

M Ű E G Y E T E M 1 7 8 2

A mitokondriális DNS mutációk hatása az oxidatív fehérje foldingra és a gyógyszertoxicitásra

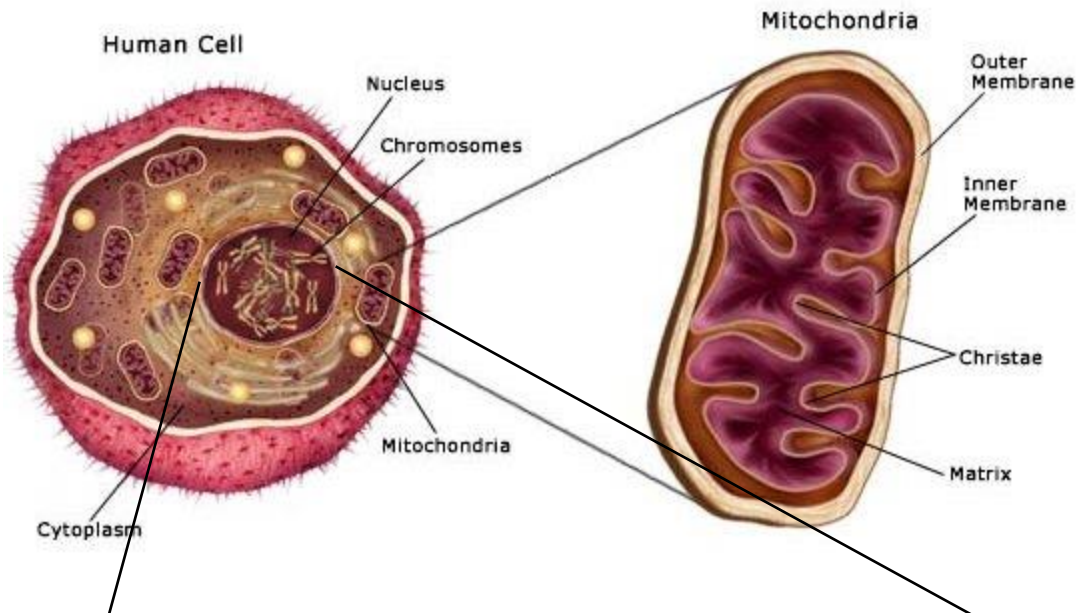
Mandl József

Semmelweis Egyetem Orvosi Vegytani
Molekuláris Biológiai és Patobiokémiai
Intézet

Szarka András

BME Alkalmazott Biotechnológia és
Élelmiszer-tudományi Tanszék
Biokémiai és Molekuláris Biológiai
Laboratórium

2015. március 21.



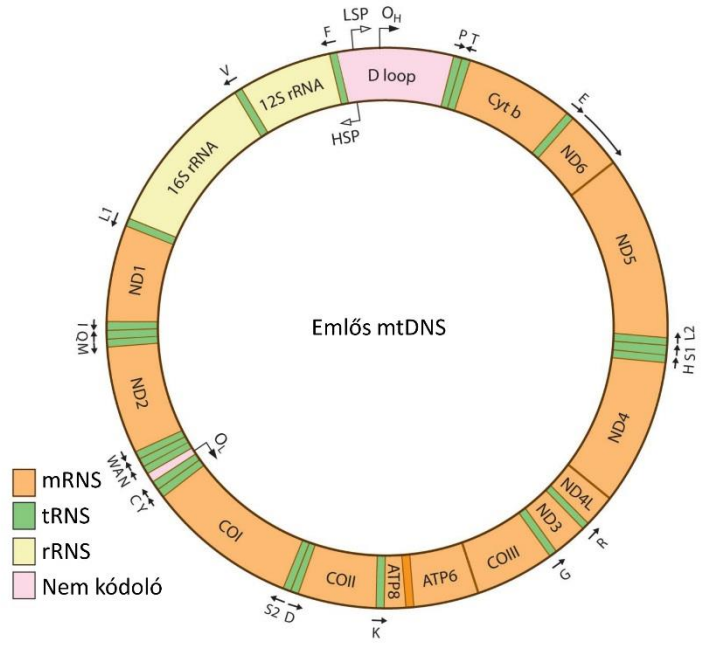
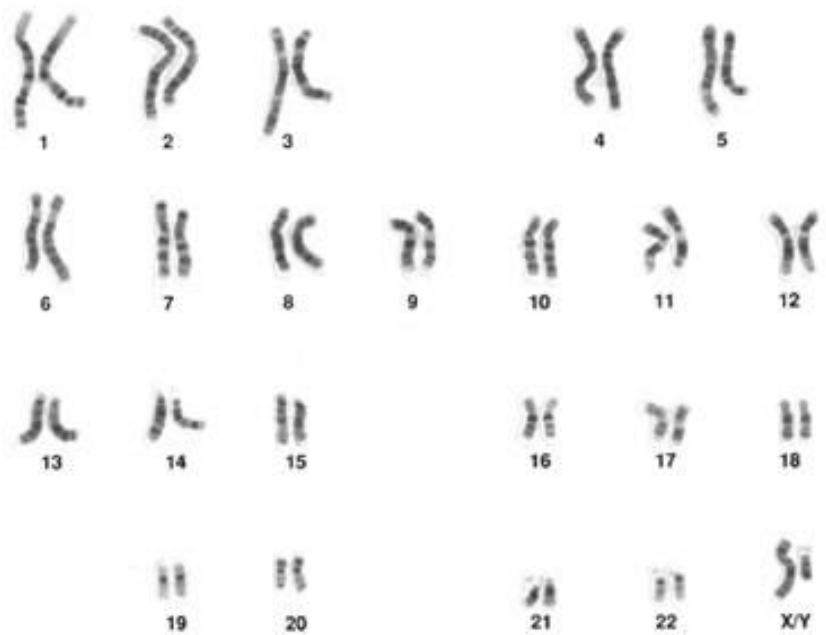
Mitokondriális genom

