

**NON-COMPACTION OF THE FAMILY LEFT VENTRICLE ABOUT TWO CASES
FROM THE SAME FAMILY AND REVIEWED LITERATURE**

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ABSTRACT

Isolated non-compaction of the left ventricle (NCVG) is a rare congenital cardiomyopathy resulting from the shutdown of normal embryogenesis of the myocardium. Its main feature is the existence of many deep heart-related ventricular trabeculations, generally located at the level of the apex of the left ventricle. Diagnosis is based on echocardiography and magnetic resonance imaging (MRI), and may be difficult in the atypical forms. The clinical presentation and the prognosis are very variable. Familial forms are not rare, ordering a family screening.^[1,2] We report the case of a young patient admitted in a context of global heart failure related to NCVG and whose screening of first-degree relatives has highlighted the same pathology in the mother who is clinically asymptomatic.

KEYWORDS: NCVG, echocardiography, magnetic resonance imaging, family screening.**I-INTRODUCTION**

Non-compaction of the left ventricle (NCVG) originally described in the child.^[1,2] and placed by the World Health Organization (WHO) in the group of "unclassified cardiomyopathies".^[3] It has been "rediscovered" in recent years, since more than 300 cases were reported, of which more than 85% after 1997. Interest of this pathology lies in its anatomical features variables that are at the origin of diagnostic criteria, based on mainly on echocardiography and MRI and finally, in its prognostic characteristics as well as in its character sometimes family.

Observation: Patient aged 25 years, without cardiovascular risk factors, admitted in a picture of predominantly right global heart failure, Rx thorax shows a cardiomegaly at 0.60. Echocardiography showed overall hypokinesia with FE at 28%, with spongy appearance of the left ventricle and aneurysm of the VG with a thrombus measuring 40 mm/ 30 mm (Figure n 1).

The patient was put on anticoagulant, and treatment of heart failure: IEC and diuretic.

II- CASE REPORT

We report the case of a son and his mother carrying the non-compaction of VG and don't the diagnosis is made by ETT and MRI.

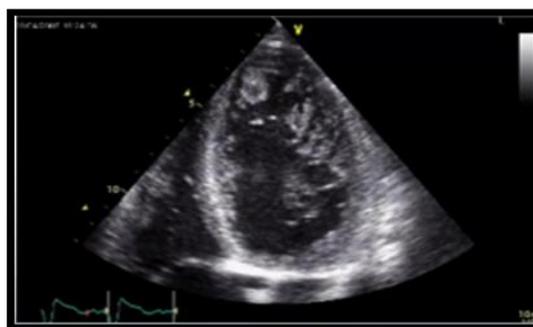


Figure 1: ETT, view 4 cavity which shows an aspect of non-compaction of VG with apical thrombus.

The family investigation revealed in the 65-year-old mother, asymptomatic clinically; same aspect of non-compaction of the predominant VG at the apex and on

the latero-median and infero-median segments with LVEF 45% (Figure n 2), an MRI was given requested and who confirmed the diagnosis for the two patients.

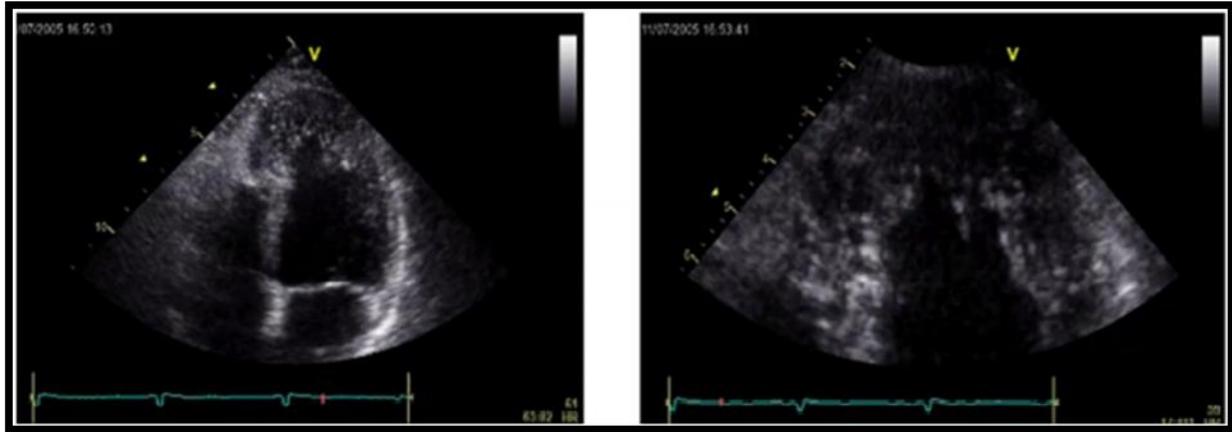


Figure 2: ETT of the patient's mother seen 4 cavity showing an aspect of non-compaction of the VG plus marked at the apex and lateral antero wall.

III. DISCUSSION

The NCVG would be the consequence of a halt to the phenomenon embryological progressive compaction of the ventricular myocardium that normally occurs between the fifth and the eighth week causing persistence, to varying degrees, prominent tabulations and deep intercedicular spaces. The main anatomical feature of the NCVG is therefore the existence of myocardial ventricular tabs numerous and deep, usually localized at the level of the apex of the left ventricle.

NCVG is a rare cardiomyopathy whose incidence is by 0,05 %.^[3,4,5] Its prevalence seems to be much higher in children than in adults. She would represent at the child the third cause of cardiomyopathy after dilated and hypertrophic heart disease.^[3] Its exact prevalence in adults is not known but seems weak. The NCVG preferentially affects young or middle-aged subjects with male predominance. The prevalence of left-hand involvement is approximately 0.014%.^[6,7] That of the right reach is unknown.

NCVG is classified as a genetic primary cardiomyopathy, with a characteristic morphological ("spongy") aspect of the VG myocardium, resulting from a cessation of normal embryogenesis. The location of the uncompacted segments is mainly in the apical and median segments of the lower and lateral walls of the left ventricle.^[8]

This pathology can progress to LV dilation and systolic dysfunction, heart failure, thromboembolic events and arrhythmia. The diagnosis of NCVG is based on three imaging examinations: echocardiography (or echocardiography with contrast medium), cardiac magnetic resonance imaging and cardiac CT, the definitive diagnosis remains based on autopsy.

Familial and non-familial cases have been described, but NCVG is frequently familial, with at least 25% of asymptomatic parents having a number of echocardiographic abnormalities.^[9] This is why a family investigation is necessary.

The prognosis for patients with NCVG is highly variable, ranging from prolonged asymptomatic course rapidly progressive heart failure (HF), which may result in the need for a heart transplant or death.

Currently, there are no recommendations for management of patients with NCVG. Treatment current NCVG is therefore that of any cardiomyopathy, based on conventional treatments for insufficiency cardiac.^[10]

There have been significant controversies about anticoagulation in patients with NCVG. The implantation of a cardiac defibrillator in these patients due to the high risk of sudden death is also extremely controversial.

A periodic inspection by Holter ECG of 24H is indicated in order to assess the risk of a possible asymptomatic arrhythmia.^[11,12]

IV. CONCLUSION

The diagnosis of left ventricular non-compaction should be evoked in the face of unexplained heart failure in adults. NCVG may be underestimated, it may be the cause of thromboembolic events and arrhythmia. The discovery of a case must propose the realization of an ETT in the family environment given the existence of family forms.

V. REFERENCES

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