ESC European Society of Cardiology

European Heart Journal - Case Reports (2023) **7**, 1–5
https://doi.org/10.1093/ehjcr/ytac493

CASE REPORT
Heart failure

Percutaneous transluminal septal myocardial ablation was effective in hypertrophic obstructive cardiomyopathy with anomalous mitral papillary muscles: a case report

Naoko Yuzawa-Tsukada [1]*, Michifumi Tokuda[1], Takashi Miyamoto[2], and Michihiro Yoshimura[3]

[1]Division of Cardiology, Department of Internal Medicine, The Jikei University Katsushika Medical Center, 6-41-2 Aoto, Katsushika-ku, Tokyo 125-8506, Japan; [2]Division of Cardiology, Saitama Cardiovascular Respiratory Center, Kumagaya, Saitama, Japan; and [3]Division of Cardiology, Department of Internal Medicine, The Jikei University School of Medicine, Tokyo, Japan

Received 8 December 2021; first decision 10 January 2022; accepted 30 December 2022; online publish-ahead-of-print 6 January 2023

Background Abnormalities of the mitral subvalvular apparatus are not uncommon in hypertrophic obstructive cardiomyopathy (HOCM). Where invasive treatment is indicated in these patients to reduce left ventricular outflow tract (LVOT) obstruction, surgical myectomy with mitral valve repair is recommended.

Case summary In this report, we describe the case of a patient with HOCM and anomalous papillary muscle anatomy, successfully treated by percutaneous transluminal septal myocardial ablation (PTSMA).

Discussion PTSMA effectively reduced septal myocardial thickness and LVOT gradient, with only mild residual systolic anterior motion and mitral regurgitation despite anomalous papillary muscle anatomy. Upon careful anatomical evaluation, PTSMA may be a suitable therapeutic option for patients with LVOT obstruction and mitral valve abnormalities who are poor surgical candidates.

Keywords Case report • Percutaneous transluminal septal myocardial ablation • Hypertrophic obstructive cardiomyopathy • Papillary muscles abnormalities

ESC Curriculum 2.3 Cardiac magnetic resonance • 2.1 Imaging modalities • 2.2 Echocardiography • 6.5 Cardiomyopathy

Learning points

- HOCM is frequently associated with mitral valve abnormalities.
- PTSMA may be effective in HOCM with mitral valve abnormality, where the papillary muscles and tendon cords are attached to the septal myocardium, resulting in outflow tract obstruction.
- For better surgical planning, it is important to evaluate comprehensive imaging such as transoesophageal echocardiography and cardiac magnetic resonance before surgery.

* Corresponding author. Tel: +81 3 3433 2111, Email: h21ms-yuzawa@jikei.ac.jp
Handling Editor: Grigoris Karamasis
Peer-reviewers: Federica Mango; Doralisa Morrone
Compliance Editor: Marta Peverelli
Supplementary Material Editor: Jonathan Senior

Introduction

Concomitant mitral valve abnormalities are common in patients with hypertrophic obstructive cardiomyopathy (HOCM). Papillary muscle (PM) anomalies were reportedly present in up to 13% of autopsy specimens from patients with HOCM.[1] Surgical repair is usually recommended.[2] Here, we report a case of HOCM with anomalous mitral PM that was effectively treated with percutaneous transluminal septal myocardial ablation (PTSMA).

Timeline

Time	Events
7 August 2018	A patient with HOCM who presented with New York Heart Association (NYHA) III heart failure symptoms despite having undergone medical therapy visited our specialty outpatient department for the first time. On transthoracic echocardiography (TTE) at rest, the left ventricular outflow tract (LVOT) pressure gradient (PG) was not an indication for treatment. Computed tomography showed mild emphysema. Pulmonary function tests showed a decrease in pulmonary function with a 1 s rate of 63.8%.
September 2018	Exercise stress echocardiography was performed to confirm that the cause of shortness of breath on exertion was HOCM, and it showed an elevated LVOT PG, which was an indication for therapeutic treatment.
October 2018	Transoesophageal echocardiography and cardiac magnetic resonance imaging (MRI) showed the malformation of PM attached to the ventricular septal myocardium. The patient refused Morrow surgery and requested percutaneous transluminal septal myocardial ablation (PTSMA).
6 December 2018	The patient was admitted for catheterization, which confirmed the absence of significant stenosis in the coronary arteries and identified a septal branch as a target for PTSMA. We had a conference with the cardiac surgery department. It was concluded that PTSMA could be considered in this case.
8 April 2019	The patient was admitted for PTSMA. B-type natriuretic peptide (BNP) level was 1122.3 pg/mL.
10 April 2019	PTSMA was performed.
16 April 2019	BNP level regressed to 332.7 pg/mL.
18 April 2019	The patient was discharged from the hospital without any complications.
May 2019	The LVOT PG improved after PTSMA. The patient's symptom also improved to NYHA I.
May 2020	Echocardiographic findings did not change significantly from May 2019, and the LVOT PG remained at the improved level after PTSMA.

