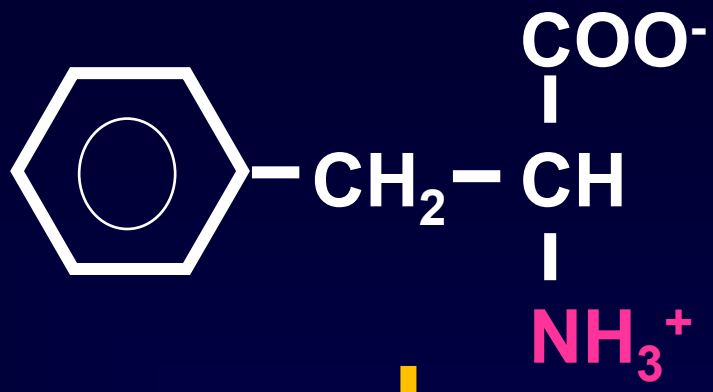
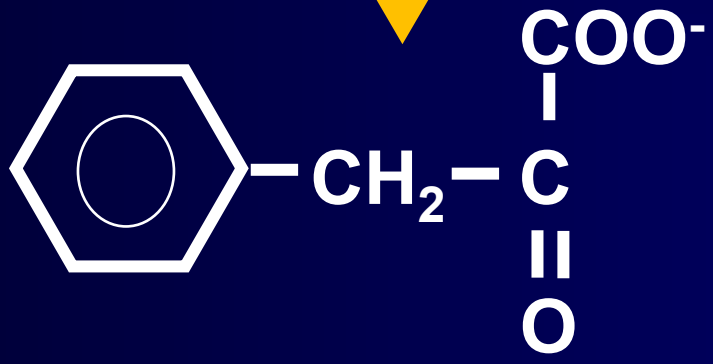
Dr Asbjørn Følling
1888 -1973




Phénylalanine




Phénylpyruvate

University of Sfax
TUNISIA

UNIVERSITÉ DE SFAX

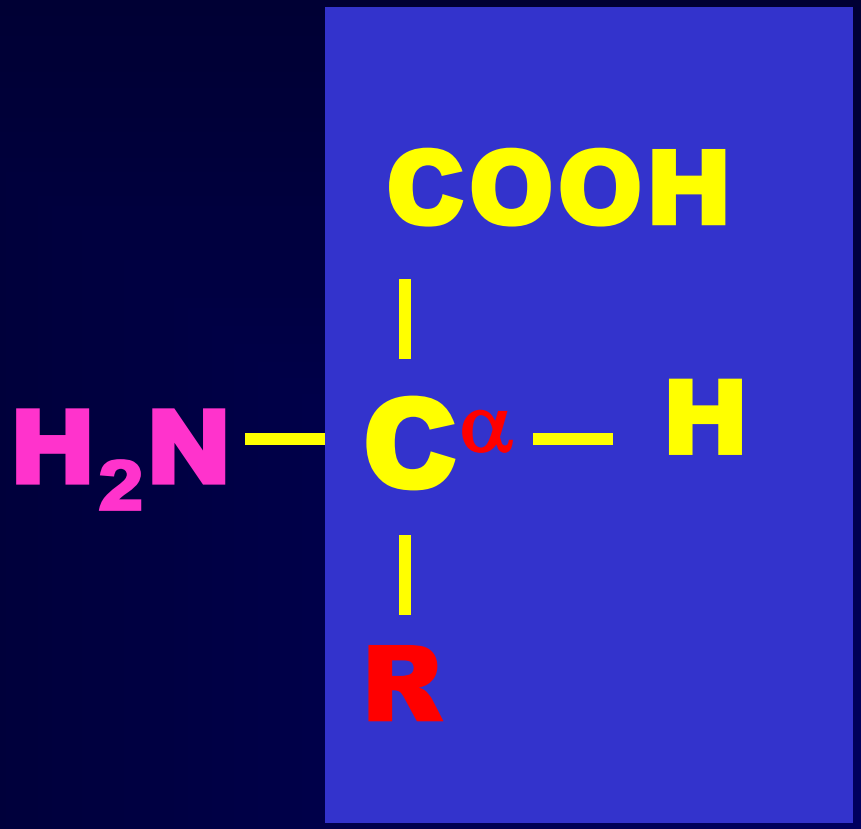
Faculté de Médecine de Sfax

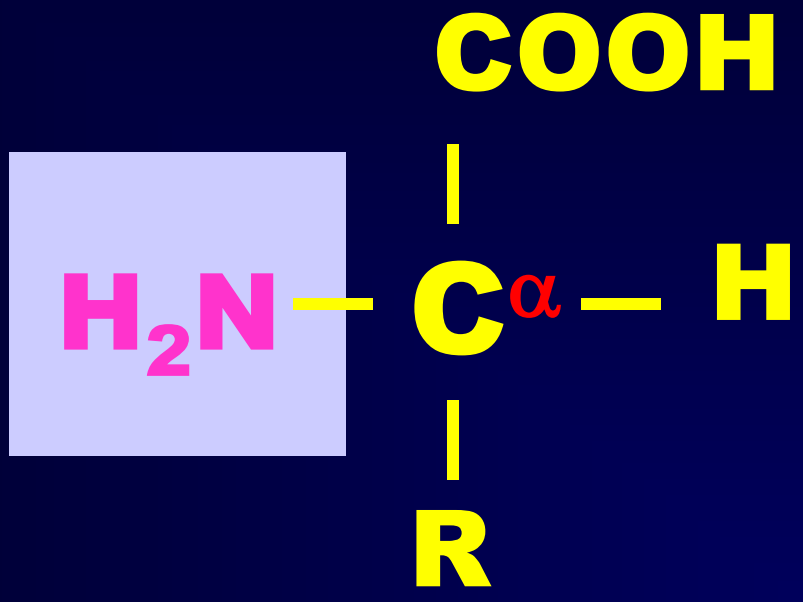
UEF 109

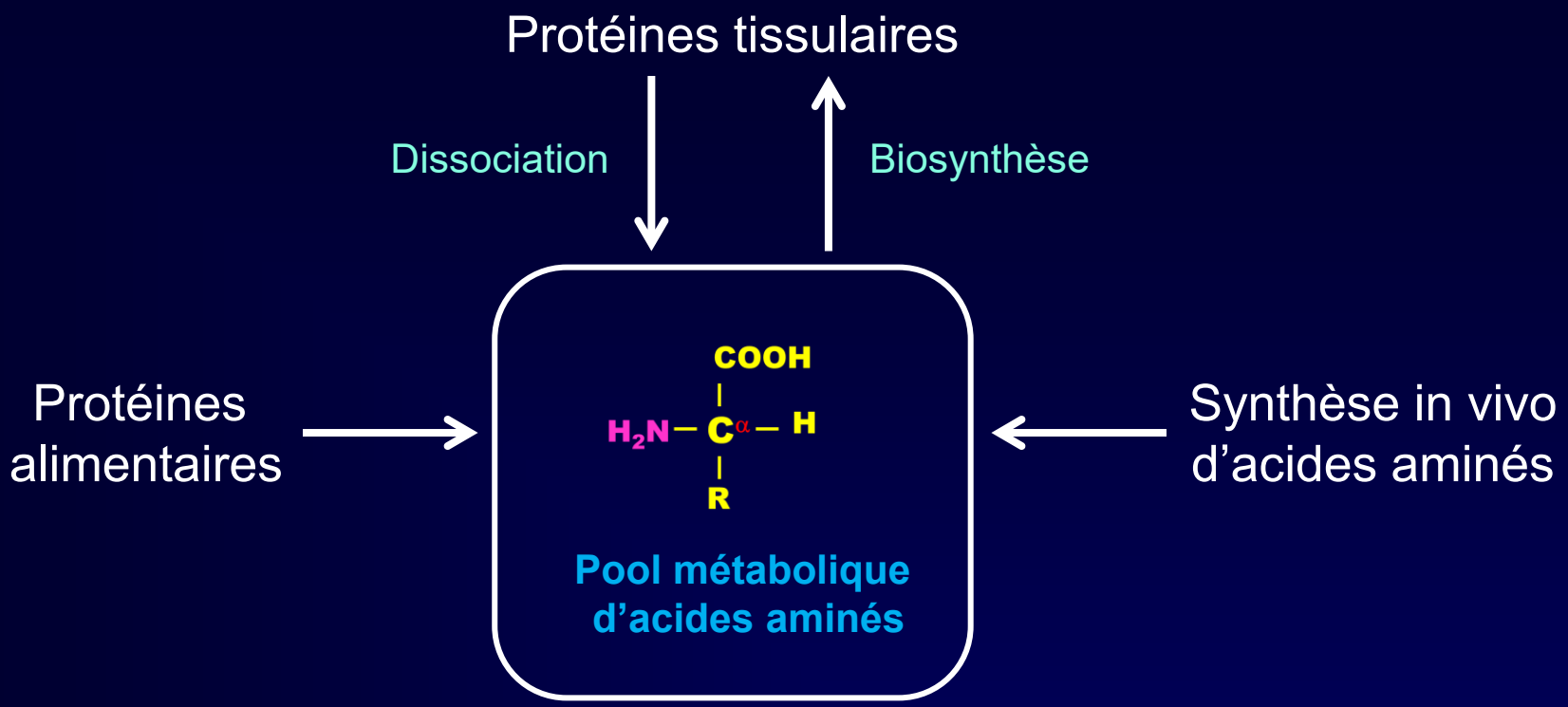
# Métabolisme des Acides Aminés

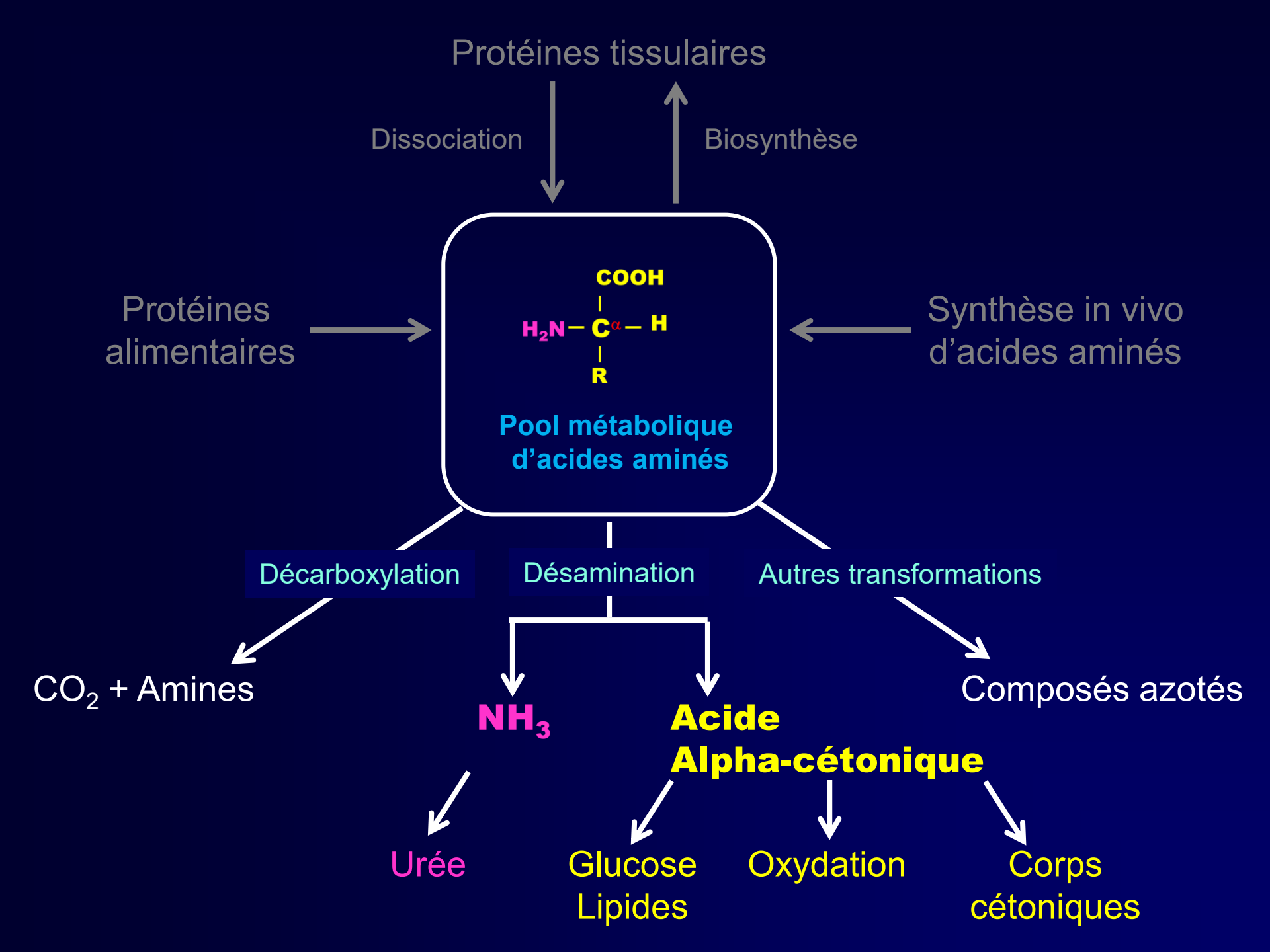
كلية الطب بصفاقس

**Kamel JAMOUSSI (MD, PHU)**
**Section de Biochimie**
**Département des Sciences de base A**

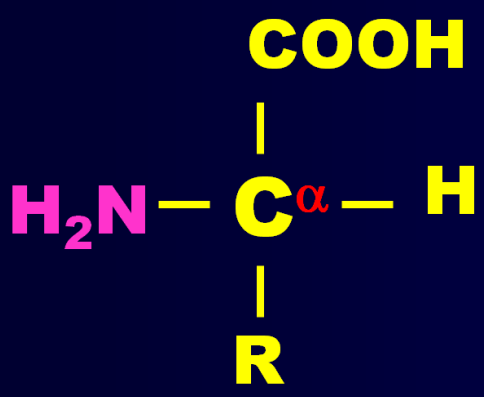

COOH
H2N
Cα
H
R

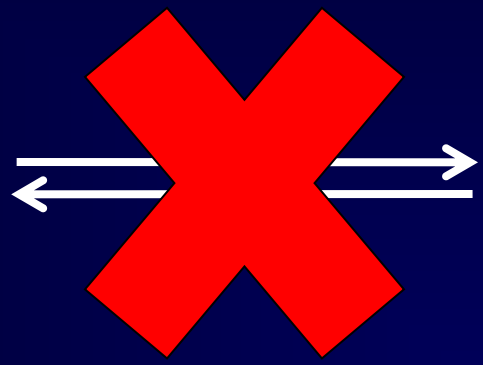

COOH
H2N
Cα
H
R


Protéines tissulaires
Dissociation
Biosynthèse
Protéines alimentaires
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Synthèse in vivo d'acides aminés

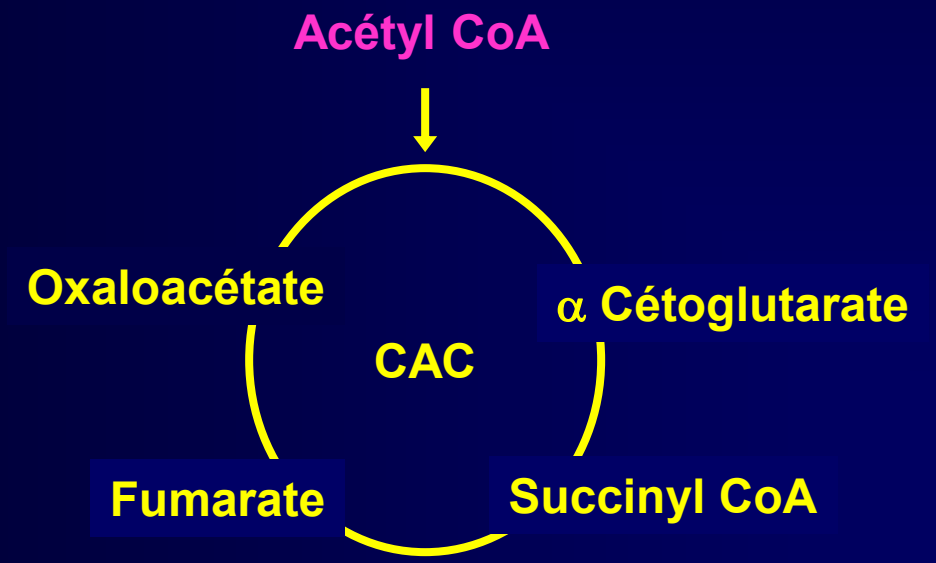

Protéines tissulaires
Dissociation
Biosynthèse
Protéines alimentaires
Synthèse in vivo d'acides aminés
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Décarboxylation
Désamination
Autres transformations
CO2 + Amines
NH3
Acide Alpha-cétonique
Composés azotés
Urée
Glucose Lipides
Oxydation
Corps cétoniques

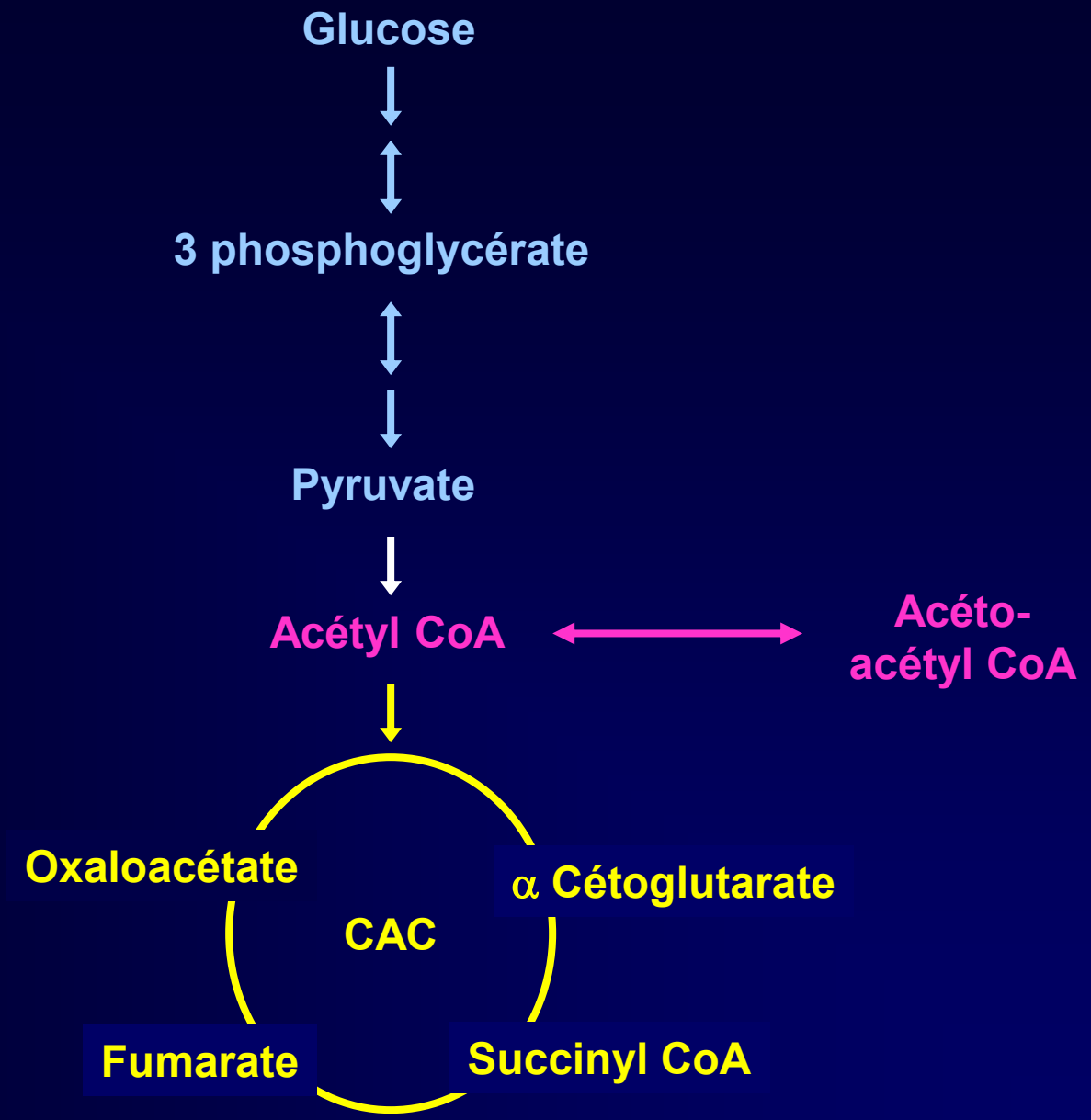

Métabolisme
Anabolisme
Catabolisme

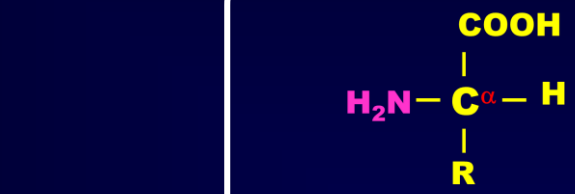


C, O, H, N…


Acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA



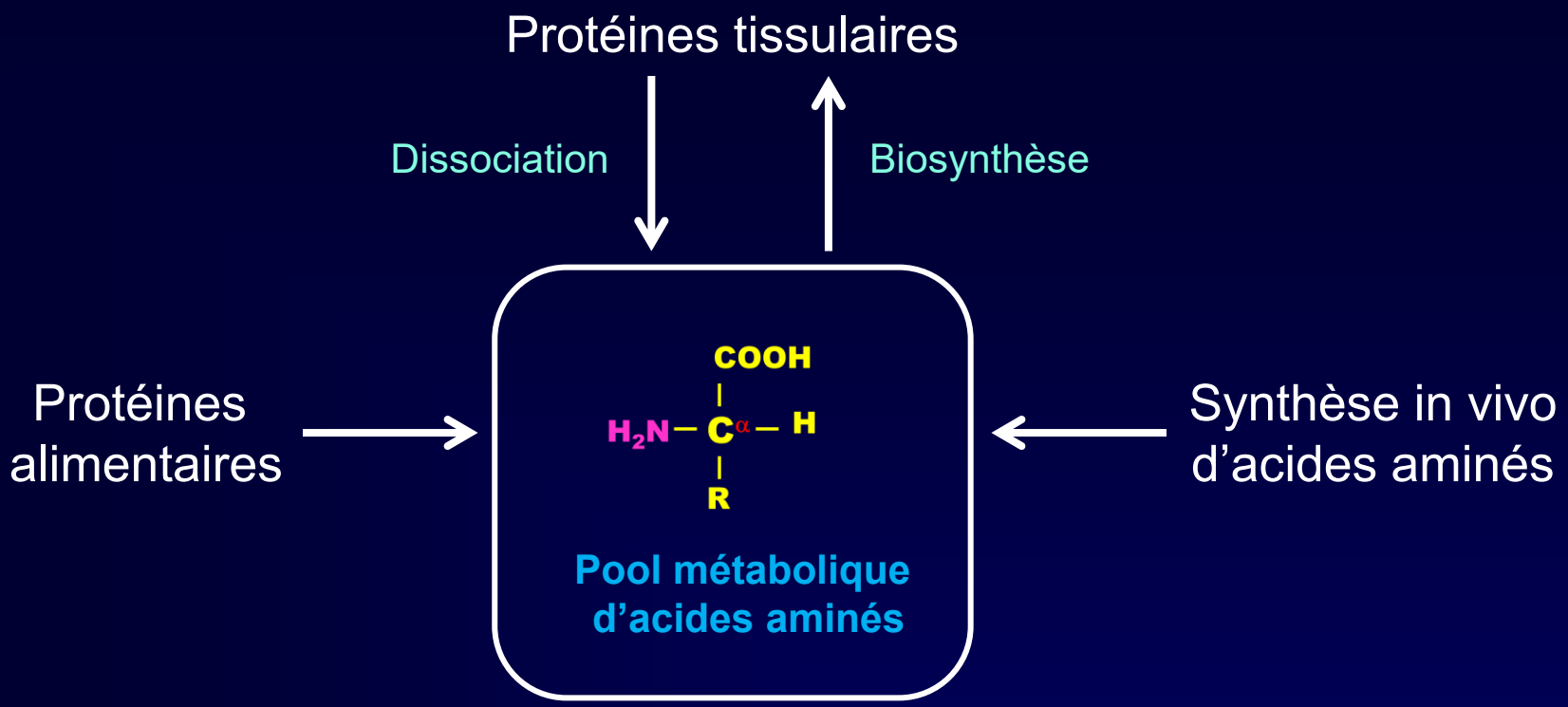

Pool métabolique
d'acides aminés

## AA indispensables

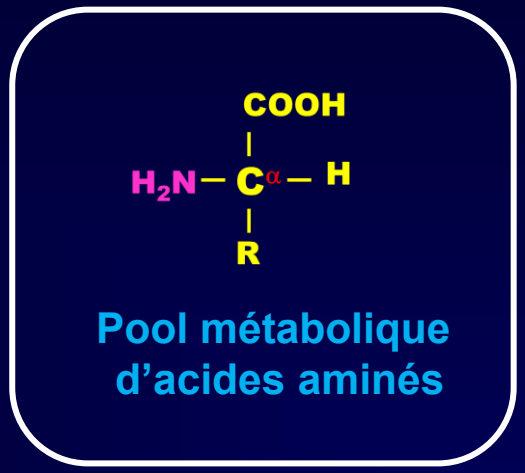
Arg
His
Ile
Leu
Lys
Met
Phe
Thr
Trp
Val

## AA non indispensables

Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln


Protéines tissulaires
Dissociation
Biosynthèse
Protéines alimentaires
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Synthèse in vivo d'acides aminés

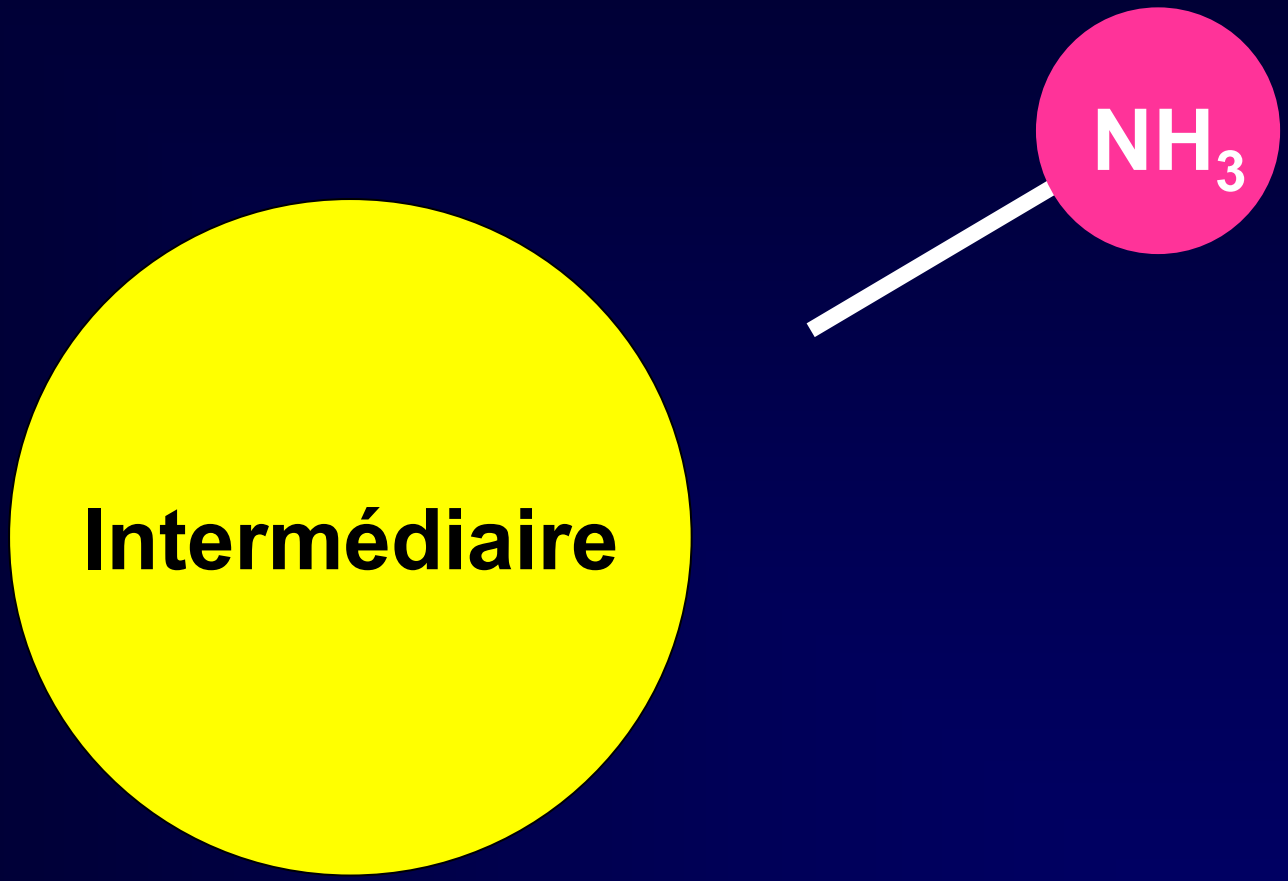
| AA indispensables | AA non indispensables |
| --- | --- |
| Arg | Ala |
| His | Asp |
| Ile | Cys |
| Leu | Glu |
| Lys | Gly |
| Met | Pro |
| Phe | Ser |
| Thr | Tyr |
| Trp | Asn |
| Val | Gln |

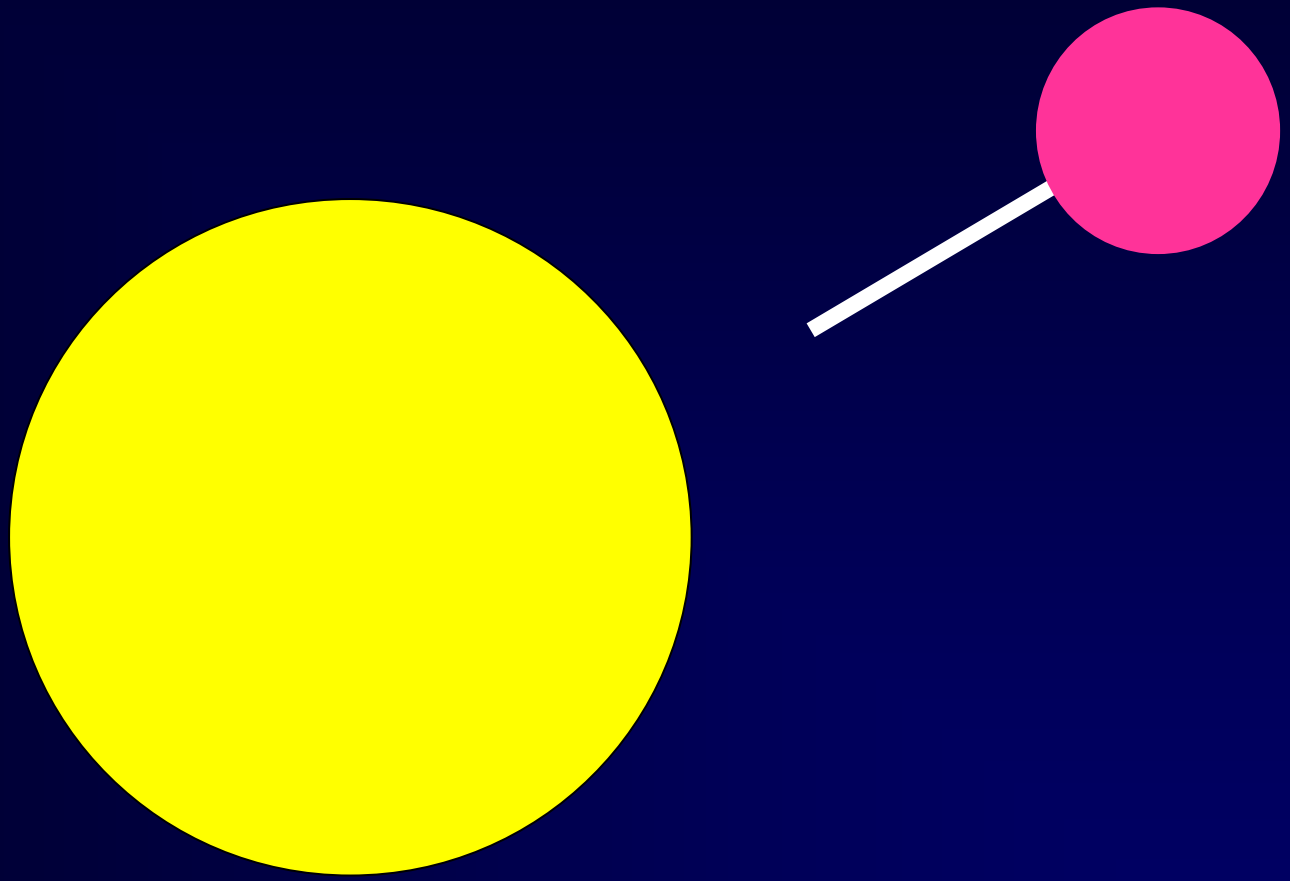


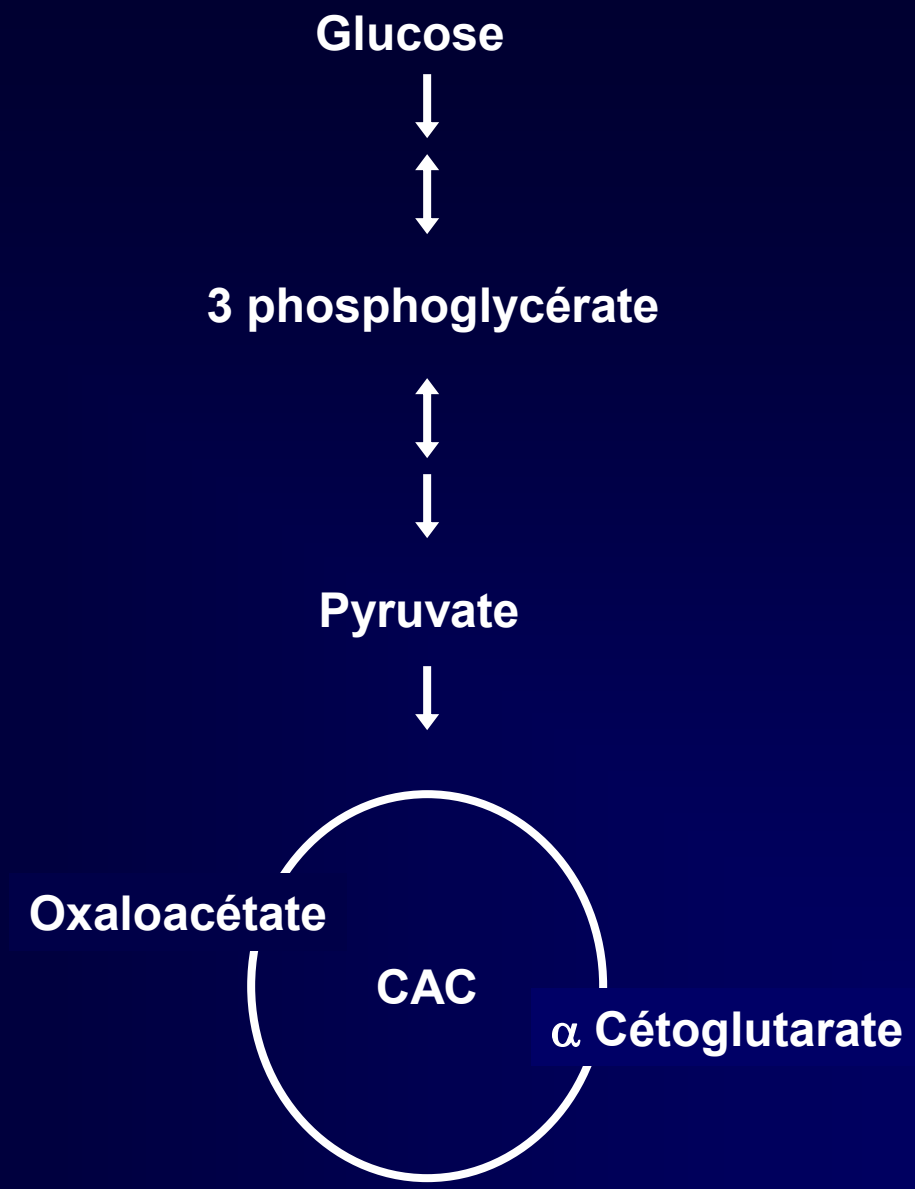

Synthèse in vivo
d'acides aminés

# AA non indispensables

Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln


$NH_3$
Intermédiaire

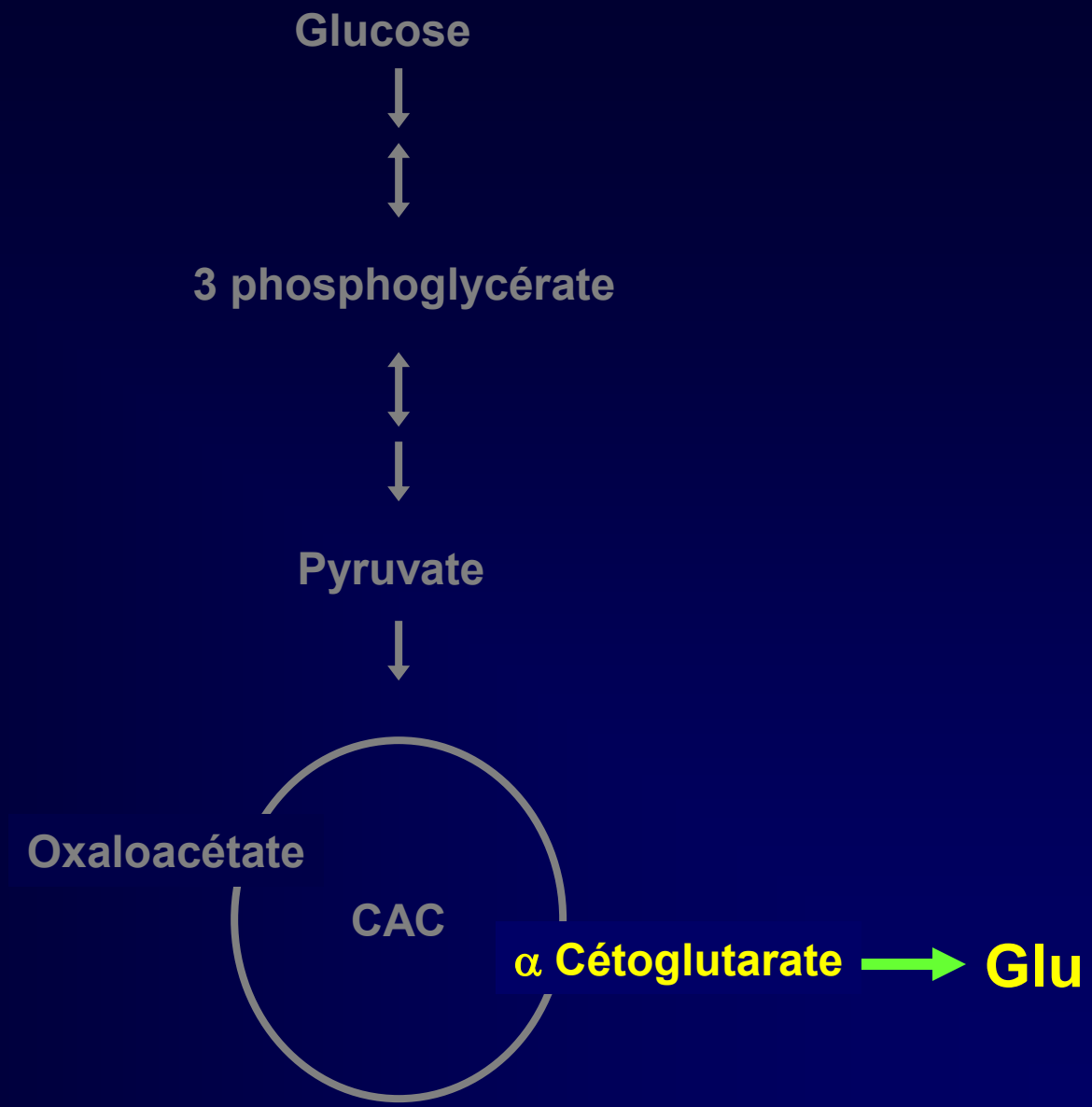


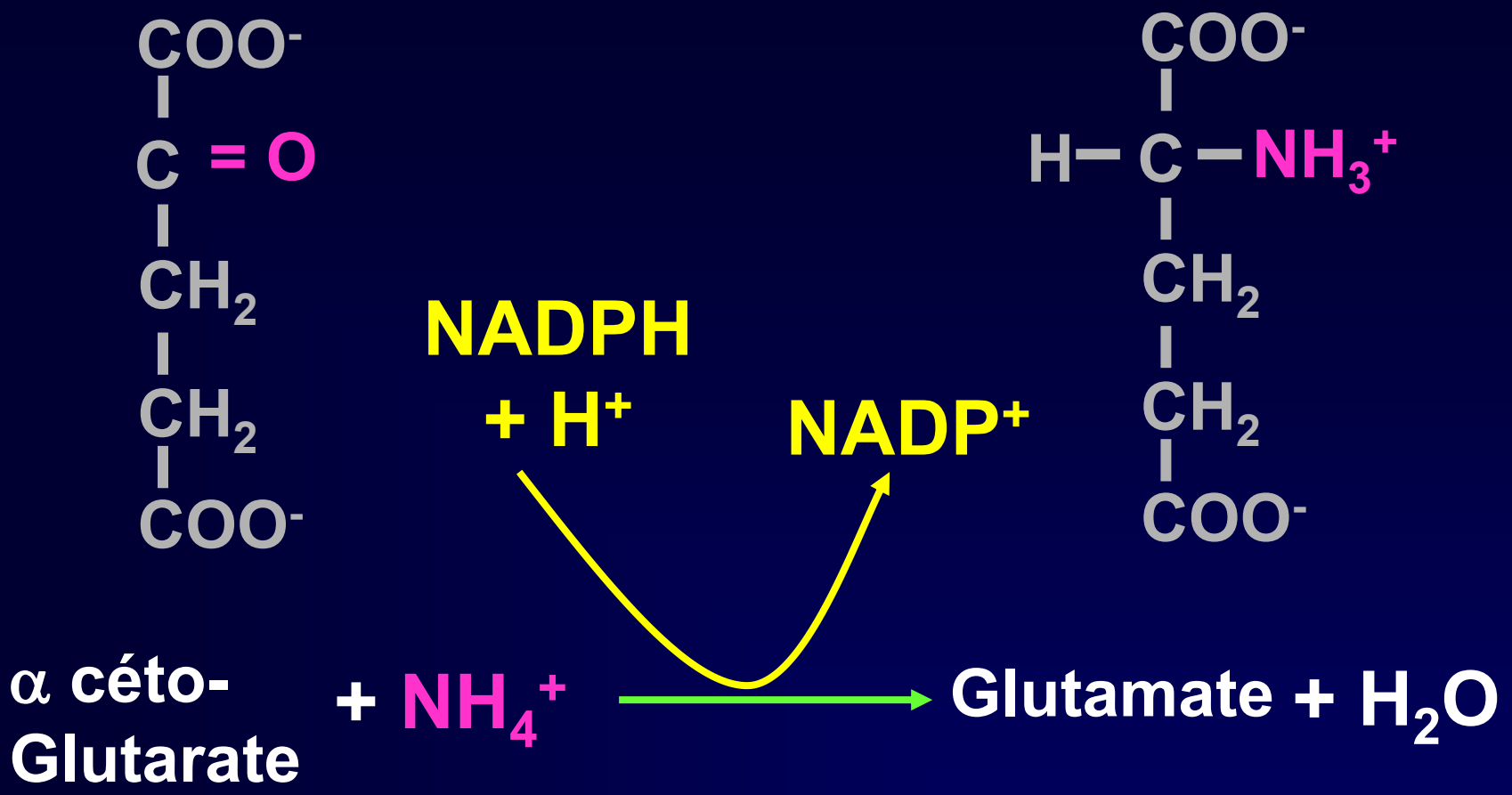

Glucose
3 phosphoglycérate
Pyruvate
Oxaloacétate
CAC
α Cétoglutarate

# AA non indispensables

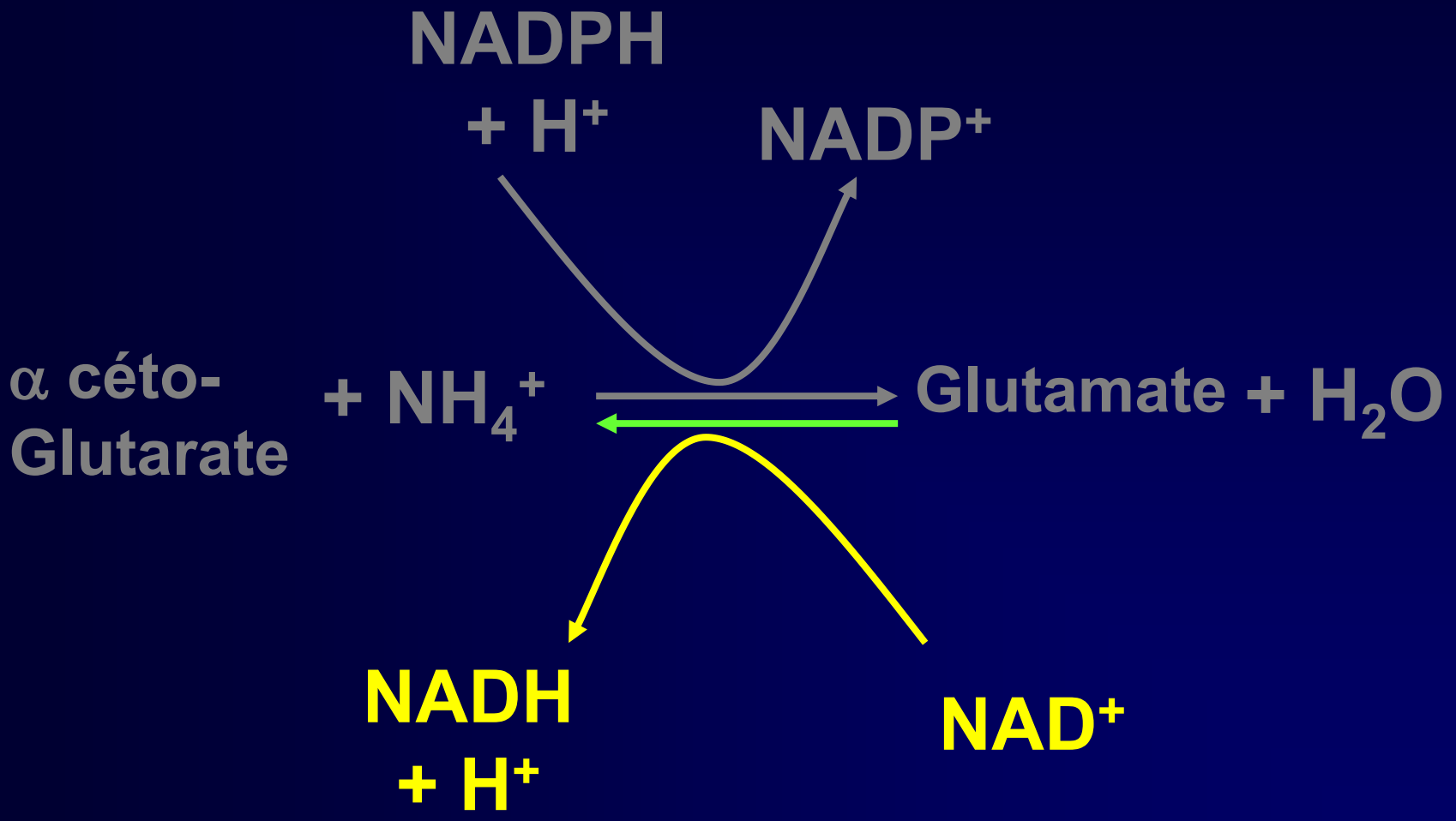
Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln

