Electrocardiograms of Ninety Patients with Acrosclerosis and Progressive Diffuse Sclerosis (Scleroderma)

By John H. Windesheim, M.D., and Thomas W. Parkin, M.D.

The records of all patients with scleroderma seen at the Mayo Clinic during the years 1949 through 1953 were reviewed. There were 90 who had electrocardiograms available for study. The diagnosis in 63 patients was acrosclerosis and 27 were diagnosed as having progressive diffuse sclerosis.

Although Heine\(^1\) described the first case of sclerodermatous heart disease in 1926, this entity did not seem to be well established until 1943, when Weiss and associates\(^2\) described and discussed the problem. Since then many articles have appeared in the literature pertaining to the clinical and pathologic aspects of the disease process known as "scleroderma." In addition, the electrocardiographic abnormalities that may occur have been described in detail. In our review of the literature it became apparent that most of the electrocardiograms were from patients with far-advanced scleroderma who died as a result of this disease process. In many instances the clinical picture indicated cardiac involvement because there were signs and symptoms of congestive heart failure. Necropsy studies have permitted a clinicopathologic correlation between the electrocardiographic patterns and the disease process within the heart. It therefore seemed to us that this particular aspect of the problem had been well documented.

However, we have been unable to find many articles pertaining to the incidence of normal and abnormal electrocardiograms in a large series of patients with all stages of scleroderma but particularly those with acrosclerosis. Before proceeding with a report of our study we should like to summarize our review of the literature with regard to the electrocardiogram in patients with scleroderma.

Review of the Literature

Weiss and associates\(^2\) discussed 9 cases with cardiac involvement. Electrocardiographic tracings were recorded in 8 of these patients and all were found to be abnormal. There were 3 tracings showing premature ventricular beats, 3 with low voltage, 2 with "partial bundle-branch block," 2 with left bundle-branch block, 2 with left ventricular hypertrophy, and 1 with atrial fibrillation. The electrocardiogram of Mathisen and Palmer's\(^3\) patient with proved cardiac involvement showed extrasystoles, left axis deviation, atrioventricular conduction of 0.24 second, slurred low-voltage QRS, and diphasic T with depressed RST in lead I. East and Oram's\(^4\) patient also had abnormal electrocardiographic findings with widened P-R interval, occasional atrial premature beats, and slurred widened QRS complex. Complete heart block and ventricular extrasystoles later developed.

Another case of sclerodermatous heart disease with necropsy findings was described by Spain and Thomas.\(^5\) The only abnormal electrocardiographic findings were a P-R interval of 0.21 to 0.28 second and a QRS interval of 0.08 to 0.11. Gil,\(^6\) in a study of 8 patients with sclerodermatous heart disease, found electrocardiographic changes in 7.

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TABLE 1.—Age and Sex Distribution of Ninety Patients with Acrosclerosis and Chronic Progressive Sclerosis (Scleroderma)

<table>
<thead>
<tr>
<th>Age, yr</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Number</td>
<td>Per cent</td>
<td>Number</td>
</tr>
<tr>
<td>0-9</td>
<td>0</td>
<td>0.0</td>
<td>1</td>
</tr>
<tr>
<td>10-19</td>
<td>1</td>
<td>3.4</td>
<td>1</td>
</tr>
<tr>
<td>20-29</td>
<td>0</td>
<td>0.0</td>
<td>8</td>
</tr>
<tr>
<td>30-39</td>
<td>4</td>
<td>13.8</td>
<td>16</td>
</tr>
<tr>
<td>40-49</td>
<td>6</td>
<td>20.7</td>
<td>15</td>
</tr>
<tr>
<td>50-59</td>
<td>8</td>
<td>27.6</td>
<td>13</td>
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<tr>
<td>60-69</td>
<td>9</td>
<td>31.1</td>
<td>7</td>
</tr>
<tr>
<td>70-79</td>
<td>0</td>
<td>0.0</td>
<td>0</td>
</tr>
<tr>
<td>80-89</td>
<td>1</td>
<td>3.4</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>29</td>
<td>100.0</td>
<td>61</td>
</tr>
</tbody>
</table>

Youngest 9 yr. 9 yr. 9 yr.
Oldest 87 yr. 69 yr. 87 yr.
Mean 53 yr. 43 yr. 46 yr.

