

Pulmonary Arterial Hypertension in a Patient Presenting to a Tertiary Care Hospital in Kolkata : Case Report

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Summary

Pulmonary artery hypertension (PAH) is a progressive condition where pulmonary vascular resistance and pressure leading to right heart failure and death as right ventricle becomes enlarged and weakened. In this case study PAH was developed for the patient from aortic stenosis. The author also discusses specific causes, pathophysiology, clinical manifestations, treatments and complications of PAH. PAH is defined as increase a mean pulmonary pressure of at least 25mm of Hg at rest. It is a life threatening disease that affects pulmonary arteries that carry blood from heart to the lung. PAH related to aortic stenosis increases morbidity and mortality.

Incidence of PAH is common throughout the world. PAH is a heterogeneous, haemodynamic, and pathophysiological state. The occurrence and consequences of disease is a significant burden in India and other developing countries. It is observed that the incident occurs approximately 1-4 persons per million populations per year in industrialised countries (Chopra et al, 2012).

Literature Review

In 2007, a retrospective observational study depicted that 626 patients with severe aortic stenosis had PAH and the pressure was measured by Doppler echocardiography. The result presented that out of 626 patients, 119 (19%) had severe PAH, defined as pulmonary artery systolic pressure 60 mmHg. Patients with severe pulmonary hypertension had a smaller aortic valve area ($p < 0.0001$), a lower left ventricular ejection fraction ($p < 0.0001$) indicating a higher filling pressure and a higher prevalence (Kapoor, 2008). It was also revealed that the survival rate of PAH may vary depending upon age, gender and exercise capacity. The survival rate is high after one year of occurrence of PAH whose complication stimulated from lung disease. These findings were also supported by WHO study (Organization Group, 2016). The said study concluded that 68 percent, 48 percent and 34 percent patients suffering from PAH survived for 1 year, 3 years, and 5 years, respectively.

Cardiovascular disease (CVD) is a leading cause of death and disability and the absolute magnitude of CVD morbidity and mortality is staggering. Most of the public health attention being focused on atherosclerotic cardiovascular disease, but the disease condition of PAH remains largely unnoticed.

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It is also mentioned that PAH is prevalent in the developing world in a much larger magnitude as compared to the western world. Evaluations of disease prevalence is very difficult in the developing world because of geographic, economic, socio-cultural, and ethnic-diversity along with regional variations in healthcare infrastructure. No published estimations are available on the expected degree of the incidence of PAH in India.

Classification and Causes of Pulmonary Arterial Hypertension

According to WHO classification, there are 5 groups of PAH

WHO Group I – Pulmonary arterial hypertension (PAH): Idiopathic PAH; Drug- and toxin-induced; Associated conditions: HIV infection, portal hypertension, congenital heart diseases.

WHO Group II – Pulmonary hypertension secondary to left heart disease: Systolic dysfunction; Diastolic dysfunction; Valvular heart disease.

WHO Group III – Pulmonary hypertension due to lung disease, chronic hypoxemia: Chronic obstructive pulmonary disease (COPD); Interstitial lung disease; Sleep-disordered breathing; Alveolar hypoventilation disorders.

WHO Group IV – Chronic arterial obstruction: Chronic thromboembolic pulmonary hypertension (CTEPH); Other pulmonary artery obstructions.

WHO Group V – Pulmonary hypertension with unclear or multifactorial mechanisms: Haematologic diseases.

Pathophysiology

The pathophysiology of PAH can vary depending upon the aetiology. According to WHO classification of PAH, there is variety of reasons behind the PAH disease process. If secondary hypertension is considered it may

be due to cardiac disease that is explained in WHO classification II. In this classification, there are systolic and diastolic dysfunction along with valvular heart disease. In this case, the patient was having aortic stenosis along with grade II diastolic dysfunction. These can create pressure and volume overload that leads to change pulmonary vasculature. In the present case study, changed pulmonary vasculature leads to increase vascular resistance that progress increased right ventricular (RV) after load and right ventricular failure (RVF). On the other hand, persistent PAH contributes to fluid overload and that leads to left ventricular failure (LVF) followed by RVF. The disease is progressive and hence, there are some compensation for counteract with pulmonary endothelium injury. To halt the progress of the disease it is required to decrease production of potent vasodilator prostacyclin and nitric oxide (NO), which inhibits platelet activation and have anti-proliferative process. It can also be stated that increased production of vasoactive compound endothelin, another compensation which effects on inflammation, vasoconstriction and hypertrophy. Beside all these compensations release of inflammatory mediators like interleukins (IL), cytokines, tissue necrotic factors (TNF) lead to more injury on pulmonary vasculature.

