have shown that pressure within the carpal tunnel is greatest when the wrist is extended, and this posture is especially associated with such household tasks as polishing, knitting, and scrubbing; indeed, it is these activities that provoke symptoms in patients with acroparaesthesiae. The occasional onset or relapse in pregnancy may be aggravated by narrowing of the carpal tunnel by fluid retention. The mechanical nature of the malady is emphasized further by the frequency with which the dominant hand is first affected, and when this does not obtain other factors such as osteoarthropathy of the wrist, trauma, or the nature of employment may be incriminated. These contributory factors are especially common when men are affected.

When surgery is employed it is most inadvisable to adopt the straight longitudinal incision along the midline of the volar aspect of the forearm. This crosses the joint flexure at right angles and pays no attention to the tension lines in skin. Nearly all the patients are women, and it is a surgical solecism to inflict a scar which will in all likelihood produce a cheloid. It is also a cosmetic impropriety to place such a scar in a position which is readily visible. A skin incision has been described which gives an adequate exposure, leaves a featureless scar, and does not in any way interfere with the function of the wrist or hand.

Most often there are no abnormal findings at operation, but in about a third of the cases the median nerve is swollen for a few inches above the transverse carpal ligament and, since this is immediately reversible, is probably determined by oedema. Less frequently there is evidence of direct compression of the median nerve by the transverse carpal ligament, and it is likely that the flattening of the nerve is caused by the duration and extent of the compression.

An analysis of patients treated by surgery shows that there is invariable immediate and dramatic relief of symptoms and that this is maintained; there is then a diminution or disappearance of any pre-existent neurological deficit as judged by clinical examination and the results of ischaemic sensory test. All returned to normal activity. Minor disadvantages include pain and swelling of the volar aspect of the wrist and aching pain provoked by pressure on the scar or by cold. The former does not last for more than a few weeks, but, because it is at times troublesome, it is best to avoid operating on both sides for bilateral symptoms at the same time unless they are very severe. Two separate operations were done on a number of patients and, after the first, symptoms disappeared on the treated side alone. All of these returned of their own accord for the second operation and this seems sufficient answer to those who maintain that the rest enforced by surgery is responsible for the cure.

It must be admitted that rest by itself often gives relief from all symptoms and leads to the disappearance of a pre-existent neurological deficit. However, none who were treated in this way were able to lead unrestricted lives, and some, despite prolonged inactivity, obtained no benefit and developed a progressive objective disability. In addition, ischaemic sensory tests showed a persistent abnormality in the majority. We consider, therefore, that surgery provides the optimum treatment unless the history is short, the symptoms relatively mild, or the patient is able to afford the luxury of prolonged unemployment.

Summary

The clinical features of 53 cases of acroparaesthesiae are described; dominant symptoms consisting of pain and paraesthesiae are almost invariably referred to the territory of the median nerve in the hand, and in the majority there is a slight objective deficit compatible with a median nerve lesion at the wrist.

Electromyography and ischaemic sensory tests lend support for localization of the lesion at the wrist.

The great predominance of female patients may be explained by the smaller size of the carpal tunnel in women and to occupational factors. In men the onset may be determined by osteoarthropathy of the wrist, trauma, or a change of employment.

Division of the transverse carpal ligament gives immediate and lasting relief of symptoms and improvement of any neurological deficit that may have existed.

In about one-third of the patients there is direct or indirect evidence of compression of the median nerve in the carpal tunnel at operation.

Rest may alone relieve symptoms, but relapse occurs in the majority of patients when they resume a normal life. Some achieve no improvement and develop a progressive neurological deficit.

