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Continuity equation is the echocardiographic method of choice to assess degenerative mitral stenosis

Maria Bonou, Apostolos-Ilias Vouliotis, Konstantinos Lampropoulos, John Lakoumentas, Maria Andriopoulou, Achileas Kotsakis, John Barbetseas

Department of Cardiology, Polyclinic General Hospital of Athens, Greece

Abstract

We present a rare case of a patient with severe, symptomatic degenerative calcific mitral stenosis (MS). Calcification of mitral valve annulus (MVA) is a frequent finding in elderly patients. It can be isolated or associated more often with mitral valve insufficiency than MS. In rare cases, it results in severe MS. An accurate measurement of MVA in degenerative calcific MS is problematic because the limiting orifice is near the annulus and not at the leaflet tips as in rheumatic MS. Continuity equation is the best echocardiographic method to assess the MVA in degenerative MS, correlating well with invasive methods. Real-time three-dimensional echocardiography is a promising tool and provides an accurate measurement of MVA in calcific MS, with a very good correlation compared to continuity equation. On the other hand, the use of pressure half-time is often inaccurate and should be avoided, while two-dimensional planimetry is difficult and not reliable in degenerative MS. The values of mean gradient and systolic pulmonary artery pressure depend on several factors and should be only supportive signs and should not be considered as surrogate markers of the severity of MS. (Cardiol J 2011; 18, 5: 577–580)

Key words: mitral stenosis, echocardiography, Doppler, valvular heart disease

Introduction

Unlike other valve diseases, the predominant cause of mitral stenosis (MS) remains rheumatic fever. The main mechanism is commissural fusion. Other anatomic lesions are chordal shortening and fusion and leaflet thickening and calcification. The calcification seen in rheumatic MS usually involves the commissures and the leaflet tissue, with only late extension to the annulus. Rarely, in only 1% of cases, MS is due to other causes such as congenital, or caused by left atrial tumors, thrombi or vegetations [1, 2].

An unusual but documented cause of severe MS is also heavy calcification of the mitral valve annulus. Rheumatic MS differs markedly from degenerative calcific MS, in which the main lesion is annular calcification resulting in limiting anatomic orifice area. This entity, common in elderly women, is a frequent cause of non-significant mitral regurgitation (MR) and is associated with coronary atherosclerosis and cerebrovascular embolic events, while severe, symptomatic valve stenosis due to this cause is extremely rare [1–4].

We present the case of a woman with severe symptomatic degenerative calcific MS and we discuss the echocardiographic criteria used in order to quantify the severity of stenosis.

Address for correspondence: Konstantinos Lampropoulos, MD, PhD, FESC, Department of Cardiology, Polyclinic General Hospital of Athens, Greece, 31, L. Porfyra str., 16673, Athens, Greece, tel: +302 108 950 863, fax: +302 108 950 863, e-mail: konlampropoulos@yahoo.gr

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Case report

A 72 year-old Caucasian woman was admitted to our hospital with symptoms of gradually increasing shortness of breath, primarily on exertion, during the last six months. She also claimed some episodes of paroxysmal nocturnal dyspnea without the pain associated with dyspnea.

The physical examination revealed a soft diastolic murmur localized at the apex. Resting electrocardiogram showed sinus rhythm with a heart rate of 80 bpm. Chest X-ray revealed mild cardiomegaly with left atrial enlargement.

Transthoracic echocardiography demonstrated normal left ventricular size and function, enlarged left atrium and normal right cavities. From the mild tricuspid regurgitation, the pulmonary arterial systolic pressure was estimated at 55–60 mm Hg. Heavy calcification was obvious in the mitral valve orifice at both the anterior and posterior aspect of the annulus. The tips were relatively free and the leaflets opened without doming restriction (Fig. 1).

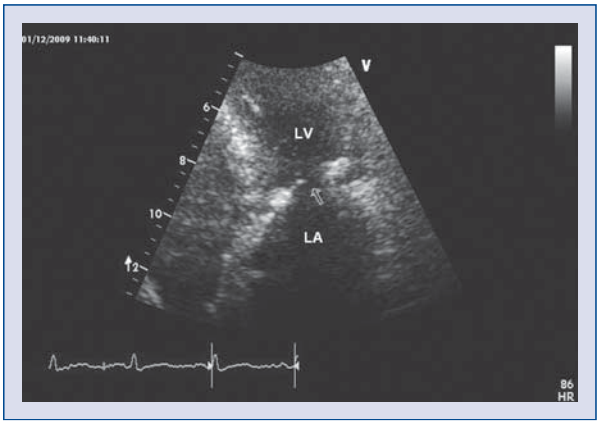

Figure 1. Two-dimensional echocardiogram. Apical four-chamber view showing the heavily calcified mitral annulus with free leaflet tips (arrow); LA — left atrium; LV — left ventricle.

