SIMPLE INTERVENTION AND PARENTS EDUCATION IN REDUCING THE MORBIDITY IN SICKLE CELL ANAEMIA

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Abstract:

The impact of simple intervention and parents education was evaluated on sickle cell anaemia. A group of 70 patients, attending sickle cell clinic at Khartoum children emergency hospital during the period June 1996 through May 200 K were enrolled. Their ages ranged between 5-18 years. The interventions were early treatment of infections, folic acid daily, parents5 education about the disease and its complications, meningitis vaccination during the epidemics and close monitoring in the sickle cell clinic.

Age, sex, residential area, frequency of hospitalization and blood transfusion requirement five years before the study and during the five years of follow up were recorded. Haematological parameters, were studied for each patient pre and post intervention.

The study showed that the number of patients is inversely related to age. Males in. this study exceeded females ($P<0.01$). 70% of the patients were living in unplanned area. Frequency of hospitalizatipn and blood transfusion were significantly reduced after the intervention ($P<0.01$). Significant differences were encountered in the values of the haematological parameters. These results show that simple intervention and parents education play a significant role in decreasing the morbidity associated with homozygous sickle cell disease.

Introduction:

Human diseases often have a well-characterized clinical course and a poorly understood molecular basis. In contrast, sickle cell anaemia (SCA) Is a disease with an unpredictable clinical course and a well characterized molecular defect in which substitution of thymine for adenine in the glutamic acid DNA codon (GAG ,GTG), results in turn, in substitution of B6 valine for glutamic acid[1]. SCA and its yariants constitute the most common lethal hereditary diseases in human. As a major cause of illness, sickle cell disease concerns not only the paeditricians and haematologests but also obstetricians, orthopedists, ophthalmologists, urologists, and surgeons [2]. The disease results in significant morbidity and mortality for those affected [3]. The symptoms and signs of SCA are exacerbated in times of crisis, characterized by tissue infarction or worsening anaemia. Prompt medical intervention is required in these distressing situations to provide relief and comfort to the' patients [4] Patients, particularly children [5,6]' have increased susceptibility to infections [5,7] As the leading cause of morbidity and mortality in SCA, infection calls for vigorous antibiotic therapy [4] Environmental factors although rather ill defined, there is no doubt that they are quantitatively and qualitatively important. In tropical countries, it is commonly noticed that the incidence of crises tends to increase during the rainy season and that exposure to cold weather or to a cold bath may trigger a crisis.

Nutrition is certainly important, for instance because SS patients have an increased folic acid requirement. The socioeconomic status is also certainly important. Patients belonging to the upper strata in general do better, perhaps mainly because they live in a more protected

environment, are likely to be diagnosed earlier, and tend to receive more continuous medical supervision and more prompt medical attention when they need it [8].

Successful bone marrow transplantation may relieve the symptoms by changing the phenotype of red cell precursors in the bone marrow to that of the donor marrow [9] .Use of the procedure has been limited by the availability of compatible donors, and the considerable cost. Such a procedure is unlikely to be applicable on a population basis especially in countries with limited resources where most sickle cell disease occurs. Gene therapy may offer hope of cure in the future. An alternative approach is the assessment of simple measures to diminish morbidity, improve survival, and prevent the high early mortality of the disease [10] These include prophylactic penicillin[11,12] parental education, earlier intervention, and coordinated management in sickle cell clinics[13,14]

In Sudan SCA is a public health problem in western and southern parts [15] The years 1984-85 showed massive influx of migrants from the west and south to the capital, Khartoum. Migrant tribes among whom SCA is prevalent are distributed in camps around the town or live in informal quarter's [16]' In the past there were no special services offered to our patients with SCA. They used to be seen at the different referred paediatrics clinics. They suffer problems of access to the medical centres, early diagnosis, long term follow up, family screening and counseling. Most of our patients are from the low socio-economic displaced population. More over they have wrong believes that the disease is transmitted through breast milk, and foods like eggs, and meat. cause abdominal pain and distention for the child with the disease (personal

observations). The economic, psychological and medical consequences of this haematologic disease are of urgent importance and can not be egnored. To offer services and collect data oprospectively concerning the natural history of sickle haemoglobin disorders, the sickle cell clinic was established in 1996.

