

# Survival to the age of 87 years in a woman with unoperated tetralogy of Fallot

Ty K. Subhawong<sup>1\*</sup>, Oleg Teytelboym<sup>2</sup>

1. The Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins School of Medicine, Baltimore, MD, USA

2. The Department of Radiology, Johns Hopkins Bayview Medical Center, Baltimore, MD, USA

\* **Correspondence:** Ty K. Subhawong, M.D., The Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins School of Medicine, 601 N Caroline Street, Baltimore, MD 21287, USA  
(✉ [tsubhaw1@jhmi.edu](mailto:tsubhaw1@jhmi.edu))

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## ABSTRACT

Tetralogy of Fallot is the most common cyanotic congenital heart defect, affecting approximately 2700 infants per year born in the United States. The natural history of the tetralogy reflects the adverse physiologic consequences of the underlying structural abnormalities, with only approximately 3% of uncorrected patients surviving past age 40. In this case report, we report an 87-year-old woman with unoperated tetralogy of Fallot, who we believe to be the oldest and only second octogenarian described in the literature.

## CASE REPORT

### CASE REPORT

An 87 year old woman presented to the emergency department with an acute onset of confusion, aphasia, and right hemiparesis of the face and limbs. Prior to this episode, she had been able to perform all activities of daily living. She had been followed by a cardiologist and had known uncorrected Tetralogy of Fallot, for which she had refused repair. Physical exam revealed a 6/6 holosystolic murmur. MRI demonstrated acute infarcts in the left basal ganglia and parietal lobes (figures 1 and 2). Old infarcts in the posterior left and right frontal lobes and posterolateral right lateral hemisphere were also found. Contrast enhanced coronary CT (figures 3-5) demonstrated right aortic arch, a large ventricular septal defect (VSD) with overriding aorta, and right ventricular outflow tract (RVOT) narrowing with right ventricular hypertrophy. The main pulmonary artery was enlarged to 3.3 cm, and the right atrium was dilated. Nonobstructive coronary artery disease was present. Echocardiogram confirmed the above findings. Additionally, a 1.9 x 1.4 cm echogenic focus in the

inferior vena cava (figure 6) was thought to represent thrombus. Approximately three months after initial presentation, the patient was residing in a nursing home, with residual cognitive deficits and both expressive and receptive aphasia.

### DISCUSSION

Tetralogy of Fallot, first described in 1888 by the French physician Etienne-Louis Arthur Fallot, is the most common cyanotic congenital heart abnormality, affecting approximately 2700 infants per year born in the United States (1), and accounting for 10% of cardiac defects detected in infancy (2). The tetralogy is classically comprised of four components: large ventricular septal defect, an aorta that overrides the left and right ventricles, obstruction of the right ventricular outflow tract, and compensatory right ventricular hypertrophy. Additional anomalies occur with varying frequencies, such as a

right aortic arch (25%), atrial septal defect (10%), and coronary anomalies (10%) (3). The presence of the large VSD permits equalization of pressures across the right and left ventricles, with resultant right-to-left shunting and cyanosis due to higher resistance to flow in the RVOT (3). Since the Blalock-Taussig procedure was described in 1945 (4), surgeons have been able to offer palliative or definitive repairs to patients from infancy to adulthood (1; 5). Previously used palliative treatments involving the anastomosis of a systemic artery to a pulmonary artery have given way in the modern era to complete surgical repair of the VSD and RVOT obstruction at an early age, with less than 3% operative mortality reported in young patients (3). Young patients can expect favorable outcomes, with long-term data demonstrating that 30-year actuarial survival rate for patients with corrected tetralogy of Fallot was 90 percent of the expected survival rate (1).

Betranou, et al., reported survival without surgery to be 66% at 1 year of age, 40% at 3 years, 11% at 20 years, 6% at 30 years, and 3% at 40 years (6). Since 1929, when the first report of a patient with uncorrected tetralogy of Fallot surviving into late adulthood was made (7), there have been various papers in the world literature of surprising longevity in such patients, several of whom lived into their seventh and eighth decades (8-10). Several years ago, an 86 year old man with uncorrected tetralogy of Fallot was reported as the oldest known surviving case (8). The ability of such patients to survive into senescence putatively owes to a lesser degree of RVOT obstruction, leading to a well-balanced bidirectional shunt or acyanotic left-to-right shunt (8; 9). Yang, et al., reported that unoperated survivors tend to have three common features, including a hypoplastic pulmonary artery, left ventricular hypertrophy, or systemic-pulmonary artery collaterals for pulmonary blood flow (10). They hypothesize that these patients benefit from relatively slow development of subpulmonary obstruction. While no definite systemic-pulmonary artery collaterals were demonstrated, the remainder of CT findings in our patient are consistent with such physiology. The morphological features of the tetralogy are depicted well by CT, and additional anatomic detail, such as variant coronary anatomy and extracardiac vascular structure, can be of particular importance for pre-surgical planning and post-operative follow-up (11; 12). The post-surgical CT findings in corrected patients have been reviewed elsewhere (13).

