Mismatch between self-estimated and objectively assessed exercise capacity in patients with congenital heart disease varies in regard to complexity of cardiac defects

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Abstract

Aim: Regular evaluation of physical capacity takes a crucial part in long-term follow-up in patients with congenital heart disease (CHD). This study aims to examine the accuracy of self-estimated exercise capacity compared to objective assessments by cardiopulmonary exercise testing in patients with CHD of various complexity. *Methods:* We conducted a single centre, cross-sectional study with retrospective analysis on 382 patients aged 8–68 years with various CHD who completed cardiopulmonary exercise tests. Peak oxygen uptake was measured. Additionally, questionnaires covering self-estimation of exercise capacity were completed. Peak oxygen uptake was compared to patient's self-estimated exercise capacity with focus on differences between complex and non-complex defects. *Results:* Peak oxygen uptake was 25.5 ± 7.9 ml/minute/kg, corresponding to $75.1 \pm 18.8\%$ of age- and sex-specific reference values. Higher values of peak oxygen uptake were seen in patients with higher subjective rating of exercise capacity. However, oxygen uptake in patients rating their exercise capacity as good (mean oxygen uptake $78.5 \pm 1.6\%$) or very good (mean oxygen uptake $84.8 \pm 4.8\%$) was on average still reduced compared to normal. In patients with non-complex cardiac defects, we saw a significant correlation between peak oxygen uptake and self-estimated exercise capacity (spearman-rho -0.30, $p < 0.001$), whereas in patients with complex cardiac defects, no correlation was found (spearman-rho -0.11, $p < 0.255$). *Conclusion:* The mismatch between self-estimated and objectively assessed exercise capacity is most prominent in patients with complex CHD.
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According to a large epidemiological study based on data from the national network for congenital heart disease (CHD), the incidence of CHD in Germany nowadays amounts to 1%.[1] Due to progress made in the fields of early diagnostics, surgical techniques, and follow-up, the percentage of patients reaching adulthood has constantly risen over the last few decades and is now over 90%.[2–4] With higher life expectancy, new aims develop that go far beyond reaching adulthood and move focus on lifestyle and quality of life. One key aspect contributing to the improvement of quality of life in these patients is their physical well-being and fitness.[5,6]

Multiple studies have shown a reduced cardiopulmonary exercise capacity amongst patients with CHD.[3,7–11] This phenomenon can in part be traced back to consequences of the cardiac defect itself or its surgical or medical therapy, for example valve stenosis or insufficiencies,[3] stenosis of vessels as in coarctation of the aorta or Tetralogy of Fallot,[12] multiple thoracotomies,[13] or chronotropic incompetence under medical beta-blockade[14] or following Fontan procedure.[15] However, a lack of physical activity amongst these patients has also been identified as part of the reason for their reduced cardiopulmonary exercise capacity.[12,16–18]

In the past, the approach towards physical activity of patients with CHD amongst paediatric cardiologists and parents has been rather restrictive.[19,20] With improved life expectancy and a better understanding of the benefits of physical activity in these patients, a shift towards the promotion of physical fitness can be seen in current guidelines.[12,21]

In order to ensure a safe implementation of physical activity into patient's lifestyle, regular evaluation of their current physical capacity is essential. Various studies emphasise the importance of objective evaluation methods, such as cardiopulmonary exercise testing using a treadmill or bicycle protocol, which provide a non-invasive method to unveil risk factors and affect the timing of therapeutic consequences.[3,21]

The necessity of objective testing methods also results from possible misinterpretations that could generate from assessing fitness solely through patient's report. Some studies have shown

a correlation between self-estimated and objectively assessed exercise capacity, however, with a tendency of subjective estimations outmatching objective results.[5,22–24]

However, it is unclear if the problem of overestimation of physical fitness observed in existing literature concerns the population of patients with CHD as a whole, as the accuracy of self-estimated physical fitness has, to our knowledge, rarely been examined for subgroups of patients with CHD.

This study aims to examine the accuracy of self-estimated exercise capacity compared to objective assessments by cardiopulmonary exercise testing in patients with CHD. Furthermore, a possible impact of severity of cardiac defects as well of patient's gender on the accuracy of self-estimated exercise capacity will be evaluated.

