Mind your heart: health care, quality of life, and biological pathways in adults with congenital heart disease
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Chapter 7

New York Heart Association class assessment by cardiologists and outpatients with congenital heart disease: A head-to-head comparison of three patient-based versions

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ABSTRACT

Background The objective of this study was to compare three patient-based New York Heart Association (NYHA) class assessments with cardiologist assessments in outpatients with congenital heart disease (CHD).

Methods Consecutive adult outpatients completed three questionnaires in a random order: a patient-based translation of the NYHA classes, a self-constructed questionnaire based on the NYHA classes, and the Specific Activity Scale (SAS). The treating cardiologist assessed the NYHA class on the same day. Patient-cardiologist agreement was assessed by calculating percent agreement and weighted kappa. We also explored the level of agreement for patients without co-morbidity.

Results In all, 86 adults with a median age of 35.8 years – including 46 women participated. An agreement of 75.6% (weighted kappa is 0.43; \( p < 0.01 \)), 70.6% (weighted kappa is 0.44; \( p < 0.01 \)), and 74.4% (weighted kappa is 0.28; \( p < 0.01 \)) was found between the cardiologist assessment and the patient-based translation, self-constructed questionnaire, and SAS respectively. The patient-based translation equally over- and underestimated the NYHA class, whereas the self-constructed questionnaire overestimated and the SAS underestimated the NYHA class. Agreement levels for patients without co-morbidity were higher than agreement levels for the total group.

Conclusion The patient-based translation yielded adequate agreement with cardiologist-assessed NYHA class, showed equal over- and underestimation, and was easy to complete. The patient-based translation with the instruction to only consider functional impairments caused by the CHD, is recommended in future studies of outpatients with CHD.
INTRODUCTION

The mean prevalence of congenital heart disease (CHD) at birth is 7.7 per 1000 live births.\(^1\) CHD is a generic term for malformations of the heart present at birth. Three of the most common malformations are (a) the narrowing of the aorta (aortic coarctation); (b) an opening in the wall dividing the left and right heart chambers (ventricular septum defect); and (c) an opening in the wall dividing the left and right atrium (atrial septum defect). In the Netherlands, approximately 1600 children with CHD are born each year.\(^2\) At least 85% of these patients reach adulthood owing to the successes of cardiac surgery.\(^2\) Even after corrective surgery, however, most patients have residual lesions with varying effects on daily functioning, for example exercise capacity and quality of life (QoL).\(^3,4\)

In daily clinical practice, many treating cardiologists assess the exercise capacity or functional status of patients with CHD following the four New York Heart Association (NYHA) classes (Table 1). These four classes were originally developed to help physicians evaluate the effect of cardiac symptoms on patients’ daily activities, but are also increasingly used to estimate patients’ functional status in clinical trials.\(^5\) The NYHA classification has been found to be clinically useful, as it is associated with survival and QoL.\(^2,6,7\)

In large scale medical research where patients cannot be seen by a physician at each time point, it is advantageous if functional status could be assessed by patients themselves. However, the usefulness of such patient-based NYHA class assessment critically depends on its agreement with the cardiologist-based score. Since the NYHA classification is a physician-based score, the cardiologist can be seen as the gold standard.

Table 1: New York Heart Association classification

<table>
<thead>
<tr>
<th>Class</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I</td>
<td>No limitation of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, or dyspnea (shortness of breath).</td>
</tr>
<tr>
<td>Class II</td>
<td>Slight limitation of physical activity. Comfortable at rest, but ordinary physical activity results in fatigue, palpitation, or dyspnea.</td>
</tr>
<tr>
<td>Class III</td>
<td>Marked limitation of physical activity. Comfortable at rest, but less than ordinary activity causes fatigue, palpitation, or dyspnea.</td>
</tr>
<tr>
<td>Class IV</td>
<td>Unable to carry out any physical activity without discomfort. Symptoms of cardiac insufficiency at rest. If any physical activity is undertaken, discomfort is increased.</td>
</tr>
</tbody>
</table>

To our knowledge, only four studies explicitly compared patient-cardiologist assessed NYHA class.\(^8-11\) These studies included patients with heart failure. In these four studies, only Goode et al\(^9\) made a direct comparison between patient-assessment and physician-assessed NYHA class. The remaining three studies\(^8,10,11\) inferred the NYHA class from patient-reported functional class scales\(^8,10,11\) or a QoL questionnaire\(^11\). In these four studies different levels of agreement were found. Contrary to heart failure patients, CHD-patients are born with impairments. Therefore,
these patients may be more used to their limitations, which might result in a different perspective of their functional status. The objective of this study is to compare three patient-based NYHA class assessments with cardiologist-assessed NYHA class in outpatients with CHD to enable “best choice”.

