

# Longevity in the tetralogy of Fallot

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The congenital abnormality of the heart consisting of pulmonary stenosis, ventricular septal defect, and an overriding aorta was described in 1673 by the Danish scholar, Nicolas Stensen (Warburg, 1942). Fallot's (Fallot, 1888) description included right ventricular hypertrophy as an essential part of the malformation. Stensen had made no mention of this, possibly because he reported on the heart of the foetus.

Maud Abbott (1936) found in her series of 1,000 cases of congenital heart disease that the average life expectancy in Fallot's tetralogy was 12 years, and only six years in cases with pulmonary artery atresia. In one report on 200 living patients (Donzelot, de Balsac, Emam-Zadé, Escalle, and Metianu, 1949) with Fallot's tetralogy, 13 were between the ages of 21 and 30 years and only one over the age of 40. In another report (Soulié, Vernant, Andreuzzi, and Magnoli, 1956) on 144 cases of this malformation, there were eight patients between the ages of 20 and 30, two between 30 and 40, and only two over the age of 40 years. Since the famous case of a musician, reported by White and Sprague (1929), who died in his sixtieth year, 26 other cases have been reported with a survival of more than 40 years. There were four patients between 60 and 70 years (Bain, 1954; Bowie, 1961; Brumlik, 1937; Marquis, 1956), 12 between 50 and 60 years (Abraham, Atkinson, and Mitchell, 1961; Ansell and Reiser, 1957; Bedford, 1956; Campbell, 1958; Feigin and Rosenthal, 1943; Lian and Fleury, 1949; Miller, 1952; Rosenthal, 1956; Soulié *et al.*, 1956; Strandell, 1939; White and Sprague, 1929), and 11 patients between 40 and 50 years of age (Báguena and Tormo, 1951; Civin and Edwards, 1950; Donzelot *et al.*, 1949; Feigin and Rosenthal, 1943; Fisher, Wilson, and Theilen, 1962; Guyot, 1945; Marquis, 1956; Middleton and Ritchie, 1947; Soulié *et al.*, 1956; Volini and Flaxman, 1938; Wood, 1956).

## CASE REPORT

F. H. D., a 57-year-old man, was admitted to hospital because of attacks of shivering, fever, and increasing shortness of breath.

As a schoolboy he had loved cricket and played sometimes two games of football a day. At the age of 10 he was told at King's College Hospital, London, that he had 'heart trouble'. He was rejected by the army medical board when he tried to volunteer in 1915, because of his 'heart'.

For the last 27 years he had worked as a fettler, a moderately heavy job consisting of trimming and lifting heavy metal castings in a foundry. He was married, had one daughter, was a moderate beer drinker, and smoked 10 cigarettes a day.

He had been very well until the summer of 1955, when he noticed shortness of breath on walking up a hill on his way to work.

On admission in 1957, there was no dyspnoea at rest, no clubbing, but central cyanosis. His blood pressure was 170/100 mm. Hg, and pulse 80, regular. There were prominent A-waves in the jugular venous pulse; the heart apex was 10 cm. in the fifth intercostal space; and a loud, long systolic murmur, accompanied by a thrill, was maximal in the third and fourth intercostal space; the pulmonary second sound was soft and single. Laboratory findings included a haemoglobin of 14.86 g./100 ml. The electrocardiogram showed right ventricular preponderance and incomplete right bundle branch block. A radiograph of the chest showed considerable cardiac enlargement (mainly of the left ventricle) and a marked increase in the size of the left pulmonary artery but no peripheral plethora. The probable diagnosis was Fallot's tetralogy; other diagnoses were staphylococcal septicaemia, carcinoma of the bladder, and chronic pyelonephritis.

Between August 1957 and September 1959 he had four general anaesthetics for a cystoscopy, underwent a course of deep radiotherapy for bladder carcinoma, had a perinephric abscess drained under local anaesthesia but refused several offers of cardiac catheterization.

