

## ACUTE ILLNESS PROTOCOLS

### 3-HYDROXYMETHYLGLUTARYL-CoA (3-HMG CoA) LYASE DEFICIENCY

#### *INTRODUCTION*

3-HMG CoA Lyase deficiency is one of several defects in the degradation pathway of leucine (a major branched-chain amino acid). Most of the defects produce metabolic ketoacidosis but ketones are absent or low despite acidosis and hypoglycemia in 3-HMG CoA Lyase deficiency. Thus, this is a cause of **hypoketotic hypoglycemia**. The pathway is as follows: