Editorial

The Syndrome of Apical Systolic Click, Late Systolic Murmur, and Abnormal T Waves

DEVELOPMENT of new diagnostic techniques often brings to our attention conditions previously described but rarely recognized and little understood. So it is with this syndrome, reported in bits about once every generation from 1887 till the past decade. Then the pieces began to fall into place, and now several articles are appearing every year, each with more information and larger numbers of subjects.

The syndrome is characterized by distinctive auscultatory and electrocardiographic features. Typically in a healthy and asymptomatic young woman one hears at the apex a click near midsystole, followed by a late systolic murmur. The heart and lungs on roentgenograms appear normal; so it is a surprise to find inverted T waves in the left precordial leads, aV\textsubscript{F}, and leads II and III. What does it all mean?

The diagnostic tests which combined to shed light on these findings were selective angiocardioangiography and phonocardiography, both extracardiac and intracardiac, with use of pharmacological agents, such as amyl nitrite and phenylephrine, and the Valsalva maneuver. The intracardiac phonocatheter localized the click and murmur to the left atrium and region of the mitral valve.\(^1\) The pharmacological and physiological maneuvers showed that the murmur behaves as does a murmur of mitral regurgitation.\(^2\)\(^3\) Selective left ventricular cineangiocardioangiography showed ballooning late in systole of the mitral valve (usually the posterior leaflet) into the left atrium, coincident with a regurgitant puff of contrast medium into the atrium. Thus, the diagnosis was established of mild mitral regurgitation in the latter part of systole.\(^2\)\(^4\)\(^5\)

Why are the T waves abnormal? This is still not adequately explained. Early in the 1960's little attention was paid to this change. Abnormally inverted T waves were commented on in some patients and attributed to pericardial disease or myocardial ischemia. But as new reports appeared, each mentioning these same findings, it came to be appreciated that the T-wave findings were part of the syndrome.\(^6\)\(^7\) Unusually prominent U waves have also been noticed.\(^6\)\(^7\)

The reason(s) why the mitral valve should leak in just this way has not been fully clarified either. Malfunction of the chorda-papillary muscle units, deficiency of the striated muscle of the mitral valve, and postsurgical changes of the valve have been invoked. Our experience has focused interest on the left ventricle, because the first girl with this syndrome whom we studied in 1960 by intravenous angiocardioangiography showed partitioning of the left ventricle into two chambers with each systole but not during diastole. Later, selective left ventricular angiocardioangiograms in her and in the other patients (all girls) whom
we have studied demonstrated in each what appears to be an abnormal systolic contraction ring, most marked in the postero-inferior portion.\(^8\),\(^9\) It could be that as the attachment of the papillary muscle is lifted by this contraction into the cavity of the ventricle, the chordae slacken so that the posterior leaflet of the mitral valve balloons and permits some reflux of blood. Tautening then of the chordae does not accomplish apposition of the two leaflets; so regurgitation continues. The click has been shown to coincide with maximal billowing of the valve\(^5\) and has also been attributed to tautening of slack chordae.\(^10\) Perhaps the T-wave changes could be attributed to regional disturbances in coronary blood flow at the site of the unusual contraction. U waves have been considered to represent delayed repolarization of papillary muscles and related structures. The double-peaked systolic contraction in the apex cardiogram, with the nadir corresponding to the click, that Kesteloot and Van Houte\(^2\) noted may be the extracardiac evidence of this unusual left ventricular contraction.

In several series, including our own, patients with Marfan’s syndrome have appeared, for reasons as yet unclear. Most patients, whether children or adults, have continued asymptomatic; only one postmortem study has been reported.\(^11\) This confirms the early opinion that the condition is benign. In contrast to the old notion, however, that these findings are “innocent” or extracardiac is the demonstration that bacterial endocarditis can occur.\(^12\) Prophylaxis at times of predictable risk is indicated.

Further studies will provide a full explanation of the etiology and pathogenesis of this not uncommon syndrome. We may even be able to name it properly. Until then we can list, as in the title of this editorial, its most prominent features, or we might shorten the name slightly to what seems at present to be the least common denominator: “late systolic dysfunction of the mitral apparatus.” In our local parlance, it is just “the click syndrome.”

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References
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