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## Case Report

## Left ventricular outflow tract obstruction with abnormal papillary muscles

Kotaro Oe (MD)<sup>a,\*</sup>, Tsutomu Araki (MD, FJCC)<sup>a</sup>, Miho Ohira (MD)<sup>a</sup>,  
Tetsuo Konno (MD, FJCC)<sup>b</sup>, Masakazu Yamagishi (MD, FJCC)<sup>b</sup><sup>a</sup> Division of Internal Medicine, Saiseikai Kanazawa Hospital, Kanazawa, Japan<sup>b</sup> Division of Cardiovascular Medicine, Kanazawa University Graduate School of Medicine, Kanazawa, Japan

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## ABSTRACT

A 65-year-old man with a history of hypertension was admitted to our hospital with fainting and syncope. He had experienced recurrent syncope since 20 years of age. On admission, systolic heart murmur was audible at the apex of the heart. Echocardiography revealed anteriorly displaced papillary muscles (PMs), elongation of the anterior mitral valve leaflet (AML), and systolic anterior motion (SAM) of the AML. Color Doppler imaging showed accelerated flow with a pressure gradient (PG) of 56 mmHg at the left ventricular outflow tract (LVOT). Cardiac magnetic resonance imaging revealed mild asymmetric septal hypertrophy and multiple accessory PMs. Cine images clearly demonstrated SAM and LVOT obstruction due to anteriorly displaced PMs. Based on these findings, the patient was diagnosed as having hypertrophic cardiomyopathy and LVOT obstruction due to abnormal PMs. Oral administration of bisoprolol (2.5 mg/day) was initiated, because the patient rejected surgical treatment. Follow-up echocardiography revealed a gradual decrease in the LVOT-PG to 24 mmHg, and no episodes of fainting or syncope have recurred for 2 years after the initiation of bisoprolol.

<Learning objective: Abnormal papillary muscle (PM) is an unusual cause of left ventricular outflow tract (LVOT) obstruction, and cardiac magnetic resonance (CMR) imaging has been reported to be useful for diagnosis of abnormal PM. Abnormal PMs with LVOT obstruction are usually treated by surgical correction, and therefore, reports on medical treatment are limited. We report a case of LVOT obstruction due to abnormal PMs, which was accurately diagnosed by CMR imaging and successfully treated with a beta-blocker.>

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## Introduction

Abnormal papillary muscle (PM) is an unusual cause of left ventricular outflow tract (LVOT) obstruction [1], and is often found in patients with hypertrophic cardiomyopathy (HCM) [2]. Cardiac magnetic resonance (CMR) imaging has been reported to be useful for diagnosis of abnormalities of the PM and mitral valve in HCM patients [3–5]. Abnormal PMs with LVOT obstruction are usually treated by surgical correction [6,7], and therefore, reports on medical treatment are limited [8]. In the present report, we describe the case of an HCM patient associated with LVOT

obstruction due to abnormal PMs, which was accurately diagnosed by CMR imaging and successfully treated with a beta-blocker.

