The Hemoglobin E Syndromes. I.
Hemoglobin E in Eti-Turks

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With the technical assistance of Kamil Tanrikulu

HEMOGLOBIN E has been found with variable frequency among peoples of southeast Asia, such as the Thais, the Burmese, the Malaysians, the Indonesians, the Bengalis, and Filipinos and Veddas of Ceylon. Sporadic cases have been encountered in other countries. In 1955 an example of the hemoglobin E trait was discovered among 131 Eti-Turks. In another survey, one instance of sickle-cell-hemoglobin E disease and one of hemoglobin E trait were encountered in one family among 155 Eti-Turks. An extensive study of the above-mentioned family was therefore carried out in Mersin and Kelahmed, a village in Tarsus. Among 19 members examined, two had sickle-cell-hemoglobin E disease, 11 the hemoglobin E trait, three sickle cell trait, and three were normal. This experience prompted us to determine the incidence of hemoglobin E in Eti-Turks, particularly among the inhabitants of Kelahmed.

METHODS

The sickling test of Daland and Castle was used. Paper electrophoretic analysis of hemoglobin solutions was made on a Spinco model apparatus (approximately 8 to 10 Gm. per cent solution of oxyhemoglobin, pH 8.6, ionic strength 0.05, 250 volts, duration six to seven hours).

RESULTS

Blood was obtained from 150 persons comprising two groups. The first group consisted of 100 unselected and apparently normal individuals living in Kelahmed village; their ages ranged from 4 to 76 years. Only 3 to 4 members of each family were investigated. None of these individuals was related to the previously investigated family known to harbor the hemoglobin E gene. The second group consisted of unrelated and apparently healthy people belonging to the Eti-Turk community and living in some cities in southern Turkey. Among these 150 subjects, one instance of sickle-cell-hemoglobin E disease and two individuals with hemoglobin E trait were encountered in the same family. The results of this electrophoretic investigation are summarized in table 1.

The results of this investigation and those of other studies performed previously among Eti-Turks indicate that the incidence of hemoglobin E in Eti-Turks is low, approximately 1.37 per cent (table 2).
Table 1.—The Distribution of Hemoglobin Types in 150 Eti-Turks

<table>
<thead>
<tr>
<th>Hemoglobin types</th>
<th>Number</th>
<th>Per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>104</td>
<td>59.44</td>
</tr>
<tr>
<td>A + S</td>
<td>41</td>
<td>27.3</td>
</tr>
<tr>
<td>A + E</td>
<td>2</td>
<td>1.3</td>
</tr>
<tr>
<td>S + E</td>
<td>1</td>
<td>0.66</td>
</tr>
<tr>
<td>S</td>
<td>2</td>
<td>1.3</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>150</td>
<td></td>
</tr>
</tbody>
</table>

Table 2.—The Incidence of Hemoglobin E in Eti-Turks

<table>
<thead>
<tr>
<th>Group</th>
<th>No. tested</th>
<th>No. with Hgb. E</th>
<th>Per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mersin and vicinity9</td>
<td>131</td>
<td>1</td>
<td>0.76</td>
</tr>
<tr>
<td>Mersin and Tarsus11</td>
<td>155</td>
<td>2</td>
<td>1.27</td>
</tr>
<tr>
<td>Kelahmed village and other cities</td>
<td>150</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>436</td>
<td>6</td>
<td>1.37</td>
</tr>
</tbody>
</table>

COMMENT

The Eti-Turks belong to a separate and distinct community in southern Turkey. Among them the gene for sickle-cell-hemoglobin has previously been reported to be 13.3 per cent, but in this and a recent more extensive survey a higher incidence was found (table 1 in reference 18). On the other hand, the carriers of the thalassemia gene have been encountered infrequently in this community. The present investigation and the other electrophoretic studies performed among Eti-Turks have shown that hemoglobin E occurs next in frequency to hemoglobin S. As was mentioned above, hemoglobin E has been found almost exclusively in peoples of southeast Asia, India and Ceylon. With the exception of Indians, the incidence of hemoglobin S among these peoples is low. Therefore, the occurrence of hemoglobin E among Eti-Turks is of anthropologic significance.

The very low frequency of cDe in Eti-Turks indicates a virtual absence of African ancestry in this community. The other blood group frequencies such as the ABO, MNS, Rh, Lutheran, Kell and Duffy do not differ greatly from the values commonly found in Europe.

Considering all these data, it seems reasonable to postulate that a relationship exists between Eti-Turks and some people of India.

SUMMARY

During an electrophoretic study of the hemoglobin from 150 persons belonging to the Eti-Turk community, two individuals with hemoglobin E trait and one case of sickle-cell-hemoglobin E disease were discovered. The results of this investigation combined with those of other electrophoretic studies performed previously in this community indicate that the incidence of hemoglobin E among Eti-Turks is 1.37 per cent.
The importance of the occurrence of hemoglobin E in this community has been discussed, and a possible relationship between Eti-Turks and some people of India has been postulated.

ADDENDUM

Since submission of this manuscript, we have had the opportunity of estimating the per cent of hemoglobin E component in an individual with hemoglobin E trait reported in this paper, by starch gel electrophoresis. Hemoglobin E was 34.3 per cent, and hemoglobin A was 65.7 per cent.

SUMMARIO IN INTERLINGUA

In le curso de un studio electrophoretic del hemoglobina de 150 subjectos pertinente al communitate del eti-turcos, duo casos del tracto de hemoglobina E e un caso de morbo de hemoglobina E e cellulas falciforme esseva discoperite. Le resultatos del presente investigation, combine con illos de altere studios electrophoretic que esseva previemente effectuate in iste mesme communitate, indica que le incidentia de hemoglobina E inter le eti-turcos es 1.37 pro cento.

Le importantia del occurrente de hemoglobina E in iste communitate es discutite. Un relation possibile inter le eti-turcos e certe populos de India es postulate.

REFERENCES

10. —, and Lehmann, H.: The first observation of sickle cell-hemoglobin E dis-
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