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ORIGINAL ARTICLE

Risk of delayed diagnosis in young patients with left ventricular non-compaction – a potential benefit of magnetic resonance imaging

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Abstract

BACKGROUND: Left ventricular non-compaction (LVNC) is a rare form of cardiomyopathy resulting from a disorder of endomyocardial morphogenesis associated with significantly increased risk of cardiovascular morbidity and premature mortality. Despite the widespread use of echocardiography, LVNC is commonly overlooked, often due to lack of knowledge about this disorder.

METHODS AND RESULTS: A complex diagnostic process and follow-up was analysed in 24 patients diagnosed with LVNC between March 2002 and February 2016 (16 boys, 8 girls; age at presentation 9 days – 18 years; follow-up 2–7 years). 17 patients were initially overlooked and followed-up for different diagnoses. After retrospective evaluation by a senior specialist in paediatric cardiology, LVNC was identified in 3 patients initially diagnosed with dilated cardiomyopathy, 11 patients followed-up with various forms of arrhythmias, and 3 patients with congenital heart disease. The diagnosis of LVNC was confirmed using magnetic resonance imaging in all patients. The classical triad of complications - heart failure, ventricular arrhythmias and systemic embolic events - was not confirmed in this study, electrocardiographic findings were abnormal in 87.5% of patients. Isolated non-compaction of the left ventricular myocardium was a dominant form of non-compaction.

CONCLUSIONS: The high variability of morphological findings and clinical manifestations of LVNC results in frequent overlooking of this disorder. Therefore, it is important to make the specialists more familiar with this condition and its pathology. Magnetic resonance imaging represents a conducive method to make correct diagnosis of LVNC under several specific conditions, particularly in case of non-conclusive echocardiographic finding.

INTRODUCTION

Left ventricular non-compaction (LVNC) or "spongy myocardium", is a rare congenital cardiomyopathy that can be diagnosed at any age. It was initially described by Chin and colleagues in 1990 in a paediatric series, and it is characterised by the presence of a thin compacted epicardial layer and an extensive non-compacted endocardial layer with prominent trabeculation and deep recesses, which communicate with the left ventricular cavity but not with the coronary circulation (Chin et al. 1990; Shemisa et al. 2013; Jenni et al. 2007; Reynen et al. 1997). It has been proposed that LVNC results from the arrest of the compaction process during the second month of embryological development, when segments of spongy myocardium fail to transform into compact, mature musculature (Reynen et al. 1997; Borges et al. 2003; Murphy et al. 2005).

The spectrum of morphological findings is very wide, ranging from hearts with a nearly absent compacted layer and an almost exclusively trabecular component, to hearts with prominent trabeculae and deep alternating recesses, but a well-represented compacted layer (Rooms et al. 2015). Although the usual site of involvement is the left ventricle, the right ventricle and septum can be affected as well (Sert et al. 2013). Non-compacted myocardium occurs either in an isolated form with absence of other cardiac anomalies or it is associated with congenital heart diseases, neuromuscular disorders, metabolic syndromes, connective tissue diseases, and haemolytic disorders (Alter & Maisch 2007; Digilio et al. 1999; Madan et al. 2010; Stöllberger & Finsterer 2006). The estimated prevalence of LVNC is 0.01% in adults and 0.14% in paediatric patients, and the clinical manifestations of this condition are highy variable, ranging from a complete lack of symptoms to disabling congestive heart failure, arrhythmias, and systemic thromboembolism (Weiford et al. 2004; Moon et al. 2006).

So far, the systematic studies on the diagnostic aspects and therapeutical outcomes of LVNC in pediatric population have been rather scarce. Moreover, due to high variability of morphological findings and clinical presentations, patients may be often overlooked and the correct diagnosis of LVNC is delayed (Cahil & Penicka 2014). Therefore, we aimed to provide a detailed summary of our experience with complex diagnostic process and follow-up in 24 young patients from childhood to adulthood diagnosed with LVNC with focus on the risk of misdiagnosis and potential benefit of magnetic resonance imaging, particularly in patients with inconclusive echocardiographic finding.

