CONGENITAL HEART DISEASE

Health related quality of life and health status in adult survivors with previously operated complex congenital heart disease

M Kamphuis, J Ottenkamp, H W Vliegen, T Vogels, K H Zwinderman, R P Kamphuis, S P Verloove-Vanhorick

Objective: To examine the impact of previously operated complex congenital heart disease on health related quality of life and subjective health status and to determine the relation between these parameters and physical status.

Setting: Patients were randomly selected from the archives of the paediatric cardiology department, Leiden University Medical Centre, Leiden, The Netherlands, and approached irrespective of current cardiac care or hospital of follow up.

Patients: Seventy eight patients with previously operated complex congenital heart disease (now aged 18–32 years) were compared with the general population.

Main outcome measures: Health related quality of life was determined with a specifically developed questionnaire [Netherlands Organisation for Applied Scientific Research Academic Medical Centre (TNO-AZL) adult quality of life (TAAQOL)] and subjective health status was assessed with the 36 item short form health survey (SF-36). Physical status was determined with the objective physical index, Somerville index, and New York Heart Association functional class.

Results: Health related quality of life of the patients was significantly worse than that of the general population in the dimensions gross motor functioning and vitality (p < 0.01). Correlations between health related quality of life and physical status were poor. Patients had significantly worse subjective health status than the general population in the dimensions physical functioning, role functioning physical, vitality, and general health perceptions (p < 0.01). Correlations between subjective health status and physical indices were weak.

Conclusion: Adult survivors with previously operated complex congenital heart disease experienced limitations only in the physical dimensions of health related quality of life and subjective health status. Objectively measured medical variables were only weakly related to health related quality of life. These results indicate that, when evaluating health related quality of life, dedicated questionnaires such as the TAAQOL should be used.

Advances in surgery and intensive care have increased the life expectancy of children with complex congenital heart disease. Therefore, it is important to assess the long term results of treatment of these patients, not only in medical terms (for example, morbidity and mortality) but also in terms of health related quality of life and subjective health status. Since a longer life does not necessarily mean a better life, knowledge of the emotional response to health status problems may allow identification of areas of daily life that need to be improved (for example, by medical care or by provisions in social or infrastructural care management). Moreover, patients’ satisfaction with the medical consultation may increase if physicians assess a patient’s general wellbeing, as well as functional status.  Some studies have explored the subjective health status of patients with congenital heart disease in adulthood but none have examined health related quality of life using a clear definition. In addition, correlating health related quality of life with the patient’s physical condition may increase our understanding of the concept of health related quality of life.

Therefore, in a group of patients with previously operated complex congenital heart disease, the aim of the study was, firstly, to compare patients’ health related quality of life and subjective health status with those of the general Dutch population (age and sex matched) and, secondly, to relate health related quality of life and subjective health status to physical status.

METHODS

The study was approved by the local medical ethical committee and informed consent was obtained from each participant. A cross sectional design was used and information on medical follow up was searched for retrospectively.

Definitions

Health related quality of life

Although there is no universal definition for health related quality of life, there is growing consensus that it should be assessed multidimensionally, including physical health, psychological state, and social relationships. Moreover, quality of life and health status are distinct constructs; quality of life is determined not only by the patient’s health status problems but also by the emotional response to these problems. To measure quality of life according to this definition, quality of life...
life questionnaires have been developed for children (Netherlands Organisation for Applied Scientific Research Academic Medical Centre (TNO-AZL) child quality of life TACQOL questionnaire) and for adults (TNO-AZL adult quality of life (TAAQOL) questionnaire). Both questionnaires have been validated in the general population as well as in patients with various chronic diseases, such as minor congenital heart disease, and thoroughly validated. Fig 1 shows an example of the format of the questionnaires. Health related quality of life is used since non-health factors such as income or housing (which may also contribute to quality of life) were not taken into account. For clinicians health related quality of life is most relevant.

The 36 item short form health survey (SF-36) is widely used and thoroughly validated. It measures subjective health status, posing such questions as “What are your limitations?” or “Do you feel healthy?” in contrast to “How do you feel about certain limitations?” in the TAAQOL.

