Original Article

Healthcare status of adults with pulmonary hypertension due to congenital heart disease

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Background: In the long-term course of treated and untreated congenital heart defects (CHD), pulmonary hypertension (PH) is one of the most relevant complications. Since PH carries a high risk for mortality and morbidity, it is important to improve the status of healthcare and medical knowledge regarding the affected patients. Therefore, this study aimed to determine the current medical care status, health-related knowledge, and specific counseling needs of adults with various forms of CHD (ACHD) who are at increased risk of developing PH, as well as those with manifest PH.

Methods: In this retrospective cross-sectional study, a representative sample of 803 ACHD were analyzed. Patients were split into three groups based on CHD: low risk for PH, at risk for pre- or post-capillary PH, and manifest PH. Data collection took place between September 2017 until February 2018 in a tertiary care center for ACHD. Healthcare status and specific needs for information or advice were analyzed using a questionnaire designed by our group. The state of knowledge of the patients was assessed by comparing this questionnaire and the corresponding medical records.

Results: Both patients with manifest PH (n=51) and patients at risk to develop PH (n=629) were insufficiently informed about their health status, specific care structures available to them, and patient organizations. About 50% of the patients had specific counseling needs, especially regarding physical capability and sports, daily stress, rehabilitation measures, and pregnancy. Only 47.8% of patients with manifest PH were aware of suffering from PH (P<0.001). In particular, the patients had large knowledge deficits regarding comorbidities related to their health condition.

Conclusions: PH is a quantitatively and qualitatively underestimated residuum or sequela of CHD that significantly affects outcome and prognosis in ACHD. Multidisciplinary, structured, and specific counseling of affected individuals with corresponding risk constellations is urgently needed. A prerequisite for this is closer collaboration between primary care physicians (PCPs), such as general practitioners, family physicians, internists, or general cardiologists, and ACHD specialists. Targeted patient counseling and care could have a positive impact on the level of awareness of those affected and favorably influence their prognosis.

Keywords: Congenital heart defects; pulmonary hypertension (PH); healthcare status; counselling needs; CHD

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Introduction

Congenital heart defects (CHD) are the most common inborn single-organ anomalies (1,2). In the long-term course of treated and untreated CHD, pulmonary hypertension (PH) is one of the most relevant residua, sequelae, or complications that can affect quality of life as well as morbidity and mortality (3). PH develops in an estimated 5–10% of patients with CHD and accounts for 34–42% of all PH cases, exact numbers being uncertain (4,5). CHD that may be complicated by PH include, in particular, primary left-right shunt lesions, cyanotic heart defects with increased pulmonary flow, congenital obstructions of the left heart, and anomalies of the pulmonary artery.

According to previous World Conferences or Symposia on Pulmonary Hypertension (6), PH can be classified into five main groups [1: pulmonary arterial hypertension (PAH), 2: PH due to left heart disease, 3: PH due to lung disease, 4: PH due to chronic blood clots in the lungs, and 5: PH due to unknown causes]. CHD are associated in particular with PH falling into groups 1 and 2 of this classification, but increasingly also in the remaining three. Moreover, PH in association with congenital shunt lesions can be allocated to clinically defined groups, including Eisenmenger syndrome, correctable or non-correctable left-to-right shunts, PAH occurring incidentally with CHD, and PH after CHD repair. In addition, there are special situations in CHD, such as Fontan circulation and segmental forms of PAH (7-9).

Since PH carries a high risk of mortality and morbidity, it is important to improve access to PH-specific healthcare for patients with CHD. There is a gap in knowledge regarding the awareness level of patients and their primary care providers about their own disease and, the appropriate level of care needed. In addition, there is a great need for targeted medical advice (10-12).

Therefore, this study aimed to determine the medical care status, health-related knowledge, and specific counseling needs of adults with CHD and manifest PH, and those who are at an increased risk of developing PH. We present the following article in accordance with the STROBE reporting checklist (available at https://cdt.amegroups.com/article/view/10.21037/cdt-22-281/rc).

Methods

Study cohort

This cross-sectional, questionnaire-based study was part of a recent nationwide survey about the status of care for adults with CHD (ACHD) throughout Germany ("VEmaH study"), which included more than 4,000 patients (13). Within this survey, a subgroup of 803 ACHD was analyzed (Figure S1). For patients in this subgroup, clinical records were analyzed and combined with the patient-reported VEmaH-questionnaire.

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 of Munich (No. 157/16 S). Participating patients completed an informed consent form. Guidelines on good pharmacoepidemiological practice (GPP) and data protection guidelines were followed.

Patient inclusion

Inclusion criteria were presence of CHD, participation in the preceding VEmaH study, age over 18 years, and enrollment as a patient at the German Heart Center Munich. Patients for whom data were collected between September 2017 and February 2018 were included in the study in the order in which they presented to the investigating institution.

For the analysis, patients were categorized into one of three subgroups: (A) CHD with low risk for PH, (B) CHD with increased risk to develop either pre- or post-capillary PH, and (C) CHD with manifest PH. Allocation to collective (C) was made according to the established PH criteria valid at that time (7,14). Cases in which the assignment was ambiguous were allocated in discussion of two experienced CHD cardiologists based on their clinical experience and according to data from the literature (*Table 1*).

Questionnaire

For this study, it was not possible to use a standardized and validated questionnaire to explore the "real world data" on patients' health status and/or health care. Therefore, in collaboration with the Chair of Behavioral Epidemiology at the Technical University of Dresden and the German Heart Center Munich as a tertiary care center for ACHD, a dedicated questionnaire was developed (Appendix 1). The questionnaire contains items on sociodemographic conditions, CHD, care providers for medical problems in general, and, for CHD-related problems, patient counseling needs and knowledge of specific care structures.

