INTRODUCTION TO MITOCHONDRIA

Background

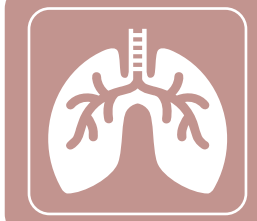
Prehistoric origin when bacteria and eukaryotic cells formed a symbiotic relationship

Like bacteria – they contain their own DNA and have an outer and inner membrane and are capable of fusion and fission

Hundreds to thousands per cell depending on the energy need

Role

GENERATE ATP FOR ALL CELLULAR PROCESSES USING BYPRODUCTS OF CARBOHYDRATE, FAT AND PROTEIN METABOLISM AND THE ELECTRON-TRANSPORT-CHAIN



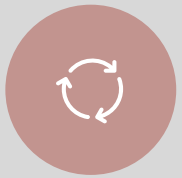
CREATE AND COMBAT REACTIVE OXYGEN SPECIES

PARTICIPATE IN APOPTOSIS AND CELL DEATH CASCADE

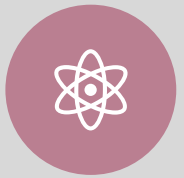
MODULATE THE INFLAMMASOME

Essential Organelle Functions

ATP Synthesis via the Electron Transport Chain



Fatty Acid Oxidation



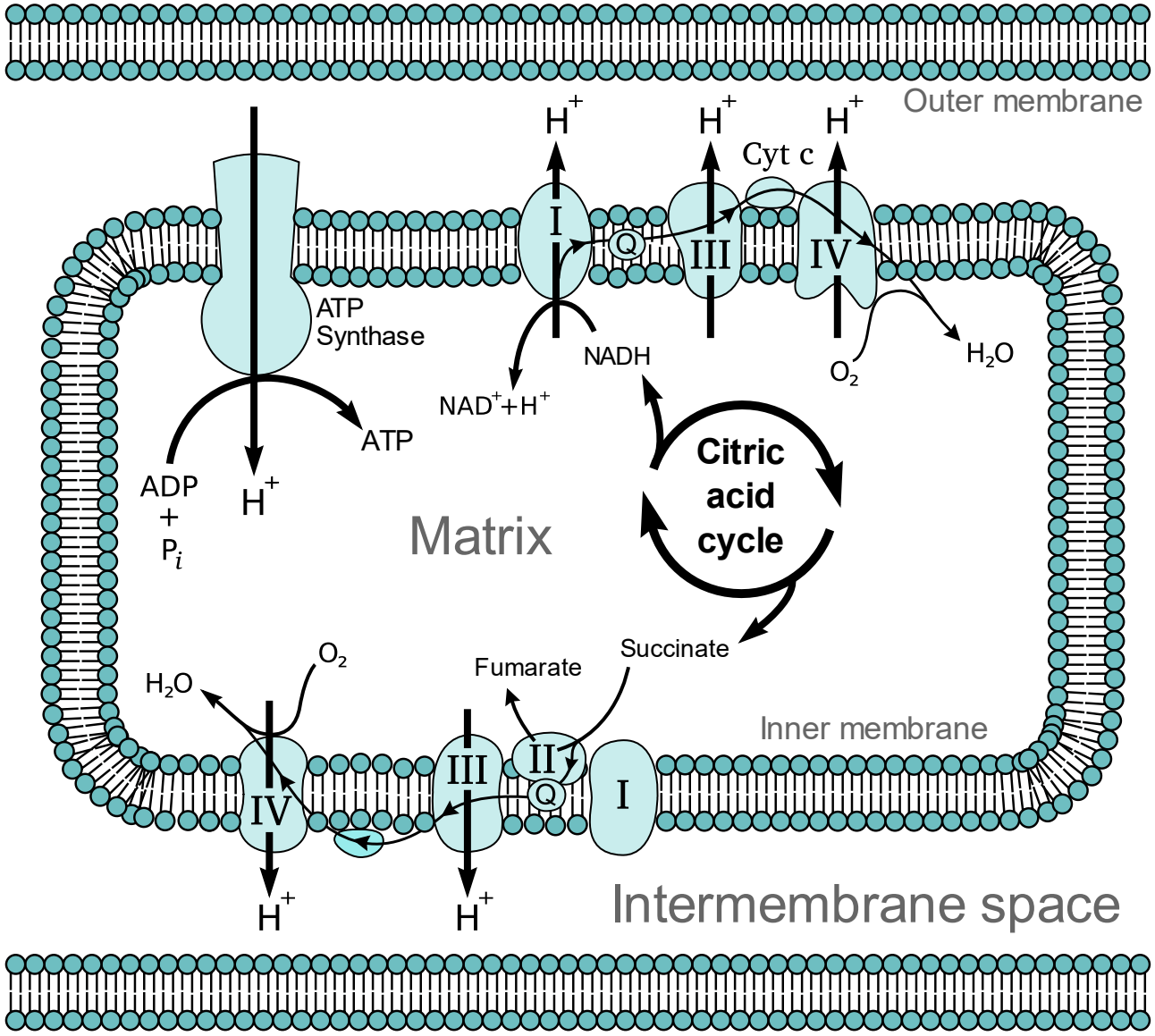
TCA Cycle

Transport of Metabolites and Ions

