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Brief Report

Longevity in the setting of tetralogy of Fallot: survival to the 84th year

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Abstract A patient with tetralogy of Fallot suffered minimal disability until the age of 45 years, when she developed bacterial endocarditis complicated by hemiplegia. She remained well, but became markedly polycythaemic and, at the age of 50 years, underwent surgical correction to reduce the risk of further thromboembolic incidents. She continued in remarkably good health until her death from lobar pneumonia in her 84th year. We present the post-mortem findings, since as far as we are aware this is the longest recorded survivorship of a patient with tetralogy of Fallot.

Keywords: Cyanotic congenital heart disease; pulmonary stenosis; geriatrics

WITHOUT SURGICAL AMELIORATION, PATIENTS with Fallot's tetralogy seldom survive to adult life. Over nine-tenths die before the age of 40 years,[1] and survival over 50 years is very rare.[2] In 1929, Paul White reported the case of a noted musician, known to have tetralogy, who died at the age of 59 years.[3] Then, in 1937 Brunlik, as cited by Marquis,[4] reported survival to the age of 62 years. The record for longevity was then raised to 69 years in 1954.[5] By 1956, however, Marquis[4] found that only 10 cases had been reported with survival over the age of 40 years, adding two further cases, a male of 47 years and a female of 64 years. The fourth sexagenarian was then recorded in 1999, a male who presented with acute ischaemic heart disease at 67 years, and was only then found to have Fallot's tetralogy. He suffered cardiac arrest shortly afterwards, and the cardiac anomalies were confirmed at autopsy.[6] None of these patients had undergone surgical correction, as most of them lived prior to the era of surgical intervention.

The first septuagenarian survivor was a male who had undergone construction of a palliative Blalock–Taussig shunt procedure at the age of 44 years, who developed ischaemic heart disease in later life, and had a radical repair of his congenital cardiac anomaly, together with a saphenous vein graft to the right coronary artery, at the age of 72 years. He died three months later from the effects of wound infection, disordered coagulation, and renal failure.[6]

We now present the case of a female who survived to her 84th year, and died 33 years after corrective surgery. Of the whole population of the United Kingdom born in the year of her birth, 1919, only one-third of the females, and one-fifth of the males, would have survived until 2003.[7] The number of these with tetralogy of Fallot, therefore, must be exceedingly low, but with the passage of time, more and more patients with the anomaly would have experienced both the specific benefit of increasingly sophisticated surgery, and the general rise in the expectation of life-span. It is probable that survival into advanced age of those with tetralogy of Fallot may eventually become commonplace.

Case report

N.S. was born in 1919, after an uneventful pregnancy and a normal full-term delivery. She was noted to be cyanosed at birth, and a cardiac murmur was first heard at the age of two years. There was no known family history of congenital cardiac disease. She had a normal childhood, and was able to play games at

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school, but was also said to have become cyanosed on exertion. She was seen at the National Heart Hospital at the age of 15, 25, and 29 years, with a presumptive diagnosis of tetralogy of Fallot. On the last of these consultations, in 1949, construction of the Blalock–Taussig shunt was considered, but was not thought to be advisable on account of her continuing good health.

In 1953, at the age of 34 years, she developed a duodenal ulcer, which was confirmed by a barium meal X-ray. Three years later, she had two episodes of melaena, and a pyloroplasty was performed. Following this, she remained well, with only minimal exertional dyspnoea, until the age of 45 years, when she was admitted to hospital because of progressive fatigue, lassitude, and pyrexia. She was then found to have subacute bacterial endocarditis, which was complicated by a sudden onset of hemiplegia. She made a good recovery, and was able to walk without too much disability.

There was little change in her cardiac symptoms and, when reviewed at the age of 49 years, she had marked central cyanosis, clubbing of the fingers and toes, a jugular venous pressure raised to 2 centimetres above the sternal angle at 45°, and a grade 2/4 ejection systolic murmur at the left sternal edge, following the first sound. She also had marked polycythaemia, with a haemoglobin of 142%. Radiography showed a small heart, with a bay in the middle segment of the left cardiac border, suggestive of infundibular stenosis, and the lungs were slightly underfilled. Cardiac catheterisation showed equal pressures in the two ventricles, peripheral arterial desaturation, and a right-to-left shunt at ventricular level. Angiography demonstrated valvar pulmonary stenosis with a distorted right ventricular outflow tract, a small right ventricle, prolapse of the aortic leaflet of the mitral valve, with slight mitral reflux, and a ventricular septal defect. Surgical correction was advised in view of the risk of further thrombotic incidents consequent to the persistently high haemoglobin.

In 1969, at the age of 50 years, correction was performed under normothermic by-pass. The ventricular septal defect was about 1.5 centimetres in diameter, there was a long narrow muscular narrowing of the infundibulum, a thick dome-shaped pulmonary valve with a narrow central orifice, and a hypoplastic pulmonary trunk. The ventricular septal defect was closed, and the infundibulum, pulmonary valve, and pulmonary trunk were incised, opened across, and oversewn with a Dacron patch. Following the operation she enjoyed good health, adopted three children, and engaged in normal activities. On review at the age of 77 years, she was found to be in remarkably good and stable condition for her age, in spite of the residual hemiplegia, with just mild dyspnoea on exertion and slight peripheral oedema.

