Alicov et al first reported in 1976 a subtype of hypertrophic obstructive cardiomyopathy (HOCM) with mid-ventricular obstruction, which is a rare subtype of HOCM, comprising 1% of hypertrophic cardiomyopathies. We present an unusual, pediatric case of mid-ventricular hypertrophic obstructive cardiomyopathy (MVHOCM).

**Case Report**

At the age of 10 years, the patient first presented to hospital with severe chest pain, which occurred early in the morning and disappeared spontaneously with bed rest. The family history was not significant. Based on the echocardiographic findings, he was diagnosed as having hypertrophic cardiomyopathy (HCM) and was prescribed 30 mg of propranolol daily. Four months later, he was admitted for further evaluation: blood pressure was 118/78 mmHg, pulse rate 80 beats/min and respiratory rate 20 breaths/min; grade 3/6 harsh systolic murmur at the lower left sternal border. Electrocardiogram showed a mean frontal QRS axis of –60° and a deep S wave in V1; chest X-ray showed neither cardiomegaly nor pulmonary venous congestion. Two-dimensional echocardiography showed asymmetric septal hypertrophy, most marked at the mid-ventricular level, and papillary muscles coming into contact at the mid-ventricular level during systole. The thickness of the interventricular septum and of the left ventricular posterior wall was 15 and 10 mm, respectively. There was no evidence of systolic anterior motion of the mitral valve. Color Doppler echocardiography showed formation of a stenotic jet into the body of the left ventricle, and a peak systolic gradient of 100 mmHg was recorded by continuous-wave Doppler echocardiography. MVHOCM was suspected, and a significant mid-ventricular pressure gradient of 89 mmHg (left ventricular apex 193/end-diastolic pressure (EDP) 12 mmHg, left ventricular outflow 104/EDP 10 mmHg, aorta 89/47/mean 63 mmHg) was confirmed by cardiac catheterization. We finally diagnosed MVHOCM from the left ventriculogram, which demonstrated a hourglass appearance at systole (Fig 1). The coronary angiogram was normal. A right ventricular endomyocardial biopsy specimen showed hypertrophic cardiomyocytes and augmentation of interstitial collagen fiber. Plasma $\beta$-galactosidase activity was 13.8 nmol·h$^{-1}$·ml$^{-1}$ (normal, 4.8–17.6). The patient did not have the characteristic skin manifestation of Fabry’s disease. An exercise stress $^{99}$mTc tetrofosmin myocardial scintigram disclosed no obvious perfusion defect, but the second $^{123}$I-[]-methyl iodophenyl pentaadecanoic acid (BMIPP) myocardial scintigram (Fig 2A, at the age of 11 years and 11 months) showed a reduced uptake by the septal and inferior walls. He was put on restricted exercise with 30 mg/day of propranolol and continued for approximately 2 years without complaint. The chest pain and dyspnea on exertion reappeared at the age of 12 years and 3 months. He sometimes became pale, which was considered to be indicative of presyncope. The second $^{123}$I-BMIPP myocardial scintigram (Fig 2B, at

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**Effective Disopyramide Treatment in a Boy With Mid-Ventricular Hypertrophic Obstructive Cardiomyopathy**

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A 14-year-old boy with mid-ventricular hypertrophic obstructive cardiomyopathy (MVHOCM) first presented at the age of 10 years with severe chest pain. Two-dimensional echocardiography disclosed marked hypertrophy at the mid-portion of the ventricular septum, and left ventriculography showed an hourglass appearance at systole. He was initially treated with propranolol, but the chest pain and dyspnea on exertion worsened at the age of 12 years. After disopyramide was started, the chest pain disappeared and the degree of the pressure gradient at the mid-ventricular level was reduced. There was also significant improvement on a $^{123}$I-[]-methyl iodophenyl pentaadecanoic acid (BMIPP) myocardial scintigraphy.

