Case of Ventricular Noncompaction: The Crypts and the Blood

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A 28-year-old man presented with progressive dyspnea, orthopnea, and edema of the lower extremity. On examination, he was noted to have jugular venous distention, bibasilar crackles, and an S3 gallop. Signs and symptoms of heart failure prompted echocardiography, which revealed severe left ventricular systolic dysfunction with an ejection fraction of 30%. Marked left ventricular trabeculae were also identified. The prominent trabeculae and deep intertrabecular recesses (crypts) that communicate with the ventricular cavity are the hallmark of isolated left ventricular noncompaction syndrome.

Noncompaction syndrome is a congenital form of cardiomyopathy in which there is an arrest of compaction of the loose meshwork that makes up the fetal myocardial primordium. The syndrome is rare; in one series of 37,000 echocardiographic examinations performed over 10 years, only 17 cases (0.05%) of noncompaction were identified. Patients with isolated left ventricular noncompaction are at risk for developing heart failure, ventricular arrhythmia, and embolic complications; thus, early diagnosis is important. Before warnings from the Food and Drug Administration limiting their use, echocardiographic contrast agents were often used to enhance these trabeculae and recesses by intensifying blood density. A supplemental video (linked to the full-text version of this article at www.mayoclinicproceedings.com) shows how echocardiographic agents can be used to enhance blood flow within the ventricular trabeculae (hence, the subtitle, The Crypts and the Blood).

Ultimately, this patient was treated for heart failure and discharged with prescriptions for an angiotensin-converting enzyme inhibitor, β-blocker, and diuretic. He has since done well.


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