The objective of this study is to show the impact of simple interventions, regular follow up and close monitoring in the sickle cell clinic and parents education on the morbidity in SCA pre and post intervention.

Materials and Methods:

Patients:

Children with SCA of ages ranging between 5-18 years attending sickle cell clinic at children emergency hospital, Khartoum, were managed and followed up in this study. Patients' enrollment was begun on June 1996 through May 2001. A total of 70 patients whose haemoglobin electrophoresis showed the presence of Hb SS were investigated and Subjected to a detailed history including hospital admissions and blood transfusions in the previous five years. Physical examination was done initially and clinical manifestations were recorded at every visit. They were followed for five years. They used to come monthly for follow up and encouraged attending at any time if sick on the clinic day or any time to the emergency department.

Laboratory:

The clinic perform cellulose acetate haemoglobin electrophoresis, haemoglobin level, packed cell volume and white blood count for all

patients on entry to the study, during the period of follow up when recommended and at the end of the study. The methods used are standard [17]

Interventions

The patients were receiving folic acid daily. Early institution of antibiotics in suspected infections. Parents were educated about the disease and its complications. They were also taught to manage mild episodes of painful crises at home using paracetamol, to palpate the spleen and to examine for pallor. Avoidance of predisposing and precipitating causes of crisis and protection against varying weather conditions were emphasized. Awareness that unusual pallor or other symptoms, or vague ill health should cause a visit to the sickle cell clinic for early detection and treatment of otherwise serious complications. Parents were informed that their children need good nutrition with high calories and proteins and their wrong believes were corrected. All the patients received meningitis vaccination during the epidemic of 1999.

Statistical Methods:

The data were statistically analyzed. Mean and standard deviation were calculated for the age. Chi square test was used for the sex difference. The average steady state values and standard deviations of the hematological parameters pre and post intervention were calculated using student^5 s t-test. P value of <0.05 was considered statistically significant.

Results:

TAPLE1

Age & Sex Distribution of the 70 Patients attending Sickle Cell Clinic, June 1996, Through May 2001.

Age (years)	Sex				Total	
	Males		Females			
	N0	%	N0	%	NO	%
5>10	28	74	23	72	51	73
10-18	10	26	9	28	19	27
Total	38	54	32	46	70	100

Mean Age = 9.6 ±3.4 years Males are more affected than females (P< 0.01)

TAPLE 2

Distribution Of The Patients According To Residential Area, Sickle Cell Clinic Study June 1996 Through May 2001.

Residential Area	NO	%
Planned	21	30
Unplanned	49	70
Total	70	100

SCA Patients are More Commonly from Unplanned Residential Area.

TAPLE 3

Age-Specific Hospital Admissions incidence spills in Patients With SCA, Sickle Cell Clinic Study June 1996, Through May 2001.

Age (years)	No Of Patients	Hospital Admissions Incidence Spills			
		Pre-Intervention		Post-Intervention	
		No of Hospital Admissions	Incidence Spills	No of Hospital. Admissions	Incidence Spills
5>10	51	186	3.6	83	1.6 1
10-18	19	83	4.3	36	1.8
Total	**70**	**269**	**3.8**	**119**	**1.7**

Incidence of hospital admissions spills is reduced post intervention (P<0.01)

TAPLE 4

Age-Specific Blood Transfusion Incidence Spills In-Patients with Sickle Cell Anaemia, Sickle Cell Clinic Study June 1996 Through May 2001.

