

LEG ULCERS IN ADULT SICKLE CELL DISEASE PATIENTS IN BENIN CITY NIGERIA

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ABSTRACT

Background: Chronic leg ulceration is the most common cutaneous manifestation of sickle cell disease. In this study we observed adult sickle cell disease patients with leg ulcer.

Methodology: In this observational study, patients with sickle cell disease at University of Benin Teaching Hospital and Sickle Cell Centre Benin, from July 2008 to June 2009 were recruited. After informed consent 273 patients were selected but only 250 (125 males and 125 females) were interviewed and completed a pre-tested questionnaire. Data included socio-demographic information, history of previous leg ulcers and current ulcers confirmed by examination. In those currently with leg ulcers, haemoglobin phenotype, stable haemoglobin (calculated as average haemoglobin in the absence of vaso-occlusive crises in past 3 months, site, diameter and status of leg ulcers were documented. Data were analyzed using SPSS version 11.

Results: In sickle cell disease patients leg ulcers were found currently in 24(9.6%) and previously in 56(22.4%), while in sickle cell trait it was 0% and 2(0.8%) respectively. Leg ulcers were more prevalent in males than females (M:F=3:1), in those with lower stable haemoglobin 6.1–8g/dl (58.4%), age range of 18-25 years (66.7%), around left lateral ankle (66.7%) and with average diameter of 5-10cm (58.4%). Aetiology was mainly traumatic (58.4%) with 33.4% as traditional traumatic scarification. Greater than one year recurrence was 50%, and ulcers were septic in 83.4%.

Conclusion: A lower stable haemoglobin, ignorance and poverty play important role in worsening chronic leg ulcers in sickle cell disease. Improved health education, prompt medical and surgical intervention could improve these leg ulcers.

KEY WORDS: Leg ulcer, Sickle cell disease, Haemoglobin.

INTRODUCTION

Sickle cell anaemia is an inherited haemolytic anaemia resulting from inheritance of haemoglobin S (HbS).¹ Chronic leg ulcer is defined as a defect in the skin below the level of knee and above the foot persisting for six or more weeks.² Leg ulcers are the most common cutaneous manifestation of sickle cell disease. Their incidence is reported from 8% to 75%.^{3,4} The aetiology of leg ulcers in SCD is not known but is thought to be due to micro-thrombi in the small capillaries of the legs resulting in ischaemia.⁴⁻⁶ This obstruction is said to up regulate integrins which promote platelet aggregation and adherence to the endothelium with additional obstruction of the vessels. Other associated factors include release of injurious cytokines by interaction of granulocytes with sickle cells, thrombocytosis, anaemia, lower levels of fetal haemoglobin, antithrombin deficiency, trauma, possession of certain HLA such as HLA

B₃₅, CW₄, males, presence of lupus anticoagulants and living in certain geographic areas.^{3,4,7-9}

Therapy for SCD leg ulcers comprise of prevention, treatment of existing ulcers and infection. Prevention includes the use of properly fitting shoes, prompt treatment of minor injuries, leg elevation and salt restriction. Treatment include topical dressing, systemic medication with zinc, and hydroxycarbamide intervention.¹⁰⁻¹² Interventions such as autologous split thickness skin grafts are advocated for recalcitrant ulcers.¹³

Previous studies on leg ulcers in the index centre were retrospective on children. This was an observational study of adult SCD patients with leg ulcers.

MATERIAL AND METHODS

Patients with sickle cell disease attending clinics at the university of Benin teaching hospital (UBTH) and Sickle cell centre Benin City from July

2008 to June 2009 were randomly recruited into the study. After informed consent a total of 273 patients were selected but only 250 SCD patients (125 males and 125 females) were interviewed and completed a pre-tested questionnaire. Data collected included their socio- demographic information, history of previous leg ulcers and current ulcers (confirmed by examination). In those SCD patients currently with leg ulcers, haemoglobin phenotype, stable haemoglobin (calculated as average haemoglobin in the absence of vaso-occlusive crises (VOC) or other crises in the past 3 months routine clinic visits were confirmed from their case notes) sites, diameter and status of leg ulcers were documented. Data were analyzed as frequency and percentage using the SPSS version 11 package and results presented in table, bar charts and graph.

