Original Research Article

Clinical Profile of Pulmonary Hypertension: An Underdiagnosed Disease

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Abstract

Introduction: Pulmonary hypertension is a complex disease that presents with a broad spectrum of morphological and hemodynamic findings of varying severity, which has a major impact on morbidity and mortality.
Objective: To study the clinical and epidemiological profile of pulmonary hypertension in a tertiary care center.
Method: All patients admitted to Tertiary care Hospital, who met PAH criteria on echocardiography, between August 2017 to January 2020 were included. A detailed history was taken along with a physical examination done on each patient. Exercise capacity was measured using a 6-minute walk test along with oxygen saturation at rest and immediately after a 6-minute walk. All relevant investigations were performed.
Result: During the study period, 100 patients were admitted with pulmonary hypertension with an average male to female ratio was 1.1: 1, with the maximum incidence in the patient between 31 to 40 years (range-18 yr. & above). Dyspnoea (94%) was the commonest symptom followed by palpitation (58%), chest pain (57%) and easy fatigability (48%). The mean 6-minute distance walked by patients was 251±110 m. Immediately following 6-minute walk, SO_2 remained unchanged (or decrease by <5%) in 61.3% but appreciably decreased (> 5% absolute reduction) in 38.6%. Major etiological factors observed were primary respiratory illness (group 3) seen in 34% followed by primary left heart disease (group 2) seen in 31%; about 17% belong to category group 1.

Keywords: Pulmonary Hypertension
Introduction, tertiary care center

Pulmonary hypertension is a complex disease that presents with a broad spectrum of morphological and hemodynamic findings of varying severity, which has a major impact on morbidity and mortality.
The global prevalence of PAH is hard to estimate because accurate diagnosis of PAH is difficult and there is no single causative factor as it may represent the final common pathway for multitude disease.

In developing country like ours, prevalence will also likely increases as newer associations with PAH emerge, including dialysis and the metabolic syndrome and as widespread access to echocardiography identifies PAH earlier and in more individuals.
This study had recorded the clinical characteristics and demographics of PAH cases in a single center study that hopefully could be a stepping stone in the development of a national registry.

Materials and Methods

This study was conducted in Tertiary care Hospital in the period between August 2017 to January 2020, who met PAH criteria on echocardiography.
It included 100 patients diagnosed with PH who were admitted in hospital. It is important to note that although current guidelines recommend right heart catheterization (RHC) for the diagnostic evaluation of patients with PAH, the diagnostic approach in this study was established mainly in the presence of clinical suspicion based on the accepted definitions and echocardiography recording high pulmonary artery systolic pressure (PASP > 40 mmHg) as the RHC wasn't available.
All the data were recorded in Microsoft excel. Statistical analysis of the compiled data was analyzed using Microsoft excel band Epi Info™ and presented in proportion and mean values. Proportions were analyzed using the Chi Square test, error value set at <5%.

Results

The total number of the studied patients was 100 with a mean age of 49 ± 16 years, among them 53 males (53%) and 47 males (47%). A majority of patients belonged to the age group of 31-40 years (28%).
The clinical characteristics of the recorded patients have illustrated in Table 1from which it is evident that breathlessness was the presenting symptom in the overwhelming majority of the patients (94%) followed by palpitations (58%) and chest pain (57%), easy fatigability (48%), swelling of feet (32%), cough (31%) and syncope (9%).
On precordial examination, 84% of patients had the loud pulmonary component of the second heart sound and 30% of patients had an audible pansystolic murmur.

Table 1: The clinical characteristics of the recorded patients

	Percentage (%)
Pulmonary symptoms	
Breathlessness	94
Palpitations	58
Chest Pain	57
Fatigability	48
Swelling of feet	32
Syncope	9
Cough	31
Clinical examination	
Hypertension	21
Irregular pulse	12
Central cyanosis	11
Clubbing	23
Elevated jugular venous pulse	51
Lower limb edema	32
Wheezes	15