Medical records were reviewed for healthcare-related information, sociodemographic parameters, relevant

Table 1 Subgroup analysis of adults with CHD assigned into three groups: low risk of PH, increased risk for pre- or post-capillary PH, and manifest PH

Demographic parameters	Low risk of PH (n=118)	Increased PH risk (n=634)	Manifest PH (n=51)
Age (years)	38.5±12.9	34.4±11.4	42.4±12.0
Sex (female:male) (%)	54.2:45.8	44.3:55.7	54.9:45.1
BMI (kg/m^2)	26.0±16.1	25.0±4.0	24.0±5.4
Type of CHD [n]	PFO [34]	Aortic valve disease [86]	VSD [13]
	Ebstein anomaly [29]	d-TGA [81]	d-TGA [7]
	PS [27]	CoA [71]	PA + VSD [5]
	Aortic aneurysm [14]	TOF [69]	ASD [4]
	MVPS [13]	VSD [56]	AVSD [4]
	IAA [1]	ASD [49]	ccTGA [2]
		ccTGA [27]	Mitral valve disease [2]
		Connective tissue disease [23]	CoA [2]
		AVSD [19]	Ebstein anomaly [2]
		TA [17]	PAPVC [2]
		PA + VSD [15]	TOF [2]
		DIV [12]	TA [2]
		Mitral valve disease [12]	DIV [1]
		Cardiomyopathy, congenital [11]	DORV (Fallot-Type) [1]
		PDA [10]	DORV + TGA [1]
		AVSD, partial [9]	AVSD, partial [1]
		Subaortic stenosis [9]	
		PA + IVS [8]	
		PAS [7]	
		DORV (Fallot-Type) [7]	
		TAC [6]	
		DORV + TGA [5]	
		HCM [4]	
		PAPVC [4]	
		TAPVC [4]	
		HLHS [1]	
		Others* [12]	

*, others = complex, other: n=5; arrhythmias, congenital: n=3; coronary artery anomaly: n=3; comorbidities, extracardiac: n=1. CHD, congenital heart defects; PH, pulmonary hypertension; BMI, body mass index; PFO, patent foramen ovale; VSD, ventricular septal defect; d-TGA, dextro-transposition of the great arteries; PS, pulmonary stenosis; CoA, aortic coarctation; PA, pulmonary atresia; TOF, tetralogy of Fallot; ASD, atrial septal defect; MVPS, mitral valve prolapse; AVSD, atrio-ventricular septal defect; IAA, interrupted aortic arch; ccTGA, congenitally corrected transposition of the great arteries; TA, tricuspid atresia; PAPVC, partial anomalous venous return; DIV, double inlet ventricle; DORV, double outlet right ventricle; PDA, patent ductus arteriosus; TGA, transposition of the great arteries; IVS, intact ventricular septum; PAS, pulmonary arterial stenosis; TAC, truncus arteriosus communis; HCM, hypertrophic cardiomyopathy; TAPVC, total anomalous pulmonary venous return; HLHS, hypoplastic left heart syndrome.

medical history, type of leading CHD, existing residua/sequelae, and surgical/interventional status. In addition, specific counseling needs were determined. To evaluate each patient's knowledge regarding diagnosis and comorbidities related to his/her health condition, the patient-reported answers on the VEmaH-questionnaire (http://www.vemah.info) and the clinically validated physicians' diagnoses were compared.

Statistical analysis

Continuous data are presented as mean ± standard deviation, and categorical or interval-scaled variables as absolute numbers or percentages. The kappa statistic was used as a measure of agreement to statistically compare responses in both questionnaires. The Landis and Koch classification was used to interpret the kappa coefficients (15). Group differences were assessed using *t*-test, Kruskal-Wallis test, and Chi-Squared-test.

Data analysis was performed using IBM SPSS 25.0 (IBM Inc., Armonk, NY, USA). Tests for significance were performed two-sided, and P values were based on an α level of 0.05. All statistical analyses were performed on pseudonymized data. As multiple responses were allowed for some questions, the number of responses received may differ from the total number of study participants surveyed.

Results

Study sample and patient characteristics

A total of 803 patients were included in the study. Of these, n=384 (47.8%) were female. The mean age was 35.4±11.9 years (range, 18–86 years). The patients were assigned to three different risk groups: those with low risk of developing PH, those with an increased risk of developing PH, and those with manifest PH.

Significant differences existed between the manifest-PH group and the increased-risk-of-PH group. In the manifest-PH group, age was significantly higher (P<0.001) and there were significantly more women (P<0.05). In the entire cohort, the number of cardiac and non-cardiac comorbidities was remarkable, particularly regarding heart failure, cardiac arrhythmias, and non-cardiac comorbidities, such as arterial hypertension, aortopathies, thromboembolic events, and metabolic disorders. People with manifest PH represented the worst functional class (more people FC-3), with a higher proportion in an unfavorable functional class and (as expected) the largest proportion of ES patients. This group also had the highest percentage of patients with heart failure as well as arrhythmias, either in the form of ventricular or, more commonly, supraventricular arrhythmias. Among the non-cardiac comorbidities, hyperuricemia was common in this group. *Tables 1,2* present relevant medical parameters, comorbidities, and surgical/interventional status of the included ACHD by subgroups.

Specific counseling needs of ACHD

The need for advice was high in both the entire cohort (n=485; 60.4%) and in all three individual groups (*Table 3*). The most frequently expressed demand for advice was about physical performance/sports activity, resilience in everyday life, as defined by Sisto *et al.* (16), and retirement planning.

The need for advice was lowest for obtaining a driver's license and for forms of education (school, university, and work).

There were significant differences between the groups. Patients with increased PH risk (P=0.007) or patients without PH risk (P=0.01) had more than three times the need for counseling regarding pregnancy compared with patients with manifest PH.

Patients with manifest PH, on the other hand, had a significantly higher need for counseling regarding rehabilitation, compared with patients with increased PH risk (P=0.05).

