## Metabolism of Individual Amino Acids:

### 1- Glutamate and Glutamine:

There are three enzymes that are essential for synthesis of nutritionally non-essential AA. These includes glutamate dehydrogenase, glutamatine synthetase, and aminotransferase.

Nutritionally non-essential Amino acids are synthesized from amphibolic intermediates of citric acid cycle.

α-ketoglutarate + NH4 + NADP $\xrightarrow{\text{Glutamate dehydrogenase}}$ Glutamate

**Glutamate + NH4** $\xrightarrow{\textbf{glutamine synthase}}$ **Glutamine**

**Glutamate + Pyruvate** $\xrightarrow{\textbf{ALanin aminotransferase}}$ Alanin + α-ketogl.

Glutamate + oxaloacetate $\xrightarrow{\text{Aspartate aminotransferase}}$ Aspartate + α-ketoglu

Glutamine $\xrightarrow{\text{Glutaminase}}$ Glutamic A + NH4

There is no inherited disorder in metabolism of these two amino acids.

2- Aspartate and Aspargine:

Aspartate is synthesized from oxaloacetate (intermediate of citric acid cycle) in presence of glutamate by transamination reaction(aspartate aminotransferase).

Aspartate + Glutamine $\xrightarrow{\text{Aspargine synthase}}$ Aspargine + glutamate

Aspargine $\xrightarrow{\text{Asparginase}}$ Aspartate + NH4

Also there are no metabolic disorders( inborn error of metabolism) in aspartate and aspargine.

## Functions of Glutamate and Aspartate

1- Most active amino acids

2- Both are glucogenic

3- n-acetyl glutamate and aspartate both participate in urea formation

4- Glutamic acid is a component of glutathione, which is very important for maintenance of sulfhydral containing enzymes in reduced form.

5- Aspartate is involved in the synthesis of purines and pyridamines.

6- Also a component of folic acid

7- Glutamate is very important in transamination, deamination, transamidation and interconversion of other amino acids.

By decarboxylation of glutamate neurotransmitter called GABA, which is a regulator of CNS in human being can be formed.

Glutamic Acid by $\xrightarrow{\text{decarboxylase}}$ GABA(gaba amino butyaric acid

## 3- Proline and Hydroxyproline:

Proline is the only amino acid that doesn't contain amino group.

It is synthesized in our body by the following chain reactions that need the presence oxygen, vitamin-C and iron($Fe^{+2}$):

Glutamic acid $\xrightarrow{\text{Glut. dehydrogenase and NADH}}$ L-G-γ semialdehyde → by dehydration→ ∆1- pyroline S- carboxylate → by dehydrogenase (DH) and in presence of NADH → Proline

There is no specific t-RNA for hydroxyproline and even when proline come in the diet as hydroxyproline, it is not incorporated into protein synthesis, only post transitionally.

## 4- Arginine and Ornithine:

Both of them are intermediates of urea cycle. Ornithine formed by hydrolysis of arginine by arginase.

Arginine $\xrightarrow{\text{Arginase}}$ Ornithine

Ornithine + α-ketoglut. $\xrightarrow{\text{ornithine }\Delta\text{-aminotransferase}}$ Glutamic acidγ-semialdehyde → Gl. A → α-ketoglutarate → CAC

Arginine is a semi-essential amino acid, it can be synthesized in small amounts in the body for short periods that are enough to maintain body health in adult but not adequate for growing children.

## 5- Glycine

This amino acid has no R-group, but instead glycine has two H atoms.

Glyoxylate + Glutamic acid $\xrightarrow{\text{Glycine aminotransferase}}$ Glycine + α-ketoglut.

Serine + Tetrahydrofolate $\xrightarrow{\text{methyl transferase}}$ Glycine + methylene H4F

Co2 + NH3 + B6 $\xrightarrow{\text{Glycine synthase- methylene H4 folate}}$ Glycine

**Catabolism of Glycine:**

Glycine $\xrightarrow{\text{Glycine Oxidase( oxidative deamination)}}$ Imino acid (-N=C)

Imino acid $\xrightarrow{\text{H2O}}$ NH4 + Glyoxalate

Glyoxalate $\xrightarrow{\text{Oxidation}}$ Oxalate (COOH-COOH)

Glyoxalate $\xrightarrow{\text{Decarboxylation}}$ Formate (HCHHO)

## Disorders of glycine metabolism:

1- Primary hyperoxaluria

Deviation of much amount of glyoxalate into oxalate rather than formate lead to form crystals in urine and result in nephrocalcinosis and early mortality from renal failure or hypertension.

2- Glycinuria: Excretion of large amount of glycine in urine due to inherited defect in the renal tubules reabsorption.

**Functions:**

1. glycine along with participate in heme synthesis
2. component of glutathione (glutamyl-cystinyl-glycine)
3. involved in purine synthesis
4. participate in detoxification reactions
5. component of creatine phosphate(energy storage of muscle only)

Creatine = lysine + glycine + methionine

**6- Serine**

It is a non-essential, glycogenic amino acid, and it is an important part of phospholipids and phospho-proteins.

## Functions:

- Serine can donate its methylene group and form glycine(-CH2).
- By serine dehydratase it is converted to Imino acid(-N=C)

Serine $\xrightarrow{\text{Serine dehydratase}}$ Imino acid $\xrightarrow{\text{H2O (hydrolysis)}}$ Pyruvic A + $NH4^+$

Pyruvate can enter the gluconeogenesis cycle to form glucose.