In this context, overall above-mentioned incidents are predominant factors of the disease progressions for this case.

Discussion

History of the patient: A 54 years old male patient was admitted in tertiary care hospital on 24 March 2018 under cardiology team with complaints of shortness of breathing, oedema in periphery and scrotal edema. Patient was admitted on 18 March 2018 with same complaint. There he developed cardiac arrest and was put on ventilator, and released from ventilator the next day. In this hospital, the patient developed left ventricular failure and haematuria.

Past medical history: The patient was a known case of diabetic mellitus type II since 25 years and hypertension since 7-8 years. He had family history of diabetic mellitus type II with his mother and sister.

Surgical history: The patient had a past surgical history of PTCA (stenting on distal RCA and proximal mid LAD) in 2017 due to double vessels disease (DVD).

Physical Examination

Vital sign assessment: BP - 130/90 mm Hg; Pulse - 75bpm; Respiration - 23/min; SPO₂ - 78%.

Inspection

Chest shape-normal; Pallor - No abnormality (N/A)detected; Neck vein distension - N/A; Central cyano-

sis - N/A; Peripheral cyanosis - N/A; Clubbing of nails - N/A

Palpation

Pulse rate - Normal; Pulse rhythm - Regular; Pulse volume - Bounding; Extremities- warm; Capillary refill time- < 3 sec; Oedema- present in upper and lower extremities , grade3; Peripheral pulses- Feeble

Percussion: No abnormality (N/A) detected

Auscultation

S₁ and S₂ - present; Heart murmur - Audible at the site of second intercostal space along the right sternal boarder; Air entry bilateral; Breath sound - Crepitation present on both lung.

Patient was having aortic stenosis and grade II diastolic dysfunction. According to WHO gradation said patient was under group II pulmonary artery hypertension

Investigations

- Echocardiography –mild concentric LV hypertrophy, regional wall motion abnormality, LV systolic dysfunction with EF=50%, grade II diastolic dysfunction and group II PAH.
- ECG: Reveals RVH, right axis deviation, depressed ST segment and T wave inversion.
- Chest X-ray: Lung congestion both side
- USG: Hepatomegaly with alter echotexture, Gall bladder wall mild thickened, increased echogenicity of pancreas, altered echogenicity of kidney with prominence of pyramid.
- USG KUB: Chronic renal parenchymal disease, prostate 15 gm.
- Laboratory investigations (Table 1)
- In ABG: PH-7.37,PCO₂-49.1mm of hg, PO₂ - 71.2 mm Hg

Diagnosis: Moderate pulmonary artery hypertension

Treatment: Different studies reveal that PAH cannot be cured, but the goal of the treatment is to restore right ventricular function and improvement of patient's symptoms.

Maintaining hydration is a challenging part of the treatment along with vasopressor or, inotropic support, vasodilator to reduce patients symptoms.

In the present case study, as the patient was having history of HTN, he used to get tablet Amlodipin 5 mg, BD for reducing cardiac contractility, tablet Prazopress 5 mg BD for dilating blood vessels and decreasing resistance. Moreover, as there is a lack of O₂ supply to myocardium and chances of developing an-