Biosynthesis of Iron-Sulfur Clusters and Cofactors

Mitochondrial Gene Expression and Translation

Mitochondrial Protein Import and Processing



THE 5-COMPLEX ELECTRON TRANSPORT CHAIN

Mitochondria Need a Dual Genome

MITOCHONDRIAL DNA

NUCLEAR DNA

Nuclear DNA

Inherited from each parent

>1500 genes needed for mitochondrial formation and function

More than 250 of these genes are linked to human disease

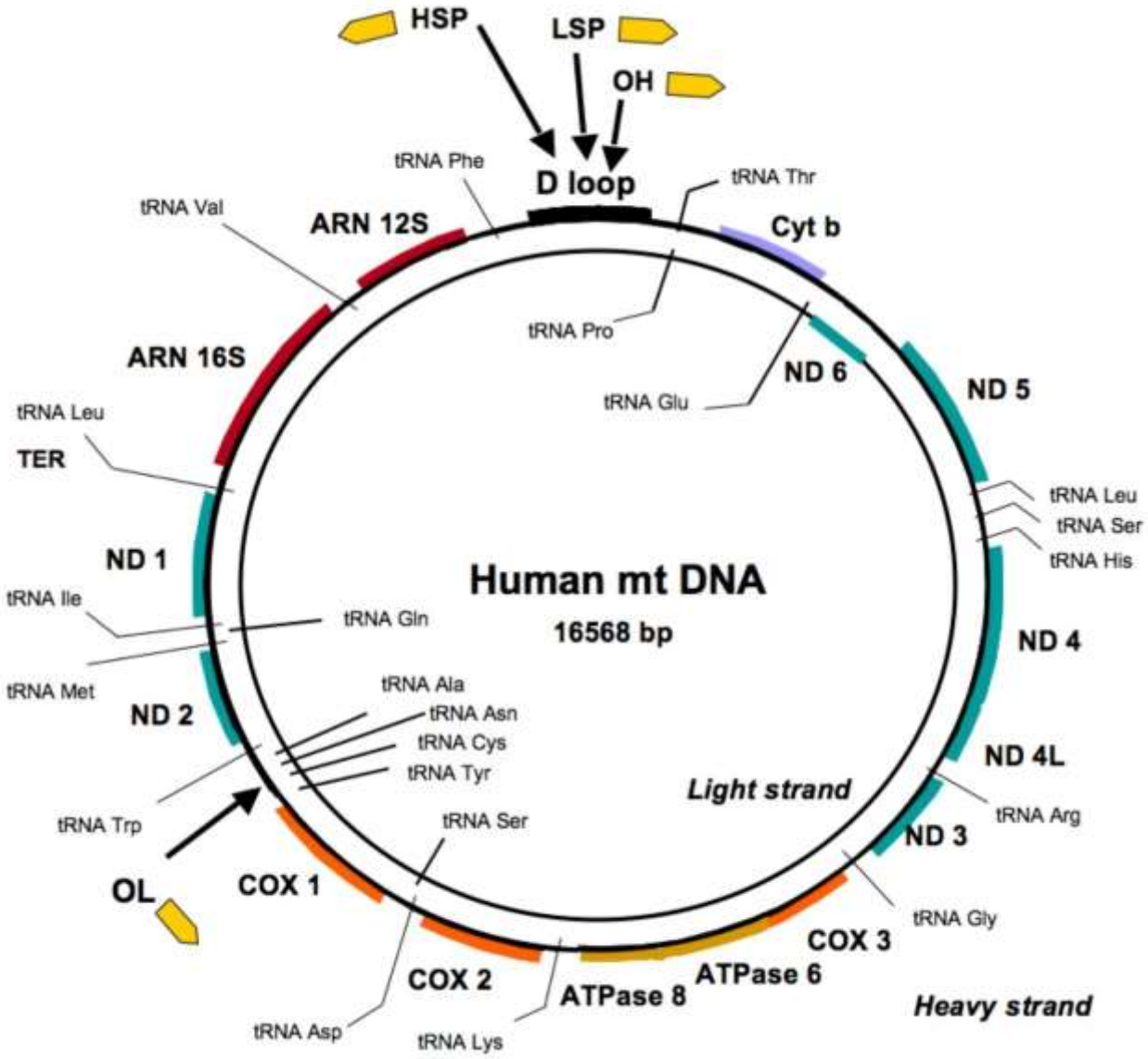
Mitochondrial DNA (mtDNA)

TRANSMITTED FROM MOTHER TO CHILD

CIRCULAR WITH 16,569 BASE PAIRS

CODE FOR SOME ETC STRUCTURAL COMPONENTS, RRNA AND TRNA

MULTIPLE SLIGHTLY VARIED COPIES PER MITOCHONDRIA



Heteroplasmy

- Varied copies of mtDNA are inherited by each cell
- These varied copies of mtDNA in a cell impact function based on the integrity of their mtDNA code
- Each cell/organ may then have variable energy output