Research Article

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Outpatient specialist clinics for pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension in the Nordic countries

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Abstract

Pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension are rare conditions that require complex interventions by multidisciplinary teams. The European Society of Cardiology (ESC)/the European Respiratory Society (ERS) 2015 guidelines included recommendations for pulmonary hypertension (PH) referral centers including minimum number of patients, staff, facilities, and network. The aim of the present study was to investigate how the PH-specialist centers in the Nordic countries are presently organized. A descriptive, questionnaire was sent to all PH-specialist centers in the Nordic countries in 2018. Sixteen of 20 PH-specialist centers completed the questionnaire. Seven centers (43%) followed less than 50 patients and three centers (19%) followed 125 patients or more. All had a physician or nurse attending or available at the clinic and eight had support staff such as physiotherapists, counsellors, dieticians, or psychologists directly connected to the center. Twelve centers were available by telephone five days or more per week. Nine centers offered a nurse-led outpatient clinic and of those, six had nurses delegated to make protocol-led changes in pulmonary arterial hypertension-specific treatment. Half of the centers had cooperation with a patient organization. All centers except one used international guidelines to guide care and treatment. More than half of the Nordic PH-specialist centers adhered to the ESC/ERS 2015 guidelines recommendations for volumes and staff in 2018, but there is potential for improvement. However, when formulating recommendations of patient volumes in guidelines, the situation for the geographical large but sparsely populated Nordic countries needs to be considered.

Keywords

access to care, chronic illness, collaborative management, multidisciplinary care, rare diseases

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Introduction

With improvements in medical treatment and the availability of advanced technical equipment for diagnosis and follow-up, the survival rate in some earlier highly fatal diseases has improved.[1] This paradigm shift, from keeping the patient alive to chronic care, demands extensive knowledge in aspects of treatment, care, and advanced technology. It has led to the development of specialist outpatient clinics in several medical areas.[2]

Pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) are both rare, debilitating and life-threatening diseases where new targeted treatments have improved the situation for these patients.[3] At the same time, the care for these patients has become even more complex. Complex investigations at the time of diagnosing as well as side effects and cost of treatment make it crucial that the patients are cared for at designated PH-specialist centers.[3,4] According to recommendations in the European Society of Cardiology (ESC)/the

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Table 1. Facts about the Nordic countries.

Country	Inhabitants (millions)	Area (km^2)	Inhab/km^2	Distance north to south (km)	Human development index	PH-expert centers
Denmark	5.8	42,931	135	368	0.925	2
Finland	5.5	338,449	16	1 157	0.895	5
Iceland	0.3	103,000	3.3	300	0.921	1
Norway	5.3	385,208	14	1 752	0.949	6
Sweden	10.1	447,435	9	1 572	0.913	6

European Respiratory Society (ERS) 2015 guidelines,[3] these specialist centers should involve a multidisciplinary team, including specialist physicians and nurses as well as radiologists, psychological and social work support, and appropriate on-call expertise. To enable more efficient use of resources,[5] and as a response to the patient's desire to have more access to outpatient clinics,[6–8] healthcare has moved toward more outpatient specialist care. Thus, PAH and CTEPH expert teams and specialist outpatient centers are emerging.[9] At these centers, nurses provide information, support, and training to patients and their relatives as well as monitor symptoms and treatment adverse reactions.[9,10]

The ESC/ERS 2015 guidelines also suggest how a PH-specialist center (in guidelines denoted PH referral center) should be organized with regard to a minimum number of patients with PAH or CTEPH followed at the center, number of referrals, facilities, and other services.[3] While this approach toward expert centers with high experience is commendable, its applicability has not been investigated. The Nordic countries exhibit similar socioeconomic status, education levels, and tax financed healthcare systems with a mix of publicly and privately owned healthcare facilities. Together, they have a total population of 27 million people spread over a large surface area (Table 1, Fact sheet).[11] Based on this context, the aim of the present study was to investigate how the PH-specialist centers in the Nordic countries are presently organized.