# Acide Glutamique


Glucose
3 phosphoglycérate
Pyruvate
Oxaloacétate
CAC
α Cétoglutarate
Glu



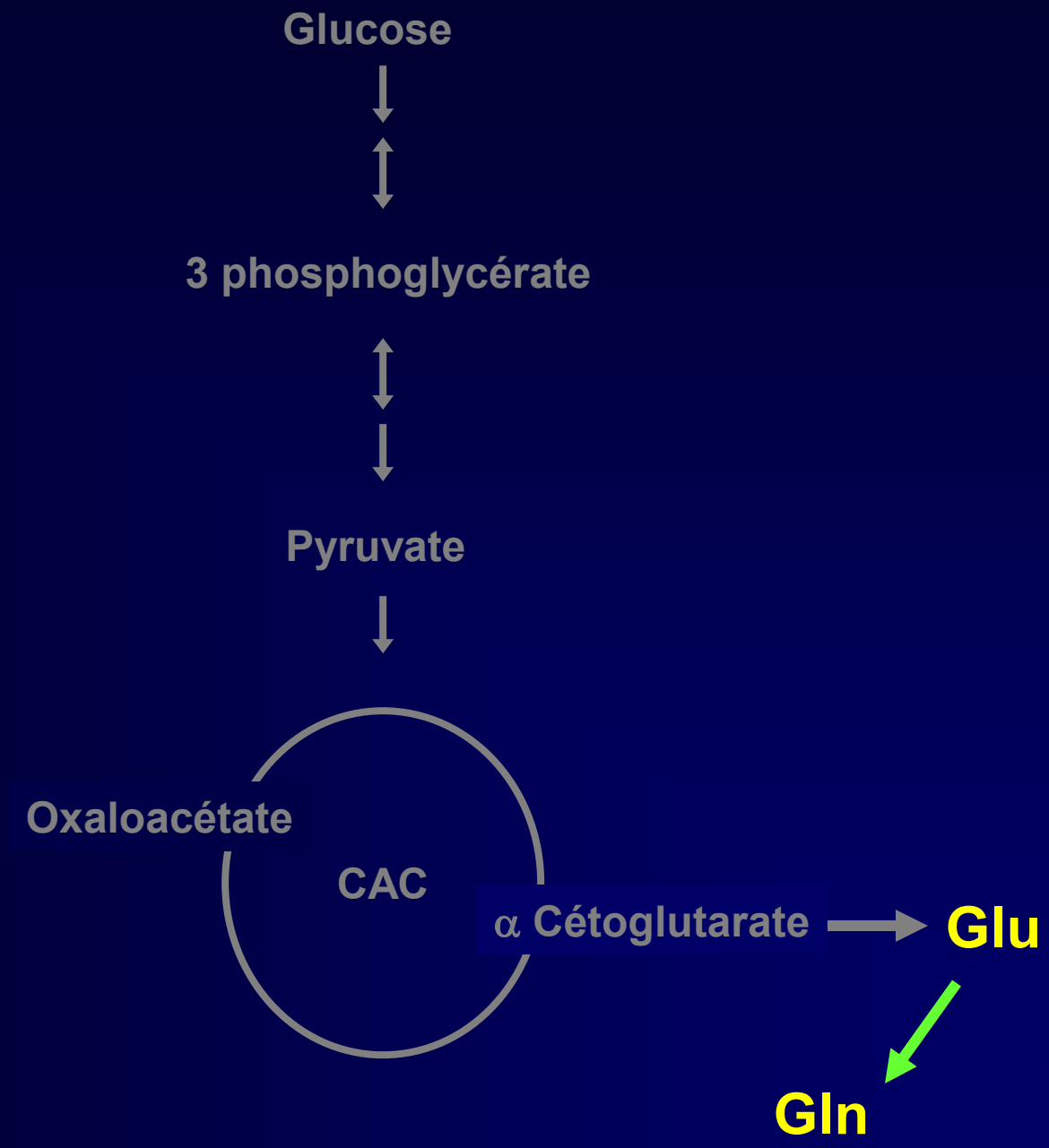

## Glutamate déshydrogénase

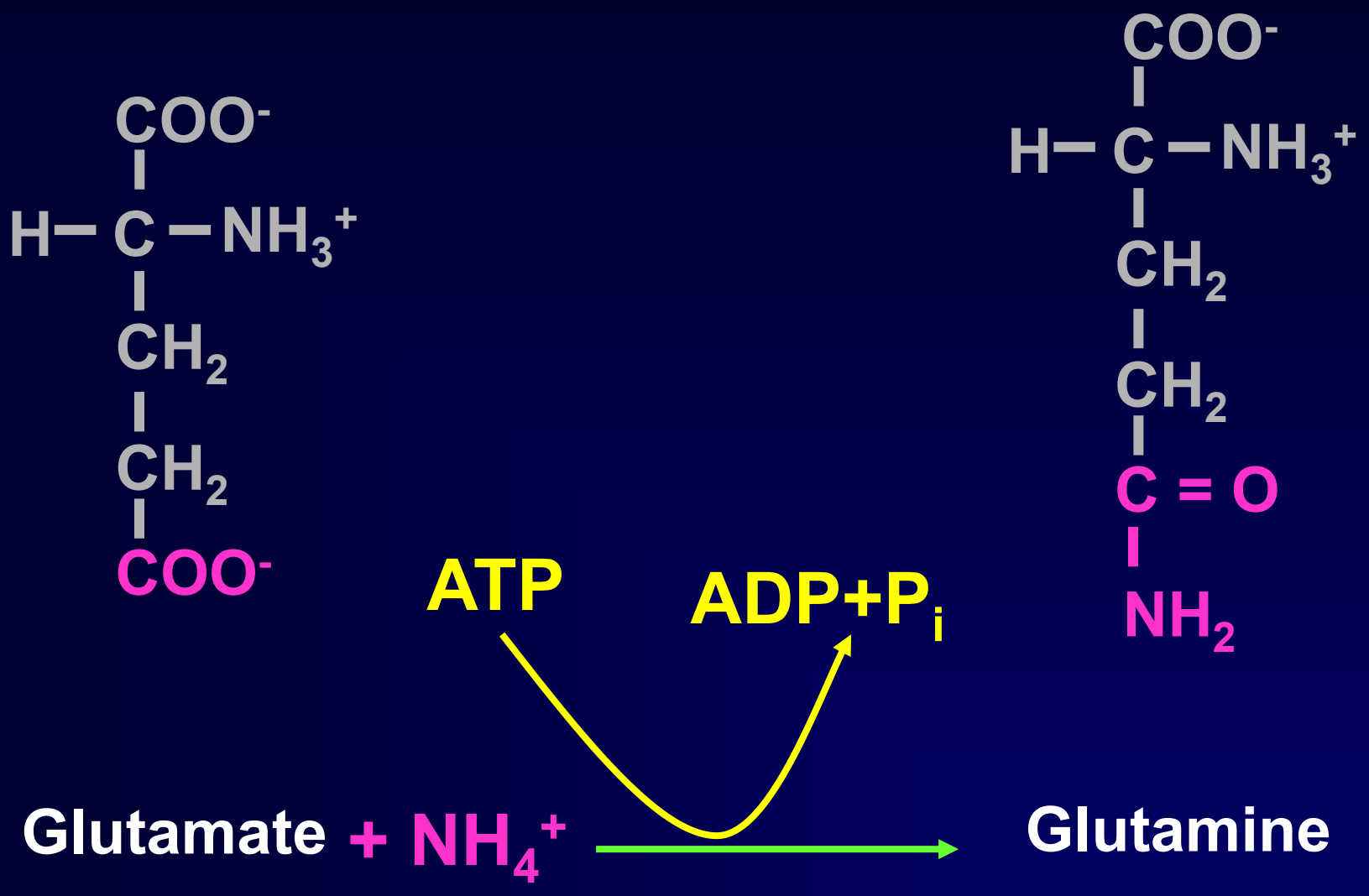

NADPH
+ H+
NADP+
α céto-
Glutarate
+ NH4+
Glutamate + H2O
NADH
+ H+
NAD+

# AA non indispensables

Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln

# Glutamine


Glucose
3 phosphoglycérate
Pyruvate
Oxaloacétate
CAC
α Cétoglutarate
Glu
Gln



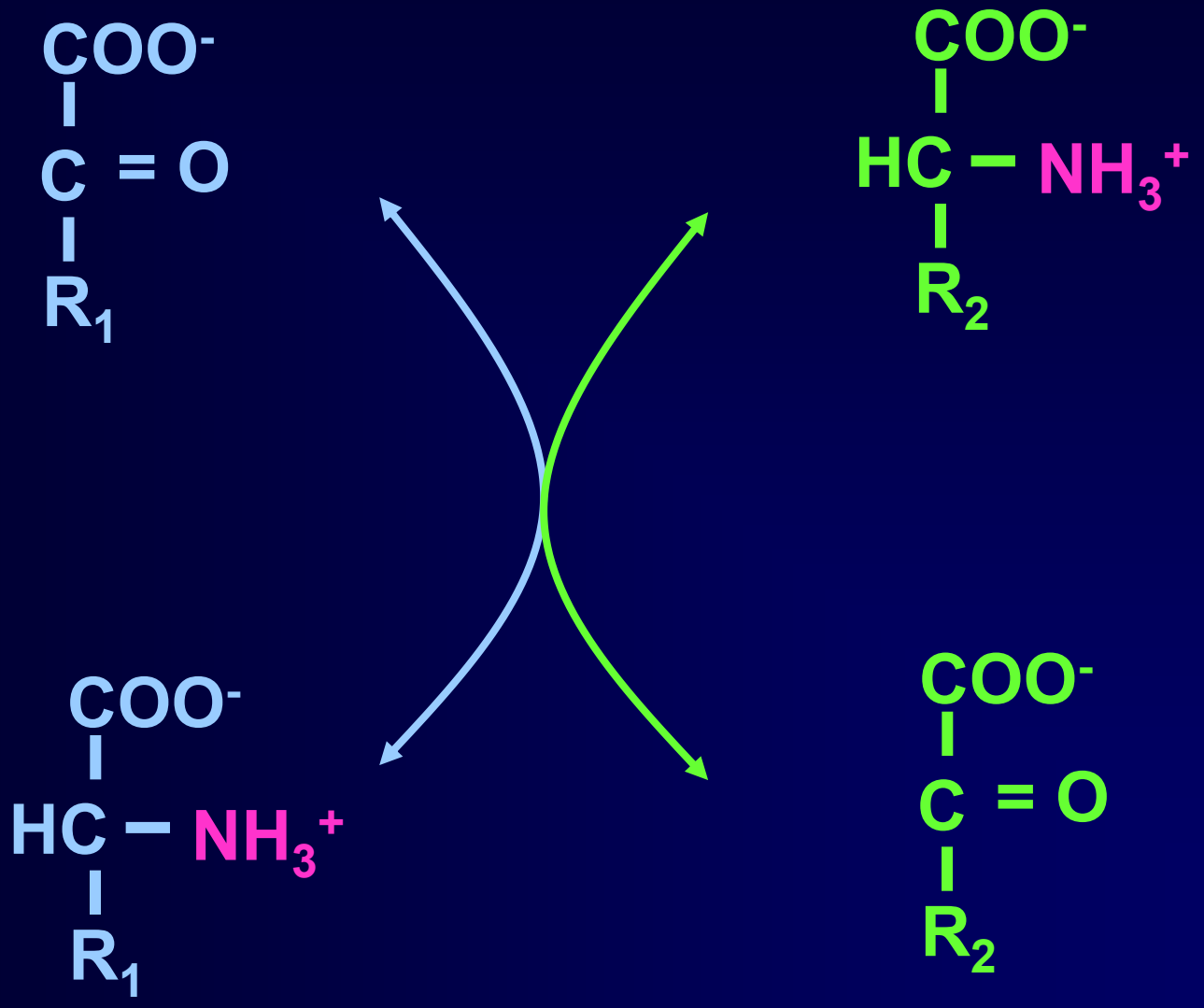

## Glutamine synthétase

# AA non indispensables

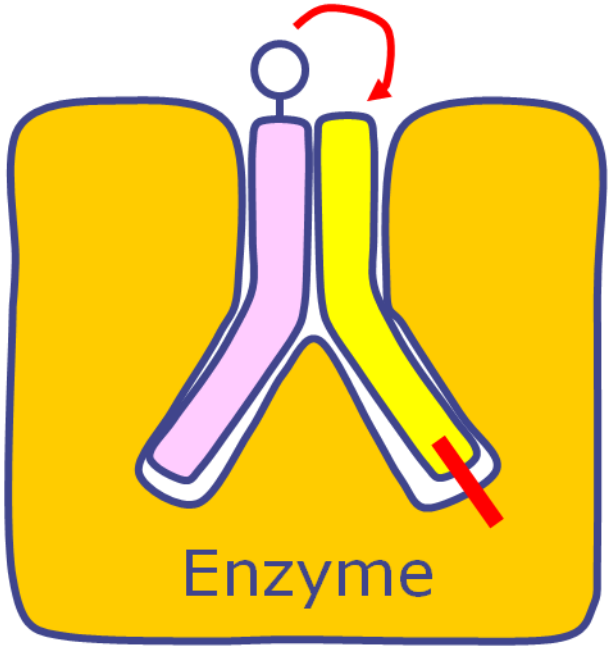
Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln

# Alanine et Aspartate

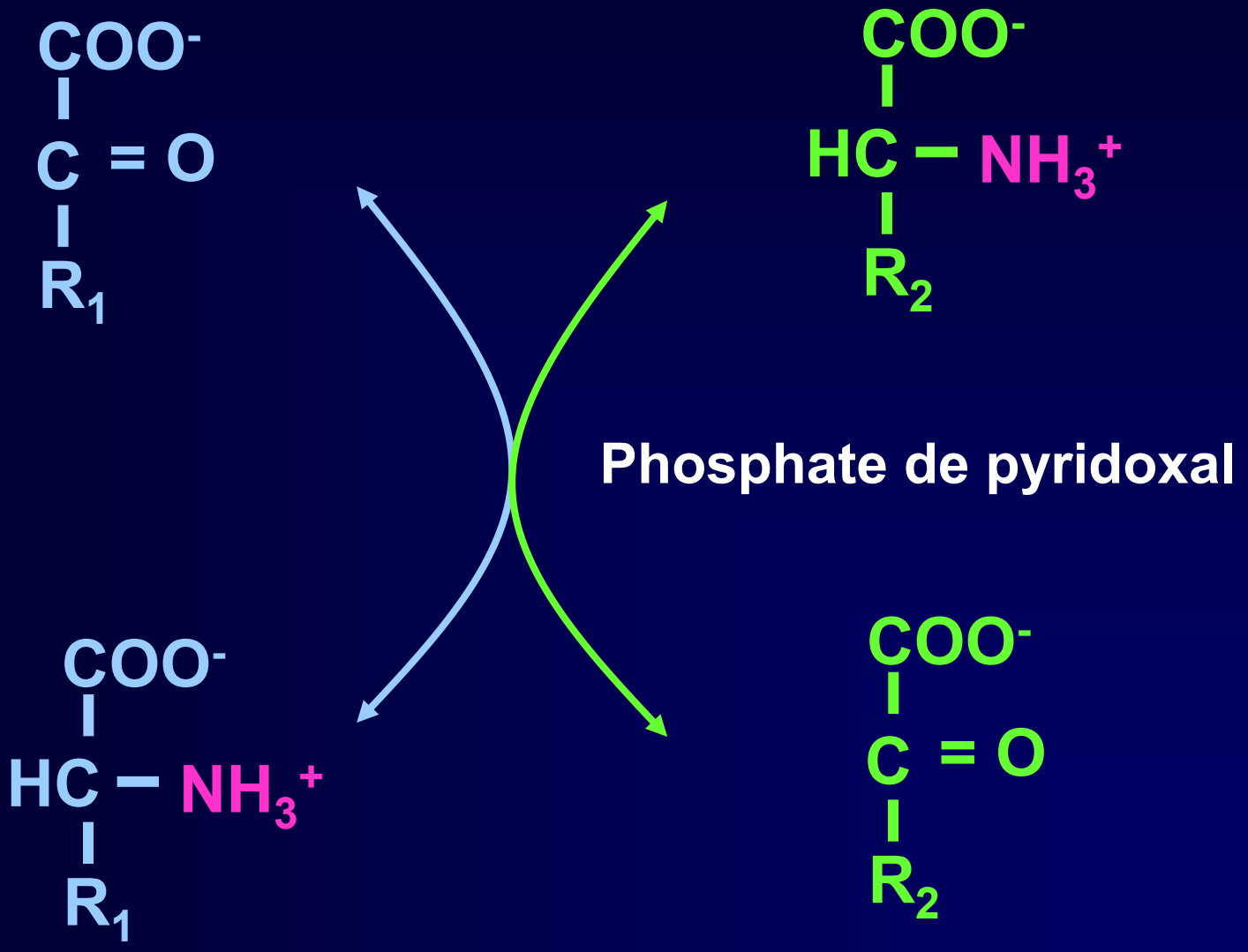
# Transamination

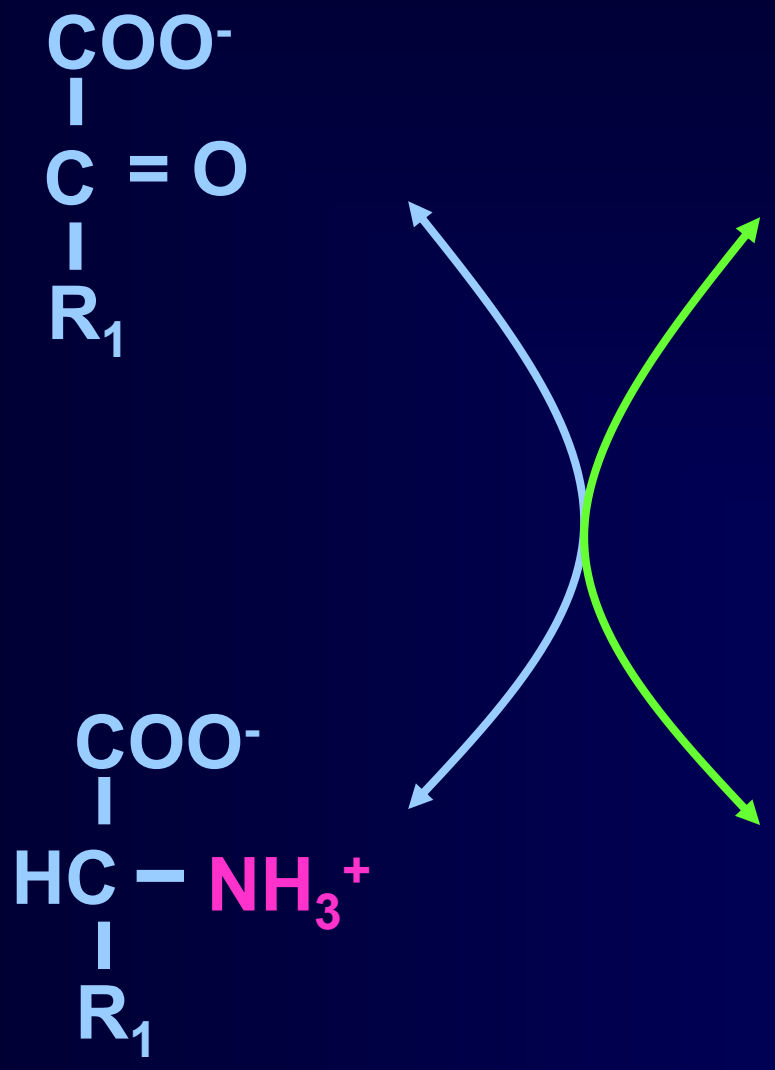


Transfert de groupement



# Transamination





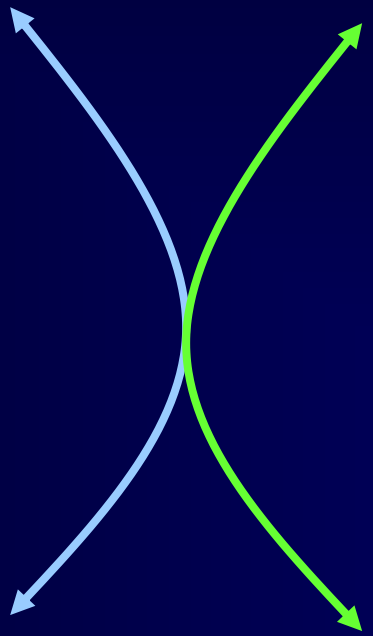

**Glu**

**α céto-Glutarate**

# Alanine

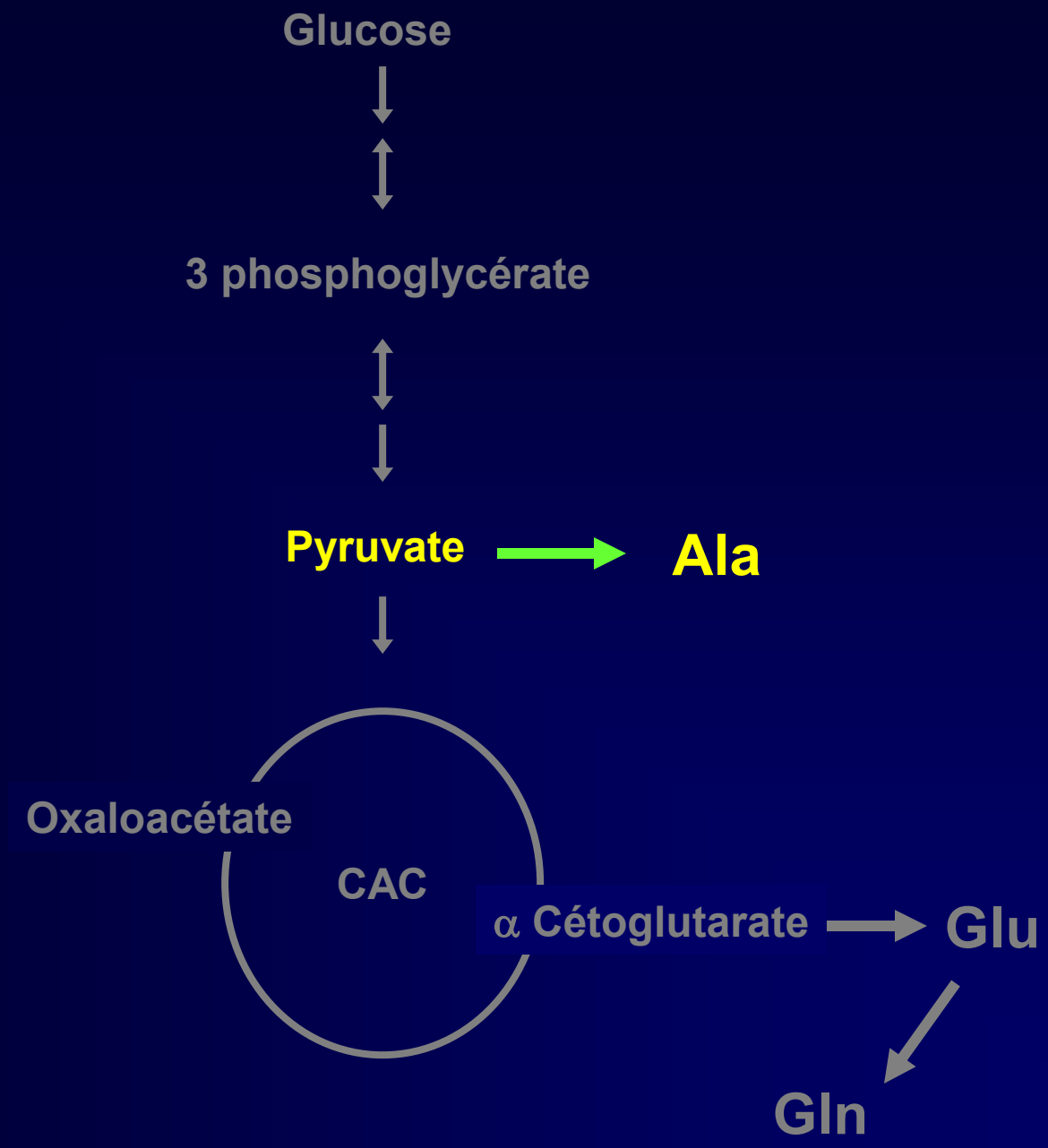
Pyruvate

Glu



Ala

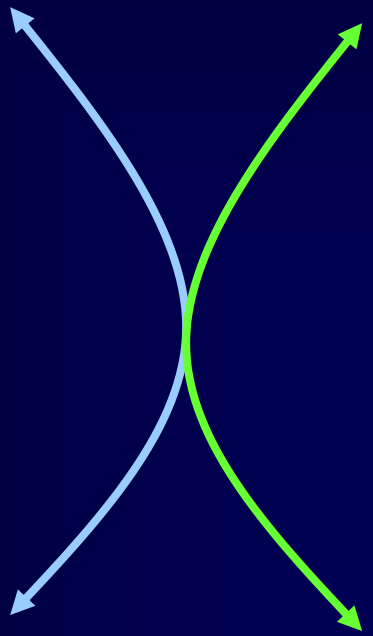
α céto-Glutarate


Glucose
3 phosphoglycérate
Pyruvate
Ala
Oxaloacétate
CAC
α Cétoglutarate
Glu
Gln

# Aspartate

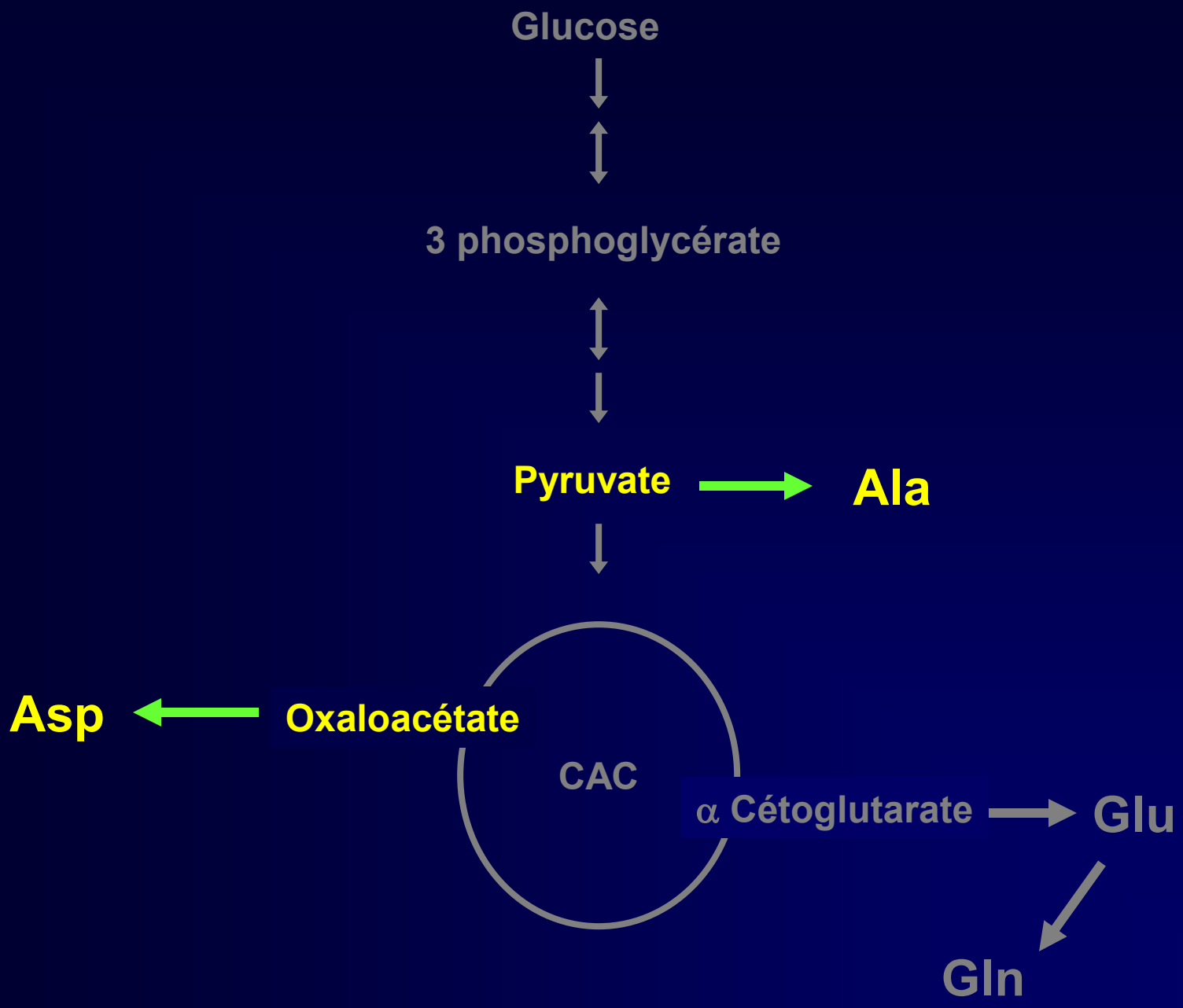
Oxalo-
acétate

Glu



Asp

α céto-
Glutarate


Glucose
3 phosphoglycérate
Pyruvate
Ala
Asp
Oxaloacétate
CAC
α Cétoglutarate
Glu
Gln

# AA non indispensables

Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln

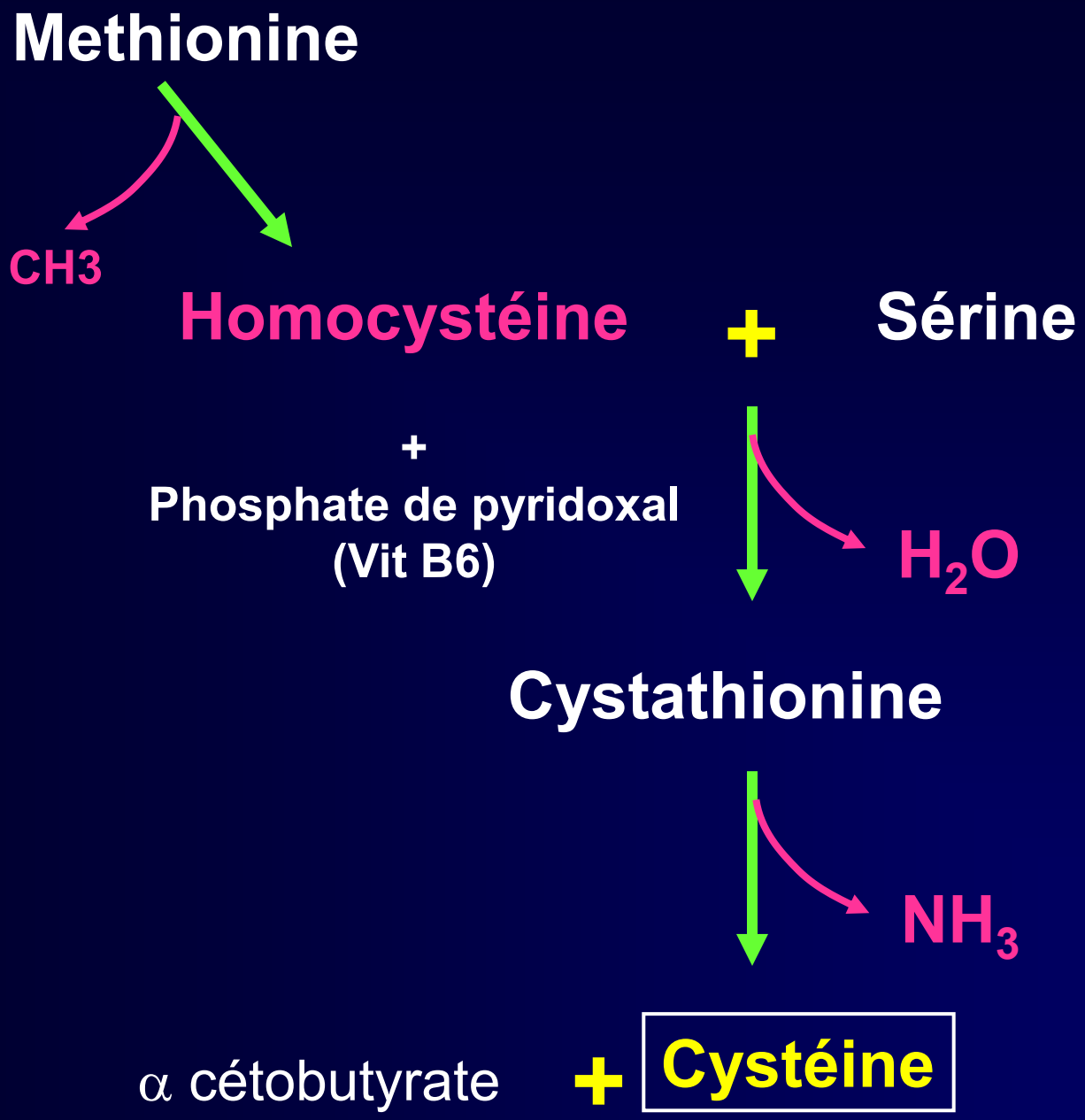
# AA non indispensables

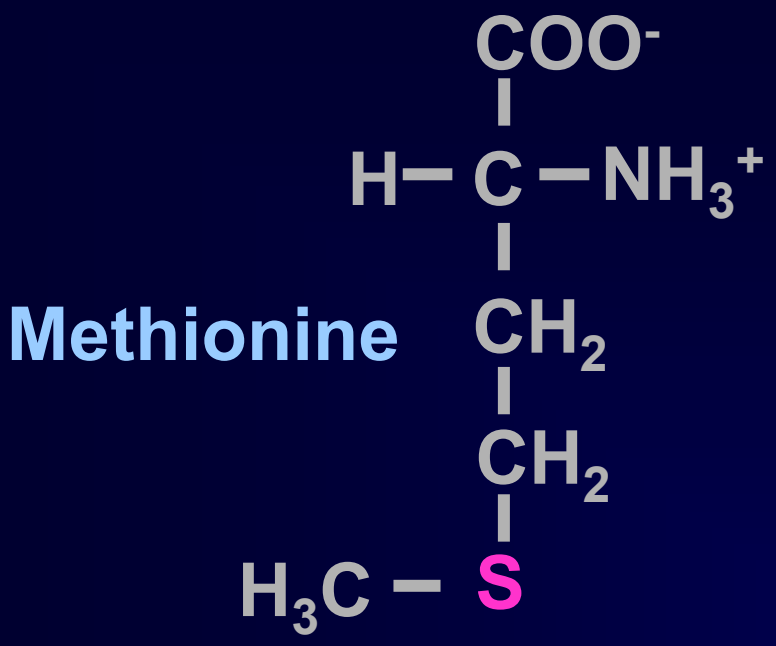
Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln

# AA non indispensables

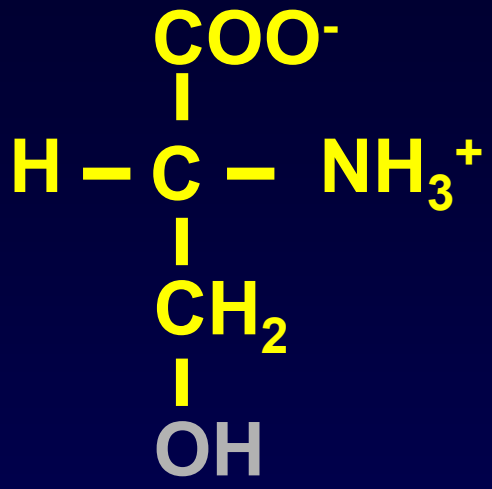
Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln

# Cystéine

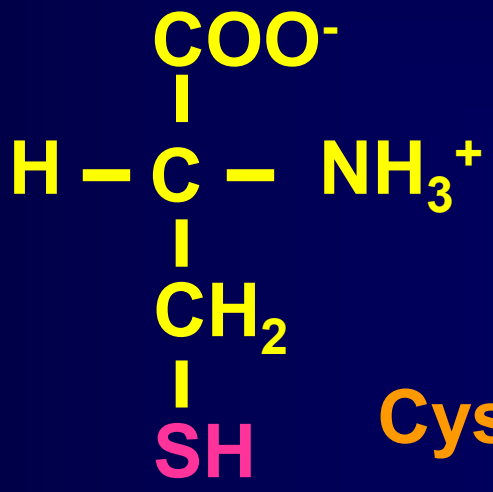

Methionine
CH3
Homocystéine
+
Sérine
+
Phosphate de pyridoxal
(Vit B6)
$H_2O$
Cystathionine
$NH_3$
α cétobutyrate
+
Cystéine



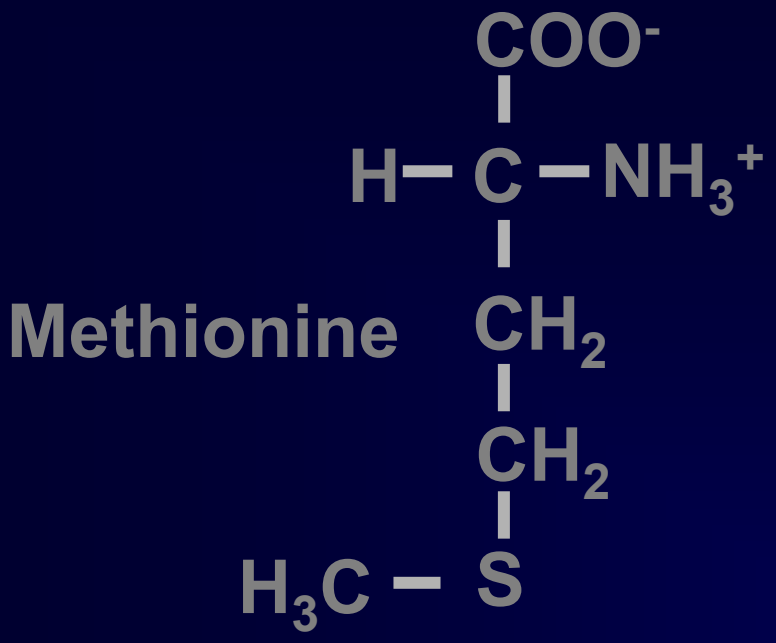

Methionine



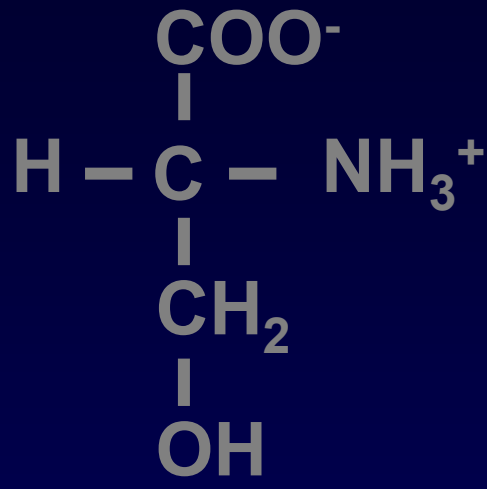

sérine




Cystéine

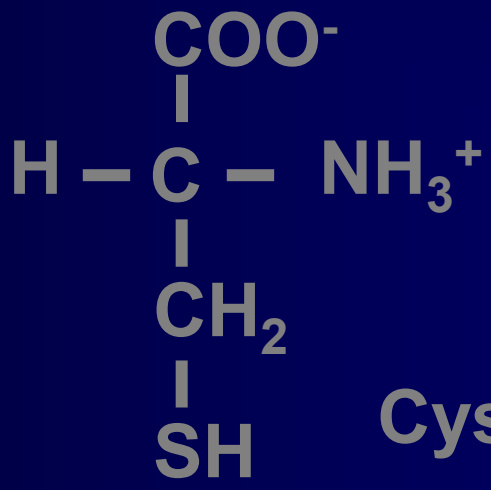



Methionine



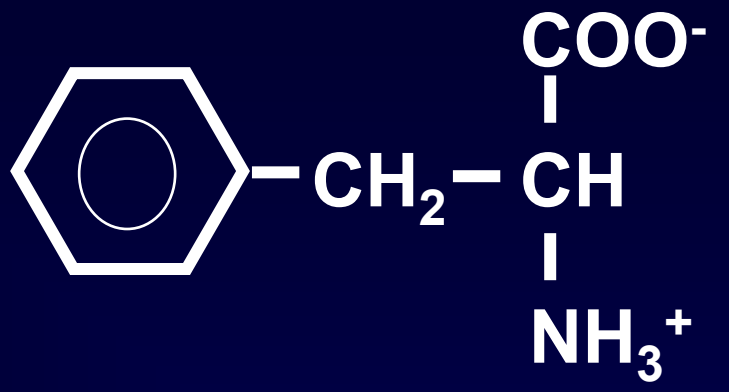

sérine

## Transulfuration




Cystéine

# Tyrosine




Phénylalanine

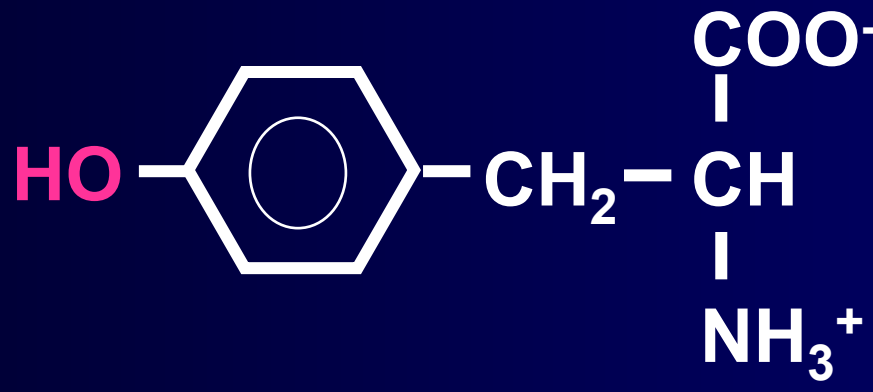
$O_2$

$H_4$Bioptérine

Phénylalanine Hydroxylase

$H_2O$

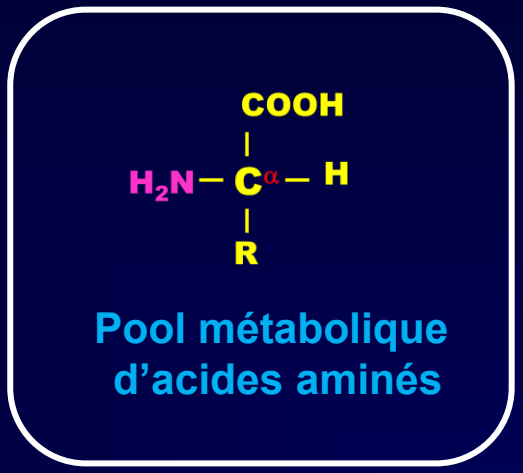
$H_2$Bioptérine




Tyrosine

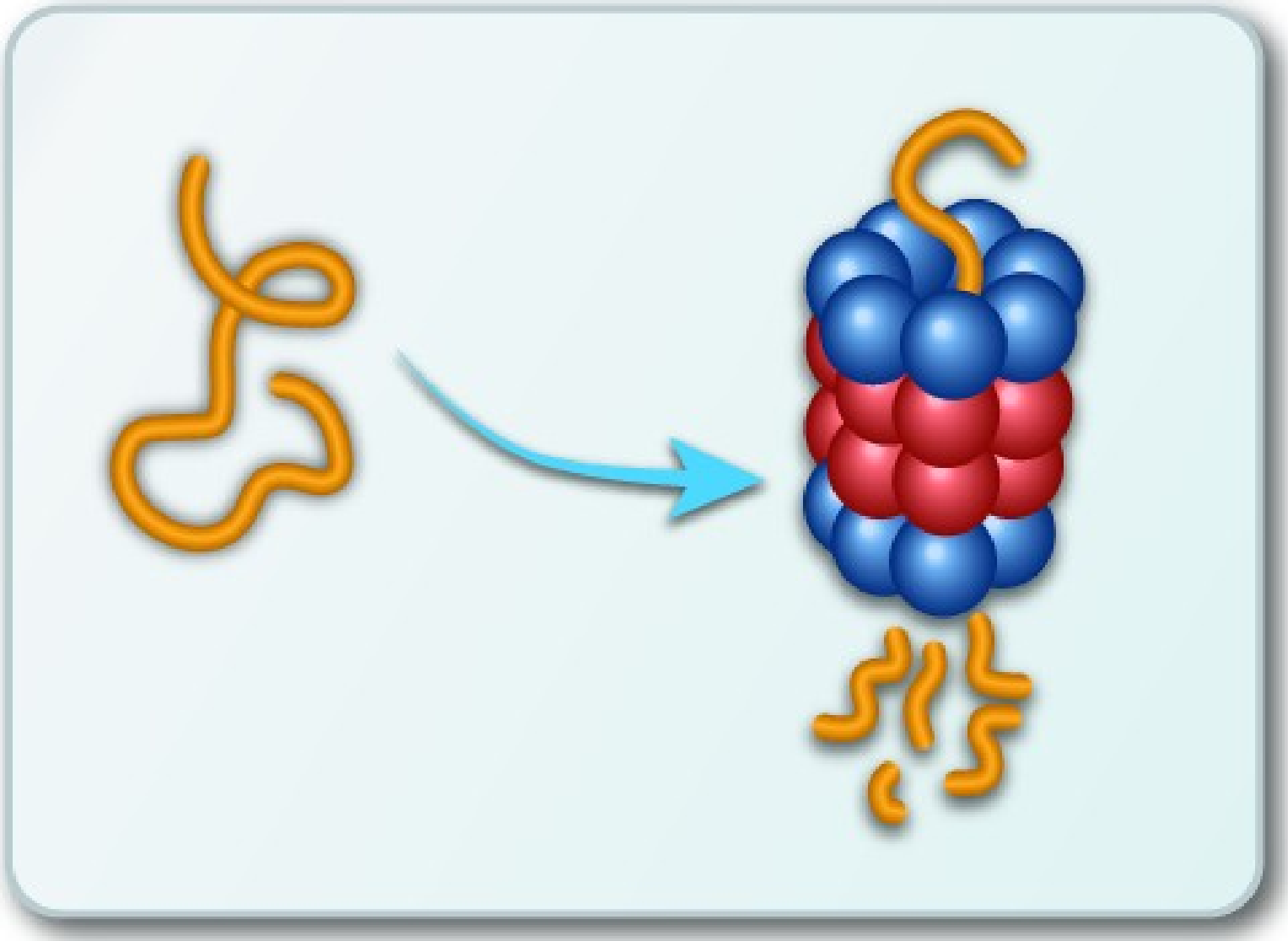
Protéines tissulaires

Dissociation



# Dégradation des protéines

- Vitesse variable
- Processus régulé



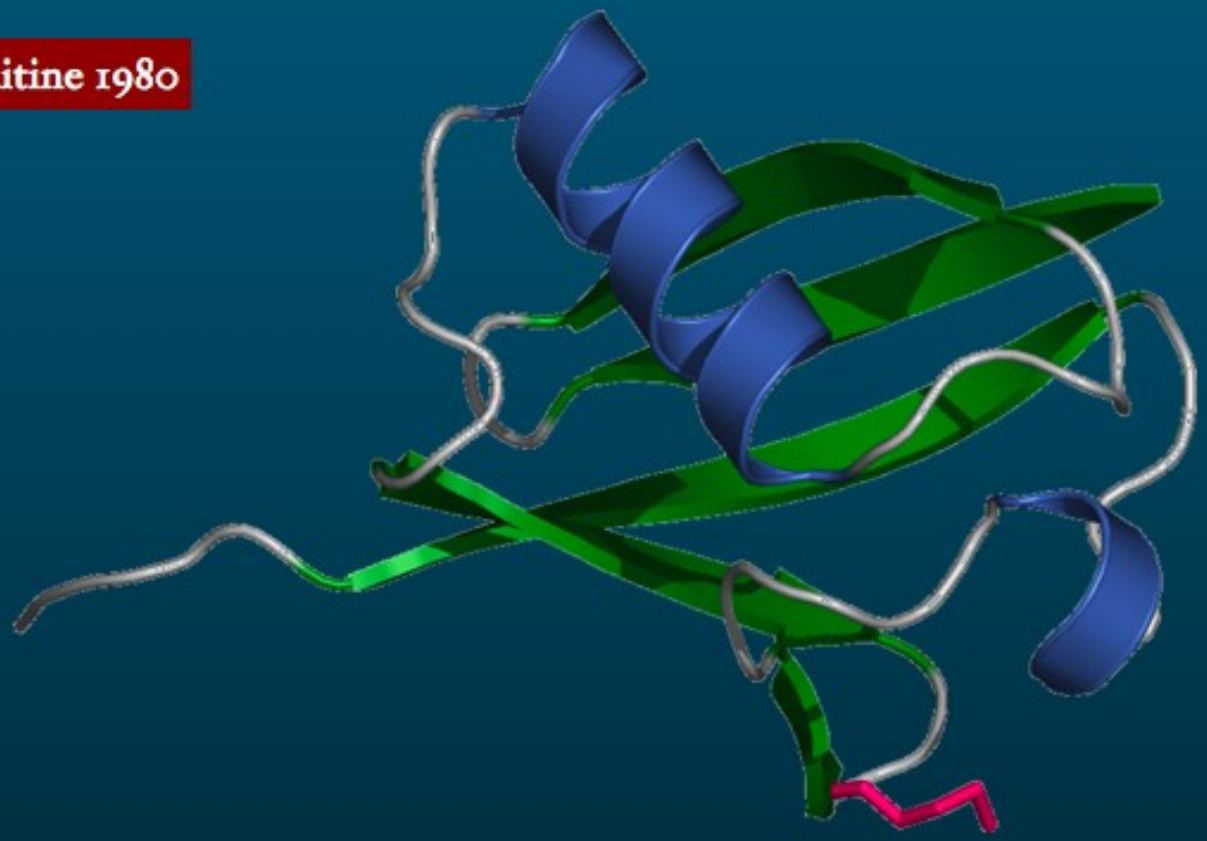
**Protéasome**

# Protéasome

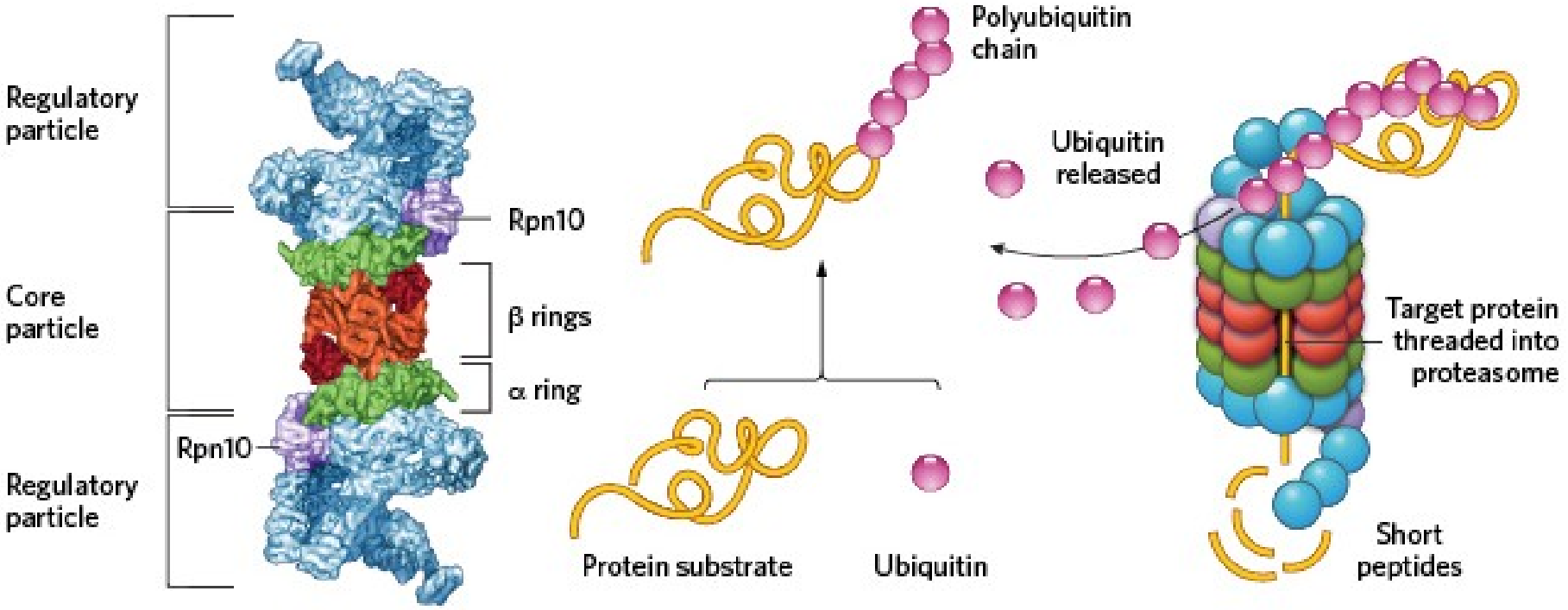

ATTENTION
DEFECTUEUX
Commentaires
Date
Autorisation

# Ubiquitine

Ubiquitine 1980

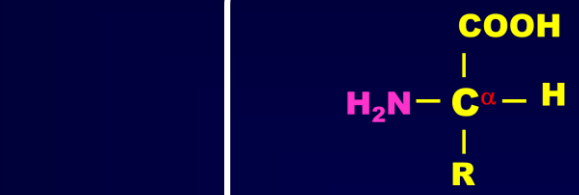


Prix Nobel de Chimie 2004 : Aaron Ciechanover, Avram Hershko, Irvin Rose


Regulatory particle
Core particle
Regulatory particle
Rpn10
β rings
α ring
Rpn10
Polyubiquitin chain
Ubiquitin released
Target protein threaded into proteasome
Protein substrate
Ubiquitin
Short peptides

