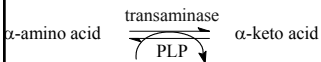
Protein Catabolism

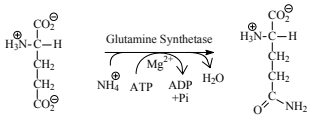
Two Amino Acids are Used to Transport Nitrogen



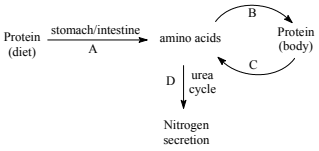
- Transaminase
- Nonspecific for one pair, specific for the other pair
  - Pyr-Ala
  - .αKG-Glu

are the two most common

- Gln synthetase particularly important to the brain



Overview of Protein Catabolism



Proteases are zymogens

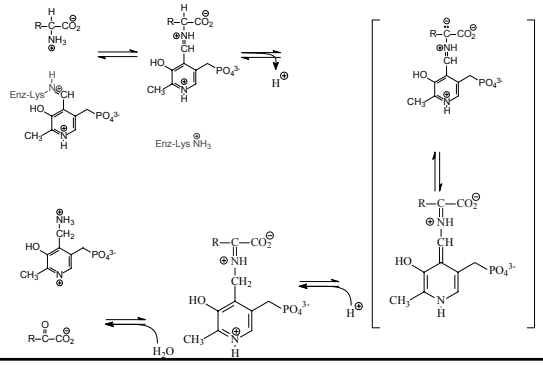
Excess amino acids are not stored

Normal protein turnover 1-2% of body protein per day

~75-80% amino acids reused; lose 30-40 g/day

Not excreted as amino acids or ammonia

Transamination Mechanism



Methods of Nitrogen Excretion

- Uricotelic (birds, reptiles)
- Ammonotelic (fish)
- Ureotelic (mammals)

Urea Cycle

Importance

means of excreting nitrogen in nontoxic form

Location

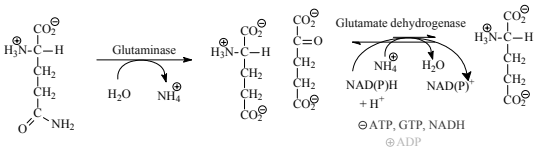
Mitochondrial matrix *and* cytoplasm of liver and kidney

Reactions

Energetics

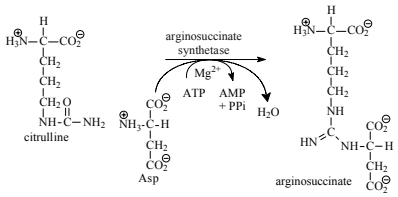
Regulation

Intracellular Ammonium Generation



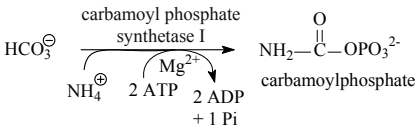
- Glutaminase mitochondria only
- Liver quantitatively more important
- Oxidative deamination of Glu Only
- Glu DH can use NAD or NADP

Arginosuccinate Synthetase



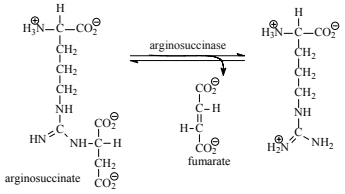
- Cytoplasmic enzyme

Carbamoyl Phosphate Synthetase I



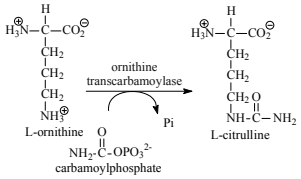
- Liver Mitochondrial enzyme
- RDS of urea cycle
- Activated by (requires) NAcGlu

Arginosuccinase



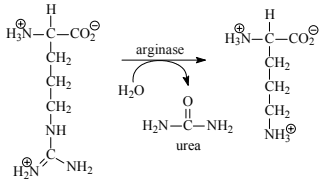
- Enzyme restricted to liver and kidney
- Fumarate converted back to asp

Ornithine Transcarbamoylase



- Mitochondrial enzyme
- Transported to Cp after synthesis

Arginase

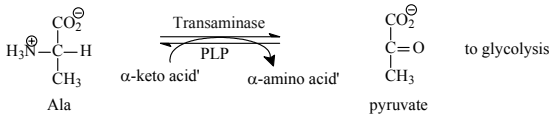


- Primarily liver enzyme
- Ornithine transported back into mitochondrion by ornithine-citrulline antiporter

Regulation

- GluNAc as described earlier occurs by changing ATP affinity
- 10-20–fold change in enzyme levels based on "nitrogen balance" i.e. how much protein is consumed relative to needs

