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Editorial

Pulmonary Hypertension in Indonesia: An Urgent Call to Close the Gaps in Diagnosis and Care

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Abstract

Pulmonary Hypertension (PH) is a progressive disease marked by elevated pulmonary arterial pressure greater than 20 mmHg at rest, as measured by right heart catheterization. Despite the advancement of diagnostic and treatment in PH, diagnostic delay has been a challenge in Indonesia due to a lack of PH awareness, a fragmented referral system, limited diagnostic facilities, and PH-specific therapy, which leads to high mortality and rehospitalization. Guideline-directed combination therapy of PH is markedly constrained, with only PDE-5i and oral prostacyclin analogues covered by the Indonesian National Health Insurance (BPJS). True prevalence and incidence remain unknown nationally. Community support, a patient-based organization, also plays a crucial role in raising PH awareness, including promoting therapy adherence among PH patients. Creating PH centers is also in urgent need to execute an integrative and comprehensive management of PH. To close the gaps of diagnosis and care of PH patients, Indonesia should establish nationwide PH registries, expand access to echocardiography and catheterizations, broaden BPJS coverage on PH combination therapy, and develop PH centers and strengthen *Yayasan Hipertensi Pulmonal Indonesia* (YHPI, Pulmonary Hypertension Association of Indonesia) on addressing social burdens of PH patients.

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Introduction

Pulmonary hypertension (PH) remains a complex, progressive syndrome marked by elevated pulmonary arterial pressure and high morbidity and mortality. Diagnostic delay is a global issue, but its magnitude in Indonesia is especially alarming. Despite major advances in diagnostics and therapy worldwide, Indonesia continues to face structural barriers that cause significant delays in PH detection and treatment. International studies demonstrate that PH is frequently diagnosed 2–4 years after symptom onset[1], with a strong association with more advanced right ventricular dysfunction, higher mortality, and diminished long-term survival.[2] In Indonesia, these challenges are exacerbated by fragmented referral systems, a lack of routine screening, limited catheterization facilities, and restricted access to therapeutic options. Local data from the COHARD-PH registry, Indonesia's first adult congenital heart disease-related (CHD) PH registry, confirms that 77% of adults with CHD show echocardiographic signs of PH at first presentation, of which 66.9% are confirmed as pulmonary arterial hypertension (PAH) by right-heart catheterization (RHC).[3] These data reflect decades of underdiagnosis and delayed PH referral in children and adults.

Definition and Classification

Pulmonary hypertension is defined hemodynamically as a mean pulmonary arterial pressure (mPAP) greater than 20 mmHg at rest, measured by RHC.[4-5] The current clinical classification classifies PH into five groups: (1) PAH; (2) PH associated with left heart disease; (3) PH associated with lung disease and/or hypoxia; (4) PH associated with pulmonary artery obstruction with Chronic Thromboembolic Pulmonary Hypertension (CTEPH) as its core; and (5) PH with unclear or multifactorial mechanisms. While this classification is universal, Indonesia faces difficulties with the classification due to limited diagnostic facilities.

Group 1 PAH in Indonesia is dominated by CHD-associated PAH. In the COHARD-PH registry, which enrolled 1,012 adults with septal defects and shunt lesions over a seven-year period, the majority of congenital heart lesions were secundum Atrial Septal Defects (ASD; 73.4%). Among patients with CHD-related PAH, 89.3% had ASD and 18.7% had already progressed to Eisenmenger syndrome at the time of diagnosis.[3] These data underscore the consequences of late CHD recognition and missed opportunities for early defect closure. Another important spectrum of group 1 PAH is idiopathic PAH, heritable PAH, associated PAH with drug and toxin, persistent PH of the newborn, and associated with several diseases, including connective tissue disease, HIV infection, portal hypertension, and schistosomiasis. Most of them are difficult to detect, either due to limited facilities for genetic and supported examination, as well as their high cost.

Group 2 PH is likely the most common PH subtype in Indonesia, given the high prevalence of hypertension, coronary disease, rheumatic heart disease, and heart failure. International data report 50-70% of PH prevalence in HFpEF and valvular heart disease.[4] However, systematic data are lacking, and few cardiac registries in Indonesia exclude this group from their registry.[6] This results in under-recognition of group 2 PH and limited understanding of its true burden.

Group 3 PH is highly relevant in Indonesia because of the country's substantial burden of Tuberculosis (TB), post-TB lung disease, Chronic Obstructive Pulmonary Disease (COPD), and interstitial lung disease. Post-TB lung disease, in particular, is increasingly recognized globally as a contributor to PH, yet PH screening in this population is not routinely performed in Indonesia.[6] Local data from Persahabatan Hospital reports that there were 30% PH among COPD patients examined by echocardiography, which needs to be confirmed by RHC.[7] This data is in line with our unpublished data of 14% TB-related-PH incidence in our hospital. Moreover, Hoeper et al reported a fairly large proportion of 30-48% of group 3 PH across the globe.[8] Therefore, we believe that this group might potentially become the largest number of PH in Indonesia.

