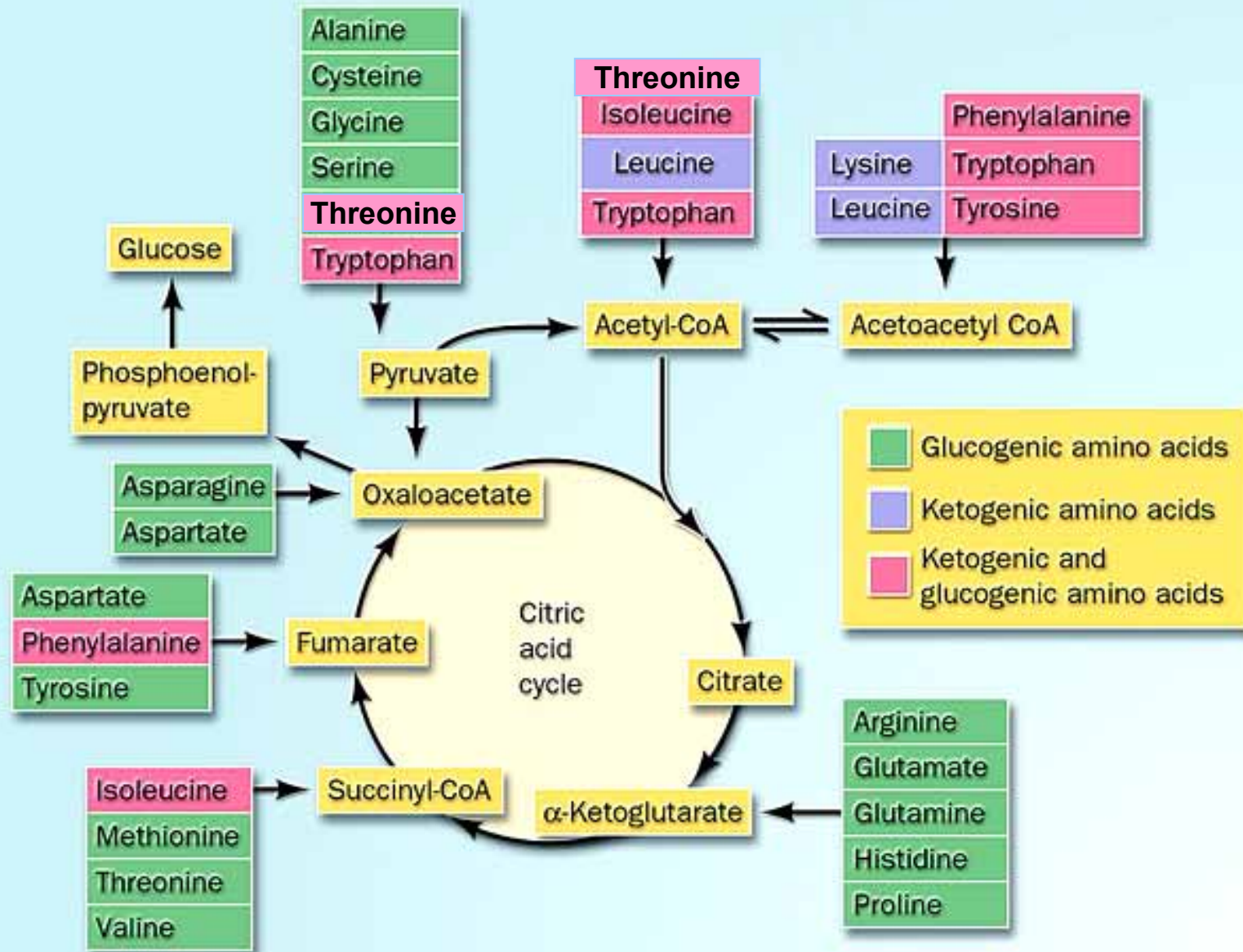


Aminosavak

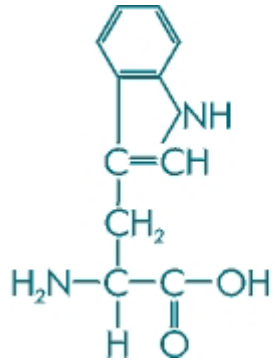
A szénlánc sorsa



Lebontási sorozatok

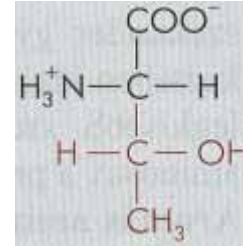
- | | | | |
|----|-----------------------------------|---|-----------------|
| 1. | Piroszölősav csoport | } | nem citrátkörös |
| 2. | Acetoacetyl-CoA csoport | | |
| 3. | α - keto-glutársav csoport | | |
| 4. | Szukcinil-CoA csoport | } | citrátkörös |
| 5. | Fumársav csoport | | |
| 6. | Oxálecetsav csoport | | |

1. Piroszőlősavvá alakuló aminosavak

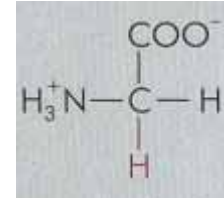


triptofán

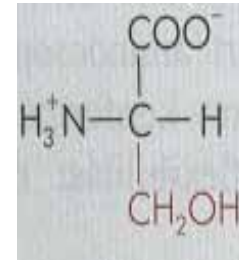
treonin



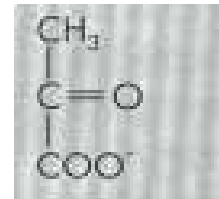
glicin



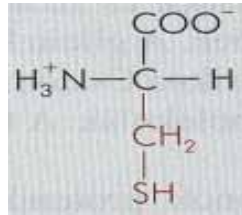
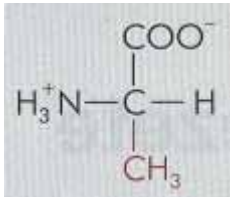
szerin



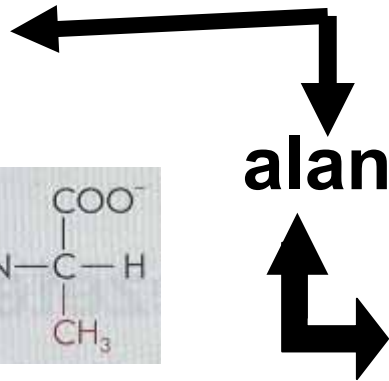
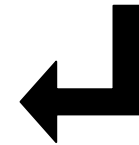
piroszőlősav



