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CONGENITAL HEART DISEASE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Tetralogy of Fallot With Aortic Origin of the Pulmonary Artery and Uncorrected Right Aortic Arch

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ABSTRACT

This clinical vignette describes the first case of a woman in his 40s with a set of congenital anomalies given by tetralogy of Fallot associated with aortic origin of the left pulmonary artery and uncorrected right aortic arch. All of these entities have a poor probability of survival in adulthood. (J Am Coll Cardiol Case Rep 2024;29:102277) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

BACKGROUND

Tetralogy of Fallot is the most frequent cyanotic congenital heart disease in the United States. It has good survival when early surgical correction is performed; however, without surgical correction, survival decreases over the years.[1,2]

The anomalous origin of a pulmonary artery from the aorta is rare among congenital heart diseases, being much less frequent the origin of the left pulmonary artery.[3]

There are few published cases of tetralogy of Fallot associated with an anomalous aortic origin of the pulmonary artery, which makes presentation of this case in an adult patient fascinating.

CLINICAL VIGNETTE

A female patient in her 40s with history of tetralogy of Fallot without surgical correction consulted for a 10-year history of medium exertion dyspnea in relation to atypical chest pain. The symptoms were exacerbated 1 year prior to the consultation. She was being managed with carvedilol and aspirin and used supplemental oxygen at night.

At the initial assessment, the patient's blood pressure was 105/56 mm Hg, oxygen saturation was 80% to 85% of ambient air at rest, and 75% in exertion. Her weight was 41 kg, her height was 165 cm, her body mass index was 15.8 kg/m^2, and clubbing was observed. At auscultation, rhythmic heart sounds were found, with a holosystolic murmur predominantly in the pulmonary focus and at the left parasternal level between the third and sixth intercostal space without evidence of fremitus. A transthoracic echocardiogram was performed; it documented findings compatible with tetralogy of Fallot, with dilation and signs of right ventricular overload,

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ABBREVIATIONS AND ACRONYMS

TAPSE = tricuspid annular plane systolic excursion

tricuspid annular plane systolic excursion (TAPSE) of 14 mm, fractional area change of 35%, and preserved left ventricular systolic function with an ejection fraction of 60%.

A right heart catheterization was performed (Figure 1). It revealed a pulmonary transvalvular gradient of 71 mm Hg, with evidence of pulmonary hypertension with a pulmonary artery systolic pressure of 61 mm Hg, diastolic pressure of 15 mm Hg, and mean pulmonary artery pressure of 31 mm Hg. Right ventricular pressures were measured, revealing an end-systolic pressure of 132 mm Hg and end-diastolic pressure of 5 mm Hg. In addition, oximetry was performed in right cavities; it documented an oximetric jump of 3.5% volume percent from the right atrium to the right ventricle, with a pulmonary to aortic flow ratio of 1.0:1.0, and a pulmonary vascular resistance of 10 Wood units.

Pulmonary and aortic arteriography was performed with right and left ventriculogram documenting dysplasia with severe pulmonary stenosis, with dilatation and hypertrophy of the ventricle, aorta overrides by 50% of the aortic diameter with perimembranous ventricular septal defect; these findings are compatible with tetralogy of Fallot. During pulmonary arteriography, absence of left pulmonary branch was documented. An aortogram was subsequently performed documenting an anomalous origin of left pulmonary artery from the aorta with right aortic arch. A CT angiogram with three-dimensional reconstruction was performed and confirmed the previous finding. Medical management with sildenafil, aspirin, metoprolol, and atorvastatin was indicated. In follow-up, the patient has had no clinical deterioration, with adequate tolerance to medication.

In conclusion, tetralogy of Fallot and aortic origin of pulmonary artery are a rare pathology, and the benefits of both medical and surgical interventions in adult patients are not clear.

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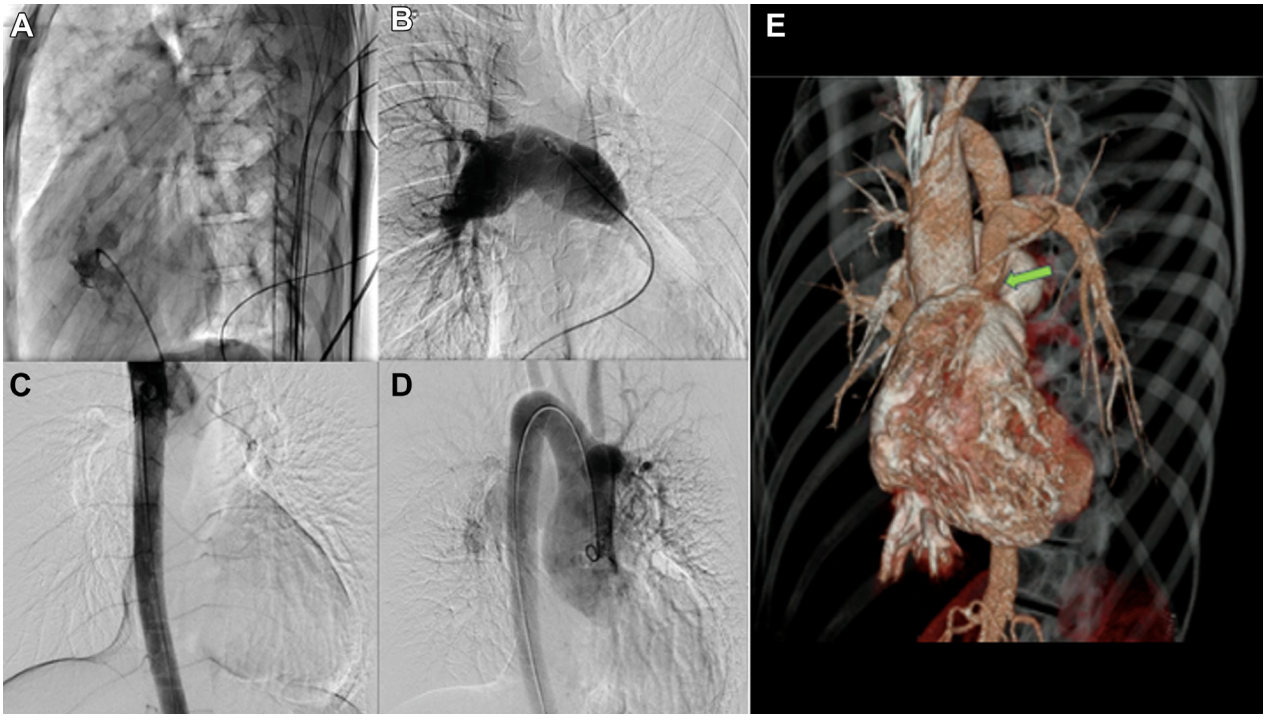
FIGURE 1 Pulmonary and Aortic Arteriography and CT Angiogram With 3-Dimensional Reconstruction



Pulmonary arteriography evidence of an interventricular communication (A), absence of the left pulmonary branch (B), right aortic arch (C), and aortic origin of the left pulmonary artery (D). (E) CT angiogram with three-dimensional reconstruction shows aortic origin of the left pulmonary artery (arrow).

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