Diagnosis

In the present study, patients with previously operated complex congenital heart disease were approached, excluding those who had undergone anatomical corrective surgery. All patients included in this study had undergone a non-anatomical correction—that is, partial or complete cavopulmonary connection, venous switch for transposition of the great arteries, or a correction with the use of allogeneous tissue (for example, Rastelli correction, insertion of a mechanical valve). The main common characteristic was that postoperatively none of the participants had an anatomically normal heart as described by the segmental analysis, as opposed to patients after anatomical corrective surgery (for example, operated ventricular or atrial septal defects, coarctation of the aorta, patent ductus arteriosus, uncomplicated tetralogy of Fallot, total anomalous pulmonary venous connection, and uncomplicated pulmonary or aortic stenosis).

Patient selection

To prevent selection bias, patients were selected from the archives of the department of paediatric cardiology, Leiden University Medical Centre, Leiden, The Netherlands, which has complete information on patients seen from 1950 onwards. From these files, 4383 patients were born between 1968 and 1982. Of these, in the year 1999, 2280 were randomly selected. Then, 251 patients with operated complex congenital heart disease were included and further studied irrespective of current cardiac surveillance. The latest medical information and home addresses were obtained from the most recent medical file, general practitioners, or local authorities. Among the 251 included patients, 123 (49%) had died. Others (n = 41) were excluded for the following reasons: they were not living in the Netherlands or did not speak Dutch n = 28 (11%), they had learning disabilities n = 12 (5%), or they had participated in a pilot study n = 1 (0.5%). One patient was lost to follow up. The remaining 86 patients were invited to participate in the study; of these, 78 patients agreed to participate (91%). Table 1 lists the diagnoses and gives the number of patients per diagnosis or operation.

Measurements

Questionnaires on health related quality of life and subjective health status were completed.

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**Table 1** Diagnosis of 78 patients previously operated on for complex congenital heart disease (number of patients is given in parentheses)

<table>
<thead>
<tr>
<th>Main diagnosis</th>
<th>Main operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Systemic right ventricle (39)</td>
<td>Senning (12) or Mustard (10)</td>
</tr>
<tr>
<td>TGA (22)</td>
<td>Senning (3) or Mustard (2) with PS repair</td>
</tr>
<tr>
<td>TGA + PS (5)</td>
<td>Senning (5) or Mustard (1) with VSD closure and PS repair</td>
</tr>
<tr>
<td>TGA + VSD + PS (6)</td>
<td>VSD closure with PS repair (2)</td>
</tr>
<tr>
<td>LTGA + VSD + PS (2)</td>
<td>VSD closure and ASD closure (1)</td>
</tr>
<tr>
<td>LTGA + VSD + ASD (1)</td>
<td>VSD closure (1)</td>
</tr>
<tr>
<td>LTGA + VSD (1)</td>
<td>Tricuspid valve replacement (1)</td>
</tr>
<tr>
<td>LTGA + PS (1)</td>
<td>Tricuspid valve replacement and conduit (1)</td>
</tr>
<tr>
<td>AV discordance + DORV + PS (1)</td>
<td></td>
</tr>
<tr>
<td>2. conduit or mechanical prosthesis (23)</td>
<td></td>
</tr>
<tr>
<td>PA + VSD (1)</td>
<td>Rastelli (3), correction with other conduit (B)</td>
</tr>
<tr>
<td>AaS (4); AVSD (1); MI (1)</td>
<td>Mechanical prosthesis (6)</td>
</tr>
<tr>
<td>TGA + VSD + PS (4)</td>
<td>Rastelli (4)</td>
</tr>
<tr>
<td>TGA + VSD (1)</td>
<td>Arterial switch operation with conduit(1)</td>
</tr>
<tr>
<td>TrArt (1)</td>
<td>Correction with conduit (1)</td>
</tr>
<tr>
<td>3. Univentricular connection (11)</td>
<td></td>
</tr>
<tr>
<td>TA (8); DLV (2); hypoplastic LV (1)</td>
<td>Fontan (11)</td>
</tr>
<tr>
<td>4. Palliative operation- shunt (5)</td>
<td></td>
</tr>
<tr>
<td>AVSD (1)</td>
<td>Glenn shunt (1)</td>
</tr>
<tr>
<td>DLV (1)</td>
<td>Blalock-Taussig shunt (1)</td>
</tr>
<tr>
<td>Univentricular heart (1)</td>
<td>Modified Blalock-Taussig shunt (1)</td>
</tr>
<tr>
<td>TA (1)</td>
<td>After Glenn shunt: AV shunt axillary (1)</td>
</tr>
<tr>
<td>TA + TGA (1)</td>
<td>After banding AP: PH (1)</td>
</tr>
</tbody>
</table>

AaS, aortic stenosis; AP, pulmonary artery; ASD, atrial septal defect; AV, atrioventricular; AVSD, atrioventricular septal defect; DLV, double inlet left ventricle; DORV, double outlet right ventricle; LTGA, congenitally corrected TGA; LV, left ventricle; MI, mitral insufficiency; PA, pulmonary atresia; PH, pulmonary hypertension; PS, pulmonary stenosis; TA, tricuspid atresia; TGA, simple transposition of the great arteries; TrArt, common truncus arteriosus; VSD, ventricular septal defect.
higher scores indicating better subjective health status.

