Abstract:

Amyloidosis is a term used for a whole group of diseases caused by deposition of a substance called amyloid into different tissues. Amyloid may be produced by a range of pathologic processes. Heart affliction is typical for only several types of amyloidoses. Heart involvement is then the patient`s prognosis major limiting factor. Diagnosis of heart amyloidosis is difficult especially for nonspecific symptoms and nonspecific findings obtained during common diagnostic procedures. The aim of this thesis was to evaluate usefulness of novel diagnostic methods, namely cardiac magnetic resonance with gadolinium enhancement and a simplified echocardiographic evaluation of left ventricular longitudinal strain, in diagnosing amyloid cardiomyopathy. In our first study we examined 22 patients with light chain amyloidosis by echocardiography and also with cardiac magnetic resonance with late gadolinium enhancement. We compared morphologic and functional parameters acquired by magnetic resonance examination, which is considered a gold standard for morphologic and functional measurements, with values obtained by echocardiographic measurement. Afterwards we evaluated the presence and eventually pattern of late gadolinium enhancement during cardiac magnetic resonance exam. From acquired data we conclude that the measurement of ejection fraction of left ventricle as well as one-dimensional morphologic characteristics, namely interventricular and posterior wall thickness and end-diastolic left ventricular diameter, are comparable between echocardiography and cardiac magnetic resonance examinations. Nonetheless in evaluation of left ventricular mass and volumes significant differences between these two types of measurements were found. Echocardiography overestimates left ventricular mass and underestimates left ventricular end-diastolic volume in comparison to cardiac magnetic resonance measurements in patients with light chain cardiac amyloidosis. From our work we conclude that the global patterns of gadolinium enhancement, either subendocardial or transmural, homogeneous or heterogeneous are typical for light chain amyloid cardiomyopathy. These patterns were found in 78% of these patients. In the second part of this thesis we evaluated measurement of relative apical sparing of longitudinal strain of left ventricle from a by us proposed simplified approach of this measurement only from apical four

chamber echocardiographic projection. We analysed 60 patients with diffusely thickened heart walls with reconcilable thickness of these walls caused by light chain amyloidosis, Fabry disease and arterial hypertension. Relative apical sparing of longitudinal strain of left ventricle evaluated only from apical four-chamber echocardiographic projection is significantly higher in patients with amyloid cardiomyopathy than in other investigated types of cardiac hypertrophy, but with large overlap of acquired values. According to multivariate analysis we conclude that the proposed simplified approach of measurement of relative apical sparing of longitudinal strain only from apical four chamber projection may be used but only with other characteristic markers of amyloid cardiomyopathy to differentiate light chain amyloid cardiomyopathy from other types of left ventricular hypertrophy, especially hypertrophy due to arterial hypertension or Fabry disease. Novel imaging methods namely cardiac magnetic resonance with gadolinium enhancement and longitudinal strain echocardiographic analysis can be used to improve our noninvasive differential diagnostic accuracy in diagnosing light chain amyloid cardiomyopathy.

Key words :

cardiac amyloidosis, echocardiography, strain, relative apical sparing of longitudinal strain, magnetic resonance, late gadolinium enhancement