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# Hyperventilation - precipitated cerebrovascular accident in a patient with sickle cell anaemia

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**Summary**
Hyperventilation exercise during electroencephalography precipitated a recurrence of right hemiplegia and aphasia in a patient with Hb SS disease. Although recovery of function started within hours of the event, full recovery has not occurred six months after. Hyperventilation provocative test during electroencephalography should be discouraged in patients with sickle cell anaemia.

**Keywords:** *Sickle cell anaemia, cerebrovascular accident stroke, hyperventilation, electroencephalogram*

**Résumé**
L'exercise de hyperventilation an cour de l'electroencephalographie a subitement provoque un retour de hemiplegie et d'aphasie du cote droit d'un patient atteint de maladie de hemoglobine de drepanocytase.Bien que la reprise de function a commence en quelques heures durant l'incident, le plein recouvrement ne s'est product qu'apres six mois.L'essai d'hyperventilation declenche a dessein au cour de l'electroencephalographie devrait etre decourage aupres des patients atteints d'anemie a hematies falciformes.

## Introduction

The request for electroencephalogram (EEG) in patients with sickle cell anaemia is not uncommon, as seizure disorder is one of the central nervous system complications of this disease [1]. Hyperventilation for 2-4 minutes is a routine provocative test conducted during EEG recording in most centers. Although there have been references in the literature to the development of cerebrovascular accident (CVA) in patients with Hb SS disease secondary to hyperventilation [2,3,4] this complication has not been recorded in our center. We did not until now discourage this provocative test in our patients. We report here our experience with one patient, in the hope of alerting doctors looking after patients with sickle cell anaemia to the potentially detrimental effect of hyperventilation in these patients.

## Case report

U.D. a 13-year-old girl with sickle cell anaemia presented in the hospital on the 27th of April, 1999 with a 2 - day history of low grade fever and recurrent seizures. At the onset of this illness, she had had several episodes of repeated unsynchronised brief jerky movements of all 4 limbs, each episode lasting about 1 minute and with intervals of about 10 minutes. The seizures finally stopped after an intramuscular injection of 4 ml of Paraldehyde given by a nurse. The following day, two similar seizure episodes occurred lasting 5 minutes and 15 minutes with interval of one hour in between. There were no seizures on the day of presentation. Her mother, since the onset of the fever, had placed the child on oral chloroquine and paracetamol. She was an old patient of the paediatric haematology unit of the University College Hospital, a diagnosis of sickle cell anaemia having been made at the age of 5 ½ months.

She had had several episodes of malaria and bone pain crises in the past. She was admitted to hospital once on account of severe anaemia (haematocrit 11%) for which she was given packed red cell transfusion.

In the 2 years preceeding the present hospitalisation, she had had 6 episodes of sudden right hemiplegia which lasted from a few hours to a week in each case. Partial exchange transfusion had been given during the second episode which had lasted 7 days. Total recovery of function followed the first 4 episodes. The fifth episode which occurred in November 1998 was associated with slurred speech (which resolved after 2 days) and left her with a residual mild right hemiparesis, power in the right upper and lower limbs being grade 4. She could write slowly with the right hand and could walk and even run though with a hemiplegic gait.

An electroencephalogram was ordered on the day of presentation of current illness. She was reported to have been very cooperative during the procedure and performed the hyperventilation provocative exercise with vigor. Immediately following the EEG record, she could not get off the couch, the right arm and right leg being limp. She was also aphasic. Power in the right - sided limbs was 0 while it was 5 on the left. Speech resumed 4 hours later and was normal by the next day. Power in the right upper limb increased to grade 3 within a few hours and to grade 4 by the next day. Power in the right lower limb increased to grade 3 after 1 week.

The EEG record showed prominent β activity bilaterally with frequent sharp waves especially in the right parieto-occipital region. Hyperventilation resulted in slowing of the record with exaggeration of the previous abnormalities. Paroxysmal discharges were noted especially on the right side.

Malaria parasites were found in the blood film on the day of presentation and these cleared with a full course of chloroquine. The haematocrit on presentation was 21%. A partial exchange transfusion was performed using Hb A blood. Post-transfusion haematocrit was 35%. She was placed on Carbamazepine and has been seizure free in the last 6 months. She is also receiving active physiotherapy. Six months after the episode described above, full recovery

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has not been achieved as evidenced by the persistent poor handwriting and dragging of the right leg, both of which are worse than the pre-episode state.

**Discussion**

Hyperventilation is known to induce EEG slowing as a result of cerebral hypoxia caused by constrictriction of cerebral vessels which in turn results from reduction in arterial $PCO_2$ [5]. The induced hypoxia may precipitate abnormal and paroxysmal discharges typical of epilepsy on the EEG record. Such abnormal discharges provoked by hyperventilation which help with diagnosis may otherwise be absent in the interictal record even in a known epileptic. The EEG record of this patient is typical of that of a patient with seizure disorder especially affecting the left side of the brain: it is however not diagnostic of CVA.

Hypoxia favours intravascular sickling in patients with sickle cell anaemia and when this is combined with constriction of the cerebral vasculature, a cerebral ischaemic attack can be easily precipitated especially when vessel disease is already present.

Although there have been few reports in the literature of CVA precipitated by hyperventilation in patients with Hb SS disease [2,3,4], we have not seen any such cases at our center. The paediatric haematology unit of the University College Hospital, Ibadan, Nigeria looks after a large group of children with sickle cell anaemia (about 1,000 actively attending the clinic). Seizure disorder is a recognized CNS complication of sickle cell disease and is seen in some of our patients. EEG record with hyperventilation is routinely ordered in these patients including the present case who had had a previous EEG test. we have however not until now seen a patient develop complications from the procedure.

It is notable that, like most of the patients reported in the literature [2-4], our patient had had previous episodes of stroke. The risk of stroke precipitated by hyperventilation, although uncommon as judged by the paucity of reports in the literature, is higher in patients with previous episodes of CVA. However, one of the cases reported by Allen *et al.* [3] had no previous central nervous system impairment before the hyperventilation-precipitated CVA. Also, there have been recent reports of extensive CNS vascular disease existing in patients with sickle cell anaemia even when such patients are asymptomatic [6,7]. Considering the fact that recovery of motor function in this patient is yet to reach the pre-attack level even 6 months after the event, we at our center have now decided to omit the hyperventilation exercise during EEG recording in patients with sickle cell anaemia; whether or not they have had previous episodes of CNS involvement. This decision is prudent in view of the fact that the CVA may result in a permanent deficit or even death of the patient. In view of the previous episodes of stroke in the reported patient, it may be argued that the later stroke was coincidental with the hyperventilation rather than being caused by it. However the temporal relationship of the 2 events cannot be overlooked. It is well documented that the majority of CVA in patients with sickle cell disease is secondary to marked narrowing of the lumen of the large cerebral arteries due to thickening of the intima and media layers [8]. Arterial spasm provoked by hyperventilation can then easily result in complete occlusion of the vessel and manifestation of stroke. Omission of the hyperventilation exercise during EEG recording may cause a slight increase in the number of negative studies in epileptics but this is a small price to pay for adverting the potentially fatal complication of stroke - rarity of this complication not withstanding. We advice that other centers exercise the same caution when dealing with patients with sickle cell anaemia.

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