Prevalence and incidence of pulmonary arterial hypertension: A systematic review of the literature for the Global Burden of Disease Study

Authors: Sophia Emmons-Bell, Alie Boon Dooley, Catherine Johnson, Gregory Roth

**Background:** The Global Burden of Disease (GBD) study is a multinational effort to produce comparable, consistent measures of disease burden for populations, but pulmonary arterial hypertension (PAH) is not included. An increasing number of studies describe the epidemiology of PAH. We sought all data on population-based prevalence, incidence, and mortality to estimate the global burden of PAH for the GBD study.

**Methods:** We searched the Global Index Medicus (GIM), including PubMed and other databases, for key words between 1980 and 2020, identifying population-representative prevalence, incidence, and mortality rates of clinically diagnosed PAH supported by diagnostic testing. We excluded PAH groups 2-5, referral center cohorts, or study samples less than 50.

**Results:** The search returned 10,098 hits, of which 56 were included: 13 for prevalence, 16 for incidence, and 50 for case fatality rate (23 contained >1 measure). The mean age of patients was 47 years, with 67.5% female. 27 countries from 6 GBD super-regions were represented. Reported prevalence ranged from 0.37 cases/100,000, derived from a referral center of French children, to 4.9 cases/100,000, derived from a Swedish register. Reported incidence ranged from 0.07 cases/100,000 person-years, derived from a study in the UK and Ireland, to 0.56 cases/100,000 person-years, derived from an Israeli clinic. All studies with sex-specific estimates reported higher levels in females than males. Studies varied in their approach to reporting age-sex categories, and in sampling procedures.

**Conclusions:** Reported PAH prevalence, incidence, and mortality rates varied widely by location and study. Harmonization of methods for PAH registries would improve efforts at disease surveillance. Results of this search can be used to inform planned GBD estimation of PAH, and help to quantifying its global burden, thereby guiding global efforts to prioritize and treat this often-overlooked condition.