MATERIALS AND METHODS

Study population and procedure
Consecutive patients, who attended one of four cardiologists of the congenital heart outpatient clinic of the Academic Medical Center from March to June, 2007, were asked to participate in this study. Patients who were not literate in Dutch were excluded.

Patients with various confirmed CHD completed three questionnaires assessing the NYHA class preceding their visit to the cardiologist. The treating cardiologist completed the NYHA class assessment on the same day as the patient-assessments and was blinded to the patients’ responses. Since no ethical approval is required for completion of the self-report questionnaires under Dutch law, the medical ethics committee exempted this study from ethical approval. This study is conducted in full accordance with the principles of the “Declaration of Helsinki” as amended in Tokyo, Venice, and Johannesburg.

Measures

Socio-demographic and clinical data
Sex, birth date, employment status and presence of co-morbidity—such as diabetes, renal diseases, hypertension, chronic obstructive pulmonary diseases, rheumatoid arthritis, chronic allergies, chronic back pain, limitation in the use of arm or leg, or “other illnesses”—were measured through self-report. Primary diagnosis was extracted from the CONCOR database, a nationwide registry for patients with CHD.12

New York Heart Association class assessment

Patient-based translation
We directly translated the four classes into patient-based statements (see Appendix). Class II, for example, was formulated as “I am slightly limited in performing physical activities. I do not experience any symptoms at rest, but ordinary physical activities cause extraordinary fatigue, palpitation or dyspnea”. Patients were asked to choose the statement that was most applicable to him or her. The NYHA classes were directly derived from the answers.

Self-constructed questionnaire
The self-constructed questionnaire was devised with the help of four expert cardiologists and consisted of 11 questions concerning possible physical limitations as a result of the following cardiac symptoms: fatigue, dyspnea and palpitation. We queried the presence of each of the symptoms separately at three different levels of exertion: heavy (exemplified by running, doing
sports, biking with adverse wind, climbing a flight of 20 steps), ordinary (exemplified by climbing a flight of three steps, walking, dressing) and at rest or when performing the slightest exertion (exemplified by standing up, reading a book, talking). For example, “Do you experience palpitations during regular physical activities (walking, climbing three steps, showering, getting (un)dressed)?”. For all questions a “yes” or “no” response option was used. The final question assessed whether discomfort of possible symptoms at rest increased when any physical activity was undertaken.

Specific Activity Scale
The Specific Activity Scale (SAS) consists of five stem questions, of which three have a different number of sub questions (4-8), each addressing a different number of example activities (1-5). The SAS is based on the metabolic expenditure values of activities that a patient reports he or she can or cannot do, and classifies patients into one of the four functional classes. The SAS is available in the original article by Goldman et al. The SAS functional classification system is comparable to the NYHA classification system. The SAS was translated into Dutch. Minor cultural adaptations that left the structure intact and that did not affect the NYHA scoring included the deletion of four examples that are inapplicable to or too specific for Dutch adults (i.e., roller skating, hang washed clothes, bowl, and push power lawn mower); and the transformation of weight (pounds) and speed (miles) into the international system of unit (i.e., kilograms and kilometers).

Pilot, order and debriefing questions
Two pilot studies were conducted to test the appropriateness of the wording of the three questionnaires. Improvements were made to the questionnaires, wherever needed.

The order of the three questionnaires was counterbalanced to avoid order effects. Thus, there were six different sets of questionnaires, which were alternately administered to the participating patients.

After completing the three questionnaires, the patients were asked two additional debriefing questions: “In your opinion, which questionnaire describes your physical functioning best?” and “Which questionnaire did you find easiest to answer?”.

Cardiologist-assessed New York Heart Association class
The standard definition of the NYHA classification (Table 1) was used by the treating cardiologists to assess patients’ NYHA class following regular clinical guidelines.

