Twenty-to-thirty-seven-year follow-up after repair for Tetralogy of Fallot

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Abstract

Objective: To describe the long-term prognosis after repair of Tetralogy of Fallot with pulmonary stenosis beyond 20 years. Methods: One hundred and eighty-five patients underwent corrective repair of Tetralogy of Fallot at Rigshospitalet in Copenhagen between January 1960 and July 1977. Ninety seven patients had undergone a palliative operation prior to Tetralogy of Fallot repair. All the 125 patients who were discharged from the hospital were traced through the population register and the patients alive July 1997 were contacted by mail and/or telephone and questioned about use of medicine, professional status, family status and ability to perform sport activities. Results: Sixty patients died in hospital and 125 patients, 78 males and 47 females, were discharged alive. Among operative survivors, median age at operation was 12.8 years (range 0.4–41 years). Thirteen patients required a reoperation, the main indication was failed VSD closure. There were 16 late cardiac deaths, out of which seven were sudden and unexpected and three were in immediate relation to reoperations. One hundred and nine patients were alive at follow-up. The mean follow-up time was 25.5 years (range 20–38 years). Sixteen percent used cardiac drugs, 89% were, or had been, working normally (all professions from academics to hard manual labors were represented), 53% (64% of women) had given birth after the repair and 51% performed sport activities regularly. No patients were lost to follow-up. Conclusions: The vast majority of the patients seemed to live normal lives 20–37 years after Tetralogy of Fallot repair. Late deaths were cardiac in origin, including sudden death from arrhythmias. The number of late reoperation has been low. Considering the natural history of the disease, Fallot repair has proven to be a beneficial procedure even including the very early experience short after introduction of open heart surgery. © 1999 Elsevier Science B.V. All rights reserved.

Keywords: Congenital heart disease; Tetralogy of Fallot; Surgery; Long term follow-up; Quality of life

1. Introduction

Tetralogy of Fallot is a rather common complex cardiac malformation with an incidence of 0.1/1000 live births. Without surgical intervention, patients had a 1 year survival rate of 66%, 49% after 2 years and only 10–15% after more than 20 years [1,2]. Today the awareness and knowledge of this natural history of Tetralogy of Fallot without cardiac surgery is fading. The natural history was dramatically changed with the introduction of palliative procedures by Blalock and Tausig in 1945 [3] and radical repair by Lillehei in 1954 [4] and Kirklin in 1955 [5]. The new ‘natural’ history with cardiac surgery is yet to be written.

Most previous papers have presented long-term follow-up with a mean follow-up of 5–20 years [6–12].

In 1982, the 14–19-year follow-up of our early series was published [13]. The object of the present study was to present the 20–37-year follow-up of our material.

2. Material and methods

All patients who underwent and survived corrective repair of Tetralogy of Fallot with pulmonary stenosis at our institution between January 1960 and July 1977, were included in the study. Patients with pulmonary atresia were excluded. Patient data were collected from patient records, including the operation notes and the perfusion atresia were excluded. Patient data were collected from patient records, including the operation notes and the perfusion records. To ascertain whether patients were alive, all patients were traced through the national population register. The patients who were still alive at the follow-up (1997) could all be located and contacted by mail and/or telephone and questioned about use of medicine, working ability, family status and ability to perform sport activities. The study was approved by the local ethical research committee.
2.1. Surgery

One hundred and eighty five patients underwent corrective repair of Tetralogy of Fallot with pulmonary stenosis at our institution between January 1960 and July 1977.

Prior to Tetralogy of Fallot repair, 97 patients had undergone a palliative surgical procedure. Seventy four had received a Blalock-Taussig shunt, nine had received pulmonary valvulotomy, 11 had received a Waterston-shunt, six had received a Brocks infundibulotomy, three had received a Pott-shunt, and four had received a Barrett pleurodesis. Sixteen patients had received more than one palliative procedure. Three patients had been explored via a thoracotomy, without any attempt of palliation performed.

Repair including relief of the right ventricular outflow obstruction and closure of the VSD was performed with

techniques and approaches corresponding to time. The VSDs were closed with a transventricular approach, always by use of patch. Muscle removal was generally more generous than today. A RV/LV of less than 0.75 was accepted.

About one-third of the patients received a transannular right ventricular outflow patch (pericardium or dacron) while three patients received valved conduits (one Polystan® pulmonary valve conduit and two Hancock conduits).

2.2. Statistics

Survival data have been evaluated using Kaplan–Meier product limit method. Comparisons between groups have been made using a log-rank test or chi square test, where appropriate.

