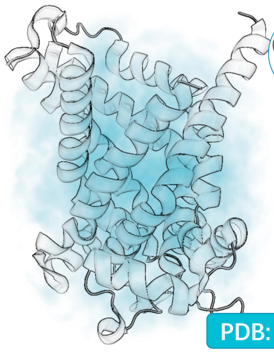
AAC1

ADP/ATP Carrier

Inner Membrane



PDB: 1OKC

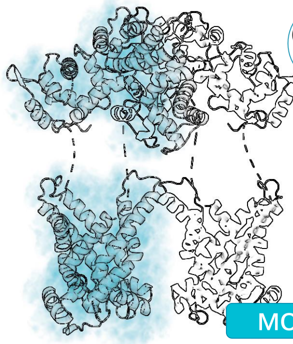
Without this tiny transport protein we'd be unable to get any ADP to make ATP, or to export the ATP that had been made!

Amino Acids: 298
Mutations: 115
Publications: 913
Chromosome: 4
MBU Rating: 66

Citrin

Aspartate-Glutamate Carrier

Inner Membrane



MODEL

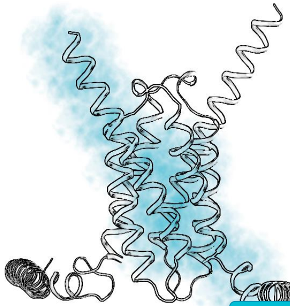
Also known as the aspartate-glutamate carrier, this mitochondrial transporter can be turned on/off with calcium!

Amino Acids: 678
Mutations: 377
Publications: 800
Chromosome: 7
MBU Rating: 54

MPC1

Mitochondrial Pyruvate Carrier 1

Inner Membrane



MODEL

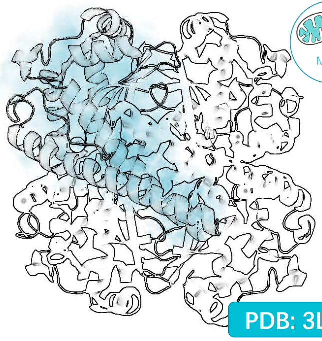
MPC1 is one of the two subunits which makes up a fully-functioning mitochondrial pyruvate carrier.

Amino Acids: 130
Mutations: 43
Publications: 283
Chromosome: 6
MBU Rating: 45

SOD2

Superoxide Dismutase 2

Matrix



PDB: 3LSU

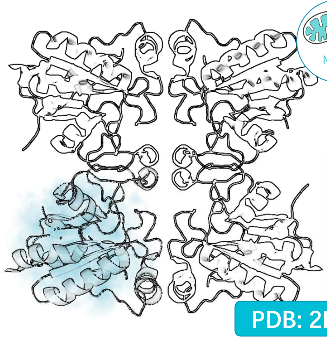
SOD2, or superoxide dismutase, is responsible for removing dangerous reactive oxygen species. Each one has four subunits.

Amino Acids: 222
Mutations: 189
Publications: 4,995
Chromosome: 6
MBU Rating: 29

GPx1

Glutathione Peroxidase 1

Matrix



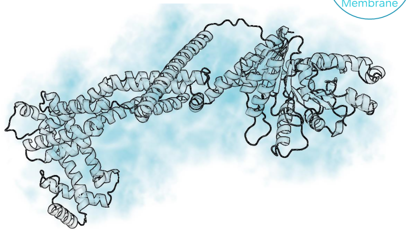
PDB: 2F8A

Glutathione peroxidase, or GPx1, removes dangerous hydrogen peroxide (bleach!) by reducing it to water.

Amino Acids: 203
Mutations: 124
Publications: 2,741
Chromosome: 3
MBU Rating: 11

OPA1

Inner Membrane



MODEL

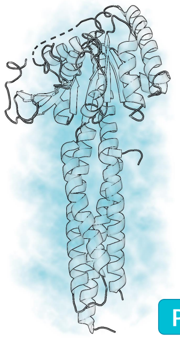
OPA1 plays a key role in the regulation of mitochondrial fission (when they divide) and fusion (when two mitos join together)

Amino Acids: 960
Mutations: 534
Publications: 1,583
Chromosome: 3
MBU Rating: 52

MFN1

Mitofusin 1

Outer Membrane



PDB: 5GNS

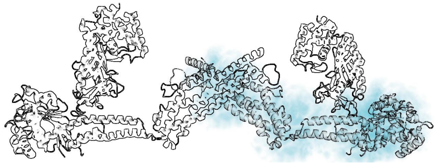
Mitofusin 1, or MFN1, helps the outer membrane of two different mitochondria to join together.

Amino Acids: 741
Mutations: 350
Publications: 852
Chromosome: 3
MBU Rating: 38

Drp1

Dynamin-Related Protein 1

Outer Membrane



PDB: 5WP9

Drp1 in the cytoplasm is recruited to the outer mitochondrial membrane to allow mitochondria to divide (fission).

