

# ‘Superimposed miracles’: an extremely rare case of a septuagenarian with an unrepaired tetralogy of Fallot: a case report

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## Background

Although the life expectancy of patients with tetralogy of Fallot (TOF) is comparable to that of the general population due to advancements in surgical intervention, if untreated, patients with TOF may die during their childhood. However, it has been anecdotally reported that a small number of patients with unrepaired TOF survived into their senescence.

## Case summary

A 71-year-old man with a history of multiple heart failure admissions was referred to our institute after successful cardiopulmonary arrest resuscitation. Transthoracic echocardiography showed the overriding of the aorta on a large ventricular septal defect and right ventricular hypertrophy, along with severe pulmonary stenosis (PS), all of which indicated unrepaired TOF. Computed tomography revealed a patent Blalock–Taussig shunt, which was constructed at the age of 19 years. Coronary angiography revealed multivessel coronary stenoses. Although radical intracardiac repair was not performed due to his multiple comorbidities, his heart failure symptoms were significantly improved owing to proper medication titration. One year following discharge, the patient was well and enjoyed playing golf.

## Discussion

Specific anatomical, functional, and haemodynamic characteristics may be required for the long-term survival of patients with TOF. Pulmonary stenosis should be initially mild to guarantee pulmonary flow during childhood and adolescence, and gradual PS exacerbation should be paralleled with systemic-to-pulmonary collateral development or an extracardiac shunt. Moreover, reduced left ventricular compliance may act as a balancing factor against a right-to-left shunt. The presence of all of these special requirements may have contributed to the unusual survival for this patient.

## Keywords

Tetralogy of Fallot • Haemodynamic balance • Longevity • Natural history • Miracles • Case report

## ESC Curriculum

4.8 Pulmonary stenosis • 5.6 Ventricular arrhythmia • 7.3 Critically ill cardiac patient • 9.7 Adult congenital heart disease

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## Learning points

- Specific anatomical, functional, and haemodynamic characteristics may be required for the long-term survival of patients with unrepaired tetralogy of Fallot.
- Pulmonary stenosis (PS) should be initially mild, and gradual exacerbation of PS should be paralleled with systemic-to-pulmonary collateral development or an extracardiac shunt.
- Reduced left ventricular compliance may act as a balancing factor against a right-to-left shunt.
- The presence of all of these special requirements may have contributed to the unusual survival of the patient in this study.

## Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, accounting for ~7% of all congenital heart diseases.<sup>1</sup> Intracardiac surgical repair is the definitive treatment for TOF and is generally recommended to be performed at the time of diagnosis. If untreated, most patients with TOF typically die during childhood. According to previous reports on the natural course of unrepaired TOF, only 66% of untreated patients lived to the age of 1 year, 24% to 10 years, and 6% to 30 years, and as few as 3% survived to the age of 40 years.<sup>2</sup> However, it has been anecdotally reported that a small number of patients with unrepaired TOF may survive into their senescence.<sup>3–7</sup> Although some anatomical, functional, and haemodynamic characteristics are speculated to be responsible for the long-term survival, the definitive contributing factors remain uncertain.

Herein, we report the extremely rare case of a patient with an unrepaired TOF with only a palliative Blalock–Taussig shunt who, at the time of this report, was alive and in his 70s.