Age (years)	No of Patients	Blood Transfusions Incidence Spills			
		Pre- Intervention		Post-Intervention	
		No. of Blood Transfusions	Incidence Spills	No of Blood Transfusions	Incidence Spills
5>10	51	148	2.9	62	1.2
10-18	19	59	3	26	1.3
Total	70	207	2.9	88	1.2

Incidence of blood transfusions spills is reduced post-intervention (P<0.01)

TAPLE 5

Haematological Parameters in Patients with Sickle Cell Anaemia Pre & Post-Intervention I

Parameters	Pre-Intervention	Post-Intervention	P. value
Hb(g/dl)	6.4+0.713	7.3+0.823	<0.01
PCV(Vl)	0.21+0.031	0.25+0.056	<0.01
WBC(x 10^9dl)	9.4±6.82	7.6±3.24	<0.05

Haematological Parameters Improved Post-Intervention

The results of the age and sex distribution in the SCA patients are presented in table 1. Males are more affected than females (P<0.01). Mean age of the patients is 9.6±3.4 years.

A significant difference in the residential area is shown in table 2. 70% of the patients are living in unplanned area.

An age-specific frequency of hospital admissions and blood transfusions is shown in table 3&4. Statistically significant differences were evident in the frequencies of hospitalizations, which has been reduced from 3.8 to 1.7 spills/patient. The best ^ reduction is obtained in the age group 5>10 years and the least is in the age group 10-18 years. Incidence of blood transfusion of all cases has changed from 2.9-1.2 spills/patient.

The results of the haematological parameters are presented in table 5. The major statistically significant differences are in the haemoglobin and the PCV, which were higher post interventions (P<0.01). The WBC has decreased significantly (P<0.05).

Discussion:

In this study the number of patients in the different age groups is inversely proportional to he age. The mean age before intervention is 9.6±3.4 years. This age is the mean age of survival .for those who escaped death. Previous reports of SCA in the young have emphasized the importance of age on the mortality (8J9)' The males are significantly more affected than females."

70% of patients are living in unplanned residential area, where more exposure to infections and environmental hazards. 30% are living in the planned area. This big difference is either due to the fact that, those who live in the planned area go to private health services or could be explained by the population number/ethnic variation or both.

This study showed improved health of children with SCA and reduced morbidity. The number of transfusion requirements and extent of hospitalization were considerably reduced in all age groups. All the patients in this study are of school age (5-18 years). Hospitalization in this age group before intervention was 3.8 spills/ patient, which has been reduced to 1.7 spills/patient. Simple intervention has decreased the lost school days, burden on the families and hospitals. It appears that early treatment with antibiotic plays a role in lowering the incidence of serious infections associated with SCA. A study described Saudi patients with SCA showed reduced morbidity and mortality following use of pneumococcal vaccine and prophylactic penicillin[20] A Jamaican study showed improved survival of childrenw with sickle cell disease and decreased morbidity .following successful trial of pneumococcal prophylaxis [21]?. Although mortality in the

United States decline before wide spread use of penicillin, possibly reflecting greater awareness, earlier diagnosis, and more prompt treatment( ) Intervention in this study has reduced infections and crisis leading to hospitalization.

Haematological parameters, like the haemoglobin level and PCV are higher post intervention in all age groups with statistically significant difference. This incidence can be explained by good nutrition, folic acid prophylaxis and decrease haemolysis. A significant difference was observed in the level of WBC, which was significantly lower in the post intervention compared to pre intervention values. It is possible that the leukocytosis in pre intervention reflects greater bone marrow activity as a result of increase haemolysis and infection. These results are similar to a Saudi study results [20].

Differing methods of intervention between our study and other's weaken comparability between them. Most of the studies described association between prophylactic measures and mortality. In this study we did not comment on mortality as the period of follow up is short.

In conclusion, the results of this study advocate early institution of antibiotic treatment for febrile episodes, parental education about the disease, supportive and prolonged care to improved survival of patients with SCA. Special attention should be given to school age children. Services offered to patients with SCA should not be centralized. Further follow-up is needed to determine the effect of these parameters on the mortality of our patients with SCA.

Recommendations:

1-Active and early detection methods should be adopted for early reaching

of SCA cases especially in slumps and unplanned areas.

2- Sickle cell clinics to be established in hospitals where there are paediatricians.