Materials and methods

Study design and study population

We conducted a single centre, cross-sectional study with retrospective analysis on children and adults with CHD who underwent cardiopulmonary exercise testing at the German Heart Institute Berlin between March, 2014 and November, 2015. Data of 604 consecutive cardiopulmonary exercise tests were collected as part of our patient's regular checkup, out of which 382 patients aged from 8 to 68 years, with CHD, given consent, completed questionnaire prior to testing and reaching formal criteria of exhaustion during exercise testing were included in this study.

Patients were grouped according to their diagnosis and complexity of their cardiac defect, as explicitly stated under results, along with the demographic data of each group.

For all patients enrolled in this study informed consent was obtained. The study was reviewed and approved by the Charité Universitätsmedizin Berlin Ethics Committee (entry number EA2/106/14).

Exercise testing protocol

We used a continuous ramp bicycle protocol adapted to the standards of the American Heart Association.[21] The test started with 2 minutes of rest followed by 2 minutes of minimally loaded pedaling (<6 watts), after which the incremental phase of exercise, adapted to an individual load maximum estimated for each patient, lasted over 8–12 minutes until reaching formal criteria of exhaustion. Finally, the test ended after another 3 minutes of unloaded cycling. Objective criteria of reaching peak oxygen uptake at full exhaustion used were: maximal heart rate of at least 85% of expected values (220/minute – age), or heart rate of at least 180/minute regardless of age, and respiratory exchange ratio (calculated as ratio of carbon dioxide exhaled to oxygen uptake per unite time) of at least 1.0.[25–27]

We calculated peak oxygen uptake in relation to body weight as well as percentage of expected values using age and gender specific formulas. For adult patients, formulas by Cooper and Storer were used.[28] For patients aged 17 and younger, expected values were calculated using formulas by Cooper and Weiler-Ravell.[29] Our study population was then grouped according to percentage of expected peak oxygen uptake, labelling exercise capacity of oxygen uptake less than 50% of expected peak values as strongly reduced, between 50 and 65% as reduced, between 65 and 85% as slightly reduced, and above 85% as normal, in adaption to existing guidelines.[21]

Questionnaire

Our questionnaire was based on epidemiologic studies conducted by Robert Koch Institute on health status of German general public.[30,31] Two versions of the questionnaire were provided, one for children up to 11 years and one for older patients, differing only in the wording, either addressing the parents or the patients themselves. One item specifically developed for this study asked "How would you judge your physical exercise capacity?", with possible answers very good, good, moderate, bad, and very bad. The questionnaires were filled out prior to exercise testing on the same day.

Statistics

Statistics were calculated on SPSS Version 22.0 (SPSS Inc., Chicago, IL, USA) using non-parametric testing. In all analysis at least one variable was not in normal distribution, therefore non-parametric tests were used. Distribution was analysed using histograms and Kolmogorov-Smirnov tests. Differences between two groups were verified by Mann–Whitney U-test. For more than two groups the Kuskal–Wallis test was applied.

Correlations were calculated as partial rank-correlations. Since this procedure is not provided by SPSS 22.0 by default, a workaround provided by IBM was used.[32]

All tests were calculated two-sidedly. Results were viewed as statistically significant with p-values lower than 0.05. This level of significance was determined in advance.

Results

Patient's demographics and categorisation of cardiac defects

382 patients with an age median of 27 years (range 8–68 years) were in included in our analysis, out of which 196 (51.3%) were male, 186 (48.7%) female.

