



## Chronic Thromboembolic Pulmonary Hypertension

Authors

**Maurya PK<sup>1</sup>, Ali MA<sup>1\*</sup>, Sureshkumar U<sup>1</sup>, Nair S<sup>1</sup>, Potdar PV<sup>1</sup>**

<sup>1</sup>Department of Respiratory Medicine, MGM Medical College, Kamothe, Navi Mumbai, India

\*Corresponding Author

**Dr M Arif Ali**

Email: [marifali87@gmail.com](mailto:marifali87@gmail.com), Mobile No-9890327036

### Introduction

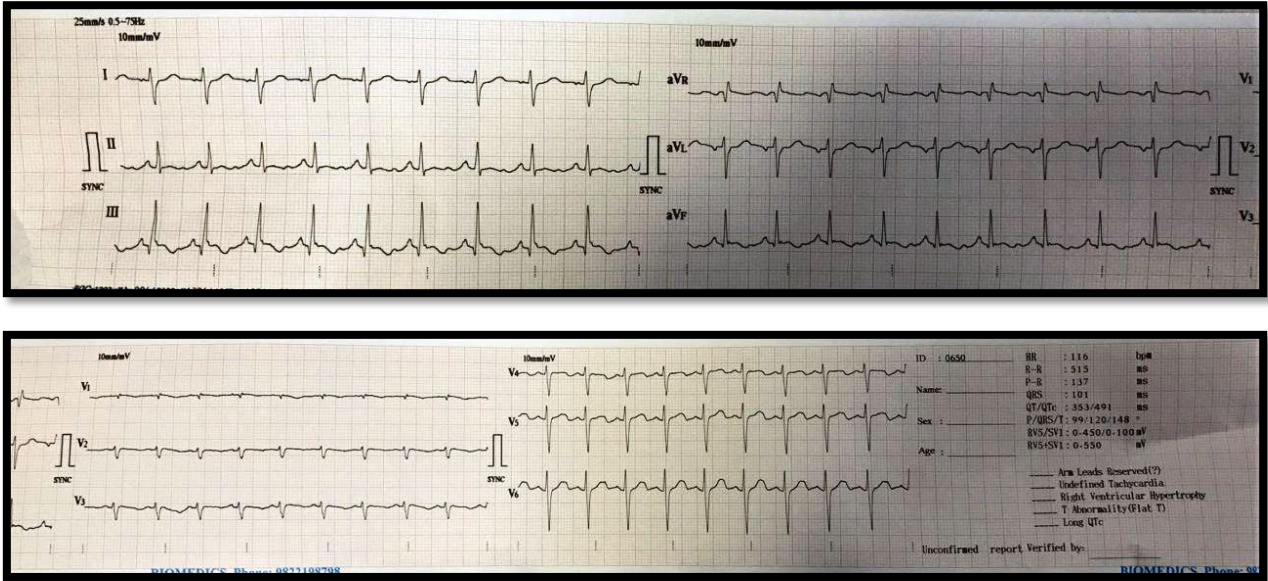
Chronic thromboembolic pulmonary hypertension is characterized by chronic obstruction of major pulmonary arteries by organized thromboembolic material. Untreated chronic pulmonary thromboembolism can result in pulmonary hypertension and right heart failure. It is the only form of pulmonary hypertension potentially curable by surgical or catheter based interventions. While early diagnosis is the key to successful treatment, CTEPH remains largely undiagnosed.