Scores range from 0 to 100, with higher scores indicating better quality of life.

Subjective health status
The SF-36 was used to measure subjective health status. This questionnaire is based on 35 items divided into eight domains (the number of questions per domain is indicated in parentheses): physical functioning (4), role functioning physical (4), bodily pain (2), general health perceptions (5), vitality (4), social functioning (4), daily activities (4), sexual activity (2), vitality (4), happiness (4), depressive moods (4), and aggressiveness (3). For each item, the frequency of occurrence of a health status problem is assessed. If such a problem is reported, the emotional reaction to this problem is also determined. Fig 1 shows an example of the format of the question. The reference period is formulated as “the last month”. The two questions of the domain sexual activity were not appropriate for the present study since the occurrence of problems was compared with one month previously (“Were you less sexually active than a month ago?”). Because such questions are only relevant when an intervention has taken place, this domain was not considered. Scores of each subscale are normalised to a scale ranging from 0 to 100, with higher scores indicating better quality of life.

Health related quality of life
The TAAQOL is a Dutch questionnaire developed by TNO and the Leiden University Medical Centre. Psychometric performance (reliability and validity) of the TAAQOL is satisfactory. It consists of 45 questions divided into 12 domains. Each domain contains two to four questions (the actual number per domain is given in parentheses): gross motor functioning (4), fine motor functioning (4), pain (4), sleeping (4), cognitive functioning (4), social functioning (4), daily activities (4), sexual activity (2), vitality (4), happiness (4), depressive moods (4), and aggressiveness (3). For each item, the frequency of occurrence of a health status problem is assessed. If such a problem is reported, the emotional reaction to this problem is also determined. Fig 1 shows an example of the format of the question. The reference period is formulated as “the last month”. The two questions of the domain sexual activity were not appropriate for the present study since the occurrence of problems was compared with one month previously (“Were you less sexually active than a month ago?”). Because such questions are only relevant when an intervention has taken place, this domain was not considered. Scores of each subscale are normalised to a scale ranging from 0 to 100, with higher scores indicating better quality of life.

Table 2 New York Heart Association (NYHA) functional class for patients with cardiac disease and ability index of Somerville

<table>
<thead>
<tr>
<th>NYHA classes</th>
<th>Ability index of Somerville (range 1–4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>No limitations of physical activities. Ordinary activity does not cause undue anginal pain, dyspnoea, fatigue, or palpitations.</td>
</tr>
<tr>
<td>II</td>
<td>Slight limitations of physical activities. Comfortable at rest, but ordinary activity results in anginal pain, dyspnoea, fatigue, or palpitations.</td>
</tr>
<tr>
<td>III</td>
<td>Marked limitations of physical activities. Comfortable at rest, less than ordinary activity causes anginal pain, dyspnoea, fatigue, or palpitations.</td>
</tr>
<tr>
<td>IV</td>
<td>Unable to carry on any physical activities without discomfort. Symptoms of cardiac insufficiency or anginal syndrome may be present even at rest. If any physical activity is undertaken discomfort is increased.</td>
</tr>
</tbody>
</table>

Figure 2 Outcome of physical status in the three indices (n = 78).

Statistical analyses
Sample size was based on results of the pilot study in which a mean difference of 11 units (on the 0 to 100 scale) was found between patients and the general population on the SF-36 vitality scale. Similar effect sizes are reported in the American SF-36 manual for similar conditions. The sample size (n = 80) was chosen to have 80% power for such a difference. With this sample size there is at least an 80% power to detect a correlation coefficient of 0.30 or more.

The participants’ health related quality of life (as measured by the TAAQOL) and their subjective health status (as measured by the SF-36) were compared with those of a random sample of the general Dutch population using analysis of variance with adjustment for age and sex. The sample of the general Dutch population comprised 831 persons for the TAAQOL and 361 for the SF-36. After comparison, the data were summarised as mean values and 95% confidence intervals.

