REFERENCES

1. Denny P. The varicella-zoster paradox: what it means for eye M.D.s. EyeNet 2012 Apr. [Accessed 3 Dec. 2014]. Available from http://www.aao.org/publications/eyenet/201204/comprehensive.cfm.
2. Federal Aviation Administration. Guide for aviation medical examiners. Washington (DC): Federal Aviation Administration; 2014. [Accessed 3 Dec. 2014]. Available from http://www.faa.gov/about/office_org/headquarters_offices/avs/offices/aam/ame/guide/.
3. Forbes HJ, Bhaskaran K, Thomas SL, Smeeth L, Clayton T, Langan SM. Quantification of risk factors for herpes zoster: population based case-control study. BMJ. 2014; 348:g2911.
4. Hales C. Herpes zoster rates are increasing, but why? 2014. [Accessed 3 Dec. 2014]. Available from http://www.medscape.com/viewarticle/822982.
5. Lee KG, Cheng MO. Varicella-zoster infection with secondary bacteremia and extensive facial abscesses. Med J Malaysia. 2012; 67(5):529.
6. Naval Aerospace Medical Institute. U.S. Navy aeromedical reference and waiver guide. Pensacola (FL): Naval Aerospace Medical Institute; 2014. [Accessed 3 Dec. 2014]. Available from http://www.med.navy.mil/sites/nmotc/nami/arwg/Pages/AeromedicalReferenceandWaiverGuide.aspx.
7. U.S. Air Force. Air Force waiver guide. Wright-Patterson AFB (OH): U.S. Air Force School of Aerospace Medicine; 2014. [Accessed 3 Dec. 2014]. Available from http://www.wpafb.af.mil/afrl/711hpw/usafsam.asp.
8. Army US. Standards of medical fitness. Washington (DC): Department of the Army; 2011. Army Regulation 40-501. [Accessed 3 Dec. 2014]. Available from http://armypubs.army.mil/epubs/pdf/r40_501.pdf.
9. Varicella and herpes zoster vaccines: WHO position paper, June 2014. Wkly Epidemiol Rec. 2014; 89(25):265–287.

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You are the flight surgeon on call for the day at a joint undergraduate pilot training base and also a squadron medical element for one of the flying squadrons. During your quiet lunch time the airman at the front desk rushes in to inform you he has one of your pilot students with complaint of chest pain and shortness of breath. Upon initial examination, you find a pale and sweating 24-yr-old athletic-built man, alert and oriented, slightly out of breath but calm. He and his wife were practicing some P90× routines at the base gym minutes ago when he developed some palpitations and then chest tightness. Past medical history is significant only for flying waiver granted for premature ventricular contractions (PVCs) and rare atrial ectopy. An echocardiogram 2 yr ago showed normal ejection fraction (EF) and cardiac chamber sizes. He normally has asymptomatic bradycardia. The patient has taken no medications or supplements. He has no medication allergies or environmental allergies. The ensuing examination reveals heart rate of 235, blood pressure 149/100, temperature 97.8°F. Electrocardiogram (EKG) shows supra ventricular tachycardia. Thankfully, your staff had the foresight to call Emergency Medical Services when the patient first presented with complaint of chest pain, as your clinic is small and a good 45-min drive from a cardiac center. During his transfer to the nearest and trusted civilian cardiac center in town, the patient develops ventricular tachycardia (VT) and a lidocaine infusion is started during the ambulance transport. That same evening, when you call the cardiac center to follow up, you discover that upon arrival your patient degenerated into ventricular fibrillation and required immediate defibrillation. He is for the moment stable in the cardiac intensive care unit.

1. The evening of the incident, your squadron commander is concerned and wants information. What is your best next step?

A. Ask the civilian cardiac center to fax updated information.
B. Call the patient's wife to express concern and to find out more about what happened.
C. Go to the hospital and visit the patient and meet his treating physicians.
D. Assure your squadron commander that all will be fine as the patient is stable.

ANSWER/DISCUSSION

1. C. The best way to find information from a civilian hospital, in a case like this, is to go there and talk to the patient's treating physicians. The patient and his family would be highly appreciative of a visit from the squadron physician. In addition, you can introduce yourself as the primary care manager to the hospitalists/specialists, who would likely prefer to communicate with the primary care manager to ensure the patient will have seamless care and follow-up.