## Timeline

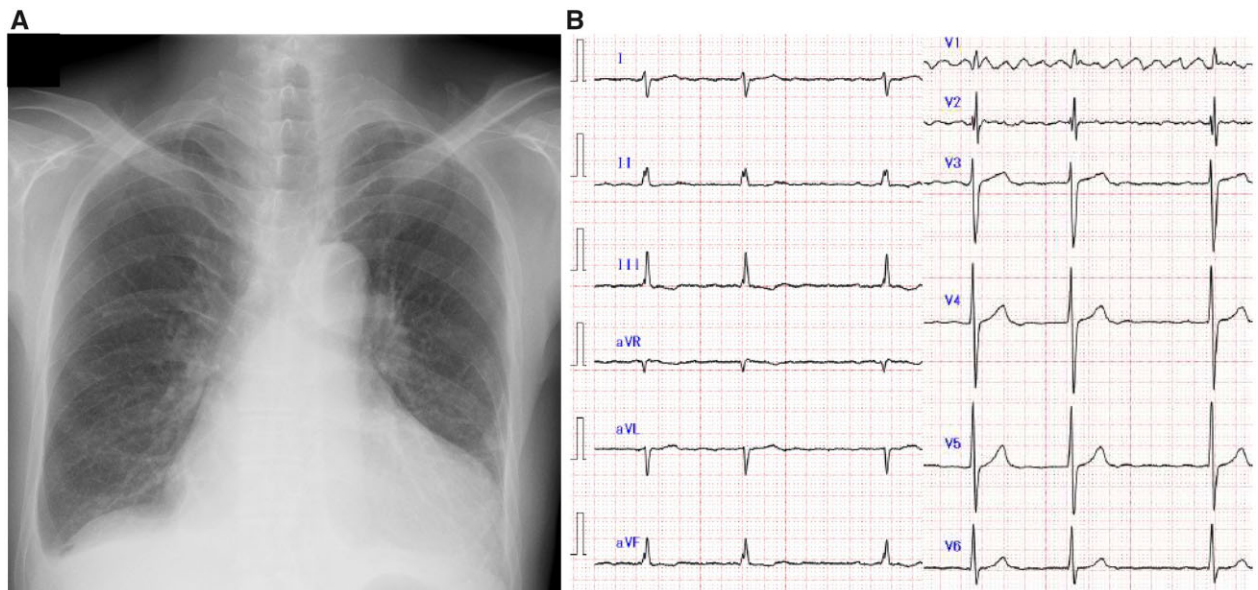
Time	Events
19 years of age	The patient was diagnosed with tetralogy of Fallot.
21 years of age	He underwent palliative Blalock–Taussig shunting. Although radical intracardiac surgery was subsequently recommended, he refused further surgical treatment.
50 years of age	At the age of 50 years, he was admitted due to decompensated heart failure for the first time.
70 years of age	He suffered acute decompensated heart failure again at the age of 70 years.
71 years of age	Nine months after the 2nd heart failure admission, he was readmitted for heart failure.
Initial presentation	Two months after the 3rd heart failure admission, he went into cardiopulmonary arrest due to ventricular fibrillation. Owing to prompt bystander cardiopulmonary resuscitation, he completely recovered without any sequelae.
Admission	Cardiac catheter examination findings showed severe pulmonary stenosis. Coronary angiography revealed multivessel stenosis. For the secondary prevention of ventricular arrhythmia, a subcutaneous implantable cardioverter-defibrillator was implanted. After obtaining a normal sinus rhythm by electrical cardioversion, the introduction of $\beta$ -blocker and titration of oral diuretics, his haemodynamics significantly improved.
After discharge	One year after discharge, his heart failure symptoms improved to New York Heart Association functional class II.

## Case presentation

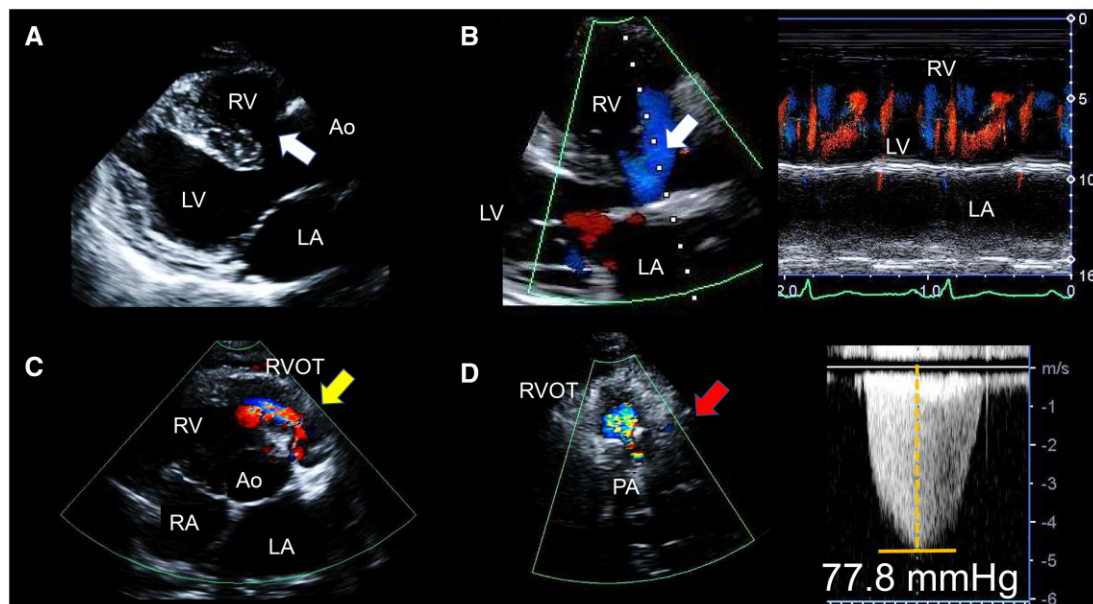
A 71-year-old man treated for chronic kidney disease, paroxysmal atrial fibrillation (AF) and chronic heart failure experienced cardiopulmonary arrest due to unexpected ventricular fibrillation (VF). Fortunately, owing to prompt bystander cardiopulmonary resuscitation, the patient completely recovered without any neurological disabilities. Upon admission in the previous institution, serum potassium levels were revealed to be significantly decreased to 3.3 mEq/L; therefore, it was speculated that VF was induced by incidental hypokalaemia. The patient was subsequently referred to our institution for further evaluation.