Basic medical care for general, CHD-associated, and CHD-independent medical problems

For general medical (i.e., not primarily CHD-related) problems, ACHD consulted a general practitioner, family physician, internist, or general cardiologist as their primary care physician (PCP) in 84.1% (675/803) of the cases. Of the 803 ACHD included, the PCPs were aware of their patient's CHD in 762 (94.9%) cases. No significant group differences were found (increased-PH-risk group: 95.4%; manifest-PH group: 90.2%; low risk group: 94.3%). However, according to the patients, in 9.8% of CHD patients with manifest PH, the PCP was unaware of the CHD.

In more than half of the patients (448 patients; 55.8%) the PCP was also the first caregiver for problems directly related to the CHD. Again, however, no statistical differences were found between the groups. Nevertheless, in patients at increased risk of PH, the PCP was more often the first contact for problems related to the CHD, compared with the other groups (increased-PH-risk group:

Table 2 Comorbidities of adults with CHD with or without risk for PH

Anamnestic parameters	Overall, n (%)	Low risk for PH, n (%)	At risk for PH, n (%)	Manifest PH, n (%)
Functional class according to Perloff				
I/II	759 (94.8)	118 (96.7)	608 (96.7)	33 (66.0)
III	40 (5.0)	4 (3.3)	21 (3.3)	15 (30.0)
IV	2 (0.2)	0	0	2 (4.0)
Cyanosis (O_2 <90%)				
No	759 (94.9)	118 (96.7)	610 (97.1)	31 (62.0)
Yes	30 (3.8)	1 (0.8)	11 (1.8)	18 (36.0)
Unknown	11 (1.4)	3 (2.5)	7 (1.1)	1 (2.0)
Ventricular function (echocardiography-Doppler)				
Normal	705 (88.5)	117 (96.7)	551 (87.9)	37 (75.5)
Moderate reduced	70 (8.8)	3 (2.5)	56 (8.9)	11 (22.4)
Severe reduced	9 (1.1)	0	8 (1.3)	1 (2.0)
Unknown	13 (1.6)	1 (0.8)	12 (1.9)	0
Infective endocarditis				
No	761 (95.1)	119 (98.3)	597 (94.9)	45 (90.0)
Yes	24 (3.0)	0	20 (3.2)	4 (8.0)
Unknown	15 (1.9)	2 (1.7)	12 (1.9)	1 (2.0)
Heart failure				
No	724 (90.6)	117 (95.9)	575 (91.7)	32 (64.0)
Yes	55 (6.9)	2 (1.6)	40 (6.4)	13 (26.0)
Unknown	20 (2.5)	3 (2.5)	12 (1.9)	5 (10.0)
Arrhythmias (general)				
No	603 (75.3)	95 (77.9)	477 (75.8)	31 (62.0)
Yes	198 (24.7)	27 (22.1)	152 (24.2)	19 (38.0)
Atrial arrhythmias				
No	624 (78.0)	97 (79.5)	498 (79.3)	29 (58.0)
Yes	169 (21.1)	24 (19.7)	125 (19.9)	20 (40.0)
Unknown	7 (0.9)	1 (0.8)	5 (0.8)	1 (2.0)
Ventricular arrhythmias				
No	714 (89.8)	115 (94.3)	560 (89.9)	39 (78.0)
Yes	72 (9.1)	6 (4.9)	55 (8.8)	11 (22.0)
Unknown	9 (1.1)	1 (0.8)	8 (1.3)	0
Arterial hypertension				
No hypertension (<140/<90 mmHg)	540 (67.6)	76 (62.3)	434 (69.2)	30 (60.0)
Currently under therapy	163 (20.4)	29 (23.8)	116 (18.5)	18 (36.0)
Mild hypertension (140–159/90–99 mmHg)	56 (7.0)	10 (8.2)	45 (7.2)	1 (2.0)
Moderate hypertension (160–179/100–109 mmHg)	16 (2.0)	5 (4.1)	10 (1.6)	1 (2.0)
Unknown	24 (3.0)	2 (1.6)	22 (3.5)	0

Table 2 *(continued)*

Table 2 *(continued)*

Anamnestic parameters	Overall, n (%)	Low risk for PH, n (%)	At risk for PH, n (%)	Manifest PH, n (%)
Aortopathy				
No	676 (84.4)	101 (82.8)	530 (84.3)	45 (90.0)
Yes	125 (15.6)	21 (17.2)	99 (15.7)	5 (10.0)
Thromboembolic events				
No	652 (81.5)	82 (67.2)	535 (85.2)	35 (70.0)
Unknown	52 (6.5)	3 (2.5)	44 (7.0)	5 (10.0)
Phlebothrombosis	12 (1.5)	2 (1.6)	9 (1.4)	1 (2.0)
Pulmonary embolism	3 (0.4)	0	1 (0.2)	2 (4.0)
Transient ischemic attack, reversible ischemic neurologic deficit, or stroke	65 (8.1)	30 (24.6)	29 (4.6)	6 (12.0)
Peripheral artery disease	16 (2.0)	5 (4.1)	10 (1.6)	1 (2.0)
Hyperuricemia				
No	353 (44.2)	55 (45.1)	280 (44.7)	18 (36.0)
Yes	48 (6.0)	3 (2.5)	30 (4.8)	15 (30.0)
Unknown	398 (49.8)	64 (52.5)	317 (50.6)	17 (34.0)

CHD, congenital heart defects; PH, pulmonary hypertension.