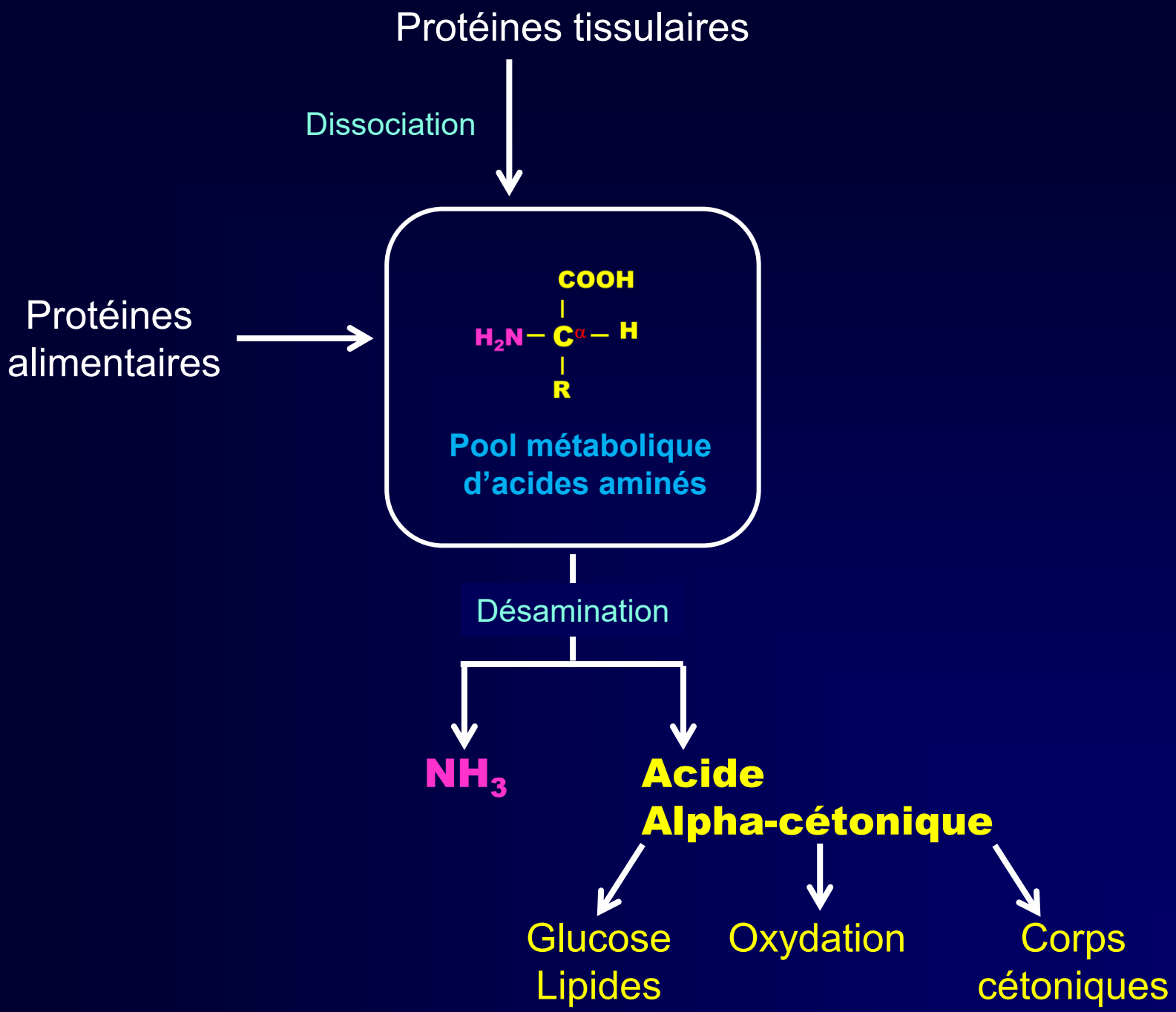
Protéines tissulaires

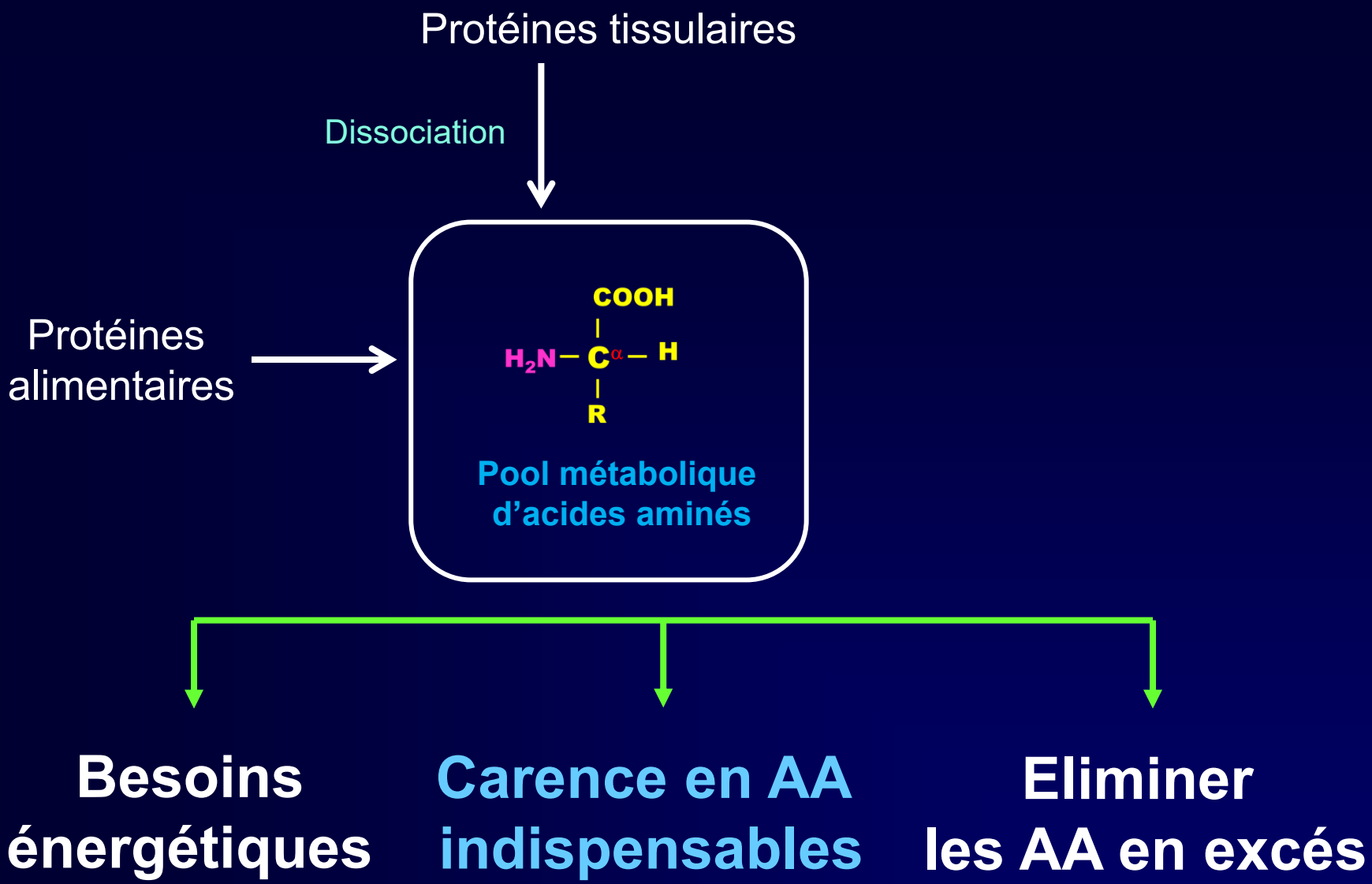
Dissociation




**Pool métabolique d'acides aminés**

Renouvellement des protéines


Protéines tissulaires
Dissociation
Protéines alimentaires
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Désamination
NH3
Acide Alpha-cétonique
Glucose Lipides
Oxydation
Corps cétoniques


Protéines tissulaires
Dissociation
Protéines alimentaires
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Besoins énergétiques
Carence en AA indispensables
Eliminer les AA en excés

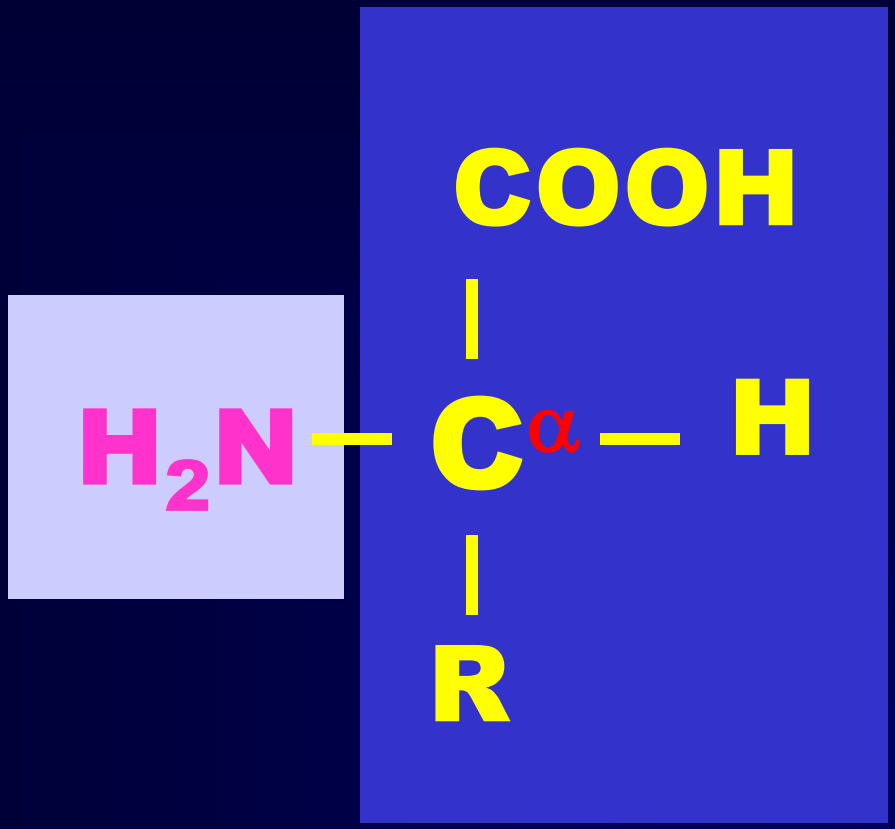
## AA indispensables

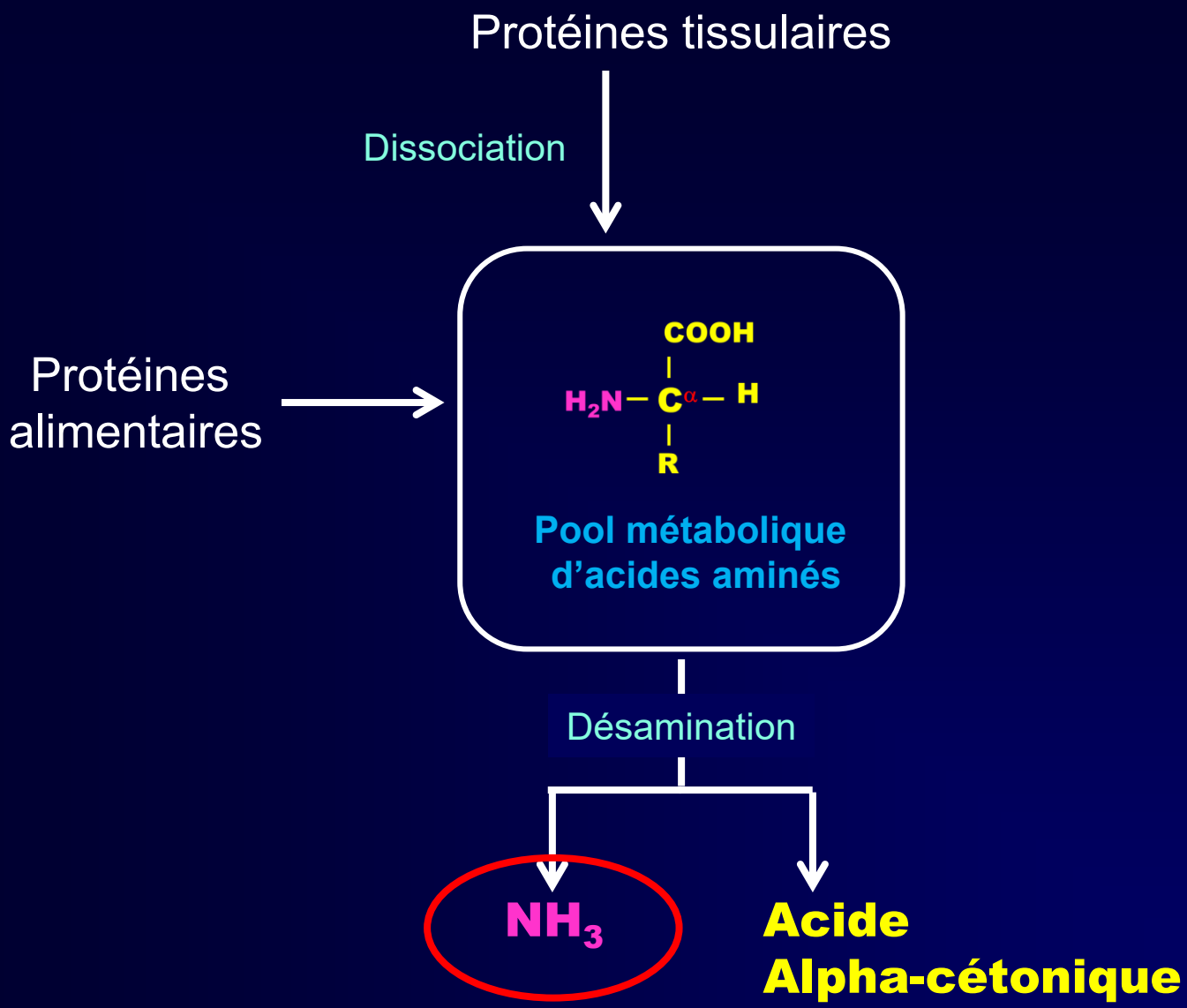
Arg
His
Ile
Leu
Lys
Met
Phe
Thr
Trp
Val

## AA non indispensables

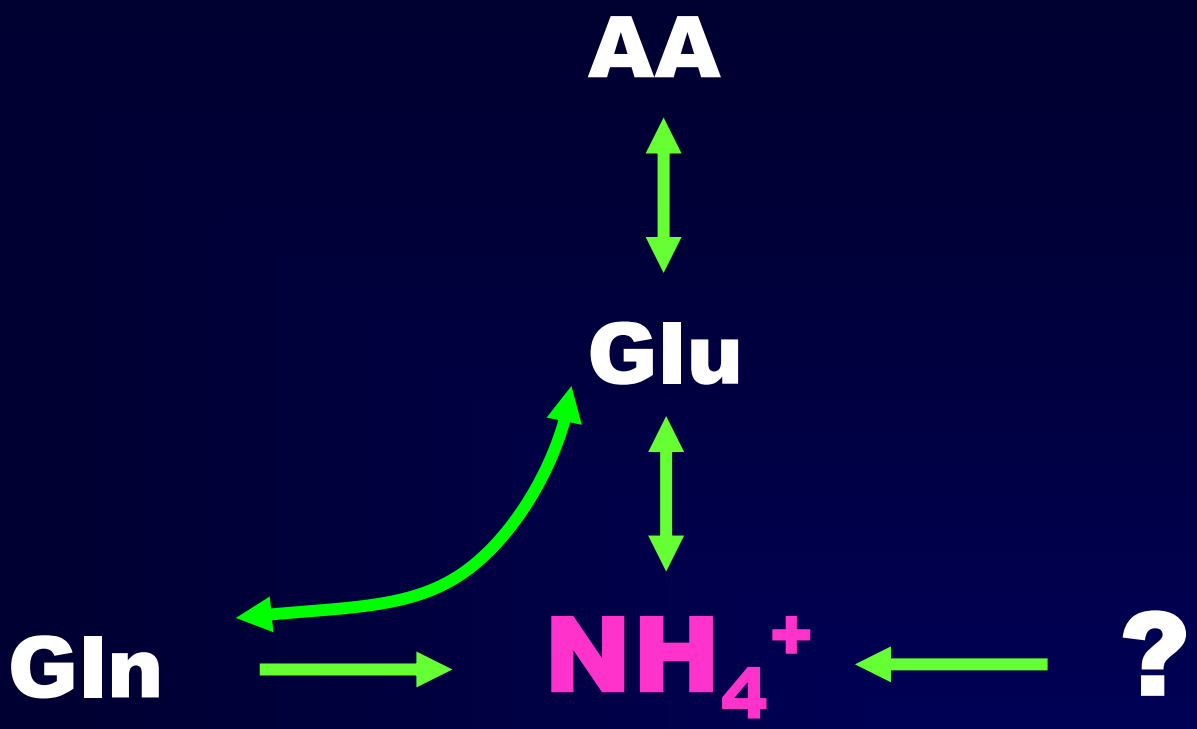
Ala
Asp
Cys
Glu
Gly
Pro
Ser
Tyr
Asn
Gln

Hyp
Hyl


COOH
H2N
Cα
H
R

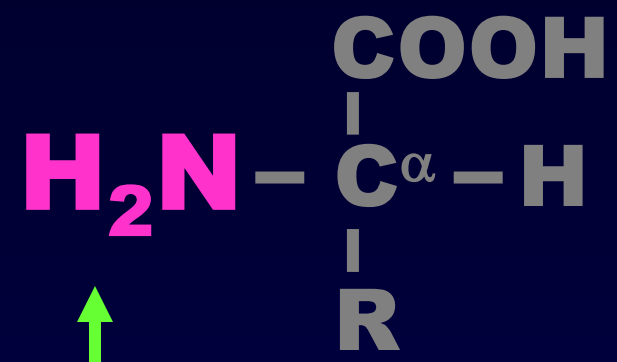

Protéines tissulaires
Dissociation
Protéines alimentaires
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Désamination
NH3
Acide Alpha-cétonique

# Flux et métabolisme de l ’azote


AA
Glu
Gln
$NH_4^+$
?

