



## TETRALOGY OF FALLOT IN ADULT; CASE OF UNPAIRED TETRALOGY OF FALLOT IN A 66 YEAR OLD MALE

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**ABSTRACT...** Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease in children but occurs rarely in adults. The extent of cyanosis depends on the balance of systemic and pulmonary vascular resistance, which depends on the severity of right ventricular outlet obstruction.<sup>3</sup> The more severe the obstruction, the more blood flows into the left side causing desaturation and cyanosis. The survival rate of patients who receive surgical full correction is about 86% at 32 years follow-up and 85% at 36 years follow up.

### Key words:

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### INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease in children but occurs rarely in adults. Its etiology is still not clear but its embryogenesis involves malalignment of the outlet septum resulting in ventricular septal defect, pulmonary outflow tract stenosis and aortic override.<sup>1,2</sup> In addition, right ventricular hypertrophy is noted secondary to pulmonary stenosis. Clinical presentation consists of cyanosis, clubbing of the fingers, polycythemia, and exertional dyspnea. Cyanosis and polycythemia may be noted in the newborn.

The extent of cyanosis depends on the balance of systemic and pulmonary vascular resistance, which depends on the severity of right ventricular outlet obstruction.<sup>3</sup> The more severe the obstruction, the more blood flows into the left side causing desaturation and cyanosis. Therefore, the more severe the pulmonary stenosis, the more protection from lung disease is noted. Mild pulmonary stenosis may present with mild cyanosis or even acyanosis, termed pink TOF or acyanotic TOF. Patients with this condition may have lung disease and may expire in early childhood if no repair or palliative surgery is performed.<sup>1,4</sup>

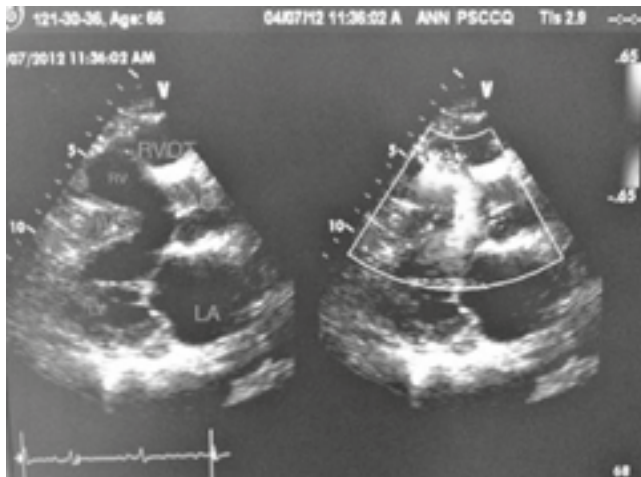
The survival rate of patients who receive surgical full correction is about 86% at 32 years follow-up and 85% at 36 years follow up.<sup>2</sup> Survival rates of un-operated TOF patients older than 10 years is about 30%, older than 20 years 11%, older than 30 years 6% and older than 40 years only about 3%.<sup>1</sup> We report here a male TOF patient diagnosed at 66 years of age, who was still asymptomatic and not diagnosed.

### Case History

We present here a case of 66-year-old asymptomatic male patient who was referred from a community hospital to our cardiology department for evaluation of congenital heart disease which was found incidently while pre-operative physical assessment of Inguinal Hernia repair. He presented with a 2-month history of Inguinal swelling which was diagnosed as a Inguinal hernia otherwise asymptomatic in the past and enjoying good social life with 8 children. Physical examination revealed well built old man, with regular pulse 76 bpm, blood pressure 120/70 mmHg, O<sub>2</sub> sat of 95% on room air and respiratory rate 22 breaths/min. Cyanosis and clubbing of the fingers were absent. The lungs were clear to auscultation and a ejection systolic murmur, grade 2/6 at left upper sternal border

was noted. No lower leg pitting edema was noted. Laboratory data revealed Hgb 13.6 g/dL, Hct 38.3% and rest of the data including liver and renal functions were normal. The chest x-ray revealed cardiomegaly with right sided Aortic Arch and bilateral lung fields were clear. The 12-lead electrocardiogram demonstrated normal sinus rhythm with right bundle branch block and right ventricular hypertrophy. Holter monitoring for 24 hrs had shown no abnormality except occasional premature ventricular contractions.

Echocardiography (Figure-1) showed a infundibular and valvular pulmonary stenosis estimating a pressure gradient of 115 mmHg with right ventricular annulus of 1.7 cm and a large ventricular septal defect at the perimembranous outlet to the subarterial portion, sized 2 cm with predominantly L to R shunt. There was significant right ventricular hypertrophy with mild Tricuspid regurgitation, mild left ventricular hypertrophy and over-riding of Aorta with Normal ejection fraction.

