

中文題目：慢性血栓栓塞性肺動脈高壓的診斷：從一位55歲漸進性呼吸困難的女性

英文題目：Diagnosis of Chronic Thromboembolic Pulmonary Hypertension in a 55-year-old female with Progressive dyspnea

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### **Abstract:**

A 55-year-old female with a prior history of hypertension and old CVA presented to the ER with dyspnea on exertion and lower legs edema on January 20. At our ER, the initial pulse oximeter showed oxygen saturation of 88% on room air. After serial survey, right pulmonary trunk embolism was diagnosed in the chest CT and she was admitted for anticoagulation treatment. Echocardiogram in January 2015 showed borderline LV systolic function (LVEF 47%) without pulmonary hypertension. Then she received oral anticoagulation treatment in the OPD.

However, progressive dyspnea still mentioned and echocardiogram in May 2015 showed pulmonary hypertension (estimated PASP 60 mmHg). Lung function test was checked and mild restrictive ventilator defect was found. Then V/Q scanning (Tc99m DTPA aerosol ventilation scintigraphy) was arranged and it showed residual segmental perfusion reduction in the RML/ partial RLL, related prior embolism. Under the suspicion of chronic thromboembolic pulmonary hypertension (CTEPH), right heart catheterization and coronary angiography were performed in June 2015. Pulmonary hypertension (mean PA 57mmHg) was confirmed and pulmonary angiogram demonstrated right middle and lower pulmonary artery filling defect while the coronary angiogram showed normal results. Otherwise, autoimmune, hematologic, systemic or metabolic disorders were excluded after serial laboratory tests. Finally, CTEPH was diagnosed and the Six-Minute Walk Test (6MWT) was performed with a value of 226 meters only.

We had consulted cardiovascular surgeons for assessment of pulmonary endarterectomy but an inoperable condition was told. Then we tried to apply for riociguat (a direct stimulator of sGC, which increases intracellular cGMP, inhibits endothelial dysfunction, and vascular remodeling) and then approved successfully. After sequential titrating dosage, this patient got much improvement in dyspnea and 6MWT.

### **Conclusions:**

1. Diagnosis of CTEPH is still challengeable in clinical practice.
2. The introduction of recent FDA-approved medication, Riociguat, will facilitate its treatment and patients' clinical outcome.