Statistical analysis
For the patient-based translation of the NYHA class, the scores were mapped directly to a NYHA class. For the self-constructed questionnaire, the NYHA classes were calculated by following an algorithm designed after consulting an expert cardiologist and following clinical guidelines in assessing NYHA class. Patients were categorized as NYHA class I if they answered negatively to all questions, indicating that they were not at all physically limited. Patients who indicated to be physically limited at heavy exertion were rated as NYHA class II. Patients limited at ordinary
exertion were rated as NYHA class III. Patients were categorized in NYHA class IV, if they indicated to experience at least one of the three cardiac symptoms at rest, and the experienced discomfort at rest increased when any physical activity was undertaken. In all, 38 patients (44.2%) completed the self-constructed questionnaire inconsistently. For example, patients rated that they experienced cardiac symptoms at ordinary, but not at heavy exertion. For these 38 patients, NYHA class was blindly assessed by one of the cardiologists (BJMM) by manually rating the answers. For the SAS we followed the original scoring procedure as developed by Goldman et al.13

The association between patient- and cardiologist-assessed NYHA class was calculated by the Spearman rank correlation coefficient and was interpreted as small (if smaller than 0.30), medium (if ranging from 0.30 to 0.50), or large (if bigger than 0.50).14 To assess agreement between patient- and cardiologist-assessed NYHA class, we calculated percent agreement and weighted kappa, which was interpreted as slight (if smaller than 0.20), fair (if ranging from 0.21 to 0.40), moderate (if ranging from 0.41 to 0.60), or substantial (if bigger than 0.61).15,16 Weighted kappa was used, as the inclusion of a weight variable enabled the calculation of kappa in SPSS, despite the unequal range of scores across types of raters (i.e. cardiologists and patients).17 Since co-morbidity is known to affect self-reported health, we also explored the level of agreement for patients without co-morbidity.

RESULTS
Patients
A total of 86 adult outpatients with a congenital malformation of the heart participated. The median age was 35.8 years, and more than half of the patients were women (53.5%). Most patients worked at least part-time (74.4%). Patients were primarily diagnosed with Marfan syndrome (26.7%), aortic coarctation (16.3%), valve malformation (15.1%), or Tetralogy of Fallot (12.8%) (see Table 2). Fifteen patients (17.4%) categorized into “other congenital heart defects”, including 11 different diagnoses, for example, Eisenmenger’s syndrome, Ebstein’s syndrome, and atrium septum defect. A total of 56 patients (65.1%) reported to have no co-morbidity, whereas 23 patients (26.7%) reported one co-morbidity. The most common co-morbidities were hypertension (9.3%), chronic back pain (5.8%), chronic obstructive pulmonary diseases (2.3%), and rheumatoid arthritis (2.3%). The number of co-morbidities were distributed across the NYHA classes as follows: class I included 27.9% (17 patients), class II included 54.5 % (12 patients), and class III included 33.3% of the patients (one patient) who had one or more co-morbidities.

Comparison of patient- and cardiologist-assessed New York Heart Association class
Patient-cardiologist agreement and association for each questionnaire are presented in Table 3. The agreement between the patient-based translation and the cardiologist assessment was 75.6%. The patient-based translation correlated highly (Spearman rank correlation coefficient is 0.54), and agreed moderately (weighted kappa is 0.43) with the cardiologist-assessed NYHA class. In 11 cases, NYHA class assessed by the patient-based translation was overestimated, as patients
reported a higher NYHA class compared with the cardiologist assessment, whereas in ten cases NYHA class was underestimated by patients. When calculating agreement, including only patients without co-morbidity (56 patients), the percentage agreement increased from 75.6% to 82.1%, and weighted kappa from 0.43 to 0.51.

