

# Surgical Treatment of Tetralogy of Fallot with Giant Pulmonary Artery Aneurysm in an Adult

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## INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect with a prevalence of 0.5 per 1000 live births; TOF represents approximately 9% of all congenital heart defects. Survival rates for adults with repaired TOF in childhood are now excellent [Knowles 2012]. The population of surviving adult patients with unrepaired TOF has been steadily increasing and recent advances in operative techniques have allowed successful surgical treatment of these patients. This case report describes a successful surgical repair of TOF with giant pulmonary artery aneurysm in a 26-year-old male.

## CASE PRESENTATION

A 26 year-old male with no known history of cardiac disease presented with dyspnea and cough. There was no family history of congenital heart disease. On admission, he had a blood pressure of 110/60 mmHg, pulse 110 beats per minute, and a hematocrit of 56%. Oxygen saturation was 78% on room air, with clubbing and cyanosis present. Serum chemistry values were all within normal limits. Height and weight were 150 cms and 50 kgs (body mass index of 22.2). The patient was assessed at NYHA functional Class III. A 12 lead ECG showed sinus rhythm, right bundle branch block and right ventricle hypertrophy. On auscultation, the first and second heart sounds were normal, with blowing systolic murmurs (4/6) and thrill. Mean PA pressure was 21 mmHg by right heart catheterization. Chest xray showed coeur en sabot (boot-shaped) configuration of the cardiac silhouette and left pulmonary artery enlargement (Figure 1). Transthoracic echocardiography (TTE) revealed classic components of TOF: a 21 mm ventricular septal defect (VSD)

with no shunting, right ventricular outflow tract obstruction (RVOTO), aortic override and right ventricular hypertrophy. The mean gradient between the right ventricle (RV) and pulmonary artery (PA) was 70 mmHg. The main pulmonary artery and right and left pulmonary arteries (RPA, LPA) were markedly dilated (Figure 2). CT showed an ascending aortic diameter of 4.9 cm. A McGoon ratio of 6.0 was calculated but was determined to be inaccurate due to aneurysmatic right and left pulmonary arteries. Right heart angiography revealed major aortopulmonary collateral arteries. CT angiography also demonstrated a markedly enlarged left pulmonary artery (Figure 3).

## SURGICAL TECHNIQUE

Following full median sternotomy and cannulation of the distal ascending aorta, hypothermic cardiopulmonary bypass (CPB) was initiated. Antegrade cold blood cardioplegia was infused every 20 minutes during aortic cross clamp time. Pulmonary arteriotomy was performed longitudinally, from the main pulmonary artery to the left hilar area (Figure 4). The pulmonary valve was absent. Ventriculotomy was performed for VSD closure using a Dacron patch. After surgical correction of the intracardiac defect, a 20 mm Contegra® bovine jugular vein conduit graft was used to achieve right ventricular to pulmonary artery continuity. Triangular segments of the anterior wall of each branch of the pulmonary artery and part of the anterior wall of the main pulmonary artery were excised. The annulus of the pulmonary artery was measured using Hegar dilators. The ascending aorta had aneurysmatic dilatation of 5 cm; the sinus of Valsalva was 6 cm. Therefore, the decision was made to remodel the ascending aorta with aortic linear plication and wrapping technique (Figure 5) [Ozcan 2013]. Total cardiopulmonary bypass time, including cross clamp time, was 240 minutes; the patient was weaned from CPB without difficulty. Postoperatively, TTE showed minimal pulmonary valvular insufficiency and stenosis, a mild degree of tricuspid insufficiency, no residual VSD, LVEF of 70%, and an ascending aortic diameter of 2.6 cm. Hospital length of stay was 8 days, with 3 days in the intensive coronary unit, and was uneventful. Postoperative improvement was excellent and the patient's physical condition appeared generally healthy (NYHA class I-II).

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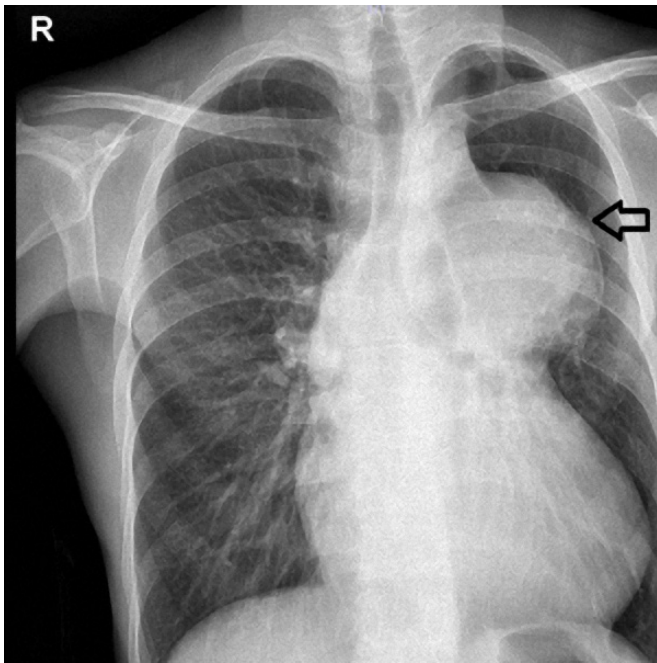


Figure 1. Coeur en sabot (boot-shaped) configuration of the cardiac silhouette and left pulmonary artery enlargement.

