Original Article

Quality of life in adults with congenital heart disease with and without pulmonary hypertension: a comparative study

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Background: Pulmonary vascular disease and pulmonary hypertension (PH) belong to the most relevant complications of congenital heart disease (CHD) in the long-term course. Although PH might lead to a severely impaired quality of life (QOL), there are no current studies comparing QOL in adults with CHD (ACHD) with and without PH. Therefore, this study aimed to systematically examine QOL in ACHD with and without PH in order to generate a more differentiated understanding of their overall health-status and to employ newly gained findings into general care of this particular patient population.

Methods: In this comparative cross-sectional study, a representative sample of 803 adults with various forms of CHD with or without PH was analyzed. Data collection took place between September 2017 until February 2018 in a tertiary care center for ACHD. Medical data was retrieved from medical records. QOL was assessed using the EQ-5D-5L questionnaire. Descriptive methods, chi-square and t-tests were used to compare QOL of patients with and without PH.

Results: Of 803 patients, 752 patients had no PH [93.6%; 47.3% female; mean age: 34.9±11.83 (range, 18–86) years], 51 were identified with manifest PH [6.4%; 55.8% female; mean age: 41.9±12.17 (range, 21–69) years]. PH patients showed significantly worse overall QOL [mean no PH: 86.78±13.30 (8.2–100) *vs.* mean PH: 81.79±12.77 (43.6–100); 95% confidence interval (CI): 1.43 to 9.46; P=0.013], worse crosswalk index [mean no PH: 93.29±12.86 (–11, 100) *vs.* mean PH: 89.73±11.77 (47–100); 95% CI: 0.08 to 7.56; P=0.043], and worse VAS [mean no PH: 80.12±16.50 (15–100) *vs.* mean PH: 72.56±16.80 (40–100); 95% CI: 3.29 to 12.17; P=0.004]. However, after adjusting for age, only the VAS stayed significant (P=0.039). The QOL within the dimensions mobility (P<0.001), self-care (P=0.002), and usual activities (P=0.007), were significantly decreased in PH patients. Overall, anxiety and depression (11.08%) as well as pain and discomfort (11.03%) were the most impaired dimensions in both patient groups.

Conclusions: Since QOL is a significant predictor of outcome, PH-CHD patients need an early provision of psychosocial, health promoting support in addition to dedicated care and targeted PH treatment. It is therefore pivotal to timely identify unique psychosocial impairments in order to enhance quantity and QOL in this particularly vulnerable patient population.

Keywords: Congenital heart defects; eisenmenger; fontan; pulmonary hypertension (PH); quality of life (QOL)

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Introduction

Pulmonary vascular disease and pulmonary hypertension (PH) are among the most relevant complications associated with congenital heart defects (CHD) and responsible for a high morbidity and mortality (1,2). Although epidemiological data are scarce, the estimated prevalence of pre- or postcapillary forms of PH in adults with CHD (ACHD) is ranging from 3.2% to 28% (3-5). The clinical classification of PH in CHD (PH-CHD) comprises the Eisenmenger Syndrome (ES), PH associated with a predominant systemic-to-pulmonary shunt, PH associated with a small defect, and PH associated with a repaired defect, as well as special forms such as pulmonary vascular disease in Fontan circulation (6). Only recent advances in research and treatment of PH have led to increasing survival rates (7). Although it is well known that patients with idiopathic forms of PH are at risk for a severely impaired quality of life (QOL) (8), reports on impaired QOL due to PH-CHD are still limited and not routinely employed in general care of this particular patient population. Additionally, previous studies have shown that QOL can be an important determinant of health outcomes in patients with PH (9). Although the relationship between QOL and clinical outcomes is not fully elucidated, QOL has been identified as a prognostic marker in the prognosis, survival rate and treatment response of PH patients (9). Consequently, the management goals of PH-CHD should be expanded from prolonging mere survival to improving QOL.

