TORSADE DE PONTES AND COMPLETE HEART BLOCK IN FAMILIAL CARDIOMYOPATHY

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SYNOPSIS

We describe two patients with familial congestive cardiomyopathy associated with trifascicular disease and complete heart block. In one patient recurrent Stoke-Adams attack occurred due to Torsade de Pontes probably precipitated by hypokalaemia. Patients with congestive cardiomyopathy and trifascicular disease are prone to sudden death. This is frequently attributed to conduction disturbances. In our patient ventricular tachycardia was the precipitating cause.

CASE REPORT

A 40-year-old Chinese female was admitted into the Coronary Care Unit with a history of 3 syncopal episodes on the day of admission. On admission she was drowsy and hypotensive. The electrocardiogram showed complete heart block with a wide QRS escape rhythm at a rate of 46 beats per minute (Figure 1). Soon after admission the patient developed torsade de pontes which degenerated into ventricular fibrillation causing a Stoke Adams attack. She was cardioverted and an emergency temporary transvenous pacemaker inserted. Subsequently she gave a history of "heart disease" for the last 13 years with 2 previous syncopal episodes. She had been told that her heart rate was slow but had previously refused a permanent pacemaker. She was dyspneic on moderate exertion and was treated with a thiazide diuretic and potassium supplements. There was no history of ischaemic chest pain. There were no other significant medical illnesses. Clinical examination revealed moderate cardiomegaly without any overt evidence of cardiac failure. There were no cardiac murmurs. The chest X-ray showed moderate cardiomegaly (Figure 2). The potassium was 2.2 mEq/litre. The other electrolytes were normal. Cardiac enzymes were normal. A M-mode and 2D-scan revealed left ventricular dilatation with features of congestive cardiomegaly. The potassium deficit was corrected and a permanent VVI pacemaker implanted. She has been seen regularly for 3 years without any further syncopal episodes.

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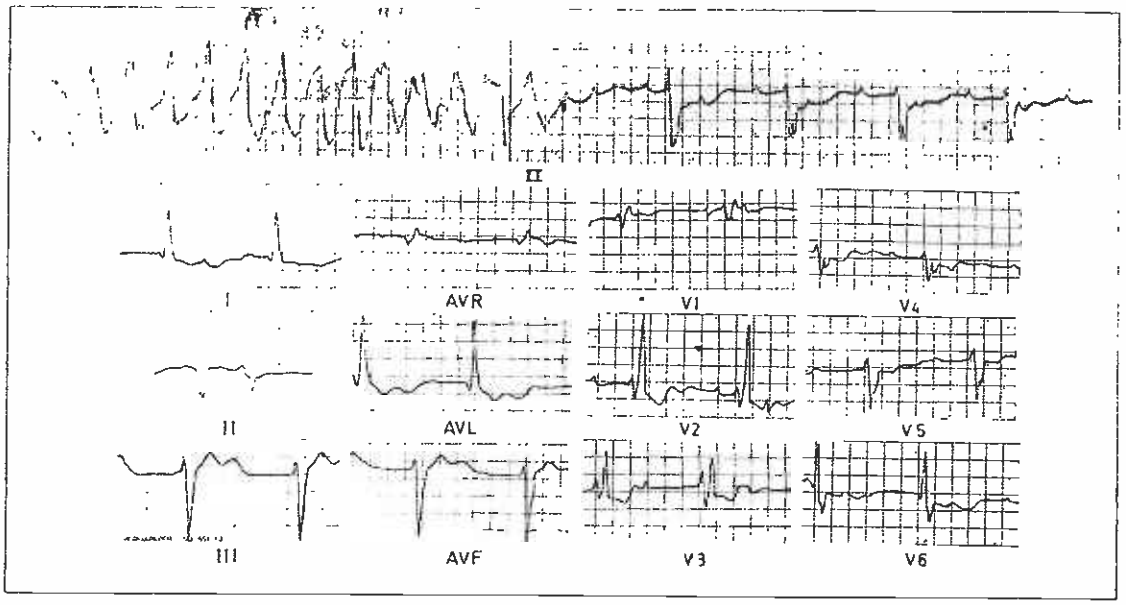


FIGURE 1
The rhythm strip 11 shows multiform ventricular tachycardia terminating spontaneously. The 12 lead electrocardiogram shows complete AV dissociation with a wide QRS escape rhythm.

