JACC: CARDIOVASCULAR IMAGING, VOL. 10, NO. 9, 2017
SEPTEMBER 2017:1076–84

**Joseph A. Ladapo, MD, PhD***
**John M. Pfeifer, MD**
**Alana A. Choy-Shan, MD**
**James M. Pitcavage, MSPH**
**Brent A. Williams, PhD**

*David Geffen School of Medicine at UCLA
Department of Medicine
Division of General Internal Medicine and Health Services Research
911 Broxton Avenue, 1st Floor
Los Angeles, California 90024
E-mail: jladapo@mednet.ucla.edu

http://dx.doi.org/10.1016/j.jcmg.2016.09.015

Please note: This research was funded by the National Heart, Lung, and Blood Institute (K23 HL116787; J.A. Ladapo, PI). The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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## The Pickelhaube Sign

### Novel Echocardiographic Risk Marker for Malignant Mitral Valve Prolapse Syndrome

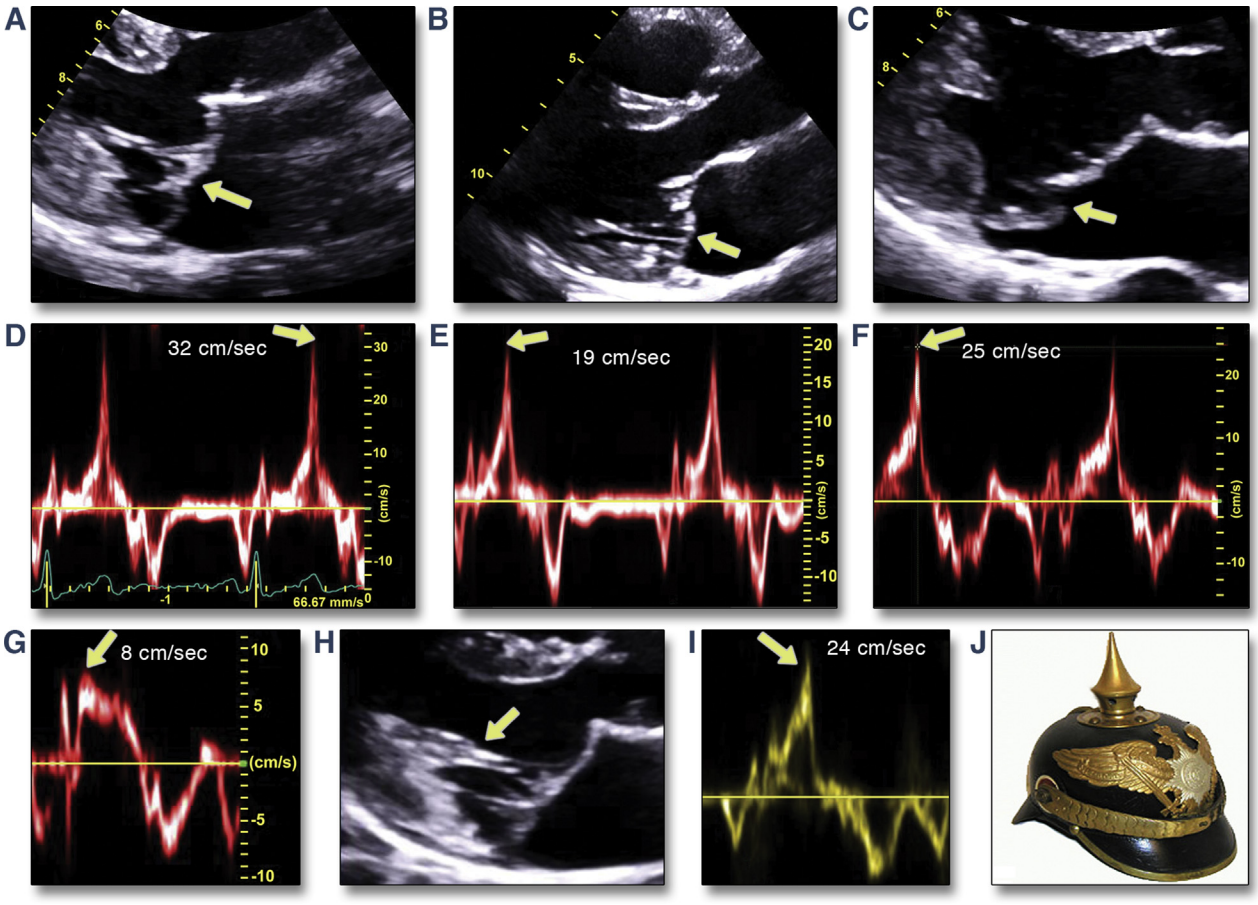
Natural history of mitral valve prolapse (MVP) in the community is widely heterogeneous (1). Prognosis in MVP ranges from normal life expectancy to subsets with high morbidity/excess mortality, including sudden cardiac death (2). Although MVP is identified in a significant number of patients with sudden cardiac death, especially young women (3), and clinical and echocardiographic markers for adverse outcomes have been described (1-3), debate about MVP leading to sudden cardiac death survives in contemporary cardiology. To this end, we report our observation of a novel echocardiographic marker of high risk in patients with myxomatous mitral valve disease (MMVD) and bileaflet MVP (BMVP).