Materials and methods

Study design and data collection

A descriptive, questionnaire-based study included all PH-specialist centers in the Nordic countries during 2018 (Table 2). A satellite center to Umeå in Sundsvall, responsible for follow-up investigations, was also included.

The questionnaire contained 34 closed, dichotomous or multiple choice questions inspired by other questionnaires[12,13] and was presented in English to all centers.

Information regarding the study, the questionnaire, and a pre-paid return envelope were mailed to a contact nurse at each Nordic PH-specialist center. Following two reminders within three months, a total of 16 (80%) of the PH-specialist centers completed and returned the questionnaires (Table 2). All centers were invited to complete the questionnaires on a voluntary basis and with guaranteed confidentiality. The submission of written answers to the questionnaires was considered as consent.

Results

Participating centers and organization

Sixteen of 20 Nordic PH-specialist centers answered the questionnaires. Fourteen centers were located at university hospitals and two at community hospitals (Table 2). The centers were located at a cardiology clinic (n = 12), a pulmonary clinic (n = 3), or combined cardiology and pulmonary clinic (n = 1) and were started between 1995 and 2018 (Table 2). Nine centers offer a nurse-led outpatient clinic and of those, six had delegated responsibility to nurses to make protocol-led changes in oral PAH-specific treatment and five to change iv, sq, or inhaled prostacyclin dosages (Table 3). Twelve centers could be contacted by telephone five days or more per week, this included voice mail with same-day call-back.

All centers had patients with CTEPH enrolled and all but one also included patients with PAH (Table 3). Three centers included patients from all five WHO PH classification groups (ESC/ERS guidelines 2015). Seven centers had 50 patients or less enrolled at the outpatient clinic and three had more than 125 patients enrolled (Fig. 1). Two-thirds of the centers had 5 outpatient visits or less per week (Fig. 1) and received up to 10 phone calls from patients or relatives every week (Table 3). One center reported more than 30 calls from patients and relatives per week. A physician and nurse was attending or available at all outpatient visits while the availability of other staff groups included in the multidisciplinary team varied between centers (Fig. 2(a)). Nine centers had multidisciplinary meetings at least once a month (Table 3).

Patient care and information

All centers except one reported using international guidelines and/or national guidelines/local directives to guide care and treatment (Table 3). Two centers used both disease-specific and generic patient-reported outcome instruments while five used one or the other.

Table 2. Participating centers and start of PH-specialist centers/outpatient clinics.

Country Center	Start of center (year)	University hospital
Denmark		
Copenhagen[c]	1995	X
Aarhus[a,b]	2000	X
Finland		
Helsinki[a,b,c]	2018	X
Oulo	–	X
Tampere	2007	X
Iceland		
Reykjavík	–	X
Norway		
Tromsø	2004	X
Trondheim	2006	X
Ålesund	1998	
Sweden		
Gothenburg[c]	1999	X
Linköping	2004	X
Lund[c]	2013	X
Stockholm[a]	2007	X
Sundsvall	2014	
Umeå	2005	X
Uppsala[a]	2007	X

Two PH-centers in Norway (Oslo and Bergen) and two in Finland (Kuopio and Turku) did not return the survey.
[a]Performs pulmonary endarterectomy.
[b]Performs balloon pulmonary angioplasty.
[c]Performs heart/lung transplant.

Nurses at all centers provided verbal information about the disease and disease-specific treatment to the patients. Twelve centers used written information to educate patients and eight centers purposely included family or next of kin (Fig. 2(b)). Eight centers included the patient organization in the education while three centers reported having no standardized patient education at all.

Education and research

All centers except one reported that the nurses had received local and/or industry sponsored training about the disease and treatment and three centers also reported disease-specific university training for nurses (Table 3). Seven centers participated in investigator-initiated research studies or clinical trials and 10 centers had designated research coordinators available either as one of the nurses at the PH-specialist center or as a separate position (Table 3).