Group 4 CTEPH remains vastly underestimated. International registry reports prevalence of 26-38 cases/million adults.[4] Ventilation–perfusion (V/Q) scanning, the key screening tool for CTEPH, is not widely available, and high-quality CT pulmonary angiography is limited to selected tertiary hospitals. Moreover, there is a lack of experts who are able to interpret these advanced imaging techniques. As a result, many potentially treatable CTEPH cases are never recognized.[6]

Epidemiology

Diagnostic delay is a major epidemiological challenge. In a large multinational survey, Small and colleagues reported a mean total delay of 17 months from first PAH symptoms to confirmed diagnosis, with patients seeing nearly three physicians on average and over 40% receiving at least one misdiagnosis.[1] Misdiagnoses frequently included asthma, COPD, and heart failure, reflecting the non-specific nature of PH early symptoms and limited awareness of PAH among primary care physicians.

The Indonesian situation is likely worse. In the COHARD-PH registry, adults with CHD typically presented in their mid-30s; a large proportion had been asymptomatic for decades and only sought care once dyspnea on exertion, fatigue, or cyanosis became limiting.[3] At first enrollment, 77.1% already had echocardiographic signs of PH, and 66.9% met invasive hemodynamic criteria for PAH.[3] These data point to a large, long-standing reservoir of undetected PAH that only becomes visible when the disease is advanced.

Nationally, the true prevalence and incidence of PH and PAH remain unknown. Indonesia does not yet have a nationwide PH registry, and the national health survey (RISKESDAS) does not include PH indicators.[4] Existing data come predominantly from single-center cohorts, such as COHARD-PH, and a limited number of specialized referral hospitals. Another single-center unpublished cohort registry in Universitas Indonesia Hospital reported 167 PH patients with distribution of 35% group 1 PH, 32% group II PH, 25% group III PH, and 7,1% group IV PH. A coordinated national registry effort by the Indonesia Pulmonary Hypertension (INA-PH) working group is currently taking place.

Diagnostic Limitations

Right-heart catheterization remains the gold standard for diagnosing and phenotyping PH for guiding treatment decisions. In Indonesia, however, access to RHC is limited to a small number of tertiary centers, and even in those, catheterization laboratories are often dominated by coronary interventions.[3,6] Facilities to measure pulmonary vascular resistance, perform vasoreactivity testing, and conduct serial hemodynamic monitoring are not uniformly available.

Transthoracic echocardiography is the main screening tool for PH, but access is uneven across health facilities in Indonesia. Operator expertise varies, and standardized PH-focused protocols, as recommended in international guidelines, are not consistently used.[6] Many patients with exertional dyspnea in primary care settings never undergo echocardiography at all.

Comprehensive risk stratification requires biomarkers such as NT-proBNP, exercise testing (6-minute walk distance), and detailed imaging of right ventricular structure and function, in addition to RHC hemodynamics. In the COHARD-PH registry, NT-proBNP was significantly higher in CHD-PAH compared with CHD without PAH (median 774 vs 121 pg/mL), and worse WHO functional class correlated with lower oxygen saturation, shorter walking distance, and more adverse hemodynamics.[3] Yet NT-proBNP assays and structured 6-minute walk testing are still not universally available across Indonesian centers.

Mortality

Pulmonary hypertension mortality varies across the globe. Recent global trend of PH mortality from 54 countries during 2001 to 2019 reports an average of 3-year crude mortality (per 100.000) of 1.6, with Georgia as the highest death of 16.51 and Nicaragua as the lowest of 0.13. This study also reported a global reduction of 3 years' crude mortality from 2.1 in 2001 to 1.6 in 2019. This reduction is due to the advancement of PH diagnosis and treatment. However, delayed diagnosis has direct prognostic implications for mortality. In a recent study of PAH patients, Kubota and colleagues demonstrated that longer time from symptom onset to diagnosis correlated with higher BNP, worse right ventricular function indices, and a significantly higher rate of death or PAH-related hospitalization during follow-up.[2] Patients diagnosed within three months of symptom onset were more likely to have lower mortality compared to those with delayed diagnosis.[2] These findings echo the broader international experience that untreated or late-treated PAH carries a poor long-term survival, particularly for patients diagnosed in WHO functional class III–IV. In Indonesia, CHD-PAH patients with worse functional class in COHARD-PH had lower oxygen saturation, shorter walking distance, higher NT-proBNP, and more severe hemodynamic derangements[3], all of which are established predictors of mortality.

Studies from the United States and Europe high-

light the superiority of apprenticeship-based subspecialty education in achieving procedural competency and in preparing for independent practice.[6,13] The Accrediation Council for Graduate Medical Education (ACGME) and the European Society of Cardiology (ESC) frameworks require fellows to demonstrate mastery in procedural skills, clinical decision-making, and academic output before board certification.

