JACC Vol. 15, No. 5
April 1990:1183–7

CASE REPORTS

Sudden Death Due to Isolated Acute Infarction of the His Bundle

THOMAS N. JAMES, MD, FACC, LEROY RIDDICK, MD*

Galveston, Texas and Mobile, Alabama

A 29 year old black man considered to be in good health died suddenly and unexpectedly. At postmortem examination there were no significant gross abnormalities, all drug screens were normal and sicklemia was not present. Special studies of the cardiac conduction system demonstrated isolated acute infarction of the His bundle, with no similar evidence of myocardial infarction anywhere else in the heart. The atrioventricular (AV) node artery was moderately narrowed, but its branch supplying the His bundle was occluded >95% by focal fibromuscular dysplasia. There were no other significant coronary lesions or other abnormalities in the heart.

(J Am Coll Cardiol 1990;15:1183–7)

Apoplexy of the heart can be just as fatal as apoplexy of the brain (1). At either site the specific tissue injured is critical in determining whether survival is to be expected. In the heart one can consider the His bundle the Achilles heel of the conduction system because its sudden destruction creates a special vulnerability to sudden death. Other sites within the conduction system, or even in the working myocardium, may lead to electrical instability of varying degrees of gravity and even death. However, acute destruction of the His bundle not only interrupts normal atrioventricular (AV) conduction, but also simultaneously affects one of the principal points of origin for AV junctional escape rhythms (2).

We recently had the opportunity to examine the heart of a young man who died suddenly and unexpectedly and we found acute infarction that was confined exclusively to the His bundle. This report describes that case including the responsible vascular lesion and relates these findings to analogous cases previously studied.

Case Report

A 29 year old black man with no known history of medical problems was sitting on the edge of the bed one morning tying his daughter's shoes when he suddenly grabbed the back of his head, moaned and collapsed unconscious. Emergency help was summoned and arrived quickly, but despite extensive resuscitative efforts for 1 h, he died. His wife, an observant detective for the Mobile Police Department, said that he had appeared well that morning and she knew of no recent illness or complaint of any nature.

Autopsy findings. At postmortem examination there was congestion in the lungs, liver, spleen and other organs. The brain weighed 1,520 g and was swollen sufficiently to cause some herniation, but there was no intracranial hemorrhage, encephalitis or other inflammatory disease and there were no focal intracerebral lesions. Except for modest generalized cardiac enlargement (heart weight 410 g), there were no gross abnormalities of the heart; the valves were normal and pliable, all major coronary arteries were widely patent and the myocardium was free of necrosis and fibrosis at gross examination. There was no evidence of trauma. Results of toxicologic examinations of blood and urine, using gas chromotography and mass spectrometry, were normal.

Conduction system histology. Because of the possibility of a fatal cardiac arrhythmia, special examination of the cardiac conduction system was performed. Methods for this purpose have been published previously (3,4). The entire AV node and His bundle were serially sectioned at 8 μm. There was focal fibromuscular dysplasia of the sinus node artery, narrowing its lumen about 50%, but the tissue of the sinus node was normal. Surrounding nerves and ganglia were essentially normal.

A striking abnormality was present in the His bundle, which was virtually destroyed by hemorrhagic infarction (Fig. 1). In addition to the hemorrhage and small areas of necrosis within the His bundle, there were a few focal accumulations of leukocytes. The involved tissue extended from the midpoint of the undivided His bundle down to the

From the World Health Organization Cardiovascular Center and the Department of Medicine and Department of Pathology, University of Texas Medical Branch, Galveston, Texas and the *Alabama Department of Forensic Sciences and the Department of Pathology of the University of South Alabama College of Medicine, Mobile, Alabama. This work was supported by the Pegasus Fund of the University of Texas Medical Branch, Galveston.

Manuscript received October 16, 1989, accepted November 7, 1989.

Address for reprints: Thomas N. James, MD, Office of the President, University of Texas Medical Branch, Galveston, Texas 77550-2774.