**Key Words:** Disopyramide; $^{123}$I-[]-methyl iodophenyl pentaadecanoic acid (BMIPP) myocardial scintigraphy; Mid-ventricular hypertrophic obstructive cardiomyopathy; Pediatrics

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Fig 1. Left ventriculogram demonstrating the hourglass appearance. The arrow shows stenosis at the mid-ventricular level. (Left) End diastole, (Right) end systole.
the age of 13 years) showed a severely reduced uptake, mainly in the septal wall. The dose of propranolol was increased to 60 mg/day, but was not effective, so 300 mg/day of disopyramide was added, which was followed by a dramatic improvement in the symptoms. We also noticed that the pressure gradient at the mid-ventricular level, estimated by continuous-wave Doppler echocardiography, reduced from 105 (at the age of 12 years and 3 months) to 64 mmHg (at the age of 13 years and 9 months), 5 months after the disopyramide was started. The third scintigram (Fig 2C, at the age of 13 years and 9 months), taken after the treatment of disopyramide had begun, showed an improved uptake in the septal wall. There has not been evidence of left ventricular hypokinesia for 4 years and 2 months; the patient is now 14 years old and doing well on 60 mg of propranolol and 300 mg of disopyramide daily.

Discussion

We have described a rare case of MVHOCM in a child, which responded to disopyramide. The diagnosis was established by the hourglass appearance on the left ventriculogram and the pressure gradient at the mid-ventricular level. The anatomic causes of mid-ventricular obstruction are thought to be: (1) hypertrophy of the papillary muscles; (2) associated with apical infarction; or (3) a membrane-like structure. In the present case, there was significant hypertrophy of the antero-lateral papillary muscle, which made contact with the left ventricular wall, creating a sphincter-like effect and the subsequent mid-ventricular obstruction. Apical infarction was not apparent on the myocardial blood-flow scintigram.

HCM is usually, but not always, associated with microscopic evidence of myocardial fiber disarray. The biopsy specimen of this case, however, did not have this characteristic feature and so our diagnosis of MVHOCM was made from the combination of clinical features and histology. Some myocardial storage diseases (Fabry’s disease, amyloid heart disease, glycogen storage disease) can mimic the full diagnostic picture of HCM, but can be ruled out by normal plasma β-galactosidase activity, absence of amyloid, and absence of damage caused by the deposition of glycogen granules, respectively.

Propranolol, a negative inotropic agent, is used to decrease the degree of outflow obstruction in HCM, but was not effective in this case. After disopyramide was started, the symptoms disappeared. Disopyramide, a class Ia antiarrhythmic agent, not only has a negative inotropic effect but also an improving effect on left ventricular diastolic function\textsuperscript{10,11} and we consider that it was both these effects that led to the significant improvement in the patient’s symptoms.

It is well known that \textsuperscript{123}I-BMIPP scintigraphy is useful for detecting the abnormal myocardial fatty acid metabolism in HCM\textsuperscript{12,13}. \textsuperscript{123}I-BMIPP is a branched-chain fatty acid in which a methyl group is introduced to the β-position of the carboxyl group. It accumulates in the myocardium and is retained for a long time. In the present case, the \textsuperscript{123}I-BMIPP scintigram taken after the treatment of disopyramide had begun showed a significant improvement in the septal wall and we speculate that it was because of an improvement in the myocardial ischemia. That is to say, we consider that slight ischemia, not detectable with \textsuperscript{99m}Tc tetrofosmin myocardial scintigraphy, was present and that the abnormal fatty acid metabolism was caused by the ischemia. There is a close relationship between CD36 deficiency and reduced myocardial uptake of \textsuperscript{123}I-BMIPP in HCM,\textsuperscript{4} but in the present case flowcytometric analysis showed that the CD36 molecule expression was positive on both platelets and monocytes.

Adult patients with MVHOCM are at a high risk of developing segmental or diffuse left ventricular hypokinesia\textsuperscript{15} but this is less well known in children with MVHOCM because of its rarity in this age group; the present patient has not progressed to left ventricular hypokinesia during the past 4 years and 2 months.

To our knowledge, this is the first reported pediatric case of MVHOCM. We propose that disopyramide should be used in children with MVHOCM to decrease the degree of mid-ventricular obstruction leading to myocardial ischemia, and that \textsuperscript{123}I-BMIPP myocardial scintigraphy should be used during the follow-up of all patients with MVHOCM.

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