

a child who had been treated by direct operation late in the disease, and the other a West Indian with extensive disease extending over nine vertebral bodies. These 21 patients have since readmission recovered and maintained their recovery with the exception of the West Indian, who is completing his treatment. Only one patient in the operative series has died of tuberculosis—a woman who was reported to have developed tuberculous meningitis three years after her discharge from Black Notley. Another patient died of non-specific pneumonia three years after discharge. The mortality rate of less than 2% for this disease on follow-up compares very favourably with the mortality rate before the introduction of antibiotics, which for most centres on five-year follow-up was approximately 30%.

The results obtained by constitutional therapy, antibiotics, and direct operation on the lesion are very reassuring, and patients have been allowed to return to full activities soon after leaving hospital. Children have been permitted to return to ordinary school and to take part in games within a short time. A spinal brace is worn by some patients on discharge, but seldom for longer than four months. A summary of the occupations to which operated patients have returned is shown in Table VII.

TABLE VII.—Particulars Concerning Employment of 120 Patients Treated by Direct Operation for Vertebral Tuberculosis

Children (16)	
Pre-school age .. .. .	2   Attending special school .. 4
Attending ordinary school ..	10
Adults: Full Employment (86)	
Household duties .. .. .	33   Ship's fireman .. .. . 1
Engineering .. .. .	14   Lorry driver .. .. . 1
Office .. .. .	18   Dock labourer .. .. . 1
Salesman .. .. .	2   Butcher .. .. . 1
Carpenter .. .. .	2   Sailor .. .. . 1
Teaching .. .. .	4   Various .. .. . 7
Porter .. .. .	1
Adults: Employed Part-time (9)	
Household duties .. .. .	5   Various .. .. . 4
Adults: Not Employed (9)	
In mental hospital .. .. .	4   Unemployed .. .. . 4
Paralysis due to stroke .. .. .	1

The average duration of treatment of the patients treated by direct operation was 9.7 months. These figures show that, by the direct operative treatment of tuberculosis of the spine, good and relatively speedy results can be obtained, with the early return of the patient to full employment. With a two-rib costotransversectomy approach the operation is safe if done with precision and deliberation, with the aid of preliminary constitutional and antibiotic therapy, and with a sufficient antibiotic cover. By this route an adequate access can be obtained to two or three vertebral bodies. For more extensive lesions the transpleural route advocated by Hodgson and Stock (1956) may well be preferable if facilities are available for its safe performance. After the debridement of the vertebral lesion, whatever may be the route of access, the vertebral defect may be reinforced with cancellous bone chips taken from the posterior part of the iliac crest, though in the smaller lesions this procedure is not strictly necessary. If the transpleural approach has been used, an anterior strut graft may be inserted by the technique of Hodgson and Stock, and by preventing the onset of deformity this may be of great benefit to patients with an extensive lesion.

### Summary

The results are given of a combination of constitutional treatment, antibiotics, and direct operation

for the treatment of certain patients suffering from tuberculosis of the spine. The selection of suitable cases for this form of operative treatment depends, *inter alia*, on the situation of the vertebral lesion. Previous experience had shown that the prognosis was worse for tuberculosis of the lower thoracic spine than for disease in the lumbar region. Experience with the direct operation has been most encouraging; the recovery rate is more rapid and certain, and the return of function is assured in most patients. On the other hand, patients with disease in the lumbar region not operated on also did well. Tuberculosis of the spine can now be rendered a curable disease with a good prognosis.

I am grateful to Sir Heneage Ogilvie, Mr. S. L. Higgs, and Mr. Ronald Reid for their help in this work. I am indebted to Dr. Jack Dixon, whose skilled anaesthesia has made the operation safe, and to Dr. Franklin Wood, whose early interest in the possibilities of bone tomography contributed materially to the success of the operations. I also thank the Editor of the *Annals of the Royal College of Surgeons of England* for permission to use Table I.