Table 3 Specific need for counseling, in absolute numbers and percent (by subgroups)

Counseling need	Overall, n (%)	Low risk for PH, n (%)	At risk for PH, n (%)	Manifest PH, n (%)
Healthcare insurance	226 (28.1)	36 (29.5)	175 (28.6)	15 (30.0)
Life insurance	233 (29.0)	32 (26.9)	184 (30.4)	17 (34.0)
Retirement insurance	265 (33.0)	36 (30.8)	212 (35.2)	17 (34.7)
Pension	167 (20.8)	19 (25.0)	137 (21.8)	11 (32.4)
Resilience, everyday life	315 (39.2)	53 (43.1)	243 (38.6)	19 (37.3)
Profession	213 (26.5)	38 (30.9)	164 (26.1)	11 (21.6)
Education	50 (6.2)	12 (9.8)	37 (5.9)	1 (2.0)
Ability to fly	127 (15.8)	18 (14.6)	100 (15.9)	9 (17.6)
Driver's license	37 (4.6)	9 (7.3)	28 (4.5)	0
Genetics	163 (20.3)	28 (22.8)	125 (19.9)	10 (19.6)
Physical performance, sports	354 (44.1)	54 (43.9)	276 (43.9)	24 (47.1)
Rehabilitation measures	197 (24.5)	34 (27.6)	145 (23.1)	18 (35.3)
Pregnancy	202 (25.2)	37 (30.1)	160 (25.4)	5 (9.8)
Other	20 (2.5)	4 (3.3)	14 (2.2)	2 (3.9)

PH, pulmonary hypertension.

Table 4 Healthcare status of patients with CHD (by subgroups)

Healthcare parameter	Overall, n (%)	Low risk for PH, n (%)	PH risk, n (%)	Manifest PH, n (%)
ACHD cardiologist known	145 (18.1)	22 (18.0)	116 (18.4)	7 (14.0)
Pediatric cardiologist known	242 (30.2)	36 (29.5)	193 (30.7)	13 (26.0)
ACHD center known	225 (28.1)	32 (26.2)	174 (27.7)	19 (38.0)
No ACHD center known	317 (39.6)	56 (45.9)	244 (38.8)	17 (34.0)
Referral for CHD-specific problems	260 (32.5)	42 (34.4)	196 (31.2)	22 (44.0)
Referral for conditions/problems affecting CHD	43 (5.4)	7 (5.7)	30 (4.8)	6 (12.0)
No referral	402 (50.1)	59 (48.4)	324 (51.5)	19 (38.0)

CHD, congenital heart defects; PH, pulmonary hypertension; ACHD, adults with congenital heart defect(s).

56.4%; manifest-PH group: 52.9%; low risk group: 53.7%).

Referral to an institution that specializes in CHD because of a medical problem

Overall, 50.1% (n=402) of all included ACHD had never been referred to a CHD specialist by their PCP. Compared with the manifest-PH group, the other two groups were significantly less likely to be referred to a specialized institution [increased-PH-risk group: 51.5%; manifest-PH group: 38.0%; low risk group: 48.4%); P<0.05]. Remarkably, only 44.0% (n=22) of the patients with manifest PH reported that a PCP had ever referred them to a CHD specialist in the past because of cardiac problems related to their CHD. There were even fewer referrals in the increased-PH-risk group, in which only 31.2% (n=196) of patients had been referred to a CHD specialist because of cardiac problems related to their CHD.

Referral rates were even worse for problems or medical conditions that may affect CHD progression (increased-PH-risk group: 4.8%; manifest-PH group: 12.0%; low risk group: 5.7%). Looking at healthcare delivery, significant differences regarding patient referral to CHD-certified specialists were observed. CHD patients with manifest PH, who experienced problems with their CHD (P=0.027) or diseases that could affect the course of their CHD (P=0.014) were significantly more likely to be referred to a specialist than the high-risk or low-risk population.

Patients' knowledge about targeted treatment for ACHD in Germany

All patients were asked about their awareness of certified facilities for the provision of ACHD care. The majority of the affected patients had no knowledge of certified and accredited pediatric cardiologists, adult cardiologists, specialized hospitals, or cardiac centers for ACHD (*Table 4*). In all groups, patients were more aware of pediatric cardiologists with ACHD certification than non-pediatric cardiologists with ACHD certification.

Among the patients low risk of PH, 18.0% (n=22) knew an accredited cardiologist, and 29.5% (n=36) knew an accredited pediatric cardiologist (11.5%). Among patients at risk of PH, 18.4% (n=116) were aware of an ACHD cardiologist or pediatric cardiologist (30.7%; n=193). Among the manifest-PH group, 14.0% (n=7) were familiar with a specialized and accredited cardiologist and 26.0% (n=13) were familiar with a pediatric cardiologist (12.0%). Only 28.1% of the study population knew about specialized and accredited ACHD centers. On this measure, there was no significant difference between the groups.

Patient knowledge about patient organizations for ACHD in Germany

The majority of patients surveyed were insufficiently informed about dedicated patient organizations. Among the group of patients without risk of developing PH, 27.6% (n=34) were adequately informed. Among the patients at risk of PH, 36.4% (n=229) answered "yes" to knowledge of peer-support groups, while, among the patients with manifest PH, 45.1% (n=23) were informed about the support groups. Patients with manifest PH were significantly better informed about patient organizations than patients without risk of developing PH (P<0.05).

Table 5 Kappa-values of health-related level of knowledge by patients (overall and subgroups)

Test parameter	Overall, (Kappa value)	Low risk for PH, (Kappa value)	At risk for PH, (Kappa value)	Manifest PH, (Kappa value)
CHD diagnosis	0.663**	0.669**	0.653**	0.678**
Heart failure	0.249**	0.326**	0.064**	0.301**
PH*	0.478**	–	–	–
Infective endocarditis	0.239**	–	0.199**	0.485**
Cardiac arrhythmias	0.406**	0.369**	0.415**	0.343**
Mental abnormalities	0.036**	0.008	0.047**	–0.016
Thrombosis	0.051**	0.077**	0.043**	–0.033

Interpretation of κ-values: 0 corresponds to the expected, random (i.e., no) agreement, whereas 1 corresponds to a perfect agreement (physician and patient give the same/correct answer). According to the Landis *et al*. classification, κ<0 was designated as "poor", κ=[0–0.20] as "somewhat", κ=[0.21–0.40] as "sufficient", κ=[0.41–0.60] as "moderate", κ=[0.61–0.80] as "considerable", and κ=[0.81–1.00] as "(almost) perfect" agreement. *, PH was treated as a constant for statistical analysis; therefore, group-specific analysis was not possible; **, P<0.05. PH, pulmonary hypertension.