RESULTS

Two hundred and fifty patients with SCD were enrolled in this study. HbSS patients made up 88% of the patients studied compared with 12% for HbSC. A previous history of leg ulcers was recorded in 56 (22.4%) and 2 (0.8%) of HbSS and HbSC respectively. Twenty four HbSS patients currently have leg ulcers 24 (9.6%), no HbSC patient had leg ulcers during this study. Leg ulcers were more common in males (ratio 3:1) than females. The age distribution of patients with leg ulcers as shown in fig 1 were mainly between 18-25 years (66.7%). Multiple ulcers were mainly single 12 (41.7%) and the left ankle was more commonly affected (66.7%). Ulcers were more common on the lateral aspects of the legs 98.9% with diameter mainly of 5-10cm (58.4%). Most patients with leg ulcers had a steady

Table I: Sickle cell disease and leg ulcers.

SCD	Frequency		Hx of previous leg ulcer		Current leg ulcers	
	n=250	%	n=250	%	n=250	%
HbSS	220	88.0	56	22.4	24	9.6
HbSC	30	12.0	2	0.8	—	—
Total	250	100.0	58	23.2	24	9.6

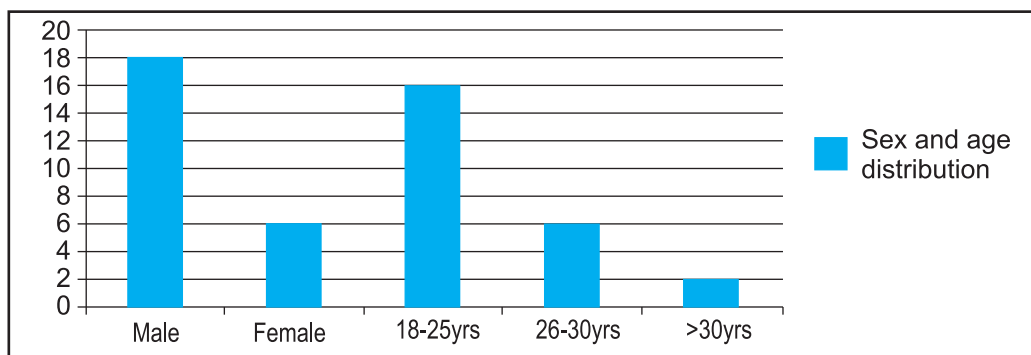


Fig. 1: Sex and age distribution of sickle cell disease patients with leg ulcers.

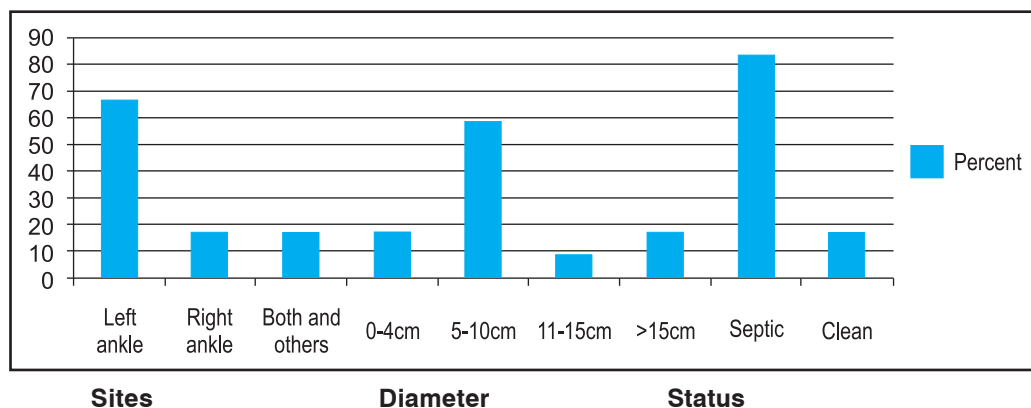


Fig. 2: Site, diameter and status of leg ulcers in sickle cell disease.



Fig. 3: Leg ulcer in a patient with sickle cell disease.

