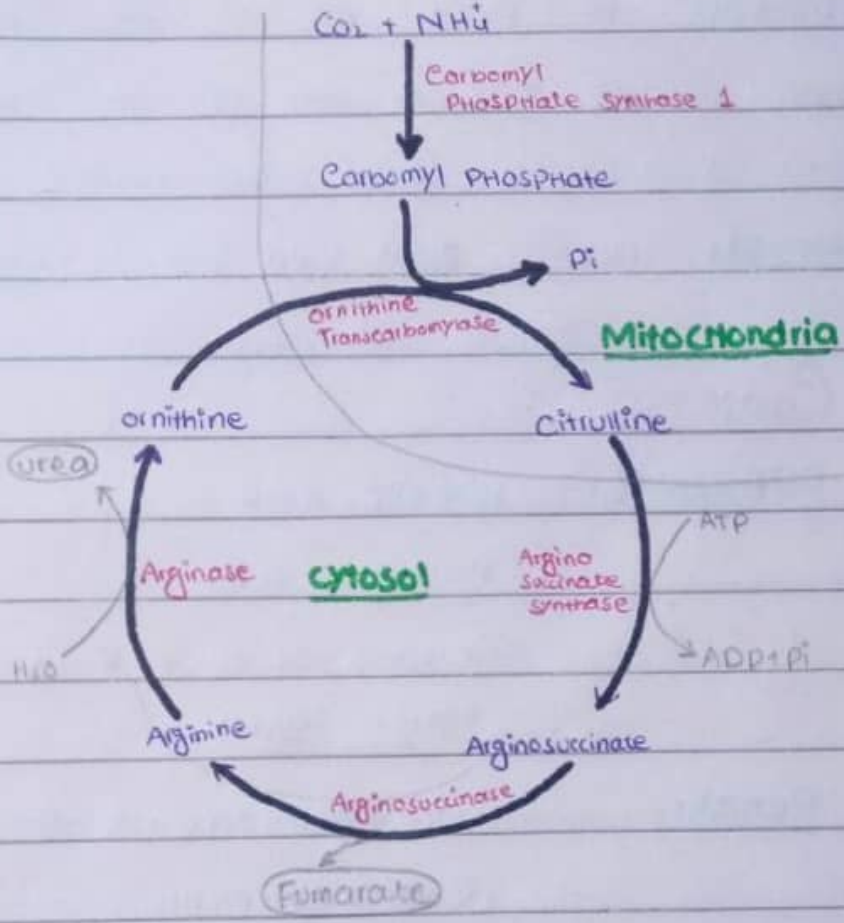
Protein

Urea Cycle and its Regulation: Urea is the end product of protein metabolism.

Urea synthesis is occur in Liver.



Regulation: The first step is catalyzed by the CPSI which is rate limiting step in urea synthesis.

- It is allosterically Activated by the N-Acetyl-Glutamate NAG is Synthesized from Acetyl-CoA & Glutamate by the Hydrolyze Enzyme.
- ↑ High Conc. of NAG ↑ NAG which ↑ urea synthesis
- Protein rich meal also ↑ NAG which enhance urea synthesis.

Uremia : Increase in Blood urea level due to renal Failure is caued uremia.

Azotemia :

Increase is Blood urea or other nitrogenous Compund which may or may not be associated with renal disease is caued Azotemia.

Normal : Normal Blood urea level 10-40 mg/dl

Causes :

PRERENAL : Diabetic Coma
Prolonged Fever
leukemia or other Blood disorder
Major Surgery

Renal : Acute glomerulonephritis
Chronic Nephritis
Polycystic Kidney

Post Renal : Obstruction in Urenary Tract.

Sources of Ammonia in our body :

1) Glutamic Acid
2) Glutamine
3) Purines & Pyramidine
4) Amino Sugars
5) Deamination

Creatinuria : ↑ output of Creatine in urine

Causes: 1) Muscular Atrophy
2) Starvation
3) DM
4) Hyperthyroidism

→ Removal of H_2O from creatine forms creatinine.