After three consecutive visits at the civilian hospital, you finally piece together the results of diagnostic tests: a cardiac lab work-up including creatine phosphokinase and troponin is normal, a coronary angiogram is normal, a transthoracic echocardiogram shows a dyskinetic right ventricle with overall EF of 35%, with enlarged (34-mm) parasternal long-axis (PLAX) view right ventricular (RV) outflow tract corrected for body size. Mid-RV diameter is in normal range. Cardiovascular magnetic resonance (CMR) imaging reveals dyssynchronous RV contraction, increased RV end-diastolic volume, and depressed RV EF of 34%. During electrophysiological (EP) testing when VT is induced, patient's EKG shows multiple QRS morphologies.

2. What is your diagnosis?

A. Right ventricular outflow tract (RVOT) tachycardia.
B. Dilated cardiomyopathy (DCM) with predominant RV dysfunction.
C. Myocarditis.
D. Arrhythmogenic right ventricular cardiomyopathy (ARVC) or arrythmogenic right ventricular dysplasia.

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ANSWER/DISCUSSION

2. D. RVOT tachycardia is an idiopathic type of VT with repetitive monomorphic VT.[9] RVOT tachycardia typically has a much more benign prognosis than ARVC and can often be treated with radiofrequency ablation.[9] In EP, programmed premature stimulation induced VT happens in a majority of ARVC patients versus a minority of patients with RVOT tachycardia.[14] The induction of VTs, with different QRS morphologies, is characteristic of ARVC.[14] The patient did not have marked and progressive left ventricular (LV) dilatation, both features rare in ARVC.[17] Myocarditis usually has cardiac biomarker elevations and ultrasound would likely also show LV dilation.[2] The patient did not have a marked (> 33-mm) RV dilatation, which is common in dilated cardiomyopathy with predominant RV dysfunction.[4,7] It is also rare to have sudden cardiac arrest/death (SCD) as the initial symptom of dilated cardiomyopathy.[1]

The diagnosis of ARVC can be difficult. The clinician must be aware of such a differential diagnosis and willing to order multiple diagnostic tests, procedures, or referrals to support the diagnosis. The diagnosis of ARVC should be considered for a young survivor of SCD, particularly SCD occurring during exercise. The patient's tests have satisfied two major criteria for definitive diagnosis of ARVC using the 2010 Task Force criteria: two-dimensional (2D) echo showing regional RV dyskinesia with PLAX RVOT enlargement and CMR showing dyssynchronous RV contraction and RV EF $\leq$ 40%.[10,15]

For ARVC, definite diagnosis requires two major, or one major and two minor, or four minor criteria from different categories (global or regional dysfunction and structural alterations, tissue characterization of wall, repolarization abnormalities, arrhythmias, family history).[10]

Under the global or regional dysfunction and structural alterations category that is applicable in this case, the criteria are largely based on either 2D echo or CMR.[10] For major criteria, 2D echo should show regional RV akinesia, dyskinesia, or aneurysm and one of the following (end diastole): 1) PLAX view RVOT $\geq$ 32 mm (corrected for body size); 2) parasternal short-axis view RVOT $\geq$ 36 mm (corrected for body size); or 3) fractional area change $\leq$ 33%. CMR should show regional RV akinesia or dyskinesia or dyssynchronous RV contraction and one of the following: 1) ratio of RV end-diastolic volume to body surface area $\geq$ 110 ml/m^2 (male) or $\geq$ 100 ml/m^2 (female); or 2) RV EF $\leq$ 40%. Additionally, a major criterion is fulfilled if RV angiography shows regional RV akinesia, dyskinesia, or aneurysm.

For minor criteria, 2D echo should show regional RV akinesia or dyskinesia and one of the following (end diastole): 1) PLAX RVOT $\geq$ 29 to < 32 mm (corrected for body size); 2) parasternal short-axis RVOT $\geq$ 32 to < 36 mm (corrected for body size); or 3) fractional area change > 33% to $\leq$ 40%. CMR should show regional RV akinesia or dyskinesia or dyssynchronous RV contraction and one of the following: 1) ratio of RV end-diastolic volume to body surface area $\geq$ 100 to < 110 ml/m^2 (male) or $\geq$ 90 to < 100 ml/m^2 (female); or 2) RV EF > 40% to $\leq$ 45%.