### Case Report

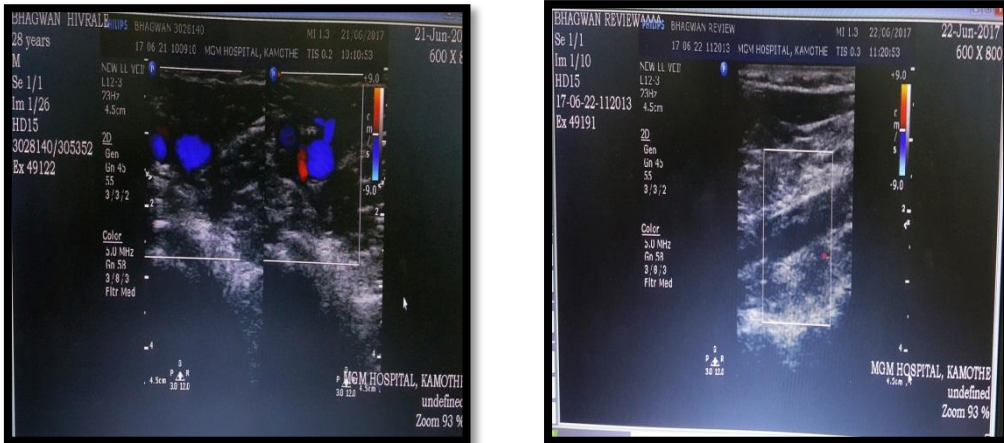
A 28 year old gentleman hailing from Nanded, farmer by occupation, current smoker, presented with complaints of breathlessness since 4 years associated with chest pain since 2 years, bilateral pedal edema more on the right side since 3 years. Retrospectively history of road traffic accident resulting in fracture and subsequent hospitalization was found. On examination patient was having sinus tachycardia, tachypnea with a raised JVP and positive hepato-jugular reflux. On cardiovascular examination loud P2 was auscultated. Routine investigation showed tachycardia on ECG, thrombocytopenia on blood

profile. Cardiac enzymes were within normal limit. Pulmonary function test was suggestive of mild airflow limitation with good bronchodilator response. 2D ECHO showed moderately dilated RA and RV with Grade I TR and IVC Dilation 1.7 cm and moderate PAH (PASP by TR jet 59 mm hg). Venous Color Doppler Study of left lower limb showed a 3 cm Baker cyst noted in popliteal fossa. Right lower limb popliteal vein lumen occluding thrombus noted in popliteal vein at popliteal fossa, approx. 5 cm compressible, extending to the origin of posterior tibial vein. Arterial doppler within normal Limits. HRCT chest showed dense calcification in the sub-carinal region, mosaic attenuation seen in both lungs right more than left side. As per Wells criteria for PE, score was 6.5 indicating a high chance of pulmonary embolism. CT Pulmonary Angiogram showed large saddle embolus noted in the right and left main pulmonary artery with calcification within. Perfusion defect in both lungs, right more translucent than left. Fibrotic nodule present in the left lower lobe supreme segment. Blood serum markers were negative for ANA, anti-ds DNA, anti-Scl 70, anti-CCP, RA Factor, CRP titers, d-Dimer.

