immunofluorescence[2] or immunoperoxidase stains.[3]

Characteristic radiologic changes in the vertebral column associated with multiple myeloma are frequently absent. Pure osteopenia without specific features occurs in 32% of cases, and the spine is radiologically normal in 18%.[4] A complete skeletal survey, bone marrow examination, and appropriate serum and urine protein investigations are required to exclude the diagnosis of myeloma.

A drastic loss in height has been reported with severe renal osteodystrophy. Uncontrolled tertiary hyperparathyroidism in a man undergoing dialysis was associated with a decrease in height of 28 cm.[5] Hence the term "the shrinking man". Earlier diagnosis of myeloma in our patient could probably have prevented the dramatic skeletal changes. The disease responded well to chemotherapy, and during treatment there was no further loss of height.

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Long-term management of arrhythmogenic right ventricular dysplasia

R.J. SEVICK,* B MED SCI
D. GEORGE WYSE,† MD, PH D, FRCP[C]

Arrhythmogenic right ventricular dysplasia (ARVD) was first described as a clinical entity by Frank and colleagues in 1978.[1] Since then, about 40 cases have been reported.[2,3]

From the electrophysiology unit, cardiology division, department of medicine, Foothills Provincial General Hospital, Calgary, and the University of Calgary

*Medical student supported by a summer studentship from the Alberta Heritage Fund for Medical Research
†Senior clinical investigator of the Alberta Heart Foundation

Reprint requests to: Dr. D. George Wyse, Department of medicine (cardiology), Faculty of medicine, University of Calgary, 3330 Hospital Dr. NW, Calgary, Alta. T2N 1N4

The condition is characterized by an unusual myopathy restricted to the right ventricle, generally associated with recurrent ventricular tachycardia. The tachycardia usually has a pattern of left bundle branch block, suggesting that the arrhythmia originates in the right ventricle; it can usually be reproduced in the electrophysiology laboratory. Echocardiography and angiography will also show that the right ventricle is dilated and hypokinetic. The cause of ARVD is still unknown, although the condition may be congenital.[4] Many of its features, particularly the fact that survival can be lengthy, remain unclear. This report describes how the dysplasia presented as near-miss sudden cardiac death in a 17-year-old boy and how it has been managed successfully over the last 4 years.

Case report

A 17-year-old boy from rural Al-

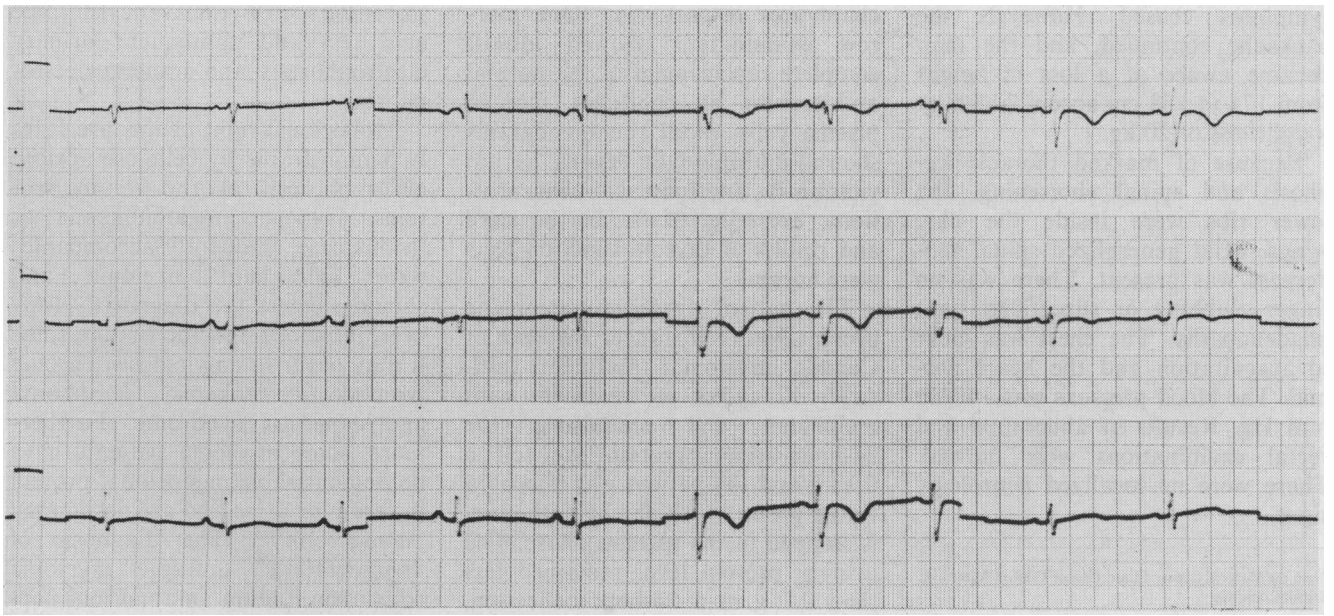

FIG. 1—When patient was not receiving medication 12-lead electrocardiogram showed mild rightward deviation of QRS axis, nondiagnostic T-wave changes and notched S-wave in lead V_1.

berta was brought to our hospital several hours after resuscitation following cardiac arrest. He had previously been well and active. His family and perinatal histories, like that of his childhood illnesses, were unremarkable with respect to cardiac disease. The patient had no chronic illness. Ten years earlier he had had surgery for strabismus; a congenital cataract was present at that time. The year before the current admission a benign laryngeal tumour had been excised.