Case presentation

A 62-year-old male patient with HOCM visited our specialty outpatient department complaining of NYHA III symptoms despite having received medical therapy (bisoprolol 5 mg/day, diltiazem hydrochloride 100 mg/day). He had no leg oedema but a systolic murmur was detected. The apex of the heart was located at the fifth intercostal space on the anterior axillary line. The patient did not take any other drugs for HOCM, and he had no significant medical history other than HOCM. His BNP level was 1122 pg/mL (reference, <18.4 pg/mL). We could not use cibenzoline, which is known to prolong the QTc interval, because electrocardiography showed a prolonged QTc interval of 486 ms, narrow QRS, and regular sinus rhythm. The cardiothoracic ratio was 53% with no significant radiographic findings. Computed tomography showed mild emphysema, and pulmonary function tests revealed a 1 s expiratory rate of 63.8%.

Resting TTE revealed systolic anterior motion (SAM) due to LVOT obstruction and mild mitral regurgitation (MR). The LVOT PG was 45 mmHg, which was not an indication for treatment (*Figure 1A*, Supplementary material online, *Video S1*). Therefore, exercise stress echocardiography was performed to confirm that HOCM was the cause for shortness of breath on exertion. The pre-loading LVOT PG of 78 mmHg (*Figure 1B*) increased immediately to 84 mm post loading (2 min, 5.4 metabolic equivalents), and MR worsened to a moderate grade (*Figure 1C*, Supplementary material online, *Video S2*). Furthermore, pulmonary hypertension was exacerbated, and patient developed shortness of breath, presumably due to HOCM. Transoesophageal echocardiography revealed an abnormal PM adhesion to the septal myocardium (*Figure 2A*). Moderate-to-severe MR due to SAM has also been observed. Cardiac MRI showed two parts of the anterior PM, one of which was attached to the septal myocardium (*Figure 2B and C*). The PM thickness was 7.3 mm, and the cross-sectional area was 98.7 mm^2. We explained that surgical intervention was the first-line treatment in this case, but the patient refused. As per the patient's request, we chose PTSMA despite a PM anomaly.

Patient was hospitalized and catheterization was performed, which confirmed that there was no significant stenosis in the coronary artery, and identified the septal branch as the target of PTSMA. After consultation with the department of cardiac surgery, it was concluded that PTSMA could be considered in this case.

PTSMA was performed as follows.[3] A temporary pacemaker was placed because of the high risk of developing a new periprocedural atrioventricular block (AVB), and a catheter was placed in the LV to continuously monitor the LVOT gradient. On the simultaneous pressure measurement, a significant PG was observed between the LV inflow and outflow; the peak PG was 17 and 57 mmHg, respectively. We targeted the 1st and 2nd septal branches (S1 and S2, respectively; *Figure 3*). First, a balloon catheter was inserted into the selected septal branch. Contrast medium was injected through the lumen to confirm septal branch occlusion using a balloon. Next, agitated radiographic contrast was injected to identify the responsible septal myocardium on the echocardiogram. Finally, ethanol was infused until the myocardium turned completely white and an acoustic shadow appeared on the echocardiogram. The PM attached to the septum did not turn white in colour. The total ethanol usage was 9.5 mL (4.4 mL for S1, 5.1 mL for S2). After balloon catheter removal, angiography was repeated to determine whether the target septal branch was occluded and the left anterior descending coronary artery was patent. After PTSMA, the aorta–LV PG decreased from 103 to 20 mmHg at rest, 197 to 91 mmHg during the Valsalva manoeuver, 199 to 132 mmHg according to the Brockenbrough–Braunwald–Morrow sign, and 114–102 mmHg after nitroglycerin loading (see Supplementary material online, *Table S1*). The patient was discharged 8 days post procedure without complications (e.g. complete AVB).