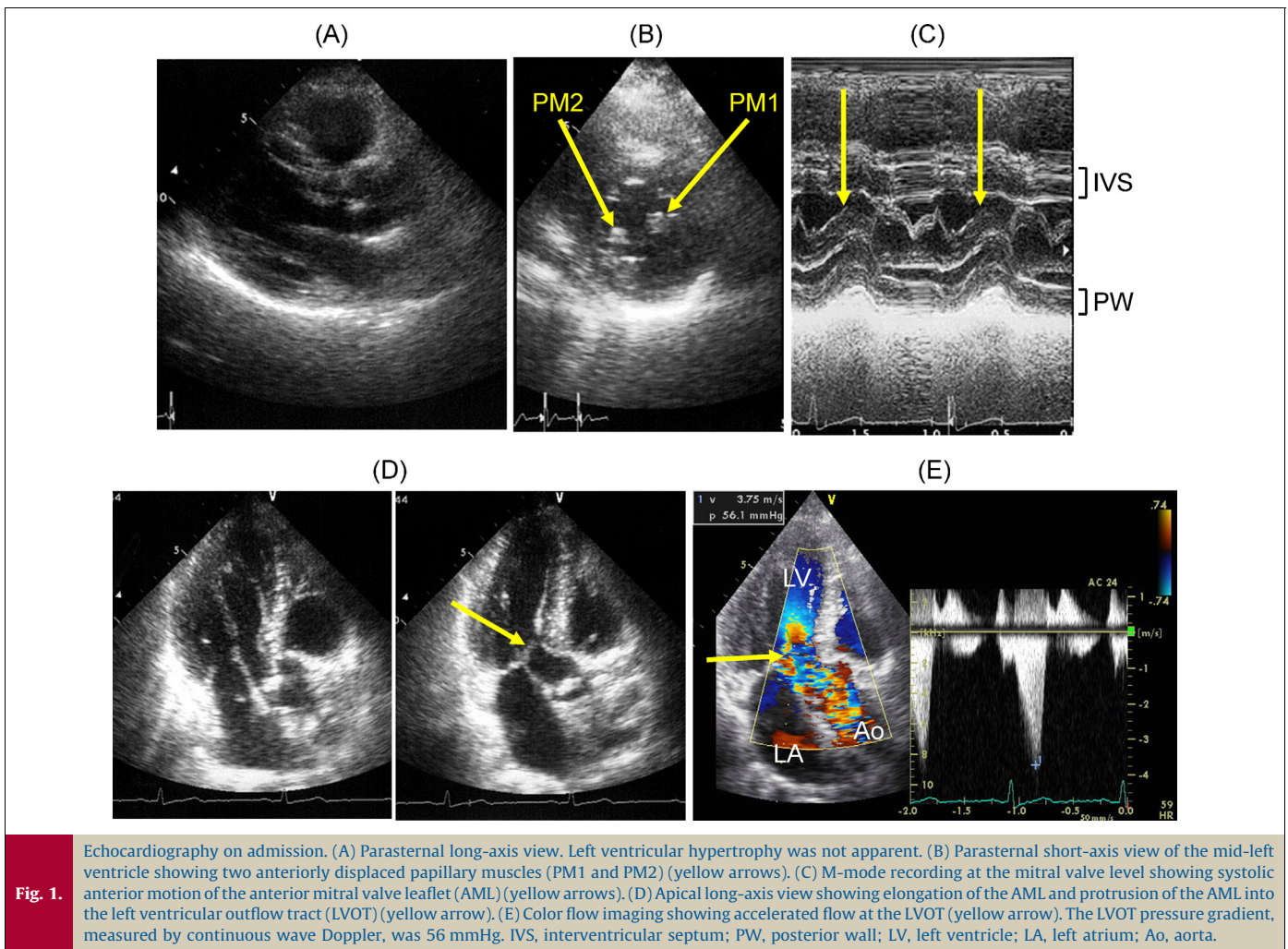
## Case report

A 65-year-old man with a history of hypertension was admitted to our hospital with fainting and syncope. The patient had experienced recurrent syncope since 20 years of age, and was diagnosed with epilepsy at 49 years of age. Treatment with anticonvulsants was soon discontinued because of skin rash. He had been taking amlodipine (5 mg/day) for hypertension. On admission, his blood pressure was 158/93 mmHg, pulse rate was 61 beats per minute, and systolic heart murmur of grade 4/6 was audible at the apex of the heart. Chest radiography showed mild cardiomegaly, and electrocardiography showed high voltage and mild ST-segment depression. Transthoracic echocardiography

\* Corresponding author at: Division of Internal Medicine, Saiseikai Kanazawa Hospital, Akatsuchimachi Ni 13-6, Kanazawa 920-0353, Japan.

Tel.: +81 76 266 1060; fax: +81 76 266 1070.