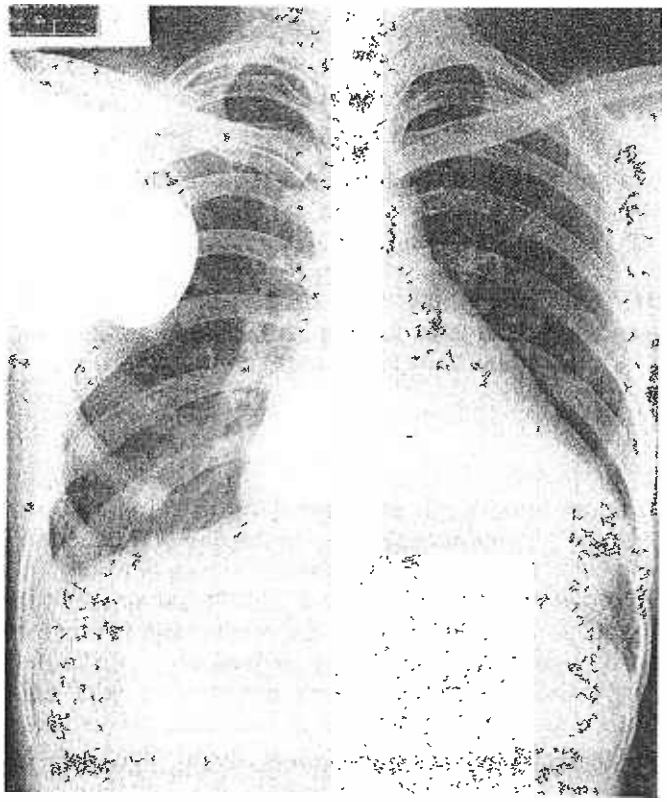
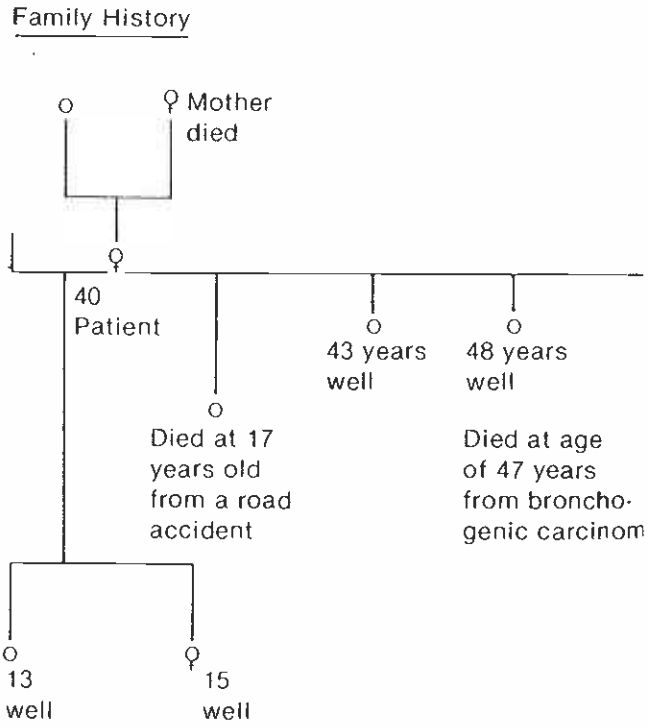


FIGURE 2
The chest roentgenogram shows gross cardiomegaly and the permanent pacemaker.

The patient has 4 brothers; two had noncardiac deaths. The other 2 brothers are farmers and asymptomatic but have refused to be examined. The patient has 2 children who have normal clinical examination and electrocardiograms. The patient's mother presented at the age of 66 with congestive cardiac failure and completed heart block with a wide QRS escape rhythm of approximately 45 beats/min. Chest X-rays showed gross cardiomegaly with pulmonary congestion. A temporary pacemaker was inserted and she



improved with treatment of her heart failure. The atric ventricular dissociation improved spontaneously witl the electrocardiogram showing sinus rhythm witl right bundle branch block, left anterior hemi-block an a prolonged PR interval. She refused a permanen pacemaker implantation and was treated with isc prenaline and diurectics. She had 4 other admission for heart failure with an additional episode of atric ventricular dissociation requiring temporary cardia pacing.

DISCUSSION

Familial transmission of primary myocardial diseas is well documented in idiopathic hypertrophic cardic

myopathy. It is less common to have congestive cardiomyopathy occurring within families (1-4). A recent series on congestive cardiomyopathy found only 2% of cases were familial (5). Genetic transmission has been suggested to be Mendelian dominant with maternal transmission (6). The presence of the disease in our patient and mother is consistent with this mode of transmission.

Although patients with congestive cardiomyopathy frequently have intraventricular conduction defects or bundle branch blocks (4, 5) complete heart block is less common (7, 8). This was present in our patient and her mother. During complete heart block in the patient and mother the escape rhythm was slow with wide QRS complexes indicating a distal escape pacemaker. During periods of atrio-ventricular conduction the electrocardiograms showed bundle branch blocks and prolonged PR intervals. These suggested the presence of disease in the distal conduction system. Myocardial fibrosis is present in variable amounts in idiopathic congestive cardiomyopathy (9). A recent case report (10) has shown the presence of degeneration of bundle branches of the AV conduction tissue in a patient with familial infra-Hisian block. This could conceivably be the cause of distal conductive disease in our patient.

Ventricular ectopy is common in patients with congestive cardiomyopathy (1-5). These patients have an increased tendency to sudden death. The mode of sudden death is usually attributed to heart blocks.

Ventricular tachycardia is unusual. In our patient torsade de pontes degenerating into ventricular fibrillation caused a Stoke Adams attack. Ventricular tachycardia was never documented in the mother despite similar conduction defects. We feel that the hypokalaemia in the presence of complete atrio-ventricular dissociation was the precipitating cause of the ventricular tachycardia in our patient. As patients with congestive cardiomyopathy are frequently treated with diuretics, greater care must be taken to maintain electrolyte homeostasis in these patients.

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