

*Case Report*

Uncorrected Tetralogy of Fallot Surviving in His Fourth Decade: A Case Report

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ABSTRACT

Tetralogy of Fallot (TOF) is one of the most common forms of cyanotic congenital heart disease (10% of all cases). The defect is in the development of foetal heart. The anterior displacement of conotruncal septum causes unequal divisions of conus leading to ventricular septal defect (VSD), pulmonary stenosis, overriding of aorta and ventricular hypertrophy. Though TOF is common in paediatric population, surviving adult cases without any surgical intervention is a rarity.

The authors reported this case because there are only a few reports of patients with an uncorrected tetralogy of Fallot reaching an advanced age. The anatomical adaptive changes against the disease entity are our area of interest.

Key words: congenital heart defect, tetralogy of Fallot, conotruncal septal defect, adult CHD

INTRODUCTION

Tetralogy of Fallot (TOF) is one of the most common congenital heart diseases in paediatric population. ^[1] Survival of this condition into adulthood is rare. The basic pathologies are mixing of oxygenated with deoxygenated blood and abnormal flow of blood in lungs, both due to faulty developmental process during embryogenesis. From anatomical point of view the problem lies in anterior deviation of the conotruncal septum resulting in mal alignment of outflow pathway of heart. The final outcome consists of pulmonary stenosis (PS) of varying degree, ventricular septal defect (VSD), overriding of aorta along with right ventricular hypertrophy

(RVH). ^[2] The child presents early with cyanosis, exertional dyspnoea, polycythemia, later develops clubbing of fingers. The extent of cyanosis is related with prognosis of patient and varies with balance between pulmonary and systemic resistance. The survival rates of un-treated TOF patients decrease radically with increasing age i.e. 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%. ^[3] Here we present a case of TOF surviving in his fourth decade without any medical or surgical interventions.

CASE REPORT

A 40 year old man presented with chest pain, easy fatigability, breathlessness on mild exertion and palpitations (lasting for 5-10 minutes) since childhood. His complaints were aggravated on a recent attack of respiratory infection for which he was brought to the hospital to seek medical advice. He had associated cough and headache. There was no history of paroxysmal nocturnal dyspnea (PND), ankle or abdominal swelling. He never attended school or played sports. There was neither past history of stroke or TIA, peripheral vascular disease nor any hospital visits.

General examination revealed an asthenic plethoric patient, centrally cyanosed with grade 4 digital clubbing. His pulse was regular with a rate of 80 beats/min. Blood pressure was 110/70 mmHg. On examination of the precordium the apex was localized in the 5th left ICS in the midclavicular line with a left parasternal heave. On auscultation there was grade 3/6 ejection systolic murmur in the left lower parasternal area. The chest was bilaterally clear.

His blood parameters revealed secondary polycythemia with hematocrit 79.4% and hemoglobin 25%.

Chest radiography revealed an enlarged cardiac silhouette with left sided aortic arch and hypoplastic pulmonary arteries.

The electrocardiography (ECG) showed sinus rhythm, normal PR interval, and right axis deviation, right ventricular hypertrophy, inverted T-waves in anterior chest leads.

Echocardiography showed a large malaligned VSD, gross aortic override (aortic outlet diameter of 40mm, hypertrophied septal wall measuring 21.5mm. There is gross right ventricular hypertrophy, hypoplastic pulmonary artery of 10mm, severe infundibular stenosis with right ventricular outflow tract peak gradient of 70 mmHg. There was also left ventricular hypertrophy with left ventricular posterior wall measuring 13mm.

Multiplanar MRI images revealed an overriding aorta, a large VSD, right ventricular hypertrophy with a hypoplastic main pulmonary artery (see fig-). Multiple retrotracheal periesophageal vascular structures probably hypertrophied and hyperplastic bronchial arteries were seen at the carina level suggesting a possibility of aorto-pulmonary collateral circulation.



Figure 1
Chest radiogram AP view showing
Enlarged cardiac silhouette & hypoplastic
Pulmonary arteries.

