

Arrhythmic mitral valve prolapse

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Summary

Arrhythmic mitral valve prolapse is a rare problem of a usually benign condition of the mitral valve. Mitral annular disjunction and the pickelhaube sign are important features of myxomatous mitral valve prolapse, which are linked to ventricular arrhythmias or even sudden cardiac death. The prolapsing mitral valve can cause structural changes (fibrosis) in the papillary muscles and the posterolateral myocardial wall, which are a possible substrate for arrhythmias. Therefore it is important to look for these signs in patients with mitral valve prolapse.

Keywords: mitral valve prolapse, mitral annular disjunction, pickelhaube sign, sudden cardiac death

About 1–3% of the general population are affected by mitral valve prolapse. It is defined as a >2 mm superior displacement of the mitral valve leaflet(s) during systole with or without mitral regurgitation. Generally it is regarded as a benign condition, but the outcome is widely heterogeneous. Since the 1980s several complications have been reported, such as progressive mitral regurgitation requiring mitral valve surgery, atrial fibrillation, strokes in sinus rhythm, congestive heart failure, infective endocarditis, ventricular arrhythmias and even sudden cardiac death [1, 2]. The association between sudden cardiac death and mitral valve prolapse has been discussed in several publications and is being increasingly recognised.

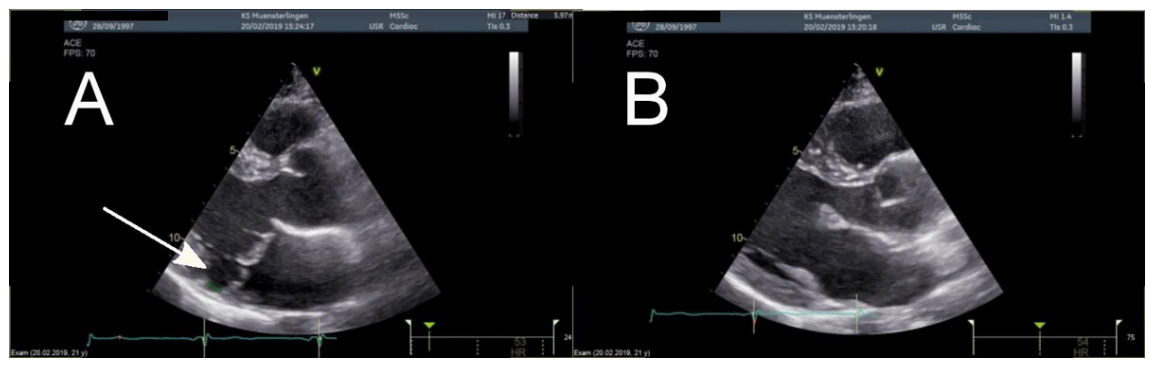
But what are the clues for the diagnosis of the arrhythmic mitral valve prolapse? Basso et al. [3] demonstrated for the first time the magnetic resonance imaging and histologi-

cal evidence of a possible arrhythmic substrate in sudden cardiac death patients: fibrosis of the papillary muscle and the inferobasal left ventricular wall suggesting myocardial stretch and friction by the prolapsing mitral leaflet. Bileaflet prolapse and myxomatous mitral valve disease (MMVD) are more responsible for producing myocardial stretch.

There are two important echocardiographic signs for MMVD and the risk for malignant ventricular arrhythmias:

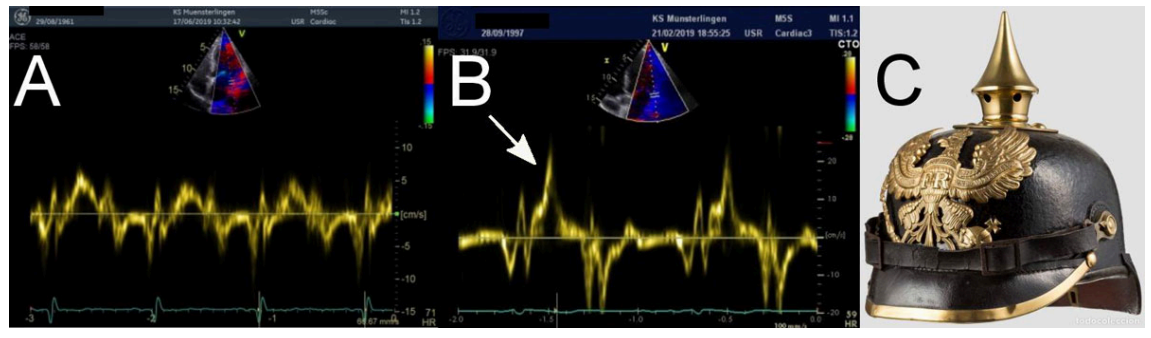
- Firstly, mitral annular disjunction, which is defined as a detachment of the roots of the annulus from the ventricular myocardium (fig. 1), producing a gap of 2–10 mm during systole. It is typically under the P1 and P2 scallop, not seen in the surgeon's view from the left atrium and positively correlated with the burden of ventricular premature beats (as a result of the amount of stretch?). Mitral annular disjunction is seen in patients with more prolapsing segments, more bileaflet disease and greater volume of prolapse, which are all components of diffuse myxomatous disease. In this way mitral annular disjunction is specific for MMVD [4].
- Secondly, the Pickelhaube sign, which is defined as a high-velocity mid-systolic spike in the tissue Doppler velocity profile of the lateral mitral valve annulus (fig. 2). This phenomenon appears when the prolapsing posterior leaflet stretches the posteromedial papillary muscle, causing a sharp traction on the adjacent posterobasal left ventricular wall. Usually the velocity surpasses 16 cm/s [5].

Figure 1: Mitral annular disjunction (MAD). Transthoracic echocardiography parasternal long axis view demonstrating mitral annular disjunction in a case with bileaflet mitral valve prolapse in systole (A). The gap between the mitral annulus and the left ventricular muscle wall is 6 mm (arrow). During diastole the MAD is not seen. (B) A frame-by-frame analysis is very useful to visualise the gap.



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Figure 2: Pickelhaube sign. (A) Normal tissue Doppler velocity profile of the lateral mitral valve annulus. (B) The pickelhaube sign is characterised by a very high, spiked, velocity profile of the lateral mitral valve annulus usually exceeding 16 cm/s, in this example >20 cm/s (arrow). (C) The pickelhaube was a Prussian military or police helmet of the 19th century with a spike.



Mitral annular disjunction and the pickelhaube sign are thought to be noninvasive echocardiographic markers associated with sudden cardiac death in myxomatous mitral valve prolapse. Furthermore, these phenomena are part of a recently proposed risk stratification for sudden cardiac death in MMVP [6]. Many questions regarding the evaluation, monitoring and therapy of these patients are open and will be discussed in the future.

In conclusion, arrhythmic mitral valve prolapse is a rare but difficult problem of a quite common and mostly benign condition of the mitral valve. Mitral annular disjunction and the pickelhaube sign are two important features of the myxomatous mitral valve disease. Therefore, it is probably important to look for these signs linked to unexplained sudden cardiac death of young patients with mitral valve prolapse.

Disclosure statement

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