Original Research Article

Study of the clinical and etiological profile of pulmonary hypertension

Priya V Patil[1], Geetanshu Goel[2*], Sujeet Kamtalwar[3]

[1]Associate Professor, [2,3]Assistant Professor, Department of Medicine, Grant Government Medical College, Sir JJ Group of Hospitals, Byculla Mumbai, Maharashtra, INDIA.
Email: charugoel2020@gmail.com

Abstract

Background: Pulmonary hypertension (PH) refers to the presence of high pulmonary vascular pressure and can be result of variety of different underlying disorders. PH is defined as increase in mPAP of 25mmhg or more. There is a vast difference between the etiologies of pulmonary hypertension as quoated in the western literature and there are only few Indian studies done to evaluate the spectrum of causes of Pulmonary Hypertension in our Indian setting. This study was carried out at our institution in the medicine department as inspite of being a tertiary care centre with super speciality departments most of the cases are treated by medicine department, right from diagnosis to the treatment. We have a large number of patients (inpatient/outpatient) who have pulmonary hypertension due to wide spectrum of causes including cardiac like congenital and valvular heart diseases and pulmonary like chronic obstructive and interstitial lung diseases. Hence a systematic study to obtain the pattern and etiological spectrum was undertaken with focus on presentation and clinical profile, 2D echo findings, severity of disease and the classification of pulmonary hypertension. **Methodology:** It was a prospective observational study carried out in patients admitted at a tertiary care center. **Results:** The most common aetiology of pulmonary hypertension in our study was valvular heart disease WHO class II (95/163, 58.28%) ;followed by COPD WHO class III (36/163, 22.08%) ;followed by Pulmonary Arterial Hypertension WHO class I (8/163, 4.91%); followed by Chronic Thromboembolic Pulmonary Hypertension WHO class IV (8/163, 4.91%). Only 3/163 cases (1.84%) belonged to WHO class V. 75 patients had moderate grade of PH (46.01%), 48 patients (29.45%) had mild grade and 40 patients (24.54%) had severe grade of PH on 2D echo.
Key Word: pulmonary hypertension.

***Address for Correspondence:**
Dr. Geetanshu Goel, Assistant Professor, Department of Medicine, Grant Government Medical College, Sir JJ Group of Hospitals, Byculla Mumbai INDIA.
Email: charugoel2020@gmail.com
Received Date: 15/06/2018 Revised Date: 10/07/2018 Accepted Date: 04/08/2018
DOI: https://doi.org/10.26611/1021724

Access this article online

Quick Response Code:

Website: www.medpulse.in

Accessed Date: 16 August 2018

INTRODUCTION

PH has previously been called an orphan disease, that is, a condition that affects few individuals and is overlooked .Although rare, the concept that PH is overlooked cannot be considered to be the case today. Indeed, a number of recent important discoveries have improved our understanding of the disease, helped guide patient management, and laid foundations for future research the medical profession, health care systems, and pharmaceutical companies. PH is defined as an increase in m PAP of 25mm hg or greater. The manifestation of pulmonary hypertension is nonspecific, leading to delays in evaluation and diagnosis.[1]The symptoms of pulmonary hypertension can also include fatigue, weakness, angina, syncope, and abdominal distention. Symptoms at rest are reported only in very advanced cases[6,7]. The clinical suspicion of pulmonary hypertension should arise in any case of dyspnea without overt signs of specific heart or lung disease or in patients with underlying lung or heart disease whenever there is increasing dyspnea unexplained by the underlying disease itself. 2. Pulmonary hypertension can sometimes be suspected when abnormal clinical findings such as loud P2, murmur of TR, jugular venous distension, central cyanosis with normal lung

How to cite this article: Priya V Patil, Geetanshu Goel, Sujeet Kamtalwar. Study of the clinical and etiological profile of pulmonary hypertension. *MedPulse International Journal of Medicine.* August 2018; 7(2): 51-56. https://www.medpulse.in/Medicine/

examination and other findings as electrocardiographic, chest radiograph, or echocardiographic findings are detected in the course of procedures performed for other clinical reasons.