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SICKLE-CELL–THALASSAEMIA DISEASE IN SOUTH TURKEY

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When both the genes controlling the formation of normal adult haemoglobin (haemoglobin A) are replaced by alleles responsible for the sickle-cell haemoglobin variant (haemoglobin S), sickle-cell anaemia (S.C.A.) results. If only one A gene is thus replaced, normally no disability results, the AS heterozygote being a symptomless carrier of the sickle-cell trait (S.C.T.) (Neel, 1947, 1949; Beet, 1949; Pauling *et al.*, 1949). The thalassaemia gene is not an allelomorph of those responsible for the formation of haemoglobin A and its variants, but its effect is a suppression of haemoglobin A formation. If an AA homozygote inherits two thalassaemia genes a severe anaemia results—thalassaemia major. If he is heterozygous for the thalassaemia gene the condition is very much less severe or even symptomless—thalassaemia minor (Gatto, 1942; Wintrobe, 1942; Dameshek, 1943; Valentine and Neel, 1944; Chini, 1946; Silvestroni and Bianco, 1946a).

If, however, an AS heterozygote inherits a single thalassaemia gene, suppression of haemoglobin A formation with a subsequent preponderance of haemoglobin S may produce a state which resembles S.C.A. This modified S.C.A. is called sickle-cell–thalassaemia disease (S.C.Th.D.) or microdrepanocytic disease (Silvestroni and Bianco, 1946b, 1952, 1955; Powell *et al.*, 1950; Gatto and Purrazzella, 1951; Wasserman *et al.*, 1952; Sturgeon *et al.*, 1952; Neel *et al.*, 1953; Humble *et al.*, 1954; Singer *et al.*, 1955; Edington and Lehmann, 1955).

The cells are small and hypochromic, and there are many target cells among them. The resistance towards lysis by hypotonic saline solution is increased (Caminopetros, 1938). Haemoglobin A may be virtually absent, or may be so decreased that it is difficult to establish its presence by the usual electrophoretic techniques.

Foetal haemoglobin (haemoglobin F) is usually present in S.C.Th.D., whereas it is not found in S.C.T. This haemoglobin is under a genetical control which is independent from that of haemoglobin A and its variants, and also from that of thalassaemia. The persistence of haemoglobin F at high concentration beyond the age of 4 months—at which time it has normally disappeared—is a common feature of thalassaemia major, and F is also often found—though at relatively low concentration—in thalassaemia minor (Liquori, 1951 ; Rich, 1952). When haemoglobin F is demonstrated in an AS heterozygote it usually indicates the inheritance of a thalassaemia gene, and the observation helps towards establishing the diagnosis of S.C.Th.D. Target cells at considerable frequency can be an indication of the presence of the thalassaemia gene. Before they are accepted as such, other conditions associated with target cells must be excluded. The most important are other haemoglobinopathies and severe cirrhosis of the liver. In milder disorders of the liver they may be found occasionally but rarely occur in large numbers, and when they are found in patients with liver disease they coincide with a strikingly increased diameter of the red cells.

We record the occurrence of S.C.Th.D. among the Eti-Turks in Southern Turkey, and in addition report that we have found that the heterozygous possession of a sickling and a thalassaemia gene can express itself either as a severe anaemia closely resembling S.C.A. or as a mild anaemia, or, lastly, that it can fail to give rise to any symptoms. In this latter case it can be discovered accidentally in the course of a survey or when a family of a known S.C.Th.D. patient is studied.

One of us (Aksoy, 1955) found that there was a high sickling incidence in the Eti-Turks, a small Arabic-speaking population enclave near Mersin, in Southern Turkey. We found that in some instances a child suffering from S.C.A. had only one sickling parent. The investigation of such families led to the discovery of the thalassaemia gene among them, and resulted in an alteration of the diagnosis of S.C.A. to one of S.C.Th.D.

Methods

Haemoglobin was determined on a Hellige haemoglobinometer. Reticulocytes and platelets were estimated in a wet preparation, using Dameshek's platelet solution (see Wintrobe, 1951). Osmotic fragility was determined according to Daland and Worthley (see Ham, 1951). The sickle-cell test was performed by incubation of cells with sodium metabisulphite (Daland and Castle, 1948). Foetal haemoglobin was determined according to Singer *et al.* (1951). Target cells were accepted as a possible indication of the presence of the thalassaemia gene only when they were found at considerable frequency, and after the exclusion of other possible causes such as cirrhosis of the liver. Electrophoresis of the haemoglobin was performed on solution freed from stroma by shaking with toluene and subsequent centrifugation. (For details of electrophoretic technique see Lehmann and Smith, 1954.)

