

Carnitine Shuttle

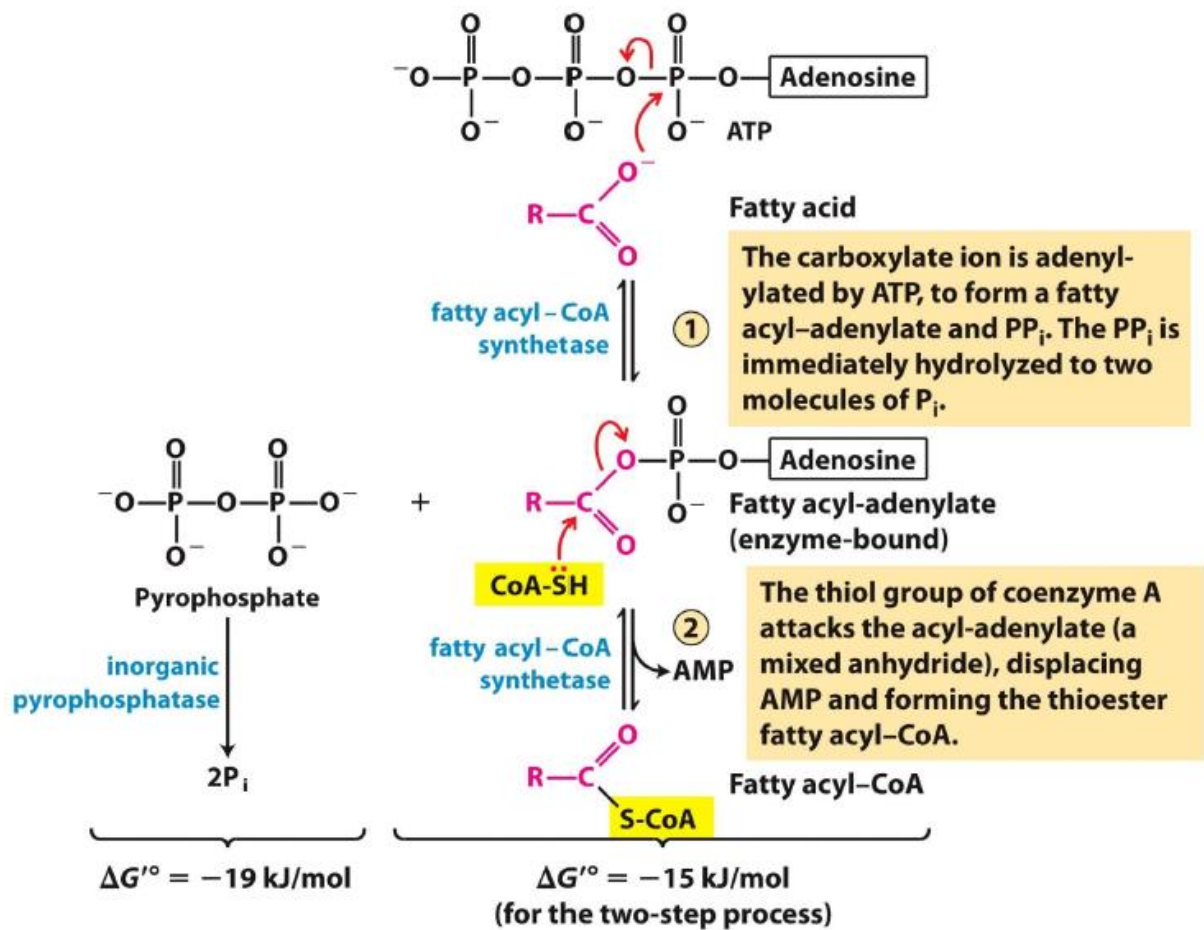


Figure 1: Conversion of a fatty acid to a fatty acyl-CoA. The conversion is catalyzed by fatty acyl-CoA synthetase and inorganic pyrophosphatase. Fatty acid activation by formation of the fatty acyl-CoA derivative occurs in two steps. The overall reaction is highly exergonic.