3- A mass media health education for parents should be carried regularly.

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References

1.Richard Steingart, MD. Management of patients with sickle cell disease. The medical clinics of North America, volume 76. Number 3. May 1992.

2.Helen M. Ranney. University of California, San Diego. The spectrum of sickle cell disease. Hospital Practice, January 15. 1992.

3.Hoppe-C; Styles-L;Vichinsky-E. The natural history of sickle cell

disease. Curr-opin-pediatr. 1998 Feb; 10(1): 49-52.

4.0kpala-l. The management of crisis in sickle cell disease. Eur-J-Haematol. 1998 Jan; 60(1): 1-6.

5.Barrett-Connor E. Bacterial infection and sickle cell anaemia: an analysis of 250 infections in 166 patients and a review of the literature. Medicine (Bait) 1971;50: 97-112.

6.0verturf GD, Powars dr, Baraff LJ. Bacterial meningitis and septicemia in sickle cell disease. Am J Dis Chiidh 1977; 131: 784-7.

7. Hand WL, King HL. Serum opsonization of Salmonella in sickle anaemia. Am J Mod 1978; 64: 388-95.

8. Lucio Luzzatto. Sickle Cell Anaemia in Tropical Africa. Clinics in Haematology-Vol. 10: No. 3, October 1981.

9. Johnson FL, Look AT, Gockerman J, Ruggiero MR, et al. Bone marrow transplantation in a patient with sickle cell anaemia. N Engl. J Med 1984; 311: 780-3.

10. Thomas AN, Pattison C Serjeant GR. Causes of death in sickle-cell disease in Jamaica. BMJ 1982; 285: 633-5.

11. John AB, Ramlal A, Jackson H, Maude GH, Waight Sharma A, Serjeant GR. Prevention of pneumococcal infection in children \vith homozygous sickle cell disease. BMJ 1984; 288: 1567-70.

12. Gaston MH, Verter JI, Woods G, Pegelow C, Kelleher J, Presbury G.etal. Prophylaxis with oral penicillin in children with sickle cell anaemia. N Engl. L Med 1986; 314: 1593-9.

13. Davies SC, Luce PJ, Win AA, Riordan JF, Brozovic M. Acute chest syndrome in sickle-cell disease. Lancet 1984; 1: 36-8.

14. Mallouh AA, Asha MI. Beneficial effect of blood transfusion in children with sickle cell chest syndrome. Am J Dis Child 1988; 142: 178-82.

15. Vella F. Haemoglobin S and sickling in Khartoum province. Trans Roy SOC Trop. Med Hyg. 1966,60:48-52.

16. Sayyid H. Hurreiz & Elfatih A. Abdel Salam. Ethnicity, Conflict and National Integration in the Sudan. Institute of Africa and Asian Studies University of Khartoum, 1989.

17.Dacie, Sir John V. Practical haematology. 6th Edi. Edinburgh (etc.): Churchill Livingstone, 1984.

18. Murphy V, and Haywood, L. Survival analysis by sex, age group and haemolysis in sickle cell disease. J. Chronic Dis. 1981; 34: 313-319.

19. Powars D. Natural history of sickle cell disease-the first ten years. Semin Hematol. 1975; 12: 267-285.

20. M. A. F. El-Hazmi, H.M. Bahakim, M. A. Babikar, A. M. Al-Swailem, and A.S. Warsy. Symptom-free Intervals in Sicklers: Does Pneumococcal Vaccination and penicillin Prophylaxis Have a Role? Journal of Tropical Pediatrics. 1990;Vol.36: 56-62.

21. Allison Lee, Peter Thomas, Lena Cupidore, Beryl Serjeant. Improved survival in homozygous sickle cell disease: lessons from a cohort study. BMJ. 1995; 311: 1600-2

22. Powards D, Overturf G, Weiss J, Lee S, Chan I. Pneumococcal septicemia in children with sickle cell anaemia. Changing trend of survival. Jama 1981; 245: 1839.