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Outcome of holistic care in Nigerian patients with sickle cell anaemia

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Summary Holistic care of patients with sickle cell anaemia (HbSS) was carried out in a dedicated support group and clinic in Lagos. This paper examines the outcome of this initiative using mortality, hospital admission and blood transfusion rates from inception in April 1988 to December 1995. Patients with sickle cell disorder and their families were admitted to the Sickle Cell Club and its associated Sickle Cell Clinic. All patients and parents were counselled on recruitment and were regularly followed up within an interactive family friendly environment. Other measures included preventive health and nutritional education, prompt treatment of illness and free supplies of vitamin supplements, malarial prophylactic and other necessary medication. The records of consecutive patients with HbSS were reviewed for this study. Over the study period, the number of subjects increased from 290 in 1988 to 1223 in 1995. The mortality rate fell from 20.6% in 1988 to 0.6% in 1995 ($P < 0.0001$); the number of hospital admissions fell from 350 (119%) in 1988 to 30 (4%) in 1995 ($P < 0.0001$); the number of patients transfused with blood fell from 260 (90%) in 1988 to 25 (2%) in 1995 ($P < 0.00001$). We conclude that the provision of well-organized holistic care can significantly reduce illness and deaths and improve the quality of lives of people living with HbSS in developing countries.

Keywords Blood transfusion rates, holistic treatment, morbidity and mortality rates, sickle cell anaemia, sickle cell disease, sickle cell disorder

Introduction

Sickle cell anaemia (HbSS) is the commonest severe inherited disorder of humans. Of the estimated 231 000 children born with HbSS in 1996, 84% were born in sub-Saharan Africa (WHO, 1994). This subregion has some of the poorest countries in the world, in many of which, poverty is further compounded by conflict. Consequently, most of these children do not receive adequate health care and hardly therefore survive childhood (Fleming *et al.*, 1979). Their disastrous health profile only serves to fuel the myths and stigma associated with sickle cell disorder in many communities in Africa. There is therefore an urgent need to improve the survival and quality of life for affected children in this subregion not only for the sake of families with affected members, but also as a strategy for combating ignorance and stigmatization within the communities.

In most of these countries, health care facilities are inadequate and access to good health care is limited to the relatively few persons who are able to afford fee-based private practices. Following the introduction of sickle cell genetic counselling to Nigeria in 1986 (Akinyanju & Anionwu, 1989), it became possible, for the first time ever, to provide basic holistic care to hundreds of affected persons and their families. The medical department of the Nigerian Ports Authority (NPA) in Lagos took up the challenge and trained many of its nurses in counselling on sickle cell disorder. The nurses, thus empowered, run a specialized Sickle Cell Clinic and an associated branch of a local support and advocacy group known as the Sickle Cell Club in the Apapa District of Lagos. The holistic care delivered in the clinic between April 1988 and December 1995 and its outcome are presented in this paper as a guide to what can be achieved in the care of people living with sickle cell disorder in developing countries with limited resources.

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Patients and methods

Patients with sickle cell disorder were referred by doctors and nurses to the NPA Sickle Cell Clinic and its associated Sickle Cell Club, if they were ill and were dependants of employees of the Nigerian Ports Authority or if they lived in or close to the Apapa District of Lagos. Consecutive patients with HbSS who were seen from inception on 4 April 1988 to 31 December 1995 were recruited into the study.

The diagnosis of HbSS was confirmed by a positive sickling test and the presence of Hb S only, on cellulose acetate electrophoresis at pH 8.9 (Dacie & Lewis, 1984). Clinics as well as Club meetings were held at the NPA Medical Centre on two alternate Saturdays of each month. The Medical Centre is a primary care facility with three consulting rooms, 25 general hospital beds, a medical laboratory, a 24-h manned emergency room but with no facilities for blood transfusion.

Nurses carried out the initial assessment of the patients and referred all new patients and others with deserving symptoms or signs to the duty medical officer for necessary management. Patients requiring hospital admission for pain control, rehydration and treatment of relatively minor ailments were admitted to the NPA Medical Centre. When necessary they were referred to the associated secondary care Railway Hospital or to the tertiary care Lagos University Teaching Hospital for ambulant or inpatient care including blood transfusion.

Patients and parents or guardians were counselled at their initial attendance or as soon as possible afterwards. Counselling was important to identify wrong beliefs and misconceptions, correct them and provide accurate information. Thereafter, they received regular health and nutritional education during meetings of the Sickle Cell Club. Prescribed drugs were dispensed free of charge and the importance of adhering to prescriptions, keeping clinic appointments and avoiding recognised triggers of sickle cell crisis, was repeatedly emphasized. Patients were encouraged to attend regularly and defaulters were visited at home by nursing staff or other parents.

