LONG-TERM EVALUATION AFTER TOTAL CORRECTION OF TETRALOGY OF FALLOT

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Cardiac functions of 28 patients long after correction of tetralogy of Fallot (TOF) were examined using non-invasive and invasive methods. For comparison, patients were grouped in 2 different ways: those with a right ventricular systolic pressure below 50 mmHg (Group A, n = 10) and above 50 mmHg (Group B, n = 18) in the early post-operative stage; those having an ability to complete the treadmill test (Group I, n = 14) and those without it (Group II, n = 10).

The following results were obtained by our study performed more than 10 years after surgery: 1) Group A showed no differences in all parameters studied as compared with Group B. 2) As compared with the normal subjects (control group, n = 20), the patients had a larger cardio-thoracic ratio (CTR) and a larger right/left ventricular dimension ratio (RVD/LVD), but showed no differences in percent fractional shortening (%FS), mean velocity of circumferential fiber shortening (mVcf), corrected ejection time (ETc) and ejection fraction (EF). 3) As compared with Group I, Group II had a larger CTR, a larger RVD/LVD ratio, and continued higher levels of the right/left ventricular systolic pressure ratio (RVP/LVP) and right ventricular pulmonary systolic pressure gradient (PG), but showed no differences in %FS, mVcf, ETc, EF and pulmonary artery pulse pressure/pulmonary arterial systolic pressure ratio. 4) Eight patients, all belonging to Group II, had ventricular arrhythmia (Lown's grade 3 or 4). 5) Severe pulmonary regurgitation (PR) was noted in Group II.

At a later time long after the correction of TOF, RVP, PG, PR and arrhythmia are considered to be important parameters for evaluating the cardiac status in this disorder.

RECENT progress in cardiovascular surgery can benefit most infants with tetralogy of Fallot (TOF) by effective hemodynamic correction and by satisfactory protection from heart blocks, and many kinds of complications can be prevented by well-controlled operative procedures. Therefore, many institutions have succeeded in decreasing the operative mortality during corrective surgery from 16% in 1960 to 3% in 1980.6

We, the Second Department of Surgery, Niigata University School of Medicine, Niigata, Japan, have also achieved satisfactory results and the mortality rate was 9% during the 5 years from

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Japanese Circulation Journal Vol. 47, September 1983 1047

At present, infants with TOF can live a reasonably normal life up to adulthood with the assistance of the various recently developed medical aids. However, this satisfactory results has already produced new problems concerning the management of patients who have reached adulthood following corrective surgery of TOF.

Thus, the present study is designed to examine invasively and non-invasively the hemodynamics, the occurrence of arrhythmia and the exercise ability for adult patients more than 10 years after the correction of TOF, and to clarify the correlations among them.

SUBJECTS AND METHODS

Our departments have seen 120 adults patients more than 10 years after the corrective surgery which was performed between 1965 and 1970. We obtained permission to examine 28 of these patients. Corrective surgery had been carried out an average of 13 years before this study, with a range of 10 to 16 years. The patients included 11 males and 17 females, and their average age was 22 years with a range of 11 to 42 years. Twenty healthy subjects underwent an exercise test as the control group (10 males, 10 females, average age 25).

They were examined using both the conventional methods (clinical history, physical findings, laboratory data, chest X-ray, electrocardiography, mechanophono cardiography and echocardiography) and some special methods, such as dynamic electrocardiography (Holter's system), treadmill exercise test (Ellestad's method), cardiac catheterization and angiography.

The cardio-thoracic ratio (CTR) was calculated from a chest X-ray film. Arrhythmia was analyzed from grade 0 to 5 according to Lown's classification using Holter's ECG. The corrected ejection time (ETc) was calculated from carotid arterial waves.

Echocardiograms were obtained using a Toshiba SSH-11A or an SSL-53M electron sector type ultrasonic tomograph. From the right ventricular dimension (RVD) and the left ventricular dimension (LVD) at the end-diastole on an M-mode echocardiogram, RVD/LVD x 100% was calculated. In addition, percent fractional shortening (%FS), mean velocity of circumferential fiber shortening (mVcf) and the ejection fraction (EF) were measured.

Cardiac catheterization was performed twice: a few days after surgery (24-72 hours) and much later (10 years after). The right ventricular systolic pressure (RVP), left ventricular systolic pressure (LVP), the right/left ventricular systolic pressure ratio (RVP/LVP x 100, %), pressure gradient between the right ventricle and the pulmonary artery (PG) and the pulmonary arterial regurgitant index (PRI = pulmonary artery pulse pressure/pulmonary arterial systolic pressure x 100, %) were calculated. Pulmonary arteriograms were used to estimate the degree of pulmonary valve regurgitation.