She remained well until 2003, when she became breathless with right-sided pleuritic pain and clinical signs of pneumonia. She died peacefully one week later at the age of 83 years, 33 years after the surgical correction of her congenital cardiac malformation.

Post-mortem examination

This was limited to the heart and lungs. There was confluent consolidation throughout most of the upper lobe of the right lung, consistent with lobar pneumonia. This was subsequently confirmed histologically. Elsewhere the lungs showed extensive oedema.

The heart was moderately enlarged, and measured 13 by 15 by 8 centimetres. The atriums were usually arranged, and the systemic and pulmonary veins were normally connected. The atrioventricular and ventriculoarterial connections were concordant. The leaflets of the tricuspid valve were slightly thickened, and the tendinous cords were rather rigid. The orifice of the valve was slightly narrowed to 2 centimetres in diameter. The macroscopic appearances were nonspecific, and were not suggestive of old rheumatic fever. The cavity of the right ventricle was normal apart from scarring from infundibular resection. There was an old, fully epithelialised, patch closing a perimembraneous ventricular septal defect, and an old, partly calcified Dacron patch enlarging the anterior aspect of the outflow tract. This was measured at 4 by 2 centimetres, crossing the pulmonary valve and extending into the pulmonary trunk.

The pulmonary valve had three incomplete leaflets, which were deformed, irregular, and nodular. Part of the anterior leaflet was missing, and the valve was thus incomplete. It was also moderately stenosed to 1 centimetre in diameter (Fig. 1).

The pulmonary trunk and the right and left pulmonary arteries were slightly dilated, to 2.5 and 2.0 centimetres diameter respectively. The mitral valve was normal. The left ventricle was normal, its free wall being 1.2 centimetres thick. The patch closing the subaortic, perimembranous septal defect enclosed a deep recess above the body of the ventricular septum, as typically seen in the presence of a dextraposed aorta. The leaflets of the aortic valve were thickened, but otherwise normal. The arterial duct was closed. There was very little atheroma in the aorta, its branches, or the coronary arteries, which was probably the reason for her longevity.

Discussion

Although suffering from a right-to-left interventricular shunt and restricted pulmonary arterial perfusion, our patient remained remarkably free from significant cardiac disability for 50 years. It is not possible

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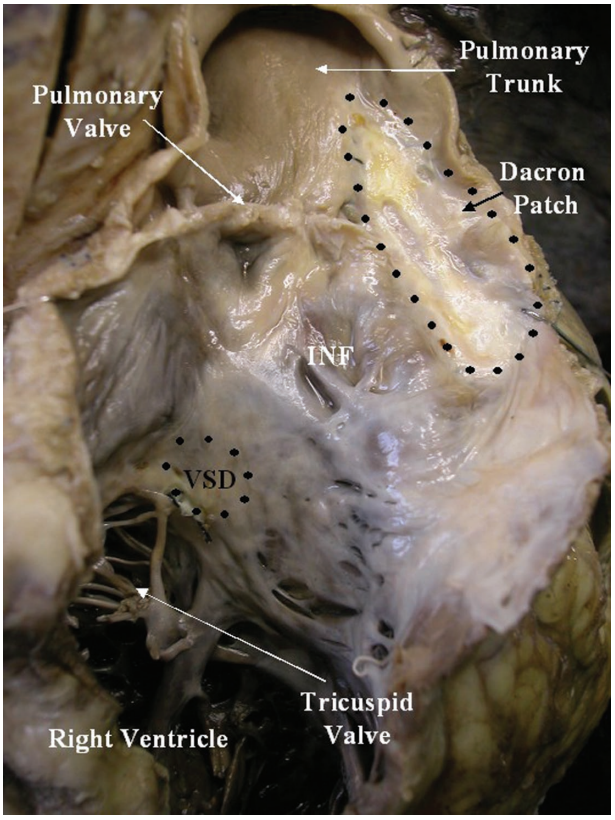

Figure 1.
The right ventricular outflow tract and the pulmonary trunk have been opened to display the dysplastic pulmonary valve, the site of the infundibulum resection (INF), the site of the repaired ventricular septal defect (VSD), and the site of the Dacron patch used to enlarge the outflow tract. The extent of the two latter sites is indicated by the dotted outlines.

to speculate on how long she may have survived without the surgical correction. Post-operative survival for a further 33 years is ample testimony to the technical excellence of the surgery, even though the pulmonary valve remained incompetent. This experience sends a clear message that reparative surgery should continue to be pursued in older patients with tetralogy of Fallot, even though increasing age at repair is associated with decreasing rate of late survival.[1]

Epilogue

Although our patient was born with a major cardiac malformation, and suffered a disabling hemiplegia at 45 years, she had a long, active, and happy life. She was deeply appreciative of the medical and surgical attention that had made this possible, and this was reflected in her dying wish that, after her death, her heart should be removed and used to further medical education. We feel privileged to have been able to contribute to this objective, and hope that this report will be of interest to all those concerned with the management of congenital cardiac disease, and that it may give encouragement and hope to other patients and their families.

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