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CASE REPORT

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A simple case of bifascicular block, or is there more than meets the eye?

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There are many neuromuscular diseases which have cardiac manifestations. These are largely infiltrative myopathies, at times having a predilection for the conducting tissues, at others, largely involving the moycytes. The authors describe a 52-year old man with myotonic dystrophy type I and syncopal episodes.

The patient presented to an outpatient neurology clinic for a regularly scheduled appointment after a few years of being lost to follow up. He related having experienced two syncopal episodes in the preceding month. On one occasion he rose from the bed to use the bathroom but apparently passed out before reaching the bathroom, he was found by family members who heard the fall; there were no stigmata of seizure activity and the patient rapidly recovered. The other episode also took place shortly after rising to cross a room. Upon hearing this, the neurologist ordered an ECG (Fig. 1) which revealed a sinus rhythm with right bundle branch and left anterior fascicular block as well as borderline first degree AV block.

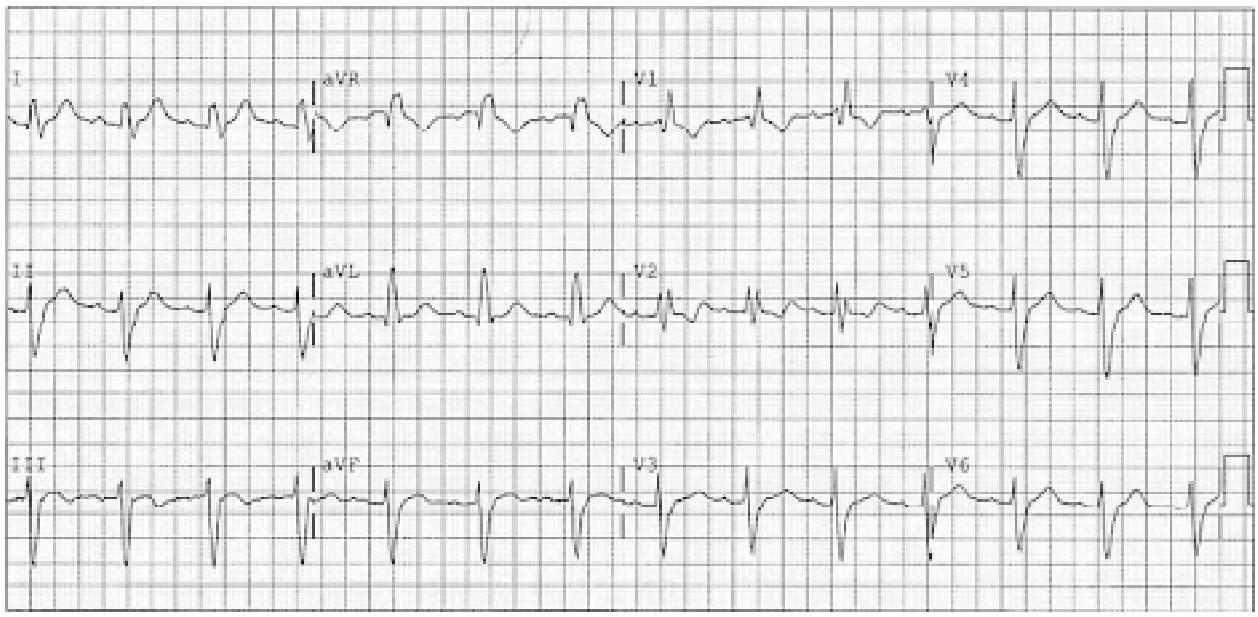

Figure 1. Twelve lead electrocardiogram demonstrating a sinus rhythm with right bundle branch block and a marked left axis deviation consistent with left anterior fascicular block.