Within the group of patients, the overall association between health related quality of life and subjective health status on the one hand and the objective physical index, the Somerville index, and the NYHA class on the other was quantified using canonical correlation analysis to reduce the number of statistical tests. With this method, the maximum association between two sets of variables is quantified. When the canonical correlation was significant, pairwise Spearman rank correlations were used to quantify the association between the various indices.

Since the distribution of a number of these variables was skewed, all statistical analyses were performed on rank transformed data. A probability value of p ≤ 0.01 was considered significant because many statistical tests were done. SPSS for Windows version 10.07 (SPSS Inc, Chicago, Illinois, USA) was used to perform statistical calculations.
RESULTS
Patient characteristics
There was an overall response rate of 91%. Medical data were available for all 78 participants and questionnaires for 76 of them. There were 44 men and 34 women, with an average age of 24.3 years (range 18–32 years). At the time of the study, 46 participants had a job for more than 12 h/week (59%; two patients were receiving a partial disability benefit), 15 were receiving a full or partial disability benefit (19%), 11 were still on sick leave or unemployed. Participants lived with a partner n = 34 (43%) or with their parents n = 31 (40%), had their own house n = 7 (9%), or lived in a student (or other) house n = 6 (8%).

For n = 44 men the average age was 24.3 years (range 18–32 years); 45 patients (58%) had had an interventional procedure (such as Rashkind septostomy or banding of the pulmonary artery). The mean age at first operation or intervention was 2.2 (range 0–24) years; 45 patients (58%) had undergone a previous palliative procedure. The patients were in NYHA class II and were thus experiencing slight limitations in physical activities.

Health related quality of life: patients compared with the general population
Fig 3 gives the average TAAQOL scores of the patients and of the general population sample. The patients had lower health related quality of life in almost all dimensions and scored significantly lower on gross motor functioning (p < 0.001) and vitality (p < 0.01).

Health related quality of life related to physical status: within the patient group
Several health related quality of life dimensions correlated significantly with the objective physical index, the Somerville index, and the NYHA class (table 3). Canonical correlation analysis showed that the first canonical correlation was significant.

Table 3  Spearman’s r correlation for medical indices correlated with the TNO-AZL adult quality of life questionnaire (TAAQOL) and the 36 item short form health survey (SF-36) for 76 patients previously operated on for complex congenital heart disease

<table>
<thead>
<tr>
<th>Questionnaire</th>
<th>Objective physical index</th>
<th>Somerville index</th>
<th>NYHA class</th>
</tr>
</thead>
<tbody>
<tr>
<td>TAAQOL</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gross motor functioning</td>
<td>-0.33*</td>
<td>-0.53**</td>
<td>-0.77**</td>
</tr>
<tr>
<td>Daily activities</td>
<td>-0.23</td>
<td>-0.46**</td>
<td>-0.45**</td>
</tr>
<tr>
<td>Vitality</td>
<td>-0.12</td>
<td>-0.42**</td>
<td>-0.47**</td>
</tr>
<tr>
<td>Sleep</td>
<td>-0.25</td>
<td>-0.38**</td>
<td>-0.43**</td>
</tr>
<tr>
<td>Social functioning</td>
<td>-0.10</td>
<td>-0.30*</td>
<td>-0.35*</td>
</tr>
<tr>
<td>Pain</td>
<td>-0.03</td>
<td>-0.24</td>
<td>-0.36**</td>
</tr>
<tr>
<td>Cognitive functioning</td>
<td>-0.30*</td>
<td>-0.33*</td>
<td>-0.32*</td>
</tr>
<tr>
<td>Depressive moods</td>
<td>-0.17</td>
<td>-0.24</td>
<td>-0.31*</td>
</tr>
<tr>
<td>Fine motor functioning</td>
<td>-0.18</td>
<td>-0.22</td>
<td>-0.29*</td>
</tr>
<tr>
<td>Happiness</td>
<td>-0.19</td>
<td>-0.06</td>
<td>-0.22</td>
</tr>
<tr>
<td>Aggressiveness</td>
<td>-0.03</td>
<td>-0.15</td>
<td>-0.19</td>
</tr>
</tbody>
</table>

| SF-36                        |                          |                  |            |
| Physical functioning         | -0.42**                  | -0.64**          | -0.87**    |
| Mental health                | -0.36**                  | -0.16            | -0.2       |
| Social functioning           | -0.24                    | -0.54**          | -0.45**    |
| Role functioning physical    | -0.28                    | -0.48**          | -0.42**    |
| General health               | -0.25                    | -0.41**          | -0.47**    |
| Vitality                     | -0.25                    | -0.31*           | -0.45**    |
| Bodily pain                  | -0.22                    | -0.32*           | -0.45**    |
| Role functioning emotion     | -0.24                    | -0.20            | -0.22      |

