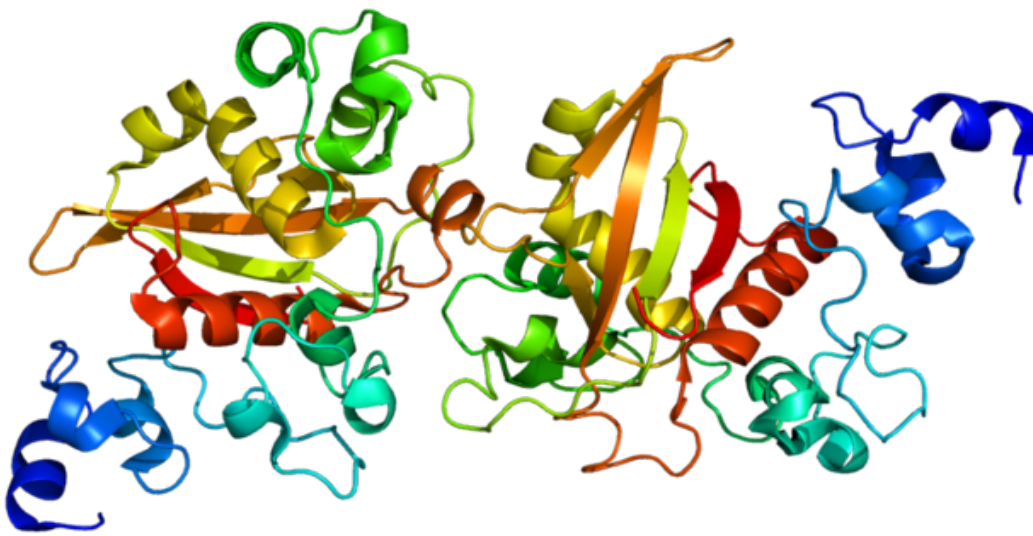


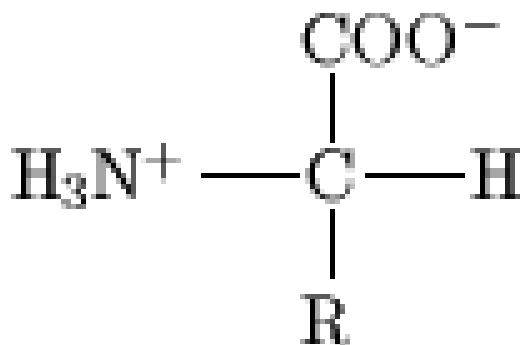
## Metabolism of Amino Acids

[See online here](#)

The biochemical processes that form part of the amino acid metabolism all support the synthesis and breakdown of amino acids. In the following article, the three most important reactions of the metabolism, i.e. transamination, deamination, and decarboxylation, are explained in a compact overview, providing you with the perfect preparation for your upcoming exams.



## Definition of Amino Acids



[Image:](#) General structure of amino acid. By Ppfk, License: [CC BY-SA 3.0](#)

Amino acids are a part of the building blocks that make up proteins. Amino acids form polymers with **peptide bonds**. These **polymers** are better known as proteins and represent, next to carbohydrates and fats, a vital nutrient and body component. Since amino acids, as suggested by their name, include a nitrogenous amino group in their base frame (NH<sub>2</sub>, NH, N), proteins are an important **nitrogen donor**. Nitrogen is

essential for many compounds and functions. This is why proteins, although they are an important food component, are only partially interesting for the energy supply (even though they still provide a significant part of the total energy amount). Rather, the organism needs them as an amino acid supplier. New **proteins** can be built with the amino acids supplied by proteins and those that are present in the organism.

## Proteins

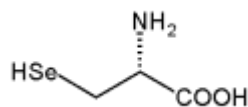
### Functions of proteins

Proteins serve as:

- **Building material:** e.g., keratin in the finger and toenails, tissue, muscles, bones, and skin hair
- **Transport vehicles:** e.g., albumin in the blood circulation, they give cells their shape
- **Signaling substances:** e.g., the G protein in signal transduction
- **Instruments:** e.g., enzymes for synthesis and breakdown of structures

Proteins are so elementary that their construction manual is codified in the deoxyribonucleic acid (DNA) of our genetic makeup. It is only with their help that an organism can be built in a functioning and operative manner because the necessary tools and builders are always proteins. They make up at least 20% of the human body. Amino acids are so important that they must not be squandered. Therefore, the constant breakdown accompanied by the rebuilding processes of the body requires an ingenious recycling system: the amino acid metabolism.

### Synthesis of proteinogenic amino acids



[Image:](#) Selenocysteine. By Teuteul, License: [public domain](#)

At least 1,000 different proteins exist, but for the human organism, mainly proteinogenic amino acids are of importance. **Proteinogenic amino acids**, comprising 20–21 amino acids, form the basic modules of proteins. Sometimes, the semi-essential amino acid **selenocysteine** is also counted as a proteinogenic amino acid which makes for the number 21. The following table highlights the 20 proteinogenic amino acids with a major characteristic included:

Protein	Characteristic
Lysine	Positively charged
Arginine	Positively charged
Aspartate	Negatively charged

Glutamate	Negatively charged
Serine	Polar
Threonine	Polar
Asparagine	Polar
Glutamine	Polar
Histidine	Polar
Tyrosine	Polar
Alanine	Hydrophobic
Valine	Hydrophobic
Leucine	Hydrophobic
Isoleucine	Hydrophobic
Proline	Hydrophobic
Phenylalanine	Hydrophobic
Tryptophan	Hydrophobic
Cysteine	Hydrophobic
Methionine	Hydrophobic
Glycine	Does not have a side chain

Similar to the alphabet, where different combinations of letters form new words, those 21 amino acids can form many different proteins, which perform many different tasks. Amino acids find their way into the organism packed as proteins in, e.g., soy, milk or meat.

## Transport proteins

Through the mechanisms of digestion, the amino acids are cleaved and enter the blood circulation on specific transporters. Next, they are channeled into body cells, again with the help of **transport proteins**. Specific transport proteins facilitate diffusion or actively transport specific substances in the body through membranes. Transport proteins can be a channel/pore that makes holes in membranes or carriers that open 1 side of the membrane. The metabolization of amino acids arriving postprandially occurs mainly in the liver and kidneys. Also, the regular breakdown of physiological structures (e.g., muscle protein) constantly supplies free amino acids so that a permanent pool of free

amino acids is always present. For an amino acid in the amino acid pool, there are 3 traveling possibilities:

1. Integration into structural and tissue proteins for the regeneration of tissue structures
2. Catabolism: deamination (removal of the amino group) and oxidation of the carbon frame releasing carbon dioxide (CO<sub>2</sub>) and adenosine triphosphate (ATP), or conversion into energy storages such as glycogen or lipids and urea synthesis from the amino group
3. Anabolism: synthesis of nitrogenous compounds such as purine bases, creatine, or adrenalin

The **3 most important reactions in the amino acid metabolism** are the **transamination**, **deamination**, and **decarboxylation**. Depending on the current state of metabolism, the existing amino acids are either regrouped or broken down entirely.