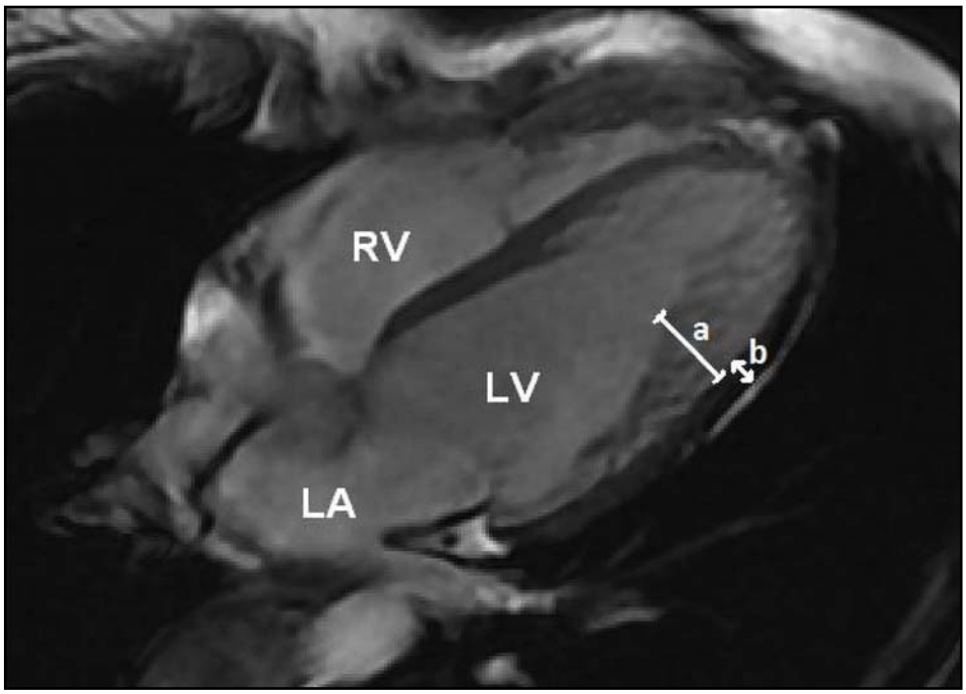

Fig. 1. Left ventricular trabeculations visualised using magnetic resonance imaging. LA – left atrium, LV – left ventricle, RV – right ventricle, a – non-compacted layer of the myocardium, b – compacted layer of the myocardium.

Tab. 1. Congenital heart defects in children with diagnosis of noncompaction left ventricle.

Total LVNC group	ASD	VSD	PDA	PS	CoA	TOF
n=24	n=3	n=2	n=1	n=1	n=1	n=1

LVNC – left ventricle non-compaction, ASD – atrial septal defect, VSD – ventricular septal defect, PDA – patent ductus arteriosus, PS – pulmonary artery stenosis, CoA – coarctation of the aorta, TOF – tetralogy of Fallot.

PATIENTS AND METHODS

The patient group included 24 patients diagnosed with LVNC between March 2002 and February 2016 (16 boys, 8 girls) at the age at presentation ranging between 9 days and 18 years (mean: 11.5 years) and the duration of follow-up 2 to 7 years (mean: 6.5 years).

Transthoracic echocardiography was performed using Aloka ProSound F75 (Aloka Co., Ltd., Japan). Magnetic resonance imaging (MRI) examination was done using Magnetom AVANTO SQ 1.5 T (Siemens AG, Germany) with specific cardiologic hardware and software. From the apical four-chamber view, the left ventricle was divided into three levels: the base at the level of mitral valve, papillary muscle level and apex. Two-dimensional echocardiograms were reviewed for severity of non-compaction, left ventricular systolic function and the presence of other congenital heart disease. Non-compacted to compacted myocardium ratio (NC/C) was evaluated using echocardiography and MRI in all 24 patients. The diagnosis of LVNC was based on the presence of characteristic multiple excessively prominent trabeculation associated with deep intertrabecular recesses, and on the presence of the maximum end-systolic (echocardiography) and/or end-diastolic (MRI) NC/C > 2 (Fig. 1). In addition, localization of ventricular noncompaction was evaluated. Twelve-lead electrocardiogram recordings (ECG) and Holter monitoring were evaluated in all patients for the screening for potential conductive abnormalities and arrhythmias.