Table 2: Patient characteristics

<table>
<thead>
<tr>
<th>Patient characteristics</th>
<th>Total (86 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (Median; range)</td>
<td>35.8 (19-64)</td>
</tr>
<tr>
<td>Sex (female)</td>
<td>53.5 (46)</td>
</tr>
<tr>
<td>Work</td>
<td></td>
</tr>
<tr>
<td>Full time</td>
<td>40.7 (35)</td>
</tr>
<tr>
<td>Part time</td>
<td>33.7 (29)</td>
</tr>
<tr>
<td>Inability to work</td>
<td>5.8 (5)</td>
</tr>
<tr>
<td>Unemployed</td>
<td>3.5 (3)</td>
</tr>
<tr>
<td>Retired</td>
<td>2.3 (2)</td>
</tr>
<tr>
<td>Other</td>
<td>12.8 (11)</td>
</tr>
<tr>
<td>Diagnosis</td>
<td></td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>26.7 (23)</td>
</tr>
<tr>
<td>Other congenital heart defects</td>
<td>17.4 (15)</td>
</tr>
<tr>
<td>Aorta coarctation</td>
<td>16.3 (14)</td>
</tr>
<tr>
<td>Valve malformation</td>
<td>15.1 (13)</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>12.8 (11)</td>
</tr>
<tr>
<td>Transposition of the Great Arteries</td>
<td>7.0 (6)</td>
</tr>
<tr>
<td>Ventricle Septum Defect</td>
<td>4.7 (4)</td>
</tr>
<tr>
<td>Number of co-morbidities</td>
<td></td>
</tr>
<tr>
<td>No co-morbidity</td>
<td>65.1 (56)</td>
</tr>
<tr>
<td>One co-morbidity</td>
<td>26.7 (23)</td>
</tr>
<tr>
<td>Two co-morbidities</td>
<td>5.8 (5)</td>
</tr>
<tr>
<td>Three or more co-morbidities</td>
<td>2.3 (2)</td>
</tr>
<tr>
<td>Cardiologist-assessed NYHA class</td>
<td></td>
</tr>
<tr>
<td>Class-I</td>
<td>70.9 (61)</td>
</tr>
<tr>
<td>Class-II</td>
<td>25.6 (22)</td>
</tr>
<tr>
<td>Class-III</td>
<td>3.5 (3)</td>
</tr>
<tr>
<td>Class-IV</td>
<td>0.0 (0)</td>
</tr>
</tbody>
</table>

Note: Data is presented in percentage (numbers). NYHA = New York Heart Association. Other congenital heart defects = including for example Eisenmenger’s syndrome, Ebstein’s syndrome, and atrium septum defect.

The agreement between the self-constructed questionnaire and cardiologist assessment was 70.6%, with a high correlation (Spearman rank correlation coefficient is 0.59) and moderate agreement.
(weighted kappa is 0.44). The self-constructed questionnaire led primarily to overestimation of patient-assessed NYHA class (22 overestimations versus three underestimations). A similar increase in agreement levels was seen when agreement was calculated for only those patients without co-morbidity (from 70.6% to 78.2% and weighted kappa from 0.44 to 0.53).

The SAS agreed in 74.4% of the cases with the cardiologist assessment. There was a moderate correlation (Spearman rank correlation coefficient is 0.40) and a fair agreement (weighted kappa is 0.28). The SAS led to underestimation in 18 cases, and in only four cases to overestimation compared with the cardiologist-assessed NYHA class. Again, agreement levels between the SAS and the cardiologist were calculated for patients without co-morbidity, showing a small increase in agreement percentages (from 74.4% to 78.6%) and a decrease in weighted kappa (0.28 to 0.18).

As shown in Table 3, in two occurrences there was maximal discrepancy between the patient and cardiologist, that is, a patient rated himself/herself in class IV, whereas the physician rated the patient in class I. Inspection of the data identified that the same patient was involved in both occurrences. Additional analyses showed that this patient reported to have one co-morbidity, that is, “other illness”.

Table 3: Patient-cardiologist agreement and association per questionnaire

<table>
<thead>
<tr>
<th>Cardiologist-assessed NYHA class</th>
<th>Patient-based translation</th>
<th>Self-constructed questionnaire</th>
<th>Specific Activity Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>I</td>
<td>II</td>
<td>III</td>
</tr>
<tr>
<td>Cardiologist-assessed NYHA class</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Agreement</td>
<td>75.6%</td>
<td>Spearman rank correlation coefficient is 0.54</td>
<td>Weighted kappa is 0.43</td>
</tr>
<tr>
<td>(86 patients)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(85 patients)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Note: Data is presented as frequencies, unless stated otherwise.</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Debriefing questions
Eighteen (20.9%) and eight (9.3%) patients did not answer the two debriefing questions, respectively. Four (4.7%) and one (1.2%) patient(s), chose all three questionnaires. The two debriefing questions were thus completed following the instruction by 89.5% and 74.4% of the patients, respectively. The distribution of their preference is given in Table 4. Patients reported the SAS as the questionnaire best describing their functional status followed by the self-constructed questionnaire, and patient-based translation, respectively. Both the patient-based translation and SAS were reported as easiest to complete, followed by the self-constructed questionnaire.

