Diagnosis and Treatment of Chronic Thromboembolic Pulmonary Hypertension

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Chronic thromboembolic pulmonary hypertension (CTEPH) is the one of the PH causes, in which pulmonary thrombus or emboli is produced and organized chronically (not changed for more than 3 months with anticoagulant therapy) and result in stenosis or obstruction of the pulmonary arteries. The 5-year survival was less than 50% only with anticoagulants when mean pulmonary artery pressure (mPAP) is over 30 mmHg. CTEPH patients in Japan have been reported to progress from acute pulmonary embolism in only 5% and female dominant different from western counterparts. Most patients with CTEPH present with chronic dyspnea on exertion, are diagnosed as PH with echocardiography, suspected as CTEPH with V/Q scan, and are finally diagnosed with pulmonary angiography including right heart catheterization. The established treatment evidenced wit improved prognosis is pulmonary endarterectomy (PEA), which requires expert skill in surgeons, is not indicated in 20 to 40 percent of patients for high age, comorbidity or predominant peripheral lesions and sometimes leaves the lesions unable to be treated. To compensate for these drawbacks, the catheter-based treatment called BPA (Balloon Pulmonary Angioplasty) or PTPA (Percutaneous Transluminal Pulmonary Angioplasty) has been developed in Japan since 2006 with improvement of the techniques decreasing the complications such as PA injuries with the catheters and reperfusion pulmonary edema, which made this treatment abandoned in 2001 in USA. BPA or PTPA has been reported from Japanese groups to be almost compatible to PEA in outcomes, medical costs and mortality, although it needs several sessions and admissions to attain the same clinical results and overcomes the above-mentioned shortcomings of PEA. The newly developed pulmonary vasodilators are also the effective adjunct of treatment. Nowadays combining these treatments are leading to an accomplishment of better morbidity and mortality in patients with CTEPH.