Electrophysiological Studies in Two Patients with Dystrophia Myotonica and Atrioventricular Conduction Block


SUMMARY
Two patients with dystrophia myotonica showed high-grade atrioventricular block. Both underwent electrophysiological studies which revealed sinus and A-V nodal disease with normal intraventricular conduction in 1 case and His-Purkinje conduction disease in the other. Dystrophia myotonica may, therefore, involve all parts of the cardiac conduction system and may affect the generation of cardiac impulses. Pacemaker implantation may be necessary especially if drugs such as procainamide, which in addition to controlling myotonic symptoms may aggravate conduction disorders, are to be used.

Additional Indexing Words:
His bundle electrocardiography

DYSTROPHIA myotonica is a neuromuscular disorder often affecting the myocardium and cardiac conduction system. We report 2 patients with this disorder in whom the surface electrocardiograms showed varying degrees of atrioventricular block. The sites of delay and block differed in the 2 patients.

CASE HISTORIES
1. A.C., a 58-year-old Caucasian gardener was first referred for investigation of muscular weakness in 1973. On examination, he had frontal balding, bilateral ptosis, marked muscle weakness, and wasting without fasciculation. Percussion of thenar muscles produced a myotonic reflex. Testicular atrophy was also present but there were no cataracts. Electromyography revealed the classical high frequency “dive bomber” discharges of myotonia. The electrocardiogram showed sinus rhythm with a prolonged PR interval of 0.47 sec with normal QRS complexes (Fig. 1). In 1976 he was referred for investigation of slow heart rates be-
Between 40–60 beat/min. Twenty four-hour ambulatory monitoring of the electrocardiogram showed sinus rhythm with a prolonged PR interval, intermittent 2:1 atrioventricular block and complete heart block with a junctional escape rhythm. On exercise the heart rate did not exceed 60 beat/min with 1:1 atrioventricular conduction but exercise tolerance was limited by muscle weakness. Atropine, 0.6 mg, intravenously administered during an episode of complete heart block resulted in acceleration of the junctional escape rhythm from 45 to 54 beat/min. The electrocardiographic abnormalities were not accompanied by dizziness, and a pacemaker was not implanted. His symptoms were thought to be due to the muscular weakness.

2. I.S., a 48-year-old Jewish tailor was admitted for investigation of bradycardia and a 2-year history of falls not associated with loss of consciousness. On examination, he had bilateral ptosis, frontal balding, testicular atrophy, and marked muscle wasting and weakness, with a myotonic response to thenar percussion. Electrocardiograms revealed varying degrees of atrioventricular block. During sinus rhythm there was a prolonged PR interval of 0.46 sec with right bundle branch block and a horizontal QRS axis. Atrioventricular block and complete heart block with a slow escape rhythm were also recorded. Electromyography showed the classical high frequency responses of myotonia. He was referred for electrophysiological investigation, the results of which are described below. A permanent
A pacemaker was implanted. Four weeks after implantation he was admitted with failure to pace and a slow ventricular escape rhythm of 25 beat/min. Before a temporary electrode could be inserted, he suffered an episode of ventricular fibrillation which responded to D.C. shock. The permanent electrode was found to have a threshold of 4.7 v and repositioning resulted in satisfactory pacing with a threshold of 0.7 v.

Both patients were studied in the catheter laboratory. Pacing electrodes were introduced via the right femoral vein and positioned in the high right atrium, against the septal cusp of the tricuspid valve and in the right ventricle. Intracardiac signals together with 4 surface leads were recorded at a paper speed of 100 mm/sec. The methods used in our laboratory have been described in detail elsewhere. Overdrive atrial pacing was used to assess sinus node function.