MATERIALS AND METHODS

Study design: It was a prospective and observational study.

Study centre: Study was carried out in patients admitted at a tertiary care centre.

Study period: This study was carried out over a period of 2 years (2015-2017) after obtaining permission from the institutional ethical committee.

Inclusion criteria: This prospective study was conducted on patients with age >18 years with pulmonary hypertension diagnosed over 2D echo.

RESULTS

TABLE 1: Distribution of study group as per, AGE

AGE	Frequency	Percent
13-30 Yrs	30	18.40%
31 to 40 Yrs	26	15.95%
41 to 50 Yrs	38	23.31%
51 to 60 Yrs	23	14.11%
61 to 70 Yrs	28	17.18%
Above 70 Yrs	18	11.04%
Total	163	100.00%

Table 2: Distribution of study as per sex

SEX	Frequency	Percent
Male	97	59.51%
Female	66	40.49%
Total	163	100.00%

Table 3: Symptoms

Symptoms	Frequency	Percent
CHEST PAIN	42	25.57%
PALPITATIONS	28	17.18%
DYSPNOEA	148	90.80%
FATIGUE	12	7.36%
SWELLING	39	23.93%
COUGH	41	25.15%

Table 4: Showing findings on physical examination

Physical Examination		
Pallor	100	61.35%
Icterus	10	6.13%
Cyanosis	5	3.07%
Clubbing	6	3.67%
Ascites	17	10.43%
Hepatomegaly	11	6.74%
Pedal edema	51	31.28%

Table 5: Clinical examination findings in the study population (n=163)

PARAMETERS	VALUE
Heart rate(beats/min)	92.44±23.09
Systolic blood pressure(mmhg)	119.7±17
Diastolic blood pressure(mmhg)	76±10.3
Elevated jugular venous pressure	21/163 (12.8%)
Loud pulmonary component of second heart sound	85/163 (52.14%)
Audible murmur	85/163 (52.14%)
Rhonchi/ Crepitations	53/163(32.52%)

Table 6: Shows the distribution of the study population on the basis of etiology of PH

Classification of PH	No of cases	%	TOTAL (n=163)
1. Pulmonary arterial hypertension			
1.1 Idiopathic PAH	2	1.2%	
1.4Associated with:			4.91%
1.4.1 Connective tissue disease	1	0.6%	
1.4.4 Congenital heart disease	5	3.07%	
2. Pulmonary hypertension due to left heart disease			
2.1Left ventricular systolic dysfunction			
2.3 Valvular disease	1	0.6%	58.89%
	95	58.28%	
3 Pulmonary hypertension due to lung diseases and/or hypoxia			
3.1 Chronic obstructive pulmonary disease	30	18.41%	
3.2 Interstitial lung disease	8	4.90%	
3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern – Bronchiectasis	6	3.75%	29.45%
3.4 Sleep- disordered breathing	4	2.45%	
4 Chronic thromboembolic pulmonary hypertension (CTEPH)	8	4.9%	4.9%
5 Pulmonary hypertension with unclear/multifactorial mechanisms			
Hematologic disorders	3	1.84%	1.84%

Table 7: Table showing findings on 2d echo in the study population:

2D ECHO FINDINGS	NO OF CASES	PERCENTAGE
I. Valvular heart diseases		
1. Pure mitral stenosis(MS)	32	19.63%
2. Pure mitral regurgitation(MR)	11	6.74%
3. Aortic stenosis (AS)	2	1.22%
4. Aortic regurgitation(AR)	1	0.06%
5. Multivalvular	49	30.06%
II. Septal defect/Congenital heart disease		
1. ASD	3	1.84%
2. PS	1	0.62%
3. PDA	1	0.62%
III.		
1.DCMP	1	0.62%
2.Diastolic dysfunction	8	4.91%
3.LA/LV dilation	52	31.90%
IV. RA/RV Dilated	53	32.51%
V. Isolated Pulmonary Hypertension	2	1.22%