Health-related state of knowledge

Current health-related knowledge was assessed using the following parameters: diagnosis of leading CHD, history of heart failure, PH, cardiac arrhythmias, previous infective endocarditis, mental abnormalities, and thrombosis. For this assessment, the patient-reported statement was compared with an assessment provided by an ACHD-specialized cardiologist.

In all three groups, the patient and the ACHD specialist agreed on the diagnosis of the leading CHD, with high agreement. Regarding the history of heart failure and previous infective endocarditis, sufficient agreement was seen among all three groups. The groups also differed only slightly with regard to the anamnestic presence of cardiac arrhythmias. There was little agreement, however, regarding mental abnormalities and thrombosis (both P<0.05). Overall, no significant group differences were found. The κ-values are presented in *Table 5*.

Regarding the presence of PH in all 803 patients, only a "sufficient agreement" between the patients' level of knowledge and the validated diagnosis of an ACHD cardiologist was seen (κ=0.478; P<0.001).

A total of 749 (99.6%) individuals correctly reported not having PH, while 18 (35.3%) correctly reported having PH. Falsely, 27 out of 51 (52.9%) individuals stated that they did not have PH, when in fact, they had manifest PH. Thus, more than 50% of respondents from the manifest-PH group answered incorrectly.

In the collective with increased PH risk, 9 (1.4%) persons stated that they had PH, although this was not the case.

Discussion

The data from the present study indicate that, in Germany, ACHD with PH (PH-CHD) are not adequately informed about their disease, their need for dedicated congenital cardiological follow-up care and the existing care structures. As the current study data show, not only is dedicated medical care currently not adequately ensured, but there is also a considerable need for improvement in the care these patients do receive. The inadequacy of ACHD care, however, is not just a national problem, but a global one (17).

In the present study, the care status, health-related knowledge, and specific counseling needs of PH-CHD were analyzed in a large subgroup (n=803) of the nationwide VEmaH study (n=4,008). For this purpose, patients were divided into three groups: ACHD with manifest PH, ACHD with increased risk of developing precapillary or postcapillary PH, and ACHD with no intrinsic PH risk. All patients in the manifest-PH group had severe pulmonary vascular disease or PH, and 2.2% (n=18) even had the most severe form of PH-CHD, Eisenmenger syndrome.

The manifest-PH group differed from the other two groups with respect to the sociodemographic parameters of age and sex; they were older and predominantly female. This is in accordance with data from the COMPERA registry, the Euro Heart Survey on adults with congenital heart disease, and the British PH registry, in which more women than men were documented and the prevalence of PH increased with age (10,18,19). It should be noted that although the world's largest PH registries, COMPERA and UK's REVEAL registry, include all types of PH, only 7.7%

and 10% of their study participants, respectively, have both PH and a CHD (10,18).

Analogous to the 2007 Euro Heart Survey (19), in the current study patients with manifest PH were more frequently affected by functional limitations (according to Perloff functional class) and cyanosis than patients without manifest PH. The role of PCPs in providing adequate and continuous care for these patients is a very important one, as they can set the course for appropriate diagnosis and treatment and encourage their patients to undergo regular pre- and post-treatment care (7,20). However, the present study shows severe deficits in this regard. The majority of patients surveyed sought a family physician, general practitioner, internist, or general cardiologist (PCP) as their first point of contact for both general illnesses and also for problems specifically related to their CHD. This is problematic because PCPs generally do not have sufficient knowledge about the management of CHD, especially when it is associated with PH (3,21).

Despite the importance of specialized care for CHD, both physicians and patients have been found to be insufficiently informed about targeted ACHD clinics and centers as well as pediatric cardiologists in private practice and cardiologists with ACHD certification (13,22). The number of referrals from PCPs to CHD specialists was correspondingly low, even when they knew that the patient had a CHD. Therefore, even patients with manifest PH were rarely referred to certified ACHD institutions/cardiologists for cardiac problems that could be related to the CHD or for problems or diseases that could be influenced by the CHD. The referral rates of the PH-risk and low risk comparison groups were even lower. These data raise the concern that inadequate awareness on the part of PCPs of the complexity of CHD and the need for patient management that in many cases differs from that of acquired heart disease, could have a negative impact on the morbidity and mortality of the affected patients (13,21).

To remedy this, PCPs need to be motivated to become more involved in the care of ACHD. In addition, their knowledge of the existing care structures in Germany for adults with CHD should be enhanced.

Descriptive data

Patients with CHD and low risk of developing PH have anomalies that do not affect the lung in terms of volume or pressure overload. In these types of patients, just a few cases of secondary PH are mentioned in the literature (23). ACHDs with a high risk of developing postcapillary PH suffer either from an obstruction that affects their left heart, or structural anomalies of the left ventricle or mitral valve. In these cases, the filling pressure of the left ventricle and left atrium is elevated, and this pressure will be directed passively into the capillary system of the lung. In the case of left ventricular hypertrophy, myocardial perfusion is impaired and systolic and diastolic myocardial dysfunction may occur. These patients are at risk for high diastolic filling pressure which eventually may lead to postcapillary PH (24). CHDs that confer a high risk for precapillary PH can be divided into pre-tricuspid and post-tricuspid shunt lesions. Pre-tricuspid anomalies have a left-to-right shunt, causing volume overload and recirculation to the right heart and the pulmonary arteries (25-27).

Need for specific advice regarding activities of daily life

The need for specific advice was highest regarding athletic/sports capability and resilience in everyday life in all three groups. As also reflected in the current study, many patients need more information about physical athletic capability and sports, because several studies have supported the positive impact of special sports programs for patients with PH on fitness, hemodynamic parameters, and quality of life (28).