Patients were grouped according to their cardiac defect. Each group was then classified as either complex or non-complex cardiac defects, see Table 1. The category of complex heart defects combined patients with morphological right systemic ventricle, univentricular heart, Ebstein anomaly as well as patients with uncorrected cyanotic heart defects, who could not be allocated to any other group. In the category of non-complex heart defects all patients have a biventricular anatomy with a morphologically left systemic ventricle. This partly resembled the classification system used in the PAN study of the German Competence Network for Congenital Heart Defects.[1]

Cardiopulmonary exercise capacity

Cardiopulmonary exercise capacity was determined by peak oxygen uptake in relation to bodyweight. Taking into account the whole group of 382 patients, peak oxygen uptake was 25.5 ± 7.9 ml/minute/kg, which corresponds to $75.1 \pm 18.8\%$ of age- and sex-specific reference values (data is provided as mean and standard deviation). Lowest values were found in patients with Fontan circulation, with an average peak oxygen uptake of $60.0 \pm 2.7\%$ of reference values, see Table 2. In the combined group of complex defects, peak oxygen uptake was $67.4 \pm 1.7\%$ and significantly lower compared to the group of non-complex defects with peak oxygen uptake of $78.1 \pm 1.1\%$ (U-test$_{(n1 = 107; \ n2 = 275)}$: $z = -5.089$; $p < 0.001$).

Table 1. Classification of congenital heart defects (CHDs).

Diagnosis	Description	Count (male/ female)	Age (median)
Complex defects			
Fontan circulation	Including different forms of univentricular heart syndrome after Fontan operation	30 (17/13)	21
TGA palliated by atrial switch operation and ccTGA	Transposition of the great arteries: palliated by switch operation of the atria by Senning or Mustard procedure as well as congenitally corrected TGA	43 (31/12)	34
Ebstein anomaly	Included patients with and without replacement or reconstructive surgery of the tricupid valve	26 (10/16)	33
Complex cyanotic	uncorrected cyanotic CHDs, not included in other groups	8 (6/2)	34.5
All complex defects		107 (64/43)	31
Non-complex defects			
AS	Aortic stenosis: patients with bicuspid valve were only included if a intervention took place before the age of 18	41 (23/18)	30
PS	Pulmonary valve stenosis: Included isolated PS as well as repaired pulmonary atresia without VSD	18 (9/9)	23.5
CoA	Coarctation of the aorta	28 (23/5)	32
ToF	Patients with ToF as well as patients with pulmonary atresia and VSD, both after surgical repair, partially by pulmonary valve repair or replacement as well as VSD closure	87 (43/44)	25
TGA repaired by ASO	Transposition of the great arteries repaired by arterial switch operation (ASO)	15 (12/3)	19
VSD	Ventricular septal defect	22 (7/15)	30
ASD	Atrial septal defect	8 (0/8)	32.5
AVSD	Atrioventricular septal defect (complete and incomplete) with biventricular anatomy	10 (3/7)	26
Other valvular defects	Isolated defects of the mitral or tricuspid valve	8 (3/5)	44.5
Other non-complex defects	Partial anomalous pulmonary venous connection, anomalous left coronary artery from the pulmonary artery (ALCAPA)	38 (9/29)	21
All non-complex defects		275 (132/ 143)	26

AS = aortic stenosis, ASO = arterial switch operation, AVSD = atrioventricular septal defect, ccTGA = congenitally corrected TGA, CoA = coarctation of the aorta, PS = pulmonary stenosis, TGA = transposition of great arteries, ToF = Tetralogy of Fallot, VSD = ventricular septal defect

Table 2. Peak oxygen uptake (VO2peak) in ml/minute/kg and as percentage of calculated reference values.

Diagnosis	Count	VO2peak* [ml/minute/kg]	VO2peak* [%]
Complex defects	107	22.7 ± 0.6	67.4 ± 1.7
Fontan	30	24.1 ± 1.2	60.0 ± 2.7
TGA (cc/atrial switch)	43	22.8 ± 1.0	69.9 ± 2.5
Ebstein	26	21.9 ± 1.3	75.0 ± 3.8
Non-complex defects	275	26.6 ± 0.5	78.1 ± 1.1
AS	41	28.5 ± 1.2	83.3 ± 2.9
PS	18	28.1 ± 2.0	75.8 ± 4.1
CoA	28	28.6 ± 1.4	83.6 ± 3.6
ToF	87	25.8 ± 0.9	73.9 ± 1.8
TGA (ASO)	15	29.3 ± 1.2	73.3 ± 3.3
VSD	22	23.7 ± 1.9	75.4 ± 4.4