They were encouraged to drink enough fluids to maintain adequate hydration. Guidelines for daily fluid intake were 100 ml/kg for children weighing up to 10 kg; 1.0 l + 50 ml/kg for children weighing 11–20 kg; 1.5 l + 20 ml/kg for every kg above 20. The subjects were advised to always carry water bottles around with them. In order to encourage compliance, they were taught to prepare a variety of local nutritious non-alcoholic plant-based beverages such as Zobo and soya bean milk.

Parents and guardians were taught to recognize pallor, detect enlarged spleens by abdominal palpation and treat minor ailments at home. The nurses always emphasized the need for the family to report severe or persistent symptoms early.

Drugs administered fell into the routine and the emergent categories. The routine drugs were supplementary folic acid and vitamins and those for malaria prophylaxis. The emergent ones were for the treatment of pain or infection – simple analgesics, non-steroidal anti-inflammatory drugs and antibiotics.

Malarial prophylaxis

On recruitment, 290 (23.7%) subjects were already taking pyrimethamine tablets once a week and 33 (2.7%) were taking proguanil tablets once a day. Nine hundred (73.6%) were not taking any chemoprophylactic medication. The patients taking either prophylactic drug were advised to continue to do so, while the 900 others were commenced on once weekly pyrimethamine; 12.5 mg for children aged below 12 years and 25 mg for all others.

Supplementary haematinics and vitamins

Each patient received one 5 mg folic acid tablet daily, one vitamin B compound tablet thrice daily and one 100 mg vitamin C tablet thrice daily, except those with leg ulceration who empirically received double the dose of vitamin C (200 mg thrice daily).

Immunizations

All patients gave a history of having received the State recommended immunizations comprising BCG at birth; triple vaccine – diphtheria, polio and tetanus – at ages 2, 3 and 4 months, measles vaccine at 9 months and a tetanus booster dose at school entry. Mothers had received 2–3 doses of tetanus toxoid 0.5 ml during pregnancy in order to prevent deadly neonatal tetanus in the children. Yellow fever vaccines were administered to all the members (parents and patients) who were eligible (i.e. 10 or more years since last vaccination) during and after the outbreak of yellow fever in Nigeria in 1989.

Emergent drugs

Acetaminophen (paracetamol) was the mainstay of analgesic and anti-pyretic treatment. Every patient received a supply to keep at home. When necessary they would come to report pain crisis and would then receive more potent analgesic drugs such as oral or parenteral diclofenac, tramadol or pentazocine. Fever was assumed to be caused

by infection and the febrile patient was treated with antimalarial drugs or antibiotics as suggested by the symptoms, clinical signs and the results of relevant laboratory tests of blood for malaria, white cell count, culture or urine or sputum cultures or radiological investigations.

Statistical analysis

Statistical analyses were carried out using the EPI info-version 6 software (CDC, Atlanta, GA, USA). Chi-square tests were used to compare results from two separate years in the study period. In order to assess the significance of differences between two periods of time, the contingency table was collapsed into a 2×2 table and the statistically significant value was regarded as $P \geq 0.05$.

Results

The number of patients registered increased gradually from 290 in 1988 to 1223 in 1995. At recruitment, 404 (33%) of the patients looked malnourished. The ages of patients at recruitment, as shown in Figure 1, ranged from 6 months to 62 years with a mean of 14.9 years (SD 10.6 years).

Hospital admissions

The number of admissions to hospital in 1988 was 350 (120.7%) as several patients were admitted more than once. This number reduced sharply and progressively over time and was only 30 (2.5%) in 1995 (Table 1).

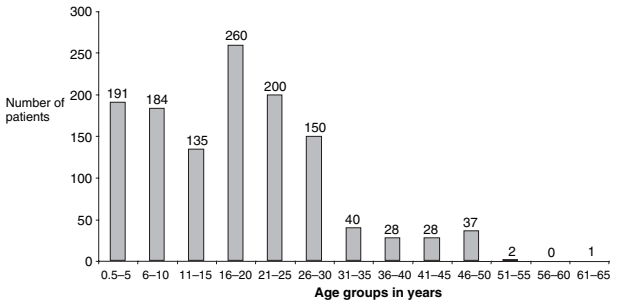

Figure 1. Ages at registration of 1223 HbSS in holistic care.

The duration of admission ranged from 3 to 35 days and it decreased progressively during the period of study, especially for long stay admissions. There was a 40% reduction in short stay admissions (<5 days), but admissions for longer than 5 days fell from 114% in 1988 to 2.8% in 1994 and 1.7% in 1995 ($P < 0.0001$). Whereas 235 of 290 (81%) patients were admitted for longer than 3 weeks in 1988, only one of 1076 (0.1%) was admitted in 1994 for that duration ($P < 0.0001$) and none was admitted in 1995 for longer than 3 weeks (Figure 2).