## Transamination of Amino Acids

One of the central reactions of the amino acid metabolism is transamination. As the name suggests, transamination refers to a **transfer of the amino group**. The process of transamination occurs through aminotransferase enzymes which can be specific for an amino acid or can cater to several amino acids that are similar in their chemical compositions. Amino acid that is currently not needed can be transformed into another amino acid that is currently needed. The reallocation of the amino group occurs via an alpha-keto acid, which has an analogous structure to **alpha-amino acids**. Alpha-keto acids only differ from alpha-amino acids in having a keto group, instead of an amino group.

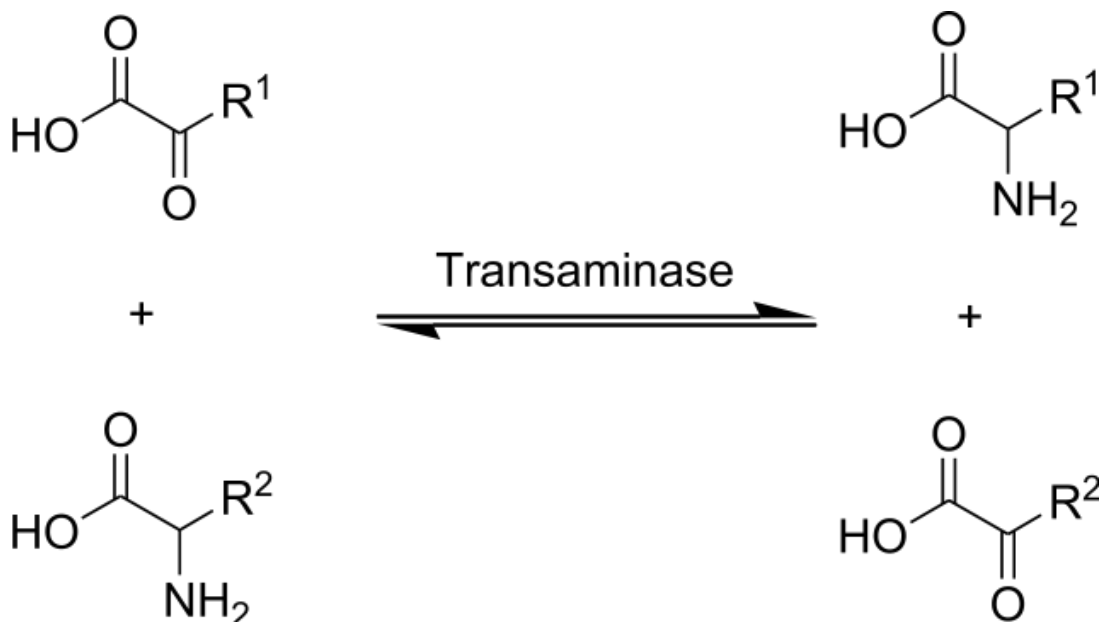


Image: Aminotransfer reaction between an amino acid and an alpha-keto acid. By Alcibiades, License: [public domain](#)

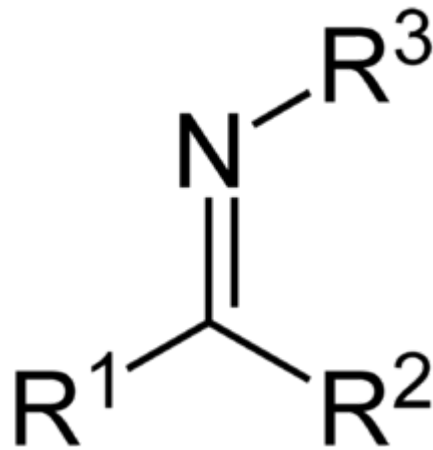


Image: Imine general structure. By Jü, License: [public domain](#)

Transamination turns the alpha-keto acid into a new amino acid available for metabolism. The responsible enzyme is called **aminotransferase (or transaminase)**. The aminotransferase, however, needs an assistant to do its work: the **pyridoxal phosphate (PLP)**. This is a coenzyme produced out of vitamin B6 (pyridoxine) by phosphorylation. PLP has an aldehyde group (H-C=O) that reacts in the transamination with the amino group of the amino acid (with the elimination of H<sub>2</sub>O). With this, a **Schiff base (R-NH<sub>2</sub>)** is formed. This reaction destabilizes the amino acid and the hydrogen atom starts to migrate which, in turn, leads to a shift of the double bond and ketamine (R-C=O) emerges from the former aldimine (H-C=O).

Then, water is added across this double bond which concludes the formation of the alpha-keto acid. PLP is reduced to PMP (pyridoxamine phosphate). The reverse reaction is also a common variation for generating another amino acid: pyridoxamine phosphate reacts with another alpha-keto acid and PLP is reconstituted.

The 2 most important transaminases are **alanine transaminase** (abbreviated as **ALT** or **ALAT**) and **aspartate transaminase** (abbreviated as **AST** or **ASAT**). ALAT catalyzes the transfer of an amino group from alanine to alpha-ketoglutarate, forming pyruvate and glutamate (which makes for its former name glutamate-pyruvate-transaminase, GPT). The ASAT transfers the amino group from aspartate to alpha-ketoglutarate, forming oxaloacetate and glutamate. Both transaminases are important diagnostic markers: increased values in a full blood count indicate a decay of cells in the liver (ALAT) and heart (ASAT and ALAT).

## Pyridoxal phosphate

The pyridoxal phosphate, or short PLP, is the most important coenzyme in the amino acid metabolism. PLP is the biologically active form of pyridoxal, the aldehyde form of vitamin B6. Vitamin B6 also appears as amine (pyridoxamine) and alcohol (pyridoxine). The derivatives of vitamin B6 can be converted into each other. They are ingested and can be found in animal-based food (pyridoxal and pyridoxine), as well as in vegetable-based food (pyridoxine).

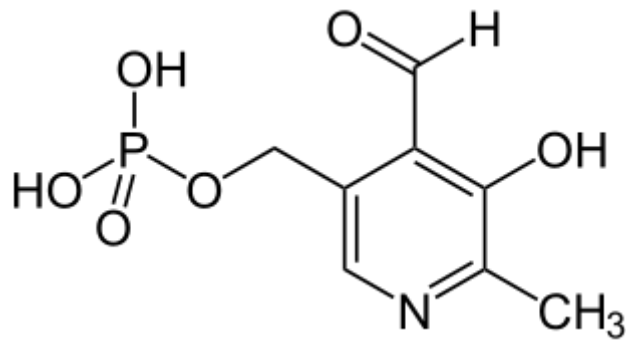


Image: Pyridoxalphosphat. By NEUROtiker, License: [public domain](#)

Wheat germs are relatively rich in vitamin B6. PLP as a coenzyme is involved in the amino acid metabolism in the reactions of transamination, decarboxylation (e.g., the formation of biogenic amines) and deamination. PLP links to a lysine residue based on the enzyme involved in the reaction, e.g., the alanine transaminase (ALT), and creates a Schiff base with the amino acid. The nitrogen of PLP contained in the pyrimidine ring has a strong electrophilic effect which leads to the shifting of the bonds. This reaction forms toxic ammonia; the concentration of the latter must be minimal at all times and it is not recommended to dispose of nitrogen. Thus the urea cycle occurs and leads to the deamination of amino acids.