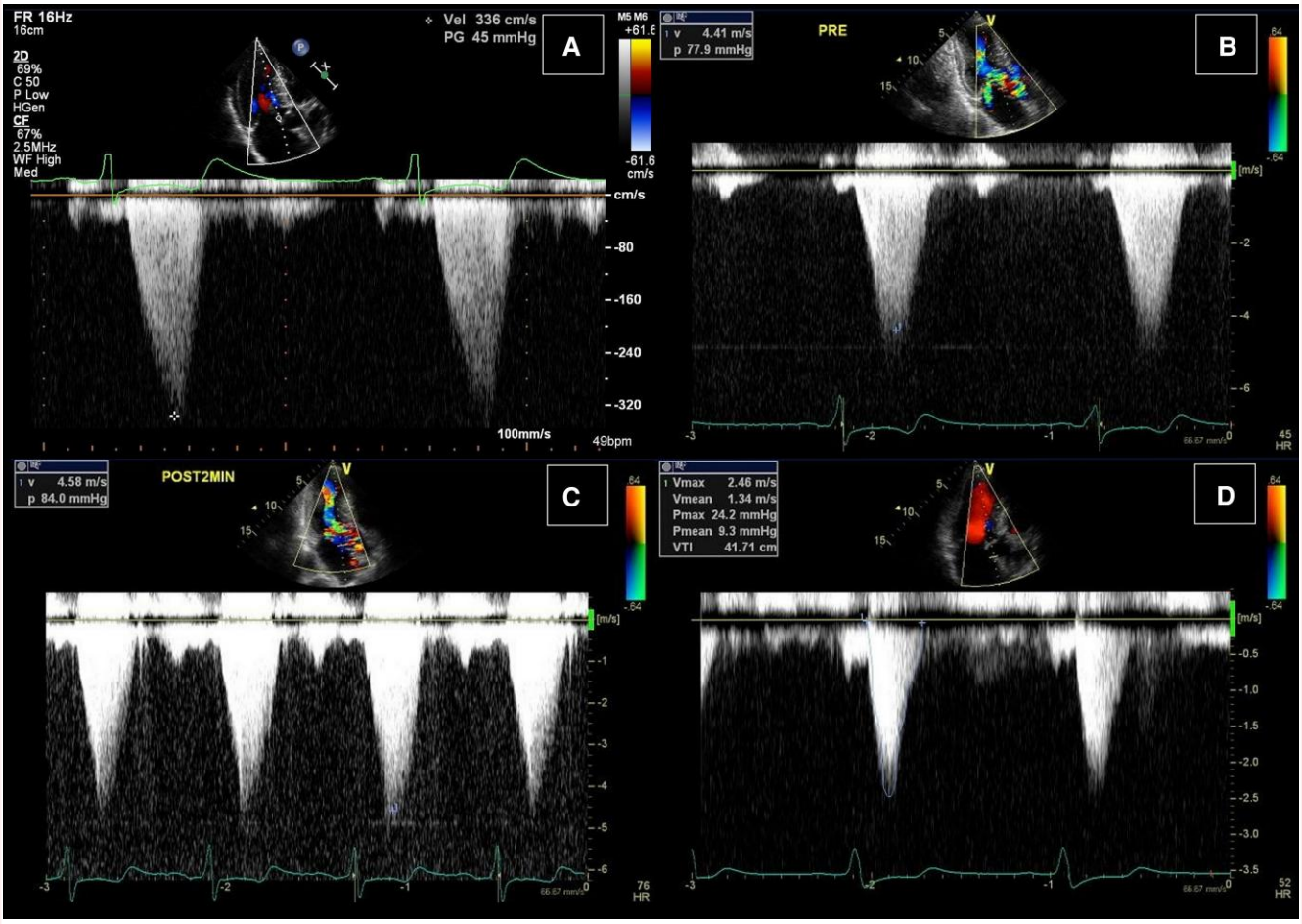

Figure 1 Four echocardiac images showing the left ventricular outflow tract pressure gradient at each time point as follows: (*A*) before the percutaneous transluminal septal myocardial ablation at rest; (*B*) before exercise; (*C*) after exercise; (*D*) after percutaneous transluminal septal myocardial ablation at rest.

