Various Types of Systolic Clicks in Patients with Muscular Subaortic Stenosis

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Summary

Four clinical cases of subaortic hypertrophic muscular stenosis are discussed. All four, in addition to a loud systolic murmur, had a loud systolic click. However, the timing of the click and its relationship to the phase of the carotid pulse were different in each case varying from close to medium and to distant (or midsystolic). The comparison of the sound tracing with the carotid tracing demonstrated that, in each case, a sudden change in acceleration was taking place. In one, the click coincided with the onset of the carotid upstroke; in another, with the first peak of the carotid pulse; in the third, with the trough between first and second peak of the pulse; in the last, with a sudden drop of the pulse at midsystole caused by sudden obstruction to flow. It is concluded that the clicks were caused by rapid changes of acceleration resulting from the abnormal aortic ejection. Thus, the study of the carotid pulse and of the sound tracing are important for a non-invasive diagnosis together with the echo study of the septum and ventricular wall.

Additional Indexing Words:
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During our study of cardiac patients by routine non-invasive methods, four cases showed outstanding systolic clicks in the presence of muscular subaortic stenosis. As the characteristics of the clicks were different and their presence was uncommon, the documents of the four patients will be briefly discussed because they are significant in regard to the mechanism of production of such clicks.

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CASE 1

S. L.—The patient was an underdeveloped, 17-year-old Mexican boy. A heart murmur had been noted by physicians for several years. Fluid had been removed from his chest many years before. Present admission was due to dull abdominal pain.

On physical examination, several congenital abnormalities were noted including a high-arched palate, several accessory nipples, and pectus excavatum. The right testis was undescended and a large mass was present in the right lower quadrant of the abdomen, obviously an undescended testis.

There was no cyanosis or clubbing. The apical impulse of the heart was in the 6th I.C.S., 1 cm outside the left midclavicular line. A thrill was palpable in early systole at the apex. Auscultation revealed a grade III/VI harsh murmur that seemed pansystolic, both at apex and at the first right I.C.S. The first sound seemed very loud and was preceded by a short presystolic murmur. The second sound was faint. The carotid pulse was brisk and of brief duration.

LABORATORY STUDIES


Phonocardiogram (recorded over 3 areas with 4 high-pass filters)—The first sound started with an a component of moderate amplitude, followed by a b component that was extremely large (ejection sound simulating on auscultation the first sound, Fig. 1A). The click was 90 msec after QRS onset and simultaneous with carotid upstroke. The second sound was small and single. There was a fourth sound at the apex that, together with the first sound, simulated a presystolic murmur if recorded with low- or medium-high-pass filters (Fig. 1B, 1C). There was a pansystolic murmur of high frequency at the apex with larger vibrations in the first part of ejection (Fig. 1D).

Apex cardiogram—Large A wave. The systolic ejection was marked by 3 rapid spikes (thrill).

Carotid tracing—There was a very rapid upstroke followed by a rapid return to the baseline. As a result, the pulse occurred during the early phase of ejection. The ejection click coincided with the upstroke of the pulse.

Echocardiogram (2D)—Left atrial enlargement (4 cm). Normal left ventricular diastolic dimensions (4 cm at base). Posterior wall = 0.6 cm. Interventricular septum = 1.8 cm. Valve openings, normal. Possible systolic anterior motion of the mitral valve (SAM). No evidence of mitral valve prolapse.
Fig. 1. Case 1—In (A) and (B) three simultaneous phonocardiograms are recorded with the electrocardiogram. From above: 3rd right, 3rd left, and 5th left interspaces. In (A), the tracings were recorded with a high-pass filter at 50 Hz; in (B), at 200 Hz. Both show well the systolic click (c) but the systolic murmur is recorded better in (B). A small fourth sound and the first sound are recorded as a murmur that seemed presystolic on auscultation because of the large click that was thought to be the first sound. In (C) there is a phonocardiogram with high-pass filter at 100 Hz and a carotid tracing. The click coincides with the upstroke of the pulse. In (D) the third derivative of a phonocardiogram is recorded with a 200 Hz filter. The tracing shows the large click and a murmur that is maximal in the first half of ejection but continues up to the second sound.
The final diagnosis was idiopathic hypertrophic subaortic stenosis.

CASE 2

M.A.—This was a 70-year-old white woman, who was first admitted because of two masses on the lateral surface of the left chest wall. She had a history of hypertension for about 15 years. Following a left mastectomy, she had several other admissions. Recently she started developing palpitation, edema of the legs, orthopnea, and occasional paroxysmal nocturnal dyspnea.

On physical examination, the blood pressure was 160/70. Examination of the heart revealed a harsh, grade III/VI pansystolic murmur over the entire precordium, best heard at the apex (5th I.C.S.) and at the 3rd right I.C.S. The first sound was of increased loudness; it was preceded by a fourth sound and followed by a loud murmur within which one could hear a loud systolic click. The second sound was decreased and single.