Discussion

The need for specialist referral centers for investigating and diagnosing PAH and CTEPH as well as for the continued, complex and chronic care for PH-patients on PAH- or CTEPH-specific treatment is obvious. This should include the availability of multidisciplinary teams, possibility to refer patients for advanced care, follow a reasonable number of patients, facilitate advanced diagnostic investigations, and participate in clinical research.[3] In the present study, all participating PH-specialist centers met at least two of the five recommendations and four centers met all

Table 3. Aggregated results from the survey on how PH-specialist centers in the Nordic countries are organized.

PH-diagnoses seen at clinic	
PAH (group 1)	13 (87)
PH at left heart disease (group 2)	7 (47)
PH at lung disease or hypoxia (group 3)	9 (60)
CTEPH (group 4)	14 (93)
PH Other (group 5)	6 (40)
Pediatric patients (<18 years)	2 (13)
Initiate and train patients to use prostacyclin at clinic[a]	13 (81)
Do you cooperate with primary care for PAH-treatment[a]	9 (56)
Nurse-led outpatient clinic[a]	9 (56)
Administrative help for the clinic[a]	14 (88)
Regular (monthly or more often) multidisciplinary meetings[a]	9 (56)
Telephone contact for patient, days/week	
<5 days	3 (20)
5 days	9 (60)
>5 days	3 (20)
Number of telephone calls from patients/week	
<10	10 (67)
11–20	2 (13)
21–30	2 (13)
>30	1 (7)
Use of international guidelines to guide care and treatment[a]	15 (94)
Use of national guidelines/local directives to guide care and treatment[a]	15 (94)
Use of disease specific patient-reported outcome measure[a]	6 (38)
Use of generic patient-reported outcome measure[a]	3 (19)
Use of patient-reported experience measure[a]	4 (25)
Do the nurses have disease specialized education[a]	15 (94)
Local training at hospital	13 (87)
University course/class	3 (20)
Industry sponsored training	11 (73)
Ongoing research studies/clinical trials at clinic	7 (47)
of those—investigator initiated	3 (43)
of those—industry/clinical trial	6 (86)
Is there a research coordinator[a]	10 (63)
Combined with position as nurse at the PH-clinic	4 (40)
Designated research coordinator (separate position)	6 (60)

PH: pulmonary hypertension; PAH: pulmonary arterial hypertension; CTEPH: chronic thromboembolic pulmonary hypertension.
Data presented as number (%).
[a]Includes Iceland.

