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

Ethan Levine, Burr Hall and Daniel Kroening

University of Rochester Medical Center, Division of Cardiology, Rochester, New York, USA

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 myocytes. 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 block and left anterior fascicular block as well as borderline first degree AV block.

![Twelve lead electrocardiogram demonstrating a sinus rhythm with right bundle branch block and a marked left axis deviation consistent with left anterior fascicular block.](image)

Address for correspondence: Ethan Levine, DO
University of Rochester Medical Center
Division of Cardiology
601 Elmwood Avenue, Box 679 C
Rochester, New York 14642, USA
e-mail: ethan_levine@urmc.rochester.edu
Received: 18.11.2006  Accepted: 3.04.2007
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 infranodal block (Fig. 3). Furthermore, with atrial pacing, infranodal Wenckebach was noted (Fig. 4). These findings, along with easily inducible ventricular tachycardia prompted the implantation of a defibrillator.
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.

References