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HOSPITAL ADMISSION OF PATIENTS WITH SICKLE CELL ANAEMIA PATTERN AND OUTCOME IN ENUGU AREA OF NIGERIA.

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ABSTRACT

Background Objective: The admission of children with sickle cell anaemia into the Paediatric ward of the University of Nigeria Teaching Hospital,(UNTH) Enugu was retrospectively reviewed to ascertain their pattern and outcome. Such data are useful in guiding future health policies on such children.

Patients and Methods: Medical files of patients with sickle cell anaemia who were admitted between May1998 and April 2002 were analyzed. Their clinical features, investigations done, diagnosis on admission, complications and subsequent outcome of admissions were noted.

Results: More children were admitted during the rainy season.($p<0.05$) Male : Female ratio was 1.2:1. More children(70.4%) failed to comply with treatment of which a statistically significant number came from the lower social class ($p=0.001$). Infections were the commonest cause of admission (69.6%). Severe anaemia was the commonest complications (39.4%). Mortality occurred in 8.5%.

Conclusions: Intensive counseling of patients, with the involvement of Social workers and health visitors, is strongly advocated. There is need for antibiotic prophylaxis for all children with sickle cell anaemia.

Key words: Sickle cell anaemia, Hospital admission, Pattern and Outcome. *(Accepted 30 May 2006)*

INTRODUCTION:

Sickle cell disease is a genetically inherited abnormality of haemoglobin[1] in which valine replaces glutamic acid in the 6th position of the B chain in the haemoglobin molecule.[2] The consequence is the formation of haemoglobin S (HbS). In its deoxygenated form, Hb S comes out of solution forming long crystals called tactoids which distort red cells. Patients who are homozygous for HbS are said to have sickle cell anaemia. About 80 95% of their haemoglogin is HbS, the remainder being Hb F and HB A_2. [2] Cells which contain homozygous Hb S are susceptible to premature destruction as such a cell has a red cell life span of 8 25 days.[3] This premature destruction results in chronic haemolysis. Besides, the formation of tactoids distorts the red cell membrane and hence, affects the microcirculation. The blood viscosity increases hence promoting venous stasis. This may occur spontaneously or secondary to dehydration or infection among other factors. Tissue death with or without infarction occurs.[4] Clinically, the child presents with mild to severe systemic malfunction. While some of the problems may be medically managed at home, others and probably the very severe cases, are managed in hospitals where professionals with expert knowledge in various specialties are found. Such admissions are on the increase since parents are increasingly becoming health and hospital conscious.[5] As has been noted by some workers, common reasons for admitting these children inlude malaria infection,[6] bacteria infections[7] or painful crisis[8] For example, it was noted that painful crisis accounted for 80 90% of sickle cell related admissions in the United Kingdom and the United States of America. However, painful crisis accounted for 8.8% of such admissions in Jamaica. This is due to the fact that, in that country, only very severe cases were admitted into the hospital while the less painful ones were observed in a day care facility. Besides, it is known that mortality in sickle cell anaemia may be due to acute chest syndrome,[9] septicaemia [10]or acute splenic sequestration.[11] It is against this background that we attempted to review the pattern and outcome of

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admissions of patients with sickle cell anaemia over a four year period as seen at the University of Nigeria Teaching Hospital (UNTH) Enugu. It is believed that such data and findings will be useful in guiding future health policies or their implementation with regard to the management of a child with sickle cell anaemia.

MATERIALS AND METHODS

The medical files of patients with sickle cell anaemia who were admitted into the Paediatric wards of UNTH Enugu between May 1988 and April 2002 were retrieved and analyzed. These children had been previously registered in our Paediatric Sickle Cell Clinic and are on regular folic acid and proguanil anti malarial prophylaxis. They were routinely seen at this clinic at intervals of four to ten weeks the more stable the child, the longer the appointment.

Information sought for, in the files, included age, gender, socioeconomic class as described by Oyedeji,[12] diagnosis on admission and associated complication. Others included full blood count with attention on the white cell count, presence of malaria parasites and haemoglobin level on admission. Results of urine examination, liver function and blood culture were reviewed. The treatment received before presenting to the hospital for admission as well as those who had blood transfusion, on admission, were noted. The outcome of their admission was documented. Compliance with treatment was determined by reviewing the patients' ability to attend the clinic regularly, as requested, as well as taking their drugs and following advice given during pre- admission clinic visits.

Statistical analysis was done on the information retrieved from the patients' medical notes using the SPSS statistical package.

Student t- test and Chi square test were used to test for associations.

RESULTS:

During the study period, there were three thousand, seven hundred and two (3,702) admissions into the Paediatric wards of which eighty three (83) patients had sickle cell anaemia. This represents 2.2% of the entire admissions. Of these patients with sickle cell anaemia, fifty one (61.4%) were admitted between May and October (rainy season) while the remaining 32 (38.6%) had their admission between November and April (dry season) - $t = 3.3, p < 0.05$.