In light of this, the aim of this study was to (I) systematically assess QOL in PH-CHD patients and (II) compare findings to patients with other forms of CHD with normal pulmonary artery pressures and resistance. Obtained findings should generate a differentiated understanding of the psychological situation in PH-CHD patients and may open up productive avenues for psychosocial, health promoting interventions to further enhance both, QOL and longevity in this particular patient population. We present the following article in accordance with the STROBE reporting checklist (available at https://cdt.amegroups.com/article/view/10.21037/cdt-22-284/rc).

Methods

Study population

The present comparative study was part of a recent nationwide survey about the healthcare situation of ACHD throughout Germany ("VEmaH study"), in which more than 4,000 patients were enrolled (10). Within this survey, a consecutively recruited subgroup of 803 ACHD was analyzed (Figure S1). Data collection took place from September 2017 until February 2018. For inclusion, patients had to fulfill the following criteria: (I) confirmed diagnosis of CHD, (II) participation in the preceding VemaH study, (III) age over 18 years, and (IV) enrollment as a patient at the German Heart Center Munich. Patients were categorized into two subgroups: (A) CHD without PH, and (B) CHD with manifest PH. Allocation to collective (B) was done according to the established PH criteria valid at that time (6,11). The categorization was conducted by experienced CHD cardiologists based on the clinical experience and according to data from the literature.

Measures

Medical records were reviewed for healthcare related information, sociodemographic parameters, relevant medical history, type of leading CHD, existing residua/sequelae, and surgical/interventional status.

For the assessment of QOL, the European Quality of Life 5-dimensional questionnaire (EQ-5D-5L) was used. The EQ-5D-5L provides a tool for the measurement of a patient's perceived health status. It has been validated for self-administration with high psychometric properties (12). The questionnaire consists of two sections: a descriptive system questionnaire (EQ-Index) and a visual analogue scale (EQ-VAS). The EQ-Index refers to five dimensions: three for functional aspects (mobility, self-care, routine activities), and the other two for physical and mental well-being (pain/discomfort and anxiety/depression). Patients are asked to indicate the level of their perceived impairments on a 5-point Likert scale (ranging from moderate to severe problems). Responses are represented as single-

digit numbers expressing the severity of impairment on each dimension. A single weighted index score (EQ-5D index) can be calculated using population reference scores. The EQ-VAS indicates a patient's overall health state on a continuous scale ranging from 0 ("The worst health you can imagine") to 100 ("The best health you can imagine"). It is used as a quantitative indicator of a patient's perceived generic health status.

The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013) and approved by the local ethics committee of the Technical University Munich (157/16 S). All participants completed an informed consent form. Guidelines on good pharmacoepidemiological practice (GPP) and all relevant data protection guidelines were followed.

Statistical analysis

Data analysis was performed using IBM SPSS 25.0 (IBM Inc., Armonk, NY, USA). All statistical evaluations were pseudonymized. Descriptive measures were calculated for sociodemographic sample characteristics. Group differences were assessed using *t*-tests, and chi-square. Continuous data was expressed as mean ± standard deviation (SD), categorical or interval scaled variables as absolute numbers or percentages. The crosswalk-index-value of the EQ5D-5L was calculated using the German value set (13). The 95% confidence interval (CI) is displayed as lower- and upper-bound. P values and tests for significance were performed two-sided, on an α-level of 0.05.

Results

A total of 803 patients (female: n=384, 47.8%) was included in the study. The mean age was 35.4±11.9 years (range, 18–86 years).

According to the allocation criteria for the different risk groups, 752 patients [47.3% female; mean age 34.9±11.83 years (range, 18–86 years)] had no PH due to their CHD. In total, 51 patients were diagnosed with manifest PH [55.8% female; mean age: 41.9±12.17 years (range, 21–69 years)]. The two subsets differed significantly in age (P<0.001) but not in sex (P=0.254). *Table 1* shows relevant medical parameters, comorbidities, and surgical/interventional status of the included ACHD by subgroups.