## Deamination of Amino Acids

When an oversupply of nitrogen in the form of amino acids exists, it needs to be disposed of somewhere in the body. **Deamination** is the process that carries out this breakdown of amino acids. However, this process releases free cytotoxic ammonia which has to be quickly metabolized to urea. This urea synthesis, which requires a lot of energy, takes place in the liver. For this to happen, the excess nitrogen needs to be transported from the periphery to the liver. Three central amino acids are involved in this transport: alanine (synthesized from pyruvate), glutamine (synthesized from aspartate), and aspartate (synthesized from oxaloacetate).

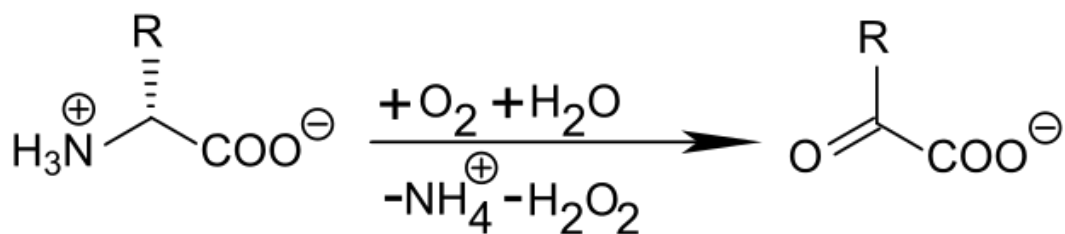


Image: Desamination of a D-amino acid with D-amino acid oxidase. By Kuebi, License: [public domain](#)

Deamination can be divided into 3 different reactions:

- Oxidative deamination
- Hydrolytic deamination
- Eliminative deamination

## Oxidative deamination

As in transamination, a Schiff base is formed with a dehydrogenase; more specifically, oxidation turns the amino group into an **imino group (C=N)**. Electron acceptors are the coenzymes  $\text{NAD}^+$  or  $\text{NADP}^+$  that are reduced in this process to  $\text{NADH/H}$  or  $\text{NADPH/H}$  respectively. By adding water, the imino group is converted into an alpha-keto group which releases ammoniac ( $\text{NH}_3$ ).

**Example:** Glutamate dehydrogenase reaction with glutamate dehydrogenase: glutamate - imino acid - alpha-ketoglutarate. In the liver, glutamate from the cytosol is taken into the mitochondrion, where oxidative deamination occurs, under the influence of the enzyme **L-glutamate dehydrogenase**, located in the mitochondrial matrix.

## Hydrolytic deamination

In hydrolytic deamination, water reacts with the amino group. Thereby, a **hydroxyl group (OH-group)** is irreversibly attached and the amino group is eliminated in the form of ammoniac. In the glutaminase reaction from glutamine to glutamate, the responsible enzyme is glutaminase. The same reaction occurs from asparagine amino acid to aspartate (the responsible enzyme is coincidentally called asparaginase).

## Eliminative Deamination

Small amino acids, such as serine or cysteine, can also be released through eliminative deamination of their nitrogen (in form of ammoniac) and by the elimination of water, or hydrogen sulfide for sulfurous amino acids. For this reaction, once again PLP is necessary as a partner. This **hydration** creates a double bond and, after the following **hydrolysis**, another alpha-keto acid.

## Decarboxylation of Amino Acids

When a carboxyl group is cleaved from an amino acid, 1 amine and  $\text{CO}_2$  are released as side products. The reaction is catalyzed by the enzyme **decarboxylase** using PLP as a partner. The resulting amines fulfill important functions in the body which is why they are called **biogenous amines**.

A well-known representative is **histamine** which is formed through decarboxylation from the basic amino acid histidine. The responsible enzyme is accordingly called histidine decarboxylase. Histamine is an important mediator and plays a vital role in, e.g., immediate hypersensitivity reactions. Further well-known biogenous amines relevant for metabolism are, e.g., **GABA** (gamma-aminobutyric acid from glutamine acid) and **dopamine** (from 3,4-dihydroxyphenylalanine).

## Central Amino Acids

There are 4 amino acids and their keto acids that can be described as the central hub of the amino acid metabolism because they are of special significance and the most important metabolic pathways take place via these amino acids:

- Glutamate and alpha-ketoglutarate
- Glutamine and alpha-ketoglutarate
- Alanine and pyruvate
- Aspartate and oxaloacetate

## Glutamine, glutamate, and alpha-ketoglutarate

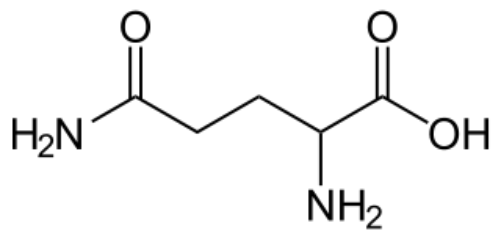


Image: Structure of glutamine. By NEUROtiker, License: [public domain](#)

Glutamine is the amino acid that is found most often in the blood plasma. Glutamine transports nitrogen to all the cells of the body that need it, e.g., synthesis of purine and pyrimidine in the nucleotide biosynthesis. Glutamine is also needed as an amino acid donor; it is deaminated in the kidneys to gain free ammoniac.

The ammoniac is secreted in the proximal tubule and neutralizes acids in the urine, thereby forming ammonium ions (NH<sub>4</sub><sup>+</sup>). In this process, alpha-ketoglutarate, which can be directly channeled into the citric acid cycle, is formed from glutamine via glutamate. This is killing 2 birds with 1 stone, as alpha-ketoglutarate refills the citric acid cycle when oxaloacetate has been pulled out of it for gluconeogenesis.

## Alanine and Pyruvate

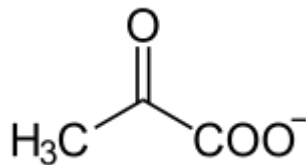


Image: Structure pyruvat. By NEUROtiker, License: [public domain](#)

The amino acid alanine is the counterpart to the alpha-keto acid pyruvate, i.e. to its salt (pyruvic acid, alpha-ketopropion acid). Amino acids can be channeled into the citric acid cycle and decomposed into CO<sub>2</sub> and energy via pyruvate and alanine. Another possibility is the synthesis of glucose from pyruvate in gluconeogenesis or metabolization of pyruvate to lactate under anaerobic conditions.