The patient's EKG (**Fig. 1**) 2 yr prior to his VT episode (**Fig. 2**) shows suspicious inverted T waves in the precordial leads (Fig. 1). After defibrillation, his EKG (**Fig. 3**) shows some notching between QRS and T waves, suggestive of Epsilon waves.

The patient was stable during his hospital stay and was discharged with a tentative diagnosis of ARVC pending a second opinion at a renowned and highly recommended institution. He was fitted with an external defibrillator and went home with no medications. At the second opinion, ARVC is confirmed by another CMR imaging, using the 2010 Task Force criteria. Genetic testing shows a desmosomal mutation, one among many possible genetic links that are associated with ARVC.[16]

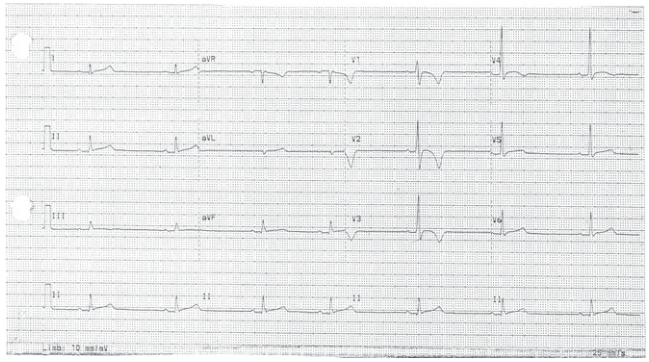

Fig. 1. EKG 2 yr prior to VT episode.

3. What is the recommended treatment?

A. Restriction from physical activities other than activities of daily living.
B. Automatic implantable cardioverter defibrillator (AICD) implantation.
C. Adding a beta blocker if heart rate/blood pressure allow and no significant side effects.
D. All the above.

ANSWER/DISCUSSION

3. D. ARVC can progress, and there is strong association between exercise and the induction of ventricular arrhythmias.[8,12] Any activity that causes symptoms of palpitations or presyncope is not advised.[5] Beta blocking agents may give a favorable risk benefit ratio and can be used empirically in patients with ARVC as tolerated.[5] An AICD is indicated for primary prevention for high-risk patients (younger patients, history of syncope, LV involvement, and those with penetrant ARVC genetic mutations) and for secondary prevention of sudden cardiac death in those who previously experienced sustained ventricular arrhythmia.[3,15]

The repeat CMR shows a reduced EF in both RV and LV. After a second EP study, the patient receives ablation of two inducible PVC foci on his RV and insertion of an AICD. He continues to work

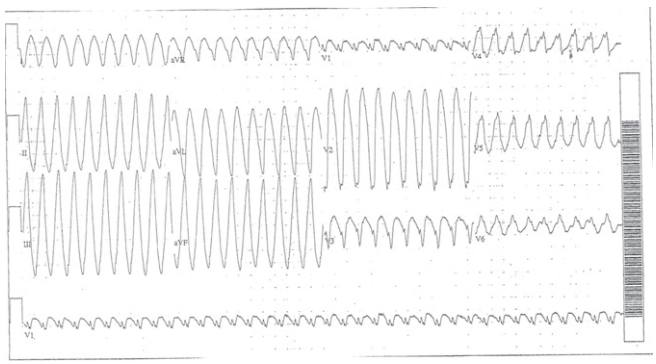

Fig. 2. EKG when presenting with symptoms.

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administratively and his AICD does not activate until 6 mo later, one time for light tennis playing and the other for mowing the lawn (AICD intervention and shock for ventricular fibrillation for both these events). Follow-up AICD interrogation reveals infrequent PVCs (4% of the time) with rare PVC couplets and triplets. Once the patient limits all sort of exertional activities, he has no further AICD firing. He goes to an AICD support group and checks in with his cardiologist regularly. He remains hopeful for a long and productive life and wishes to remain on active duty.

4. What is his aeromedical disposition?

A. Permanently disqualified from flying duty.
B. Waiver to return to flying status, Categorical Flying Class II (qualifies rated officers for duty in certain restricted aircraft categories).
C. Retain for active duty, without flying duties.
D. Monitor for another 2 yr, then reevaluate.

