Typical cases, complications and difficulties in the diagnosis of non-compact myocardium in children by MRI and CT.

Poster No.: P-0020
Congress: ESCR 2015
Type: Scientific Poster
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Keywords: Cardiac, Echocardiography, CT-Angiography, MR, Contrast agent-intravenous, Diagnostic procedure, Congenital, Tissue characterisation, Image verification

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Purpose

Left ventricular non-compacted cardiomyopathy (LVNC) is a rare disease. It is morphologically characterized by increased left ventricular (LV) trabeculation and deep intertrabecular recesses communicating with the LV cavity. The disease usually has genetic or inherited pattern. But clinical manifestations and severity of clinical course often do not correlate with the severity of morphological changes. It is well known that prominent LV trabeculations may be found in up to 20-30% of patients without LVNC (in athletes, patients with LV overload due to valvular or congenital heart diseases, hypertrophic or dilative cardiomyopathy).

Diagnosis of LVNC is on clinical and imaging manifestations of the disease. Cardiac imaging modalities (echocardiography, cardiac MRI, in some cases - low-dose CT) play an important role in diagnosis of LVNC. Some thresholds based on the ratio of the noncompacted to the compacted myocardial tissue have been proposed for diagnosis of LVNC. For cardiac MRI the mostly used diagnostic criteria of non-compact myocardium is the ratio 2.3 or more of non-compact layer to compact in diastole (in presence of two-layered structure with compacted-noncompacted layers - Petersen et al., 2005) in one of the three long projections or percentage of LV trabeculated mass above 20% (Jacquier et al. 2010). But all these criteria are based on studies with rather small number of patients (from 7 to 62) and therefore they still seem to controversial.
Methods and Materials

The poster presents 18 cases of children and teenagers (age range 7 months - 16 year old) with diagnosis of LVNC and LV hypetabeculation of secondary origin. Patients were examined with cardiac echocardiography and cardiac MRI (1.5 tesla scanner), in 3 cases- with low-dose cardiac CT. Diagnosis of LVNC was based on Petersen's criteria and clinical manifestations of the disease.
Results

Poster presents a spectrum of clinical cases with true LVNC and cases when hypertrabeculations have secondary or incidental origin, being combined with other cardiac diseases. Typical cases of LV non-compaction are shown on Fig. 1-4. These patients were asymptomatic but they had typical signs of LVNC by echocardiography and MRI. One more patient with LVNC had died from fatal pulmonary thromboembolism (Fig. 5). A patient with a large atrial septal defect, RV dilation and trabecular layer on top of the LV, also had MR images, suggestive of LVNC, but most probably in this case hypetrabelculation of LV had secondary origin (Fig. 6). Fig. 7 shows advantages of MRI over echocardiography in visualisation of non-compacted myocardium. Fig. 8 demonstrate a rare case of congenital aneurysm of LV combined with LV non-compaction (an attempt of surgical correction of the aneurysm had been done). Case 9 demonstrates combination of LV hypetrabeuctions with fibrosis of IVS from the right side of unknown origin. Existence of right-sided NC has been discussed in the literature, but such cases are difficult to discriminate from ARVC. Such case is shown on fig.10. Secondary hypertrabeculation of LV in patient with chronic myocarditis and cardiomegaly is depicted on fig.11. In some patients with LVNC LGE MRI shows hyperenhancement of non-compact layer due to fibrotic changes (fig. 12). A patient with severe ARVC also had typical manifestations of LVNC (fig.13) and it stayed unclear - was it a case of one single cardiomyopathy or two different ones. Case 14 shows a case of chronic myocarditis with LV dilatation with marked fibrotic changes in the myocardium, where non-compaction was probably seen due to thinning of the LV myocardium with preservation of trabeculations. Last 4 cases show combination of LV non-compaction (of secondary origin) with primary and secondary cardiomyopathies. Fig.15 demonstrates LV secondary hypertrabeculation of LV in patient with dilative cardiomyopathy (DCM). Fig.16 and 17 demonstrate LV hypertrabeculation (not fulfilling criteria for LVNC) in patients with (probably) restrictive cardiomyopathies. Last case (fig.18) shows LV hypertrabeculation in in 8-year-old patient with mitochondrial encephalomyopathy. At the age of 1.5 years he suffered from an ischemic stroke. The cause of stroke could not be established. Dilatation of LV with boundary hypertrabeuctions in combination with decreased EF could be a risk factor for thrombi formation and cerebral embolism.
Fig. 1: Girl 2 years. A. MRI cine short axis. B. MRI cine 4ch. C. MRI LGE short axis. D. MRI LGE 4ch. Noncompact layer at the apical segment of LV. No thrombus and fibrosis.
Fig. 2: Boy 15 years. A. US parasternal long-axis view. B. US parasternal long-axis view. C. MRI cine short axis. D. MRI cine 4ch. E. MRI LGE short axis. F. MRI LGE 4ch. Noncompact layer at the apical segment of LV. No thrombus and fibrosis.

