3-Hydroxy-3-Methylglutaryl-CoA Synthase Deficiency

3-Hydroxy-3-Methylglutaryl-CoA Synthase (HMG-CoA Synthase) Deficiency is a rare metabolic disorder that is caused by a lack of the HMG-CoA Synthase enzyme which is needed to produce ketone bodies. Ketone bodies are chemicals made by the body when there is not enough insulin in the blood and it must break down fat instead of glucose for energy. They provide chemical energy to the brain and other organs during long periods without food (fasting).

**Synonyms**

Alternative names for this condition are:

- HMG-CoA Synthase Deficiency
- HMGCS Deficiency
- Mitochondrial HMG-CoA Lyase Deficiency

Further information about this condition is available from Climb.

**Disclaimer**

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