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# Carnitine: A Dietary Compound and its Synthesis in Human Body

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## INTRODUCTION

Carnitine is a quaternary ammonium compound that plays a crucial role in the metabolism of fatty acids. It is a nutrient that is naturally synthesized in the human body, but it can also be obtained through dietary sources such as red meat, dairy products and some vegetables. The synthesis of carnitine occurs mainly in the liver and kidneys, but other tissues like the brain and muscles can also produce it.

The primary function of carnitine is to transport fatty acids from the cytoplasm to the mitochondria, where they undergo beta-oxidation to produce energy. The fatty acids cannot cross the inner mitochondrial membrane on their own because of their hydrophobic nature and that's where carnitine comes in to play. It binds to the fatty acid molecule, forming acylcarnitine, which can then be transported across the membrane *via* the Carnitine Palmitoyltransferase (CPT) system. Carnitine metabolism is a complex process that involves multiple enzymes and transporters. Let's take a closer look at each step.

## DESCRIPTION

### Synthesis of Carnitine

The biosynthesis of carnitine requires three amino acids: Lysine, methionine and glycine. These amino acids are converted into gamma butyrobetaine, which is then hydroxylated to form L-carnitine. The enzymes involved in this process include gamma butyrobetaine dioxygenase, 4-N-trimethylaminobutyraldehyde dehydrogenase and 3-hydroxy-N-trimethylaminobutyrate.

### Carnitine Transport

Carnitine transport occurs *via* the plasma membrane carnitine transporter Organic Cation Transporter Number 2 (OCTN2) in various tissues like the heart, skeletal muscles and kidneys. This transporter is an active transport system that moves carnitine from the extracellular fluid into the cells. Once inside the cells, the cytosolic enzyme Carnitine Acyltransferase (CAT) attaches the carnitine molecule to a fatty acid, forming acylcarnitine. The acylcarnitine molecule can now be transported across the inner mitochondrial membrane by the CPT system.

### Carnitine Palmitoyltransferase System

The CPT system is responsible for transporting the acylcarnitine molecule across the inner mitochondrial membrane. There are two types of CPT systems: CPT1 and CPT2. CPT1 is located on the outer mitochondrial membrane and catalyzes the transfer of the acyl group from acyl-CoA to carnitine, forming acylcarnitine. CPT2, on the other hand, is located on the inner mitochondrial membrane and catalyzes the transfer of the acyl group from acylcarnitine back to acyl-CoA.

### Beta-oxidation

Once inside the mitochondrial matrix, the acylcarnitine molecule undergoes beta-oxidation, a series of enzymatic reactions that break down the fatty acid molecule to produce energy. Beta-oxidation occurs in four steps: Oxidation, hydration, oxidation and thiolysis. The end products of beta-oxidation are acetyl-CoA, NADH and FADH2, which are used in the tricarboxylic acid cycle to produce ATP.

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### Carnitine Deficiency

Carnitine deficiency can result from a genetic defect in the biosynthesis of carnitine or a defect in the transport of carnitine into cells. The symptoms of carnitine deficiency include muscle weakness, fatigue, hypoglycemia and cardiomyopathy. The treatment of carnitine deficiency involves oral supplementation of L-carnitine, which can improve muscle strength.

## CONCLUSION

Carnitine metabolism is a vital process that plays a significant role in the oxidation of fatty acids to produce energy in various tissues of the human body. The biosynthesis of carnitine involves three amino acids and its transport occurs *via* the OCTN2 transporter. The CPT system is responsible for transporting acylcarnitine across the inner mitochondrial membrane and beta-oxidation occurs in the mitochondrial matrix. Carnitine deficiency can result in various symptoms and its treatment involves oral supplementation of L-carnitine. Understanding the complexities of carnitine metabolism is essential in the development of therapies for various metabolic disorders.