AA

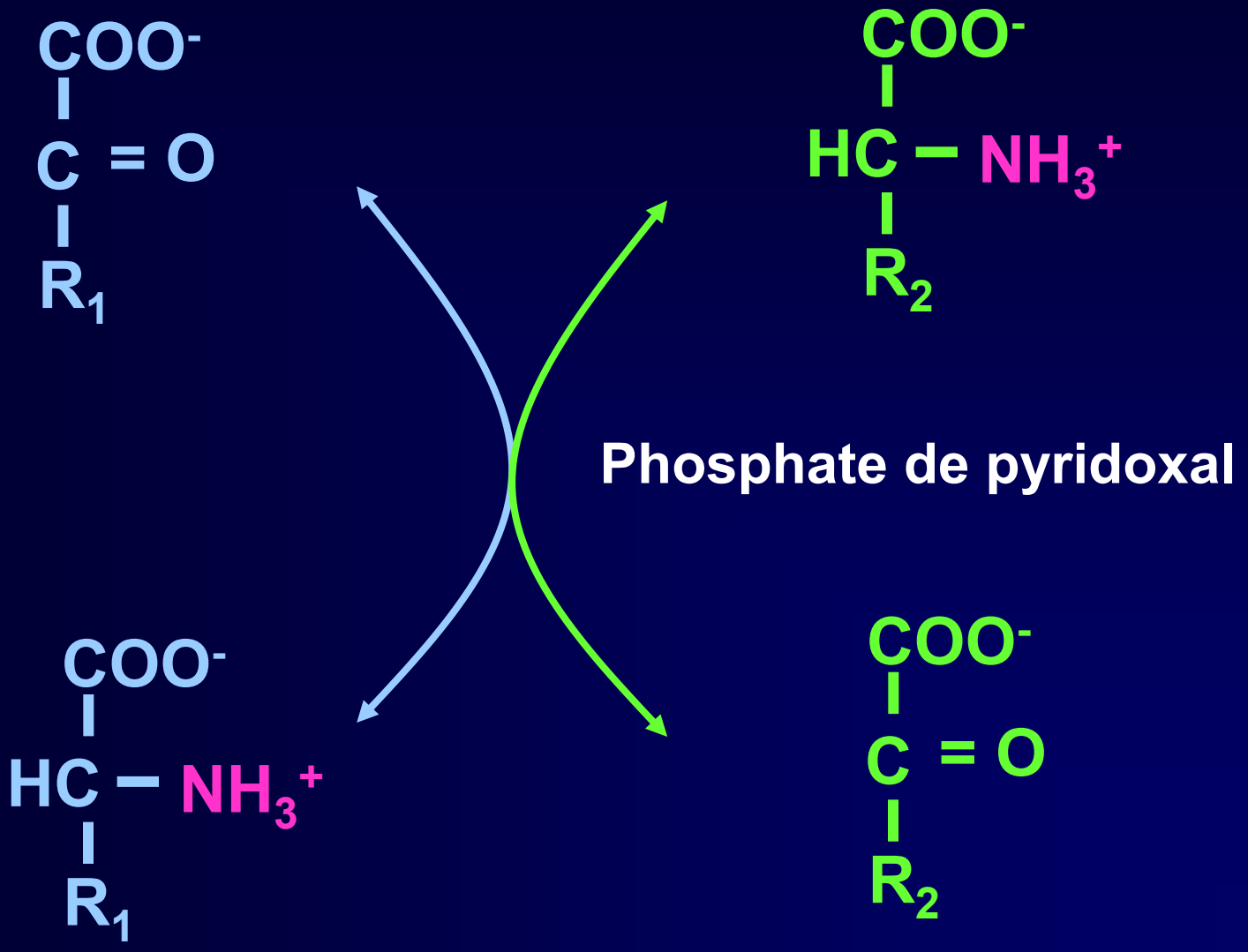
Glu

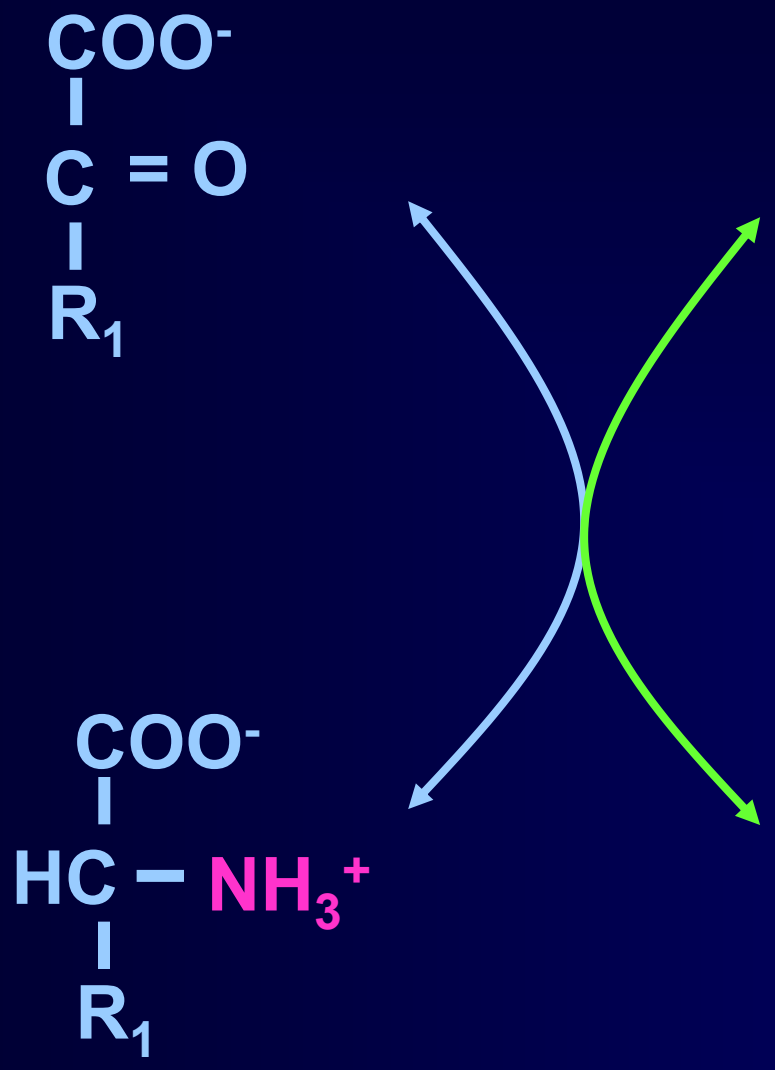



Transamination

Glutamate

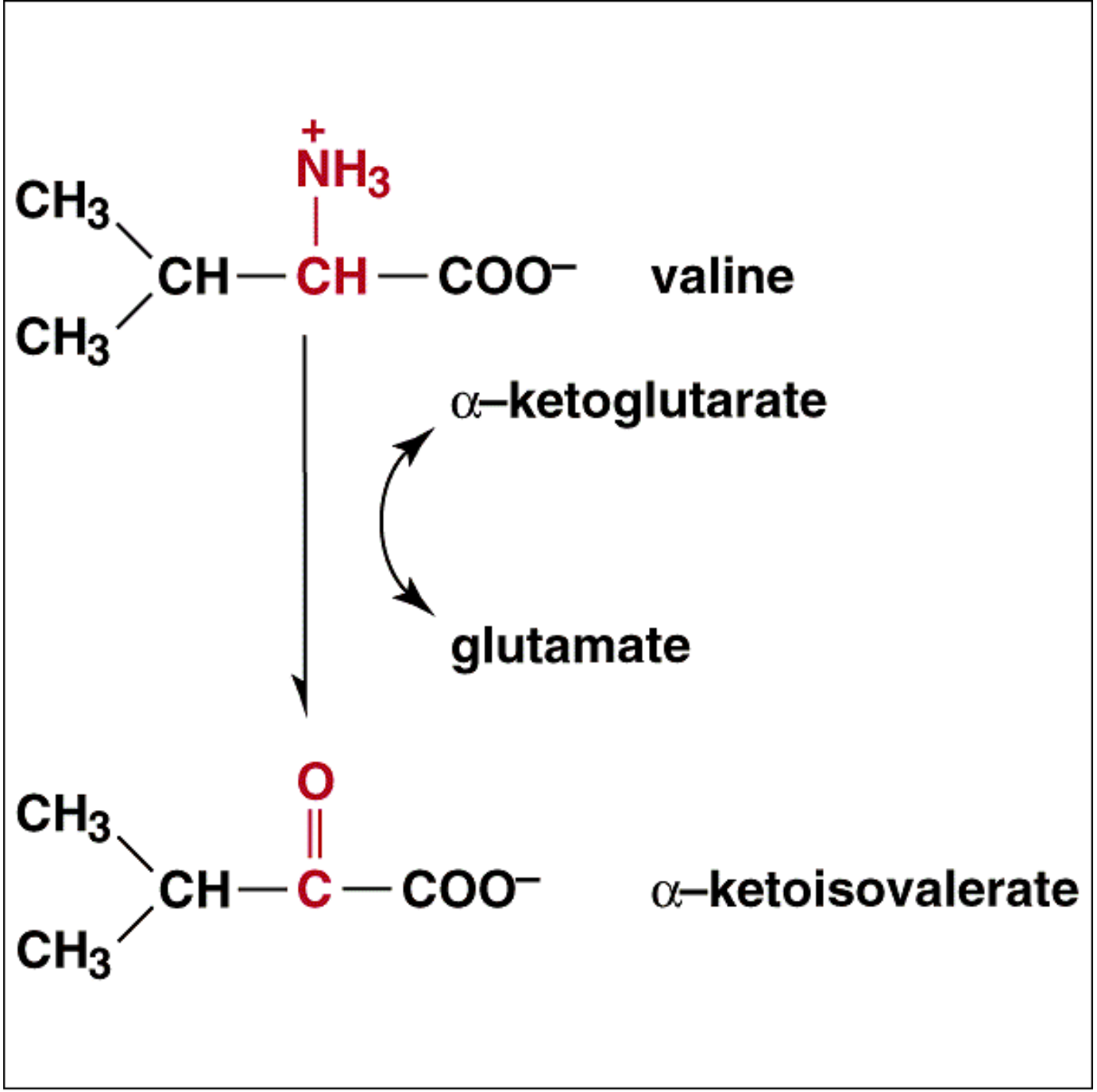
# Transamination

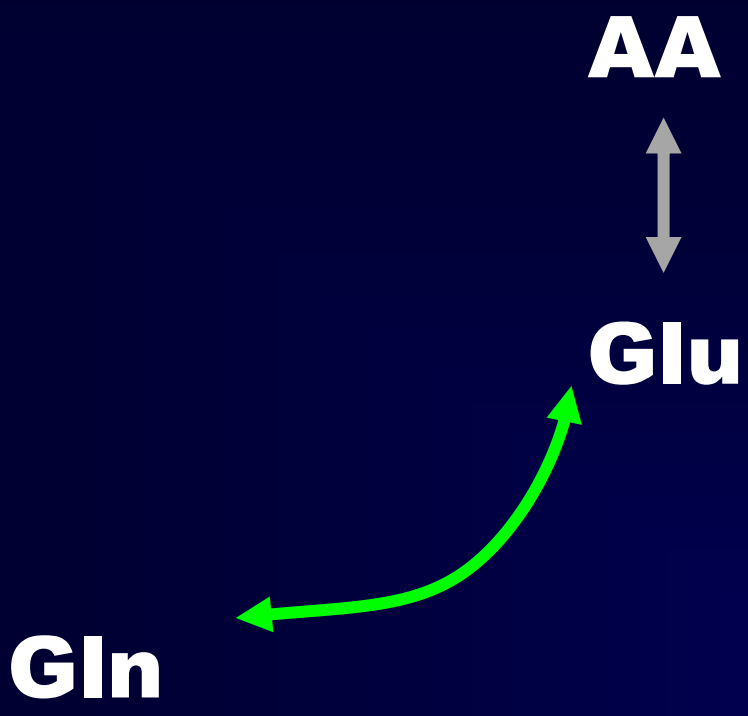




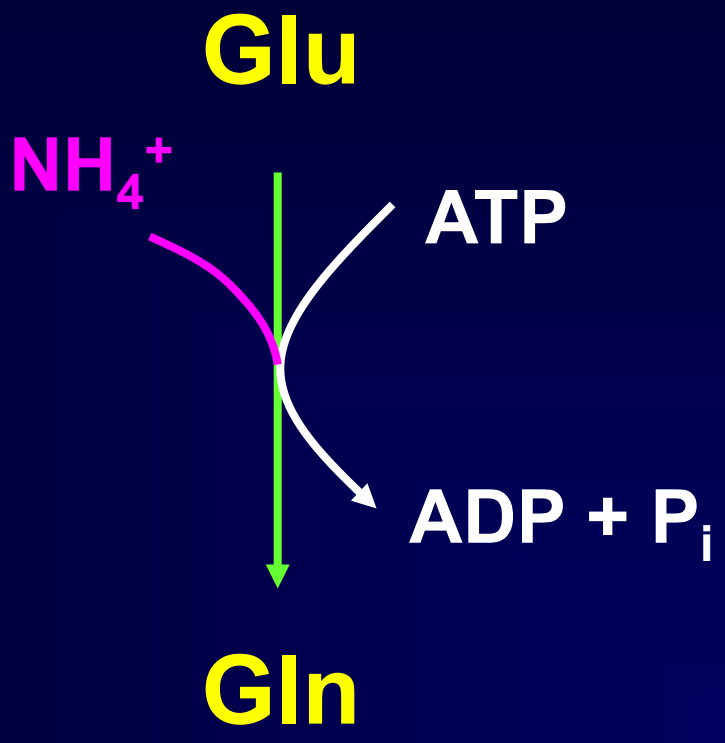

**Glu**

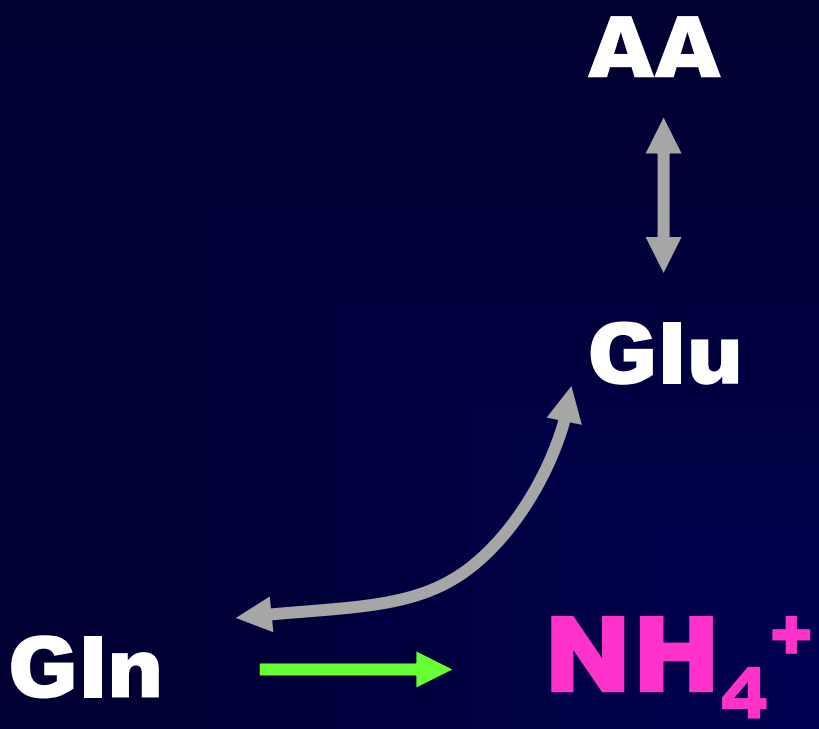
**α céto-Glutarate**



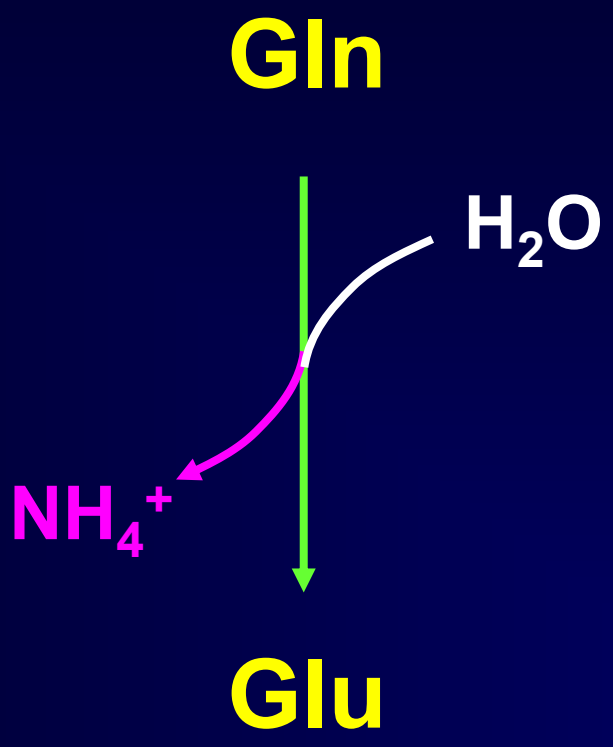

AA
Glu
Gln

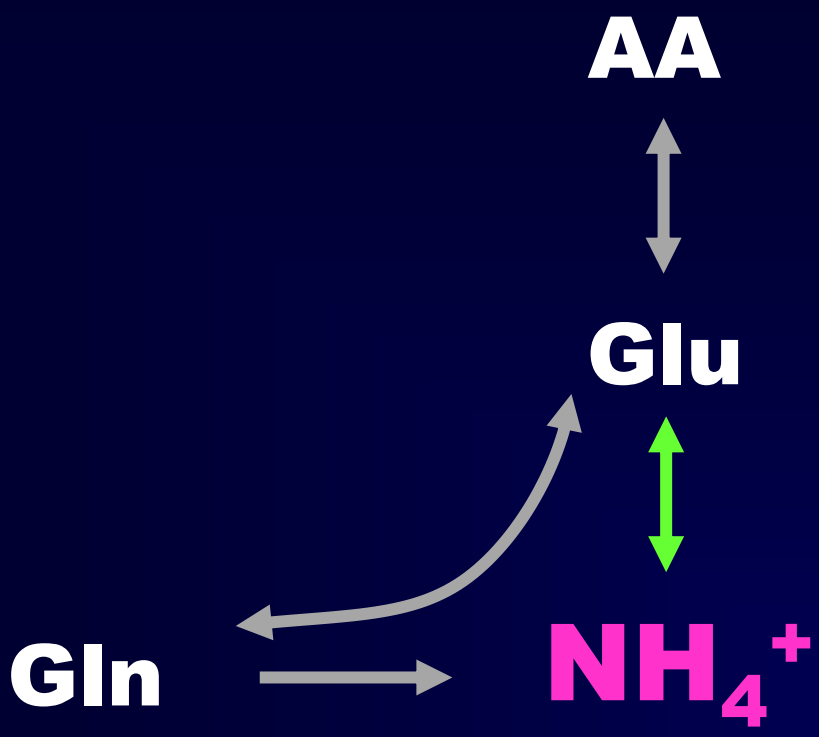
# Glutamine synthétase

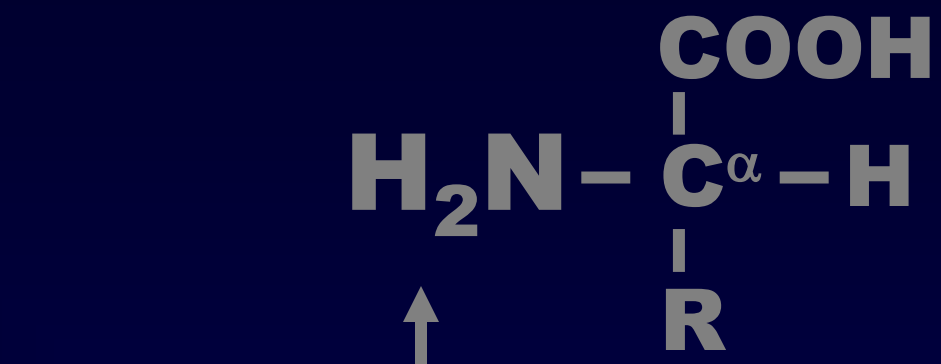



AA
Glu
Gln
$NH_4^+$

# Glutaminase




AA
Glu
Gln
$NH_4^+$




Transamination

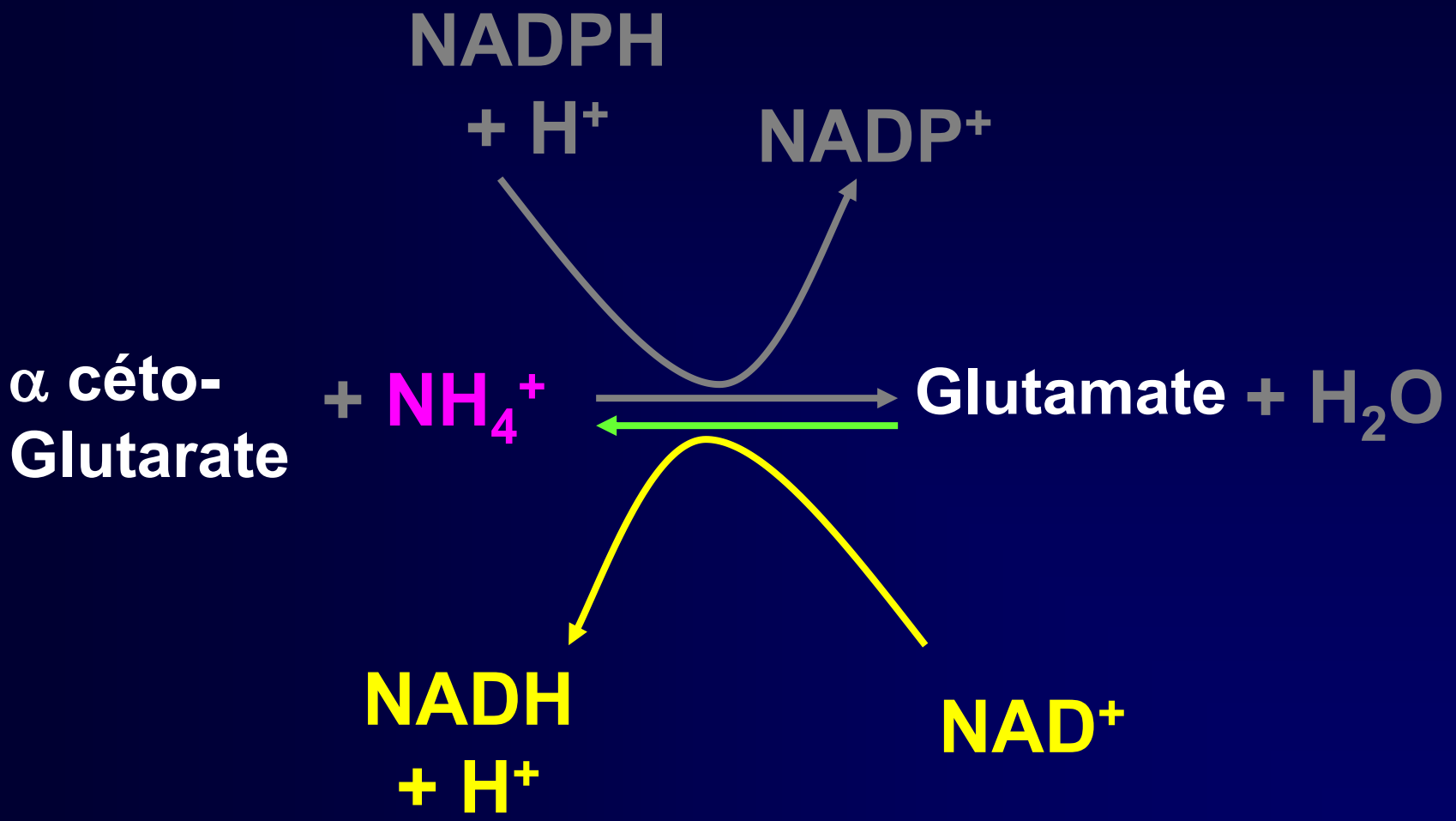
**Glutamate**

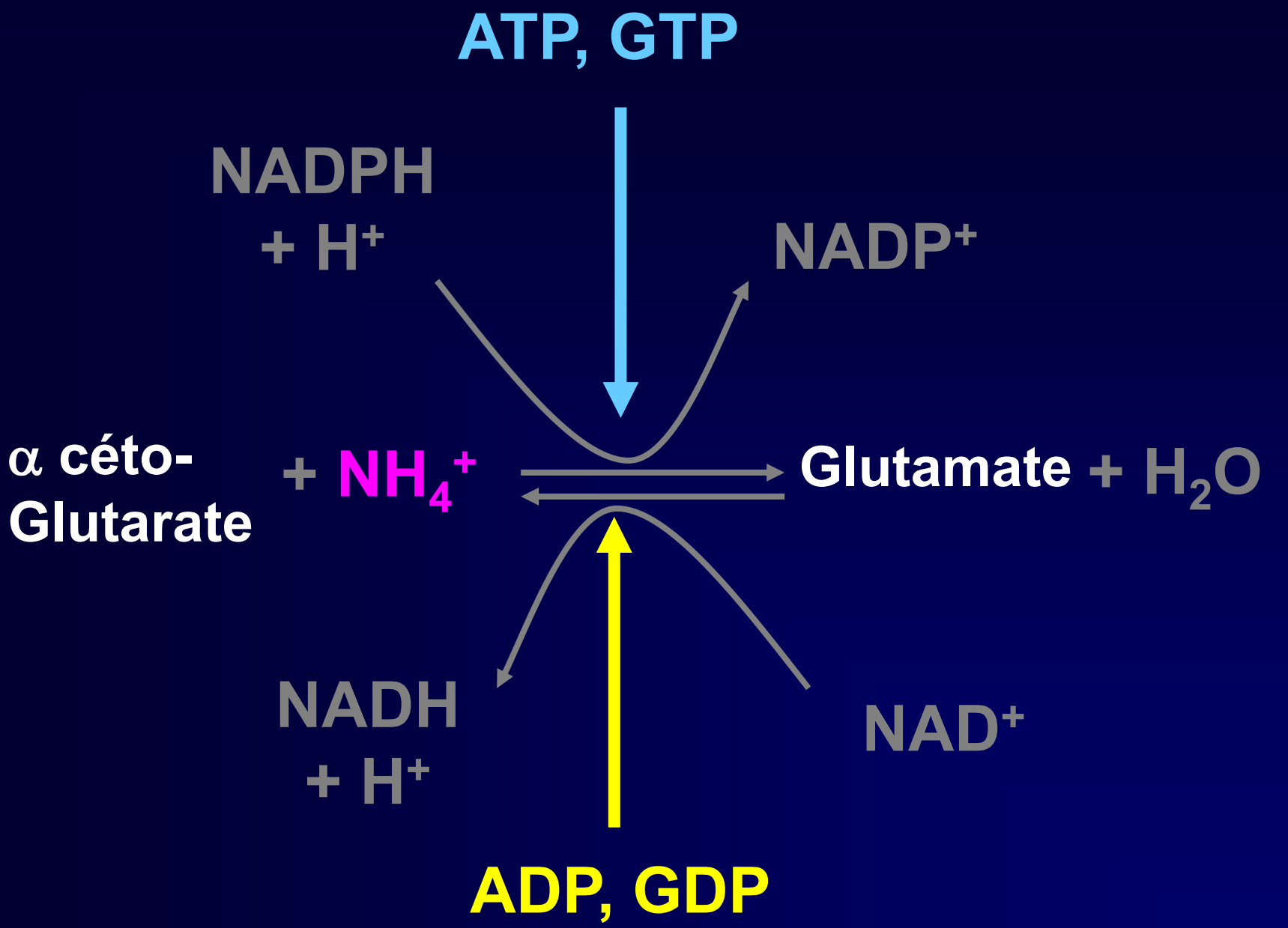
Désamination
Oxydative

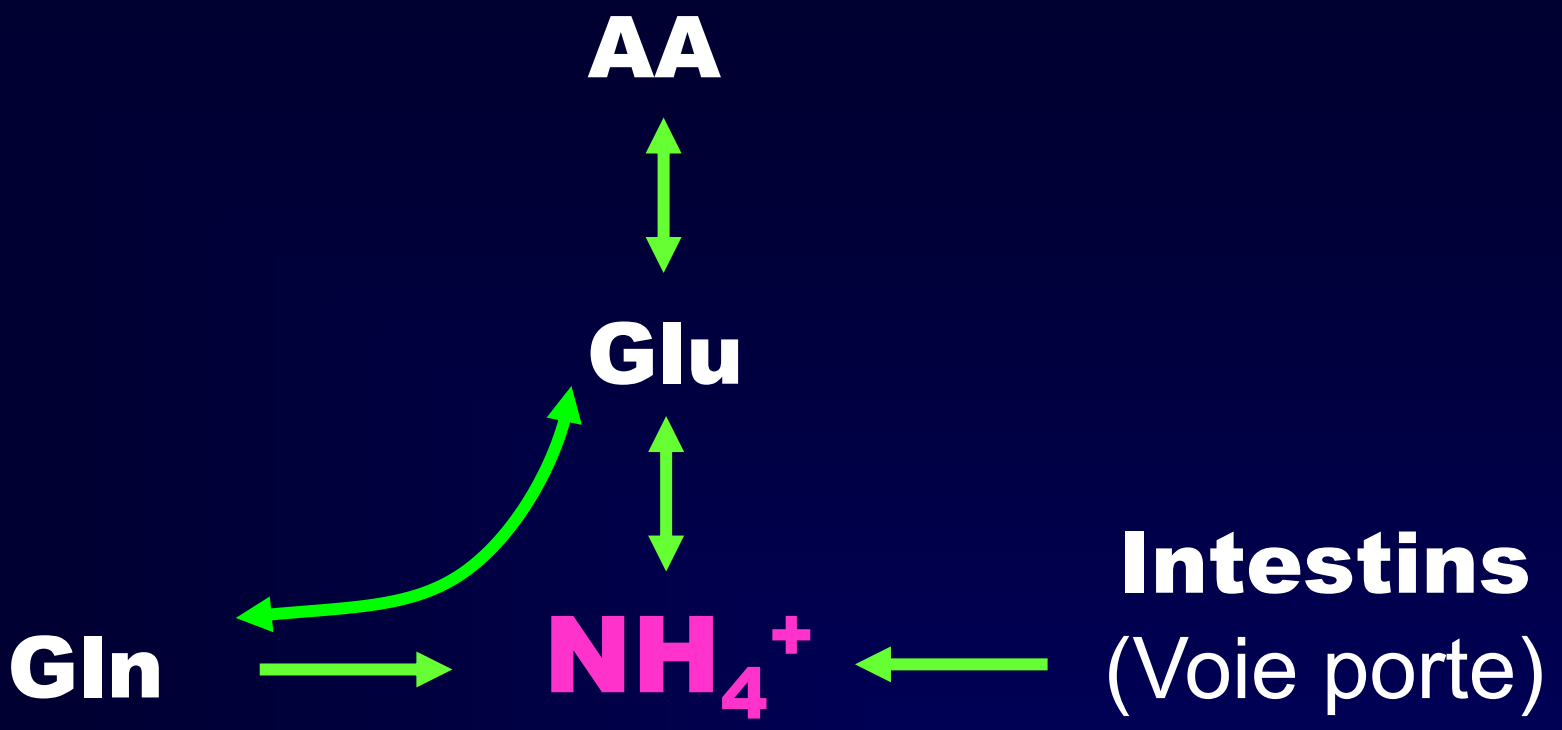
Glutamate
déshydrogénase

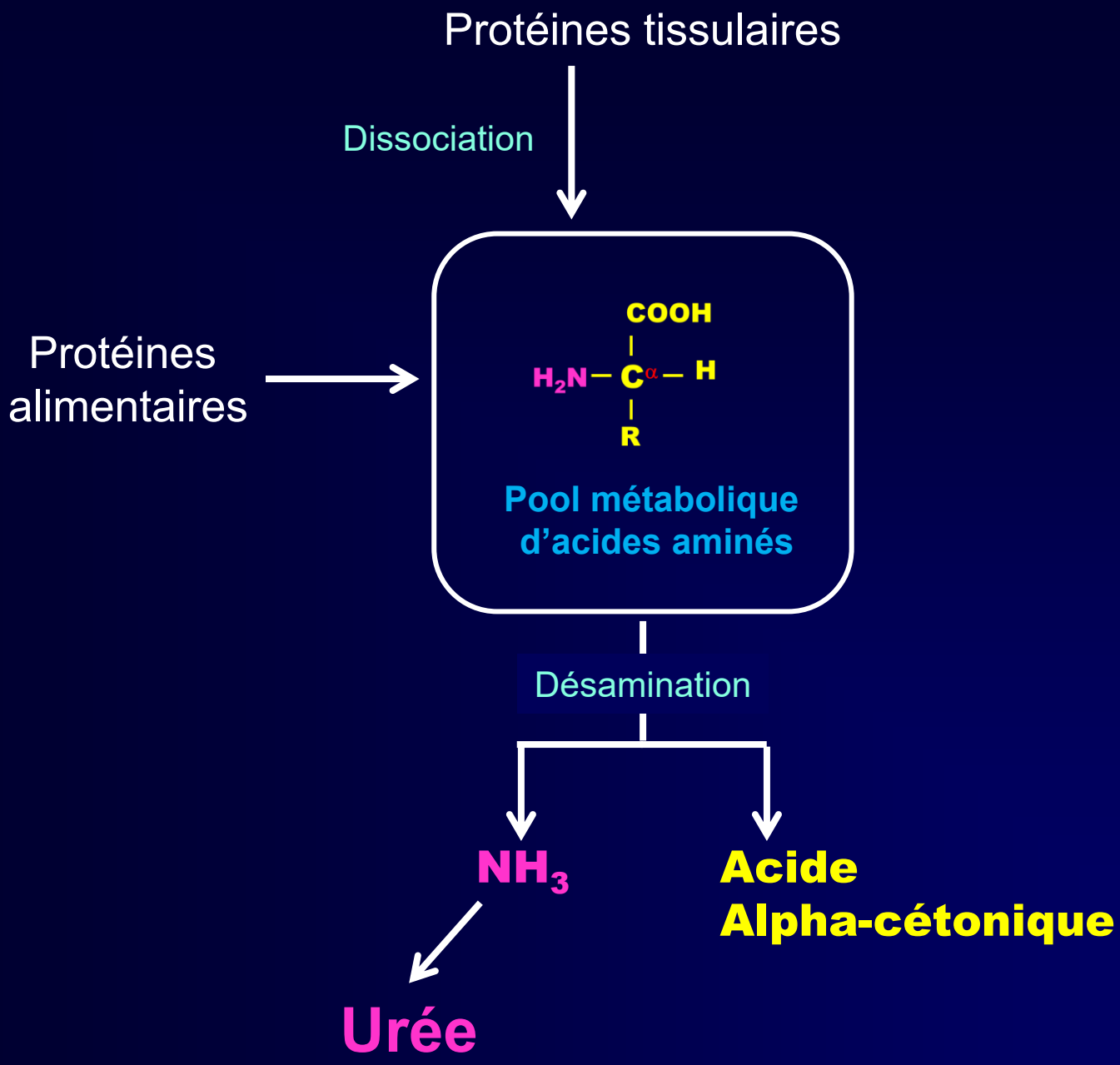
$NH_4^+$

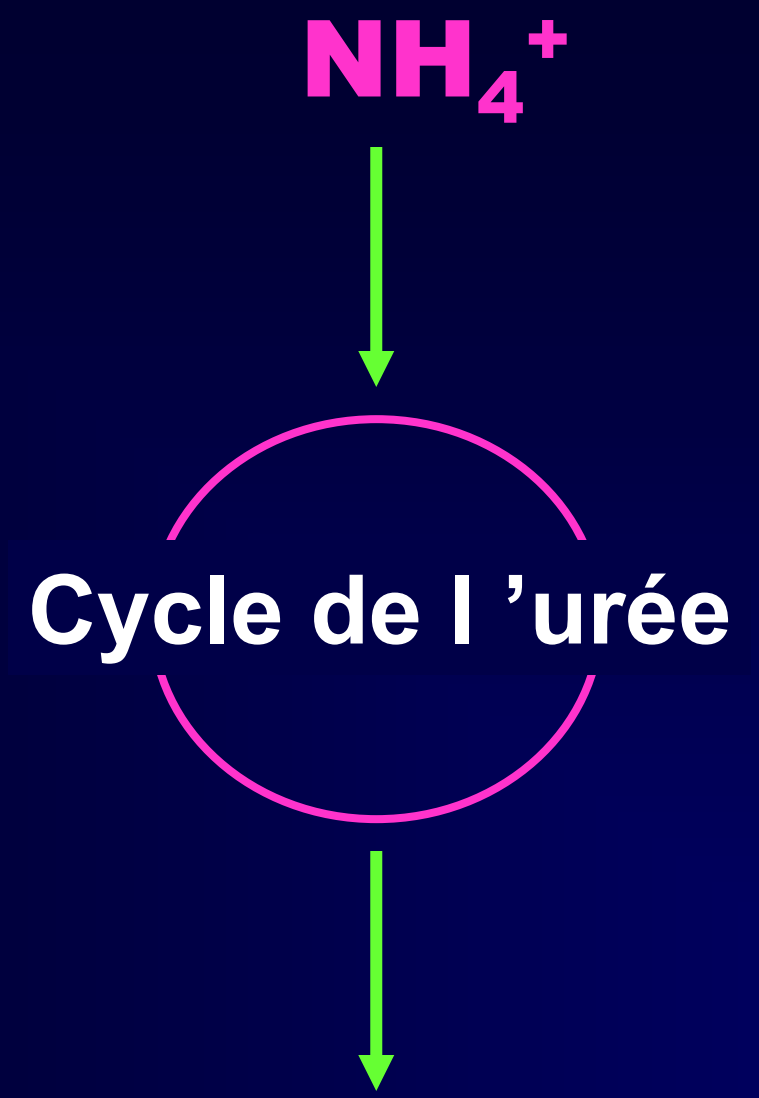
# Glutamate déshydrogénase

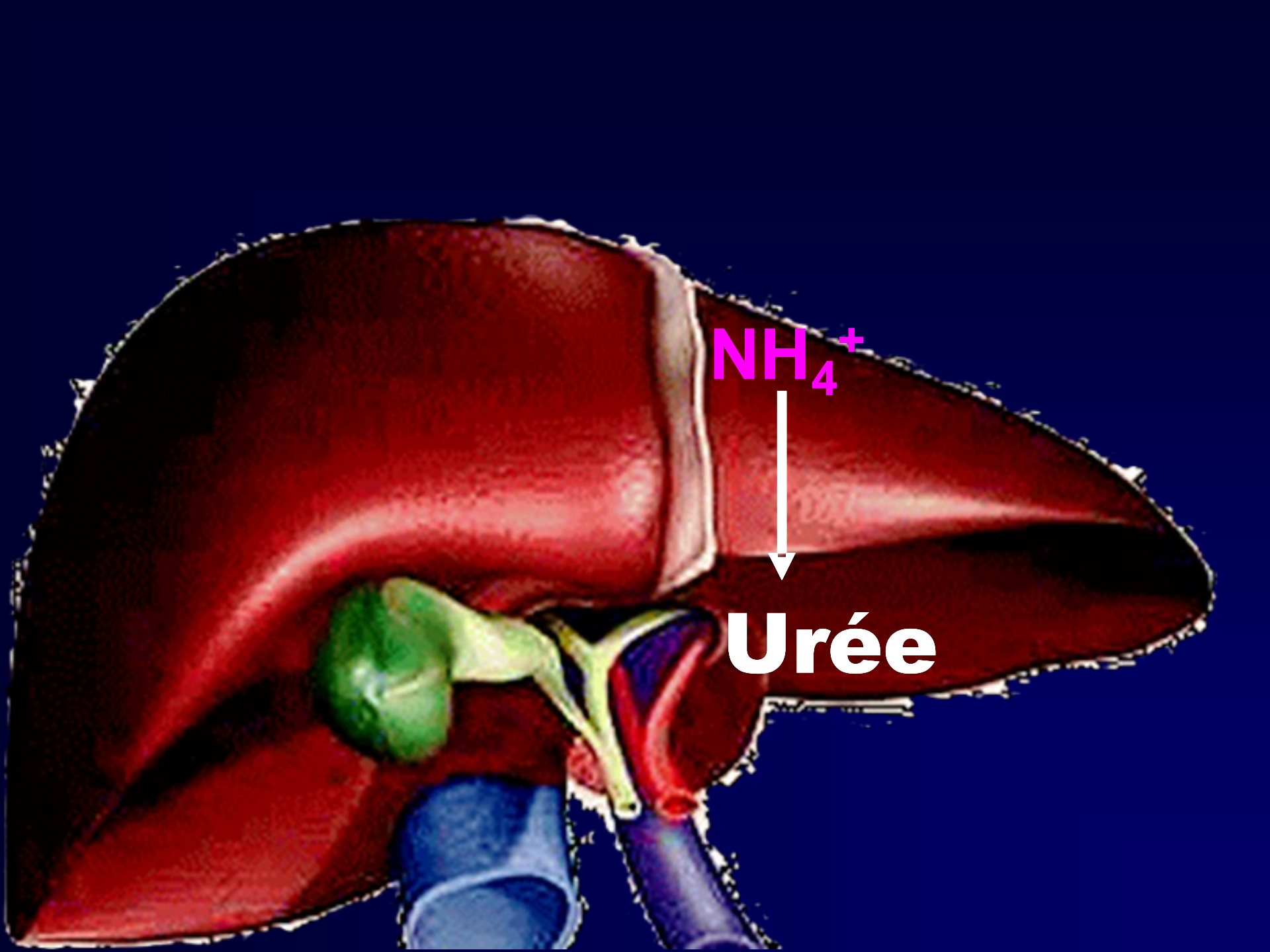



ATP, GTP
NADPH
+ H+
NADP+
α céto-
Glutarate
+ NH4+
Glutamate + H2O
NADH
+ H+
NAD+
ADP, GDP


AA
Glu
Gln
$NH_4^+$
Intestins
(Voie porte)
# Bactéries


Protéines tissulaires
Dissociation
Protéines alimentaires
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Désamination
NH3
Acide Alpha-cétonique
Urée


$NH_4^+$
Cycle de l ’urée
Urée


$NH_4^+$
Urée

Protéine

↓

AαA

αcétoglutarate

↓

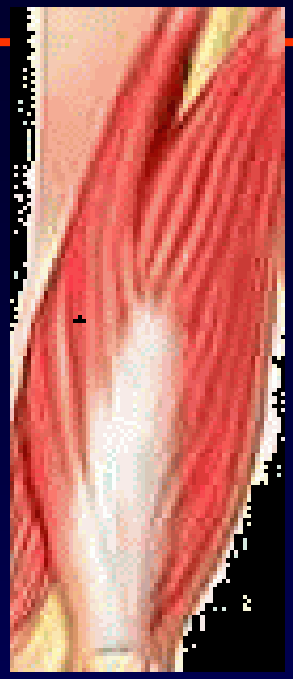
AαC

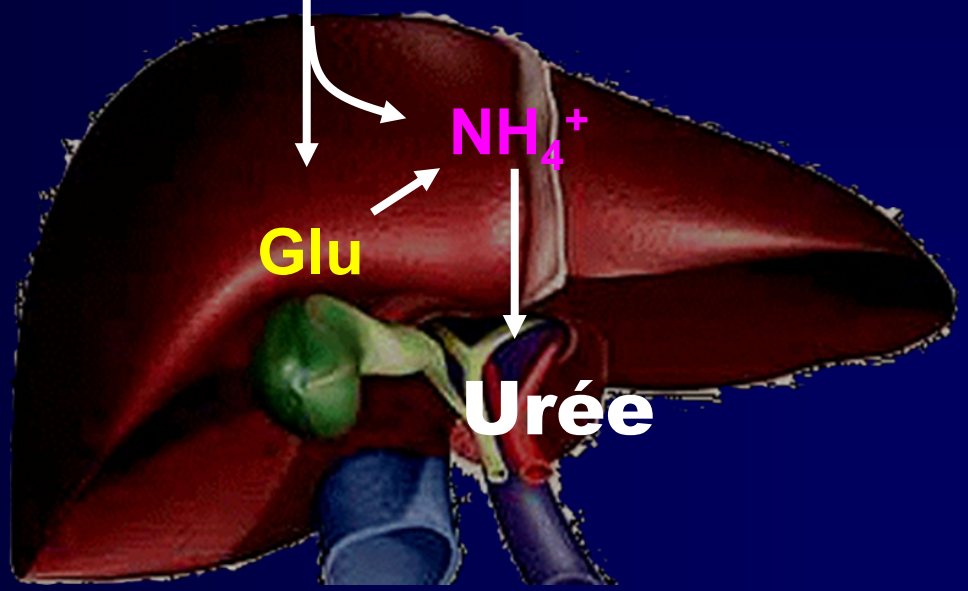
Glu

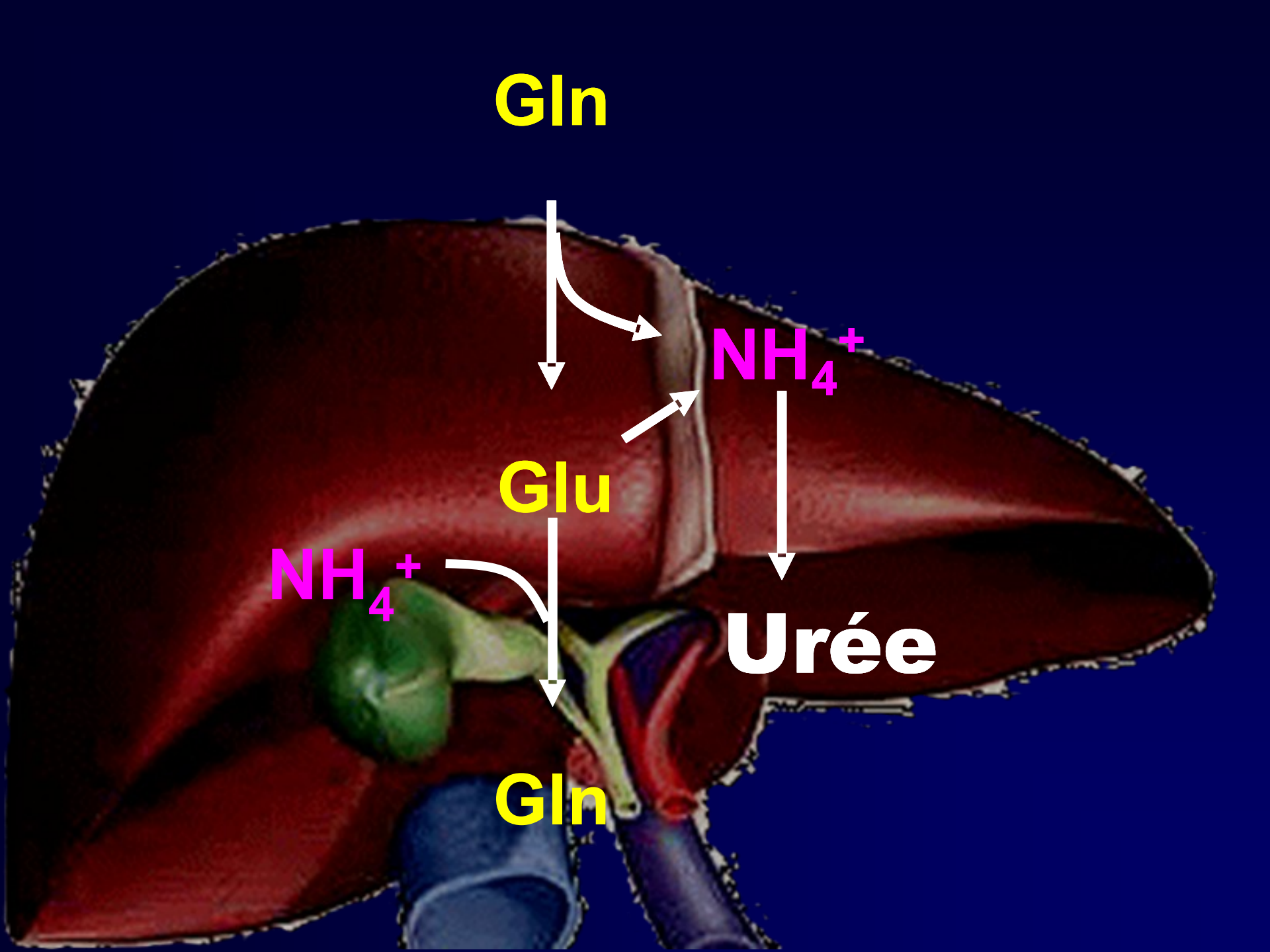
$NH_4^+$

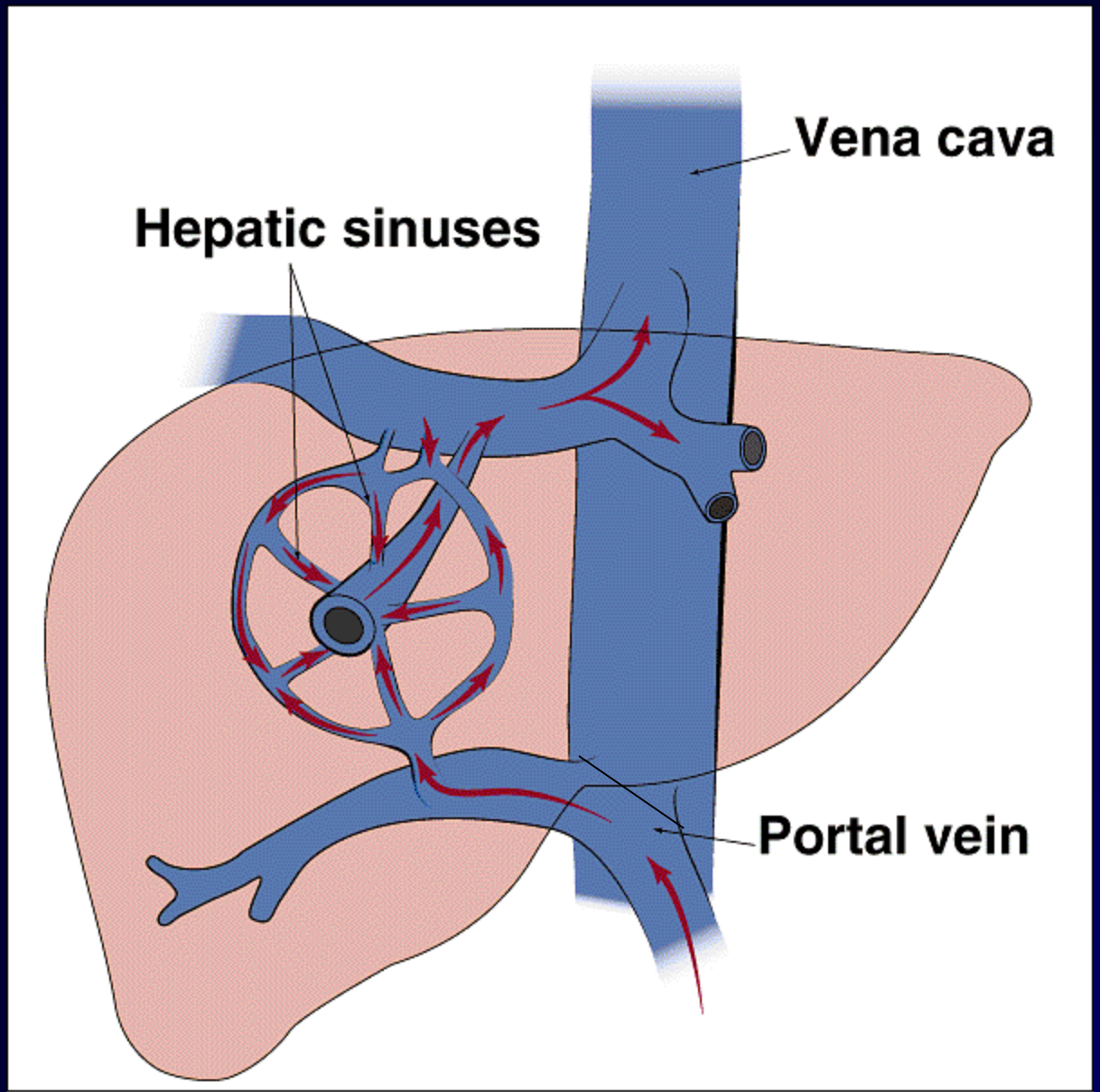
↓

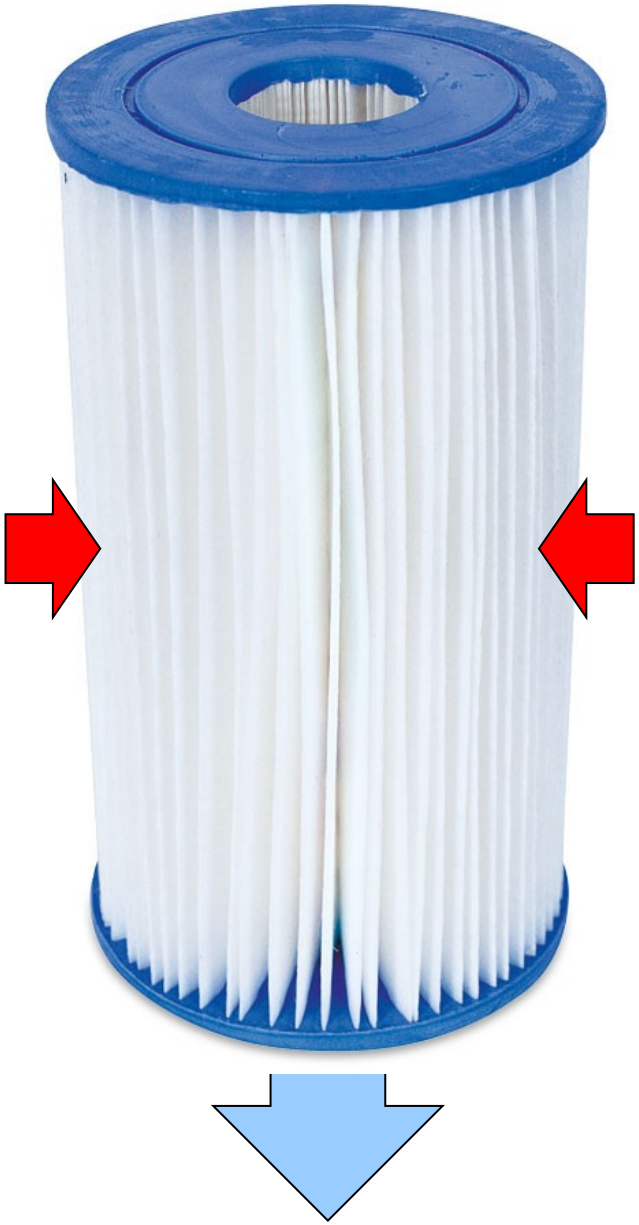
Gln

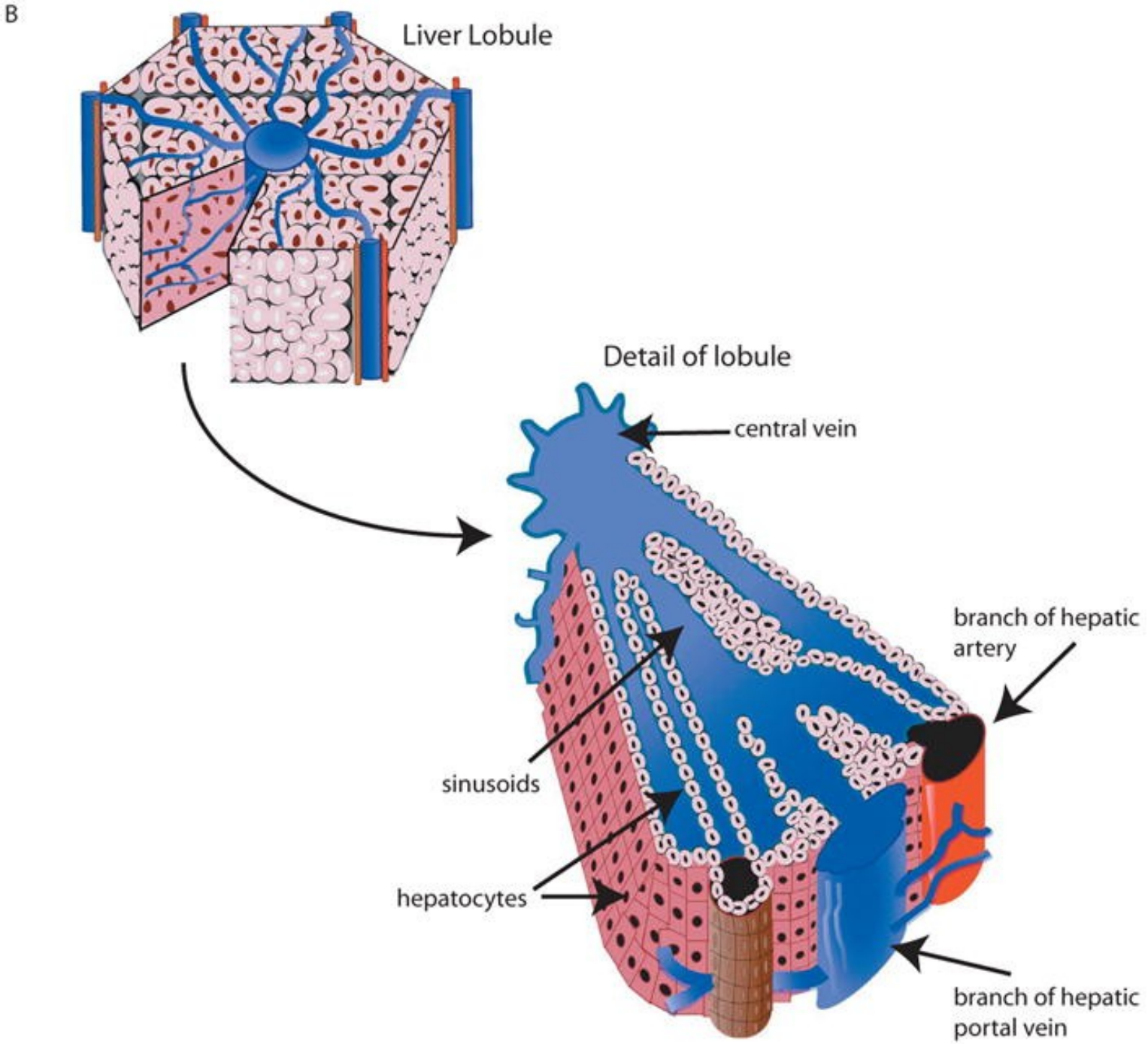




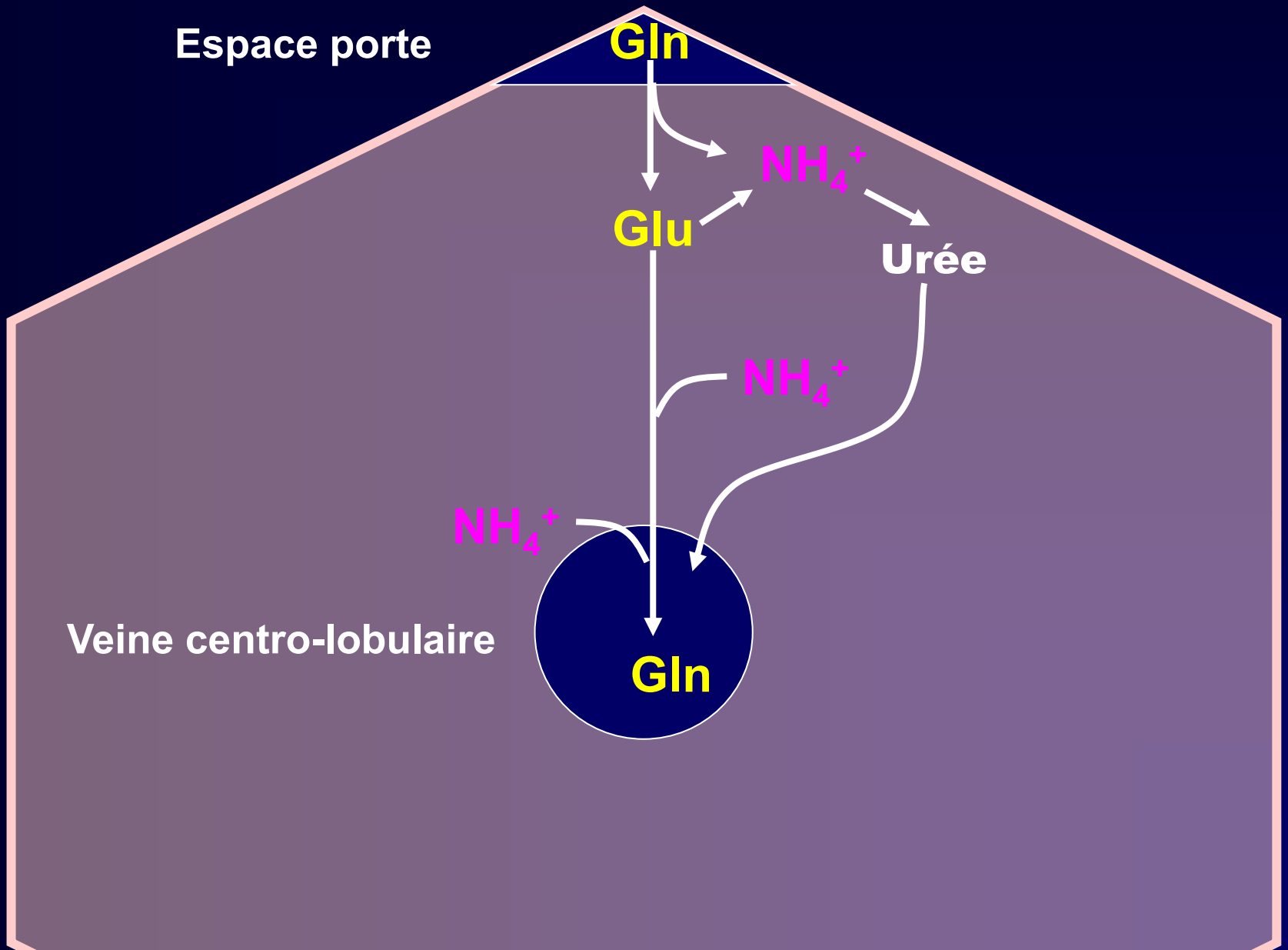

Gln
$NH_4^+$
Glu
$NH_4^+$
Urée
Gln

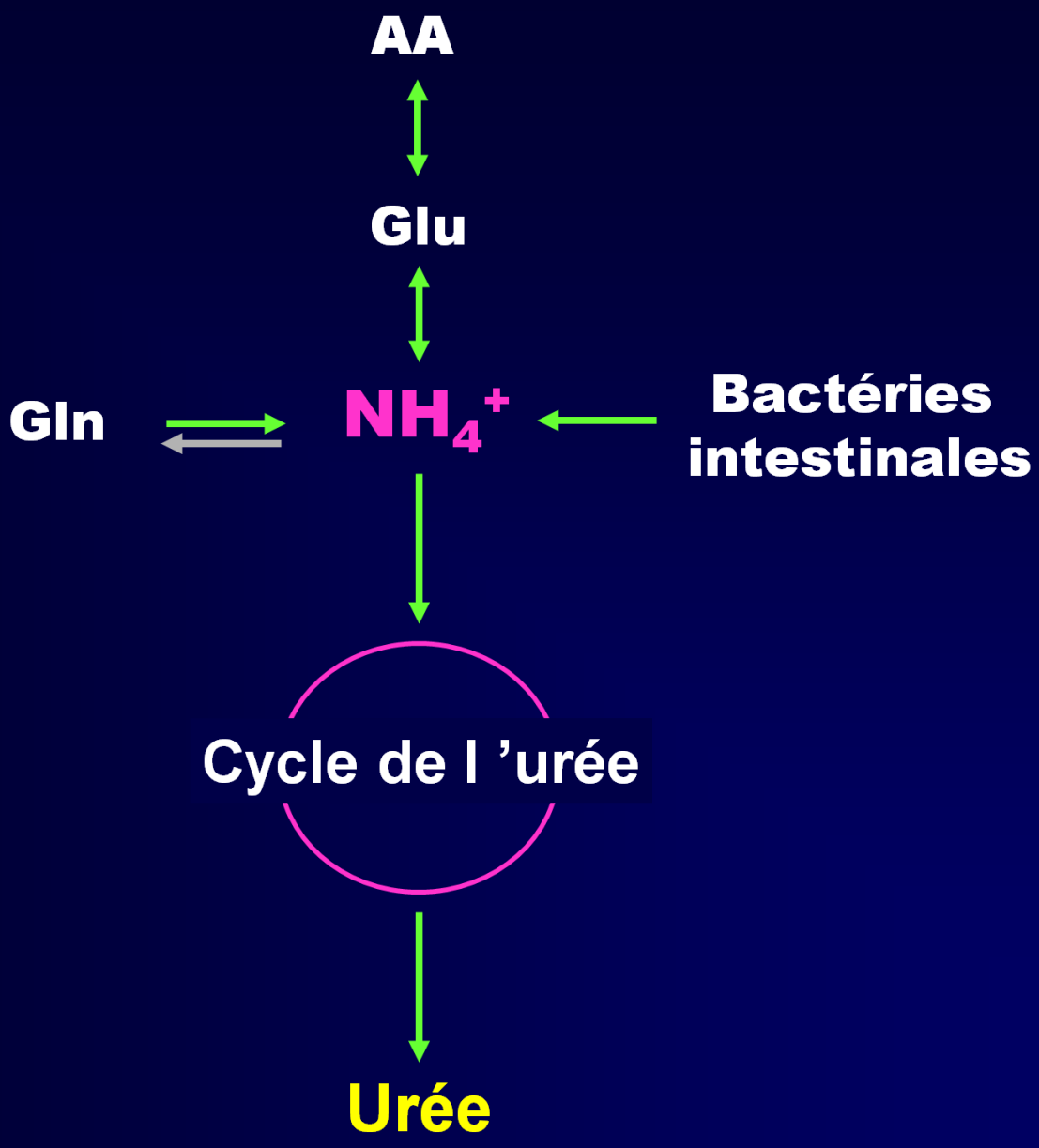





B
Liver Lobule
Detail of lobule
central vein
branch of hepatic artery
sinusoids
hepatocytes
branch of hepatic portal vein

# Lobule Hépatique

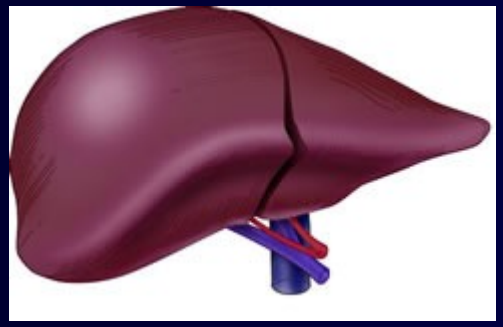



AA
Glu
Gln
$NH_4^+$
Bactéries intestinales
Cycle de l 'urée
Urée

AA

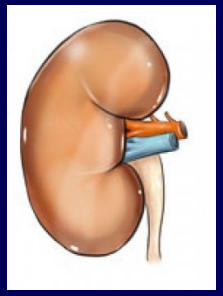
Glu

Gln $\rightleftharpoons$ $NH_4^+$ ← Bactéries intestinales



Cycle de l ’urée

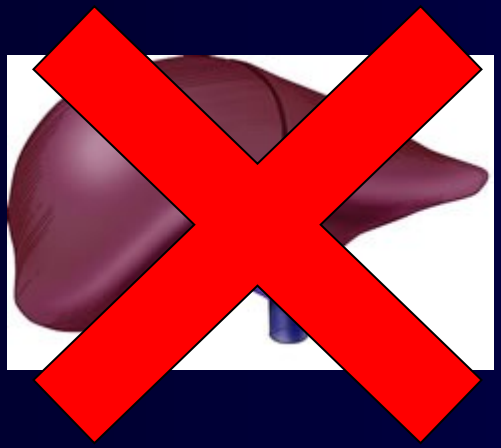
Urée



AA

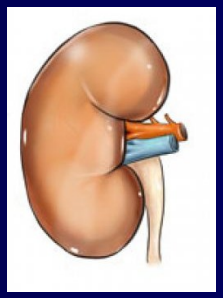
Glu

Gln $\rightleftharpoons$ $NH_4^+$ ← Bactéries intestinales



Cycle de l ’urée

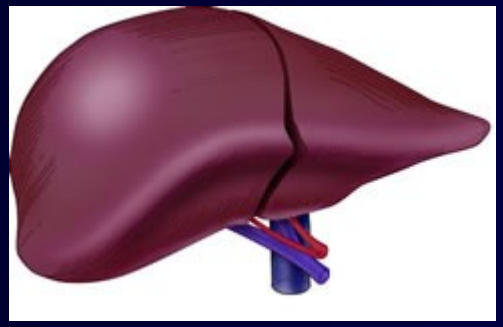
Urée



AA

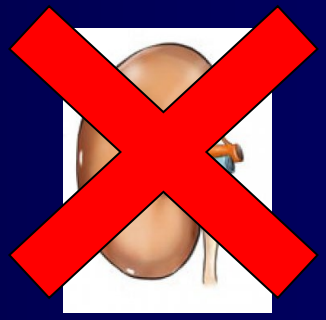
Glu

Gln ⇌ $NH_4^+$ ← Bactéries intestinales



Cycle de l ’urée

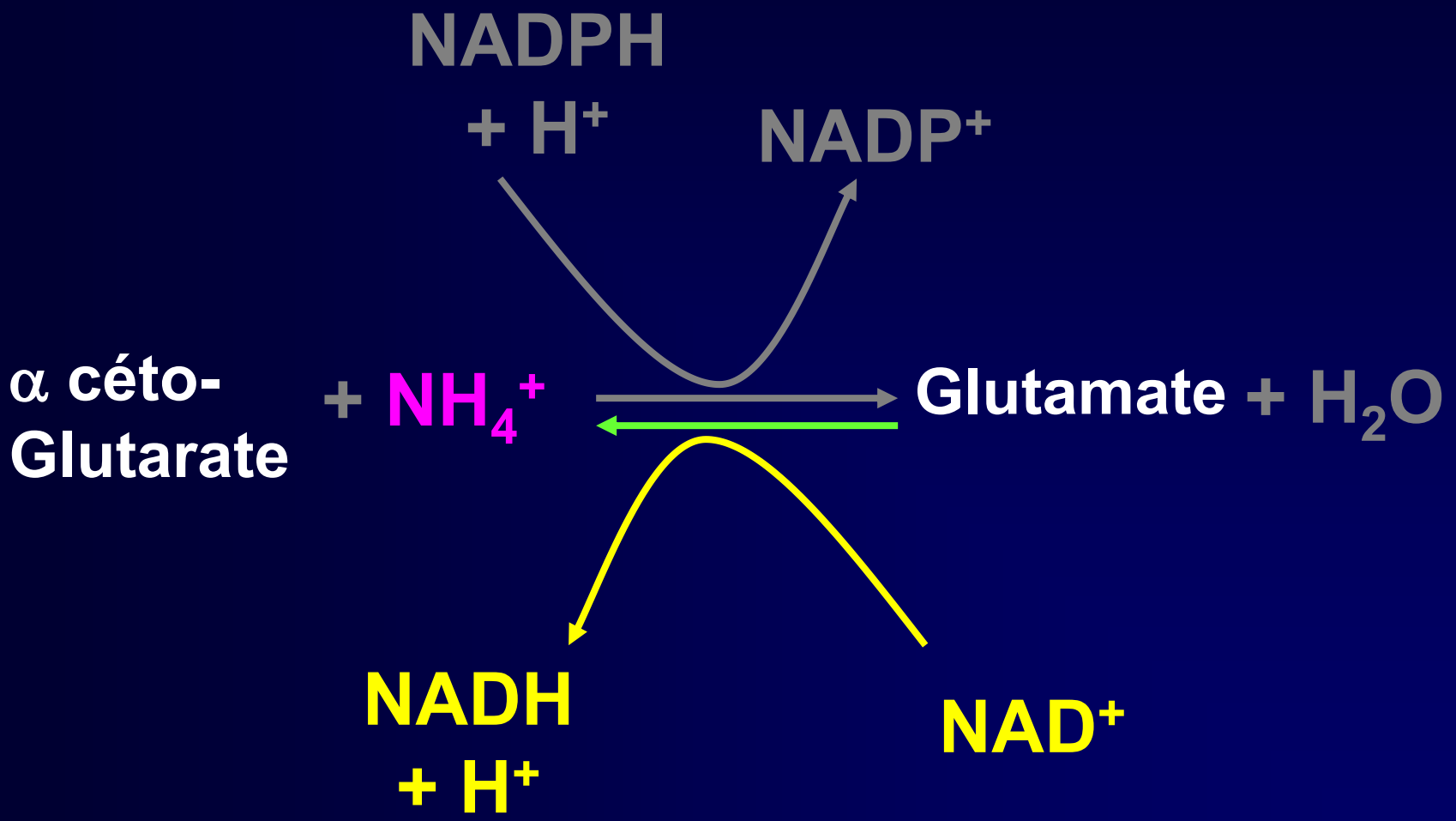
Urée




QUIZ
TIME

- La désamination oxydative est la réaction qui désamine quel acide aminé ?