0735-1097/90/$3.50

JACC Vol. 15, No. 5
April 1990:1183–7

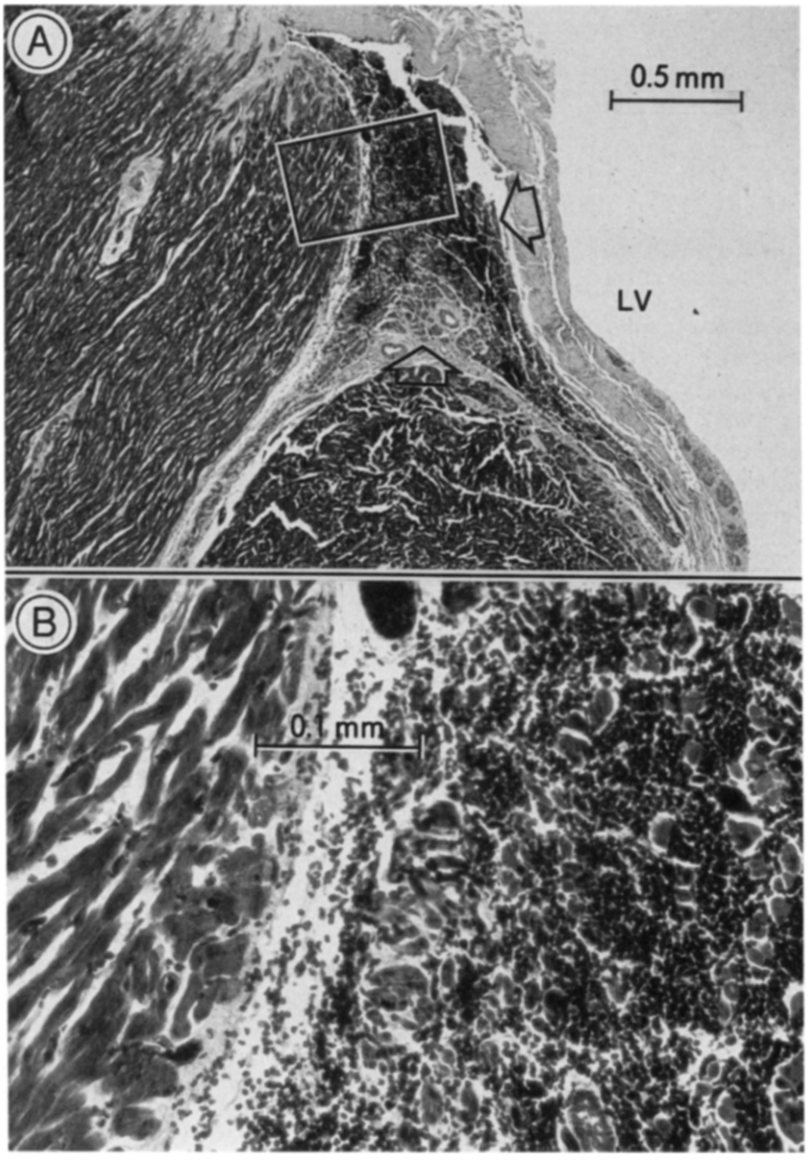

Figure 1. Photomicrograph illustrating acute isolated infarction of the His bundle (arrows in A). The boxed area in A is seen at higher magnification in B, where necrotic myocytes are seen scattered within the hemorrhagic His bundle. All stains are the Goldner trichrome and all magnifications are indicated with reference bars. LV = left ventricular cavity.

divisions into right and left bundle branches, a distance of about 1.5 mm. Except for a small focus of hemorrhage in the proximal left bundle branch, the two bundle branches were histologically normal. Within the AV node there were a few foci of old fatty degeneration and some focal fibrosis, but no recent lesions. The nutrient arteries supplying the AV node and His bundle normally enter the posterior margin of the AV node, originating from the major coronary artery that crosses the crux; in 90% of human hearts this is the right coronary artery (5), as it was in this case. The left circumflex artery terminated at the obtuse margin of the left ventricle.

Near the posterior margin of the AV node its nutrient artery divided into several small branches, one of which was determined by serial sectioning of this region to be the vessel supplying the His bundle (Fig. 2). There was moderate narrowing of the main artery to the AV node, but the artery supplying the His bundle was >95% occluded by focal fibromuscular dysplasia. There was no local inflammation nor any embolus or thrombosis of these or any other coronary branches. There were no sickled erythrocytes.