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Fig. 4: Boy 16 years. US parasternal short-axis view. B.US modified apical 5-chamber view. C. US apical 4-chamber view. D. MRI cine short axis. E. MRI cine 4ch. F. MRI LGE 4ch. Noncompact layer at the apical segment of LV. No thrombus and fibrosis.

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**Fig. 5:** Girl 8 years. A. US modified parasternal long-axis view. B. MRI cine short axis. C. MRI cine coronal axis. D. Autopsy photograph of LV. Noncompact layer at the apical and middle segment of the LV. Between trabeculae visualized thrombus (arrow).

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Fig. 6: Girl 7mounts. A. US parasternal long-axis view. B. US parasternal long-axis view. C. MRI cine short axis. D. MRI cine 4ch. E. MRI LGE short axis. F. MRI LGE 4ch. Noncompact layer at the apical segment of LV. No thrombus and fibrosis. Short arrows - large atrial septal defect with RV dilation. Long arrows - enhancement artifact from the abnormal movement of the interventricular septum.

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**Fig. 7:** Boy 15 years. A. US parasternal short-axis view. B. US parasternal long-axis view. C. MRI cine short axis. D. MRI cine 2ch. E. MRI IR-T1 short axis. F. MRI LGE 4ch. Noncompact layer at the apical and middle segment of the LV. An US differentiation noncompact trabecular layer is difficult. No thrombus and fibrosis.

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Fig. 8: Girl 14 years. A. US parasternal short-axis view. B. US parasternal long-axis view. C. US parasternal long-axis view. D. CT short axis. E. CT short axis. F. CT 3ch. Noncompact layer at the apical and middle segment of LV. Postoperative complications aneurysm on the lateral wall of LV.

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**Fig. 9:** Girl 10 years. A. MRI cine 4ch. B. MRI cine 2ch. C. MRI cine short axis. D. MRI IR-T1 short axis. E. MRI IR-T1 FS short axis. F. MRI LGE 4ch. G. MRI LGE short axis. H. MRI LGE short axis. I. MRI LGE short axis. Spherization of LV. Non-compact layer in apical segment of LV. Massive structural changes of septum (fibrosis) (arrowheads) with areas of fat transformation (long arrows).

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**Fig. 11:** Boy 16 years. A. US parasternal short-axis view. B. US parasternal long-axis view. C. US apical 4-chamber view. D. MRI cine short axis. E. MRI cine 4ch. F. MRI LGE 4ch. Noncompact layer at the apical and middle segment of LV. LV and RV dilatation. Inflammation in the epicardial layers of LV (arrows).

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**Fig. 12:** Boy 14 years. A. MRI cine 4ch. B. MRI LGE short axis. C. MRI LGE 2ch. Noncompact layer at the apical and middle segment of LV. LA dilatation. No thrombus. Trabecular fibrosis (arrows).
Fig. 13: Boy 10 years. A. MRI cine 4ch. B. MRI cine short axis. C. MRI LGE 4ch. D. MRI LGE short axis. ARVD. Arrows - fatty transformation. Noncompact layer at the apical, medium and basal segment of LV. No thrombus and fibrosis.
**Fig. 14:** Girl 11 years. A. MRI cine short axis. B. MRI cine 4ch. C. MRI LGE short axis. D. MRI LGE 4ch. Post myocarditis LV dilatation (LV EDV 240ml). Arrows - fibrotic changes. No noncompact layer, normal trabecular/chordal lateral LV wall, no trabecular layer on the apical segment. Due to thinning of the lateral LV wall (4mm) ratio of trabecular/chordal (10mm) layer to the compact can be increased artificially.

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Fig. 15: Girl 16 years. A, B, C. MRI cine short axis on three different levels in the apical segment of the LV. D. MRI cine 4ch. E. MRI LGE short axis. F. MRI LGE 4ch. DCM (LV EDV 160ml). Short arrows - local trabecular layer. Long arrow - fibrotic changes.

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**Fig. 16:** Girl 10 years. A. CT 4ch. B. CT short axis. C. MRI cine 4ch. D. MRI cine short axis.

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Fig. 17: Girl 14 years. A CT 4ch. B. CT short axis. D. MRI cine 4ch. D. MRI cine short axis.

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Fig. 18: Boy 8 years. A. MRI cine short axis, apical segment. B. MRI cine 2ch. C. MRI cine 4ch. Arrows - trabecular layer.

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Conclusion

Typical cases of LVNC are easy diagnosed by echocardiography or MRI. However, quite often different cardiac diseases (e.g. cardiomyopathies, myocarditis, CHD) accompanied with increased trabeculation of LV. In order to avoid hyperdiagnosis of LVNC, cardiac MRI is needed. In asymptomatic patients with some minor areas of non-compacted myocardium or increased trabeculations usually there is no need for treatment, but special care should be taken about cases with true LVNC.

On the large scale, this demonstration of clinical cases with LVNC and LVNC-like manifestations of different cardiac diseases stresses the much discussed in the literature need for further improvements and refinements of diagnostic criteria for LVNC are needed.
References