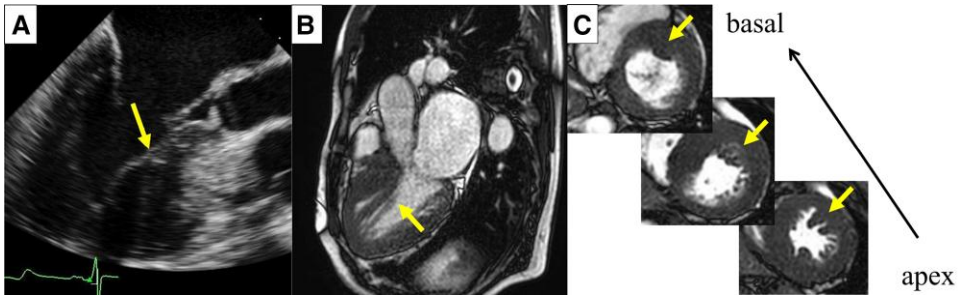

Figure 2 (*A*) Transoesophageal echocardiography. (*B*) Cardiac magnetic resonance imaging long-axis image. (*C*) Cardiac magnetic resonance imaging short-axis image. All images demonstrate a papillary muscle structural abnormality (arrow).

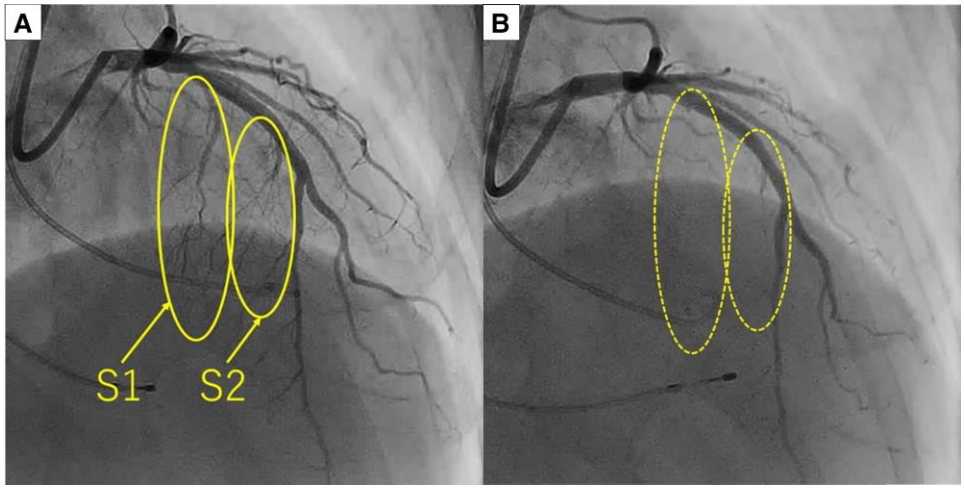

Figure 3 (*A*) Coronary angiography before the percutaneous transluminal septal myocardial ablation. We ablated the two septal branches (S1 and S2). (*B*) Coronary angiogram taken after percutaneous transluminal septal myocardial ablation.

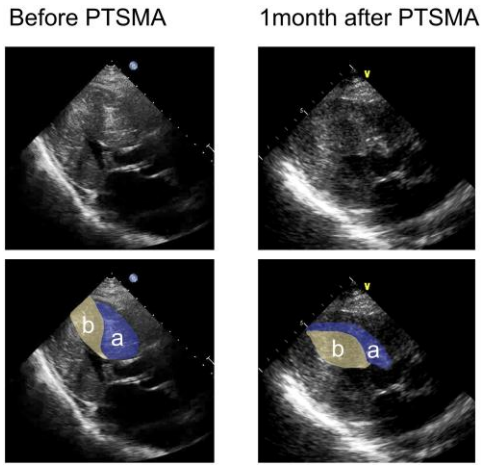

Figure 4 Before percutaneous transluminal septal myocardial ablation and 1 month after percutaneous transluminal septal myocardial ablation. The septal myocardium (a) is smaller, whereas the thickness of the papillary muscle (b) attached to the septal myocardium has not changed.

TTE performed one month post-PTSMA showed that the SAM persisted, but regressed to a mild state (see Supplementary material online, *Video S3*). MR due to SAM also improved to a trivial grade (see Supplementary material online, *Video S4*). The interventricular septal thickness decreased from 21 to 18 mm (*Figure 4*). Although the PM adherent to the septum persisted and moved to the LVOT during systole, its gradient decreased from 84.0 to 24.2 mmHg due to the reduced septal myocardial thickness (*Figure 1D*). The BNP level improved from 1122.3 to 332.7 pg/mL at 1 week post treatment and 58.0 pg/mL at 1 year post treatment. The patient's symptoms improved to NYHA Class I.