16569 bp 37 gén

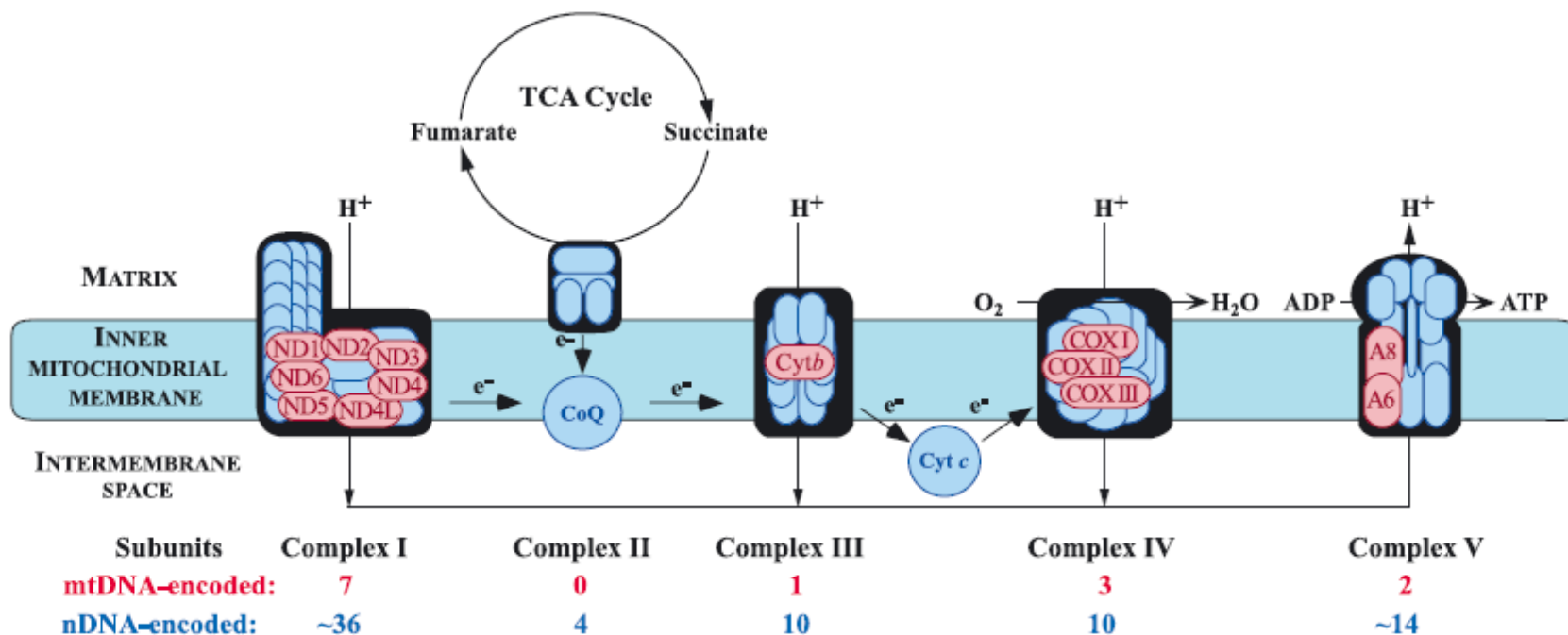
13 gén fehérjét kódol

2 gén rRNS-t

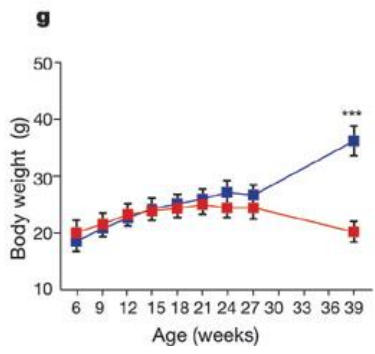
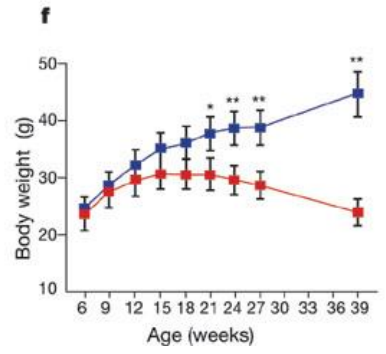
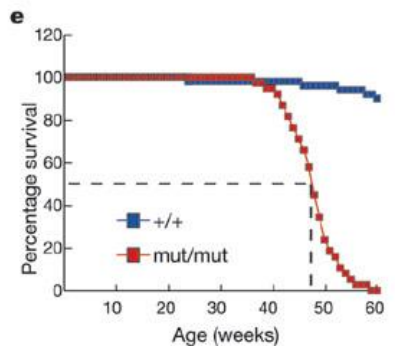
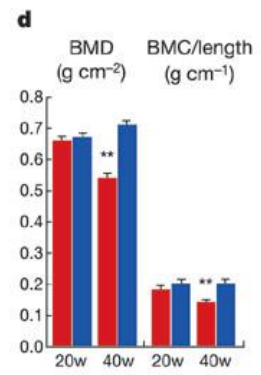
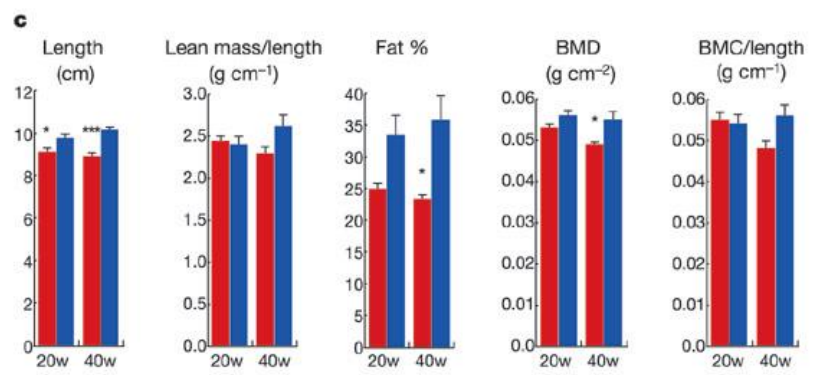
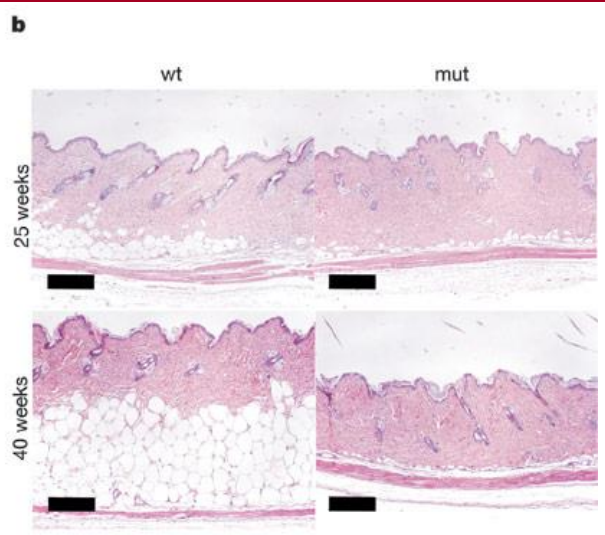
22 gén tRNS-t



1988 első mtDNS mutáció (betegség leírása)