Of the 83 admissions, only seventy one medical files (86%) were retrieved and subsequently analyzed. There were 38 males(M) and 33 females(F) with a M:F of 1.2:1. Their ages ranged from 13 months to 21 years (mean 90.4 +11.6 months). Compliance with treatment occurred in 50 patients (70.45%). The children were admitted for a period of 6 to 41 days- (mean 15 +3 days). Six (8.5%) of the 71 admitted children died. Of the children that died, four had defaulted to follow up visits for ten months or more. Majority of the children had received some form therapy at home which included oral antibiotics, analgesics, folic acid and multivitamins.

Table 1A displays the socio economic status (SES) of the 71 analyzed children with sickle cell anaemia. It is noted that only 8 children (11.2%) from the upper socio-economic status i.e SES 1 and II, Fifty patients (73.2%) were either in the middle SES III or low social classes SES IV and V.

Table1B shows that twenty six children (36.6%) were aged 5 years and below, twenty patients (28.2%) were between 6 and 10 years of age while the remaining 25 children were above 10 years of age.

Table II examines the interval between their last clinic attendance and hospital admission. Only 21 patients (29.6%) were seen within 3 months prior to admission and hence could be said to have complied with attendance and follow-up management since they regularly abided by the advice given to them during their follow up visits.

Table III reviews the diagnosis of the patients on admission. Eighteen children (25.4%) were admitted for various crisis namely hyper haemolytic (5), Hepatic (5), and vaso-occlusive (8) crises. The other 53(74.6%) children were admitted for other reasons. Of the latter group, bacterial infections were the commonest cause of admission followed by malaria. These infections involved the bone, lungs joints or muscles. The diagnosis of bacterial infections was based on their clinical presentation as well as the presence of toxic granulated neutrophils in the blood film. No organism was cultured from the patients on whom blood cultures were done.

Table IV evaluates their associated complications. Severe anaemia (haemoglobin of less than 5gms/dl) occurred in 28 patients (39.4%). Other complications included chronic osteomyelitis with

or without fractures in 8 patients and pleura effusion among others. Their initial haemoglobin values as well as the frequency of blood transfusion, while on admission, are shown on Tables VA and VB. It is noteworthy that 29 children (40.8%) required more than one blood transfusion.

Table 1A: Socio economic classes of children Admitted with sickle cell anaemia

Social class	Number	Percentage
I	3	4.3
II	5	7.0
III	21	29.6
IV	14	19.7
V	17	23.9
Unstated	11	15.5
Total	**71**	**100**

Table IB: **Age distribution of the 71 Children**

Age Range, (Years)	M	F	Total	%
1- 5	14	12	26	36.6
6 - 10	12	8	20	28.2
11-15	9	7	16	22.5
>16	3	6	9	12.7
Total	**38**	**33**	**71**	**100**

Table II: **Interval between last clinic visit and hospital admission**

Duration in Months	Number	Percentage
0 -3	21	29. 6
4 -7	17	23.9
8 - 11	10	14. 1
12- 15	9	12. 7
16 - 19	5	7.0
>20	9	12. 7

Table 111 : **Diagnosis of the patients o admission**

Diagnosis	Number
Osteomyelitis	14
Pneumonias	13
Severe malaria	12
Septic arthritis	3
Pyomyositis	4
Hyperhaemolytic crisis	5
Septicaemia	2
Hepatic crisis	5
Vaso occlusive crisis	8
Priapism	2
Others*	15

*** Some patients had more than one diagnosis
*Others included cellulitis, acute glomerular nephritis (AGN), obstructed hernia, carvenons sinus thrombosis etc.

Table Iv: **Associated Complications in the 71 Patients**

Complications	Number	Percentage
- Severe anaemia	28	39.4
- Chronic Osteomyehtis with or Without fractures	8	11.3
- Pleural effusion	2	2.8
- Epilepsy	3	4.2
- 6th Cranial nerve palsy	1	1.4
- Aseptic necrosis of the femoral head	2	2.8
- Pseudo membranous colitis	1	1.4
- Erectile dysfunction	1	1.4
- Arthropathy	3	4.2

Table VA: **Haemoglobin values of the children at the beginning of admission**

Hb gms.dl^{-1}	Number	Percentage (%)
0 - 2.4	1	1.4
2.5 - 4.9	27	38.0
5 - 7.4	23	32.4
>7.5	14	19.7
Not stated	6	8.5
Total	**71**	**100**

Table VB: **Frequency of blood transfusion during the admission**

No of. Transfusions patients.	Number of (%	Percentage
0	19	26.8
1	23	32.4
2	17	23.9
3	9	12.7
>4	3	4.2
Total	**71**	**100**