A comparison of the subscales of the EQ-5D-5L for the different risk groups can be found in *Table 2*. In general, most patients presented without symptoms in any of the five dimensions. The highest degree of impairment could be seen in the dimensions of pain/discomfort as well as in anxiety/depression and for PH-CHD patients in usual activities (*Figure 1*). For the dimension of mobility (P<0.001), self-care (P=0.002) and usual activities (P=0.007), significant differences between the groups were found. Only for pain/discomfort (P=0.235) and anxiety and depression no significant group differences were detected (P=0.166).

Significant differences were found between the patient groups for all QOL measurement dimensions (P<0.05). PH-CHD patients demonstrated worse QOL than patients without PH. Comparison between the patient groups showed significant differences in terms of overall QOL [mean no PH: 86.78±13.30 (8.2–100) *vs.* mean PH: 81.79±12.77 (43.6–100); 95% confidence interval (CI): 1.43 to 9.46; P=0.013], the crosswalk index [mean no PH: 93.29±12.86 (–11, 100) *vs.* mean PH: 89.73±11.77 (47–100); 95% CI: 0.08 to 7.56; P=0.043], and the VAS [mean no PH: 80.12±16.50 (15–100) *vs.* mean PH: 72.56±16.80 (40–100); 95% CI: 3.29 to 12.17; P=0.004] (*Table 3*).

Discussion

To date, this is the largest cross-sectional, single-center study to investigate psychosocial implications of PH-CHD. The present study provides important information about the QOL of adults with PH-CHD that should be taken into account in the complex care of this special group of patients.

Although targeted PH treatment has become a cornerstone in the management of affected CHD patients (14), present findings indicate that their general well-being in terms of QOL remains markedly impaired. The study contributes to the paucity of knowledge on this topic by employing the EQ-5D-5L which allows a highly valid and reliable evaluation of QOL for both, clinical and research settings (12). Present findings provide evidence that patients with PH-CHD are at particularly high risk for a diminished QOL and major mental, psychosocial and physiological impairments. Physical impairment, struggle to cope with everyday life and debilitating psychiatric symptoms were the most commonly affected domains in patients, comparable to recently published results of an international multicentre study (12). Since diminished QOL was found to predict mortality in PH-CHD patients (14), there is an urgent need for optimisation of care in order to advance both, quality and lifespan in this particularly vulnerable patient population.

Table 1 Comorbidities of adults with CHD with or without PH

Anamnestic parameters	Overall		No PH		Manifest PH	
	N	%	n	%	n	%
Functional class according to Perloff						
I/II	759	94.8	726	96.7	33	66.0
III	40	5.0	25	33.3	15	30.0
IV	2	0.2	0	0	2	4.0
Cyanosis (O_2<90%)						
No	759	94.9	728	97.1	31	62.0
Yes	30	3.8	12	1.6	18	36.0
Unknown	11	1.4	10	1.3	1	2.0
ES						
No	778	97.3	746	99.5	32	64.0
Yes	22	2.8	4	0.5	18	36.0
Ventricular function (Echo)						
Normal	705	88.5	668	89.3	37	75.5
Moderate reduced	70	8.8	59	7.9	11	22.4
Severe reduced	9	1.1	8	1.1	1	2.0
Unknown	13	1.6	13	1.7	0	0
Infective endocarditis						
No	761	95.1	716	95.5	45	90.0
Yes	24	3.0	20	2.7	4	8.0
Unknown	15	1.9	14	1.9	1	2.0
Cardiac decompensation						
No	724	90.6	692	92.4	32	64.0
Yes	55	6.9	42	5.6	13	26.0
Unknown	20	2.5	15	2.0	5	10.0
Atrial arrhythmias						
No	624	78.0	595	79.3	29	58.0
Yes	169	21.1	149	19.9	20	40.0
Unknown	7	0.9	6	0.8	1	2.0
Arrhythmias						
No	603	75.3	572	76.2	31	62.0
Yes	198	24.7	179	23.8	19	38.0
Ventricular arrhythmias						
No	714	89.8	665	90.5	39	78.0
Yes	72	9.1	61	8.3	11	22.0
Unknown	9	1.1	9	1.2	0	0

