THE ELECTROCARDIOGRAM IN PROGRESSIVE MUSCULAR DYSTROPHY

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Received February 28, 1958

From a survey of recent reports it is increasingly apparent that myocardial involvement is a fairly frequent sequel in progressive muscular dystropy. It has been estimated that the heart is affected in from 50 to 85 per cent of cases of progressive muscular dystrophy, and more commonly when this is of the childhood type (Berenbaum and Horowitz, 1956). Recent figures, provided by the Muscular Dystrophy Society of the United States have revealed well over 200,000 cases, and since cardiovascular complications are very likely, the problem would appear to be important, particularly in the differential diagnosis of obscure forms of heart disease, now grouped under the term isolated or idiopathic myocarditis. Nothacker and Netsky (1950) reported lesions in the cardiac muscle identical with lesions found in the skeletal muscles. Others (Bevans, 1945; Weisenfeld and Messinger, 1952; Rubin and Buchberg, 1952) have described similar cardiac involvement by the dystrophic process. Histological examination revealed an increase in connective tissue with atrophic muscle cells and fatty replacement, especially in the right ventricle. No descriptions of significant coronary artery disease have appeared. Although pathological studies have shown that the disease is not related to disturbances of blood supply and that the myocardial lesions are very similar to those seen in the skeletal muscle, they have provided little or no information with regard to the ætiology of the dystrophic process.

The clinical manifestations of heart disease in progressive muscular dystrophy are non-specific according to Rubin and Buchberg (1952). These authors and others (Weisenfeld and Messinger, 1952; Berenbaum and Horowitz, 1956) have described tachycardia, cardiac arrhythmias, cardiac enlargement in the absence of the usual causes, congestive failure, and electrocardiographic abnormalities of no specific pattern. Among a number of reports about the cardiographic changes, Weisenfeld and Messinger (1952), in a study of 44 cases, reported that tachycardia, abnormalities of the P wave, and tall R waves in V1 and V2 were of frequent occurrence, and this is the nearest approach to the description of any specific pattern. Rubin and Buchberg (1952) confirmed the high percentage of cardiac involvement in progressive muscular dystrophy, and found that very few of these patients had any clinical evidence of heart disease, although, some did present with congestive failure and others with disturbing arrhythmias and tachycardia: abnormal Q waves and a short P–R interval were the features emphasized in their review of 33 cases. A variety of electrocardiographic abnormalities has been described and these are much more frequent than the clinical manifestations of cardiac involvement, and are likely to precede, often by a considerable time, the appearance of clinical heart disease. This study of 38 cases pays particular regard to the incidence and characteristic features of these abnormalities in progressive muscular dystrophy.

Materials. Through the co-operation and assistance of the Muscular Dystrophy Association of London, Ontario, 38 cases of progressive muscular dystrophy have been studied, 28 in the juvenile group and 10 in the adult or facio-scapulo-humeral group. All had been carefully reviewed by

* This work was supported by the Ontario Heart Foundation and the Defence Research Board of Canada.

the neurological consultants and accepted as progressive muscular dystrophy of the pseudo-hypertrophic or facio-scapulo-humeral types. The two groups are considered separately because of the very obvious differences found in the electrocardiogram of the juvenile as compared to the adult type. Furthermore, such a division appears reasonable when one considers the neurological manifestations of the disease. The juvenile or pseudo-hypertrophic groups show a less localized and more widespread type of involvement and are, therefore, more likely to include the myocardium than the more localized facio-scapulo-humeral groups, which present as a clinical problem later in life.

Results

Pseudo-hypertrophic Muscular Dystrophy. In this group there were 28 cases. At the time of this study, the average age was 16, with a range of 5–37 years. The average age for onset of symptoms was 6 years with a range of 0–14 years. Twenty-two cases were confined to a wheeled chair or very limited walking and six were ambulatory. Chest deformities were not very common, nor of advanced degree, but were present in 9 cases—6 with funnel chest and 3 with scoliosis. Five of the 28 revealed clinical evidence of heart disease or cardiac involvement: in two of them the findings were very suggestive of associated congenital heart disease, and the remaining three gave a history of dyspnœa, and one of these had congestive failure (Fig. 1). Systolic murmurs were present in eight cases. Blood pressure was found to be within the normal range with one exception where it was 154/100. The heart rate was from 65 to 135 and contrary to previous reports tachycardia was not a feature in this series, though five were slightly above the upper limits of normal for their age.