ANSWER/DISCUSSION

4. A. Sustained VT or any duration of nonsustained VT with associated hemodynamic symptoms is disqualifying for all United States Air Force (USAF) flying classes and for air traffic control/ground-based control/missile operation duties without waiver recommendation.[19] In the U.S. Navy Aeromedical Reference and Waiver Guide, sustained or unsustained VT is disqualifying and a waiver is not recommended.[13] For the Army, a flying waiver will not be recommended for those with nonsustained VT greater than 11 beats, sustained VT greater than 30 s in length, greater than 4 VT\episodes per evaluation, or VT with associated structural heart disease.[18]

According to the USAF Waiver Guide,[19] there are two primary military aeromedical concerns for individuals with cardiomyopathy. The first is the risk of sudden incapacitation. The risk of sudden death, arrhythmias, and/or thromboembolic events is generally correlated with the overall degree of cardiac dysfunction. ARVC and hypertrophic cardiomyopathy are more likely to be associated with potentially sudden incapacitating symptoms. Secondly, even mild degrees of myocardial dysfunction may be incompatible with military aviation duties due to an associated reduction in exercise tolerance, the need for complex medical therapy, and the need for frequent access to specialized medical care. Specifically, standard-of-care medical therapy for cardiomyopathy usually involves multiple hemodynamic, vasoactive, chronotropic, and diuretic medications that may alter physiological responses to the military aeromedical environment such that aviators cannot perform their usual duties without an undue increase in risk to themselves, the crew, or the mission. Device therapies for cardiomyopathies are not waiverable due in part to the unacceptably high complication rates associated with the devices themselves.[11] According to the Federal Aviation Administration, AICD is disqualifying.[6] The USAF Medical Standards Directory states that pacemakers or implantable cardioverter defibrillators are disqualifying for service.*

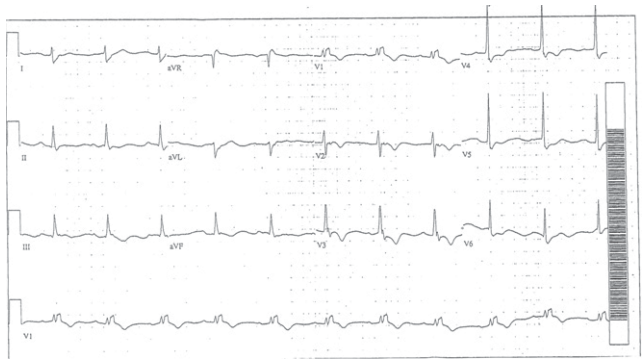

Fig. 3. EKG after defibrillation.

The patient underwent a Medical Board, which recommended an early retirement from the Air Force. The Department of Veterans Affairs initially advised him he would not qualify for veteran services/benefits because of his "preexisting condition." This thankfully resolved after much coordination between his military physicians and the Department of Veterans Affairs.

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REFERENCES

1. Abelmann WH, Lorell BH. The challenge of cardiomyopathy. J Am Coll Cardiol. 1989; 13(6):1219–1239.
2. Caforio AL, Pankuweit S, Arbustini E, Basso C, Gimeno-Blanes J, et al. Current state of knowledge on aetiology, diagnosis, management, and therapy of myocarditis: a position statement of the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases. Eur Heart J. 2013; 34(33):2636–2648.
3. Corrado D, Calkins H, Link MS, Leoni L, Favale S, et al. Prophylactic implantable defibrillator in patients with arrhythmogenic right ventricular cardiomyopathy/dysplasia and no prior ventricular fibrillation or sustained ventricular tachycardia. Circulation. 2010; 122(12):1144–1152.
4. Dec GW, Fuster V. Idiopathic dilated cardiomyopathy. N Engl J Med. 1994; 331(23):1564–1575.
5. European Heart Rhythm Association, Heart Rhythm Society, Zipes DP, Camm AJ, Borggrefe M, Buxton AE, et al. ACC/AHA/ESC 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology/American Heart Association Task Force and the European Society of Cardiology Committee for Practice Guidelines (Writing Committee to Develop Guidelines for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death). J Am Coll Cardiol. 2006; 48(5):e247–346.
6. Federal Aviation Administration. Item 36. Heart. III Aerospace medical disposition. Other cardiac conditions. In: Guide for aviation medical

* U.S. Air Force. Section H: heart and vascular USAF medical standards, H18. In: Medical standards directory. Washington (DC): Department of the Air Force; 2014:25. [Accessed 15 Nov. 2014]. Available for those with access from https://kx2.afms.mil/kj/kx4/FlightMedicine/Documents/Forms/ShowFolders.aspx?RootFolder=%2fkj%2fkx4%2fFlightMedicine%2fDocuments%2fMedical%20Standards%20Directory%20%28MSD%29&FolderCTID=0x0120004DEB19A0C597EF4794DF99094B5AD8FC.

