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REFERENCES

REPLY

We do appreciate the valuable comments from Drs. Almeda and Calvin regarding our article (1) and are happy to provide, as a response to their questions, some further data from the FRISC II study. Drs. Almeda and Calvin wonder how often electrocardiogram (ECG) changes suggestive of posterior wall injury (i.e., ST-segment depression in the anterior precordial leads) were present on admission in patients with occlusion of the left circumflex artery as the culprit lesion. Unfortunately, the database did not contain information about ST-segment depression in each of the 12 individual leads, but only whether ST-segment depression was present in anterior, lateral or inferior leads. However, in patients with occlusion of the left circumflex artery as the culprit lesion, ST-segment depression was located exclusively in the anterior leads in only 5% (4/75). The ST-segment depression in the anterior leads together with ST-segment depression in the lateral or inferior leads was present in 13% (10/75) of patients. Thus, as pointed out by Drs. Almeda and Calvin, to be able to rapidly identify patients with occlusion of the left circumflex artery we need better diagnostic techniques than the standard 12-lead ECG.

Drs. Almeda and Calvin also ask about the outcome in patients with “normal” (<50% stenosis) coronary arteries and an elevated troponin T level (tnT). In the invasive cohort, 57 patients had “normal” coronary arteries and an elevated tnT; the median tnT level among those was 0.23 µg/l (10th to 90th percentile, 0.02 to 0.73). Among those 57 patients, 2 patients (3.5%) suffered a myocardial infarction (MI) and none died during the one-year follow-up. Thus, this was much lower than the event rates (approximately 17% MI and 4% death) seen in the patients with corresponding tnT levels (the two middle quartiles, tnT 0.01 to 0.63 µg/l) in the noninvasive cohort. In fact, the event rates among those with “normal” coronary arteries and an elevated tnT were not significantly different from those seen in the 101 patients with “normal” coronary arteries and no tnT elevation (one MI and no deaths). These data further support the important point that Drs. Almeda and Calvin make, namely that “troponin elevations are, in and of itself, not equivalent, and that it is not merely the troponin elevation, but rather the etiology behind the elevation, that makes the most difference clinically.”

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REFERENCE

Late-Onset Primary LVH: HCM Versus Cardiac Fabry Variant

In their article that appeared in the August 2001 issue of JACC, Maron et al. (1) suggest that left ventricular hypertrophy (LVH) can be a late manifestation of hypertrophic cardiomyopathy (HCM) due to cardiac myosin-binding protein C (MyBP-C) mutations. However, the echocardiographically documented LVH developing in their midlife in patients with MyBP-C mutations cannot be attributed for certain to that genetic abnormality unless a histologic demonstration of HCM can be provided by LV endomyocardial biopsy.

In fact, penetrance of MyBP-C mutations in large series ranges between 37% and 71% even in patients over 50 years of age (2–4), and other genetic disorders that give rise to LVH in middle-aged patients also exist, such as the cardiac variant of Fabry disease (5,6). This last entity, although X-linked, can occur in both male and female patients (7), can account for up to 9% of patients with nonobstructive HCM (8,9) and can be diagnosed simply by assessment of the alpha-galactosidase A enzymatic activity in the peripheral blood. If Fabry disease was present even in a proportion of patients with presumed HCM, those patients could now benefit from galactose infusion (10) and/or enzyme replacement therapy (11).