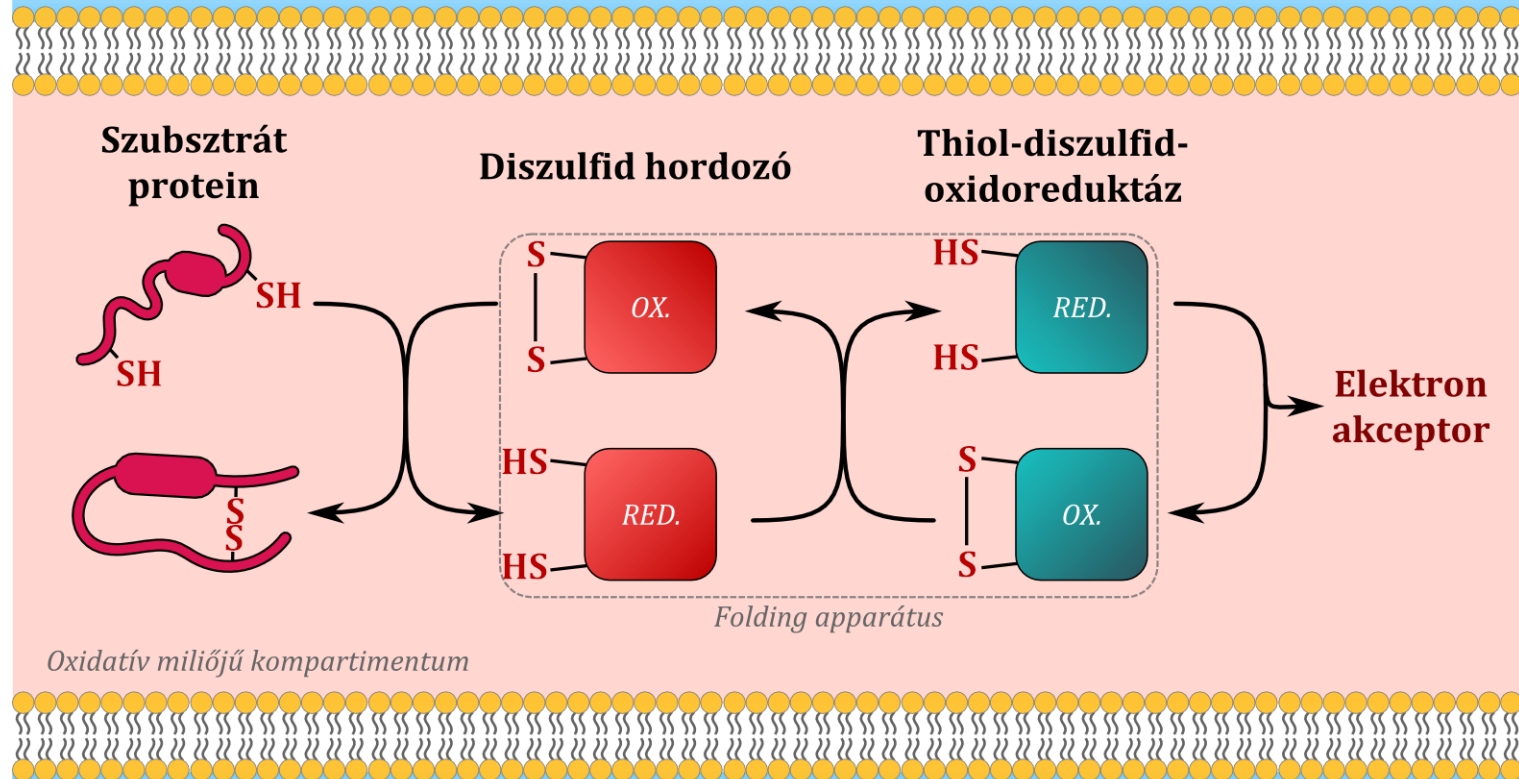


Életkorral felszaporodó mtDNS mutációk

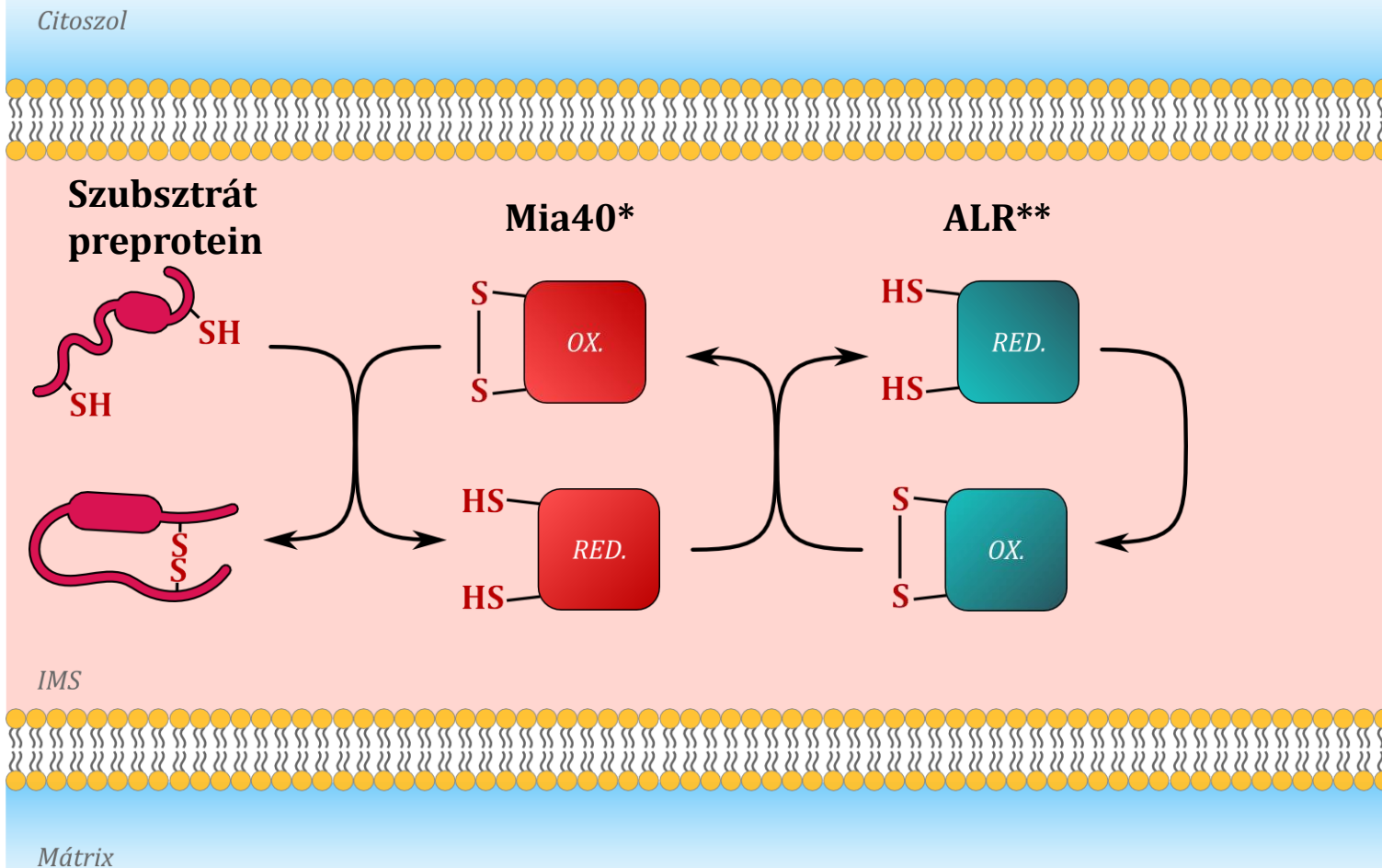


Forrás:
Trifunovic et al
 Nature 2004. 429:417-423.

