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RISK FACTORS AND CAUSES OF MORTALITY AFTER TOTAL CORRECTION FOR TETRALOGY OF FALLOT*

*Argun Saylam MD**, *Rüstem Olga MD***, *Coşkun İkizler MD****, *Ali Ertuğrul MD*****, *Aydın Aytaç MD******

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Mortality rates following total correction for tetralogy of Fallot (TOF) have decreased from 50% to close to 10% as a result of developments in surgical technique, better understanding of surgical anatomy, and improvement in cardiopulmonary bypass methods[1,2]. This article discusses risk factors and causes of mortality following total correction for TOF based on our clinical experience and world literature on the subject.

Material, Methods and Findings

The first total correction for TOF in our clinic was performed in 1963[1]. Since then and up to June 1979, the authors of this paper have performed 322 operations of total correction for TOF, with 47 deaths following the intervention (14.5%). Complementary data on those 47 cases are shown in Table I.

The patients were subjected to open heart surgery using the Kay-Cross disc oxygenator in earlier years and a disposable bubble oxygenator with DeBakey roller pumps more recently. Hypothermia of 32°C and anoxic cardiac arrest were used during total correction in the majority of the cases, except for a few earlier patients. Infundibular resection and mobilization were performed after right ventriculotomy, and the ventricular septal defect (VSD) was closed using interrupted sutures with a teflon patch slightly smaller than the size of the VSD. Details of the surgical techniques are given in a previous article[1]. The mean duration of anoxic arrest was 36 minutes, and that of extracorporeal circulation was 44 minutes.

* From the Hacettepe University Institute of Child Health, Ankara.
** Lecturer in Pediatric Thoracic and Cardiovascular Surgery, Hacettepe University Institute of Child Health.
*** Associate Professor of Pediatric Thoracic and Cardiovascular Surgery, Hacettepe University Institute of Child Health.
**** Professor of Pediatric Cardiology, Hacettepe University Institute of Child Health.
***** Professor of Pediatric Thoracic and Cardiovascular Surgery, Hacettepe University Institute of Child Health.

TABLE I

COMPLEMENTARY DATA IN 47 CASES OF EXITUS

	No. cases
Male patients	29
Female patients	18
Cyanosis	47
Pink tetralogy	None
Age : 0 - 4	10
5 - 9	23
10 - 14	8
15 - 19	5
20 and above	1
Use of outflow patch	7
Previous shunt operation (Blalock)	6
Additional pathologic findings:*	
Atrial septal defect (secundum)	4
Hypoplastic left ventricle	4
Pulmonary valvular atresia	2
Patent foramen ovale	2
Endocardial calcification	2
Absence of superior vena cava	1
Coronary artery anomalies	2
Pectus excavatum and carinatum	1

* Infundibular stenosis was associated with valvular pulmonary stenosis in 7 cases.

Causes of the 47 deaths are outlined in Table II. Cardiac failure and low cardiac output syndrome were the most common. Seven patients died in the operating room (2.1% of total cases), 38 during the early postoperative period (11.8%) and 2 during the late postoperative period (0.6%).

Discussion

There is no definite agreement concerning the optimum age for total correction of TOF. The classic position holds that the best time is when the patient is over 5 years old, particularly between the ages of 6 and 10[1,3]. We share this opinion on the basis of our clinical experience, and the great majority of our subjects were between 5 and 10 years old. Daily et al[4] stated that the total correction should be deferred until after the age of 2 years in asymptomatic patients and performed at any age in

symptomatic cases. Some recommend total correction even in infants and small children, except in individuals with hypoplastic pulmonary artery (ies) or anomalous distribution of the coronary arteries (such as the left anterior descending branch originating from the right coronary artery) which cause difficulty in total correction at early ages[4-9]. The technique of deep hypothermia and circulatory arrest plays an important role in the total correction in infants and small children[5,6,8]. Total correction in adolescents and adults is not a common practice, although mortality rates in older age groups are not reported to be significantly higher than in children (15.6% in our series of 32 older cases) [10]. This implies that advanced age is not a contraindication for the total correction of TOF.

Mortality in total correction generally takes place in the operative and early postoperative periods. Late mortality rates are very low, ranging from 2 to 3.4%[2,11,12] (0.6% in our series), and are mainly due to sudden arrhythmias and other unrelated causes.

Stone heart, a well-defined entity which occurs especially in cases of severe aortic stenosis[13], has not been previously observed in cyanotic congenital heart disease. We encountered 3 such cases (Table II) in this series, all dating to the time when hypothermia was not used during open heart surgery.