LABORATORY STUDIES

Electrocardiogram—Sinus rhythm. Increased QRS voltage. Possible left atrial enlargement.

Phonocardiogram—Normal first sound, small and single second sound. Large fourth sound. With high-pass filtration, a large fourth sound was recorded (Fig. 2A). There was a pansystolic murmur of high frequency with superimposed diamond-shaped murmur. At midsystole, there was a large, complex click of an amplitude that was three times greater than that of the first sound. The click was recorded with any filter (from 25 to 400) (Fig. 2B) and occurred 220 msec after the QRS onset and 100 msec from carotid upstroke.

Carotid tracing—Rapid upstroke and immediate rapid downward drop; following this, there was a second rise with a rounded contour. The click coincided with the drop between first and second pulsation (Fig. 2D).

Echocardiogram (2D)—Mitral valve: reduced EF slope; no evidence of mitral valve prolapse; anterior motion in systole. Left atrial and ventricular dimensions within normal limits. Interventricular septum=2.0 cm. Posterior left ventricular wall at base=1.2 cm.

Final diagnosis: asymmetric septal hypertrophy with subaortic stenosis; systolic anterior motion of the mitral leaflets.

CASE 3

J. M.—This was an 80-year-old black man, who was admitted to the
hospital complaining of exertional dyspnea, orthopnea, and occasional precordial pain. In previous admissions to another hospital, chronic obstructive pulmonary disease had been diagnosed. A permanent artificial demand pacemaker had been installed in 1977 because of a slow pulse.

On physical examination, rapid pulse (120/min) and normal blood pressure were recorded. On auscultation of the heart, the first sound seemed
preceded by a fourth sound at the apex while the second sound was small and split at the base on both sides of the sternum.

LABORATORY STUDIES

Electrocardiogram—Junctional tachycardia with aberrant conduction. Right bundle branch block and left anterior hemiblock.

Phonocardiogram—There was a fourth sound in the low frequency (inaudible) range. The first sound was small and simulated a fourth sound on auscultation. The second sound was split at the base and was followed by a small click (opening snap?). The most prominent feature was a large systolic click (180 msec after onset of QRS and 60 msec after carotid upstroke). This was preceded and followed by small vibrations and simulated the first sound on auscultation (Fig. 3A).

Apex cardiogram—Lifting systolic impulse, non diagnostic.

Carotid tracing—Double peaked but rapid impulse; the first peak coincided with the prominent systolic click (Fig. 3B).

Echo (M-mode)—Increased E-septal separation and slow DE slope. Early-systolic closure of aortic cusps consistent with mitral regurgitation. No evidence of mitral valve prolapse. Systolic anterior motion of the leaflets. Interventricular septum=1.5 cm. Left ventricular wall thickness=1.0. Left ventricular diastolic dimension=6 cm (base).
Fig. 4. *Case 4*—Section (A) shows the sound tracings over the three areas, recorded at high frequency (200 Hz). Section (B) shows the apical phono (above), the right carotid phono (center), and the right carotid pulse. The latter has a sudden drop at midsystole, followed by a flat portion in late systole. The click (c) coincides with the sudden drop of the pulse tracing.
Conclusion—Asymmetric septal hypertrophy. Probable old inferior wall infarct. Enlarged left ventricle with dysfunction.

CASE 4

G. B.—This was a 90-year-old white woman with a long history of hypertension and a recent fracture of the right hip. She never experienced either dyspnea or precordial pain.

On physical examination, the pulse was regular, 74/min; the blood pressure, 114/60. There was no engorgement of the jugular veins. The cardiac apex was located in the 6th left I.C.S., 2 cm outside the midclavicular line. There was a grade 3, harsh, pansystolic murmur, best heard at the apex radiating toward the left axilla, and also heard at both the lung bases. There was no pedal edema and no abnormality of the other organs.

LABORATORY STUDIES

Electrocardiogram—Sinus bradycardia (55/min), normal PR (0.20). Left anterior hemiblock. Evidence of left ventricular hypertrophy and dilatation.

Phonocardiogram—First sound of normal amplitude. Second sound of decreased amplitude and wide splitting. Small fourth sound of low frequency. There was a large pansystolic and diamond-shaped murmur, best recorded at the apex and at the 3rd right I.C.S. In the high frequency, the murmur had a sharp decrease in the second part of ejection (Fig. 4A), often after a large click (Fig. 4B-c) (320 msec from QRS onset and 160 msec from carotid upstroke).

Apex cardiogram—Tall pulsation, non diagnostic.

Carotid tracing—Rapid upstroke followed by a rapid downstroke at midsystole, and then a flat portion before the incisura. The large click coincided with the drop of the pulse (Fig. 4B).