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examiners. Washington (DC): Federal Aviation Administration; 2014:83-84. [Accessed 1 Dec. 2014]. Available from https://www.faa.gov/about/office_org/headquarters_offices/avs/offices/aam/ame/guide/media/guide.pdf.
7. Foale R, Nihoyannopoulos P, McKenna W, Kleinebenne A, Nadazdin A, et al. Echocardiographic measurement of the normal adult right ventricle [see corrections]. Br Heart J. 1986; 56(1):33–44. Corrected in: Br Heart J. 1987;57(4):396; Note: Klienebenne A [corrected to Kleinebenne A].
8. James CA, Bhonsale A, Tichnell C, Murray B, Russell SD, et al. Exercise increases age-related penetrance and arrhythmic risk in arrhythmogenic right ventricular dysplasia/cardiomyopathy-associated desmosomal mutation carriers. J Am Coll Cardiol. 2013; 62(14):1290–1297.
9. Le Guludec D, Gauthier H, Porcher R, Frank R, Daou D, et al. Prognostic value of radionuclide angiography in patients with right ventricular arrhythmias. Circulation. 2001; 103(15):1972–1976.
10. Marcus FI, McKenna WJ, Sherrill D, Basso C, Bauce B, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the task force criteria. Circulation. 2010; 121(13):1533–1541.
11. Murray D, Kruyer B, Van Syoc D. Cardiomyopathy (Mar 11). In: Air Force waiver guide. Wright-Patterson AFB (OH): U.S. Air Force School of Aerospace Medicine; 2014:149-154. [Accessed 26 Nov. 2014]. Available from http://www.wpafb.af.mil/afrl/711hpw/usafsam.asp.
12. Nava A, Bauce B, Basso C, Muriago M, Rampazzo A, et al. Clinical profile and long-term follow-up of 37 families with arrhythmogenic right ventricular cardiomyopathy. J Am Coll Cardiol. 2000; 36:2226–2233.
13. Naval Aerospace Medical Institute. 3.24. Ventricular tachycardia. In: U.S. Navy aeromedical reference and waiver guide. Pensacola (FL): Naval Aerospace Medical Institute; 2014. [Accessed 26 Nov. 2014]. Available from http://www.med.navy.mil/sites/nmotc/nami/arwg/Documents/Wavier%20Guide%20Nov%202014/Complete_Waiver_Guide_141121.pdf.
14. Niroomand F, Carbucicchio C, Tondo C, Riva S, Fassini G, et al. Electrophysiological characteristics and outcome in patients with idiopathic right ventricular arrhythmia compared with arrhythmogenic right ventricular dysplasia. Heart. 2002; 87(1):41–47.
15. Romero J, Mejia-Lopez E, Manrique C, Lucariello R. Arrythmogenic right ventricular cardiomyopathy (ARVC/D): a systematic literature review. Clin Med Insights Cardiol. 2013; 7:97–114.
16. Sturm AC. Genetic testing in the contemporary diagnosis of cardiomyopathy. Curr Heart Fail Rep. 2013; 10(1):63–72.
17. Thiene G, Corrado D, Basso C. Arrhythmogenic right ventricular cardiomyopathy/dysplasia. Orphanet J Rare Dis. 2007; 2(1):45.
18. Army US. 4-15a(13). Heart and vascular system. In: Standards of medical fitness. Washington (DC): Department of the Army; 2008:44. Army Regulation 40-501. [Accessed 26 Nov. 2014]. Available from http://armypubs.army.mil/epubs/pdf/r40_501.pdf.
19. Van Syoc D. Ventricular tachycardia (Jul 14). In: Air Force waiver guide. Wright-Patterson AFB (OH): U.S. Air Force School of Aerospace Medicine; 2014:1030-1035. [Accessed 26 Nov. 2014]. Available from http://www.wpafb.af.mil/afrl/711hpw/usafsam.asp.

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