


# Imaging findings in uncorrected tetralogy of Fallot and pulmonary atresia with major aortopulmonary collateral arteries and septic embolism

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## Abstract

Tetralogy of Fallot (TOF) is one of the most common congenital heart malformations comprising a ventricular septal defect, right ventricular outflow tract obstruction, right ventricular hypertrophy, and overriding aorta. A rare variant includes pulmonary atresia and major aortopulmonary collateral arteries. Altered hemodynamics within the functional single-ventricle results in turbulent flow and predisposes to endocardial vegetation formation which may consequently lead to thromboembolic events. We present a rare case of an adult survivor of uncorrected TOF with pulmonary atresia.

## Keywords

Heart, congenital heart malformation, tetralogy of Fallot, CT angiography, infection

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## Introduction

Tetralogy of Fallot (TOF) is one of the most common congenital heart malformations comprising a ventricular septal defect, right ventricular outflow tract obstruction, right ventricular hypertrophy, and overriding aorta. A rare variant includes pulmonary atresia and major aortopulmonary collateral arteries.

## Case report

We report a case of a 39-year-old female patient with uncorrected TOF and pulmonary atresia presenting to our emergency department with fever, elevated infection parameters, and hemorrhagic macular lesion on the middle finger of the right hand. Patient history revealed a Blalock–Taussig shunt performed for palliation in 1983 due to pulmonary atresia with subsequent spontaneous closure. Corrective surgery failed in 1998. Coronary angiography performed in 1997 was normal.

On admission, a contrast-enhanced multidetector computed tomography (CT) of the chest and abdomen was performed to rule out focal thoracic or abdominal infection. The CT study demonstrated an

absent pulmonary artery, a perimembranous ventricular septal defect (VSD) of 1.8 cm diameter, an atrial septal defect (ASD), and right ventricular hypertrophy (RVH). Furthermore, major aortopulmonary collateral arteries (MAPCA) arising from the descending aorta, an obliterated ductus arteriosus, and a dilatation of the aortic root (diameter 5.0 cm) were observed (Fig. 1). The patient demonstrated multiple pulmonary and splenic emboli (Fig. 2).

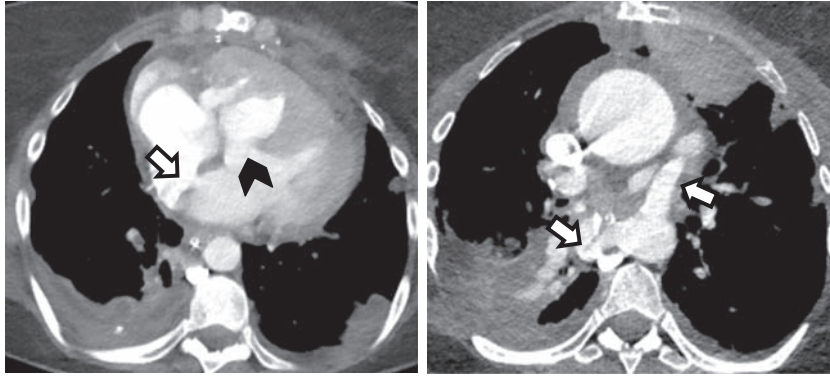
Echocardiography revealed an ostium secundum type ASD, a VSD, a moderately reduced left ventricular ejection fraction (45%), and two large vegetations on the tricuspid valve. Blood cultures were positive