**Results**

1. **A.C.** The sinus rate at the time of study was 49 beat/min with 1:1 atrioventricular conduction (Fig. 2a). The A-H interval was prolonged at

![Diagram](image)

**Fig. 2. Intracardiac electrograms from patient A.C. (a) and patient I.S. (b).**

- **HRAE** = high right atrial electrogram
- **RVAE** = right ventricular apex electrogram
- I, AVF, V1, V6 = standard surface leads
- A = atrial deflection
- H = His bundle deflection
- V = ventricular deflection

(Paper speed 100 mm/sec)
465 msec with a normal H-V interval of 40 msec. Atrial pacing of 55 beat/min resulted in complete atrioventricular block. Sinus node recovery times after pacing at 100, 130, and 160 beat/min for 2–5 min revealed a maximum value of 2.9 sec. Atropine, 0.6 mg, given intravenously resulted in an increase of the sinus rate to 60 beat/min with shortening of the A-H interval and continued 1:1 conduction. Sinus node recovery times were reduced to a maximum of 1.6 sec.

2. I.S. The sinus cycle showed alternation consistent with ventriculophasic arrhythmia in the presence of 2:1 atrioventricular block (Fig. 2b). The A-H interval was normal at 105 msec but the H-V interval was grossly prolonged at 290 msec. Conduction block of alternate impulses was below the site of recording of the His potential. Sinus node recovery times were normal. Atropine, 0.6 mg, given intravenously had no effect on atrioventricular conduction intervals and the sinus rate increased minimally without loss of the ventriculophasic alternation.

Discussion

Disorders of cardiac conduction are common in dystrophia myotonica occurring in up to 68% of cases in 1 series. These abnormalities include prolongation of the PR interval and variable degrees of intraventricular conduction delay. The site of delay cannot be ascertained from the surface electrocardiogram because the PR interval is a measure of total atrioventricular conduction time. Uemura and colleagues reported 3 patients, 2 of whom were found to have prolonged A-H intervals indicating intra-atrioventricular nodal delay, and 1 patient with H-V prolongation indicating delay below the site of His bundle recording. In both patients reported here, there was intermittent high grade atrioventricular block in addition to atrioventricular conduction delay. The site of block was in the same part of the conduction system as the site of delay in both cases.

The risk of syncopal episodes is high in patients with high degree atrioventricular block, especially if the block occurs below the His bundle. Such episodes may be due to intermittent complete atrioventricular block with a slow escape rhythm or ventricular tachyarrhythmias, both of which were observed in our patient (I.S.) with His-Purkinje delay and block. The differing responses to atropine in our patients are consistent with the different levels of delay and block. Atropine shortens the effective refractory period and conduction time of the atrioventricular node, but has no effect on the His-Purkinje system. The prolonged sinus node recovery time in patient
A.C. may suggest the presence of additional sinus node involvement.

These observations indicate the extent of conduction system disease in these patients and is consistent with histological studies\(^{(11,10-13)}\) which have demonstrated varying degrees of involvement of the atrioventricular node, the intraventricular conduction system and the sinus node.\(^{(10)}\) Our experience is similar to that of Uemura et al\(^{(8)}\) whose patient with H-V prolongation and bundle branch block also suffered from intermittent complete atrioventricular block and ventricular tachyarrhythmias. The use of procainamide to treat the symptoms of myotonia\(^{(14)}\) in patients with His-Purkinje involvement may be hazardous because it may have a depressent effect on the intraventricular conduction system.\(^{(15)}\)

The extent of cardiac involvement in patients with dystrophia myotonica may be difficult to determine clinically. Symptoms of weakness and repeated falling may be due to skeletal muscle disease or cardiac involvement. The associated intellectual impairment may make the patient's account of his symptoms unhelpful. In addition, symptoms due to severe skeletal muscle involvement may mask underlying cardiac problems. The progressive nature of this disease requires early recognition of electrocardiographic abnormalities. Ambulatory monitoring of the electrocardiogram and intracardiac electrophography may be of value in assessing the site and severity of conduction system disease. The necessity for permanent pacing should be assessed in relation to the symptoms associated with bradycardia and the extent of conduction system disease. Progressive disease of the His-Purkinje system in patients with dystrophia myotonica may require long term pacing.\(^{(16)}\)

References

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