Alanine - Pyruvate

1) CO<sub>2</sub> + ATP

Or 2) Glucose

Or 3) Lactate

When alanine is broken down in the muscle, it 1st travels through the bloodstream to the liver. There, the synthesis of glucose in gluconeogenesis occurs, based on pyruvate obtained from transamination. The glucose travels through the bloodstream to the muscles and provides energy supply in the muscle cell through the breaking down of glucose in glycolysis.

This again yields pyruvate. Pyruvate receives the amino group of an amino acid from the



breakdown of muscle protein via the transamination reaction. The alanine arrives in the liver where it is transaminated to pyruvate again and is now available for glucose synthesis in gluconeogenesis. The enzyme that is responsible for the back and forth transfers of amino groups is called **alanine aminotransferase (ALT, ALAT)**.

## Aspartate and Oxaloacetate

Another important reactional pair is the set of 2 **amino acids, aspartate, and oxaloacetate (alpha-keto pyruvic acid)**. Their transformation is catalyzed by the enzyme aspartate aminotransferase (AST, ASAT) whose concentration can also be determined as a diagnostic marker in the blood count. The ASAT catalyzes the transfer of the amino group from aspartate to alpha-ketoglutarate, forming oxaloacetate and glutamate. This is why this enzyme was formerly known as glutamate oxaloacetate transaminase (GOT).

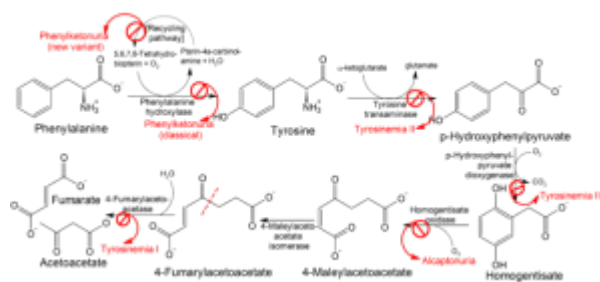


Image: Pathophysiology of metabolic disorders of phenylalanine and tyrosine. By LHcheM, License: [CC BY-SA 3.0](https://creativecommons.org/licenses/by-sa/3.0/)

Increase of ASAT in the blood can indicate diseases of the liver and heart because this enzyme is present in large numbers in hepatocytes and the heart muscle cells. So, when these cells are destroyed due to pathological changes, the ASAT presence in the blood increases. Oxaloacetate can participate in the citric acid cycle or gluconeogenesis. Glutamate can be further transformed into alpha-ketoglutarate or, for instance, be used in oxidative deamination for ammoniac synthesis in the kidneys.

## Serine Family

**Two main paths lead to serine:**

1. From 3-phosphoglycerate (connection to glycolysis)

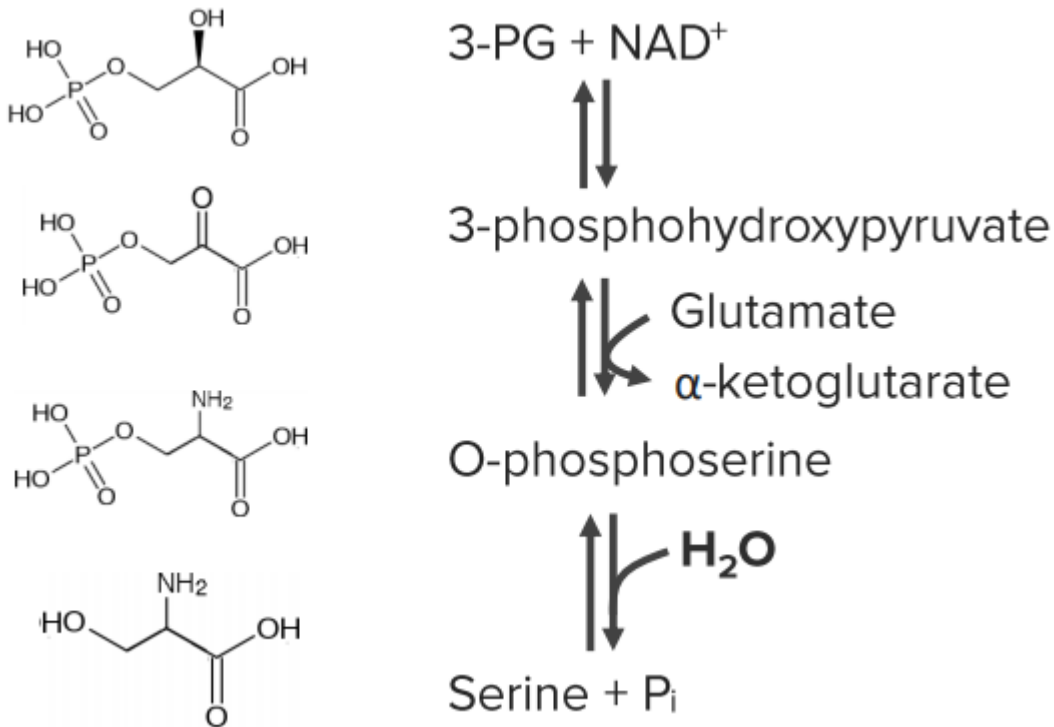


Image by Lecturio

2. Exchanging carbon with glycine and folates (important for folate recycling)

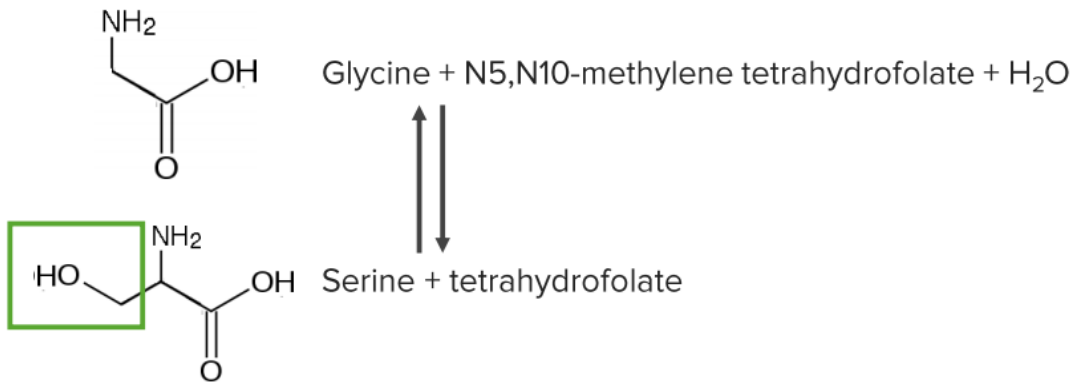


Image by Lecturio

Cysteine metabolism	Multiple ways of making cysteine	Primary means tied to methionine catabolism
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## Methionine catabolism

## Cysteine metabolism & health

Homocystinuria is a genetic disease. It most commonly involves the mutation of cystathionine β-synthase. Problems can include:

