# MEDICAL PRACTICE

## *Hospital Topics*

# Screening cord blood for sickle haemoglobinopathies in Brent

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### Abstract

**Between 1981 and 1983, 3165 consecutive specimens of cord blood were tested at the Central Middlesex Hospital for the presence of an abnormal haemoglobin: the incidence of sickle cell trait was 2·8%, of HbC trait 0·9%, and the overall incidence of an abnormal haemoglobin at birth was 6·9%. Five babies with homozygous sickle cell disease, three with HbSC, and three with either HbCC or HbC β thalassaemia were detected. Twenty two per cent of the mothers were of Afro-Caribbean origin. The cost of the test was 30p.**

**An H6000 blood count was carried out on 1000 consecutive cord blood samples. The mean red cell volume was 97·95 (SD 3·67) fl. Thirteen cord blood samples had a mean cell volume below 85 fl, and all contained Hb Barts. In addition, six samples with a mean cell volume between 86 and 92 fl also showed Hb Barts on electrophoresis. The overall incidence of Hb Barts was 2·1%.**

**These results indicate that the incidence of HbSS and HbSC on neonatal screening in Brent is similar to that found in the urban areas of North America and that the number may be predicted from the number of births to mothers of Afro-Caribbean origin.**


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### Introduction

The need to screen the newborn for sickle cell disease has not been established in Britain. The two British papers on newborn screening[1 2] dealt with populations containing less than 10% of Negroes, and the number of cases detected was so small that the programmes were not cost effective. Reports from America[3 4] and Jamaica[5] indicate that in areas where a high proportion of the newborn are black such screening yields a high number of babies with sickle cell disease and justifies energetic public health measures. Recent papers from America[3 4] show that screening combined with parental education and prophylactic administration of penicillin may significantly reduce infant mortality from the disease. The expected death rate during the first year of life, before these measures, is 13-14%.[6-9]

At the Central Middlesex Hospital, London, 59% of births are to non-European mothers, Afro-Caribbean mothers representing 22% of the total. Of the Afro-Caribbean births, 28% were to west African mothers and 72% to west Indian mothers. We therefore started a programme of cord blood screening in November 1981 and report the results of the first two years.

### Materials and methods

Cord blood was collected by midwives at birth and sent to the haematology laboratory within 24 hours during weekdays and within 48 hours during weekends. The mother's name, hospital number, and ethnic origin were noted.

Cord blood was subjected to electrophoresis on cellulose acetate, as

described.[5] After July 1983, H6000 blood counts were carried out on all cord blood samples.

The cost of each electrophoresis was 30p; this included reagents, cost and depreciation of equipment, and technical time.

## Results

A total of 3165 specimens of cord blood were tested during the two years. Five babies with homozygous sickle cell disease (SS), three with SC disease, and three with CC or C $\beta$ thalassaemia were diagnosed. Table I compares the incidences of HbS and HbC trait, as well as sickle cell disease, at the Central Middlesex Hospital and other published locations. Table II lists other abnormal haemoglobins encountered among the last 1000 births. During the two years 27 babies were found to have HbF only on initial testing; one of the babies was later found to be HbSC.

The mean cell volume for AF cord blood on H6000 was 97·95 (SD 3·67) fl; for AFS, AFC, SF, SCF, and CF it was 98·80 (4·83) fl. As expected, cord blood samples containing Hb Barts had an appreciably lower mean cell volume—85·75 (4·84) fl. Table III shows the relation between mean cell volume and Hb Barts.

The overall cost of detecting sickle cell disease was £118·68 for each case of major sickle haemoglobinopathy (3165 × 30p/8 cases).

