Arrhythmic Mitral Valve Prolapse

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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 heterogenous. Since the 1980s several complications are reported, for example progressing mitral regurgitation requiring mitral valve surgery, atrial fibrillation, stroke's in sinus rhythm, congestive heart failure, infective endocarditis, ventricular arrhythmias and even sudden cardiac death [1, 2]. The association between sudden cardiac death (SCD) and mitral valve prolapse (MVP) has been discussed in several publications and is of increasing awareness.

But what are the clues for the diagnosis of the arrhythmic mitral valve prolaps? Basso et al. [3] demonstrated for the first time the MRI and histologic evidence of a possible arrhythmic substrate in SCD 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 producing myocardial stretch. There are two important echocardiographic signs for the myxomatous mitral valve disease and the risk for malignant ventricular arrhythmias [3].

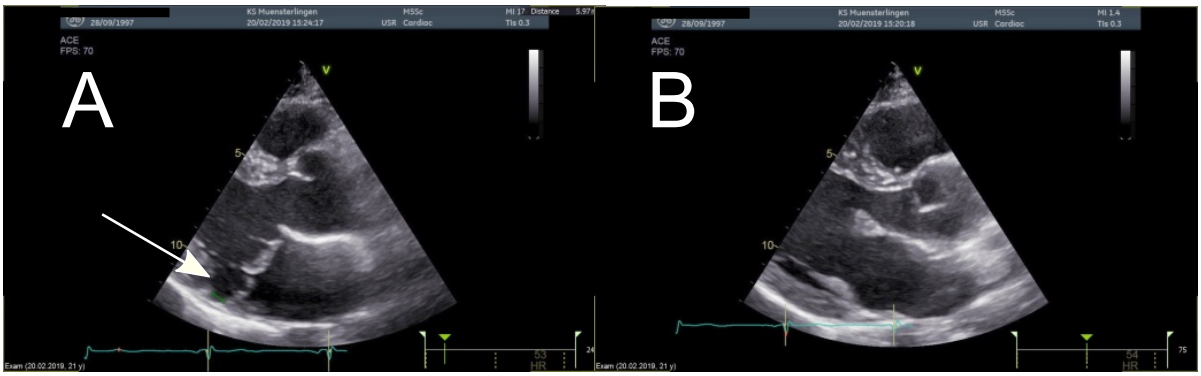


Figure 1: **Mitral Annular Disjunction (MAD)**
Transthoracic echocardiography parasternal long axis view demonstrating mitral annular disjunction in a case with bileaflet mitral valve prolapse in systole **(A)**. The gap between the mitral annulus and the left ventricular muscle wall is 6 mm (arrow).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

Firstly, the mitral annular disjunction (MAD) is defined as a detachement of the roots of the annulus from the ventricular myocardium (Figure 1), producing a gap of 2-10 mm during systole. It is typically under the P1 and P2 scallop, not seen by the surgeons view from the left atrium and positive correlated to the burden of VPB's (as a result of the amount of stretch?). MAD is seen in patients with more segments prolapsing, more bileaflet disease and greater volume of prolaps, which are all components of diffuse myxomatous disease. In this way MAD is specific for MMVD [4].

Secondly, the Pickelhaube Sign is defined as a high-velocity mid-systolic spike in tissue Doppler velocity profile of the lateral mitral valve annulus (Figure 2). This phenomenon appears when the prolapsing posterior leaflet stretches the posteromedial papillary muscle causing a sharp traction of the adjacent posterobasal left ventricular wall. Usually the velocity surpasses 16 cm/s [5].

The Mitral Annular Disjunction and the Pickelhaube sign are thought to be non-invasive, echocardiographic markers associated with sudden cardiac death of myxomatous mitral valve prolapse. Furthermore these phenomenons are part of risk stratification recently proposed [6] for SCD in MMVP. Many questions regarding the evaluation, monitoring and therapy of these patients are open and will be discussed in the future.

In conclusion, the arrhythmic mitral valve prolapse is a rare but difficult problem of a quite common and mostly benign condition of the mitral valve. The 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.

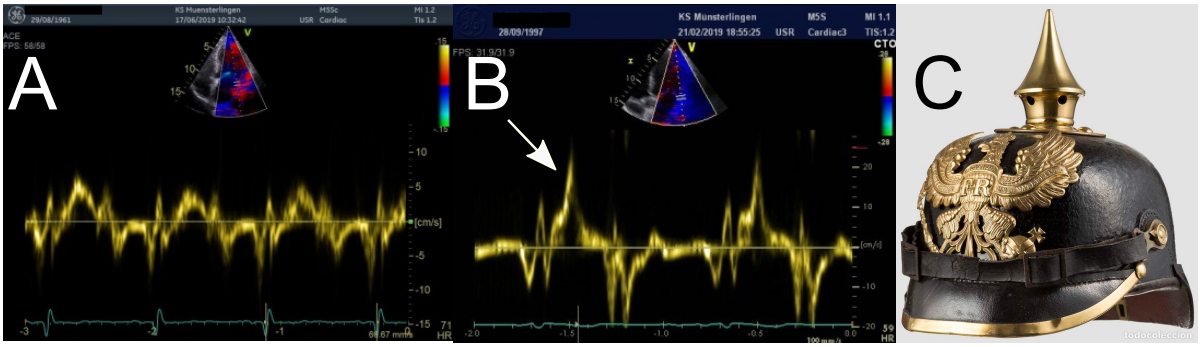


Figure 2: **Pickelhaube sign**
(A) Normal tissue doppler velocity profile of the lateral mitral valve annulus. **(B)** The Pickelhaube Sign is characterized by a very high, spicked, velocity profile of the lateral mitral valve annulus usually exceeding 16 cm/s. In this example >20 cm/s (arrow). **(C)** The Pickelhaube was a Prussian military or police hut of the 19th century with a peak.

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