

METABOLISM OF AMINO ACIDS AND PROTEINS

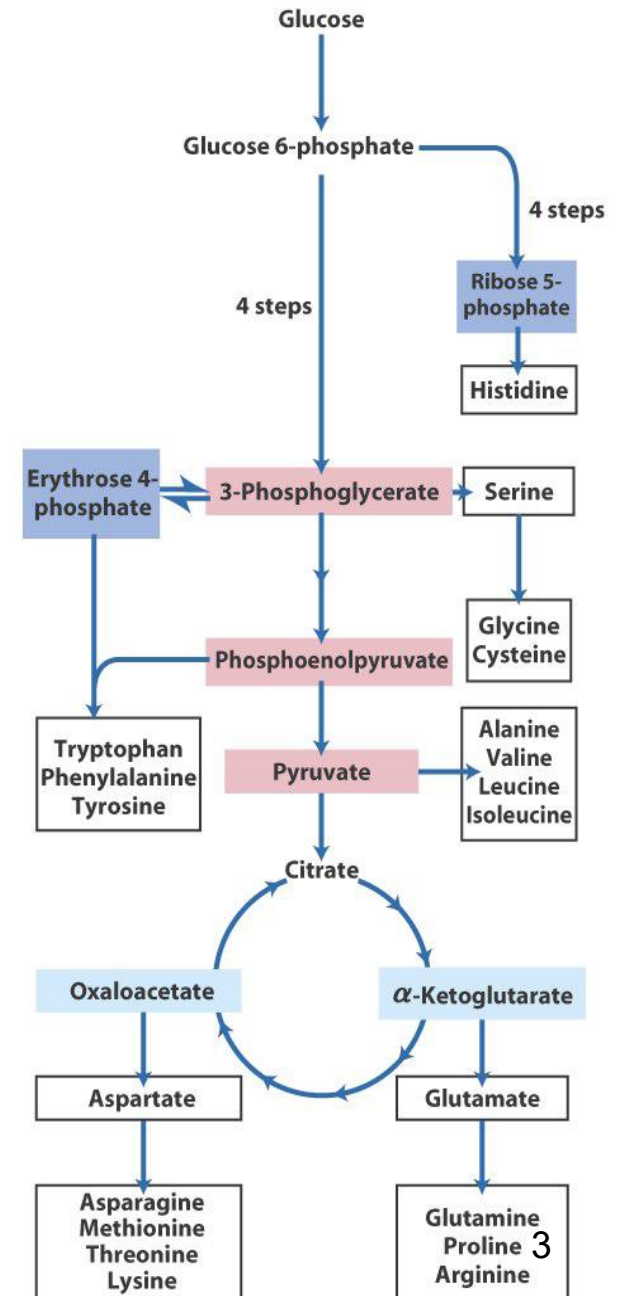
*prof.dr. zuhair maarouf hussein
biochemistry*

- Plants and bacteria synthesize all twenty amino acids, whereas, most other organisms obtain at least some of the required amino acids from their diet. Humans can only synthesize about half of the twenty amino acids.
- In general, the more complex amino acids are essential amino acids in humans as they require enzymes that have been lost from the human genome over evolutionary time.
- Concentration of total protein in serum ranges from (6 – 8) g / dl and about 0.3 g / dl higher for plasma because of the presence of fibrinogen .
- Amino acids classified as ketogenic and glucogenic according to their metabolic end product(ketogenic a.a. whose catabolism produce acetyl CoA or KBs while glucogenic a.a. whose catabolism produce pyruvate or intermediates of the CAC).

Amino Acids Biosynthesis

Twenty amino acids are used during synthesis of protein, which occurs in ribosomes. Ten of these can be synthesized in the body from glucose (NEAA). The other ten are required in the diet (EAA), cannot be synthesized in the body.