Following data were collected at the time of examination or retrospectively from the reviewed medical records: clinical manifestations at the time of presentation including heart failure symptoms, treatment with cardiac medications, surgical interventions, comorbidities, facial dysmorphisms, familial inheritance patterns, developmental delay, failure to thrive, and associated medical illnesses. The familial screening based on echocardiographic examination for the purpose of confirmation or exclusion of cardiomypathies was performed

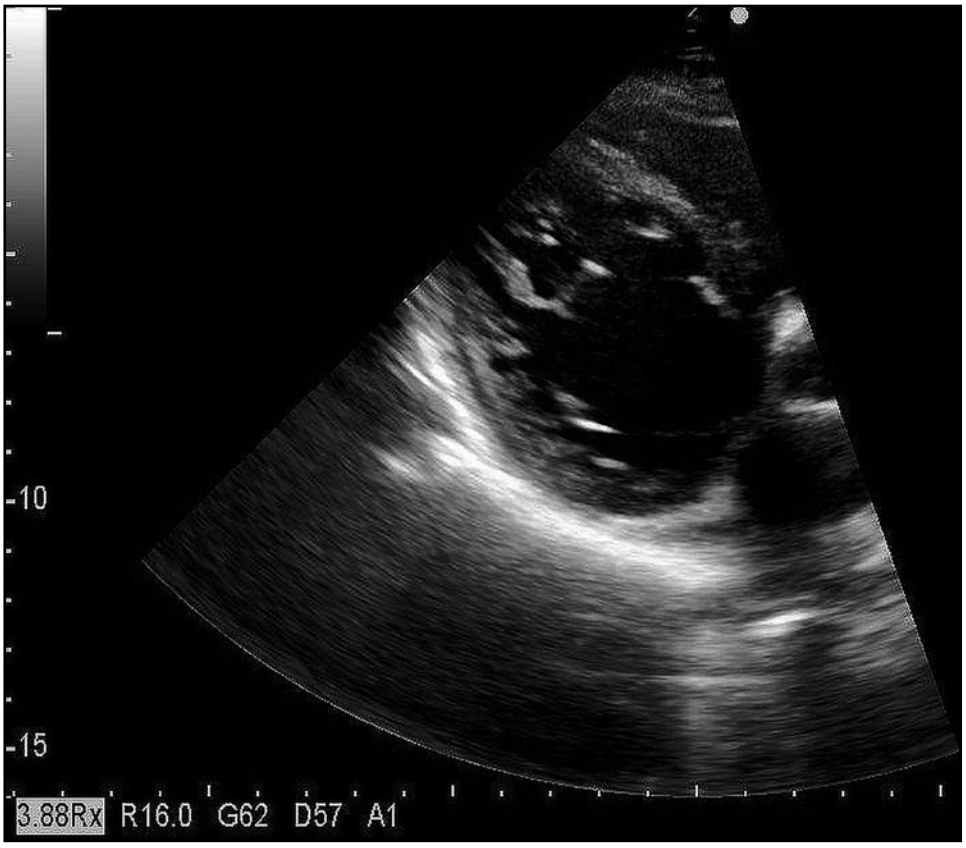

Fig. 2. Echocardiogram of the left ventricular long axis view with numerous trabeculations in the lateral wall.