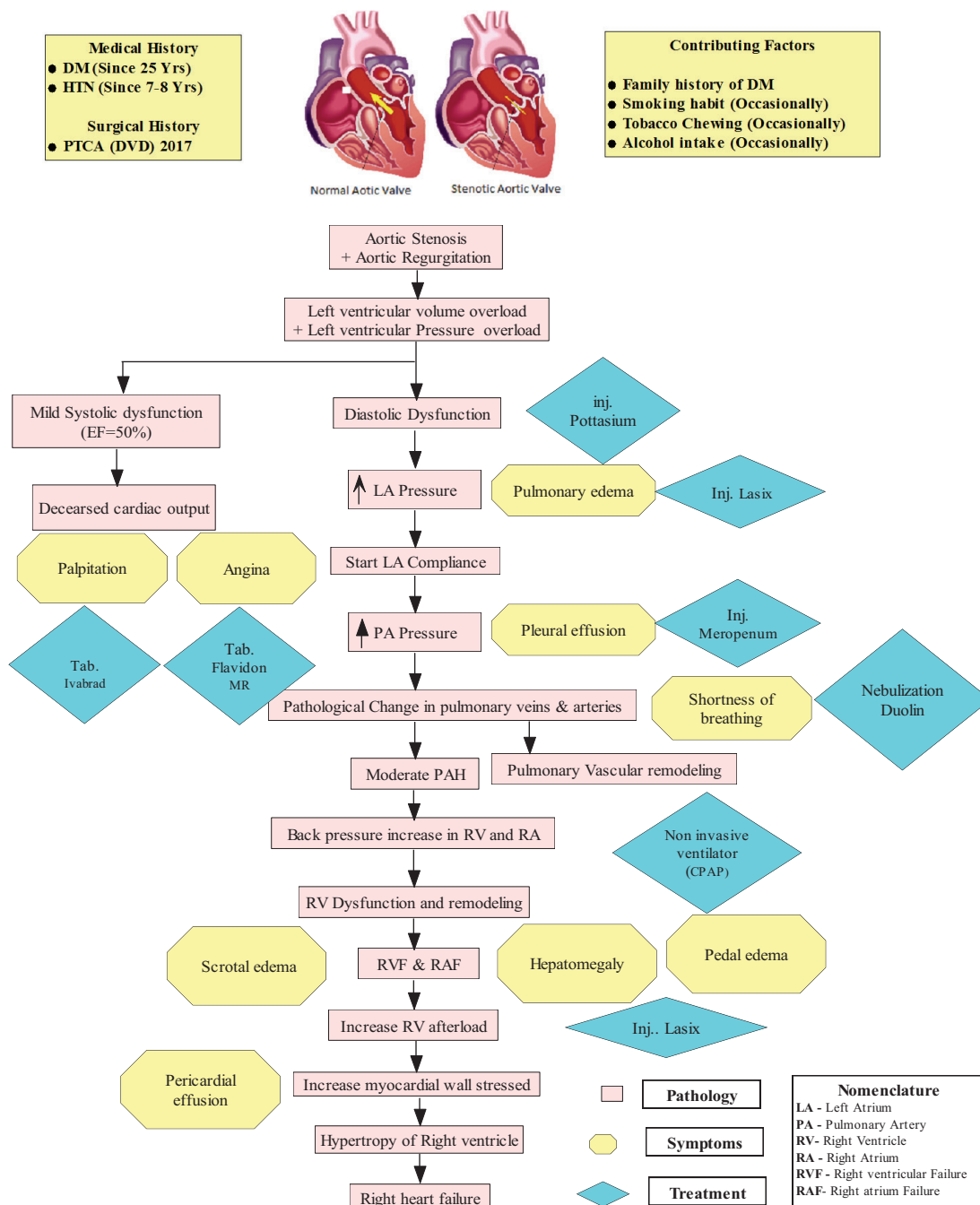


Figure 1: Clinicopathocorrelation of PAH and RVF

gina pain, patient used to get tablet Flavidon MR 35 mg BD for decreasing O₂ requirement of myocardium by improving myocardial glucose utilisation through inhibition of fatty acid metabolism.

According to our present goal of treatment for improvement of pulmonary function, tablet Winlop 5 mg OD is given for relaxing wall of the blood vessels of the lung, tablet Abflo one cap. BD is given to reduce bronchial obstruction by regulating surfactant production. Besides that to improve pulmonary congestion nebulisation Duolin is also given. In this case intermittent non-invasive ventilator therapy was sup-

ported to prevent hypoxia as well as to reduce pulmonary vasoconstriction. Inj Lasix was used to reduce fluid overload and to combat RVF. Along with that inj. KCL 40 mg I/V was given to compensate potassium lacking for diuresis. Since this patient was suffering from DM, to control this metabolic disorder, patient was given inj. Human Actrapid 3-4 U S/C BD. For prevention of infections inj Meropenum was chosen as there was stagnation of interstitial fluid and possibility of development of infection. This need based treatment helped to reduce the complications of RVF and PAH.

Table 1: Investigations Result

Investigation	Result
Haemoglobin	7.8 gm/dl
Serum urea	3.43 mg/dl
Creatinine	4.74 mg/dl
GFR	12.93 ml/min / 1.73m ²
BUN	49 mg/dl
Potassium	3.3 mmol/l
Procalcitonin (PCT)	0.21 ng/ml
Prothrombin time	19.8 Sec
Control INR	13.6
	1.62 Sec
CRP	31.5 mg/dl and day by day it was increased up to 291 mg/dl
Pro BNP	9756Pg/ml

So it can be concluded that all the above mentioned treatment was justified to improve pulmonary as well as cardiac function.