Table 4: Answers to the debriefing questions for each questionnaire

<table>
<thead>
<tr>
<th></th>
<th>Best describing (77 patients)</th>
<th>Easiest to complete (64 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient-based translation</td>
<td>23.4 (18)</td>
<td>40.6 (26)</td>
</tr>
<tr>
<td>Self-constructed questionnaire</td>
<td>26.0 (20)</td>
<td>18.8 (12)</td>
</tr>
<tr>
<td>Specific Activity Scale</td>
<td>50.6 (39)</td>
<td>40.6 (26)</td>
</tr>
</tbody>
</table>

Note: Data is presented in percentage (numbers).

DISCUSSION
This study was conducted to explore which patient-based NYHA class assessment agrees best with cardiologist-assessed NYHA class and can be used in future research contexts. The patient-based translation was found to be the best choice in assessing the NYHA class in CHD-patients given its adequate agreement, its equal over- and underestimation, and its ease of completion. The patient-based translation can be used for research purposes; however, its 75.6% agreement with the cardiologist precludes its use in the individual case. The self-constructed questionnaire also showed adequate agreement, but led primarily to overestimation of the NYHA class, whereas the SAS showed only fair agreement, and led primarily to underestimation of the NYHA class. Interestingly, for all three questionnaires agreement levels for patients without co-morbidity were higher than agreement levels for the total group.

In general, the agreement levels found in our study are higher than the agreement levels found in the four previous studies. Goode et al,9 used a direct comparison that is comparable to the patient-based translation and found an agreement of kappa is 0.28. Similar to our study, there was equal over- as underestimation of patient-assessed NYHA class. In the study by Goode et al patients were referred to the cardiologist for the first time, whereas in our study the NYHA class was assessed by the regularly treating cardiologist. Perhaps the latter cardiologists have more clinical data – for example, electrocardiography or echography – about the patient to base their rating on. Moreover, they may be better informed about possible co-morbidity and therefore might be more accurate. In addition, patients may also be better aligned to the physician owing
to experience ("training effect"). These factors may have resulted in a higher level of agreement between patient- and cardiologist-assessed NYHA class in our study.

Subramanian et al assessed patient-based NYHA class by means of the Kansas City Cardiomyopathy Questionnaire. A general nurse or a project coordinator assessed the NYHA class on average 20 days after the patient-assessment. An agreement level of 43%, with a weighted kappa of 0.28, was found. Factors likely to have contributed to this greater discrepancy between patient-based and cardiologist-assessed NYHA class include a lower health literacy of the patients, assessment by a nurse or project coordinator instead of a cardiologist, and a time lapse of 20 days between assessments.

In the study by Kubo et al, patients with heart failure were interviewed by a physician assistant or nurse, who recorded their answers on a questionnaire. These answers were then categorized into the NYHA classes by one of three independent raters. The NYHA class assessed by the cardiologist was subsequently compared with the NYHA class scored by each of these independent raters. Results showed agreement levels ranging from 57% to 65%, with weighted kappa scores ranging from 0.55 to 0.63. Contrary to our study, rater-based patient assessment underestimated the NYHA class compared with cardiologist-assessed NYHA class. The agreement levels found by Kubo et al were lower, whereas, slightly higher kappa levels were reported compared to those found in our study. The raters of the patient-based assessments are health care professionals, they are likely more closely aligned to the cardiologists.

Ekman et al found an agreement of 32% between the SAS and cardiologist-assessed NYHA class. Kappa was not calculated. Similar to our study, the SAS scores primarily underestimated the NYHA class compared with cardiologist assessment. Similar to the study conducted by Goode et al, patients were referred to an outpatient heart failure clinic for the first time. The discrepancy between the SAS and cardiologist-assessed NYHA class might be explained by similar reasons formulated to explain the results of Goode et al.

On a general note, CHD-patients are born with their impairments, and as a consequence have visited a cardiologist their entire life, contrary to heart failure patients. This might result in better alignment with their cardiologist in the interpretation of their symptoms and the assessment of the NYHA class. Moreover, both the patient and the cardiologist assessed the NYHA class on the same day, possibly further increasing agreement levels. In addition, in all four studies the presence of co-morbidity was not assessed and its influence on the level of agreement was therefore not explored. Co-morbidity is relevant for heart failure patients as they are generally in the higher age ranges.

