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Tetralogy of Fallot - A Case Report

Atufa Tahur[1], Omera Begum[2], Dr. Sujala Aakaram[3]

[1]Pharm.D (Intern), Sarojini Naidu Vanita Pharmacy Maha Vidyalaya , Secunderabad, Mareedpally, Telangana 500017, India

[2]Pharm.D (Intern), Sarojini Naidu Vanita Pharmacy Maha Vidyalaya , Secunderabad, Mareedpally, Telangana 500017, India

[3]Assistant Professor, Sarojini Naidu Vanita Pharmacy Maha Vidyalaya , Secunderabad, Mareedpally, Telangana 500017, India

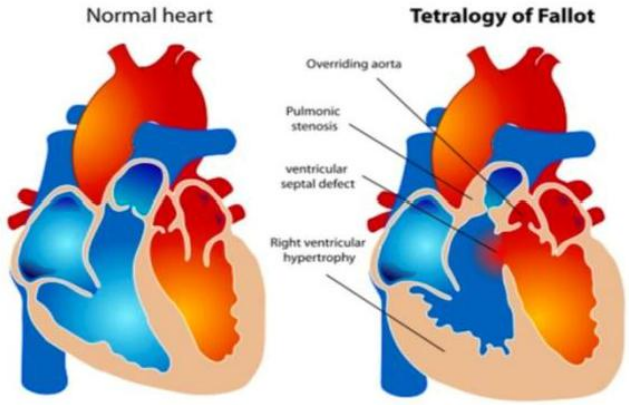
Corresponding Author Email: *atufatahur1234[at]gmail.com*

Abstract: ***TOF is a cardiac abnormality consisting of a tetrad of heart defects (Ventricular Septal Defect, Pulmonary Stenosis, Overriding Aorta and Right Ventricular Hypertrophy).Various diagnostic test to confirm TOF includes chest x-ray, echocardiogram, ECG, pulse oximetry test, MRI, Cardiac catheterization. In this case the patient was reported with complains of sudden onset of shortness of breath (SOB) along with sweating since 1 year. The confirmatory test of TOF in this patient was ECG, 2D echo, CT-coronary angio. Post confirmation of TOF, the patient underwent an uneventful ICR (Intra Cardiac Repair) surgery and hence the overall QoL of patient has been improved.***

Keywords: TOF, ICR, tetrad of heart defects, shortness of breath, sweating.

1. Introduction

Tetralogy of fallot is a cyanotic congenital heart disease, comprises of four different heart defects as, ventricular septal defect (VSD), right ventricular outflow tract (RVOT) obstruction, overriding aorta and right ventricular hypertrophy.[1]



- **Ventricular septal defect** - is a defect or hole in the septum which separates the ventricles of the heart. A septum is a barrier that prevents the mixing of blood from both the sides of ventricles.[2]
- **Right ventricular outflow tract obstruction** - is the narrowing of the pulmonary valve which obstructs the blood outflow from the right ventricles to the lungs (pulmonary stenosis).
- **Overriding aorta** - in TOF, the aorta is displaced and found between the left and right ventricle.Which causes deoxygenated blood to flow into the aorta instead of the pulmonary artery.[3]
- **Right ventricular hypertrophy** - is the thickening of the muscular walls of the right ventricle, which occurs because the right ventricle is pumping at high pressure and can contribute to obstruction of blood flow through the pulmonary valve.[2]

TOF has prevalence of 3 cases per 10,000 live births as estimated world-wide. Regardless of its low prevalence, it is the most frequently occurring congenital heart defect (CHD).TOF illustrates 5-10% of all CHD in the general population of 0.8% of global birth prevalence.[4]

2. Signs and Symptoms

Severity of TOF symptoms varies and depends on degree of blood flow obstruction and includes:

- cyanosis (bluish coloration of skin due to low oxy-haemoglobin levels)
- shortness of breath (mainly during eating or exercise)
- fainting
- fatigue
- heart murmur
- failure to gain weight
- irritability
- clubbing
- squatting
- prolonged crying in infants

Tet spell- caused by rapid decrease in the levels of oxygen in blood and commonly seen in young infants (2 to 4 months old). [5]