There was incomplete right bundle-branch block in 3, complete right bundle-branch block in 1, and "myocardial changes" in 3. Hutcheson\(^7\) stated that, of 8 cases of scleroderma studied, all had clinical or laboratory evidence of cardiac involvement. One patient showed right bundle-branch block, and another had low voltage and premature ventricular beats. True love and Whyte\(^8\) thought that the heart was probably involved in 1 of their patients. The electrocardiogram showed negative T\(_2\) and T\(_3\); and R\(_4\) varied from one complex to another, sometimes well marked, at other times, nonexistent. Hurly and associates\(^9\) presented 1 case with marked congestive failure secondary to scleroderma-tous involvement of the heart. The electrocardiogram showed low voltage in the limb leads, first-degree heart block, low R waves in leads V\(_1\) to V\(_4\), and low T waves in the precordial leads. In a later tracing there was disappearance of the R wave in V\(_1\) to V\(_4\) and depressed S-T segment in V\(_5\) and V\(_6\).

Three cases of scleroderma with involvement of the heart, proved at necropsy, were discussed by Goetz.\(^{10}\) The electrocardiogram of one patient showed atrial fibrillation with multifocal extrasystoles and low voltage, the second had inverted T\(_2\) and T\(_3\), whereas the third patient had an electrocardiogram within normal limits.

Barritt and O'Brien\(^{11}\) reported 2 cases with involvement of the heart by generalized scleroderma. One patient had cardiac failure and possible pericardial effusion, but the other had no cardiorespiratory symptoms or signs. The electrocardiogram of the former demonstrated depressed S-T segments over the left ventricle with flat T waves. That of the latter patient showed inverted T waves in V\(_1\) to V\(_4\).
Another case was presented at a clinicopathologic conference in the Barnes Hospital. The electrocardiogram showed only left ventricular strain and sinus tachycardia, although marked myocardial fibrosis was found at necropsy.

Beigelman and associates noted no diagnostic electrocardiographic changes in 14 patients with progressive systemic sclerosis, although all had some cardiorespiratory symptoms. They stated that most findings were nonspecific, including T-wave inversion, prolonged Q-T interval, low voltage, and various arrhythmias, principally premature beats.

Serial electrocardiograms in another patient with generalized scleroderma showed low voltage, supraventricular premature beats, and transient episodes of right bundle-branch block.

Mustakallio and Sarajas collected electrocardiograms in 7 cases of scleroderma without regard to their cardiac status and found only nonspecific changes. They concluded that, since none of their cases showed low amplitude of multiple leads, the sclerodermatous process, if present in the heart at all, would not be very extensive.

Boyd and associates reviewed 63 cases of scleroderma. The electrocardiographic studies in an unspecified number of these patients showed both right and left axis deviation and frequent conduction defects, and in several instances bundle-branch block was present.

In a recent review by Leinwand and associates of a large group of patients with scleroderma, abnormal electrocardiograms were obtained in 5 of 51 patients so examined and 20 showed minor changes of possible significance. It was noted that postmortem examination of the heart showed more extensive changes than anticipated, even in patients with normal electrocardiograms. There were few patients who did not reveal some charac-
ELECTROCARDIOGRAMS IN ACROSCLEROSIS AND SCLEROSIS

Fig. 1. Electrocardiograms of 3 patients with acrosclerosis. That of patient 1, who also had mitral stenosis, reveals atrial fibrillation. The tracing of patient 2 is indicative of left ventricular hypertrophy and that of patient 3 reveals a minor intraventricular conduction defect with low-amplitude T waves.

METHODS

We reviewed the records of all patients with scleroderma seen at the Mayo Clinic during the years 1949 through 1953. All cases were included whether or not the patients had symptoms or signs referable to the cardiovascular system.

The records of 205 consecutive patients were reviewed. Each patient had been seen by a member of the Section of Dermatology or of the section dealing with peripheral vascular diseases or of both. The records of 105 patients indicated that no electrocardiogram had been taken. There remained a group of 100 patients who had electrocardiograms available for study. The diagnosis in 63 patients was acrosclerosis. Of the remaining 37 patients, 27 were diagnosed as having progressive diffuse sclerosis and 10 were not used in our survey because of some unusual features about the problems that left the final accurate diagnosis in doubt.

