Left Ventricular Noncompaction

A 55-year-old man with a history of biliary colic necessitating stent placement presented to the emergency department with exertional chest pain. Physical exam was consistent with fluid overload. Preliminary laboratory data revealed minimally increased cardiac enzymes and an elevated creatinine.

The patient underwent a transthoracic echocardiogram (TTE), which demonstrated a markedly dilated left ventricle, severely reduced systolic function, and diffuse hypokinesis with an ejection fraction of 27%. At the apex of the apical four-chamber view in end systole (A), the ratio of the noncompacted left ventricular myocardium (between arrows) to compacted myocardium (between arrowheads) exceeds 2:1, consistent with echocardiographic diagnostic criteria for left ventricular noncompaction (LVNC).1 A four-chamber fast imaging employing steady-state acquisition (FIESTA) image through the apex in end diastole from an EKG-gated cardiac MRI (B) confirms the elevated ratio consistent with LVNC. No delayed myocardial enhancement was seen on post-contrast sequences.

A short-axis FIESTA image with a large field of view shows the findings of LVNC in a different plane (white double-headed arrow), as well as numerous left renal cysts (black arrows), pancreatic ductal dilatation (black arrowheads), and pneumobilia (white arrow heads) associated with patient’s known autosomal dominant polycystic kidney disease (ADPKD) and prior biliary stent placement.

LVNC is a congenital disorder of myocardial development with genetically heterogeneous defects associated with diverse protein-related gene mutations.1 Imaging criteria of LVNC are based on determining the ratio of the noncompacted to compacted myocardium, with the midlateral, apical, and midinferior segments most commonly involved.1 There is association between ADPKD and LVNC genetic disturbances.2

REFERENCES