

# WikiLectures: Causes of pathological conformation of proteins

## 5-Aminolevulinic acid

It is formed by the condensation of glycine and succinyl-CoA via the intermediate  $\delta$ -aminolvalerate (2-amino-3-oxo-1,6-hexanedioic acid), which is subsequently decarboxylated (removes CO<sub>2</sub>) to form 5-aminolevulinic acid.

The reaction is catalyzed by aminolevulinate synthase (ALA-synthase, ALAS), located in the mitochondrial matrix. Pyridoxal-5-phosphate acts as a cofactor, forming a temporary Schiff base with glycine. Enzyme activity is sensitive to vitamin B<sub>6</sub> deficiency. The product of this reaction is the starting precursor of **heme**.

## Odkazy

### Související články

- Hemoglobin

### Použitá literatura

- *Incomplete citation of publication*. MATOUŠ, Bohuslav. *Základy lékařské chemie a biochemie*. Praha : Galen, 2010. ISBN 978-80-7262-702-8.