THE SICKLE-CELL PHENOMENON IN SOUTH AFRICA*

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Sickle-cell anaemia was first described in Negroes in 1910, and since then the disease itself and the trait have been intimately associated with Negroes. Thus, Wintrobe,[2] in his definition of sickle-cell anaemia, says that it is 'essentially peculiar to Negroes', while Whitby and Britton,[3] in their definition, say that it occurs 'almost exclusively in Negroes'. Thompson,[4] in his book, says 'with very rare exceptions it (sickle-cell anaemia) occurs only in the Negro race', and Dacie,[5] in his book *The Haemolytic Anaemias*, says 'The sickle-cell phenomenon and sickle-cell anaemia are almost entirely confined to the blood of Negroes'.

One object of this article is to show that the racial incidence of sickling in South Africa does not correspond with the views quoted. At the same time we wish to consider in greater detail the incidence of the condition in South African Indians and the means of preventing it.

At the outset there is a semantic difficulty which requires clarification. What is a Negro? There is no doubt that this term is used in different ways in different places. Even in our laboratory differences of opinion existed. Help was therefore sought in the *Oxford English Dictionary*, which says that a Negro is 'an individual (esp. a male) belonging to the African race of mankind, which is distinguished by a black skin, black woolly hair, flat nose and thick protruding lips'. Further help was obtained from the *Encyclopaedia Britannica*, which, under the heading 'Negro', says: 'the designation of a member of the negroid race, one of the three major groupings of mankind, the others being the caucasoid and the mongoloid'. We can therefore safely conclude that the native people of South Africa, locally called Bantu, are Negroes. We also conclude that many who are called Negroes, especially in the USA, are not Negroes but persons of mixed racial blood.

CLINICAL MATERIAL

King Edward VIII Hospital, in Durban, is a university hospital which caters for a large non-White population. In broad terms it can be said that about one-third of the patients are Indian and two-thirds Bantu, or, to use the words of the *Encyclopaedia Britannica*, one-third are Caucasoid and two-thirds are Negroes. It is a hospital of some 2,000 beds and has about 90,000 admissions annually. In addition some 630,000 persons are treated as outpatients each year. The haematology section of the hospital laboratory receives anything up to 400 specimens each day. This is the source of our material.

RESULTS

Over a period of 10 years, but mainly in the last few years, we have found the numerical and racial distribution of the sickle-cell phenomenon shown in Table I. It should be noted that there was some selection in favour of Indians, because Indian families are easier to trace. Allowing for this, however, we find that sickling occurs about 5 times more frequently in Indians than in Bantu. Thus, despite the fact that we receive vastly more specimens from Bantu than from Indians, we have discovered the sickling phenomenon more frequently in Indians than in Bantu. This surprising discovery led to a consideration of the Indian people of South Africa and the origin and distribution of the sickle gene among them.

TABLE I. RACIAL DISTRIBUTION OF SICKLE-CELL PHENOMENON

Condition	*Indian*	*Bantu*
Sickle-cell anaemia	18	3
Sickle-cell trait	24	1
Total	42	4

DISCUSSION

The Indians in South Africa

The socio-economic situation in Natal in the middle of the 19th century was such that cheap labour was required for the sugar plantations. India satisfied that requirement. A system of indentured labour was organized and the first group of indentured Indians arrived in 1860. Except for a period between 1866 and 1874 they continued to arrive at irregular intervals until 1911. This was the main source of the Indians in South Africa. Others, described as 'passenger Indians' came at their own expense and entered the country under the immigration laws then in force. The vast majority of South African Indians are Hindus, but in addition there are large numbers of Moslems. This division of the Indian people into Hindu and Moslem groups is the most apparent. The Moslems can often be distinguished by their personal appearance and by their dress. Traditionally the men wear the fez and the women wear ejars. The Hindu men, by contrast, do not wear the fez and their women by tradition wear saris. This mode of division, while often precise in particular, is fallacious in general: many South African Moslems are converts from Hinduism, the fez is going out of fashion and many Moslem women wear saris. Furthermore, this classification is not comprehensive, because, in addition to Hindus and Moslems, there are other religious groups, e.g. Christians, Buddhists, Jains and Zaroastrians among the Indians.

A more precise, yet still not completely satisfactory, method of classification is based on language. By this means Indians may be divided into Tamil-, Telugu-, Hindustani-, Gujarati- and Urdu-speaking peoples. The Tamils and the Telugus originated in the South of India. They intermarry freely, there being no social sanction against such intermarriage. They tend, however, to marry into their own groups and often into their own families. Thus, inbreeding often occurs. The Hindustani-, Urdu- and Gujarati-speaking people originated in Central and North India. These, too, tend to marry into their own groups and there is strong social sanction against marriage with either Tamil or Telugu.

In an attempt to pin-point the incidence of the sickling phenomenon by the use of the linguist classification we ran into difficulties. For example, some people who ought to

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speak Telugu, in fact speak Tamil; furthermore, in modern times these Indian languages are giving way to English as the lingua franca of the South African Indians. We then considered using the family name. This also led to difficulties as the Indian system of naming is sometimes difficult, and often impossible, to grasp. Ordinarily a man's children will take their father's name and on marriage the daughters will take the name of their husbands. Thus far the system parallels the usual European system. The sons, however, on marriage may (or may not) use their first names as family names; thus each son may establish a family with a name different from his father's. To add to the confusion, pet-names or even nicknames are sometimes given as official names; sometimes a family name, if it denotes an inferior caste, will be replaced by a more favourable name, and, on being converted to Christianity, it is not uncommon to relinquish the previous name and adopt a biblical name. Nevertheless, in the hope that it might prove useful, we list in Table II the names of Indian families in which we have discovered the sickling phenomenon. Included, also, is an educated guess at the linguistic group and religion.

