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IMAGE FOCUS

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Isolated left ventricular apical hypoplasia

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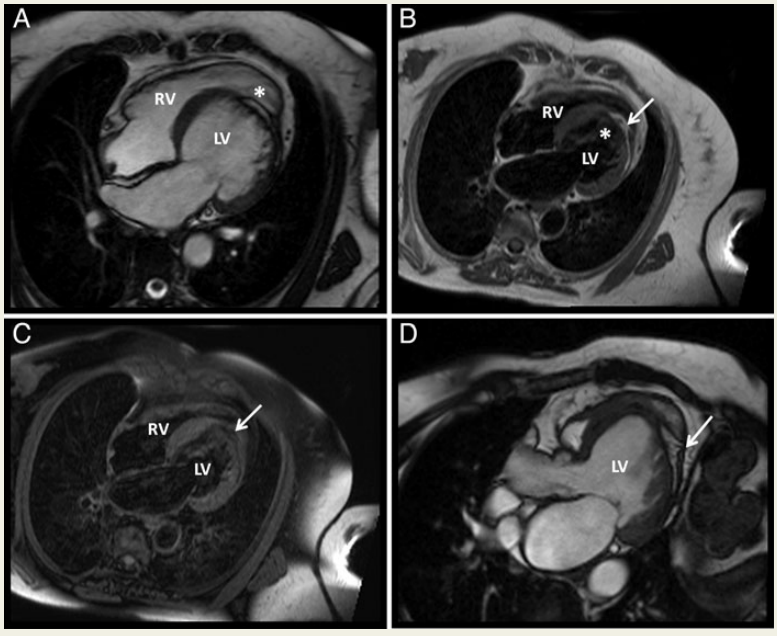

A 66-year-old woman with hypertension, dyslipidaemia and history of stable angina was referred for cardiological assessment after a transthoracic echocardiogram raising the suspicion of right ventricular arrhythmogenic dysplasia. The patient reported atypical chest pain. The 12-lead electrocardiogram showed sinus rhythm with a complete left bundle brunch block. The echocardiogram was repeated and demonstrated mild left ventricular (LV) systolic dysfunction with apical hypokinesia, abnormal interventricular septal motion and elongated right ventricle (RV). A myocardial perfusion scan showed a small fixed apical attenuation defect and absence of reversible ischaemia. To clarify diagnosis, cardiac MRI was performed and demonstrated the four phenotypical features of isolated LV apical hypoplasia: a bizarre LV with spherical configuration, truncated apex, interventricular septum bulging to the right and impaired systolic function (ejection fraction of 48%; *Panel A* and Supplementary data online *Video S1*); elongated RV wrapping around the deficient LV apex (*, *Panel A;* Supplementary data online *Video S1*); anomalous origin of anterior LV papillary muscle from the LV apex (*, *Panel B*); and replacement of the LV apex with fatty material [arrows, *Panels B* and *C*, which represent T1-weighted black-blood images without and with fat suppression (STIR) in four-chamber view, respectively], in this case with extension to the mesocardium of anterior, lateral and inferior midwall segments and continuity with the epicardial fat (arrow, *Panel D*, which represents cine-balanced steady-state-free precession in three-chamber view).

Isolated LV apical hypoplasia is a rare entity and MRI provides an excellent method to completely characterize the disease and evaluate the extent of myocardial fat replacement.

Supplementary data are available at *European Heart Journal – Cardiovascular Imaging* online.