Detection of pulmonary arteriovenous malformation by contrast echocardiography in pediatric hereditary hemorrhagic telangiectasia


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Background.— In hereditary hemorrhagic telangiectasia (HHT), assessment of pulmonary arteriovenous malformations (PAMs) may be difficult in pediatric patients. The aim of this study was to assess the reliability of contrast echocardiography in a pediatric population presenting with HHT.

Patients.— We prospectively studied 22 pediatric patients presenting with HHT. All these patients underwent transthoracic contrast echocardiography (TTCE) and low-dose thoracic computed tomography (CT). Each TTCE examination was performed using second harmonic imaging, allowing to improve the quality of 2-dimensional imaging. The contrast protocol consisted of the injection of agitated 5% glucose solution through an upper extremity vein. We used the classification proposed by Barzilai et al.: grade 0 means no opacification of the left ventricle after the first three cardiac cycles following contrast appearance in the right atrium, grade 1 means minimal opacification; grade 2, moderate; grade 3, extensive opacification without outlining the endocardium; and grade 4, extensive opacification with clear endocardial definition. We considered CT as normal when no PAMs or only one microscopic PAMs was detected.

Results.— Mean age of the population was $11 \pm 5$ years (12 male). A PAM was detected in 10 patients (45%) by CT. TTCE was feasible in all pediatric patients. Using TTCE, a grade 0 was found in four patients, a grade 1 in seven patients, a grade 2 in five patients, a grade 3 in six patients and no patient had a grade 4. In case of grade 0 or 1, no patient had a significant PAMs, whereas for grade 2 and 3, all patients excepted one had PAMs. The sensitivity and specificity of TTCE for the detection of PAMs was respectively 100% and 92%.

Conclusion.— Detection of PAMs by TTCE is feasible in pediatric patients presenting with HHT. The reliability of TTCE is high in this specific population. Low-grade classification could presumably allow to avoid CT irradiation in pediatric patients.