Discussion

There can be little doubt that the isolated acute infarction of the His bundle in this case was responsible for sudden death, but there are several unanswered questions. Why were there no recognized preceding symptoms, given the chronic nature of the lesion probably responsible for narrowing the nutrient artery to the His bundle? Why was there no survivable escape rhythm? What was the significance, if any, of the slight cardiac enlargement? How often are similar abnormalities responsible for cases of sudden death? Why

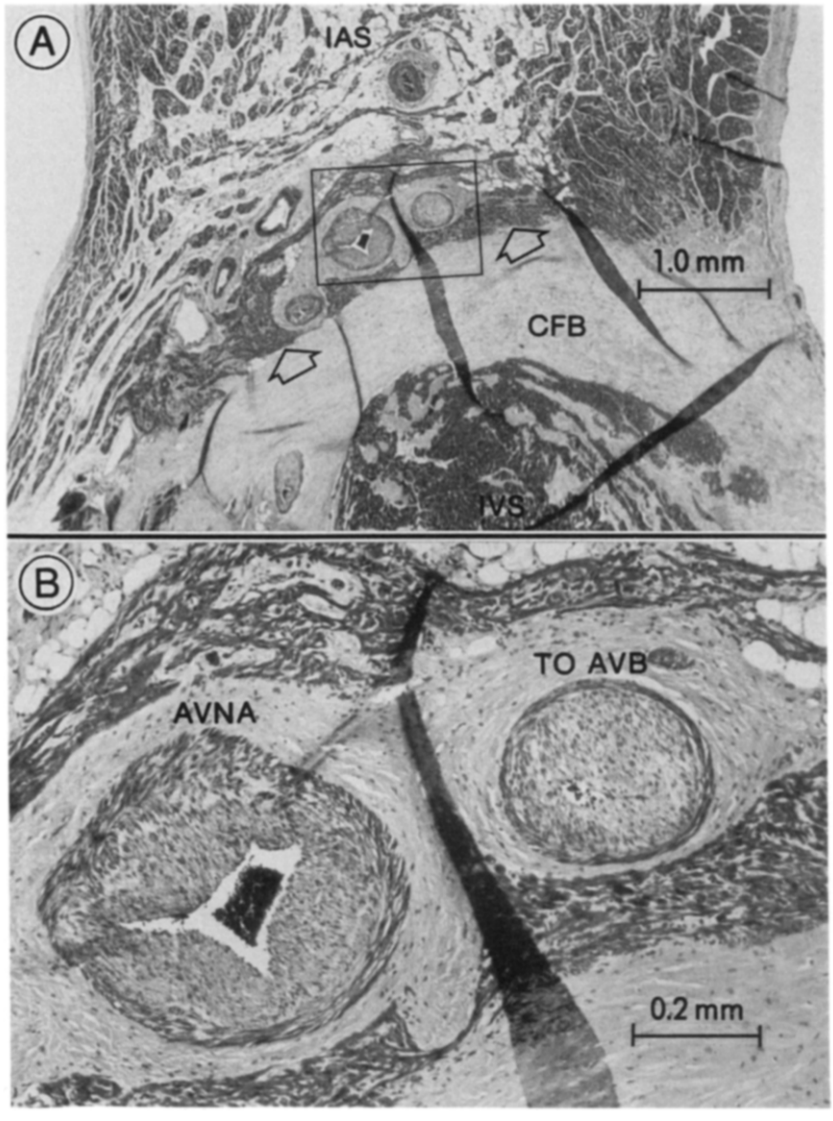

Figure 2. The area of the AV node (two open arrows in A) contains the main AV node artery and two of its branches. The boxed area in A is shown at higher magnification in B, where details of the moderate focal fibromuscular dysplastic narrowing are visible for the main AV node artery (AVNA) and its nearly occluded branch supplying the His bundle (marked "TO AVB"). CFB = central fibrous body; IAS = interatrial septum; IVS = interventricular septum.

was there cerebral edema? The following discussion will review these questions.