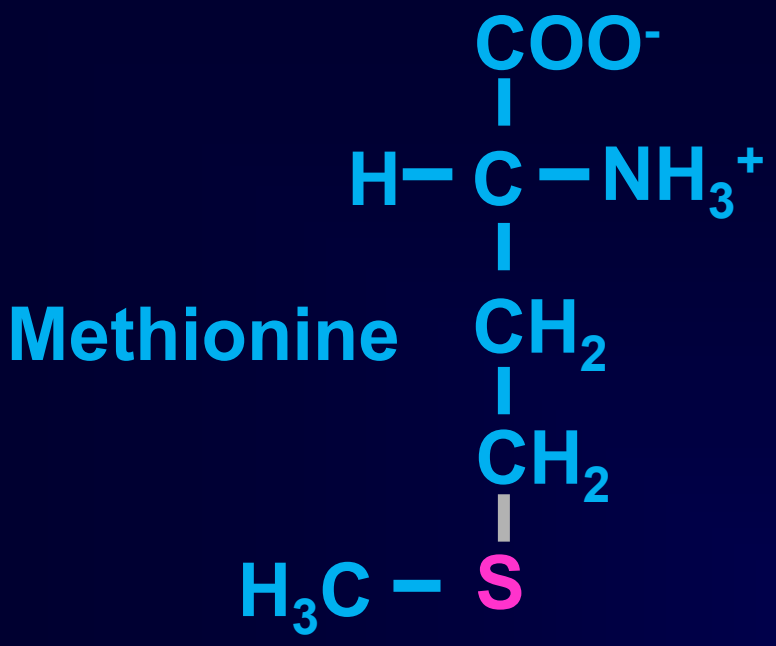
# Glutamate déshydrogénase



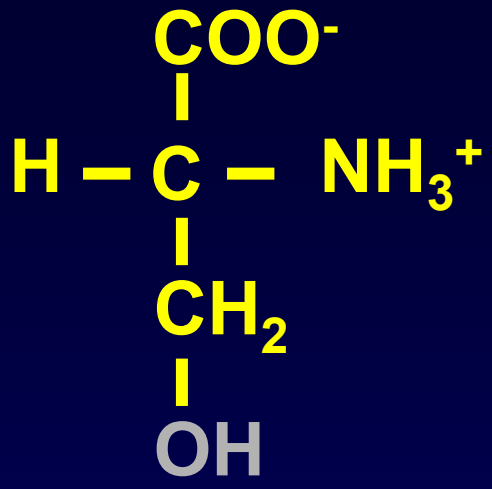
- La désamination oxydative est la réaction qui désamine quel acide aminé ?

- Le glutamate

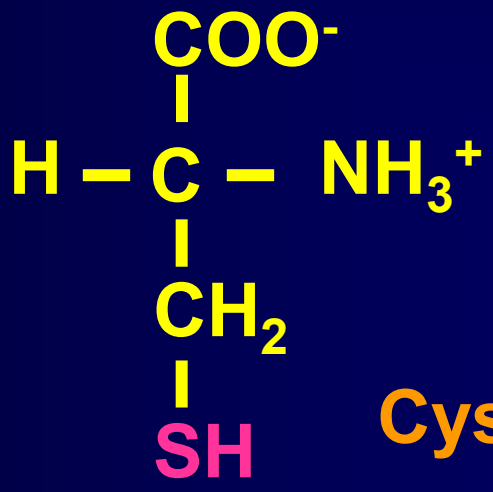
- Quels acides aminés sont précurseurs de la cystéine ?




Methionine




sérine




Cystéine

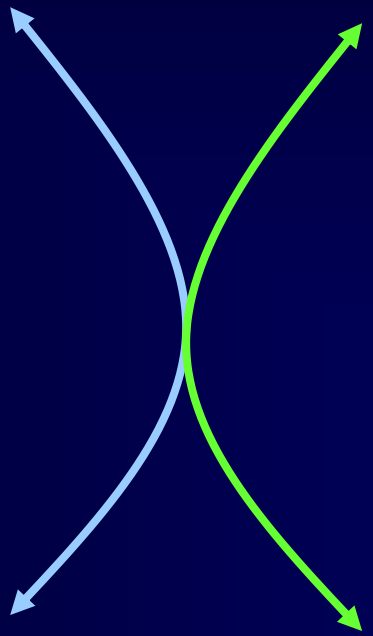
- Quels acides aminés sont précurseurs de la cystéine ?

- La méthionine & la sérine

- Le pyruvate, après transamination, permet la synthèse de quel acide aminé ?

Pyruvate

Glu



Ala

α céto-
Glutarate

- Le pyruvate, après transamination, permet la synthèse de quel acide aminé ?

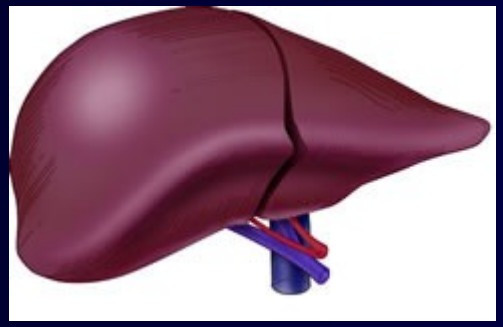
- **Alanine**

- Chez l’espèce humaine, sous quelle forme majoritaire, irréversible et atoxique circule l’azote aminé dans le sang ?
- **Urée**

AA

Glu

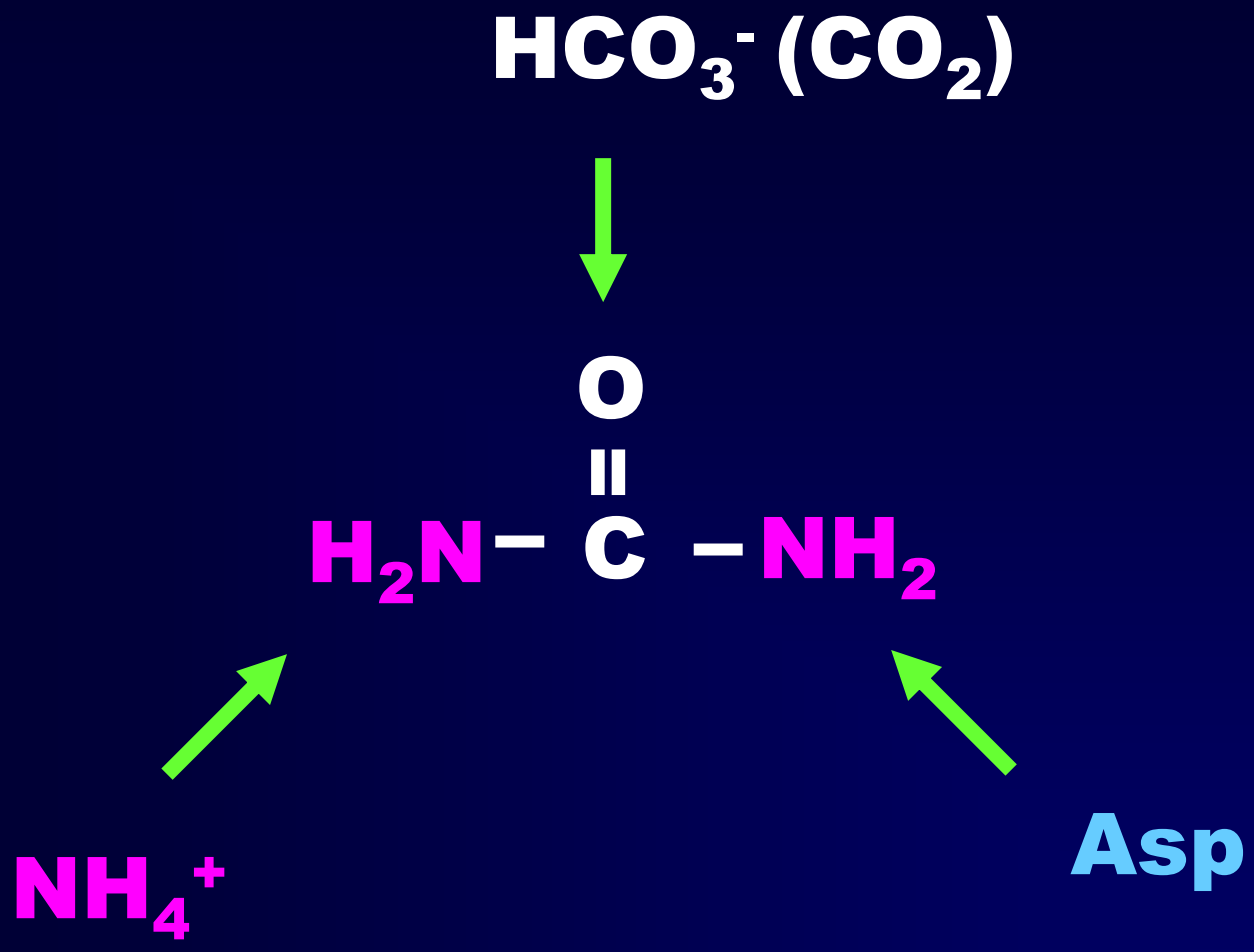
Gln ⇌ $NH_4^+$ ← Bactéries intestinales

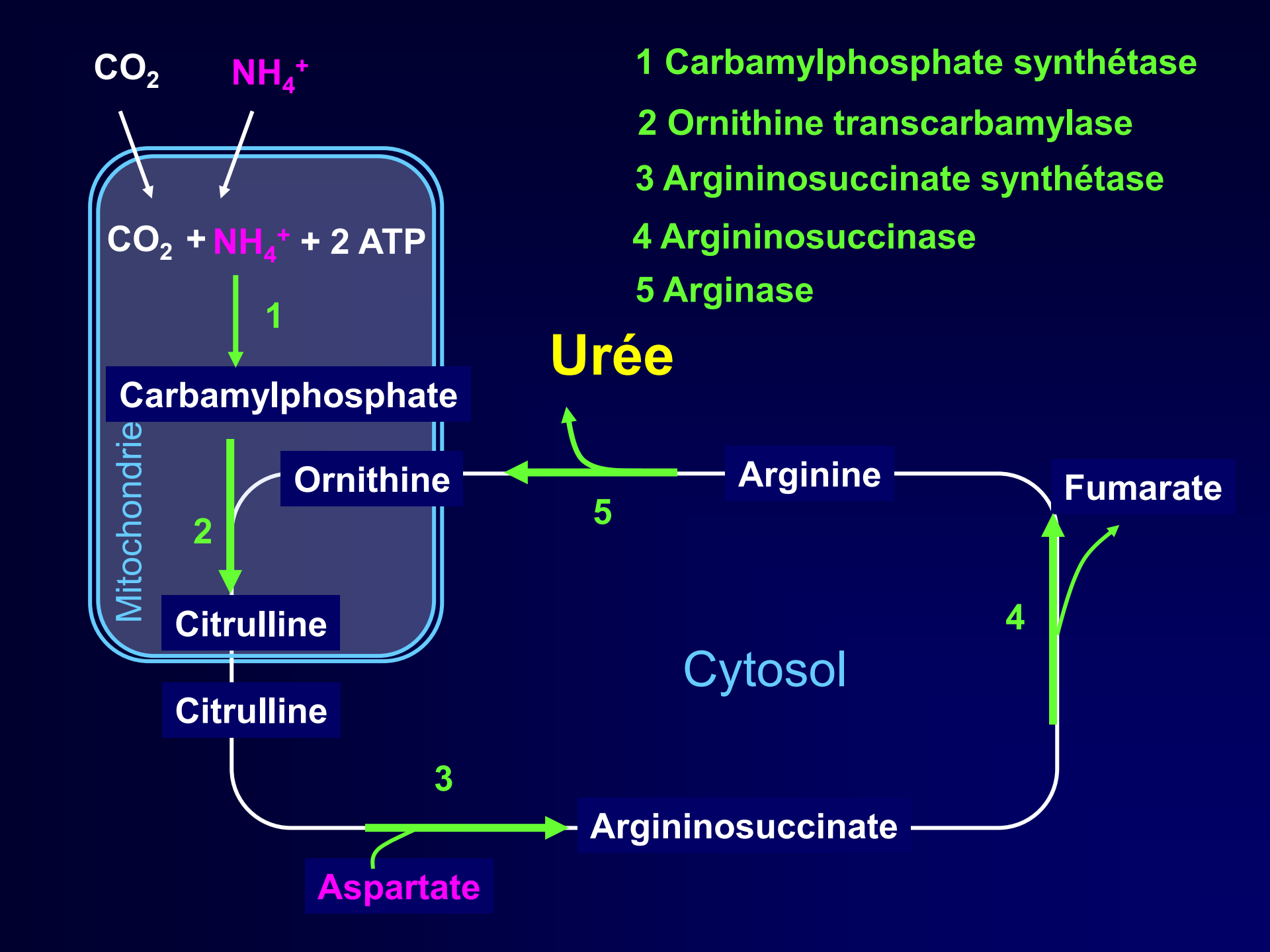


Cycle de l ’urée

Urée

# Cycle de l ’urée


$HCO_3^-$ ($CO_2$)
O
$H_2N-C-NH_2$
$NH_4^+$
Asp


$CO_2$
$NH_4^+$
1 Carbamylphosphate synthétase
2 Ornithine transcarbamylase
3 Argininosuccinate synthétase
4 Argininosuccinase
5 Arginase
$CO_2 + NH_4^+ + 2$ ATP
1
Carbamylphosphate
Mitochondrie
Ornithine
2
Citrulline
Urée
Arginine
5
Fumarate
4
Cytosol
Citrulline
3
Argininosuccinate
Aspartate

# Cycle de l ’urée

Cycle de ~~l'urée~~
l'Ornithine

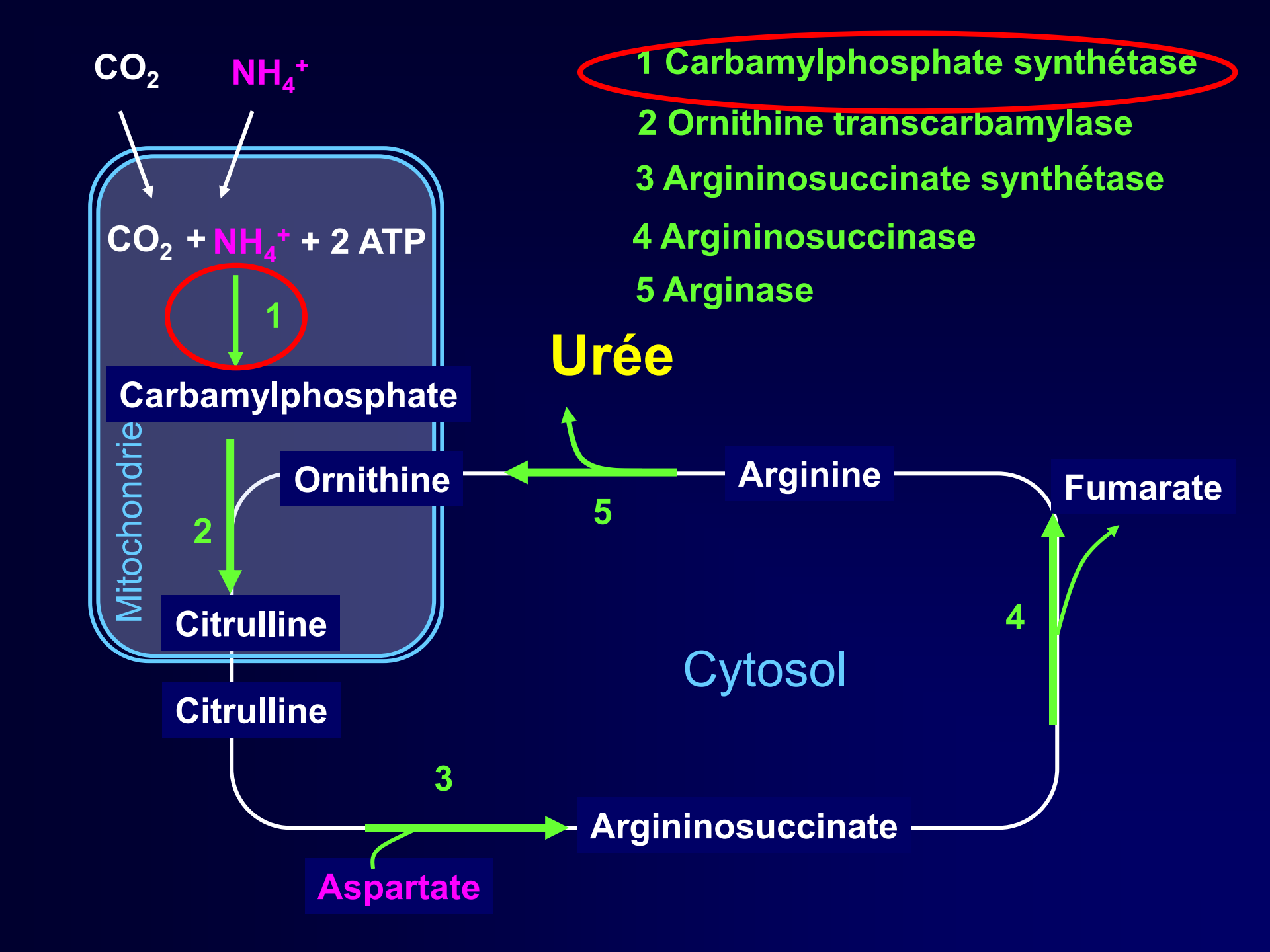
# Régulation du Cycle de l ’urée

**Allostérique**

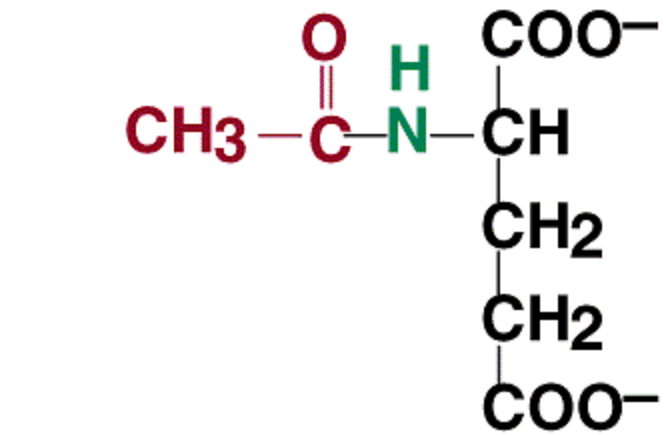
**+**

**Inductible**

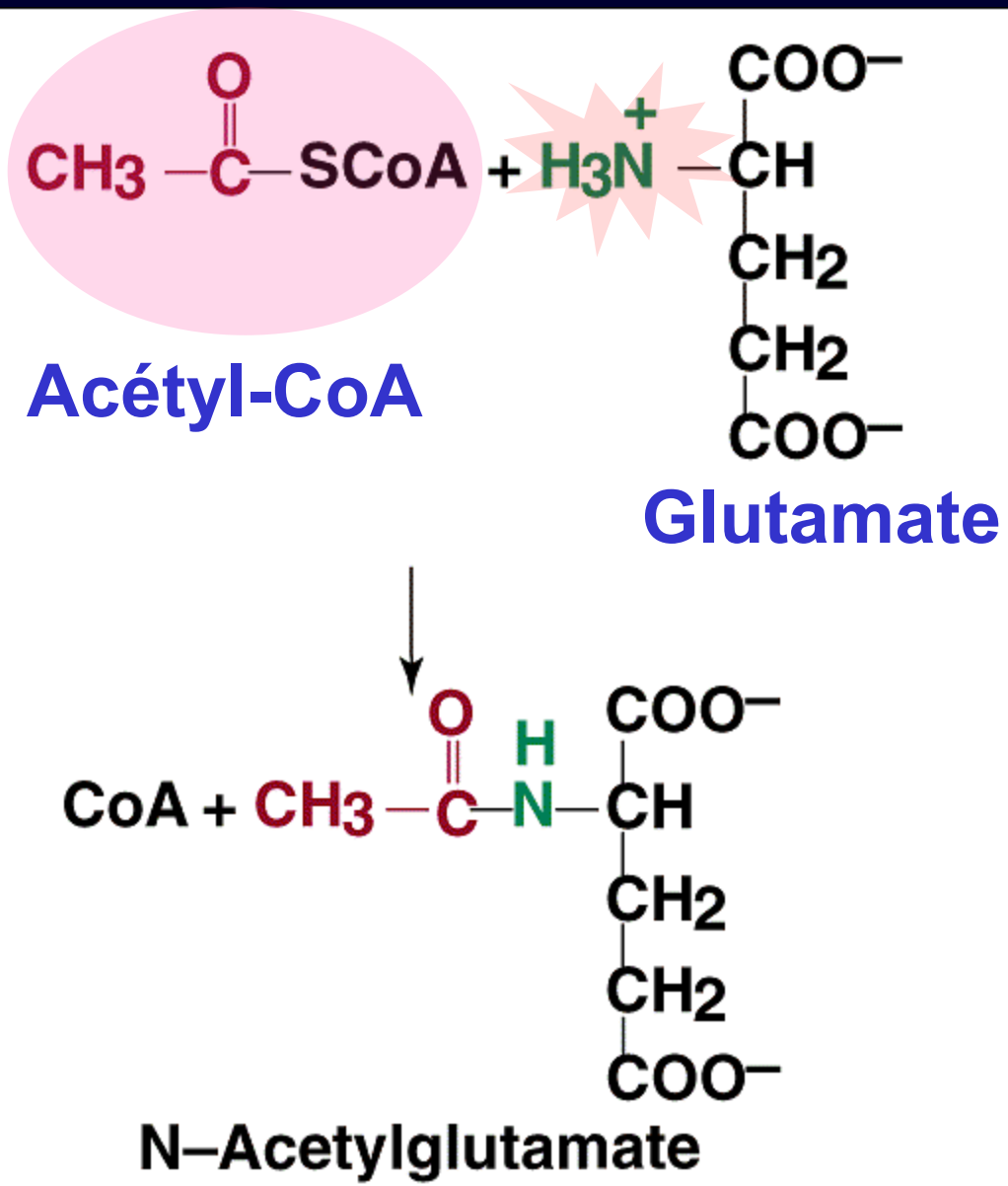
# Allostérique


$CO_2$
$NH_4^+$
1 Carbamylphosphate synthétase
2 Ornithine transcarbamylase
3 Argininosuccinate synthétase
4 Argininosuccinase
5 Arginase
$CO_2$ + $NH_4^+$ + 2 ATP
1
Carbamylphosphate
Mitochondrie
Ornithine
2
Citrulline
Urée
5
Arginine
Fumarate
4
Cytosol
Citrulline
3
Argininosuccinate
Aspartate

N–Acetylglutamate




N–Acetylglutamate


CH3 – C – SCoA + H3N – CH
O
+
COO–
CH2
CH2
COO–
Acétyl-CoA
Glutamate
CoA + CH3 – C – N – CH
O
H
COO–
CH2
CH2
COO–
N–Acetylglutamate

# Inductible

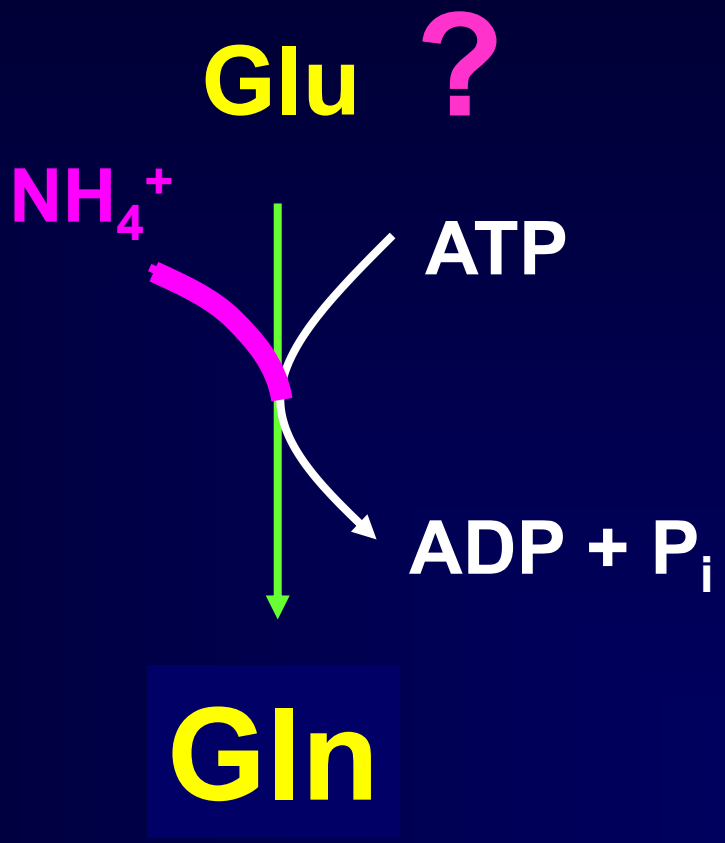
# Concentrations des enzymes

# Hyperammoniémies

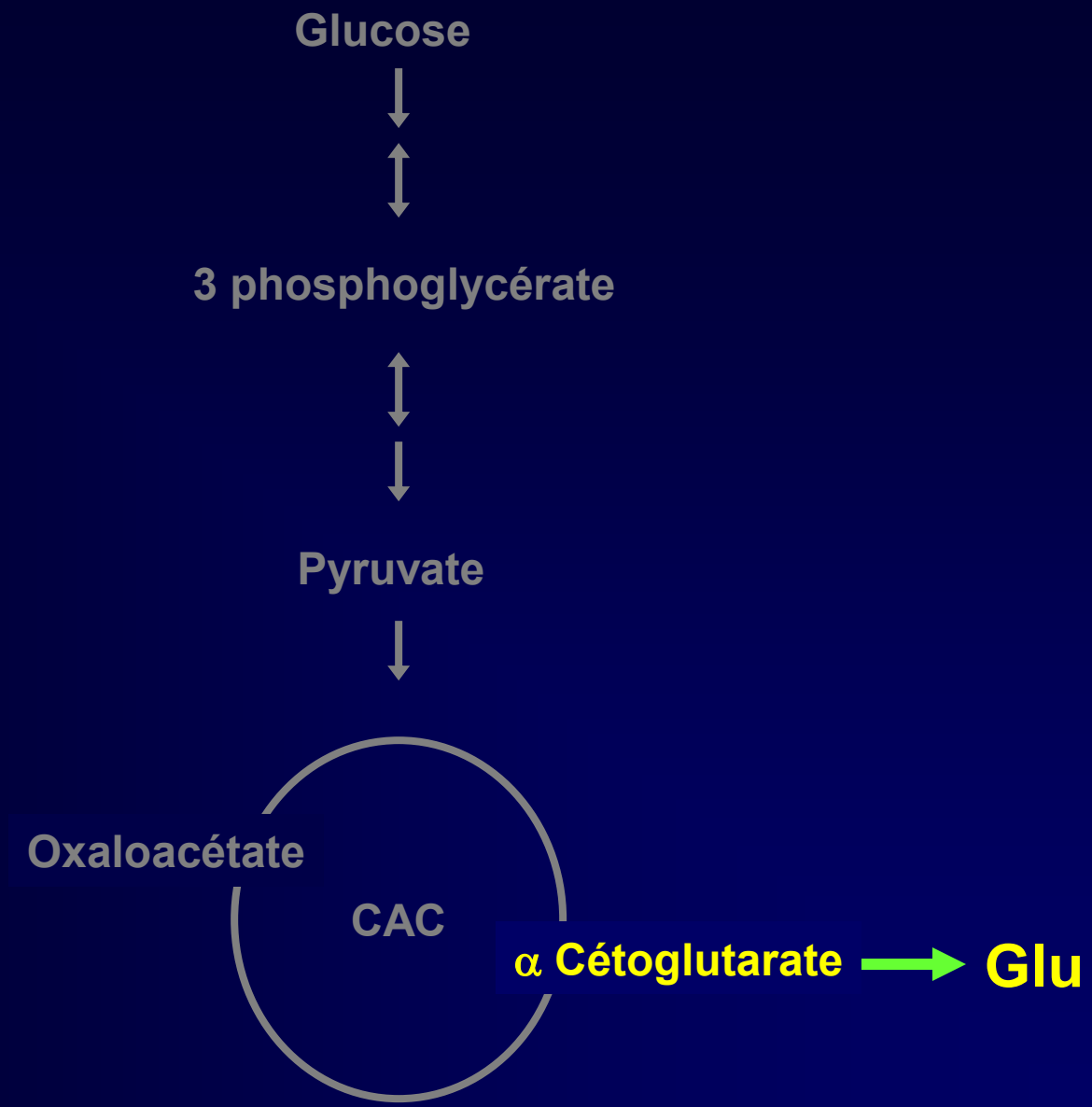
$[NH_4^+]_{plasma}$ > 40 µmol/L


TOXIC

# Glutamine synthétase

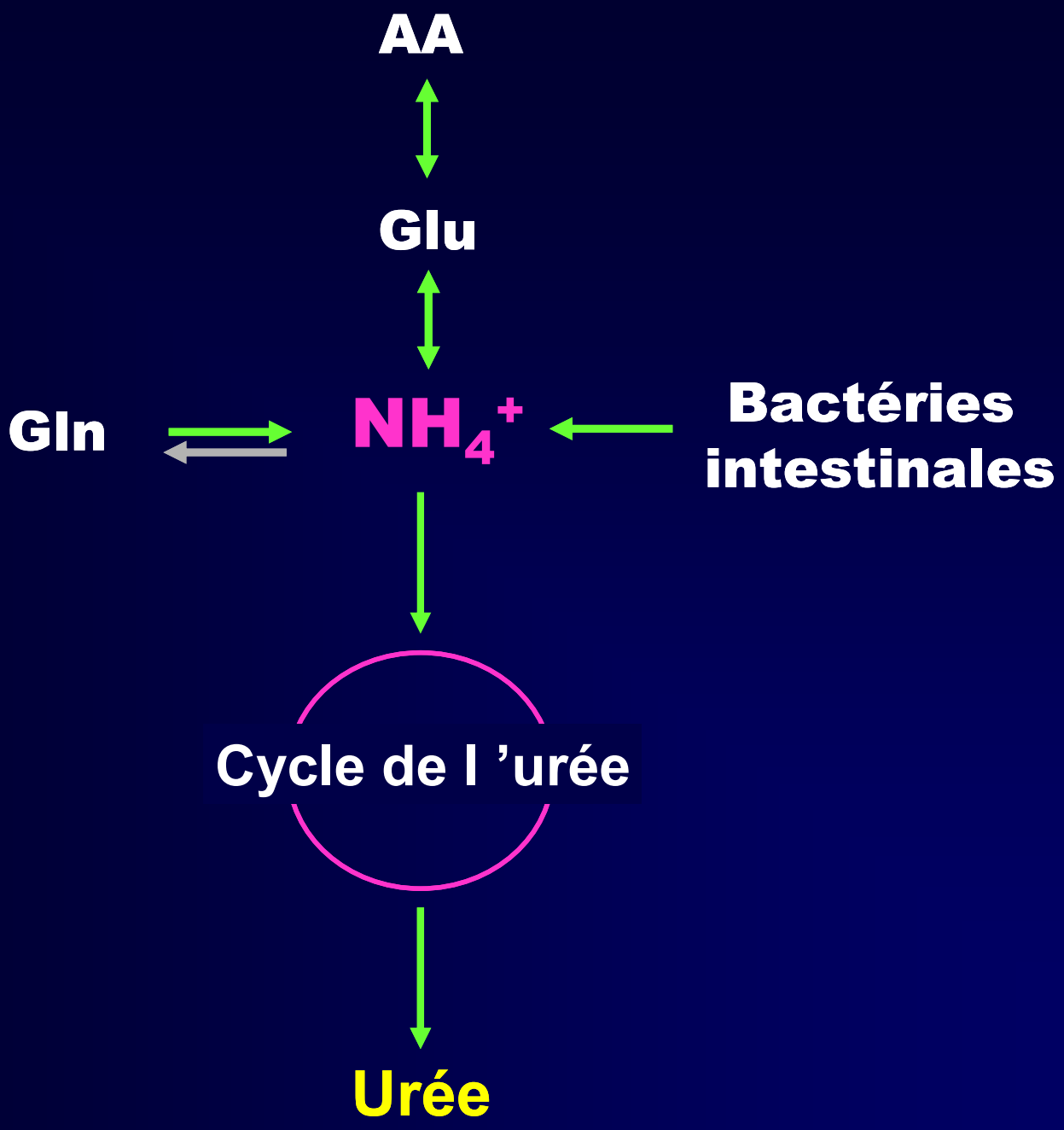


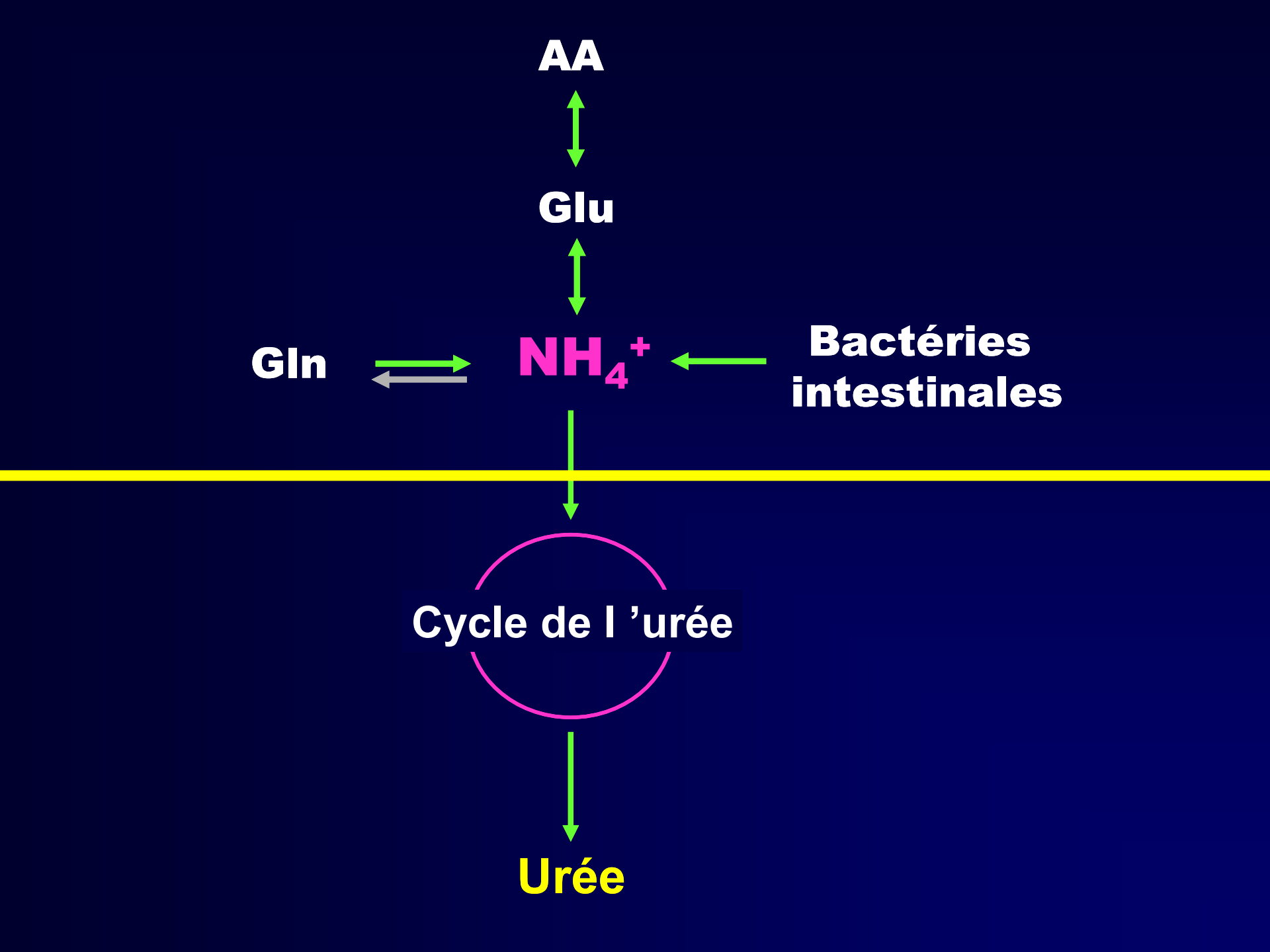

**Effet osmotique**

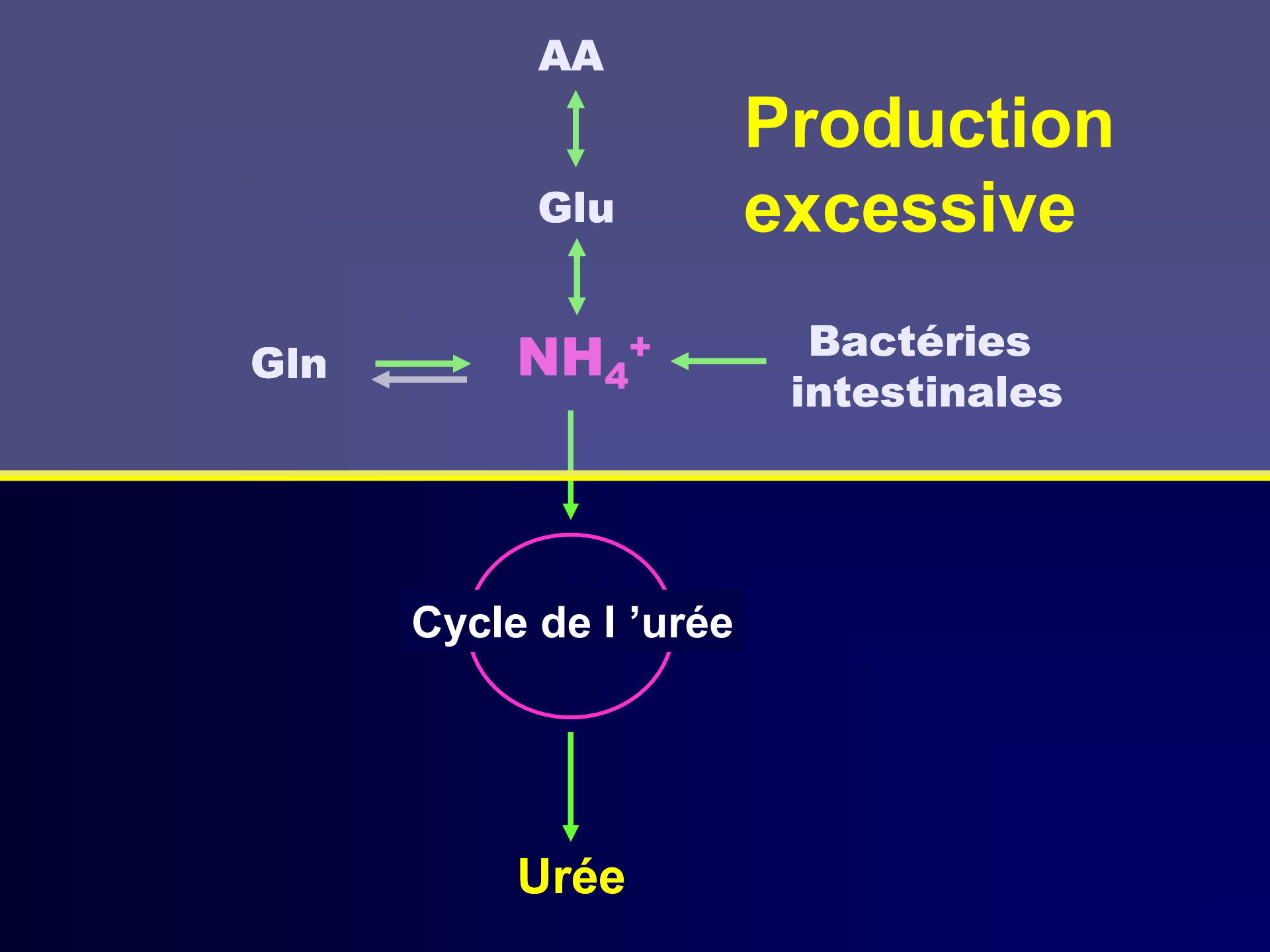

Glucose
3 phosphoglycérate
Pyruvate
Oxaloacétate
CAC
α Cétoglutarate
Glu

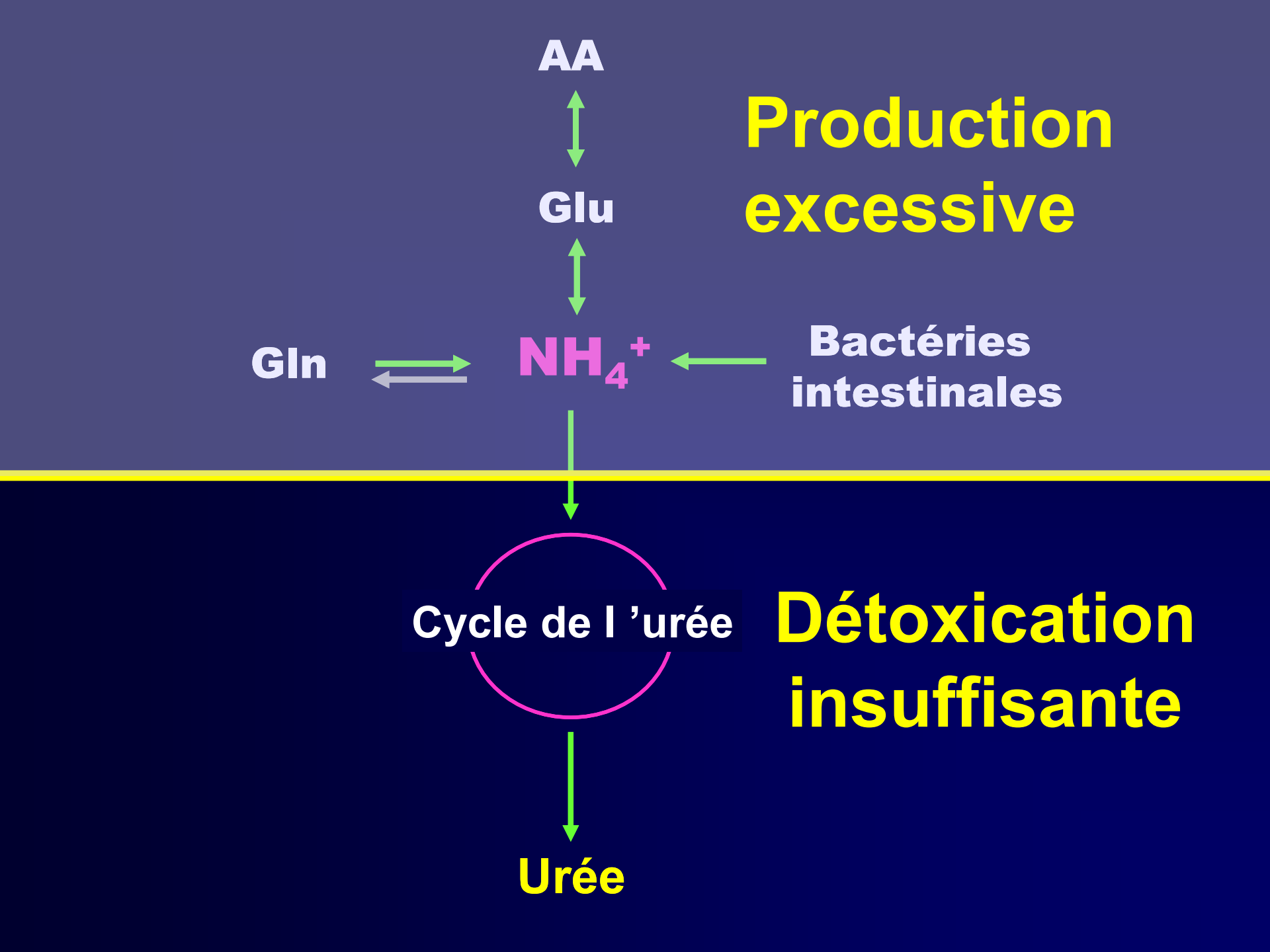
# Hyperammoniémies

Mécanismes


AA
Glu
Gln
$NH_4^+$
Bactéries intestinales
Cycle de l 'urée
Urée


AA
Glu
Gln
$NH_4^+$
Bactéries intestinales
Cycle de l 'urée
Urée


AA
Glu
Gln
$NH_4^+$
Bactéries intestinales
Production excessive
Cycle de l 'urée
Urée


AA
Glu
Gln
$NH_4^+$
Bactéries intestinales
Production excessive
Cycle de l 'urée
Détoxication insuffisante
Urée

