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Cerebral Blood Flow Velocities Measured by Transcranial Doppler Ultrasonography in Children with Sickle Cell Disease in Africa.

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Abstract

Background: Sickle cell anemia (SCA) is one of the commonest monogenic disorders, with 90% of the world's population living in sub-Saharan Africa. Cerebrovascular accident (CVA) is a major cause of morbidity, but its clinical prediction in resource rich countries has allowed effective primary and secondary prevention. Measurements of time-averaged maximum of the mean (TAMM) cerebral blood flow velocity (CBFv) in the internal carotid/middle cerebral (ICA/MCA) arteries by Transcranial Doppler (TCD) ultrasonography and of mean overnight oxyhemoglobin saturation (SpO_2) have been useful in predicting CVA. The criteria used in Western populations may not be appropriate to children living in Africa.

Aims: The aims of this study were to evaluate the TAMM CBFv in patients with SCA in Kilifi district hospital, Kenya, to assess risk factors associated with high ICA/MCA TAMM CBFv and to examine any association with neurological complications.

Study design: This was a cross sectional descriptive study, where TCD ultrasonography was performed on all SCA patients attending the outpatient clinic at CGMR-C, Kilifi, Kenya in 2002. Previous data from 1990 and follow-up data from 2004 were included.

Results: In 140 patients with SCA, aged 3 months to 16 years, the median ICA/MCA TAMM CBFv was 116cm/sec (SD 38, range 0–219 cms/s) compared with 97 (SD 24, range 46–190) cm/sec in 142 controls aged 2 months to 14 years (p=0.0001). 28 SCA patients (20%) had TAMM CBFv greater than and 16 (11%) had TAMM CBFv less than 2 standard deviations from the mean for controls in one or both ICA/MCA's, but only seven (5%) had a velocity above 170 cm/sec (one >200cm/sec), with the highest proportion of patients aged between 5–9 years (p=0.02). In only two of the patients with low velocities, both with previous CVA, was there no ultrasound signal from either side. 45 (32%) SCA patients had a second TCD after 2 years (two after 14 years). Of the 21 restudied who had high TAMM CBFv at baseline, 14 remained high and 2 became low. Of the 15 restudied who had low TAMM CBFv at baseline, 14 remained low and none became high. Patients with abnormal TCD had lower daytime SpO_2 oxygen saturation (p=0.01) and hematocrit (p=0.05). Abnormal TCD was also associated with lower haemoglobin level, red blood cell count and higher white cell count, but not significantly. Neurological abnormalities included history of convulsions in 25 (18%) and history of CVA in 5 (4%). Of those with CVA, maximum TAMM CBFv on either side were 157, 156, 108, 0 and 0; the last patient subsequently died. Three patients who had convulsions in the interim attended for follow-up TCD; compared with those without seizures there was a trend for a greater increase in TAMM CBFv in these patients (p=0.06).

Conclusion: Compared with the developed world, in Africa a smaller proportion of patients with SCA have conditional or abnormal TCDs or CVA, although convulsions are common. The proportion of those with low velocities, perhaps due to ICA/MCA occlusion with moyamoya, may increase with time. Further population-based studies in a birth cohort will determine whether cerebrovascular disease is rare or lethal and, together with imaging and neuropsychology, will establish whether abnormal TCD predicts neurological events in Africa.

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