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Total Correction for Tetralogy of Fallot in Adolescents and Adults*

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Intracardiac repair for tetralogy of Fallot (TOF) has been accomplished with reasonable risk in current practice, since the first successful correction of this malformation by Lillehei et al.[1] In 1955. Palliative shunts have recently lost their importance in the treatment of TOF and direct total correction regardless the age of the patients has gained interest. In the current circumstances the risks of palliative operations combined with the risks of subsequent total correction (two-stage operation) are more pronounced than the hazards of direct total correction per se. Technical impovement in operative methods such as deep hypothermia and circulatory arrest, first used by Kyoto University group (Japan) in 1967 and later advocated by Barrett-Boyes et al. in 1971 (so called "Kyoto-Barrett-Boyes technique)[2], has enabled us to perform total correction even in infants and small children.

Total correction for TOF in adolescents and adults is not a common practice, since these age groups are not optimal periods for the correction of this pathology. Review of the literature revealed only few papers regarding the total correction for TOF in these age groups.[3-8]

Case Reports

Thirty-two adult patients with tetralogy of Fallot have undergone total correction in our clinic until January 1980. They have been opera-

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ted under mild hypothermic conditions of 32° C and anoxic cardiac arrest by cross-clamping the aorta; using disposable bubble oxygenators and DeBakey roller pumps. The youngest patient was 16, and the oldest 28 years-old, mean age being 19. Ten cases had previously performed (more than 10 years ago) left Blalock-Taussig shunts, in whom this anastomosis was prepared in the extrapleural space between the pleura and the pericardium, and made ready to be ligated temporarily before the institution of cardiopulmonary bypass.

Intracardiac repair was performed via vertical right ventriculotomy. Hypertrophic muscular bands were resected and outflow tract mobilized. Valvular pulmonary stenosis (if present) was eliminated by the fracture of the fused cusps and dilatation of the pulmonary ring by Hegar's bougies; or fused cusps were incised by scalpel. Ventricular septal defect was closed with a teflon patch slightly smaller than the size of the defect using interrupted ti-cron sutures. Blalock-Taussing shunt was permanently ligated after total correction in the above mentioned 10 cases. Right ventricular outflow patch (teflon) was used only in 3 cases where the immediate postoperative right ventricular/left ventricular pressure ratio was greater than 0.9. In one of these cases outflow patch was not used during the initial operation, but 3 days later during re-operation for the low cardiac output syndrome. These 3 cases with outflow patch reconstruction did not have previous Blalock-Taussig shunts.

Complementary data for the above 32 cases are tabulated in Table I.

TABLE I

COMPLEMENTARY DATA IN 32 CASES

	No. Cases
Previously performed left Blalock-Taussig shunt	10(*)
Valvular pulmonary stenosis	1
Atrial septal defect (secundum)	2
Pulmonary artery coarctation	1

(*) Subclavian artery was used in 9, and dacron graft in 1. One of the shunts was not patent at the time of total correction.

Results

Five patients expired during the early postoperative period (15.6 %), one of whom was hemodynamically stable but succumbed to mediastinitis. Therefore, only 4 cases died of cardiac complications (12.5 %), 3 of whom had previously performed Blalock-Taussig shunts. Low

cardiac output was the cause of fatal outcome in 2 cases; pulmonary insufficiency in 1; and severe bradycardia and hemorrhage in 1.

There was no late mortality.

Discussion

A patient with tetralogy of Fallot may rarely live to his adulthood and even old age without any treatment. White and Sprague[9] reported the life story of a noted American musician (H. F. Gilbert) who lived to his 60 th. age. Some of such patients may even give birth to healthy children. A woman with uncorrected tetralogy of Fallot giving birth to 4 consecutive children was observed in our University Clinics.[10]

A smal number of publications regarding surgical treatment of TOF in adolescents and adults has appeared in medical journals.[3-8] It has been observed in our practice that some patients with excellent palliative operations perfoı med many years ago do not want to accept the total correction, because their daily activity is quite satisfactory.

Houchin[7] also reported a 49 year-old man undergoing total correction for TOF and myocardial revascularization simultaneously, who had an excellent palliation by Potts anastomosis 23 years ago.

Mortality rate for total correction in adults has been reported within acceptable limits. Beach et al.[3] presented an over-all mortality of 7.8 % in 64 adolescents and adults, Bender et al.[4] 11.9 % in 84 adults, and Chiariello et al.[5,6] 14.5 % in their cases.

The mortality rate in general is about 10 % in total correction for tetralogy of Fallot.[6, 11] Study of the first 156 cases in our clinic showed an over-all mortality of 12 %, where most of the patients were between 5 and 10 years old.[11]

An outflow tract patch reconstruction is necessary in subjects where the immediate postoperative right ventricular/left ventricular pressure ratio is above 0.9 as experienced in our cases. Blackstone et al.[12] recommend the use of a transannular patch reconstruction of the right ventricular outflow tract when the right ventricular/left ventricular pressure ratio is equal to/or above 0,95, measured immediately after the cessation of cardiopulmonary bypass; providing that the hemodynamic state is satisfactory and right atrial pressure is similar to/or less than the left atrial pressure. It has been accepted that the pulmonary regurgitation encountered after transannular patch reconstruction is tolerated better than the residual stenosis.[4, 6, 8]

It has been reported that the total corrections performed after the long standing presence of palliative shunts carry slightly higher mortality rates compared to those without any palliation before, owing to the ill effects of these shunts like pulmonary hypertension and pulmonary vascular disease.[6] Blalock-Taussig shunt has the least ill effects compared to other palliations such as Waterston and Potts anastomosis.[6] This point was also reflected in our series. We have 10 cases of TOF with previous shunt operations, 3 of whom expired after total correction due to various cardiac complications (30 %), but only 1 case died of cardiac complication in 22 patients (4.5 %) without previous shunt operations.

Summary

Total correction for tetralogy of Fallot in adolescents and adults is an uncommon practice, since this malformation is almost always corrected during early childhood period. Total correction performed in the late age group bears some disadvantages like fibrotic and calcific changes in some adult hearts and the ill effects of the long standing presence of palliative shunts in some cases. Although the old age carries some risk compared to early childhood, the mortality rate in this age group is within acceptable limits (8-15 %).

It is concluded that the advanced age should not be a contrindication for total correction of tetralogy of Fallot.

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