3. Causes and Risk Factors

The exact etiology of TOF is still not known. However, around 15% of people have a specific genetic abnormality, either due to defective gene or chromosome, and it can be inherited. [6] Other possible reasons that may arise during pregnancy include environmental exposure such as:

- Overuse of alcohol
- Viral infections (rubella)
- Phenylketonuria
- Taking seizures medications
- Diabetes
- Late pregnancy (>40 years age)

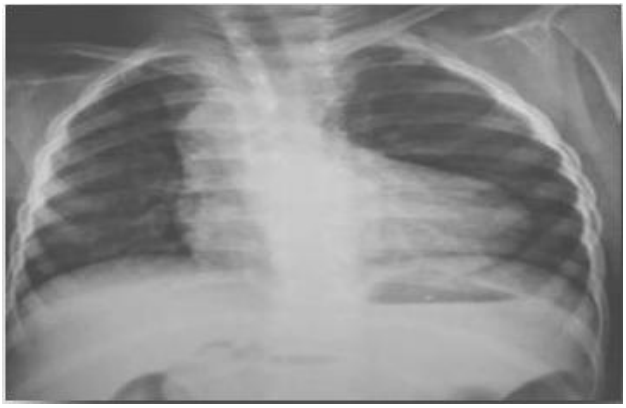
- Poor prenatal diet.[7]

4. Diagnosis

It can be observed during physical examination, when physician hears abnormal heart murmur (whooshing sound) and when the baby's skin appears blue.[8]

Tests to diagnose Tetrology of fallot include:

Pulse oximetry test - to know blood oxygen level **Chest-x-ray** - done to observe heart and lungs structure. It may indicate a boot-shaped heart (significant sign in TOF) because of right ventricle hypertrophy.



Echocardiogram: Performed to check the structure and function of heart.

Electrocardiogram (ECG): Aid to evaluate the abnormal heart beat and enlarged heart chambers.

MRI: To determine structural abnormality of heart.

Cardiac catheterization - it helps to determine the structure of heart and to select the appropriate surgical procedure.[9]

5. Treatment

Babies with tetralogy of fallot require corrective surgery carried out by a cardiologist. There is no proper growth and development of baby without treatment. The doctor will decide the most relevant surgery and time depending on your condition.[9]

Surgeries involved in tetralogy of fallot are open heart surgery to correct the defect (intracardiac repair), or a palliative (temporary) surgery that needs shunt.

Palliative shunt surgery (temporary shunt surgery):

It is performed when the baby is born prematurely or has undeveloped pulmonary arteries. In this surgery, shunt is placed between a large artery that originates from the aorta and the pulmonary artery to improve blood flow to the lungs. The shunt is removed when the baby is ready for intracardiac repair.

Intracardiac repair (ICR):
This surgery is performed by the first year of birth and rarely in adults who didn't underwent surgical repair during their childhood. The procedure involved in ICR is as follows:

The ventricular septal defect / hole is closed with a patch between the lower chambers of heart. The way out of the right ventricle is opened and the narrowed pulmonary valve (pulmonary stenosis) is repaired or replaced. To enhance the blood flow a tube is fixed between the right ventricle and pulmonary artery.

After the surgery, blood oxygen level improves and symptoms subside.[9]

Post Surgery:
The long-term survival rate improves in people who underwent tetralogy of fallot repair. Pulmonary regurgitation can be seen in an adult with TOF repair surgery and requires pulmonary valve replacement. Arrhythmias (abnormal heart rhythm) is a common complication of TOF surgery requiring medications or a procedure (ablation).[9]

Follow-Up Care:
People with tetralogy of fallot should undergo annual check-ups or imaging tests (ECG, Echo, chest-x-ray), whether or not they are suffering with problems to ensure surgery's success and to monitor for complications.[10]

6. Case Presentation

A 36 years old man with no comorbid conditions presented with chief complaints of sudden onset of shortness of breath (SOB) associated with sweating since 1 year. He has no medical reconciliation (treatment history). The patient is moderately built and nourished and is non-ethanolic and non-smoker. At admission, the patient's oxygen saturation of 97%, a regular pulse of 77 beats per minute and a blood pressure of 100/70mmHg was observed.

