A 51-year-old man presented with exertional dyspnea, especially during post-prandial periods. On physical examination the fourth sound and mid-late systolic murmurs were heard. The systolic murmur did not radiate to the neck, but it did increase with the Valsalva maneuver and in the standing position. Interestingly, an extra sound was audible between the first sound (S1) and a paradoxically split, second sound (S2) over a wide area, that is, from the second to fourth left sternal borders as well as the apex, with the intensity being loudest at the third left sternal border (Movie S1). No third sound was heard. The remainder of the examination was normal. The patient had not taken any regular medication. His mother had hypertrophic cardiomyopathy (HCM).

An electrocardiogram showed left ventricular (LV) hypertrophy with a QRS width of 115 ms. Echocardiography indicated an LV ejection fraction of 65%, asymmetric septal hypertrophy with a maximum wall thickness of 21 mm, systolic anterior motion (SAM) of the mitral valve, LV outflow tract (LVOT) obstruction with a resting pressure gradient of 92 mmHg, and mild mitral regurgitation (Movie S2). The peak pressure gradient across the LVOT was 96 mmHg during the Valsalva maneuver, and increased to 185 mmHg in a sitting position. A diagnosis of obstructive

**Figure 1.** (A) Phonocardiography at the third left sternal border shows a loud, low-pitched sound approximately 130 ms after the first sound (S1) (arrow) and systolic murmurs that end before the paradoxically split, second sound (S2) with the pulmonary component accentuated (asterisk). (B) Echocardiography shows an asymmetric septal hypertrophy with systolic anterior motion of the mitral valve; the systolic sound (arrow) coincides with the contact of the mitral leaflet with the interventricular septum (arrowhead).
Figure 2. (A) After a premature ventricular contraction with a compensatory pause, the systolic anterior motion (SAM)-associated sound increases in intensity and appears approximately 30 ms earlier (arrow), as does the mid-late systolic murmur (asterisk). Also note the attenuated aortic component of S2 after the premature ventricular contraction, probably due to increased left ventricular outflow tract obstruction. (B) The SAM-associated sound and murmur appear almost 20 ms earlier, compared with supine, in a 75° upright position; note that the S1–S2 duration is almost identical in both recordings although the heart rates are different. An increase in intensity is also seen in the murmurs (†) but not in the sounds. (C) With β-blocker therapy, the SAM-associated sounds almost disappear with a reduction of systolic murmurs, as well as the paradoxical split of S2. All three phonocardiograms were recorded at the third left sternal border on the same scale.
SAM-Associated Sound in HCM

HCM was made. Phonocardiography, which was obtained at the third left sternal border as described previously,1 recorded a low-pitched sound approximately 130 ms after S1, accompanied by systolic murmurs that did not continue to the aortic component of S2 (Figure 1A). The differential diagnosis of this uncommon systolic sound included ejection sounds, for example, as seen in a bicuspid or stenotic aortic or pulmonary valve. These conditions, however, were less likely in the present case, because these ejection sounds are generally high-pitched during early systole, and best heard at the apex or base.2 Systolic click due to prolapsed mitral valve could also be ruled out because of the absence of mitral valve prolapse. Given that the extra sound coincided with contact of the mitral leaflet with the interventricular septum or the sudden deceleration of blood flow due to SAM in the LVOT or both,3 the SAM-associated sound in the present case increased in intensity, along with louder systolic murmurs, after a premature ventricular contraction with a compensatory pause (Figure 2A). The onset of the SAM-associated sound was earlier after premature ventricular contractions than with normal conduction. Similar findings were obtained with postural change from supine to upright position (Figure 2B), although the intensity of the SAM-associated sounds was almost unchanged. After initiation of β-blockers with bisoprolol 5 mg daily, the SAM-associated sounds disappeared and the systolic murmurs decreased (Figure 2C), Follow-up echocardiography showed decreases in peak pressure gradient across the LVOT: 23 mmHg at rest, 37 mmHg at sitting, and 52 mmHg at standing. The patient continued to do well for >6 months with minimal symptoms in his daily activities.

A SAM-associated sound was first reported more than half a century earlier,4 and was estimated to occur in 11% of patients with obstructive HCM.5 This unique sound is known by various names,3 such as innocent late systolic click,4 pseudo-ejection sound,4 low-pitched mid-systolic sound,7 systolic click-like sound,8 or SAM sound.9 It is worth noting that a SAM-associated sound is not easy to hear on auscultation because it may be masked by loud systolic murmurs that are commonly heard in patients with obstructive HCM. In 10 patients with SAM-associated sounds on phonocardiography, the sound was audible only in 2.3 SAM-associated sounds are characteristically low or medium pitched, occur 70–150 ms after S1, and have a wax-and-wane pattern according to physiological conditions (i.e., enhancement with an increase in ventricular outflow tract gradient, and vice versa),3 all of which were observed in the present case. Given the dynamic changes in a SAM-associated sound, we believe that its assessment on auscultation is still of importance to understand the unique hemodynamics of obstructive HCM even in the era of advanced imaging.

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Disclosures

The authors declare no conflict of interest.

References


Supplementary Files

Supplementary File 1

Movie S1. After the first sound, an extra sound, followed by a systolic murmur, is audible at the third left sternal border.

Supplementary File 2

Movie S2. A parasternal long-axis view shows asymmetric septal hypertrophy, systolic anterior motion of the mitral valve, and left ventricular outflow tract obstruction.

Please find supplementary file(s):