Case 1

A 40-year-old housewife, known to be anaemic for several years, was admitted to Mersin State Hospital because of lassitude and gastric discomfort. Some months prior to admission she tired easily and suffered from moderate osteoarticular pains. She also complained of palpitation on exertion and of irregular fever.

Physical examination revealed a well-developed and well-nourished but pale woman—height 5 ft. 7 in. (170 cm.), weight 11 st. 11½ lb. (75 kg.). The sclerae were subicteric. The spleen was barely palpable, liver not enlarged. A systolic murmur was heard over the apex. Blood pressure was 125/85 mm. Hg. There was no adenopathy. *X*-ray examination of the skull showed moderate thickening of the diploe and mild trabecular striation on frontal and parietal bones. There were no abnormalities of long bones and hands. The sickling test was positive. The electrophoretic pattern of the haemoglobin was AS ; haemoglobin F was absent. The haematological findings are summarized in Tables I and II. The direct and the indirect Coombs test were negative. Features suggesting the presence of a thalassaemia gene were the appearance of the red cells, target cells, and lowered osmotic fragility. A family study revealed thalassaemia minor in one sibling and in one daughter, the husband being normal. It was concluded that this was a mild case of S.C.Th.D.

Details of Family Study (see Fig. 1).—The daughter in whom thalassaemia minor was diagnosed was 17 years old:

TABLE I.—*Haematological Data on Five Patients with Sickle-Cell Thalassaemia Disease*

	1	2	3	4	5
R.B.C./10^6 c.mm.	2·40	2·50	2·90	2·55	2·55
Haemoglobin (g. per 100 ml.)	7·5	6·7	7·25	7·5	7·8
W.B.C./c.mm.	9,800	16,000	12,700	7,400	19,000
Reticulocytes (%)	15	35	20	10	13·2
Platelets/c.mm.	946,000	135,000	160,000	357,000	780,000
Packed cell volume (%)	23	23	28	24	24
M.C.V. (cu. μ)	95	92	96	94	94
M.C.H. (%)	31	27	25	29	31
M.C.H.C. (%)	33	29	26	31	32
Neutrophils (%)	46	54	57	63	66
Band forms (%)	2	7	1	8	10
Metamyelocytes (%)	—	2	—	2	2
Eosinophils (%)	23	1	8	1	—
Basophils (%)	1	—	—	1	—
Monocytes (%)	1	7	1	12	4
Plasma cells (%)	—	2	—	—	—
Lymphocytes (%)	27	27	33	12	18
Nucleated red cells/100 W.B.C.	—	4	3	2	11
Anisocytosis	+++	+++	++	+++	+++
Polychromasia	++	+	±	+	++
Poikilocytosis	±	—	—	—	±
Target cells (%)	8·7	18·2	9·6	19	27·8
Sickle ,, (%)	—	2·2	—	0·2	0·4
Sickling test	+	+	+	+	+
Foetal haemoglobin (%)	0	1·3	7·8	5·9	1·9
Osmotic fragility (% NaCl)	0·4–0·2	0·42–0·15	0·40–0·20	0·36–0·16	0·38–0·16
Serum bilirubin (mg./100 ml.)	2·3	3·2	2·6	3·6	3·8
van den Bergh reaction	Indirect	Indirect	Indirect	Indirect	Indirect

TABLE II.—*Differential Counts of Sternal Marrow Smears in Three Patients with Sickle-Cell-Thalassaemia Disease*