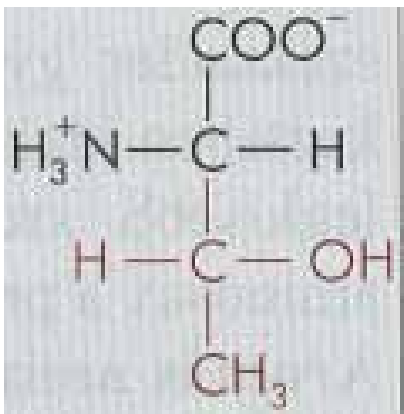
alanin



cisztein

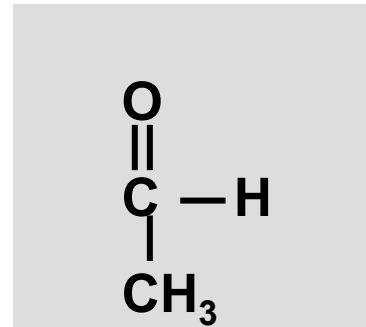


Treonin lebontása



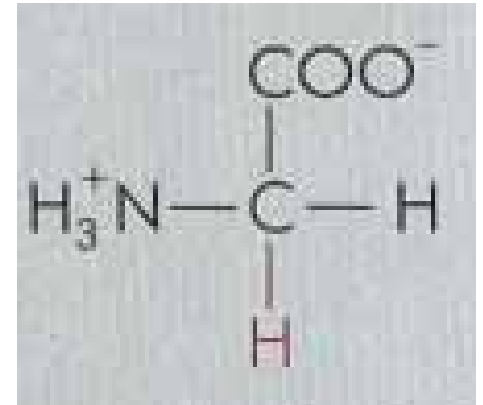
treonin

**acetaldehid-
liáz**



acetaldehid

+



glicin

Treonin dehidratáz



propionát



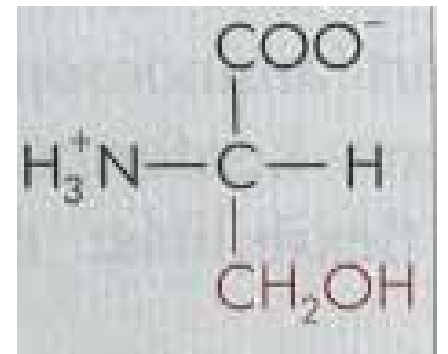
Szukcinil-koA

**Acetaldehid
dehidrogenáz**



acetát

szerin-hidroximetil-transzferáz

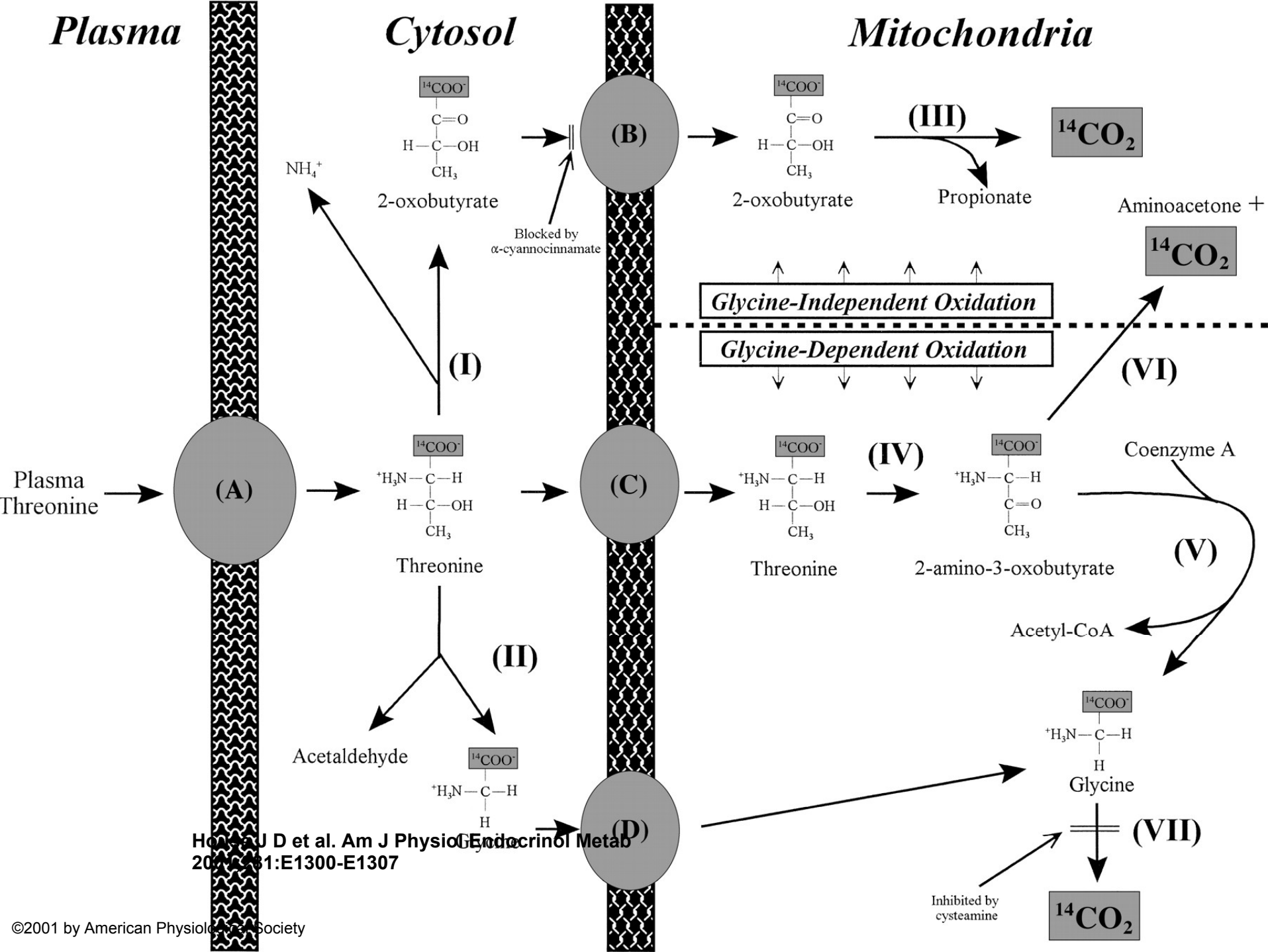


szerin

Plasma

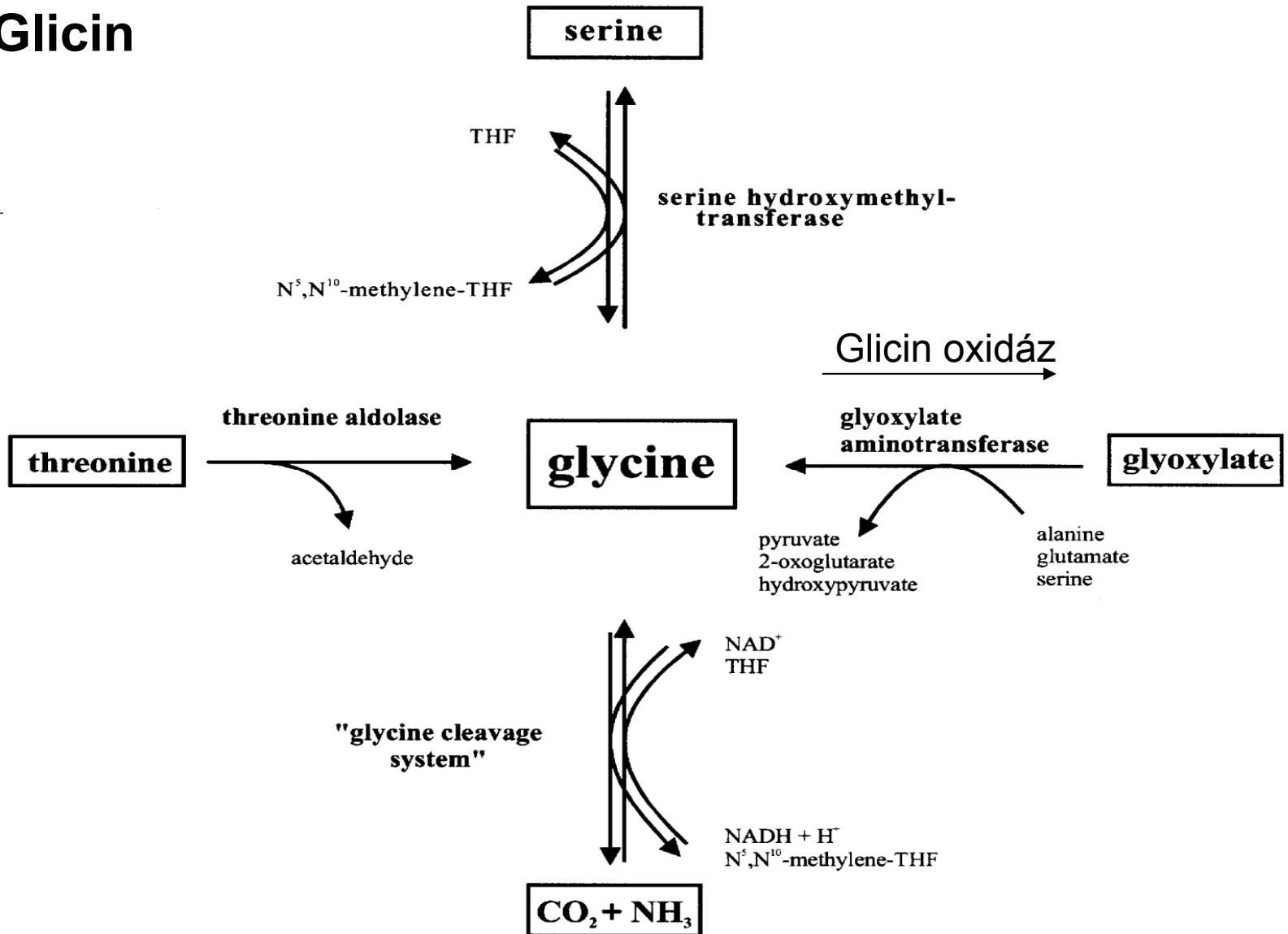
Cytosol

Mitochondria



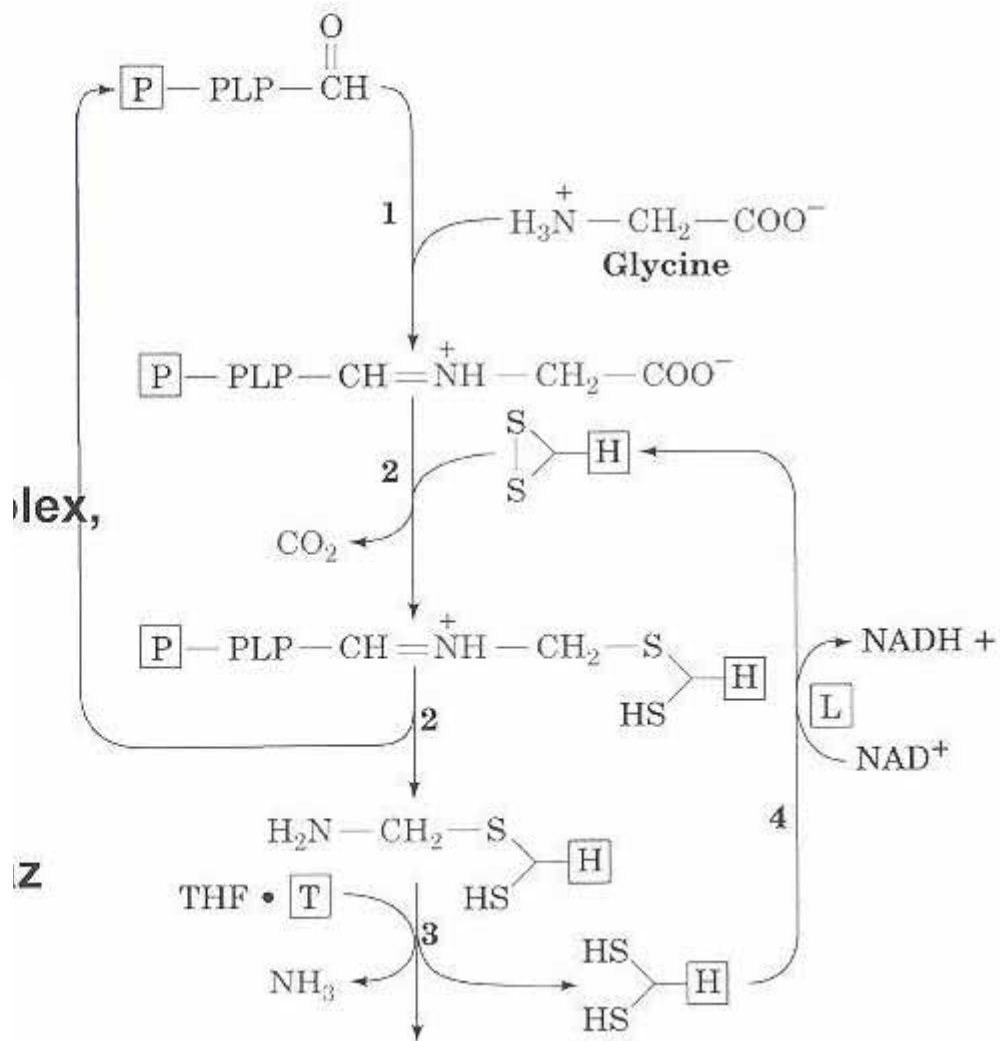
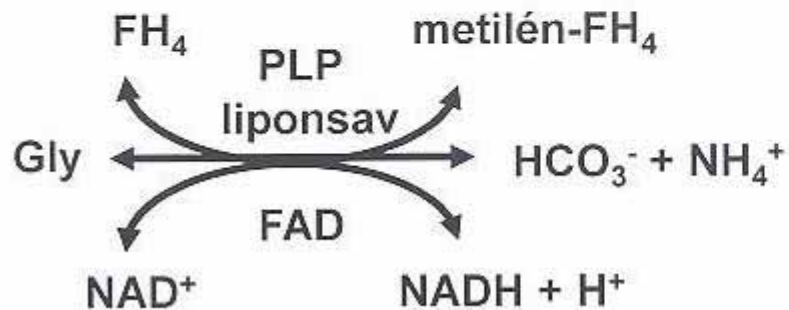
Hong J D et al. Am J Physiol Endocrinol Metab 2001; 281:E1300-E1307

Glicin



cin

Glicin szintetáz: reverzibilis enzim, szintézis is történhet



m komplex.

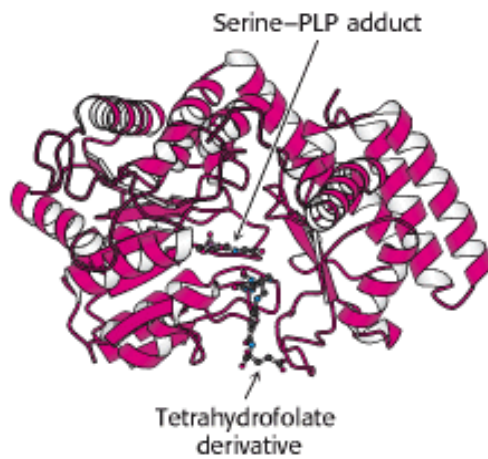
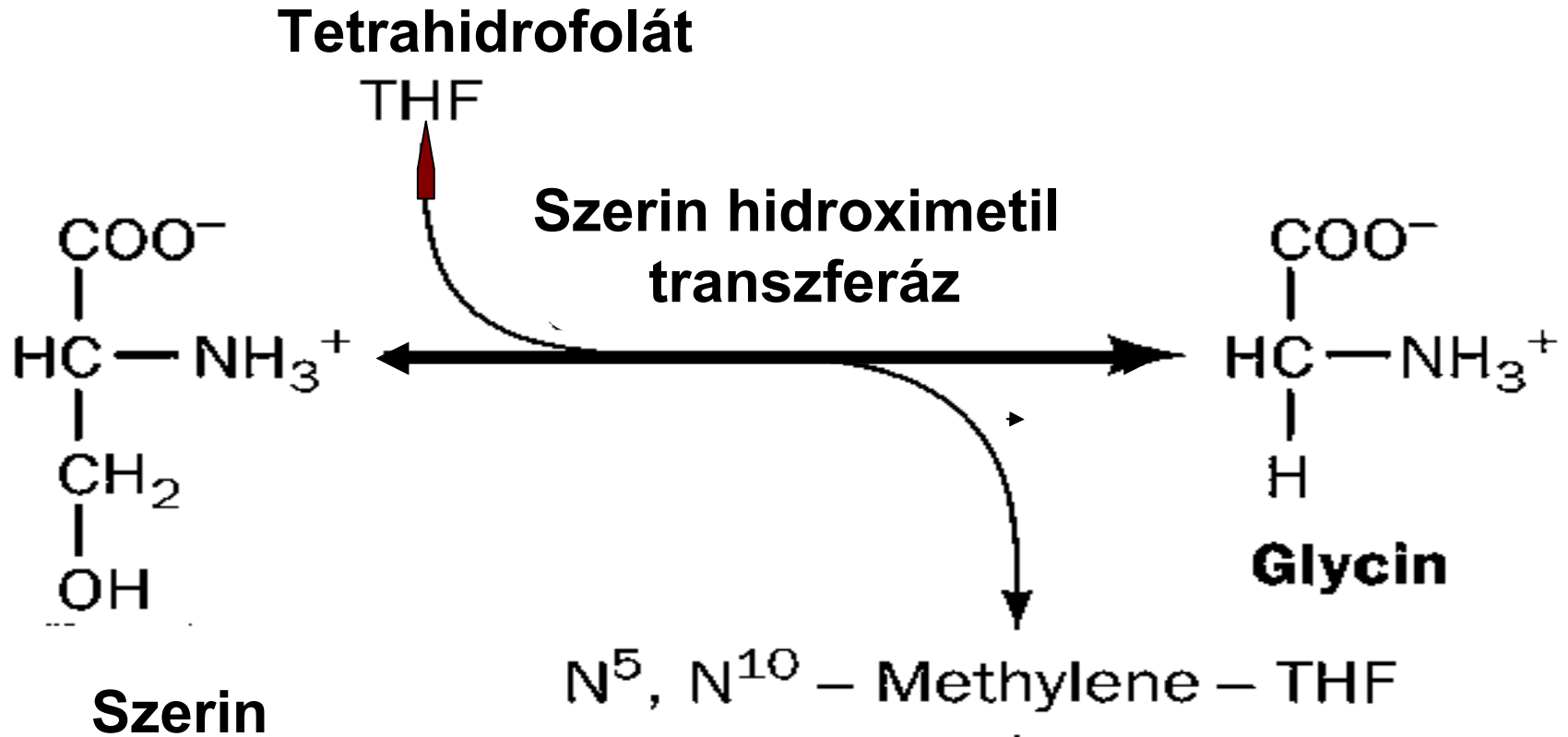
hasonló:

zim PLP-vel működik.

zim prosztetikus csoportja lipoamid.

zim prosztetikus csoportja FAD, koenzime

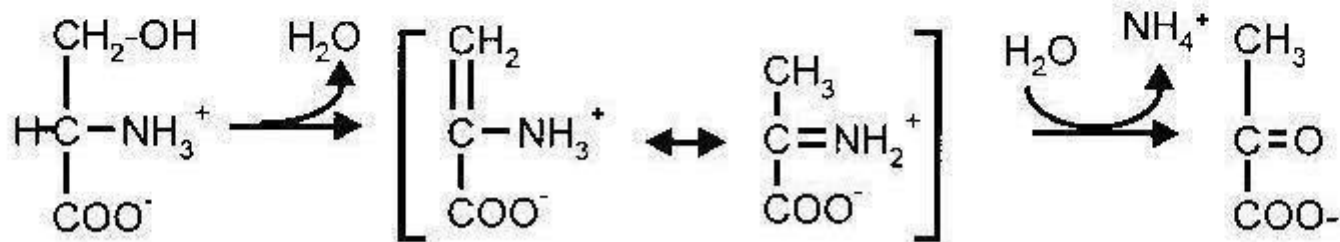
Glicin



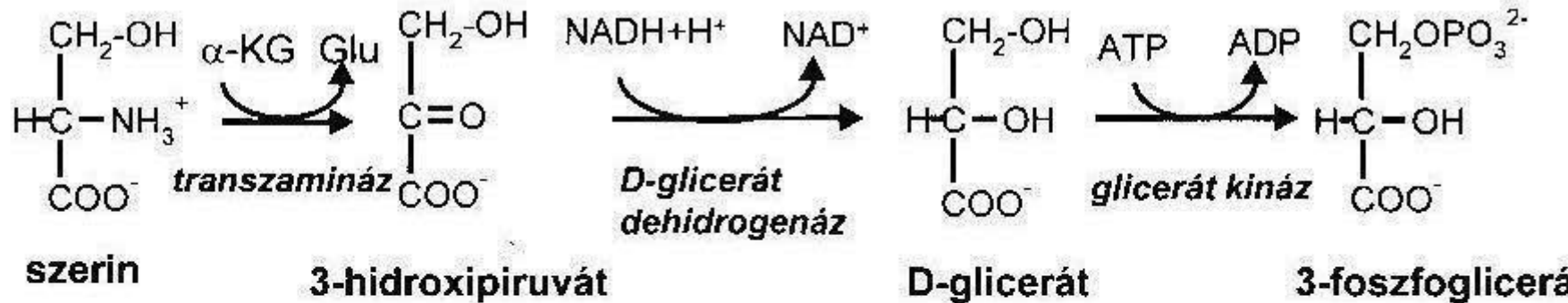
Szerin lebontása

e

Szerin-dehidratáz:



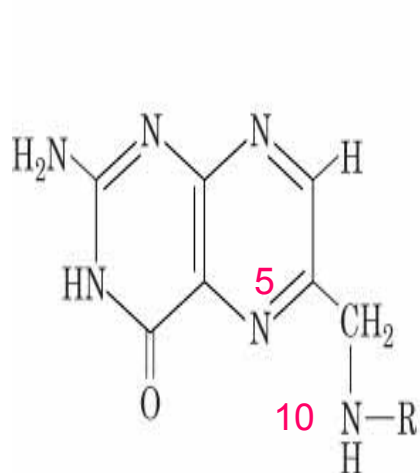
Glükoneogenesis irányába:



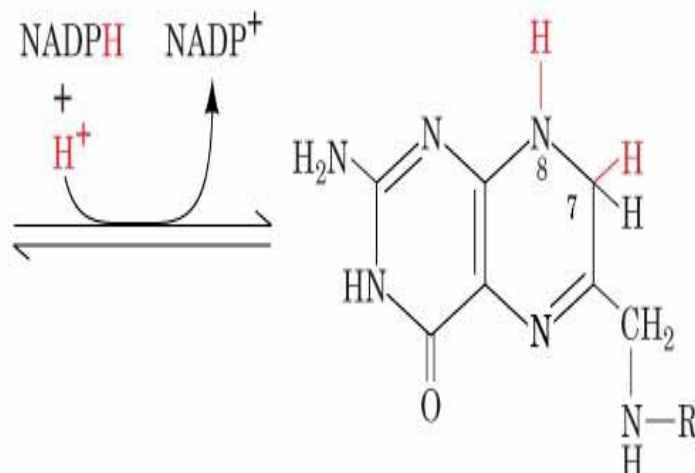
*Szerin-
hidroximetiltranszferáz:*
Szerin glicin átalakulás