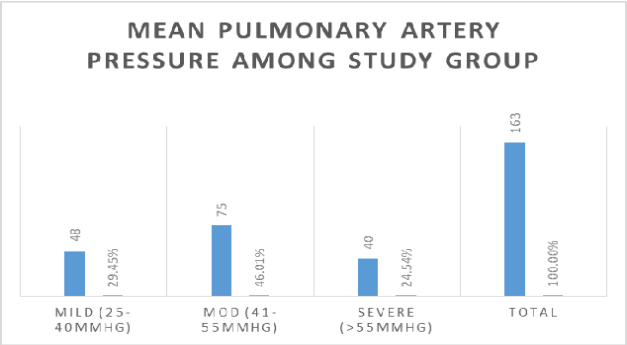

Figure 1

DISCUSSION

Table no 1 shows distribution of the patients as per age group. The maximum number of patients in the study group belonged to age group 41-50 years of age (38/163, 23.31%). The mean age of the patients enrolled under this study was 48.48±17.57 years. It was almost similar to the studies done earlier.Hence, in our study it was seen that pulmonary hypertension is a disease of middle aged to old age people as compared to younger population.

Table 2 shows distribution of study group according to sex. The current study was dominated by male population (males – 59.51% and females- 40.49%). Male to female ratio was 1.46: 1.

Table 3 shows common presenting symptoms in the study population and their frequency.

The most common presenting symptom was dyspnoea on exertion which was seen in148(90.80%) of the patients which was followed by chest pain 42 (25.57%) cases, cough 41 (25.15%) cases, swelling (especially of the lower limbs) 39 (23.93%) cases, palpitations 28 (17.18%) cases and fatigue 12 (7.36%) cases. Dyspnoea is the commonest symptom as it reflects underlying aetiology. 58.8% of cases in this study had left heart disease and 29.45% had lung diseases. Hence dyspnoea was commonest symptom, which could be due to pulmonary congestion/ oedema in left heart diseases or congenital heart diseases or lung parenchymal pathology in lung diseases.

Table 4 shows various findings on physical examination Pallor was the most common physical finding in the study population. 100 (61.35%) patients had pallor on examination. Pedal edema was present in 51 (31.28%) patients which was suggestive of the failing right ventricle secondary to pulmonary hypertension in this group of patients. Clubbing was found in 6 (3.67%) patients and all had interstitial lung disease. Thus clubbing is an important clue for presence of underlying cause of pulmonary hypertension being Either UIP/IPF especially in association with other signs and relevant clinical examination Cyanosis was seen in 5 (3.07%) cases of the study group and all had obstructive airway disease. Thus cyanosis is an important clue for the presence of underlying cause of pulmonary hypertension being chronic obstructive airway disease due to presence of polycythemia in these patients.

Table 5 shows findings of study population on clinical examination JVP was raised in around 21 (12.88%) patients which is one of the markers of failing right ventricle. Loud P2 was present in about 85 (52.14%) % of the patients. It is one of the important clinical finding of presence of pulmonary hypertension. In 85 (52.14%) of the patients there was presence of audible murmurs. Hence on auscultation, there can a definite clue of underlying heart disorder especially of valvular heart disease. In our study, Mid Diastolic Murmur, Pan systolic Murmur were heard which were suggestive of Mitral Stenosis, Mitral Regurgitation respectively. Ejection Systolic Murmur was also present which itself is an indicator of underlying pulmonary hypertension. Rhonchi/ Crepitations were present in total number of 53 (32.52%) cases. It was either seen in patients having underlying lung pathology or in patients having left sided heart diseases. Presence of rhonchi and coarse crepitations were seen in cases of COPD, course and mid inspiratory crepts were seen in cases of ILD whereas bilateral fine basal crepts were seen in left ventricular failure.

