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## **Magnetic resonance imaging in diagnosing fibrotic and cicatricial heart alterations in children**

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*The article provides an analysis of capabilities of magnetic resonance imaging with delayed contrast enhancement in diagnosing fibrotic alterations of varying severity. A hyperintense signal in the setting of myocardial delayed contrast enhancement programs indicates alterations of myocardial structure. Localization, severity and hyperintense signal size facilitate correct diagnosis and prognosis of complications development.*

**Keywords:** cardiac magnetic resonance imaging, delayed contrast enhancement, myocardial ischemia, myocardial fibrosis, cicatricial myocardial alterations, children.

## **INTRODUCTION**

Post-ischemic myocardial alterations are rarely observed in children. However, coronary bed structural anomalies causing myocardial ischemia, myocardites and cardiomyopathies may result in fibrous rearrangement of myocardium and, ultimately, in cicatricial myocardial alterations. Fibrous and cicatricial alterations may, in their turn, cause derangement of electrical conductivity and cardiac rhythm, myocardial remodeling and development of complications [1].

Cardiac magnetic resonance imaging (MRI) is an important method of appraising structural alterations of the heart. MRI allows diagnosing and appraising spread of the pathological process at such diseases as ischemic heart disease, cardiomyopathies, myocardites, storage diseases etc. [2].

The main method of appraising structural alterations of the myocardium is the so called delayed contrast enhancement consisting in different storage and leaching of a contrast medium on the basis of gadolinium salts from interstitial spaces of normal and structurally altered myocardia. The unaltered ("normal") myocardium appears black 7-20 minutes after administration of a contrast medium, whereas the structurally altered myocardium appears light [3].

Mild ischemia affects primarily endocardial layers. If ischemia takes a severer course, the pathological process affects the whole cardiac wall, thus causing a transmural infarction. That is why MRI visualizes post-ischemic alterations as hypertensive areas spreading from endocardial to epicardial layers and corresponding to any coronary artery's circulation [2].

Myocardites primarily involve central parts in the pathological process, which is why MRI visualizes a hypertensive signal deep inside the myocardium characteristic of inflammatory or fibrotic alterations [2]. Thus, ischemic alterations spread from the endocardium, whereas damage of the median layers and the epicardium supports non-ischemic etiology [4, 5].

This article is aimed at presenting clinical cases demonstrating capabilities of MRI with delayed contrast enhancement for diagnosing structural alterations of the myocardium in children.

## **PATIENTS AND METHODS**

The article presents cases of the 3 patients treated at the cardiology unit of the pediatrics research institute of the Scientific Center of Children's Health (Federal State Budgetary Institution); 2 of the children had myocardites of different severity and one patient underwent a surgery due to anomalous origin of the left principal coronary artery from the pulmonary trunk.

The examination procedure involved echocardiography (EchoCG) and cardiac MRI with delayed contrast enhancement in all the children. MRI was performed using an apparatus with field strength of 1.5 T. We used the following modes: CINE images FIESTA 2-ch, 4ch and short axis (TR = 4.0, TE = 1.7, slice thickness – 8 mm). A perfusion appraisal program would be launched at contrast medium administration (TR = 8.8, TE = 2.4, slice thickness – 10 mm) and after 10-15 minutes of delayed contrast enhancement along three axes (2D MDE 2-ch, 4-ch and short axis; TR = 5.8, TE = 1.7, slice thickness – 8 mm). Time of inversion (TI) was selected on an individual basis with program 2D MDE TEST (TR = 5.8, TE = 1.7, slice thickness – 8 mm). The contrast medium was used in the dose of 0.2-0.4 mmol/kg of body weight.

## **RESULTS AND DISCUSSION**

### **Patient 1**

Girl, 6 years of age. Dyspnea, anorexia, low body weight gain and physical developmental delay had been observed since the first postnatal month. Edemas and acrocyanosis developed at 3 months of age. The examination-based diagnosis was "Anomalous origin of the left coronary artery from the pulmonary trunk". Surgical correction of the vascular malformation was performed at the age of 6 months; it consisted in reimplantation of the left coronary artery in the ascending aorta and plastic repair of the pulmonary artery trunk with a xenopericardial patch. Condition stabilization was achieved in the postoperative period; no active complaints are reported.