He was readmitted on 16 March 1962 with increasing dyspnoea and swelling of the ankles following an attack of severe chest pain three weeks previously, which lasted about 10 hours, causing him to lose a day's work. Recently he had been working as a clerk. On examination he was an adequately nourished man; his weight was 64.5 kg. (142 lb.) and height 170 cm. (5 ft. 7 in.). He was dyspnoeic at rest and cyanosed, with prominent blue ears and nose but no clubbing of the fingers or toes. There was a plum-sized, irregular, hard tumour in the left parotid region. The pulse was 52 and regular at the wrist, 104 at the

apex with obvious 'coupling'. The blood pressure was 164/80 mm. Hg. The heart apex beat was 12.5 cm. in the fifth left intercostal space. There was a slight left parasternal heave, and a loud, long, rough, systolic murmur best heard over the third and fourth left interspace, accompanied by a thrill.  $P_2$  was single and soft, and the jugular venous pressure was 8 cm. above the sternal angle. A tender, enlarged liver was palpable 8 cm. below the right costal margin, with pitting oedema of the entire lower limbs and sacral area, but only a few crepitations at both lung bases. Laboratory investigations included the following: haemoglobin 18.2 g./100 ml. (123%); white cell count 7,600/c.mm. The urine contained a moderate amount of albumin. An electrocardiogram showed alternate ventricular extrasystoles and prominent PII, raised S-T segments and large Q waves in I, aVL, and chest leads, indicating recent extensive myocardial infarction. On chest radiography, the transverse diameter of the heart and the hilar vessels were markedly enlarged but the peripheral vessels were within normal limits. He was treated with bed rest, low sodium diet, and diuretics; digitalis therapy was withheld because of 'coupling' and digoxin treatment at home. On the fourth day in hospital he collapsed, became deeply cyanosed, almost black, and was dead two minutes later.

**NECROPSY** The heart weighed 710 g.: it showed dilatation of both ventricles and the right atrium. The average thickness of the right ventricular wall was 13 mm., which equalled that of the left. The left-sided aorta overrode a circular ventricular septal defect 30 mm. in diameter, so that two-thirds of the aortic opening appeared to be continuous with the right ventricle (Fig. 1). The pulmonary valve presented a 'fish-mouth' opening measuring  $10 \times 7$  mm. at the apex of a dome-shaped diaphragm, formed by the

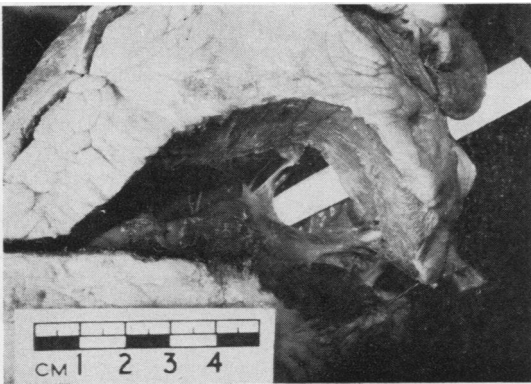


FIG. 1. View of the right ventricle showing marked hypertrophy of its wall and the large ventricular septal defect. (The marker passes from the aorta into the left ventricle.)

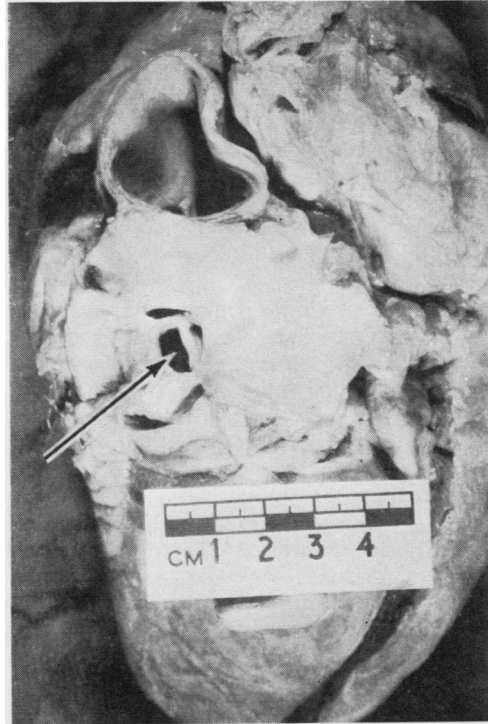


FIG. 2. Superior view of the heart demonstrating post-stenotic dilatation of the pulmonary artery with the arrow pointing to the 'fish-mouth' opening of the stenosed pulmonary valve.

fusion of three thickened pulmonary valves (Fig. 2). Small, hard vegetations were noted on the ventricular surface of the pulmonary valves, but the aortic and mitral valves showed no abnormality. The main pulmonary artery was dilated. The ductus arteriosus and foramen ovale were closed. The venae cavae, pulmonary veins, and bronchial arteries were normal. Moderate atheromatous changes were observed in the aorta and coronary arteries. The anterior descending branch of the left coronary artery was occluded by a thrombus and atheroma. There were changes of a fairly recent myocardial infarction involving the anterior wall of the left ventricle. The lungs showed severe congestion. Several large depressed scars were noted on the outer surface of the enlarged left kidney. The bladder showed a few mucosal haemorrhages but no evidence of carcinoma.