Table 1 *(continued)*

Table 1 *(continued)*

Anamnestic parameters	Overall		No PH		Manifest PH	
	N	%	n	%	n	%
Arterial hypertension						
No hypertension (<140/<90 mmHg)	540	67.6	510	68.1	30	60.0
Currently under therapy	163	20.4	145	19.4	18	36.0
Mild hypertension (140–159/90–99 mmHg)	56	7.0	55	7.3	1	2.0
Moderate hypertension (160–179/100–109 mmHg)	16	2.0	15	2.0	1	2.0
Unknown	24	3.0	24	3.2	0	0
Aortopathy						
No	676	84.4	631	84.0	45	90.0
Yes	125	15.6	120	16.0	5	10.0
Thromboembolic events						
No	652	81.5	617	82.4	35	70.0
Unknown	52	6.5	47	6.3	5	10.0
Phlebothrombosis	12	1.5	10	1.3	1	2.0
Pulmonary embolism	3	0.4	1	0.1	2	4.0
TIA, RIND or stroke	65	8.1	59	7.9	6	12.0
Peripheral artery disease	16	2.0	15	2.0	1	2.0
Hyperlipidemia						
No	332	41.6	307	41.0	25	50.0
Yes	69	8.6	60	8.0	9	18.0
Unknown	398	49.8	382	51.0	16	32.0
Hyperurikemia						
No	353	44.2	335	44.7	18	36.0
Yes	48	6.0	33	4.4	15	30.0
Unknown	398	49.8	381	50.9	17	34.0
Diabetes mellitus						
No	342	43.2	321	43.3	21	42.9
Type I	0	0	0	0	0	0
Type II	25	3.2	21	2.8	4	8.2
Unknown	424	53.6	400	53.9	24	49.0
Other (extracardiac) comorbidities						
No	400	49.9	378	50.3	22	44.0
Yes	401	50.1	373	49.7	28	56.0

CHD, congenital heart disease; PH, pulmonary hypertension; ES, Eisenmenger Syndrome; TIA, transient ischemic attack; RIND, reversible ischemic neurological deficit.

Table 2 Different dimensions of QOL from the patient's perspective in patients with and without manifest PH

EQ-5D-5L dimension	Overall, n (%)	No PH, n (%)	Manifest PH, n (%)	95% CI	P value
Mobility				(–0.46, –0.04)	<0.001
No problems	613 (83.1)	580 (84.5)	33 (63.5)		
Slight problems	72 (9.8)	59 (8.6)	13 (25.0)		
Moderate problems	38 (1.4)	32 (4.7)	6 (11.5)		
Severe problems	10 (5.1)	10 (1.5)	0		
Extreme problems	5 (0.7)	5 (0.7)	0		
Self-care				(–0.51, –0.04)	0.002
No problems	696 (94.6)	652 (95.2)	44 (86.3)		
Slight problems	26 (3.5)	19 (2.8)	7 (13.7)		
Moderate problems	6 (0.8)	6 (0.9)	0		
Severe problems	6 (0.8)	6 (0.9)	0		
Extreme problems	2 (0.3)	2 (0.3)	0		
Usual activities				(–0.57, –0.06)	0.007
No problems	547 (74.7)	518 (76.1)	29 (56.9)		
Slight problems	104 (14.2)	93 (13.7)	11 (21.6)		
Moderate problems	58 (7.9)	48 (7.0)	10 (19.6)		
Severe problems	19 (2.6)	18 (2.6)	1 (2.0)		
Extreme problems	4 (0.5)	4 (0.6)	0		
Pain/discomfort				(–0.42, 0.02)	0.235
No problems	484 (66.2)	457 (67.3)	27 (51.9)		
Slight problems	165 (22.6)	148 (21.8)	17 (32.7)		
Moderate problems	67 (9.2)	60 (8.8)	7 (13.5)		
Severe problems	13 (1.8)	12 (1.8)	1 (1.9)		
Extreme problems	2 (0.3)	2 (0.3)	0		
Anxiety/depression				(–0.55, 0)	0.166
No problems	469 (64.2)	442 (65.0)	27 (52.9)		
Slight problems	176 (24.1)	163 (24.0)	13 (25.5)		
Moderate problems	56 (7.7)	48 (7.1)	8 (15.7)		
Severe problems	28 (3.8)	25 (3.7)	3 (5.9)		
Extreme problems	2 (0.3)	2 (0.3)	0		

QOL, quality of life; PH, pulmonary hypertension; EQ-5D-5L, European Quality of Life 5 Dimensions—5 Levels questionnaire; CI, confidence interval.