Blood transfusion requirement

In 1988, 260 of 290 (90%) subjects were transfused with a total of 320 units of blood at an average of 1.23 units of blood per patient. The blood transfusion rate decreased over time, such that, in 1995 only 25 of 1223 (2%) patients were transfused with a total of 27 units of blood ($P < 0.0001$) at an average of 1.08 units per patient (Figure 3).

Mortality rate

Table 2 shows the mortality rate during the study period. In keeping with the reduction in hospital admission there was a steady decline in mortality rate from 20.7% in 1988 to 0.6% in 1995 ($P < 0.0001$).

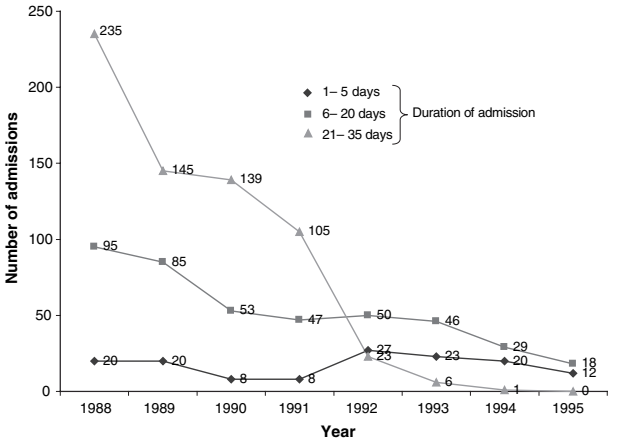

Figure 2. Duration of hospital admission in patients receiving holistic care.

Table 1. Hospital admission rates in HbSS patients in holistic care

	1988	1989	1990	1991	1992	1993	1994	1995
Total number of admissions	350	250	200	166	100	75	50	30
Number of patients	290	765	769	889	934	1054	1076	1223
Admission rate (%)	120.7	32.7	26	18.7	10.7	7.1	4.6	2.5

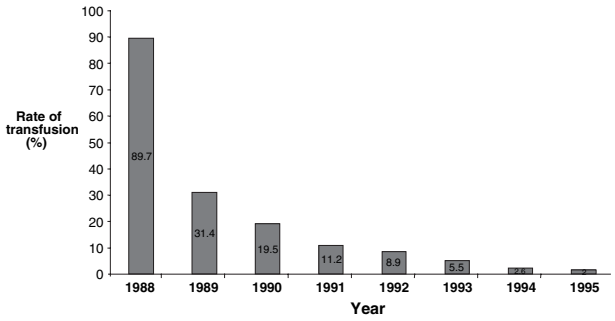

Figure 3. Blood transfusion in HbSS patients receiving holistic care.

Discussion

The results obtained clearly show that the morbidity and mortality rates were progressively and significantly reduced throughout the period of study. It is remarkable that these results were achieved without the use of attested prophylactic measures such as daily oral penicillin (Gaston *et al.*, 1986), pneumococcal vaccination (Kaplan, Sarnaik & Schiffman, 1986) and daily oral hydroxyurea (Charache *et al.*, 1995).

This should in no way suggest that these prophylactic measures are unnecessary but rather that significant improvement in outcomes is possible without them, especially in deprived populations. It is conceivable that their addition to our regimen might have further improved the outcomes, particularly in the very young children who are the most susceptible to pneumococcal sepsis (Kabins & Lerner, 1970; Seeler, Metzger & Mufson, 1972). Although previous work had suggested that pneumococcal sepsis was relatively unimportant in children with sickle cell anemia in Africa (Akinyanju & Johnson, 1987; Akuse, 1996) this may be an artifact due to underreporting as other publications from the same continent had demonstrated the contrary (Eeckels, Gatti & Renoirte, 1967; Nottidge, 1983). Underreporting may arise from the fulminant course and rapid fatality of the infection (Hallock, David & Marshall, 1970; Kabins & Lerner, 1970; Seeler *et al.*, 1972) which may occur before presentation to hospital, the difficulty in recovering the organism (Noah *et al.*, 1976) and of its isolation in some of our laboratories.

These results also suggest that much improvement is possible without hydroxyurea which should be encouraging to many health workers in Africa where hydroxyurea is usually unavailable and too expensive for most patients.

The significant reduction in blood transfusions is in keeping with the other indices of improvement in the health status of the patients. Malaria has been associated with severe anaemia in children with HbSS (Akinyanju & Johnson, 1987) and the introduction to all subjects of chemoprophylactic drugs may have contributed to the reduction of malaria and consequent reduction in the requirement for blood transfusion.

It may also be argued that some of the reduction may be the result of increasingly better judgement by the doctors responsible for ordering blood transfusion. However this would probably not be a major factor as the transfusions were not given at the NPA Medical Centre but only at the Railway Hospital or the Lagos University Teaching Hospital to which patients with severe anaemia were referred. An important benefit of the low transfusion rate would be the prevention of serious blood transmissible disease such as hepatitis, HIV and the reduction in iron overload.