- Musculoskeletal anomalies

- Eye anomalies: cataracts, glaucoma, retinal detachment
- Intellectual disabilities
- Seizures
- Vascular disease
- Cystinuria — an unrelated genetic disease
- The inability of kidneys to reabsorb cysteine leads to high levels of cysteine, ornithine, and arginine in urine
- Frequent kidney stones also occur

## Other cysteine metabolisms

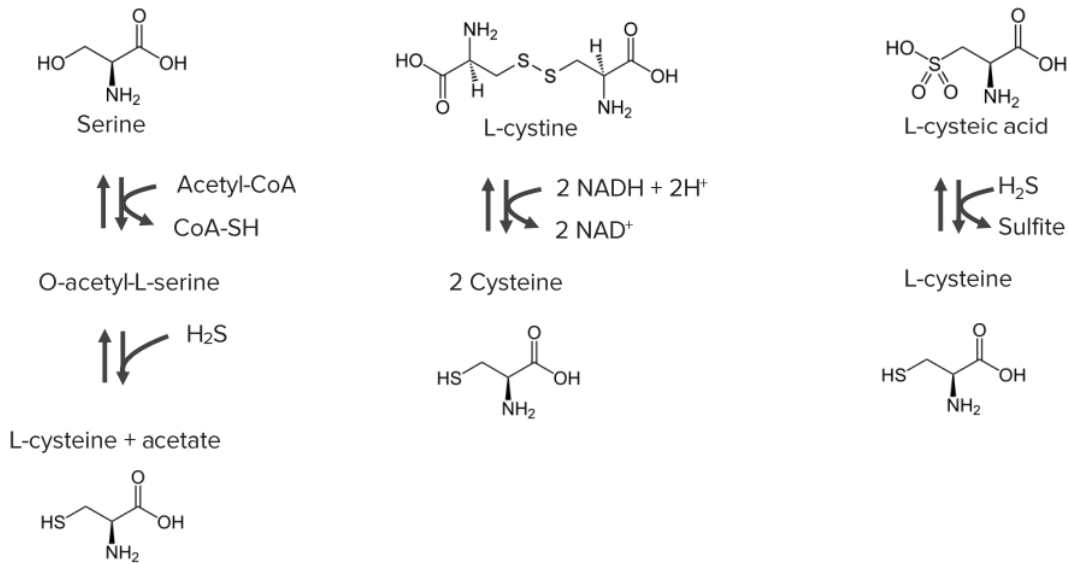


Image by Lecturio

## Selenocysteine metabolism

- Sometimes called 21st amino acid
- Not specified directly in genetic code
- Uses stop codon with an unusual structure
- Synthesized from serine on transfer ribonucleic acid (tRNA)



Image by Lecturio

# Aspartate Family

- All family members arise from aspartate
- Aspartate can be made from 1 of them — asparagine
- Numerous paths lead to aspartate

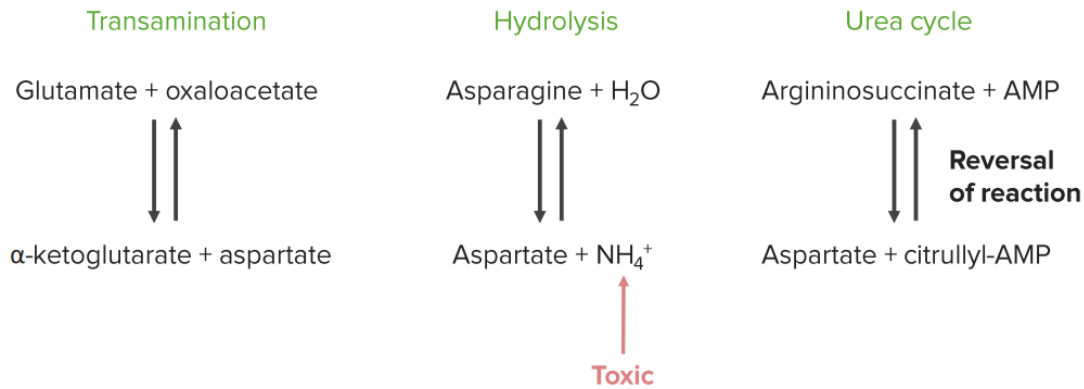


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## Asparagine metabolism

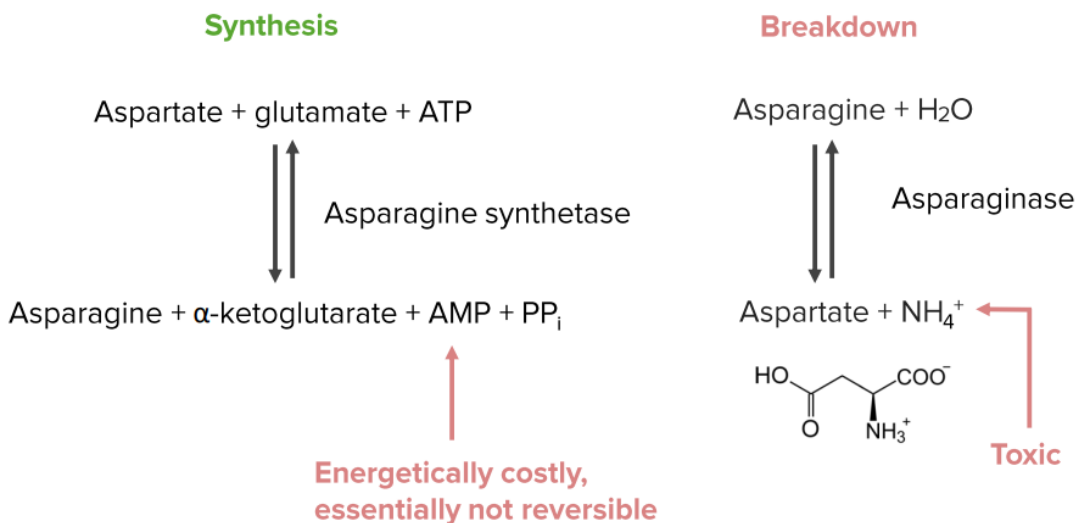


Image by Lecturio

## Methionine metabolism

- Breakdown overlaps with cysteine metabolism
- Complicated synthesis from aspartate

### Steps to the process

1. Phosphorylation
2. Oxidation and dephosphorylation
3. Oxidation
4. Creates homoserine

5. Succinylation
6. Cysteine replacement of succinate
7. Loss of pyruvate & ammonium ion
8. This creates homocysteine
9. Methylation of homocysteine by N5-methyl folate creates methionine (requires vitamin B<sub>12</sub>)

Homocysteine can be converted to methionine by the alternate path on the right.