Az oxidatív fehérjefolding általános sémája



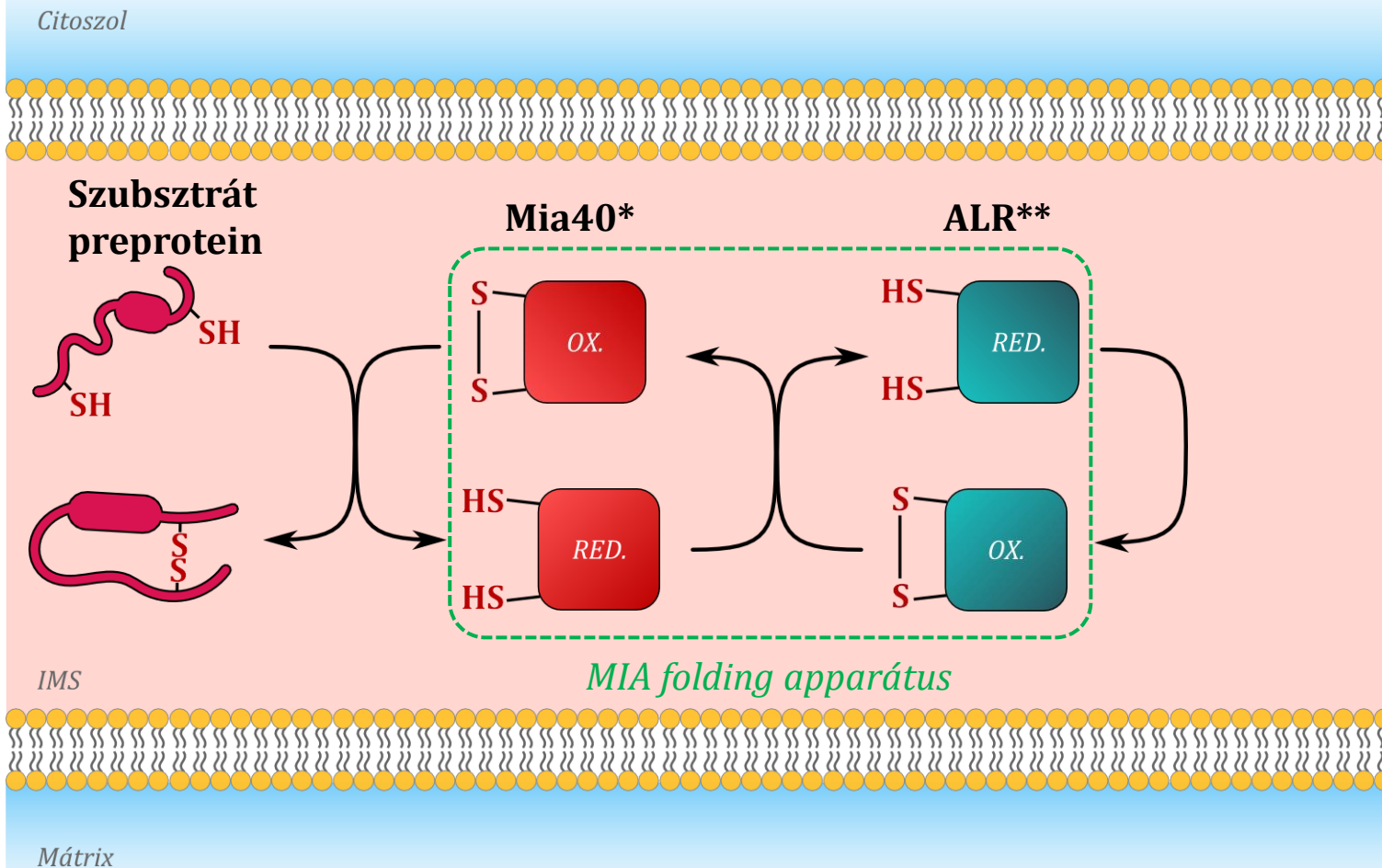
Oxidatív fehérjefolding a mitokondriális IMS-ben