# Hyperammoniémies

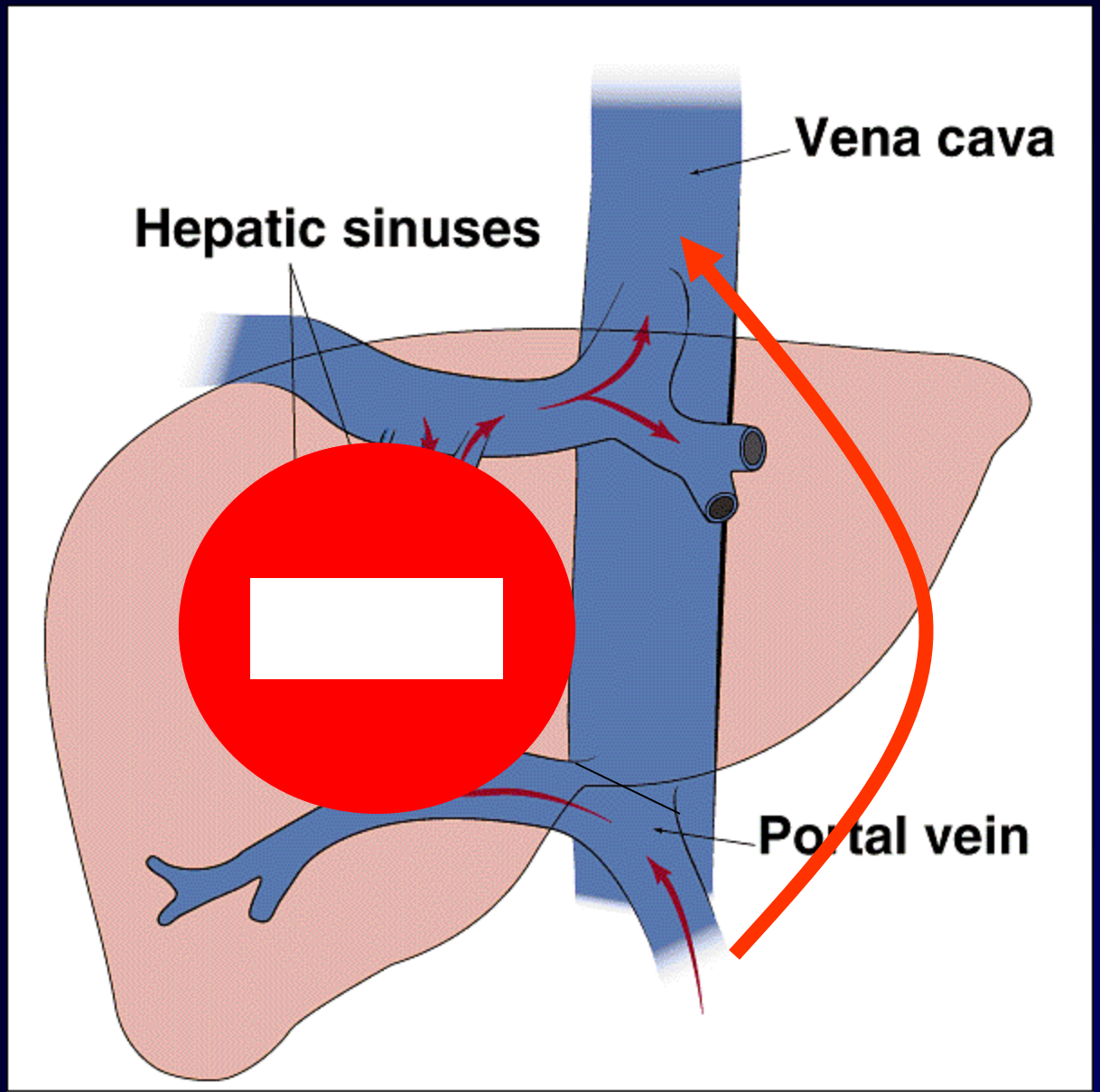
**Acquises**

**Primitives**

# Hyperammoniémies

**Acquises**

Primitives



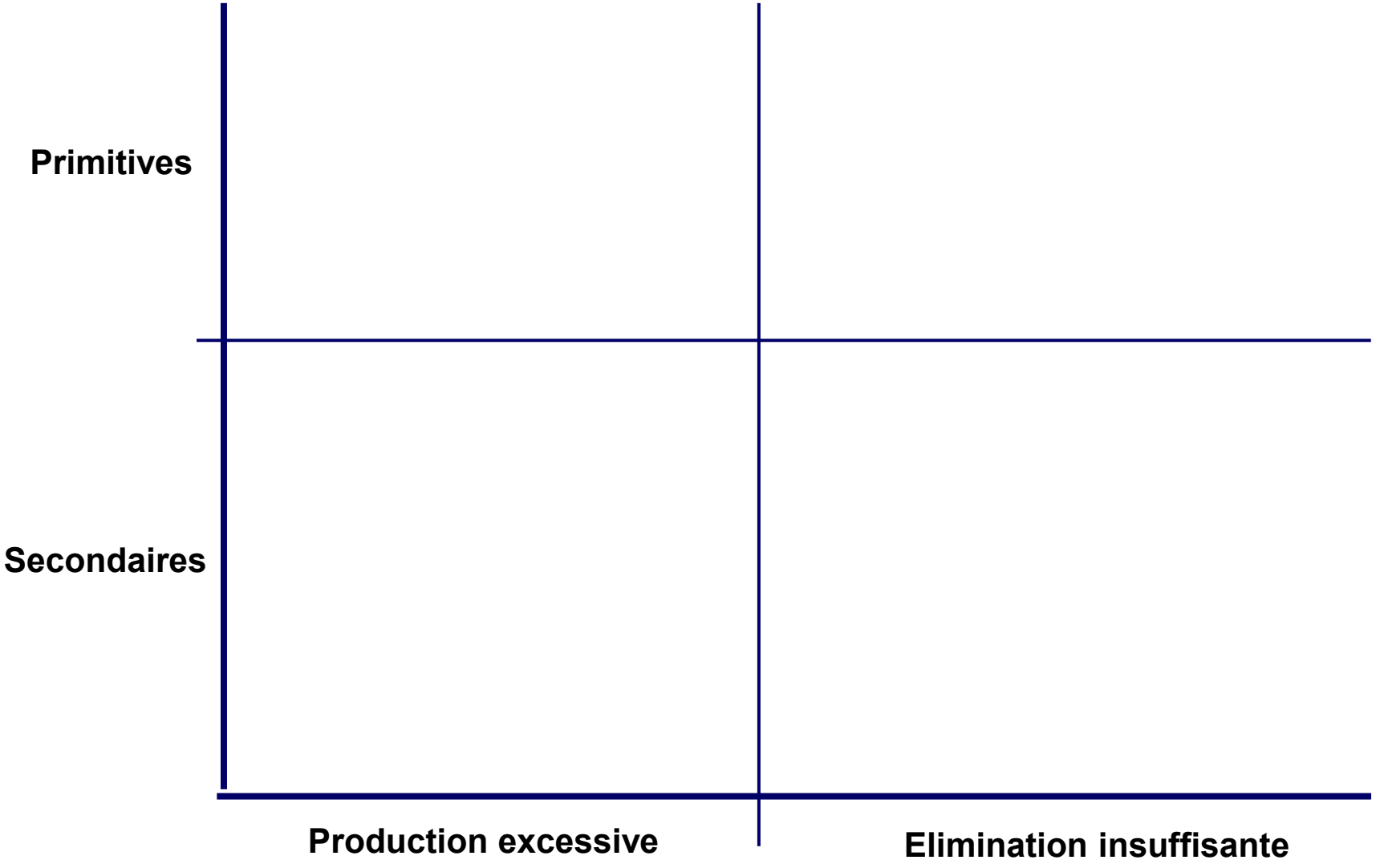
# Hyperammoniémies

Acquises

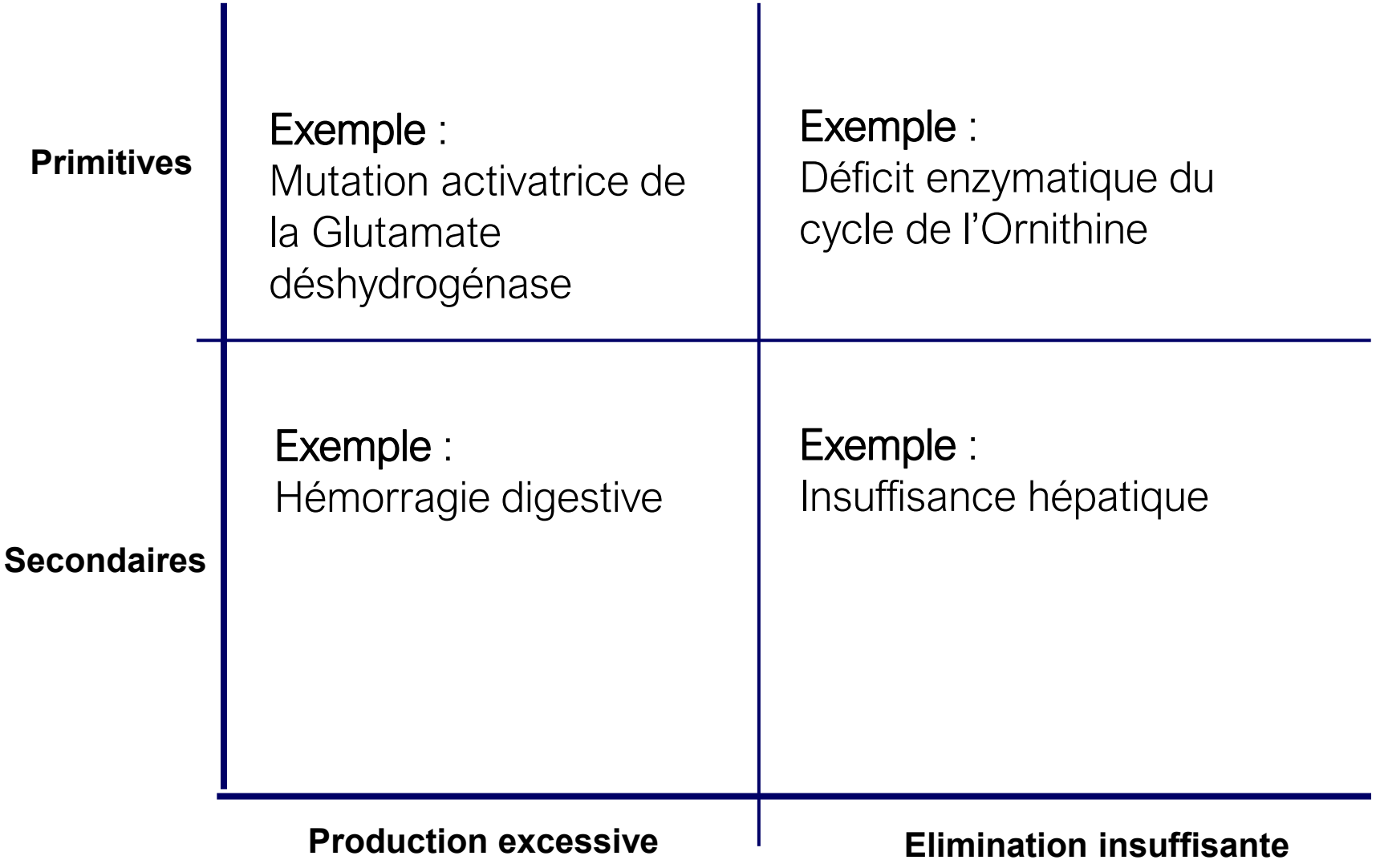
**Primitives**

| | |
|---|---|
| Hyperammoniémie de type 1 | déficit en carbamylphosphate synthétase |
| Hyperammoniémie de type 2 | déficit en Ornithine transcarbamylase |
| Citrullinémie | déficit en arginosuccinate synthétase |
| Acidurie argininosuccinique | déficit en argininosuccinase |
| Hyperargininémie | déficit en Arginase |

# Hyperammoniémies



# Hyperammoniémies

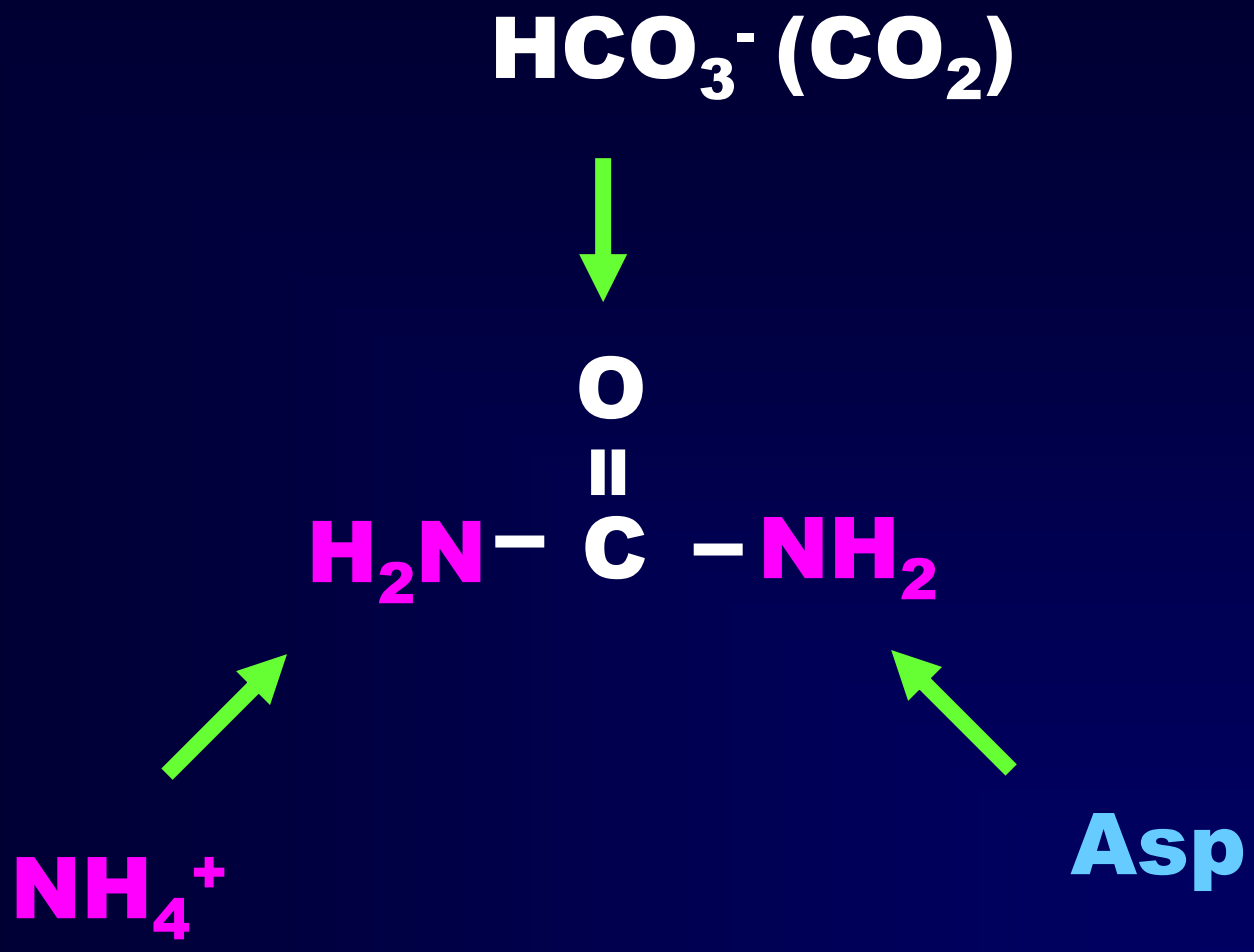


# Traitement des hyperammoniémies

?


QUIZ
TIME

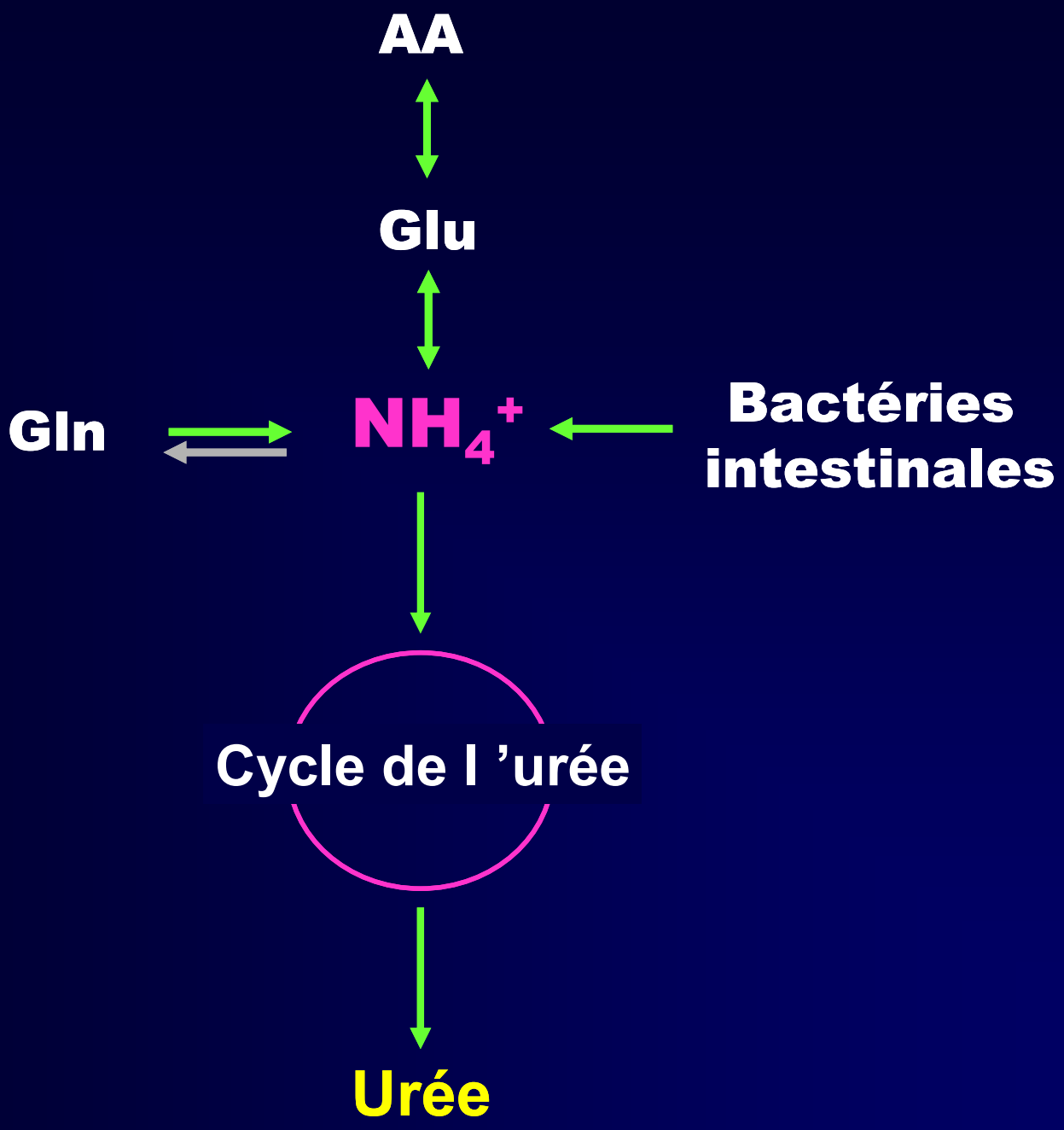
- L’Urée permet d’éliminer combien d’azote aminé ?


$HCO_3^-$ ($CO_2$)
O
$H_2N-C-NH_2$
$NH_4^+$
Asp

- L’Urée permet d’éliminer combien d’azote aminé ?

- 2

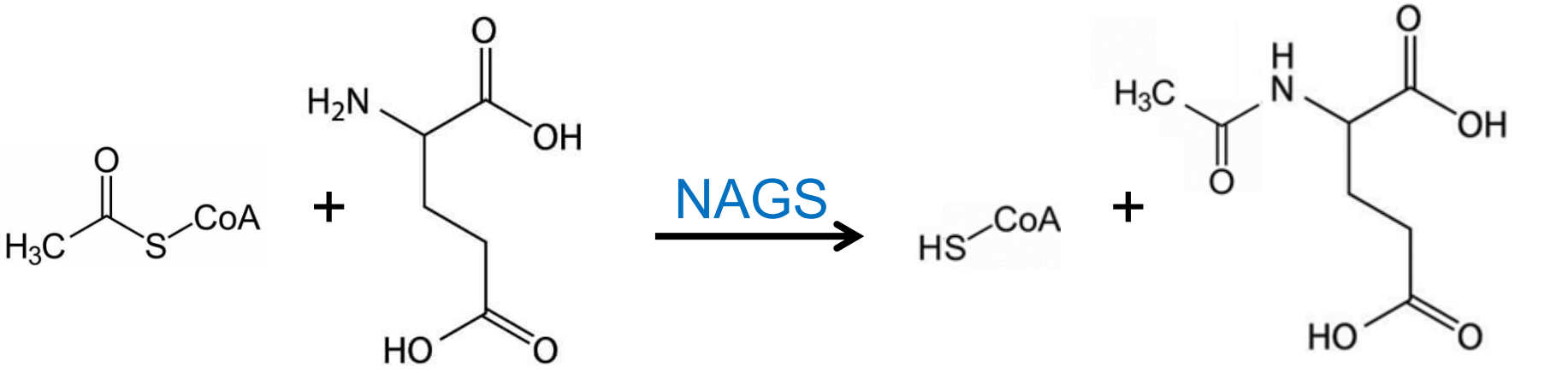
- La **production** d'urée augmente dans les situations suivantes :
  A. Insuffisance hépatique aigue
  B. Hyper-catabolisme protidique
  C. Insuffisance rénale
  D. Hypertension portale avec shunt porto-cave
  E. Régime riche en viande


AA
Glu
Gln
$NH_4^+$
Bactéries intestinales
Cycle de l 'urée
Urée

- La **production** d'urée augmente dans les situations suivantes :
  A. Insuffisance hépatique aigue
  B. Hyper-catabolisme protidique
  C. Insuffisance rénale
  D. Hypertension portale avec shunt porto-cave
  E. Régime riche en viande

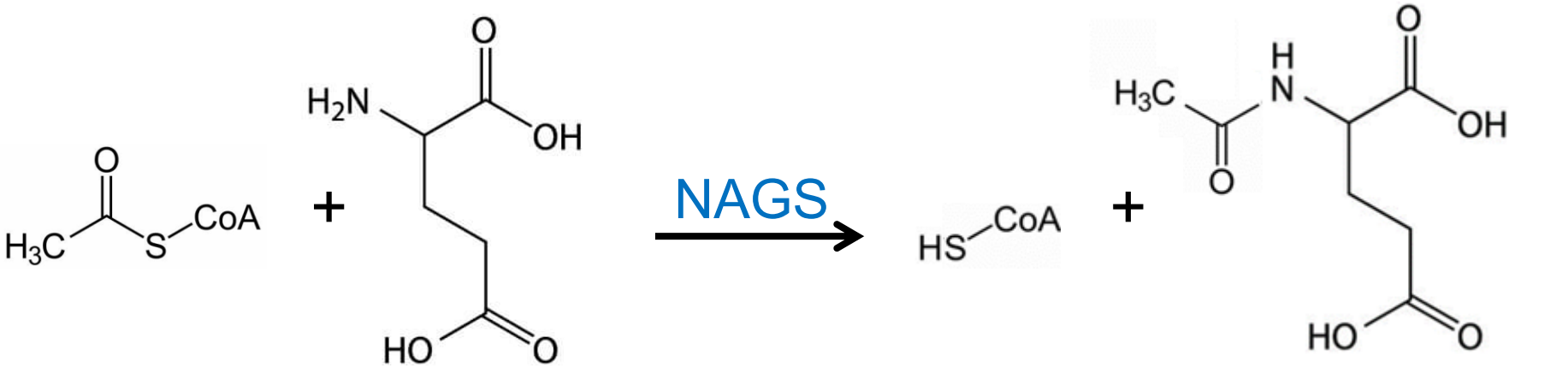
- B, E (l'urée augmente dans l'insuffisance rénale mais par défaut d'élimination)

Quelle serait la conséquence d'un déficit enzymatique de la N-Acétyl-Glutamate Synthétase (NAGS) ?



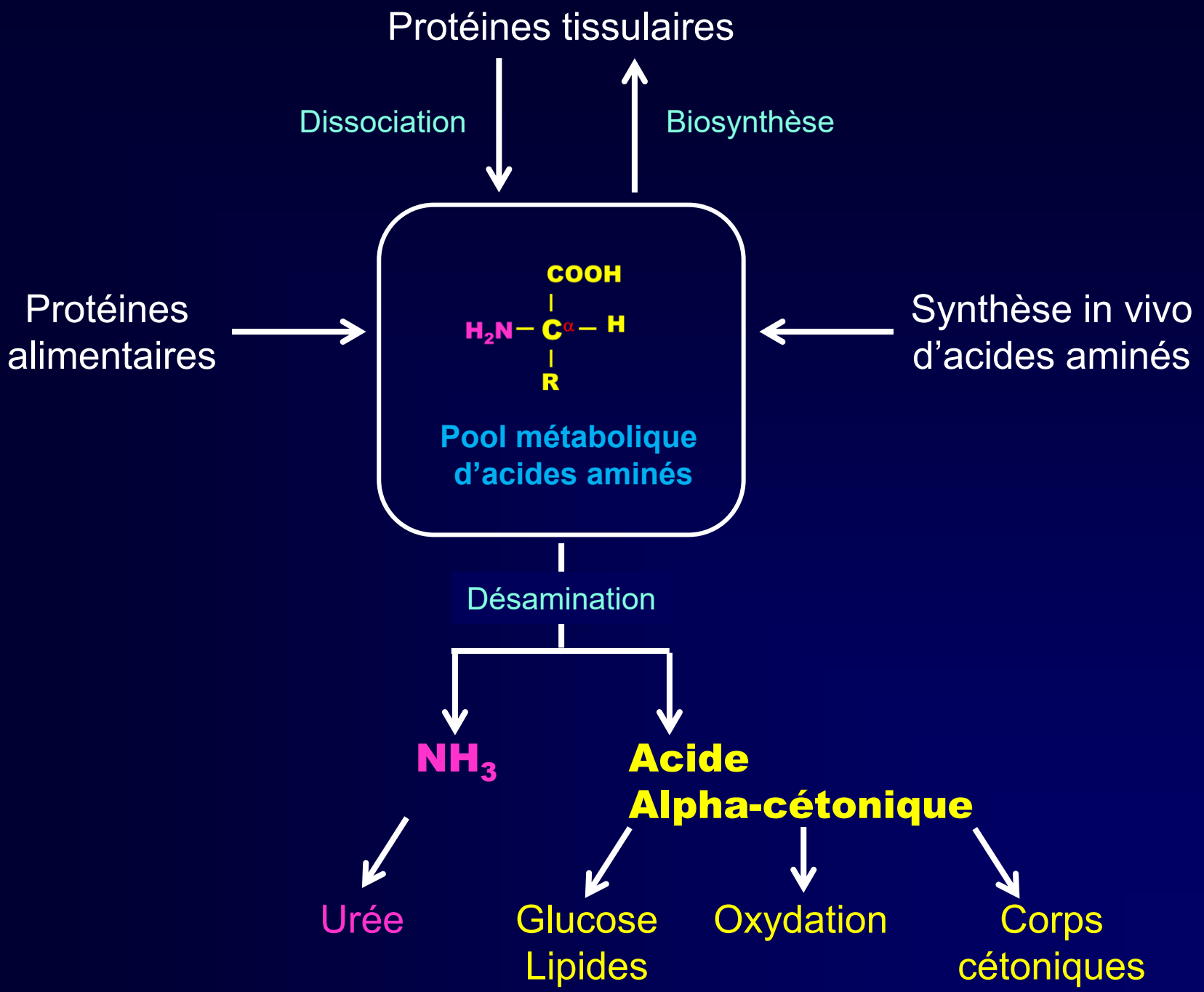

Acétyl-CoA | Acide Glutamique | HS-CoA | Acide N-Acétyl-Glutamique

Quelle serait la conséquence d'un déficit enzymatique de la N-Acétyl-Glutamate Synthétase (NAGS) ?

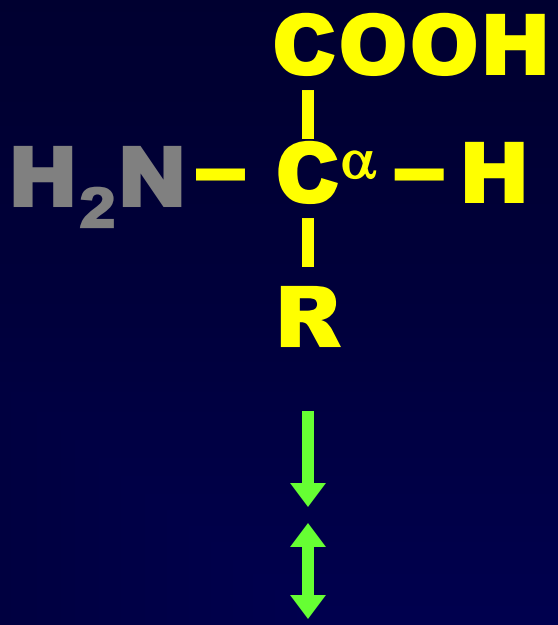



Acétyl-CoA | Acide Glutamique | HS-CoA | Acide N-Acétyl-Glutamique

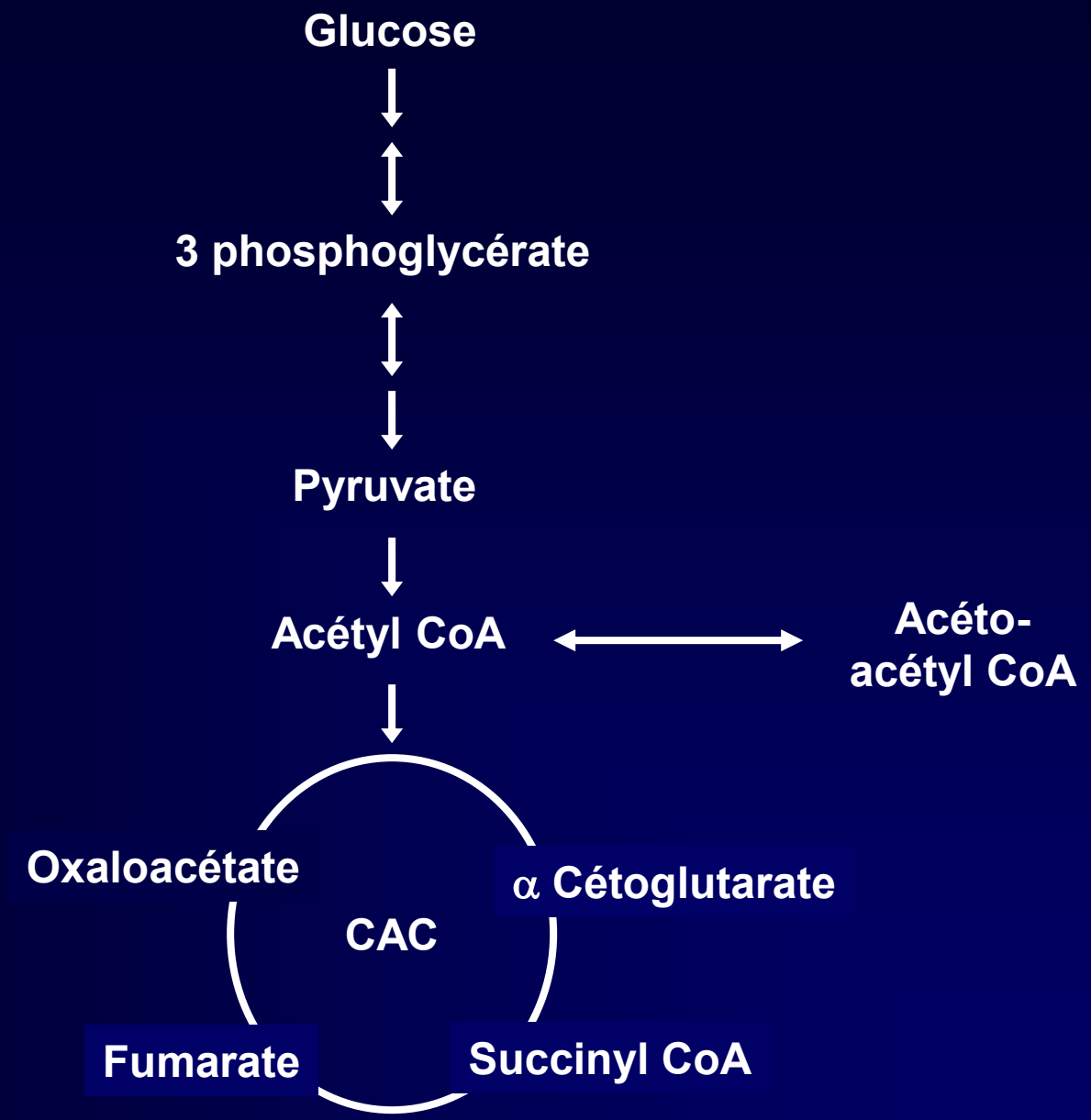
- Diminution de l'activité du cycle de l'urée avec risque d'hyperammoniémie

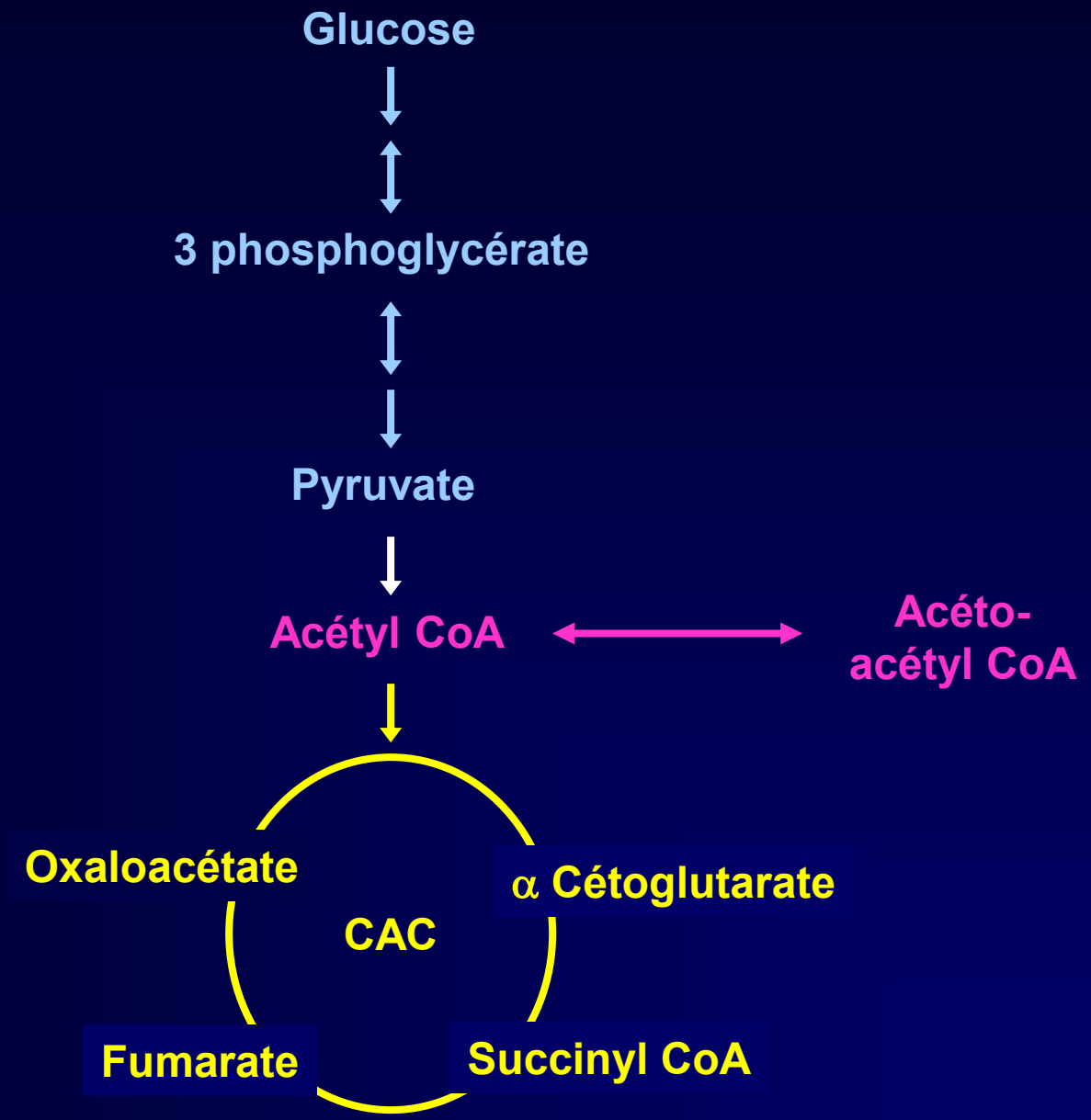

Protéines tissulaires
Dissociation
Biosynthèse
Protéines alimentaires
Synthèse in vivo d'acides aminés
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Désamination
NH3
Acide Alpha-cétonique
Urée
Glucose Lipides
Oxydation
Corps cétoniques

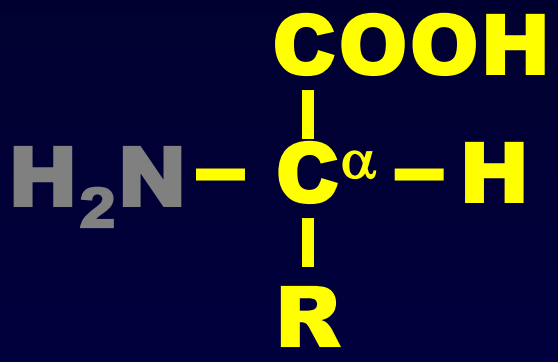
# Transformation du squelette carboné des acides aminés




