

## CAUSATIVE STUDY OF PULMONARY ARTERIAL HYPERTENSION

Mayanglambam Bijoy<sup>1</sup>, Nameirakpam Dhanachand Singh<sup>2</sup>, Thounaojam Rameshchandra<sup>3</sup>, Thokchom Sachin Deba Singh<sup>4</sup>, Salman Chambugong Marak<sup>5</sup>, P. Shamjeih Phom<sup>5</sup>, Posa Vishnu Theja<sup>5</sup>, Johny Lalmanpuia Tlau<sup>5</sup>

Received : 20/09/2024  
Received in revised form : 04/11/2024  
Accepted : 19/11/2024

**Keywords:**

Echo Doppler, Dilated Cardiomyopathy, Chronic Obstructive Pulmonary Disease, Cor Pulmonale.

Corresponding Author:

**Dr. Salman Chambugong Marak,**  
Email: [saldiegomaraq@gmail.com](mailto:saldiegomaraq@gmail.com)

DOI: 10.47009/jamp.2024.6.6.68

Source of Support: Nil,  
Conflict of Interest: None declared

*Int J Acad Med Pharm*  
2024; 6 (6); 346-348



<sup>1</sup>Associate Professor, Department of Medicine, Regional Institute of Medical Sciences, Imphal, India

<sup>2</sup>Assistant Professor, Department of Cardiology Regional Institute of Medical Sciences, Imphal, India

<sup>3</sup>Senior Resident, Department of Cardiology, Jawaharlal National Institute of Medical Sciences, Imphal, India

<sup>4</sup>Professor and Head of Department of Cardiology, Regional Institute of Medical Sciences, Imphal, India

<sup>5</sup>Junior Resident, Department of General Medicine, Regional Institute of Medical Science, Imphal, India

### Abstract

**Background:** Pulmonary arterial hypertension (PAH) is one of the most devastating and progressive disorder of the cardiopulmonary condition associated with poor survival and considerable morbidity. It is a disease characterized by increase pulmonary vascular resistance, eventually leading to right ventricular enlargement, right heart failure and increased mortality. Echo Doppler is an important non-invasive diagnostic technique and also a tool which can indicate the ventricular remodelling and diastolic dysfunction induced by PAH. The aim is to study the causes of Pulmonary arterial Hypertension. **Materials and Methods:** A single prospective observational study was conducted to study the etiologic profile of patients of PAH from September 2023 to April 2024 in Outpatient department (OPD) and Ward of Regional Institute of Medical Sciences (RIMS) and Jawaharlal Nehru Institute of Medical Sciences (JNIMS) Hospital, Manipur. Echo doppler was used as main modality to diagnose and grade PAH. **Result:** A total of 500 patients suffering from PAH were enrolled in our study to find out the associated cause of the 500 patients that were enrolled, the total number of male and female patients were 220 and 280 respectively, with the Male: Female ratio being 1:1.3. Most of the patients in our study belong to the age group 60 -69 years (175 patients) followed closely by  $\geq 70$  years (174 patients) with patients ranging from 17 years to 80 years. Most common cause of PAH was Dilated Cardiomyopathy(DCM) with 135 patients (27%), Chronic obstructive pulmonary disease (COPD) 112 (22.4%), Cor pulmonale 73 (14.6%), and others including HIV infections. Our study is a single prospective observational study designed to know the etiologies of patients of PAH. The median age being  $65.6 \pm 1.4$  years and DCM being the most common cause with 135 patients seen in the age group 60 -69 more in males and second most common being the COPD with 112 patients. Others like Cor pulmonale, Rheumatic heart disease, Congenital heart disease(CHD), Myocardial infarction(MI), Ischemic heart disease(IHD), Hypertension(HTN), Diabetes mellitus(DM), Chronic kidney disease (CKD) etc also led to the contributing factor to cause PAH. **Conclusion:** Pulmonary arterial hypertension (PAH) is a devastating cardiovascular condition with relatively poor quality of life which predisposes to significant morbidity and mortality. Echo doppler imaging is an important tool for diagnostic purpose.