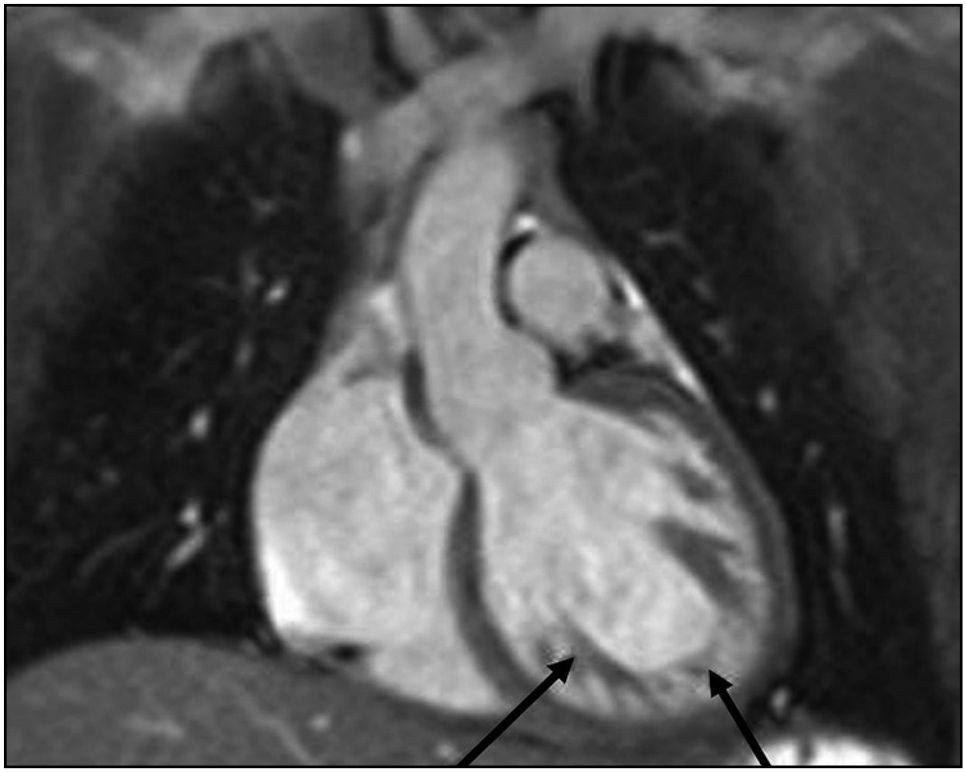

Fig. 3. Cardiac magnetic resonance imaging of the non-compacted (arrows) and compacted segment in left ventricular wall (long axis view). Coronary trufiFISP (bright blood sequence).

in all the first degree relatives of patients suffering from LVNC diagnosis.

The study was approved by the Ethical Committee institutional review board and all of the subjects and parents of minors gave informed consent to be involved in the study.

RESULTS

Among the total group of 24 patients diagnosed with LVNC, 17 patients have been initially followed-up for different diagnoses, which were based on ECG findings and echocardiographic evaluation without use of MRI, and thus, the diagnosis of LVNC was delayed. After retrospective evaluation by a senior specialist in pediatric cardiology, LVNC was found in 3 patients initially diagnosed with dilated cardiomyopathy, 11 patients followed-up with various forms of arrhythmias, and 3 patients with congenital heart disease. Afterwards, the diagnosis of LVNC was confirmed using MRI in all patients. The mean time period from the first evaluation until the LVNC diagnosis was made was 23 months. During this period, three standard echocardiograms were performed and 7 children (29 %) were completely asymptomatic.

Previous clinical examination revealed heart failure in 6 patients, hydrops fetalis in 1 patient, severe forms of arrhythmia in 2 patients, incidental finding of cardiac murmur in 7 patients, and disturbances of rhythm in 9 patients. Clinical comorbidities included failure to thrive (3 patients), developmental delay (3 patients), facial dysmorphism (1 patient), pericarditis and myocarditis (2 patients), and thyroiditis (1 patient). Neurologic abnormalities including seizure disorder and encephalopathy occurred in 2 patients. Only one case of familial occurrence of cardiomyopathy in the first–degree relatives was identified.