Prognosis

The patient was having clinical stability for a particular period of time, but his condition became worsen after day 12 (i.e. 4.4.18) from the day of admission. Con-

ventional therapeutic, nursing management and meticulous vigilance on physical and diagnostic findings help the patient to stay remain stable, which is observed from previously mentioned physical and diagnostic findings as well as therapeutic regimen. On the other hand, patient was potential risk for developing different conditions with compensatory stage, as he had lung involved, severe oedema, along with PAH.

On the day 13 (i.e. 5.4.18) there was sudden onset of drowsiness and bradycardia. After cardiopulmonary resuscitation patient was intubated and was on ventilator support (PRVC mode). Again for this case, high Pro BNP revealed patient was on cardiac compensation. Other inotropic supports were started like inj. Dopamin and inj Noradrenalin to limit compensation for heart failure. It was identified that from PAH and RVF patient was developed multi organ failure as renal function was limited, patient had neurological deficit (GCS-E1V1M2) and had systemic sepsis. All these findings support that PAH and RV dysfunction may be largely mediated remodelling and subsequent inflammations.

Complication: Cor pulmonale is a condition that results from PAH, which causes the right side of the heart to

enlarge because of the increased work required to pump blood against high resistance through the pulmonary vascular system. This causes right sided heart failure (Fig 1).

Nursing Implications

Timely nursing assessment and proactive nursing care is needed to analyse and evaluate the case through history taking and physical examinations. In this case patient was diagnosed grade II pulmonary hypertension with right ventricular failure after analysing all the investigations and from patient's symptoms.

After recognising all the data, problem solving will be the next step. In this regard the goals of the problem solving are: to restore pulmonary and cardiac functions, to reduce fluid overload and to maintained nutritional balance. Table 2 presents the nursing implications to reach the defined goals:

We have to focus on cost-benefit by evaluating beneficial and harmful effect of the management process. In this case, therapeutic modalities adopted had beneficial

Table 2: Nursing Management

Diagnosis	Nursing implications
Decreased cardiac output related to cardiac failure as evidenced by multi-organ failure	- Continuous haemodynamic monitoring is maintained - Pulse rate, depth and volume are checked. - Assessed tissue perfusion - Assessed renal functions and intake output chart - Neurological functions-GCS, strength of limbs, pupillary reactions are checked
Fluid volume overload related to right heart failure as evidenced by presence of oedema	- Factors and physiological parameters associated with increased fluid volume and electrolyte loss are assessed - Assessed degree and location of oedema - According to fluid volume, physiological parameters I/V fluid are maintained - Prescribed medication is given along with patient's progress
Impaired gas exchange related to altered pulmonary vasculature as evidenced by patient was on mechanical ventilation	- Respiratory support is provided through mechanical ventilator (PRVC MODE) - Respiratory status is monitored through respiratory rate, lung auscultation. 45 degree elevation of the bed is maintained to prevent ventilator associated pneumonia - Ventilator modes are monitored and manipulate according to hemodynamic conditions, chest X-ray, ABG and SpO₂ analysis. - Early and aggressive chest physiotherapy is provided to reduce use of ventilator
Fear for death for unknown outcome of the disease process related to poor prognosis	- Continuous observation is done - Knowledge level and readiness for learning is determines - Assessed coping strategies - Extensive description is given orally to the relative regarding patient's day by day survival and explained their queries about patient - Responses of all instructions is evaluated - Holistic care is provided through collaboration with others: physiotherapist, dietitian and others supporting staff. - According to the patient's physiological status and investigation medical management is altered to prevent complications

effect for the patient.

Despite the discussion of progression this case cannot be concluded. Here prognosis was keenly observed and immediate actions were taken in any altered situations as needed. Therefore a critical care nurse plays a central role for survival through all the ladders of care.

Conclusion

The present case study deals with a patient aged 54 years with history of diabetic mellitus and PTCA. From several investigations and through physical examination he has been diagnosed as grade II PAH with aortic stenosis (AS) and RVF. The study also shows how the case was developed from PAH and how RVF advanced from PAH. Therefore, patient was treated as a critically ill patient having the larger clinical pathway. The impact of treatment was based on focused investigations i.e. CXR, ECHO, ECG, ABG analysis etc. Constant supervision, keen observation and heartiest endeavour of all health care personnel were admirable.

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