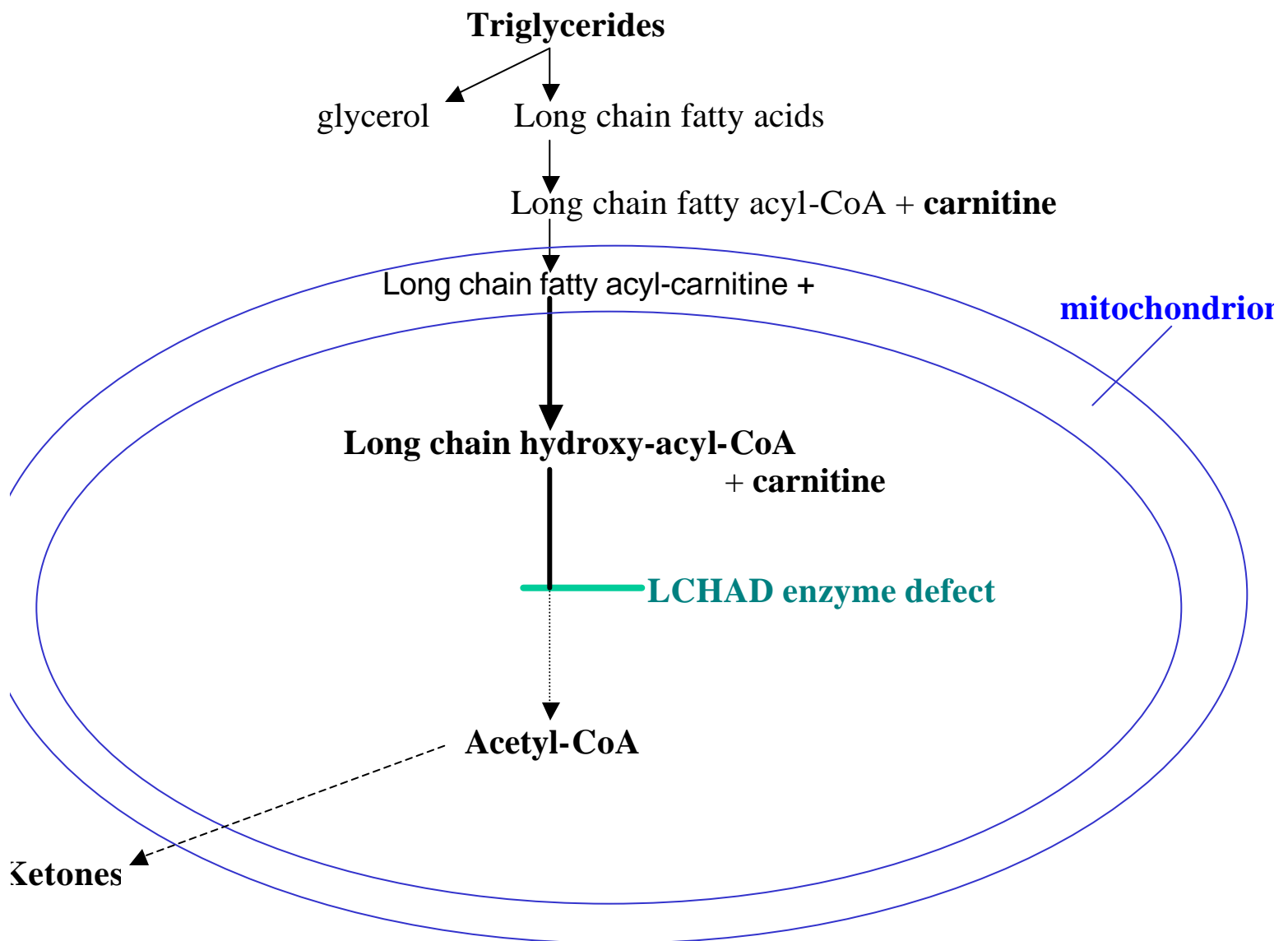


**ACUTE ILLNESS PROTOCOL  
FATTY ACID OXIDATION DISORDERS  
LONG CHAIN HYDROXY Acyl-CoA DEHYDROGENASE DEFICIENCY  
(LCHADD)**

**PATHOPHYSIOLOGY**

Below is the fatty acid  $\beta$ -oxidation metabolic pathway indicating the LCHADD block.

**Long chain hydroxy-acyl Co-A dehydrogenase deficiency (LCHADD)**



The pathophysiological process begins with reduced glucose intake as a result of, a fasting state or increased energy needs from a catabolic state (infection, stress, fever, etc...) not sufficiently satisfied by caloric intake. The resulting hypoglycemia leads to mobilization of free fatty acids (FFAs) which enter the mitochondria via the carnitine cycle. In the mitochondria, as shown in the diagram above, the fatty acids in the hydroxy form are normally oxidized to acetyl-CoA which is used to produce the ketones that can supply the energy needs to compensate for the lack of adequate glucose. A deficiency of LCHAD however, prevents this. The block at LCHAD also results in the accumulation of fatty acid intermediates that inhibit gluconeogenesis (thus preventing endogenous glucose production), have a toxic effect on the liver and produce metabolic (including lactic) acidosis. Muscle, particularly myocardium, requires a lot of energy and, therefore, becomes functionally impaired resulting in lethargy, hypotonia and cardiomyopathy.