There is disagreement concerning the effects of the right ventricular outflow patch on morbidity and mortality after total correction for TOF[1]. We observed in our clinical practice that transannular patch repair of the outflow tract increases right heart failure during the early postoperative course by inhibiting the effective right ventricular contractions and causing pulmonary regurgitation, thus playing a role in the increased morbidity and mortality[1]. Similar results have been observed by Irisawa et al[14] and Rocchini et al[15]. We therefore prefer limited use of the outflow patch and use patch reconstruction of the outflow tract when the right ventricular/left ventricular pressure ratio is above 0.9 immediately following total correction. Chiariello et al[3] reported the use of outflow patch in 57% of their cases and stated that residual pulmonary insufficiency is tolerated better than residual pulmonary stenosis. The controversy concerns the use of the outflow patch in TOF. Artificial outflow patches should be used (teflon, dacron) instead of pericardium because of the risk of aneurysm formation in the latter[3]. We believe that the use of an outflow patch is not necessary if the immediate postoperative right ventricular/left ventricular pressure ratio is below 0.9. Residual stenosis in patients with this ratio is tolerated easily during the postoperative course, as shown by the study on late hemodynamic and clinical results of our patients[16].

Severe cyanosis is a risk factor in TOF. Cyanotic children develop increased collateral circulation, disseminated intravascular coagulation and a tendency towards postoperative bleeding. Increased collaterals and large bronchial arteries in cases with pulmonary vascular hypoplasia have been reported as leading to important

TABLE II

CAUSES OF MORTALITY IN 47 CASES

	No. cases
Operative Mortality (2.1%):	
Stone heart	3
Relaxed "dying" myocardium	2
Low cardiac output	1
Uncontrollable bleeding	1
Hospital Mortality (11.8%):	
Low cardiac output	11
Hypoplastic left ventricle	4
Pulmonary vascular insufficiency	3
Renal failure	3
Massive bronchial hemorrhage	3
Mediastinitis	3
Decerebration	2
Severe gastrointestinal bleeding	2
Rupture of right ventricle-hemorrhage	1
Heart block not responsive to pacemaker	1
Recurrent VSD	1
Paralytic ileus	1
Malignant hyperpyrexia	1
Coronary artery anomaly	1
Sudden arrhythmia (ventricular fibrillation)	1
Late Mortality (0.6%):	
Infectious hepatitis	1
Possible rupture of right ventricular aneurysm	1

pulmonary bleeding and dysfunction after total correction[17]. Pulmonary intravascular thrombosis occurs in cyanotic patients causing pulmonary vascular resistance[18]. Cyanotic tetralogies carry higher risks than acyanotic (pink) cases. We have no mortality in pink tetralogies.

Another possible risk factor for total correction in TOF is a previous shunt operation. There are several ill effects of long-standing palliative procedures. Selmonosky et al[19] stated that atresia of the right ventricular outflow tract might occur after systemic-pulmonary shunts. He also stated that the need for inserting an outflow patch during total correction is more frequent in such cases, but this could not be

demonstrated in our patients with long-standing presence (more than 10 years) of Blalock-Taussig shunts[10]. In our series of adult patients, none with previous palliative procedures needed right ventricular outflow patch reconstruction[10]. Chiariello et al[3] reported a slightly higher mortality rate in total correction of patients with previous shunts than in ones without any palliation. This is because of the ill effects of these shunts, including pulmonary hypertension and pulmonary vascular disease, which likewise were reflected in our series of adolescent and adult subjects[10].

Blalock-Taussig anastomosis has the fewest side effects compared to other types of palliations, such as Waterston and Potts anastomoses[3,4]. These side effects consist mainly of pulmonary hypertension and pulmonary vascular disease, encountered especially in the long-standing presence of these anastomoses.

Closure of some of the palliative shunts during the total correction is difficult and prolongs the duration of cardiopulmonary bypass and the operation. Closure of Potts anastomosis is complicated, and for this reason the shunt has recently been abandoned. Angulation of the pulmonary artery may occur in the Waterston shunt, causing some technical difficulties during its closure[20]. Hence, when a palliation is needed[3,4], the Blalock-Taussig anastomosis should be chosen, as is the practice in our clinic.

Shunting procedures have their own mortality rates, which, added to the mortality of total correction *per se,* lead to a high overall death rate in two-stage correction of TOF. The current mode of treatment in TOF is therefore the direct total correction (one-stage operation). Palliative procedures are still advised in cases of pulmonary vascular hypoplasia and/or pulmonary vascular disease where the patient cannot withstand direct total correction[9,21,22]. Pulmonary vascular bed usually develops 2-4 years following shunt operations[22], but as the duration of the shunt increases so does the incidence of pulmonary vascular disease[23]. We observed this fact in our adult subjects with long-standing presence of palliative procedures (more than 10 years) where mortality resulted mainly from the diseased pulmonary bed[10]. In our practice, we generally prefer that the time lapse between the shunt procedure and the total correction not exceed 2 to 3 years, which gives enough time for the development of the pulmonary vascular bed. A small left ventricle, evaluated by angiocardiography and echocardiography, is also an indication for a palliative procedure. Patients in this situation should be re-evaluated later and subjected to total correction in case the left ventricular cavity develops after the shunt operation.

