patients sent for echocardiography during a specified period of time.¹ Reasons for presentation were innocent heart murmurs, a family history of congenital heart disease, rule-out of cardiac side effects of non-cardiac medications, among others.

4. Patient populations. While comparing study populations, we would be interested to know the age of the patients and relatives studied by Fazio et al. Presumably most of them were adults. By which criteria did patients enter the registry; was it hospital admissions only? How many NCVM patients were entered with and without congenital heart disease? NCVM subpopulations may carry a different cardio-vascular risk.¹

5. Incidence. We do not know the total number of patients entered into the highly specialized Italian registry. But certainly, the number of NCVM patients entered in 1 year (>230) is remarkable; as is the number of first-degree relatives detected with NCVM (48/31). It has been repeatedly stated that NCVM appears to have been previously under-diagnosed all together.¹,²,³

Pretty much all existing data on NCVM has been prone to a selection bias. We do not know the prevalence and the natural history in a truly non-selected population. Based on existing data, including our own study and the one by Fazio et al., the occurrence of CHF in NCVM is concerning. It will be highly interesting to learn about details of the investigation by Fazio et al. addressing first-degree relatives and future similar data. The outlook may be more encouraging in incidental or familial discovery of NCVM.³ Unfortunately, even such populations are prone to a selection bias.

References


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We thank Efthimiadis et al. for their comments on our article concerning the improvement of systolic myocardial function in the lateral (free) wall in hypertrophic obstructive cardiomyopathy (HOCM) patients, which was studied using CMR tissue tagging and 3D strain analysis. In a previous work, we have demonstrated that in symptomatic patients with HOCM, left ventricular remodelling after ASA occurs early and progresses on mid-term follow-up, and total left ventricular mass reduction exceeded septal mass reduction. The remote mass reduction was correlated with the LVOT pressure gradient reduction, and thus we concluded that myocardial hypertrophy in HOCM is, at least in part, afterload-dependent and reversible and is not exclusively caused by the genetic disorder.

In this article, we have studied in a subgroup of patients the regional changes in septal, adjacent, and remote systolic myocardial function by calculating the shortening index, a combined strain parameter reflecting myocardial contraction. We have demonstrated for the first time that reduction in symptomatic HOCM patients achieved by ASA not only was associated with a significant reduction in myocardial mass, but also with an improvement of intramural systolic myocardial function in the lateral (remote) wall, supporting the concept of reversed LV remodelling.

Previously, our group had demonstrated that contrast-enhanced CMR allowed detailed evaluation of size and location of septal myocardial infarction induced by ASA, and that the infarction size was correlated with clinical indexes of infarct size. In this study, we have demonstrated that contrast-enhanced imaging data derived in ~60% of the study-group pre-ASA after administration of gadolinium-DTPA contained only small pre-existing foci of delayed myocardial hyperenhancement, representing myocardial fibrosis and other pathological changes in the myocardial wall (e.g. disarray, inflammation, oedema, myolysis, and necrosis). Compared with these hyperenhanced area-size-assessed pre-ablation, the infarct-size induced by ASA was 10-fold larger. In this respect, the induced myocardial infarct after septal ablation therapy enlarges the already existing arrhythmic substrate in HOCM patients. However, an electrophysiology report in high-risk patients after ASA has not indicated an increased arrhythmic substrate necessitating higher rates of implanting defibrillators. Although ventricular tachycardia and sudden death have been reported after ASA, these clinical features characterize the natural course hypertrophic cardiomyopathy irrespective of therapeutic LVOT gradient reduction. Further studies are necessary to evaluate the long-term effects of ASA with respect to ventricular arrhythmias and sudden cardiac death. Our goal in the near future must be developing additional tools to identify the high-risk HOCM patients, regardless of a potential intervention for LVOT obstruction, in whom defibrillator implantation is justified.

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