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Screening For Psychosocial Dysfunction in Children With Sickle Cell Anaemia

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SUMMARY

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In order to find out whether sickle cell anaemia (SCA) is associated with increased risk for psychosocial disorders in Nigerian children, a psychosocial screening questionnaire, the paediatric symptoms checklist (PSC), was administered to parents of 412 children (mean age 9 ± 2.7 years) comprising 204 with SCA (patient group) and 208 with no haemoglobinopathy (control group). Mean PSC score for children with SCA was 19 ± 8.2 and controls 13.3 ± 6.5; median 18 and 12 respectively. Sample mean difference in PSC score between children with SCA and controls was 4.8, 95% confidence interval 3.3 to 6.3; ($p < 0.001$). Within groups, there was no significant difference between scores for boys and girls (SCA, $p > 0.1$; Control $p > 0.7$). However, mean score for boys with SCA (19.1 ± 8.2) was significantly higher than for boys in control group (13.3 ± 6.3) and for girls, SCA sample mean score of 17.3 ± 8.8 was higher than 13.0 ± 6.2 for control girls ($p < 0.001$). Thirty children with SCA (14.7%) and two (0.96%) controls scored ≥28 (cut off score for children with psychosocial problems) ($p < 0.001$). It is concluded that SCA is associated with psychosocial dysfunction in children. Therefore, health care programmes for children with SCA should include routine psychiatric evaluation

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Introduction

SICKLE cell disease is the commonest genetic disorder that affects people of the black race. The sickle gene occurs at high frequencies in

Nigeria, ranging from 25-30% in some communities where it is estimated that up to 2% of all newborns have sickle cell anaemia (SCA)[1]. With a better understanding of the biology of this condition and the general improvement in the care of affected children, many of them will survive to become adults. Sickle cell anaemia is a life-long chronic illness and in childhood, it manifests with frequent episodes of aches and pains, recurrent infections, frequent hospitalization with loss of school time, chronic fatigue, retarded growth and physical deformities.[2]

Physicians caring for children with long-term chronic illnesses, such as sickle cell anaemia, are often advised to give greater attention to the emotional well being, behaviour and social adjustment of these children because it is believed that they are at risk for psychosocial dysfunction.[3] In fact, substantial evidence has accumulated over the past 30 years to indicate that chronic illness in children causes an increase in emotional disorders, both for the children themselves and possibly for their parents and siblings as well.[4 5 6]

However, empirical studies on the psychosocial problems of children with sickle cell anaemia are surprisingly scanty, particularly, in Nigeria.[7 8] The little information available on this subject has been derived from a few studies carried out in the United States some of which have given inconsistent results.[9 10] Other authors have commented on the methodological weakness in some of those studies.[11] Moreover, it is not certain whether conclusions about children with a chronic illness in a different cultural milieu can be applied to Nigerian children.

The purpose of the present study was to define the mental health status of children with sickle cell anaemia and to find out whether emotional problems are associated with this condition in Nigerian children. We report here our preliminary findings in a screening for psychosocial dysfunction among children with SCA and controls with no haemoglobinopathy.

Materials and Methods

The subjects were 412 children (182 boys and 230 girls) aged 6-16 years. Children in this age group were chosen because we are aware that many stresses and most psychosocial disorders become manifest during the school age. Also, the instrument used in the study was most appropriate for the age group. The samples were drawn from two groups of children attending the paediatric haematology clinic and a general out patient clinic in the University of Benin Teaching Hospital (UBTH). A four factor index (father's level of education, occupation, employment and income status) was used to stratify the subjects according to socio-economic status (SES). Accordingly, arbitrary scores of 3,2,1 and 0 were assigned to the factors as follows, 3 points for completed post secondary education (University/Technical), occupation in professional or senior executive cadre (Administrative/Technical), continuous employment in the last 12 months, and income not less than ₦770.4 p.a. (GL. 12); 2 points for completed secondary education (grammar or technical/vocational), occupation rated as skilled (Senior Clerical/Technical) continuous employment for not less than 6 months in the last year, and income not less than ₦3,864 (GL.08); 1 point for completed primary school, occupation ranked as clerical, skilled or semi skilled manual, continuous employment for