Medical history taking revealed that the patient had a heart murmur throughout his childhood, yet this matter was not further evaluated. At the age of 19 years, the patient was diagnosed with TOF and underwent palliative Blalock–Taussig shunting at the age of 21 years. Although radical intracardiac surgery was recommended, the patient refused further surgical treatment. For a while, he worked and enjoyed playing golf without any heart failure symptoms; however, he was eventually admitted due to heart failure at 50, 70, and 71 years of age.

Upon referral, his blood pressure was 114/58 mmHg, heart rate was 69 b.p.m., and oxygen saturation was 89% (ambient air). On visual inspection, he had lip cyanosis and clubbing; however, no peripheral oedema was observed. Cardiac auscultation revealed a harsh systolic ejection murmur in the 2nd to 4th intercostal space of the left sternal border. Laboratory findings revealed polycythaemia with a haemoglobin level of 18.0 g/dL and a haematocrit of 52.6%. His serum creatinine levels were 1.8 mg/dL, and brain natriuretic peptide concentration was significantly increased at 697 pg/mL. Chest radiography showed a 'boot-shaped' heart with bilateral pleural effusion (*Figure 1A*), and electrocardiography revealed an AF rhythm (*Figure 1B*). Transthoracic echocardiography showed the overriding of the aorta on a large ventricular septal defect (VSD) that was 32 mm in



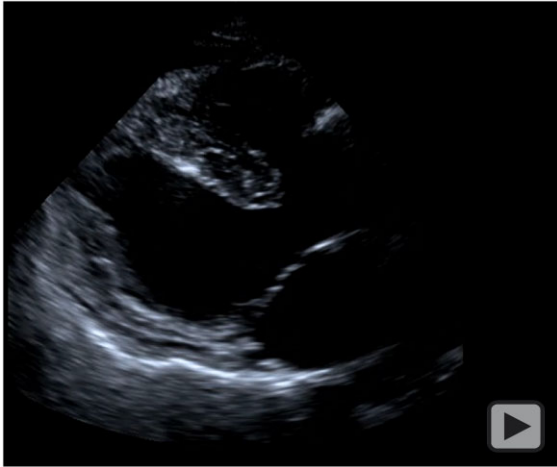
**Figure 1** Chest radiography and electrocardiography in a 71-year-old man. Chest radiography revealed significant cardiomegaly and bilateral pleural effusion (A). Electrocardiography revealed atrial fibrillation (B).



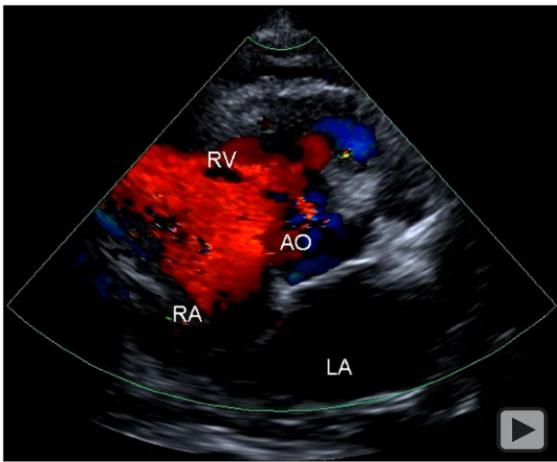
**Figure 2** Transthoracic echocardiography in a 71-year-old man. Overriding of the aorta on a large ventricular septal defect (VSD, white arrow) was observed (A). On colour Doppler M-mode echocardiography, shunt flow through ventricular septal defect was observed to be bidirectional (B). A turbulent colour Doppler signal was observed from the right ventricular outflow tract to the pulmonary valve, suggesting significant pulmonary stenosis (C). A continuous wave Doppler echocardiogram revealed a pressure gradient at the stenotic pulmonary valve of 77.8 mmHg (D). Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle; RVOT, right ventricular outflow tract; VSD, ventricular septal defect.