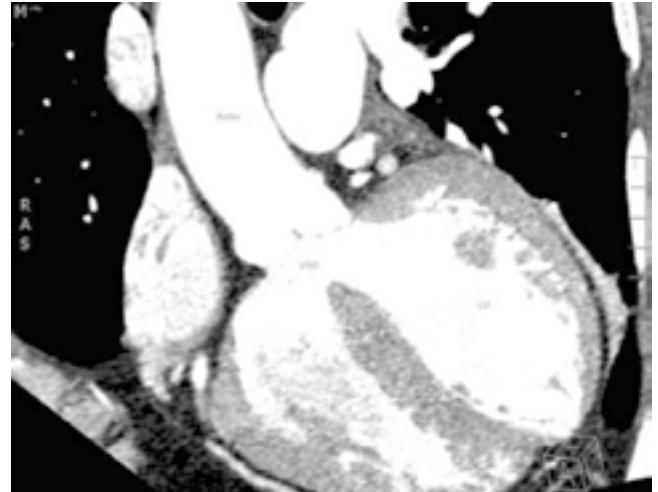


**Figure-1. Two-dimensional, para-sternal long axis echocardiographic images demonstrating clearly all features of this tetralogy of Patient with left to right shunt, IVS- interventricular septum, RVOT- right ventricle outlet tract obstruction, AO- aorta, RV- right ventricle, LA- left atrium, LV- left ventricle)**

Computed tomography (CT) angiogram (Figure-2) was done to rule out any coronary abnormalities demonstrated a peri-membranous ventricular septal defect, overriding of the aortic root, severe narrowing of the right ventricular infundibular portion, right ventricular hypertrophy

with Post-stenotic dilatation of main pulmonary artery and normal coronary arteries pattern.

Patient underwent for inguinal hernia repair successfully and uneventfully.



**Figure-2. CT angiogram of same patient showing clearly the Aortic over-riding valve (PV) and dilated main pulmonary artery (MPA).**

## DISCUSSION

TOF is rarely detected in a 66-year-old patient asymptomatic patient. Subhawong TK reported in 2009, a 87 year old lady, the oldest surviving unrepaired TOF patient.<sup>5</sup> With the extensive literature search, survival beyond age of 60 is very rare and reported cases are not more than single digit number.<sup>6,7</sup> Previously, congenital heart disease was thought of as a pediatric problem, and we have almost no experience of making a diagnosis of a new case or taking care of a congenital heart disease patient in adulthood. Over the past few decades, there has been a revolution in caring for children with congenital heart disease. Advances in diagnosis and surgery have made it possible to repair most defects, even those once thought to be hopeless. Many people with these defects are now reaching adulthood and we must continue to take care of them.<sup>8</sup>

Without operation, few patients with ToF reach adulthood with an average life expectancy of 12 years. 10% may survive to their 30s but only 3% reach their 40s or older.<sup>1</sup> There are three main factors which determine the longevity in natural survivors with unoperated ToF. Firstly,

a hypoplastic pulmonary artery with slow progression of subpulmonary obstruction which is present at birth.<sup>4,7</sup> secondly, LVH as seen in this patient which we don't know since how long it is there but presumably this acts by delaying of shunting from the right to left ventricle. LVH may be a late finding in the natural history of Fallot and any beneficial effect may not be seen until adult life.<sup>4</sup> Such a fine haemodynamic balance is clearly rare as seen in our patient. The third factor has been extracardiac shunts including patent ductus arteriosus or systemic to pulmonary artery shunting via internal mammaries<sup>8</sup>, although our patient did show any evidence of this kind.

There are few studies on the outcome and benefit of late surgical repair. The Mayo series followed up 30 patients who had total correction of ToF between the ages of 40 to 60 years<sup>4</sup> The operative mortality was 3% with long term survival rate at 5 years and 10 years postoperatively of 92% and 74% respectively.<sup>4</sup> However, higher age at the time of surgery has been reported as a risk factor for impaired long-term survival in one large independent study from the Mayo Clinic<sup>4</sup> but there is a recent case report of TOF surgically corrected by mayo clinic at the age of 83.<sup>9</sup> this case was diagnosed at the age of 78, the patient underwent complete repair of TOF due to New York Heart Association (NYHA) class IV heart failure, progressive cyanosis, and poor quality of life. After a challenging perioperative course, the patient eventually obtained an excellent result from his surgery and able to perform activities of daily living with NYHA class II at 3 years after surgery.

In summary, a TOF patient diagnosed at age 66 is rare, having said that survival rates of un-operated


TOF patients older than 40 years only about 3%<sup>1</sup>, this patient is among those lucky survivors. With the given physiology and asymptomatic, after discussion with patient the decision has been made not to operate in this case.

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