As PH-CHD patients during and after pregnancy are at high-risk, with an overall mortality up to 25–56% in severe forms (such as Eisenmenger syndrome) (29,30), patients usually choose not to get pregnant. Moreover, most of them are taking a targeted PAH-medication, which may be accompanied with a contraindication against pregnancy (e.g., in endothelin antagonists). Accordingly, the need for counseling regarding pregnancy was significantly lower in patients with manifest PH than in the other two groups. Education programs about pregnancy in CHD-PH should be established for women at risk and obstetricians to provide appropriate counseling that lives up to the individual needs of affected women. Additionally, specific advice is required regarding all forms of rehabilitation, especially in patients with manifest PH.

The current data showed a considerable need to improve patients' knowledge in all three groups. Many patients with PAH-CHD indicated a broad and individualized need for specific counseling. The counseling needs of patients with manifest PH did not differ from those of patients who were only at increased risk of developing PH.

Since management recommendations for patients with "PH without CHD" may differ fundamentally from those

for PH-CHD (13), ACHD with PH should be counseled by, or at least in cooperation with, certified ACHD specialists. For this, it is of considerable importance to increase awareness among PCPs of ACHD-certified clinics/centers and ACHD (as well as pediatric) cardiologists.

Healthcare status

Despite the high demand for CHD-related information, there is a dearth of awareness regarding specialized centers for CHD. Such centers exist throughout Germany and cover the needs of all three included patient groups. Patients with no intrinsic risk for PH had the least awareness of appropriate care structures, although they constitute the patients who could benefit most from information about a healthy lifestyle and health promotion. Interestingly, among patients the level of awareness regarding pediatric cardiologists was higher than that regarding adult cardiologists. This may be related to the early establishment of a relationship between the patients and their doctors, during childhood. A considerable problem is the fact that many patients with CHD feel subjectively well and have a high quality of life due to early and successful treatment, which often results in avoiding medical advice and not attending regular medical follow-up visits. By contrast, patients with manifest PH are better informed about centers for ACHD and are more likely to receive multidisciplinary care if cardiac or non-cardiac comorbidities develop.

For the future, optimized treatment requires a close cooperation between PCPs, CHD specialists, and specialized centers for ACHD. Special programs should help educate and inform patients about their disease in order to detect symptoms of PH early and optimize therapeutic management while improving patients' prognosis (31).

State of knowledge

There are considerable deficiencies among the patients' levels of health-related knowledge. While the patients were often familiar with their diagnoses, they were lacking in knowledge about any comorbidities that might be present. The poor state of knowledge regarding thromboembolic events and psychiatric disorders can be explained by two different hypotheses: thromboembolic events are not a common word in regular language, so patients may not have known the meaning and did not know about the diagnosis. The state of knowledge of psychiatric disorders is not truly lacking, but because of the negative stigmata of psychiatric problems, patients did not want to admit to the diagnosis. Psychiatric disorders such as anxiety and depression are very common, affecting 53% of patients with PH, so knowledge in this area needs improvement (32).

Only about half of the patients with manifest PH stated that they suffered from PH. Other studies also revealed significant knowledge gaps in ACHD regarding their disease, although the patients' level of knowledge is considered an important factor in achieving adequate health-related behavior (33,34). Therefore, multidisciplinary counseling is necessary to improve the level of knowledge.

Limitations

The present study recruited a remarkably large sample of ACHD with PH. However, some limitations must be considered when interpreting the current results. The present study is a subgroup analysis of the data from the original VEmaH study. Due to the survey modalities, not all 4,000+ patients could be included in the survey. As participation was voluntary, it is not possible to determine why some patients chose to fill out the study questionnaire while others rejected participation. Patients were permitted to utilize help from another person in completion of the questionnaire, which may bias the data. A selection bias should be considered, as all the questionnaires were handed out at a tertiary care center. Hence, it can be assumed that the study collective differs from the general ACHD population with respect to severity and overall awareness of lifelong necessary follow-up. It can be assumed that, in the real world, ACHD without specific follow-up are even less aware of CHD-related issues and existing ACHD resources, than patients who are in continuous follow-up in a tertiary care center. A key limitation of this study was that participants might have misunderstood medical terminology in the questionnaire, leading them to be misclassified as unaware of a condition, despite our best efforts as outlined in the methods. Finally, the generalization of the conclusions to the primary care of ACHD in other countries is only possible to a limited extent.

Conclusions

PH is an underestimated complication in a broad variety of CHD. Since PH is a common complication with significant morbidity and mortality, all available options should be utilized to improve long-term prognosis through early diagnosis and adequate treatment (21,35). Management

and therapy must always take into account CHD-specific features (7). The presented data show an urgent need for improved awareness among patients and PCPs to facilitate early and correct diagnosis, as well as lifelong, regular follow-up of ACHD with increased risk for or already-existing PH by ACHD specialized cardiologists or pediatric cardiologists. A close collaboration between PCPs and certified ACHD institutions is essential.

To embrace the specific needs of ACHD patients, therapy management should be directed by experienced specialists, ideally in close cooperation with the PCP.

