

**AUTHOR:**Gerald J. Maarmarman¹ **AFFILIATION:**¹CARMA: Centre for Cardio-Metabolic Research in Africa, Tygerberg Medical School, Stellenbosch University, Cape Town, South Africa**CORRESPONDENCE TO:**

Gerald Maarmarman

EMAIL:

gmaarmarman@sun.ac.za

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A philosophical perspective on pulmonary hypertension: What is 'rare'?

Pulmonary hypertension (PH) is a fatal disease and public health concern.¹ The global prevalence of PH is not known¹ and a major focus is to establish registries in order to determine the actual prevalence of PH per country². PH prevalence is largely subject to aetiology, geographical region and the tools used to make a diagnosis (e.g. echocardiography or right heart catheterisation).¹ In Africa, the prevalence of PH that is secondary to HIV differs from its prevalence that is secondary to rheumatic heart disease or schistosomiasis. For example, PH prevalence in HIV is approximately 14%³, while the prevalence can be 1% or 10% in schistosomiasis⁴. In comparison with a world population of some eight billion people, the relatively 'low' number of people who have been diagnosed with or who have succumbed to PH has triggered the assumption that it is a rare disease, which is how it is also reported throughout the literature.

However, let us consider the following. In one study of 277 people living with HIV, 18 were diagnosed with PH.⁵ In a recent systematic review and meta-analysis of studies with 42 642 people living with HIV from 17 countries, the overall PH prevalence was 8.3% in adults – a total of about 3 540 people with PH.³ Surely this is not 'rare' disease if one considers that this is ultimately the number of people who might die due to PH? To add an interesting dimension, during the 2019 festive season in South Africa, approximately 600 people died in motor accidents (based on public reports of the National Minister of Transport). The count of these deaths is always met with shock, sadness, and calls for urgent action for prevention – and rightly so. However, that fewer deaths are reported for car accidents compared to other causes (e.g. infectious diseases that kill millions of people), does not make the one 'rare' and the other not. More importantly, simply because fewer people have died from one disease than from another, does not make that one disease less important. PH causes morbidity and mortality, period, and should be considered a formidable health threat.

PH is a clinical complication of many diseases that are highly prevalent in South Africa. Altogether, millions of South Africans suffer from heart disease, tuberculosis, HIV and schistosomiasis.⁶ Thus, many people who currently have these diseases may later develop and succumb to PH, especially considering that the symptoms of PH are often misperceived, and patients may remain undiagnosed or are diagnosed too late. This may be considered an oversimplification or romanticised exaggeration of a poorly understood reality. However, the matter should be settled: PH is not a 'rare' disease⁷ because it has already cost many lives and, from a patient's perspective, the identification of a disease as 'rare' is irrelevant, because their own clinical reality takes precedence⁸. From a philosophical viewpoint, I believe that we should challenge the persistent portrayal of PH as 'rare'.

Suggesting that PH is a rare disease has negative consequences for PH research and public health efforts. The use of the term 'rare' may create several negative impressions in the PH research/clinical sector, two of which I highlight here. First, describing PH as a 'rare' disease may create the impression that more research funding should not be funnelled towards PH research. Second, it can create the erroneous impression that it is not necessary to make targeted therapy available and affordable to all patients (those with and without medical aid insurance, and those in developing countries). As the clinical and research community, and other stakeholders, we should begin to consider that PH might become a greater health concern in South Africa, given the high prevalence of diseases or risk factors, like heart disease, tuberculosis, HIV and schistosomiasis. It is perhaps time that we reflect on this matter with due diligence and we ask ourselves this important philosophical question: *What is rare?*

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