Non-compaction Cardiomyopathy: Prevalence, Prognosis, Pathoetiology, Genetics, and Risk of Cardioembolism

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Abstract
Left ventricular non-compaction (LVNC) is thought to arise from arrest of the normal process of trabecular remodeling or “compaction” that takes place during embryonic life and is characterized by the presence of a two-layered ventricular wall, with a compact epicardial layer and a non-compacted endocardial layer. It is an uncommon condition that can occur isolated or in association with other disorders, including congenital heart anomalies and mitochondrial or musculoskeletal disorders. Both familial and sporadic forms are recognized, and several responsible genes have been identified, although only a minority of patients can be successfully genotyped. The diagnosis is usually made by echocardiography, but cardiac magnetic resonance imaging has been used increasingly. Management is mainly empirical and directed at the major clinical manifestations: heart failure, arrhythmias, and systemic embolic events. This article will review the major features of LVNC and present new trends in the diagnosis and management of this intriguing condition.

Keywords
Left ventricular non-compaction, Pathogenesis, Epidemiology, Genetics, Clinical presentation, Diagnosis, Management, Natural history, Prognosis