

Two essential cardiac disorders improvement in patients with hypertrophic cardiomyopathy

Sudden cardiac death and heart failure are the most serious complications of hypertrophic cardiomyopathy (HCM). To prevent these serious complications, 2 essential cardiac disorders in patients with HCM must be corrected. These are left ventricular (LV) hypertrophy and LV diastolic dysfunction. In addition, the prognosis for HCM patients with an LV pressure gradient (hypertrophic obstructive cardiomyopathy, HOCM) is known to be worse in those without an LV pressure gradient (hypertrophic nonobstructive cardiomyopathy, HNCM). Maron and Spirito mentioned in their review that there is no intervention capable of inducing LV hypertrophy regression in patients with HCM without causing a clinically unfavorable end-stage disease. In addition, Harris et al. reported that the interval between identifying end-stage disease and death or transplantation is short.

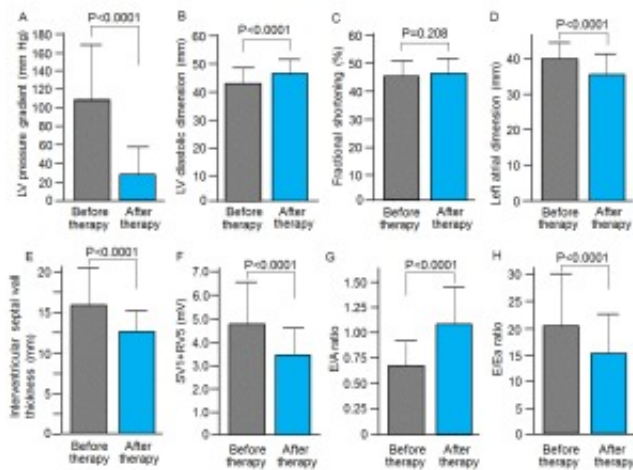


Fig. 1.

Recently, we reported that long-term treatment with cibenzoline improved the LV pressure gradient, LV diastolic dysfunction, and LV hypertrophy in patients with HOCM without causing serious complications. Figure 1 indicates the study results. (A, LV pressure gradient change; B, LV size; C, LV systolic function; D, left atrial size; E and F, extent of LV hypertrophy; G and H, extent of LV diastolic dysfunction). As shown in figure 1, the LV pressure gradient decreased, LV diastolic dimensions increased and were close to normal, LV systolic function remained normal, left atrial size decreased and was close to normal, LV hypertrophy regressed (improved) in both echocardiographic (E) and electrocardiographic (F) assessments, and LV diastolic dysfunction also improved. Figure 2 shows the change in electrocardiogram results, following cibenzoline treatment, in a patient with HOCM. As shown in Figure 2, the voltage in each lead decreased markedly, reflecting LV hypertrophy regression. The fact that the fractional shortening observed in the patients in this study was normal to hyper-dynamic, and remained unchanged during the study, is

very important. This finding indicates that LV hypertrophy regression is not the result of the advancement of LV remodeling (deterioration of LV function), but is the result of LV reverse remodeling (recovery to a better condition) associated with cibenzoline therapy. To our knowledge, this is the first report to elucidate LV hypertrophy regression in patients with HOCM undergoing medical treatment. In addition, the patients' QTc intervals were significantly shortened as a result of the treatment. The QT interval is a surrogate marker for predicting serious adverse drug effects, syncope, or death due to torsade de pointes.

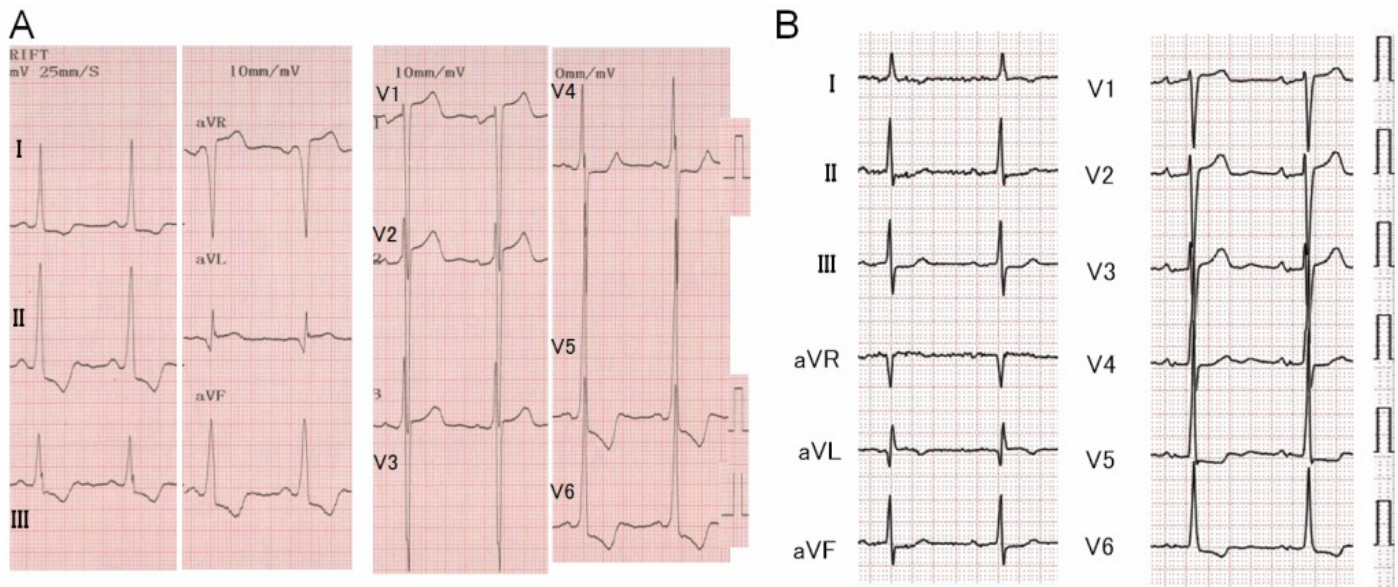


Fig. 2.

To avoid the potentially serious complications associated with cibenzoline treatment, patient's renal function, age, etc. must be considered. We believe that cibenzoline treatment avoids the development of heart failure in patients with LV hypertrophy and LV diastolic dysfunction associated with HCM.

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