Amino Acids: 736
Mutations: 220
Publications: 2,638
Chromosome: 12
MBU Rating: 58

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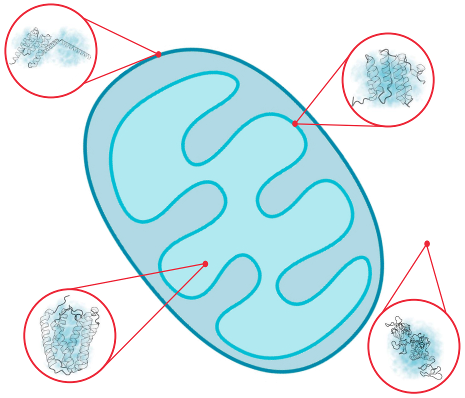

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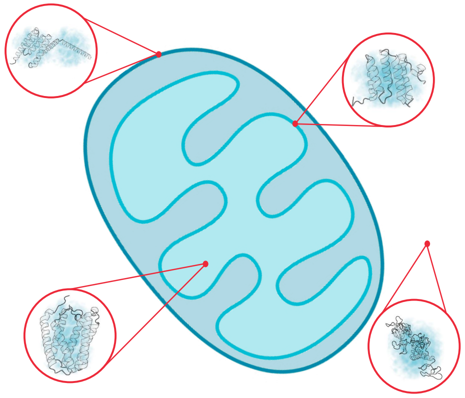

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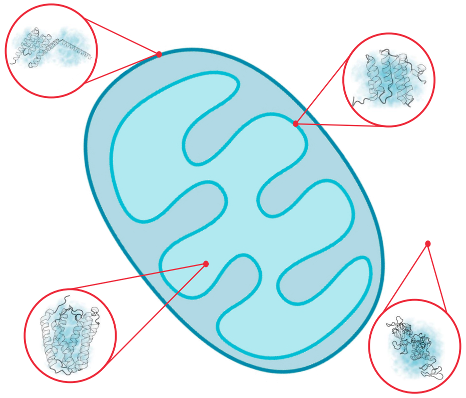

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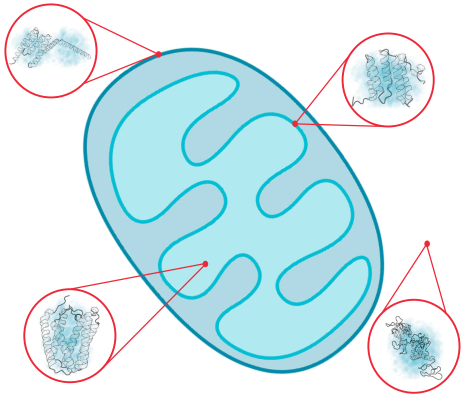

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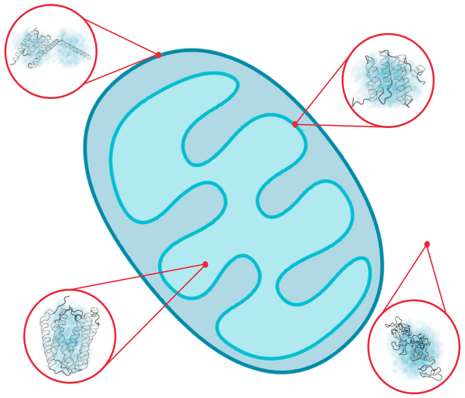

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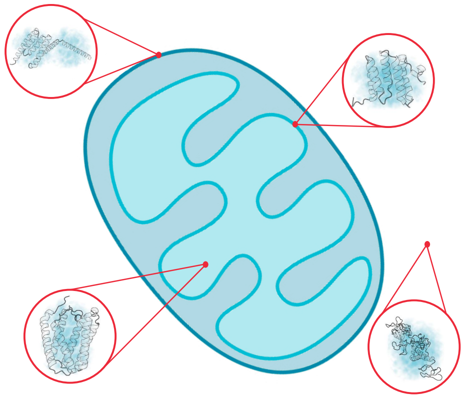

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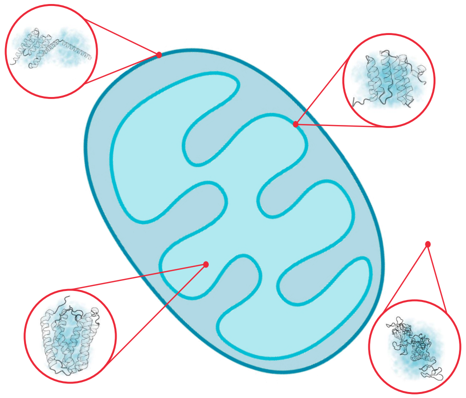