Determining at what age a patient's surgical risk outweighs the benefits of operating is controversial and must be made in the context of the patient's clinical circumstances and personal desires. A retrospective analysis of surgical repair in adulthood (mean age 29 years) of 52 patients with tetralogy of Fallot or pulmonary atresia and VSD documented a 15% early post-operative mortality (14). Other studies have demonstrated slightly lower early mortality rates, and arguably, with appropriate population selection, improvements in functional status and the potential for long-term survival outweigh operative risks (15). In addition to primary cardiac morbidities such as biventricular failure and sudden cardiac death (16; 17), adults with cyanotic congenital heart disease are known to develop a compensatory erythrocytosis as a result of chronic hypoxemia. The consequent elevation in blood viscosity may predispose these patients to an increased risk of cerebrovascular accidents or venoocclusive disease (18-

20), although these risks are not as well established as in the pediatric population (21; 22). Interestingly, a case of paradoxical embolus to the brain from an IVC thrombus has been reported in the pediatric literature (23); that a similar mechanism was responsible for the symptoms suffered by our patient is a compelling possibility.

#### TEACHING POINT

Adult patients with uncorrected tetralogy of Fallot rarely live beyond the age of 40, with this case of an 87 year old unoperated patient being the oldest and only second octogenarian reported. Morbidity in such patients may stem not only from primary cardiac events, but also secondary neurological deficits incurred by their underlying disease. Along with echocardiography, CT serves as an important modality in investigating congenital cardiac abnormalities of such patients, particularly in defining extracardiac anatomy.

#### REFERENCES

1. Murphy JG, Gersh BJ, Mair DD, Fuster V, McGoon MD, Ilstrup DM, McGoon DC, Kirklin JW, Danielson GK. Long-Term Outcome in Patients Undergoing Surgical Repair of Tetralogy of Fallot. *N Engl J Med*. 1993 Aug 26;329(9):593-599.
2. Pinsky WW, Arciniegas E. Tetralogy of Fallot. *Pediatr Clin North Am*. 1990 Feb;37(1):179-92.
3. Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. Second of two parts. *N Engl J Med*. 2000 Feb 3;342(5):334-42.
4. Blalock A, Taussig HB. Landmark article May 19, 1945: The surgical treatment of malformations of the heart in which there is pulmonary stenosis or pulmonary atresia. By Alfred Blalock and Helen B. Taussig. *JAMA*. 1984 Apr 27;251(16):2123-38.
5. Hu DC, Seward JB, Puga FJ, Fuster V, Tajik AJ. Total correction of tetralogy of Fallot at age 40 years and older: long-term follow-up. *J Am Coll Cardiol*. 1985 Jan ;5(1):40-4.
6. Bertranou EG, Blackstone EH, Hazelrig JB, Turner ME, Kirklin JW. Life expectancy without surgery in tetralogy of Fallot. *Am J Cardiol*. 1978 Sep ;42(3):458-66.
7. White PD, Sprague HB. Tetralogy of Fallot: Report of a case in noted musician, who lived to his 60th year. *JAMA*. 1929 ;92787-790.
8. Alonso A, Downey BC, Kuvin JT. Uncorrected tetralogy of Fallot in an 86-year-old patient. *Am J Geriatr Cardiol*. 2007;16(1):38-41.
9. Makaryus AN, Aronov I, Diamond J, Park CH, Rosen SE, Stephen B. Survival to the age of 52 years in a man with unrepaired tetralogy of Fallot. *Echocardiography*. 2004 Oct ;21(7):631-7.
10. Yang X, Freeman LJ, Ross C. Unoperated tetralogy of Fallot: case report of a natural survivor who died in his 73rd year; is it ever too late to operate? *Postgrad Med J*. 2005 Feb ;81(952):133-4.
11. Leschka S, Oechslin E, Husmann L, Desbiolles L, Marincek B, Genoni M, Pretre R, Jenni R, Wildermuth S, Alkadhi H. Pre- and

Postoperative Evaluation of Congenital Heart Disease in Children and Adults with 64-Section CT. *Radiographics*. 2007 May 1;27(3):829-846.

12. Wang X, Wu L, Sun C, Liu C, Chao B, Han B, Zhang Y, Chen H, Li Z. Clinical application of 64-slice spiral CT in the diagnosis of the Tetralogy of Fallot. *European Journal of Radiology*. 2007 Nov ;64(2):296-301.

