

# A Case of Chronic Thromboembolic Pulmonary Hypertension in Association with Deep Vein Thrombosis and Pulmonary Embolism: A Case Report of a Young Female in Bangladesh

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## Abstract:

Chronic thromboembolic pulmonary hypertension (CTEPH) is classified as Group 4 Pulmonary Hypertension (PH) in current categorization by WHO. The initial suspicion with clinical presentation, then screening with imaging and confirmation with Right heart catheterization (RHC) are the steps of diagnostic algorithm for CTEPH. We report the case of a 35 years old female presented with progressive swelling of left lower limb for 2 months and shortness of breath for 1 and half months. Examination revealed respiratory rate 28 breaths/min,

pitting edema of left leg, left parasternal heave and palpable P2. Doppler study of both lower limbs and Computed tomography pulmonary angiography (CTPA) confirmed the diagnosis of deep vein thrombosis of left leg (DVT) and pulmonary embolism (PE), respectively. The echocardiography suggested presence of pulmonary hypertension. We approached the patient with conservative management and discharged her home, once she improved, with advice of life-long anticoagulation.

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## Background:

World Symposium on Pulmonary Hypertension (WSPH) categorized CTEPH as group 4 PH.<sup>(1-6)</sup> The epidemiology of CTEPH is variable across the ethnicities; the commonest presentation at an average age of 63 years, without any gender-specific predisposition.<sup>(2,3,7)</sup> The disease entity was underrepresented due to lack of availability of gold-standard investigations in the past.<sup>(1,8)</sup> With the growing consciousness among the physicians and availability of diagnostic tools, cases of CTEPH have been frequently reported in the recent years.<sup>(1)</sup> The reported incidence rate is 0.9 cases per million.<sup>(3)</sup> The

long-standing CTEPH progresses to right heart failure with high mortality rate.<sup>(2,3)</sup>

An analysis of the data from international registry of CTEPH patients evidenced a history of acute pulmonary embolism (PE) and a history of vein thrombosis in 74.8% and 56.1% of them, respectively.<sup>(6)</sup>

We present the case of a 35-year-old female with DVT with chronic pulmonary embolism whose physical findings and echocardiogram suggestive of pulmonary hypertension.

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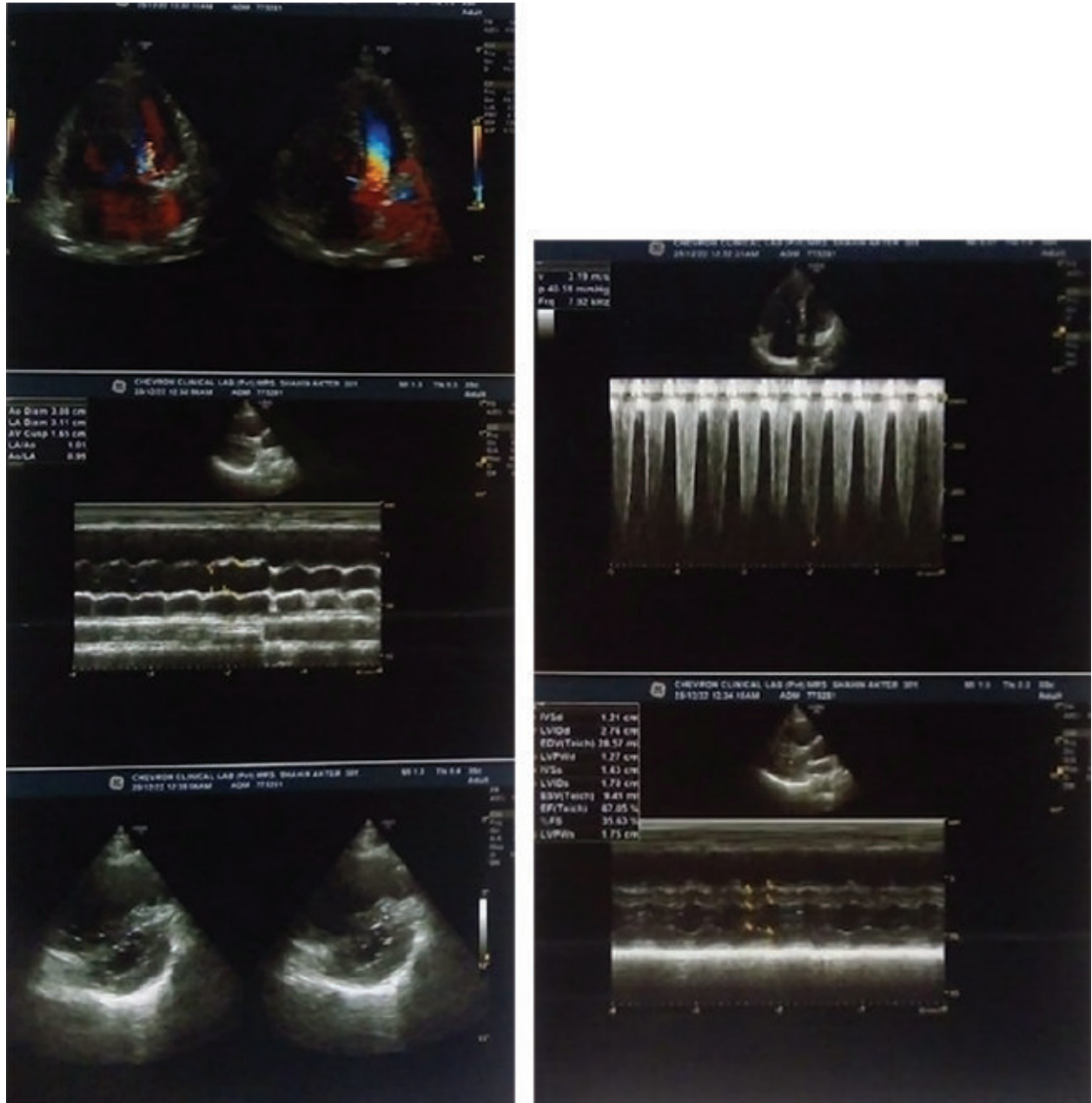
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**Case presentation:**

