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Current Management and Treatment Result of PAH

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[II-S13-01]Current Management and Treatment Result of PAH in China

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Pulmonary hypertension (PH) is a debilitating disease of the pulmonary artery branches characterized by increased pulmonary arterial pressure and pulmonary vascular resistance. It is often associated with progressive right heart failure and a poor prognosis. In the past 20 years, tremendous progress has been made in understanding the pathogenesis of PH and developing many new therapeutic approaches for PH. However, PH still remains a worldwide health problem not only in the Western countries but also in Asian-Pacific countries including China. The objective of this article was to review and highlight the current status regarding the prevalence, diagnosis and treatment of PH in China.The research on PH in China can be traced back to the early 1970s. The National Collaboration Group of Pulmonary Heart Diseases, organized by physicians and researchers in the field of cardiovascular disease and pulmonary vascular physiology, was established in 1973. The early research on pulmonary circulation in the 1970s was focused on pathogenic mechanisms, and the diagnosis and treatment of COPD, PH associated with high altitude and hypoxia, and cor pulmonale. Since the 1980s, the researches on PH have been established as one of the National Key Technology R&D Programs in China. Four Five-Year Plans have been implemented and made remarkable contributions to the innovation of research techniques and the improvement of clinical management of PH in China.China has a relatively large population of PH patients than in the United States and European countries, but the epidemiologic study is limited. A study from one of the national leading cardiovascular center in China has shown that among 106,640 hospitalized patients, 7,085 (6.6%) of the patients were diagnosed with PH. In the patients with PH, there were 65.9% whose PH originated from congenital heart diseases (CHDs), 22.6% from left-sided heart diseases, 5.7% from thrombotic diseases, 3.8% from IPAH, 0.9% from respiratory diseases, 0.6% from connective tissue diseases (CTDs), 0.5% from pulmonary vasculitis, and 0.03% from portal hypertensive diseases. PAH related to CHD is still the most common type of PH in China. The prevalence of PAH-CHD in China is much higher than previously reported in other registries in European countries. The prevalence of PAH associated with congenital systemic-to-pulmonary shunts in Europe and North America has been estimated to range between 1.6 and 12.5 cases per million adults, but the precise prevalence of PAH associated with CHD in China is still unknown.At present, three separate signaling pathways (the endothelin, nitric oxide and prostacyclin pathways) are known to be involved in the pathogenesis of PH. The drugs used for treatment of PH mainly consist of endothelin receptor antagonists,

phosphodiesterase-5 inhibitors and prostacyclin analogues. In Europe and the United States a variety of drugs have already been approved for PAH treatment in the past 20 years, whereas the era of advanced therapy for PAH in China was initiated since 2006. Inhaled Iloprost and Bosantan were approved for PAH treatment in 2006 and 2007 respectively. Before 2006, many drugs including calcium channel blockers, leukotriene receptor antagonists and traditional Chinese medicine like Qian-Hu and Ginsenosides were used for the treatment of respiratory diseases and PH. Recently, Chinese FDA approved Ambrisentan and Remodulin for treatment of PAH. Whereas Sildenafil, Tadalafil, Vaidenafil have been used off-label for patients with PAH in China. Macitentan and Riociguat still are not available in China now. Many clinical researches in China by using advanced therapy for PAH including Bosentan, Ambrisentan, Sildenafil, Tadalafil and Iloprost, have shown an effect on the patients with IPAH, PH due to COPD or PAH related to CHD including children, respectively. The advanced therapy for PAH can increase the 6MWD, improve the NYHA function class, and reduce the pulmonary vascular resistance. Recently, Chinese physicians have actively participated in many international clinical trails for treatment of PAH, like SERAPHIN trail, PATENT trail, CHEST trail and MAESTRO trail et al.Chinese physicians and researchers have made significant progress in the clinical management of PH, and participated in international associations and societies devoted to the research, treatment, and prevention of pulmonary vascular diseases and PH in the past 10 years, further progress remains necessary to improve outcomes of PAH patients.