

ABSTRACT

This study of Acid-base balance in adults with Sickle Cell Anaemia was undertaken because of the reports of severe metabolic acidosis during "Painful Crises". Metabolic acidosis has been incriminated as the cause of these "Painful Crises" and it has been claimed that alkali treatment can prevent and even abort these painful episodes. It was therefore possible that a defect in urinary acidification could explain this tendency to develop acidosis with its serious consequences.

The results of Acid-base parameters during the steady state showed a mild respiratory alkalosis which is a non-specific finding in patients with severe anaemia.

The response to oral NH_4Cl loading revealed a slight but significant defect in urinary acidification (minimum pH in SCA 5.38 against 4.83 for controls).

Titrateable Acid excretion was reduced but the urinary NH_4^+ though also reduced was normal when related to the urine pH. The glomerular filtration rate was normal. These findings are compatible with the syndrome of Incomplete Renal Tubular Acidosis.

The administration of oral neutral phosphate resulted in a marked increase in titrateable acid excretion but the defect in urinary acidification persisted. A maximal acidifying stimulus (Na_2SO_4 infusion) produced intense urinary acidification in both normal controls (minimum pH 4.55) and patients with SCA (minimum pH 4.59). Since the sulfate infusion is a known test of distal tubular acidification, a gradient type defect (Distal RTA) was ruled out. The threshold for bicarbonate excretion was reduced in 3 of 6 patients and it was therefore suggested that these patients have a form of Proximal Renal Tubular Acidosis due to defective bicarbonate reabsorption.

There was no evidence of metabolic acidosis during "Painful Crises". This would support our belief that Alkalis are of little use in the treatment or prevention of "Painful Crises", at least in our population.