DISCUSSION

Though retrospective studies are easy to handle, they have problems of providing incomplete data especially on some relevant variables. For example, in 6 of the admitted patients the haemoglobin levels on admission were not documented. Bamgboye and Jegede[13] had observed on similar problems as associated with retrospective studies. However, this study revealed that there were more admissions

During the rainy months. These are the cold and damp periods of the year. This is similar to the findings by Amjad et al[14] as well as studies from Jamaica as documented by Redwood et al[15] But, neither the observation by Slovis et al[16] nor Seeler[17] in Chicago could substantiate any seasonal variation in hospital admission among patients with sickle cell anaemia. It is possible that the practice of wearing warm clothings during the cold months may have minimized the effect of cold on those children. Such practices are not commonly observed in our environment and hence our sicklers are maximally exposed to the adverse effects of cold on their health.

There are only eight children (11.3%) of the 71 patients analyzed who were from the upper social classes (1 and II). It is possible that children of this social class attend private hospitals as they could afford the attendant high medical bills. Besides, children from this social group may have been seeking medical treatment as early as possible hence limiting further treatment to home management except in situations where hospital admission is inevitable. This health seeking behaviour may explain the mild course of the disease in the high social group as observed by some workers[6,13] Lesi[18] had also stressed the positive role of education and better socio-economic conditions on the survival of patients with sickle cell anaemia. This acquisition and use of knowledge explains the better understanding and subsequent responsiveness to medical care among parents in the upper social class.

Compliance with treatment was poor in 50 patients (70.4%). Of this number, 38 patients are from the low social class (IV and V). There is a significant influence of poor compliance on hospital admission among patients in the low social class. (P= 0.001). While reviewing the characteristics, in patients, that affect their attendance at the general out patient clinic (OPC), McClure and his colleagues[19] observed that low social class and poor housing, among other factors contributed to non-attendance to OPC. This is similar to Lesi's[18] view on the survival of these patients.[13] Such patients are easily lost to follow up as has been the view of Ohene Frampong et al.[20]

Of the 71 children, 33 children (46.5%) last attended the clinic eight months or more before admission. These are children who require counseling and frequent home visits in order to minimize the adverse effect of a poor clinic attendance on survival of these children. . Such children are not in touch with their clinicians until their clinical condition worsens-a situation that contributed to four of the six deaths that were recorded in this review.

As has been noted by Andrews et al,[21] the future health of an ill child is determined by his ability to comply with both his appointment and treatment. Counseling these patients or their parents is vital so as to improve their perceptual responses to their illness. This response is determined by their knowledge and experience of the disease and is low in people from low social group[13] Bamgboye and Jegede[13] have emphasized the positive influence of a sound education on clinic visits.

Infections were the commonest cause of hospital admission. These infections involved the bone, muscles, joints, lungs or liver. This is similar to the observations by Mahrajan et al[7] but contrasts with the findings of Konotey Ahulu[6] who noted that malaria was the commonest cause of admission. It is possible that our use of proguanil for malaria prophylaxis has influenced the place of malaria as a common cause of admission in this environment. Some reasons have been given for the susceptibility to infection in children with sickle cell anaemia. This included immune deficit opsonin defect[22,23] and impaired splenic function.[2] Other factors include neutrophil dysfunction, impaired cell mediated immunity, impaired phagocytosis and the occurrence of tissue ischaemia which provides a suitable environment for the occurrence and persistence of infection.[24] Though no organism was isolated in some of the cultures done, probably, because the patients were already on antibiotics before presenting at the clinic, it is necessary to educate patients on the use of prophylactic antibiotics in these children. When properly used, this practice will reduce the prevalence of infective complications. This practice of home treatment, before presenting to hospital, has been documented.[25]

These children, sometimes, require urgent blood transfusion as their haemoglobin levels can fall very rapidly. Above a third of the children presented with very low haemoglobin of less than 5gms/dl. This finding is similar to that noted by Okoroma[26] in Enugu. The indications for blood transfusion were mainly due to severe anaemia which was as low as 2.4gm/dl in one child . However, blood transfusion was considered in children whose haemoglobin levels, on admission, were observed to be falling in the presence of clinically suspected ongoing infection. Most transfused children came from the low social class and had not complied to treatment and follow up visits. Four of the children had central nervous system affectation and might have benefited from hypertransfusion programme.

Styes and Vichinsky[27] observed that this programe was able to shorten the duration of hospitalization in 4 and 13 patients with intractable pain and cerebrovascular accident respectively. It is pertinent to note that most of the children who did not receive blood transfusion complied with appointment and treatment, hence it was easier to monitor their progress during their visit to the clinics.

There is, therefore, the need to review our own services. There should be close involvement by the home visitor and social worker with regard to monitoring our sickle cell patients. More emphasis should be placed on the non-compliant patients as the children tend to have increasing morbidity with time when they are not regularly reviewed.

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