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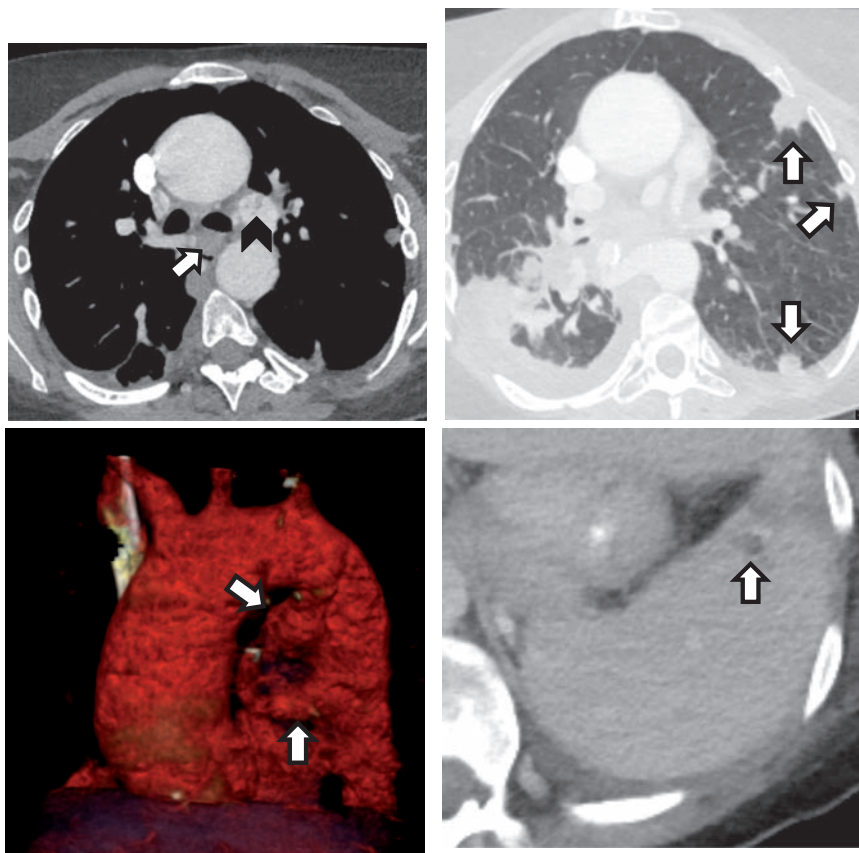
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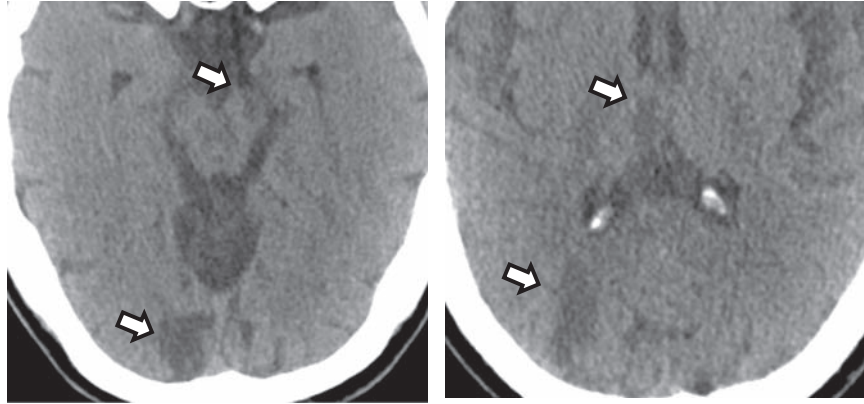
**Fig. 1.** Axial contrast-enhanced CT images. The image on the left shows a large VSD with a diameter of 1.8 cm (arrow head) and an ASD (arrow). MAPCA from the descending aorta (right). Note the absent pulmonary trunk, right ventricular hypertrophy and multiple, peripherally located, wedge-shaped lung lesions representing pulmonary emboli.