Treatment included furosemide (4 patients), digitalis (4 patients), angiotensin-converting enzyme inhibitors (3 patients), and beta-blockers (2 patients). Four patients required inotropic agents to augment cardiac output. Antithrombotic prophylaxis was performed in patients with reduced LV function. Three patients had associated congenital cardiac abnormalities treated by cardiac surgeries or interventions: in the first case a repair of ventricular septal defect, in the second case complete repair of Fallot's tetralogy, and in the third case treatment of the coarctation of the aorta. A summary of all the diagnosed congenital heart defects is presented in Table 1. During the period of follow-up, none of the patients experienced systemic embolic events and there was no single case of sudden death.

ECG/Holter monitoring findings

21 patients were noted to have ECG and/or rhythm abnormalities: left ventricular hypertrophy (6 patients), prolonged QTc (2 patients), intraventricular conduction delay (2 patients), right bundle branch block (6 patients), Wolff–Parkinson–White syndrome(2 patients), Lown-Ganong-Levine syndrome (3 patients), left anterior

Tab. 2. Echocardiographic and magnetic resonance imaging findings in patients with LVNC.

	Echocardiography	Magnetic resonance imaging
NC/C ratio (-)	2.94 (2.3 - 4.0)	2.86 (2.2 - 3.8)
Ejection fraction (%)	55 (39 - 68)	50 (25 - 60)
LVIDd (cm)	4.21 (3.5 – 4.9)	4.55 (3.6 – 5.1)
LVIDs (cm)	2.68 (2.23 – 3.2)	2.8 (2.2- 3.4)
LVM (g)	145 (115 – 171)	147 (119 – 183)
LVMI (g/m^{2,7})	30.1 (26.1 – 36.8)	30,56 (24.4 – 35.7)

LVNC - left ventricular non-compaction; NC/C ratio - non-compacted to compacted myocardium ratio; LVIDd, left ventricular internal dimension in diastole; LVIDs, left ventricular internal dimension in systole; *LVM*, left ventricular mass; *LVMI*, left ventricular mass index

fascicular block (2 patients), and arrhythmias including ectopic atrial rhythm (4 patients), supraventricular tachycardia (5 patients), and ventricular tachycardia (2 patients). The severe form of arrhythmia with congestive heart failure and ECG abnormalities typical for cardiomyopathy on the left precordial leads was found in three newborns with isolated form of LVNC.

Echocardiographic and cardiac MRI findings

The 2-D echocardiography and cardiac MRI revealed left ventricular (LV) trabeculation and intertrabecular recessescommunicating with the ventricular cavity in all patients (Fig. 2, 3). Non-compaction was more prominent in the myocardialsegment of the LV apex and posterior wall. LVNC involved mid lateral wall in 12 patients, the LV apex and mid lateral wall in 10 patients, and the LV apex alone in 2 patients. None of the patients had right ventricular involvement, intracardiac thrombi or embolic phenomena at the time of diagnosis or during the follow-up. The data from echocardiographic and MRI evaluation of NC/C ratio and ejection fraction are presented in Table 2.

DISCUSSION

This study presents a detailed description of complex diagnostic process including clinical presentation, comorbidities, ECG/Holter, echocardiographic and MRI findings, as well as findings from follow-up in 24 pediatric patients with LVNC, out of whom in 17 the diagnosis was initially overlooked. Re-evaluation by a senior specialist in pediatric cardiology and confirmation of the findings using MRI lead to accurate diagnosis in all the patients with previously borderline inconclusive results.

The first line modality for LVNC diagnosis is two-dimensional echocardiography with Doppler imaging. LVNC is typically characterised by two-layered myocardial structure with a thin compacted outer (epicardial) band, a much thicker non-compacted inner (endomyocardial) layer, and deep myocardial trabeculae, particularly in the apex and free wall of the left ventricle (Fig.2). It is important to note that for an accurate diagnosis, the physician must be familiar with the condition and must apply very specific diagnostic criteria. In our group, 17 out of 24 patients were initially overlooked and followed-up with different diagnoses. The delay in correct diagnosis in our patients may be explained mostly by the severity of cardiac symptomatology and by the lack of experience of the clinicians making the diagnosis. The majority of our misdiagnosed patients were followed-up with various forms of arrhythmias, fewer with dilated cardiomyopathy, and the undiagnosed LVNC was associated also with the presence of congenital heart disease. Other studies reported that LVNC was mostly misdiagnosed as dilated cardiomyopathy (Alehan 2004; Ergul et al. 2011).