A number of features of each patient’s problem were noted and given consideration in order to evaluate changes in the electrocardiogram more accurately. The age and sex distribution is listed in Table 1. Special note was made of abnormalities on chest roentgenograms, the arterial blood pressure, history of Raynaud’s phenomena and the duration of such, if present, symptoms and signs referable to the cardiovascular system, ingestion of such drugs as digitalis and quinidine at the time when the electrocardiograms were taken, and the presence of cerebral, pulmonary and esophageal involvement.

All of the patients had electrocardiograms consisting of standard leads I, II, and III and precordial leads V1, V6, and Vn. A few also had the unipolar extremity and precordial leads V1 through Vn.
Clinical data pertaining to the 8 patients with abnormal electrocardiograms are shown in table 3. Five of these patients (cases 1, 2, 3, 4, and 5) were thought to have acrosclerosis. Their electrocardiograms are shown in figures 1 and 2. There was nothing at the time of the clinical examination to suggest sclerodermatous heart disease, but 1 of the patients had mitral stenosis, which could of course account for the atrial fibrillation. Three patients (cases 6, 7, and 8) had progressive diffuse sclerosis. Their electrocardiograms are shown in figure 3. There was clinical evidence to suggest cardiac failure in 2 of them (cases 6 and 7). These 2 patients had electrocardiograms showing low-amplitude QRS complexes with gross T-wave abnormalities in multiple leads, which usually indicate the presence of an extensive cardiac disease process.

**RESULTS**

Eighty-two patients had normal electrocardiograms, and 8 tracings were considered abnormal. Only 5 of the 63 patients with acrosclerosis and 3 of the 27 patients with progressive diffuse sclerosis had abnormal electrocardiograms. The abnormalities are listed in table 2. It is to be noted that such patterns can be found in a wide variety of cardiac diseases and that in themselves they are not really pathognomonic of a specific heart lesion and must be interpreted with respect to all the aspects of the patient’s problem.

**DISCUSSION**

When one reviews the literature on scleroderma, it is immediately apparent that many aspects of the problem remain unknown. This applies particularly to the etiology of the disease. Another problem that appears pertains to the terminology used in the literature. Such terms as "sclerodactyly," "scleroderma," "acrosclerosis," "acroscleroderma," "generalized scleroderma," "diffuse scleroderma," and "progressive systemic sclerosis" are used. There are different opinions as to whether one is justified in separating the patients into different categories. The different classifications may be simply a matter of definition and the patients may have the same disease but simply different degrees thereof. Nevertheless, we separated our patients into 2 groups, acrosclerosis and progressive diffuse sclerosis. The term "acrosclerosis" refers to those patients with or without Raynaud’s phenomena who have scleroderma limited to the face, chest, and extremities. When the disease process extends over wide regions of the body, the term "progressive diffuse sclerosis" is used. All but 3 of the 58 patients with acrosclerosis and normal electrocardiograms had Raynaud's phenomena. Of the 5 patients
with acrosclerosis and abnormal electrocardiograms, 4 had Raynaud’s phenomena. There were 3 instances of incomplete right bundle-branch block of the type belonging to group I (as defined by Barker and Valencia18) in the patients with acrosclerosis. Such incomplete right bundle-branch block patterns do not necessarily represent organic heart disease, for the pattern might be due to physiologic late activation of the base of the right ventricle or crista supraventricularis.

There was no significant correlation between the known duration of scleroderma and the types of electrocardiographic patterns. It might be noted that the known duration of the scleroderma in the 2 patients with electrocardiographic patterns indicating old infarctions of the anterior wall of the left ventricle was 8 months and 2 years. We use the term “infarction,” realizing however that such patterns simply indicate a loss of viable myocardium (transmural) beneath the exploring electrodes. It is known that several disease processes affecting the myocardium other than infarctions due to insufficiency of the coronary circulation can produce similar patterns; for example, amyloidosis, hemochromatosis, and scleroderma. Without gross and microscopic examinations of the heart we cannot be certain about the exact lesions present.

