COMMENTARY This commentary provides editorial perspectives on the report which follows

Pathologic Findings in a Patient Dying with Ventricular Tachycardia

In this issue of Chest (see page 24), James and MacLean present the pathologic findings in a 17-year-old male athlete with recurrent syncpe, documented ventricular tachycardia, and sudden cardiac death. The patient's brother had a somewhat similar history and also died suddenly. Pathologic examination in the present case revealed some fatal dispersion of the A-V node, a right-sided His bundle, and evidence of inflammatory neuritis and ganglionitis (most prominent around the area of the sinus node).

Before commenting upon the pathologic features described by James and MacLean, we would like to make some clinical observations that we feel are relevant to this case. Examination of the 12-lead electrocardiogram during ventricular tachycardia (kindly supplied to us by Drs. James and MacLean) revealed a wide QRS tachycardia (rate approximately 200 beats/min) with a QRS of left bundle branch pattern. These findings were consistent with ventricular tachycardia originating from the right ventricle. In our experience, electrocardiographic prediction of the site of origin of ventricular tachycardia in the absence of chronic ischemic heart disease (based upon QRS morphology of ventricular tachycardia), is a reliable guide to the ventricle of origin (ventricle of origin diagnosed by endocardial and/or epicardial mapping). This is in contrast to prediction of site of origin of ventricular tachycardia occurring in ischemic heart disease, where electrocardiographic prediction is considerably less reliable.

Assuming that the case reported by James and MacLean is in fact an example of recurrent right ventricular tachycardia, one can question whether this diagnosis has any special implications. Recently, Fontaine et al[8,9] have described a group of patients with "arrhythmogenic right ventricular dysplasia." This group of patients has been afflicted with recurrent right ventricular tachycardia proved by epicardial mapping, and sometimes cured with surgical attack upon the site of origin of the dysrhythmia. Characteristic features of "arrhythmogenic right ventricular dysplasia" include delayed activation of portions of the right ventricle (occasionally diagnosed from the routine surface electrocardiogram, more frequently diagnosed from the amplified and computer average surface electrocardiogram), and discernible with endocavitary and epicardial mapping of the right ventricle. Right ventricular angiography in this group of patients reveals localized or generalized right ventricular wall motion abnormality, and pathologic examination reveals dysplastic changes in the right ventricle.[8,9] Arrhythmogenic right ventricular dysplasia appears to be related to Uhl's disease, which is characterized by partial or complete absence of the myocardium of the right ventricle with replacement by fibroelastic and adipose tissue. We are currently examining the heart of a young woman who died with right ventricular tachycardia complicating Uhl's disease. The patient of James and MacLean did not have arrhythmogenic right ventricular dysplasia or Uhl's disease either clinically or pathologically.

Our laboratories have been aware of a strong association of recurrent right ventricular tachycardia and absence of clinically manifest organic heart disease (as opposed to left ventricular tachycardia, where organic heart disease is the rule).[1,8-9] We have noted great variability in the clinical manifestations of recurrent right ventricular tachycardia without organic heart disease.[9] Our patients have fallen into a number of categories including: 1) sporadic paroxysmal sustained ventricular tachycardia (usually reentrant based upon electrophysiologic criteria); 2) repetitive, nonsustained ventricular tachycardia (usually not reentrant utilizing electrophysiologic criteria); and 3) exercise-provoked right ventricular tachycardia (not reentrant by electrophysiologic criteria, but inducible with catecholamine infusion). Based upon QRS morphology, we would classify the presently reported patient of James and MacLean as an example of recurrent right ventricular tachycardia without apparent complicating organic heart disease. The history of sudden death in the brother is probably clinically relevant and certainly intriguing.
Our understanding of the group of patients with recurrent right ventricular tachycardia and absence of organic heart disease is limited. The results of electrophysiologic studies in the various groups are very briefly summarized in the preceding paragraph. Right ventricular angiography has usually revealed normal findings (no evidence of "arrhythmogenic right ventricular dysplasia" or Uhl's disease).8

Our reported pathologic data are limited to one fatal case,10 which shared some features with the presently reported case of James and MacLean. These shared features included anatomic abnormality of AV nodal architecture (our case demonstrated an AV node which was partially engulfed in the central fibrous body), and a right-sided His bundle.

In our case there was focal infiltration of mononuclear cells in the approaches to the SA node, approaches to the AV node and the atrial preferential pathways. The nerve ganglia were not involved. We did not believe that this constituted sufficient evidence of myocarditis in the conduction system in our case. In addition there were recent and old scars in the ventricular myocardium which we believed were secondary to longstanding tachycardia.

The pathologic findings described by James and MacLean in this issue are of great interest. Pathologic demonstration of degenerative and inflammatory neural changes is most intriguing. The hypotheses proposed by James and MacLean are persuasively presented. However, there was another shared feature in their case and in our recently reported case.10 Both had obvious pathologic abnormalities, but in both cases, it was certainly not immediately apparent how the pathologic features related to the clinically defined arrhythmia in a direct cause-and-effect relationship.

It is our opinion that further intense scrutiny of patients with recurrent ventricular tachycardia and no apparent heart disease is mandatory before the pathophysiology of these arrhythmias will be completely clarified. This scrutiny should include careful clinical analysis, noninvasive and invasive cardiovascular evaluation, detailed electrophysiologic study (in the catheterization laboratory and in the operating room when appropriate), and detailed pathologic examination when the opportunity is available. It is hoped that continued correlation of function (electrophysiology) and structure (pathology) will provide important data concerning the pathophysiology of arrhythmia. Understanding of the pathophysiology is crucial to developing rational therapy.

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REFERENCES
7 Bharati S, Rosen KM, Lev M. Unpublished data