diameter (Figure 2A, white arrow, Video 1) and right ventricular (RV) hypertrophy, along with pulmonary stenosis (PS; Video 2), all of which were indications of unrepaired TOF. On colour Doppler

echocardiography, a bidirectional shunt flow through the VSD was clearly observed (Figure 2B). Moreover, the pulmonary valve was markedly calcified, making leaflet mobility significantly restricted

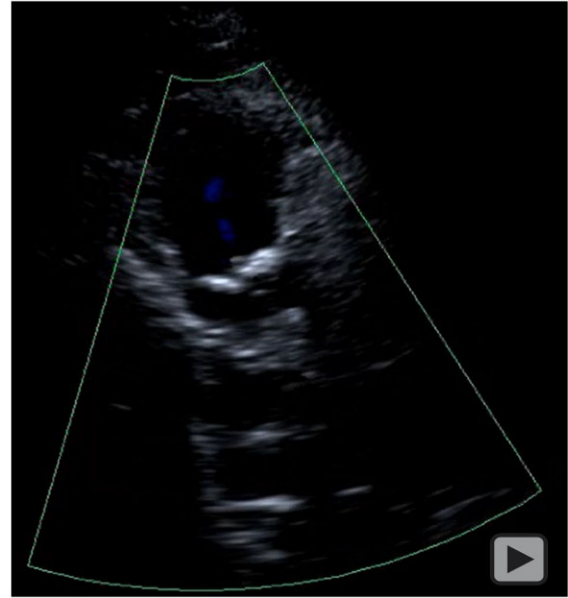


**Video 1** Parasternal long-axis view showing the overriding of the aorta on a large ventricular septal defect.



**Video 2** Parasternal right ventricular inflow-outflow view shows the narrowing of the right ventricular outflow tract and a significant colour convergence, suggesting significant stenosis.

(Video 3). Continuous wave Doppler echocardiography showed a significant pressure gradient of 77.8 mmHg across the pulmonary valve, confirming the presence of severe valvular PS (Figure 2C and D). Left ventricular (LV) ejection fraction decreased to 46.8%, and the base-to-mid posterior and inferoseptal wall motion abnormality was observed. To assess the detailed intra- and extracardiac anatomy of this patient, the patient underwent multidetector-row computed tomography, and the findings of the acquired images were consistent with the characteristics of unrepaired TOF. The findings revealed a large VSD with aortic overriding (Figure 3A, white arrow), severe PS (Figure 3B, red arrow), and RV hypertrophy. Although major aortopulmonary collateral arteries were absent, a

