Isolated noncompaction cardiomyopathy (NCCM) - 2010

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The entity was first described in 1984. (25 years ago.)This is an unclassified primary genetic cardiomyopathy, is now attracting increased attention.

NCCM is an extremely rare cardiomyopathy, not fully clarified. Since first description in 1984, few clinical studies were done.

The pathogenesis of NCCM is thought to involve a genetically determined disturbance of the myocardial compaction process during fetal endomyocardial morphogenesis. Is assumed to occur as an arrest of the compaction process during the normal development of the heart.

Between weeks 5 to 8 of human fetal development, the ventricular myocardium undergoes gradual compaction with transformation of the relatively large intertrabecular spaces into capillaries while the residual spaces within the trabecular meshwork gradually flatten or disappear. In the case of NCCM, the spaces within the intertrabecular meshwork persist while no other cardiac abnormalities exist. Although there is substantial evidence supporting the developmental hypothesis, other pathogenetic processes responsible for NCCM have been discussed. It can be assumed that NCCM will be better understood in the future as the molecular genetic basis of cardiomyopathies will be further unravelled.

Echocardiography is the diagnostic method of choice.

The diagnosis is based on the following echocardiographic criteria:

1. The presence of at least 4 prominent trabeculations and deep intertrabecular recessesas visualized by color Doppler imaging;

2. Blood flow from the ventricular cavity into the intertrabecular recesses

3. Typical bilaminar structure of the affected portion of the left ventricular myocardium. It is a typically show a two-layered structure with an endsystolic ratio greater than two between the noncompacted subendocardial layer and the compacted subepicardial layer;

4. The segments of noncompacted myocardium mainly involve the apex and the inferior mid and lateral mid of the left ventricular wall

5. Absence of coexisting cardiac abnormalities. It is not accompanied by any other cardiac anomalies.

NCCM can also be diagnosed with magnetic resonance imaging of the heart. Magnetic resonance imaging using modern gradient echo sequences has also been shown to diagnose NCCM accurately.

The clinical severity of NCCM is variable; its manifestations include:

1. Heart failure is the most common presenting condition.
2. Thromboembolic events
3. Arrhythmias including ventricular tachycardia and atrial fibrillation.

The treatment is symptom-based. Patients with symptomatic NCCM have a poor prognosis.

Its molecular genetic basis is not yet fully clear, and the same is true of its diagnosis, treatment, and prognosis. Further study of these matters is needed.

The establishment of a registry, which was initiated by the "Arbeitsgemeinschaft Leitende Kardiologische Krankenhausärzte (ALKK)" recently, may provide further clues for diagnosis, risk stratification, and management of this disease.