Intermédiaires amphiboliques


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA

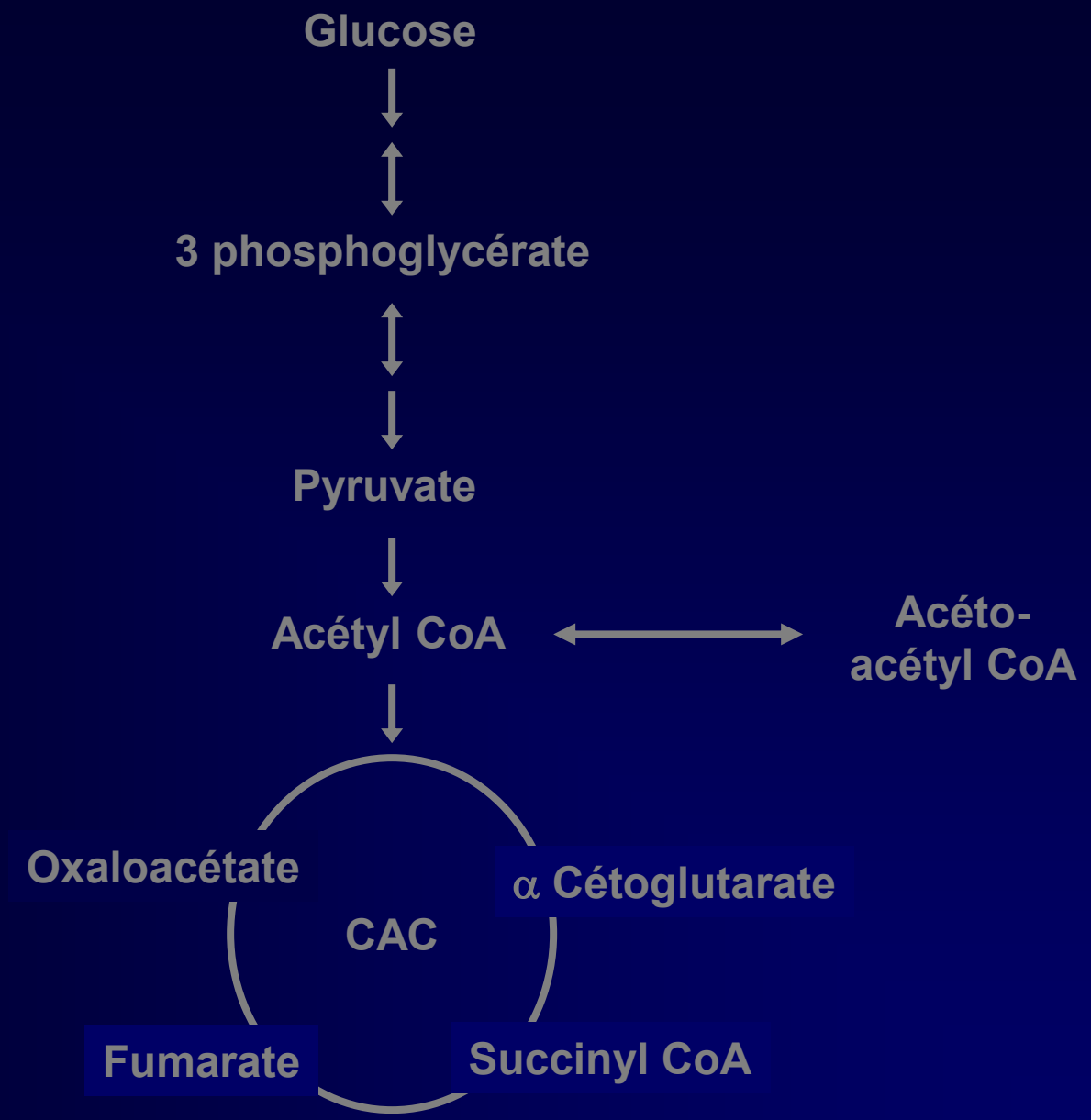


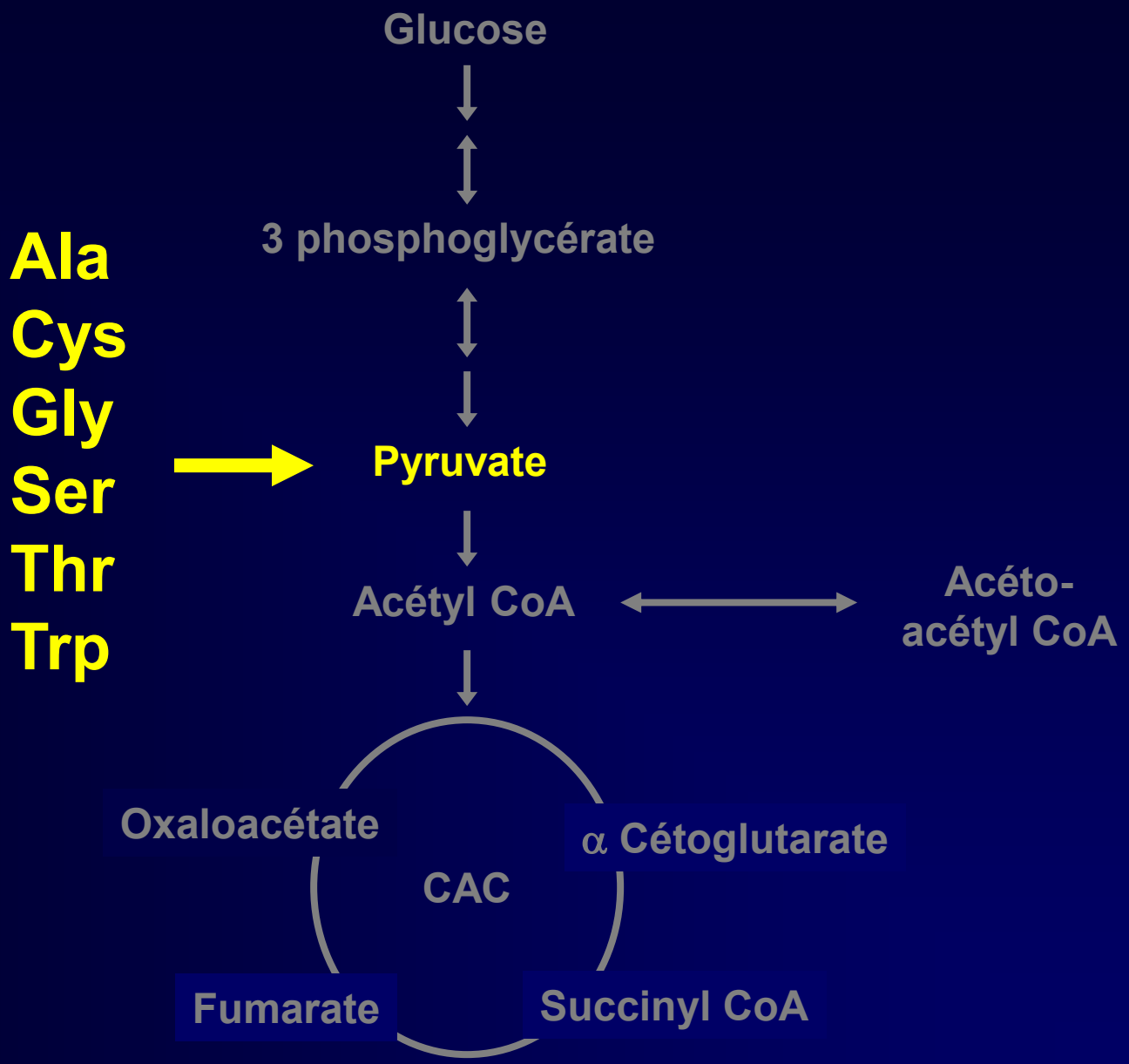

Intermédiaires amphiboliques

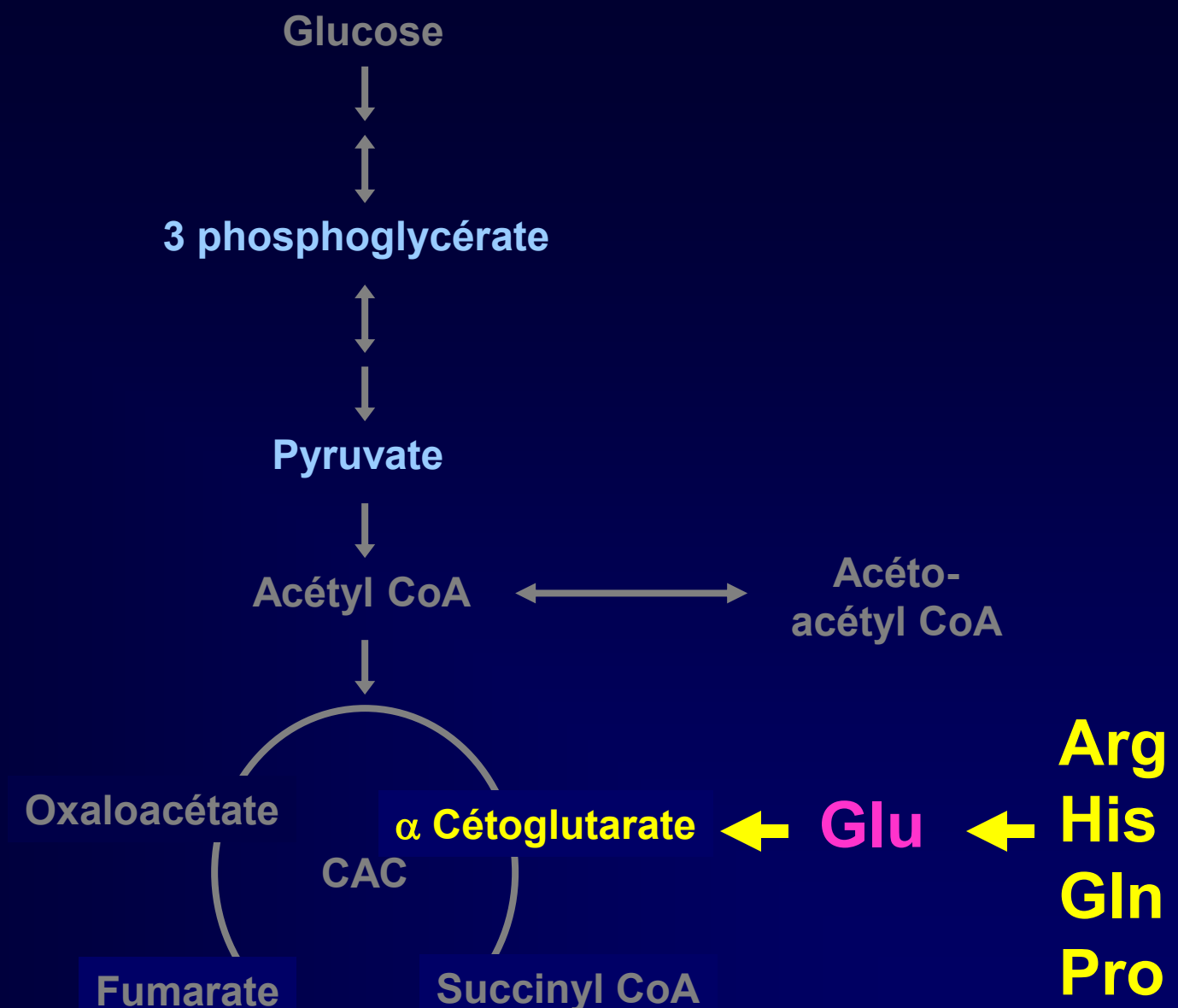
Glucogènes

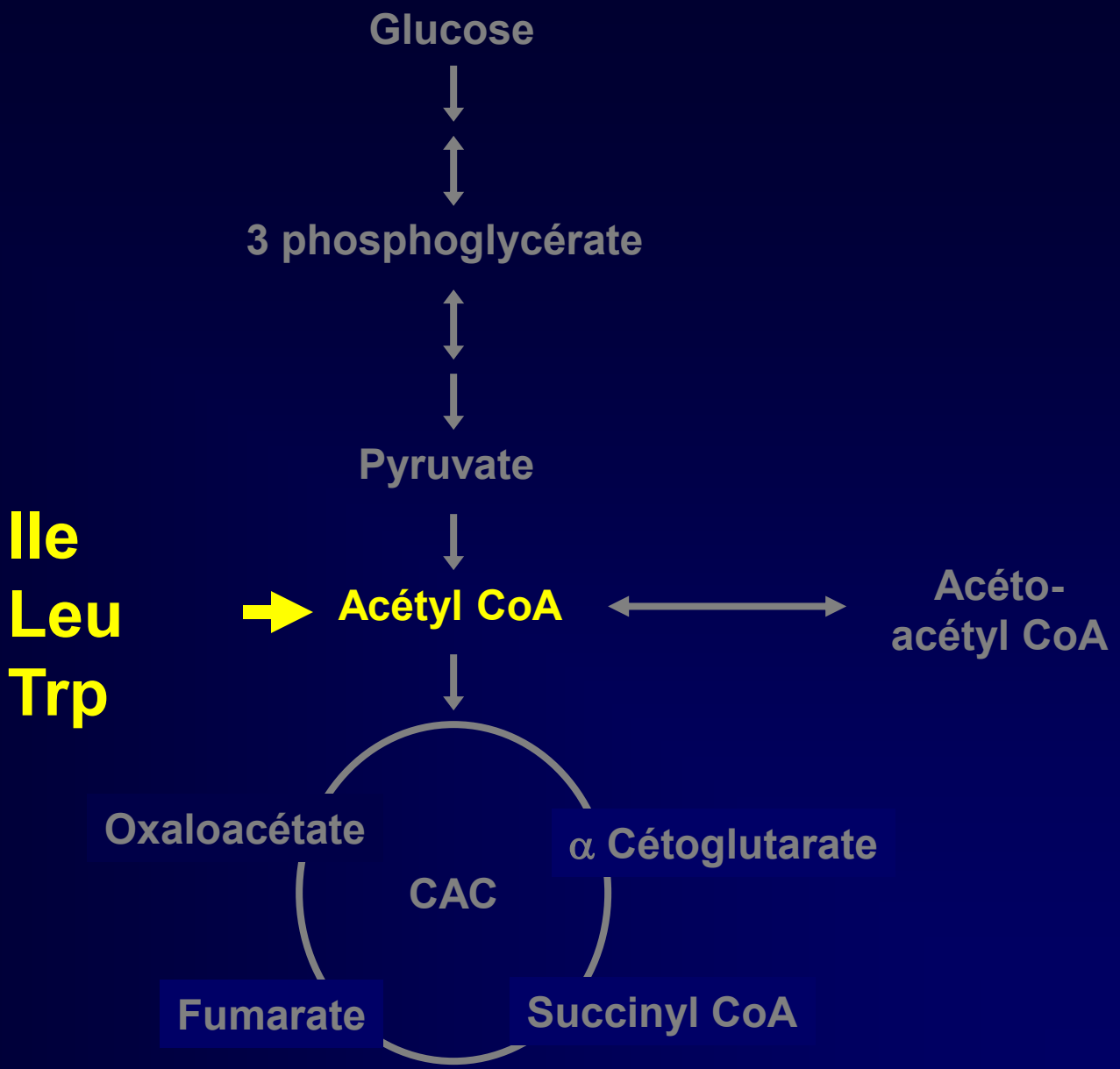
Cétogènes

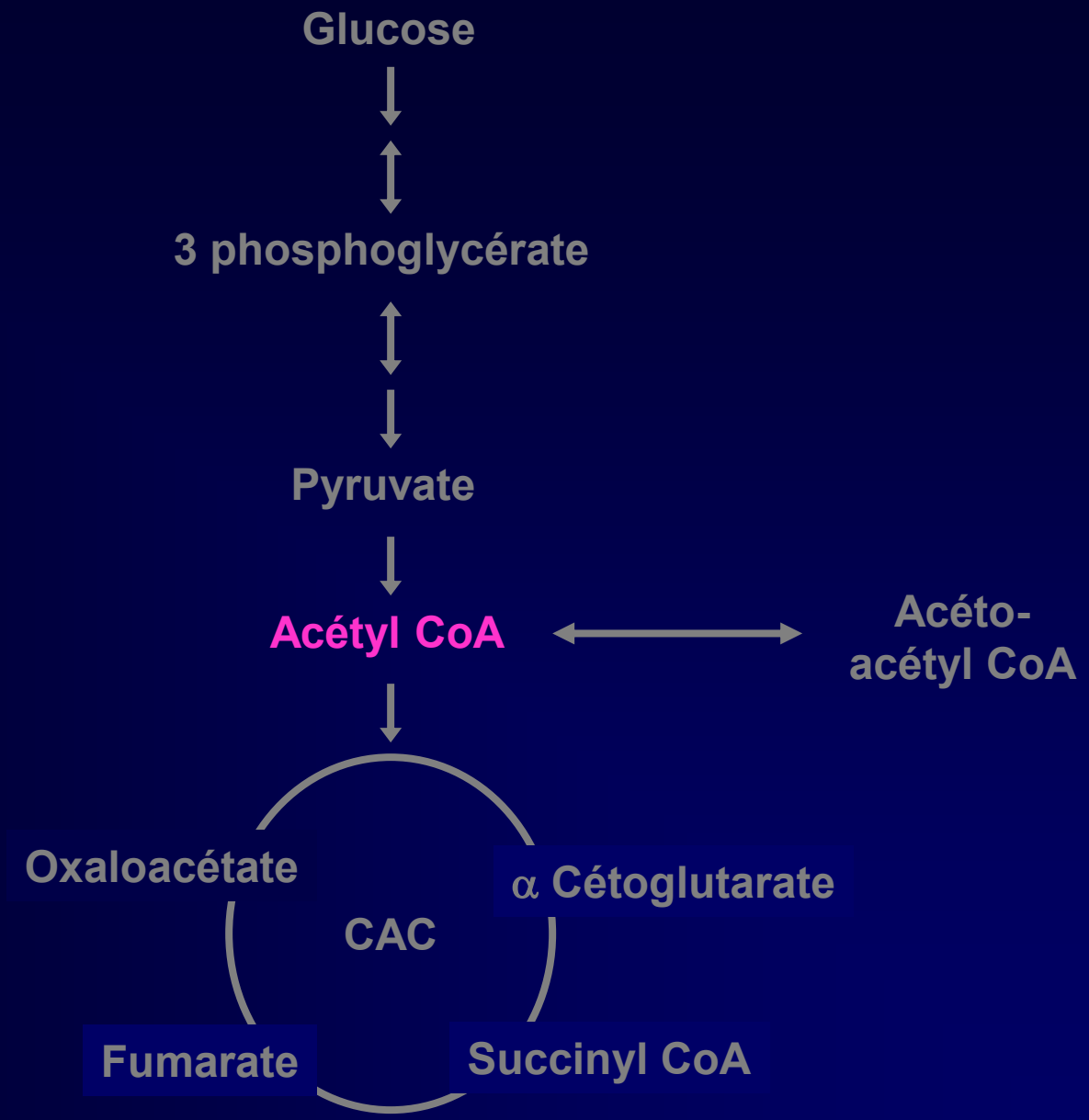
Glucogènes
Cétogènes

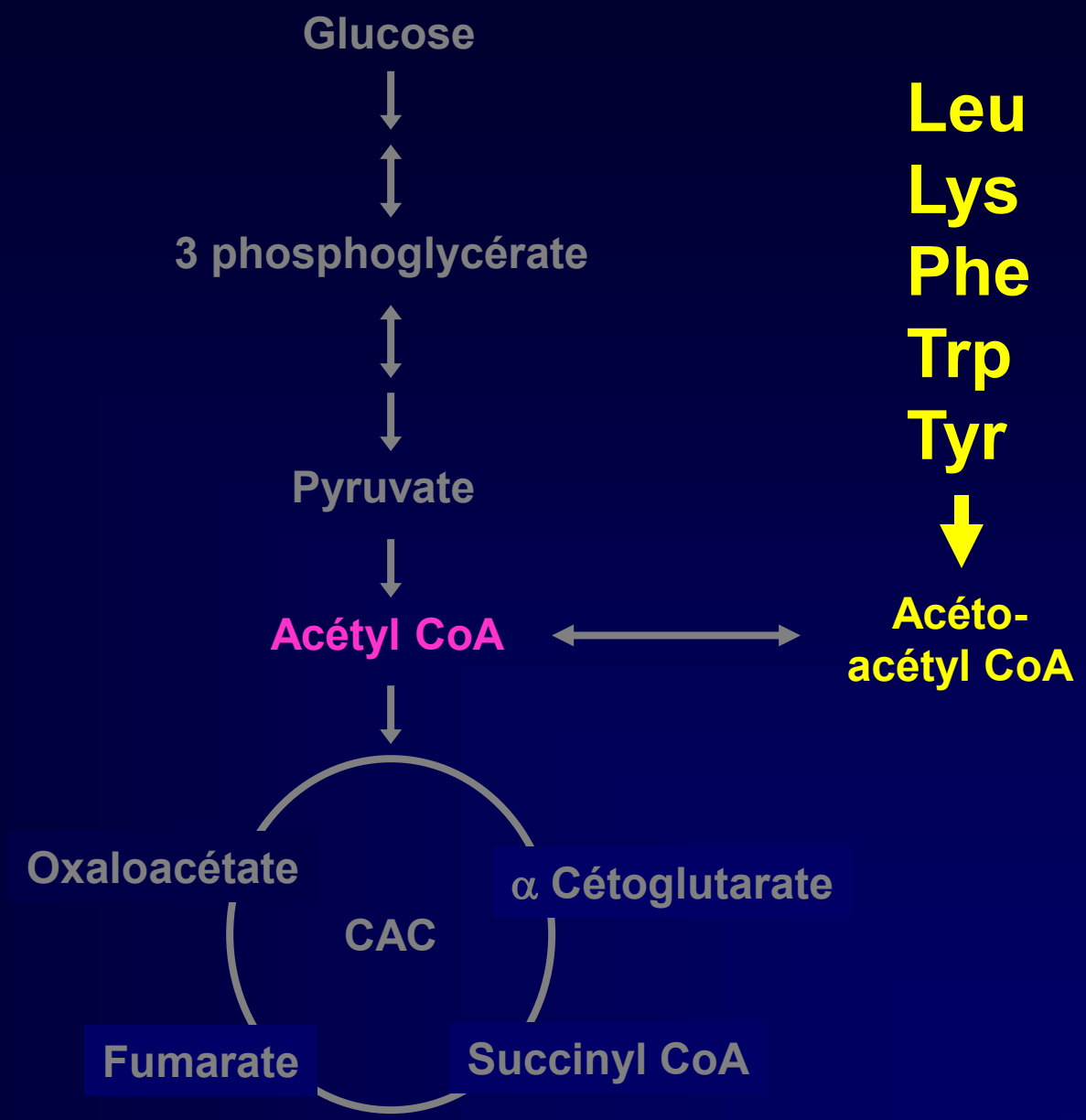

Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA


Glucose
3 phosphoglycérate
Ala
Cys
Gly
Ser
Thr
Trp
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA
Glu
Arg
His
Gln
Pro


Glucose
3 phosphoglycérate
Pyruvate
Ile
Leu
Trp
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Leu
Lys
Phe
Trp
Tyr
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA

Glucogènes

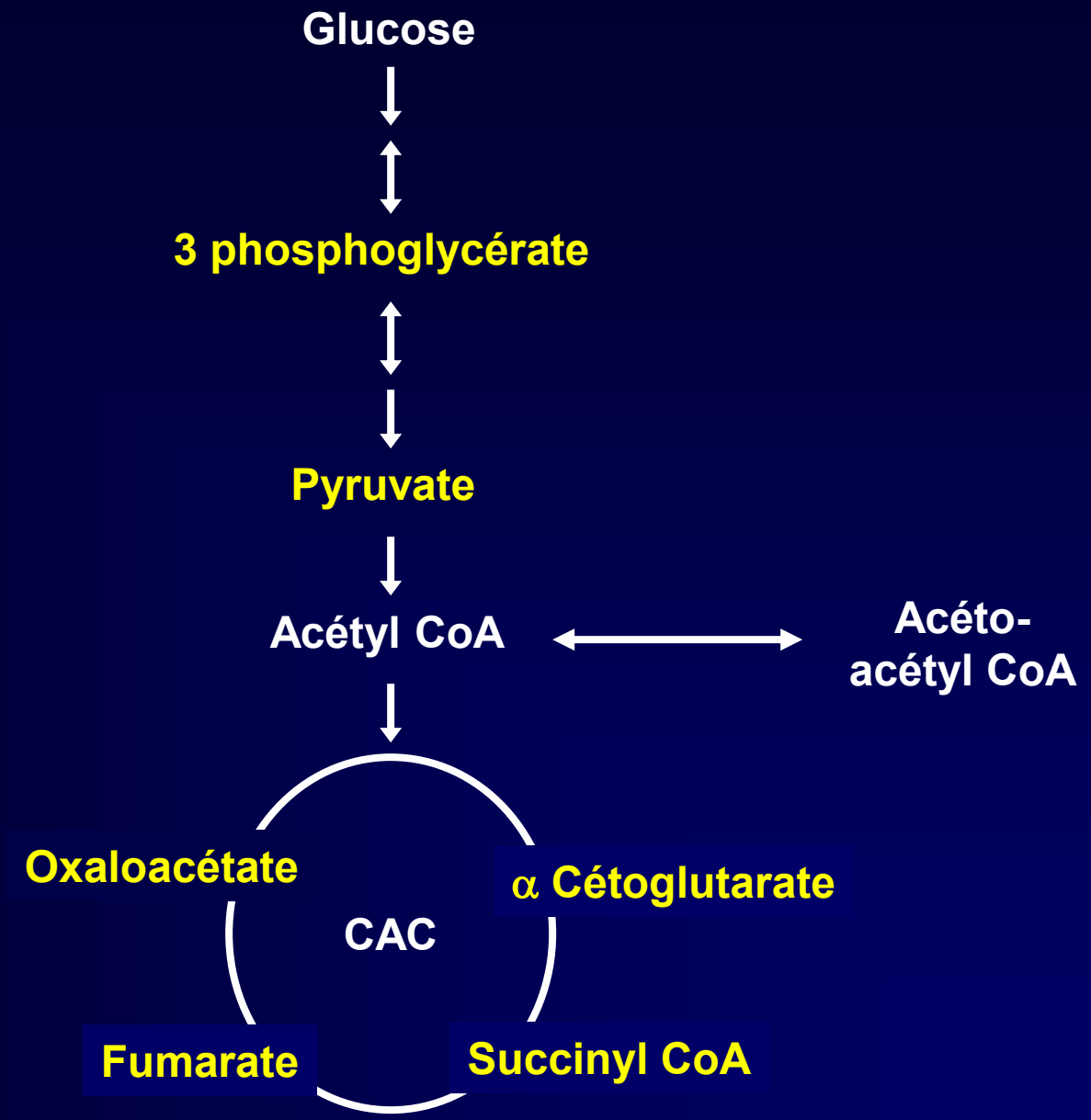
Cétogènes

Glucogènes
Cétogènes

**Glucogènes**

Cétogènes

Glucogènes
Cétogènes


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA

**Glucogènes**

**Ala**
**Arg**
**Asp**
**Cys**
**Glu**
**Gly**
**His**
**Met**
**Pro**
**Ser**
**Thr**
**Val**

**Cétogènes**

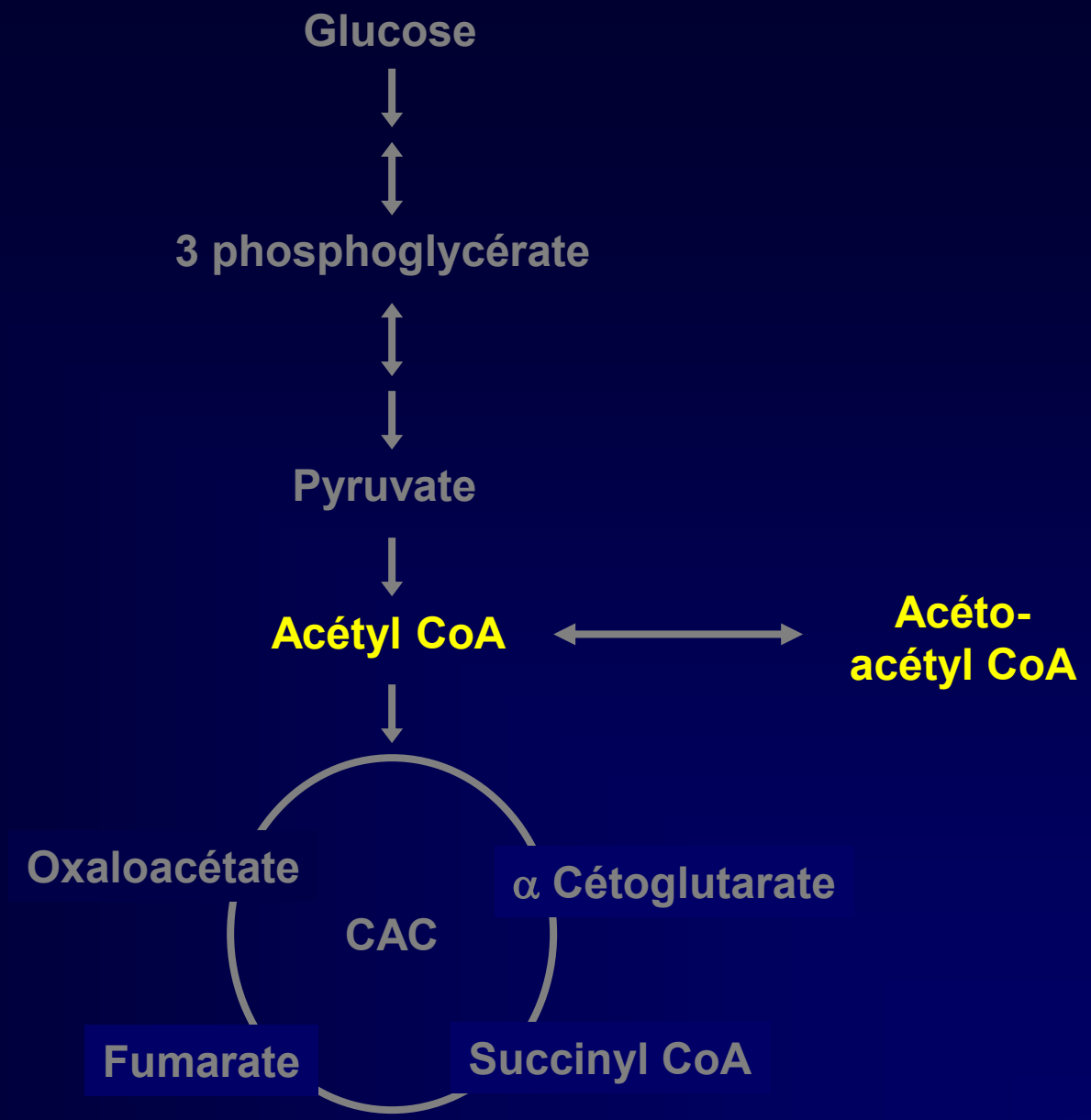
**Glucogènes**
**Cétogènes**

| Glucogènes | Cétogènes | Glucogènes Cétogènes |
| --- | --- | --- |
| Ala | | |
| Arg | | |
| Asp | | |
| Cys | | |
| Glu | | |
| Gly | | |
| His | | |
| Met | | |
| Pro | | |
| Ser | | |
| Thr | | |
| Val | | |

Glucogènes

Cétogènes

Glucogènes
Cétogènes


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA

## Glucogènes

Ala
Arg
Asp
Cys
Glu
Gly
His
Met
Pro
Ser
Thr
Val

## Cétogènes

Leu
Lys

## Glucogènes Cétogènes

| Glucogènes | Cétogènes | Glucogènes Cétogènes |
| --- | --- | --- |
| Ala | Leu | |
| Arg | Lys | |
| Asp | | |
| Cys | | |
| Glu | | |
| Gly | | |
| His | | |
| Met | | |
| Pro | | |
| Ser | | |
| Thr | | |
| Val | | |

## Glucogènes

Ala
Arg
Asp
Cys
Glu
Gly
His
Met
Pro
Ser
Thr
Val

## Cétogènes

Leu
Lys

## Glucogènes Cétogènes

Ile
Phe
Trp
Tyr

## Glucogènes

Ala
Arg
Asp
Cys
Glu
Gly
His
Met
Pro
Ser
Thr
Val

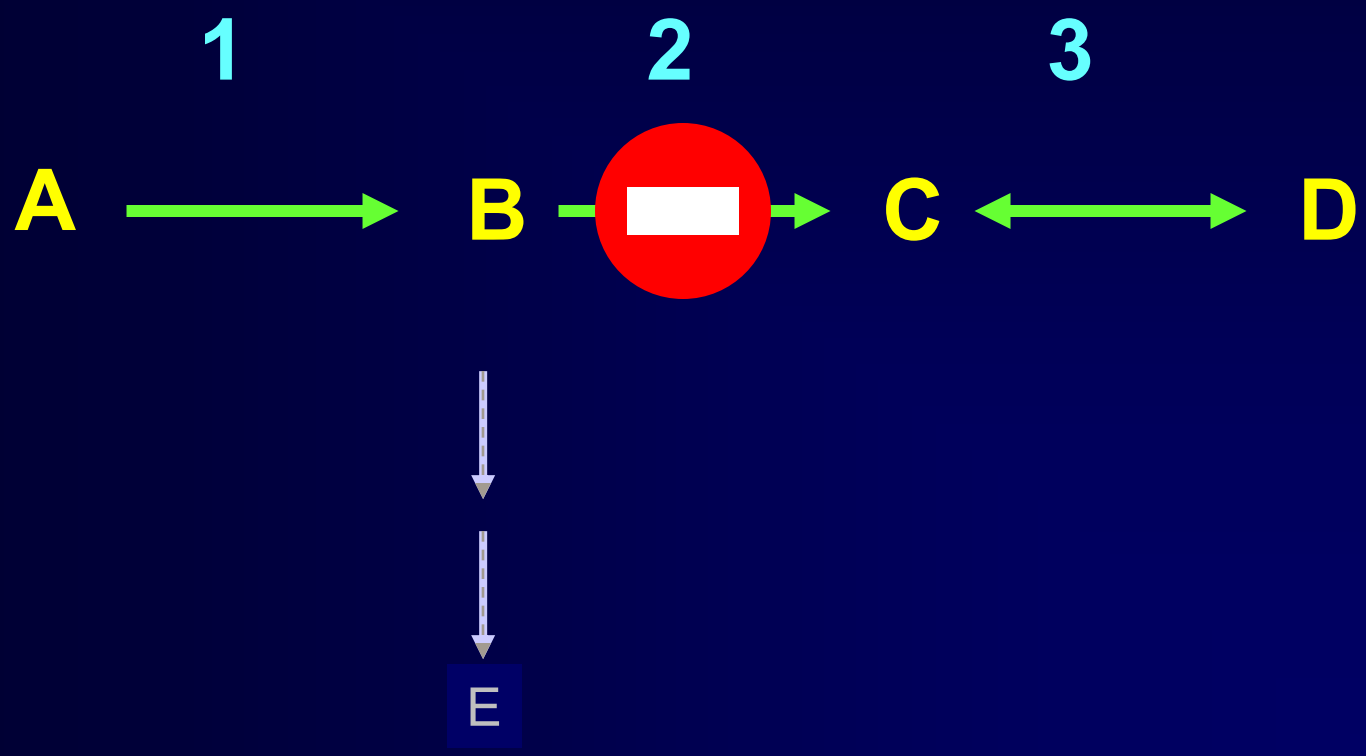
## Cétogènes

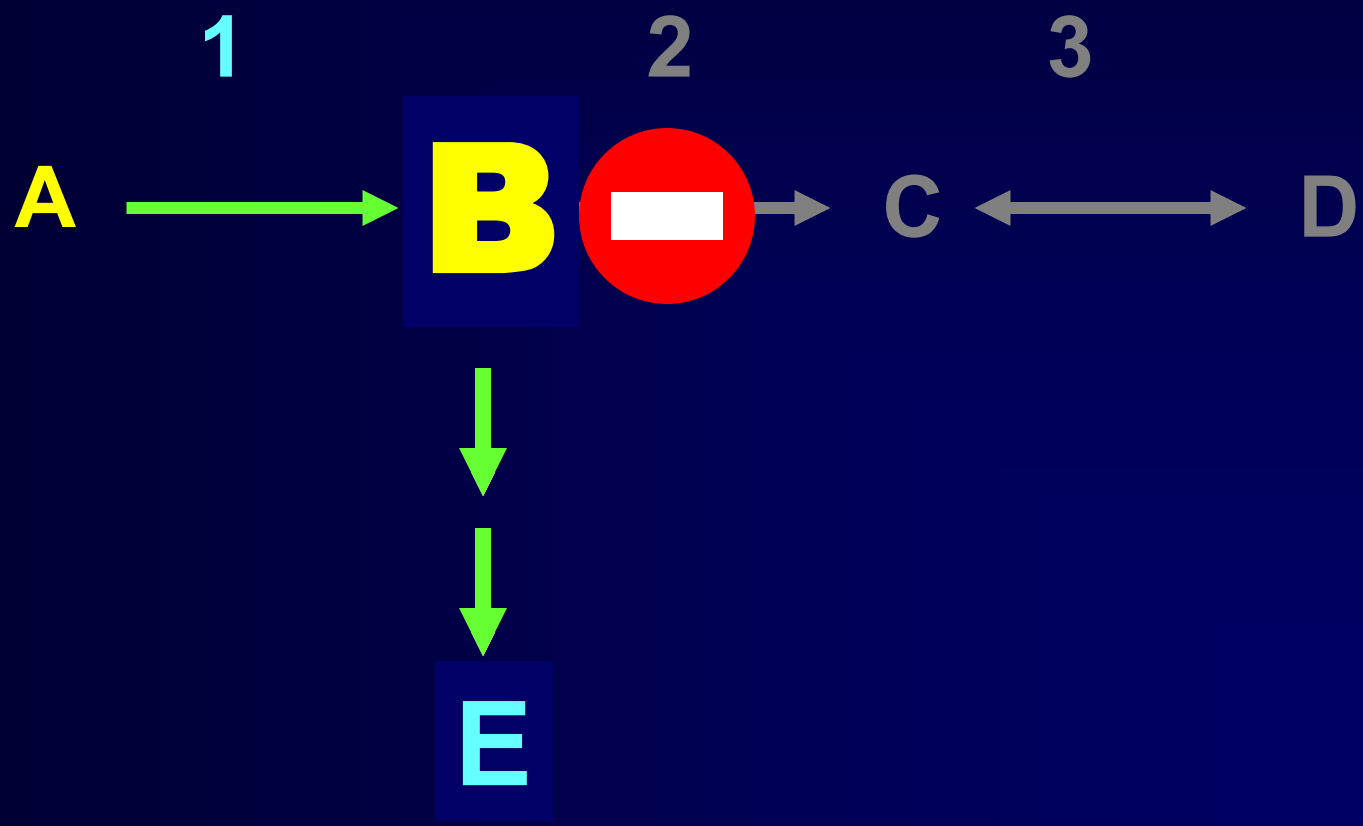
Leu
Lys

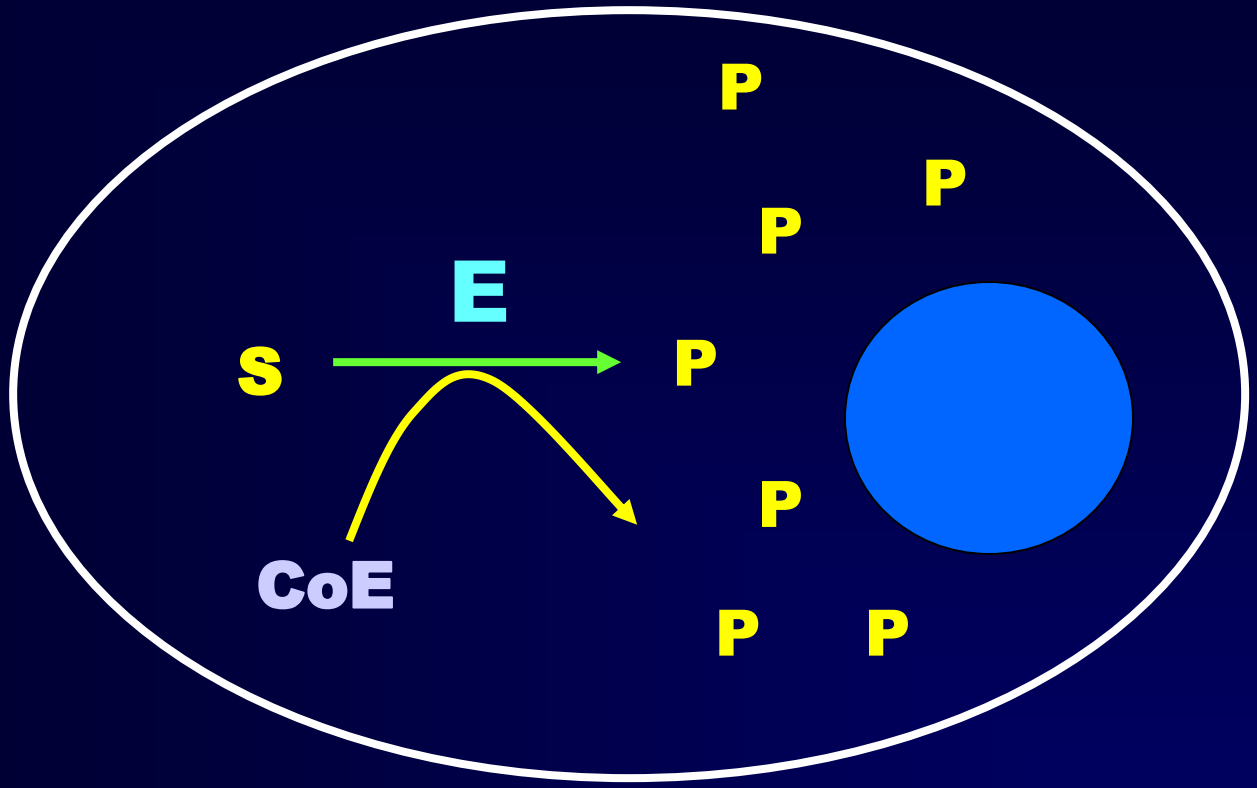
## Glucogènes Cétogènes

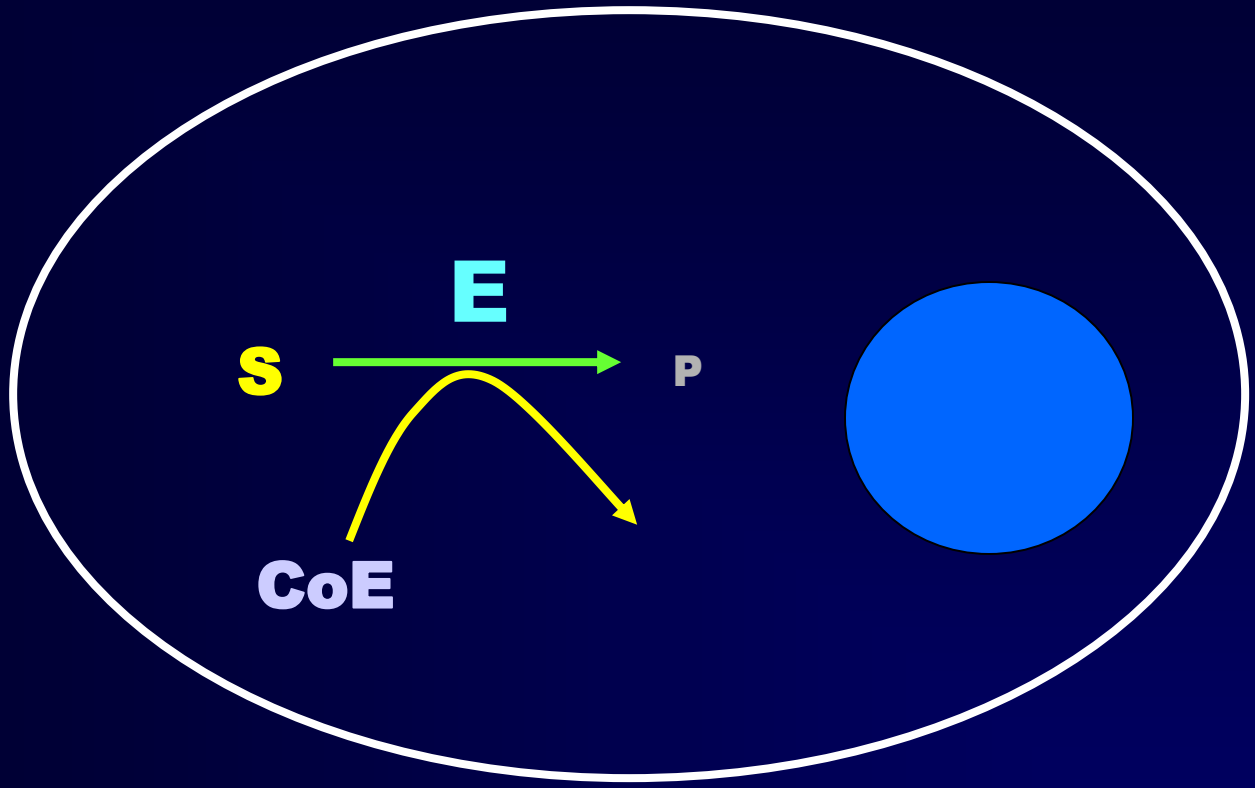
Ile
Phe
Trp
Tyr

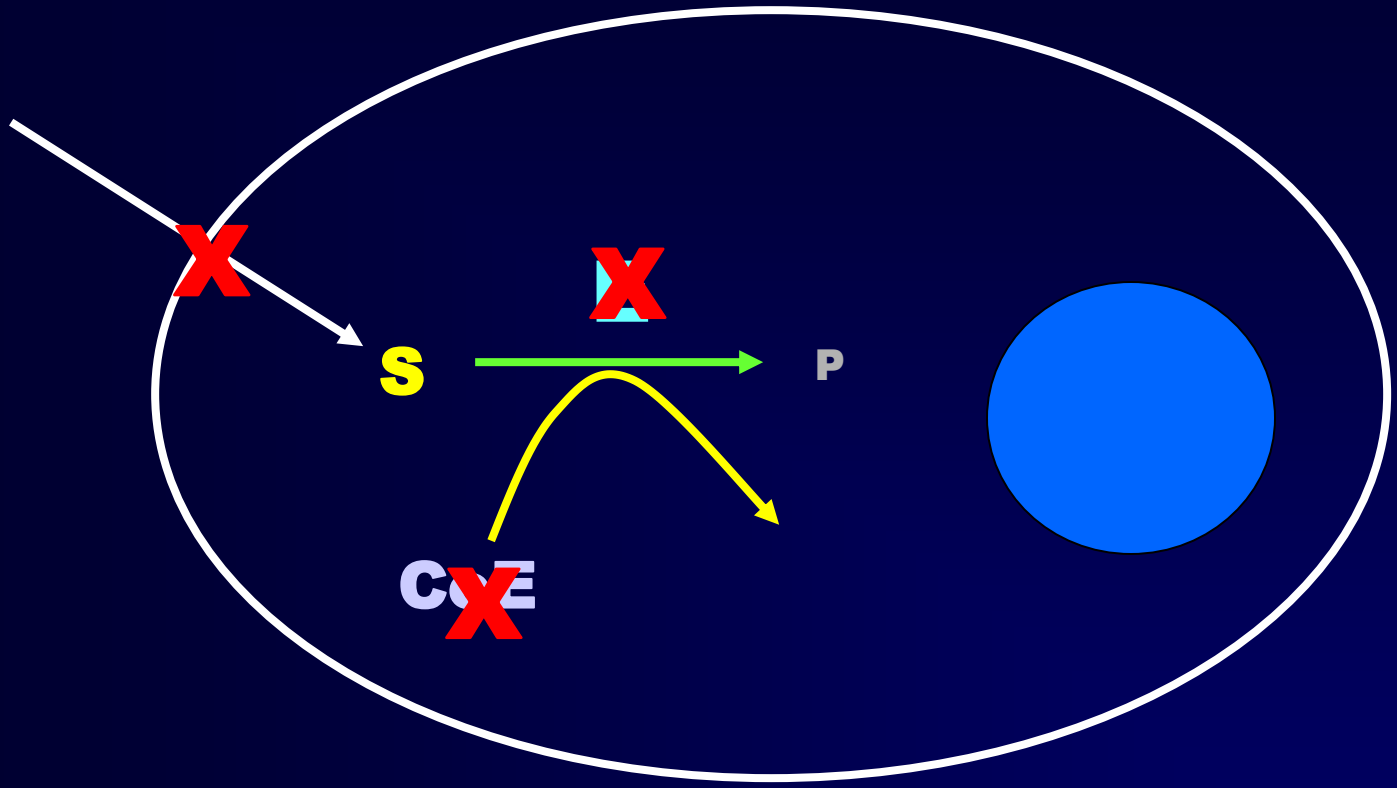
# Erreurs innées du métabolisme des acides aminés


1
2
3
A
B
C
D
E


1
2
3
A
B
C
D
E


P
P
P
P
E
S
P
P
CoE
P
P


E
S
P
CoE

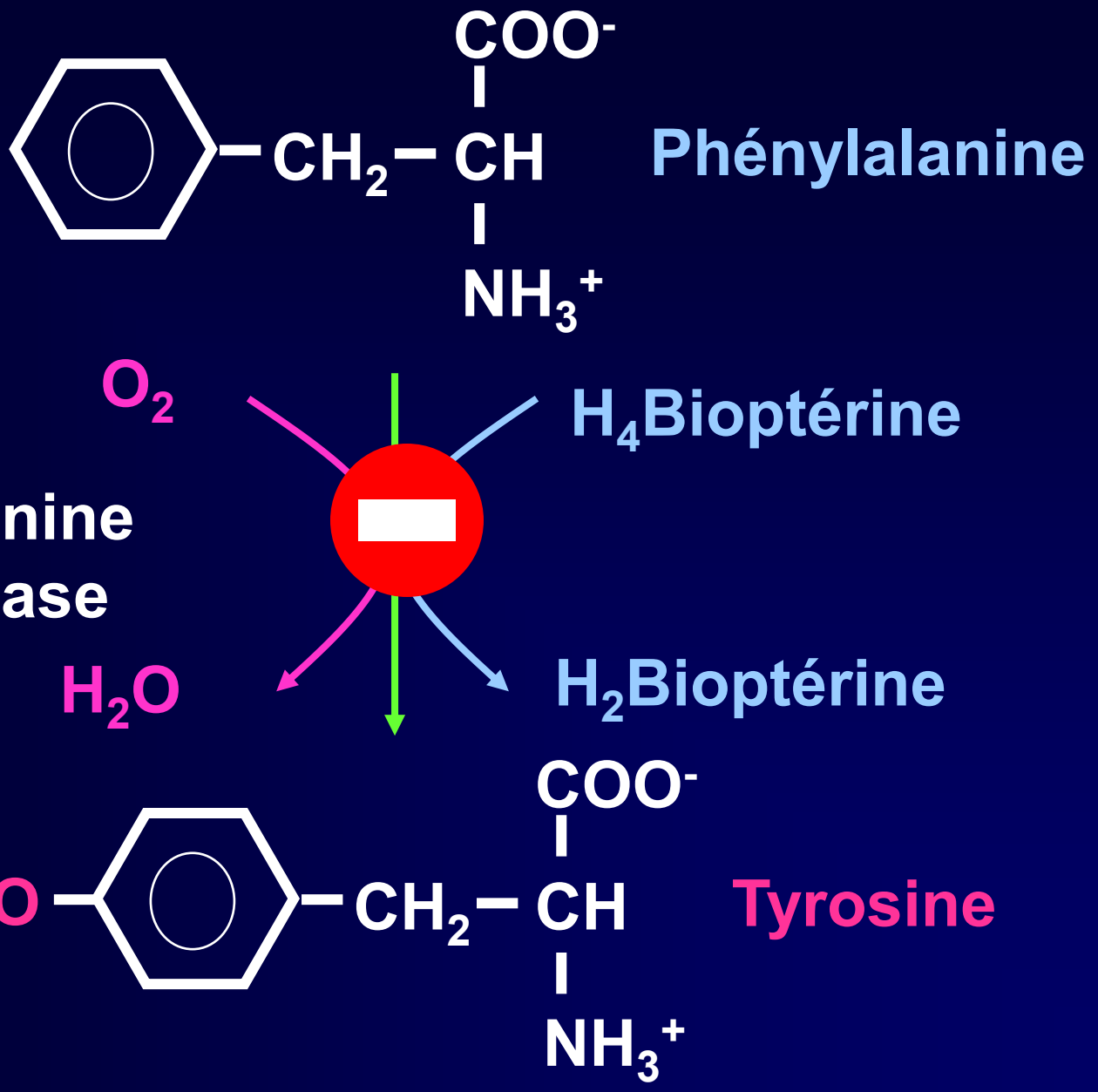

X
X
S
P
C
E
X

**Nourrisson**
**2 semaines**
**Urines :**

**acidephényl pyruvique ++**
acide phényl acétique ++
acide phényl lactique ++

**Sang :**

**Phénylalanine ++**
**Tyrosine -**

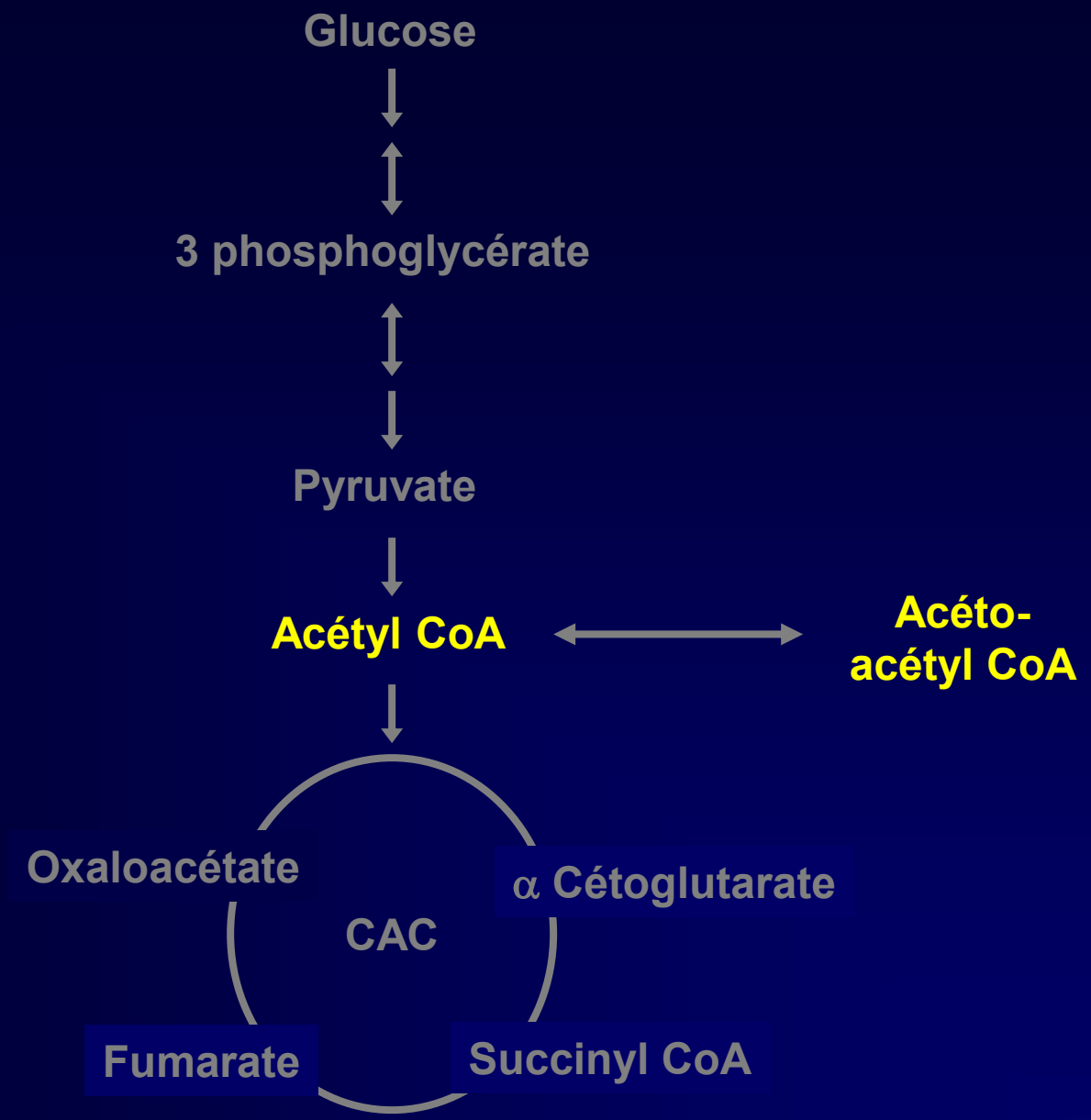

COO-
CH2
CH
NH3+
Phénylalanine
O2
H4Bioptérine
Phénylalanine
Hydroxylase
H2O
H2Bioptérine
COO-
HO
CH2
CH
NH3+
Tyrosine

Dr Asbjørn Følling
1888 -1973

# Phénylcétonurie

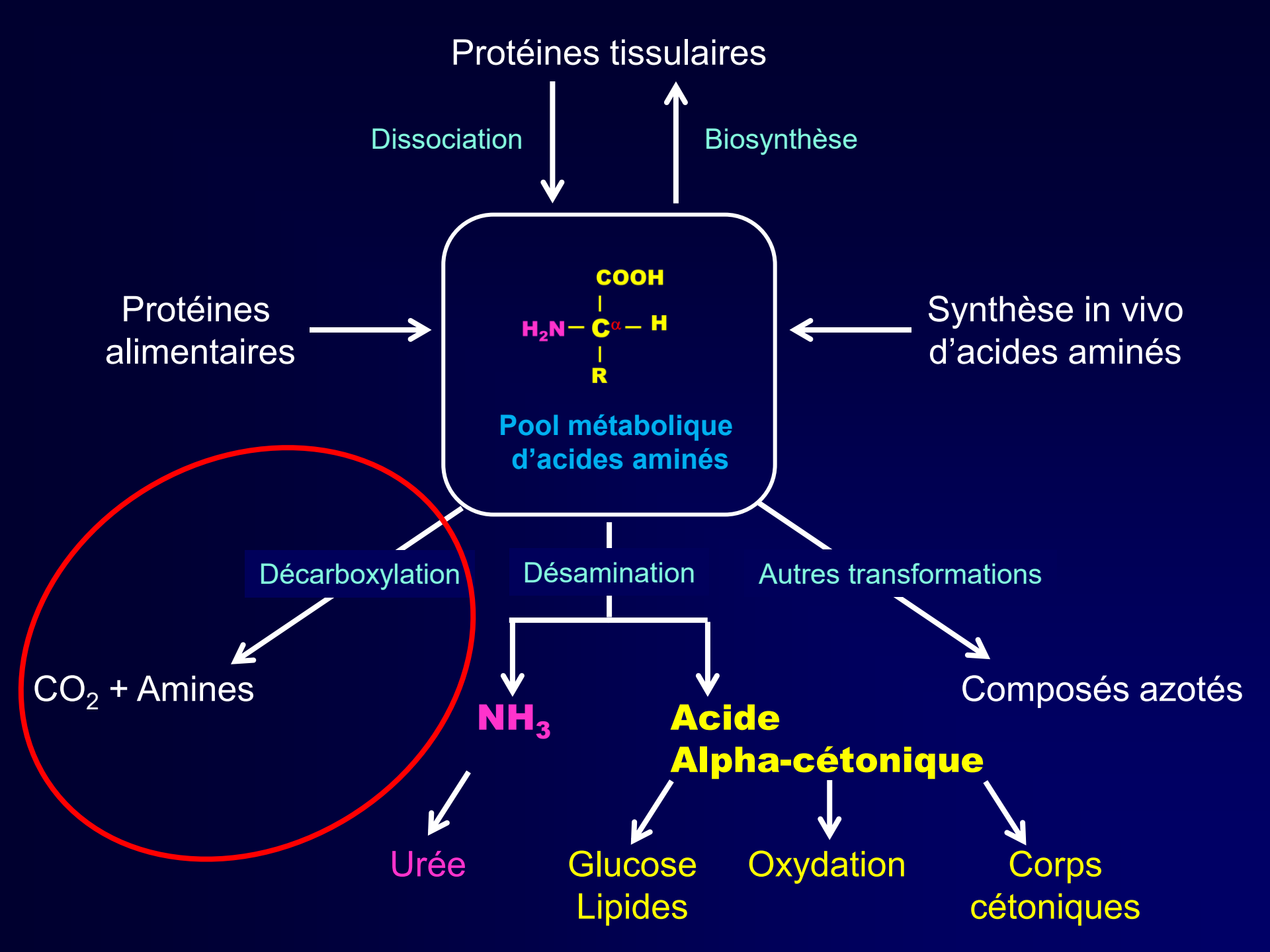

QUIZ
TIME

- Un acide aminé « CETOGENE » est un AA qui aboutit à la formation de quels intermédiaires ?


Glucose
3 phosphoglycérate
Pyruvate
Acétyl CoA
Acéto-
acétyl CoA
Oxaloacétate
α Cétoglutarate
CAC
Fumarate
Succinyl CoA

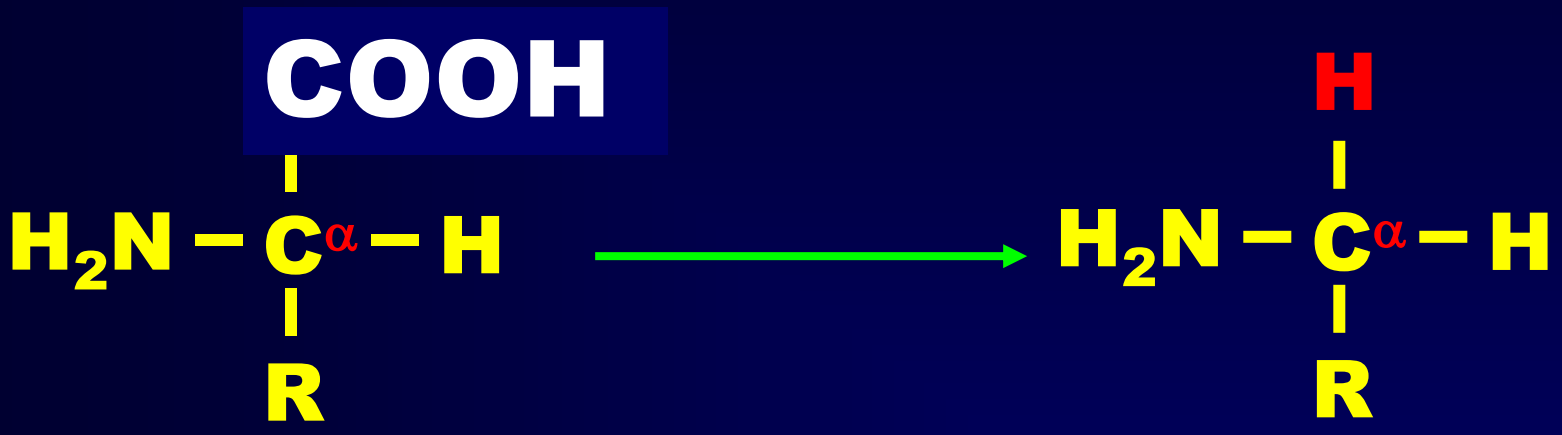
- Un acide aminé « CETOGENE » est un AA qui aboutit à la formation de quels intermédiaires ?