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Supplementary

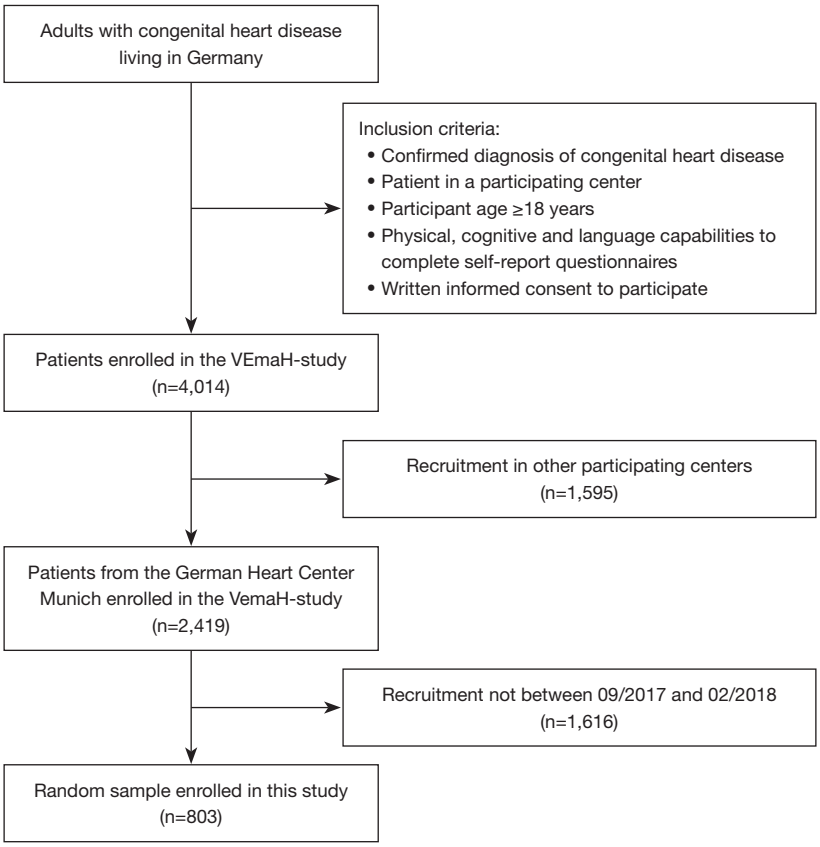

Figure S1 Flow chart for patient selection.

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Appendix 1

VEmaH Questionnaire in its Original Form

VEmaH is an abbreviation for a study conducted throughout Germany. Translated, this stands for "Study on Health Care Provision Research in Adults with Congenital Heart Defects (ACHD)". The present study was the first study to explore the “real world data” on the health status and/or the provision of health care in ACHD. As no standardized and validated questionnaire for this purpose exists, a particular questionnaire was specifically developed in cooperation with the Chair of Behavioral Epidemiology at the Technical University of Dresden and the German Heart Center Munich, a tertiary care center for ACHD. This questionnaire contains questions related to sociodemographic status, underlying congenital heart defect, co-morbidities, details about care providers for medical problems in general and CHD-related problems, individual demands of the patients for counselling, knowledge of specific care structures, and problems with the structure or level of care provided from the patient’s perspective. The questionnaire was completed either directly during the stay at the hospital or online on the homepage of the study (http://www.vemah.info).

The questionnaire, which is only available in German at this time, is reproduced here in its original form:

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Beginn des Fragebogens

1. Ihr **Alter**: _____ Ihr **Geschlecht**: ❑ männlich ❑ weiblich Ihre **Postleitzahl**: _____

2. Sie **leben** in einer:
   - ❑ Großstadt (> 100.000 Einwohner)
   - ❑ Mittelstadt (> 20.000 – 100.000 Einwohner)
   - ❑ Kleinstadt (5.000 – 20.000 Einwohner)
   - ❑ Landgemeinde (< 5.000 Einwohner)

3. Welche Form von **angeborenen Herzfehlern** haben Sie?
   - ❑ Aortenisthmusstenose
   - ❑ Aortenklappenstenose/ Aortenklappeninsuffizienz
   - ❑ Atrioventrikulärer Septumdefekt
   - ❑ Fallot´sche Tetralogie
   - ❑ Hypoplastisches Linksherzsyndrom
   - ❑ Persistierender Ductus Arteriosus Botalli
   - ❑ Pulmonalklappenstenose/ Pulmonalklappeninsuffizienz
   - ❑ Transposition der großen Arterien
   - ❑ Univentrikuläres Herz
   - ❑ Ventrikelseptumdefekt
   - ❑ Vorhofseptumdefekt
   - ❑ Ich habe mehrere Herzfehler, nämlich: _____
   - ❑ Einen anderen Herzfehler, und zwar: _____

4. Leiden Sie an einer der folgenden **Erkrankung**?

Marfan - Syndrom	Ja ❑	Nein ❑	Weiß nicht ❑
Ehlers - Danlos - Syndrom	Ja ❑	Nein ❑	Weiß nicht ❑
Turner - Syndrom	Ja ❑	Nein ❑	Weiß nicht ❑
Morbus Fabry	Ja ❑	Nein ❑	Weiß nicht ❑

5. Leiden Sie unter einer der folgenden **typischen Begleit- oder Folgeerkrankungen Ihres Herzfehlers**?
   - ❑ Herzschwäche
   - ❑ Herzrhythmusstörungen
   - ❑ Herzinnenhautentzündung (Endokarditis)
   - ❑ Koronare Herzerkrankung
   - ❑ Veränderungen im Blutbild
   - ❑ Weiß nicht
   - ❑ Gerinnungsstörungen
   - ❑ Psychische Einschränkungen
   - ❑ Thrombosen
   - ❑ Lungenhochdruck
   - ❑ Neurologische Komplikationen
   - ❑ Nein, ich leide an keiner Begleit-/ Folgeerkrankung

6. Wer ist Ihr **erster Ansprechpartner bei allgemeinmedizinischen/gesundheitlichen Problemen**, die **nicht** in Zusammenhang mit Ihrem Herzfehler gebracht werden, und welche Fachrichtung hat dieser Arzt?
   - ❑ Allgemeinarzt
   - ❑ Praktischer Arzt
   - ❑ Internist
   - ❑ Eine andere Fachrichtung, und zwar: _____

7. Führt dieser niedergelassene Arzt auch eine **Zusatzbezeichnung**? Wenn ja, welche?
   - ❑ Kardiologie
   - ❑ Gastroenterologie
   - ❑ Hämatologie
   - ❑ Angiologie
   - ❑ Pneumologie
   - ❑ Endokrinologie
   - ❑ Rheumatologie
   - ❑ Nephrologie
   - ❑ Keine Zusatzbezeichnung
   - ❑ Weiß nicht
   - ❑ Eine andere Schwerpunktbezeichnung, und zwar: _____