- Methionine modified in bacteria for use in translation
- Occurs after methionine acts on initiator tRNA

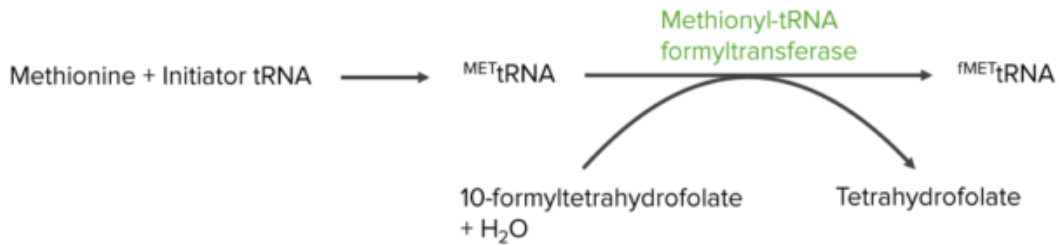


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## Threonine Metabolism

First 3 steps are the same as methionine metabolism, creating homoserine

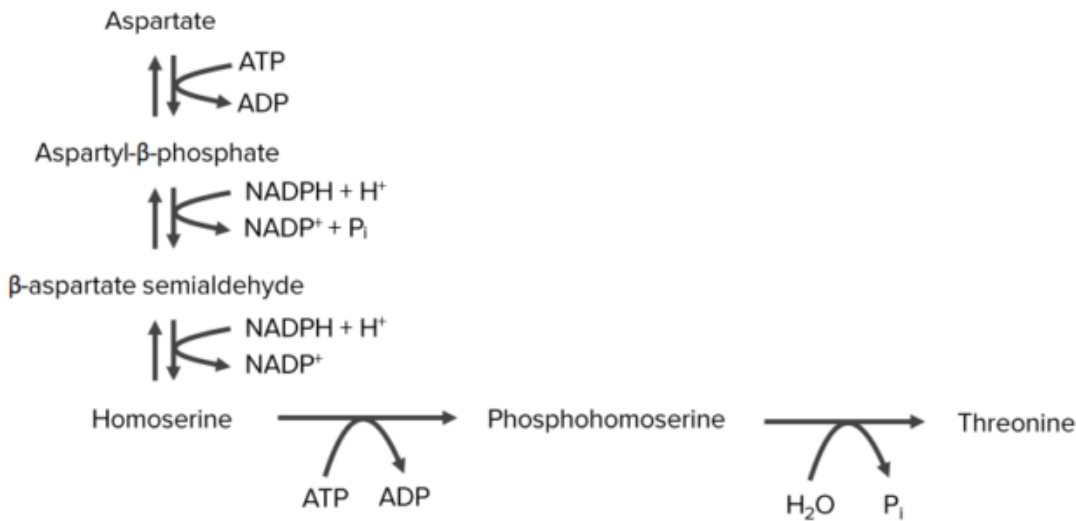


Image by Lecturio

## Lysine metabolism

- First 2 reactions are the same as threonine and methionine
- Nine steps in total
- Lysine is 1 of the most post-translationally modified amino acids, especially in histones
- Hydroxylation of lysine is important for making strong collagen
- Deficiency of 1 of the enzymes in the lysine degradation pathway, α-

aminoadipic semialdehyde synthase, leads to hyperlysinemia — accumulation of lysine in the blood

## Aromatic Family

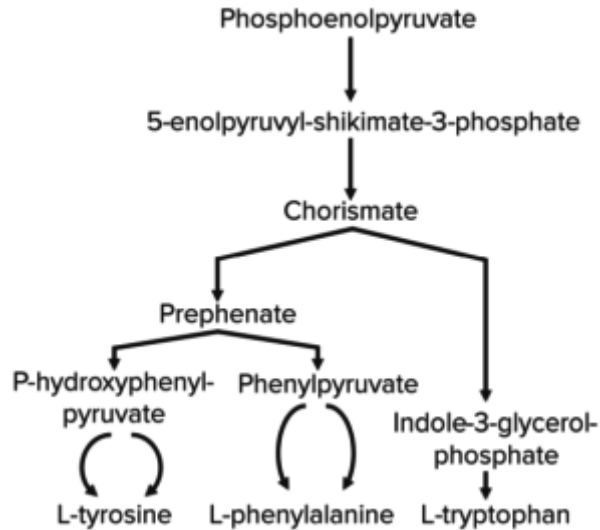


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Tryptophan, phenylalanine, and tyrosine

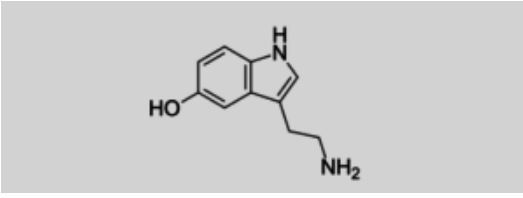
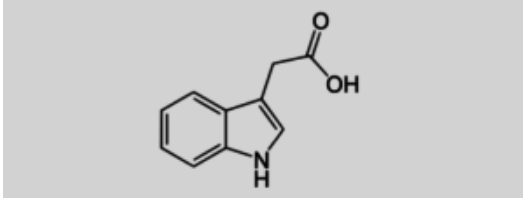
- Each derived from phosphoenol-pyruvate and erythrose-4-phosphate
- Synthesis pathways complex
- Each involves shikimic acid and chorismic acid
- Phenylalanine and tyrosine pathways overlap
- Hormones and neurotransmitters are made from each of these amino acids

## Tryptophan

- Interesting regulation of synthesis in bacteria
- Attenuation — all 5 genes on 1 operon
- When tryptophan is high, transcription of operon aborts early
- When tryptophan is low, transcription of operon continues through all genes

### Molecules made from tryptophan

Melatonin	Niacin
<p>Image by Lecturio</p>	<p>Image by Lecturio</p>
<ul style="list-style-type: none"> <li>• Circadian rhythm sensing</li> <li>• Affects mood, sleep, and blood pressure</li> <li>• Production affected by blue light</li> </ul>	<ul style="list-style-type: none"> <li>• Vitamin B<sub>3</sub></li> <li>• Nicotinamide derived from it — part of NAD<sup>+</sup>/NADH and NADP<sup>+</sup>/NADPH</li> <li>• Deficiency leads to pellagra</li> </ul>

Serotonin	Auxins
 <p data-bbox="316 443 459 465">Image by Lecturio</p>	 <p data-bbox="860 443 1003 465">Image by Lecturio</p>
<ul style="list-style-type: none"> <li>• Neurotransmitter</li> <li>• Causes vasoconstriction, enhanced memory/learning, and is a contributor to happy feelings</li> </ul>	<ul style="list-style-type: none"> <li>• Indole-3-acetic acid most important</li> <li>• Stimulate cell division and rooting in plants</li> </ul>

## Phenylalanine (PHE)

- Essential amino acid and precursor of tyrosine
- PHE hydroxylase catalyzes the formation of tyrosine from PHE
- Deficiency of the enzyme PHE hydroxylase causes phenylketonuria
- High PHE levels cause damage to the brain
- Treatable by reducing PHE levels
- Nutrasweet contains PHE