*p<0.01; **p<0.001
significant \( r = 0.768 \); in particular the NYHA class was the most important correlate of gross motor functioning. The pairwise Spearman correlations (table 3) showed the following pattern: health related quality of life correlated weakly with the objective physical index but significantly with gross motor functioning and cognitive functioning. The strongest relations were between the TAAQOL dimensions and the NYHA class.

**Subjective health status: patients compared with the general population**

The SF-36 dimensions physical functioning, role functioning physical (\( p < 0.01 \)), vitality, and general health perception were significantly lower (\( p < 0.001 \)) in patients than in the general population (fig 4).

**Subjective health status related to physical status: within the patient group**

Almost all SF-36 dimensions correlated significantly with the three physical indices (table 3). Canonical correlation analysis showed that the NYHA class was the strongest correlate of the SF-36 dimensions and of physical functioning in particular (\( r = 0.864 \)). The patterns for pairwise Spearman correlations were the same as those for health related quality of life, with the strongest relations between subjective health status dimensions and the NYHA class (table 3).

**DISCUSSION**

This study evaluated health related quality of life and subjective health status in adults with previously operated complex congenital heart disease and determined the relation between these parameters and physical status.

**Patients compared with the general population**

Comparison between the patients and the general population showed significantly lower scores in the patient group in the physical dimensions only: gross motor functioning and vitality (in the TAAQOL) and physical functioning, role functioning physical, general health perceptions, and vitality (in the SF-36). Thus, both physical function and appraisal of physical dysfunction were significantly lower in the patient group.

There was no significant difference in any of the scores for the non-physical dimensions (social functioning, role functioning emotional, daily activities, depressive moods, etc) between patients and the general population. Some studies on social, psychological, and behavioural outcome in children and adolescents with heart disease have reported problems in these fields, whereas others show normal social behaviour and successful educational and occupational achievements in this group of patients, thus supporting the promising outcome of the present study. These latter studies indicate that, over time, patients may adapt to their disease and that denial may even help to normalise functioning.

To improve the patients' long term quality of life, physical aspects must be taken into account.

Firstly, appropriate exercise should be encouraged to avoid patients imposing unnecessary restrictions on themselves, sometimes caused when patients receive insufficient information from the specialist. In addition, parents should be informed that overprotection can have a negative impact on the physical activities of the child. Counselling should therefore, be based on a thorough clinical assessment together with the use of specific recommendations. Another way to improve physical capacity is to offer exercise training. For example, in adult male patients with chronic heart failure, specific cardiac training has been shown to improve physical capacity and quality of life, as in healthy subjects. In addition, training in children with various congenital heart diseases has been shown to improve physical abilities. However, in patients with previously operated complex congenital heart disease the value of exercise training should be further studied, focusing on long term effects in adulthood.

Prevention of postoperative complications may also avoid poor outcome of the physical dimension of health related quality of life, but this topic is beyond the scope of the present study.

**Relation to physical status**

Relating physical indices to the dimensions of health related quality of life and subjective health status showed weak correlations. The objective physical index correlated less with health related quality of life and subjective health status than the index of Somerville and the NYHA class. This means that variables such as cyanosis, cardiac failure, or arrhythmia (the best indicators for the objective severity of the disease) do not correlate with the patient's health related quality of life or subjective health status. This is a surprising finding, but other reports on these correlations show the same pattern: factors other than the complexity of the disease (for example, parental attitudes and perceptions, mother's concern, and social support) play a more important part in psychosocial outcome.

Correlations between all the quality of life dimensions and the Somerville index were significantly weaker than those between the quality of life dimensions and the NYHA class. Thus, although the Somerville index was specifically devised to take into account the adjustment and perception of patients with congenital heart disease, its usefulness for this purpose was not confirmed in the present study. The NYHA class correlated significantly and relevantly with the dimensions gross motor functioning (TAAQOL) and physical functioning (SF-36) (Spearman’s \( r = 0.77 \) and \( r = 0.87 \), respectively). When focusing solely on these dimensions, the NYHA class correlations, mother’s concern, and social support) play a more important part in psychosocial outcome.