Planimetry measurement of the mitral orifice using two-dimensional (2D) echocardiography was not feasible because of the poor acoustic window and the existence of severe annular calcification. Continuous-wave Doppler of the transmitral flow is depicted in Figure 2. The mean mitral valve gradient was 13 mm Hg, consistent with severe MS.

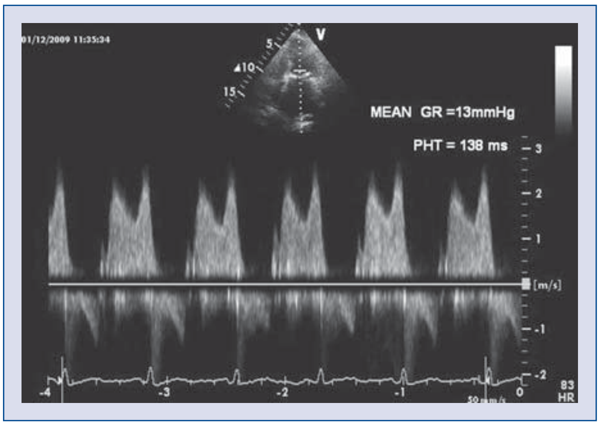

Figure 2. Transmitral Doppler derived mean gradient (MEAN GR) and pressure-half time (PHT).

Pressure half-time (PHT) derived mitral valve area (MVA) was calculated using the following formula: MVA = 220/PHT. PHT is obtained by tracing the deceleration slope of the E-wave on Doppler spectral display of transmitral flow. In our case, PHT was 138 ms (Fig. 2). Thus, MVA = 220/138 = 1.6 cm^2, suggesting mild mitral stenosis.

In this particular case of degenerative MS, we also estimated the MVA using the continuity equation: MVA = $\pi \times (D2/4) \times$ (VTI aortic/VTI mitral), where D = 2 cm was the diameter of the left ventricular outflow track (LVOT) and VTI (velocity time interval) aortic = 15 cm and VTI mitral = 52 cm. VTI aortic was measured from the LVOT velocities recorded by pulsed Doppler and VTI mitral from the MS jet velocities by continuous-wave Doppler. Thus, MVA = $3.14 \times (2^2/4) \times (15/52) = 0.91$ cm^2, suggesting severe stenosis.

Discussion

Mitral annulus calcification is a degenerative calcific process of the mitral valve support ring. It is a common finding in the elderly, especially in women, often with coexisting hypertension, coronary artery disease and diabetes mellitus [2, 4]. It essentially involves the posterior annulus, with minimal extension to leaflets and chordae. This entity can be isolated causing few or no hemodynamic consequences, or associated with MR rather than MS. Generally it is not associated with clinically important flow obstruction [3, 5]. Akram et al. [6] found that out of 70 patients reporting as having MS, 18.5% had calcific mild to moderate MS rather than rheumatic MS.

Nevertheless, severe degenerative MS is extremely rare and occurs when calcification extends to the basis of mitral leaflets resulting in narrowing of the diastolic flow area, without tips restriction unlike rheumatic MS [5, 6]. Reduction of normal mitral annular dilatation during diastole and the addition of impaired anterior mitral leaflet mobility appeared to be possible mechanisms for the significant functional stenosis caused by calcification of mitral valve annulus [5, 7]. It is more frequently encountered in patients with advanced renal disease requiring dialysis [8].

An accurate measurement of MVA in degenerative calcific MS is problematic. In our case, there was a discrepancy in the measurements of the MVA using echocardiographic methods.

Transvalvular mean gradient is not the best marker of the severity of MS since it is related to the MVA but it is also influenced by a number of other factors, the most important being heart rate, cardiac output and associated MR [3]. The values of mean gradient and systolic pulmonary artery pressure should be only supportive signs and cannot be considered as surrogate markers of the severity of MS.