C1 intermedierek szállítása

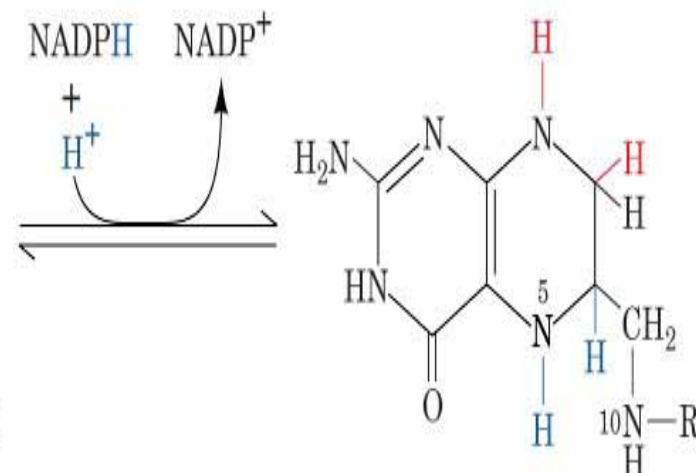
FOLSAV



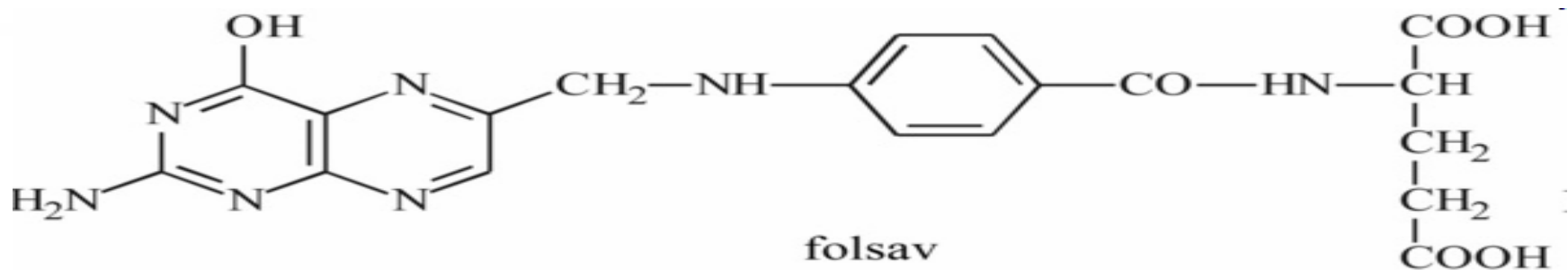
Folát



Dihidrofolát



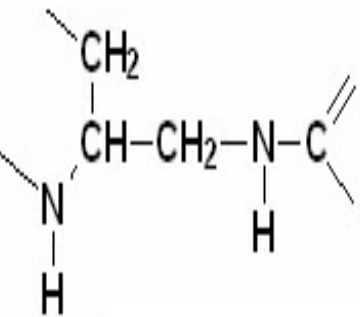
Tetrahidrofolát



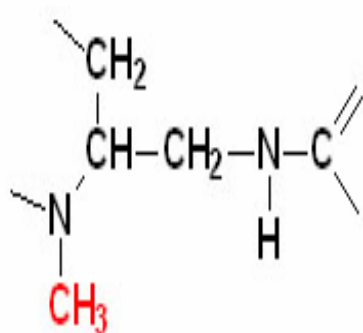
PTERIDIN

p-AMINO BENZOESAV

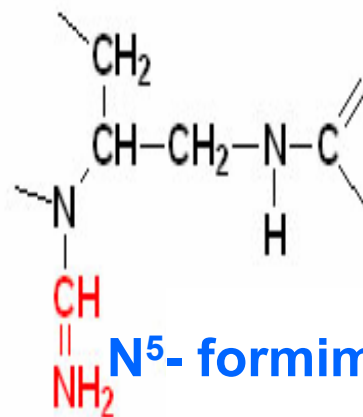
GLUTAMÁT



THF

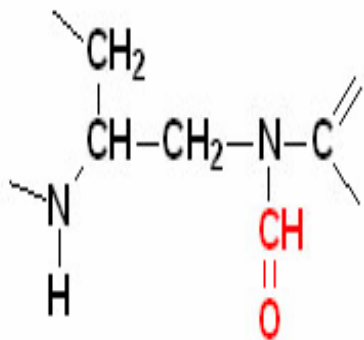


N⁵-metil THF

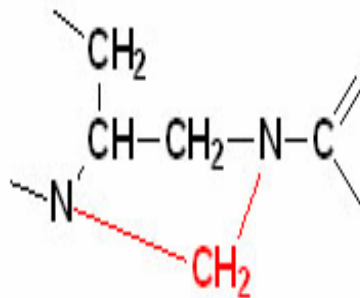


N⁵-formimino THF

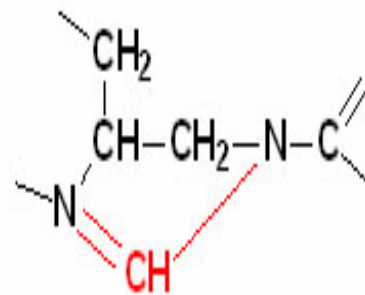
C1 intermedierek



N¹⁰-formil THF



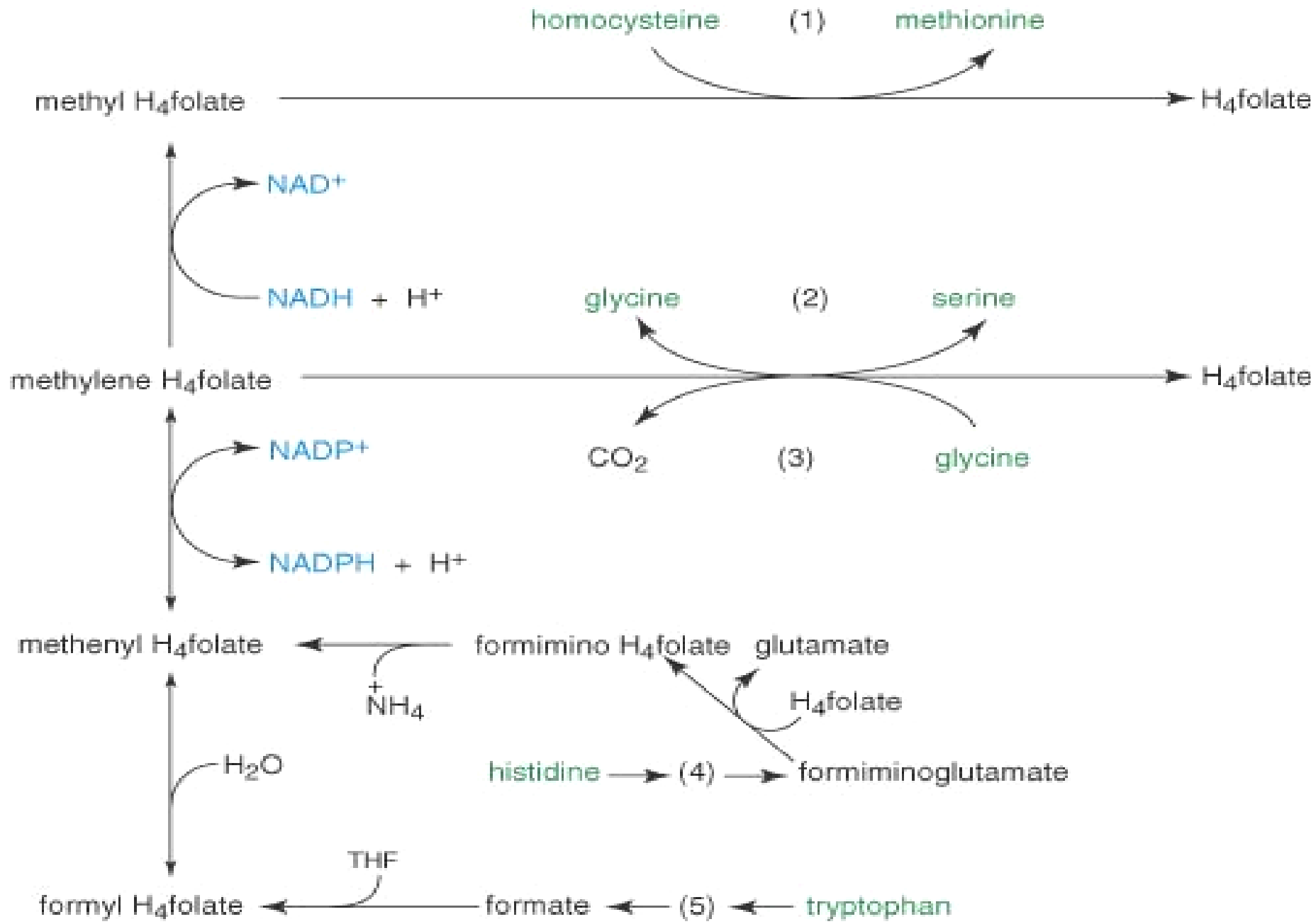
N⁵,N¹⁰-metilén THF



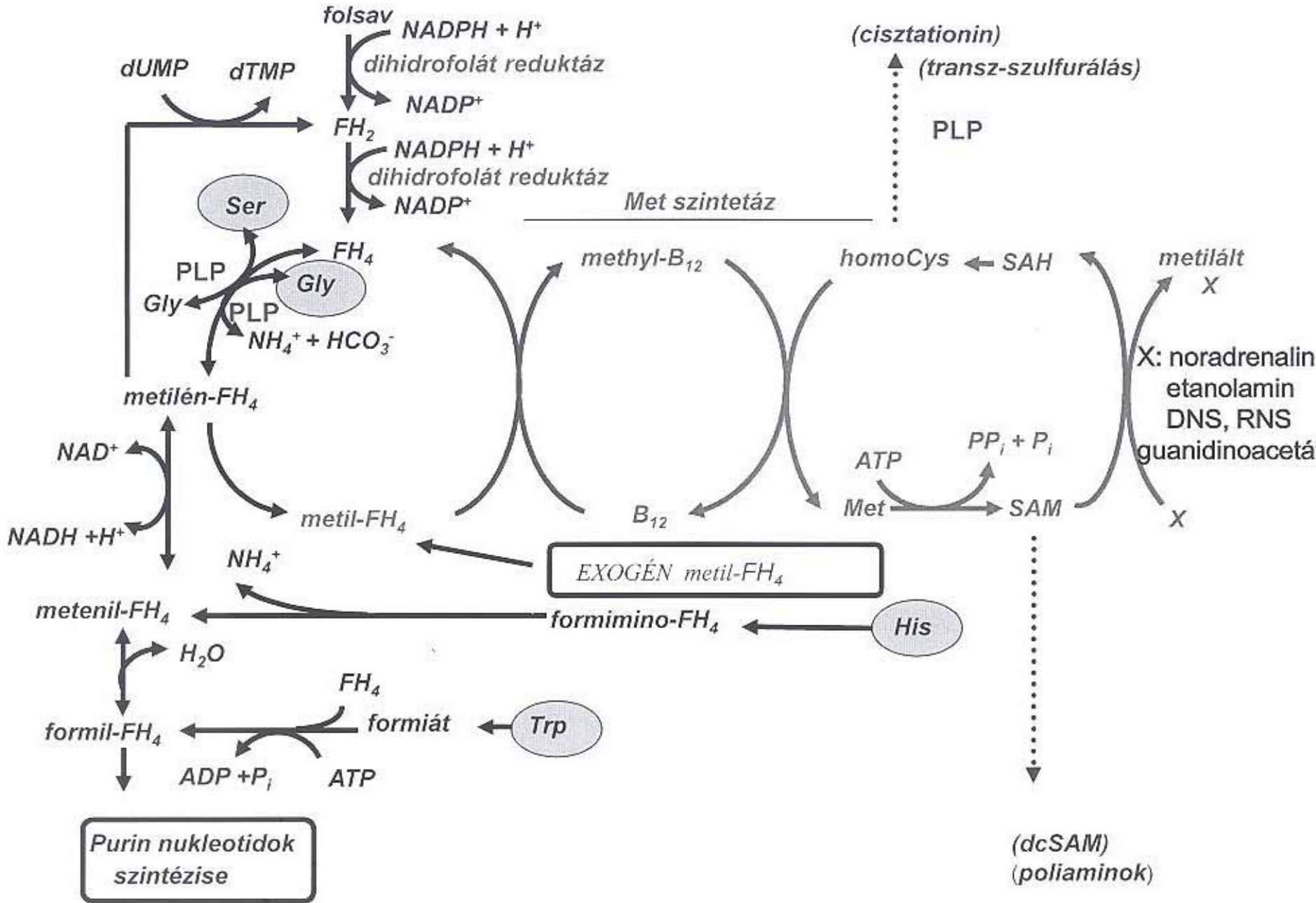
N⁵,N¹⁰-metenil THF

Oxidációs állapot	csoport
Legkevésebé oxidált („CH ₃ OH”)	-CH ₃ , metil
Közbülső („CH ₂ =O”)	-CH ₂ ⁻ , metilén
Legoxidáltabb („HCOOH”)	-CH=O, formil
	-CH=NH, formimino
	-CH=, metenil

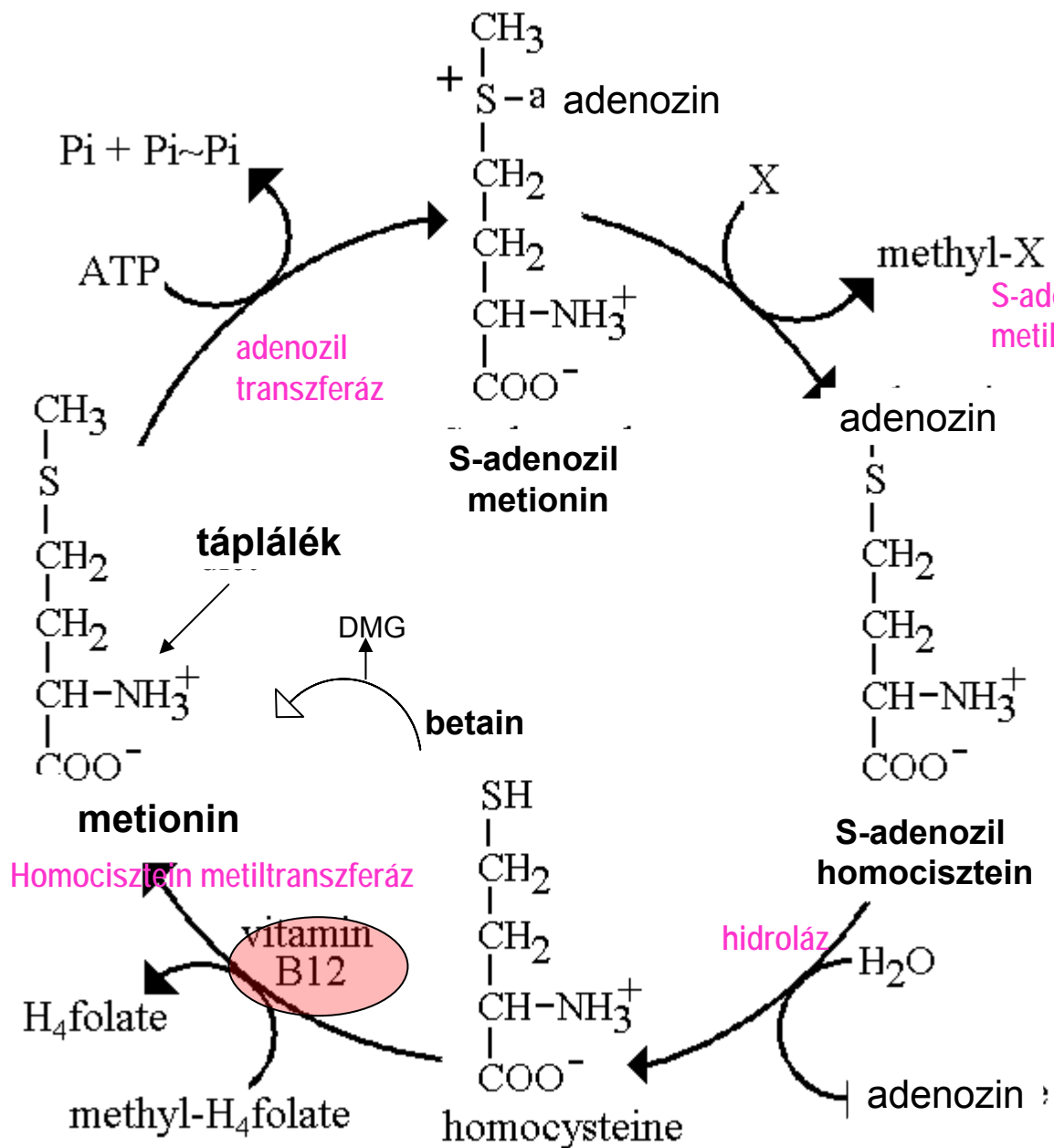
A THF szerepe az aminosav anyagcserében



C₁ egységek keletkezése és felhasználása, transzmetilálás

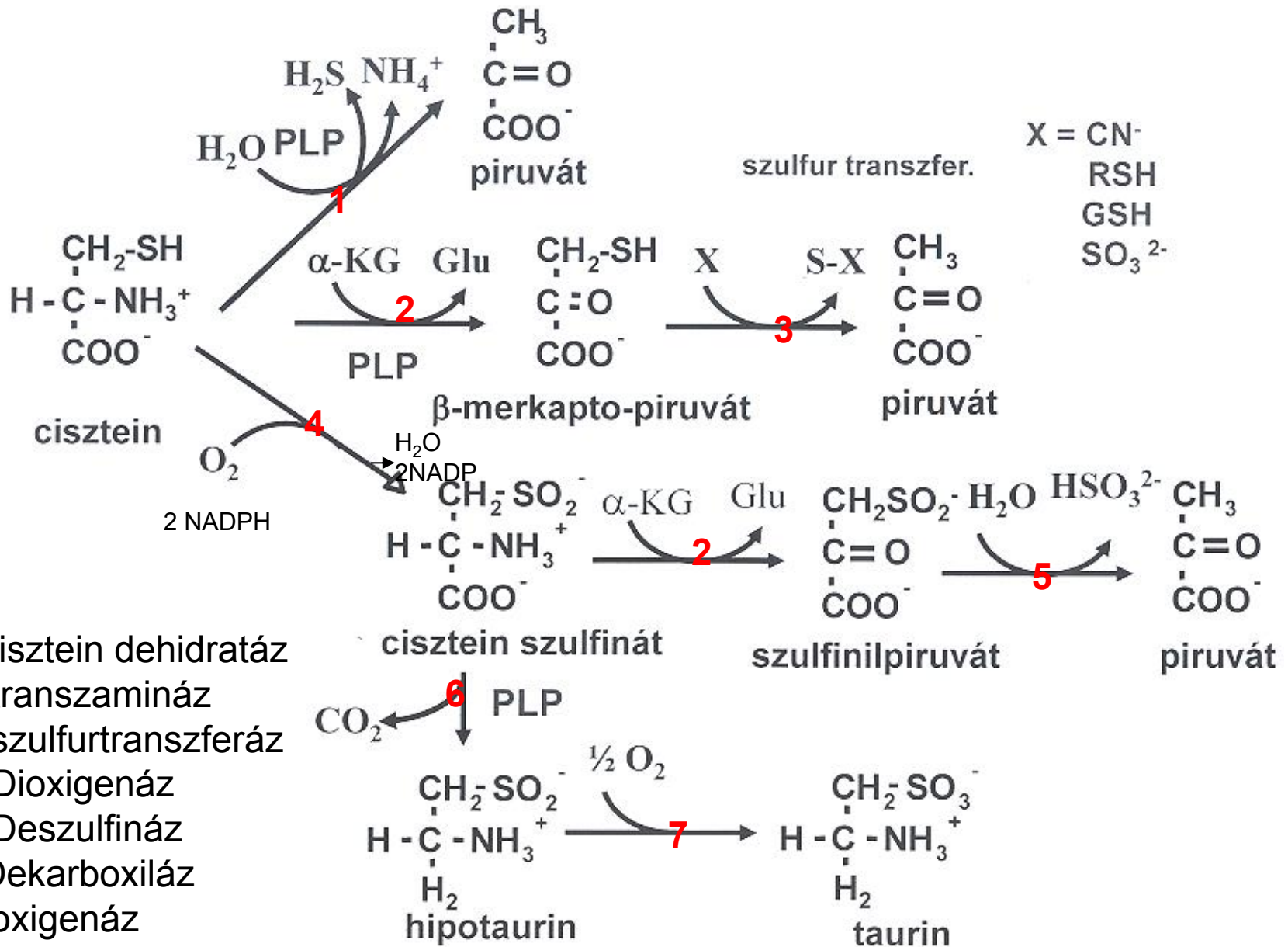


Aktivált metil ciklus



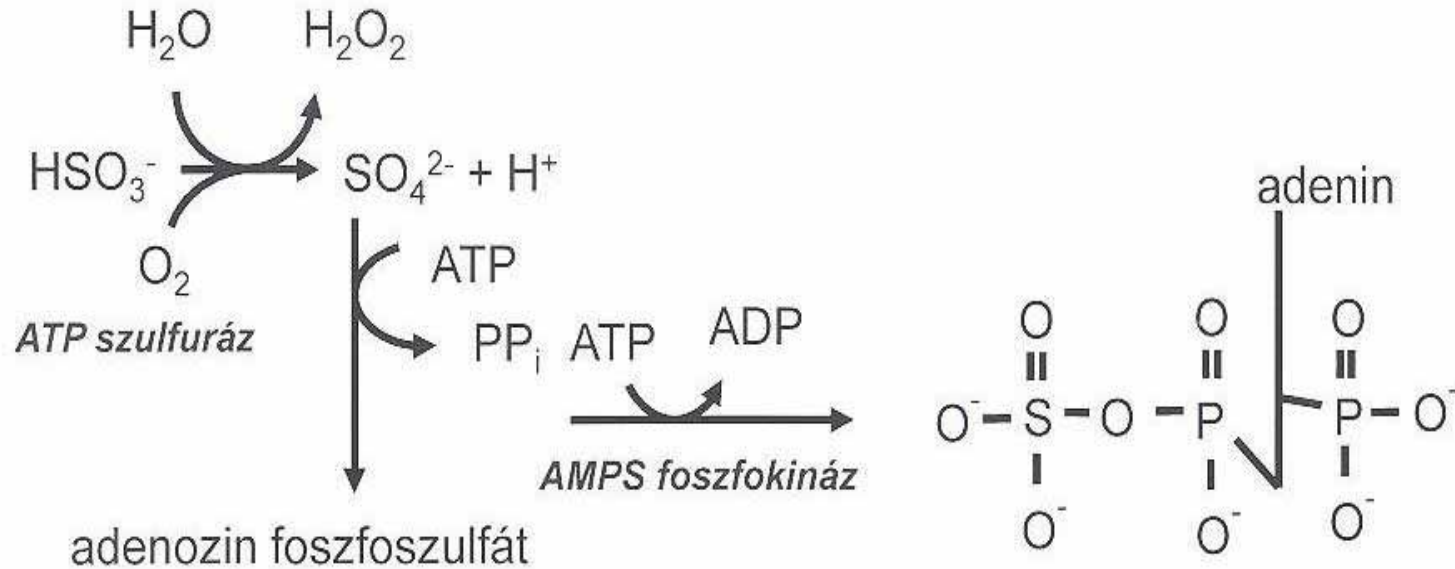
•**SAM**: metil csoport donor a szintetikus reakciókban, DNS, RNS, proteinek metilációja, foszfatidilkolin bioszintézis kreatin, adrenalin, karnitin bioszintézise