Although there were a number of T wave abnormalities of a non-specific type, from flat to negative in the limb and lateral præcordial leads, the most prominent abnormality was found in the R wave amplitude in lead V1 and in the presence of abnormally deep Q waves in the limb and lateral chest leads (Fig. 2). Contrary to previously reported studies we found no significant variation from the normal in the P waves, P–R interval, cardiac rhythms, QRS, or Q–T duration. Since one expects large R waves in the younger age groups a very careful study of the Q and R V1 wave amplitudes was carried out in accordance with the standards described by a number of authors (Lepeschkin, E., 1951; Ziegler, 1951; Amer. H. Assoc., 1956; Nadas, 1957) (Fig. 3).

In these twenty-eight cases the following electrocardiographic features were noted.

The rate and rhythm were unchanged in most patients, but one had nodal rhythm, four ventricular extrasystoles, and one a shifting pacemaker.

The P wave amplitude was high in six cases and low in one; the remainder were normal.

The P–R interval and QRS duration were within normal limits in all of these cases.

Axis deviation occurred in eight cases, being left in four and right in four.

In 20 of the 28 cases, abnormally deep Q waves were present in the limb and/or lateral chest leads, V4, V5, and V6.

All had tall R waves in lead V1, 20 showing abnormally tall R waves, with an additional three at the upper limit of normal, in this lead (Fig. 3).

The R wave duration in lead V1 ranged from 0·02 to 0·08 sec., being 0·02 in one, 0·03 in eleven, 0·04 in ten, 0·05 in one, 0·06 in three, and 0·08 in two cases.

An RSR′ pattern in lead V1 was observed in four cases.

Low voltage was observed in three cases in the limb leads.

Most cases showed no variation in the S–T segment but this was depressed in lead V2 in one and elevated in V2 in one. S–T depression was noted in leads III and aVF in one other case.

T wave abnormalities ranging from flat to negative waves were noted in the limb and/or lateral V chest leads in 13 cases. A Q1T1 pattern occurred in two and a QIIITIII pattern in two.

In 25 cases U waves were present, of which all were positive and 9 very prominent.

To sum up the electrocardiographic findings. Twenty-five of the 28 were considered to be abnormal, two questionable (possibly normal), and only one completely normal in all respects.

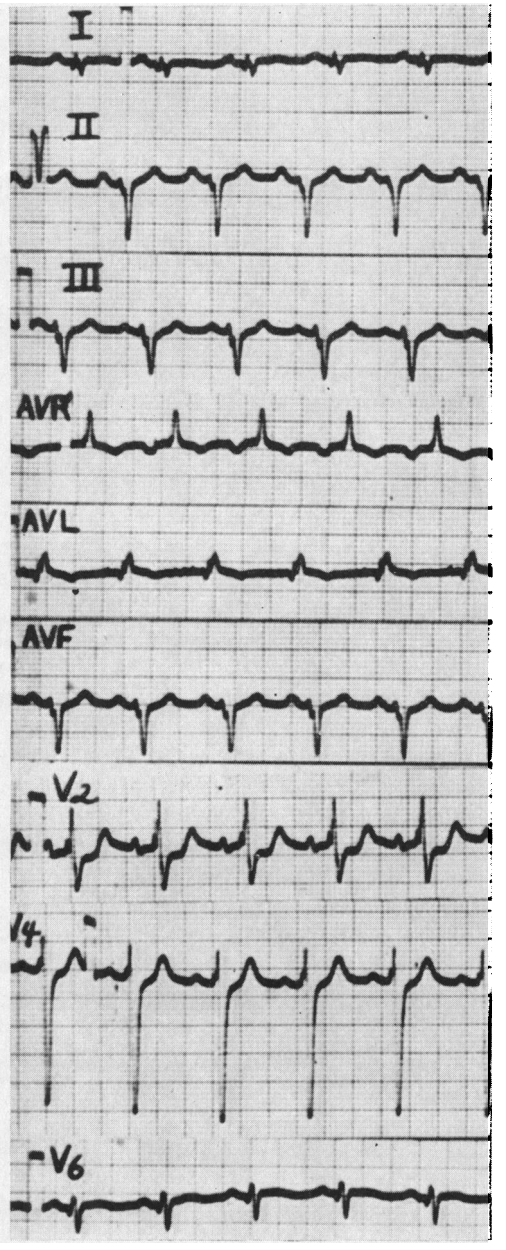

FIG. 1.—Man, aged 29, with classical pseudo-hypertrophic muscular dystrophy. Admitted with congestive failure, with cardiac enlargement, a mid-diastolic gallop rhythm, but no cardiac murmurs.

