Diagnosis of chronic thromboembolic pulmonary hypertension after acute pulmonary embolism

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ABSTRACT Chronic thromboembolic pulmonary hypertension (CTEPH) is the most severe long-term complication of acute pulmonary embolism (PE). Untreated CTEPH is fatal, but, if diagnosed in time, successful surgical (pulmonary endarterectomy), medical (pulmonary hypertension drugs) and/or interventional (balloon pulmonary angioplasty) therapies have been shown to improve clinical outcomes, especially in case of successful pulmonary endarterectomy. Early diagnosis has however been demonstrated to be challenging. Poor awareness of the disease by patients and physicians, high prevalence of the post-PE syndrome (i.e. persistent dyspnoea, functional limitations and/or decreased quality of life following an acute PE diagnosis), lack of clear guideline recommendations as well as inefficient application of diagnostic tests in clinical practice lead to a reported staggering diagnostic delay >1 year. Hence, there is a great need to improve current clinical practice and diagnose CTEPH earlier. In this review, we will focus on the clinical presentation of and risk factors for CTEPH, and provide best practices for PE follow-up programmes from expert centres, based on a clinical case.