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## STANLEYVILLE I AND II

### TWO NEW VARIANTS OF ADULT HAEMOGLOBIN

BY

#### PAULETTE DHERTE

*Belgian Congo Provincial Medical Laboratory,  
Stanleyville, Belgian Congo*

#### J. VANDEPITTE, M.D.

*Department of Bacteriology, Lovanium University,  
Leopoldville, Belgian Congo*

#### J. A. M. AGER, M.B., B.S.

*Department of Pathology, St. Thomas's Hospital, London*

AND

#### H. LEHMANN, M.D., Sc.D., M.R.C.P., F.R.I.C.

*Department of Pathology, St. Bartholomew's Hospital,  
London*

In the course of a survey of haemoglobin types seen in the antenatal clinic at Stanleyville, a number of women were discovered whose red cells did not sickle but whose haemoglobin behaved on electrophoresis and chromatography like a mixture of haemoglobins A and S—that is, the abnormal haemoglobin was haemoglobin D. This was confirmed by solubility tests after Itano (1953) where the solubility of the haemoglobin was that of AD samples and not of AS samples.

In addition two abnormal haemoglobins were found to resemble haemoglobin D in some respects but to differ in others.

### Techniques Employed

Sickle-cell test .. .. .	Daland and Castle (1948)
Paper electrophoresis .. .. .	{ Hanging-strip method according to Lehmann and Smith (1954) Alkaline: barbitone buffer, pH 8.6 Acid: phosphate buffer, pH 6.5
Alkali denaturation .. .. .	1-minute test of Singer, Chernoff, and Singer (1951)
Cold denaturation .. .. .	Rigas, Koler, and Osgood (1956)
Solubility .. .. .	Itano (1953)
Chromatography .. .. .	Huisman and Prins (1955)

“Stanleyville I”

In one woman belonging to the Lugbara tribe paper electrophoresis at pH 8.6 demonstrated haemoglobin A and a haemoglobin variant amounting to 25% which moved slightly faster than haemoglobin S or D and resembled haemoglobin L or P by that method (Fig. 1). The red cells did not sickle. There was no foetal haemoglobin, and the haemoglobin mixture was highly soluble when tested according to Itano (1953): >5 g./l. in 2.24 M phosphate buffer, and 1.8 g./l. soluble in 2.58 M phosphate buffer. The abnormal haemoglobin was not resistant to alkali-denaturation but was resistant to cold-denaturation. Her husband and four of her five children showed haemoglobin A only, but one daughter aged 12 showed the same haemoglobin composition as her mother. Details regarding their blood are given in Table I. Whereas on paper electrophoresis at alkaline

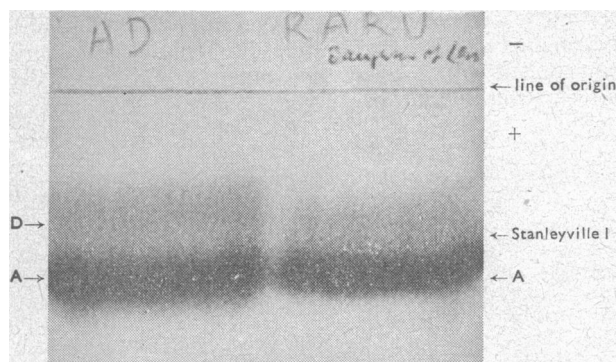


FIG. 1.—Comparison by hanging-strip paper electrophoresis at pH 8.6 of haemoglobin A plus D and A plus Stanleyville I. The new haemoglobin moves faster than haemoglobin D and more slowly than haemoglobin A; its position resembles that in which L and P would be found.

TABLE I.—Red Cells in Carriers of Haemoglobin Stanleyville I

	Count c.mm.	Hb/100 ml.	P.C.V. %	M.C.V. $\mu^3$	M.C.H. $\mu\mu\text{g.}$	M.C.H.C. %	Appearance of Film
Propositus	4,180,000	12.5	39.5	95	30	32	Normal
12-year-old daughter	4,200,000	12.4	40	96	30	32	„

pH the abnormal haemoglobin resembled L or P, it could be distinguished clearly from these two by chromatography on amberlite resin at pH 6. By this technique haemoglobin P does not separate properly from haemoglobin A, haemoglobin D moves more slowly than A, and haemoglobin L moves more slowly still. The abnormal component in the blood of the propositus and her daughter moved between L and P.