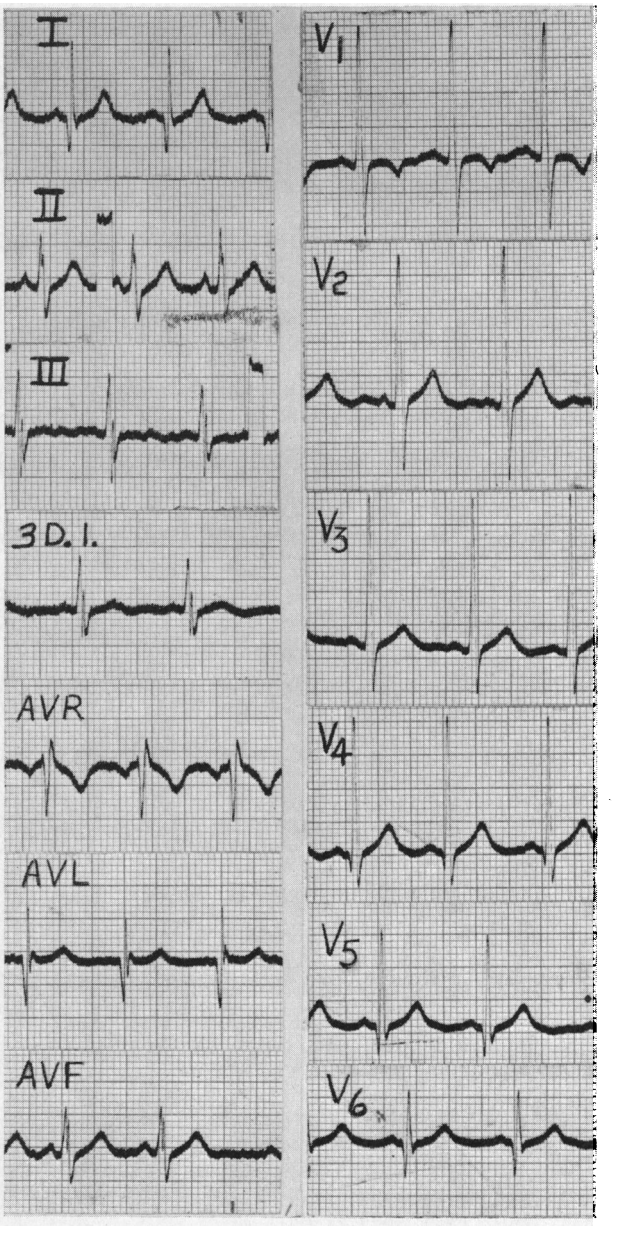

FIG. 2.—Boy, aged 13, with classical pseudo-hypertrophic muscular dystrophy. No clinical cardiac manifestations. Note deep Q waves in leads I and V6 and tall R in lead V1, typical of this series.

The major and most consistent finding was the presence of abnormally deep Q waves and abnormally tall R waves in lead V1.

Facio-scapulo-humeral Group. In the ten cases the average age at the time of this study was 44 years (range 14–67 years). The average age of onset of symptoms was 16 years. Five patients were