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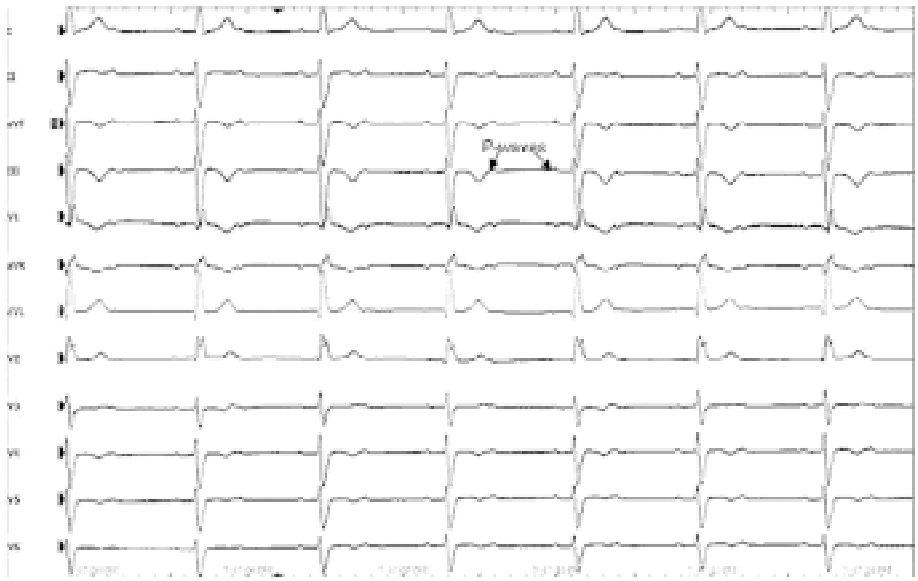

Figure 2. Standard 12 lead ECG displayed at 25 mm/s. Note 2:1 AV block with one P wave located at the terminal portion of the T wave.

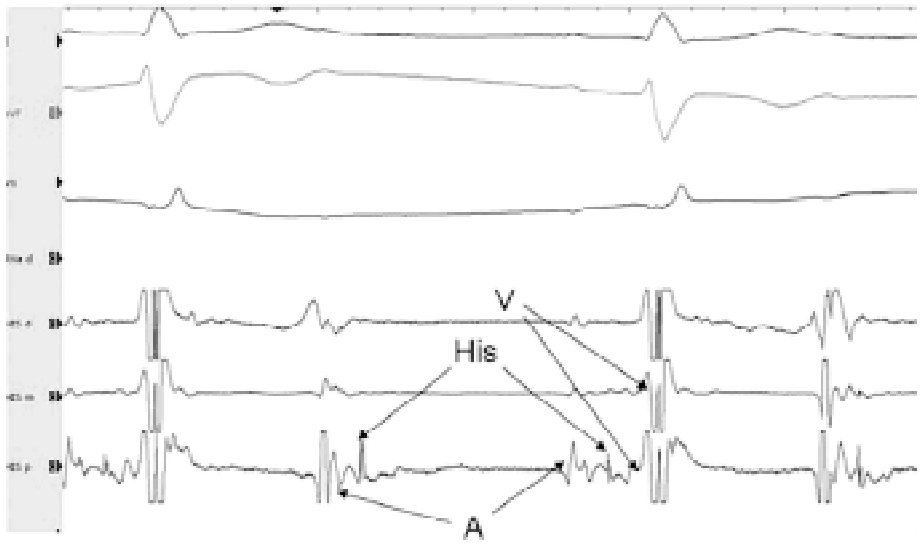

Figure 3. Three surface leads and intracardiac electrograms recorded during sinus rhythm at 100 mm/s. Note that the second atrial electrogram is followed by a His signal which fails to excite the ventricle, proof of block below the His; A — atrial electrogram, His — His electrogram, V — ventricular electrogram.

Given the high prevalence of His-Purkinje disease in patients with myotonic dystrophy the patient was urgently referred to the electrophysiology clinic where a repeat ECG was essentially unchanged. The patient had experienced no further episodes; however a strong suspicion of infranodal disease prompted the scheduling of an EP study.

On the day of EP study the patient presented to the EP lab in 2:1 AV block (Fig. 2), again without further episodes or symptoms. Intracardiac recordings confirmed the presence of infranodal disease as evidenced by infrahisian block (Fig. 3). Furthermore, with atrial pacing, infranodal Wenckebach was noted (Fig. 4). These findings, along with easily inducible ventricular tachycardia prompted the implantation if a defibrillator.