TABLE II. FAMILIES WITH SICKLING PHENOMENON

Name	*Linguistic group*	*Religion*
Applesamy	Telugu	Hindu
Chengiah	Telugu	Hindu
Chinniah	Telugu	Hindu
Chetty	Tamil	Hindu
David	?	Christian
Genasen	Tamil	Hindu
Govindamah	Tamil	Hindu
Latchigadu	Telugu	Hindu
Lazarus	?	Christian
Lutchman	Telugu	Hindu
Moonsamy	Telugu	Hindu
Nagiah	Telugu	Hindu

It is important to realize that during the period of Indian immigration, both men and women entered the country; indeed, sometimes whole families arrived. This accounts, in part at least, for the low incidence of miscegenation between Indians and the other races already present in South Africa when they arrived. Even at the present time there is no legal sanction against marriage between Indian and Bantu; nevertheless such unions seldom occur. If the present-day South African Indians who show the sickle-cell phenomenon did not receive the gene from Bantu ancestors, the question of its origin arises. After considering the available evidence, including our own observations, we conclude that they did not receive the gene in Africa but that they brought it with them from India. Indeed, in at least one of our cases of sickle-cell anaemia both parents (who incidentally were first cousins) were born in India. We find ourselves in agreement with Lehmann,[6] who postulates that the gene originated in the Middle East and that from this common origin it spread eastwards to India and southwards to Africa. We further believe that the gene whose products we are finding among South African Indians came directly from India during the period of immigration already described.

Clinical Considerations

It seems safe to say that sickle-cell anaemia cannot be diagnosed at the bedside by the ordinary methods of clinical medicine. Certainly none of our cases was diagnosed by these methods. This is completely understandable. Sickle-cell anaemia has been described as a disease of multiple infarcts. As such, an unlimited variety of clinical presentations is possible. Thus, among our cases we have had diagnosed rheumatoid arthritis, non-specific arthritis, peptic ulceration, intestinal obstruction, typhoid fever and, probably most significant of all, neurosis. One patient was under treatment for several months, in the course of which he had two abdominal operations before the true condition was recognized. For diagnosis, therefore, recourse must be had to the laboratory, when, if the condition has been suggested to the pathologist, the problem is easily solved. Sometimes, even if the condition has not been suggested, it can be recognized by finding the characteristic cells in the ordinary routine blood film. If these cells are not found, then often the features of haemolytic anaemia are present, and this would lead to a further investigation of the case, in the course of which the sickling test would be done. The sickle-cell trait can only be recognized in the laboratory and then only when it is specifically sought.

Sickle-cell anaemia may, for practical purposes, be regarded as a disease due to a recessive gene. True, the gene manifests itself in the heterozygote and thus is entitled to be called dominant or co-dominant, but the manifestation is not sickle-cell anaemia, it is sickling of the red cells in abnormal conditions or reduced oxygen tension. Viewed as a recessive gene it is all the more sinister, as the carrier state is not immediately recognizable. This is why it can propagate so readily. In communities where inbreeding often occurs, a social-medical problem is created and this is the situation among the Indians in South Africa. Writing on recessive genes, Payling-Wright[7] says: 'It is this large reservoir of potentially detrimental genes in the general population that provides genetical justification for most of the marriage prohibitions found in Leviticus and for about one-third of those in the table of affinity in the Book of Common Prayer'.

In the light of these clinical and genetical considerations we can say that the sickling phenomenon is far more common than the figures given here would suggest. Furthermore, it is increasing yearly. We would therefore appeal to clinicians to suspect the possibility of sickle-cell anaemia among Indians more often than heretofore, to suspect it particularly in cases of chronic anaemia which do not respond to treatment, and to suspect it in cases of 'neurosis'. We also suggest that relatives of known cases be checked for sickling and advised accordingly.

The Prevention of Sickle-Cell Anaemia

As the gene which determines sickle-cell anaemia is, for practical purposes, recessive, there is no reasonable means of preventing its propagation. All we can hope to do is to prevent two such genes coming together in the same person. Towards this end the following are recommended:

1. All persons found to have sickle-cell anaemia should be informed of the genetic implications of their condition. We have found that this is understandable even by illiterate patients. In addition they ought to be provided with a wristband on which the words 'sickle-cell anaemia' would be inscribed. This simple procedure would save time, money and perhaps unnecessary operation.

2. The relatives of all cases of sickle-cell anaemia should be tested for the sickle-cell phenomenon and, when it is found, the significance of it should be explained. In particular the danger of marrying a near relative should be stressed. Those found with the trait should be provided with a wristband bearing the words 'sickle-cell trait'.

3. A law prohibiting the marriage of closely related persons might be introduced, or any such existing law might be more strictly enforced.

SUMMARY

The commonly quoted racial incidence of sickle-cell anaemia and sickle-cell trait does not apply in Natal. In Natal, both occur more commonly in the Indian than in the Bantu (Negro) population. The origin of the gene for this condition is discussed and it is concluded that it came, not from Africa as might have been supposed, but from India. The difficulties of recognizing the sickling phenomenon are stressed and methods for overcoming them are suggested. Recommendations for the prevention of sickle-cell anaemia are also discussed.

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ADDENDUM

Since this article was accepted for publication a further 7 cases of sickle-cell trait have been discovered in an Indian family named James.

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