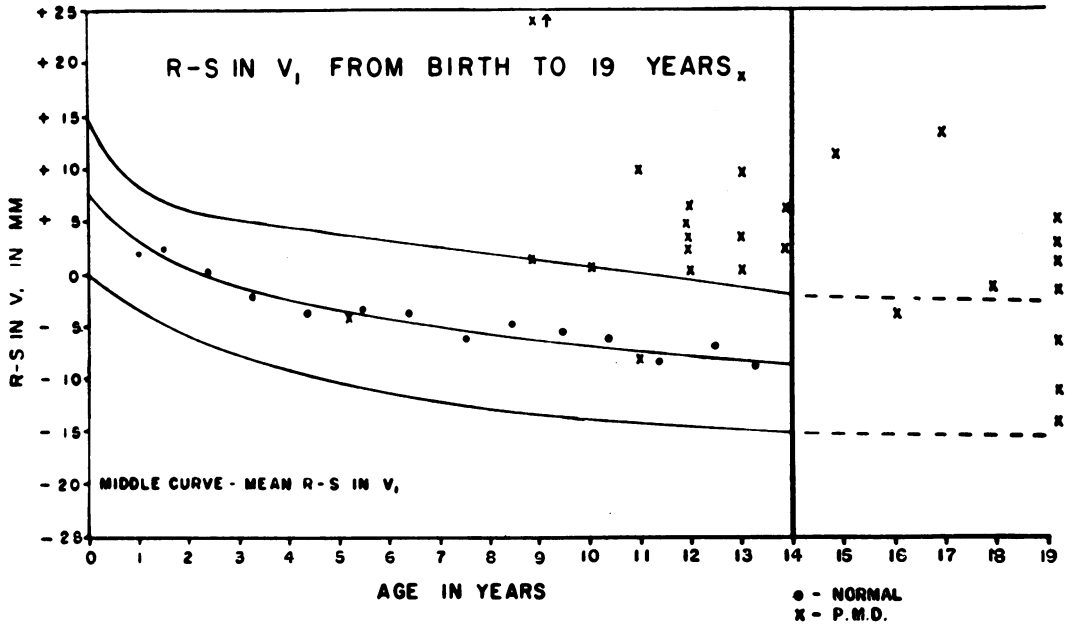

FIG. 3.—The algebraic sum of R and S waves in lead V1 for the normal (Nadas, 1957) and for twenty-eight cases of pseudo-hypertrophic muscular dystrophy from this study.

ambulatory and five confined to bed or wheel chair. There were five examples of left axis deviation in the electrocardiograms of this group. There were, however, only two cases with significant cardiographic abnormalities. One had rheumatic heart disease with mitral stenosis and auricular fibrillation, and the other, a man aged 67, showed classical left bundle-branch block, and second degree A-V block which was quite likely due to coronary sclerotic heart disease. The high incidence of large R in V1 and deep Q waves noted in the juvenile group was not apparent. There was only one case in which the R in V1 approached the upper limit of normal, and this occurred in a boy aged 14, the youngest patient of the group. One woman, aged 67, presented deep Q waves in lead III and AVF, together with well-marked left axis deviation.

DISCUSSION

From previous studies it is apparent that muscular dystrophy may involve the myocardium as well as the skeletal muscle. The two most commonly recognized types of progressive muscular dystrophy are the pseudo-hypertrophic form of childhood and the facio-scapulo-humeral type with onset at puberty. According to published reports the incidence of cardiac manifestations varies considerably, electrocardiographic changes being the earliest and most common manifestation. Walton and Nattrass (1954) in an excellent review of the myopathies reported normal findings in 36 out of 48 cases of the Duchenne type of muscular dystrophy, but only the standard limb leads and lead CF4 were recorded. In our study over 70 per cent of the juvenile pseudo-hypertrophic group revealed abnormal electrocardiographic patterns of a specific nature as compared with the facio-scapulo-humeral group who showed no such patterns and, in fact, little or nothing abnormal. In our series of cases the most interesting and significant findings were confined to the Q waves, and the R wave amplitude in lead V1 (Fig. 2).

From this study it is apparent that the earliest cardiac manifestations in the pseudo-hypertrophic cases occur in the electrocardiogram (Fig. 2 and 3). Furthermore, no such changes were noted in the facio-scapulo-humeral group. In view of the current clinical and pathological concept of these two groups of progressive muscular dystrophy it seems reasonable to expect myocardial

changes more frequently and earlier in the pseudo-hypertrophic juvenile group. The findings in this study are in keeping with this view, indicating more widespread disease involving the myocardium in the juvenile group. Furthermore, in view of the high incidence of abnormally tall R waves in lead V1 (and deep Q waves) it would appear that the electrocardiograph should be of value in the diagnosis of muscular dystrophy and also in differentiating the two groups.

Although no pathological studies have been carried out on these cases there appears to be no reason to suspect right ventricular hypertrophy as the mechanism producing tall R waves in lead V1. A more reasonable explanation would appear to be associated with myocardial damage disturbing the initial QRS vector. The major abnormality appears to be a persistence of an infantile vector loop pattern which in the age groups studied is abnormal by all the accepted standards (Fig. 3).

Summary

Abnormally deep Q waves and tall R waves in lead V1 were present as a specific feature in 70–75 per cent of the electrocardiograms in 28 cases of pseudo-hypertrophic muscular dystrophy.

In ten cases of facio-scapulo-humeral muscular dystrophy the Q and R V1 patterns were normal.

Clinical manifestations of cardiac disease were rare in this study.

The specific electrocardiographic abnormality in pseudo-hypertrophic muscular dystrophy appears to be the persistence of an infantile QRS pattern.

Sincere appreciation and thanks are expressed to the Muscular Dystrophy Association, London, Ontario, for their co-operation and assistance in providing patients for this study.

We are indebted to W. B. Saunders, Philadelphia, for Fig. 3, which is a modification of a diagram illustrating the normal R and S amplitudes from Nadas (1957).

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