less than 6 months in the last year and income not less than ₦1800 (GL.04); and zero for less than 6 years and of primary education, occupation rated as unskilled manual, no regular employment in the last 12 months and income less than ₦1800 p.a. The scores were summed and total score was used to determine the SES class as follows, 12 points class I, 8-11 points class II, 4-7 points class III and 0-3 points class IV.

Patients Group

Two hundred and four children (92 boys and 112 girls) in whom the diagnosis of SCA had been made on paper electrophoresis of the haemoglobin constituted the patient group. Their ages ranged from 6 years 3 months to 16 years 5 months and they were attending the paediatric haematology clinic routinely at intervals ranging from 2 – 3 months. In order to minimise the effects of any acute anxiety response that may be associated with current or recent pain crisis, we adopted the following criteria for selection of the children: (i) diagnosis of SCA was made not less than 6 months before the study; (ii) the child was in a steady state at the time of study, (iii) age was 6 – 16 years, (iv) the child has not been hospitalised in the preceding 3 months, and (v) the child had been attending school for at least, 6 months. Twenty five (12%) of the children came from families in SES class 1 (high), thirty seven (18%) from class II, one hundred and six (53%) from class III and seventy six (37%) from class IV (Low).

Control Group

For comparison, 208 children (90 boys and 118 girls) were chosen from 400 consecutive first attenders at the general out patient clinic (GPC) from 1st April through 30th September, 1987, that were aged 6 – 16 years and met certain predetermined conditions. Haemoglobin electrophoresis pattern was not always available for these children, so, we adopted the following criteria for their selection: (i) child had been attending school for at least 6 months, (ii) had not been hospitalised in the preceding 3 months and (iii) presenting illness had not lasted longer than one week. We excluded all children with past history of blood transfusion, jaundice and recurrent bone pains, as well as those in whom the attending senior medical officer (formerly a Registrar in the Department of Child Health) suspected the diagnosis of a haemoglobinopathy. We reasoned that in our environment, it was unlikely that a child with SCA would lack a past history of some suggestive symptoms by 6 years of age. The ages of the children ranged from 6 years 8 months to 16 years 3 months. Seventeen (8%) of these children came from families in SES, class 1, forty four (21%) from class II, one hundred and twenty (58%) from class III and twenty seven (13%) from class IV.

Screening Instrument

The questionnaire used for the study was a revised version of the paediatric symptoms check list (PSC), a brief psychosocial screening questionnaire with 35 item that identifies children with psychosocial problems. Items in the questionnaire include questions based on major diagnostic categories listed in the American Psychiatric Association DSM-III[12] and some items from other instruments that in the opinion of many paediatricians, clinical psychologists and child psychiatrists, are considered most suitable for identifying children with emotional problems. Parents are requested to

rate each of the items in PSC as 'often' 'sometimes' or 'never' present. The ratings are scored 2, 1 and 0 respectively and scores on individual items are summed for a total score. Paediatric symptoms check list has been found to have a high degree of internal consistency and a satisfactory test-retest reliability.[13] The revised PSC has been well validated and is known to correlate with the widely used child behaviour check list (CBCL) of Achenbach and Edelbrock.[14] A total score of 28 on the revised PSC has been recommended as the cut-off score for detecting children with psychosocial disorders.