Of the 278 patients at our institution who fulfilled myxomatous BMVP published criteria (3), 21 were referred to our specialized valve clinic for further characterization (quantification) of the severity of mitral regurgitation (MR) and further management options. These referrals were generated by the primary cardiologists who had initially evaluated the patient. All underwent careful interrogation of lateral and medial annuli by pulsed-wave tissue Doppler from the apical window. Extent of mitral prolapse, leaflet thickness, medial and lateral wall Doppler velocity, MR severity, left ventricular ejection fraction, left atrial size, and 12-lead electrocardiograms were analyzed. Cardiac magnetic resonance (CMR) imaging studies were reviewed with particular focus on delayed gadolinium enhancement of posterobasal myocardium and of the papillary muscles.

Patients were divided into 2 groups based on tissue Doppler: those with a spiked systolic high-velocity signal ≥16 cm/s (Group 1) and those <16 cm/s (Group 2). Among 21 patients with MMVD and BMVP (median age, 51.6 ± 12.3 years; 71% female), 12 patients were in Group 1 (47.4 ± 9.5 years; 75% female) and 9 in Group 2 (57.1 ± 14.0 years; 67% female). Group 1 patients with distinctive spiked signal (≥16 cm/s) had 8 malignant (combined ventricular tachycardia/ventricular fibrillation) events compared with 2 malignant events in Group 2 patients (67% vs. 22%; $p < 0.08$ [Fisher exact test]). Of the 12 patients in Group 1, a total of 5 patients had an implantable cardioverter defibrillator implanted versus none in Group 2. Delayed gadolinium enhancement was noted only in Group 1 (2 of 6; 33%), out of the 11 patients who underwent CMR. Inverted or biphasic T waves in the inferior leads were observed in 6 patients in Group 1 and 2 patients in Group 2 (p = NS). There was no difference between the 2 groups in severity of prolapse, leaflet thickness, medial annulus Doppler velocity, MR severity, or left ventricular ejection fraction. This observation of high-velocity systolic signal with tissue Doppler imaging resembling the "Pickelhaube," a spiked helmet, was thus an indicator of a malignant phenotype of MMVD and BMVP (**Figure 1**).

We hypothesize that the tugging of the posteromedial papillary muscle in mid-systole by the myxomatous prolapsing leaflets causes the adjacent posterobasal left ventricular wall to be pulled sharply toward the apex, resulting in the observed spiked configuration of the lateral annular velocities (Pickelhaube sign). It has been suggested that this mechanical traction of the papillary muscles and posterolateral left ventricular wall is arrhythmogenic with early electrical dysfunction being recognized during electrophysiological studies even in the absence of gadolinium enhancement on CMR (4). Endocardial friction lesions in the inferolateral mural endocardium are additional mechanisms for ventricular arrhythmias in MMVD and MVP (2,4). Recent CMR studies (5) have shown delayed gadolinium enhancement in these areas, suggesting fibrosis. Our novel finding adds to the emerging risk markers of arrhythmogenic MVP syndrome (2-5), suggesting the

**FIGURE 1** Pickelhaube Sign




Transthoracic echocardiography demonstrating myxomatous bileaflet mitral valve prolapse **(arrows)** in cases 1 **(A)**, 2 **(B)**, and 3 **(C)**. High-velocity mid-systolic spike (lateral annulus, 32 cm/s) in cases 1 **(D)**, 2 (mid-systole, 19 cm/s) **(E)**, and 3 (late systole, 25 cm/s) **(F)**. **(G)** Normal medial annulus systolic velocity, case 1. **(H)** Tugging of the posteromedial papillary muscle by prolapsing leaflets **(arrow)**, case 4. **(I)** Late-peaking systolic tissue velocity spike of 24 cm/s, case 4. **(J)** Pickelhaube, spiked German military helmet (reprinted with permission from the collection of Peter Suciu).

possibility that this spiked tissue Doppler velocity profile may be a risk marker for malignant arrhythmias in patients with MMVD and BMVP.

