Database of Pulmonary Hypertension in the Polish Population (BNP-PL): design of the registry

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Introduction Pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) are rare types of pulmonary hypertension (PH). Consequently, patient registries are key instruments that provide data for clinical research and improve patient care and healthcare planning.1

Currently, most of our knowledge on PAH and CTEPH epidemiology, management, and treatment outcomes come from registries originating in the Western populations.2-5 However, a global view on the epidemiology of PH reveals important geographical differences.1

The Database of Pulmonary Hypertension in the Polish population (Baza Nadciśnienia Płucnego [BNP-PL]) (ClinicalTrials.gov identifier, NCT03959748) is the first multicenter and prospective registry of adult and pediatric patients with PAH and CTEPH created in any of the Central-Eastern European countries. In the present report, we describe the design of the registry.

Objectives of the BNP-PL registry The BNP-PL registry is an initiative of the Working Group on Pulmonary Circulation of the Polish Cardiac Society in cooperation with Polish PH reference centers6-9 to assess prospectively the epidemiology, clinical course, and disease management of patients with PAH and CTEPH. The specific objectives of the study are shown in Supplementary material, Table S1.

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Methods  Organization of the BNP-PL registry  An invitation to participate in the study was sent to Polish centers (22 adult and 8 pediatric centers) that were accredited to treat PAH or CTEPH by the Polish National Health Fund (Narodowy Fundusz Zdrowia, NFZ), the only health-care payer in Poland. Of the invited centers, all of the adult and pediatric centers accepted the invitation and were formally involved in the project. Design of the registry is shown in Figure 1 and is further described in Supplementary material, Methods S1.

The protocol of the study was reviewed and accepted by the Bioethical Committee of Physicians and Dentists Chamber in Kraków.

Participants  Patients with PAH and CTEPH older than 3 months of age and treated in the participating centers are enrolled to the BNP-PL registry to form 4 separate arms as shown in Figure 1A. Patients with both newly and previously diagnosed PAH or CTEPH are eligible. Newly diagnosed is defined as diagnosis established after March 1, 2018 (termed “incident cases”). Patients who were diagnosed earlier are classified as “prevalent cases” (Figure 1B). For the purpose of the study, we use the most recent definitions of PAH and CTEPH as recommended by the European Society of Cardiology (Supplementary material, Methods S2).10

Patient records are submitted by local coordinators to a dedicated internet platform created and managed by the Informatics Centre of the Jagiellonian University Medical College, which ensures system integrity and data protection.

Cohort size and study duration  Enrollment to our study started on March 1, 2018 and is planned to continue at least until February 28, 2023. We plan to collect the follow-up data for a minimum of 5 years from the enrollment of the last patient. As our registry is mostly descriptive, the study size is not prespecified.

Prevalence and incidence  The prevalence of PAH and CTEPH will be calculated separately for adults (≥18 years of age) and children (≥3 months and <18 years of age) as the number of cases per 1,000,000 inhabitants, and the incidence, as the number of new cases of PAH or CTEPH per 1,000,000 inhabitants each year. Data for the number of adults and children living in Poland will be acquired from the publications of Statistics Poland (https://stat.gov.pl)

Baseline assessment  In incident cases, the baseline assessment includes data acquired at the diagnosis of PAH or CTEPH, including the first prescribed treatment (Supplementary material, Table S2). We also collect information about the first symptoms and the time elapsed from the first symptoms to diagnosis. In prevalent cases, the baseline assessment includes data obtained at the most recent visit after March 1, 2018 and also from the most recent right heart catheterization. We also record the date of diagnosis and patient’s functional class at that time. The date of PAH or CTEPH diagnosis is defined as the date of the first right heart catheterization11 fulfilling the hemodynamic criteria for precapillary PH.

Follow-up assessment  Follow-up data (Supplementary material, Table S3) will be recorded in the database every year between September 1 and October 30, starting in 2019. Accordingly, follow-up data will include the most recent information on patients’ status before September 1 each year, including changes in treatment, major clinical events and outcomes, and results of additional tests performed in a patient in the last 12 months.

Treatment  Medical therapy in patients with PAH and CTEPH in Poland is reimbursed by the NFZ in a structured program with specific inclusion and exclusion criteria. This program is subject to change along with new drug approvals.

Discussion  Bias and generalizability of the results  Enrollment of both prevalent and incident patients may bias the outcome analysis as data on patients with prevalent cases who did not survive until the study enrollment will not be

Figure 1  Design of the Database of Pulmonary Hypertension in the Polish Population (BNP-PL) registry. A – four arms of the BNP-PL registry including adults and children with pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH). Two additional arms of the CTEPH group represent patients treated with balloon pulmonary angioplasty (BPA) and pulmonary endarterectomy (PEA). B – time points of enrollment and follow-up of prevalent and incident cases.
Comparison with other registries on pulmonary hypertension

In their recent systematic literature search, Skride et al. identified 11 prospective registries performed in patients with PH from Europe. Most of them were national (4 single-center and 6 multicenter registries) and only 1 with CTEPH were followed for a median of 33 months.

Due to specific organization of treatment of patients with PAH and CTEPH in Poland in centers accredited by the NFZ, we believe that we will enroll almost all affected individuals in our analysis. In that way, our results will be representative of the recently diagnosed patients with PAH or CTEPH and of patients who lived with PAH or CTEPH long enough to be represented in the registry. Still, our data will not be generalizable to patients with PAH or CTEPH who have not been yet diagnosed.

Conclusions

The BNP-PL registry is designed to show the epidemiology and characteristics of PAH and CTEPH in a large Central-Eastern post-communist European country with a relatively short history of availability of modern PAH diagnostics and therapies.

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST

None declared.

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