	Case 1	Case 2	Case 5
Cellularity	++++	+++	++++
M./E. ratio	1:1·4	1:1	1:0·8
Promyelocyte (%)	—	0·5	0·5
Myelocyte (%)	5	10	14
Metamyelocytes (%)	7	10	11
Band form (%)	12	9·5	20
Segm. poly. (%)	7	16·5	6
Eosinophil myel. (%)	2	—	—
,, band (%)	6	—	—
,, segm. (%)	1	0·5	—
Lymphocyte (%)	—	2·5	—
Reticulum cell (%)	—	—	1
Plasma cell (%)	1	1	5
Proerythroblast (%)	10	2·5	6
Basophilic normoblast (%)	—	10	3·5
Polychromatic ,, (%)	10	18·5	13
Orthochromatic ,, (%)	20	18·5	15·5
Polychromatic macroblast (%)	9	—	2
Orthochromatic normoblast (%)	10	—	0·5
Mitosis (%)	20	15	10

Hb. 10.9 g./100 ml; R.B.C., 4,200,000 per c.mm.; 7.4% target cells; P.C.V., 39%; osmotic fragility, 0.4–0.3% NaCl; haemoglobin F absent; fine osteoporosis of parietal and occipital bones.

Case 2

A 12-year-old boy came to the out-patient department of Mersin State Hospital because of lassitude, irregular fever, and malaise. He had a history of infective hepatitis. He developed normally until 2 years of age, when irregular fever, anaemia, and malaise appeared. Malaria was diagnosed, but treatment with quinine, mepacrine, liver extract, and iron gave no relief. Latterly complaints became more severe.

Physical examination revealed a normally developed well-nourished pale boy. The sclerae were subicteric; spleen not palpable; liver enlarged two fingerbreadths below costal margin. Striking frontal bossing was present. *X*-ray examination of skull showed considerable thickening of diploe and mild striation of frontal, parietal, and occipital bones; there was, however, no "hair-on-end" appearance and the outer table had not disappeared. Long bones showed coarse striation around elbow-joints. The sickling test was positive and the blood smear showed sickle cells. The electrophoretic pattern of the haemoglobin was AS; haemoglobin F was 1.3% (not significant). The haematological findings are summarized in Tables I and II. Features suggesting the presence of a thalassaemia gene were the appearance of the red cells as a whole, 18.2% target cells, and a lowered osmotic fragility. A family study revealed one other sibling with S.C.Th.D. (Case 3) and the presence of the thalassaemia gene in the father, both parents being S.C.T. carriers.

Case 3

A 10-year-old girl, sister of Case 2, was first seen in 1954 suffering from lassitude, irregular fever, malaise, and anaemia. She was treated for malaria. Physical examination revealed a normally developed well-nourished though pale girl. The sclerae were slightly subicteric; spleen three fingerbreadths and liver two fingerbreadths below costal margin. Mild frontal bossing was present. *X*-ray examination of skull showed moderate thickening of diploe and mild trabecular striation around elbow-joints. The sickling test was positive. The electrophoretic pattern of the haemoglobin was AS; haemoglobin F was 7.8%.

The haematological findings are summarized in Table I. Features suggesting the presence of a thalassaemia gene were the appearance of the red cells as a whole, 9.6% target cells, a lowered osmotic fragility, and the presence of haemoglobin F.

Details of Family Study of Cases 2 and 3 (see Fig 1).—Both parents and one sibling were AS heterozygotes. All had less than 1% foetal haemoglobin. The appearance of the father's blood film suggested the possession of the thalassaemia gene (8.6% target cells), but there was no anaemia. The thymol turbidity and the gamma-globulin content of the serum were not raised; the pseudocholinesterase was within normal limits. It was concluded that the father was a symptomless heterozygote for the thalassaemia and the sickling gene. The ABO, rhesus, MNS, Lutheran, Kell, and Duffy blood groups of the parents and their three children were determined by Dr. A. E. Mourant and Miss E. W. Ikin at the M.R.C. Blood Group Reference Laboratory; there was no evidence of non-paternity.

Case 4

A 13-year-old girl was brought to the out-patient department of Mersin State Hospital because of palpitation, lassitude, and irregular fever. She had been ill from childhood and there had been bone pains with irregular fever now and then, but there had been no swelling of joints.