A cisztein lebontása



- 1: cisztein dehidratáz
- 2: transzamináz
- 3: szulfurtranszferáz
- 4: Dioxigenáz
- 5: Deszulfínáz
- 6: Dekarboxiláz
- 7: oxigenáz

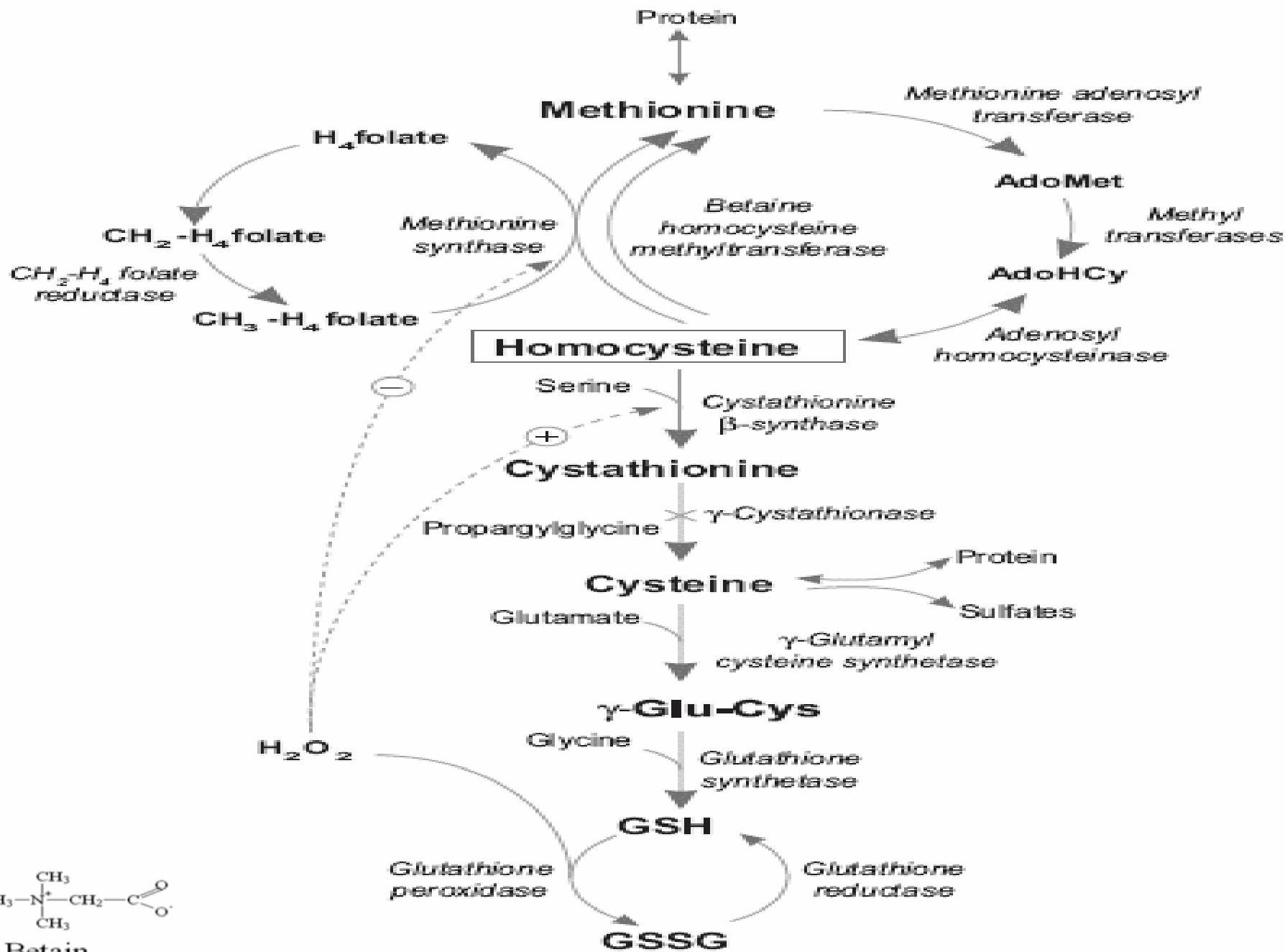
Szulfid ion sorsa, PAPS szintézise és felhasználása



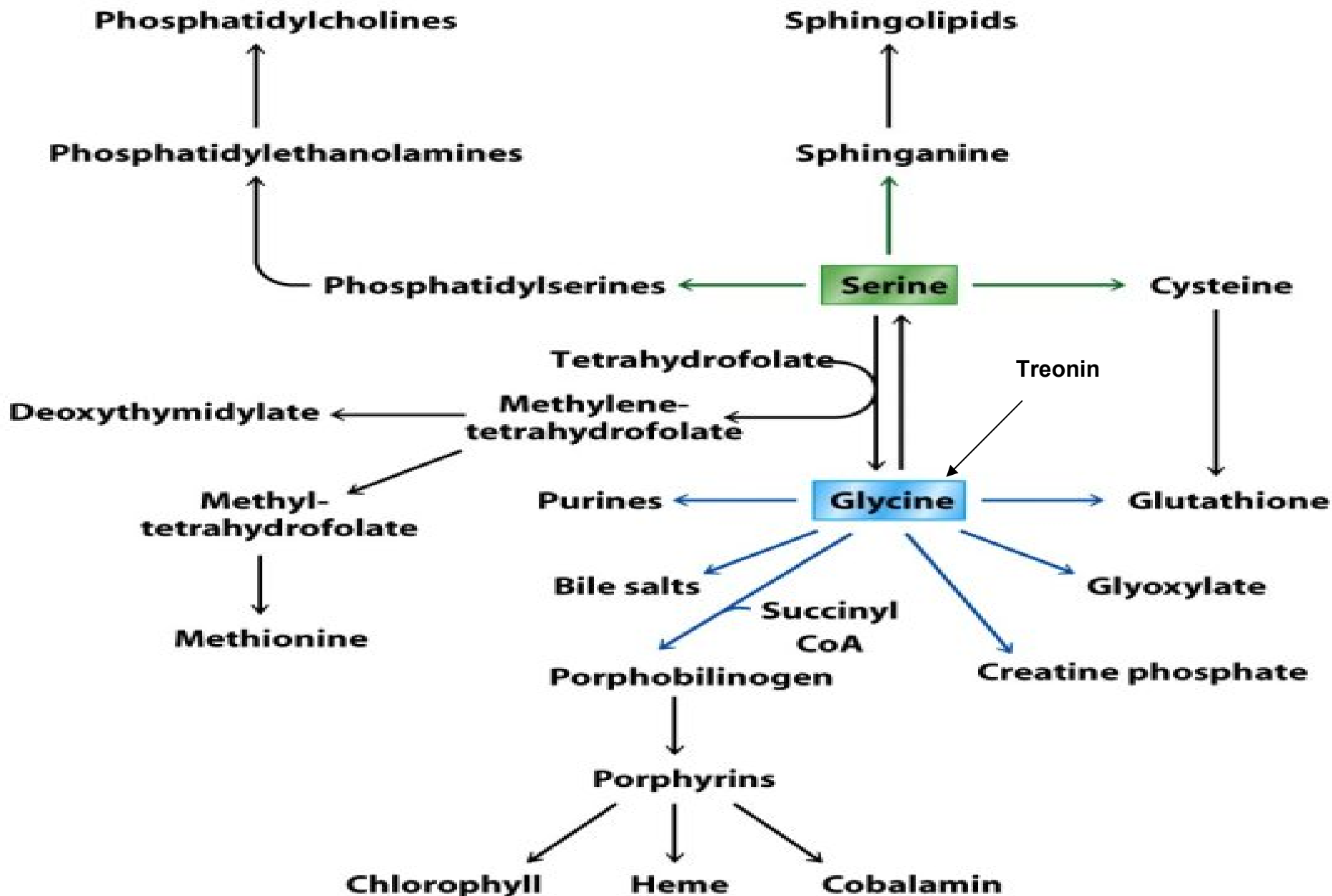
- SO_2^{2-} szulfínát
- SO_3^{2-} szulfid
- SO_4^{2-} szulfát

(AMPS)

foszfoadenozin foszfoszulfát
(PAPS)

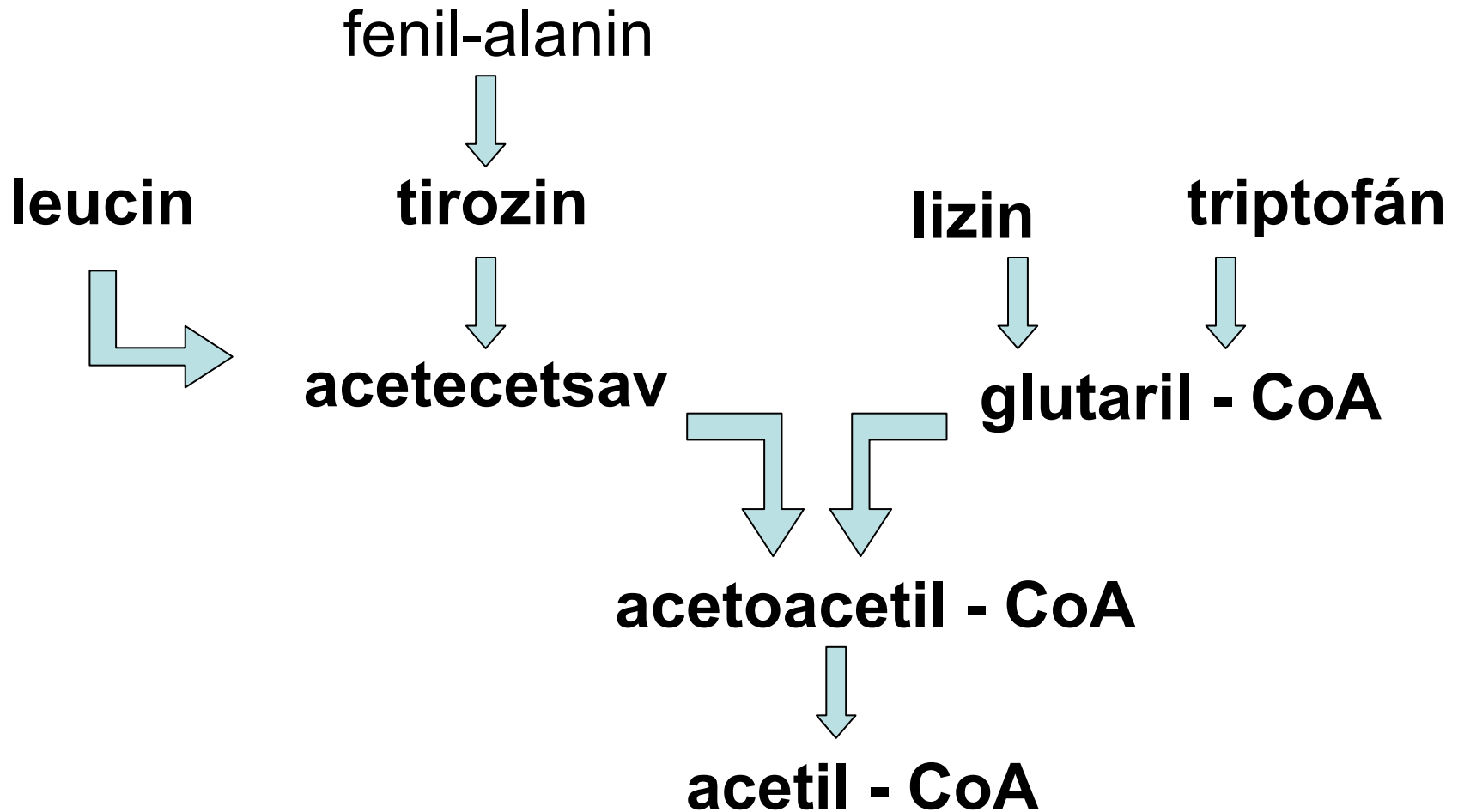


Szerin és a glicin szerepe



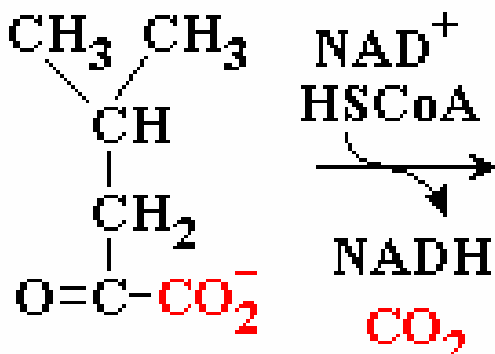
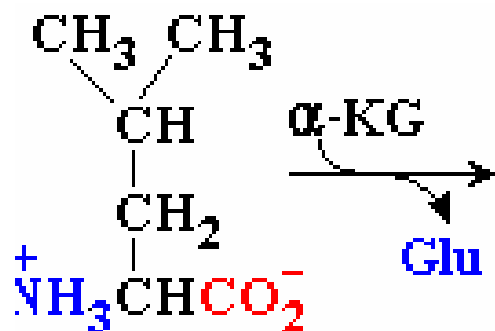
2. Acetoacetyl-CoA csoport lebontása

(fenil-alanin és tirozin lebontása, katecholaminok szintézise)

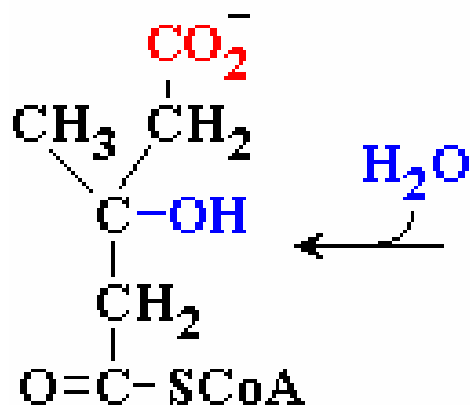
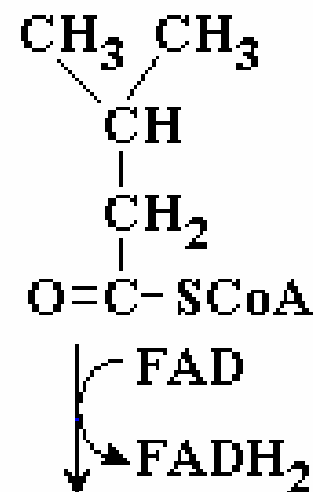