The two-fold measure of the EQ-5D-5L revealed differences in QOL values depending on the type of measurement applied. Apparently, the overall VAS score indicated significantly worse QOL scores than the descriptive value stemming from the specific factor descriptions. One explanation for this discrepancy could be due to differences in the QOL coverage of both measures (15). It can be assumed that the descriptive system encourages a patient to examine QOL from several angles while the VAS picks up a one-dimensional and therefore generic view of perceived somatic health restrictions imposed by PH. Therefore, the average reduction in VAS scores was considerably greater in PH-CHD than among CHD patients without PH.

Overall, however, it is noticeable, particularly on the basis of *Figure 1*, that patients with manifest PH-CHD have the lowest QOL across all five dimensions. This also becomes evident in the calculation of overall QOL.

Independent of the presence of PH, all included patients reported impairments mostly within the dimensions anxiety, depression (11.08%), pain and discomfort (11.03%). This is in line with a former large-scale study of Andonian *et al.*

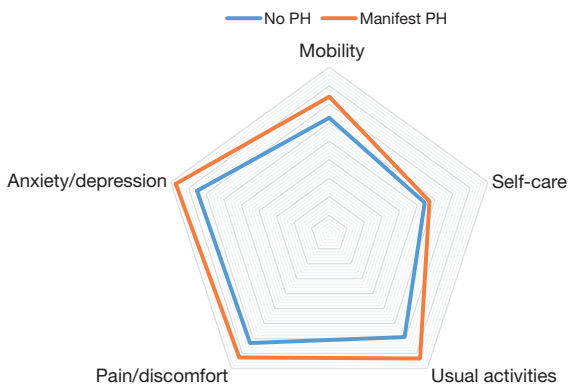

Figure 1 Dimensions of QOL in patients with CHD with and without PH. PH, pulmonary hypertension; QOL, quality of life; CHD, congenital heart disease.

investigating 4,000 patients with CHD (15), as well as a study by Berghammer *et al.* (16), who found that ACHD present significant impairment in the perception of their physical and mental well-being. Accordingly, emotional distress as well as problems related to functional aspects (pain and discomfort) were higher in PH-CHD as compared to the general public *per se*. A further increased prevalence of depression and anxiety in PH-CHD patients has been well documented previously (17-19). Potential risk factors for the development of anxiety or depression in this patient population include socioeconomic factors, such as financial burden due to treatment or unemployment, as well as emotional burden due to the prognostic uncertainty and continuous deterioration of their condition (20). In addition, patients with PH showed significantly higher impairment in the domains of mobility, self-care and usual activities. These findings might be explained by functional restrictions, such as decreased exercise capacity, dyspnoea and limitations in day-to-day activities (8,21).

Limitations

To date, the present study represents the largest mono-center study to comprehensively assess QOL in PH-CHD patients. A representative comparison group was surveyed with the same assessment tools to offer a point of reference. However, some limitations must be considered when interpreting the current results.