USG abdomen, TSH, serum creatinine, LFT, blood urea, RBS, PT, APTT, CUE, serum electrolytes, chest-x-ray tests were done and revealed normal. Whereas, electrocardiogram (ECG), echocardiogram, CT coronary angiogram were performed and found to be abnormal as follows:

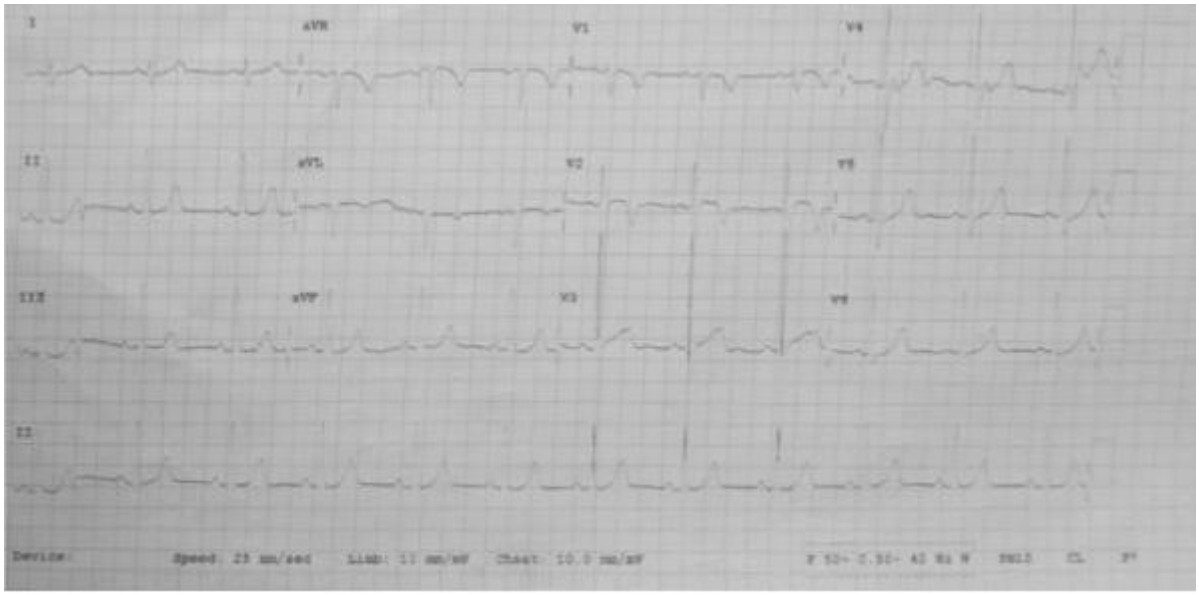


Figure 1: ECG showing left ventricular hypertrophy

Electrocardiaogram describing ventricular premature complex, left atrial enlargement, abnormal R-wave progression, left ventricular hypertrophy, abnormal T-wave, probable ischemia.