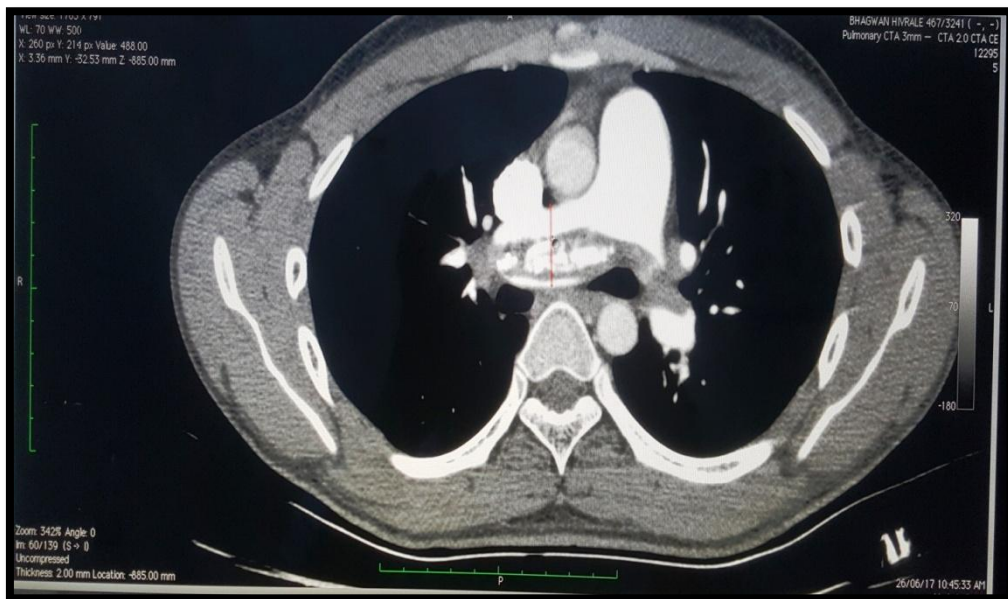
ECG



Lower Limb Doppler



CTPA



## Discussion

Incidence of CTPEH is 0.5-3.0% (CTEPH) in pulmonary hypertension cases which is recently documented to complicate 3.8% of acute pulmonary embolic events<sup>[3]</sup>

Following an initial episode of PE, almost 30% patients who develop CTPEH have no documented history of acute DVT or pulmonary embolism which hampers the diagnosis.

Even in patients receiving appropriate treatment for an acute pulmonary embolism incomplete resolution occurs in a significant proportion of patients, placing them at risk of developing CTEPH<sup>[5][6][7]</sup>.

Trauma to the lower limb can increase the risk of DVT which complicates as mild PE and can later lead to CTPEH.

Mortality of CTPEH is high in patients with mean PAP of more than 50 mm Hg

Treatment of choice is Pulmonary Thromboendarterectomy. Bosentan treatment in patients with inoperable CTEPH resulted in improvements in hemodynamics, exercise tolerance (6 min walk test) and functional class<sup>[4]</sup>.

Medical management is undertaken in patients who despite of an accessible thrombus choose not to undergo surgery. Other indications for medical management are a very distal thrombus or severe PAH leading to high risk during surgical intervention.

Usually first line medical management is anticoagulant therapy. In our case we used LMWH – Enoxaparin. Thrombolysis wasn't indicated. PDE – 5 inhibitors like sildenafil have shown limited benefits. Gradually the patient was shifted to oral anticoagulants, warfarin in this case, dose adjusted according to the PT/INR values. PT/INR maintained between 2 –3. Patient was discharged on oral anticoagulants.

## References

1. Michael Grippi; Jack Elias; Jay Fishman; Allan Pack; Robert Senior; Robert Kotloff

"Fishman's Pulmonary Diseases and Disorders"

2. Suqiao Yang, 1,2,3,\* Yuanhua Yang, 1,2,3,\* Zhenguo Zhai, 1,2,3,\* Tuguang Kuang,1,2,3 Juanni Gong, 1,2,3 Shuai Zhang, 1,2,3Jianguo Zhu,4 Lirong Liang, 2,3 Ying H. Shen,5 and Chen Wan "Incidence and risk factors of chronic thromboembolic pulmonary hypertension in patients after acute pulmonary embolism" J Thorac Dis. 2015 Nov; 7(11): 1927-1938.doi: 10.3978/j.issn.2072-1439.2015.11.43
3. Pengo V, Lensing A W, Prins M H. *et al* Incidence of chronic thromboembolic pulmonary hypertension after pulmonary embolism. N Engl J Med 2004;350:2257–2264.
4. Hughes R, George P, Parameshwar J. *et al* Bosentan improves inoperable chronic thromboembolic pulmonary hypertension. Eur Respir J 2006;19:138–143
5. Remy-Jardin M, Louveigny S, Remy J. *et al* Acute central thromboembolic disease: post therapeutic follow-up with spiral CT angiography. Radiology 1997;203:173–180
6. Ribeiro A, Lindmarker P, Johnsson H. *et al* Pulmonary embolism. One-year follow-up with echocardiography Doppler and five-year survival analysis. Circulation 1999;100:1280–1283
7. Wartski M, Collignon M A. Incomplete recovery of lung perfusion after 3 months in patients with acute pulmonary embolism treated with antithrombotic agents. J Nucl Med 2000;41:1043–1048.