Unoperated tetralogy of Fallot: case report of a natural survivor who died in his 73rd year; is it ever too late to operate?

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The case history is described of a man in his 73rd year who was one of the oldest surviving patients with uncorrected tetralogy of Fallot (ToF) before succumbing with renal failure. Factors contributing to his longevity included small pulmonary arteries and presumed slow development of subpulmonary obstruction together with moderate concentric left ventricular hypertrophy—features previously seen in long term survivors. Less than 3% of all patients with uncorrected ToF survive beyond their 40s but late operative repair is still a valuable option. Practicalities of renal dialysis in the presence of an intracardiac shunt are considered.

Abbreviations: ToF, tetralogy of Fallot; RVH, right ventricular hypertrophy; LA, left atrium; RA, right atrium; LV, left ventricle; RVOT, right ventricle outflow tract; VSD, ventricular septal defect; LAD, left anterior descending.
but postmortem autolysis prevented microscopic interpretation.

DISCUSSION

Without operation, few patients with ToF reach adulthood with an average life expectancy of 12 years. Ten per cent may survive to their 30s but only 3% reach their 40s or older. There are three main reasons for the longevity in natural survivors with unoperated ToF. Firstly, a hypoplastic pulmonary artery with slow development of subpulmonary obstruction. The pulmonary annulus was 13 mm in this patient, comparable to that reported in the other published cases (age 45, PVD 10 mm), (age 77, PVD 9 mm). A second common feature is that of LVH as seen in this patient (16 mm); presumably this acts by delaying of shunting from the right to left ventricle. LVH may be a late development in the natural history of Fallot and any beneficial effect may not be seen until adult life. In this case hypoplasia of the pulmonary artery (seen in up to 50% of all Fallot’s) may have been sufficient early on to reduce pulmonary blood flow; then the subpulmonary stenosis needs to have also developed slowly over decades while his left ventricular hypertrophied concomitantly. Such a fine haemodynamic balance is clearly rare. The third finding in other cases has been extracardiac shunts including patent ductus arteriosus—reported in the oldest survivor who died aged 77—or systemic to pulmonary artery shunting via internal mammarys. There are few studies on the outcome and benefit of late surgical repair. The Mayo series followed up 30 patients who had total correction of ToF between the ages of 40 to 60 years. The operative mortality was 3% with long term survival rate at 5 years and 10 years postoperatively of 92% and 74% respectively. In this case, surgical repair would have been possible at the age of 66 years with low operative mortality and is likely to have prevented his subsequent systemic venous congestion. Nevertheless his personal wishes were not for operation—but given similar circumstances, operation would be recommended. Preoperative catheterisation also would have disclosed his coronary artery disease, which was unrecognised and untreated and probably had a role in his terminal decline. It is always important to consider detailed reinvestigation of any new elderly patient given a label of inoperable congenital heart disease as new approaches may be available to palliate the condition. He remained in sinus rhythm for a long time given his atrial dimensions; this undoubtedly improved his longevity and is probably a testament to amiodarone. His rapid decline was heralded by the change to atrial fibrillation resulting in pre-renal failure—which was not a consequence of his longstanding congenital heart disease. Dialysis in patients with intracardiac shunts requires antibiotic prophylaxis at the time of line insertion, use of a blood filter downstream from the bubble trap on the venous limb of the dialysis circuit to avoid paradoxical embolism together with avoidance of rapid fluid removal, which may precipitate hypotension and exacerbate shunt reversal.

Learning points

- Few patients with tetralogy of Fallot survive into adulthood without operation
- Unoperated survivors have three common features: hypoplastic pulmonary artery with moderately slow development of subpulmonary obstruction, left ventricular hypertrophy, or systemic-pulmonary artery collaterals for pulmonary blood flow
- Late operative intervention is associated with low mortality—it may never be too late to operate
- Full re-investigation of older patients labelled as “inoperable” congenital heart disease may sometimes permit late palliation
- Renal dialysis in the presence of an intracardiac shunt requires bubble filters to prevent paradoxical embolism

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