## Tyrosine (TYR)

- Not essential if PHE present
- The precursor of catecholamines — L-dopa, L-dopamine, norepinephrine, and epinephrine
- Donates electrons to reduce chlorophyll in photosystem 2
- Forms radical in ribonucleotide reductase

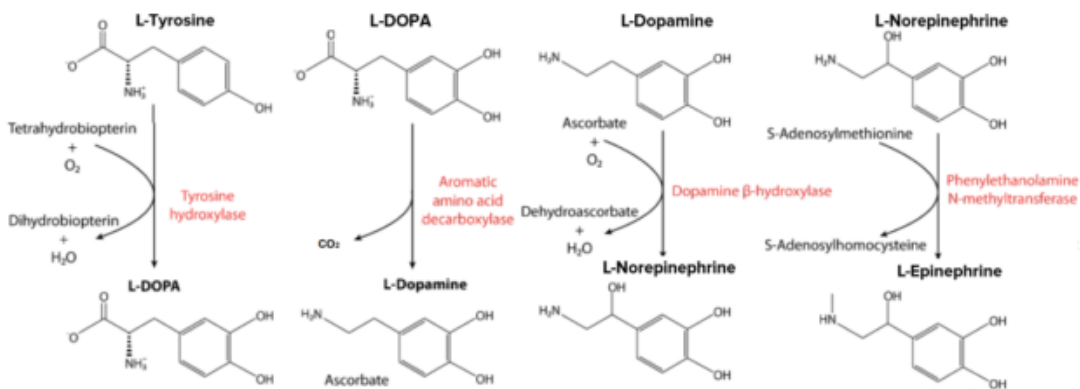
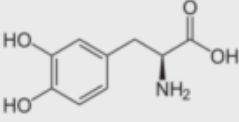
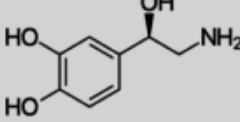
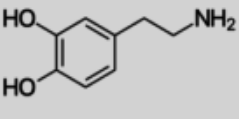
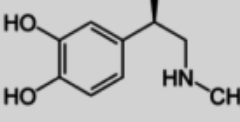


Image by Lecturio

## Tyrosine metabolism

L-DOPA	Norepinephrine
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 <p>Image by Lecturio</p>	 <p>Image by Lecturio</p>
<ul style="list-style-type: none"> <li>• Precursor to dopamine</li> <li>• Crosses blood-brain barrier</li> <li>• used to treat Parkinson's disease</li> </ul>	<ul style="list-style-type: none"> <li>• Hormone and neurotransmitter</li> <li>• Works through noradrenergic receptors <ul style="list-style-type: none"> <li>• Fight or flight response</li> </ul> </li> <li>• Increases heart rate and blood pressure</li> </ul>
<p><b>Dopamine</b></p>	<p><b>Epinephrine (adrenalin)</b></p>
 <p>Image by Lecturio</p>	 <p>Image by Lecturio</p>
<ul style="list-style-type: none"> <li>• Neurotransmitter</li> <li>◦ Inhibits norepinephrine release in blood vessels — acts as a vasodilator</li> <li>◦ Reduces insulin production in the pancreas</li> <li>◦ Deficiency causes Parkinson's disease</li> <li>◦ Links to schizophrenia and attention deficit hyperactivity disorder (ADHD)</li> </ul>	<ul style="list-style-type: none"> <li>• Hormone</li> <li>• Actions similar to norepinephrine <ul style="list-style-type: none"> <li>• Fight and flight response</li> </ul> </li> <li>• Increases heart rate and blood pressure</li> </ul>

Tyrosine is a precursor of the thyroid hormones:

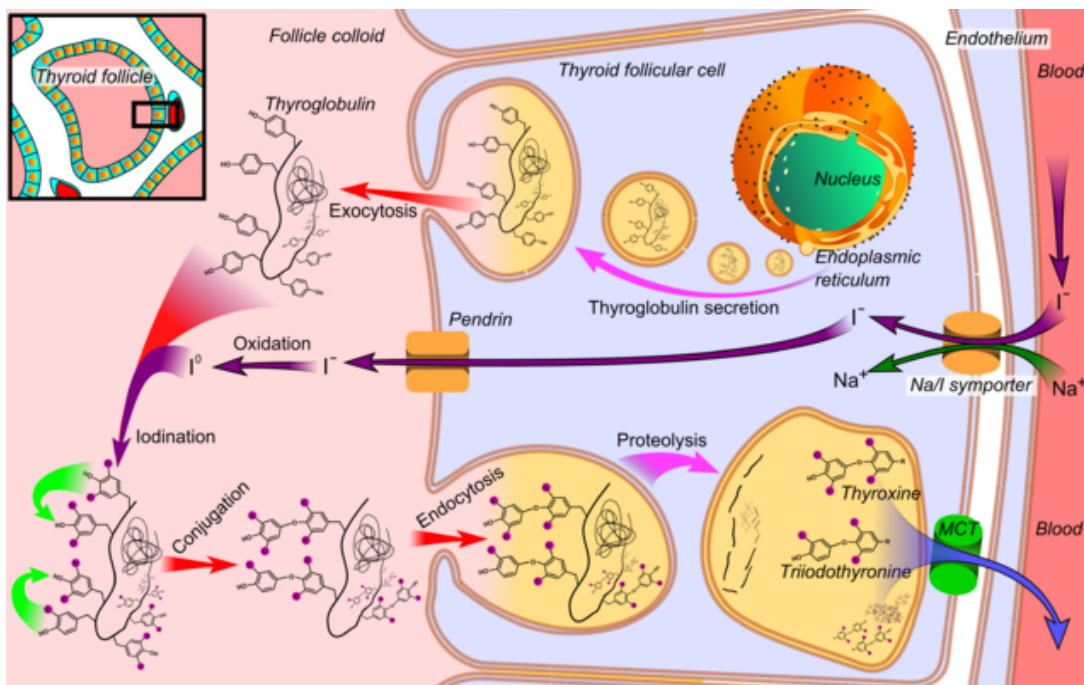


Image: Thyroid hormone synthesis. By Haggström, Mikael, License: [CC0](https://creativecommons.org/licenses/by/4.0/)

## Tyrosine metabolism & disease



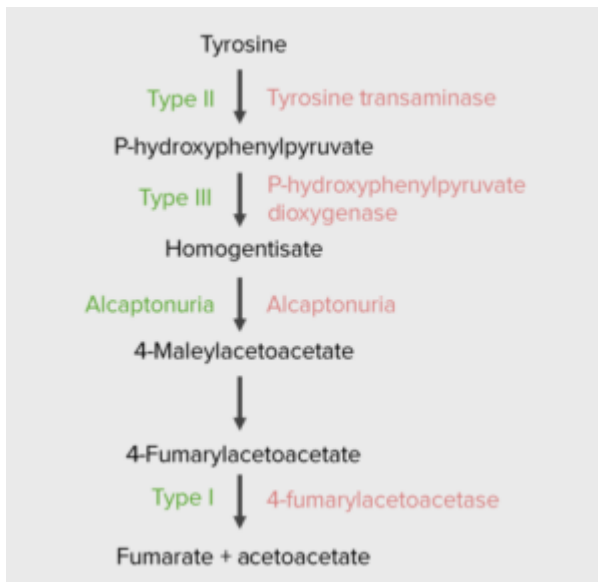


Image by Lecturio

## Tyrosinemia — problems with tyrosine catabolism

- Type 1
- Type 2
- Type 3
- Alcaptonuria — black urine disease

## Treatments

- Restricted TYR/PHE diet
- Liver transplant

## Pyruvate Family

### Alanine metabolism

- Most easily produced from pyruvate — transamination
- A byproduct of catabolism of valine, leucine, and isoleucine
- Glucose-alanine cycle