Table 6.shows the distribution of the study population on the basis of Etiology of PH which is one of the important aim of our study. According to the classification of pulmonary hypertension, 58.89% of the cases were due to left heart diseases(class II) which was followed by PH due to lung diseases and/or hypoxia(class III) with 29.45% of the patients.4.9% each of the patients belonged to PAH(class I) and CTEPH(class IV) group. Only 1.84% of the total population enrolled in the study had PH due to hematologic disorder (class V) which was due to sickle cell anaemia. Thus in our study incidence of class I pulmonary hypertension was much lower than class II,III. This was quite different from western studies where class I was most common. 8 patients (4.91%) had class I pulmonary hypertension which is Pulmonary Arterial Hypertension. Out of these, 2 patients had Idiopathic PAH. The diagnosis was done after ruling out cardiac and pulmonary causes in both the cases. One case had an underlying Connective Tissue Disorder. Patient presented with history of rash over the cheeks, difficulty in breathing and painful digits. Reynolds phenomenon was present on examination. ANA was positive and ANA Blot was done which was positive for RO, DsDNA AND U1-snRNP.Hence, patient had Pulmonary Arterial Hypertension secondary to MCTD. Rest of the 5 cases had Pulmonary Arterial Hypertension due to congenital heart disease. 3 patients had an underlying OS-ASD diagnosed on 2D Echo. 1 patient had PDA with 2Decho suggestive of severe PAH with reversal of shunt (Eisenmenger phenomenon present). 1 patient had pulmonary arterial hypertension due to Pulmonary Stenosis. Patient had valvular and infundibular type of pulmonary stenosis with RA/RV dilated on 2D Echo. In a study conducted by *Morjaria s et al*.[8] it was seen that years of stenosis can result in sub endocardial hypertrophy causing significant outflow

obstruction and resulting in right ventricular pressure overload and pulmonary hypertension. 96 patients (58.89%) had class II Pulmonary Hypertension which is due to an underlying left heart disease.[9] 1 patient had DCMP in whom pulmonary hypertension developed due to systolic dysfunction of left ventricle. All the chambers were dilated with EF 20%. 95 cases had pulmonary hypertension secondary to valvular heart disease. All these valvular lesions were Rheumatic in origin. Out of 95 patients, 32patients (19.63%) had pure mitral stenosis, 11(6.74%) had pure mitral regurgitation, 2(1.22%) had aortic stenosis, 1(0.06%) had aortic regurgitation and remaining 49 cases (30.06%) had multivalvular heart disease. 48 patients (29.45%) had class III Pulmonary Hypertension out of which 30(18.41%) patients had Chronic Obstructive Airway Disease, 8 patients (4.90%) had Interstitial Lung Disease, 6 patients (3.75%) had bronchiectasis and remaining 4 patients (2.45%) had Obstructive Sleep Apnea.

Table 7 also highlights other findings on 2d echo in patients with pulmonary hypertension. It was seen that 8 patients (4.91%) had diastolic dysfunction on 2d echo. It is also one of the factors responsible for pulmonary hypertension[10,11]. It comes under class II of classification of pulmonary hypertension. However, it was seen that the main cause of pulmonary hypertension in these cases was either an underlying lung or heart disease. It can be said that diastolic dysfunction also significantly could have contributed to pulmonary hypertension in these cases. However it was not the sole factor responsible for pulmonary hypertension as they had other causes. From this, it can be concluded that in a given patient, etiology of pulmonary hypertension could be multifactorial with more than one factor responsible collectively towards the pathophysiology of pulmonary hypertension.