Cystinuria : ↑ Excretion of cystine in urine

Normal : 25-40 times

There is also Excretion of other amino acids
i.e Arginine, Lysine, ornithine

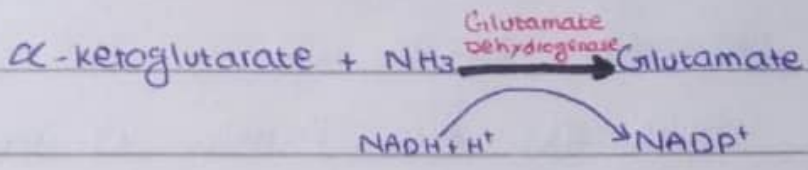
Homocysteineuria : ↑ urinary level of Homocystine and
↑ Plasma Homocysteine &
↑ Methionine & ↓ Plasma cystine.

Defective Enzyme is Cystathionine β synthase
Intellectual disabilities
Skeletal Abnormalities
Dislocation of Lense
Risk of Developing thrombi

It is a Autosomal recessive disease.

Adverse Effect of Hyperammonia :

1) Slurred speech
2) Blurred vision / Eyes
3) Hepatic Coma
4) Mental retardation
5) ATP Deplation

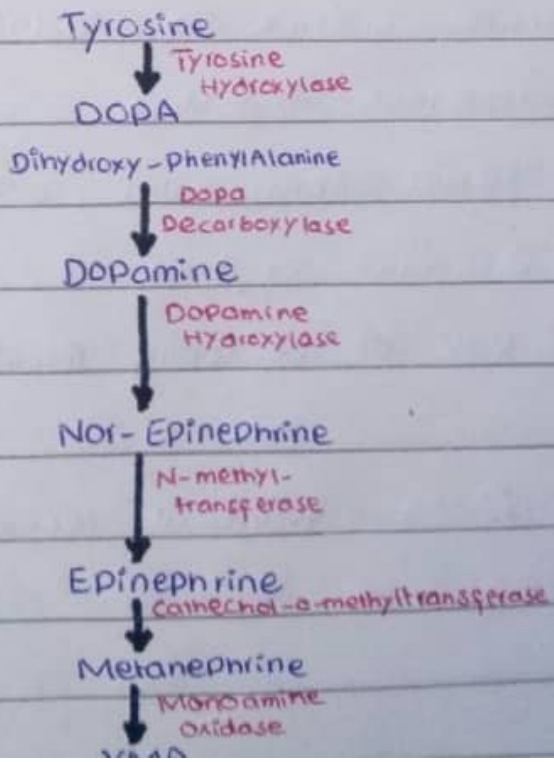


Accumulation of Ammonia shift toward right equilibriu

There is more utilization of α-ketoglutarate and it is very intermediate in TCA cycle.

It is ATP depletion impairement in ATP form acric

Synthesis of Catecholamines :



3) on standing Homogentisate oxidase get oxidize to corresponding quinone which polymerize and give Black color urine.

Diagnose:

Urine Black on standing

Benedict Test

Ferric chloride Test

Treatment:

NO specific Treatment But minimal Protein intake with phenylalanine Less than 500 mg/day.

Albinism: It is due to defect of Tyrosine metabolism. Defective Enzyme is Tyrosinase. "No melanine"

Result is fully or partly absence of Pigments from Hair & Skin

Photophobia, Nystagmus, ↓ visual acuity

Maple Syrup urine disease: It is also known as Branched chain ketonuria.

It is a metabolic disorder of Branched chain amino acid. The smell of affected person is Maple Syrup or Burn Sugar.

PHenyKetoNuria : It is due to deficiency of Enzyme Phenylalanine Hydroxylase an Autosomal recesive gen

→ The net outcome is that PhenylAlanine is not Converting into tyrosine.

→ Accumulation of phenylalanine in brain impairs Synthesis and metabolism of Aromatic amino acid.

→ There is also defect in myelin formation resulting in Hypopigmentation.

Features : 1) Failure of walk & Talk

2) Tremors & Convulsion

3) Mental retardation

Diagnose :

↑ Blood Phenylalanine

Ferric Test

Gauthers Test

Treatment :

low Phenylalanine in diet and ↑ Tyrosine.

Alkaptonuria :

The defective Enzyme is Homogentisate oxidase.

Homogentisate oxidase accomulaced into tissues, Blood then it is Excreted into Urine.

