



**Figure 1.** Parasternal long axis view of tetralogy of Fallot. Ascending aorta is overriding the interventricular septum. A large ventricular septal defect (VSD) is indicated by an arrow.

RV, right ventricle; LV, left ventricle; LA, left atrium; Ao, aorta; VSD, ventricular septal defect



**Figure 2.** Cardiac magnetic resonance imaging frame from a long axis shows a large aortic root overlying the ventricular septal defect (arrow). Right ventricle (RV) is enlarged and trabeculated

There are some case reports showing unoperated TOF in elderly (2-6). Adults develop progressive dyspnea and cyanosis. The severity of cyanosis is related to the degree of the right-to-left shunting. Transthoracic echocardiography usually provides a comprehensive description of the intracardiac anatomy, and physiologic information for repair of TOF (7). Especially for emergency physicians, the diagnosis of right ventricular outflow obstruction is required in the Emergency Department.

The distinctive and outstanding feature of this case includes the rare association of multiparous pregnancy with cyanotic congenital heart disease (i.e. uncorrected tetralogy of Fallot). The patient had remained asymptomatic for much of her life in the course of unrepaired TOF. To our knowledge, this is the first case of uncorrected TOF presenting after multiparous pregnancy in our country. When uncorrected, TOF pregnancy presents serious risks, including maternal and fetal mortality (8). Successful pregnancy in women with uncorrected TOF is most closely related to the degree of cyanosis (9). The hemodynamic changes during pregnancy may affect the course of the tetralogy of Fallot by increasing cardiac output and reducing systemic vascular resistance and blood pressure. These changes may worsen the patient's hemodynamic status but some instances, such as the presence of an extracardiac shunt, like patent ductus arteriosus (PDA), or systemic to pulmonary artery shunts, which contribute to blood supply to pulmonary circulation, and development of left ventricular hypertrophy may act as balancing factors against the right to left shunt (10). In our patient, cyanosis developed gradually. The potential factor contributing to longevity in this case is probably gradual development of the right ventricular outflow tract obstruction over a long period time.

It is well known that pregnancy in cyanotic congenital heart diseases carries a high incidence of miscarriages, premature births, and low birth weights. The incidence of miscarriages and fetal and maternal deaths is increasing in women with uncorrected congenital heart defects. There are two case reports presenting undiagnosed or uncorrected TOF in pregnant women (11, 12). Nevertheless, women with surgically corrected TOF are now able to have more successful pregnancies. This feature was demonstrated in the trials. Meijer et al. determined the risk of pregnancy in women with surgically corrected tetralogy of Fallot (TOF) and found that all women with a corrected TOF can lead their lives without restrictions and the pregnancy is well tolerated (13).

In conclusion, this case highlights the fact that patients with unrepaired TOF may still present in our clinics and challenge health care providers. Patients with TOF in pregnancy need special care by a team consisting of an obstetrician, cardiologist, and cardiovascular surgeon. Although our patient has not been under multidisciplinary assessment, there is a strong need for carefully coordinated multidisciplinary efforts to maintain a long-term follow-up in these women.

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