Crackles	57

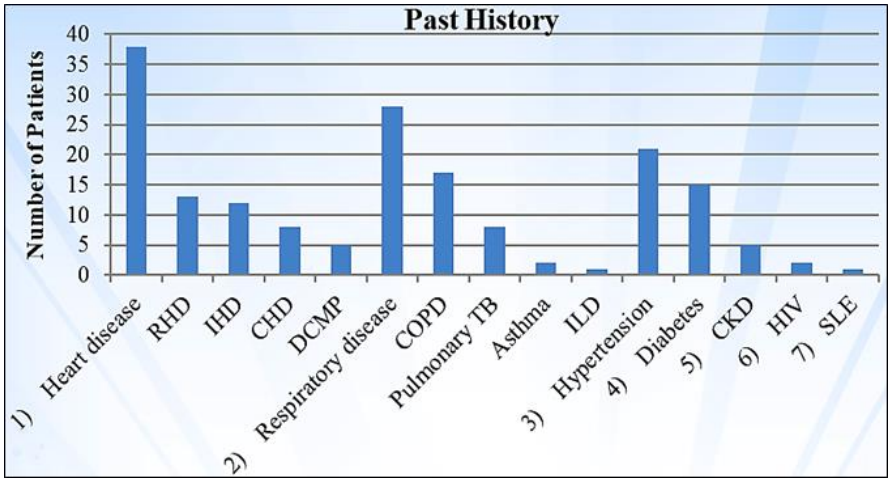
Cardiovascular examination	
Loud	S2
Intensity	84
Heave	58
Left 2nd space dullness	60
S3 gallop	29

The mean 6 minute distance walked by patients was 251±110 m. Immediately following 6-minute walk, SO_2 remained unchanged (or decrease by <5%) in 61.3% but appreciably decreased (> 5% absolute reduction) in 38.6% (Table 2).

Table 2: Pulse oximetry and 6-minute walk distance

Parameter	**Value**
SaO_2 at rest (%)	
≥90%	81%
81-89%	19%
6-minute walk distance (meters)	
Overall	248±112 m
After excluding the 12 patients who could not walk at all	251±110 m
SaO_2 after walk-absolute change from baseline	
Unchanged (same or <5% decrease)	54 (61.3%)
Decreased (> 5%)	34(38.6%)

History of heart disease was present in 38% of patients, while of respiratory illness and chronic kidney disease were present in 28% and 5% of patients respectively. 2 patients were retroviral positive and 2 patients were observed in post-partum period (2nd and 4th month respectively). 1 patient in the study was diagnosed with scleroderma.

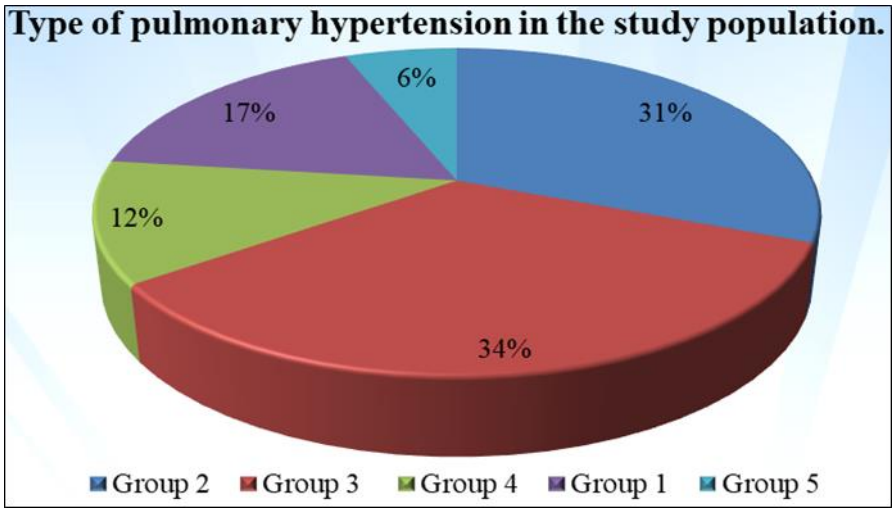


Among the 100 ECG reports collected two predominant findings were noticed equally in the form of right axis deviation and P pulmonale in 44%. The most commonly observed findings on chest X-rays were cardiomegaly (61%) and hilar prominence (63%).

Echocardiography assessment revealed an elevated right ventricular systolic pressure (RVSP) with an estimated mean of 59 ± 13mmHg, while mean REVF was 46% and mean RVEDV was 57.95mm3 and RVESV was 38mm3. It is important to note that some of the patients had more than one abnormality detected.