Accurate measurement of MVA is essential in the evaluation of patients with MS. The gold standard method has been invasive evaluation using the Gorlin equation to derive a calculated valve area [3]. However, cardiac catheterization is an invasive method, it has several limitations and should be restricted to the rare cases where echocardiography is inconclusive or discordant with clinical findings.

In clinical practice, 2D and Doppler echocardiography has been the usual method to determine indirectly the MVA by PHT or by direct planimetry. The PHT derived MVA is limited and often inaccurate in patients with calcific annular MS because there are frequently coexisting conditions, such as hypertension and diabetes mellitus, that impair left ventricular compliance and thus alter left atrial to left ventricular pressure difference [9]. Therefore the PHT would be shorter as a result of a more rapid equilibration of left atrial and ventricular pressure, resulting in an overestimation of the MVA, which was our experience in this case. On the other hand, not all prolonged PHTs indicate MS. Patients with abnormal myocardial relaxation have a prolonged PHT without valve stenosis. Hence, according to the EAE/ASE recommendations for echocardiographic assessment of valve stenosis, the use of PHT in degenerative calcific MS may be unreliable and should be avoided [3].

Planimetry using 2D is considered as the reference measurement of MVA. It is a direct and relative hemodynamic-independent method without the need for geometric or mathematical assumptions [3]. However, planimetry and PHT methods have a better correlation with anatomical valve area in younger patients with rheumatic MS than in calcific annular MS [6]. In the case of degenerative MS, planimetry is difficult and mostly not reliable. This may be related to both the presence of heavy calcification and the fact that the limiting orifice is near the annulus and does not present an area for planimetry at the leaflet tips as in rheumatic MS [3, 10]. The PHT derived MVA has limitations in patients with calcific annular MS in whom there are coexisting conditions, such as hypertension and diabetes mellitus, that impair left ventricular compliance and thus alter left atrial to left ventricular pressure difference. Therefore, the PHT would be shorter as a result of a more rapid equilibration of left atrial and ventricular pressure, resulting in an overestimation of the MVA, which was our experience in this case.

The continuity equation method is an independent standard for measuring the effective MVA in degenerative MS, correlating well with invasive methods [3, 10]. On the other hand, it is technically demanding and requires multiple measurements, increasing the impact of errors. This is why it is not recommended for routine use in MS but may be useful in certain patients, as in our case, when standard measurements are inconclusive [3].

Recent reports demonstrate that real-time 3D echocardiography provides an accurate measurement of MVA in both rheumatic and degenerative MS [10, 11]. Chu et al. [10], used 3D color-planimetry to measure MVA in calcific MS and compared this method to PHT derived MVA, considering continuity equation as an independent standard. Real-time 3D echocardiography had a greater correlation and agreement to MVA by continuity equation than MVA by PHT. This reflects the tubular valve geometry of mitral valve orifice in these patients [10]. 2D planimetry has limitations, particularly in regard to positioning the correct image plane orientation. This is even more pronounced in degenerative calcific MS because the limiting orifice is near the annulus and not at the leaflet tips as in rheumatic MS. In addition 2D planimetry is limited to those patients in whom parasternal images of high quality can be obtained [10, 11]. 3D echocardiography overcomes these limitations, enables appropriate orientation and provides the optimal plane of the smallest mitral valve orifice. In addition, 3D planimetry is not limited to the parasternal view and allows MVA measurements from an apical view [10, 11].

Other echocardiographic findings in degenerative MS, as reported by Akram et al. [6], are higher mitral annulus calcification severity score, lower frequency of significant MR, milder degree of MS, higher frequency of concomitant aortic stenosis and higher left ventricular mass compared to rheumatic MS. The last finding may reflect the high prevalence of hypertension in this group of patients.

Replacement of the mitral valve in the presence of extensive annular calcification can be extremely difficult and challenging. Calcification interferes with suture placement and prevents proper insertion of a prosthesis, increasing the possibility of paravalvular leakage and valvular dehiscence. Aggressive debridement of a calcified annulus may lead to atrioventricular groove rupture, injury of the circumflex coronary artery or thromboembolic events [12, 13]. Several surgical techniques have been proposed to deal with MS due to extensive calcification of mitral annulus. In patients with heavy circumferential annular calcification, these approaches can often be unfeasible. In these cases, mitral prosthesis can be implanted at the intra-atrial level [13].

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