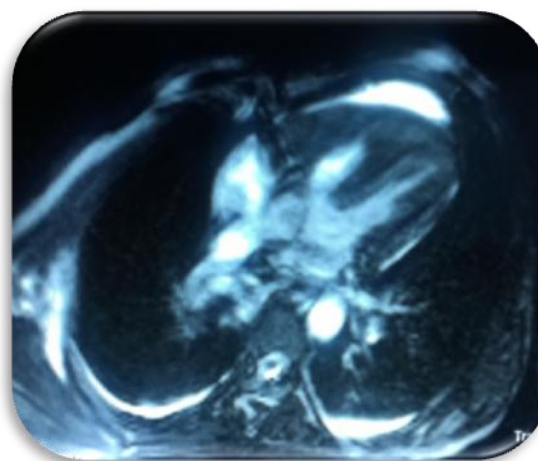


Figure 2
Multiplanar MRI showing large VSD



Figure : 3
MRI showing large VSD
Periesophageal vascular structures

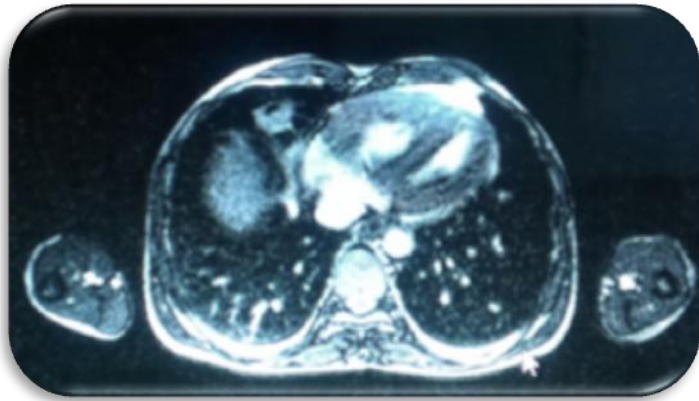


Figure: 4
MRI showing multiple retro tracheal



Figure: 5
Echocardiography showing
hypoplastic pulmonary artery

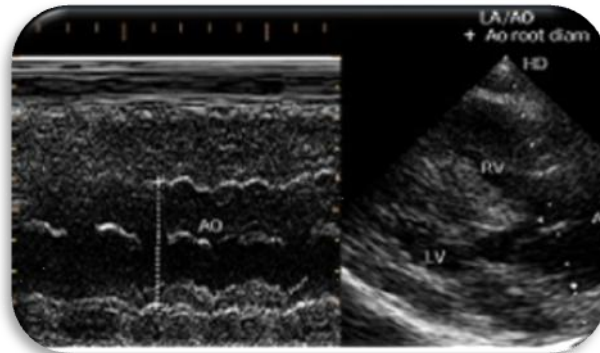


Figure: 6
Echocardiography showing overriding of aorta

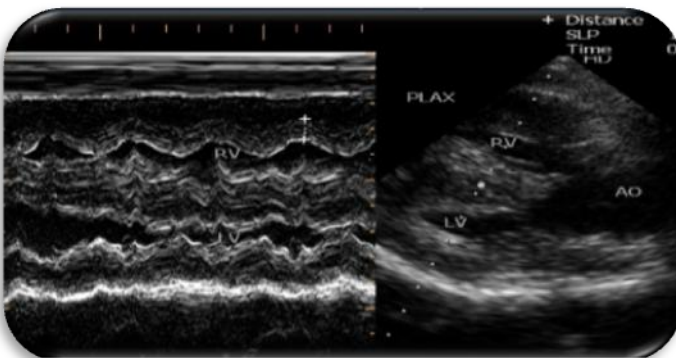


Figure: 7
Echocardiography showing gross right
Ventricular hypertrophy

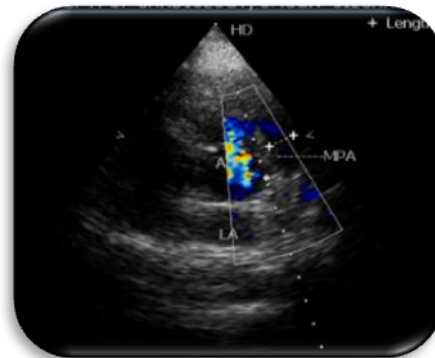


Figure: 8
Echocardiography showing
severe infundibular stenosis

DISCUSSION

The events during 5th week of embryogenesis lead to formation of ventricular septum and also the outflow tract. First pairs of opposing ridges appear in the truncus, growing towards aortic sac they twist around each other. After complete

fusion they form the aortico pulmonary septum dividing truncus into aortic and pulmonary channel. Similarly conus cordis is divided by two approaching cushions into outflow tract of right and left ventricle. [4] TOF is due to anterior displacement of this conotruncal septum leading to unequal

divisions of conus.

The affected child suffers from different complications of cyanosis, thrombocytosis leading to cerebral thrombosis, brain abscess, and bacterial endocarditis. Heart failure is also not a rare finding. [5] As a result these patients suffer from high mortality. TOF patients reaching adult age is a rarity but in those who ultimately survive to adult age (without any corrective procedure) some adaptive changes are seen. Three main factors have been identified for the longevity in natural survivor with unoperated TOF. [6]

Firstly a hypoplastic pulmonary artery with slow development of sub-pulmonary obstruction.

A second common feature is that of left ventricular hypertrophy (LVH), presumably this acts by delaying of shunting from the right to left ventricle.

The third finding in other cases has been extra-cardiac shunts including patent ductus arteriosus (PDA) or systemic to pulmonary shunting via major aorto-pulmonary collaterals (MAPCAs).

In our patient we found two of these mechanisms present.

There was hypo plastic pulmonary artery with gradual development of infundibular stenosis.

And there was hyperplasia and hypertrophy of bronchial arteries suggestive of aorto pulmonary collateral circulation.

These two factors contributed to the survival of our patient in adulthood.

Complete surgical correction is now the standard treatment of TOF. Mortality is about 2.5% to 8.5% in adults. Overall survival of surgically treated adult patients is acceptable and the great benefit of complete repair at this age is the functional

improvement. [7]

CONCLUSION

In summery this adult TOF case is reported for better understanding of the embryological and anatomical factors contributing to the disease entity.

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