There were 27 patients classified as having progressive diffuse sclerosis. Fourteen of the 24 patients with normal electrocardiograms had Raynaud’s phenomena.

The sclerotic process is known to affect many organ systems of the body such as the skin, lungs, heart, gastrointestinal tract, and kidneys. The clinical course and manifestations are varied. It is well established now that the heart may be affected and some patients can die as a result of this involvement.

The pathologic changes characterizing sclerodematous heart disease have been described many times.10, 13, 14, 19, 20 It is not our intention to discuss such pathologic changes, but
a few remarks in this regard seem appropriate. The pericardium and myocardium may show varying degrees of fibrosis. The increase in connective tissue in the myocardium may vary from slight interstitial cellular fibrosis to large dense (hyaline) scars. It is of interest to note that the pericardium may be affected as well as the myocardium.19, 21

Such lesions in the heart should theoretically produce a variety of changes in the electrocardiogram22 depending upon the location and extent of the pathologic process. However, one would expect a number of different types of other disease processes that affect the heart to be capable of producing the same types of electrocardiographic abnormalities.

From the review of the literature it is apparent that the electrocardiograms of patients with scleroderma may be normal or may present a variety of abnormalities, such as arrhythmias, a decrease in amplitude of the QRS complexes in multiple leads, depression of S-T segments, decrease in amplitude or inversion of T waves, and disturbances in atrioventricular and intraventricular conduction. It is also to be noted that postmortem examinations have revealed sclerodermatous changes in the heart in patients with normal electrocardiograms.17

**Summary**

The electrocardiograms of 90 patients with scleroderma were reviewed. There were 63 patients with atherosclerosis, of whom 5 had abnormal electrocardiograms (8 per cent). These abnormalities consisted of patterns suggesting an anterior wall scar (2), atrial fibrillation (1), left ventricular hypertrophy (1), and a minor intraventricular conduction defect with T-wave change (1). There were 27 patients with progressive diffuse sclerosis, of whom 3 had electrocardiographic abnormalities (11 per cent), consisting of low-amplitude QRS complexes with gross T-wave changes (2), and complete right bundle-branch block (1). The 2 patients with the low-amplitude QRS complexes and T-wave changes in multiple leads had clinical evidence of congestive heart failure.

**Summario in Interlingua**

Esseva scrutiniate le electrocardiogrammas de 90 patientes con scleroderma. Le serie includeva 63 patientes con atherosclerosis, e 5 de istes (8 pro cento) havéva electrocardiogrammas anormal. Le anormalitates consisteva de configurationes que suggereva cicatrice del pariете anterior (2 casos), fibrillation atrial (1 caso), hypertrophia sinistro-ventricular (1 caso), e un minor defecto de conduction intraventricular con alteration del unda T (1 caso). Le serie includeva 27 patientes con progressive sclerosis diffuse. Tres de istes (11 pro cento) havéva anormalitates electrocardiographic, consistente de complexos QRS a basse amplitude con grossier alteraciones del unda T (2 casos) e bloco complete de branca dextere (1 caso). Le 2 patientes con complexos QRS a basse amplitude e alterationes de unda T in derivationes multiple exhibiva evidentia clinica de congestive insufficientia cardiace.

**REFERENCES**

In 1773, John Hunter had his first attack, which was graphically described by his nephew, Everard Home: "While he was walking about the room he cast his eyes on the looking-glass, and observed his countenance to be pale, his lips white, giving the appearance of a dead man. This alarmed him and led him to feel for his pulse, but he found none in either arm; the pain continued, and he found himself at times not breathing. Being afraid of death soon taking place if he did not breathe, he produced the voluntary act of breathing by working his lungs by the power of the will." In 1776 he had a second attack, and when convalescent he visited Bath. Here he was seen by his friend and pupil, Edward Jenner, of Berkeley; and one of the most interesting and sagacious letters of that distinguished man was written to Heberden, giving his diagnosis of John Hunter's case, and suggesting, for the first time, the probable association of disease of the coronary arteries with angina pectoris.—WILLIAM OSLER. Lectures on Angina Pectoris and Allied States, 1897.
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