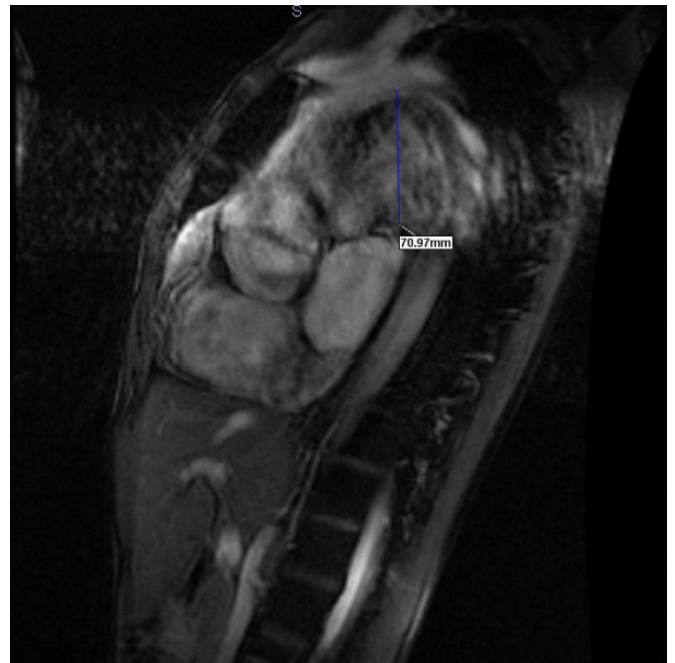


Figure 2. Image showing enlarged main pulmonary artery and a markedly enlarged right and left pulmonary arteries.

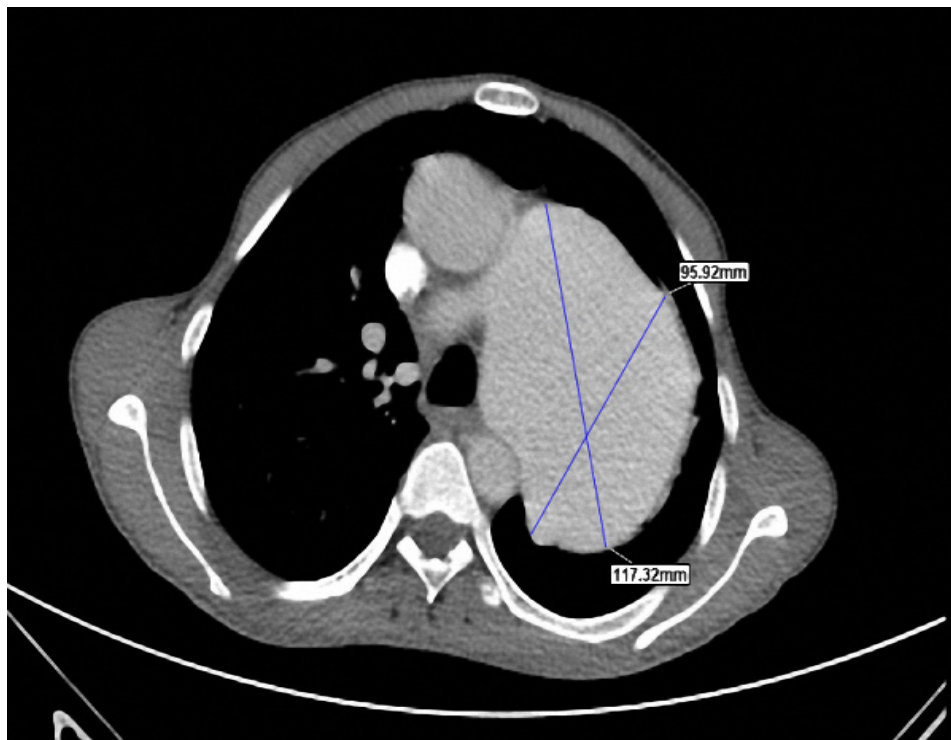


Figure 3. CT angiography showing a dilated left pulmonary artery.