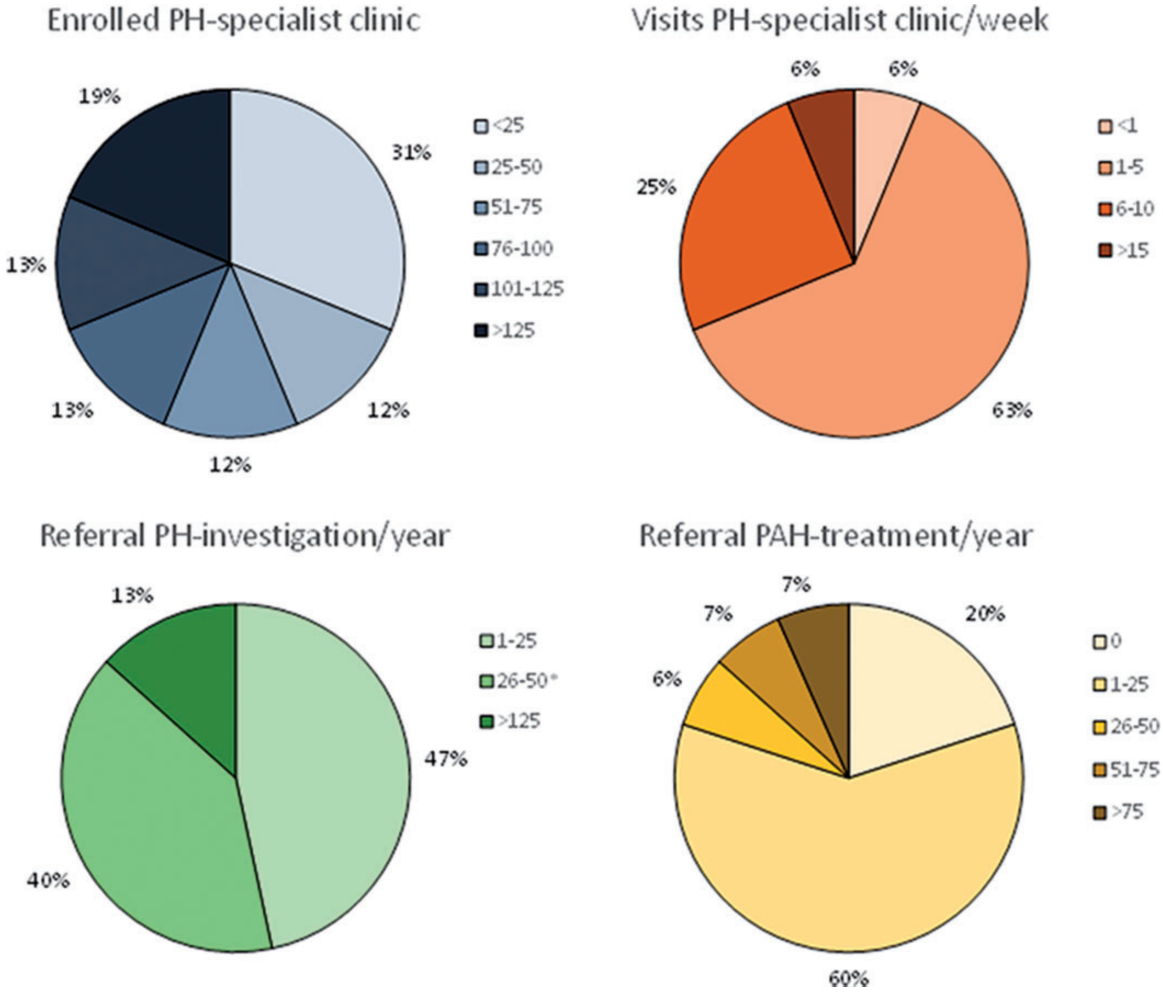

Fig. 1. Number of patients followed at and referred to the Nordic PH-specialist centers.
*no center had 51–124 patients refered to them.

five criteria. Advanced care such as lung transplantation, pulmonary endarterectomy, and pulmonary angioplasty are performed at five, five, and four centers respectively. Due to small volumes, there is collaboration between the Nordic countries and patients can be referred between countries.

All Nordic PH-specialist outpatient centers had a physician or nurse attending or available at the clinic but only half of the centers had other staff groups such as physiotherapists, counsellors, dieticians, or psychologists directly connected to the center. Nine centers had multidisciplinary team meetings at least once a month. The present survey highlights the need for improvement in the organization of the PH-specialist centers in the Nordic countries. But one should keep in mind that the ESC/ERS recommendation[3] was published just three years before this survey and it takes time to change large structures as the healthcare system. Already, more than half of the centers do adhere to the ESC/ERS recommendations for volumes and staff at the PH-specialist centers.

Worldwide estimates of PAH prevalence is 15–60 cases/million inhabitants and the incidence is 5–10 cases/million inhabitants/year; for CTEPH, corresponding numbers are 3 and 1 cases/million inhabitants/year.[3] In Sweden, the prevalence for PAH and CTEPH is 50 and 20 cases/million inhabitants and the incidence is 8 and 2 cases/million inhabitants/year, respectively.[14] The Nordic countries are sparsely populated in respect to the large geographical areas, i.e. the Nordic countries represent 13% of Europe's surface area but only 3.6% of its population (Table 1, Fact sheet),[11] and it is important to keep this in mind when formulating recommendations for future guidelines. Current ESC/ERS recommendation is that a PH-specialist center, in a country with a general population of 10 million or less, should follow a minimum of 50 patients with PAH or CTEPH and receive at least two new referrals per month with documented PAH or CTEPH. In the Nordic countries, 7 of the 16 PH-specialist centers followed less than 50 patients at their clinics and only 3 centers followed 125 patients or more. While volume play an important role in gaining experience with diagnosis, treatment, and care for a rare disease, this needs to be put in perspective of the patients need. In a German study,