## INTRODUCTION

Pulmonary arterial hypertension (PAH) is one of the most devastating and progressive disorder of the

cardiopulmonary condition associated with poor survival and considerable morbidity.<sup>[1]</sup> It is a disease characterized by increase pulmonary vascular resistance, eventually leading to right ventricular

enlargement, right heart failure and increased mortality.<sup>[2-6]</sup>

PAH may represent the final common pathway for myriad disease (ranging from collagen vascular disease to infection with HIV, Congenital heart disease or disease of the left side of heart, or respiratory system, or as an adverse reaction to several drugs) and this has compounded the problem of studying and managing the disease.<sup>[7,8]</sup>

Current management of PAH is a severity and risk based approach with early initiation of combination/parenteral therapy for high-risk patients. Even in the developed world, management of PAH is challenging due to complexity of pathology, need for invasive diagnostic studies and high-cost therapies. Management of PAH in a developing country is immensely challenging.<sup>[9]</sup>

Echo Doppler is an important non-invasive diagnostic technique for PAH patients, but also a tool which can indicate the ventricular remodelling and diastolic dysfunction induced by PAH to some extent.<sup>[10,11]</sup>

## MATERIALS AND METHODS

**Study Design:** This was a single prospective observational study designed to study the etiologic profile of patients of PAH. It was conducted from September 2023 to April 2024

**Study setting:** The study was done at Outpatient department (OPD) and Ward of Regional Institute of Medical Sciences (RIMS) and Jawaharlal Nehru Institute of Medical Sciences (JNIMS) Hospital, Manipur. Echo doppler was used as main modality to diagnose and grade PAH

**Study Population:** Total of 500 patients who attended OPD and admitted in Wards

### Inclusion Criteria

Patient who were above 18 years of age and diagnosed to have pulmonary arterial hypertension based on echo doppler imaging. Informed consent data was obtained from patients using a predesigned proforma and confidentiality maintained by assigning abbreviations

### Exclusion criteria

Patients who refused to give consent for echo doppler imaging. In addition, who have transient or acute elevation in pulmonary arterial systolic pressure (PASP) were also excluded from study

**Baseline Characteristics:** These were obtained based on detailed history and clinical examination including eliciting typical symptoms and signs. All the investigation reports and drugs being prescribed are recorded. Relevant investigation like blood examination. Chest-Xray, echocardiogram including doppler study, ECG, Computed tomography scan of chest were performed as required to establish the diagnosis and etiology method of measuring pulmonary artery systolic pressure by echo doppler study.

## RESULTS

A total of 500 patients suffering from PAH were enrolled in our study to find out the associated cause of the 500 patients that were enrolled, the total number of male and female patients were 220 and 280 respectively, with the Male: Female ratio being 1:1.3.

Most of the patients in our study belonged to the age group 60 -69 years (175 patients) followed closely by  $\geq 70$  years (174 patients) with patients ranging from 17 years to 80 years. The median age being 65.6  $\pm$ 1.4years. Age distribution of the patients are shown in [Table 1]

In Our study, the most common cause of PAH was Dilated Cardiomyopathy(DCM) with 135 patients (27%), Chronic obstructive pulmonary disease (COPD) 112 (22.4%), Cor pulmonale 73 (14.6%), and others including HIV infections. [Table 2] showing various causes of PAH with respect to sex According to severity cutoff of PAH based on echo doppler study it has been classified as Mild (35-50 mmHg), Moderate (50-70 mm Hg), Severe ( $>70$ mmHg) pulmonary systolic pressure.<sup>10</sup> In our study the most common group on echo doppler study of PAH was found to be Moderate grade (45.4%) followed by Mild grade (36.2%) and Severe grade (18.4%). [Table 3] showing severity of PAH based on echo doppler study.

**Table 1: Age distribution of study group.**

Age in years	No of patients
$\leq 29$	20
30 – 39	30
40- 49	35
50 – 59	66
60-69	175
$\geq 70$	174
Total	500

**Table 2: Causes of PAH with respect to sex**

Causes	Sex		Total
	Male	Female	
1.DCM	72	63	135
2. COPD	49	63	112
3.COR PULMONALE	14	59	73
4.RHD	9	28	37

5.CHD	11	20	31
6.HTN	17	8	25
7.MI	7	13	20
8.IHD	10	2	12
9.DM	6	8	14
10.CKD	9	2	11
11.OTHERS	16	14	20
TOTAL	220	280	500

**Table 3: Severity of PAH based on echo doppler study**

Grades	Sex		Total
	Male	Female	
Mild	83	98	181
Moderate	112	115	227
Severe	35	57	92
Total	230	270	500

## DISCUSSION

Pulmonary arterial hypertension (PAH) is a progressive disorder in which endothelial dysfunction and vascular remodelling obstruct small pulmonary arteries, resulting in increased pulmonary vascular resistance and pulmonary pressures resulting in reduced cardiac output, right heart failure, and ultimately death.