Echo (M-mode)—Left ventricular end-diastolic dimension = 3.9 cm; end-systolic = 2.6. Left ventricular inferior wall = 1.5 cm. Interventricular septum = 2.2 cm. The final diagnosis was hypertrophic cardiomyopathy with moderate subaortic obstruction.

DISCUSSION

The occurrence of systolic sounds in hypertrophic subaortic stenosis has been debated for a long time; older authors denied them. Braunwald et al1 noted early systolic sounds in 11% of 64 cases. De Joseph et al2 observed them in 19% of 42 patients; they mentioned that the systolic sounds were
delayed (even though they were still called “ejection sounds”) and occurred at the first peak of the carotid pulse. In a previous study, Snellen\textsuperscript{3} had described a loud, low-pitched systolic sound that coincided with the downstroke that followed the first peak of the carotid pulse. On the other hand, Hancock\textsuperscript{4} had described low-pitched sounds coinciding with the peak of the carotid pulse; he called them “pseudo-ejection sounds”. Tucker et al\textsuperscript{5} found 6 patients out of 90 having clicks in different phases of systole. Soon afterwards Sze and Shah\textsuperscript{6} described systolic sounds in 10 of 23 cases. These sounds were of low frequency in 8 and occurred close to the first peak of the carotid pulse.

Of course, these systolic sounds are easily differentiated from the so-called “septal snap”,\textsuperscript{7} which has been described in patients with “paradoxic septal motion” and falls in early diastole.

Our four cases were observed during the routine study of cardiac patients. They were selected because of the observation of a systolic click in the sound tracing while the echo tracing documented an asymmetric and excessive hypertrophy of the interventricular septum. Actually, it is only the graphic study of cardiac vibrations that revealed the clicks because, on auscultation the extremely loud systolic murmur dominated and the click became confused with part of the murmur or was believed to be the first sound.

It is interesting to note that the phase of the click and its relationship to the carotid pulse is different between our four cases.

Their relationship is as follows:

\textit{Case 1:} 90 msec from upstroke of QRS; coincidence between click and upstroke of carotid pulse.

\textit{Case 2:} 220 msec from upstroke of QRS; 100 msec from upstroke of carotid pulse; coincidence with trough between first and second peak of the pulse.

\textit{Case 3:} 180 msec from upstroke of QRS; 60 msec from upstroke of carotid pulse; coincidence with first peak of the pulse.

\textit{Case 4:} 320 msec from upstroke of QRS; 160 msec from upstroke of carotid pulse; coincidence with sudden drop of the pulse at midsystole.

While in the first case the click marked the onset of the murmur, in the others it occurred later, and in one marked the end of the loudest vibrations of the murmur. These clicks included vibrations of low, medium, and high frequency and thus were different from the ejection clicks of valvular aortic stenosis; this fact had been already noted by Snellen\textsuperscript{3} as well as Sze and Shah.\textsuperscript{6}

In regard to the time of occurrence, the click of Case 1 can be called an ejection click and explained with the extreme rapidity of rise of the aortic
pulse; a similar occurrence is found in some patients with syphilitic aortic insufficiency. Those of Cases 2 and 3 coincided with either the first peak or the trough between peaks of the carotid pulse. That of Case 4 occurred when the sudden constriction due to the septal contraction caused a drop of the pulse tracing at midsystole.*

It seems obvious that sudden acceleration (Case 1), swing from acceleration to deceleration or vice versa (Cases 2 and 3), and sudden deceleration (Case 4) were the causes of the clicks. This mechanism, which had been mentioned by De Joseph et al,2) Snellen,3) and Sze and Shah6) (though an alternative possibility was also suggested by the latter) has been accepted by one of us8) as a general explanation of all clicks that can be observed in heart diseases. The alternative view of Sze and Shah (impact of the anterior leaflet of mitral valve against the septum) could be modified as a deceleration of both blood and structures at the cessation of the anterior motion of the mitral; moreover it cannot apply to all clicks. For example our Case 4 has a late click that could not be explained in such manner because the anterior mitral motion had already ended at the time of the click.

From a diagnostic point of view, while the echocardiographic measurement of the thickness of both the septum and the inferior left ventricular wall is of basic importance, it should be emphasized that the phonocardiographic study and the comparison between sound vibrations and pattern of the carotid pulse are also extremely important.

The ease with which these sound and echo studies can be made and repeated is of great value because patients with IHSS may show different abnormalities in different days. Moreover, in several of them, cardiac catheterization may be avoided by the use of these methods.

References

* We are well aware that the carotid pulse has a delay over the phases of opening and closing of the aortic valve. However, the clicks that were observed were within the mentioned phases of rise or fall of the pulse because such phases have a fairly long duration.