Physical examination revealed a normal stature and a somewhat mongoloid facies. The sclerae were subicteric, and there was pallor with a brown-yellow tint to the skin. There were no leg ulcers or adenopathy. The spleen was two fingerbreadths and the liver one fingerbreadth enlarged below the costal margin. Pulse rate was 180 a minute, regular. A systolic murmur was heard over apex and pulmonary artery area. The second pulmonary sound was accentuated. Blood pressure was 110/80 mm. Hg. There were no abnormalities over the lung. *X*-ray examination of the thorax showed mild left ventricular enlargement, and prominence of the pulmonary artery. E.C.G. showed sinus tachycardia, P-mitrale, left ventricular strain, and heart in vertical position. Frontal bossing was present. *X*-ray films of the skull revealed thickening of the diploe, mild trabecular striation on parietal bone, and some osteoporosis of frontal bone. The sickle-cell test was positive, and the peripheral blood smear showed some sickled cells. The electrophoretic

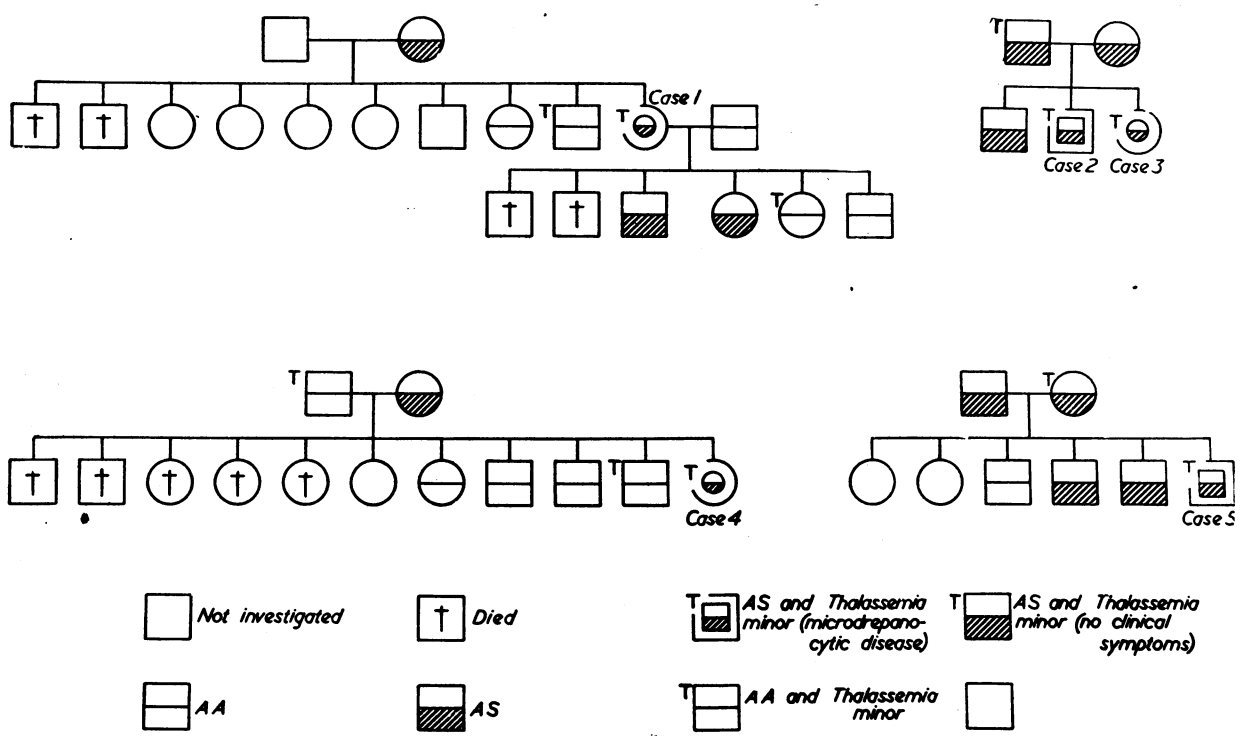

FIG. 1.—Family trees of five patients with sickle-cell–thalassaemia disease.

pattern of the haemoglobin (Fig. 2) showed most of the haemoglobin to be in the S position (80% as determined by densitometry); there was, however, a "trailing" of haemoglobin extending forward to the haemoglobin A position with a slightly greater density towards the A position than in the intervening area. Foetal haemoglobin was 5.9%. The haematological findings are summarized in Table I.