Figure 4: Subjective health status (SF-36) in 76 patients with previously operated complex congenital heart disease is compared with that of the general population (n = 361).

Horizontal axis: 36 item short form health survey (SF-36) domains.
Vertical axis: score (range 0–100).
\* \( p < 0.01 \); \* \* \( p < 0.001 \).
quality of life and subjective health status, parameters other than those routinely used in a physically oriented cardiologi-
cal setting should be included.

Remarks and study limitations
When measuring long term outcome of patients with previously operated congenital heart disease, mortality has to
be considered. In the unbiased group of 251 selected patients, 123 (49%) had died and could therefore not participate in the
study; at least nine of these latter patients were over 18 years old when they died. In addition, two of the participants died
from cardiac causes during the study period. Mortality in the age group 18–32 years is, therefore, at least 8.1% (that is, 11 of
135 patients died). Although the study found good health related quality of life in many aspects, the considerable risk of
dying after the age of 18 years has to be taken into account. In
addition, studying only surviving patients implies a positive
selection bias.

The overall response rate in this study was 91%. There was no significant difference between the participants and
non-participants in terms of sex or residence. Similarly, there
was no significant difference in response between the
subdiagnosis groups. However, there was a significant differ-
ence between age groups. The response rate in the 24–32
year age group was lower than in the 18–24 year age group
(77% and 95%, respectively; p = 0.02). Communication with
some of the non-participants indicated that older patients
may have more family or work responsibilities and thus less
time to participate in a study; how this may have influenced
our results remains, however, unclear.

Conclusions
Patients with previously operated complex congenital heart
disease experienced limitations only in the physical dimen-
sions of health related quality of life and subjective health sta-
tus. Objective medical variables correlated only weakly with
health related quality of life and can therefore not be used to
assess health related quality of life. Wagner and colleagues4
reported that “the routine use of health status measures may
enhance patients’ care”. The dedicated health related quality of
life instrument used in the present study (TAAQOL) is well
validated23 and valuable in patient care since it measures the
patients’ own health related feelings and problems in daily
life. It provides more relevant information than just measur-
ing functioning with subjective health status.12 14 17 These
results indicate that, when evaluating health related quality of
life, dedicated questionnaires such as the TAAQOL should be
used.

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general population.

Authors’ affiliations
*Department of Paediatric Cardiology, Leiden University Medical Centre, Leiden, The Netherlands
†Department of Paediatric Cardiology, Leiden University Medical Centre, Leiden, The Netherlands
‡Department of Paediatric Cardiology, Leiden University Medical Centre and TNO Prevention and Health, Leiden, The Netherlands

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Interception of aortic regurgitation by vegetation plug in a patient with infective endocarditis

A 75 year old Japanese woman with rheumatic valvar disease (aortic steno-insufficiency and mitral steno-insufficiency) was admitted because of a two month history of fever and general malaise. Eleven years previously she was diagnosed with aortic mixed valve disease and mitral mitral valve disease by echocardiography and cardiac catheterisation, which revealed grade III aortic regurgitation.

On admission the patient’s temperature was 37.1°C and blood pressure was 120/76 mm Hg, with irregular heart beats of 112 per minute because of pre-existing atrial fibrillation. A pansystolic murmur of grade 3/6 was audible at the apex. There was no early diastolic sound, because of pre-existing atrial fibrillation. A pansystolic murmur of grade 3/6 was audible at the apex. There was no early diastolic sound. A pansystolic murmur of grade 3/6 was audible at the apex.

A transeosophageal echocardiogram performed after admission revealed extensive vegetation involving the right and non-coronary cusps of the aortic valve which plugged the aortic orifice during diastole, and abolished pre-existing aortic regurgitation. Blood cultures revealed streptococcal bacteraemia (Streptococcus mitis).

The patient underwent aortic and mitral valve replacement surgery successfully. The extensive vegetations were attached to the right and non-coronary cusps; the fibrous thickening and adhesion commissures and cusps of the aortic valve were pathologically consistent with a rheumatological aetiology. Similar rheumatic findings were confirmed at the mitral valve.

K Miura
Y Seino
H Honma
y.seino@h.t.u.ac.jp

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