- Acétyl-CoA & Acéto-acétyl-CoA


Protéines tissulaires
Dissociation
Biosynthèse
Protéines alimentaires
Synthèse in vivo d'acides aminés
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Décarboxylation
Désamination
Autres transformations
CO2 + Amines
NH3
Acide Alpha-cétonique
Composés azotés
Urée
Glucose Lipides
Oxydation
Corps cétoniques

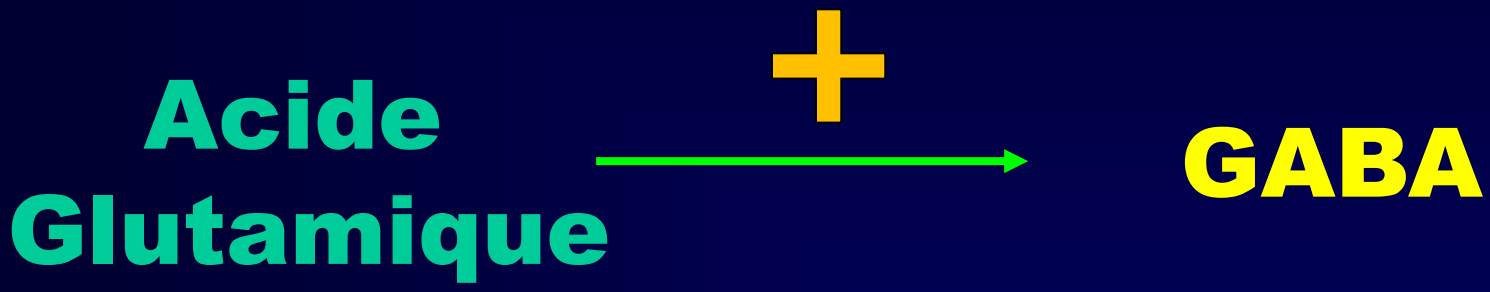
# Les Amines Biogènes

# Décarboxylase

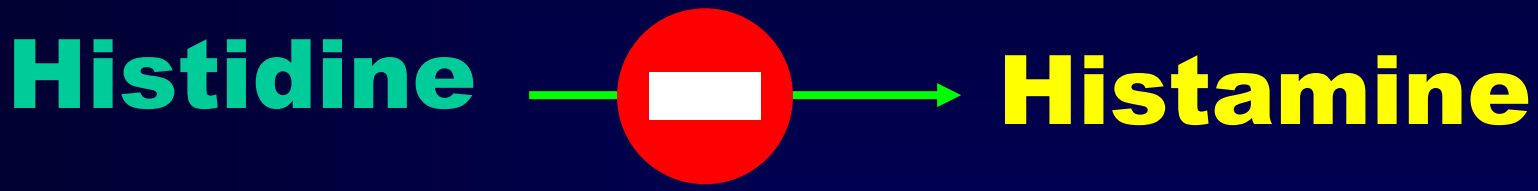



Phosphate de pyridoxal

Acide Glutamique → Acide γ aminobutyrique


Acide
Glutamique
+
GABA

| | |
|---|---|
| Gabapentine | NEURONTIN* |
| Acide valproïque | DÉPAKINE* |

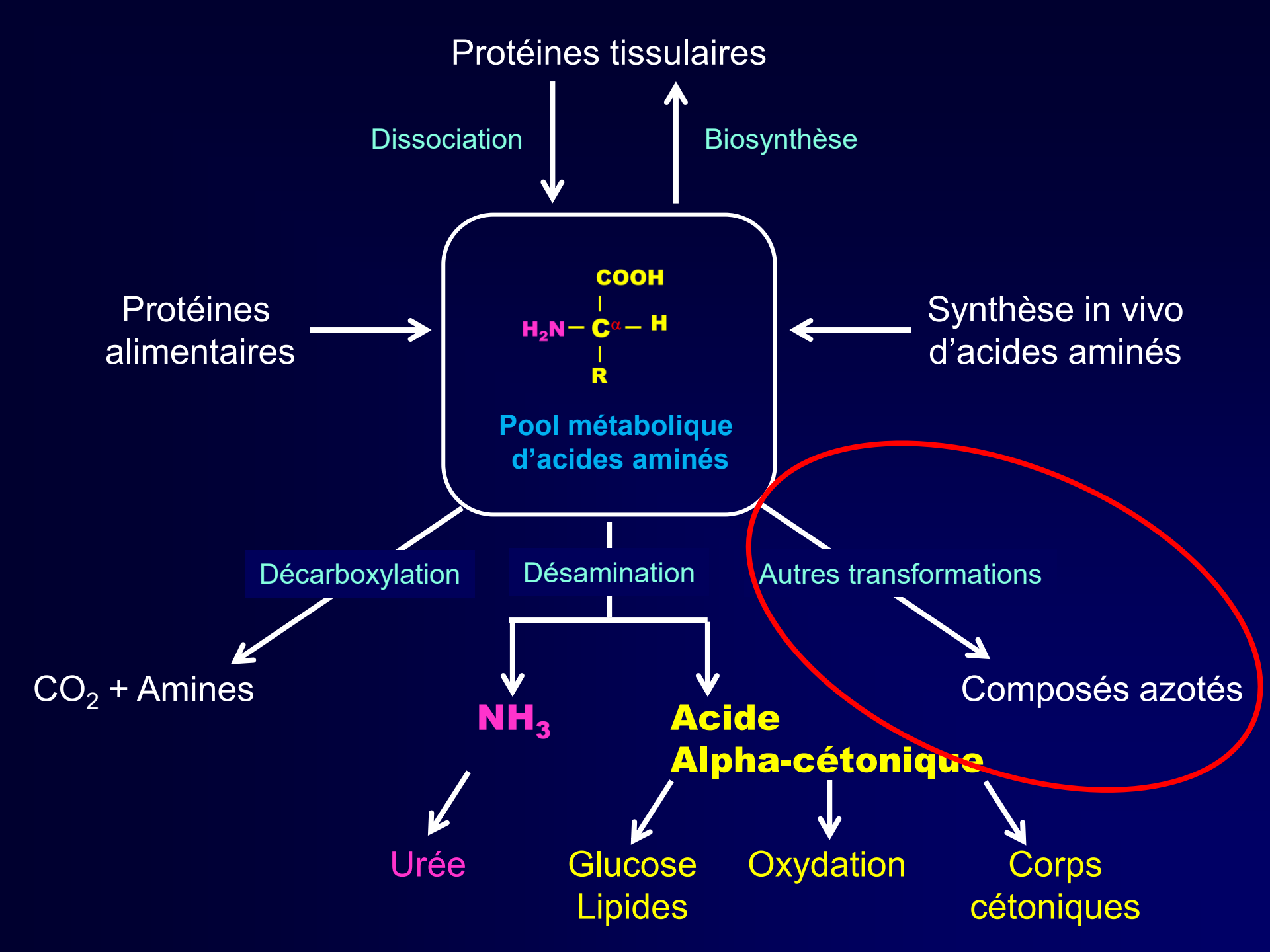

Histidine
Histamine

INFLUENCE D'UN **INHIBITEUR DE L'HISTIDINE DECARBOXYLASE** SUR L'HISTAMINO-LIBERATION PRODUITE CHEZ L'HOMME ET LA REACTION ANAPHYLACTIQUE LOCALE

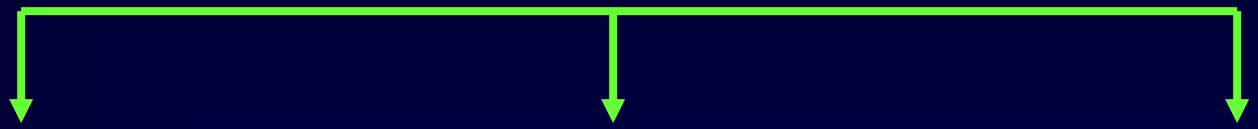
**Allergy 2007**

**Volume 20 Issue 2, Pages 77 – 83**

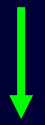
# La tritoqualine (Hypostamine®)


Protéines tissulaires
Dissociation
Biosynthèse
Protéines alimentaires
Synthèse in vivo d'acides aminés
COOH
H2N – Cα – H
R
Pool métabolique d'acides aminés
Décarboxylation
Désamination
Autres transformations
CO2 + Amines
NH3
Acide Alpha-cétonique
Composés azotés
Urée
Glucose Lipides
Oxydation
Corps cétoniques

# Tyrosine



DOPA



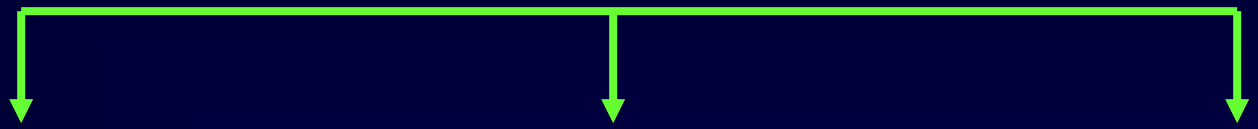
Catécholamines

Dopamine

Noradrénaline

Adrénaline

# Tyrosine



DOPA

Mélanine

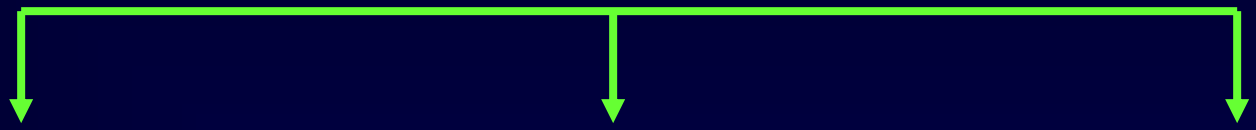
Catécholamines

Dopamine

Noradrénaline

Adrénaline

# Tyrosine



DOPA

Mélanine

Hormones Thyroïdiennes

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Dopamine

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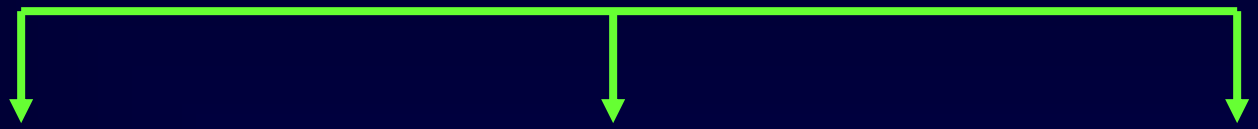
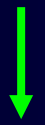
# Tyrosine



DOPA

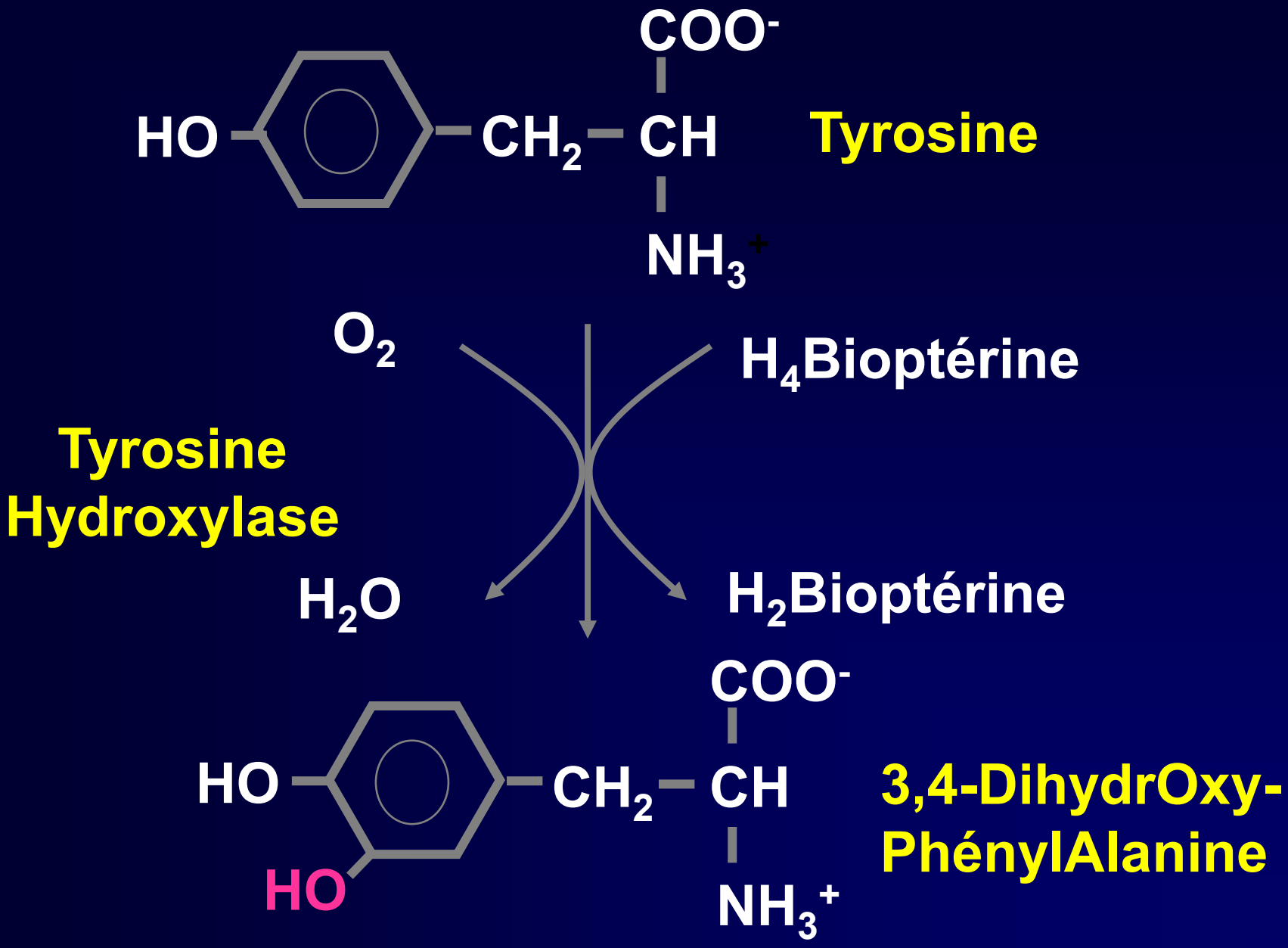


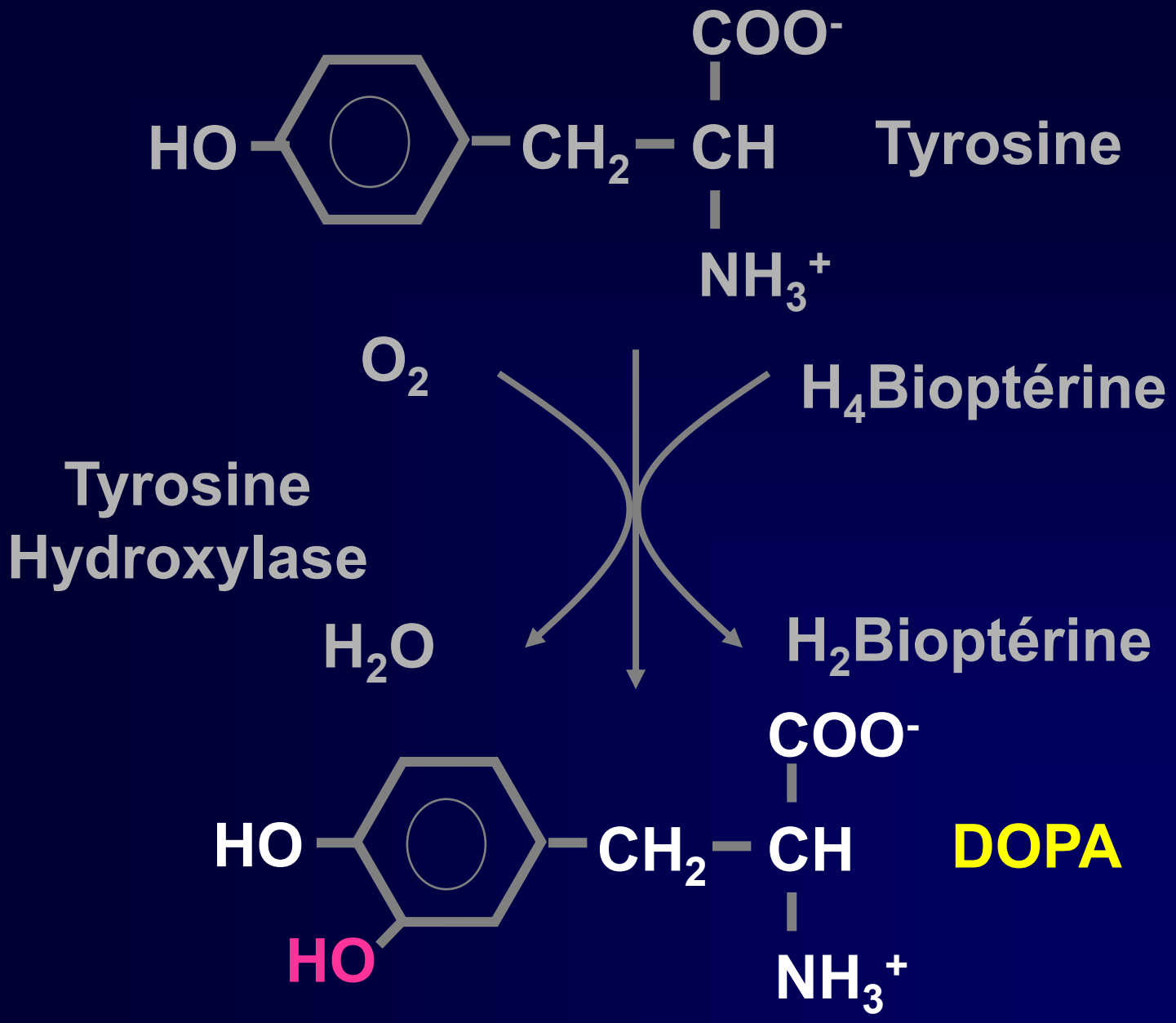
Catécholamines

Dopamine

Noradrénaline

Adrénaline


COO-
HO
CH2
CH
Tyrosine
NH3+
O2
H4Bioptérine
Tyrosine
Hydroxylase
H2O
H2Bioptérine
COO-
HO
CH2
CH
3,4-DihydrOxy-
PhénylAlanine
HO
NH3+


COO-
HO
CH2
CH
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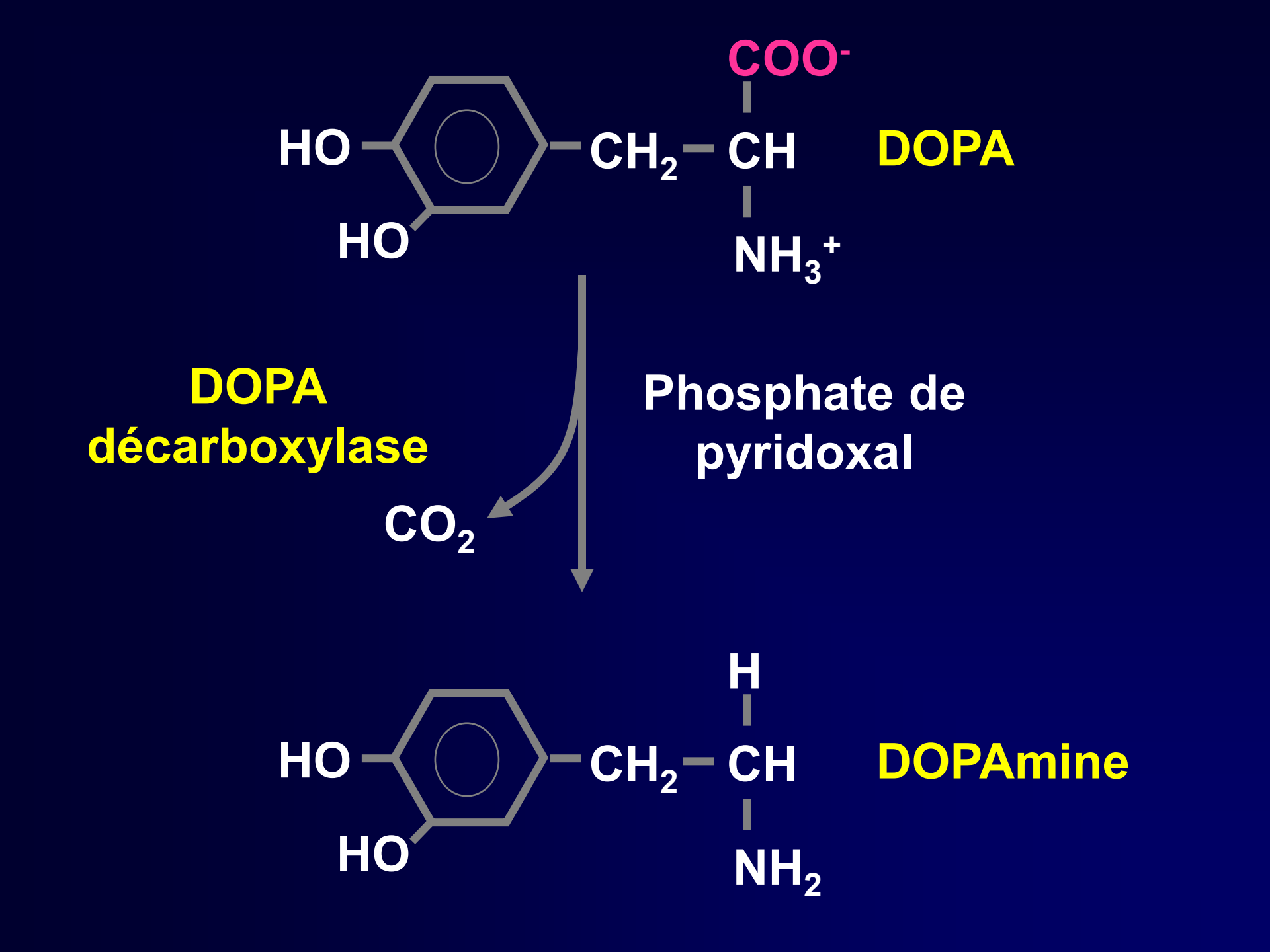
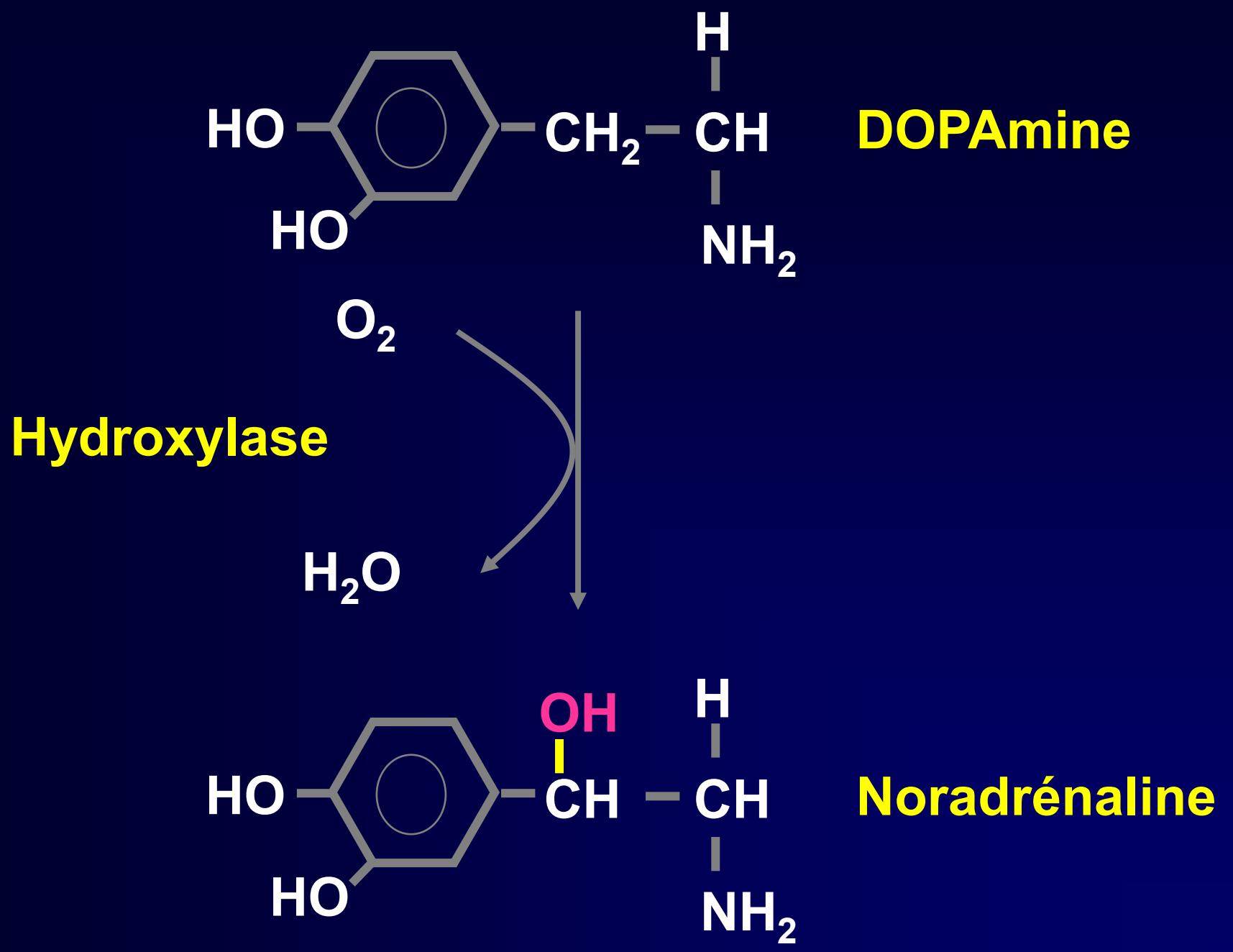
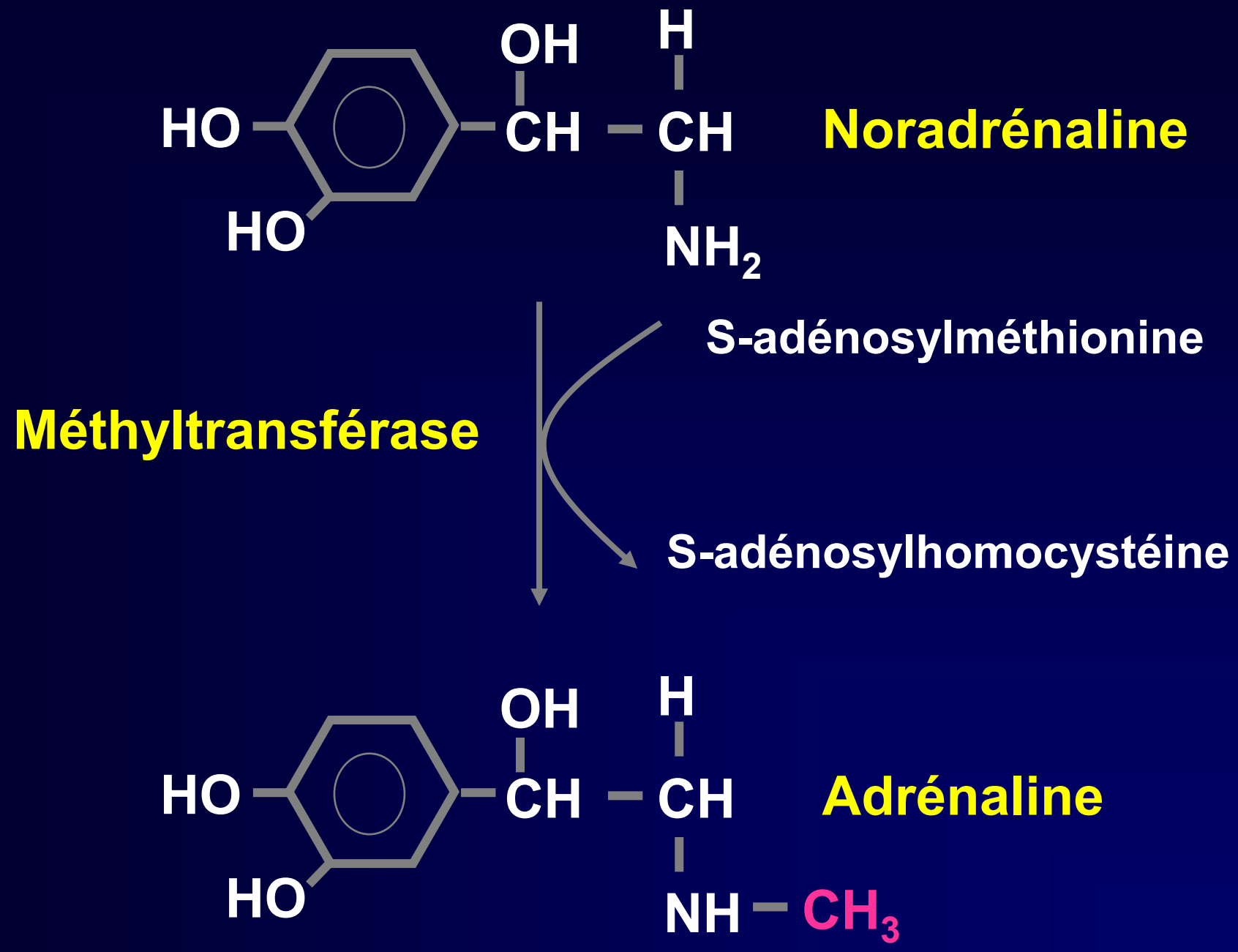
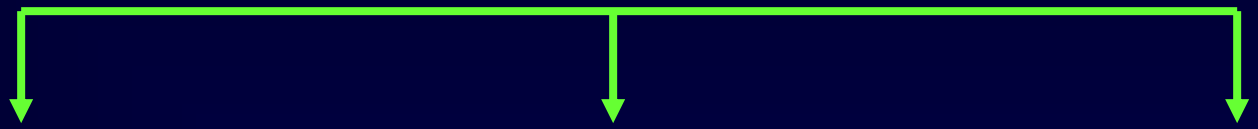
NH3+
O2
H4Bioptérine
Tyrosine
Hydroxylase
H2O
H2Bioptérine
COO-
HO
CH2
CH
DOPA
HO
NH3+


COO-
HO
CH2
CH
DOPA
HO
NH3+
DOPA
décarboxylase
Phosphate de
pyridoxal
CO2
H
HO
CH2
CH
DOPAmine
HO
NH2


HO
HO
CH2
CH
H
NH2
DOPAmine
O2
Hydroxylase
H2O
OH
HO
HO
CH
CH
H
NH2
Noradrénaline


OH
H
HO
CH
CH
Noradrénaline
HO
NH2
S-adénosylméthionine
Méthyltransférase
S-adénosylhomocystéine
OH
H
HO
CH
CH
Adrénaline
HO
NH
CH3

# Tyrosine



DOPA

Mélanine

Catécholamines

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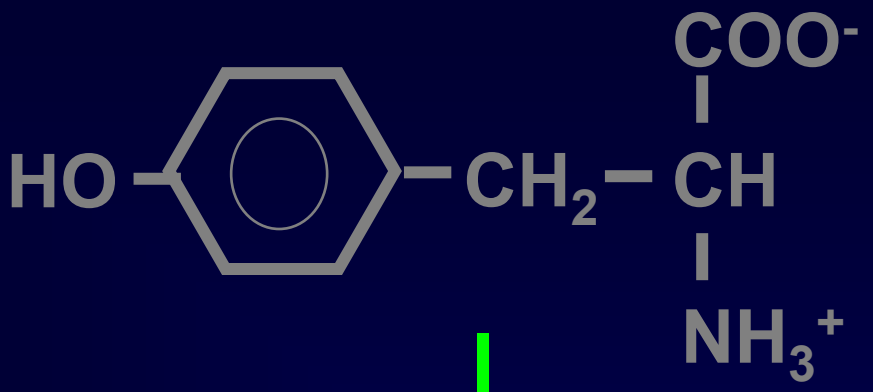
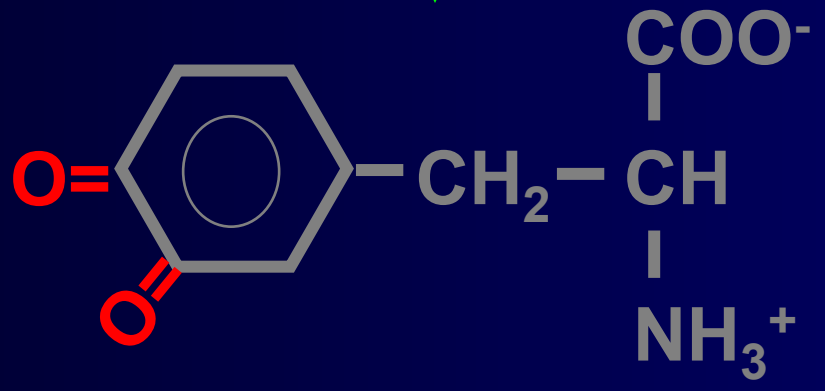



Tyrosine

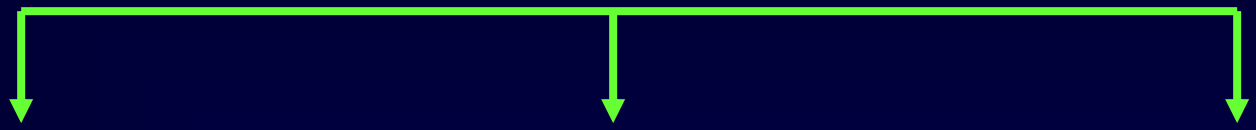
DOPA

Tyrosinase




Dopa Quinone

# Tyrosine



DOPA

Mélanine

**Hormones Thyroïdiennes**

Catécholamines

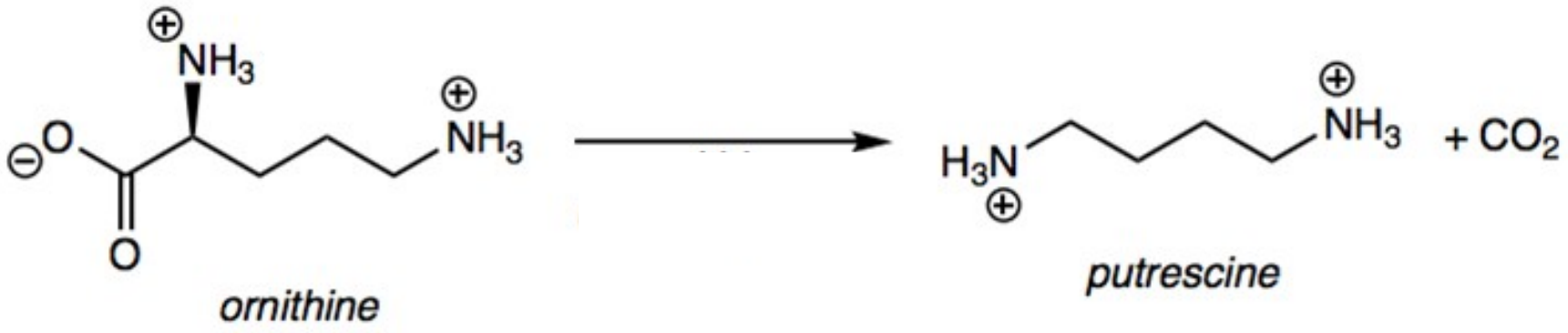
Dopamine

Noradrénaline

Adrénaline


QUIZ
TIME

L’ornithine peut-être transformée en putrescine (dont l’odeur est réellement nauséabonde)




1. Identifiez le type de réaction
2. Que représente la putrescine pour l’ornithine ?

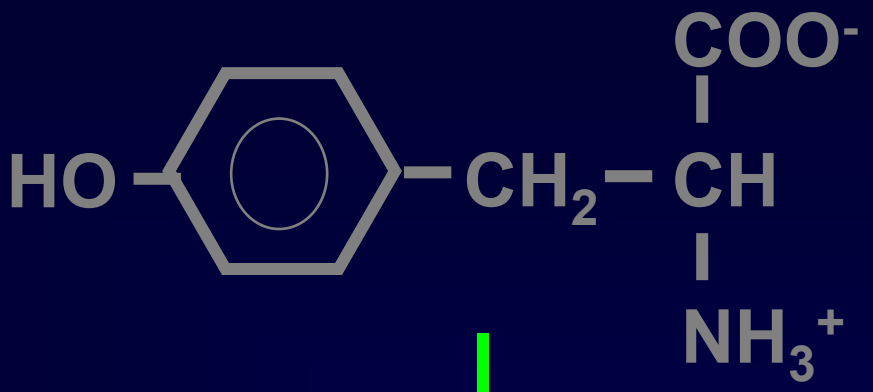
    1. Une Décarboxylation
    2. Son amine biogène

- Lequel des composés suivants n’est pas un dérivé de Tyrosine ?
  A. Hormones thyroïdiennes
  B. DOPA
  C. Noradrénaline
  D. Mélatonine
  E. Mélanine

- D

Votre patient est un albinos avec des cheveux blancs depuis la naissance.
Vous faites une biopsie de la peau de votre patient et vous cultivez ses mélanocytes.
Vous ajoutez de la tyrosine radioactive aux mélanocytes et vous constatez que la production de DOPAquinone radioactive n'est que de 1% de celle d'un sujet normal.

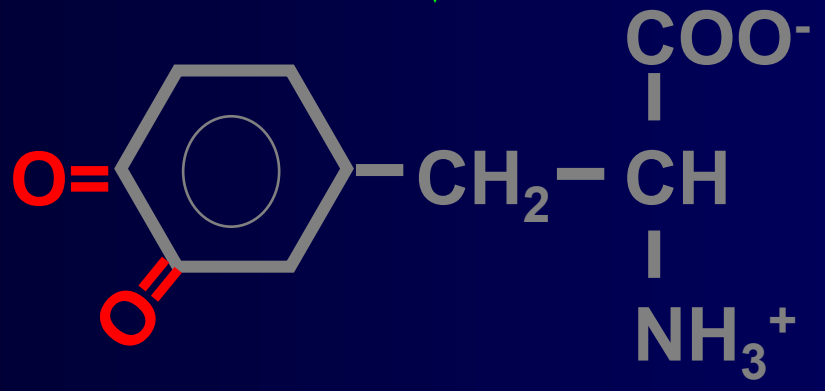
Quelle hypothèse évoquez-vous ?




Tyrosine

DOPA

Tyrosinase




Dopa Quinone

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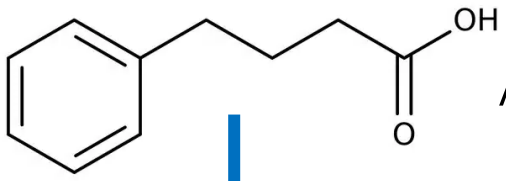
Quelle hypothèse évoquez-vous ?

- Un déficit en Tyrosinase

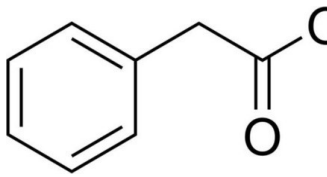
**L'acide Phénylbutyrique** est un traitement qui peut être donné dans les désordres du cycle de l'urée. Expliquez la logique de son utilisation




Acide Phényl-butyrique

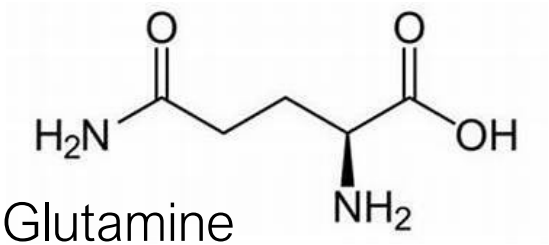
Bêta-oxydation

Si désordre du cycle de l'urée, l'ammoniaque s'accumule et la concentration de Glutamine augmente. Donner de l'acide phénylbutyrique au patient permet d'augmenter l'élimination de Glutamine

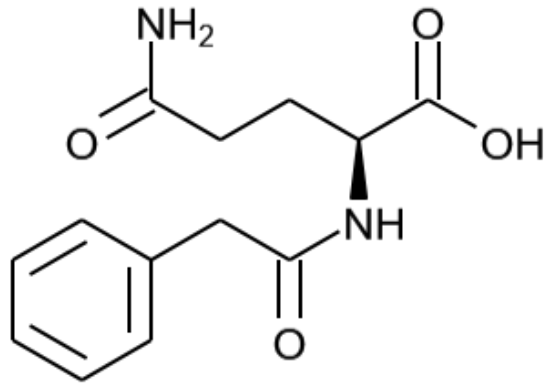



Acide Phényl-acétique

+



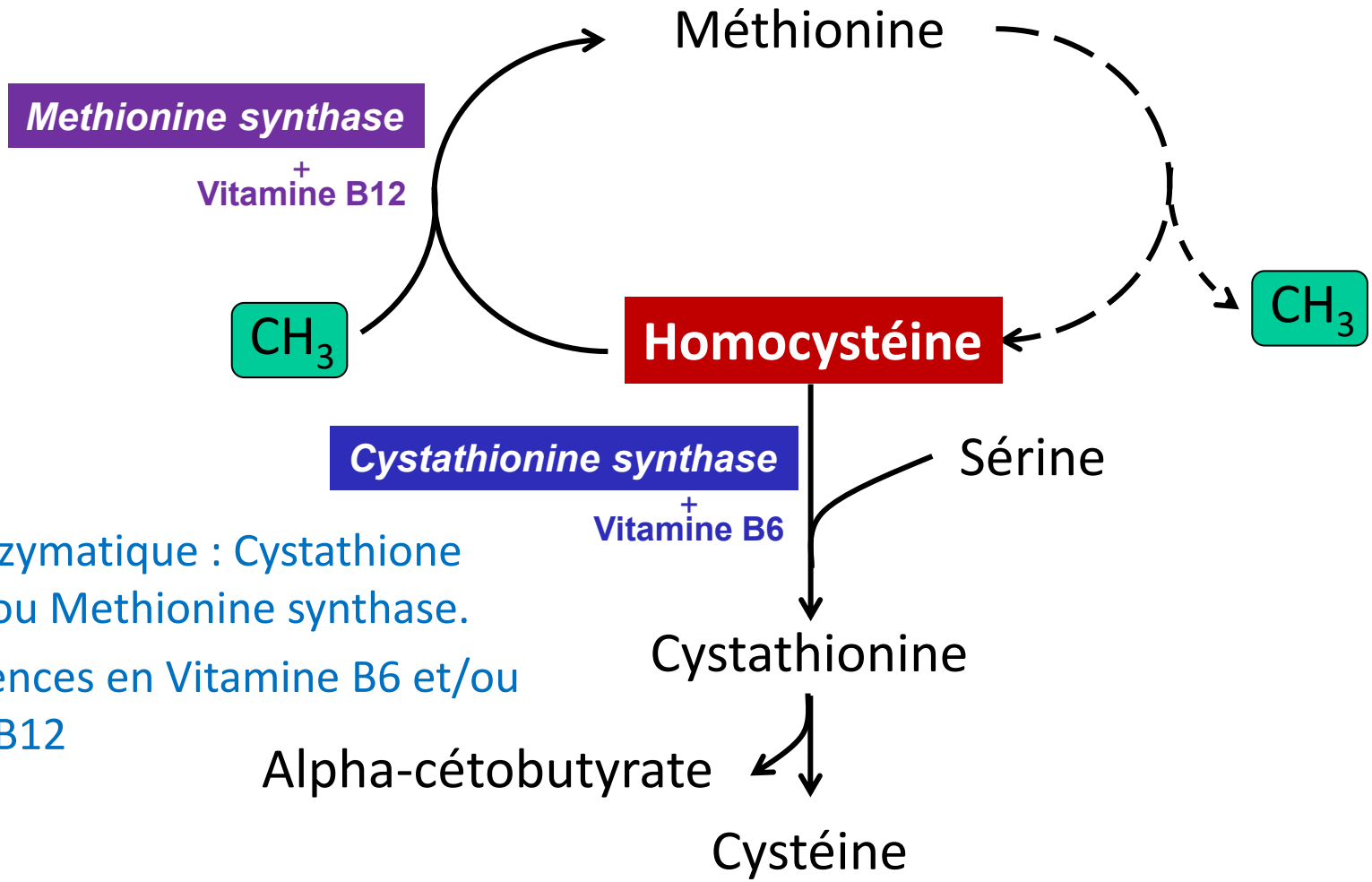

Glutamine



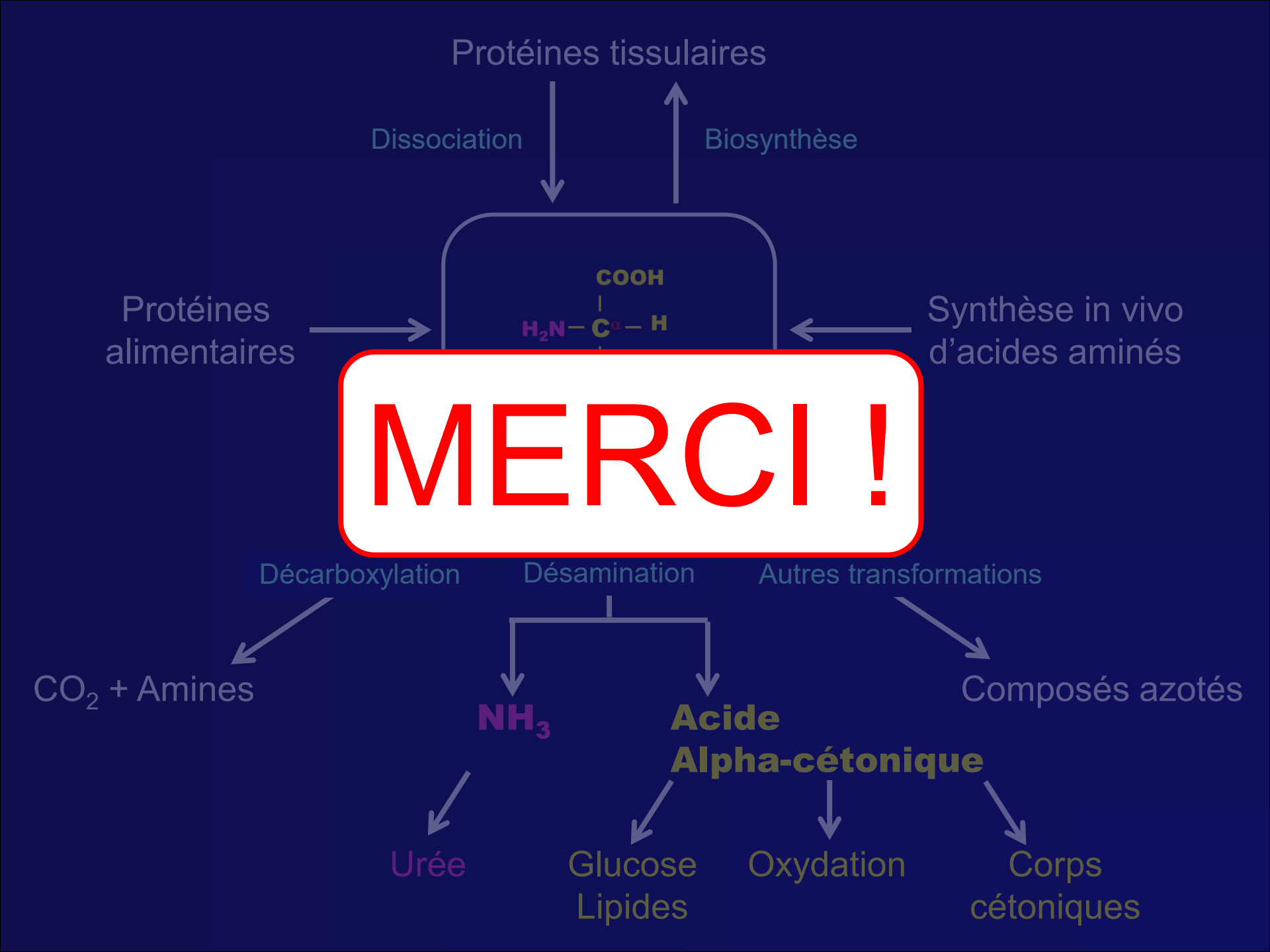

Phényl-acétyl-glutamine

*Eliminé par le rein*

L'**Homocystéine** est un acide aminé soufré de l'organisme humain. Cependant, son accumulation (hyper-Homocystéinémie) est toxique. Sur la base de son métabolisme, avancez des mécanismes d'hyper-homocyctéinémies




- Défaut enzymatique : Cystathione synthase ou Methionine synthase.
- Et/ou carences en Vitamine B6 et/ou Vitamine B12


Protéines tissulaires
Dissociation
Biosynthèse
Protéines alimentaires
Synthèse in vivo d'acides aminés
MERCI !
Décarboxylation
Désamination
Autres transformations
$CO_2$ + Amines
$NH_3$
Acide Alpha-cétonique
Composés azotés
Urée
Glucose Lipides
Oxydation
Corps cétoniques