AS = aortic stenosis, ASO = arterial switch operation, ccTGA = congenitally corrected TGA, CoA = coarctation of the aorta, PS = pulmonary stenosis, TGA = transposition of great arteries, ToF = Tetralogy of Fallot, VSD = ventricular septal defect
*Data is provided as mean and standard-deviation

Patient's self-estimated exercise capacity

In total, 382 patients rated their subjective exercise capacity according to our questionnaire, assigning it to one of five levels as stated in Table 3. In total, 31% of patients with complex cardiac defects and 43% of patients with non-complex cardiac defects rated their exercise capacity as good or very good. About 40% found their exercise capacity to be moderate, as well in patients with complex as in non-complex defects. Overall, 29% of patients with complex and 19% of patients with non-complex defects rated their exercise capacity as bad or very bad.

Comparison of self-estimated and objectively assessed exercise capacity

There were significant differences in peak oxygen uptake in percent of estimated values between the groups (Kuskal–Wallis test: $\chi^2(n = 382) = 31.425$; df = 4; $p < 0.001$). Higher values were seen in patients with higher subjective rating of exercise capacity. However, oxygen uptake in patients rating their exercise capacity as good (mean VO2peak 78.5 ± 1.6%) or very good (mean VO2peak 84.8 ± 4.8%) was on average still reduced compared to normal.

Table 3. Count, sex, age, peak oxygen uptake (VO2peak, in ml/minute/kg as well as percentage of calculated reference values), divided by subjective evaluation of own exercise capacity; p calculated using Kuskal–Wallis test for metric variables.

Subjective evaluation	Total	Very good	Good	Moderate	Bad	Very bad	p-value
Count (%) (male/female)	382 (196/186)	29 (7.6) (17/12)	123 (32.2) (73/50)	148 (38.7) (74/74)	72 (18.9) (29/43)	10 (2.6) (3/7)	
Complex (%)	107 (100)	7 (6.5)	26 (24.3)	43 (40.2)	29 (27.1)	2 (1.9)	
Non-complex (%)	275 (100)	22 (8.0)	97 (35.3)	105 (38.2)	43 (15.6)	8 (2.9)	
Age [years]*	29.4 ± 13.3	24.2 ± 2.4	25.7 ± 1.1	32.0 ± 1.1	31.5 ± 1.6	38.7 ± 5.9	<0.001
VO2peak [ml/minute/kg]*	25.5 ± 7.9	32.1 ± 2.1	28.4 ± 0.7	24.3 ± 0.6	20.8 ± 0.6	20.7 ± 2.5	<0.001
VO2peak [%predicted]*	75.1 ± 18.8	84.8 ± 4.8	78.5 ± 1.6	75.4 ± 1.5	66.1 ± 2.1	67.4 ± 4.4	<0.001

*Data is provided as mean and standard-deviation

For further analysis, patients were divided according to their diagnosis into the group of complex cardiac defects and non-complex cardiac defects. Figure 1 provides a comparison between complex and non-complex cardiac defects divided by their subjective rating of exercise capacity, with regard to their peak oxygen uptake. It shows a tendency of higher values of peak oxygen uptake with higher subjective ratings of exercise capacity, also reflecting a difference between complex and non-complex cardiac defects.

Further, we checked for correlations between peak oxygen uptake and self-estimated physical fitness. In patients with non-complex cardiac defects, we saw a significant correlation (partial spearman-rho controlled for age and sex: −0.368, p < 0.001). In patients with complex cardiac defects, no correlation was found (partial spearman-rho controlled for age and sex: −0.183, p = 0.062).

In a similar type of analysis, we abolished the separation into complex and non-complex defects and split our group of patients into males and females instead. For females as well as for males, we saw significant correlations between peak oxygen uptake and self-estimated physical fitness (partial spearman-rho, controlled for age and complexity of cardiac defects, female: −0.315, p < 0.001; male: −0.318, p < 0.001).

