



Etiology of Pulmonary Hypertension less than 40 years age in a tertiary care centre: Hundred patients tribal based population Study

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Abstract

Pulmonary Hypertension is a serious and unrelenting pulmonary vascular disorder that affects the functional quality of patients significantly and decrease the lifespan. Diagnosis at early stage may save many life as numbers of therapeutic options are available. Etiological diagnosis of PAH is very important as everyone demands separate therapeutic protocol and may curable if diagnosed in time. As the symptoms are often subtle in the early stage of disease it is imperative that physician should be aware of the manifestation of condition. A routine work up may be carried out in well equipped Hospital at periphery in many affluent and advanced countries. In developing countries like India work up can only be done in a tertiary care centre under the supervision of pulmonologist and cardiologist. In our study we had tried to focus the etiological diagnosis of PAH as availability of resources in our setup to give a brief idea in this population group. Comparative young age group were choosen as the stage of disease in this age group shows significant reversibility with proper treatment.

Keywords: *Etiology of Pulmonary Hypertension, Pulmonary Hypertension, Primary Pulmonary Hypertension, Secondary Pulmonary Hypertension.*

Introduction

Pulmonary Hypertension is a progressive disease where the Pulmonary artery continue to shrink, making the right side of the heart work harder as it makes the higher pressure needed to force blood through narrowed artery. It happens because the muscle cells in arteries devise and plug up the arteries. The vessels may also blocked by Blood clots or lung damage by systemic illness. This results in loss of alveolar space and blood vessels in lung. Consequently cardiac function is

compromised due to heavy work to be done by cardiac muscle.

WHO has divided Pulmonary Hypertension into five groups on the basis of patho physiology, clinical presentation and therapeutic options. These group includes as follows:

Group 1: Pulmonary arterial Hypertension cause unknown, Known as idiopathic Pulmonary arterial Hypertension.

Group2: Pulmonary Hypertension caused by left sided heart disease.

Group3: Pulmonary Hypertension caused by lung disease.

Group 4: Pulmonary Hypertension caused by chronic blood clots.

Group 5: Pulmonary Hypertension associated with other conditions that have unclear reasons why the Pulmonary Hypertension occurs.

Symptoms of Pulmonary Hypertension include:

Shortness of breath, Fatigue, dizziness or fainting spells, chest pressure or pain, swelling in ankles, legs and abdomen (Ascites), Bluish color in lips and skin, Racing pulse or palpitation.

Materials and Methods

In this we have studied 100 patient population in a tertiary care hospital. Population were came from merely Tribal area. Patient admitted in our hospital between January 2017 to September 2018. Teen age and young adult population less than 40 yrs were included in the study. Cross sectional study with retrospective analysis was done to search for Etiology, stage of disease and available resources. As the population mostly came from low socioeconomic status interventional approach and high cost medicines were restricted. Every patient were registered, assessment of PAH done with echocardiography. Severity of PAH and supportive evidence of etiological diagnosis was done by ECG, Echo, CT scan chest, PFT and by biochemical parameters.

Family History of PAH significant childhood medical ailment, prolonged systemic illness, long term continued medicine use were critically evaluated. With echocardiography severity of PAH done by TR gradient as per protocol as secondary evidence. No patient in our study right heart catheterization done as neither necessary nor Cathlab facility available in our setup. Most of the patient not willing for invasive procedure as a diagnostic test.

Results

We have studied total 100 patient populations. Teenage to age forty included. Lowest age was

fifteen and highest one had forty. Of the population group 67 was female and 33 was male. Most of them were came from lower socioeconomic group. Among the female population 23 patient had history of oral contraceptive use more than three years. Twenty four patients with no apparent cause of Pulmonary Hypertension was found as per our investigation protocol diagnosed as Idiopathic Pulmonary Hypertension. Out of 24 patients with IPH 19 was female and Five was male. All of them did not significant family history suggestive of such ailment. Congenital heart disease with significant shunt leads to pay as an important cause. PAH is either due to Hyperkinetic circulation, fixed or due to shunt reversal. In our population group eleven patients had congenital heart disease with significant left to right shunt. Out of them seven was female and Four was male. Three patient had VSD, Two patient had PDA, one patient had ASD, one patient had ECD. Recurrent childhood respiratory tract infection either bronchitis or pneumonia are important cause for PAH. Five patients in our studies found history of recurrent LRTI suggested by 3-4 attacks per year during childhood. Nine patient had associated systemic illness S/O rheumatological disorder. All of them were female either with SLE or systemic sclerosis and one was MCTD. No patient with Typical RA as per clinical and biochemical parameter shows PAH in our study. Secondary PAH due to rheumatic heart disease is an important cause in developing country, In our population 23 patient had significant valvular heart disease either mitral valve or Mitral and aortic valve involvement.