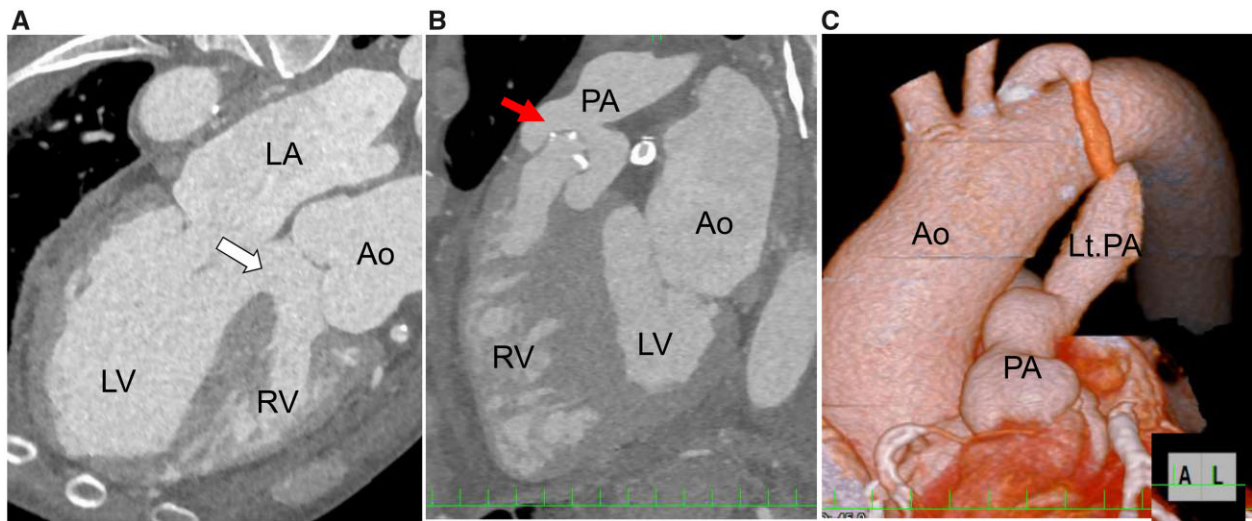


**Video 3** Right ventricular outflow view shows pulmonary valve calcification and immobility, at which the turbulent colour of the Doppler signal is clearly detected.

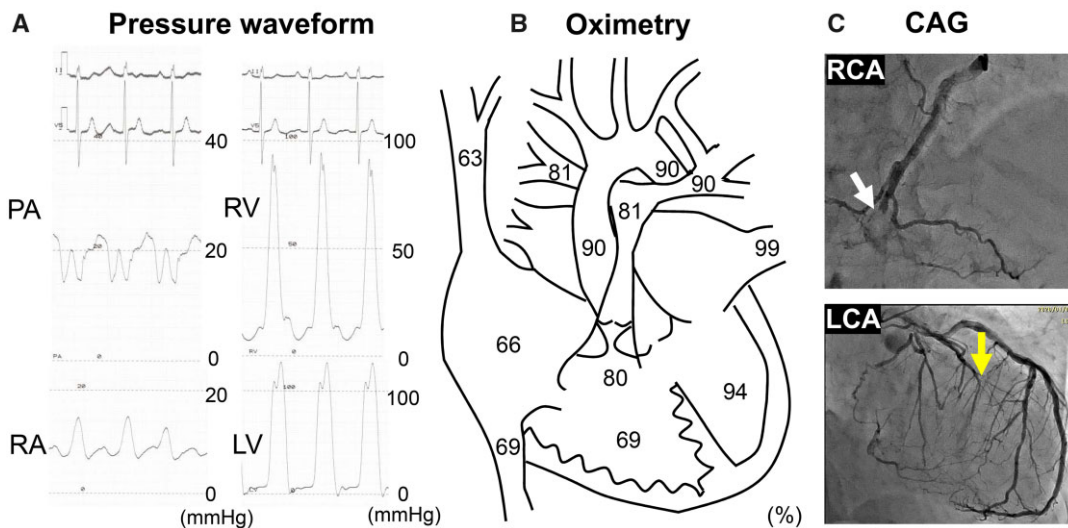
relatively restrictive Blalock–Taussig shunt was clearly observed (Figure 3C, yellow arrow). Functional assessment using cardiac magnetic resonance imaging revealed RV dilatation (end-diastolic volume, 192 mL; end-systolic volume, 146 mL) and reduced RV ejection fraction (RVEF, 24%). Cardiac catheter examination revealed a pulmonary arterial pressure of 23/14 mmHg (mean, 18 mmHg), an RV pressure of 96/7 mmHg (end-diastolic pressure, 12 mmHg), and a pulmonary vascular resistance of 1.45 wood unit. Pressure waveforms and oximetry run results are presented in Figure 4A and B, respectively. Coronary angiography showed the total occlusion of the mid-portion of the right coronary artery (Figure 4C, white arrow) and the proximal portion of the left circumflex coronary artery (Figure 4C, yellow arrow) and the critical stenosis of the diagonal branch of the left anterior descending artery.

To improve the unstable haemodynamics, electrical cardioversion was performed on the 6th day of hospitalization. To maintain a normal sinus rhythm and suppress ventricular arrhythmia, amiodarone (100 mg/day) was introduced. Because the patient had multiple comorbidities, radical intracardiac repair was not performed due to significant surgical risk. Regarding the less invasive transcatheter intervention, coronary reperfusion therapy was withheld due to the absence of inducible ischaemia on the basis of the findings of stress myocardial scintigraphy. On the other hand, because it was expected that palliative transcatheter intervention for PS could even impair exquisite haemodynamic balance for this patient, transcatheter pulmonary valvuloplasty was not applied for this case. Alternatively, for the secondary prevention of lethal ventricular arrhythmia, a subcutaneous implantable cardioverter-defibrillator was





**Figure 3** Cardiac multidetector-row computed tomographic images in a 71-year-old man. Overriding of the aorta on a large ventricular septal defect (white arrow) was clearly observed (A). The pulmonary valve (red arrow) was stenotic with dense calcification (B). A volume-rendered image showed a restrictive Blalock–Taussig shunt between the left subclavian artery and left pulmonary artery (C). Ao, Aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; VSD, ventricular septal defect.