Table 8 shows the mean pulmonary arterial pressure in study population. On 2d echo it was found that mean pulmonary arterial pressure (mPAP) was 47.34 ± 12.88mmhg. This is almost similar to what was observed in previous studies. The mean pulmonary arterial pressure was found to be higher in women as compared to men. In men mean PSAP was 46.89±12.56mmhg whereas in females it was 48±13.30mmhg.

LIMITATIONS

The following were the limitations in our study:

1. We could not do the Right Heart Catheterization studies especially in cases of Pulmonary Arterial Hypertension who are diagnosed to be having Primary Pulmonary Hypertension
2. Study protocol did not include 6 minute walk test to assess the functional severity of Pulmonary Hypertension in the patients
3. We included only patients who were admitted and discharged. We didn't include the patients who were admitted and died. Hence we couldn't assess the mortality as complication of PH.

CONCLUSION

The most common aetiology of pulmonary hypertension in our study was valvular heart disease WHO class II (95/163, 58.28%) ,followed by COPD WHO class III (36/163, 22.08%) followed by Pulmonary Arterial Hypertension WHO class I (8/163, 4.91%) followed by Chronic Thromboembolic Pulmonary Hypertension WHO class IV (8/163, 4.91%). Only 3/163 cases (1.84%) belonged to WHO class V. Hence, while treating Pulmonary Hypertension, it is very important to have an accurate diagnosis of etiology as the specific treatment depends on the underlying etiology. Thus only Idiopathic Pulmonary Hypertension is treated with pulmonary vasodilators like Sildenafil and Bosentan. However this is a rare diagnosis and other common etiology like valvular heart diseases and lung diseases needs to be ruled out.

REFERENCES

1. Simonneau G, Catzoulis MA, Adatia I, *et al*: Updated clinical classification of pulmonary hypertension. J Am Coll Cardiol 62(25 Suppl):D34, 2013.
2. Pulmonary Hypertension Registry of Kerala (PROKERALA) - Rationale, design and methods.Harikrishnan S, *et al*. Indian Heart J. 2016.
3. A Study of Etio-Clinical and Echocardiographic Profile of Patients Presenting with Pulmonary Hypertension by Asif Hasan1, Muhammad Uwais Ashraf 2, Sandeep Kumar3 ,123Department of Medicine, J N Medical College, AMU, Aligarh, UP, India.
4. Epidemiology of pulmonary hypertension: new data from the Swiss registry. Tueller C1, Stricker H, Soccal P, Tamm M, Aubert JD, Maggiorini M, Zwahlen M, Nicod L; Swiss Society for Pulmonary Hypertension.
5. Badesch DB, Champion HC, Sanchez MA, *et al*. Diagnosis and assessment of pulmonary arterial hypertension. J Am Coll Cardiol 2009; 54:S55-66.
6. Galie N, Hoeper MM, Humbert M, *et al*. Guidelines for the diagnosis and treatment of pulmonary hypertension: The Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT). Eur Heart J 2009; 30:2493-2537.

7. Voelkel NF, Gomez-Arroyo J, Abbate A, *et al*: Pathobiology of pulmonary arterial hypertension and right ventricular failure. Eur Respir J 40:1555, 2012.
8. Massive dilatation of the pulmonary artery in association with pulmonic stenosis and pulmonary hypertension.Pulm Circ. 2012; 2(2):256-7 (ISSN: 2045-8940) Morjaria S; Grinnan D; Voelkel N
9. Pulmonary Hypertension Associated with Left-Sided Heart Disease Christopher F. Barnett, MD, MPH, Teresa De Marco, MD
10. Diagnosis and management of pulmonary hypertension associated with left ventricular diastolic dysfunction by Perez VA1, Haddad F, Zamanian RT.
11. Pulmonary hypertension associated with left ventricular diastolic dysfunction by Christopher A. Fiack, MD, Harrison W. Farber, MD .Pulmonary Center, Boston University School of Medicine, Boston, Massachusetts.

Source of Support: None Declared
Conflict of Interest: None Declared