ORIGINAL ARTICLE

Adolescents with sickle cell anaemia: Experience in a private tertiary hospital serving a tertiary institution

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ABSTRACT

Background: Many adolescents with sickle cell disease (SCD) have adjustment difficulties in the transition period from paediatric care to the adult system because they find themselves in unfamiliar waters where they have to learn to manage themselves. The aim of this study is to evaluate the prevalent crises and morbidities associated with SCD in adolescents in Babcock University Teaching Hospital (BUTH), to also assess the level of knowledge of these adolescents about SCD and to determine their emotional response to the disease. **Materials and Methods:** This was a retrospective review of case notes of adolescents with sickle cell anaemia that were seen in BUTH, from May 2013 to April 2014. Data extracted from the case notes was entered into a Microsoft (MS) Excel and analysed using descriptive statistics. Results were presented in tables. **Results:** A total of 50 subjects were seen in the department during this study period. Vaso-occlusive crises in the form of bone pains (93.1%) were the commonest crises encountered. Associated morbidities were malaria 34 (85%), tonsilitis 1 (2.5%), pneumonia 1 (2.5%), leg ulcer 1 (2.5%), azotaemia 1 (2.5%) and subarachnoid haemorrhage 2 (5%). Majority (88%) had adequate knowledge about general health maintenance while knowledge on nutrition and appropriate analgesia use is still inadequate. Eleven (22%) had symptoms of depression, four (8%) had suicidal ideation while one (2%) had a history of attempted suicide. **Conclusion:** This study emphasizes the importance of psychosocial intervention as part of a comprehensive health management for people with SCD.

Key words: Adolescent, psychosocial intervention, sickle cell anaemia

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INTRODUCTION

The commonest genetic disorder among Africans is sickle cell disease (SCD).[1,2] In Nigeria, the prevalence of sickle cell trait is about 25% while the homozygous state is found in about 3% of the population.[3] Nigeria has the largest population of people with SCD, with about 150,000 births annually.[4,5]

The aim of this study is to evaluate the prevalent crises and morbidities associated with SCD in adolescents in Babcock University Teaching Hospital (BUTH), to also assess the level of knowledge of these adolescents about SCD and to determine their emotional response to the disease.

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MATERIALS AND METHODS

This was a retrospective review of case notes of adolescents with sickle cell anaemia (aged 17-24 years) that was seen in the emergency room and Haematology Clinic of BUTH, Ogun State, Nigeria from May 2013 to April 2014. The Haematology Clinic of BUTH operates once a week and receives referrals from the Babcock University, other units within the Clinical Department in the hospital and its environs. Only adolescents with haemoglobin genotype SS established by the cellulose acetate haemoglobin electrophoresis were included for this study. Those with incomplete data were excluded. Data extracted from the patients' case notes included psychological assessment, age, gender, genotype, history of blood transfusion, complications and crises.

Data was entered into a Microsoft (MS) Excel Spread sheet and analysed using descriptive statistics. Results were presented in tables.

RESULT

A total of 50 adolescents were seen in the hospital during this study period. Of these 50 (aged 17-24 years), 25 (50%)

were males while 25 (50%) were females giving a male/female ratio of 1:1. The mean age was 21.2 ± 5.2 years. Forty-nine (98%) were students while one (2%) subject a nurse [Table 1].

Vaso-occlusive crises (VOC) in the form of bone pains (93.1%) were the commonest crises encountered [Table 2]. In total, 108 episodes of bone pain were documented giving an average rate of 2.6 episodes per patient. The average duration for each episode was about 7 days.

Three (6%) adolescents gave a history of previous blood transfusions (ranging from one to two). Associated morbidities were malaria 34 (85%), tonsilitis 1 (2.5%), pneumonia 1 (2.5%), leg ulcer 1 (2.5%), azotaemia 1 (2.5%) and subarachnoid haemorrhage 2 (5%) [Figure 1].

Majority (88%) had adequate knowledge about general health maintenance while knowledge on nutrition and appropriate analgesia use is still largely inadequate [Table 3]. Eleven (22%) had symptoms attributable to depression, four (8%) had suicidal ideation while one (2%) had a history of attempted suicide.

