

Oldest survivors after repair of tetralogy of Fallot

Pia K. Schuler^a, Erwin Oechslin^b, Juraj Turina^a, Rolf Jenni^a

^a Department of Cardiology, University Hospital Zurich, Zurich, Switzerland

^b Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network / Toronto General Hospital, Toronto, Canada

Summary

We report two cases of a 83- and a 78-year-old female with tetralogy of Fallot (TOF) who underwent operation at an advanced age in their 30's, making them the oldest patients repaired for tetralogy of Fallot. The patients remained fairly asymptomatic for much of their life. Survival until the age of late 70 or early 80 is possible in a historical cohort of patients with a balanced physiology between the systemic and pulmonary circulation which allowed them to survive until surgical repair became available in the 1960s.

Key words: tetralogy of Fallot; oldest survivors; operation

Case report

We present two patients believed to be the oldest survivors after repair of tetralogy of Fallot.

The first case describes a 83-year-old woman who was diagnosed with a congenital heart defect at the age of five months. As a child she suffered from recurrent syncope. At the age of 38 an angiography revealed a large ventricular septal defect and an infundibular pulmonary stenosis. The systemic output was measured to be 6 l/min, the pulmonary flow was 4.5 l/min.

After the diagnosis of tetralogy of Fallot she was operated at the age of 38 by Åke Senning in 1962. The operative report describes a hypertrophied right ventricle (RV) with a myocardial thickness of 1.5 cm causing infundibular outflow obstruction; the pulmonary valve was well developed and valvular pulmonary stenosis was absent. The ventricular septal defect localised in a high position showed a diameter of 2 cm. The hypertrophic myocardium was resected and the right outflow tract was enlarged by a pericardial patch. The ventricular septal defect was closed by a Dacron patch. The patient tolerated the operation well despite her advanced age.

In 1996 and 2003 the patient suffered from left ventricular heart failure, but she was compensated at the time of her clinical visit in 2007. The echocardiogram at that time showed a combined pulmonary valve disease with a mean systolic gradient of 14 mm Hg and mild pulmonary insuffi-

ciency. Both ventricles were of normal size and normal systolic function. Moderate aortic regurgitation was present, but no residual VSD was documented. The ECG revealed sinus rhythm with a QRS duration of 92 msec.

The patient described mild dyspnea on exertion but she denied any other cardiac symptoms under a well established heart failure therapy. The patient is still alive.

Tetralogy of Fallot was complicated by an endocarditis with right heart failure in the second patient. In 1961, Åke Senning operated on her at the age of 34 in 1961 after therapy of her endocarditis.

The operative report describes a hypertrophied right ventricle with a myocardial thickness of 1.2 cm causing infundibular stenosis of pencil-sized lumen, which was heavily calcified. The severe stenosis across the pulmonary outflow tract was repaired with a transannular patch. The small ventricular septal defect was closed with five stitches.

Atrial flutter was diagnosed 47 years after the operation. At the age of 78 the patient was hospitalised with a metastatic uterus carcinoma and an endocarditis of the aortic valve. The echocardiogram at that time revealed severe pulmonary insufficiency without obstruction across the right outflow tract. The pulmonary annulus was calcified and the pulmonary valve cusps were thickened. The pulmonary artery showed an aneurysm measuring 4.6 cm in diameter and a dilatation of the left and right pulmonary artery. There was no residual VSD. The patient died from a cardiogenic/hypovolaemic shock after the new onset of a complete AV block in combination with a heavy vaginal bleeding.

The autopsy showed extensive eccentric hypertrophy of both ventricles with a heart weighing 675 g (BMI 26 kg/m²) 45 years after repair of tetralogy of Fallot. The

There is no conflict of interest.

Correspondence:

Pia Schuler
Clinic of Cardiology University Hospital Zurich
Raemistrasse 100
CH-8091 Zurich
Switzerland
schulerp@gmx.de

thickness of the right ventricular myocardium was 7 mm. A large aneurysm of the conus pulmonalis with a diameter of 4.5 cm and a massive dilatation of the pulmonary artery were confirmed. The pulmonary artery was fibrosed and calcified, and a persistent foramen ovale was detected.

Discussion

Tetralogy of Fallot, first described by Etienne Fallot [1] (fig. 1) in the classic paper "l'anatomie pathologique de la maladie bleue" in 1888, is the most common cyanotic congenital heart defect (6%). It comprises an inter-ventricular septal defect, right ventricular outflow tract obstruction, an overriding aorta, and a right ventricular hypertrophy (fig. 2 and 3).

Figure 1
Etienne-Louis Fallot (1850–1911).



Figure 2

A diagram showing a healthy heart and one suffering from the tetralogy of Fallot, which comprises an inter-ventricular septal defect, right ventricular outflow tract obstruction, an overriding aorta, and a right ventricular hypertrophy. (From: Mariana Ruiz Villarreal, Hamburg, Germany. Website: http://wpcontent.answers.com/wikipedia/commons/thumb/e6/Tetralogy_of_Fallot.svg/190px-Tetralogy_of_Fallot.svg.png. This image has been released into the public domain by its author, LadyofHats.)

