

ABSTRACT

Assessment of Peripheral Blood Flow in Homozygous Sickle Cell (SS) Disease

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Several factors, in particular skin cooling, are associated with the onset of painful crisis in Jamaican patients with homozygous sickle cell (SS) disease.

Since the painful crisis results from avascular necrosis of bone marrow, skin cooling may initiate a reflex shunting of blood away from bone marrow. The purpose of this study was to evaluate peripheral blood flow in SS disease and its response to physiological stimuli to elucidate the mechanisms that may initiate the painful crisis.

Forearm blood flow (\dot{Q}_F) was measured in 20 (9M,11F) controls with normal (AA) haemoglobin and in 23 (13M,10F) patients with homozygous sickle cell (SS) disease in the steady state. Measurements of \dot{Q}_F

were made using venous occlusion air plethysmography under standardised conditions, at rest, post exercise and during exposure of the contralateral forearm to cold (15°C) and heat (40°C).

Resting \dot{Q}_F in SS patients was significantly higher than in controls ($p < 0.001$). \dot{Q}_F was increased by cold and decreased by heat in controls and SS patients. Isotonic exercise increased \dot{Q}_F in the same arm. The magnitude of these changes were greater in SS subjects for all stimuli.

Mean arterial pressure measured in a subgroup of controls and SS patients changed significantly in the same direction as \dot{Q}_F but the magnitude of change was small.

Cardiac output, measured in a subgroup of SS patients, did not correlate with resting \dot{Q}_F but was inversely related to total haemoglobin concentration.

Thus, both indirect cooling and heating evoked responses which were contrary to previous findings in Caucasians. This in itself requires further investigation. Moreover, further studies are also required to assess the accentuated responses of the forearm vasculature of SS patients in response to various physiological stimuli particularly in relation to the role of indirect cooling in precipitating the painful crisis.