In this case the differential diagnosis was between S.C.A. and S.C.Th.D., as all the features suggesting the presence of a thalassaemia gene could equally well be found in S.C.A.

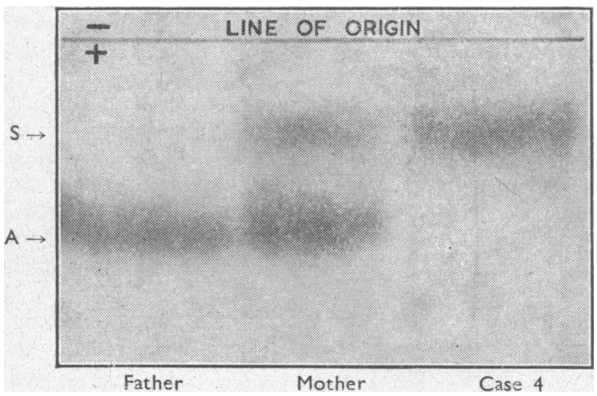

FIG. 2.—Separation of haemoglobin variants by filter-paper electrophoresis. Father, AA. Mother, AS. Daughter (Case 4), sickle-cell–thalassaemia disease.

However, electrophoresis revealed that the amount of non-sickle-cell haemoglobin exceeded the 5.9% haemoglobin F found by chemical estimation, and this suggested the diagnosis of S.C.Th.D. A final proof was brought about by family study, which showed thalassaemia minor in the father (a non-sickler) and in one sibling.

Details of Family Study.—The father, a non-sickler, had thalassaemia minor; Hb, 13.1 g./100 ml.; R.B.C., 4,850,000 per c.mm.; 16.8% target cells; P.C.V. 44%; osmotic fragility, 0.4–0.2% NaCl. *X*-ray examination of skull showed mild thickening of frontal, parietal, and occipital bones, and striation on parietal bones. The mother was a symptomless sickle-cell-trait carrier. Several siblings had died, one sister at the age of 18 years from anaemia resembling in its features that of Case 4. Two siblings showed no abnormality, but one brother aged 15, a non-sickler, showed evidence of thalassaemia minor (7.8% target cells, osmotic fragility 0.4–0.2% NaCl, mild changes in skull and long bones revealed by *x*-ray examination).

Case 5

A 23-year-old man was admitted to Mersin State Hospital with dyspnoea, orthopnoea, cough, and abdominal discomfort. He was diagnosed some three years previously in one of the medical clinics of Istanbul University as having sickle-cell anaemia. All his life he had suffered from irregular fever, bone pains, joint swellings, anaemia, and lassitude. The osteoarticular pains were intolerable, and every antirheumatic treatment was ineffective. Three years ago he had leg ulcers which lasted for about one year. Five months before admission his liver became noticeably enlarged and he had dyspnoea. He improved after rest. One week prior to admission he had three to four days' fever, and the yellow discoloration of his sclerae increased; there was abdominal discomfort and cough.