E-mail address: [kotaroee316@yahoo.co.jp](mailto:kotaroee316@yahoo.co.jp) (K. Oe).

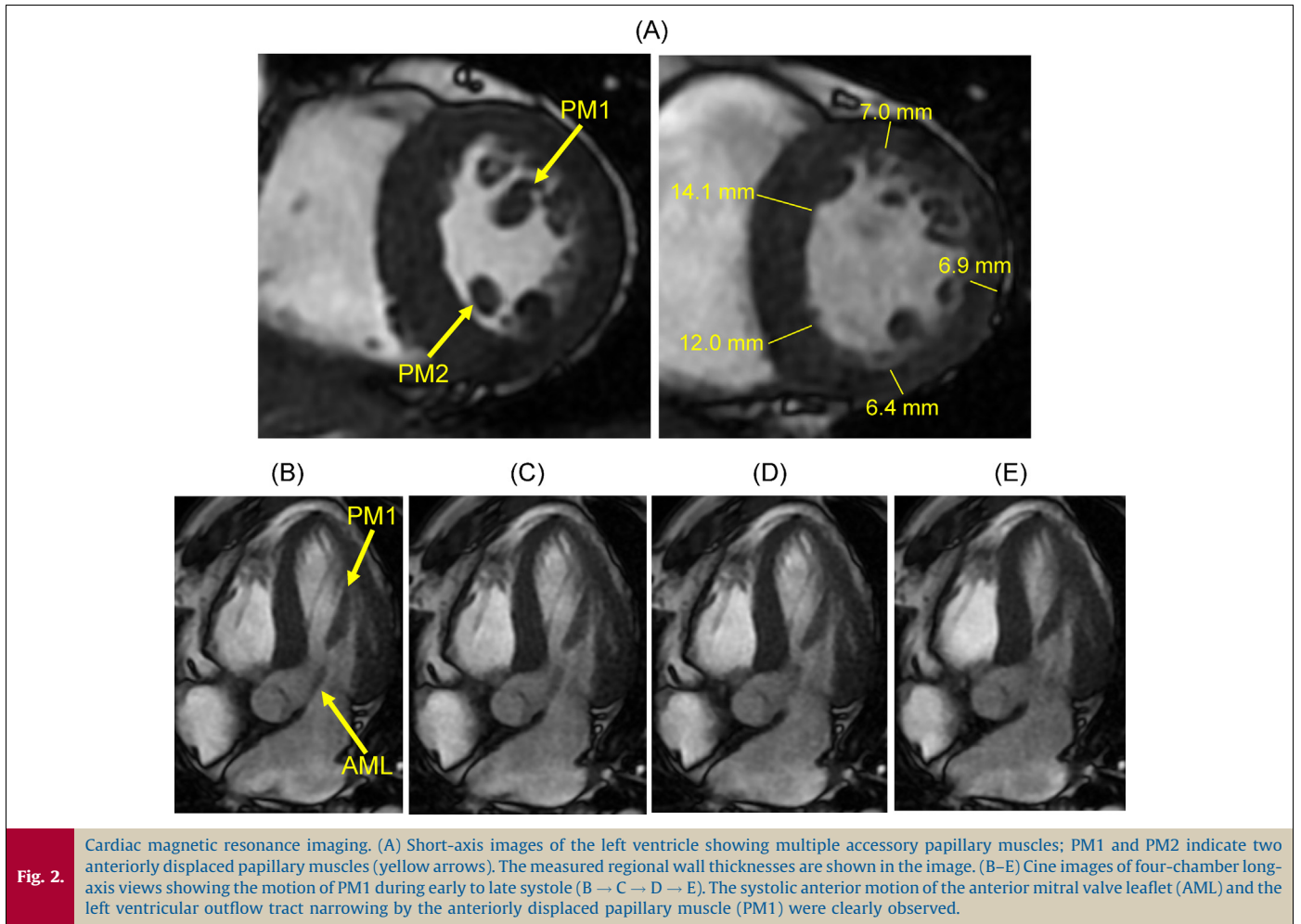


revealed normal left ventricular wall thickness (septal wall: 12 mm, posterior wall: 10 mm, septal/posterior wall thickness ratio: 1.2), dimension (end-diastolic: 41 mm, end-systolic: 22 mm), and contractility (ejection fraction: 77%) (Fig. 1A). Parasternal short-axis views of the left ventricle showed two anteriorly displaced PMs (Fig. 1B). M-mode recording at the mitral valve level showed systolic anterior motion (SAM) of the anterior mitral valve leaflet (AML) (Fig. 1C). Apical long-axis views showed elongation of the AML and protrusion of the AML into the LVOT (Fig. 1D). Color flow imaging showed accelerated flow at the LVOT and continuous wave Doppler recorded a pressure gradient (PG) of 56 mmHg (Fig. 1E). CMR imaging revealed mild asymmetric septal hypertrophy (anterior septal wall: 14.1 mm, posterior wall: 6.9 mm) which was not apparent on echocardiography, and confirmed the presence of multiple accessory PMs, some of which were displaced anteriorly (Fig. 2A). Cine images clearly displayed SAM and LVOT obstruction due to anteriorly displaced PM (Fig. 2B–E). Left ventricular pressure measured at the apex of left ventricle and LVOT showed a PG of 40 mmHg (Fig. 3). Coronary angiography showed no significant stenosis. Based on these findings, the patient was diagnosed as having HCM and LVOT obstruction due to abnormal PMs. Oral administration of bisoprolol (2.5 mg/day) was initiated, because the patient rejected surgical treatment. The daily dose of bisoprolol was maintained at 2.5 mg because of bradycardia. Follow-up echocardiography performed 6 months and 2 years after the initiation of bisoprolol revealed a gradual decrease in the LVOT-PG to 42 and 24 mmHg, respectively. The systolic heart murmur also decreased to