Leucin elágazó szénláncú as

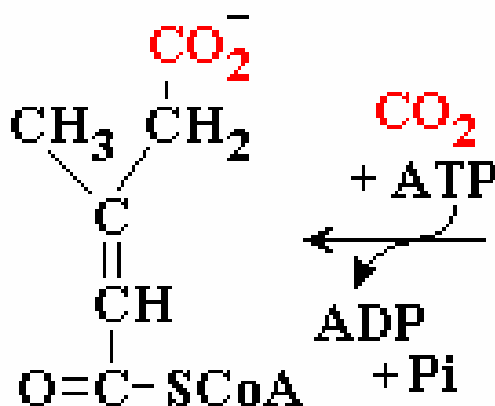
Leucin



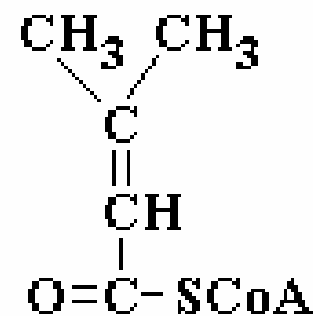
izovaleril-koA



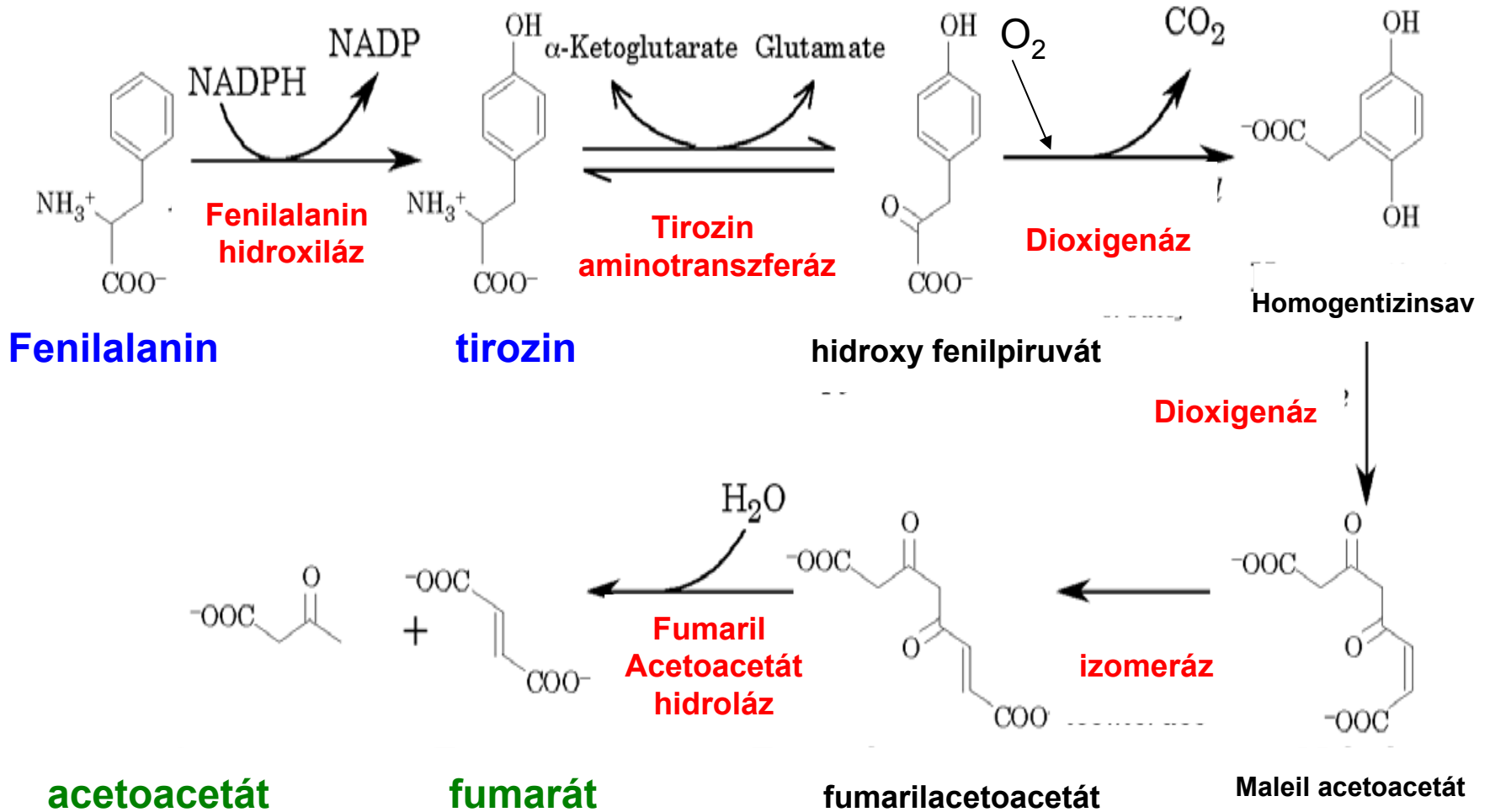
HMG-koA



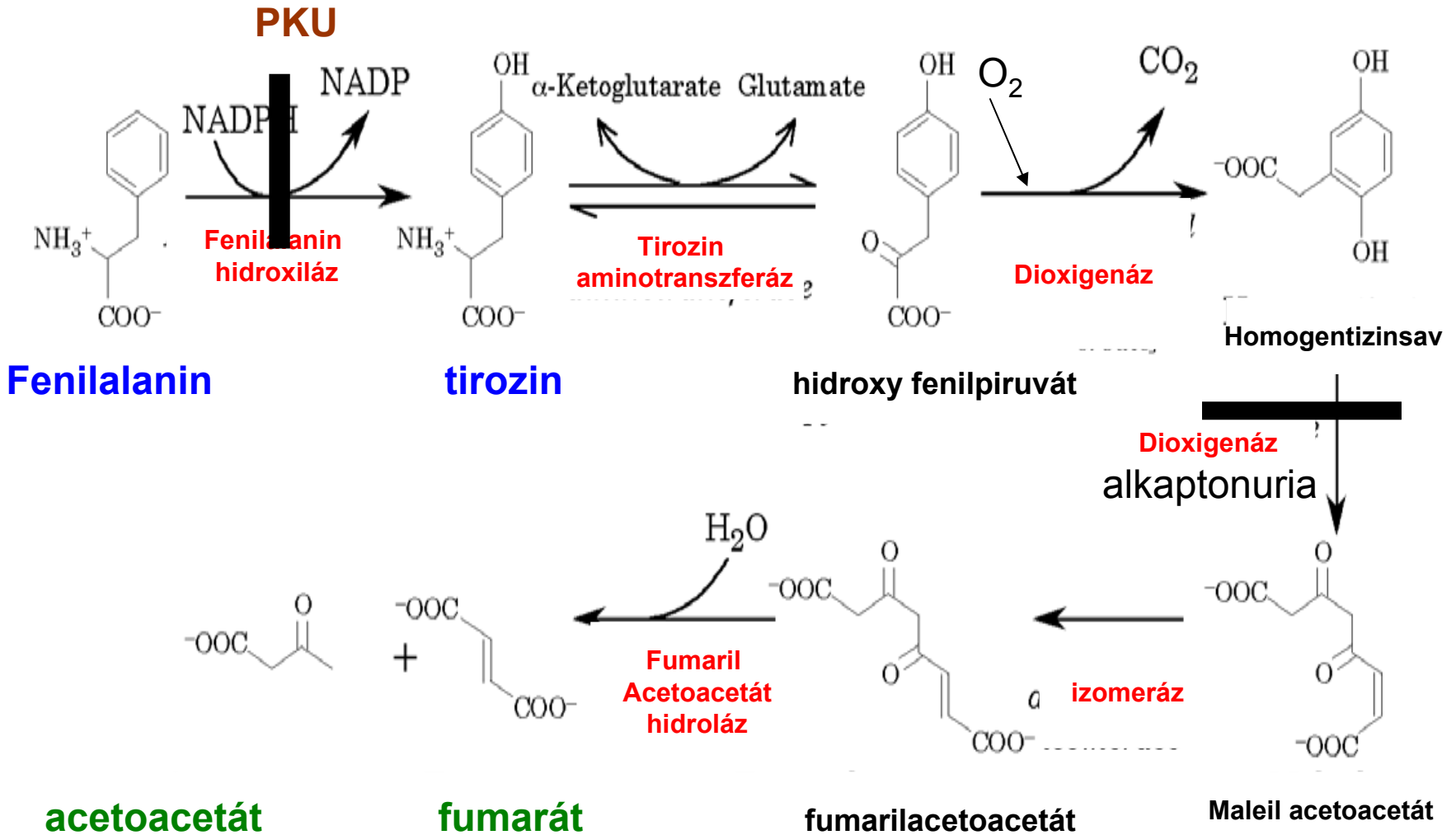
metilkrotonil-koA



Fenilalanin tirozin anyagcsere



Fenilalanin tirozin anyagcsere



Phenylketonuria (PKU)

Fenilalanin hidroxiláz
genetikus hiány;

Tetrahydrobiopterin (THB)
kofaktor elégtelenség

(trp hidroxiláz, NOS)

Fenilalanin
hidroxiláz

Több

Reakció

Transzamináz

Fenilalanin

Fenilpiruvát

(fenilketon)

THB

Hiány

Phenylketonuria

Tirozin

melanin

Tyr hidroxiláz

DOPA

dopamin

transzamináz, dyoxigenáz

Fumarát + Acetoacetát

Fenilalanin és fenilpiruvát

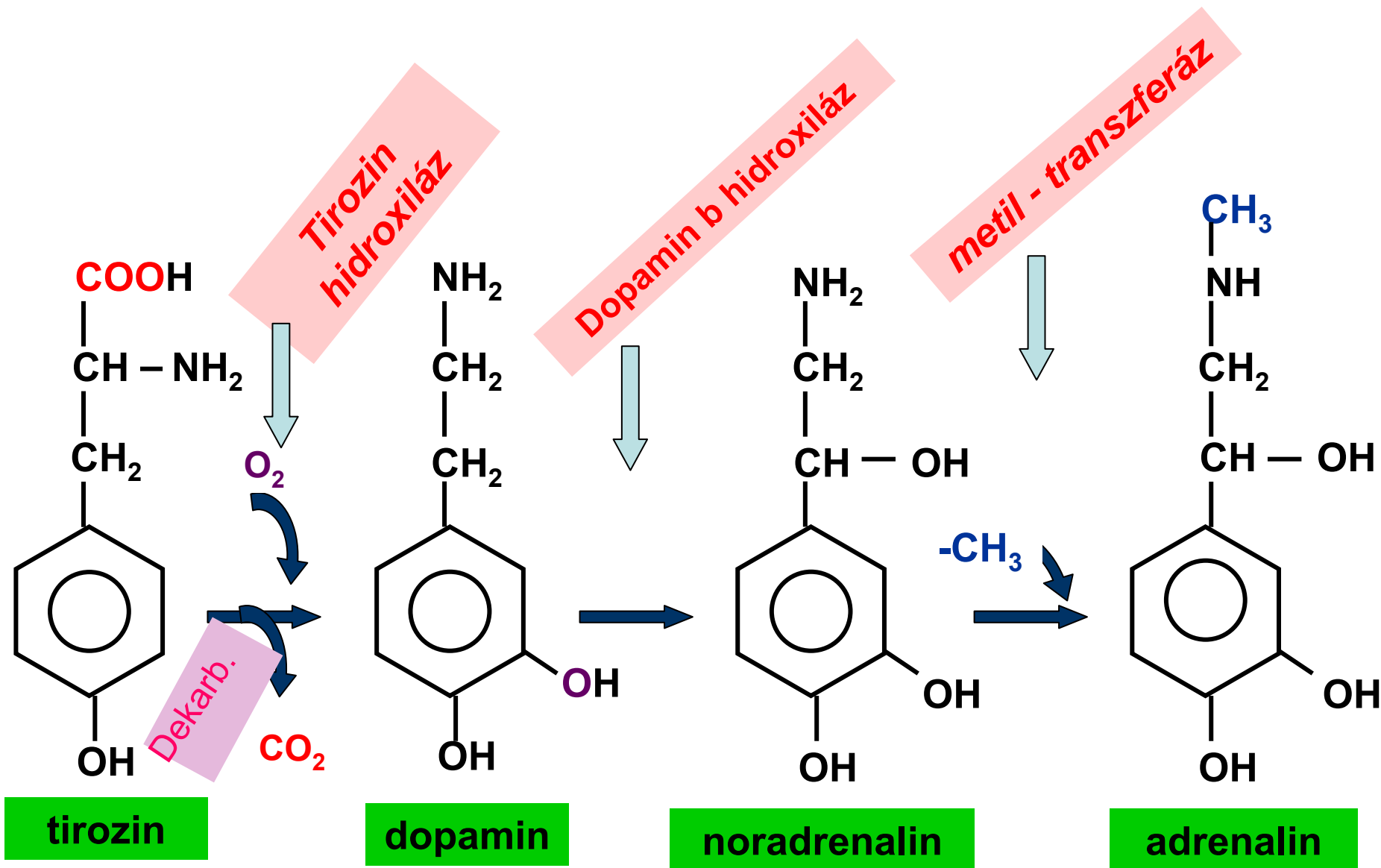
felhalmozódik a vérben, vizeletben, károsítja a myelint az idegsejtekben.

1:9 000 gyakoriság.

Mentális retardáció keletkezik, ha a kezelés nem kezdődik el közvetlenül a születés után. A kezelés során csökkenteni kell a **fenilalanin felvételt, a növekedéshez szükséges szintig.**

Fenilketonuriában a diétának feltétlenül tartalmaznia kell tirozint.

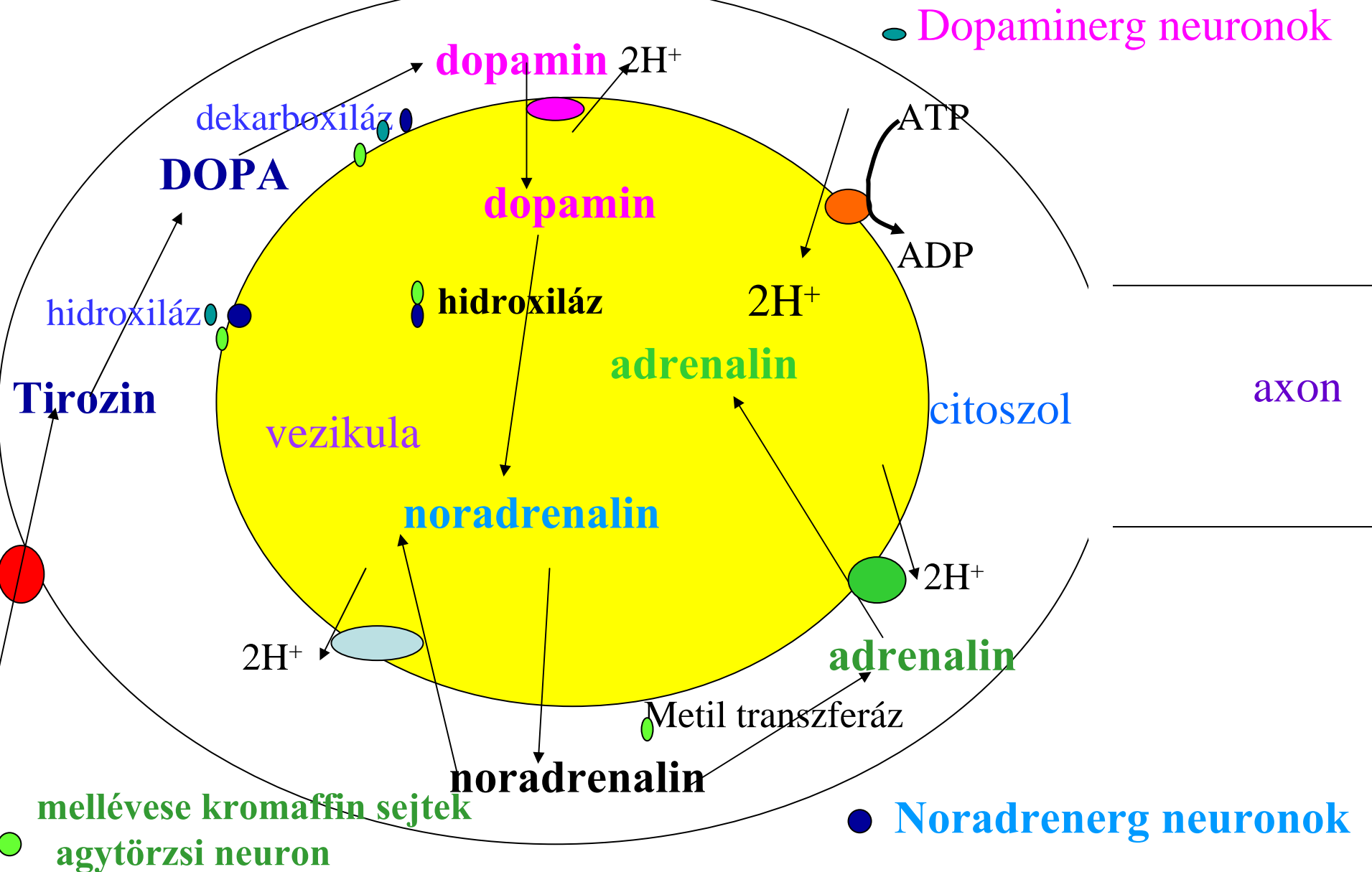
Katecholaminok



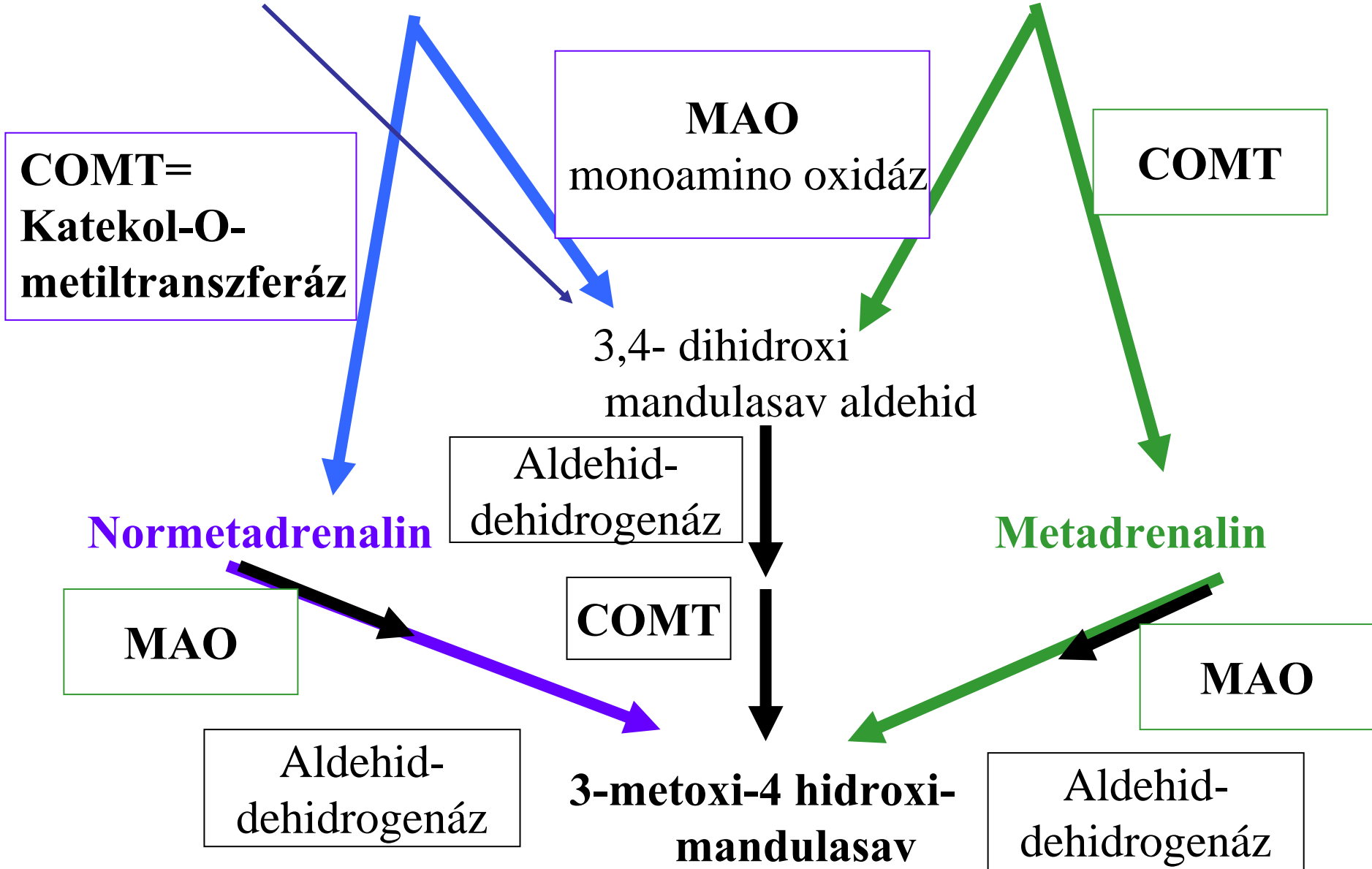
- **Tirozin hidroxiláz:** sebesség meghatározó, kevert funkciójú oxigenáz, kofaktora biopterin, Ca/calmodulin, tartós neurális aktivitás →enzimindukció ↑
- **DOPA dekarboxiláz:** alacsony K_m , nagy V_{max} . (nem specifikus) aromás dekarboxiláz
- Dopamin β hidroxiláz : kevert funkciójú oxigenáz , Cu aszkorbát, tartós neurális aktivitás →enzimindukció ↑
- Feniletanolamin metiltranszferáz: Metil donor S-adenozil metionin, kortikoszteroidok →enzimindukció ↑