EchoCG at the age of 6 years revealed small dilatation of the left ventricular cavity and Simpson's ejection fraction reduction down to 42%. Systolic volume remained unaltered. Aneurism-like protrusion of interventricular septum (IVS), increase in echogenicity of papillary muscles and mitral regurgitation, grade 1, were revealed. Fibrotic alterations of papillary muscles and IVS were suspected on the basis of the EchoCG, which is why cardiac MRI was performed. MRI revealed thinning of the apical segment's anterior wall. Delayed contrast enhancement visualized a hyperintense signal in the abovementioned area and the posterior papillary muscle corresponding to cicatricial alterations (pic. 1).

### **Patient 2**

Girl, 16 years of age. Complained of rapid fatigability and asthenia from the age of 11 years. Exertional dyspnea accompanied by cyanosis of the nasolabial triangle had been observed since 14 years of age. Diagnosis established at the local inpatient hospital – "Sporadic hypertrophic cardiomyopathy complicated by arrhythmogenic syndrome". The girl was prescribed symptomatic drug treatment. The girl continued to complain of stabbing heartaches at exertion and fatigability after discharge.

Examined at the Scientific Center of Children's Health at the age of 16 years. The condition at admission was considered severe due to pronounced cardiac failure. 24-hour (Holter) ECG monitoring revealed numerous extrasystoles and episodes of bi- and trigeminy. EchoCG results: Simpson's ejection fraction – 33%, impaired diastolic function and uneven wall thickening of the left ventricle, increased myocardial echogenicity.

MRI with delayed contrast enhancement revealed numerous areas with hyperintense signal diffusely located deep inside the myocardium of all the cardiac chambers (pic. 2). These alterations correspond, above all, to the pronounced post-myocarditis fibrotic alterations. Given the condition's severity and high degree of structural alterations of the myocardium, the child was put in the queue for heart transplantation. The girl died of unexpected rhythm disturbance 3 months after, before she could undergo heart transplantation.

### **Patient 3**

Girl, 9 years of age. The disease set on 2 years ago with pyretic fever accompanied by girdle pains in the stomach. The girl was hospitalized when the condition aggravated; 4-fold increase in the level of isozyme creatine phosphokinase (CPK-MB), increase in the level of serum troponin and ECG signs of intramural anterior wall myocardial infarction. EchoCG revealed ejection fraction's reduction down to 46% and hypokinesis of the left ventricular posterior wall migrating to the ventricular apex and septal areas. Established diagnosis – “Acute coronary syndrome. Myocarditis. Coronaritis. HF, stage 2A”; the girl was prescribed antibacterial, antiviral and symptomatic treatment, in the setting of which the child's condition improved, body temperature normalized and cardiac pains disappeared. The girl was discharged from hospital with improved condition and positive dynamics of laboratory instrumental examination results.

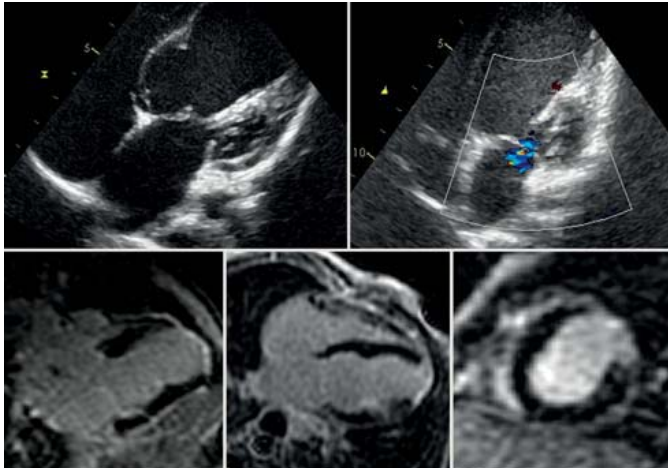
EchoCG at the age of 9 years revealed moderate left ventricular wall hypertrophy. MRI with delayed contrast enhancement revealed hyperintense signals in basal and middle segments of median septal myocardium layers; this indicates fibrotic alterations of the cardiac muscle resulting from myocarditis (pic. 3).