8. Ist diesem **Arzt bekannt**, dass Sie einen **angeborenen Herzfehler haben**?
   - ❑ Ja ❑ Nein ❑ Weiß nicht

9. Wer ist Ihr erster Ansprechpartner bei Problemen in **Zusammenhang mit Ihrem angeborenen Herzfehler**, und welche **Fachrichtung** hat dieser Arzt?
   - ❑ Allgemeinarzt
   - ❑ Praktischer Arzt
   - ❑ Internist
   - ❑ Eine andere Fachrichtung, und zwar: _____

10. Führt dieser niedergelassene Arzt auch eine **Zusatzbezeichnung**? Wenn ja, welche?
    - ❑ Kardiologie
    - ❑ Gastroenterologie
    - ❑ Hämatologie
    - ❑ Angiologie
    - ❑ Pneumologie
    - ❑ Endokrinologie
    - ❑ Rheumatologie
    - ❑ Nephrologie
    - ❑ Keine Zusatzbezeichnung
    - ❑ Weiß nicht
    - ❑ Eine andere Schwerpunktbezeichnung, und zwar: _____

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11. Handelt es sich bei dem Arzt, den Sie bei **allgemeinmedizinischen/gesundheitlichen Problemen** und bei **Problemen in Zusammenhang mit Ihrem angeborenen Herzfehler** aufsuchen, **um denselben Versorger**?
    ❑ Ja ❑ Nein

12. Welchen **Versicherungsstatus** haben Sie aktuell?
    ❑ Gesetzliche Krankenversicherung ❑ Private Krankenversicherung ❑ Keine ❑ Weiß nicht

13. Besteht aus Ihrer Sicht ein Bedarf an **spezifischer Beratung** für Patienten mit angeborenen Herzfehlern bezüglich folgender Themen?

Krankenversicherung	❑ Ja	❑ Nein	❑ Weiß nicht
Lebensversicherung	❑ Ja	❑ Nein	❑ Weiß nicht
Alterssicherung	❑ Ja	❑ Nein	❑ Weiß nicht

14. Welchen **Grad der Behinderung** haben Sie? _____ (in 10er Schritten von 0-100)

15. Besteht aus Ihrer Sicht der **Bedarf an spezifischer Beratung, vor allem** hinsichtlich **Behinderung und folgender Themen**?
    ❑ Ja, bezüglich Rente ❑ Ja, bezüglich Schwerbehindertenausweis ❑ Nein

16. Werden Ihnen regelmäßig **sehr teure Medikamente** verordnet?
    ❑ Ja, Medikamente wegen Lungenhochdruck ❑ Ja, Gerinnungshemmer ❑Nein
    ❑ Weiß nicht ❑ Sonstige, und zwar: _____

17. Hat Ihr Hausarzt **Probleme bei der Verordnung** Ihrer Medikamente? (z.B. auf Grund hoher Kosten?)
    ❑ Ja ❑ Nein ❑ Weiß nicht ❑ Ich nehme keine Medikamente

18. Bitte bewerten Sie mit Schulnoten Ihre aktuelle V**ersorgungslage in Zusammenhang mit Ihrem Herzfehler!**
    ❑ Sehr gut ❑ Gut ❑ Befriedigend ❑ Ausreichend ❑ Mangelhaft ❑ Ungenügend

19. Bitte bewerten Sie mit Schulnoten Ihre aktuelle allgemeinmedizinisch-ärztliche V**ersorgungslage!**
    ❑ Sehr gut ❑ Gut ❑ Befriedigend ❑ Ausreichend ❑ Mangelhaft ❑ Ungenügend

20. Besteht aus Ihrer Sicht ein **Bedarf an spezifischer Beratung** für Patienten mit angeborenen Herzfehlern bezüglich folgender Punkte? Wenn ja, bitte ankreuzen! (Mehrfachantworten möglich)
    ❑ Rehabilitationsmaßnahmen ❑ Bildungsformen (Schule, Studium, Beruf)
    ❑ Berufsfähigkeit ❑ Belastbarkeit im Alltag
    ❑ Führerscheinerwerb ❑ Flugtauglichkeit
    ❑ Leistungsfähigkeit, sportliche Betätigung ❑ Genetische Beratung
    ❑ Ernährung und Bewegung ❑ Prävention
    ❑ Schwangerschaft ❑ Sonstige, und zwar: _____

21. Ist Ihnen bekannt, dass es **zertifizierte Kliniken/ Zentren** für **Erwachsene mit angeborenen Herzfehlern** gibt? (Mehrfachantworten möglich)
    ❑ Ja, und zwar niedergelassene Kinderkardiologen mit EMAH-Zertifizierung
    ❑ Ja, und zwar niedergelassene Kardiologen mit EMAH-Zertifizierung
    ❑ Ja, und zwar zertifizierte EMAH-Schwerpunktkliniken, EMAH-Zentren
    ❑ Nein, mir sind keine zertifizierten Kliniken/Zentren für EMAH bekannt

22. Hat Sie Ihr niedergelassener Arzt in der **Vergangenheit** an eine **EMAH-zertifizierte Institution** überwiesen?
    ❑ Ja, bei kardialen Probleme in Zusammenhang mit meinem Herzfehler
    ❑ Ja, bei Problemen/ Erkrankungen, deren Verlauf von meinem Herzfehler beeinflusst werden kann
    ❑ Nein, ich wurde noch nie in eine EMAH-Institution überwiesen

23. Sind Sie über die **spezifischen Versorgungszentren ausreichend informiert**?
    ❑ Ja ❑ Nein ❑ Weiß nicht

24. Sind Ihnen **Selbsthilfeorganisationen für EMAH** bekannt? (z.B. Bundesverband JEMAH e.V., Deutsche Kinderherzstiftung, Bundesverband herzkranker Kinder e.V., Herzkind e.V.)
    ❑ Ja ❑ Nein ❑ Weiß nicht

25. Sind Sie bereit an einer vertiefenden **Befragung** teilzunehmen?
    ❑ Ja, **bitte kontaktieren Sie mich** unter folgender Emailadresse: _____
    ❑ Nein danke, ich bin an einer Befragung nicht interessiert

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