Some additional pathologies, such as anomalous systemic venous return, coarctations of the pulmonary artery, apical cardiac diverticulum, atrio-ventricular canal, anomalous distribution of the coronary arteries, left ventricular obstruction and cor triatriatum, also add some risks to total correction by prolonging the duration of cardiopulmonary bypass and anoxic cardiac arrest[24-28]. Unrecognized patent

foramen ovale or atrial septal defect during the total correction may worsen the patient's post-operative prognosis by the presence of residual shunts[28]. The right atrium of every patient undergoing total correction through the right auricula should therefore be routinely checked just before the insertion of caval cannulas[1,28]. Accidental division of an abnormal coronary artery (commonly the left anterior descending branch originating from the right coronary artery) can lead to post-operative myocardial infarction and low cardiac output syndrome[26,28,29]. In one of our cases with TOF, the left anterior descending branch originated from the pulmonary artery and the left circumflex branch from the aorta, and the patient died of myocardial damage and low cardiac output syndrome during the early post-operative period. Autopsy revealed old and new areas of myocardial infarction[27]. In addition to total correction, aorto-left anterior descending coronary artery bypass using saphenous vein graft had also been performed in this case, and the vein graft was found to be patent at autopsy[27].

Surgically-produced atrioventricular block, the common cause of death in earlier years, is rare in current practice. Temporary blocks due to edema around the sutures close to the conduction system are treated with steroids, isoproterenol and temporary pacemakers[30]. We routinely insert epicardial electrodes during every total correction for TOF to be used in case of dysrhythmia after the operation. The need for permanent pacemakers after total correction is very rare (one case in our surviving patients).

Pulmonary hypertension after total correction due to recanalized ventricular septal defect (VSD), partial absence of the pulmonary arterial tree, peripheric pulmonary artery coarctations and previous pulmonary vascular resistance are also risk factors affecting mortality following total correction[21,31].

Cardiac failure and low cardiac output syndrome are the most common causes of death after total correction. Possible etiologies in cardiac failure are tabulated in Table III.

Some of the residual pathologies after total correction may necessitate re-operation. Donahoo et al[32] reported 15 cases of re-operation in a series of 343 patients (4.3%). The indications for re-operation were: recurrent VSD (9 cases), tricuspid regurgitation (2 cases), infection of the right ventriculotomy (3 cases) and residual right ventricular hypertension (1 case). VSD is the most common cause of residual pulmonary hypertension and cardiac failure requiring re-operation[15,32]. Tricuspid insufficiency after total correction for TOF is rarely encountered and may be the result either of injury to the tricuspid mechanism (primary type) or secondary to right ventricular dilatation (secondary type)[32]. Tricuspid valve replacement may be needed in primary type residual tricuspid valve insufficiency[32].

TABLE III

CAUSES OF CARDIAC FAILURE AFTER TOTAL CORRECTION

1. Inadequate relief of right ventricular outflow tract obstruction, immediate right ventricular/left ventricular pressure ratio after total correction being above 0.9.
2. Underdeveloped pulmonary vascular bed.
3. Narrow pulmonary arterial tree (hypoplasia, coarctations, etc.).
4. Pulmonary regurgitation due to the use of right ventricular outflow patch (transannular patch).
5. Postoperative tricuspid or aortic insufficiency due to inadequate surgical technique.
6. Recurrent VSD.
7. Postoperative pulmonary hypertension and increased pulmonary vascular resistance.
8. Hypoplastic left ventricle.
9. Severely cyanotic patients with increased collaterals.
10. Uncorrected additional anomalies (atrial septic defect, coronary artery anomalies, etc.).
11. Prolonged duration of extracorporeal circulation and anoxic arrest.
12. Unexpected sudden cardiac arrhythmias.
13. Dysrhythmias not responsive to pacemaker.
14. Previous shunt operations.
15. Infection of the prosthetic materials used during the total correction and bacterial endocarditis.
16. Poor postoperative care.

Summary

Of 322 patients undergoing total correction for tetralogy of Fallot by the authors of this article, 47 died (14.5%). Operative mortality was 2.1%, hospital mortality 11.8% and late mortality 0.6%. Cardiac failure and low cardiac output syndrome were the most common causes of death after total correction. Risk factors and causes of mortality after total correction for tetralogy of Fallot are reviewed.

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