### **CONCLUSION**

Cardiac MRI is a safe method of X-ray diagnostics which allows appraising structure of the myocardium and determining the spread of fibrotic and cicatricial alterations of the cardiac muscle. Visualization of myocardial structural damage areas contributes to detection of the cardiac rhythm disturbance cause, sites for ablation and other interventions [1]. According to the studies, the MRI-based fibrosis score correlates with intensity of clinical symptoms, prognosis and rate of complications in patients with structural alterations of the myocardium [5-8].

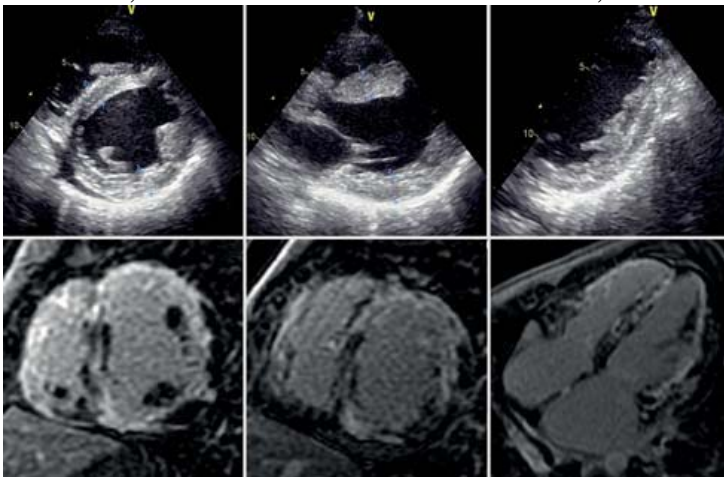
It ought to be mentioned that cardiac MRI is not used for diagnosing acute ischemic disorders. Moreover, the MRI-detected alterations are non-specific and usually merely indicate damage of the normal myocardial structure. However, delayed contrast enhancement visualizes localization, size and shape of the hyperintense signal; this allows assuming the cause of alterations in a patient with known clinical pattern of the disease [2, 8]. Differentiation between ischemic and non-ischemic nature of structural alterations is important for selecting treatment and rehabilitation tactics on any stage of patient management.

**Pic. 1.** EchoCG and cardiac MRI with delayed contrast enhancement (patient 1):  
 1A – EchoCG, apical 4-chamber view; 1B – EchoCG, apical 4-chamber view; 1C – MRI, 2D MDE 4-ch; 1D – MRI, 2D MDE 4-ch; 1E – MRI, 2D MDE short axis.



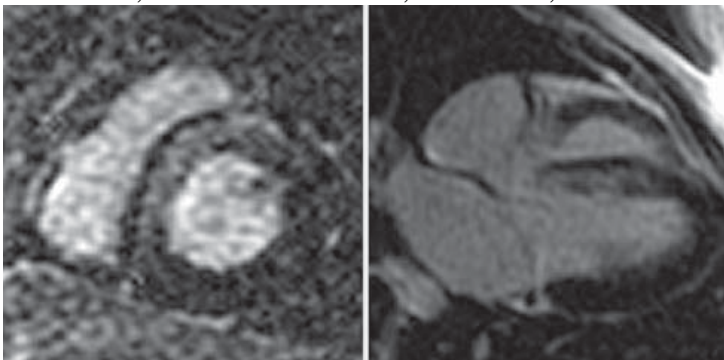
*Note.* The area of cicatrical alterations in the left ventricular apex is shown with arrows.

**Pic. 2.** EchoCG and cardiac MRI with delayed contrast enhancement (patient 2):  
 2A – EchoCG, parasternal view, short axis (papillary muscles); 2B – EchoCG, parasternal view, long axis (left ventricle); 2C – EchoCG, modified apical 2-chamber view; 2D – MRI, 2D MDE short axis; 2E – MRI 2D MDE short axis 4-ch; 2F – MRI, 2D MDE 4-ch.



*Note.* Numerous diffuse fibrotic areas in myocardia of both ventricles are shown with arrows.

**Pic. 3.** Cardiac MRI with delayed contrast enhancement (patient 3):  
 3A – MRI, 2D MDE short axis; 3B – MRI, 2D MDE 4-ch.



*Note.* Fibrotic areas in the interventricular septal myocardium are shown with arrows.

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