**Figure 4** Intracardiac pressure waveforms and oximetry run results in a 71-year-old man. Pulmonary arterial pressure was 23/14 mmHg (mean, 18 mmHg), and right ventricular pressure was 96/7 mmHg (end-diastolic pressure, 12 mmHg), indicating severe pulmonary stenosis (A). The oximetry run results during the cardiac catheter examination are shown (B). Coronary angiography showed the total occlusion of the mid-portion of the right coronary artery (white arrow) and the proximal portion of the left circumflex coronary artery (yellow arrow) (C). CAG, coronary angiography; LCA, left coronary artery; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RCA, right coronary artery; RV, right ventricle.

implanted. After obtaining a normal sinus rhythm,  $\beta$ -blocker use and oral diuretic titration, his haemodynamics significantly improved. Eventually, the patient was discharged on the 55th day of hospital admission.

One year after discharge, his heart failure symptoms improved to New York Heart Association functional class 2.

## Discussion

Previous investigators have reported that few patients with TOF can survive into their adulthood with an average life expectancy of 12 years.<sup>2</sup> Nevertheless, only a few patients reportedly able to survive into old ages.<sup>8,9</sup> According to previous

sporadic case reports, some anatomical, functional, and haemodynamic requirements may be the reason behind such a prolonged survival. First, PS should be initially mild, which guarantees that pulmonary flow is sufficient for systemic oxygenation during childhood to adolescence. Second, exacerbation of PS should be paralleled with systemic-to-pulmonary collateral development or an extracardiac shunt, which compensates for the reduced pulmonary blood flow.<sup>10</sup> Third, reduced LV compliance could mitigate the right-to-left shunt.<sup>7</sup> In this case, PS might initially be mild during childhood, allowing the normal growth of the pulmonary vascular bed and the left side of the heart and guaranteeing the systemic supply of oxygenated blood flow. Moreover, pulmonary hypoperfusion due to progressive development of PS could be well compensated by Blalock–Taussig shunting, which was miraculously timed in this case. Conversely, it has been reported that prolonged systemic-to-pulmonary shunting through Blalock–Taussig shunt could lead to pulmonary overcirculation and adverse effects on the pulmonary vascular bed. Hofschire et al.<sup>11</sup> histologically analysed the pulmonary vasculature in 36 patients with congenital heart diseases who underwent Blalock–Taussig shunting.<sup>11</sup> They reported that half of the patients developed obvious obstructive pulmonary disease following 8 years of shunting. Of note, severe histological damage was observed in 50% of patients who developed obstructive vascular changes.<sup>11</sup> These results suggest that the Blalock–Taussig shunt can initially be beneficial for patients with pulmonary hypoperfusion, yet prolonged shunting may have adverse effects on the pulmonary vasculature. However, in this case, because the Blalock–Taussig shunt has remained restrictive, the pulmonary vascular bed may have miraculously avoided overcirculation and the consequent obstructive pulmonary vascular damage, or the shunt may have supplied appropriate blood flow for pulmonary circulation, allowing the patient to survive into senescence. Furthermore, in this case, impaired LV compliance due to multivessel ischaemia might have acted as a balancing factor against right-to-left shunting, possibly preventing systemic hypoxia and the resultant hypoxic end-organ damage.

## Conclusions

We encountered an extremely rare case of a patient with unrepaired TOF who, at the time of this report, was surviving through his 70s with only a palliative Blalock–Taussig shunt. To survive senescence, several important anatomical, functional, and haemodynamic requirements should be simultaneously fulfilled. Although it is speculative, it may be argued that ‘Superimposed miracles’ were the factors contributing to the prolonged survival for this special case.

## Lead author biography



After graduating from Kobe university in 2013, I went into the path of cardiology after 2 years of residency. After studying general cardiology for 3 years, I have conducted research activities focusing on echocardiography for 3 years at Kobe University.

## Supplementary material

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for the submission and publication of this case report, including images and associated text, has been obtained from the patient in line with the COPE guidance.

**Conflict of interest:** None declared.

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