A mixture of haemoglobins A, P, L, and of the abnormal variant separated into three fractions in the following order: A+P, the abnormal haemoglobin, and L (Fig. 2). On the other hand, a mixture of haemoglobins A+S or A+D and the haemoglobin of the propositus separated into two fractions only, one with the mobility of haemoglobin A and one with that of haemoglobin S or D (Fig. 3). This

hitherto undescribed haemoglobin variant, which on paper electrophoresis at alkaline pH behaves like haemoglobin L or P, but which on chromatography at pH 6 behaves like haemoglobin S or D, is named, provisionally, haemoglobin “Stanleyville I.”

The propositus was a Lugbara, and her family all live in the Aru territory in the extreme north-east of the Belgian Congo. The Lugbara are Nilotes, and the haemoglobin described as D by Vandepitte in 1956, which, however, moved, as the illustrations show, somewhat faster than D on alkaline electrophoresis, was found in four members of a family belonging to the Lago tribe, who are Nilotes from the same part of the Belgian Congo.

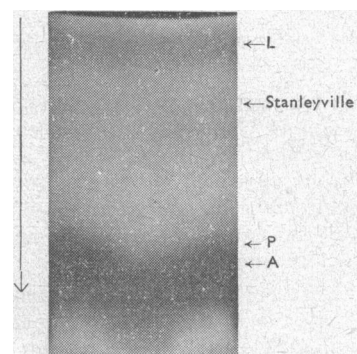


FIG. 2.—Results of ion-exchange chromatography of a mixture of haemoglobins A plus Stanleyville I, A plus P, and A plus L. Haemoglobin P does not separate from A, while Stanleyville I and L separate from A and from each other.

“Stanleyville II”

Another haemoglobin resembling haemoglobin D has been seen in two unrelated families.

The first propositus belonged to the Budu tribe. Her husband and four of her five surviving children showed haemoglobin A only, but one child—a daughter aged 6 years—showed the same haemoglobin composition as her mother. The Budu are considered Bantu; they live in the north-east of the Belgian Congo and are surrounded by Nilotic tribes. The other propositus was not discovered in the course of the survey of pregnant women, but was a boy aged 8 who was admitted to hospital for albuminuria. Subsequently his mother was found to show the same haemoglobin composition as her son. She came from the River Uele region in the north-east of the Congo, where the population is of mixed Bantu-Nilotic origin. She was of mixed Greek-African origin.

The blood of these four individuals did not sickle and there was no foetal haemoglobin present. Table II shows details regarding their red cells. The abnormal haemoglobin was not resistant to alkali-denaturation but was resistant to cold-denaturation. On electrophoresis at alkaline pH on paper and starch, haemoglobin A was found, and also a second fraction which was

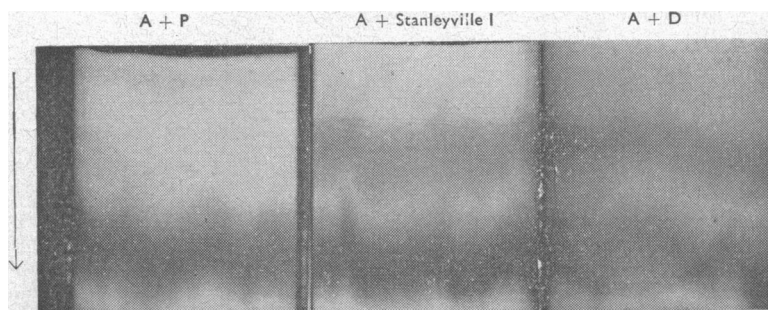


FIG. 3.—Comparison by ion-exchange chromatography of haemoglobins A plus P, A plus Stanleyville I, and A plus D. Although on paper electrophoresis at alkaline pH Stanleyville I moves faster than D and resembles P (Fig. 1), on chromatography it has the mobility of D.