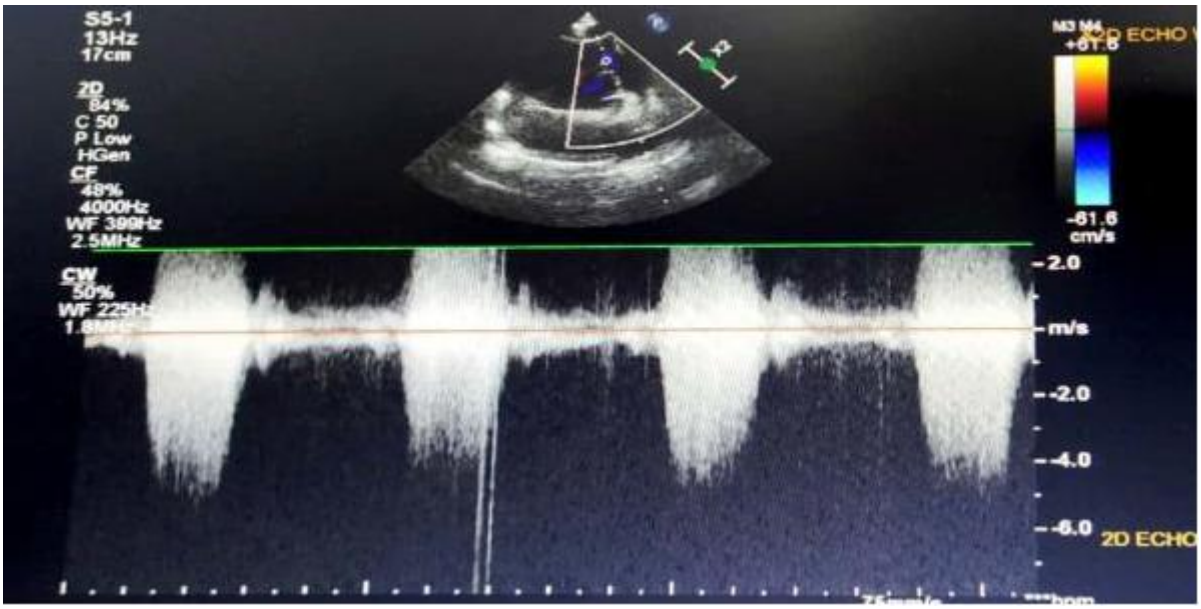


Figure 2 (a) : Echocardiogram showing abnormalities pre surgery

The abnormal results found in (figure-2a) echocardiogram pre surgery includes CHD, large sub-aortic VSD with L…>R shunt, severe RVOT obstruction (102mmHg).

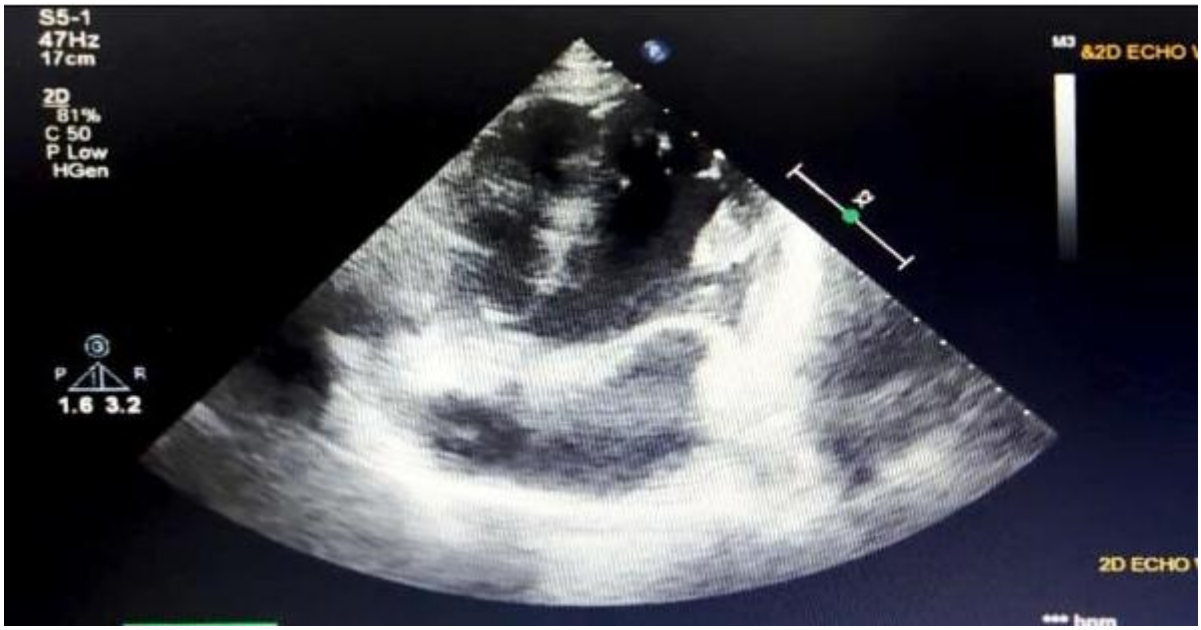


Figure 2 (b): Echocardiogram showing normal results post surgery

The result found in (figure-2b) echocardiogram post surgery: s/p VSD surgical closure intact IVS patch with no residual shunt, trivial MR/TR.