The patient's condition was now dominated by coma and normotensive pulmonary edema. The first electrocardiogram (ECG) showed numerous multifocal ventricular premature beats, including salvoes of unsustained ventricular tachycardia and nondiagnostic ST-segment and T-wave changes. The QT interval was not significantly prolonged. The pulmonary edema cleared rapidly with medical management (mechanical ventilation, insertion of a Swan–Ganz catheter and administration of digitalis and a diuretic); the patient's neurologic status gradually improved too, leaving no evidence of permanent brain damage.

After the patient's recovery the pulse was irregular, the jugular venous "a" wave was prominent, and the components of the second heart sound did not completely merge to a single sound in expiration. The following investigations yielded normal results: posterior–anterior and lateral roentgenography; complete and differential blood counts; biochemical and immunologic analysis of the blood; radionuclide lung scanning; culture of sputum, urine, blood and stool; and determination of virus antibody titres in blood obtained during the acute and convalescent stages of the illness. Multiview chest roentgenography after a barium swallow suggested that there was slight enlargement of the right ventricle, while M-mode echocardiography showed that the ventricle's diameter was at the upper limit of normal and that there was "minimally abnormal septal motion". Right- and left-sided cardiac catheterization gave normal results, but angiography was not done at this time because the patient had had a period of rigor following a test injection of the contrast medium Renografin (diatrizoate meglumine and diatrizoate sodium). A test for serum antibodies against rat myocardium gave a positive result. Treatment with disopyramide seemed to control the rhythm disturbances. The patient did well and was discharged with a prescription for 800 mg/d of disopyramide. The tentative diagnosis was myocarditis of uncertain cause.