The number of explanations given by various authors as to the cause of this prolonged survival probably illustrates our ignorance in the matter. The statement of Soulié *et al.* (1956) that 'the cause of the long survival is found mainly in the structural fault' is easily accepted; but evidence to

support it is difficult to find. In the analysis of 21 previously reported patients with Fallot's tetralogy who survived to over the age of 40 years, Bowie (1961) found good correlation between the intensity of cyanosis and the degree of clubbing. She was able to find some relation between cyanosis and clubbing on the one hand and reduction of exercise tolerance on the other, but was unable to demonstrate any relation between the severity of symptoms, the length of survival, or the degree of structural deformity. Attempts to find the cause of long survival in individual cases have been unsatisfactory. The lack of symptoms, but not the prolonged survival, in our case may be related to the mildness of the pulmonary stenosis: further support for this view is provided by three patients (Bain, 1954; Marquis, 1956; Miller, 1952) with lumina of pulmonary outflow tracts only half the size of that of our patient; they all survived to the age of 55 years or more. In a man of 44 (Báguena and Tormo, 1951) a patent ductus arteriosus (because of the co-existence of pulmonary artery atresia) explained survival, but not why it was prolonged. No other case exhibited a patent ductus arteriosus, but Feigin and Rosenthal (1943) thought that their second case had benefited from one until recently; although at necropsy it was represented by a lumenless fibrous strand 10 mm. long and 1 mm. in diameter. Bowie (1961) found five cases where the aorta arose more from the right than from the left ventricle and one where it arose entirely from the right (Volini and Flaxman, 1938); these observations disprove Bain's view that it is the degree of overriding of the aorta that determines long survival. Middleton and Ritchie (1947) emphasized wide bronchial arteries and vascular pulmonary adhesions as providing a significant part of the pulmonary blood flow; it remains unanswered how much blood is carried by this collateral circulation and what part of it eventually becomes oxygenated. Blood pressure recordings are available in 15 cases of prolonged survival with this malformation, varying from 230/160 to 110/80 mm.Hg, with an average systolic pressure of 144 and diastolic pressure of 94 mm.Hg. It is unlikely that this small increase in the diastolic pressure was responsible for any significant rise in the pulmonary blood flow. Feigin and Rosenthal suggested that their patient suffered from Eisenmenger's complex (where prolonged survival is not rare) with the addition of rheumatic pulmonary stenosis at some later date. Although not supported by histological evidence, it is an ingenious idea and could perhaps be extended to similar malformations where pulmonary valve

stenosis is produced by bacterial endocarditis. (Our patient was hospitalized in 1957 with septicaemia!) Lian and Fleury (1949) thought that these patients lived to an advanced age because they avoided physical exertion and lived sheltered lives; the amount of exercise involved in occupations followed by some of the patients is against this suggestion, e.g., miner (Feigin and Rosenthal, 1943), pilot and racing driver (Miller, 1952), labourer (Civin and Edwards, 1950), labourer (Volini and Flaxman, 1938), mother of two and a housewife (Rosenthal, 1956), and foundry worker (present case). The only patient (Fisher, Wilson, and Theilen, 1962) who underwent surgery to improve the circulatory defect had a Blalock operation done when he was 32; he did well for 10 years but died at the age of 44.

Perhaps one can go no further than to admire the capacity of adaptation of the human body to severe structural faults until more thorough studies, including cardiac catheterization, pressure measurements, and gas analyses, on a larger number of similar patients are completed.

#### SUMMARY

Nowadays, because the enthusiasm for surgical intervention in Fallot's tetralogy prevails, there will be less opportunity to study the natural history of this malformation. The case of a man suffering from Fallot's tetralogy, who died of coronary thrombosis in his sixty-second year, is reported. Possible causes of prolonged survival with this malformation are discussed with reference to 27 previously reported patients who have survived to the age of 40 years or more.

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