Follow-up of Cardiac Fabry Disease Treated by Cardiac Resynchronization Therapy

Yuko Fukuda, MD, PhD, Tetsuari Onishi, MD, PhD, Atsushi Suzuki, MD, PhD, Hidekazu Tanaka, MD, PhD, Koji Fukuzawa, MD, PhD, Akihiro Yoshida, MD, PhD, Hiroya Kawai, MD, PhD, and Ken-ichi Hirata, MD, PhD, *Kobe, Japan*

INTRODUCTION

The cardiac variant of Fabry disease is associated with significant morbidity and early death. We report the case of a 60-year-old man who was admitted with severe dyspnea at rest. He developed heart failure with severe left ventricular (LV) systolic dysfunction, sustained atrial flutter, and frequent nonsustained ventricular tachycardia. He was diagnosed with the cardiac variant of Fabry disease and was treated with flutter ablation, optimal medical therapy, and cardiac resynchronization therapy (CRT) with a defibrillator. Three years after CRT, he is totally asymptomatic and is able to perform activities of daily living and work with no limitations. This report highlights the role of advanced echocardiography to evaluate LV function in patients with the cardiac variant of Fabry disease.

We previously reported a case of cardiac variant of Fabry disease treated with CRT.[1] Three years of follow-up on this rare case will be detailed, as excellent images were successfully recorded along with the clinical course. This report highlights the role of advanced echocardiography to evaluate LV function in patients with cardiac variant of Fabry disease.

CASE PRESENTATION

A 60-year-old man who had received a diagnosis of hypertrophic cardiomyopathy 13 years previously (Figure 1) was admitted to our hospital with severe dyspnea at rest (New York Heart Association class IV). He had developed heart failure with severe LV systolic dysfunction with an LV ejection fraction of 20% (Videos 1 and 2), sustained atrial flutter (Figure 2), and frequent nonsustained ventricular tachycardia. Dyssynchrony in the left ventricle was observed as a time delay of 164 msec between

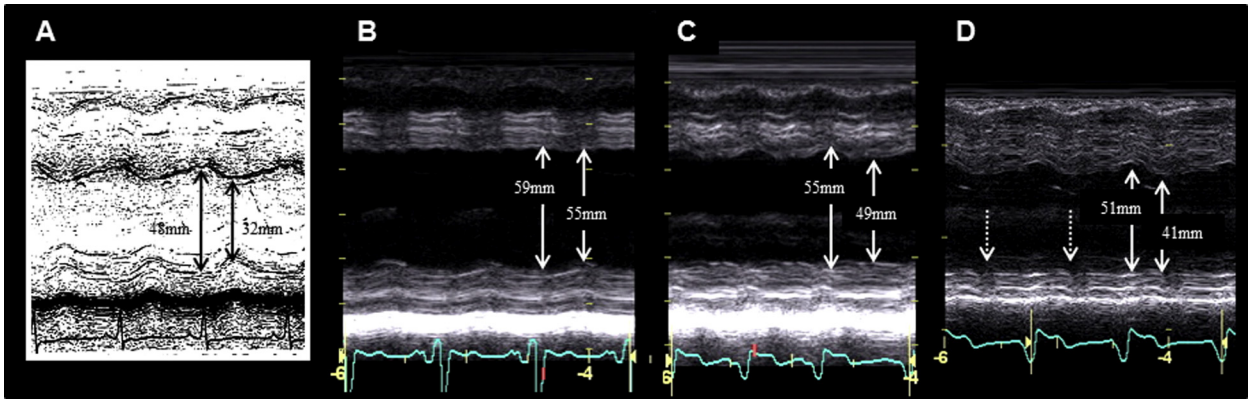

Figure 1 M-mode echocardiograms. *Arrows* indicate LV end-diastolic and end-systolic dimensions. Normal contraction with LV hypertrophy 13 years before admission **(A)**, dilated left ventricle with abnormal wall motion before CRT **(B)**, decrease in LV dimensions and slight improvement in LV contraction 1 week after CRT **(C)**, and normalized LV dimensions and further improvement in LV contraction 3 years after CRT **(D)** were seen (fractional shortening 33%, 7%, 11%, and 20%, respectively). Of note, posterior wall motion deteriorated (*broken arrows*).

From the Division of Cardiovascular Medicine, Department of Internal Medicine, Kobe University Graduate School of Medicine, Kobe, Japan.

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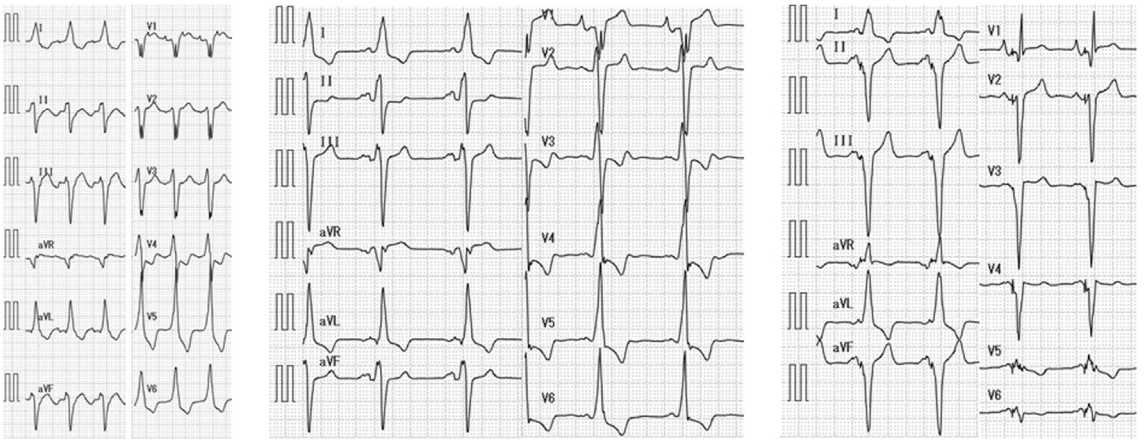