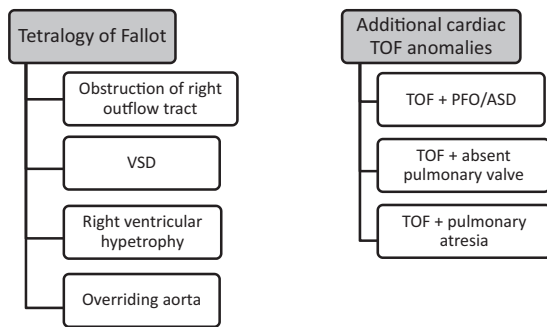
**Fig. 2.** Axial contrast-enhanced CT images showing left upper lobe pulmonary artery with filling defects representing wall adherent thrombi (arrow head) and reactive hilar lymphadenopathy (arrow, upper left), multiple peripherally located pulmonary lesions (upper right). 3D volume-rendered image demonstrating several MAPCA from the descending aorta (lower left). Wedge-shaped peripheral splenic defects representing splenic emboli (lower right).

for staphylococcus aureus. Targeted antibiotic treatment was initiated immediately, and the patient was referred to the cardiac surgery department for tricuspid valve repair which was performed 2 days after admission.

Due to a protracted clinical course and neurologic deterioration after surgery a cranial CT was performed. Several hypoattenuating cerebral and cerebellar lesions were predominantly localized in the right hemisphere consistent with thromboembolic events (Fig. 3).



**Fig. 3.** Non-enhanced cranial CT images showing hypoattenuating lesions in the left cerebral peduncle, in the right occipital lobe, thalamus, and cuneus (arrows), consistent with embolic infarctions.



**Fig. 4.** The four TOF-defining components (left) and three most common variants of TOF (right). TOF plus PFO/ASD also referred to as pentalogy of Fallot.

The further course of the patient was unfavorable, showing progressive cardiorespiratory insufficiency and increasing demand for vasoactive agents. Eighteen days after initial admission the patient passed away, due to cardiopulmonary failure.

## Discussion

TOF is one of the most common congenital heart malformations. It consists of an interventricular communication, a biventricular connection of the aortic root, which overrides the muscular ventricular septum an obstruction of the right ventricular outflow tract, and RVH (1). The variants of TOF include TOF with a patent foramen ovale/ASD (pentalogy), TOF with absent pulmonary valve, and TOF with pulmonary atresia (Fig. 4).

TOF with pulmonary atresia is clinically and radiologically distinct from regular TOF. It comprises 5–10% of all tetralogy complexes (2). The anatomy of central pulmonary arteries is often abnormal (3,4). MAPCA are systemic-to-pulmonary collateral

arteries representing remnants of the embryonic ventral splanchnic arteries and provide an alternative pulmonary blood supply in patients with TOF and pulmonary atresia. These embryonic vessels normally regress concomitantly with the formation of the normal pulmonary arterial system in the first weeks of gestation, whereas in patients with pulmonary atresia they persist (5).

Survival rates in TOF with pulmonary atresia without surgical repair reported in the literature are as low as 50% at 1 year of age and 8% at 10 years (6). Our case shows an adult survivor of uncorrected TOF with pulmonary atresia, the oldest survivor ever reported in the literature being 59 years old (2). As postulated by Fukui et al. the survival might be primarily dependent on the adequacy of pulmonary blood flow derived from MAPCAs (2). TOF with pulmonary atresia may lead to a more severe RVH than in non-variant TOF due to a functional single-ventricle circulation. Altered hemodynamics within this functional single-ventricle results in turbulent flow and may be more dominant in the right chamber than in the left due to the complete absence of the physiologic right ventricle outflow. Turbulent blood flow predisposes to endocardial vegetation formation on the tricuspid valve which may consequently lead to thromboembolic events. Usually emboli and vegetations from the right heart are filtered by the lung. This is not the case in TOF variants with systemic shunts, where dangerous systemic emboli to the brain and other vital organs may occur.

In conclusion, TOF with pulmonary atresia is a rare entity of congenital heart disease presenting typical findings on cross-sectional imaging including VSD, right ventricular outflow tract obstruction, right ventricular hypertrophy, overriding aorta, and MAPCA. Uncorrected blood flow in patients with palliative surgery or failed correction predisposes to endocardial

vegetations and consecutive thromboembolic events within the systemic circulation.

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