A 35-year old hypertensive female got admitted with progressive swelling of left leg for 2 months and shortness of breath for 1.5 months. The swelling was gradual in onset. Her shortness of breath can be described as initially NYHA class II, later deteriorating to NYHA class IV associated with orthopnoea, paroxysmal nocturnal dyspnoea and productive cough. On physical

examination, blood pressure 100/70mmhg, Respiratory rate 28 breaths/min, pulse rate 92 beaths/min, swollen left leg with pitting, tender edema. Precordial examination revealed palpable p2 and left parasternal heave.

Echocardiography showed dilated right atrium, right ventricle, moderate tricuspid regurgitation, systolic pulmonary arterial pressure (sPAP) 60mm Hg.



**Fig.-1:** Two-dimensional and m-mode Echocardiography suggestive of pulmonary hypertension

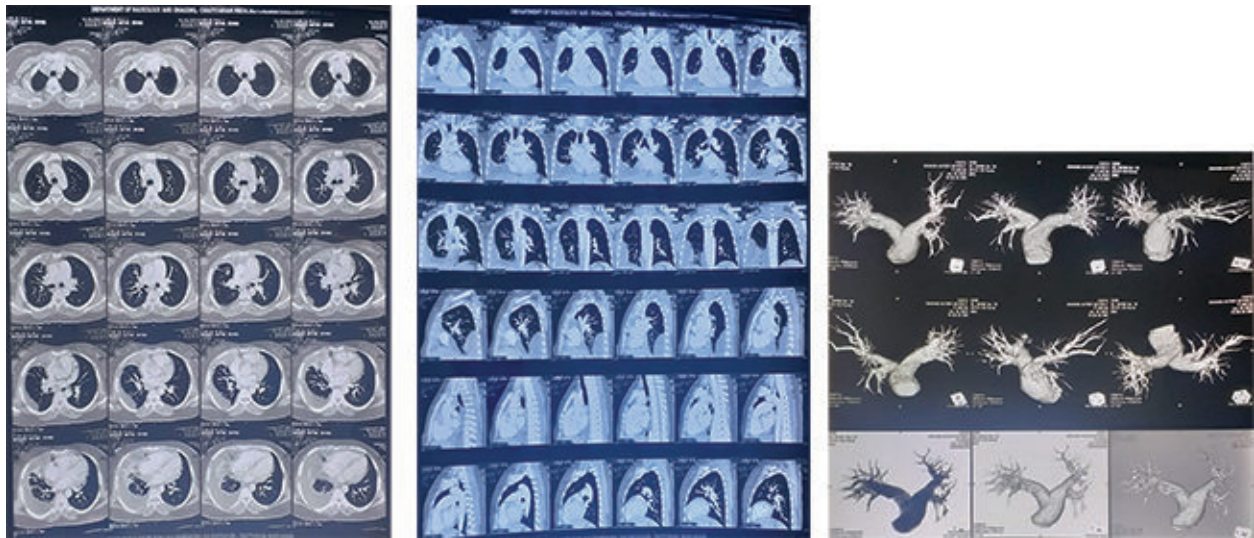
Color doppler ultrasonography of both lower limbs revealed presence of deep vein thrombosis involving the left femoral-popliteal segment with no sign of recanalization.

CTPA showed bandlike defect in right pulmonary artery (PA). Short segment filling defect within anterior segment branch of right upper lobar pulmonary artery, with webs in lower lobar PA extending into posterior segment artery with reduced calibre and poor contrast opacification and layered filling defects at PA bifurcation.