Dopamin, noradrenalin, adrenalin

szintézis

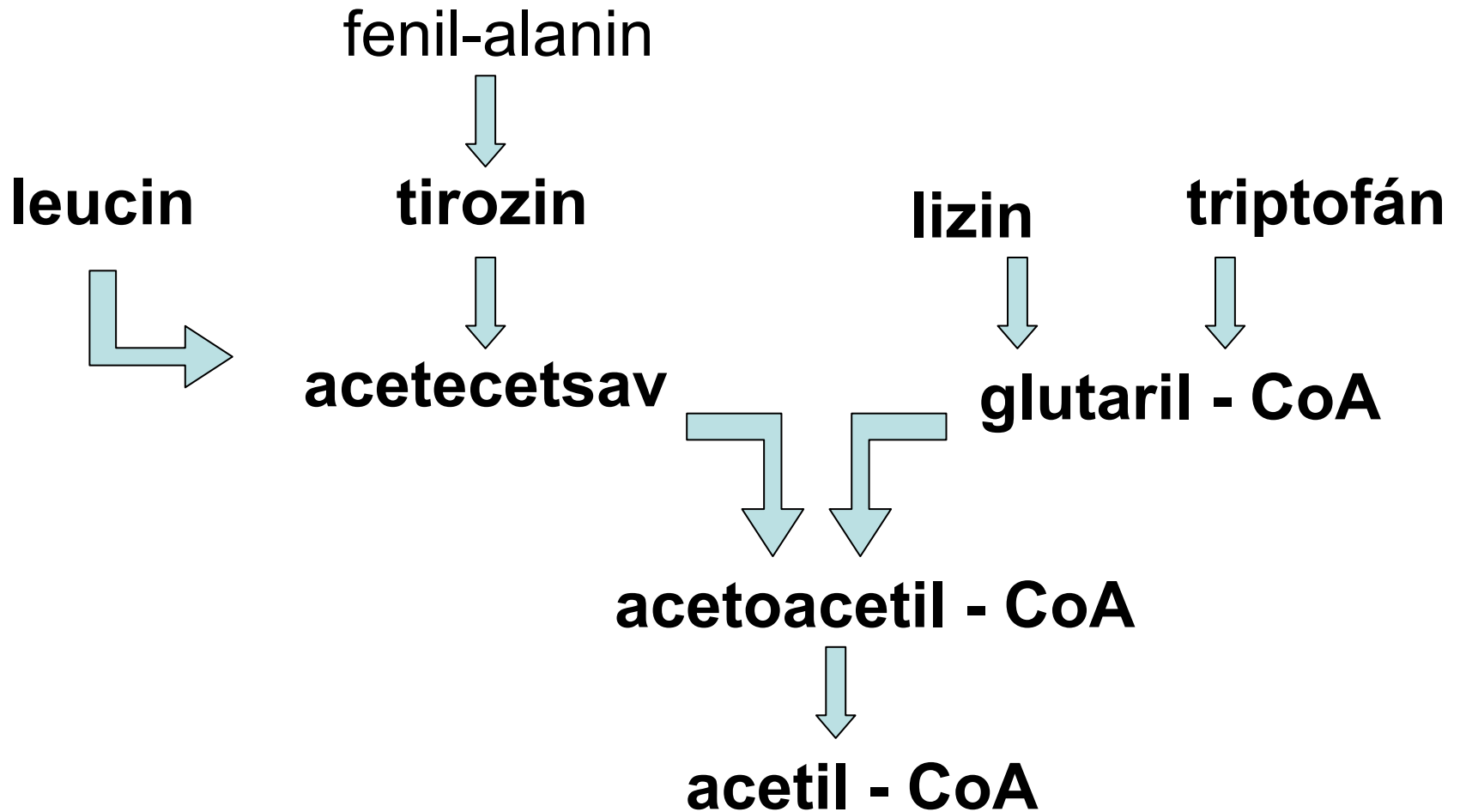


Dopamin → noradrenalin → adrenalin lebontás



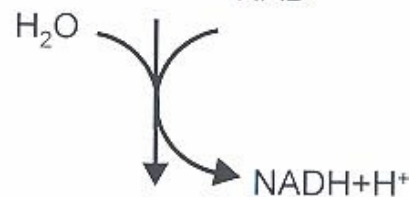
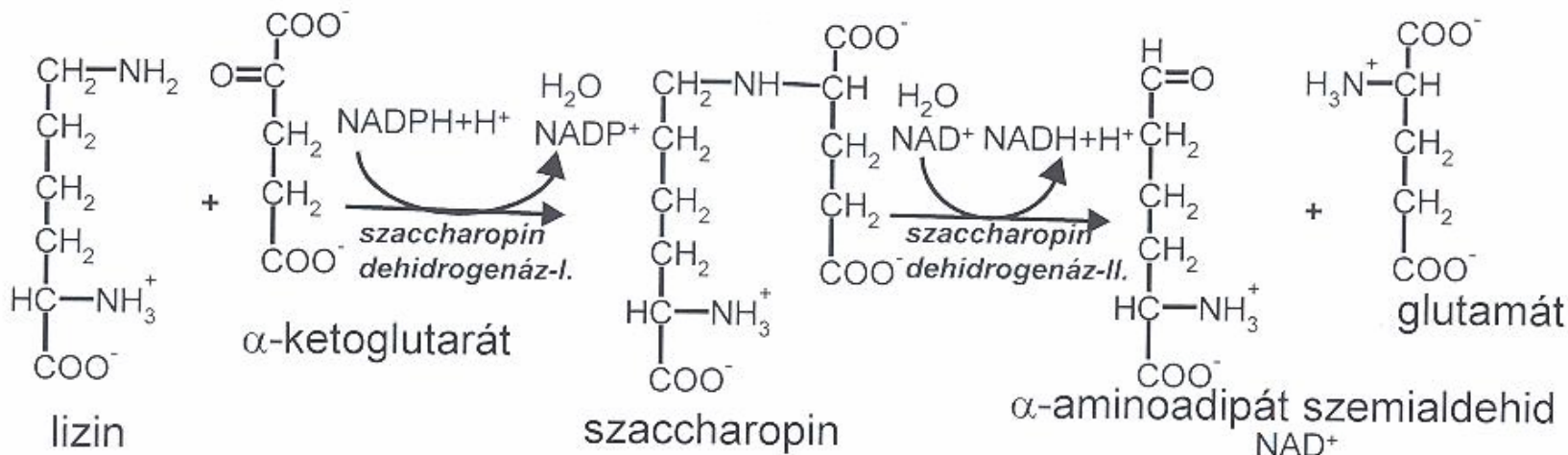
2. Acetoacetyl-CoA csoport lebontása

(fenil-alanin és tirozin lebontása, katecholaminok szintézise)



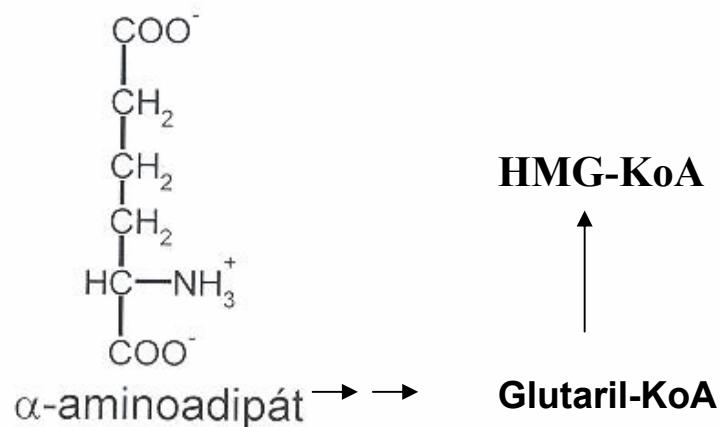
Lizin lebontása, karnitin szintézise:

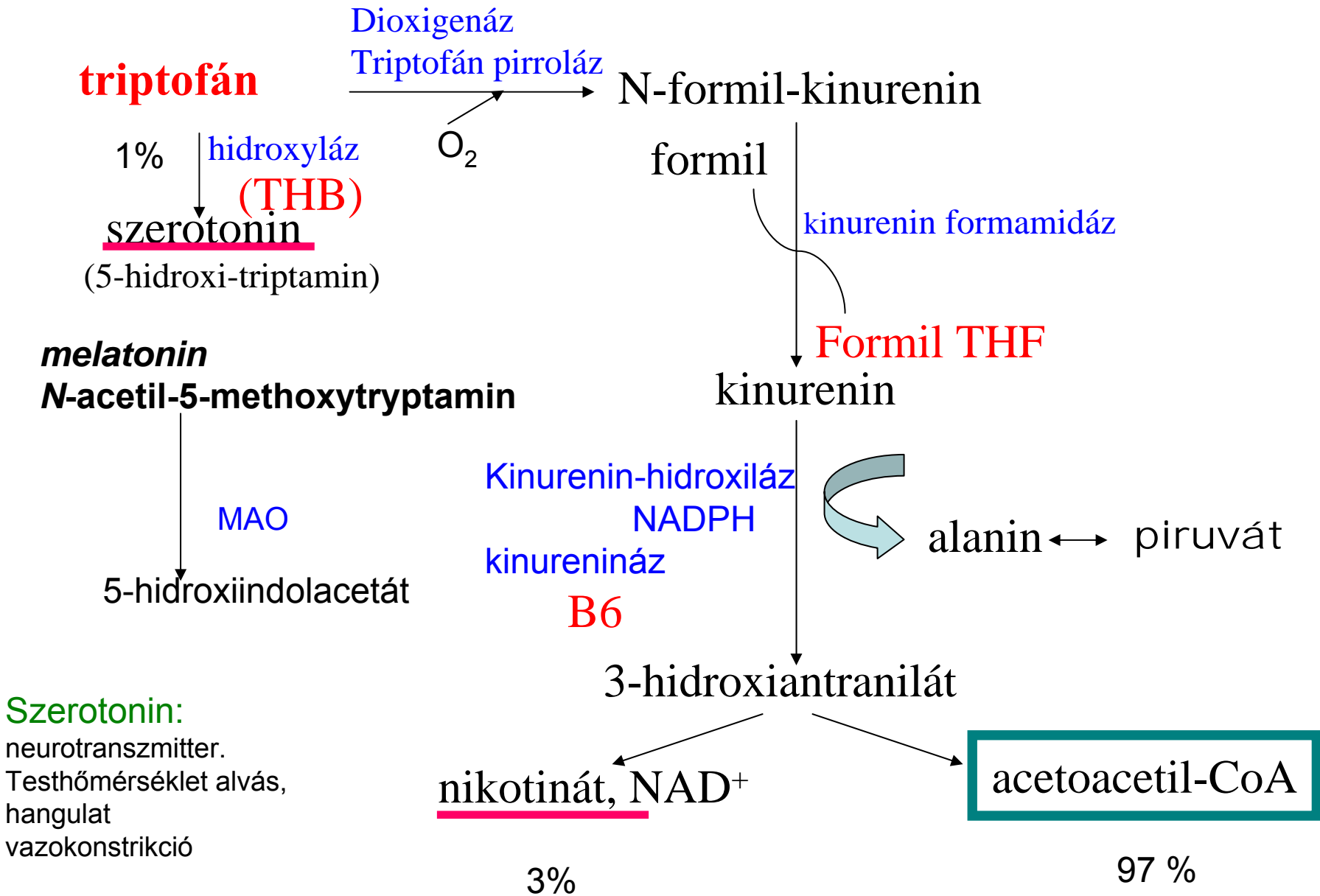
Lizinre nincs transzamináz. A lizin lebontása elsősorban a szaccharopin útvonalon történik. Az útvonal végterméke ugyanaz, mint amit ϵ -transzaminálási reakcióban kapnánk.



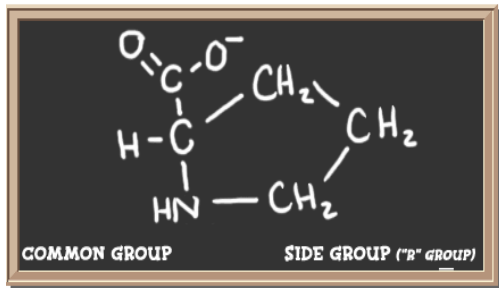
Képződik még:
VAL, Ile. Leu
lebontásban

Sorsa lehet:
Transzaminálás
Oxidatív dekarboxilezés
dekarboxilálás
FAD függő dehidrogenálás
Víz addíció
NAD függő dehidrogenálás