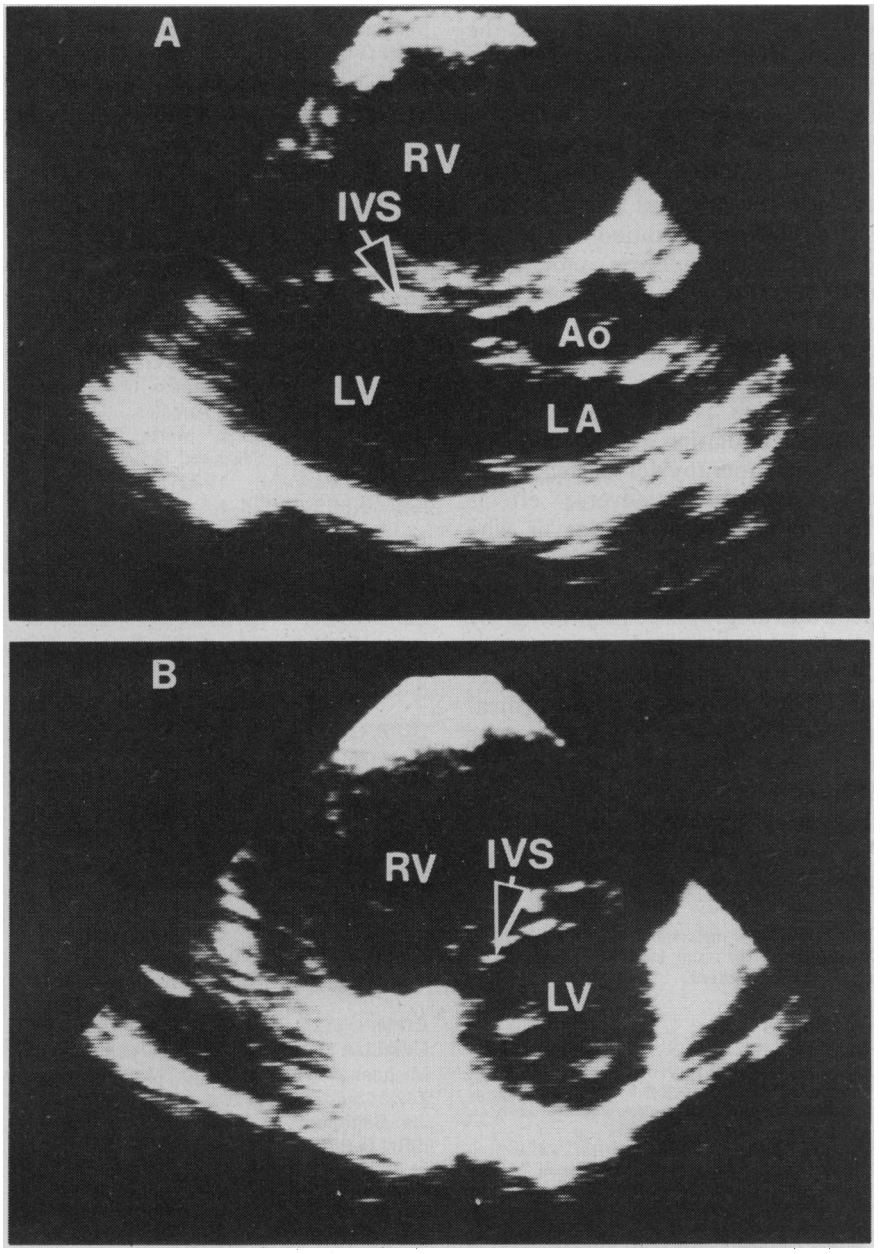

FIG. 2—Single-frame, end-diastolic, two-dimensional echocardiograms (A, long-axis view; B, short-axis view) revealed enlarged and hypokinetic right ventricle (RV). LV = left ventricle; IVS = interventricular septum; LA = left atrium; Ao = aorta.

The patient was readmitted 10 months later after ambulatory electrocardiographic monitoring had revealed asymptomatic, unsustained ventricular tachycardia. Many of the earlier laboratory investigations were repeated, without producing any additional diagnostic information. A 12-lead ECG obtained during a period when the patient had received no medication (Fig. 1) revealed mild rightward deviation of the mean QRS axis, the axis being +120°, a notched S-wave in leads V_1 to V_3 and nondiagnostic T-wave changes. Two-dimensional echocardiography revealed an enlarged and hypokinetic right ventricle (Fig. 2). This finding is always present in ARVD.[4] Several antiarrhythmic drugs (lidocaine, propranolol, quinidine, phenytoin and procainamide),

used alone or in combination, failed to suppress the arrhythmia and caused intolerable adverse effects. The patient underwent an electrophysiologic evaluation, and during invasive investigation programmed right ventricular extrastimuli repeatedly induced ventricular tachycardia with a pattern of left bundle branch block. The tachycardia was suppressed completely by a combination of disopyramide and lidocaine. Tocainide was substituted for lidocaine, and the patient was discharged taking every 8 hours 300 mg of disopyramide and 600 mg of tocainide.

For the last 3 years the patient has been successfully managed with this combination of drugs. At first his condition was checked by ambulatory electrocardiographic monitoring and in frequent visits to our arrhythmia clinic. Currently a rhythm strip is transmitted once each week by telephone,[5] and the patient is seen annually in the arrhythmia clinic and somewhat more frequently by his family physician.

Two years ago the rhythm strips started to show an increased number of ventricular premature beats, including some salvoes of three. The patient was admitted to hospital and monitored constantly. It was noted then that the arrhythmia increased in frequency in the hour before the next dose of medication. The dosage was altered to disopyramide, 300 mg every 6 hours, and tocainide, 600 mg at 6 am and midnight and 400 mg at noon and 6 pm. This resolved his cardiac problem. For the last 18 months the weekly rhythm strips have shown only infrequent unifocal ventricular premature beats.

Once in the first year of therapy the patient's leukocyte count fell to 2.1×10^9/l; this was initially attributed to the disopyramide therapy.[6] However, measles developed shortly thereafter, and following that illness the count returned to normal. The only apparent adverse reaction to the drugs he takes is an intermittent fine tremor that is easily tolerated and is felt to be due to the tocainide. A proposed trial of withdrawal of medication with continuous electrocardiographic monitoring in hospital, which would be followed by surgical treatment[2,3,7,8] if the ventricular tachycardia recurred, was refused by the patient.

Discussion

We have described a fairly typical case of ARVD, although the presentation as near-miss sudden cardiac death was unusual. Features that were characteristic of the condition included ventricular arrhythmias, a dilated and hypokinetic right ventricle, no other structural heart disease, the patient's sex, incomplete merging of the components of the second heart sound and rightward deviation of the QRS axis, with T-wave changes in the precordial leads. However, the classic epsilon wave, which, when present, is the second component of a markedly notched or biphasic T wave,[1-4] was not seen in this case.

This recently described and relatively unusual life-threatening illness has probably escaped recognition in the past. The condition should be looked for in children and young adults, particularly males, who have otherwise unexplained ventricular arrhythmias. Two-dimensional echocardiography is useful in making the diagnosis, as is programmed ventricular stimulation in devising pharmacologic therapy for patients who suffer intolerable adverse effects from antiarrhythmic drugs or who have drug-resistant arrhythmias. This case also illustrates the value of electrocardiographic monitoring by telephone in selected patients. On the basis of our experience we suggest that in some patients medical management will control the arrhythmia for extended periods. Spontaneous resolution of the arrhythmia has been reported in ARVD[3] and may yet occur in this individual.

The cooperation of the patient's primary physicians, Drs. Alam S. Khan and Larry G. Hoogeveen, is gratefully acknowledged.

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