Methods

On clinic days during the period of the study, the purpose of the investigation was explained to parents (usually mothers) of eligible subjects stressing that the investigation was optional and not part of the child's treatment, that the findings were confidential but that results for the individual child would be made available to his or her parent on request. Only those whose parents gave consent were included in the study. In the sickle cell sample all the parents approached gave consent for their children to be included in the study but 12 (5.5%) of the 220 parents approached in the control group did not agree. They gave no reason for refusing their children's participation in the study. The questionnaire was administered by one trained interviewer in English, "pidgin English" or Edo language. There was no substantial modification of the original PSC but because we felt that two items (1 and 3) "Complains of aches and pains" and "Tires easily, little energy" would tend to give sickle cell patents high scores we did not include scores on these items in the calculation of the summed scores. Furthermore, the language of some other items were slightly modified in order to convey the same idea in the local parlance. For example, item 19, "Is down on himself or herself" was changed to 'He hates himself, wishes bad things for himself.'

Statistical Methods

The significance of comparisons between mean values for the groups was evaluated by the Student's 't' test for unpaired samples. Frequency distributions were compared in appropriate contingency tables and significance of differences evaluated by the Chi-square test with Yates correction. The 95% confidence limits (CI) for differences were calculated where appropriate.

Results

The clinical characteristics of the subjects are summarized in Table I, and the distribution of their scores on PSC shown

TABLE I
Summary of Clinical Details of Patients and Control

Characteristics	SCA Patients	Controls	All Subjects
Total No of Children	204	208	412
Sex: M/F	92/112	90/118	182/230
Mean Age (Yrs) ± SD	9.2 ± 2.7	8.8 ± 2.6	9.0 ± 2.7
SES Distribution (%)			
Class I	12	8	9.9
Class II	18	21	19.7
Class III	53	58	55.3
Class IV	17	13	15.1

in Table II. Children with SCA (patient group) were comparable to those with no haemoglobinopathy (control group) in age, sex and SES distribution.

TABLE II
Distribution of Scores on PSC According to Subject Groups

PSC Score	Sickle Cell		Controls		All Subjects	
	n = 204		n = 208		n = 412	
	No	%	No	%	No	%
Below 10	28	13.7	72	34.6	100	24.3
10 – 15	60	29.4	70	33.6	130	31.6
16 – 21	54	26.5	44	21.2	98	23.8
22 – 27	32	15.7	20	9.6	52	12.6
28 – 33	16	7.8	1	0.5	17	4.1
34 – 39	14	6.9	1	0.5	15	3.6

PSC = Paediatric Symptoms Checklist

The difference between the (sample) mean ages (0.4 years) was not statistically significant ($t = 1.5$, df 410, $p>0.1$). Similarly the sex distribution between the patient and control groups was not significantly different ($X_12 = 0.08$, $P>0.25$).

Most of the children (75%) came from families in the middle class (SES 11 and 111). There was a slight excess of children from families in SES class 1 in the SCA sample (12%) over the control (8%). Similarly the proportion of children in SES class IV was slightly higher in the patient group (17%) than in the control group (13%). The differences in these distributions were, however, not significant ($X_3^2 = 4.05$, $P> 0.1$).

The main scores on PSC for children in the two groups, their median scores as well as the proportion of children in each group scoring at or above the cut off score of 28 are given in Table III. The difference between sample mean score on PSC for children with SCA and controls was 4.8, with a 95% confidence interval from 3.3 to 6.3, ($t = 6.4$, DF = 410, $P>0.001$). Half of the children in the sickle cell group scored above 18 on PSC and half of those in th control group scored below 12. Thirty children (14.7%) in the sickle cell group scored 28 or above on the PSC indicating that these children probably have psychosocial disorders, and hence, need further psychiatric evaluation. By contrast, only two (0.096%) children in the control group scored 28 on the PSC ($X_1^2 = 31.9$, $P<0.001$).

TABLE III
Scores on PSC, Mean, Median and Proportion Above Cut-off Score According to Subjects Group

PSC Score	SCA Patients n = 204	Controls n = 208
Mean ± SD	18.0 ± 8.6	13.2 ± 6.5
Median	18	12
≤ 28	30 (14.7%)	2 (0.96%)

Figures in parenthesis = percentages of total number of children in the group.