Essential Amino Acids	Nonessential Amino Acids
<i>Arginine*</i>	Alanine
<i>Histidine</i>	Asparagine
<i>Isoleucine</i>	Aspartate
<i>Leucine</i>	Cysteine
<i>Lysine</i>	Glutamate
<i>Methionine</i>	Glutamine
<i>Phenylalanine</i>	Glycine
<i>Threonine</i>	Proline
<i>Tryptophan</i>	Serine
<i>Valine</i>	Tyrosine*



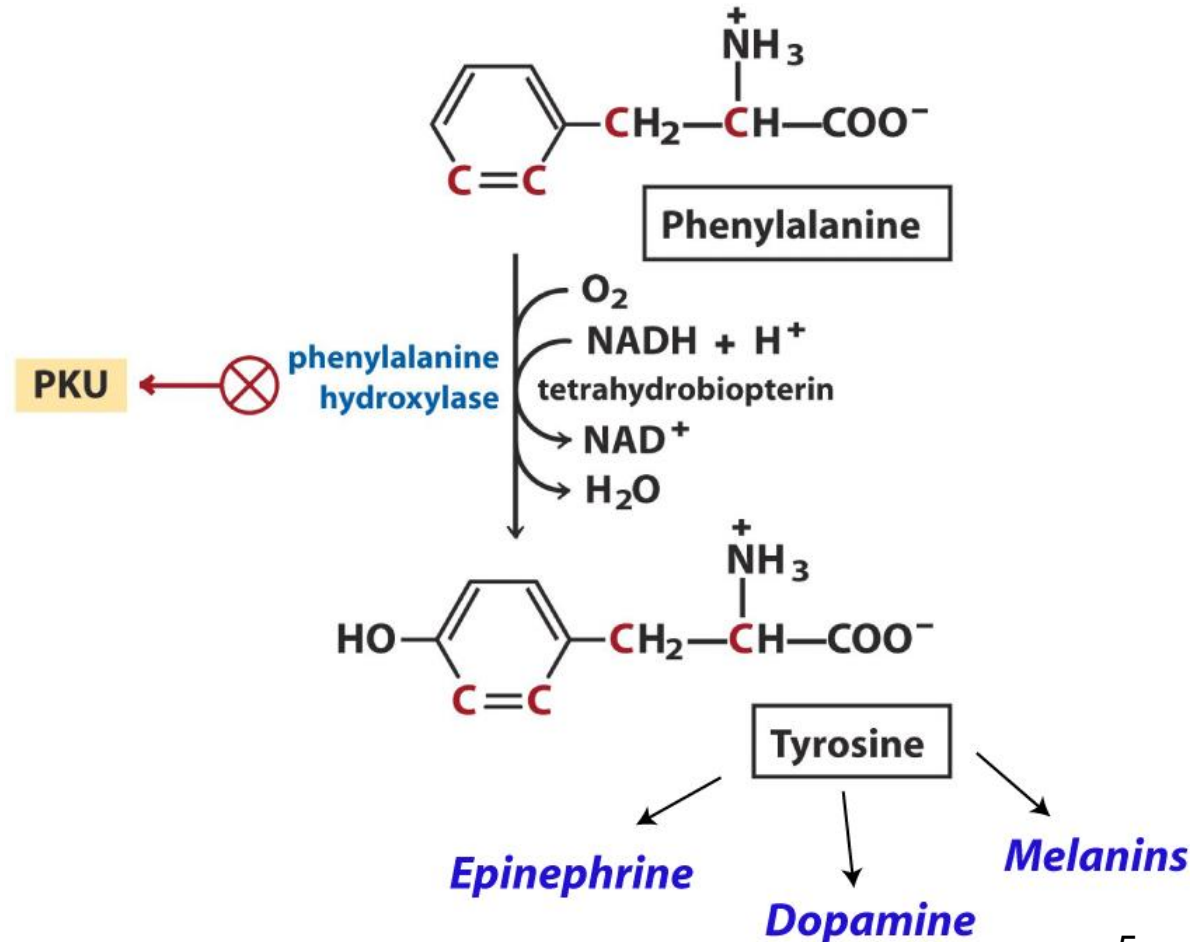
Notes :

1. **Arginine** is listed as an essential amino acid because humans require arginine in their diet to support rapid growth during childhood and pregnancy . However , arginine is actually generated from argininosuccinate in the *urea cycle* , which means that a small amount of this "essential" amino acid is made available for protein synthesis through this route .
2. The carbon skeletons of the 10 NEAA derived from glucose are produced from intermediates of the glycolysis and the CAC .
3. Three example of protein functions :
 - Catalysis (almost all chemical reactions in a living cells are catalyzed by protein enzymes) .
 - Transport (some proteins transports various substances , such as oxygen , ions , fatty acids ,.....) .
 - Information transfer for examples hormones .