13. Spevak PJ, Johnson PT, Fishman EK. Surgically Corrected Congenital Heart Disease: Utility of 64-MDCT. *Am. J. Roentgenol*. 2008 Sep 1;191(3):854-861.

14. Hörer J, Friebe J, Schreiber C, Kostolny M, Cleuziou J, Holper K, Lange R. Correction of tetralogy of Fallot and of pulmonary atresia with ventricular septal defect in adults. *Ann Thorac Surg*. 2005 Dec ;80(6):2285-91.

15. Sadiq A, Shyamkrishnan KG, Theodore S, Gopalakrishnan S, Tharakan JM, Karunakaran J. Long-term functional assessment after correction of tetralogy of Fallot in adulthood. *Ann Thorac Surg*. 2007 May ;83(5):1790-5.

16. Pelech AN, Neish SR. Sudden death in congenital heart disease. *Pediatr Clin North Am*. 2004 Oct ;51(5):1257-71.

17. Oechslin EN, Harrison DA, Connelly MS, Webb GD, Siu SC. Mode of death in adults with congenital heart disease. *Am. J. Cardiol*. 2000 Nov 15;86(10):1111-1116.

18. DeFilippis AP, Law K, Curtin S, Eckman JR. Blood is thicker than water: the management of hyperviscosity in adults with cyanotic heart disease. *Cardiol Rev*. 2007 ;15(1):31-4.

19. Perloff J, Marelli A, Miner P. Risk of stroke in adults with cyanotic congenital heart disease. *Circulation*. 1993 Jun 1;87(6):1954-1959.

20. Ammash N, Warnes CA. Cerebrovascular events in adult patients with cyanotic congenital heart disease. *J Am Coll Cardiol*. 1996 Sep ;28(3):768-72.

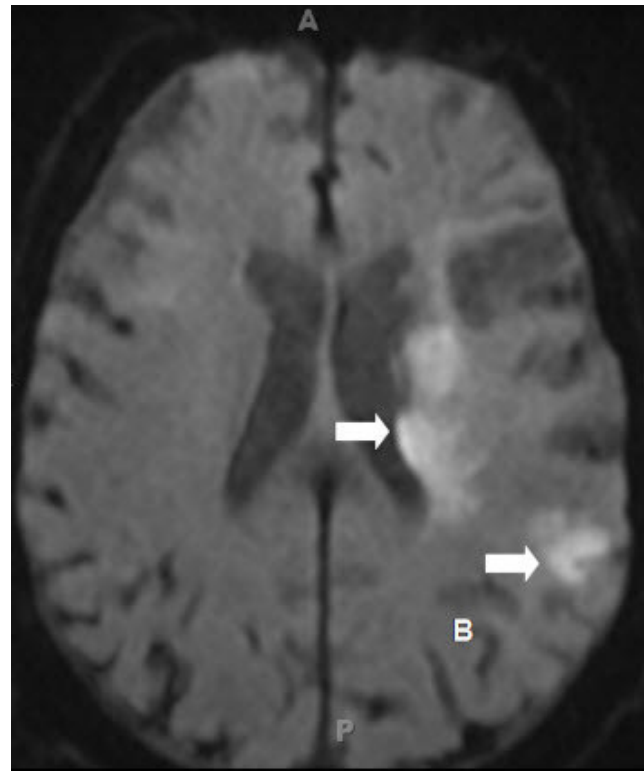
21. Amitai Y, Blieden L, Shemtov A, Neufeld H. Cerebrovascular accidents in infants and children with congenital cyanotic heart disease. *Isr J Med Sci*. 1984 Dec ;20(12):1143-5.

22. Phornphutkul M, Rosenthal M, MDAlex, Nadas ES, Berenberg M. Cerebrovascular accidents in infants and children with cyanotic congenital heart disease. *Am J Cardiol*. 1973 ;32(3):329-334.

