Vectorcardiographic Findings in Friedreich's Ataxia

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Summary

Cardiac manifestations in Friedreich's ataxia occur frequently. We have studied 17 cases of Friedreich's ataxia, of which 94% showed a disturbance of the repolarization in the scalar ECG, 29% showed evidence of left ventricular hypertrophy. These changes are not specific but typical of Friedreich's ataxia. Vectorcardiograms were taken with the Frank-system. The vectorcardiographic findings showed not only the same changes as seen in the scalar ECG, but furthermore an atypical configuration of the QRS-loop was evident, which was not recognized in the scalar ECG. According to our experience compared with the scalar ECG, the vectorcardiographic investigations can be considered a more useful method in the diagnosis of Friedreich's ataxia.

Additional Indexing Words:
Disturbance of repolarization  Frank method  Negative T-waves

FRIEDREICH'S ataxia is a rare neurologic disease. Its main sign is spinocerebellar ataxia. Thorén1) set up in 1964 following criteria for the diagnosis of Friedreich's ataxia.

1. Progressive ataxia of chiefly spinal type, with onset in childhood or youth
2. Areflexia of varying degree in the legs
3. Sensory loss
4. Skeletal deformity with pes cavus and/or scoliosis
5. Plantar reflex of extensor type

In the early stage some of these signs may be absent. However, early characteristic signs are the deformity of the feet, pes cavus and a hammer toe.

In 1863, Friedreich mentioned in his original work the cardiac disturbance. It has been known for many years that the cardiac manifestations in

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Friedreich's ataxia occur frequently. According to the literature it is estimated about 50–100%.\(^1\)–\(^3\)

**Material and Method**

We have followed 17 cases (9 male and 8 female patients), some of them over many years. If there was doubt as to the correct diagnosis, the patient was excluded from this study. The youngest patient was 20 years old, the oldest 54 years old. In all cases we could obtain ECG, phonocardiogram, and special chest X-ray films for the heart. The vectorcardiogram was taken in 9 cases with the Frank-method. The right heart catheterization was performed in 4 cases, the left heart catheterization and angiocardiography in 2 cases.

**Results**

1. **ECG findings**

   In scalar ECG (I, II, III, aVR, aVL, aVF, V₁–V₆, and additional special leads), 1 of the 17 cases had a complete normal tracing. This patient showed also no pathologic findings in any other cardiological investigations. In all remaining 16 cases we found electrocardiographic abnormalities. Rhythm disorders were present in 6 cases. Three cases showed sinus tachycardia, 1 case each with atrial fibrillation, atrial and ventricular ectopic beats. Sagittal type was confirmed in 2 cases, signs of left ventricular hypertrophy in 5 cases, signs of right ventricular hypertrophy in 1 case and left anterior hemiblock also in 1 case. The most usual ECG changes were the disturbance of repolarization in the left precordial leads, V₅ and V₆, namely flat, biphasic or inverted T waves with or without ST-depression. By further analysis of the repolarization, we found that 14 of 16 cases showed inverted T-waves, −0.02 to −1.0 mV as follows:

   *Alterations of the repolarization in 16 cases of Friedreich's ataxia*

<table>
<thead>
<tr>
<th>Grade I (flat T-waves)</th>
<th>.................</th>
<th>1 case</th>
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<tbody>
<tr>
<td>Grade II (inverted T-waves up to 0.5 mV)</td>
<td>............</td>
<td>12 cases</td>
</tr>
<tr>
<td>Grade III (negative T-waves 0.5 to 1.0 mV)</td>
<td>........</td>
<td>3 cases</td>
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<tr>
<td>Grade IV (negative T-waves greater than 1.0 mV)</td>
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2. **Vectorcardiographic findings**

   Vectorcardiographic findings are as follows. One of the 9 vectorcardiograms was normal. This was the patient, who had no pathologic findings either in ECG or in other cardiological investigations. The remaining 8 vectorcardiograms were abnormal. In 3 cases we found the signs of left ventricular hypertrophy without any other additional changes. In the re-
maining 5 cases the QRS-loop was atypically configurated, namely 1 case had an initial narrowing. The other case had an initial and terminal narrowing. One other case showed a figure of 8 configuration and the other 2 a kidney form.

In all 8 abnormal vectorcardiograms we could find a pathologic T-loop, which shows the tendency to be directed outside of the QRS-loop. Between the direction of the QRS and T-loop there was in the majority of the cases (7 out of 9) a large difference of both directions (Fig. 1). We can see these changes especially on the horizontal plane. The angle of the QRS-T-vector was also pathologic in 7 out of 9 cases (Fig. 2). The form of the T-loop was oval in 3 cases and extremely narrow in other 6 cases. Two cases of the vectorcardiograms will be demonstrated in Figs. 3 and 4.

**DISCUSSION**

Mollaret\(^4\) reported in 1929 for the first time electrocardiographic abnormalities in Friedreich's ataxia. In recent years the cardiac manifestations in Friedreich's ataxia were more and more emphasized.\(^1,2,7-9\) Generally,
the ECG showed the most pathological findings among the various cardiac investigations. Using 12 standard ECG leads Boyer and co-workers found in 1962 the pathological ECG changes in 55% of 22 patients, Thorén in 1964 in 92% of 49 cases, Graham in 1972 in all of 25 cases (children). Electrocardiographic findings include rhythm disorders (sinus tachycardia, fibrillation, supraventricular or ventricular ectopic beats), left or right ventricular hypertrophy, prominent Q wave and especially disturbance of the repolarization. Among these changes the common findings were the disturbance of the repolarization and signs of left ventricular hypertrophy.
We found in our cases the disturbance of the repolarization in 94% and the signs of the left ventricular hypertrophy in 29%. The prominent Q waves were rare in our cases (12%). So, our results were similar to the findings of Thorén’s cases. The disturbance of the repolarization could be seen in other neurologic diseases, for instance, amyotrophic lateral sclerosis or progressive muscular dystrophy. Nevertheless, we may say as follows: The above ECG changes are not specific but typical of Friedreich’s ataxia. Therefore, Friedreich’s ataxia should be thought in the differential diagnosis, when the disturbance of the repolarization in ECG exists, in the patients with
Reports on vectorcardiographic findings are very rare until now in the literature. Most authors published only case reports. They showed similar changes as ours, even though an atypical QRS-loop configuration was not yet described.

The reports of pathologic anatomic findings are also rare. Hewer investigated 27 cases post mortem. All heart muscles showed extreme interstitial fibrosis without change in the main coronary artery.

According to our opinion, the main findings of the ECG and vectorcardiogram, namely the disturbance of the repolarization and the signs of the left ventricular hypertrophy, also the atypical configuration of the QRS-loop in the vectorcardiogram would be caused by primary myocardial disease, as was shown in the case of Gach and in our cases with catheterization of the heart and angiocardiography.

**References**