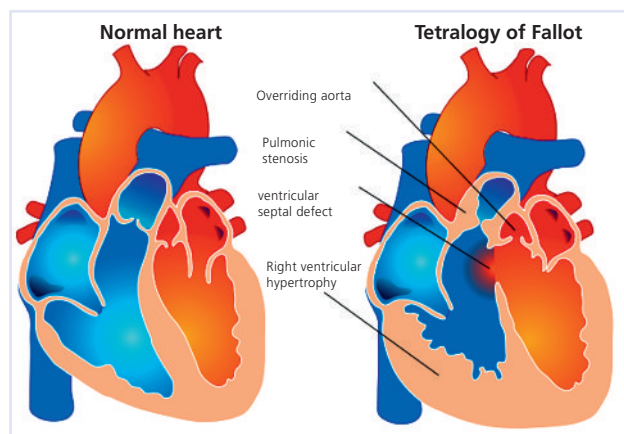
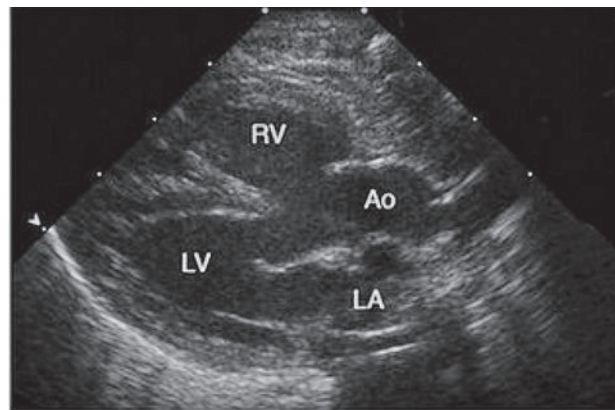


Figure 3

Echocardiogram with anterior displacement of the outflow ventricular septum that results in stenosis of the subpulmonic right ventricular outflow tract, overriding of the aorta, and an associated ventricular septal defect. (This picture was published in: Nelson Textbook of Pediatrics. Kliegman RM, Behrman RE, Jenson HB, Stanton BF, eds. 18th edition. Oxford: W. B. Saunders; 2007. Copyright © 2009 Elsevier, Oxford, UK.)

Ao = overriding aorta; LA = left atrium; LV = left ventricle; RV = right ventricle.



tract obstruction, an overriding aorta, and a right ventricular hypertrophy (fig. 2 and 3).

Helen Tausig, a paediatric cardiologist, observed that children with a patent ductus arteriosus were doing better or died after closure of the ductus arteriosus. Her idea was to create an "arti-

ficial" ductus arteriosus to improve cyanosis in these children. The first palliative anastomosis was done between the subclavian and the ipsilateral pulmonary artery to increase pulmonary blood flow (Blalock-Taussig-Anastomosis, 1945 [2]). In 1954, Lillehei and collaborators, using controlled cross circulation in a 10-month-old boy, carried out the first intracardiac repair of tetralogy of Fallot [3]. The first successful repair of tetralogy of Fallot using a heart-lung-machine was accomplished by Kirklin and associates in 1955.

Children with tetralogy of Fallot can survive until adulthood only if there is a balance between the pulmonary and systemic circulation. Thus, only few patients with tetralogy of Fallot survive into adulthood without operation. Without repair the mean life expectancy is 12 years and only 3% reach their 40s or older [4]. Unoperated survivors show three characteristics for longevity: hypoplastic pulmonary artery and moderately slow development of subpulmonary obstruction, left ventricular hypertrophy, or systemic-pulmonary artery collaterals for pulmonary blood flow [5]. Our two patients were not treated until the age of 38 and 34. It is not known if the above features of longevity were present.

There are few studies on the outcome and benefit of late surgical repair. In the Mayo series [6] 30 patients were followed after repair of tetralogy of Fallot between the ages of 40 and 60 years. The operative mortality was 3% with a 10-year-survival of 74%.

Murphy [7] showed an overall 32-year-survival rate of 86% in 163 patients after a repair of tetralogy of Fallot. The survival rate was 76% among patients 12 years old or older at the time of surgery. Nollert [8] showed

that the long-term outcome after repair of tetralogy of Fallot was influenced by cyanosis, operative experience of the surgeon and an RV outflow patch in older children. Independent predictors of long-term mortality were older age at repair, previous heart failure and a higher ratio of right ventricular to left ventricular systolic pressure after surgery (>0.5). Patients with preceding Blalock-Taussig shunt procedure had the same mortality rate as patients without a previous palliative surgery. Transannular patching has been described as a risk factor for late pulmonary valvular insufficiency and right ventricular volume overload leading to a high rate of reoperation and late mortality [9, 10], but other studies showed no differences compared to patients without a transannular patch [11].