Why were there no preceding symptoms? Two considerations may help explain the absence of recognized previous symptoms. First, we could assume that there were some symptoms, perhaps transient light-headedness or syncope followed by amnesia for the episode, that the deceased did not consider significant and that were not fortuitously observed by his wife or others. Second, we could assume that there genuinely were no preceding symptoms and that the terminal episode represented a new transient but lethal event, such as local coronary spasm or transient obstructive aggregation of platelets. Concerning spasm, the narrowing lesion did contain much smooth muscle, all organized in dysplastic array, but whether that would make it either more or less responsive to a vasoconstrictive stimulus is uncertain. A search for evidence of platelet debris or other material in the lumens of downstream branches did not reveal any. It may be that flow through the narrowed artery remained sufficient, even barely, until some finite limit was imposed by the simple gradual progression of the narrowing lesion and that the first clinical evidence of local coronary insufficiency was also the last.

Why was there no survivable escape rhythm? The site from which stable or even survivable AV junctional escape rhythms emerge has been the subject of frequent clinical and experimental studies, but there is some evidence to suggest that the site is at or very near to the point of junction between the AV node and the His bundle (2,6). If that is so, then destruction of the His bundle, as occurred in the present case, may be anticipated not only to block subsequent transmission of normal or abnormal supraventricular rhythms, but also to simultaneously destroy the site from which a locally originating escape rhythm might have emerged. Similar considerations apply to postulated terminal events associated with benign congenital polycystic tumors

JACC Vol. 15, No. 5
April 1990:1183–7

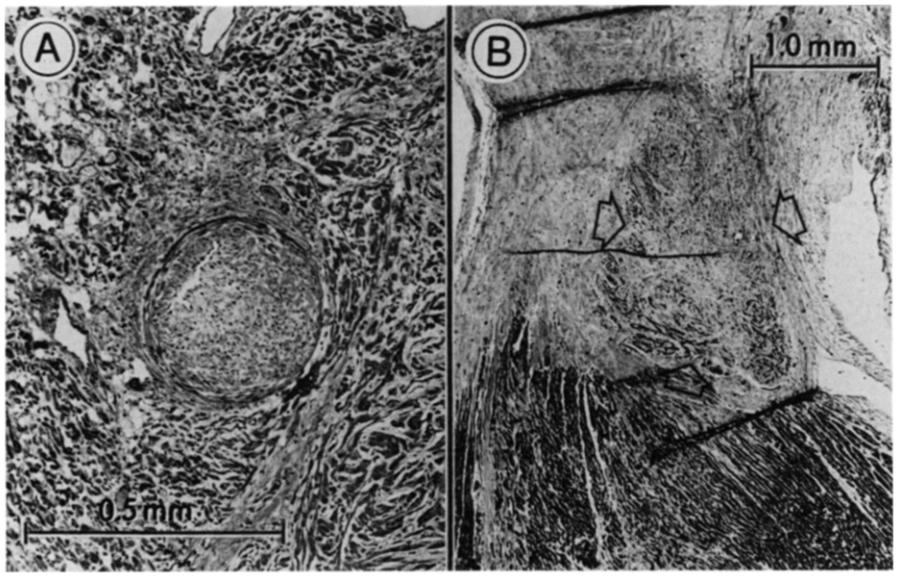

Figure 3. An example of photomicrographs analogous to a previously published report (10), showing focal fibromuscular dysplastic narrowing of the artery supplying the His bundle. Note the similarity of the artery shown in A and the artery supplying the His bundle in Figure 2. B, Chronic fibrous destruction of the His bundle (three arrows) is displayed, being the consequence of long-standing ischemia caused by the lesion in A.

of the AV node (so-called mesotheliomas) (7) or with hemorrhage in and around the His bundle from a bleeding diathesis due to leukemia and thrombocytopenia in a patient who died with heart block (8).