We excluded patients who had an incomplete correction of ventricular septal defect.

From the findings of the cardiac catheterization and the treadmill exercise test the patients were classified in 2 ways: According to the level of RVP obtained by cardiac catheterization performed early after surgery, 10 patients whose RVP could be reduced below 50 mmHg after the corrective surgery were classified as Group A, and 18 patients whose RVP remained over 50 mmHg were classified as Group B. According to the results of the treadmill exercise test, 14 patients who could complete the whole program were classified as Group I and 10 patients who could not were classified as Group II.

All data was presented as mean ± SD and were statistically analyzed using Student's t-test.

RESULTS

Effects of the Surgery

The RVP/LVP ratio early after surgery were 46 ± 16% in Group A and 74 ± 16% in Group B. However, the RVP/LVP ratio long after surgery showed no significant difference: 51 ± 25% in Group A and 52 ± 18% in Group B. No significant differences were noted either in echocardiograms, pulse waves or CTRs (Table I).

Exercise Test

The exercise test was not completed in Group II due to an occurrence of symptoms such as chest pain or hypotension. However, testing was possible for an average of 5 min. There were significant differences between Groups I and II in CTR, RVD/LVD, PG and RVP/LVP long after surgery, and all these parameters were greater in Group II (Table II). In Groups I and II, CTR was 53 ± 4 and 60 ± 6% (p < 0.01), RVD/LVD was 67 ± 15 and 85 ± 12% (p < 0.01), PG was 11 ± 7 and 34 ± 33 mmHg (p < 0.02) and RVP/LVP was 45 ± 10 and
### TABLE I  CLINICAL CHARACTERISTICS OF PATIENTS GROUPED ACCORDING TO A POST-OPERATIVE DECREASE IN RIGHT VENTRICULAR SYSTOLIC PRESSURE

<table>
<thead>
<tr>
<th>Number of patients</th>
<th>Early after surgery</th>
<th>Long after surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>RVP/LVP (%)</td>
<td>PG (mmHg)</td>
</tr>
<tr>
<td>Group A</td>
<td>46 ± 16</td>
<td>51 ± 25</td>
</tr>
<tr>
<td>Group B</td>
<td>74 ± 16</td>
<td>52 ± 18</td>
</tr>
<tr>
<td>p &lt; 0.001</td>
<td>ns</td>
<td>ns</td>
</tr>
</tbody>
</table>

Values are expressed as mean ± SD. Group A: right ventricular pressure < 50 mmHg. Group B: right ventricular pressure ≥ 50 mmHg. Abbreviations: RVP/LVP = right/left ventricular systolic pressure ratio; PG = right ventricular-pulmonary artery systolic pressure gradient; RVD/LVD = right/left ventricular dimension ratio; %FS = percent fractional shortening; mVcf = mean velocity of circumferential fiber shortening; EF = ejection fraction of the left ventricle; CTR = cardio-thoracic ratio; ns = not significant.

### TABLE II  CLINICAL CHARACTERISTICS IN PATIENTS GROUPED ACCORDING TO A RESPONSE TO TREADMILL EXERCISE TEST

<table>
<thead>
<tr>
<th>Number of patients</th>
<th>Early after surgery</th>
<th>Long after surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>RVP/LVP (%)</td>
<td>PG (mmHg)</td>
</tr>
<tr>
<td>Control group</td>
<td>20</td>
<td>–</td>
</tr>
<tr>
<td>Group I</td>
<td>14</td>
<td>74±21</td>
</tr>
<tr>
<td>Group II</td>
<td>10</td>
<td>65±16</td>
</tr>
<tr>
<td>p value (Group II vs Group I)</td>
<td>ns</td>
<td>&lt; 0.01</td>
</tr>
</tbody>
</table>

Values are expressed as mean ± SD. *p < 0.001 as compared with the control group. Group I: patients completing the test. Group II: patients unable to complete the test. PRI (pulmonary arterial regurgitant index) = pulmonary arterial pulse pressure / systolic pulmonary arterial pressure x 100, %. Other abbreviations are the same as in Table I.
TABLE III ELECTROCARDIOGRAPHIC FINDINGS IN TWO GROUPS CLASSIFIED
ACCORDING TO THE TREADMILL EXERCISE TEST