Discussion

Cardiopulmonary exercise capacity in CHD

The majority of patients with CHD today reach adulthood, a development mainly achieved by improvements in congenital heart surgery and long-term follow-up in paediatric cardiology.[2–4] With higher life expectancy, focus shifts towards patients lifestyle and quality of life, with patient's physical fitness taking a crucial role.[5,6] Cardiopulmonary exercise testing plays an important part in routine checkups in patients with CHD.[3,21] Peak oxygen uptake is regarded as the best indicator of cardiopulmonary function in children and adults.[27]

In our group of 382 patients, cardiopulmonary exercise capacity represented by peak oxygen uptake was reduced at 25.5 ± 7.9 ml/minute/kg correspondent to 75.1 ± 18.8% of age and sex-specific reference values. Reduced peak oxygen uptake in patients with CHD has previously been described by various studies.[3,7–11]

We could see that in patients with complex cardiac defects, peak oxygen uptake as percentage of reference values was significantly lower than in the group of non-complex defects. There were also differences between the groups separated by diagnosis, in which peak oxygen uptake showed a similar distribution as in the study

by Kempny et al, with lowest values seen in patients with Fontan circulation.[3]

Self-estimated exercise capacity

Comparable to existing literature, exercise capacity determined by peak oxygen uptake is reduced in our group of patients with CHD. Our next step was to examine if this reduction of objectively determined exercise capacity could also reliably be estimated by patient's subjective impression of their own physical fitness.

After division of our study population into groups in regard to their subjective rating, we observed significant differences in peak oxygen uptake between the different groups. Peak oxygen uptake tended to be higher on average in groups with higher subjective rating, as we would also expect in a normal population. This tendency was also seen in regard to peak oxygen uptake as percentage of age- and sex-specific reference values and can therefore not solely be traced back to a higher age level in groups of lower self-rated exercise capacity. Interpreting the association between subjective and objective evaluation of exercise capacity, it should be noted that peak oxygen uptake was reduced, even amongst patients perceiving their exercise capacity as good or very good. These findings correspond to results from other studies that showed a tendency towards overestimation of own exercise capacity in patients with CHD.[5,22–24] A study by Chen et al showed higher self-perceived physical condition in patients with CHD, even when compared to a healthy control group.[23]

This might have a negative impact on patient's willingness to exercise, as patients being accustomed to their reduced exercise capacity might not see any call for action.

Accuracy of self-estimated exercise capacity in regard to complexity of CHD

Most of the studies that show a tendency of overestimation of physical fitness refer to the population of patients with CHD as a whole. The main aim of this study was to examine a possible impact of severity of cardiac defects as well of patient's gender on the accuracy of self-estimated exercise capacity.

While we could see a significant correlation between self-estimated and objectively assessed exercise capacity in patients with non-complex cardiac defects, no correlation was found in patients with complex cardiac defects. No difference was found in female versus male patients.

One possible explanation for the observed overestimation could be that limitations and linked precautions have often followed through the patient's whole life. Therefore, physical capacity meets
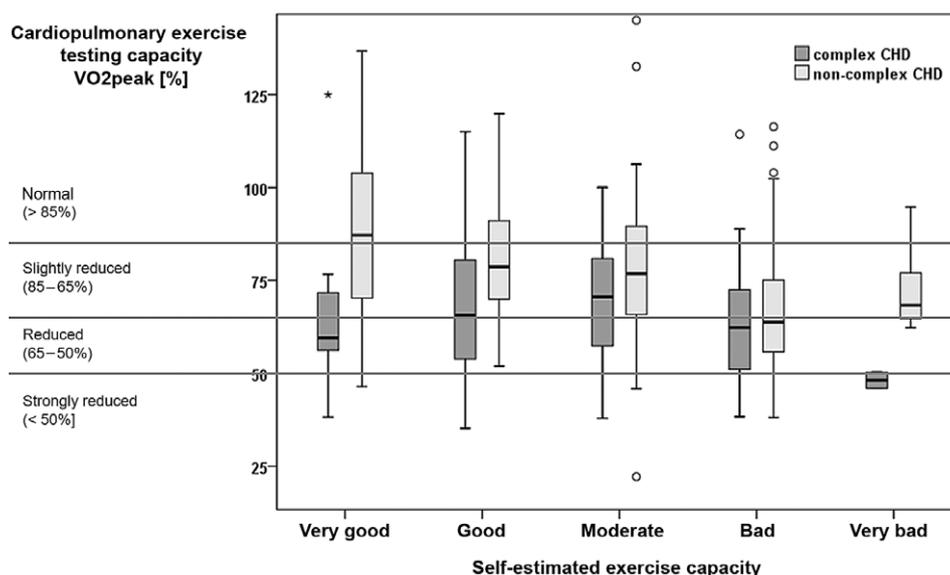
Figure 1. Peak oxygen uptake as percentage of calculated reference values and subjective rating of exercise capacity, divided by complex versus non-complex cardiac defects; data is provided as median and quartile (25–75) distribution.