Discussion

In patients with HOCM, abnormal PM causes mitral valve abnormalities and increases the resting LVOT PG.[4] In the present case, the anomalous PM and thickened septal myocardium may have caused LVOT obstruction and led to SAM and MR. The 2020 US guidelines support the use of surgery, such as modified Morrow myectomy, for patients with mitral structural abnormalities.[2] This is because PTSMA only affects a thickened septal myocardium, whereas repair of the mitral valve structure during the Morrow procedure can eliminate the causes of SAM in patients with mitral valve abnormalities. Patients with mitral valve abnormalities, including anterior PM displacement, reportedly have persistent SAM, LVOT gradient, and MR post-PTSMA.[5] To the best of our knowledge, no reports has detailed PTSMA in patients with HOCM and abnormal mitral valve structure.

In this case, PTSMA was effective because despite direct PM insertion into the septal myocardium, the reduced septal myocardial thickness improved the LVOT obstruction. As PTSMA mainly reduces the septal myocardium, its therapeutic effect in patients with HOCM and malformation depends on the PM thickness. An accurate evaluation of the PM morphology using cardiac MRI is recommended. Moreover, it is essential to assess the risk of PTSMA complications, such as chest pain, complete AVB, extensive myocardial infarction due to ethanol leakage into the left anterior descending coronary artery, and ventricular arrhythmias.[6]

BNP, whose value plateaus three months after treatment, can help evaluate PTSMA efficacy.[7] The BNP level markedly improved in the present case until 1 year post treatment.

Conclusion

In HOCM, mitral valve malformations should be thoroughly evaluated before treatment, as it may affect the treatment strategy. In the present case, PTSMA was effective despite abnormal PM attachment; however, our findings require further validation.

Lead author biography

Dr Naoko Tsukada is a 7-year cardiologist at the Jikei Universit Katsushika Medical Center. She graduated from The Jikei University School of Medicine in 2015. She has a special interest in structural heart disease.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Acknowledgement

The authors thank Dr Brian Quinn (Japan Medical Communication Inc.) for his comments on the language of the manuscript.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for the submission and publication of this case report, including images and associated text, was obtained from the patient according to COPE requirements.

Conflict of interest: None declared.

Funding: None declared.

References

1. Klues HG, Roberts WC, Maron BJ. Anomalous insertion of papillary muscle directly into anterior mitral leaflet in hypertrophic cardiomyopathy. Significance in producing left ventricular outflow obstruction. *Circulation* 1991;**84**:1188–1197.
2. Ommen SR, Mital S, Burke MA, Day SM, Deswal A, Elliott P, *et al.* 2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy: executive summary: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation* 2020;**142**:e533–e557.
3. Nagueh SF, Groves BM, Schwartz L, Smith KM, Wang A, Bach RG, *et al.* Alcohol septal ablation for the treatment of hypertrophic obstructive cardiomyopathy. A multicenter North American registry. *J Am Coll Cardiol* 2011;**58**:2322–2328.
4. Kwon DH, Setser RM, Thamilarasan M, Popovic ZV, Smedira NG, Schoenhagen P, *et al.* Abnormal papillary muscle morphology is independently associated with increased left ventricular outflow tract obstruction in hypertrophic cardiomyopathy. *Heart* 2008;**94**:1295–1301.
5. Delling FN, Sanborn DY, Levine RA, Picard MH, Fifer MA, Palacios IF, *et al.* Frequency and mechanism of persistent systolic anterior motion and mitral regurgitation after septal ablation in obstructive hypertrophic cardiomyopathy. *Am J Cardiol* 2007;**100**:1691–1695.
6. Kuhn H, Lawrenz T, Lieder F, Leuner C, Strunk-Mueller C, Obergassel L, *et al.* Survival after transcoronary ablation of septal hypertrophy in hypertrophic obstructive cardiomyopathy (TASH): a 10 year experience. *Clin Res Cardiol* 2008;**97**:234–243.
7. Akita K, Tsuruta H, Yuasa S, Murata M, Fukuda K, Maekawa Y. Prognostic significance of repeated brain natriuretic peptide measurements after percutaneous transluminal septal myocardial ablation in patients with drug-refractory hypertrophic obstructive cardiomyopathy. *Open Heart* 2018;**5**:e000786.