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PULMONARY HYPERTENSION STILL AN 'ORPHAN LUNG DISEASE' IN PAKISTAN

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Contribution
FA conceived the idea of editorial and MIK conducted literature review, and drafted manuscript.

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In our country when it comes to diagnosis and treatment of Pulmonary Hypertension (PH) especially Pulmonary Arterial Hypertension (PAH), it seems that it is still an Orphan Lung disease. Whether it is data about its prevalence in our country, available diagnostic services or treatment options, one may easily feel the scarcity in all mentioned areas.

Most current classification of PH, categorizes it into 5 groups.[1] However this categorization requires availability of diagnostic tools especially right heart catheterization data to classify and know the severity of PH and to offer targeted treatment to patients who belong to group 1 PH i.e. Pulmonary Arterial Hypertension (PAH).

Studies done about PH prevalence in Pakistan have mainly focused on data in selected patients group e.g. in patients with Chronic Obstructive Pulmonary Diseases (COPD) or Systemic Lupus Erythematosus (SLE).[2,3] All of these studies relied on diagnosis without using the diagnostic bench mark i.e. Right heart catheterization. Moreover, effects of targeted treatment for PAH have also been studied only in selected group of patients e.g. COPD.[4,5]

While above work by the learned colleagues has to be appreciated, it emphasizes the need to,

- Spread the awareness about PH more so to reduce inadvertent use of targeted treatment which may prove detrimental outside group 1 PH and should only be considered at specialized centers.
- Develop specialized centers with required diagnostic services especially Lung function and anatomical evaluation and Right heart catheterization.
- Establish PH registries (at least one in each province) which can play a vital role to collect data and ensure effective evidence based patient management.

This may be an uphill task however the right direction to move forward.

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