grade 2/6. In addition, no episodes of fainting or syncope recurred for 2 years after the initiation of bisoprolol.

## Discussion

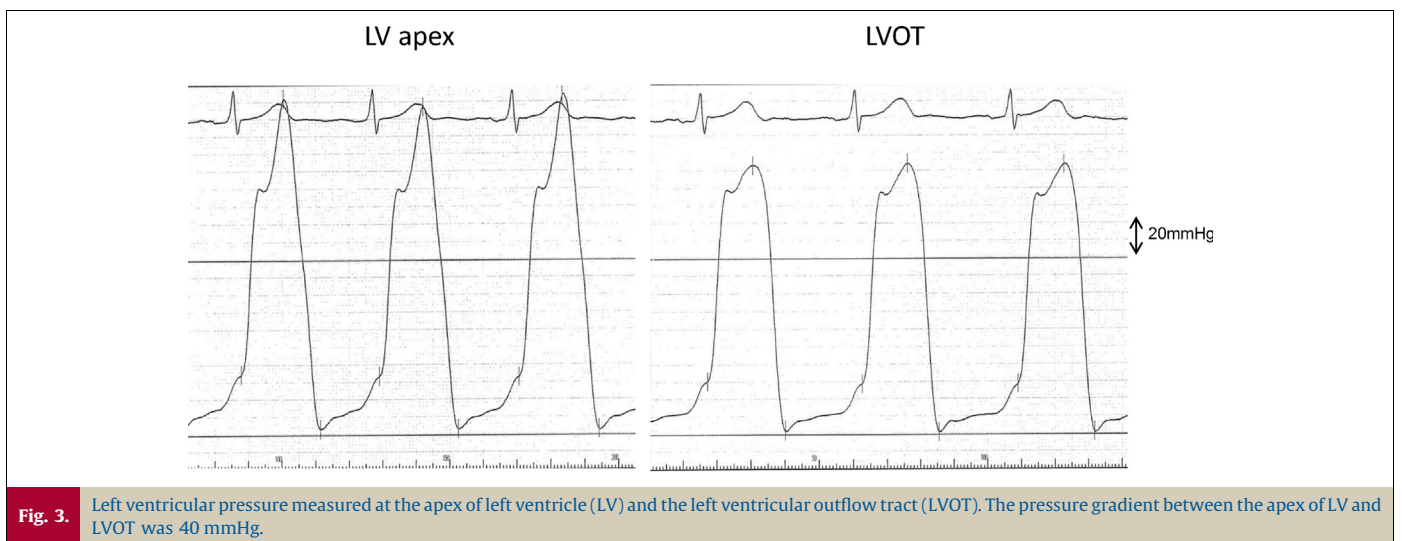
Hypertrophic obstructive cardiomyopathy is a common cause of LVOT obstruction, whereas abnormal PM, although unusual, has been reported to cause LVOT obstruction [1]. Roberts and Cohen classified abnormal PM morphology into the following groups: single PM, accessory PMs, abnormally large (or small) and malpositioned PM, and insertion of the PM directly into the mitral leaflet [9]. In the present case, the number of PMs was high, and a few of these PMs were anteriorly displaced; thus, the patient was diagnosed as having accessory PMs and malpositioned PMs. Although left ventricular hypertrophy was not apparent on echocardiography, CMR imaging revealed mild asymmetric septal hypertrophy; thus, the patient was diagnosed as having HCM. Previous studies reported that abnormalities of the PM and mitral valve were often found in patients with HCM [2], and CMR was useful for diagnosis of these abnormalities [3–5]. Harrigan et al. evaluated PM morphology in 201 patients with HCM using CMR imaging, and indicated that PM number and mass were increased in HCM patients, and anteriorly displaced PMs were associated with LVOT obstruction [3]. Furthermore, Kwon et al. assessed the relationship between abnormal PM morphologies and LVOT obstruction in 56 patients with HCM using CMR, and noted that patients with anteroapically displaced PMs had higher LVOT-PG,