Our patients reached advanced age although they were in their 30s at the time of surgical repair and the second patient suffered from previous heart failure and received a transannular patch.

Nollert [8] analysed the long-term survival in patients after repair of tetralogy of Fallot. He looked at 490 survivors in a 36-year follow-up. Mortality was linear in the first 25 years after the operation and equalled 0.24%/year. Thereafter, the mortality risk increased from 0.24%/year to 0.94%/year. During the follow-up 42 patients died, most of them from cardiac causes ($n = 26$, 62%). The most common cause of death was sudden death ($n = 13$) and congestive heart failure ($n = 6$). Nieminen [13] examined the causes of late death after paediatric cardiac surgery in 6024 patients and confirmed the findings from Oechslin [14]. Most patients died due to cardiac causes. The principal mode for cardiac related death was sudden cardiac death (18%) followed by heart failure death (13%) and other cardiovascular deaths (9%); perioperative death (20%) was also common.

Oechslin [10] looked at sixty adults who underwent reoperation between 1975 and 1997 after previous repair of tetralogy of Fallot. The main reason for reoperation in the long-term complications was the right ventricular outflow tract. Less common indications were ventricular septal patch leaks and severe tricuspid regurgitation. Of these 60 adults two deaths occurred during follow-up. One man died 8 months after reoperation due to severe pulmonary hypertension and pulmonary vascular disease. The other man died 7.7 years after reoperation due to end-stage heart failure.

Mid-term survival and functional improvement after reoperation were excellent.

Our two patients belong to the era of the first patients undergoing repair of tetralogy of Fallot and to our knowledge are the oldest survivors: not only the medical team, but also the patients were pioneers who have agreed to proceed with a surgical procedure without any outcome data. Survival until the age of late 70 or early 80 is possible in a historical cohort of patients with a balanced physiology between the systemic and pulmonary circulation which allowed them to survive until surgical repair became available in the 1960s.

References

- 1 Fallot ELA. Contribution à l'anatomie pathologique de la maladie bleue (cyanose cardiaque). *Marseille Méd.* 1888;25:77–93, 138–158, 207–223, 270–286, 341–354, 403–420.
- 2 Blalock A, Taussig HB. The surgical treatment of malformation of the heart in which there is pulmonary stenosis or pulmonary atresia. *JAMA.* 1945;128:189–202.
- 3 Lillehei CW, Cohen M, Warden HE, Read RC, Aust JB, Dewall RA, et al. Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects; report of first ten cases. *Ann Surg.* 1955;142(3):418–42.
- 4 Betranou EG, Blackstone EH, Hazelrig JB, et al. Life expectancy without surgery in TOF. *Am J Cardiol.* 1978;42:458–66.
- 5 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;81(952):133–4.
- 6 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;5(1):40–4.
- 7 Murphy JG, Gersh BJ, Mair DD, Fuster V, McGoon MD, Ilstrup DM, et al. Long-term outcome in patients undergoing surgical repair of tetralogy of Fallot. *N Engl J Med.* 1993;329(9):593–9.
- 8 Nollert G, Fischlein T, Bouterwek S, Böhmmer C, Klinner W, Reichart B. Long-term survival in patients with repair of tetralogy of Fallot: 36-year follow-up of 490 survivors of the first year after surgical repair. *J Am Coll Cardiol.* 1997;30(5):1384.
- 9 Klinner W, Reichart B, Pfaller M, Hatz R. Late results after correction of tetralogy of Fallot necessitating outflow tract reconstruction. Comparison with results after correction without outflow tract patch. *Thorac Cardiovasc Surg.* 1984;32(4):244–7.
- 10 Oechslin EN, Harrison DA, Harris L, Downar E, Webb GD, Siu SS, et al. Reoperation in adults with repair of tetralogy of Fallot: indications and outcomes. *J Thorac Cardiovasc Surg.* 1999;118(2):245–51.
- 11 d'Udekem Y, Rubay J, Shango-Lody P, Ovaert C, Vliers A, Caliteaux M, et al. Late homograft valve insertion after transannular patch repair of tetralogy of Fallot. *J Heart Valve Dis.* 1998;7(4):450–4.
- 12 Katz NM, Blackstone EH, Kirklin JW, Pacifico AD, Barger LM Jr. Late survival and symptoms after repair of tetralogy of Fallot. *Circulation.* 1982;65(2):403–10.
- 13 Nieminen HP, Jokinen EV, Sairanen HI. Causes of late deaths after paediatric cardiac surgery: a population-based study. *J Am Coll Cardiol.* 2007;50(13):1263–71. Epub 2007 Sep 10.
- 14 Oechslin E, Harrison D, Connelly M, Webb, Siu SS. Mode of death in adults with congenital heart disease. *Am J Cardiol.* 2000;86:1111–6.