The defective features of CT coronary angiogram observed are s/o sub-aortic ventricle septal defect measuring 1.4cms in diameter. Overriding of aorta. Narrowing at right ventricular outflow tract (RVOT) with max diameter of 9mm. Thickening of pulmonary valves with decrease in diameter of the lumen at the wall.

The impression of ECG, 2D echo and CT coronary angio has established the diagnosis of TOF. Following the confirmatory diagnosis of TOF, the patient was adviced to undergo ICR (intracardiac repair) surgery.

After 3 months of follow up, the patient is able to carry out regular activities with no complaints of symptoms and complications.

7. Discussion

TOF is considered as the most common form of cyanotic congenital heart disease. Without performing surgical procedure long-term survival with TOF is rare and is often linked with well developed left ventricle.[11]

TOF has prevalence of 3 cases per 10,000 live births as estimated world-wide. Regardless of its low prevalence, it is the most frequently occurring congenital heart defect (CHD).TOF illustrates 5-10% of all CHD in the general population of 0.8% of global birth prevalence.[4]

Based on patients symptoms, physician adviced the following diagnostic tests:

Chest-x-ray - done to observe heart and lungs structure. It may indicate a boot-shaped heart (significant sign in TOF) because of right ventricle hypertrophy. **Echocardiogram** - performed to check the structure and function of heart.

Electrocardiogram (ECG) - aid to evaluate the abnormal heart beat and enlarged heart chambers.

MRI, **Pulse oximetry test**, **Cardiac catheterization:**
Surgeries involved in tetralogy of fallot are open heart surgery to correct the defect (intracardiac repair), or a palliative (temporary) surgery that needs shunt.

Palliative shunt surgery (temporary shunt surgery):
In this surgery, shunt is placed between a large artery that originates from the aorta and the pulmonary artery to improve blood flow to the lungs.The shunt is removed when the baby is ready for intracardiac repair.[9]

Intracardiac repair (ICR):
The section involves a combination of methods to repair all four defects.[6]

The ventricular septal defect / hole is closed with a patch between the lower chambers of heart. The way out of the right ventricle is opened and the narrowed pulmonary valve (pulmonary stenosis) is repaired or replaced. To enhance the blood flow a tube is fixed between the right ventricle and pulmonary artery.

After the surgery, blood oxygen level improves and symptoms subside.[9]

Procedure done in patient:

- Midline Sternotomy – surgical procedure in which vertical inline incision is made along sternum.
- Bicaval Cannulation - aorta bicaval cannulation, blood returning from body through SVC and IVC enter to separate canulae, which drain passively into an open reservior.
- Aortic Cross-clamp – used in cardiac surgery to clamp the aorta and separate the systemic circulation from the outflow of the heart.
- Arterograde Cardioplagia – is administered into small cannula placed in the ascending aorta or directly into the coronary ostia.

- Cava Snugged- absent of SVC.
- RA opened – right atrium is opened.
- VSD- (when it is not closed, oxygen-rich blood in left chamber can mix with oxygen-poor blood) this condition has been treated with glutaraldehyde and peri-cardial patch closer with 5-0 prolene

RVOT muscle bundle resected – no 20 Hegar's dilator passed. RA closed in two layers, deairing done, X-clamed removed, haemostasis done, two ventricular and two atrial pairing wires.chest closed with

8. Conclusion

We presented a case of patient who was diagnosed with TOF during his childhood for which shunting was done. Later, at the age of 36 he was suffering with complaints of chest pain associated with sweating since 1 year. Following the complaints of patient the shunt has been removed and successful ICR surgery was performed. Post surgery 5 months follow up was done and no complains of any symptoms and complications noted. Hence, the overall quality of life of patient is improved.

Consent

The verbal consent from the patient was obtained along with diagnostic reports for the publication of this case report.

Conflict of interest

Case is collected in March 2022 by Atufa Tahur et.al . This is an open access case report and the author has no conflict of interest to declare. We clarify that the submission is original work and it is not under review at any other publications.

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