Inborn Errors of Metabolism: Genetic Disease

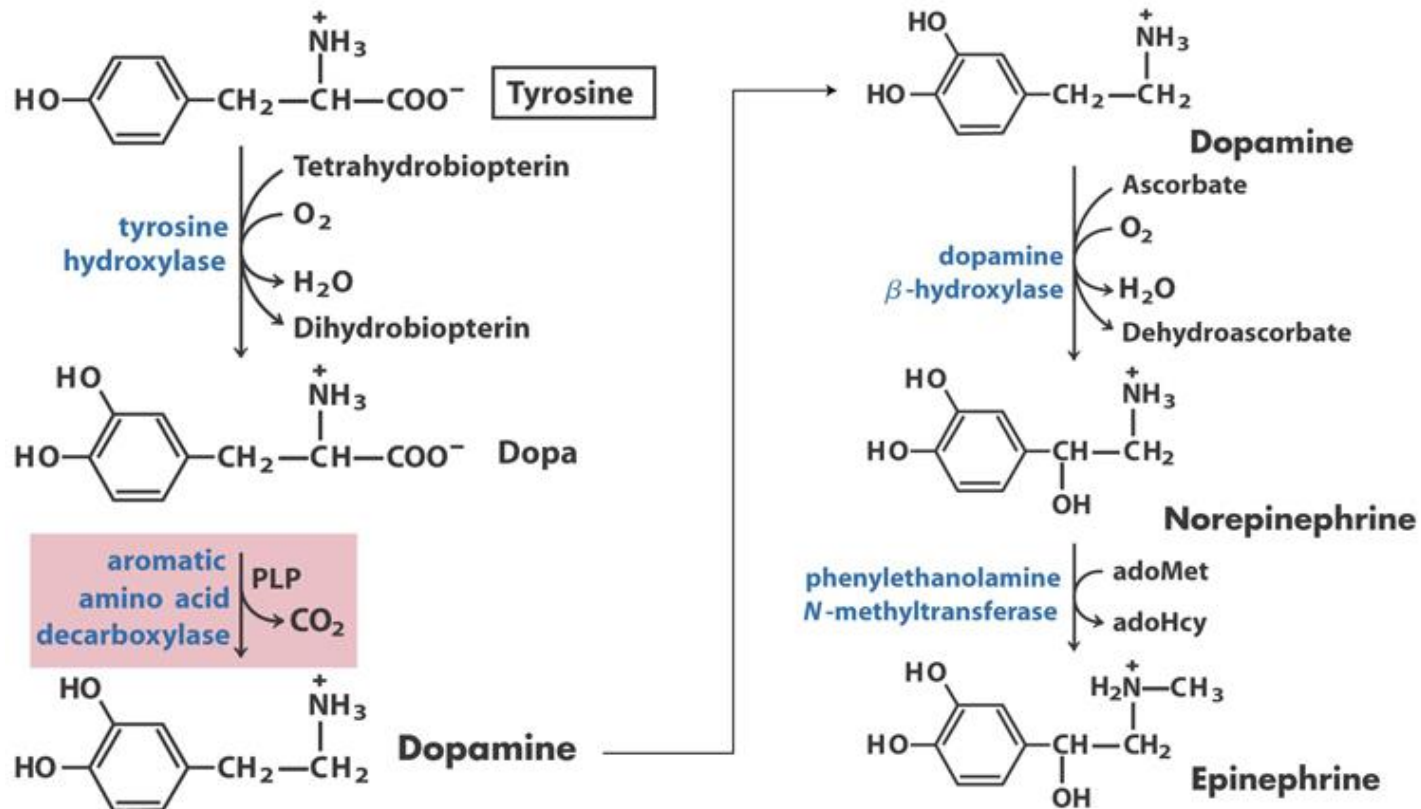
A genetic defect in the gene encoding phenylalanine hydroxylase is responsible for the metabolic disease **phenylketonuria (PKU)**.

Defect in phenyl alanine hydroxylase lead to PKU in which patients suffered from mental retardation, psychoses and eczema. In parkinson's disease, dopamine level in the CNS are decreased because deficiency of cells that produce dopamine or deficiency in conversion of dopa to dopamine.