<table>
<thead>
<tr>
<th>Number of patients</th>
<th>iRBBB</th>
<th>CRBBB</th>
<th>Bifascicular block</th>
<th>Lown’s grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group I</td>
<td>14</td>
<td>3</td>
<td>11</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>8 6 0 0 0 0 0</td>
</tr>
<tr>
<td>Group II</td>
<td>10</td>
<td>0</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>2 0 0 2 4 2 0</td>
</tr>
</tbody>
</table>

Group I: patients completing the test.
Group II: patients unable to complete the test.
iRBBB = incomplete right bundle branch block; CRBBB = complete right bundle branch block

62 ± 22% (p < 0.01), respectively. The first 2 parameters were significantly higher in both groups as compared with the control group (p < 0.001).

On the other hand, %FS (39 ± 5 and 36 ± 7%), mVcf (1.3 ± 0.2 and 1.3 ± 0.3 circ/sec), EF (70 ± 5 and 65 ± 9%) and ETC (0.407 ± 0.015 and 0.403 ± 0.039 sec) did not show any significant differences between Groups I and II. In addition, these parameters did not show any significant differences as compared with the control group.

The PRls in Groups I and II were 84 ± 16% and 76 ± 14%, respectively, showing no significant difference. However, pulmonary arteriography revealed a high degree of regurgitation in 4 cases of Group II.

ECG Examination: In Group I, an incomplete right bundle branch block was observed in 3 cases, and a complete right bundle branch block was noted in all the remaining cases. In Group II a complete right bundle branch block was observed in all cases. In Group I Lown’s grade 1 arrhythmias were found in 6 cases and grade 0 in the remainder, while in Group II 6 cases had grade 4 arrhythmias, 2 cases had grade 3 and the remainder had grade 0 (Table III).

DISCUSSION

The total number of radical operations for TOF performed during the 6 years from 1965 to 1970 at the 2nd Department of Surgery, Niigata University, was 120. The mortality rate at that time was 9%. Thereafter, 7 deaths occurred at a later time after surgery, and the survival rate 10 years after the operation was 85% (102 cases).

The major problem involved in a radical operation of TOF is how to release the obstruction of the right ventricular outflow. The success of the operation has been evaluated as the lowering of the RVP/LVP to below 0.5—12

However, by dividing patients into groups according to whether or not their post-operative right ventricular pressure had declined, the present study revealed no differences between the 2 groups in the right ventricular size, arrhythmias, left heart function and reserve function for exercise. These results may be associated with various factors at the time of surgery, including right ventriculotomy and the use of patches. It is believed that the long-term hemodynamics may not always be correlated with those in the early post-operative stage even if the right ventricular pressure declines.13,14

In addition, the 2 groups classified according to their response to the treadmill exercise test were compared. No differences were found in the parameters at rest reflecting the left heart function, such as %FS, mVcf, EF and ETC. However, the RVP/LVP ratio was significantly higher in Group II, in which exercise loading had to be halted due to the occurrence of symptoms. It was also found that the degree of pulmonary arterial stenosis, which presumably constitutes one of the important factors for long-term hemodynamics, was higher in Group II. However, Group II included 4 cases with no pulmonary arterial stenosis and no arrhythmia. The PRls of these cases ranged from 88 to 100% (95% on the average) and all of their pulmonary arteriograms showed a high degree of regurgitation. Thus, pulmonary arterial valve regurgitation may be an important long-term factor.

No difference in LVD on M-mode echocardiograms was observed between the control group and the patient group, and the high values of the RVD/LVD ratio were caused by the enlargement of RVD (1.6 ± 0.4 and 3.3 ± 0.8 cm, respectively), so that the enlargement of the CTR is at-

Japanese Circulation Journal Vol. 47, September 1983
Long-term Evaluation of TOF

...tributable mainly to the right ventricle.

In Group II, ventricular arrhythmia and bifascicular block attributable to the right ventricular lesions were noted in many cases and these cases are thus considered to require careful observation in the future.

Although it is generally thought that invasive follow-up inspections are not always advisable, non-invasive inspections are said to be insufficient to determine the degrees of pulmonary arterial stenosis and regurgitation as well as the levels of the right ventricular pressure.

In the present study, post-operative cardiac function was examined using chest X-ray, 24-hour ECG, carotid arterial waves, treadmill exercise test, echocardiography and cardiac catheterization.

For evaluating cardiac function in TOF long after surgery, examinations not only of the right ventricular pressure, but also of pulmonary arterial stenosis, regurgitation and arrhythmia are important.

REFERENCES
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