*: Mitochondrial intermembrane space Import and Assembly protein 40

** : Augmenter of Liver Regeneration

Oxidatív fehérjefolding a mitokondriális IMS-ben

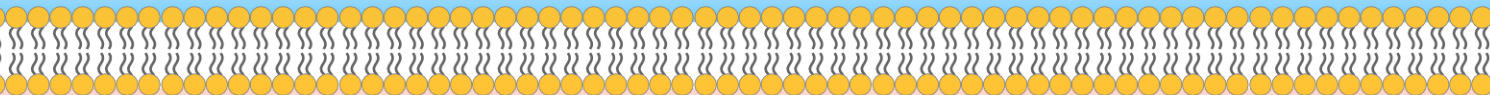


*: Mitochondrial intermembrane space Import and Assembly protein 40

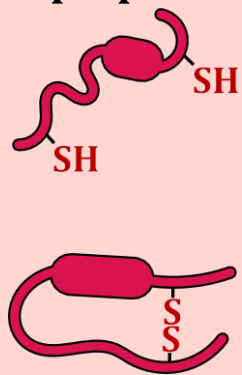
** : Augmenter of Liver Regeneration

Oxidatív fehérjefolding a mitokondriális IMS-ben

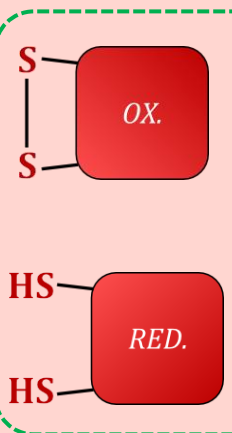
Citoszol



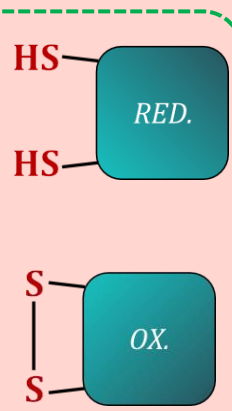
Szubsztrát preprotein



Mia40



ALR



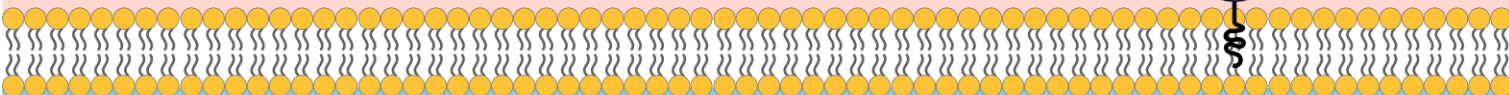
MIA folding apparátus

Cyt-c

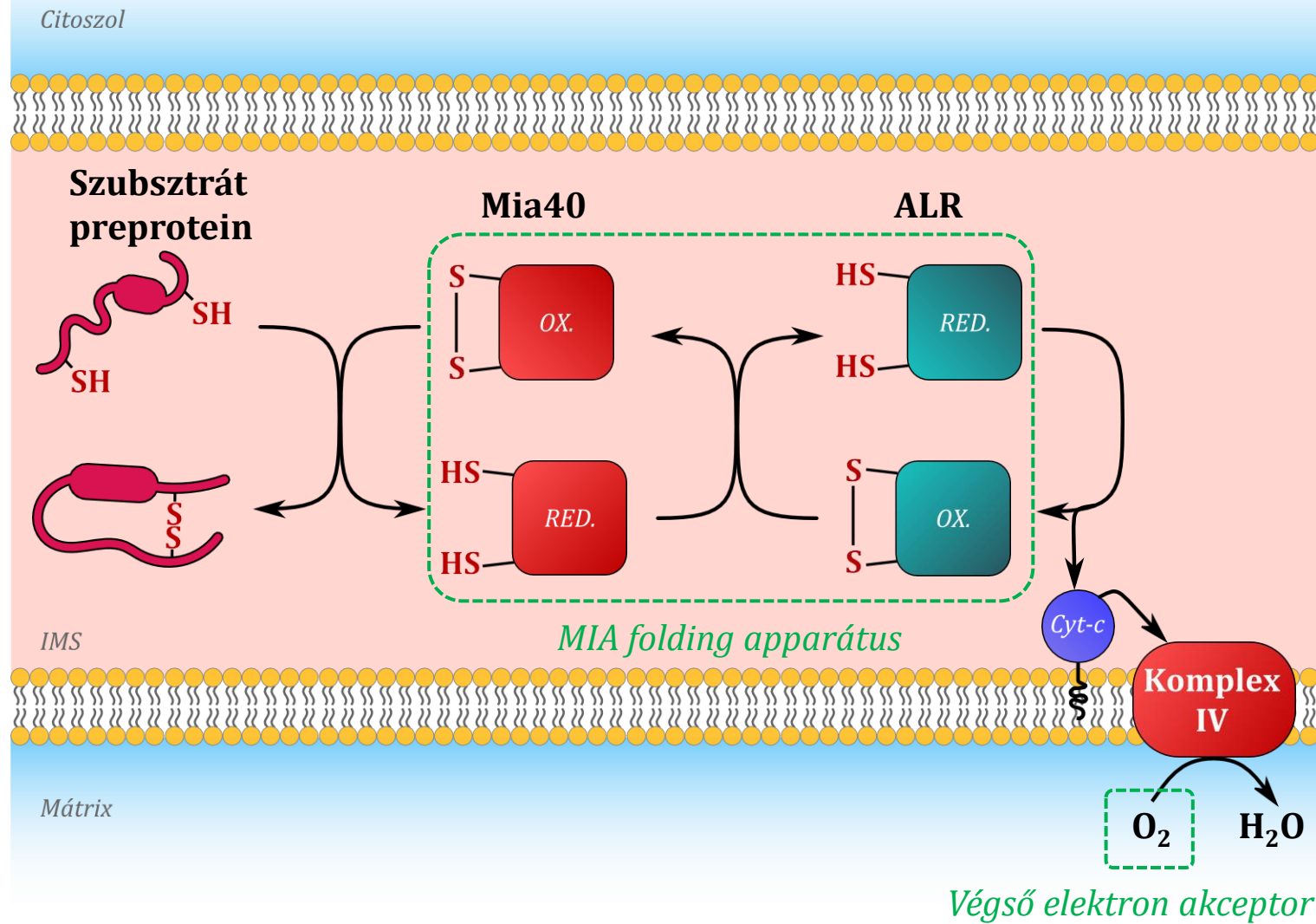


IMS

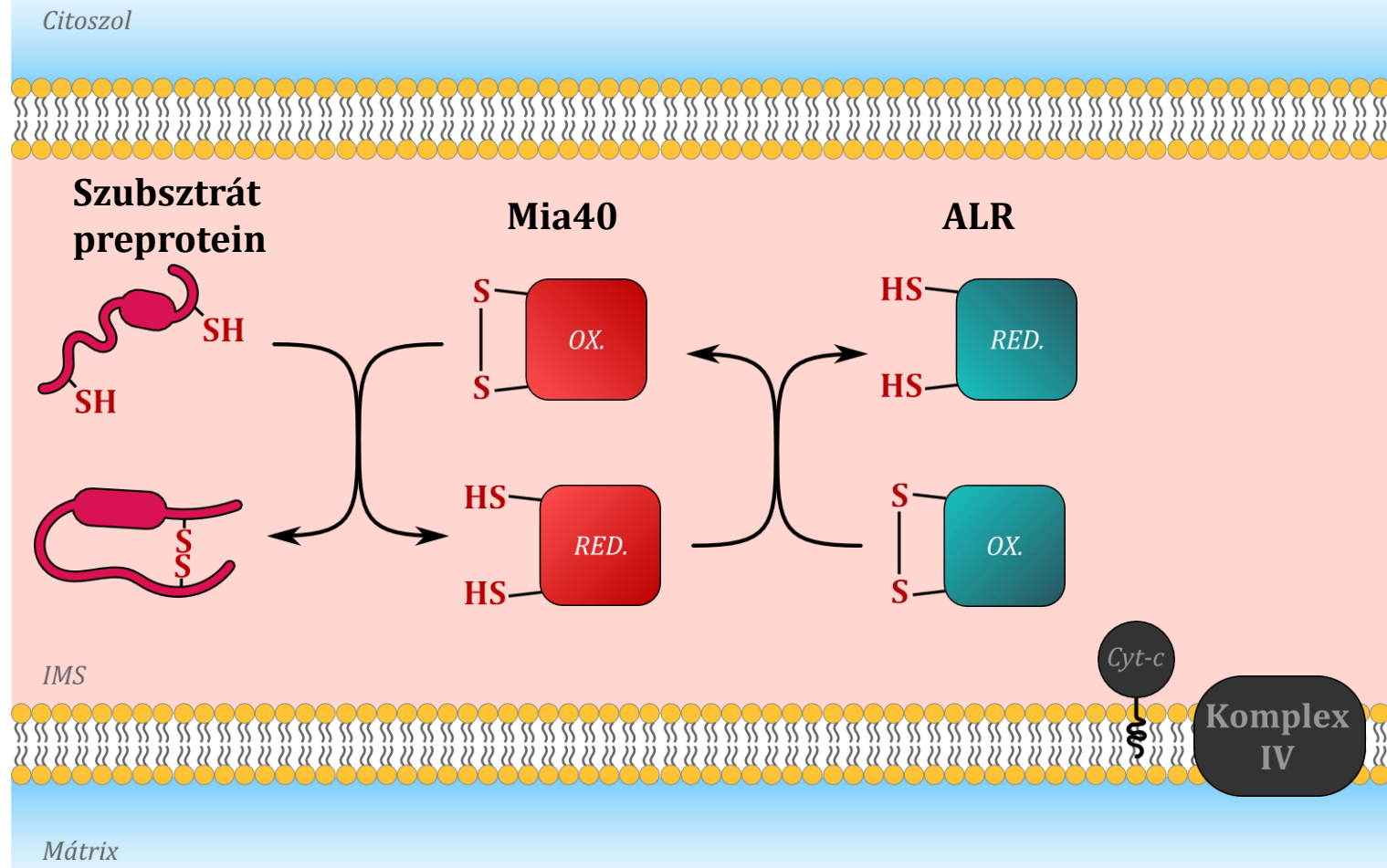
Mátrix



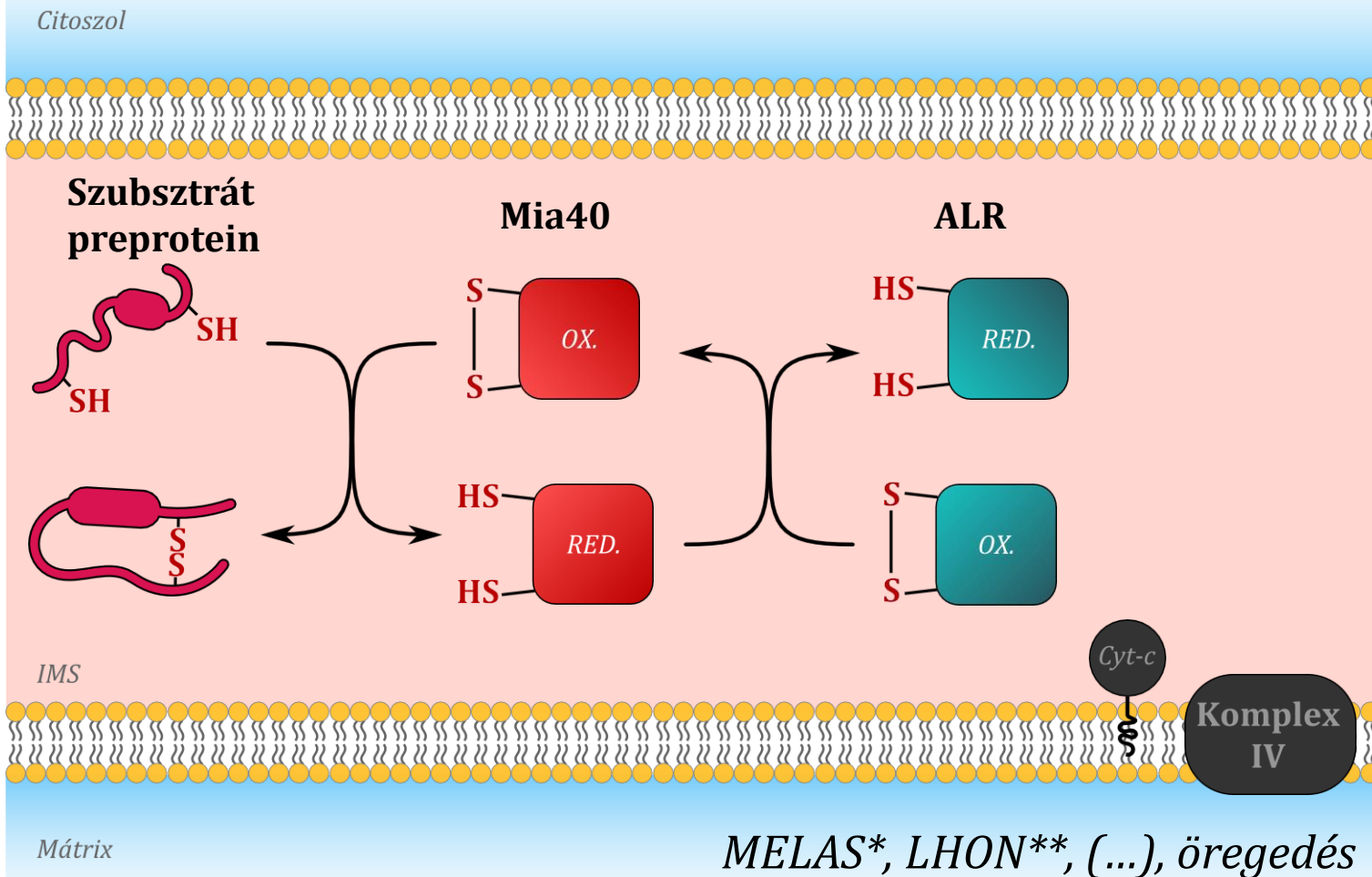
Oxidatív fehérjefolding a mitokondriális IMS-ben