Physical examination revealed an underdeveloped boy rather than man—height 5 ft. 1 in. (154 cm.), weight 7 st. 11 lb. (49.5 kg.)—with a somewhat mongoloid facies. He was pale and icteric. He was orthopnoeic and sat rigidly because of abdominal pain and distension. His temperature was 101.2° F. (38.4° C.). There was a mild inguinal adenopathy, and four scars of leg ulcers were noted. The area of cardiac dullness extended into the lung fields, the pulse rate was 84 a minute and regular, and a moderate friction rub was noted at the left sternal border. Blood pressure was 105/55 mm. Hg. The epigastrium was distended. The spleen was enlarged to two fingerbreadths below the costal margin, and the liver was tender and extended to the iliac crest. *X*-ray examination of the thorax disclosed an enormous enlargement of the heart; there was no characteristic shape. On screening no pulsations were seen over the heart. *X*-ray films of the skull showed thickening of the frontal, parietal, and occipital bones and mild striation on the parietal bone. The long bones showed coarse striation around the elbow-joint and the femur; the elbow joint had a "honeycombed" appearance. There was osteoporosis of the pelvis, femur, and tibia.

The sickle-cell test was positive, and the peripheral blood film showed some sickle cells. The electrophoretic pattern of the haemoglobin was AS; the foetal haemoglobin was 1.9% (just significant). The haematological findings are summarized in Tables I and II. Features suggesting the presence of a thalassaemia gene were the appearance of the red cells as a whole, 27.8% target cells, traces of foetal haemoglobin, and a lowered osmotic fragility.

Treatment with ouabain, salicylates, antibiotics, mercurial diuretics, low-sodium diet, and oxygen therapy resulted in prompt improvement. The pyrexia subsided and on the fifth day the friction rub was no longer heard. A mild systolic murmur became audible, and the heart silhouette measured radiographically showed a 3.5 cm. diminution in size. Salicylates and antibiotics were discontinued after two weeks. Treatment with digitalis and mercurial diuretics was continued, and the size of the liver diminished to about four fingerbreadths below the costal margin.

Details of Family Study (see Fig. 1).—As in Cases 2 and 3, both parents were AS heterozygotes. Two of the three siblings investigated were also AS heterozygotes. The father, aged 50, showed no other abnormalities in his blood. The mother, aged 40, however, had numerous target cells (10.5%) in her blood smear. Liver-function tests were not carried out. The serum thymol turbidity was not raised, and there was no obvious evidence of liver disease or of any other condition except thalassaemia which could be responsible for this high incidence of target cells. It was concluded that she was a symptomless heterozygote for both the sickling and the thalassaemia gene.

Discussion

The community of the Eti-Turks is small, and it is remarkable that we should have seen no fewer than five cases of S.C.Th.D. over a period of six months. The Eti-Turks resemble in this respect other communities living around the Mediterranean shore. It is well known that thalassaemia minor by itself may be completely symptomless. This variable effect of one thalassaemia gene is well reflected in the varying severity of the S.C.Th.D. seen in our series. Contrary to the findings of the other writers, none of our five patients had a lowered M.C.V., and the M.C.H.C. was significantly lowered in only two. Nucleated red cells were always present in four, and three showed sickled cells in the peripheral smear. Foetal haemoglobin was absent in two, present in traces in one, and significantly raised in two. All had a lowered osmotic fragility of their red cells—a feature both of S.C.A. and of thalassaemia major—but the conspicuous changes seen in the *x*-ray films of the skeleton were similar to those seen in thalassaemia major rather than to the milder ones of S.C.A.

In addition to the five patients, two individuals were discovered who were heterozygous for the thalassaemia and sickling genes but did not seem to suffer from S.C.Th.D. This suggests that the same thalassaemia gene does not suppress the haemoglobin A formation equally in different members of a family, and that its penetrance may possibly depend on the particular A gene an individual carries.

Summary

Five cases of sickle-cell–thalassaemia disease were seen in four Eti-Turk families. In addition two individuals were discovered who were heterozygous for the thalass-

aemia and sickling genes but were not anaemic. The incidence of the haemoglobinopathy among the Eti-Turks is equalled only in Sicily.