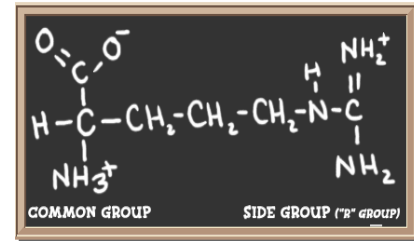




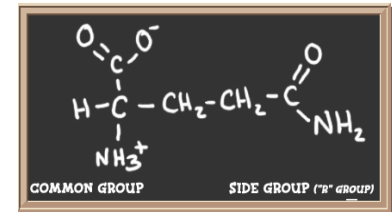
3. α - keto-glutársav csoport lebontása



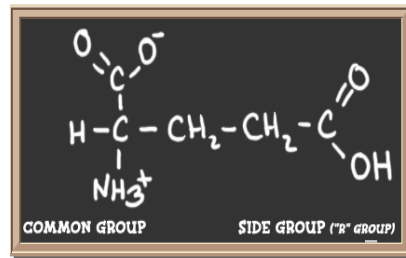
prolin



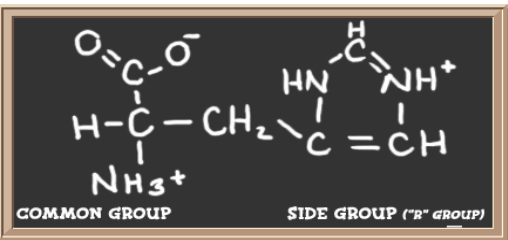
arginin



glutamin

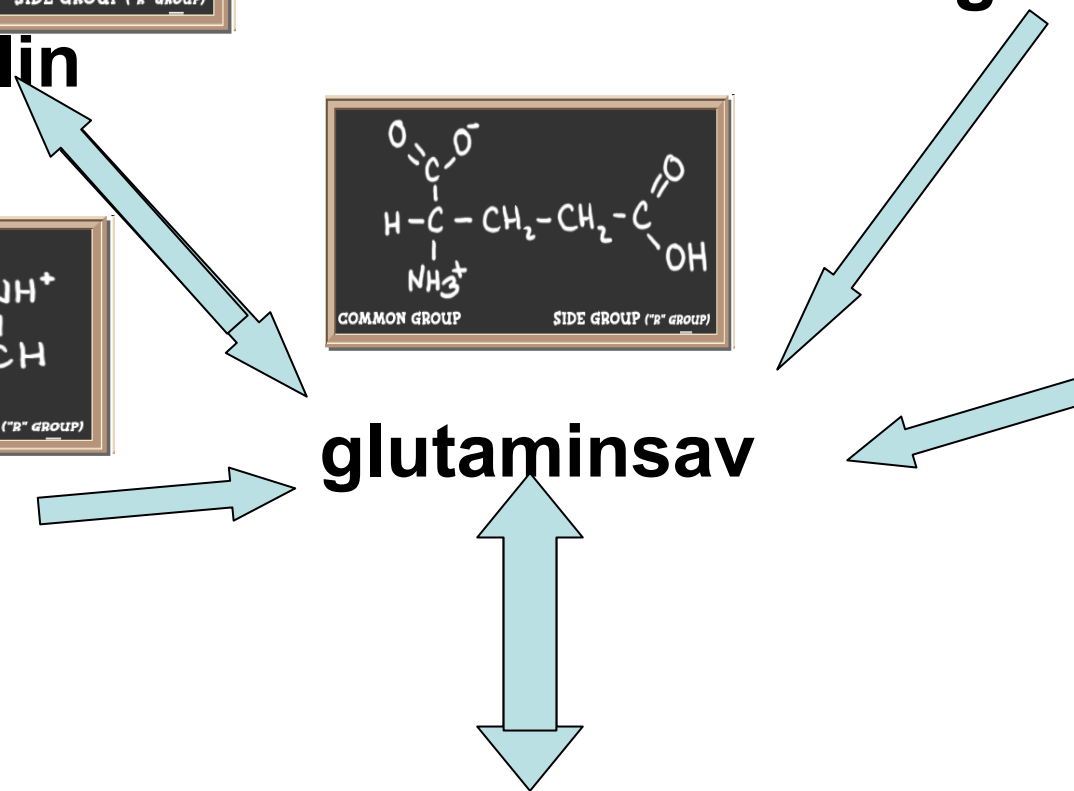


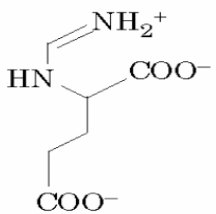
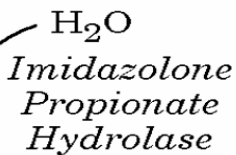
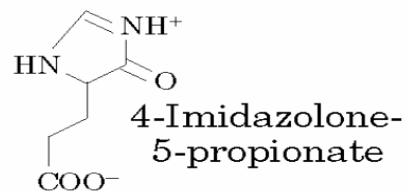
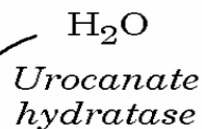
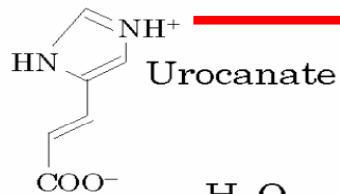
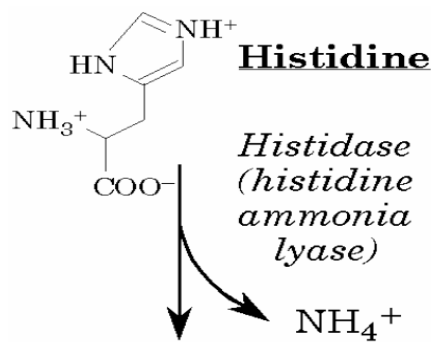
glutaminsav



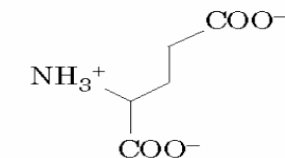
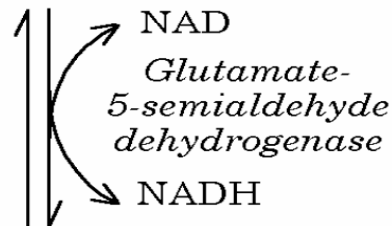
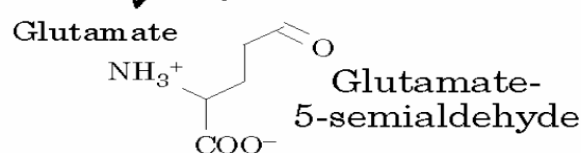
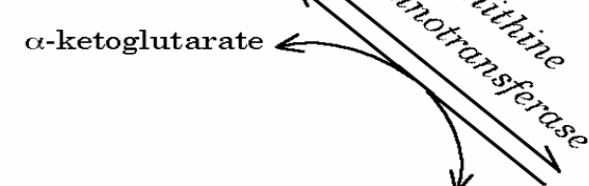
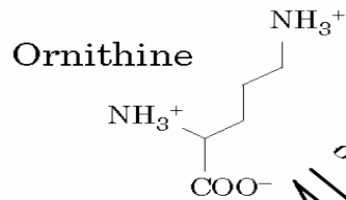
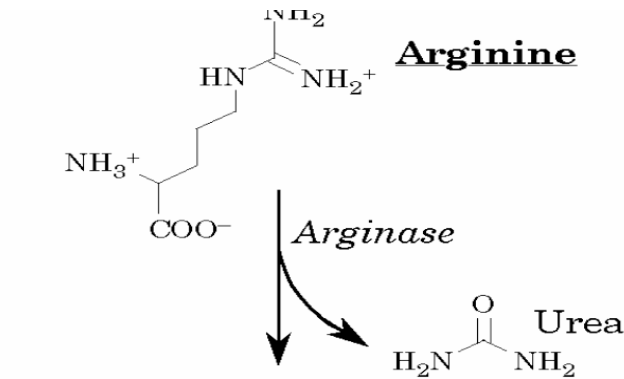
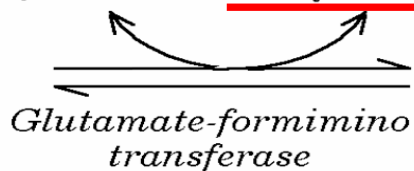
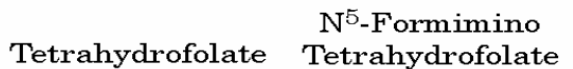
hisztidin

α -keto-glutársav

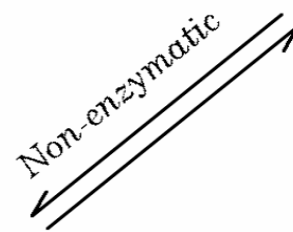
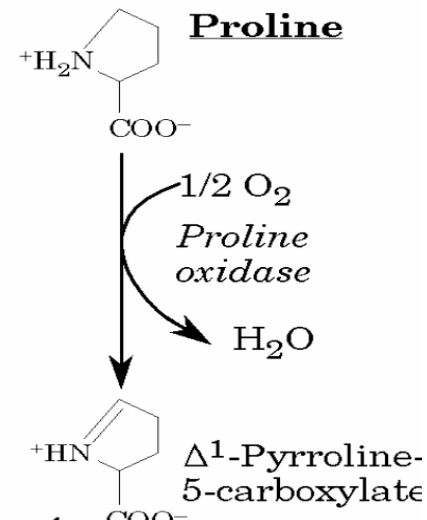




N-Formimino glutamate

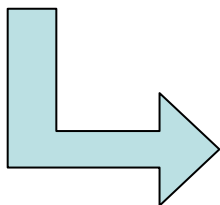


Glutamate

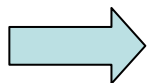


Szukcinil-CoA csoport lebontása

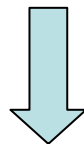
treonin



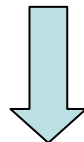
valin



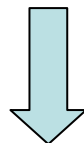
metionin



propionil - CoA

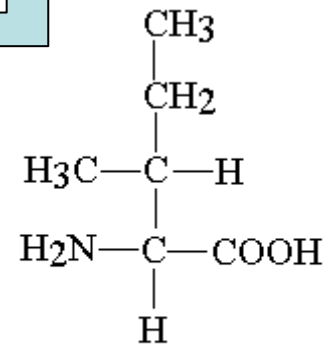
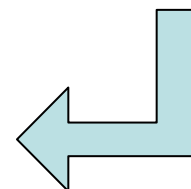