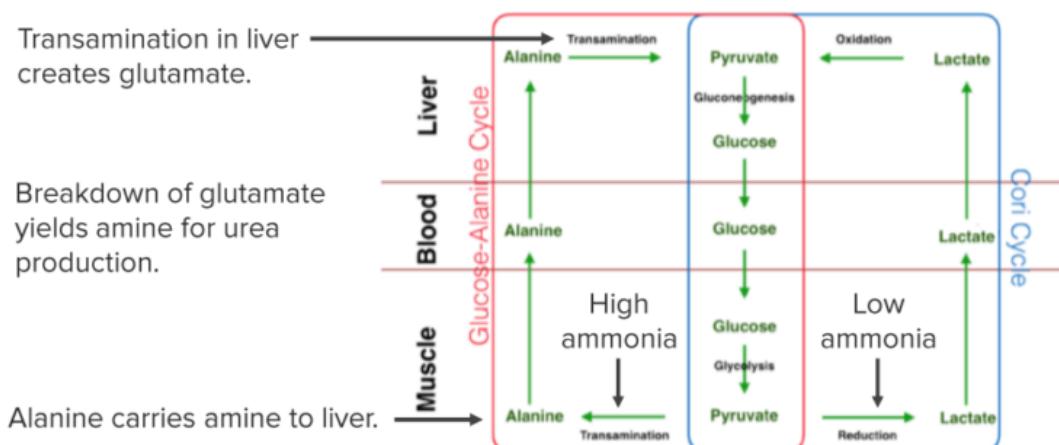


Image: Glucose-alanine-cycle — important for removing ammonia. By Lecturio

## Leucine (LEU)/Valine (VAL)/Isoleucine (ILE) metabolism

- Branched-chain amino acids (BCAAs)
- Several common steps
- Start with decarboxylation and attachment of 2 carbon piece to TPP (thiamin pyrophosphate)
- Valine and leucine pathways involve attachment of 2 carbon piece to pyruvate
- Isoleucine pathway involves attaching 2 carbon piece to  $\alpha$ -ketobutyrate
- Penultimate products —  $\alpha$ -ketoisocaproate (LEU),  $\alpha$ -ketoisovalerate (VAL), and  $\alpha$ -keto- $\beta$ -methyl valerate (ILE) each is transaminated to make a final amino acid

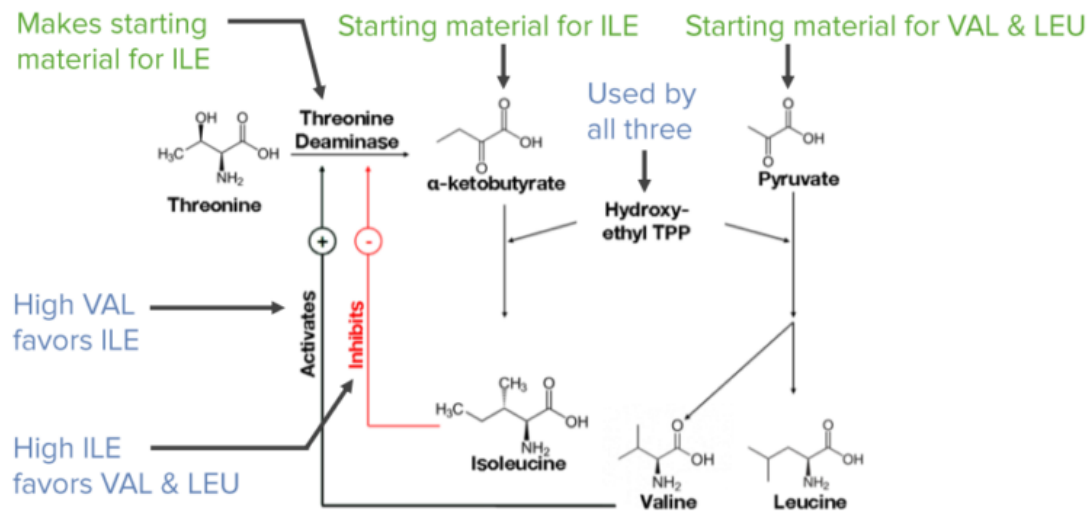


Image: Synthesis feedback is regulated through threonine deaminase. By Lecturio

- Breakdown of BCAAs occurs through the branched-chain  $\alpha$ -keto acid dehydrogenase complex
- Mutations altering this complex can result in maple syrup urine disease

## Histidine (HIS) metabolism

- The most complex of all the amino acids
- Overlaps nucleotide metabolism with ribose-5-phosphate & PRPP (phosphoribosyl pyrophosphate)
- Ten steps in the pathway
- Second enzyme of pathway (ATP-phosphoribosyltransferase) feedback inhibited by histidine

## Disorders of the Amino Acid Metabolism

Relatively common genetic metabolic disorder is **phenylketonuria, PKU** (approx. 1 in 10,000 newborns is affected). Every newborn is tested for PKU (e.g., with the **Guthrie test**) because most effects of the disease can be avoided by keeping a strict diet.

Due to a defect of the enzyme phenylalanine hydroxylase, the amino acid phenylalanine accumulates in the tissue. Instead, the body turns phenylalanine into the keto acid phenylpyruvate. This product is partly exuded – which explains the term

phenylketonuria—but partly metabolized further, building cytotoxic substrates. They cause damage, especially in the myelin sheaths of the central nervous system (CNS). This impairs the normal development of the brain. It is mainly facilitated by excess quantities of phenylalanine compared to the concentration of other amino acids being transported across the blood-brain barrier, resulting in a deficit of required metabolites. The consequence of this disease is mental retardation. The newborn screening aims to identify affected persons and give them a specific diet that is low in phenylalanine and rich in tyrosine (tyrosine becoming the essential amino acid for the affected person). This way, damages can be prevented, or at least kept at bay.

Another genetically linked disease of phenylalanine catabolism is **alkaptonuria**, caused by the defective enzyme known as homogentisate dioxygenase. It is less fatal than PKU. This condition has fewer symptoms, although large amounts of homogentisate are excreted through the urine, and laboratory tests confirm its presence when the urine sample turns black after oxidation. Alkaptonuria patients are also prone to develop a form of arthritis.

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Notes