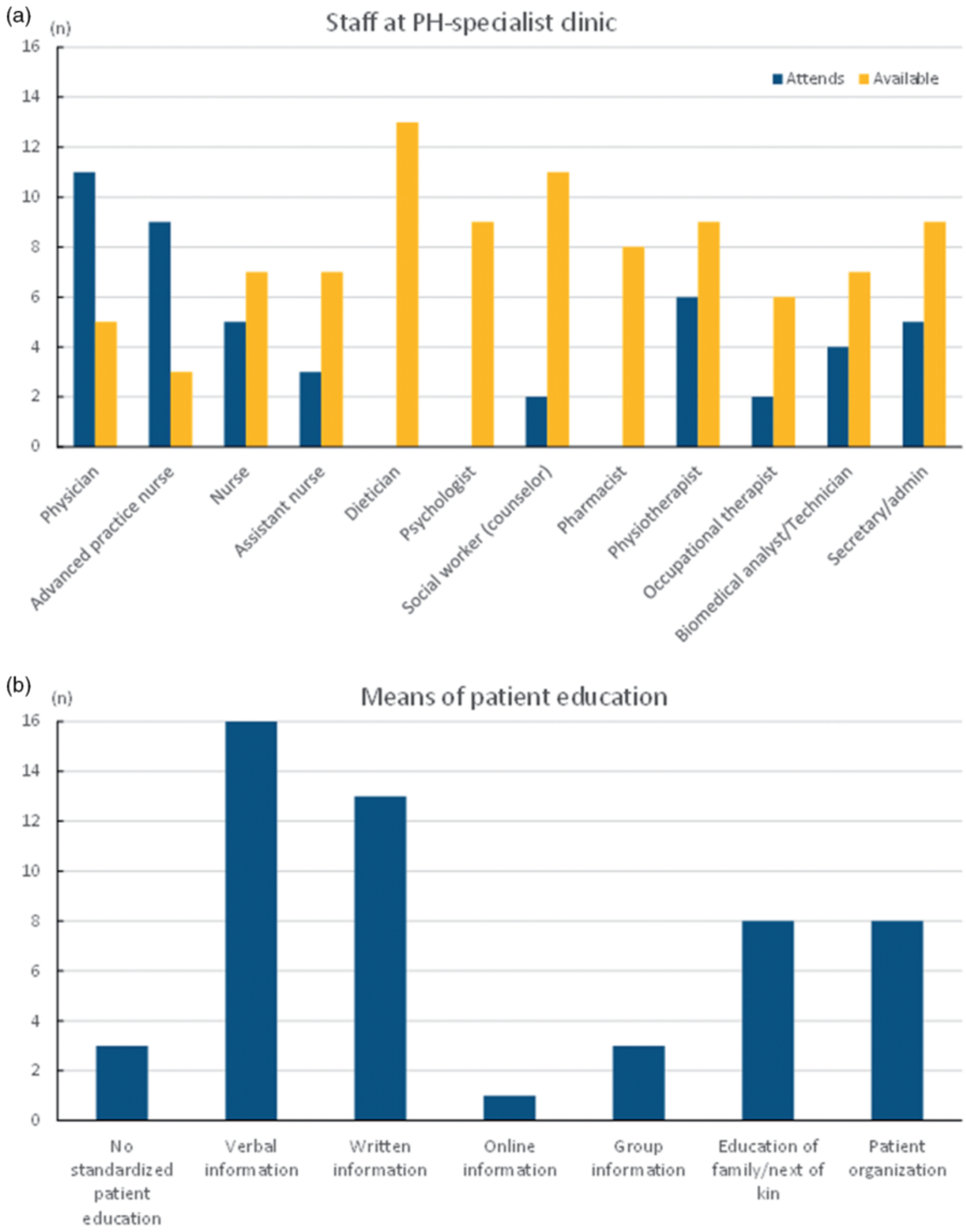

Fig. 2. (a) Staff at the Nordic PH-specialist centers. Bars in blue indicate how many centers that have the staff group attending the clinic and bars in yellow those centers where the staff group is available. (b) Means of patient education provided at the Nordic PH-specialist centers. Each center can provide more than one kind of education.

