Pulmonary arterial hypertension and autoimmune disease

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Pulmonary arterial hypertension (PAH) is a rare condition characterized by elevated pulmonary arterial resistance leading to right heart failure. PAH can be sporadic (idiopathic PAH, or primary pulmonary hypertension), familial (caused by germline BMPR2 mutations, a type II member of the TGF beta receptor superfamily), or related to other conditions including connective tissue disease, congenital heart disease, human immunodeficiency virus infection, portal hypertension, and appetite suppressant exposure. Prevalence of PAH is around 15 per million. The lack of specificity of PAH symptoms (mostly dyspnea) presumably leads to an under-diagnosis of this condition. Echocardiography is the investigation of choice for non-invasive screening. Measurement of hemodynamic parameters during right-heart catheterizing is mandatory to establish the diagnosis (mean pulmonary artery pressure > 25 mmHg and pulmonary artery wedge pressure < 12 mmHg). Acute pulmonary vasodilator testing should be performed with nitric oxide or prostacyclin during right-heart catheterization. Recent advances in the management of PAH including continuous intravenous prostacyclin infusion and endothelin receptor antagonists have improved markedly the patients' prognosis. Novel treatments such as inhaled iloprost and type 5 phosphodiesterase inhibitors have also been evaluated in this setting. Lung transplantation is the last option for patients deteriorating despite medical treatment.

Several treatments for PAH are now approved. With the exception of recent data from patients receiving prolonged epoprostenol therapy, the long-term effects of novel treatments are still unknown. There is a substantial need for long-term observational studies evaluating the different treatments in terms of survival, side effects, quality of life and costs. As head-to-head comparisons of currently approved therapies are not available, the choice of optimal treatment will be dictated by clinical experience and drug availability, as well as patient preference. The treatment of PAH has historically been restricted due to limited therapeutic options. Recent advances in our understanding of the pathophysiological and molecular mechanisms that may underlie PAH, with the subsequent availability of novel pharmacological therapies, provide renewed hope for both patients and their physicians. Advancing knowledge of

this devastating disease may ultimately lead to the development of therapies that ensure a better prognosis.