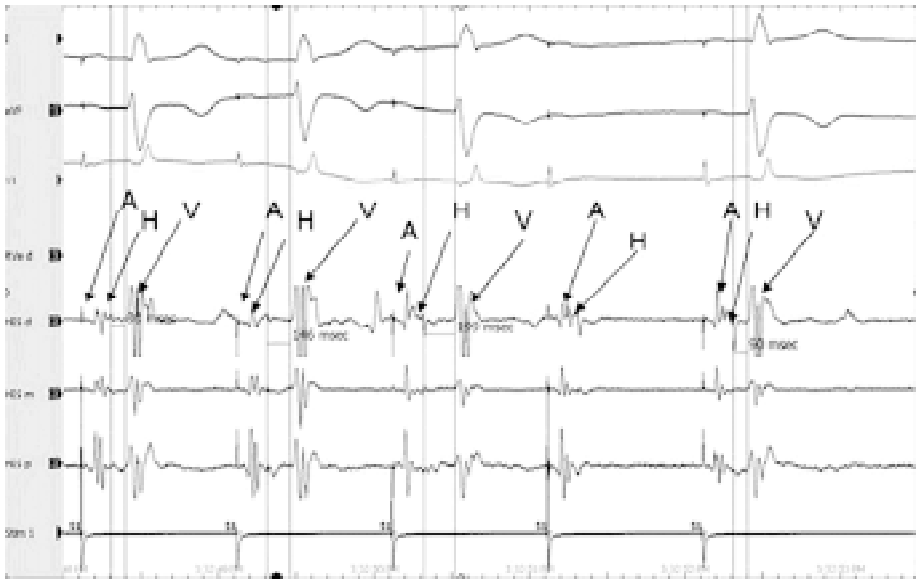

Figure 4. Again, three surface leads and the intracardiac tracings from the His catheter, this time at a speed of 150 mm/s. Note the "Stim" channel at the bottom, indicating atrial pacing (not shown). Note the relatively stable A-H interval indicating normal AV node function at this pacing rate, however the H-V interval progressively prolongs and then "drops" a beat in a Wenkebach pattern; A, H and V are as in Figure 3.

Myotonic dystrophy (type 1) is the most common neuromuscular disease with a prevalence estimated at 1/8000 live births. It is an inherited disorder, passed on in an autosomal dominant fashion with variable penetrance. On a molecular level there is a mutation in the myotonic dystrophy protein kinase gene on chromosome 19 resulting in a variable number of CTG nucleotide repeats. The major clinical features of the disease include myotonia, frontal balding, endocrinopathies, proximal limb and facial muscle weakness, developmental delay and premature cataracts [1]. The cardiac manifestations are preferential infiltration and fibrosis of the conducting system, ventricular tachycardia (particularly bundle branch reentry), syncope and sudden death [2]. Infiltrative cardiomyopathy due to myotonic dystrophy is rare.

The recommendations for the cardiac care of patients with myotonic dystrophy are understandably not as well defined as for those of many other conditions. Current recommendations for patients with myotonic dystrophy call for routine electrocardiographic screening [3]. EP study had been recommended for patients with palpitations, syncope, presyncope or evidence of AV block on the surface ECG. The standard of practice in many labs is to implant a pacemaker for patients with an H-V interval of greater than 70 milliseconds [4]. Unfortunately, pacemakers do not eliminate the risk of sudden death as they do not afford protection from ventricular tachycardia which is also well described in this cohort. The guidelines for dealing with ventricular tachycardia in these patients are less well defined. Radiofrequency ablation of bundle branch reentry (when it is the clinical tachycardia) may be effective in eliminating symptoms, however; in this patient with non bundle branch reentrant ventricular tachycardia ICD implantation is reasonable [4]. Although these patients do not classically manifest a significant cardiomyopathy, it has been described. In these patients, most experts advise standard therapy for non-ischemic cardiomyopathy.

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