patients with rare diseases rated the competence of the physician, excellent information, and easy access to medical appliances as the most important factors.[1] In addition, the patients did not consider being treated close to home was important.[1] However, while Germany is a large country, the distance from north to south is about half compared to Norway, Sweden, or Finland (Fact sheet).[11] Thus, other solutions might have to be applied in sparsely populated regions, such as the Nordic countries. Close contact with larger PH-specialist centers, including participating in multidisciplinary team meetings and close consultations using modern technologies, can likely support small expert centers in sparsely populated areas with day to day care of stable patients.

Changes and titration of medication is a task that is normally outside the nurse regular responsibility. In the present study, some nurses had received delegation to perform these tasks. The delegation is personal and based on competence and close collaboration between physicians and nurses at the PH-specialist centers. When nurses at heart failure outpatient clinics adjusted treatment dosages, it was shown to be cost-effective and resulted in fewer hospitalizations.[15,16] One might assume that the same would apply for PH-specialist centers. This is supported by a study from the United

States, where a 58% reduction in hospital admissions from 2001 to 2012 among patients with PAH was attributed to the availability of PAH-specific vasodilator therapies as well as an improvement in the outpatient care of these patients.[17]

In the present times, with the availability of internet and cell phones, we are getting used to instant contact and fast response. This is notable in healthcare as well, where the inability to properly answer the patients' queries by phone directly, as a nonqualified staff answered the calls, made the patients seek care elsewhere.[18] In Sweden, patients have earlier reported difficulties in contacting the PAH/CTEPH team between scheduled visits.[7] Therefore, it is gratifying that the present study shows that in 2018, a vast majority of the Nordic PH-specialist centers nurses could be reached five days a week or more. Notably, investigating the patients experience is a critical step toward improving the quality of care. In the coming years, the use of patient-reported experience measure should be encouraged at the PH-specialist centers.

Disease-specific education of nurses working at the PH-specialist centers was based on local training/experience or industry sponsored. While this training might fulfil the basic needs, it might vary between centers. This indicates that there is a need for standardized specialist training in this field, a problem likely found for many rare diseases that affected small patient populations. The number of staff needing this training, even including the full multidisciplinary team, is likely too small to organize certified training locally or on a country level. However, large organizations such as ESC or ERS might be able to provide certified education as part of their educational programs as they do in other diseases. The importance of integrating patients' perspectives into this education is also important to highlight.[19]

The ESC/ERS 2015 guidelines recommend that a PH-specialist center participate in clinical research, including phase II and phase III clinical trials.[3] While this is a desirable goal, it might be a slightly too optimistic goal considering number of phase II and III trials currently ongoing and the hurdles to gain funding for investigator-initiated studies involving patients with a rare disease. Less than half of the centers in the present survey reported ongoing participation in clinical trials. Maybe a more reachable recommendation and a base for research would be participating in national or international registries.

Limitations

Four centers (20%), two from Finland and two from Norway, did not return the questionnaires despite resending the form and making contact either by mail, phone, or personal meetings at congresses. The form was in English and language might have been a barrier future studies might have to address. The missing information from these centers might have affected the results and the study cannot claim to represent all the PH-specialist centers in the Nordic countries.

Conclusion

More than half of the Nordic PH-specialist centers adhered to the ESC/ERS 2015 guidelines recommendations for volumes and staff in 2018, but the organization can likely be improved to better comply with guideline recommendations. However, when formulating recommendations of patient volumes in future guidelines, the situation for the geographical large but sparsely populated Nordic countries, needs to be considered.

Contributorship

All authors conceived and designed the paper; B.K. and B.I. analyzed the data, wrote the first draft of the manuscript, and contributed to the writing of the manuscript. All authors read and approved the final version of the manuscript.

Conflict of interest

The author(s) declare that there is no conflict of interest.

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