Pulmonary arterial hypertension (PAH), defined as a mean pulmonary arterial pressure greater than 25 mmHg at rest detected by right heart catheterization (RHC), is a fatal disease caused by small pulmonary artery obstruction as a result of vascular proliferation and remodeling. PAH is characterized by markedly elevated pulmonary artery pressure and increased pulmonary vascular resistance (PVR), frequently leading to right-sided heart failure and death. The pathophysiological changes of the pulmonary arteries in PAH include endothelial injury, proliferation, and hypercontraction of vascular smooth muscle cells, migration of inflammatory cells, and activation of several intracellular signaling pathways such as Rho-kinase pathway.

The possible prognostic factors of PAH include functional class (FC), exercise capacity (eg, 6-min walking distance, cardiopulmonary exercise testing), pulmonary hemodynamics (eg, mean right atrial pressure, PVR, right ventricular end-diastolic pressure), right ventricular function (eg, cardiac output (CO), clinical evidence of right ventricular failure), and plasma concentrations of brain natriuretic peptide (BNP), endothelin-1, uric acid, and troponin. Among these prognostic factors, plasma BNP concentrations and CO are considered as the most influential prognostic ones. Furthermore, it has been recently demonstrated that cardiac index (CI) normalization in response to the treatment is an independent new prognostic factor of pulmonary hypertension when CI is reduced at diagnosis. In this issue of the Journal, Kang et al demonstrated that non-invasive cardiac magnetic resonance (CMR)-derived pulmonary artery (PA) distensibility index correlates with RHC-derived PA stiffness, PVR, and PA capacitance. They also demonstrated with multivariate logistic regression analysis...
that CMR-derived PA distensibility index could be used to predict FC, which was evaluated by 6-min walking distance, in patients with PAH (Figure).

It has previously been shown that histopathological changes in PA during PAH progression modifies the elastic properties of the pulmonary vessel wall. Furthermore, it also has been reported that PA pressure linearly correlates with PA stiffness and that PA distensibility evaluated by echocardiography or computed tomography was introduced to easily measure PA stiffness, although these methods have some limitations in their reproducibility and variability. In this issue of the Journal, the authors have emphasized that the reproducibility and variability of CMR-derived PA distensibility index were presented to obtain both invasively and non-invasively measured PA hemodynamic data within 72 h, and that the CMR-derived PA distensibility index significantly correlated with the RHC-derived PA stiffness index. The authors also have mentioned that CMR-derived PA distensibility index predicts FC evaluated by exercise capacity in patients with PAH, which is related to the clinical outcomes and prognosis.

In some institutes, PAH patients are regularly followed-up by invasive RHC to evaluate the effects of PAH treatment on pulmonary hemodynamics. As the less invasive procedures are preferable in clinical situations, this non-invasive method with CMR-derived PA distensibility index is important to evaluate the progression and prognosis in patients with PAH. From this point of view, more effective and less invasive examinations are required to be developed in the near future.

References