CHAPTER 73

CHRONIC TROMBOEMBOLIC PULMONER HYPERTENSION

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INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) is characterized pathologically by organized thromboembolic material and by altered vascular remodeling initiated or potentiated by a combination of defective angiogenesis, impaired fibrinolysis and endothelial dysfunction and classified within group 4 pulmonary hypertension (PH) (Table 1). [1]. These changes lead to PH and ultimately right ventricular failure. At 6th World Symposium on Pulmonary Hypertension (WSPH) meeting, a new threshold for PH (mean Pulmonary artery pressure (mPAP) >20 mmHg) and pre-capillary PH (combination of mPAP >20 mmHg, pulmonary arterial wedge pressure ⩽15 mmHg and pulmonary vascular resistance (PVR) ⩾3Wood Units) has been proposed by the [2]. However, we still use mPAP>25 mmHg in our clinical practice for the definition of CTEPH.

It is generally accepted that CTEPH develops in 4% of the acute pulmonary embolism cases [1]. Today, personalised treatment modalities which include pulmonary endarterectomy (PEA), balloon pulmonary angioplasty (BPA) and medical treatment are recommended for the treatment of the patients diagnosed as CTEPH. All patients should be referred to the expert centers that

Table 1. Pulmonary hypertension (PH) due to pulmonary artery obstructions

4.1. Chronic thromboembolic PH (CTEPH)
4.2.1.Sarcoma (high or intermediate grade) or angiosarcoma
4.2.2. Other malignant tumours Renal carcinoma Uterine carcinoma Germ cell tumours of the testis Other tumours
4.2.3. Non-alignant tumours Uterine leiomyoma
4.2.4. Arteritis without connective tissue disease
4.2.5. Congenital pulmonary artery stenoses
4.2.6. Parasites Hydatidos

* Adopted from Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. Eur Respir J 2019; 53: 1801913 [https://doi.org/ 10.1183/13993003.01913-2018].

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