TABLE IV
Mean Scores on PSC for Children with SCA and Controls Comparison by Sex

	Subject Group		Comparison	
	SCA	Control		
	No Mean ± SD	No Mean ± SD	t	P
Boys	92 19.1 ± 8.2	90 13.3 ± 6.3	5.0	<0.001
Girls	112 17.3 ± 8.8	118 13.0 ± 6.2	4.3	<0.001

The mean scores for the two groups of children according to sex is shown in Table IV. There was no significant difference between the scores for boys and girls either in the control or patient group, although the mean score of 19.1 ± 8.2 for boys in the sickle cell group was slightly higher than the 17.3 ± 8.8 for girls, ($t = 1.5$, $df = 202$, $p>0.1$). However, the mean score of boys in the patients group was significantly higher than the mean for boys in the control group ($t = 5.0$, df 180, $p<0.001$), while the mean score for girls in SCA group was significantly higher than that for girls in the control group ($t = 4.3$, df 228, $p<0.001$) Table IV.

Discussion

The results of this study show that symptoms of psychosocial disorders are more prevalent in children with SCA than in controls with no haemoglobinopathy. The PSC is a general screening instrument that does not classify children into specific diagnostic categories of psychiatric disorders. Hence, the study does not indicate the specific nature of psychosocial disorder associated with SCA in children; neither does it give indication of the risk for such disorders to children with SCA in the general population.

Nevertheless, our study provides quantitative evidence to support earlier impression from anecdotal reports that SCA in childhood is frequently associated with psychosocial dysfunction. Thus, SCA may increase the risk of psychiatric disorders in children. Whitten and Fischolf have extensively discussed various aspects of SCA that are stressful and can lead to maladjustment and other forms of psychosocial disorder in children.[7]. Bamishaiye, Bakare and Olatawura have drawn attention to the socio-psychological consequences of SCA in Nigeria[8], and others[15] have commented on the presence and manifestation of psychiatric disorders in children with SCA. Similarly, workers in Ghana, have described the psychosocial problems of some children with SCA[16]. A wealth of information as accumulated in the Western literature on the psycosocial effects of various chronic health conditions in children.[17–21]

We do not know of any study on the psychosocial disturbance in children with SCA that has been carried out on a large sample as in other chronic childhood illnesses. Pela and Okafor,[22] studies 30 patients (age 15 – 37 years) with SCA and found that they, like patients with mental illness, had low self-concept. Although these workers found their subjects to have a more positive attitude towards SCA than to mental illness, they commented that society has the same negative feeling towards SCA and mental illness.

The study from Detroit[9] involving 19 children with SCA paired with their siblings who had no haemoglobinopathy reported that SCA did not affect intellectual or psychological functioning in children. Kumar and others[10] found that children with SCA in California did not differ from their peer group in personal, social and total adjustment. However, the latter workers found that self-concept scores were lower in children with SCA who, surprisingly, also had lower scores on the anxiety scales. We think that discrepancies in these studies may be due to small sample sizes.

In the present study, 14.7% of the children with SCA were found to have symptoms suggestive of psychosocial dysfunction and, therefore, in need of further psychiatric evaluation. This is comparable to 12% found

by others using PSC in a group of children in a paediatric waiting room.[13] The number of children identified in our sample may not represent all those at risk since we did not include scores on two items in the questionnaire relating to symptoms known to be common among children with SCA. If we did, then the prevalence rate in the sickle cell sample would have increased to 20% and the control group unchanged.

Thus, we conclude that SCA, like other chronic illnesses, increases the risk of psychosocial dysfunction in children. There fore, psychiatric evaluation should form part of comprehensive health care programme for children with SCA.

Acknowledgements

We are grateful to Dr (Mrs) A R Kubeyinju, Senior Medical Officer, General Out-Patients Department, University of Benin Teaching Hospital for her help in the selection of patients for control and to Messrs J J Ukhuriegbe and M B S Momoh for helping in the preparation of the manuscript.

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