Doppler Echocardiography has a pivotal role in screening and diagnosis of pulmonary arterial hypertension and has been shown to correlate with invasive measurement during cardiac catheterisation. In resource limited setting, it is difficult to access cardiac catheterisation and diagnosis is often based on Doppler echocardiography and sign and symptoms of PAH.

Our study is a single prospective observational study designed to know the etiologies of patients of PAH. The median age being 65.6 ±1.4years and DCM being the most common cause with 135 patients seen in the age group 60 -69 more in males and second most common being the COPD with 112 patients .Others like Cor pulmonale , Rheumatic heart disease(RHD), Congenital heart disease(CHD), Myocardial infarction(MI) ,Ischemic heart disease(IHD), Hypertension(HTN), Diabetes mellitus(DM), Chronic kidney disease (CKD) etc also led to the contributing factor to cause PAH .<sup>[11]</sup>

The PROKERALA registry is the largest PAH registry from Asia and third largest registry in the world after the US based REVEAL registry and German based GIESSEN registry.<sup>[12,13]</sup> Valvular heart disease i.e RHD was the common etiological diagnosis in the GROUP 2 patients of PROKERALA registry. In concurrence with other international registry , Idiopathic PAH was the common followed by CHD-PAH and CTD-PAH in Group 1 PAH patients in the PROKERALA registry referral no.<sup>[12]</sup>

## CONCLUSION

Pulmonary arterial hypertension (PAH) is a devastating cardiovascular condition with relatively poor quality of life which predisposes to significant morbidity and

mortality. Echo doppler imaging an important tool for diagnostic purpose.

## REFERENCES

1. R Romberg E. Ueber slerose der lungenarterien. Deutsch Arch Klin Med. 1891; 48:197.
2. Pulmonary hypertension in clinical practice in the UK and Ireland. National pulmonary centres of the UK and Ireland. Thorax. 2008; 63(Suppl ID):ii1–ii41.
3. Michelakis ED, Wilkins MR, Rabinovitch M. Pulmonary vascular diseases. Emerging concepts and translational priorities in pulmonary arterial hypertension. Circulation. 2008; 118:1486–95.
4. Rich S, Dantzker DR, Ayres SM, Bergofsky EH, Brundage BH, Detre KM et al. Primary pulmonary hypertension: a national prospective study. Ann Intern Med.1987; 107(2):216–23.
5. D'Alonzo GE, Barst RJ, Ayres SM, Bergofsky EH, Brundage BH, Detre KM et al. Survival in patients with primary pulmonary hypertension: results from a national prospective study. Ann Intern Med. 1991; 115(5):343–9.
6. Jing ZC, Xu XQ, Han ZY, Wu Y, Deng KW, Wang H et al Registry and survival study in Chinese patients with idiopathic and familial pulmonary arterial hypertension. Chest. 2007; 132(2):373–9.
7. Rich S, Rubin L, Walker AM, Schneeweiss S, Abenheim L. Anorexigens and pulmonary hypertension in the United States: results from the surveillance of North American pulmonary hypertension. Chest. 2000; 117(3):870–4.
8. Walker AM, Langleben D, Korelitz JJ, Rich S, Rubin LJ, Strom BL, Gonin R, Keast S, Badesch D, Barst RJ, Bourge RC, Channick R, Frost A, Gaine S, McGoon M, McLaughlin V, Murali S, Oudiz RJ, Robbins IM, Tapson V, Abenheim L, Constantine G.. Temporal trends and drug exposures in pulmonary hypertension: an American experience. Am Heart J. 2006; 152(3):521–6.
9. Stricker H, Domenighetti G, Popov W, Speich R, Nicod L, Aubert JD, Solèr M; Swiss Group for Severe Pulmonary Hypertension Severe pulmonary hypertension: data from the Swiss Registry. Swiss Med Wkly. 2001; 131(23–24):346–50.
10. Cardina RL , Playford D, Lang I, Celer majer DS. State -of-the-Art Review : echocardiography in pulmonary hypertension Heart, Lung and Circulation .2019;28: 1351
11. Obokata M , Kane GC , Sorimachi H, et al : Noninvasive evaluation of pulmonary artery pressure during exercise: the importance of right atrial hypertension Eur Respir J 2020 Feb 12; 55(2): 1901617
12. . McGoon M.D., Miller D.P. REVEAL: a contemporary US pulmonary arterial hypertension registry. Eur Respir Rev. 2012 Mar 1;21(123):8–18.
13. Gall H., Felix J.F., Schneck F.K., et al. The giessen pulmonary hypertension registry: survival in pulmonary hypertension subgroups. J Heart Lung Transplant. 2017 Sep;36(9):957–967.