In our study, echocardiography and cardiac MRI was performed in all patients. Echocardiographic examination shows a good correlation with cardiac MRI in the evaluation of the left ventricle ratio of non-compacted to compacted myocardium and also in the assessment of average ejection fraction (Rovai et al.; Thuny et al. 2010). However, cardiac MRI appears to be superior to standard echocardiography in the assessment of the extent of non-compaction morphology beyond that obtained with conventional echocardiography (Thuny et al. 2010). Moreover, echocardiographic visualization of non-compacted myocardium localised in the apex can be limited in some cases; therefore, MRI becomes the method of choice to confirm or rule-out possible LVNC (Olejník et al. 2017; Nunez-Gil & Feltes-Guzmán 2012). It seems that cardiac MRI is crucial in the diagnosis of the LVNC. In this study, the diagnosis of the LVNC was confirmed by ECHO and cardiac MRI in sixteen patients. Further, three patients had ambiguous result and five patients had a different diagnosis based on echocardiographic examination and cardiologist symposium, but the diagnosis of the LVNC was confirmed by cardiac MRI in all patients. Thus, the cardiac MRI examination plays an important role in the LVNC diagnosis, especially in unclear findings from other examinations (i.e. echocardiographic examination).

Moreover, seven patients (29%) were characterized by lower systolic function of the left ventricle based on echocardiography and the decreased systolic function of the left ventricle based on cardiac MRI was in nine patients (38%). With respect to left ventricle dilata-

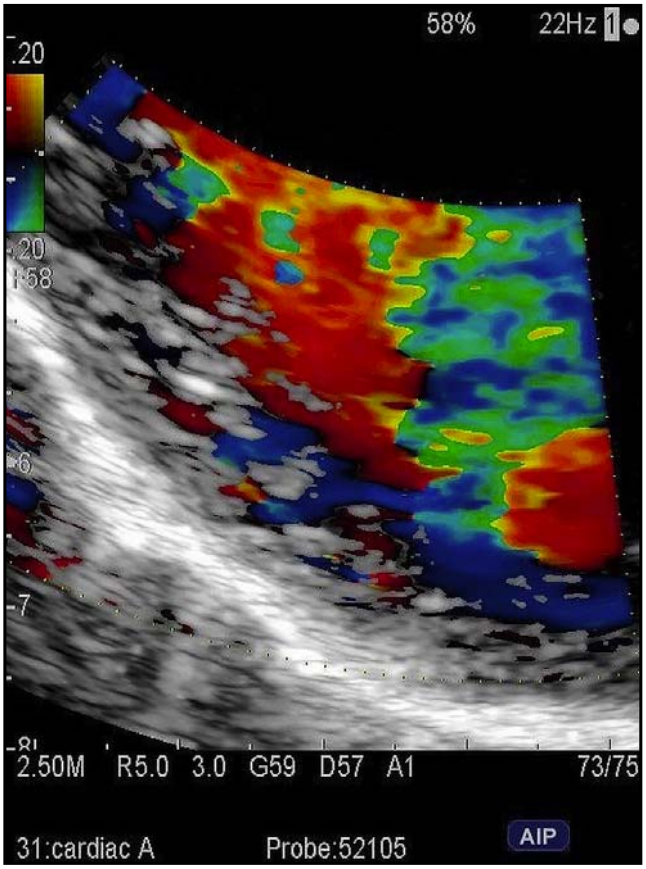

Fig. 4. Perfusion of intertrabecular recesses in the left ventricular cavity visualised using colour Doppler echocardiography.

tion, this finding was confirmed in five patients (20%) based on echocardiography as well as on cardiac MRI. Hypertrophic cardiomyopathy was not confirmed in any patient. In agreement with these findings, MRI was a conducive method to make a correct diagnosis of LVNC in our study in patients with inconclusive borderline results from echocardiography. In addition, MRI could be a more convenient method for detection of non-compacted segments in the right ventricle (Ergul et al. 2011). In the present study, there was no case of the right ventricular involvement.

