Early detection of pulmonary arterial hypertension: do not forget the right ventricle

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We read with great interest the Review by Lau *et al.* (Early detection of pulmonary arterial hypertension. *Nat. Rev. Cardiol.* doi:10.1038/nrcardio.2014.191), and we agree with the authors about the importance of early diagnosis of pulmonary arterial hypertension (PAH) to improve survival, particularly in young patients.¹ We would like to focus more attention on right ventricular (RV) adaptation and its relationship to outcomes during PAH.

Increased pulmonary pressure can occur independently of increases in pulmonary wedge pressure and pulmonary vascular resistance. In the latter instance, increased pulmonary arterial pressure strictly depends on pulmonary vascular properties.² In both cases, the increased afterload leads to several RV adaptations; this phenomenon depends not only on the severity of pulmonary vascular disease, but is also related to neuro-hormonal activation, myocardial perfusion, and cardiomyocyte metabolism.^{2,3} On the basis of RV volume dilatation or free wall thickness, we currently recognize two distinct RV patterns. Firstly, an adaptive pattern consisting of concentric remodelling (high ratio of mass to volume), increased myocardial wall thickness, preserved systolic function, and increased filling pressure. Secondly, a maladaptive pattern consisting of eccentric hypertrophy (low ratio of mass to volume), an increase in RV free wall, and concomitant systolic dysfunction.^{3,4}

Echocardiography is the most commonly used technique for screening and diagnosis of PAH; Rudski *et al.* published guidelines for its use in the assessment of the right heart.⁵ Echocardiographic assessment of the right ventricle can provide an accurate estimate of pulmonary arterial pressure based on the peak velocity of the regurgitant jet through the tricuspid valve, as well as an assessment of the RV and right atrial inflow structures.⁶ By measuring longitudinal systolic function by tricuspid annular plane systolic excursion (TAPSE) analysis, an estimation of the effective systolic function can be obtained.

Forfia and colleagues showed that, in patients with PAH, TAPSE was correlated with poor outcome—every 1 mm decrease in TAPSE was associated with an increased risk of death (HR 1.17, 95% CI 1.05–1.30, $P=0.006$).⁷ However, given the anatomical complexity of the right ventricle and the individual interpretation of echocardiography, this technique is not sufficiently accurate for routine clinical use. The anatomical position and shape of the right ventricle make 2D analysis difficult.⁸

An emerging technique for RV study is cardiac magnetic resonance (CMR). This imaging modality provides more precise information than echocardiography about RV volume and structure. Using cine CMR, we are able to detect with precision RV ejection fraction, stroke volume, and segmental and global parietal kinesis, as well as eventual dyssynchrony between the right and left ventricular chambers. The RV outflow tract and pulmonary valve morphology and dimensions can, therefore, be viewed using CMR analysis.⁹ PAH often causes an initial remodelling of the right side of the interventricular septum.⁹ Moreover patients with PAH can develop a systolic movement change of the interventricular septum, with systolic excursion towards the right side as a consequence of increased RV pressures.⁹ All these data provide important additional information about outcomes in patients with PAH, independently of pulmonary pressure values.¹⁰

We agree that early assessment of PAH is very important to begin appropriate treatment and possibly to improve outcomes. In our experience, a detailed evaluation of the right ventricle in terms of size and function seems to be much more important than single pressure measurement—assessments of RV enlargement and systolic dysfunction are pivotal for predicting outcome. Currently, an integrated algorithm including echocardiography, CMR, and cardiac CT, together with measurement of haemodynamic parameters, should lead to improved PAH characterization and risk assessment.

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Competing interests

The authors declare no competing interests.

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