Cardiac magnetic resonance imaging and echocardiography in pediatric pulmonary arterial hypertension

Editor:

We read with great interest the article “Magnetic Resonance Imaging of the Right Ventricle in Pediatric Pulmonary Arterial Hypertension” by Blalock et al.¹ They present highly valuable cardiac magnetic resonance (CMR) data on children with idiopathic pulmonary arterial hypertension (IPAH) without a secondary cause of PAH, such as congenital heart disease (CHD). It has recently been shown that in patients with chronic pressure overload the resulting right ventricular (RV) myocardial damage can be demonstrated by an impaired RV long-axis function.² Blalock and colleagues found an impaired RV function in children with IPAH. They describe an increased indexed RV end-diastolic volume (RVEDVi) and a reduced RV ejection fraction in comparison to healthy controls. For the convenience of the audience of *Pulmonary Circulation*, we want to add that their data are in agreement with results obtained in patients with PAH secondary to CHD (PAH-CHD).^{3,4} However, we find a higher RVEDVi in PAH-CHD patients, which might be related to their multiple surgical procedures. It has been shown that echocardiographic measures of systolic RV function, such as tricuspid annular peak systolic velocity and tricuspid annular plane systolic excursion, correlate with CMR data on patients with PAH-CHD.^{3,4} Considering that about half of PAH patients in childhood have underlying CHD,⁵ the CMR results of Blalock and colleagues and the respective echocardiographic results of Kassem et al.⁶ in patients with IPAH add up to our data in PAH-CHD patients and together provide a compact image of RV function in pediatric PAH patients. We want to thank the authors for addressing the need to investigate RV function parameters in children with IPAH. In the future, with increasing data in pediatric PAH patients, echocardiography, in addition to the gold standard method, CMR, may become the technique of choice for

routine assessment of systolic RV function in the follow-up of patients with PAH.

Gunther Gores, Gernot Grangl, Claus Weitzer, Martin Koestenberger, *Division of Pediatric Cardiology, Department of Pediatrics, Medical University of Graz, Graz, Austria* (martin.koestenberger@medunigraz.at, koestenbergerm@gmx.at)

REFERENCES

1. Blalock S, Chan F, Rosenthal D, Ogawa M, Maxey D, Feinstein J. Magnetic resonance imaging of the right ventricle in pediatric pulmonary arterial hypertension. *Pulm Circ* 2013;3(2):350–355.
2. Fang F, Henein MY, Yu CM, et al. Right ventricular long-axis response to different chronic loading conditions: its relevance to clinical symptoms. *Int J Cardiol* 2013;167(2):378–382.
3. Koestenberger M, Nagel B, Ravekes W, et al. Tricuspid annular peak systolic velocity (S') in children and young adults with pulmonary artery hypertension secondary to congenital heart diseases, and in those with repaired tetralogy of Fallot: echocardiography and MRI data. *J Am Soc Echocardiogr* 2012;25(10):1041–1049.
4. Koestenberger M, Nagel B, Avian A, et al. Systolic right ventricular function in children and young adults with pulmonary artery hypertension secondary to congenital heart disease and tetralogy of Fallot: tricuspid annular plane systolic excursion (TAPSE) and magnetic resonance imaging data. *Congenit Heart Dis* 2012;7(3):250–258.
5. Haworth SG, Hislop AA. Treatment and survival in children with pulmonary arterial hypertension: the UK Pulmonary Hypertension Service for Children 2001–2006. *Heart* 2009;95(4):312–317.
6. Kassem E, Humpl T, Friedberg MK. Prognostic significance of 2-dimensional, M-mode, and Doppler echo indices of right ventricular function in children with pulmonary arterial hypertension. *Am Heart J* 2013;165(6):1024–1031.