TABLE I—*Comparison of the incidence of sickle cell disease in the newborn*

| Location | Total No of births | % Black births | % of all infants | | | | |
|---|---|---|---|---|---|---|---|
| | | | AS | AC | SS | SC | CC |
| Birmingham | 43 500 | 10·0 | 1·19 | 0·46 | 0·006 | 0·006 | |
| Manchester | 7 691 | 3·8 | 0·38 | | | | |
| Jamaica | 8 000 | >95·0 | 8·60 | 2·89 | 0·30 | 0·20 | 0·003 |
| New York | 106 223 | 35·0 | 3·26 | 0·83 | 0·18 | 0·12 | 0·002 |
| Central Middlesex Hospital | 3 165 | 22·0 | 2·80 | 0·91 | 0·15 | 0·09 | 0·090 |

TABLE II—*Haemoglobin phenotype and mother's ethnic origin*

| Haemoglobin phenotype | Mother's ethnic origin | | | | |
|---|---|---|---|---|---|
| | European (n = 340) | Indian (n = 405) | Afro-Caribbean (n = 205) | Other* (n = 50) | Total (n = 1000) |
| AA | 334 | 395 | 157 | 45 | 931 |
| AS | 1 | | 21 | 1 | 23 |
| AC | 2 | 1 | 7 | | 10 |
| AD | | 1 | | | 1 |
| AE | | 2 | | | 2 |
| Barts | | 5 | 13 | 3 | 21 |
| SS | | | 2 | | 2 |
| SC | | | 1 | | 1 |
| CC | | | 1 | | 1 |
| AG | | | 1 | | 1 |
| AJ | 1 | | | 1 | 2 |
| Others | 2 | 1 | 2 | | 5† |
| Total of variants per group | 6 | 10 | 48 | 5 | 69 |
| % of variants detected per group | 1·76 | 2·47 | 23·41 | 10·0 | 6·9 |

*Far East, Cyprus, Arab countries, mixed origin.
†Two haemoglobin F only, three unknown variants.

TABLE III—*Relation between mean cell volume measured on H6000 and Hb Barts*

| | Mean cell volume (fl) | | |
|---|---|---|---|
| | <85 | 85-92 | >92 |
| Hb Barts present | 13 | 8 | |
| Hb Barts absent | | 67 | 912 |

## Discussion

The incidence of sickle haemoglobinopathies on cord blood screening was higher than the expected one case of SS per 300 and one case of SC per 400 black births. This was probably due to the high percentage of west African mothers (28%) in the antenatal population. Our numbers are not large enough to allow us to estimate the incidence of SS and SC with absolute certainty. Nevertheless, it is clear that the expected incidence of three babies with homozygous sickle cell anaemia and two with SC per 1000 births to mothers of Afro-Caribbean origin was reached and probably exceeded.

As the technique of cord blood haemoglobin electrophoresis is simple and cheap, neonatal screening should become mandatory for the population at risk. Early identification of children with sickle cell disease will help reduce mortality and morbidity from the disease in the first years of life, as has already been successfully achieved in some American cities.

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*What is the relation between humidity and the health of people working indoors?*

Although plenty has been written about the effects of high humidity on health, there is much less information on the effects of low humidity (dryness of the air). I expect that this will change as more buildings are air conditioned. Full air conditioning includes humidification but some systems do not allow for added moisture. Others are intermediate and stop humidification when the outside temperature falls below 6°C. These buildings, though modern, do not have double glazing, with the result that humidified air condenses on cold windows. In such conditions, even were the outside air fully saturated (which it generally is not), the relative humidity would fall to well below 40% when the air is warmed to a comfortable 20°C. Relative humidities of 40-60% are considered comfortable but below 30% they may lead to irritation of the nasal and bronchial airways. Whether this is simply due to dryness or some other factor such as airborne dust is not clear.[1] The mucus in the air passage is said to become desiccated, so reducing its rate of flow due to ciliary action, with consequent diminished resistance to infection.[2] Recently, a condition of low humidity occupational dermatosis has been described in people working in relative humidities of under 30%. Essentially, it is a mild chapping of the skin of the face. In large modern open plan offices and hotels people walking short distances on carpets in a dry atmosphere may generate sufficient static voltages to experience minor shocks on touching grounded objects such as filing cabinets or door handles.[3] A work room of 100 m³ volume is said to require a minimum output of one litre of water an hour.[2] The profusely growing large leaved plants often seen in modern offices and hotel lounges may, therefore, have more than simple decorative value.—W R LEE, professor of occupational health, Manchester.


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