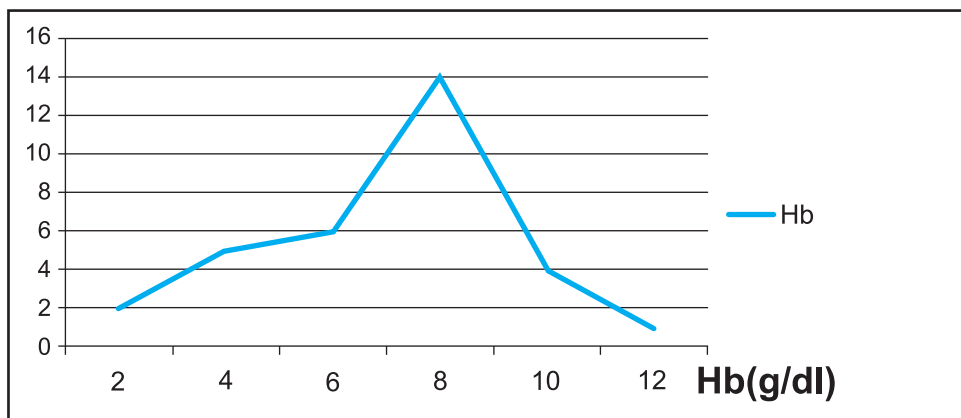
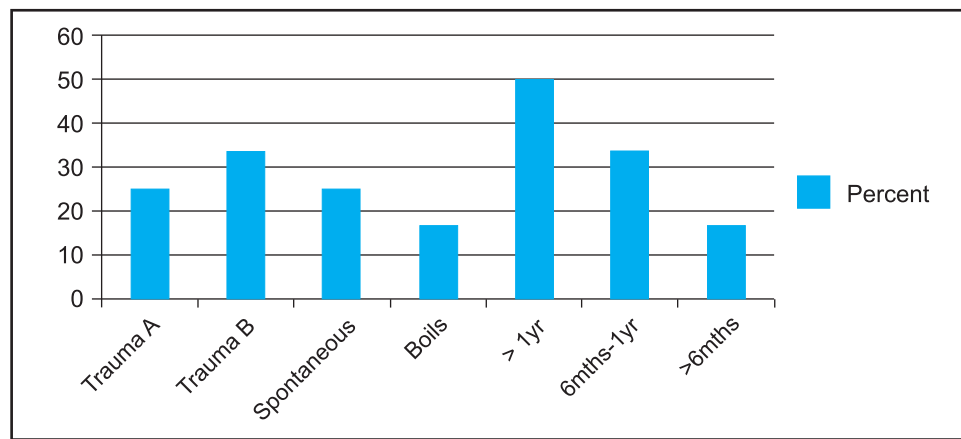


Fig. 4: Stable haemoglobin of sickle cell disease patients with leg ulcers.



Associated factors

Duration of ulcers

Trauma-A (other traumatic factors), Trauma-B (traumatic traditional scarification)

Fig. 5: Associated factors and duration of leg ulcers in sickle cell disease.

state Hb concentration in the range 6- 8 g/dl (58.4%). Most patients associated the onset of the leg ulcer with some form of trauma (traditional scarification 33.4%). Half of the patients had had the leg ulcers for more than a year, the ulcer had recurred in 75% of cases. Only 13 (15.9% previous history: current leg ulcers 11/2) of these patients have attempted surgical skin grafting and 5 (38.4%) was successful.

DISCUSSION

In this study leg ulcers were more common in patients with HbSS than HbSC in agreement with earlier studies.¹⁴ The lower prevalence of leg ulcers in HbSC patients may be due to their higher steady state Hb.¹⁵ This is also reflected in the lower frequency of leg ulcers in HbSS patients with steady state Hb higher than 10 g/dl. More males had leg ulcers in this study than females which is also similar to reports in other centres.¹⁵⁻¹⁷ This finding was at variance to an earlier finding in a retrospective study which included children from our center that reported a slightly higher female to male ratio (M;F=1:1.08).¹⁸

More than half of the SCD patients with leg ulcers were in the age group of 18-25yrs, the frequency decreased with increasing age of the patients, similar to earlier reports.¹⁵ This could be explained by the fact that this age group is very active, prone to injuries and may reflect why the ulcers are mainly on the outer parts of the ankle.

There was a positive past history of leg ulcers in 23.2% of sickle cell disease patients and 12 (9.6%) currently had leg ulcers. This finding is similar to published data both previously in our centre¹⁸ and other parts of Nigeria¹⁶ but less than reports from Jamaica (75%).³ The difference in regional data may be related to suggested genetic or environmental predisposition as earlier suggested in pathogenesis.⁹

Majority of the leg ulcers were located on the left ankle which agrees with other reports^{15,16,18} although the reason for this preference is not clear. Over half of the SCD had a diameter of 5-10cm, the large diameter of the ulcer may be a reflection of possible secondary infection of these ulcers. The most frequent associated factor was trauma, including traditional scarification which further reflects poverty and ignorance. Over half of the patients have recurrence which is a known feature of leg ulcers in SCD.¹⁵

CONCLUSION

Leg ulcers are common in sickle cell disease. Ignorance and poverty play an important role in

this chronic event worsened by lower stable haemoglobin.

Improving stable haemoglobin with agents like hydroxycarbamide, health education prompt medical and surgical intervention may reduce the incidence of chronic leg ulcers.

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