Features : 1) Joint Arthritis

2) Pigmentation of Sclera

Defective Enzyme is Branched Chain α- Ketoacid dehydrogenase

Symptoms: 1) Acidosis
2) Lethergy
3) Convulsion
4) Comma
5) Mental retardation

and Finaly death within 1 year after birth.

Metabolic Role Of Glycine:

1) It takes part in Synthesis of Heme, Purines and Creatinine
2) It Conjugated with benzoic acid in Liver to form Hippuric acid which is deroxyfying agent
3) Neurotransmitter of inhibitory Nature in CNS.
4) It is Conjugated with Colic acid & Chenodeoxycholic acid in Liver to form Glycocolic acid & Glycochenodeoxycholic acid.
5) It is Converted to other aminoacid Such as Serine which is Converted to Pyruvate.

How Nitric oxide is synthesized in body & Functi

Arginine give rise to NO:

Arginine $\xrightarrow{\text{NO synthase}}$ Citrulline + NO

Functions: 1) vasodilation

2) Platelet aggregation

3) Bacterial Action

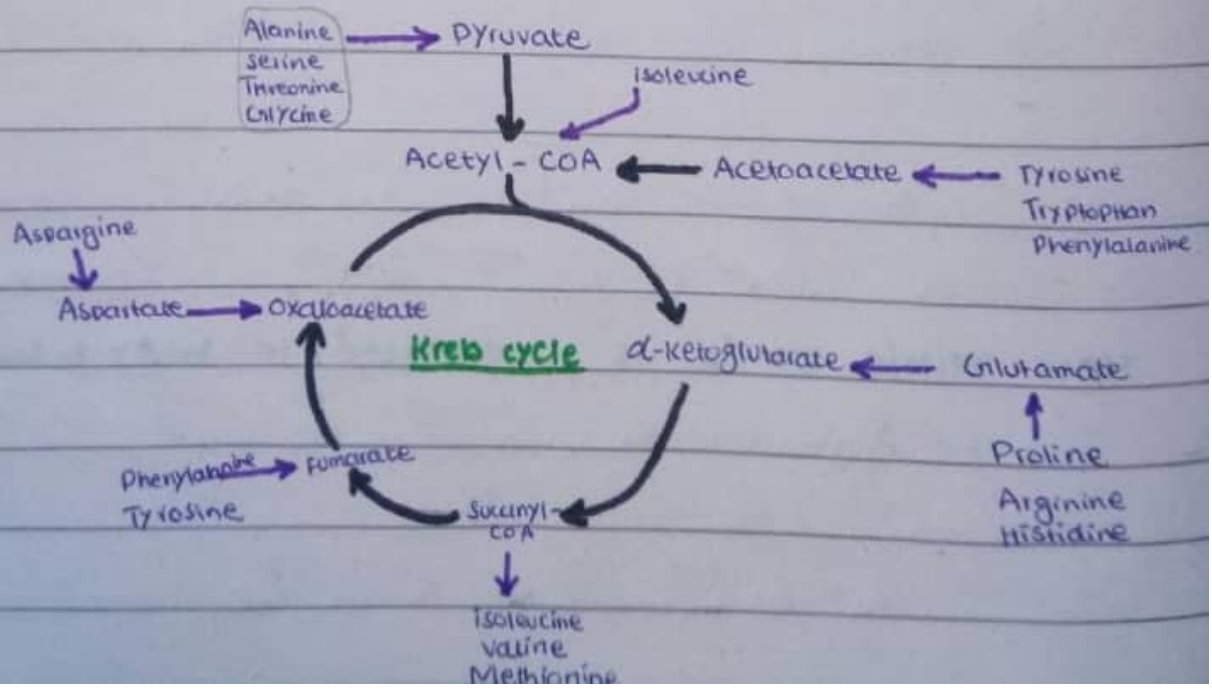
4) Neurotransmission function

5) used in Treatment of Chronic Obstructive Pulmonary Disease

Amino Acids :