Management

International guidelines recommend a risk-stratified approach to PAH therapy, with early use of combination regimens that target the endothelin, nitric oxide, prostacyclin pathways, and Activin-A signal inhibition. For most of the newly diagnosed patients, upfront dual therapy with an Endothelin Receptor Antagonist (ERA) plus a Phosphodiesterase-5 inhibitor (PDE5i) is now standard, whereas high-risk patients may benefit from initial triple therapy.

In Indonesia, however, therapeutic options are markedly constrained. There are two PAH-targeted drugs that are widely available, including a PDE-5 inhibitor (sildenafil) and oral prostacyclin analogues (beraprost), which fortunately currently reimbursed by the Indonesian National Health Insurance (BPJS). Inhaled prostacyclin analogues (iloprost) and ERA agents such as macitentan and ambrisentan require out-of-pocket payment.[6] Even further, ambrisentan is currently only accessible through a limited special-access program in Universitas Indonesia Hospital. These constraints prevent many Indonesian patients from receiving guideline-directed combination therapy.

Despite these limitations, local studies have shown that sildenafil-based regimens can improve symptoms, functional capacity, and quality of life in CHD-related PAH.[4] Structured cardiopulmonary rehabilitation and supervised exercise programs have also been shown to add benefits when combined with pharmacologic therapy.[6] Scaling these relatively low-cost interventions could provide significant improvements in outcomes, even before broader drug access is achieved.

Several types of PH patients might benefit from an interventional strategy. Patient with CHD should undergo their respective CHD repair if possible. However, they might miss the window of repair due to the development of CHD-PAH, which, unfortunately, is in line with the trend of more severe forms of CHD-PAH findings in COHARD-PH. Therefore, early screening for CHD is mandatory to prevent the development of CHD-PAH in the future.

Another type of PH that might benefit from an interventional strategy is group IV CTEPH. Robust international data revealed that the surgical procedure of Pulmonary Endarterectomy (PEA) and Interventional Balloon Pulmonary Angioplasty (BPA) reduced pulmonary vascular resistance and improved CTEPH mortality and morbidity.[10] Pulmonary endarterectomy is indicated for CTEPH with proximal thrombus in the main, interlobar, or proximal part of the segmental pulmonary artery, while BPA is indicated for cases with difficulty in performing PEA or segmental-subsegmental pulmonary artery. Despite these positive interventional outcomes, Indonesia is not yet able to offer these interventional strategies due to limited facilities and interventional experts.

Community Services and PH Centers

Beyond drugs and diagnostics, community and institutional structures play a crucial role in PH care. In Indonesia, the patient-based organization *Yayasan Hipertensi Pulmonal Indonesia* (YHPI) has emerged as an important partner in raising awareness, supporting patients and families, and promoting adherence and follow-up.[6] Such organizations help address the psychosocial burden of a chronic, life-limiting disease that is often diagnosed late.

At the health-system level, there is an urgent need to develop and accredit dedicated PH centers of excellence. International guidelines recommend that PH patients be managed in expert centers with access to RHC, advanced echocardiography, V/Q scanning or high-resolution CT, biomarker testing, exercise testing, and a multidisciplinary team including cardiology, pulmonology, rheumatology, imaging, and rehabilitation. Currently, only a handful of Indonesian institutions approach this standard.

Conclusion

Pulmonary hypertension in Indonesia remains underdiagnosed, underestimated, and undertreated. Structural limitations in diagnostic capacity and restricted access to PAH-specific therapies further amplify the burden.

Indonesia now has an opportunity and a responsibility to close these gaps. Priorities include establishing nationwide PH registries, integrating

PH indicators into national health surveys, expanding access to echocardiography and RHC, hopefully broadening BPJS coverage to include combination PAH therapy, developing PH centers of excellence, and strengthening collaboration with patient organizations. With coordinated action from clinicians, policymakers, and patient groups, it should be possible to shorten the diagnostic journey, improve access to effective treatment, and ultimately change the trajectory of PH in Indonesia.

List of Abbreviations

ABIM	American Board Internal Medicine
ACGME	Accrediation Council for Graduate Medical Education
ASD	Artial Septal Defects
BPA	Balloon Pulmonary Angioplasty
BPJS	Indonesian National Health Insurance
COPD	Chronic Obstructive Pulmonary Disease
CHD	Congenital Heart Disease
CTEPH	Chronic Thromboembolic Pulmonary Hypertension
ERA	Endothelin Receptor Antagonist
ESC	European Society of Cardiology
INA-PH	Indonesia Pulmonary Hypertension
PAH	Pulmonary Arterial Hypertension
PDE5i	Phosphodiesterase-5 inhibitor
PEA	Pulmonary Endarterectomy
PH	Pulmonary Hypertension
RHC	Right-Heart Catheterization
RISKESDAS	National Health Survey
YHPI	Pulmonary Hypertension Association of Indonesia

Conflict of Interest Statement

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Authors acknowledge that Artificial Intelligence (AI) tools were only used to assist in language editing and did not generate or alter the scientific content, analyses, or conclusions presented in this manuscript.

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