# ABSTRACT

The study was undertaken to determine the relationship between glucose 6-phosphate dehydrogenase deficiency and intravascular haemolysis in patients treated for intravascular haemolysis at General Hospital Anuradhapura and resident in Anuradhapura district. The effects of glucose 6-phosphate dehydrogenase deficiency on the severity of intravascular haemolysis was assessed. In addition, the hypothesis glucose 6-phosphate dehydrogenase deficiency offers protection from malaria was tested. The usefulness of some of the tests used to identify glucose 6-phosphate dehydrogenase status was also assessed.

In phase one of the study data contained in bed head tickets of patients treated for intravascular haemolysis during two years were studied. The majority of patients treated for intravascular haemolysis were young, with 72.5% of patients being under 30 years of age. These results indicated that patients with glucose 6-phosphate dehydrogenase deficiency spent a significantly longer time in hospital due to intravascular haemolysis ($z = 2.28$, $p < 0.05$ ). The commonest cause which precipitated intravascular haemolysis in those with glucose 6-phosphate dehydrogenase deficiency was identified by patients and according to them by medical officers who attended on them as primaquine.

The severity of haemolysis was also assessed by the degree of anaemia and the need for blood transfusions. Many of these patients underwent severe haemolysis and required blood transfusions. The case fatality rate from

intravascular haemolysis for patients treated at General Hospital Anuradhapura during the two years was 1.9%.

The identification of glucose 6-phosphate dehydrogenase deficiency and the issue of and use of the diagnosis card are considered methods by which exposure to potentially haemolytic drugs could be minimised in these patients. However, it was evident that both of these methods were not being utilised adequately. The Methaemoglobin Reduction Test which is presently in use at General Hospital Anuradhapura for determining the glucose 6-phosphate dehydrogenase status, was not found to be satisfactory as it was found to have a sensitivity of 81.9% and a specificity of 88.9%. Its predictive value of a negative was only 34.8%.

Episodes of intravascular haemolysis had been previously reported in 33% of patients and a family history of same was present in 18% of patients. Knowledge of the condition was not satisfactory in many respects and an informed change of some practices, it is believed will be able to reduce the burden of the problem. Early screening for the defect with the Fluorescent Spot Test which had a sensitivity of 100% and specificity of 100% during non acute stage screening of subjects would be an advantage. However, this method may be used for delayed determination of glucose 6-phosphate dehydrogenase status approximately four months after an acute episode in the absence of a quantitative assay at General Hospital Anuradhapura. But the introduction of a method to perform quantitative assay of glucose 6-phosphate dehydrogenase during episodes of haemolysis at General Hospital Anuradhapura requires priority attention.

Screening for the enzyme deficiency in male siblings and fathers resident in Anuradhapura showed a deficiency of glucose 6-phosphate dehydrogenase among 52.4% siblings tested, and a 35.4% of deficiency among fathers tested. However, the coverage in these two groups were inadequate to draw conclusions.

The follow up of glucose 6-phosphate dehydrogenase deficient subjects and non-deficient controls indicated the incidence density of malaria to be 0.33 cases/100 person-months among subjects and 0.41 cases/100 person-months among controls. The relative risk (95% CI) for malaria was 0.8 (0.51 - 1.27). Glucose 6-phosphate dehydrogenase deficiency appears to have a protective effect on malaria, though not significant. The preventive fraction was 19.5%.