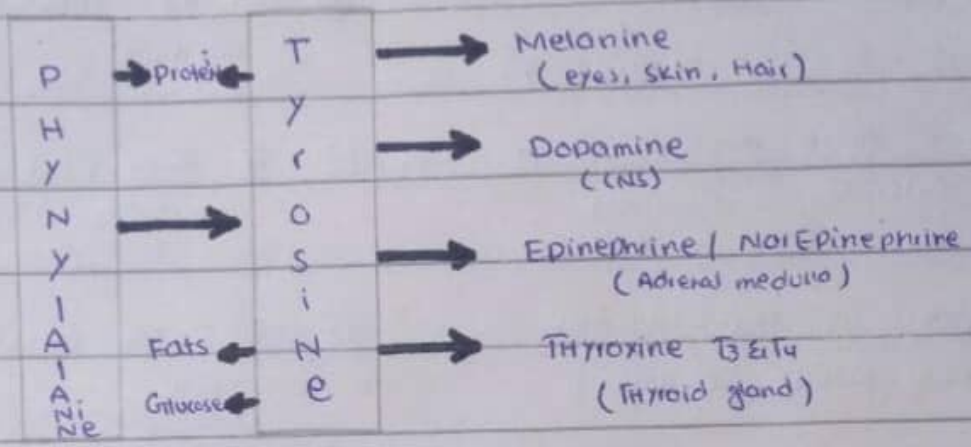
Glucogenic :	Ketogenic :	Both :
Serine	Leucine	Phenylalanine
Alanine	Lysine	Tyrosine
Threonine		Tryptophan
Hydroxyproline		Isoleucine
Glycine		
Cysteine		

Show entry point in citric acid cycle :

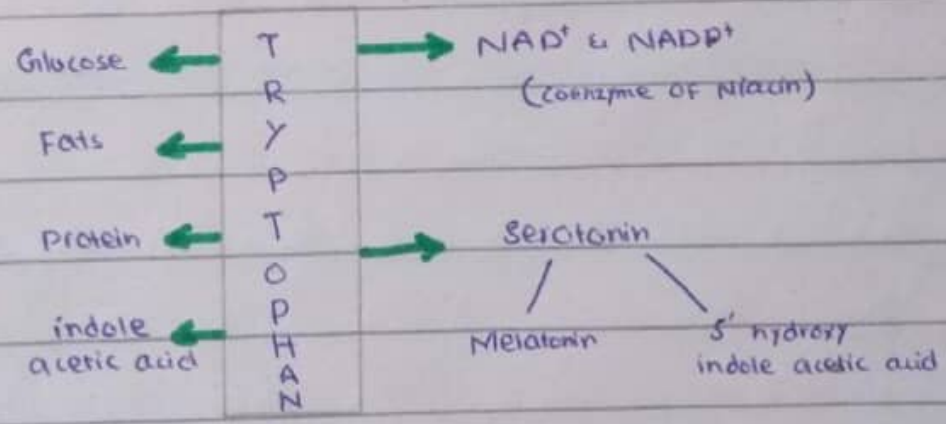


Product Derived From Followings:

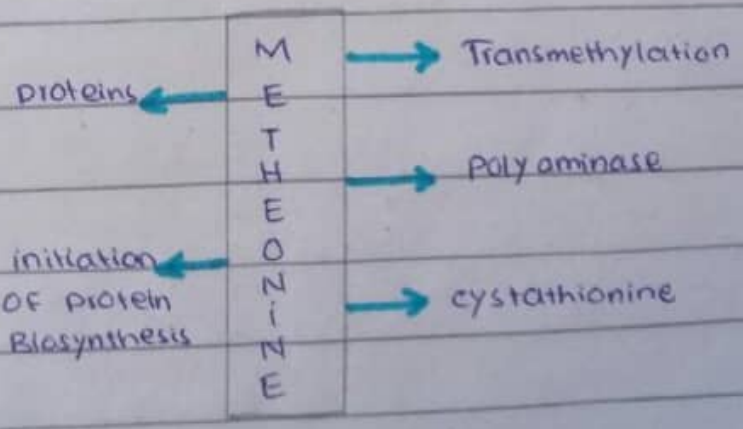
Tyrosine:



Tryptophan:



Methionine:



Function of Serotonine

) It is neurotransmitter

) It is Powerful vasoconstrictor & Cause Smooth muscle contraction in arterioles & Branches

) Involve in regulation of Cereboral Activity (Excitation)

) Motility of GIT

5) It is Happiness Hormone

6) It regulate behaviour Pattern

Function of Melatonine

1) It is involve in circad rhythms

2) It also perform Neurotrans functions

3) Has -ve effect on Ovarian cycle

4) It initiate melanocyte stim Hormone and ADH