

## **ABSTRACT**

### **Peripheral Vascular Responses in Homozygous Sickle Cell (SS) Disease**

**Junette Suzanna Mohan**

Skin cooling is the most common precipitating factor of the painful crisis in Jamaica, yet the role of changes in peripheral blood flow in response to cooling as a contributing factor to the painful crisis has not been investigated. The aim of this study was to compare the peripheral vascular response to mild cooling in patients with homozygous sickle cell disease and subjects with a normal haemoglobin (AA) genotype, and to assess how the vascular behaviour in SS patients relates to their history of painful crisis.

The effect of mild, repeated cooling of the contralateral hand on forearm and cutaneous vascular resistance was assessed in 60 SS patients and 30 AA subjects. The response to a sudden, unexpected auditory stimulus, known to activate the muscle vasodilation of the alerting stage of the "defence" reaction was also determined to test the ability of the SS patients to show muscle vasodilation. Total forearm blood flow was measured by venous occlusion plethysmography. Mean arterial blood pressure and heart rate were monitored with a Finapres™ instrument. Cutaneous red cell flux at the forearm and hand were recorded by

laser doppler flowmetry. Forearm vascular resistance and forearm and hand cutaneous vascular resistances were calculated by the ratio of mean arterial pressure to forearm blood flow and forearm and hand cutaneous red cell flux, respectively.

In response to cooling, significantly more SS patients than AA subjects showed forearm vasoconstriction which was more persistent in the SS patients. The auditory stimulus evoked forearm vasodilation in equal proportions of SS patients and AA subjects suggesting that the ability to show muscle vasodilation was normal in the SS patients. SS patients who showed forearm vasoconstriction in response to cooling had a three-fold greater frequency of painful crisis than those who showed forearm vasodilation.

These observations showed that the peripheral vascular response to cooling in SS patients was different from that in AA subjects. The higher frequency of painful crisis in the patients who vasoconstricted in response to cooling was consistent with the theory that this vascular behaviour may be an important contributing factor to the mechanism by which cooling initiates the painful crisis.

**Keywords:** peripheral vascular responses; sickle cell disease; homozygous sickle cell disease; painful crisis.