Figure 2 Twelve-lead electrocardiograms. (*Left*) Typical atrial flutter and 2:1 atrioventricular conduction with left bundle branch block (QRS duration 160 msec) was observed on admission. (*Middle*) Sinus rhythm was maintained after ablation for atrial flutter (QRS duration 160 msec). (*Right*) Electrocardiogram of biventricular pacing shows the narrowing of QRS duration (100 msec).

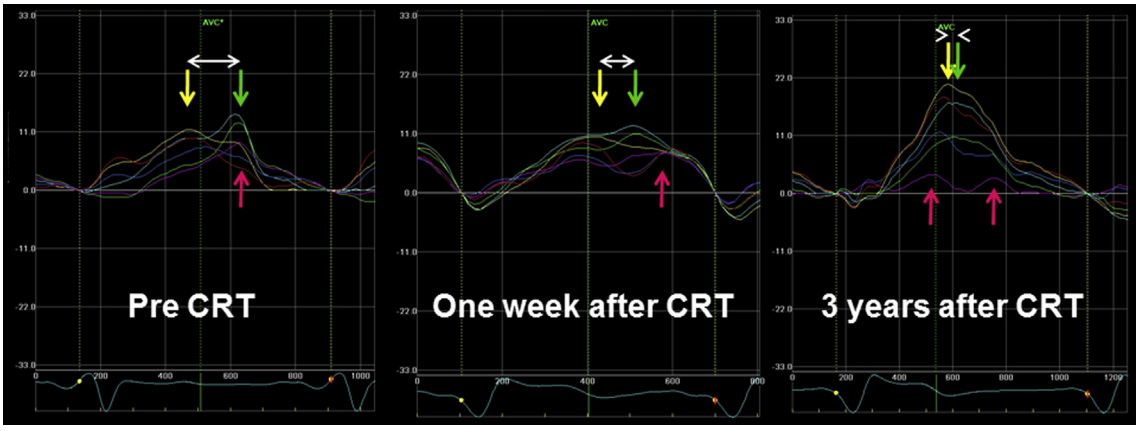

Figure 3 Speckle-tracking radial strain images in the midventricular short-axis view. The curves are color-coded by the defined myocardial regions as depicted in the figure (*yellow*, anterior septum; *light blue*, anterior segment; *green*, lateral; *purple*, posterior; *dark blue*, inferior; *red*, septum). Dyssynchrony is shown as the difference (*white arrow*) in the timing of peak systolic strain between the anteroseptal (*yellow*) and posterior wall (*green*).

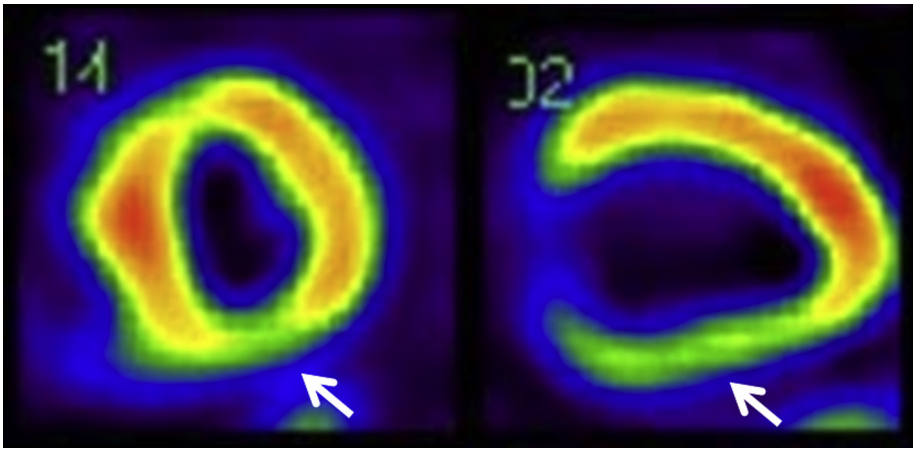

Figure 4 Single-photon emission computed tomography. LV short-axis (*left*) and long-axis (*right*) images showed reduced uptake of ^{99m}Tc-tetrofosmin at the posterior wall (*arrows*).

the anterior-septal and lateral wall peak strain before CRT[2] (Figure 3, *left*). He was treated with atrial flutter ablation, optimal medical therapy, and CRT with a defibrillator. One week after CRT, the time difference had decreased to 89 msec, suggesting reduction of LV dyssynchrony (Figure 3, *middle*). We diagnosed the cardiac variant of Fabry disease by enzyme assay and