

Dr. Prompt replacement of dietary protein by an amino acid mixture that lacks leucine, isoleucine, and valine averts brain damage and early mortality. Amar Babikir Elhussein BSc, MSc & PhD Biosynthesis of Nonessential Amino acids and Fate of Amino Acid Carbon Skeleton Introduction: ?The major product of degradation of the carbon skeleton of the amino acids are pyruvate, intermediate of TCA cycle acetyl CoA and acetoacetate. Methionine: provide methyl group for synthesis of various compounds, its sulfur is incorporated into cysteine; and the remaining carbon forms Succinyl CoA. The amino acids that are synthesized from intermediate of glycolysis are degraded to pyruvate (Serine, Glycine, Cysteine and alanine). Four amino acids converted to Propionyl CoA, Which carboxylated in reaction require Biotin to form Methylmalonyl CoA, Which is rearranged to form Succinyl CoA in reaction require vitamin B12. Plasma and urinary levels of leucine, isoleucine, valine, ?-keto acids, and ?-hydroxy acids (reduced ?-keto acids) are elevated. The enzymes glutamate dehydrogenase, glutamine synthetase, and aminotransferases occupy central positions in amino acid biosynthesis. When the amino acid is degraded, the major products are Pyruvate, Intermediate of TCA cycle, acetyl CoA or Acetoacetate. Tyrosine: Obtained from the diet or by hydroxylation of phenylalanine, is converted to Homogentisate, whose aromatic ring is opened and cleaved, forming fumarate. 3) Glycine and cysteine: Can be derived from Serine Serine biosynthesis II. Amino acid derived from TCA cycle: 1. ?Arginine can be produced in urea cycle via Ornithine which is produced by transamination of Glutamate semialdehyde. As the name implies, the odor of urine in maple syrup urine disease (branched-chain ketonuria) suggests maple syrup or burnt sugar. Ten of the nonessential amino acids contain carbon skeleton that may be derived from glucose. Four amino acids (Lysine, Threonine, Isoleucine and tryptophan), Can form acetyl CoA ?Of the 20 amino acids commonly found in proteins, 11 are not essential in the adult diets because it can be synthesized in the body. Intermediate of glycolysis serve as precursor for Serine, glycine, cysteine and alanine. B. Proline and Arginine are derived from Glutamate semialdehyde which is formed by reduction of Glutamate. The eleven nonessential amino acid is synthesized by hydroxylation of the essential amino acid Phenyl alanine. Activity II catalyzes reduction of dihydrobiopterin by NADPH, and activity I the reduction of O₂ to H₂O and of phenylalanine to tyrosine. Glycine: in a reversal of the reaction utilized for its synthesis, react with Methylene tetrahydrofolate to form serine. ii. Amino acids that are converted to intermediate of TCA cycle: ?Carbons from 4 groups of amino acids from TCA cycle intermediates, ?Valine and Isoleucine: two of three branched chain amino acids, Form Succinyl CoA. i. Valine is eventually converted to Succinyl CoA via Propionyl CoA and Methylmalonyl CoA. ii. Isoleucine is converted to Succinyl CoA after two of its carbon are released as acetyl CoA. Tyrosine: ?- ??????. Ketoglutarate. ?????? 2. 3. 4. ??? 1. 2. 3. 4. 2. 4. 5. 6. 1. 2. ??? 3. 2. 3. 2