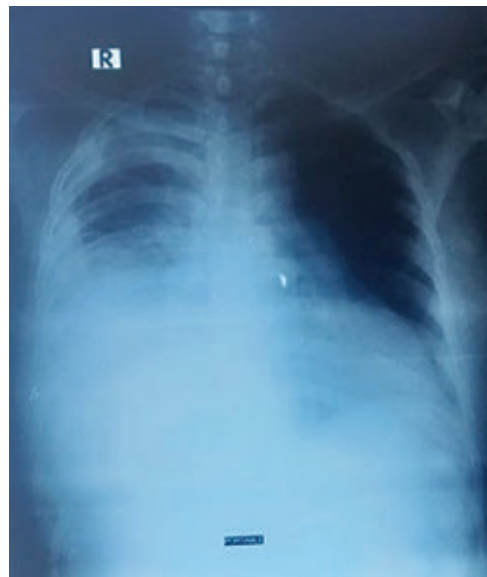
Chest radiograph showed cardiomegaly with bilateral pleural effusion more prominent on the right side.

D-dimer was raised with a value of 2.76mg/l(reference range <.50mg/l).

Due to unavailability of RHC and digital subtraction angiography (DSA) in our centre, we couldn't perform these tests. We treated the patient conservatively. The patient was discharged accordingly with the advice of lifelong anticoagulation.



**Fig.-2:** CTPA shows filling defects in right pulmonary artery and left pulmonary artery, favouring pulmonary embolism



**Fig.-3:** Chest x ray showing cardiomegaly with bilateral pleural effusion.

### Discussion:

CTEPH is a distinct entity of pulmonary hypertension that involves subsegmental, segmental, and main branches of the pulmonary arteries. (2,3) Presence of fibrotic thromboembolic materials cause an increase in pulmonary vascular resistance (PVR). (3) Pulmonary vasculature undergoes remodeling.(3,9)The condition consistent with the pre-capillary PH (mean pulmonary arterial pressure >20 mmHg, pulmonary artery wedge pressure >15 mmHg, pulmonary vascular resistance >2 Woods Unit).(1,3) Insufficient clot resolution are key pathophysiological feature of this disease.(1)

Preceding events of DVT and PE have been frequently reported with CTEPH; but not essential for diagnosis in all cases.(1-6,9) A large prospective study showed that cumulative incidence of CTEPH in patients with PE diagnosed for the first time was 11.2% at 3 months, 12.7% at 1 year, 13.4% at 2 years, and 14.5% at 3 years. (8) The other predisposing conditions include permanent intravascular devices (pacemaker, long-term central lines, ventriculoatrial shunts), inflammatory bowel diseases, essential thrombocythaemia, polycythaemia vera, splenectomy, antiphospholipid syndrome, high-dose thyroid hormone replacement, and malignancy.(1)

Initial screening for the presence of intermediate/high probability of PH with echocardiography is mandatory. Systolic pulmonary artery pressure (sPAP) >60 mmHg is a predictor of CTEPH.(1)

A ventilation/perfusion (V/Q) lung scan (preferably single-photon emission computed tomography [SPECT]) has sensitivity of 90–100% and a specificity of 94–100% in detection of CTEPH. (2) V/Q scan is indicated in patients with persistent or new-onset dyspnoea or exercise limitation following PE, also in suspected or newly diagnosed cases of PH to rule out or detect signs of CTEPH.(1)

CTPA has 100% sensitivity, 93.7% specificity in diagnosis of CTEPH and supports the technical evaluation for surgery eligibility. (1-3)The typical signs observed in pulmonary angiography are ring stenosis, reticular lesions, tortuous lesions, and complete vascular obstructions, filling defects, webs or bands in the PAs, PA retraction/dilatation, mosaic perfusion, and enlarged bronchial arteries. (1,3)

Digital subtraction angiography (DSA) is mainly used to confirm CTEPH with characterising vessel morphology and assess treatment options .(1,2)

Right heart catheterization is the gold-standard confirmatory test to establish CTEPH.(1-6)

CTEPH is a potentially treatable disease.(1,3) The current guidelines suggest multidisciplinary care and multimodal approach of combinations of pulmonary endarterectomy (PEA), Balloon pulmonary angioplasty (BPA), and medical therapies to target the mixed anatomical lesions.(1) Treatment of choice is pulmonary endarterectomy with accessible PA lesions.(3) BPA is preserved for the cases which are technically inoperable. It is also indicated in residual PH after PEA and distal obstructions amenable to BPA.

Vitamin K antagonists (VKAs) remain preferred choice till the date, particularly in patients with antiphospholipid syndrome.(1) Non-vitamin K antagonist oral anticoagulants (NOACs) have been investigated in several studies, but stronger evidences are required.(1) Treprostinil (subcutaneous injection) and Riociguat have been approved for inoperable CTEPH and the residual PH after PEA.

Long-term follow up should be emphasized. (1)

### Conclusion:

CTEPH is a rare form of pulmonary hypertension which needs appropriate recognition for management purpose. We identified a younger case in comparison with the average age of presentation in CTEPH. In spite of limitations of availability of diagnostic tools in our centre, we attempted to manage the case with the existing facilities.

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