3. Results

3.1. Activity and operative mortality

Out of the 185 patients operated with Fallot repair during the period, 60 patients (30 female, 30 male) died in hospital.
while 125 patients (78 males and 47 females) were discharged alive.

The operative activity during the following periods was:
1960±1964: 53 (27 discharged), 1965±1969: 27 (15 discharged), 1970±1974: 53 (39 discharged) and 1975±1977: 52 (44 discharged) (Fig. 1). Among operative survivors median age at operation was 12.8 years (range 5 months–41 years) (Fig. 2). At the follow-up, 109 patients were alive while another 16 patients had died, six of the reoperated.

3.2. Reoperations

Thirteen patients had been reoperated. In 10 patients, a persistent VSD was the main indication for reoperation. In one patient, VSD closure was combined with relief of persistent pulmonary valve stenosis, in one patient the VSD was combined with pulmonary valve insufficiency and both were repaired, and in one patient the VSD closure was combined with repair of tricuspid valve insufficiency. One patient was reoperated twice for VSD closure. One patient with severe pulmonary insufficiency and right ventricular dilatation received a homograft when reoperated 7 years following the second VSD closure. One patient was reoperated a second time for ASD closure and, in addition, the same patient had a pulmonary artery stenting in the catheterization laboratory.

One patient underwent an aortic valve repair for severe insufficiency. One patient was given a pulmonary stenosis repair by mono-cusp outflow patch implantation at the age of 13 and was operated for tricuspid insufficiency and persistent VSD 1 year later, finally to be heart transplanted at the age of 23, but died 3 weeks postoperatively.

Three patients died in immediate relation to the reoperation, including the heart transplant patient already mentioned. A further three of the reoperated patients died later.

3.3. Survival and causes of late death

Although previous palliative procedures were associated with increased operative mortality ($P = 0.04$ by Chi-square test), there were no differences in survival (from birth and from Fallot repair) between operative survivors who had had a palliative procedure prior to the Fallot repair and those who received the repair as the primary operation (Fig. 3). Sixteen patients had died between discharge and follow-up (1997). The causes of death were all cardiac, seven were sudden deaths probably related to arrhythmias. The time and causes of death are presented in Table 1. Three patients died early following the reoperation and another three died late.

3.4. Follow-up questionnaire (1997)

The 109 patients (45 female and 64 male) who were still alive at follow-up were all located and contacted. The mean follow-up interval for the surviving patients was 25.8 years (range: 20–37.5 years, a total of 2807 patient years). One hundred and five patients answered the questionnaire to some extent (mean age at follow-up, 38.6 years) while four did not answer.

Fifty two patients (28 female and 24 male) out of 97 who answered the question had one or more children. Two of the female patients had each given birth to a child with Tetralogy of Fallot, while all other children were healthy and none had congenital heart disease.

Seventeen patients out of 100 who answered the question were presently using some kind of cardiac medication (10 used anti-arrhythmics, 10 used diuretics, two used digoxin and two used anticoagulative medication).

Fifty patients out of 99 who answered the question performed sport activities, primarily athletics, badminton, swimming and bicycling.

Eighty nine patients out of 105 who answered the question were working in a variety of ordinary professions. All kinds of professions from academics to hard manual labor were represented. Out of the 16 who were not in employment, two were housewives, four were unemployed, one was retired, and eight received disability pension. The impression was that many of these patients did not attend regular medical check-ups.

Medical reexaminations were almost exclusively performed as a consequence of symptoms.

4. Discussion

The present series of patients after repair of Tetralogy of Fallot is one the largest series with a follow-up longer than

