Long-term survival of a patient with tetralogy of Fallot after intracardiac palliation in the pre-pump era

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The first successful intracardiac surgery with cardiopulmonary bypass (CPB) was performed by John H. Gibbon¹ on May 6, 1953, in a patient with an atrial septal defect. The first intracardiac repair of tetralogy of Fallot (TOF) was performed by C. Walton Lillehei² on August 31, 1954, with the use of cross-circulation. In Japan, the first cardiac surgery with CPB was a total correction of TOF performed by H. Manabe³ on April 18, 1956. We report the case of a successful direct-vision intracardiac palliation of TOF in the pre-pump era in which the patient survived for half a century.

Clinical Summary

On May 13, 1955, a 16-year-old girl with TOF underwent palliative surgery with selective cerebral hypothermia (SCH). Two cannulas were inserted into the right common carotid artery, 1 of which was inserted downward to aspirate arterial blood out of the body. The blood collected was cooled and then delivered to the brain through the other cannula, which was inserted upward. With crossclamping of both the aortic root and main pulmonary artery, accompanied by total caval occlusion, cardiac standstill was obtained by injection of acetylcholine into the right atrium. The right ventricular outflow tract (RVOT) was incised, followed by resection of an abnormal muscle bundle (AMB) and a pulmonary valvular commissurotomy. The RVOT was closed by a suture. Cardiac blood flow was stopped for 8 minutes. After the palliation, the patient remained in New York Heart Association (NYHA) class I to II status without cyanosis for a long time, living an almost normal life and being delivered of 2 children.

When the patient was 55 years old, she had shortness of breath with occasional cyanosis. When she was 65 years old, in 2004, she was admitted to the hospital in NYHA class III status

because of congestive heart failure accompanied by systemic cyanosis on effort. Preoperative catheterization and echocardiography were consistent with TOF. Measured pressures were 41/12 (24) mm Hg in the pulmonary artery and 48/11 (12) mm Hg in the low-pressure chamber. The pulmonary/systemic blood flow ratio was 1.5. A double-chambered right ventricle and aneurysm of the RVOT were revealed (Figure 1). Echocardiography showed that the interchamber pressure gradient (PG) was 73 mm Hg, whereas the PG between the right atrium and high-pressure chamber was 110 mm Hg. Total correction was performed with CPB. The AMBs of the RVOT and right ventricular inflow were excised, the ventricular septal defect was closed with a Dacron patch, and the RVOT was reconstructed with a polytetrafluoroethylene patch (Gore-Tex patch; W. L. Gore & Associates, Inc, Flagstaff, Ariz). Postoperative right-sided heart failure was managed successfully. Three months after the operation the patient was in NYHA class I status with a pulmonary pressure of 29/10 (19) mm Hg and right ventricular inflow pressure of 37/11 (12) mm Hg measured by postoperative catheterization (Figure 2).

Discussion

The first intracardiac surgery under SCH in Japan was performed by Kimoto and associates on January 17, 1955, in a 12-year-old boy with an atrial septal defect.⁴ In SCH, only the brain is cooled down to 17°C to 18°C by perfusing cold arterial blood to prevent cerebral damage during circulatory standstill. Kimoto and his colleagues found the possible circulatory standstill was 15 minutes at a cerebral temperature of 20°C.⁵ They reported the results of intracardiac surgery under SCH from January 1955 to April 1956. Of the 24 cases, 11 had atrial septal defects with no perioperative deaths, and 8 had TOF with 6 perioperative deaths. The patient in this case was the first of 8 cases and the first of 2 survivors. We could not find any reference in the literature to a successful correction after intracardiac palliation in the pre-pump era in elderly patients with TOF.

What was the reason for the long-term survival of our patient? The well-balanced pulmonary/systemic blood flow ratio may have been an important factor, and we speculate that the right ventricular inflow pressure, estimated to be greater than 110 mm Hg, would have prevented severe left-to-right shunt. Simultaneously, the AMB between the right ventricular inflow tract and RVOT prevented serious pulmonary hypertension. If the AMB constriction had been too severe, serious systemic

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Figure 1. Double-chambered right ventricle. An abnormal muscle bundle (AMB) separates the high pressure inflow chamber and the low pressure outflow chamber.

cyanosis would have developed earlier because a higher right ventricular pressure would have led to a right-to-left shunt. If the constriction was small, serious pulmonary hypertension would have occurred because of the severe left-to-right shunt. In addition, because an AMB constriction existed between the right ventricular inflow tract and RVOT, the pressure in the right ventricular outflow chamber was not increased, and therefore severe congestive heart failure did not develop in the patient over a long period. In the pre-pump era, it was impossible to excise the AMB completely during the short cardiac arrest, but this inability may have resulted in the favorable outcome in this case.

Figure 2. Postoperative right ventriculography.

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