independent of septal wall thickness [4]. Indeed, patients with abnormal PM and LVOT obstruction who had mild or no septal hypertrophy were reported [6–8]. Using experimental dog models, Levine et al. demonstrated that the anterior displacement of the PM could cause SAM without septal hypertrophy, suggesting that PM displacement and mitral leaflet slack may be the main causes of

SAM in HCM [10]. In the present case, septal hypertrophy was not severe, and therefore, PM displacement and AML elongation were considered to be the main cause of LVOT obstruction.

Abnormal PMs with LVOT obstruction are usually treated by surgical correction [6,7], and therefore, only a few reports have described the efficacy of medical treatments in patients with LVOT



obstruction due to abnormal PMs [8]. Izgi et al. administered metoprolol 100 mg daily to treat a patient with LVOT obstruction due to abnormal PMs; in this patient, a decrease in the LVOT-PG and symptomatic improvement was noted [8]. Similarly, in the present case, the LVOT-PG decreased and symptoms resolved after the initiation of beta-blocker therapy. Exercise stress or amyl nitrate consumption would increase LVOT-PG in patients with LVOT obstruction due to abnormal PMs [7,8], suggesting that the hemodynamic characteristics may be similar between LVOT obstruction due to abnormal PMs and hypertrophic obstructive cardiomyopathy. Beta-blockers have been considered to be the first-line therapy for hypertrophic obstructive cardiomyopathy. The findings of the present case suggest that beta-blockers might be a useful treatment for LVOT obstruction due to abnormal PMs, although this should be confirmed by future clinical studies.

### Conclusion

We report a case of LVOT obstruction due to abnormal PMs, which was accurately diagnosed by CMR imaging and successfully treated with a beta-blocker. Although transthoracic echocardiography is the most useful for diagnosis and follow-up for patients with HCM, CMR imaging could provide more detailed information about abnormal PMs and LVOT obstruction. Beta-blockers might be effective for reducing LVOT-PG in patients with LVOT obstruction due to abnormal PMs.

### Conflict of interest

None.

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