the patient's own standards and is not perceived as reduced.[5] As reduction of exercise capacity is highest in patients with complex cardiac defects,[3] these patients might also be especially accustomed to their circumstances, including to their level of physical fitness. This would suggest that due to a high level of adaption, the mismatch between self-estimated and objectively assessed exercise capacity is most prominent in patients with complex CHD.

Another explanation might result from the assumption that a sedentary lifestyle seen in patients with CHD would be more dominant in patients with complex defects. Based on this assumption, it could be argued that patients with mild defects tend to be more physically active and might therefore be more familiar with their physical abilities and limitations, while patients with complex defects might have less experience in being physically active to judge from. However, this assumed difference of physical activity in regard to the complexity of underlying cardiac defects could not be found in existing literature.[33,34]

Another aspect contributing to the mismatch between self-estimated and objectively assessed exercise capacity might become apparent from a psychological perspective. Patients with CHD show high prevalence of depressive symptoms and lower self-esteem, both affected by disease severity.[35,36] This might also partly account for the misperception of physical abilities amongst these patients, since depressive symptoms have been associated with an altered self-concept.[36] Another interesting finding in this context is that sport participation was found to be independently associated with reduced depressive symptoms in patients with CHD.[37] The study by Ko et al concluded that sport programs uniquely designed for patients with CHD may improve depression status in these patients.[37] These findings suggest that the high prevalence of depressive symptoms in patients with CHD might be connected to the inaccurate self-estimation of exercise capacity in these patients. Thus, specialised sport programs could improve both self-perception in terms of physical abilities as well as depressive symptoms. Moreover, such programs could provide a possibility to get motivated and accustomed to physical activity in a safe and controlled environment. Another benefit is feedback

experiencing the body's response to training as well as from comparison with peers that, with a weekly training interval, is way more frequent than cardiopulmonary exercise testing performed in routine checkups usually every 6 to 12 months. A training program specialised in patients with CHD has already been introduced at German Heart Centres in Berlin and other locations.[38,39]

Considering an increasing prevalence of severe CHD as seen in a large demographic study by Pfitzer et al,[40] capacities for cardiopulmonary exercise testing might have to be adapted in the future.

Conclusions

The mismatch between self-estimated and objectively assessed exercise capacity is most prominent in patients with complex CHD.

Subjective self-evaluation can contribute to but not replace objective assessments, as many patients tend to overestimate their own exercise capacity. It can still provide valuable information, as declines of perceived exercise capacity can be noticed during their everyday lives, directly and independently of checkup intervals and can speed up necessary diagnostics.

Our findings support the key role of cardiopulmonary exercise testing in evaluating physical fitness, especially in patients with complex cardiac defects.

Limitations

One difficulty resulted from our inhomogeneous study population, especially present in evaluating exercise capacity. In order to make peak oxygen consumption comparable between patients of different characteristics, it is commonly not only stated in millilitre per minute and kilogram, but also as a percentage of age- and sex-specific reference values. The formulas available for calculating these reference values stem from relatively small studies performed on healthy subjects[28,29,41,42] Most studies included only very few subjects in middle childhood or approaching retirement age, if any. Therefore, anthropometric margins were not represented and could only be extrapolated from existing data.[43] Due to various

age groups, reference values had to be calculated using multiple formulas, which might strictly speaking still not fully account for the wide range of age (8–68 years) in this study. Reference values derived from one large study including all age groups and both sexes are needed. A study of this kind was performed by Dubowy et al, however, only for treadmill testing, not covering tests performed on also commonly used bicycle ergometers.[43]

Further research on the subject, including the impact of sports intervention programs on patient's subjective experience of physical fitness, is necessary.

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