An Interpretation - Mitral Valve Prolapse Syndrome

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Mitral valve prolapse (MVP) syndrome is a mechanical phenomenon in which one or both of the mitral valve leaflets move exaggeratedly during systole, upwards and backwards, surpassing the valvar ring level (plane) [1-4] . This can happen under two circumstances, allowing its classification as either primary or secondary.

In secondary MVP, the leaflets are normal in dimension and structure. However, due to several causes, the left ventricular cavity is smaller, diminished, or the papillary muscles do not contract efficiently.

In the first case diminished left ventricular cavity the leaflets become proportionally larger in relation to the chamber, which allows them to go upwards beyond the valvar level. It may happen, for example, in interatrial communication. The defective myocardial contraction, as a consequence, will not allow the papillary muscles to draw the leaflets adequately, thus the prolapse results. This can happen in dilated myocardiopathy and myocardial ischemia.

In primary prolapse the valves are abnormal. The leaflets are thick, of great dimension and redundant. This redundancy permits exaggerated movement beyond the valvar plan (level) [1,3-5].

The ample movement of the leaflets may create the meso-systolic click (rumble) followed or not by a meso or tele-systolic murmur.

However in patients with primary prolapse, in addition to the mechano-acoustic phenomena, symptoms and signs that do not appear in secondary prolapse may be observed.

These may include asthenia [6]; deformity of the skeleton, such as dorsal kyphoscolisis *orpectus excavatum* or *carinatum*, *straight back* and decrease of posterior- anterior thorax diameter [6,7]; ogival palate [6]; articulate hypermobility and decrease of subcutaneous tissue [8]; hiatal hernia [9] is also frequent. Patients may also experience anxiety or panic symptoms [10-14] and sympathetic-parasympathetic disautonomy [10,15-18]. Patients may have complaints of atypical chest pain and palpitations. The electrocardiogram may show ventricular repolarization alterations, especially in the inferior derivatives (leads) [19]. Patients might have arrhythmias [20,21], stretching of the QT segment and its greater dispersion (spread) [22,23]. Late potentials may exist [24,25]. The literature also reports cases of sudden death [19,26,27].

These clinical manifestations associated with prolapse constitute MVP syndrome.

To date, these elements of MVP syndrome have not had a convincing explanation. It is improbable that only the mechanical phenomena, the exaggerated movement of the leaflets, can explain it [15,23].

Hypotheses have been thought of, for example the exaggerated traction of the leaflets over the papillary muscles could cause ischemia and pain [17]; the increased tension of the leaflet could deform, the atrioventricular groove affecting the circumflex artery and producing ischemia [19]. Nervous terminals in the overly tensioned leaflets could result in arrhythmia [3,17]. These explanations do not seem to me to be satisfactory. Histology and histochemistry of the leaflets of primary prolapse show alteration in collagen constitution (makeup).

An increase occurs in glycosaminoglycan content [12] ; the relation between collagens type I and III is altered [3-8,25-30]. Collagen and elastic fibers are disorganized [3,30,31]. Collagen is looser (lank-limp) than usual.

Myxoma degeneration found in valves of patients who have MVP syndrome [27-32] occurs.

It is common to find concomitant myxoma degeneration of other cardiac valves [15].

The connective tissue of the mitral valve leaflets is not isolated. It is a *continuum* with the collagen of the myocardium [25]. It is continued by the epi-, peri- and endomysium. It is one entity. A whole.

If these alterations ~~of~~ in collagen are found in the valvular leaflets, it is not absurd to think that they can are also be found, in the connective tissues of the myocardium, of which they are an extension.

Studies of ventricular function show asynergy of contraction and segmental irregularity in systole and diastole; dyscinesia [6,33-36]. Histochemical tests in biopsied fractions of the left ventricle have abnormal results that indicate myocardiopathy [37].

Histologic studies have shown myocardial fibrosis and coronary displasia [26]; focal atrophy, disorganization of fibers and adipose deposits [27], myocytic and mitochondrial degeneration or *clumping* [38] .

Today, genes have been identified that are responsible for the synthesis of 18 or 19 types of collagen all with different characteristics and functions. In the heart, types I, III and V prevail [39].

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In the myocardium, besides making up the frame for muscle cells and coronary vases with the struts between isolated cells and muscular bundles, the collagen is part of the contraction of fibers, prevents the sliding of myocytes and guarantees the supply of orderly and efficient strength; struts that connect myocytes with capillaries, tensioned in systole, prevent the folding (sticking together) of the vases and guarantee coronary flow at times of high pressure on the ventricular wall [39].

If this connective tissue becomes slack (limp), due to a genetic defect, it is possible that alterations may occur in the structure and function of the myocardium.

The hypotheses presented so far to explain arrhythmias, the electrocardiographic alterations and even the pain, in patients with MVP syndrome have not been convincing enough for me. Arrhythmias and electrocardiogram-electromechanical results-providing features of the myocardium - do not seem to me to be resulting only from structural alterations, and functions of the myxoma of the mitral valve.

On the other hand, publications that show asynergy of ventricular contraction, alterations in histology and histochemistry of the myocardium associated with mesenchyme alterations of the valve in primary MPV makes one think that the altered collagen of the valves also is found in the myocardium.

Clinical practice throughout the years, has allowed me to find, in the same individual, several symptoms and signs, apparently unconnected, such as bone deformities, astenic habits, disautonomy, anxiety, panic and MPV. This group of signs and symptoms has presented itself with a greater frequency than expected. Could this be due to simple chance?

It is possible that a casual nexus (connection) exists between them. This nexus could be the inadequate gene of the collagen not only in the valve but also in the myocardium, bones and other tissues.

The existence of myocardiopathy, along with MVP, makes it easier to understand arrhythmia, pain, asynergy of contraction and the electrocardiographic alterations found in MVP syndrome [6,27,33,34,37,38,40,41].

MVP could also be just one of the components of a generalized, ample syndrome [15], due to a defect in the development of the collagen. Other components might include cardiac manifestations, that of the skeleton, and possibly, even that of the disautonomy, the anxiety and the panic, resulting from the eventual alteration in the scarce collagen of the central nervous tissue [42,43].

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