Table 1: Socio-demographic data

Characteristic	Category	*n*	%
Gender	Male	25	50
	Female	25	50
Occupation	Student	49	98
	Nurse	1	2
Religion	Christian	48	96
	Muslim	2	4

Table 2: Type of cickle cell crises

Crises	*n*	%
Bone pain	41	93.1
Hyperhaemolytic	2	4.6
Priapism	1	2.3

Table 3: Subjects' knowledge about sickle cell anaemia

General health maintenance	Aware *n* (%)	Not aware *n* (%)
Avoid exposure to extreme weather	50 (100)	
Avoid strenuous exercise	50 (100)	
Regular hydration	50 (100)	
Avoid alcoholic beverages	50 (100)	
Regular use of routine drugs	28 (56)	22 (44)
Symptoms requiring medical advice	32 (64)	18 (36)
Appropriate use of analgesia at home	38 (76)	12 (24)
Avoid tobacco use	49 (98)	1 (2)

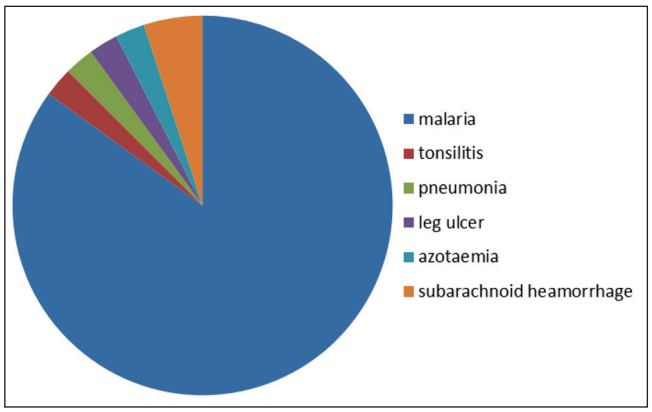

Figure 1: Morbidities associated with SCA

DISCUSSION

Previous reports have described SCD as a condition of childhood with most youths not living past their teens.[6,7] Improvement in the diagnosis and treatment of many of SCD's life-threatening conditions over the past decade has led to an extended life expectancy with many living well into their 40s.[8] Many adolescents with SCD have to deal the psychological burden of having a chronic disease, the disadvantage of being categorised as socially dysfunctional.[9,10] As a result, there is clinical consensus that added attention needs to be focused on the period from adolescence to young adulthood.

To facilitate adolescents with SCD live a longer and healthier life, they should be taught to manage their illness. LePontois and others[11-14] are of the opinion that there is need for adolescents with chronic conditions, like SCD, to develop both short- and long-term psychological and social skills needed to successfully navigate this developmental period and the medical and social systems in which they function.

Acute episodes of bone pain signalling marrow ischaemia or necrosis is pathognomonic of SCD and is the most common cause of hospital admissions. These recurrent attacks of acute pain are serious complications that often require treatment with parenteral opioids, in the Emergency Room (ER) and/or the hospital.[15-17]

This study reveals that bone pain is the commonest crises necessitating hospital admission as documented in previous studies.[18,19] The episode of pain per patient is higher than that documented by Platt *et al*.; this could be a consequence of the academic stress the subjects are experiencing. However, the average duration of pain of 7 days is consistent with literature.[15]

In this study, the commonest indication for blood transfusion was severe anaemia; this is consistent with the finding of Otaigbe in Port Harcourt, Nigeria.[20] This is however different from studies in developed countries where the indication for transfusion were variable with several studies showing a decline in complication rates among sickle cell patients.[21,22]

Nigeria is a malaria-endemic region; so, it is not surprising that the prevalent morbidity recorded in these subjects was malaria. This finding is also consistent with results from other studies.[18,23] The fact that malaria infection precipitates VOC could also account for bone pain crises being the commonest presentation of sickle cell crises recorded in this study.

It was quite interesting to note that majority of the subjects had adequate knowledge about general health maintenance; though on further questioning, many of the subjects do not actually stick to their care plan. The reluctance to use routine medications, to avoid conditions that can precipitate VOC and attend follow-up appointments is all part of the adjustment difficulties documented in chronic medical conditions like SCD.[24-26]

Consistent with the literature,[4,27-29] a significant number of the patients in this study had feelings of depression. However, it was noted that subjects who had more knowledge about their condition with strong family support had a more positive outlook about their disease; improved coping skills and better adherence to their routine medications which could be responsible for reduced hospital visits.

CONCLUSION

The study shows that the majority of adolescents who have better knowledge about their condition with strong family support have a positive perception of their illness which equates not only better control of their illness but also better quality of life with reduced hospital visits. This study emphasizes the importance of psychosocial intervention as part of a comprehensive health management for people with SCD.

The onus is on health-care providers to work closely with the family to ensure that there is a smooth transition from paediatric to adolescent care by providing as much information as possible to assist the affected adolescent in navigating this turbulent period.

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