CASE REPORT

Case Report: A forty year-survivor of Tetralogy of Fallot with pulmonary atresia and chronic pediatric shunt thrombosis; findings from cardiac CT scan [version 1; peer review: awaiting peer review]

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Abstract
Here we illustrate a rare case of Tetralogy of Fallot (TOF) with pulmonary atresia in a 40-year-old survivor patient, despite a chronic pediatric shunt thrombosis. The patient became symptomatic at 38-years-old with progressive dyspnea on exertion and short cyanosis spells. The clinical findings were unspecific except for sinus tachycardia. To reassess cardiac abnormalities and associated intrathoracic malformations, a cardiac CT-scan was performed. Here we explain the mechanism for prolonged survival as an expansion of several and huge major aortopulmonary collateral arteries. This case predicts an exceptional late outcome of untreated TOF.

Keywords
Computed tomography, cardiac imaging, Tetralogy of Fallot, Pulmonary atresia, Blalock Taussig shunt.

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Author roles: Achour A: Conceptualization, Formal Analysis, Funding Acquisition, Investigation, Methodology, Visualization; Mnari W: Supervision, Validation; Abdelali M: Visualization; Zrig A: Supervision, Validation; Miladi A: Visualization; Ben Messaoud M: Validation; Hmida B: Visualization; Maatouk M: Supervision, Validation

Competing interests: No competing interests were disclosed.

Grant information: The author(s) declared that no grants were involved in supporting this work.

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How to cite this article: Achour A, Mnari W, Abdelali M et al. Case Report: A forty year-survivor of Tetralogy of Fallot with pulmonary atresia and chronic pediatric shunt thrombosis; findings from cardiac CT scan [version 1; peer review: awaiting peer review] F1000Research 2020, 9:647 https://doi.org/10.12688/f1000research.24374.1

Introduction

Tetralogy of Fallot (TOF) is the most frequent form of cyanotic congenital heart disease. Only a few patients come of age without surgical intervention mainly in the extreme form with pulmonary atresia\(^1\). Here, we relate the case of a patient who survived until the age of 40 years without surgical curative intervention. The patient had an unusual association of chronic Blalock-Taussig shunt (BTS) thrombosis and huge major aortopulmonary collateral arteries (MAPCAs). This case provides the main role that a cardiac CT scan can lay in understanding the late outcome of these untreated cardiac abnormalities.

Case report

A 40-year-old man with “complex” cardiac disease and dyspnea was referred to our department for imaging. Old medical records with conventional angiographic studies inferred the diagnosis of TOF with pulmonary atresia, made at birth, treated by BTS. As the patient was asymptomatic, he had not consulted previously, and had been lost to follow-up. Two years ago, he began to present dyspnea on exertion, and also short cyanosis spells.

Clinical findings on presentation were unspecific except for sinus tachycardia. ECG showed biventricular hypertrophy and incomplete right bundle branch block. Echocardiography confirms the diagnosis of TOF with pulmonary atresia. To reassess cardiac abnormalities, primarily pulmonary trunk morphology and associated intrathoracic malformations, a 128-slice CT scan with ECG synchronization was performed. The scan revealed a large ventricle septal defect with overriding aorta (Figure 1, curved arrow) and right ventricle hypertrophy (Figure 1, asterisks). The pulmonary trunk was atretic, showing characteristic seagull pattern (Figure 2, arrows). The right pulmonary artery had a good diameter, while the left was significantly smaller (Figure 2). Pulmonary blood flow was given by MAPCA connecting blood vessels between the aorta and the pulmonary arteries (Figure 3, arrows). MAPCA was more numerous on the right side supplying the largest pulmonary artery. The BTS was completely thrombosed with a total heterogeneous filling defect and parietal calcifications (Figure 4, arrows); this is compatible with chronic thrombosis.

Figure 1. Four-chamber view from cardiac CT scan with ECG synchronization visualizing ventricular septal defect with overriding aorta and RV hypertrophy. Curved arrow shows the ventricular septal defect; arrows show the overriding Ao; and asterisks show the RV hypertrophy. LA, left atrium; RA, right atrium; LV, left ventricle; RV, right ventricle; Ao, aorta.

Figure 2. Multiplanar reconstruction image from a cardiac CT scan with ECG synchronization showing an atretic pulmonary trunk and hypoplasia of the LPA. Arrows show the atretic pulmonary trunk. LPA, left pulmonary artery; RPA, right pulmonary artery.

Figure 3. Cardiac CT scan with ECG synchronization visualizing collateral blood supply of the lungs showing coronal-targeted maximum intensity projection (C1) and posterior volume rendering (C2) views of the heart and mediastinal great vessel. Arrows show major aortopulmonary collateral arteries.
MAPCAs are arteries that grow to irrigate lung circulation when native pulmonary arteries are underdeveloped. MAPCAs often arise from the descending aorta but also from the aortic arch and other systemic arteries like subclavian, the carotid, or rarely the coronary arteries. Pulmonary atresia-ventricular septal defects are classified into 3 types: type A is associated with presence of the native pulmonary arteries with a pulmonary-aortic duct; type B has MAPCAs and native pulmonary arteries, such as in our case; type C has only MAPCAs, which are seen to provide the pulmonary blood without native pulmonary arteries.

Adulthood clinical presentation, as in our case, is extremely rare. Without surgical intervention, most patients die at a young age with a rate of survival of 66% at one year of age, 11% at 20 years, and 3% at 40 years. In contrast, the survival rate after surgical treatment is over 90% at 40 years old. The surgical option includes palliative forms, such as BTS, which consists in the creation of systemic to pulmonary shunt. Complete repair is the main option and associated with excellent outcome results. In this case, despite a duc tus arteriosus closure and a chronic pediatric shunt thrombosis, the patient survived at the age of 40 due to large and developed MAPCAs. The oldest survivor ever reported in the literature is 59 years old.

After a cardiac ultrasound, multidetector angiography CT scan is indicated to the assessment of TOF with pulmonary atresia mainly for associated intrathoracic malformations and to establish the precise MAPCA cartography before curative surgery. Reformatted images allow a comprehensive analysis of pulmonary artery anatomy, measurements of the lumen of ascending aorta, analysis of the origins and course of coronary arteries, study location, the size, and flow in prior shunts. The excellent spatial resolution and fast acquisition of multidetector CT make it a prime imaging tool to demonstrate even distal aortopulmonary collateral anatomy. Radiation should be reduced as low as possible in the pediatric population.

Conclusion
Untreated pulmonary atresia with a ventricular septal defect is uncommon in adults. Most patients die from serious respiratory troubles or congestive cardiac failure very early. An angiography CT scan is the main imaging tool to delineate the pulmonary arterial supply, which is essential for an appropriate surgical approach.

Consent
Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

Data availability
All data underlying the results are available as part of the article and no additional source data are required.

References
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