metilmalonil - CoA

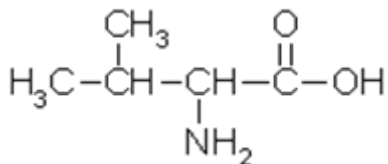


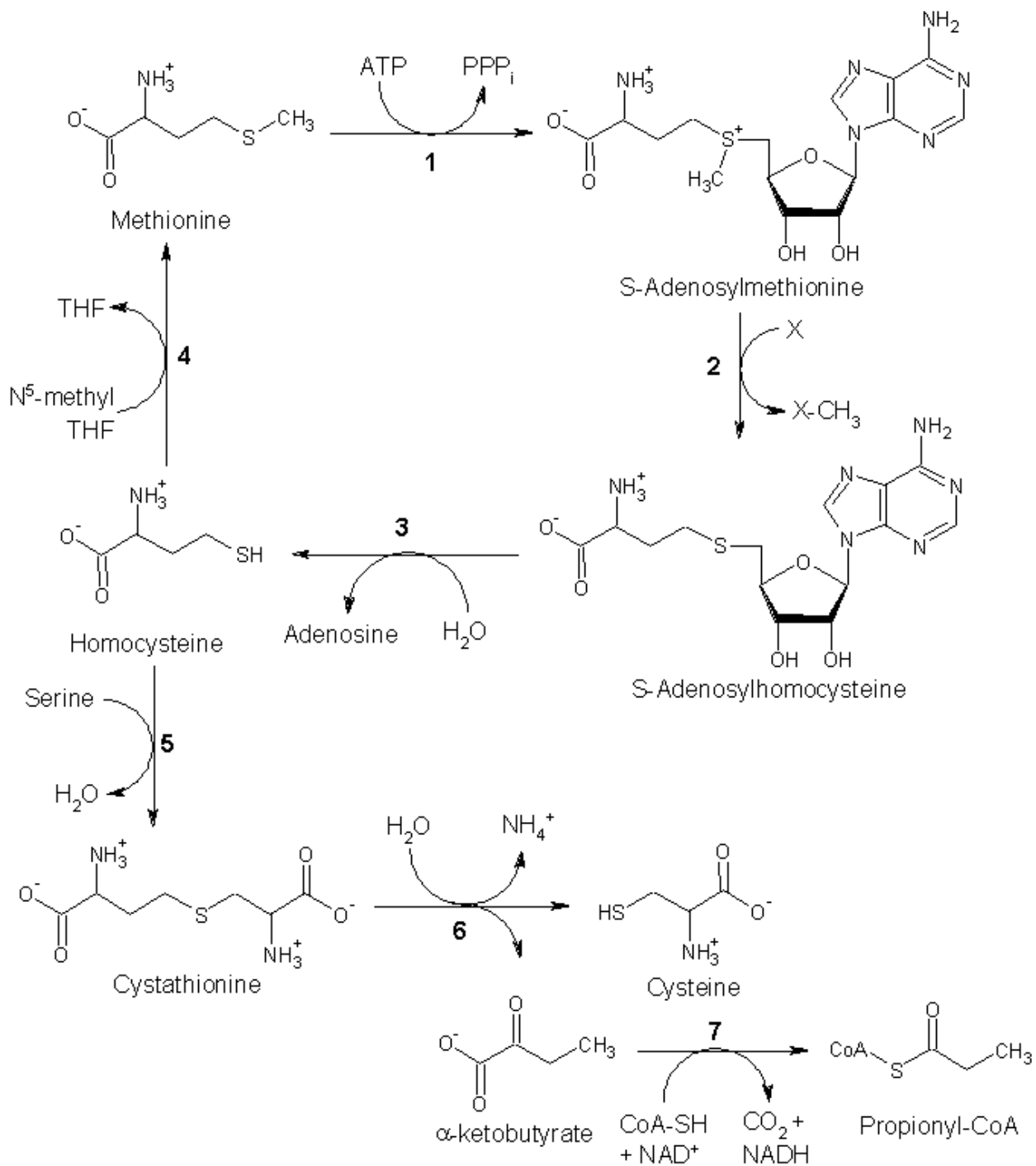
szukcinil - CoA

izoleucin

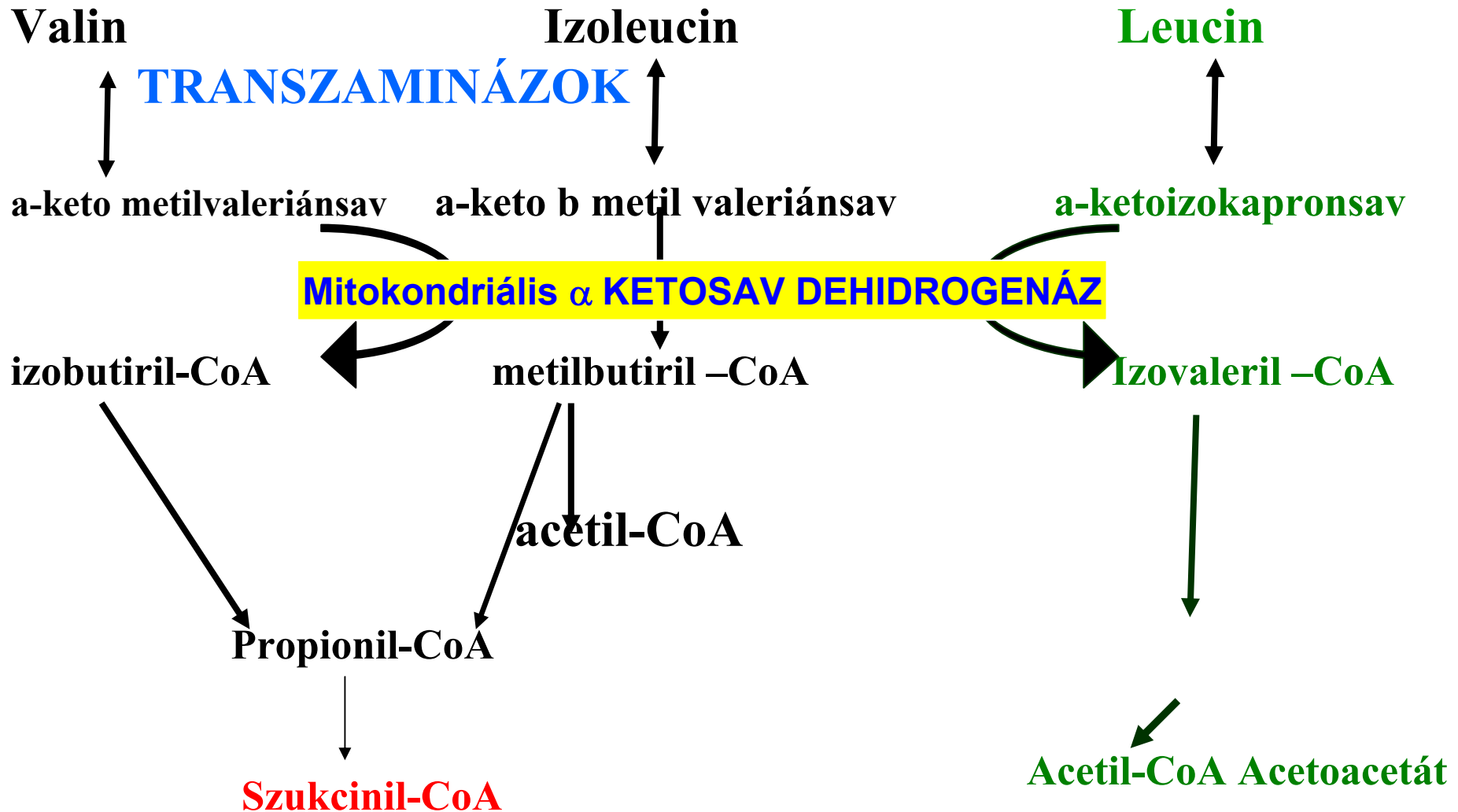


isoleucine

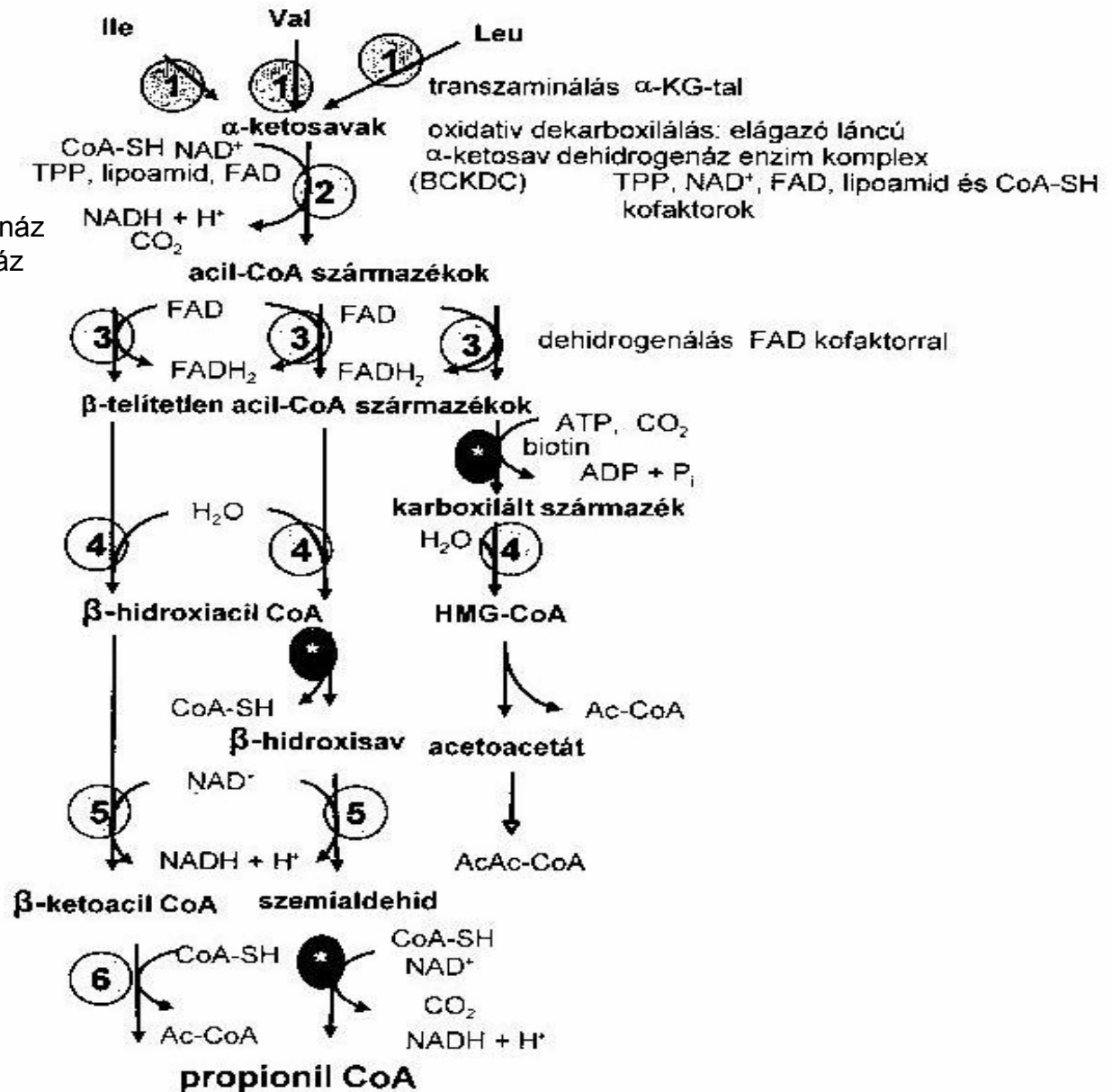




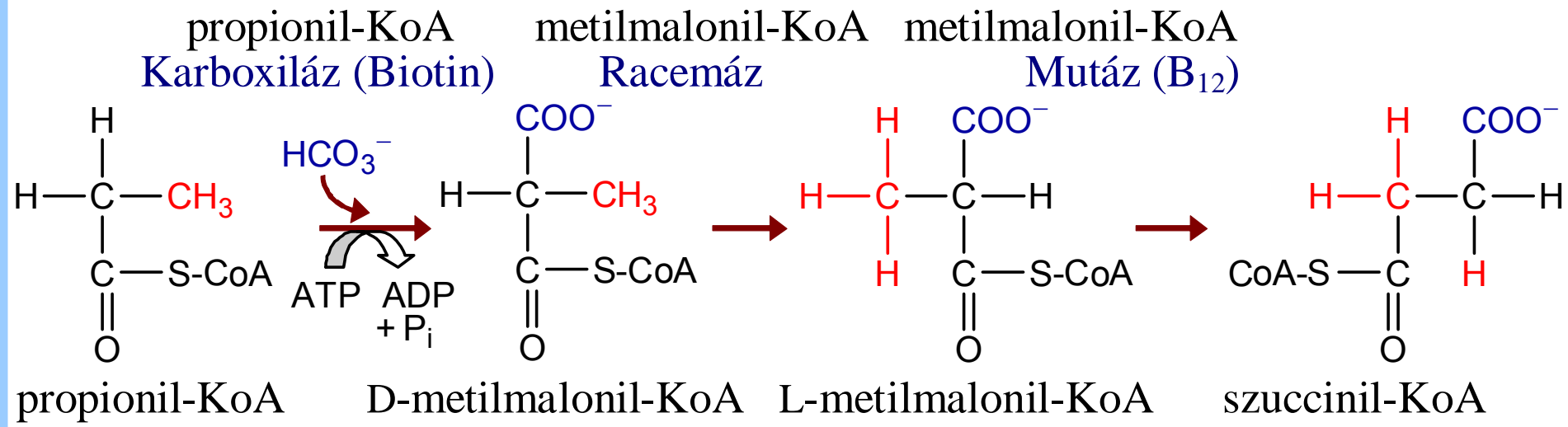
Az elágazó szénláncú aminosavak lebontása



A Val, Ile és Leu lebontásának vázlatja



1. Elágazó szénláncú aminosav transzamináz
2. Elágazó szénláncú ketosav dehidrogenáz
3. Dehidrogenáz
4. Hidratáz
5. Dehidrogenáz
6. Tioláz



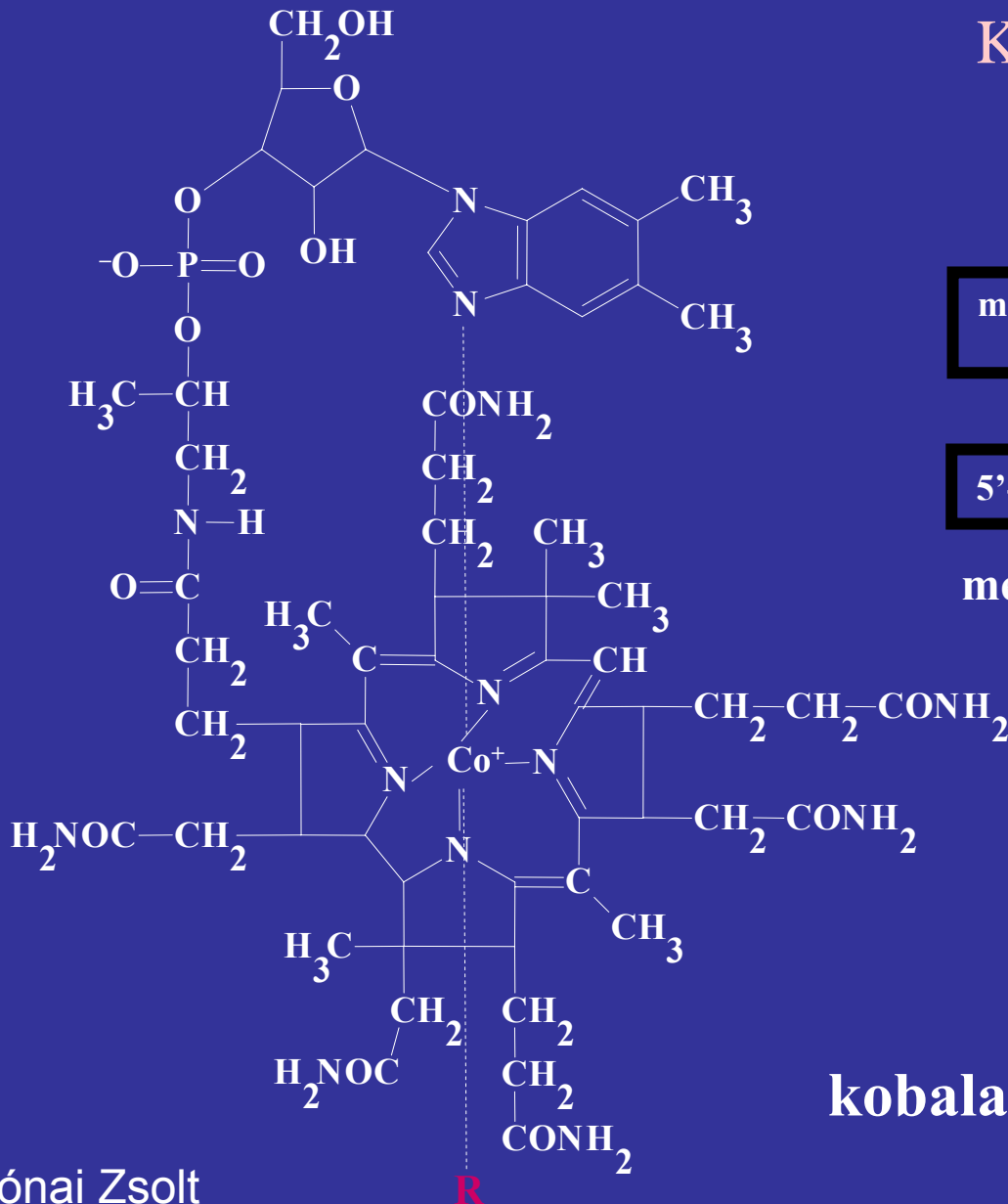
Propionil-KoA - metilmaloniil-KoA-vá alakul

Racemáz - L-izomer.

Metilmaloniil-KoA Mutáz – molekuláris átrendeződés:
 metilmaloniil-KoA elágazó lánca szukcinil-KoA lineáris lánca.

Koenzim B12 (vitamin B12+ATP, adenzilkobalamin)-
 metilmaloniil-KoA mutáz kofaktora

B₁₂-vitamin



Kobalt 2 axiális liganduma

- dimetil-benzimidazol
- felszívódás: -OH

mitokondrium

citoplazma

5'-deoxiadenozil

metil-malonil-koA mutáz

-CH₃
homoCys-metil-transzferáz

korrin váz

4 pirrol gyűrű

kobalamin

B12 vitamin szerepe

- homocisztein metil-transzferáz
- metil malonil coenzym-A mutáz (adenozilkobalamin)
- B12 vitamin hiány: **“folát csapda”** felszaporodik a THF legredukáltabb formája
- nukleotid szintézis gátlás
- sejtproliferáció gátlás
- megaloblasztos anémia
- B12 és tetrahydrofólsav együttes adása
- B12 vitamin hiány: **homocisztein-metiltranszferáz aktivitás csökkenése** emelkedett homocisztein ateroszklerózis, csökkent metionin idegrendszeri problémák

5. Fumársav csoport

- tirozin, fenilalanin, aszparaginsav

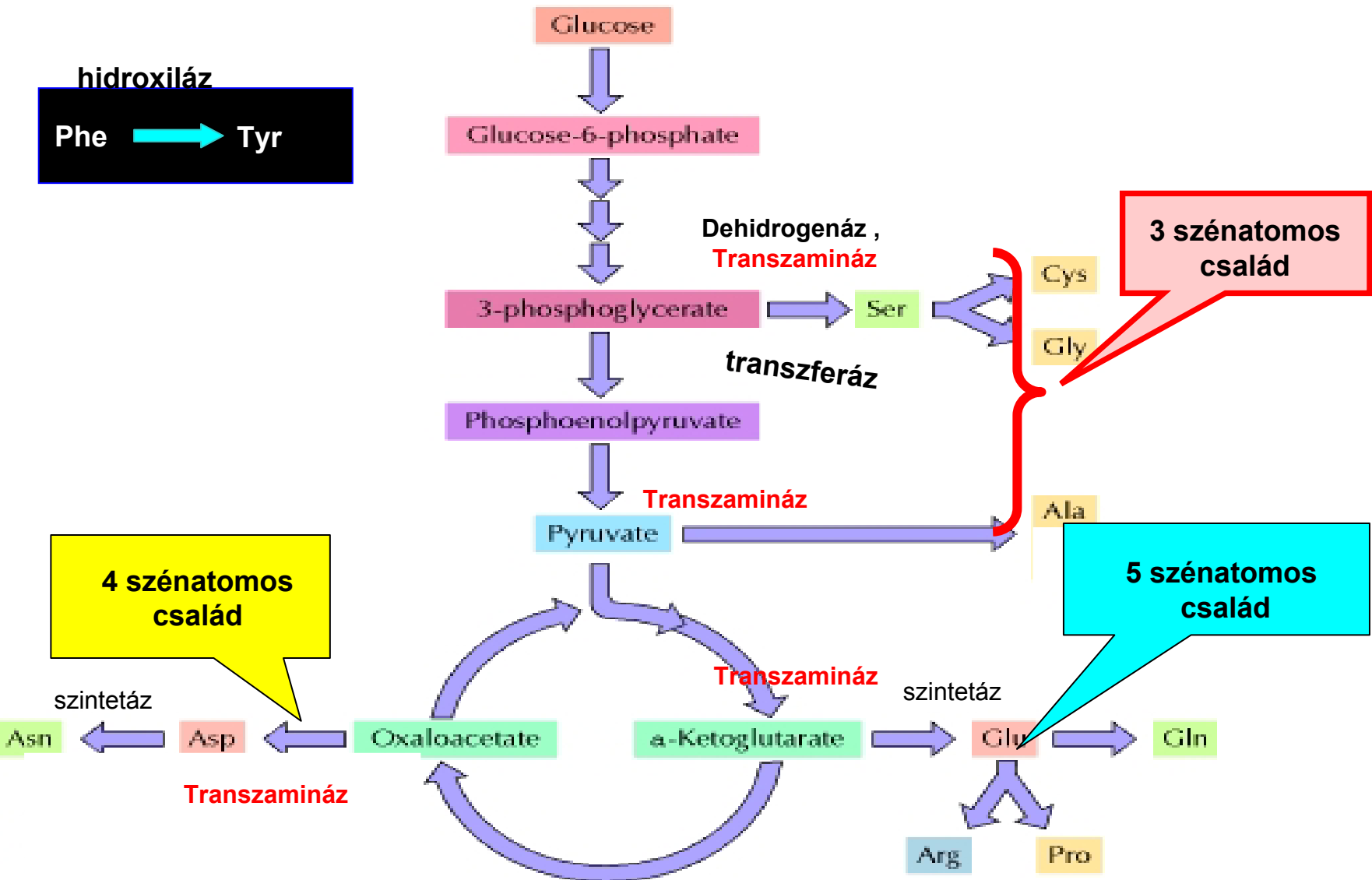
6. Oxálecetsav csoport

- aszparagin, aszparaginsav

Aszparaginsav

- Ornitin ciklus (fumársav)
- Purin nukleotid ciklus (fumársav)
- Transzaminálás (oxálecetsav)

A nem esszenciális aminosavak szintézise

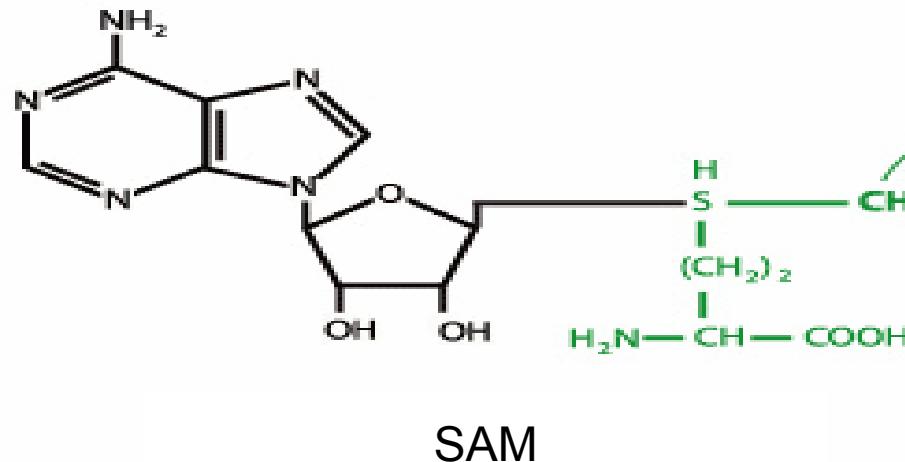
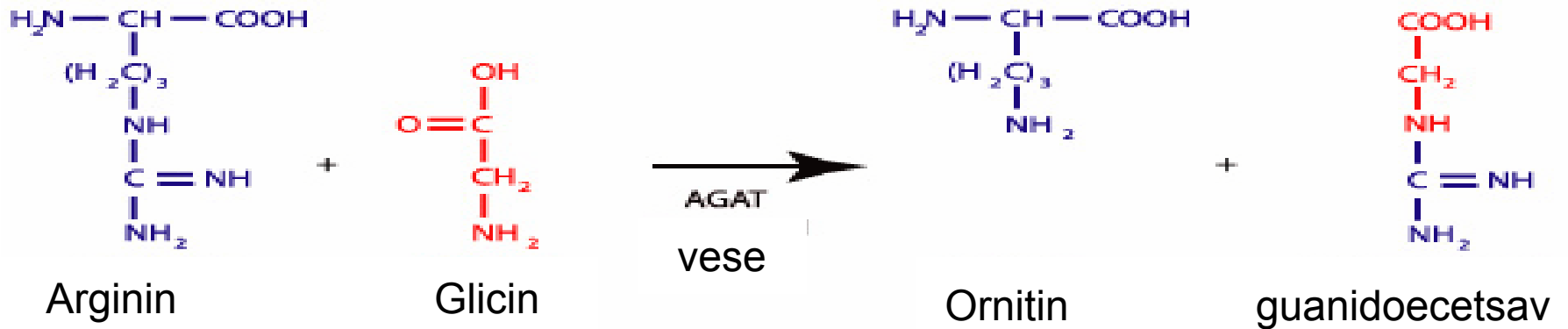


Szintézis enzimei

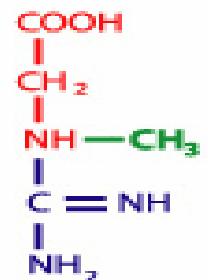
- Transzaminázok: pir **ala**
- ketoglutarát **glu**
- oa **asp**
- glu semialdehid szintetáz, transzamináz : **orn**
ureaciklus enzimei –**arg**
- glu semialdehid szintetáz, dehidratáz, reduktáz **prolin**

- Dehidrogenáz , transzamináz **ser**
- Hidroximetil transzferáz **gly**
- Hidroxiláz **tyr**

Kreatin szintézis



GAMT
máj



KREATIN

AGAT: arginin glicin amidotranszferáz
GAMT: SAM N guanidoacetát metiltranszferáz