Fourteen patient was female and nine was male. All of them had severe Mitral stenosis or Mitral regurgitation with or without Aortic valve involvement. Three patients had history of BMV procedure and two had history of MVR. Long standing DCM is an important cause of PAH. Thirteen patient in our population with DCM had significant PAH. DCM was either idiopathic, metabolic or ischemic in origin. Obstructive or restrictive lung disease is an important cause of

PAH. Though obstructive pattern mainly found in middle and advanced age group, restrictive lung disease such as ILD can affect middle age population. In our study ten patient had significant primary lung disease as diagnosed by PFT and HRCT chest had significant PAH. Rest of the group two patient had chronic liver disease associated with portal Hypertension. Three patient had diabetic nephropathy with maintenance dialysis and one patient was sero +ve for HIV.

Table

Total population-N=100
Idiopathic Pulmonary Hypertension-N=24.
Acquired Heart Disease. (RHD+Cardiomyopathy)-N=36
Congenital heart disease, N=11
PAH due to primary lung disease, N=10
PAH due to connective tissues disorder, N=9
PAH associated with portal Hypertension, N=2
PAH associated with chronic kidney disease, N=3
PAH due to recurrent Bronchiolitis and childhood LRTI, N=5

Discussion

The study is an observational study in our centre. The study was done in small cohort of patient for Two years period. As this geographic population was less highlighted in literature we were trying to found the trend of PAH here with the respect to global trend found in various literature. In our study group most common cause of PAH less than forty was secondary to acquired heart disease. Thirty six out of hundred population with PAH was due to acquired cardiac ailment. As the prevalence of RHD is high in this geographic area RHD is the predominant cause. Most patient in this area presentation late at hospital and they want to adhere with medical management. Poor socioeconomic status, lack of hygiene, awareness, ignorance and scarcity of proper health care service are important cause.

This is also applicable for heart failure patient. Patient diagnosed to have DCM develops PAH during the natural course. Most of the patient with DCM etiology remains unknown as the population group not well oriented with the disease course nor they were alert. Idiopathic PAH is an important cause of PAH as usual for other geographic area. Though more younger

population shows manifestation. Poor general health, lack of hygiene, food habits, addiction perhaps play the key role. As the population mostly are manual worker 6 min walk test results better when compare with echocardiographic parameters for level of PAH. congenital heart disease also play a major role for prevalence of PAH. In our study eleven patients was found to have PAH due to uncorrected CHD. CHD due to L->R shunt with either Hyperkinetic or fixed PAH found. Two patient had Eisenmenger syndrome were due to uncorrected large PDA.

PAH due to primary lung disease either obstructive or primary restrictive pattern is an important cause of secondary PAH. Though the manifest mainly late adult age ten patient in our population group shows PAH due to primary lung disease as diagnosed by PFT and HRCT chest. PAH due to lung involvement in rheumatological disorder is widely prevalent. Nine patient in our study group found PAH. They are now getting medicines for Rheumatological problem. Their PAH are variable during the course of illness in time to time. Portal HTN and advanced CKD cause immunological insult to the Pulmonary vascular bed causing progressive PAH. One patient in our study group was sero +very for HIV. Patient had thalassemia with history of multiple blood transfusion.

Conclusion

Etiology of PAH is diverse. Prognosis of advanced PAH is poor. Treatment cost is cumbersome and beyond the imagination of middle-class and economically challenged people. Proper diagnosis at early stage may improve survival, may cure and at least save the money of poorer pocket.

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