TABLE II.—Red Cells in Carriers of Haemoglobin Stanleyville II

	Count c.mm.	Hb/100 ml.	P.C.V. %	M.C.V. $\mu^3$	M.C.H. %#	M.C.H.C. %	Appearance of Film
1st propositus	3,450,000	10.0	32	93	29	31	Normal
1st propositus's 6-year-old daughter	4,200,000	13.1	40	96	31	33	..
2nd propositus	3,880,000	10.3	34	88	27	30	..
2nd propositus's mother	4,400,000	13.0	43.5	99	30	30	..

indistinguishable from haemoglobin S or D by this technique (Fig. 4). On paper electrophoresis at acid pH no separation from A was achieved. However, on chromatography on amberlite resin at pH 6 two fractions were seen, one moving like haemoglobin A and one moving behind haemoglobin S or D but faster than haemoglobin L.

Mixtures of this haemoglobin with haemoglobins A, S, and L formed four distinct bands (Fig. 5). The solubility of the haemoglobin of the first propositus, which consisted of 40% of this hitherto undescribed variant and 60% of haemoglobin A, was rather low: measured according to Itano (1953) it was 1.1 g./l. of

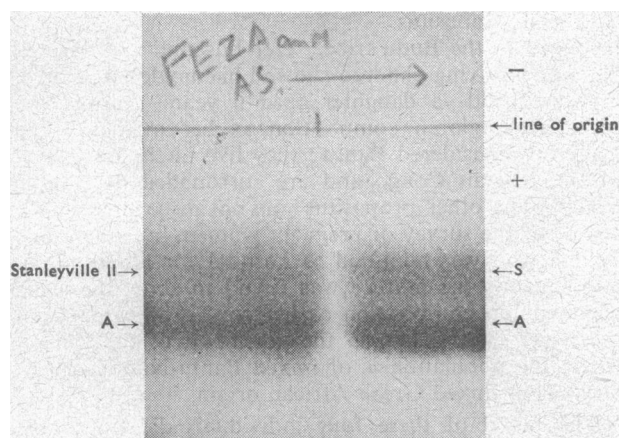


FIG. 4.—Comparison by hanging-strip paper electrophoresis at pH 8.6 of haemoglobin A plus S and A plus Stanleyville II. Haemoglobin Stanleyville II has the mobility of S or D.

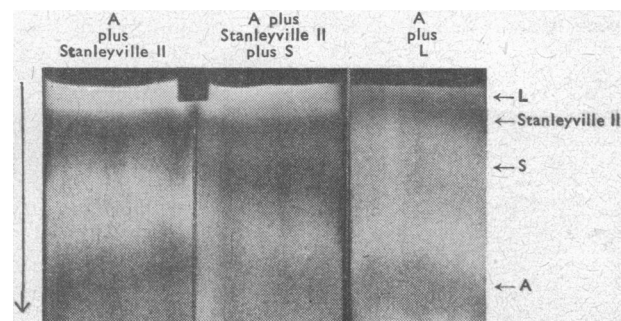


FIG. 5.—Comparison by ion-exchange chromatography of haemoglobin A plus Stanleyville II, a mixture of A plus Stanleyville II plus S, and of A plus L. Although on paper electrophoresis at alkaline pH Stanleyville II moves like S or D (Fig. 4), on chromatography it moves more slowly than S or D but not so slow as L.

2.58 M phosphate buffer. The haemoglobin of the first and of the second propositus and that of the son of the second propositus were all fully soluble in 2.24 M phosphate buffer in the test according to Itano. The haemoglobin of the daughter of the first propositus could not be tested for solubility. The haemoglobin variant described here, which on paper electrophoresis at pH 8.6 behaves like haemoglobins S or D, but which on chromatography at pH 6 differs from both by moving between haemoglobins S or D and L, we propose to name, provisionally, haemoglobin "Stanleyville II."

### Summary

Two hitherto unrecorded familial variants of adult haemoglobin are described. One, "Stanleyville I," was found in a Lugbara (Nilotic) family and is thought to be identical with one previously described when it was thought to be D, also occurring in a Nilotic family. The second, "Stanleyville II," was seen in two families, one belonging to the Budu tribe—a Bantu community surrounded by Nilotes—the other being of mixed Greek-Bantu-Nilotic origin.

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