mtDNS károsodás okozta funkcióvesztés



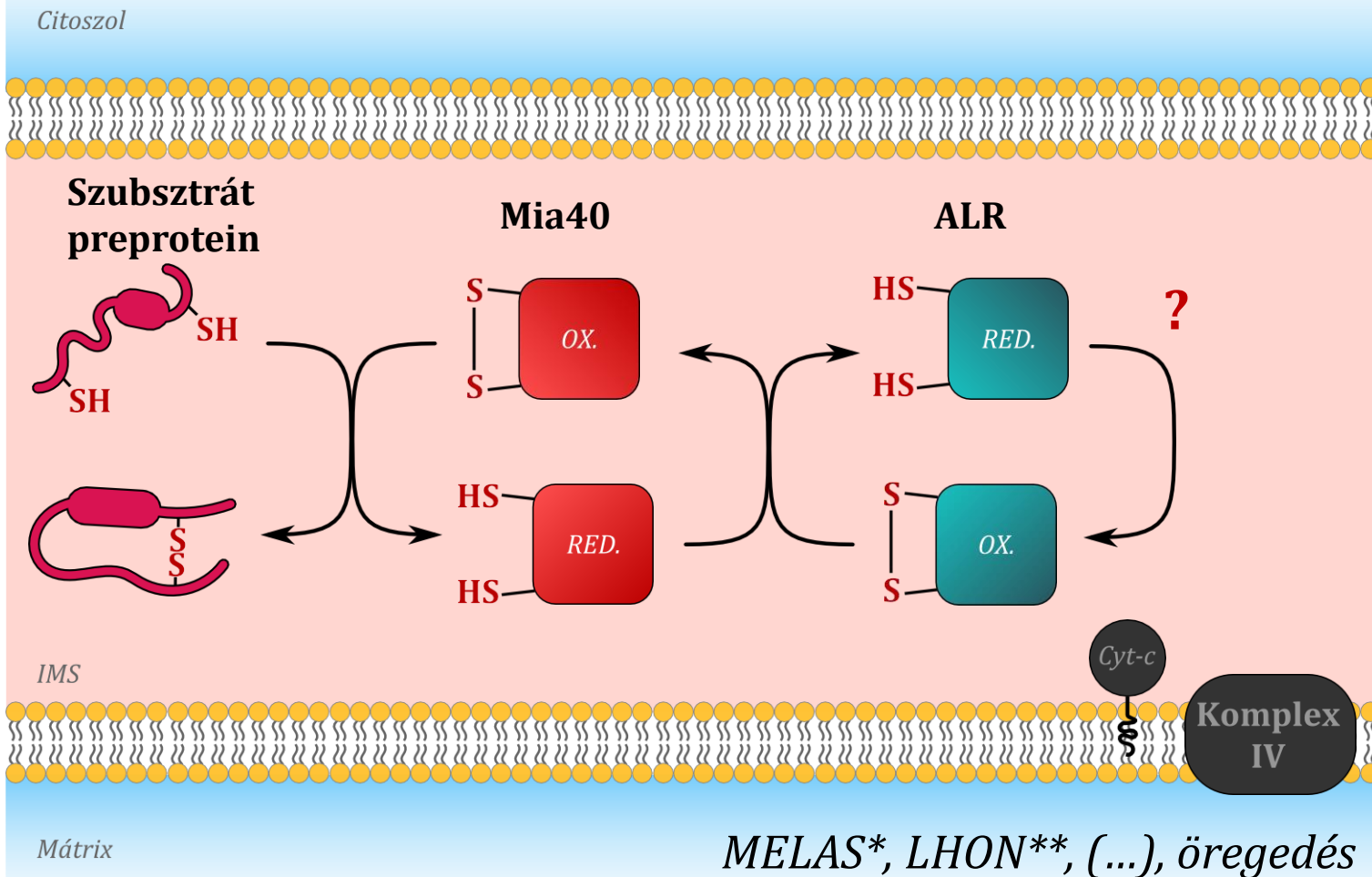
mtDNS károsodás okozta funkcióvesztés



*: Mitochondrial Encephalomyopathy, Lactic Acidosis, and Stroke-like episodes

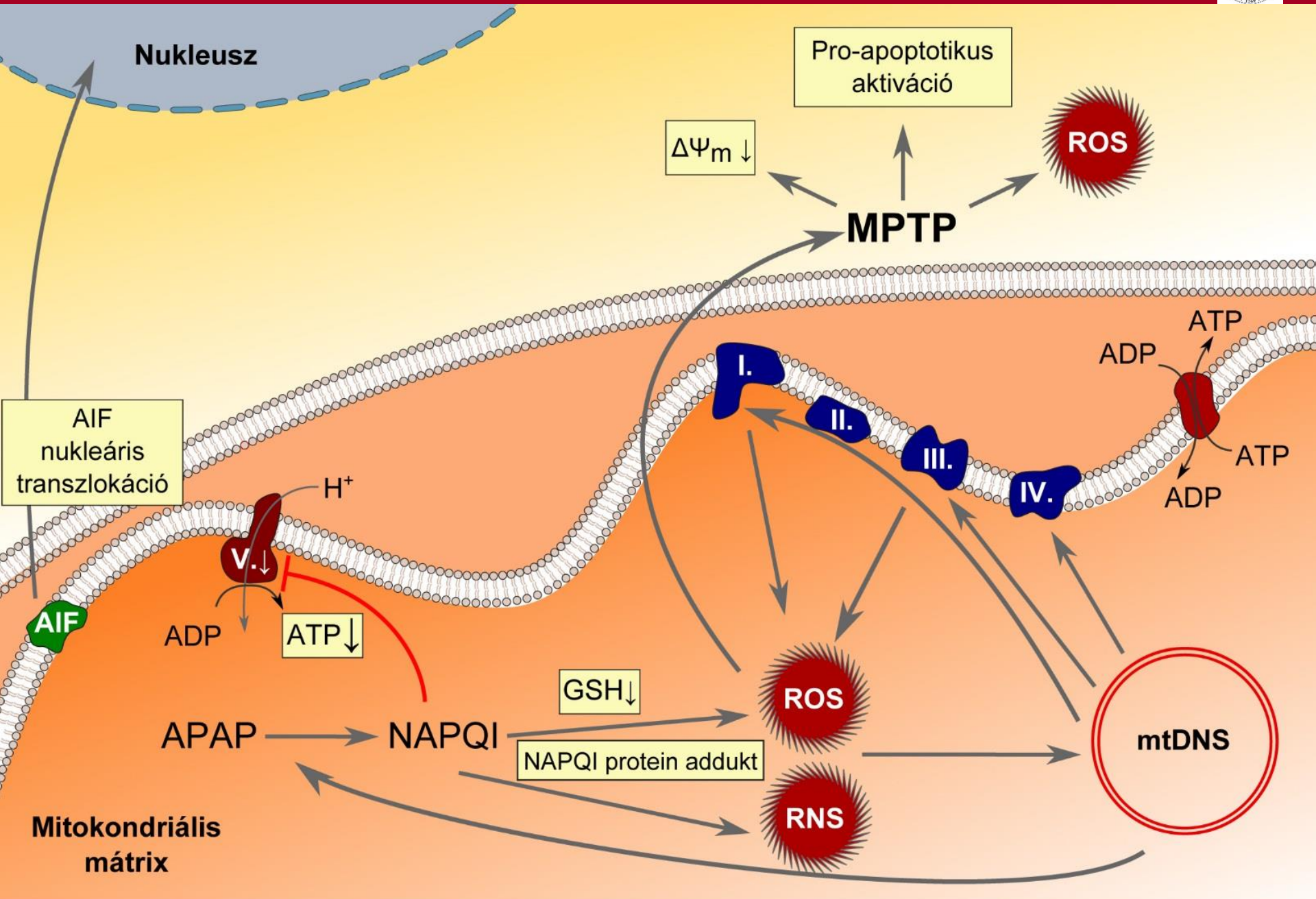
** : Leber's Hereditary Optic Neuropathy

