Tetralogy of Fallot presenting with Right Ventricular Failure

Priyam Mukherjee¹, Arijit Ghosh¹, Saubhik Kanjilal¹, Arindam Maitra¹, Tanmoy Majee¹, Pradip Saha¹, Aloke Choudhury¹, Manojit Lodha², Sibananda Dutta³, Jotideb Mukhopadhaya⁴

Abstract
A 32 year old male patient was admitted with exertional dyspnea for long duration with features of right sided failure. On examination, it seemed to be a congenital cyanotic heart disease with decrease pulmonary flow. Echocardiography showed a malaligned VSD with pulmonary stenosis with severe AR and the total regurgitation volume was going to the RV instead of going to both the ventricles leading to dilatation of right ventricle and failure.

Case Report
A 32 year old male patient presented with exertional dyspnoea for 25 yrs with exacerbation since last 6 months and h/o- exertional cyanosis since early childhood. He also noticed palpitation for last 6 months and bilateral pedal edema for one month. The infancy of the patient was uneventful without any history of squatting or cyanotic spells. There was no history of hemoptysis, headache, seizures, blurring of vision or bleeding tendency. There was no history of orthopnea/PND. The patient’s mother didn’t give any history of recurrent LRTI, failure to thrive, feeding difficulty or excessive sweating during childhood.

On examination, he was mildly cyanotic having grade 1 clubbing with raised JVP (prominent v and y descent). RV type of apical impulse was present at 5th intercostal in anterior axillary line. S2 was single and loud, best heard at 2nd left intercostal space. Ejection click heard at 2nd left intercostal space. High pitch pansystolic murmur heard at left 3rd/4th intercostal space. Ejection systolic murmur and EDM also heard over precordium.

ECG was consistent with TOF (Figure 1). X-ray chest showed huge cardiomegaly (CTR of about 80%) (Figure 2). 2D-echo and colour Doppler study showed the following features (Figure 3-6).

• Large malaligned nonrestrictive VSD with bidirectional shunt along with pulmonary stenosis.
• Dilated RA and RV
• Colour Doppler of the aorta showed severe aortic regurgitation and whole of the regurgitant volume was going towards RV instead of going to both ventricle
• Severe tricuspid regurgitation noted.

Thus, our final diagnosis was TOF with severe AR causing RV failure.

Discussion
Aortic root dilatation occurs due to increased flow through the aorta.¹ However, there are also aortic root histologic² and elastic abnormalities³ which too play role in aortic root dilatation

![Image: 12 lead ECG showing rapid transition of r wave from v1 to v2](image-url)

¹Post Doctorate Trainee, ²Assistant Professor, ³Professor and Director, ICVS, ⁴Professor and HOD Department of Medicine, IPGMEandR, Kolkata, West Bengal
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causing AR. TOF with AR causing biventricular failure is a known fact. But the regurgitant jet going solely to the right ventricle and causing RV failure is not known. In this case there is no feature of LV volume overload due to AR and such peculiar behavior of aortic regurgitation in Fallot has not been described in literature as per our knowledge. The direction of the jet was eccentric and septal malalignment was such that the whole jet was going towards RV. This is an unknown presentation of Fallot. So it is a common defect presenting in an uncommon way.

References
