Supplementary material

Kopeć G, Kurzyna M, Mroczek E, et al. Data Base of pulmoNary hyPertension in the PoLish population (BNP-PL) – design of the registry. Kardiol Pol. 2019; 77: 972-974. doi:10.33963/KP.14988.

Methods S1

Study organization and data collection

In each center, one pulmonary hypertension specialist was designated to locally coordinate the study and to be involved in the steering committee. Local coordinators are responsible for patients recruitment and data submission. They are also invited to contribute to data analysis and to give final approval to the results of the project. Since our goal is to describe current practice and outcomes, the registry is entirely observational. All data is acquired from patient records available at the treating centers. The protocol of the study does not require additional patient visits or diagnostic tests, and does not influence the management of patients.

Patient records are submitted by local coordinators to the dedicated internet platform created and managed by the Informatics Centre of the Jagiellonian University Medical College which ensures system integrity and data protection. Only authorized investigator is assigned an individual login and password to enter data into the BNP-PL database. Investigators of one center can view and edit data of patients only from the same center. Several on-line trainings were organized for the investigators to provide a general overview of the registry and to discuss potential factors which could compromise the quality of data collection. Additional explanations have been provided in the BNP-PL accompanying booklet and at the data capture forms in the electronic platform of the registry. The electronic data base automatically identifies

and discards any duplicate entries. Additional queries are generated by the data coordinating center at the Department of Cardiac and Vascular Diseases of the Jagiellonian University Medical College at the John Paul II Hospital in Poland. Sites are monitored for adherence to the protocol and resolution of data queries.

Each patient is assigned a unique patient-identifying number when enrolled in the registry. The personal data of patients enrolled from each center can be known only by investigators from the same center but are anonymized to the other investigators.

Since the accreditation conditions of the National Health Fund are very strict, centers are supervised and controlled. Consequently, the inclusion and treatment criteria must be precisely followed which assures high credibility of the acquired data.

Methods S2

Definition of PAH and CTEPH

Hemodynamic inclusion criteria include the following parameters obtained at right heart catheterization (RHC) at rest: mean pulmonary arterial pressure (mPAP) of 25 mm Hg or more, mean pulmonary arterial wedge pressure (PAWP) or left ventricular end-diastolic pressure (PVEDP) of 15 mm Hg or less, pulmonary vascular resistance (PVR) of > 3 Woods Units (WU), and a clinical diagnosis of PAH or CTEPH according to the clinical investigators' judgments, guided by the definitions of the European Society of Cardiology [S1]. Newly diagnosed patients with mPAP >20 mmHg, pulmonary vascular resistance ≥3 Wood Units and PAWP or PVEDP ≤ 15 mmHg which define pre-capillary PH as proposed at the 6th World Symposium of Pulmonary Hypertension Task Force [S2,S3] may also be recorded in our data base. However this new group of patients will be analyzed separately from the group of CTEPH and PAH patients diagnosed based on currently recommended guidelines.

Supplementary references

S1. Galie N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Heart J 2016;37:67-119.

S2. Simonneau G, Montani D, Celermajer DS, et al . Haemodynamic definitions and updated clinical classification of pulmonary hypertension. Eur Respir J. 2019;53: 1801913.

S3. Frost A, Badesch D, Gibbs JSR, et al. Diagnosis of pulmonary hypertension. Eur Respir J. 2019;53: 1801904.

Table S1. Objectives of the BNP-PL registry

1. To assess the prevalence, clinical characteristics and treatments of Polish adults and children diagnosed with PAH or CTEPH.
2. To evaluate differences in patient outcomes according to specific diagnoses.
3. To evaluate longitudinal changes in PAH and CTEPH epidemiology, clinics and outcomes.
4. To assess effectiveness of novel medical and interventional therapies in PAH and CTEPH.
5. To assess the effects of changing diagnostic criteria of pulmonary hypertension on PAH and CTEPH epidemiology.
6. To evaluate the effects of changing reimbursement policies and novel methods of treatment on therapeutic decisions.
7. To identify clinical predictors of outcomes in PAH and CTEPH patients.
8. To assess outcomes of pregnant women with PAH and CTEPH.
9. To initiate prospective studies and retrospective analysis of patients with PAH and CTEPH.

CTEPH - chronic thromboembolic pulmonary hypertension; PAH - pulmonary arterial hypertension

Table S2. Major variables collected in the BNP-PL registry for pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) patients at study entry.

Parameter	Specific categories
Date of birth	DD/MM/YYYY
Sex	Male or female
Race	Specify
Zip code	Specify
Involvement in clinical trials	Blinded/Open label
Diagnosis	Date/List according to the ESC guidelines
Patient's status	Incident/prevalent
Anthropometric data (current)	Weight, Height
Major comorbid conditions (history)	List
Major medical procedures (history)	List
Pregnancy	Yes/No
WHO functional class (diagnosis, enrollment)	I, II, III, or IV
Targeted medications (current)	List
Concomitant medications (current)	List
Laboratory measurements (current)	List
6-minute walk test (current)	Distance and Borg dyspnea score
Echocardiography (current)	List
Right heart catheterization (current)	List
ECG (current)	Major abnormalities
Socioeconomic status	Education/Work/Dependence on others

Data acquired in incident patients only	
First symptoms	List/date
First medical contact	Specialization/date
Data acquired in CTEPH patients only	
Status	Incident/Prevalent/on medical treatment/after BPA/after PEA/combinations
Diagnostic tests results	Ventilation perfusion scan, Computed tomography of PA, Arteriography, Deep vein ultrasonography
CTEPH team decision	Operable/ non-operable/ recommended treatment
Thrombophilia	Yes/No/Specify

BPA – balloon pulmonary angioplasty, ESC -European Society of Cardiology, PA – pulmonary artery, PEA – pulmonary endarterectomy, WHO – World Health Organization

Table S3. Parameters collected at follow-up visits

Death	Yes/No
Cause	List
Place	Hospital/Non-hospital
Autopsy	Yes/No/result
Hospitalization	Yes/No
Cause	List
Result	Discharge/Death

Atrial septostomy	Yes/No
Potts procedure	Yes/No
Correction of congenital heart defect	Yes/No/Type of operation
Lung transplantation	Yes/No
Change in pharmacotherapy	Yes/No; if Yes: active substance, dose, administration route, start and stop dates
Treatment side effects	Yes/No/List
Pregnancy	Yes/No
Enrollment to clinical study	Yes/No
Progression of symptoms	No/Slow/Rapid
Syncope	Yes/No
Clinical signs of right heart failure	Yes/No
New major comorbid conditions	List
New major medical procedures	List
Referral to lung transplantation centre	Yes/No
Listing for lung transplantation	Yes/No
Electrophysiology procedures	Yes/No/Type/Efficacy
Diagnostic tests (if performed)	
Laboratory tests	List
Echocardiography	List
Right heart catheterization	List
Anthropometric data	weight/height
In CTEPH patients only	
BPA	Yes/No; List

PEA	Yes/No; List

BPA – balloon pulmonary angioplasty, CTEPH – chronic thromboembolic pulmonary hypertension, PEA – pulmonary endarterectomy