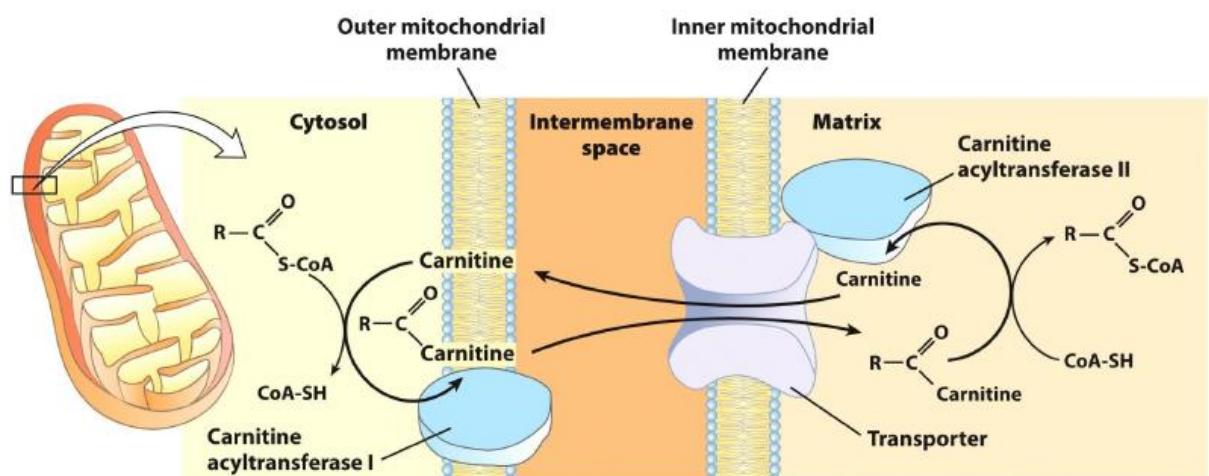


Figure 2: Fatty acid entry into mitochondria via the acyl-carnitine/carnitine transporter. In the matrix, the acyl group is transferred to mitochondrial CoA, freeing carnitine to return to the intermembrane space through the same transporter. Acyltransferase I is inhibited by malonyl-CoA, the first intermediate in fatty acid synthesis.

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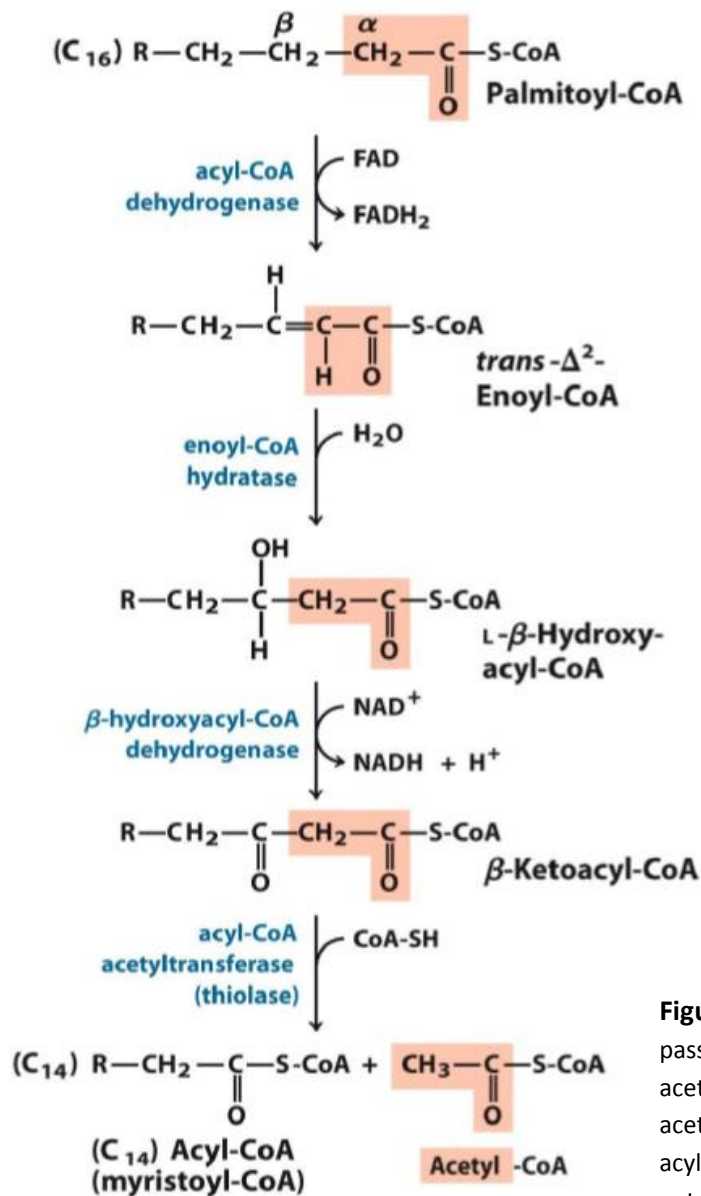


Figure 3: The β -oxidation pathway. In each pass through this four-step sequence, one acetyl residue is removed in the form of acetyl-CoA from the carboxyl end of the fatty acyl chain in this example palmitate, which enters as palmytoyl-CoA.

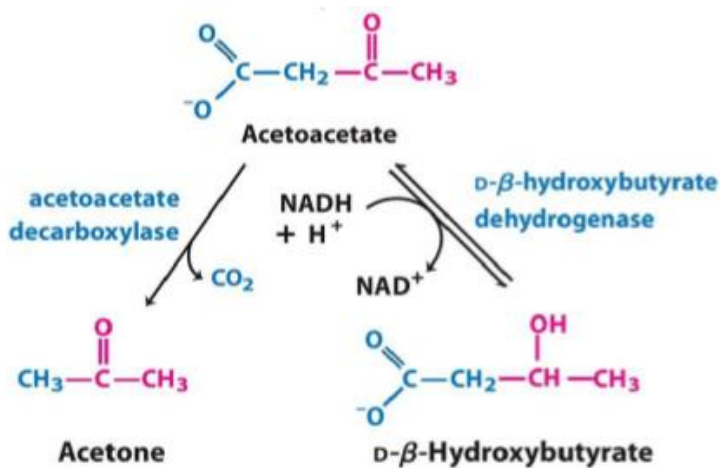


Figure 4: Formation of ketone bodies from acetyl-CoA. Healthy, well-nourished individuals produce ketone bodies at a relatively low rate.