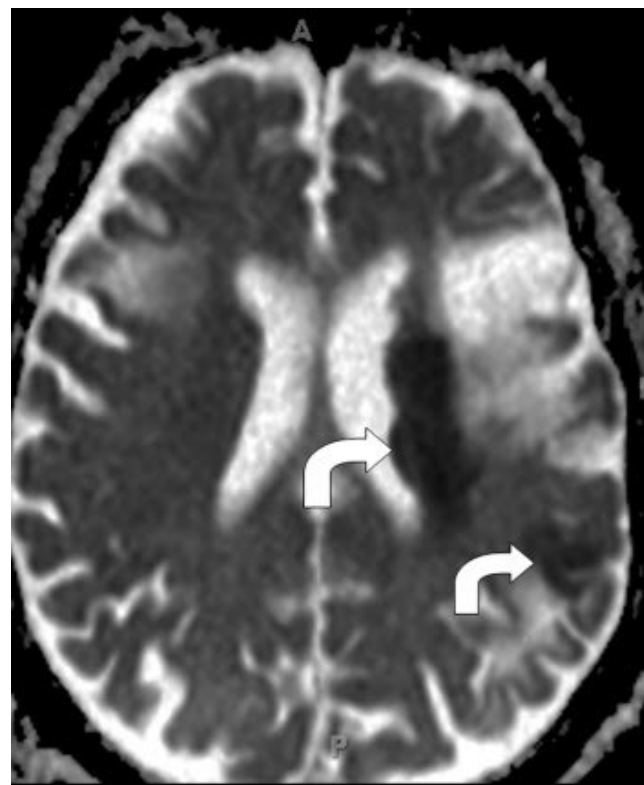
23. Geva T, Frand M, Benjamin P, Hegesh J. Cerebral embolization from an inferior vena cava thrombus in tetralogy of Fallot. *Pediatr Cardiol*. 1990 Jan ;11(1):44-6.

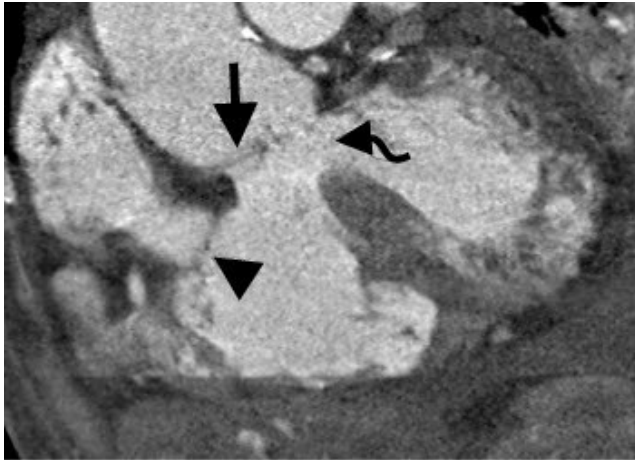
**Figure 2 (right).** Axial ADC map of brain demonstrates areas of low signal intensity (arrows) confirming acute infarct in the vascular distribution of the left middle cerebral artery.

FIGURES



**Figure 1.** 87 year old woman with tetralogy of Fallot. Axial diffusion weighted image of the brain demonstrates high signal abnormalities in the region of the left basal ganglia and left parietal lobe (arrows) consistent with restricted diffusion.





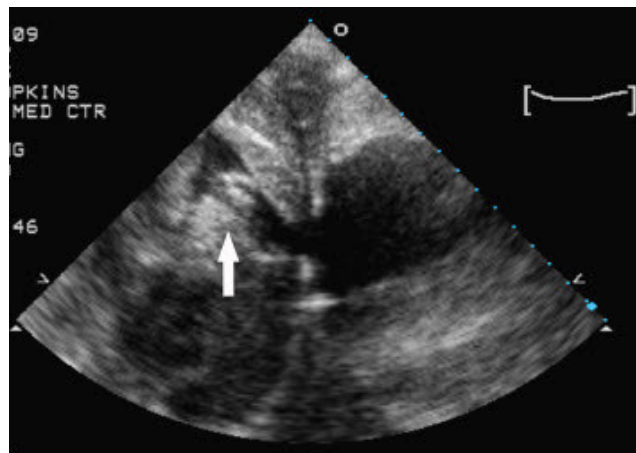
**Figure 3.** 87 year old woman with tetralogy of Fallot. Sagittal oblique reconstruction of contrast enhanced CT of the chest in arterial phase demonstrates ventricular septal defect (curved arrow), overriding aorta (straight arrow), and narrowed pulmonary outflow tract (arrowhead).



**Figure 5.** Axial contrast enhanced CT of the chest in arterial phase demonstrates normal origin of the left main coronary artery from left aortic sinus (arrowhead) and right descending thoracic aorta (straight arrow).



**Figure 4.** Axial contrast enhanced CT of the chest in arterial phase shows enlarged right atrium (curved arrow) and right ventricular hypertrophy (straight arrow).



**Figure 6.** A transthoracic echocardiogram demonstrates an echogenic filling defect (arrow) in the inferior vena cava compatible with a thrombus, presumed to be the source of an acute embolic stroke in the territory of the left middle cerebral artery (see figures 1 and 2).

**ABBREVIATIONS**

CT: computed tomography  
 IVC: inferior vena cava  
 VSD: ventricular septal defect  
 RVOT: right ventricular outflow tract  
 MRI: magnetic resonance imaging  
 ADC: apparent diffusion coefficient

**KEYWORDS**

Congenital heart disease, cardiovascular CT, tetralogy of Fallot

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