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A STUDY OF THE SURVIVAL RATE OF CASES OF SICKLE-CELL ANAEMIA

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It is now generally accepted that the phenomenon of the sickling of red cells at reduced oxygen tensions is due to the presence of greater or lesser amounts of an abnormal haemoglobin within the cells. The production of this haemoglobin is controlled by a gene which is allelic to the gene controlling normal haemoglobin production. The heterozygous inheritance of this sickle-cell gene produces the harmless sickle-cell trait, while the homozygote is manifest as sickle-cell anaemia. This anaemia is characterized by haemolysis with intermittent crises and is usually fatal in childhood. This death in childhood of the homozygotes must remove abnormal genes from the population, and the immediate question arises of how, in the face of this recurrent gene loss, the gene frequency can be maintained at the high levels at which it is found in many African tribes.

The answer seems to lie in the interaction of three main factors. Firstly, recurrent mutations must play some part, though it is highly improbable that they could provide the sole replacement of all the genes lost by childhood deaths. Secondly, it is now well established that the heterozygote sickler enjoys some advantage over the normal in respect of falciparum malaria, though the magnitude and exact mechanism of this effect are not yet clear. Thirdly, there is the fact that while most subjects with sickle-cell anaemia die in childhood, some few do survive to adult life and contribute abnormal genes to the next generation. This paper presents some evidence on this third factor as found in a single African tribe in Uganda.

The older literature on the natural history of sickle-cell anaemia has become largely invalidated by the recent discovery of other genetically controlled abnormal haemoglobins and by the description of microdrepanocytic disease. Both this latter disease and the anaemia often resulting from the double inheritance of two abnormal haemoglobins may produce a clinical picture closely similar to sickle-cell anaemia. In East Africa, however, other abnormal haemoglobins have not been found among the African tribes (Jacob, 1955 ; Roberts and Lehmann, 1955) and thalassaemia has never been detected. The same situation appears to obtain in the Belgian Congo. A high degree of validity can therefore be attached both to clinical reports in children and to simple electrophoretic surveys in adults directed towards defining the survival rate of cases of sickle-cell anaemia in these two areas. Nevertheless, published work has produced somewhat variable estimates of survival rates.

In Kenya, Allison (1954) examined 70 adult sicklers of the Luo tribe by paper electrophoresis and found two cases with the pattern of sickle-cell anaemia. He calculated that these two represented a 35% survival rate to adult life. He later suggested that the survival rate in Africa was more likely to be in the region of 20% on the basis of figures collected from the Musoma tribe of Tanganyika (Allison, 1956). In a survey of the Baamba tribe in Uganda, Lehmann and Raper (1956) found no case showing the electrophoretic pattern of sickle-cell anaemia in a random sample of 478 adults. They conclude "that the survival of sickle-cell homozygotes plays no significant part in the maintenance of the high sickling rate in the Baamba." Workers in the Belgian Congo have expressed the same opinion on the basis of clinical observations in children. The Lambotte-Legrands (1955) state that the great majority of patients die in childhood, and Vandepitte (1955) estimates that only about 1% survive to adult life. It was therefore decided to undertake an electrophoretic survey of adults of the Baganda tribe in an attempt to obtain a more exact figure for the survival rate to reproductive age.

Method

Blood samples were obtained from specimens which had been sent in to the laboratory for routine Kahn tests from the antenatal clinic and from the male V.D. clinic of Mulago Hospital. A few samples were also obtained from mothers bringing their children to the paediatric clinics. Specimens from the Baganda tribe only were examined. In all cases a sickling test was done by the *E. coli* method, and all which were positive were subject to paper electrophoresis by the method already described (Jacob, 1955). An attempt was made to see all cases showing the pattern of sickle-cell anaemia at their next clinic attendance.

Results

Altogether 3,362 specimens were examined, of which 545 were from sicklers and were subjected to electrophoresis. The source of the specimens can be seen from the Table.

Cases 291, 316, and 543 of this series of 545 sicklers showed the pattern of sickle-cell anaemia. These samples were all from the antenatal clinic, and further details are as follows.

Sources of Blood Specimens

Source of Blood	Non-sicklers	Sicklers	% Sickling
Antenatal clinic	2,090	396	15·9
Male V.D. ,, ...	651	126	16·2
Mothers in paediatric clinic ..	76	23	23·2
Total	2,817	545	16·2