What is the significance of the cardiac enlargement and cerebral edema? In the absence of focal myocardial fibrosis or inflammation or other generalized lesions in the myocardium of the ventricles or atria and in the absence of valvular abnormalities or known hypertension, we interpret the small amount of cardiac enlargement as a normal variant not of clinical significance except that any increased myocardial mass would facilitate the development of ventricular fibrillation and render it more difficult to treat (9). A corollary question may be raised about the edema of the brain, especially in view of the dying episode in which the man grabbed his head and fell unconscious. However, one of us (L.R.) has seen numerous examples of this type of edema of the brain in persons who died after prolonged resuscitative measures as were applied in the present case; in the absence of cerebral hemorrhage, encephalitis or a tumor, we attribute the cerebral edema in this case to hypoxia associated with attempted resuscitation.

How often does isolated acute infarction of the His bundle occur? No one knows how often sudden death may be explained, as in this man, but isolated acute infarction of the His bundle may be more unrecognized than rare. This suspicion is supported by the facts that the possibility is seldom considered and the appropriate region is infrequently examined. In the present case we have little doubt that most would (and both of us did) miss the lesion on gross examination. It was only because of the routine histologic study of appropriate sites, as is customary in our practice in such cases, that the distinctive abnormality was recognized.

In a previous analogous study (10) it was demonstrated that focal fibromuscular dysplastic narrowing of the artery supplying the His bundle led to chronic fibrotic destruction of the His bundle (Fig. 3). Similar findings causing sudden death have been reported by Anderson et al. (11). In one of our two reported subjects (10) there was a recognized long-standing complete heart block that was for a time successfully treated with a pacemaker, permitting "an experiment of nature" in which the effect of prolonged ischemic destruction and the consequent dense fibrosis of the His bundle could be visualized because survival had been assured with electronic pacemaking. Why neither the second patient of that report (10) nor the present patient had any recognized clinical or electrocardiographic evidence of heart block remains perplexing. However, it should be remembered that in every case of sudden death one must consider multiple contributing causative factors that interplay for the final event, most often on the basis of chance alone (12).

Conclusions. The best lesson from this case may be the recognition of what is arguably the smallest form of acute myocardial infarction capable of causing sudden death. The absence of any other area of myocardial infarction, or indeed of any other major cardiac abnormality, was distinctive. Even though the total volume of infarcted myocardium here could not have been more than 2 mm^3, it was readily demonstrable by a histologic study, which is not difficult. We urge that in every investigation of otherwise unexplained sudden death, examination of the entire cardiac conduction system be included, but especially of the His bundle and its blood supply.

References

1. James TN. De subitaneis mortibus. XXVIII. Apoplexy of the heart. Circulation 1978;57:385–91.

2. James TN, Isobe JH, Urthaler F. Correlative electrophysiological and

anatomical studies concerning the site of origin of escape rhythm during complete atrioventricular block in the dog. Circ Res 1979;45:108–19.

3. James TN. The sinus node. Am J Cardiol 1977;40:965–86.
4. James TN. Structure and function of the AV junction: the Mikamo Lecture for 1982. Jpn Circ J 1983;47:1–47.
5. James TN. Anatomy of the Coronary Arteries. Hagerstown, MD: Harper Brothers, 1961:88–102.
6. Woods WT, Sherf L, James TN. Structure and function of specific regions in the canine atrioventricular node. Am J Physiol 1982;243:H41–50.
7. James TN, Galakhov I. De subitaneis mortibus. XXVI. Fatal electrical instability of the heart associated with benign congenital polycystic tumor of the atrioventricular node. Circulation 1977;56:667–78.
8. Alpert MA, Dix JD, Hamel PC, Vu LC. Complete heart block caused by hemorrhage into the atrioventricular conduction pathway. South Med J 1982;75:601–4.
9. Kohya T, Kimura S, Myerburg RJ, Bassett AL. Susceptibility of hypertrophied rat hearts to ventricular fibrillation during acute ischemia. J Mol Cell Cardiol 1988;20:159–68.
10. James TN, Hackel DB, Marshall TK. De subitaneis mortibus. V. Occluded A-V node artery. Circulation 1974;49:772–7.
11. Anderson KR, Bowie J, Dempster AG, Gwynne JF. Sudden death from occlusive disease of the atrioventricular node artery. Pathol Annu 1981; 13:417–21.
12. James TN. Chance and sudden death—lessons from nature. In: Yu PN, Goodwin JF, eds. Progress in Cardiology. Philadelphia: Lea & Febiger, 1981:101–27.