

ABSTRACT

Whole body protein turnover and basal metabolic rate were measured in 6 patients with homozygous sickle cell disease, HbSS, and 6 normal controls.

A technique using oral prime/intermittent infusion of ^{15}N glycine as tracer, and enrichment in urinary urea and ammonia was used to measure protein turnover. The subjects received a priming dose containing $90\ \mu\text{g}\ ^{15}\text{Nkg}^{-1}$ followed, six hours later, by a continuous infusion at the rate of $5\ \mu\text{g}\ ^{15}\text{Nkg}^{-1}\ \text{hr}^{-1}$. Feeds were given intermittently along with the tracer. Plateau enrichments were mostly achieved by 9-12 hours in both ammonia and urea.

Protein turnover in the sickle cell patients was 66-81 percent more than in the normals and represented an increase in synthesis and breakdown. A theoretical increase in haemoglobin turnover accounted for 20-26 percent of the increase in turnover.

The average basal metabolic rate, as measured by oxygen consumption, was also significantly higher in sickle cell patients, despite subnormal nutritional status in 5 of the 6 patients. It was 7 percent ($P < .05$) more than in the normals and the difference was even greater when expressed in terms of body weight and muscle mass, being 22 percent, $P < 0.005$ and 20 percent ($P < .005$) respectively.

Protein turnover is an energy demanding process and with no other obvious cause, it seems that the process of protein turnover contributed to most of the increase in energy expenditure. With this assumption protein turnover accounted for 40-44 percent of the energy

expended in the sickle cell subjects compared to 27-32 percent in the normals. If the cost of protein turnover in normals is as much as 27-34 percent, this is further evidence that previous estimates based on the energy required for protein synthesis is significantly underestimated.

The significant increase in protein turnover and energy expenditure indicates that there is a higher requirement for energy in HbSS compared to normal that could be relevant to the generalised poor growth, high susceptibility to infection, poor wound healing and the general ill health observed in the patients.