First, the study was cross-sectional in nature and therefore depicts a snapshot of a patient's health status. It not possible to draw conclusions about the etiology of psychological, psychosocial and physiological effects or the development of QOL in patients with or without PH-CHD. Since there is preliminary evidence on a continuous deterioration of QOL as the condition of PH progresses, longitudinal studies designs would be feasible to obtain a thorough understanding of the disease-related burden over time. Second, selection bias in respondents willing

Table 3 QOL, crosswalk index and VAS of patients with and without manifest PH

Dimension	Overall (mean ± SD; range)	No PH (mean ± SD; range)	Manifest PH (mean ± SD; range)	95% CI	P value
Crosswalk*	93.03±12.84; [−11, 100]	93.29±12.86; [−11, 100]	89.73±11.77; [47, 100]	0.08 to 7.56	0.043
QOL overall*	86.43±13.32; [8.2, 100]	86.78±13.30; [8.2, 100]	81.79±12.77; [43.6, 100]	1.43 to 9.46	0.013
VAS*	79.61±16.62; [15, 100]	80.12±16.50; [15, 100]	72.56±16.80; [40, 100]	3.29 to 12.17	0.004

*, statistically significant (P≤0.05). PH, pulmonary hypertension; QOL, quality of life; VAS, visual analogue scale; SD, standard deviation; CI, confidence interval.

to participate appears unavoidable, such that more serious disease manifestations might be more (or less) frequent among non-responders. Third, patients with PH are significantly older than patients without PH. This condition is caused by the progressive character of the disease. Therefore, age might be an important and influencing factor on QOL besides the PH itself. Fourth, in future research, more psychosocial endpoints, such as depression, anxiety and overall emotional distress should be considered. Additionally, investigating more medical parameters, such as treatment and treatment duration, might add valuable insights on QOL in PH-CHD patients. Finally, the study was based on self-report outcomes and might be subject to recall and self-presentation bias.

Conclusions

PH is an often underestimated complication and sequel of a broad variety of CHD. Given the established negative consequences of PH on morbidity and mortality in ACHD, it is pivotal to identify unique psychosocial impairments and prevent further psychological and physical decline.

Patients would therefore benefit most from early interdisciplinary assessments and continuous health promotion, resource oriented counselling as routine component of a holistic approach to PH management.

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Footnote

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Supplementary

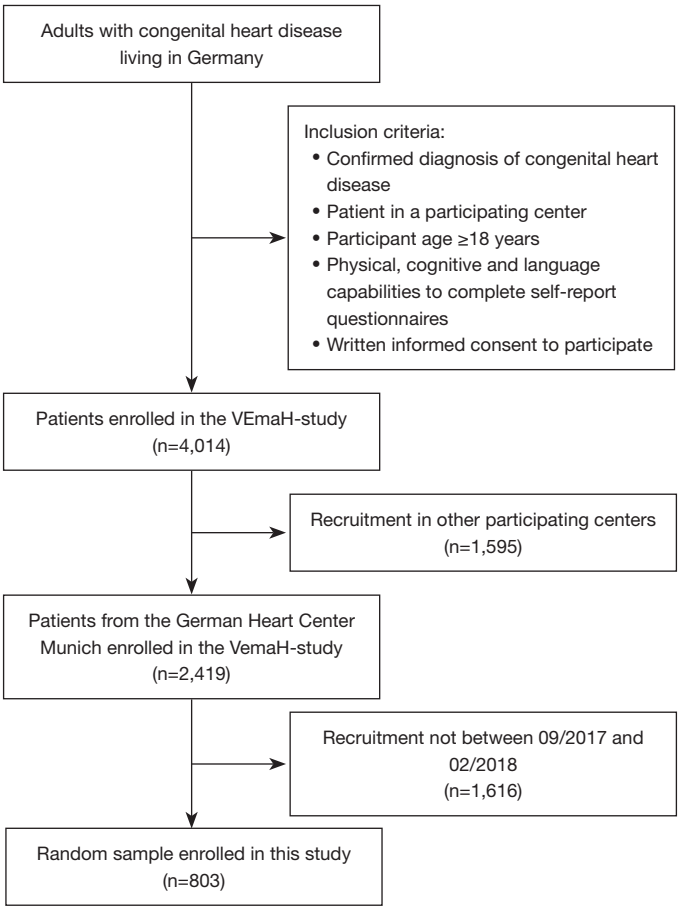

Figure S1 Flow chart for patient selection. PH, pulmonary hypertension.

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