LVNC can be isolated or associated with cardiomyopathies, congenital heart diseases and complex syndromes affecting the heart (Weiford et al. 2004; Rosa et al. 2011; Roberts et al. 2011; Cavusoglu et al. 2003; Oechslin & Jenni 2011). The knowledge that isolated ventricular noncompaction can co-exist with restrictive and hypertrophic cardiomyopathy (in addition to the dilated form) supports the concept that isolated ventricular noncompaction is a morphologic trait rather than a distinct cardiomyopathy (Biagini et al. 2006). In our study, majority of patients had isolated forms of LVNC, concomitant cardiac anomalies were less frequent. Segments involved were mid-ventricular (particularly the inferior and lateral ones) and apical with evidence of direct blood flow from the ventricular cavity into deep intertrabecular recesses visualised using colour Doppler (Fig 4).

The classical triad of complications - heart failure, ventricular arrhythmias and systemic embolic events - was not confirmed in our patients with LVNC. In our study, out of 24 patients, heart failure was present in three patients suffering from severe congenital heart disease and in two patients with severe forms of arrhythmia. Based on these findings, it seems that the classic triad of complications is more common in older patients with advanced disease and it is less frequently observed at younger age. Our results are in agreement with the previous findings indicating that the incidence of systemic thromboembolism, ventricular arrhythmia, and decreased systolic function is not as significant as originally reported by Chin *et al.* (Chin et al. 1990; Ichida et al. 1999; Pignatelli et al. 2003). In six patients with LVNC, heart failure was diagnosed in newborn period (three patients with severe forms of congenital heart defects before correction, one patients with LVNC presenting with hydrops fetalis, and two patients with severe forms of arrhythmia).

The disorders of the heart rhythm were investigated using standard resting ECG as well as 24-hours monitoring. Pathological ECG findings were confirmed in 87.5% of our patients, some of whom presented several ECG abnormalities, similarly as previously described (Chin et al. 1990). In three newborns with isolated form of LVNC, severe forms of arrhythmia with congestive heart failure were found with ECG abnormalities typical for cardiomyopathy on the left precordial leads. After treatment of arrhythmias and congestive heart failure, clinical state of the patients was improved, however, the pathological ECG changes remained unchanged. The ECG findings depend on several factors, in particular, whether LVNC occurs alone or associated with congenital heart diseases, and whether cardiac arrhythmia is present in newborn period or in older children. Whether these ECG findings have prognostic implications needs to be investigated during long-term follow-up period. Late gadolinium enhancement (LGE) showing myocardial fibrosis as a potential substrate for arrhythmias has been described in 8 (33%) our patients with LVNC.

Despite the previously documented familiar occurrence of LVNC and the assumed significant role of genetic aberrations (Weisz et al. 2010), there was only one case of cardiomyopathy identified in the first degree relatives in our study.

CONCLUSIONS

LVNC is a congenital heart disease that has been recognized with increasing frequency because of the advances in awareness and diagnostic imaging. Early presentation in infancy is associated with an increased risk of premature mortality. Despite the widespread use of echocardiography, LVNC is commonly overlooked,

often due to lack of knowledge about this disorder. Therefore, it is important to make the specialists more familiar with this condition and its pathology. MRI represents an appropriate method in case of non-conclusive echocardiographic finding, and could be a conducive procedure to make a correct diagnosis of LVNC under several specific conditions.

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DECLARATIONS OF INTEREST

The authors report no declarations of interest.

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