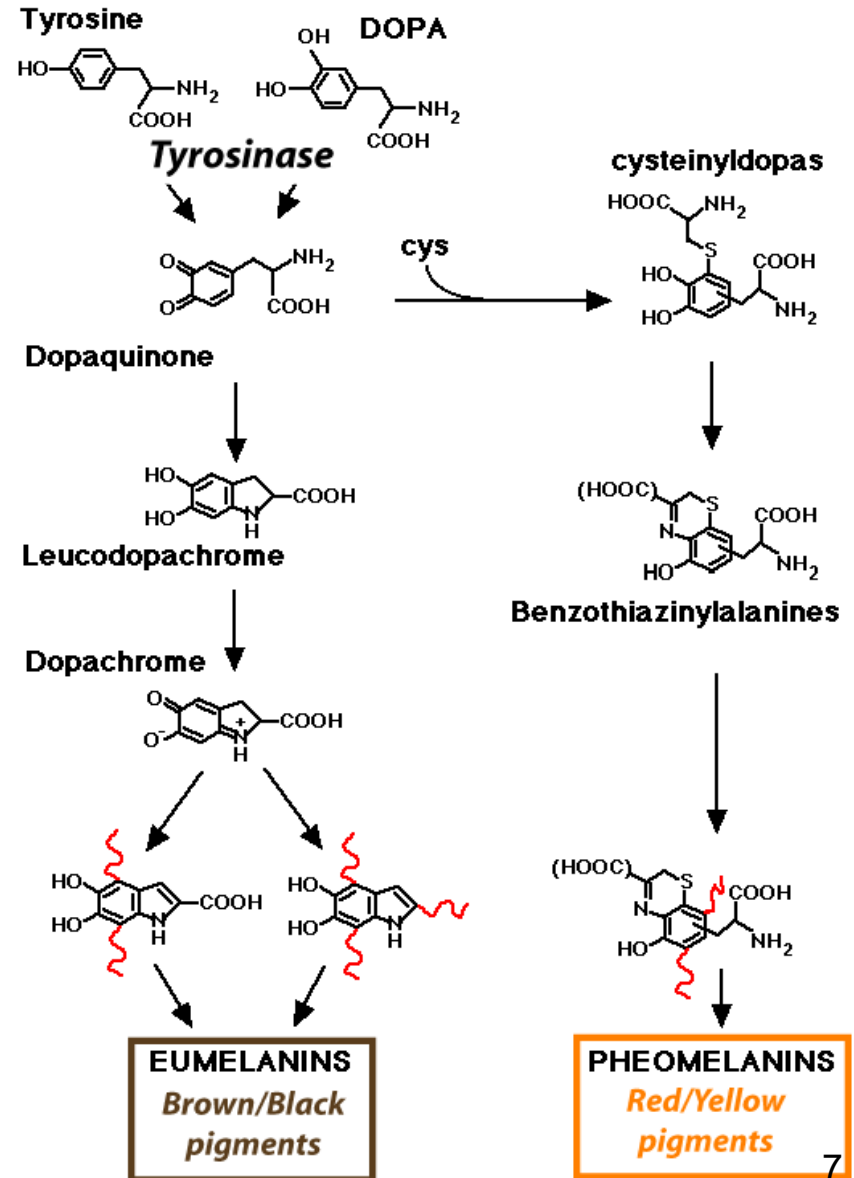
Conversion of tyrosine to norepinephrine and epinephrine in neural and adrenal cells

Many amino acids serve as neurotransmitters (e.g. glutamate, glycine) or are converted to other compounds that serve as neurotransmitters (e.g. GABA from glutamate and catecholamines from tyrosine).



Amino Acids as Metabolic Precursors

Tyrosine is also the precursor to pigment molecules called **melanin**. The two primary melanins are **eumelanin**, which are dark pigments having a brown or black color, and **pheomelanin** that have red or yellow color. The yellow color of pheomelanin pigments comes from the sulfur in cysteine that is combined with dopaquinone. A deficiency in tyrosinase will result in loss of skin and hair pigments (albinism).



Notes

1. In albinism, either the tyrosinase (Cu dependent) of melanocytes or other enzymes that convert tyrosine to melanin may be defective.
2. Parkinson's disease characterized by tremors, shuffling gait and masking face with a staring expression.
3. Decreased skin pigmentation is associated with an increased incidence of skin cancer.
4. All of the diseases related with amino acid cause mental retardation.
5. Tyrosine also precursors of thyroid hormones such as iodothyronine (T₃) and thyroxine (T₄).
6. In melanocytes of the skin, eye and hair, dopa is oxidized to quinones that polymerize forming melanin pigment.