Future research with a large cohort of patients will help elucidate the clinical implications of this observation.

**Lakshmi Muthukumar, MD**
**Faraaz Rahman, MD**
**M. Fuad Jan, MBBS, MD**
**Armaan Shaikh, DO**
**Lindsey Kalvin, RCDS**
**Anwer Dhala, MD**
**Arshad Jahangir, MD**
**A. Jamil Tajik, MD***

*Aurora Cardiovascular Services
Aurora St. Luke's Medical Center
2801 West Kinnickinnic River Parkway
Suite 840
Milwaukee, Wisconsin 53215
E-mail: publishing14@aurora.org

http://dx.doi.org/10.1016/j.jcmg.2016.09.016

Please note: The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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## Employment Status After Training in Advanced CV Imaging

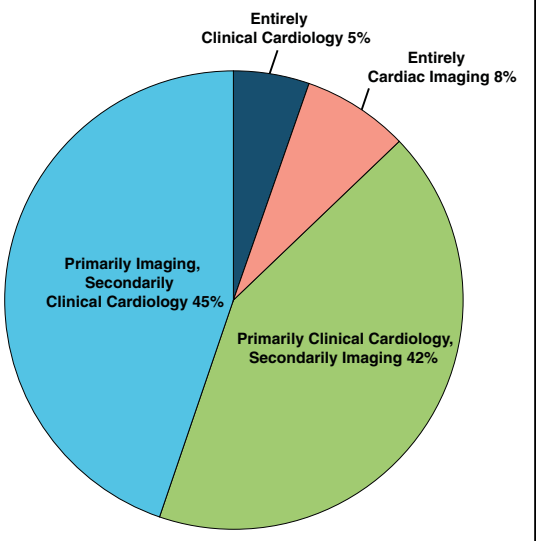
Advances in cardiovascular imaging have led to growth in the number of training programs offering advanced cardiovascular imaging (ACVI). These programs offer additional training beyond the general cardiovascular disease fellowship. As for Accreditation Council for Graduate Medical Education nonaccredited programs, there is a lack of uniformity in these training programs (1). Current documents detail the skills needed for independent proficiency (Level II training), but they largely leave advanced proficiency training requirements (Level III training) to future documents (2). This leaves program directors (PDs) with significant flexibility in developing an ACVI fellowship curriculum.

One important issue for trainees and PDs relates to the professional opportunities available to graduates of ACVI programs. Knowledge of market opportunities may help programs and applicants understand how to better craft the training experience. To that end, we aimed to characterize the state of the post-ACVI-training job environment. We created an online survey designed to identify the professional opportunities available to graduates of ACVI training programs in the U.S. The anonymous survey was administered in February of 2016 to 51 ACVI training programs listed in the American College of Cardiology database of ACVI programs (3). This survey was initiated with cooperation of the Cardiovascular Training Section. Programs were asked to answer the questions presented based on the most recent 5 years of graduates, or in the case of more recent programs, for the duration of the existence of the program.

Nineteen programs responded to the survey (37% survey response rate). These programs reported 180 graduates over the specified time period (mean 9.5 graduates per program; interquartile range: 5 to 11). Positions accepted after graduation included academic (51%), private sector (41%), further training (4%), nonclinical research (3%), and industry positions (1%). Most positions offered a mixture of clinical cardiology and imaging (45% primarily imaging and 42% primarily clinical cardiology). A minority of position were entirely imaging (8%) or clinical cardiology (5%) (Figure 1). Expertise in echocardiography was required in most positions (80%), whereas cardiac magnetic resonance, cardiac computed tomography, and nuclear cardiology expertise was required by about one-half of the positions (53%, 49%, and 45%, respectively). Programs were asked to describe the difficulty for graduates to find employment in a position that "encompassed the [imaging] elements in which the fellow had trained." As assessed by the PDs, 16% of graduates found it "very easy" to find such a position, 32% found it "somewhat easy," 32% found it "neither difficult nor easy," 21% found it "somewhat difficult," and none found it "very difficult."

Acknowledging the limitations of survey-based investigation and the response rate of our study, and the fact that this survey represents only the perspective of PDs as opposed to graduates, whose insight and experiences are critical to a more comprehensive understanding of this issue, these results offer initial evidence as to the job market for graduates of ACVI programs. There is significant heterogeneity in positions available after graduation. There is a near even split in academic versus nonacademic positions, and in positions that are defined

**FIGURE 1 Employment Type on Graduation**




Makeup of positions taken by graduates of advanced cardiovascular imaging fellowships.