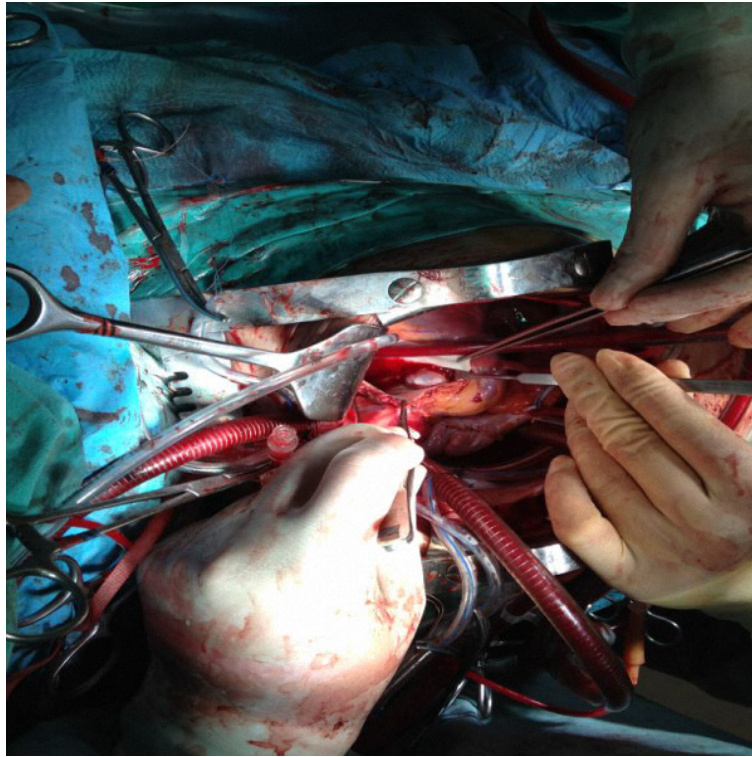


Figure 4. Pulmonary arteriotomy performed longitudinally, from the main pulmonary artery to the left hilar area.

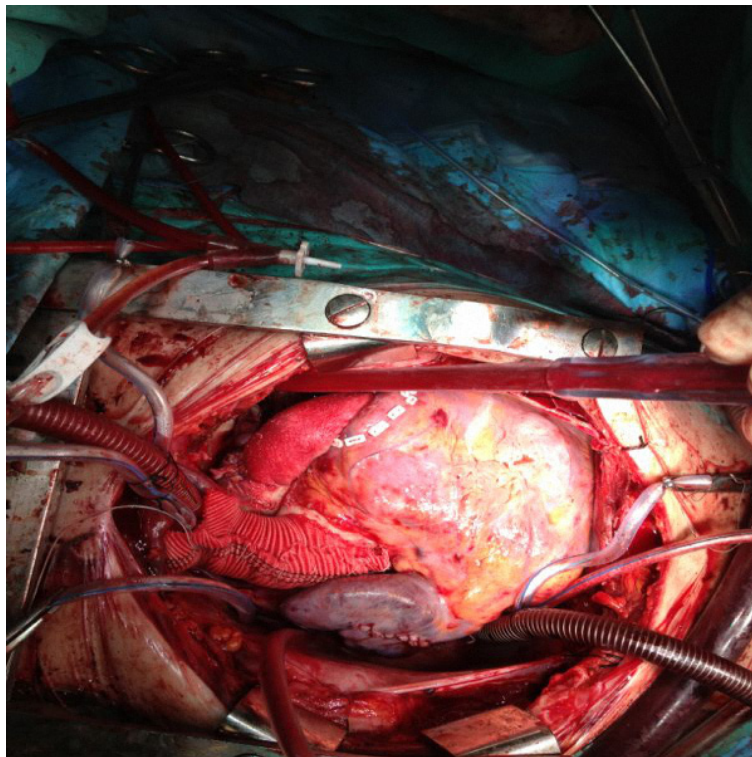


Figure 5. Remodeling of the ascending aorta with aortic linear plication and wrapping technique.

## DISCUSSION

Seventy percent of patients with TOF die before 10 years of age if not surgically treated. For those few who survive, surgical repair of TOF has been shown to improve long-term survival and life expectancy. Tetralogy of Fallot can be surgically managed successfully even in adulthood; however, the operative risk is largely determined by the experience of the cardiac surgical center [Hu 1985]. It is noteworthy that congenital heart defects can be repaired in adults by either a congenital or an adult heart surgeon, with a comparable risk of morbidity and mortality.

Aneurismal dilatation of the main pulmonary artery and its peripheral branches are rare lesions in patients with congenital heart disease, including TOF. The size of the pulmonary artery aneurismal dilatation should guide decisions on whether corrective surgical treatment is appropriate, particularly in severe cases, as with an absent pulmonary valve (which occurs in 3-6% of TOF patients) and pulmonary artery aneurysm, in which the mediastinal structures can be compressed [Conte 1997]. Dilatation of the main pulmonary artery often leads to compression and obstruction of the tracheobronchial tree or to extrinsic compression of the left main coronary artery (LMCA) [Khante 2011].

Patients with untreated TOF who survive infancy are generally those with mild-to-moderate RVOTO with mild or no cyanosis, and they typically do well for another 5-20 years.

However, this group of patients tends to eventually become symptomatic and some die of intractable RV failure as a result of chronic volume overload [Murphy 1993]. In many of these cases surgical repair is feasible and can have successful outcomes. This case study is one of a number of cases that demonstrates that cardiac transplantation is not the only surgical option available to this patient population.

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