An anomalous presentation of tetralogy of Fallot with pulmonary atresia surviving to the age of 62 years

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The following images are from a 62-year-old woman with uncorrected tetralogy of Fallot and pulmonary atresia. Her presenting symptom was ascites. Her clinical examination was remarkable for cachexia, cyanosis and clubbing of the fingers. The resting pulse was 71 b.p.m. and irregular. Her baseline blood pressure was 112/40 mmHg, and her arterial oxygen saturation was 81%. On auscultation, there was a mid-systolic murmur audible throughout the precordium. A prominent left parasternal heave was detectable. The patient had no prior history of chest pain or electrocardiographic evidence of suspected myocardial ischaemia.

Despite several invasive angiographic studies, the correct morphological diagnosis or source of pulmonary blood supply could not be identified and the presumptive diagnosis of truncus arteriosus was made. To reevaluate her morphology, a 64 detector-row computed tomography scan was performed. Although the distal coronary arteries could not be evaluated in detail due to motion artefacts caused by insufficient breath-holding and an irregular rhythm, the imaging revealed a ventricular septal defect with overriding aorta, pulmonary atresia (Panel A), right descending aorta (Panel B), and a giant coronary-pulmonary artery fistula, which arose from the left main coronary artery and tortuously terminated in the main pulmonary artery (Panels B and C; arrow). The findings were confirmed by echocardiography (Panel D; arrow). This aberrant anatomy allowed growth of the pulmonary arteries and provided sufficient pulmonary blood supply despite the atretic valve. The mechanism for prolonged survival may be the initial physiological adaptations that ameliorated the left-to-right shunting, such as the formation of a giant coronary-pulmonary fistula allowing subsequent growth of the pulmonary arteries. The patient is currently receiving conservative medical management including low-dose diuretic therapy, removal of ascites by abdominocentesis, and non-invasive positive pressure ventilation at home. She is currently asymptomatic.