Although we did not apply any specific instrument to measure the quality of lives of the patients, we believe from our observation and particularly from the comments of the parents that it was much improved in the affected persons and their families.

The supplemental vitamins and repeated general health and nutritional education would have contributed to these outcomes but we have no evidence that the patients required as much supplemental vitamins as they received. There is obviously room for fine-tuning the intervention in order to eliminate superfluous medication or else correctly attribute benefits to them.

We attribute the excellent outcome in this study to three main factors. First was the introduction of expert counselling. The introduction of training in counselling is probably the most important low cost and low technology strategy for addressing the problems of sickle cell disorder in any country, as it creates a corps of informed workers who can organize and advocate for necessary services.

Table 2. Mortality rates in HbSS patients in holistic care

	1988	1989	1990	1991	1992	1993	1994	1995
Number of deaths	60	45	22	24	18	18	11	7
Total number of patients	290	765	769	889	934	1054	1076	1223
Mortality rate (%)	20.7	5.9	2.9	2.7	1.9	1.7	1.0	0.6

Secondly, the provision of prompt access to needed care irrespective of patients' ability to pay for it. Thirdly, the commitment of the medical department and dedication of the nurse/counsellors who ran the clinic and the associated patient/parent support group, the Sickle Cell Club. The total effect of these factors was the creation of a friendly and interactive atmosphere of enlightened care and optimism.

The measures employed to achieve these results should be available to people with HbSS in other developing countries and we hope that they will serve as a model to achieve similar or even better results wherever available resources do not begin to match the scale of the problem.

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References

Akinyanju O.O. & Anionwu E.N. (1989) Training of counsellors on sickle cell disease in Africa. *Lancet* **i,** 653–654.

Akinyanju O. & Johnson A.O. (1987) Acute illness in Nigerian children with sickle cell anaemia. *Annals of Tropical Paediatrics* **7,** 181–186.

Akuse R.M. (1996) Variation in the pattern of bacterial infection in patients with sickle cell disease requiring admission. *Journal of Tropical Paediatrics* **42,** 318–323.

Charache S., Terrin M.L., Moore D.R., Dover G.J., Barton F.B., Eckert S.V., McMahon R.P. & Bonds D.R. (1995) Effect of hydroxyurea on the frequency of painful crisis in sickle cell anemia. *New England Journal of Medicine* **332,** 1317–1322.

Dacie J.V. & Lewis S.M. (1984) *Practical Haematology*, 6th edn. Churchill Livingston, London.

Eeckels R., Gatti F. & Renoirte A.M. (1967) Abnormal distribution of haemoglobin genotypes in negro children with severe bacterial infections. *Nature* **216,** 382.

Fleming A.F., Storey J., Molineaux L., Iroko E.A. & Attai E.D.E. (1979) Abnormal haemoglobins in Sudan Savanna of Nigeria. I. *Annals of Tropical Medicine and Parasitology* **73,** 161–172.

Gaston M.H., Verter J.I., Woods G., Pegelow C., Kelleher J., Presbury G., Zarkowsky H., Vichinsky E., Iyer R., Lobel J.S., Diamond S., Hoolbrook C.T., Gill F.M., Ritchey K. & Falleta J.M. (1986) Prophylaxis with oral penicillin in children with sickle cell anemia. A randomised trial. *New England Journal of Medicine* **314,** 1593–1599.

Hallock J.A., David E. & Marshall L. (1970) Pneumococcal infection in sickle cell anemia. *Journal of the American Medical Association* **212,** 629.

Kabins S.A. & Lerner C. (1970) Fulminant pneumococcemia and sickle cell anemia. *Journal of the American Medical Association* **211,** 467–471.

Kaplan J., Sarnaik S. & Schiffman G. (1986) Revaccination with polyvalent pneumococcal vaccine in children with sickle cell anemia. *American Journal of Pediatric Hematology and Oncology* **8,** 80–82.

Noah M.A., Odugbemi T.O., Oyegunle A.O. & Ogundipe O. (1976) Bacterial aetiology of acute pneumonia in Lagos children by examination of tracheal aspirates. *Nigerian Medical Journal* **6,** 319–322.

Nottidge V.A. (1983) Pneumococcal meningitis in sickle cell disease in childhood. *American Journal of Diseases of Children* **137,** 29–31.

Seeler R.A., Metzger W. & Mufson M.A. (1972) Diplococcus pneumoniae infection in children with sickle cell anemia. *American Journal of Diseases of Children* **123,** 8–10.

WHO (1994). *Guidelines for the control of haemoglobin disorders*. WHO/HDP/HB/GL/94.1.