The finding that co-morbidity affects patient-cardiologist agreement in assessing the NYHA class deserves further attention, especially since co-morbidity is common in CHD-patients. It might be hypothesized that the cardiologists filter out the impact of co-morbidity in assessing the NYHA class, whereas patients do not. The maximal discrepancy in patient and cardiologist assessment found in this study, might also be explained by the ability of the cardiologist to discriminate between the CHD and co-morbidities, since the patient involved reported to have one co-morbidity. On the
basis of the results of this study we added an instruction to the patient-based translation (see Appendix) in which we ask patients to only consider functional impairments caused by their CHD.

The results of this study raise the following question: who should be the gold standard in assessing the NYHA class, the cardiologist or the patient? Since the NYHA class is based on the patient’s subjective perceptions of disease-related restrictions in physical activity, and the patient is by definition the expert on these subjective perceptions, the patient can be considered the gold standard. In contrast, the ability of the cardiologists to filter out the impact of co-morbidity when assessing the NYHA class pleads for the cardiologist as the gold standard. One can also choose an empirical approach to this question. Future studies should examine whose subjective assessment (i.e. cardiologist or patient) is most closely aligned with an objective measure of patients’ functional status such as an exercise test (e.g. the six minute walking test). The most closely related measure can be considered the gold standard.

The limitations of this study merit attention. First, the sample size was too small to explore patient characteristics affecting the patient-cardiologist agreement. For example, it would have been interesting to compare the patients who underestimated versus overestimated their NYHA class with regard to a number of background characteristics. However, the sample size was sufficiently large to explore whether co-morbidity affects patient-cardiologist agreement levels, by examining the group of patients without co-morbidity separately. Second, the focus of this study was on outpatients. As a consequence, the distribution of NYHA classes was skewed, with most patients being categorized in physician-based NYHA class I and II and none in class IV. This may have resulted in lower Kappa levels. More importantly, the results are only generalizable to patients with NYHA classes I and II. Despite the fact that it may not be too far-fetched to expect that the simple, direct translation of the NYHA classes would also work for the two higher classes, this needs to be confirmed in future studies with patients who have poorer function, such as hospitalized patients. Third, it is important to note that the distribution of CHD was not representative for adults with CHD, since patients with the Marfan syndrome constituted the largest group. Our patient sample was therefore not representative of the population of adults with CHD. Fourth, we were unable to describe the patient sample with respect to a number of clinical characteristics, such as cardiac functioning and type of treatment. However, we did present data on type of CHD, physician-based functional class and co-morbidity, allowing for characterization of the patient sample in clinical terms.

We would also like to highlight the strengths of this study. It is the first study that addresses outpatients with CHD, compares three patient-based questionnaires in a counterbalanced way, and explores whether co-morbidity affects patient-cardiologist agreement in NYHA class assessment. A final strength of this paper is that patients and their treating cardiologists completed the NYHA assessment on the same day. This study shows that the simple and direct translation of the NYHA class, as provided in the Appendix, is a valuable patient-based tool that can be used in future studies of outpatients with CHD.

In summary, the patient-based translation with the instruction to only consider functional impairments caused by the CHD, is recommended in future studies of outpatients with CHD,
given its adequate agreement with cardiologist-assessed NYHA class, its equally over- and underestimation, and its ease of completion.

ACKNOWLEDGEMENTS

We are very grateful to all participants. Moreover, we thank all participating congenital cardiologists from the Academic Medical Centre in Amsterdam. This study is partly financed by the Interuniversity Cardiology Institute of the Netherlands.
REFERENCES


**APPENDIX**

*Patient-based translation*

Below you find four descriptions that describe different degrees in which individuals are limited in their physical functioning. Which description is most applicable to you? Please only consider the limitations that you believe are caused by your congenital heart defect.†

- □ A  I am not limited during physical activities. Ordinary physical activities do not cause extraordinary fatigue, palpitations or shortness of breath.
- □ B  I am slightly limited during physical activities. I do not experience any symptoms at rest, but ordinary physical activities cause extraordinary fatigue, palpitations or shortness of breath.
- □ C  I am considerably limited during physical activities. I do not experience any symptoms at rest, but less than ordinary physical activities cause extraordinary fatigue, palpitations or shortness of breath.
- □ D  I am unable to be physically active without experiencing discomfort. I experience one or more of the following complaints at rest; fatigue, palpitations or shortness of breath. When I am physically active, the discomfort increases.

Note: † = this instruction was adapted on the basis of this study.