Large majority of the study population had pulmonary hypertension related to hypoxemic

lung disease (group 3) and left heart disease (group 2) as major etiologic factors.
To elaborate primary respiratory illness (group 3) around 34% followed by primary left heart disease (group 2) around 31%; about 17% of patients in our study fall in group 1. Among patients with group I, the largest percentage was contributed by patients with congenital heart disease (44%) followed by patients with idiopathic pulmonary hypertension. Of the total study subjects, 3 patients were Human immunodeficiency positive and 2 patients had pre-existing connective tissue disorders of which 1 was diagnosed with SLE and the other was diagnosed with systemic sclerosis. There was no patient with familial PH. 12 patients in study group were diagnosed with chronic pulmonary thromboembolism (group 4).
6% of patients were of group 5 PH (miscellaneous causes like sarcoidosis, histiocytosis X, chronic kidney disease, haematologica etc.). Of which, 4 patients were of CKD stage 4/5and 1 patient is diagnosed with sarcoidosis in this study.



Discussion

The study reveals thatmajority of patients belonged to the middle age group of 31-40 years (28 %) with mean age was 49 ± 16 years matching with studies done by Mehrotra*et al*. (35.6 ± 14.3) years and Marc Humbert*et al*. (50 ± 15) years. However, it wasn't in accord with the Gartman*et al*. (55 ± 10) years and Fayngersh*et al*. (63 ± 10.2) years
There is no pathognomonic clinical sign of PH; clinical presentation is related either to right heart failure or to associated diseases. Persistent dyspnea on exertion is the most frequent symptom; and it is present in almost all patients even in the presence of mild hemodynamic abnormalities. The study showed that dyspnea was the most prevalent symptom being manifested by 94% patients, followed by palpitations (58%), chest pain (57%), swelling of feet (32%), cough (31%), syncope (9%) and easy fatigability (48%) showing concordance with results of Ling *et al*., Marc Humbart*et al*.
The 6-minute walk test is a submaximal exercise test that can be safely performed by patients incapable of tolerating maximal exercise testing. It is straightforward, reproducible, and does not require any equipment. The distance walked in 6 minutes has a strong independent association with mortality and correlates with functional class.
Oxygen saturation at rest and after 6-minute walk is also a useful marker of functional capacity and was assessed in our patients. In most (81%) of our patients, oxygen saturation was greater than 90% at rest and in more than half of them, it remained same or reduced by less than 5%. In about 36.8% of the patients, oxygen saturation measured by pulse oximetry was reduced by more than 5%.
Among the 100 ECG reports collected two predominant findings were noticed equally in the form right axis deviation and P pulmonale in 44% and this agreed with Bossone*et al*.
The right ventricle plays an important role in the morbidity and mortality of patients presenting with signs and symptoms of cardiopulmonary disease. However, the systematic

assessment of right heart function is not uniformly carried out. This is due partly to the enormous attention given to the evaluation of the left heart, lack of familiarity with ultrasound techniques that can be used in imaging the right heart, and a paucity of ultrasound studies providing normal reference values of right heart size and function. Unfortunately, this applies to this study since data were collected retrospectively; ECHO assessment of the right heart was defective and represents one of the limitations of this study.

In this study retrospective, ECHO assessment records revealed an elevated right ventricular systolic pressure (RVSP) with an estimated mean of 59 ± 13mmHg. It is important to note that some patients had more than one abnormality detected.

PAH due to lung diseases and/or hypoxia (group 3) constituted 34% of cases mostly chronic obstructive pulmonary disease (COPD), followed equally by patients with PAH due to left sided heart failure (31%). About 17% of patients in our study fall in idiopathic pulmonary hypertension (group 1). 12 patients of study group were diagnosed with chronic pulmonary thromboembolism (group 4).

The results didn't conform with the findings of many studies as Marc Humbert*et al.* and Rahul *et al.*, The discrepancies in the results could be attributed to many factors, firstly the variable populations, enrollment pattern and cohort size among the different studies. Secondly the profile of PH and patients characteristics varies around the world.

Limitations

The study was done on a small sample size from one part of one city which may lead to some degree of selection bias. The methodology involves use of non-invasive techniques including 2Dechocardiography, Doppler study; all of which have subjectivity and operator dependency. Furthermore, PA pressure estimated by Doppler does not have adequately proven correlation with their invasive gold standard.

Conclusion

Our study provides novel information on the clinical and epidemiological features of all types of PH. It also highlights some unique characteristics of PH. Some of the findings are quite similar to the western data but some interesting observations had also emerged. There remains a need to spread awareness about PH among the physicians and specialists since it occurs in association with myriad diseases.

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