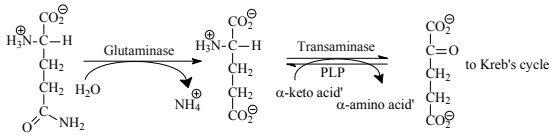
Ala



Energetics
False Claim #27 of Vegetarians

- "consuming protein uses energy"
- 4 ATP equivalents consumed in the urea cycle
- However, the carbon backbone is now available for energy use
- Carnivores obtain large amount of energy from amino acid carbon, herbivores ~10-15%

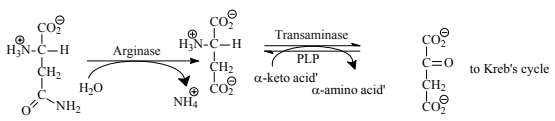
Gln, Glu



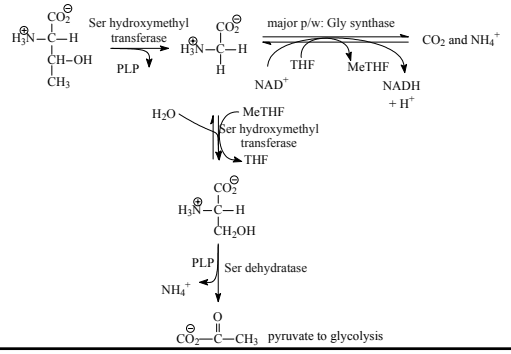
Carbon Backbone Catabolism

- Glucogenic amino acids *vs*. ketogenic amino acids
- Glucogenic are converted to metabolite of glycolysis (e.g., pyruvate) and can be converted into glucose
- Ketogenic form molecules such as acetoacetate, which can be converted to fat
- Most amino acids are glucogenic and ketogenic
- Leu is the sole amino acid which is ketogenic

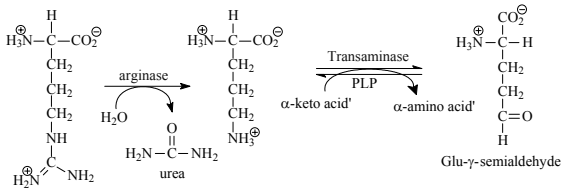
Asn, Asp



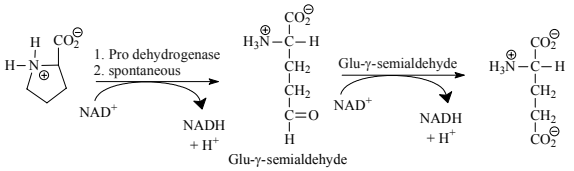
Thr, Ser, Gly



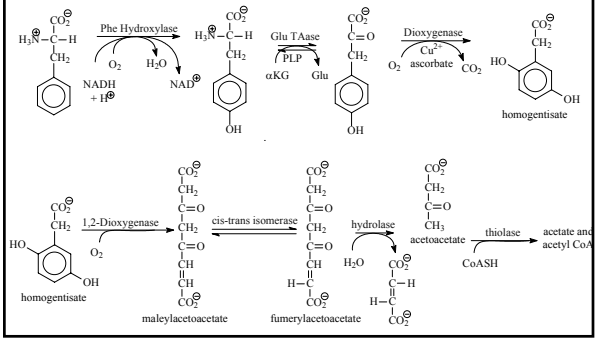
Arg



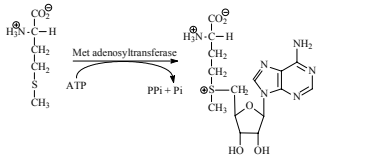
Pro

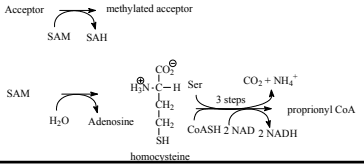


Phe

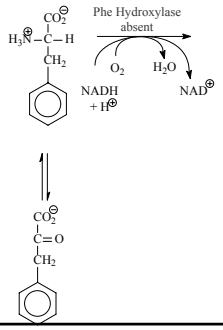


Met

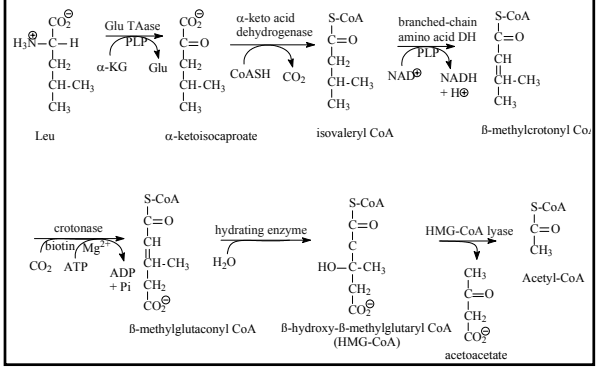




Phenylketonuria (PKU) is a Congenital Absence of Phe Hydroxylase



Leu



Maple Syrup Urine Disease

- The result of branched chain amino acid dehydrogenase
- Oxidation products of keto compounds accumulate