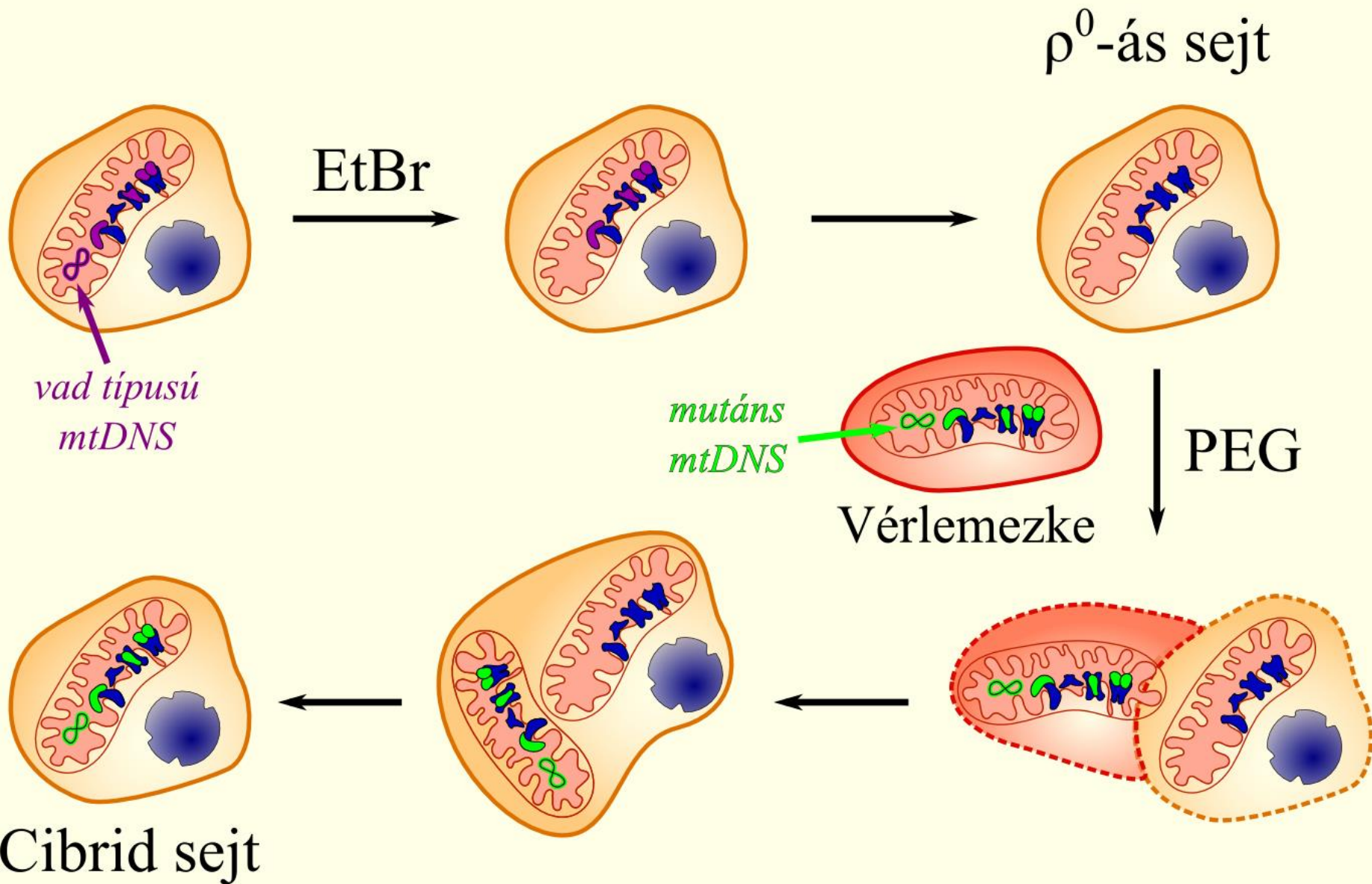
mtDNS károsodás okozta funkcióvesztés

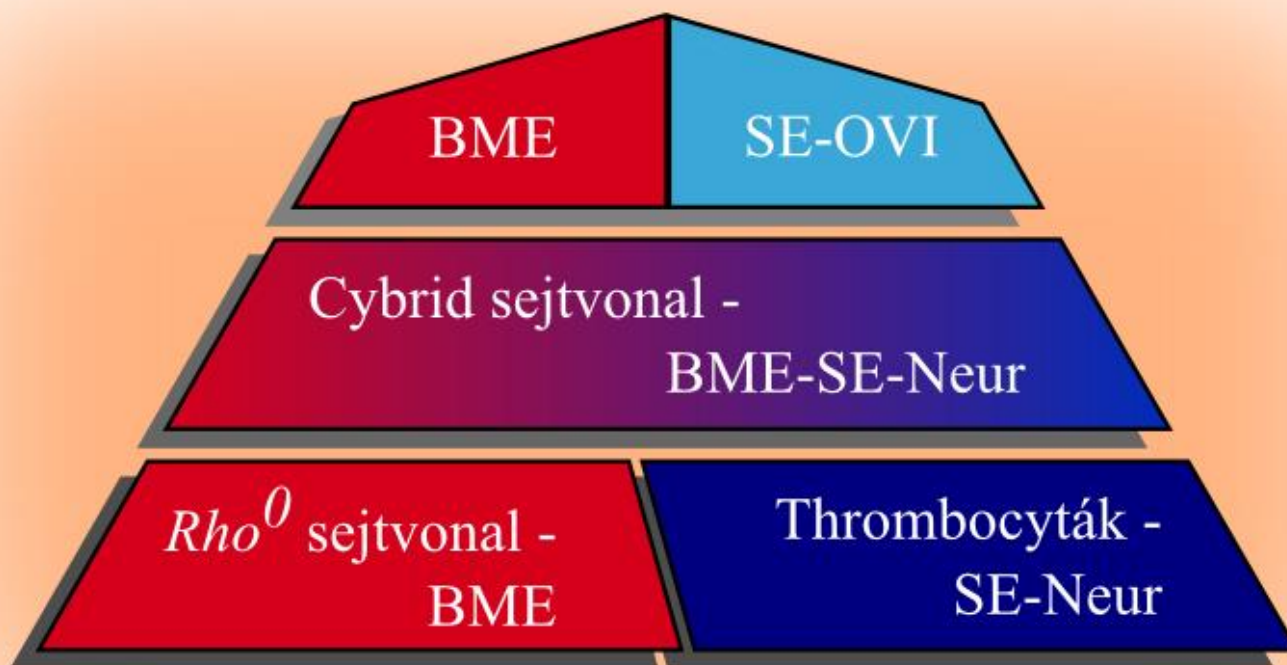


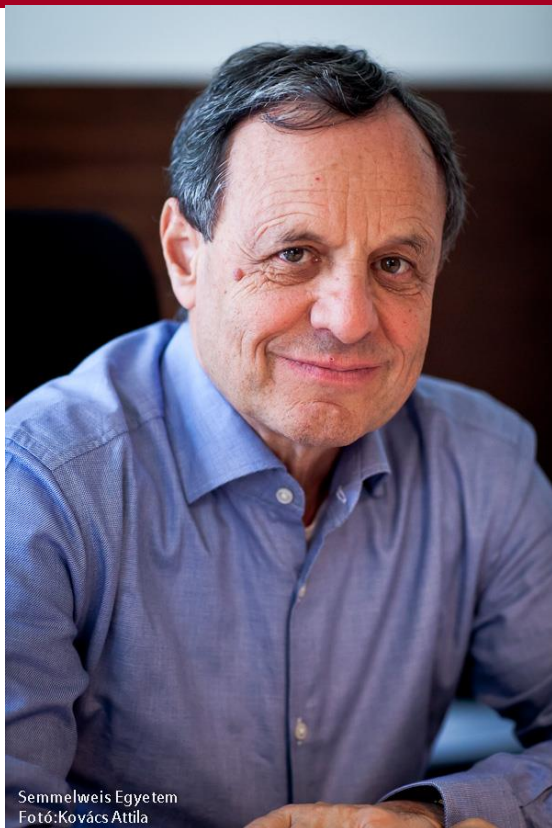
*: Mitochondrial Encephalomyopathy, Lactic Acidosis, and Stroke-like episodes

** : Leber's Hereditary Optic Neuropathy









Semmelweis Egyetem
Fotó: Kovács Attila

Mandl József

Semmelweis Egyetem Orvosi Vegytani
Molekuláris Biológiai és Patobiokémiai Intézet

**BME Alkalmazott
Biotechnológia és Élelmiszertudományi Tanszék
Biokémiai és Molekuláris
Biológiai Laboratórium**

