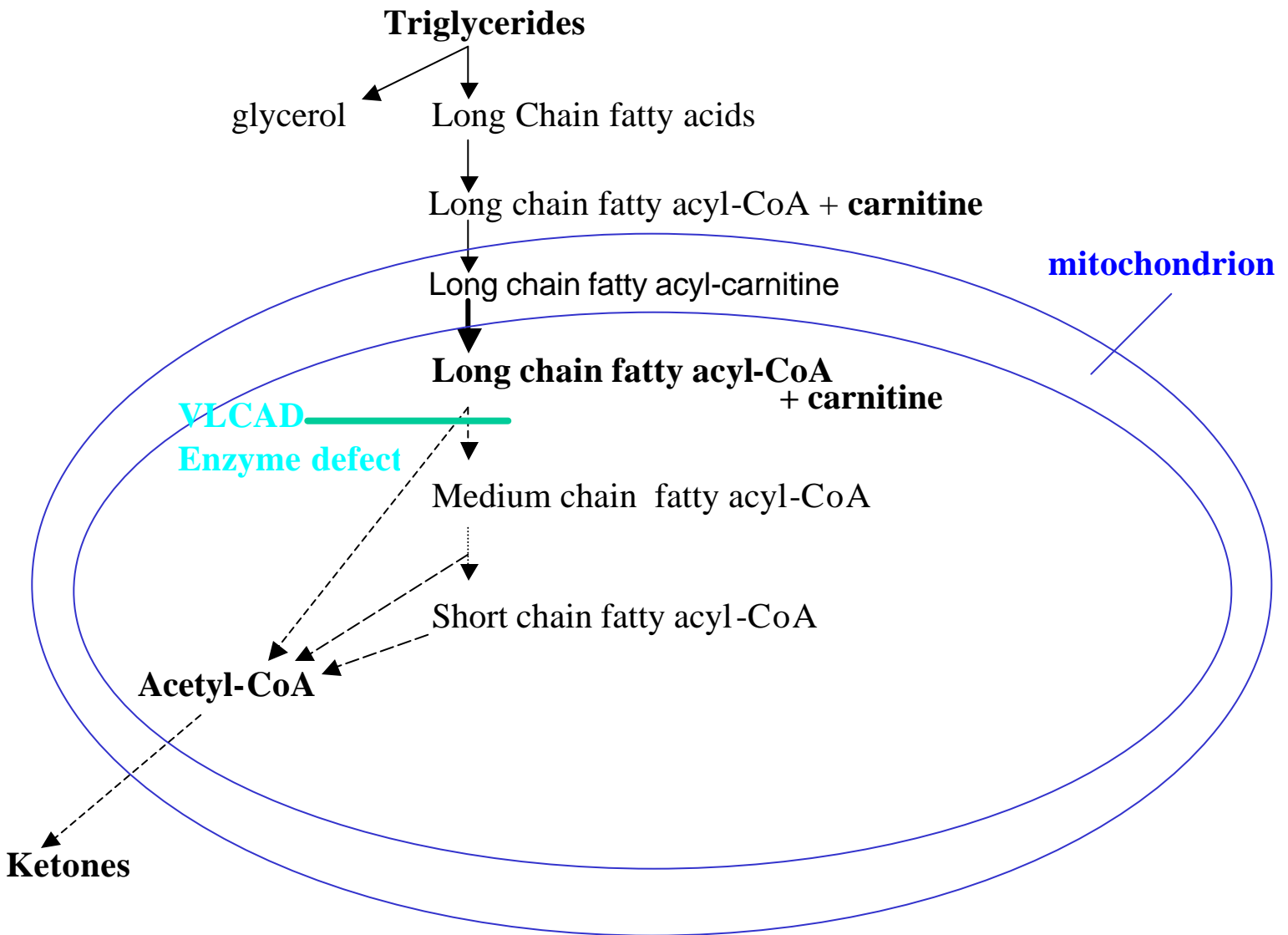


**ACUTE ILLNESS PROTOCOL
FATTY ACID OXIDATION DISORDERS
VERY LONG CHAIN ACYL CoA DEHYDROGENASE (VLCADD) DEFICIENCY**

PATHOPHYSIOLOGY

Below is the fatty acid β -oxidation pathway indicating the VLCADD block

Very Long chain acyl Co-A dehydrogenase deficiency (VLCADD)



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, etc...) not sufficiently provided for 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 acyl Co-A 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 VLCAD however prevents ketone formation. The block at VLCAD 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 acidosis. Muscle, particularly myocardium, requires a lot of energy and, therefore, becomes functionally impaired resulting in lethargy, hypotonia and hypertrophic cardiomyopathy.