<table>
<thead>
<tr>
<th>Cause of death</th>
<th>Years from repair at death</th>
<th>Age at death</th>
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<tbody>
<tr>
<td>Sudden death</td>
<td>0.5</td>
<td>5.7</td>
</tr>
<tr>
<td>Post reoperation for VSD</td>
<td>0.6</td>
<td>15.7</td>
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<tr>
<td>Post reoperation for AI, endocarditis</td>
<td>0.8</td>
<td>7.2</td>
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<tr>
<td>VF during heart catheterization</td>
<td>1.4</td>
<td>7.4</td>
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<tr>
<td>Heart failure/pneumonia</td>
<td>10.9</td>
<td>26.3</td>
</tr>
<tr>
<td>Endocarditis/subarachnoidal hemorrhage</td>
<td>13.1</td>
<td>26.9</td>
</tr>
<tr>
<td>Heart failure post HTx</td>
<td>14.7</td>
<td>23.5</td>
</tr>
<tr>
<td>Sudden death</td>
<td>16.6</td>
<td>24.6</td>
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<tr>
<td>Sudden death</td>
<td>18.2</td>
<td>33.8</td>
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<tr>
<td>Pulmonary artery embolism</td>
<td>21.6</td>
<td>52.3</td>
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<tr>
<td>Sudden death</td>
<td>23.0</td>
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<td>Sudden death</td>
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<tr>
<td>Sudden death</td>
<td>25.7</td>
<td>37.8</td>
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<tr>
<td>Sudden death</td>
<td>28.4</td>
<td>50.5</td>
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<tr>
<td>Heart failure</td>
<td>30.5</td>
<td>46.5</td>
</tr>
<tr>
<td>Post reoperation for VSD + TI</td>
<td>33.1</td>
<td>54.1</td>
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</table>
operative period but eventually all will deteriorate and will serve well in the early post-operation but probably do little or no harm. Valved conduits might serve to reduce the immediate postoperative problems, while most of them will have no long-term function but probably do little or no harm. Valved conduits (homografts and others) will serve well in the early postoperative period but eventually all will deteriorate and become obstructed and will have to be replaced with regular intervals (10–15 years or less) [17]. The outcome in this series justifies the advice to minimize pulmonary insufficiency, as suggested above with the conservative approach to outflow patching, and reserve valved conduits for patients with an additional indication usually including a suspicion of increased pulmonary vascular resistance. The study by Norgard et al. [18] of the right ventricular diastolic function following Tetralogy of Fallot repair indicates that a restrictive physiology might be beneficial to long-term outcome. In the present material, only one patient was recently given a homograft in the right ventricular outflow tract for severe pulmonary insufficiency and ventricular dilatation.

In this series, there were seven late, sudden deaths indicating an important problem to look for and try to prevent. This incidence is identical to that observed by Jonsson et al. [19] while Waien et al. [11] found a very low risk in adults with previous repair. The cause of sudden death may be heart block or tachycardia/ventricular fibrillation. A trifascicular block pattern was associated with early sudden death [20]. The risk of symptomatic arrhythmia is high when marked right ventricular enlargement and QRS prolongation develop [21]. QRS prolongation relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death [21]. Earlier repair seems to reduce the risk of late ventricular arrhythmias [19]. Deane-field et al. [22] have found late Tetralogy of Fallot repair to be associated with more arrhythmias. This might explain our high incidence of late, sudden death. Transatrial VSD closure with short transannular incision, less aggressive muscle resection in right ventricular outflow tract and less generous outflow patching have been found to be associated with reduced ventricular arrhythmias and less right ventricular dilatation/dysfunction and improved working capacity [23,24]. Diagnostic possibilities and treatment options have increased, and today the treatment includes the option of cardioverter-defibrillator implantation. Hopefully, more aggressive medical treatment of ventricular arrhythmias, will reduce the number of sudden deaths. Arrhythmias after repair of Tetralogy of Fallot also include atrial tachycardia [25].

As a consequence of the present study, the patients will be approached and offered a check-up by a qualified cardiologist. It will take a long time before all patients have been reexamined. It is possible that a few patients in the series will be in need of a reoperation, e.g. those three who primarily received valved conduits. In addition, such a follow-up program can be expected to produce additional interesting information.

5. Conclusions

The majority of survivors following repair of Tetralogy of Fallot live normal lives. Future patients may hope that an earlier, more perfect operation and a more close postoperative medical follow-up and treatment of complications will...
reduce the risk of sudden death, improve the working capacity and reduce the need for reoperation.

References


Appendix A. Conference discussion

Dr E. Baudet (Bordeaux, France): Do you consider that current myocardial protection with cardiopilia, if available at that time, could have avoided some of these late deaths?

Dr Norgaard: We expect that if the patients had been followed closer we may have been able to discover some cardiac arrhythmias and intervene. As a consequence of this study, we will offer all the patients that are still alive to be followed closely by cardiologists.

Dr C. Knott-Craig (Oklahoma, OK): With respect to the late deaths, were any of those related to exercise? How do you advise your patients whether they are able to exercise? For example, do you do an exercise Holter monitoring before allowing them to exercise, or do you just give them carte blanche ability to exercise if they feel fit enough?

Dr Norgaard: We have not pointed out any kind of restrictions to these patients. We think if the patients should really benefit from Fallot repair, they should be able to live a normal life, and at controls performed by cardiologists, cardiac arrhythmias should be discovered.

Dr Knott-Craig: Were any of the late deaths related to exercise at all?

Dr Norgaard: We don’t know what the patients were doing at the time of death.