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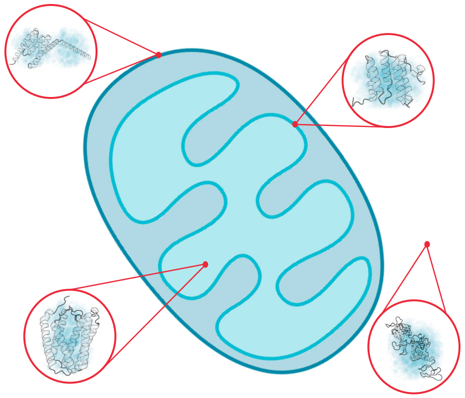

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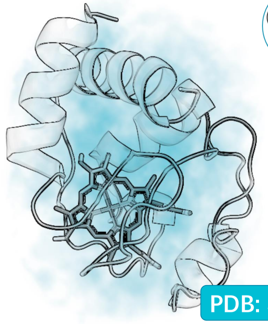
Cytochrome C

Inner Membrane



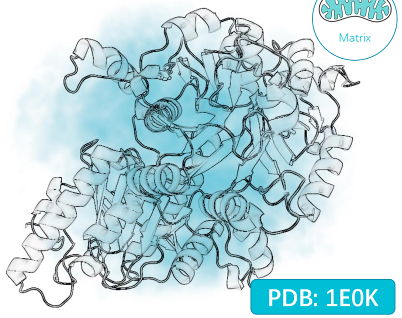
PDB: 3NWV

Each molecule of cytochrome C can carry one electron from complex III to complex IV in the electron transport chain.

Amino Acids:	325
Mutations:	29
Publications:	55,275
Chromosome:	7
MBU Rating:	48

TWINKLE

Matrix



PDB: 1E0K

Six of these subunits make up one TWINKLE helicase, which uses energy from ATP to unwind mtDNA ready for it to be copied.

Amino Acids:	684
Mutations:	329
Publications:	166
Chromosome:	10
MBU Rating:	72

PolG

DNA Polymerase
Subunit Gamma-1

Matrix

PDB: 4ZTU

Mitochondrial DNA polymerase is made up of three subunits, one of which is PolG, which does all the work to copy mtDNA!

Amino Acids:	1,239
Mutations:	658
Publications:	1,508
Chromosome:	15
MBU Rating:	59

TFAM

Transcription Factor
A

Matrix

PDB: 4NNU

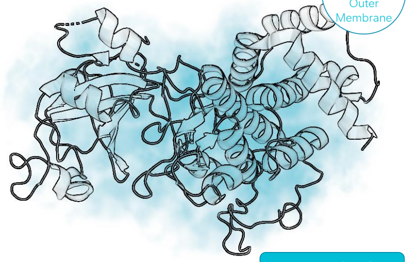
TFAM – mitochondrial transcription factor A – is needed in order to both read and replicate genes on mtDNA.

Amino Acids:	246
Mutations:	137
Publications:	1,752
Chromosome:	10
MBU Rating:	72

PINK1

PTEN-Induced Kinase
1

Outer Membrane



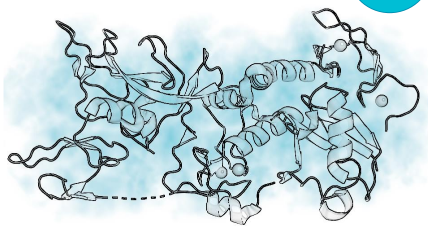
PDB: 6EQI

PINK1 tags mitochondria that are damaged and need to be removed by adding a phosphate group to the protein Parkin.

Amino Acids:	581
Mutations:	364
Publications:	2,763
Chromosome:	1
MBU Rating:	70

Parkin

Cytoplasm



PDB: 4K95

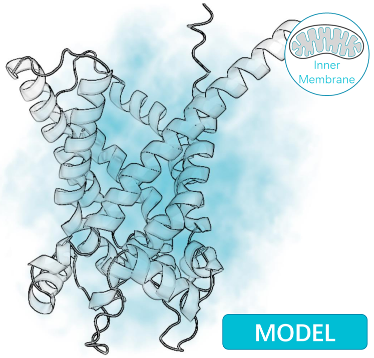
When phosphorylated by PINK1, Parkin tags damaged mitochondria. It is often mutated in Parkinson's disease.

Amino Acids:	465
Mutations:	350
Publications:	9,246
Chromosome:	6
MBU Rating:	65

UCP1

Uncoupling
Protein 1

Inner Membrane



MODEL

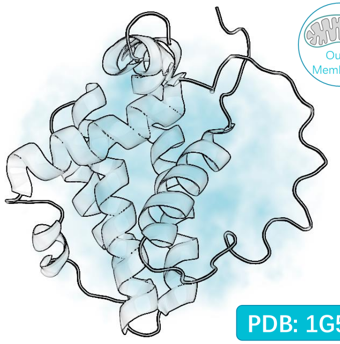
A member of the mitochondrial carrier family, UCP1 converts energy from food into heat to help keep us warm!

Amino Acids:	307
Mutations:	188
Publications:	5,121
Chromosome:	4
MBU Rating:	35

BCL2

B-Cell
Lymphoma 2

Outer Membrane



PDB: 1G5M

BCL2 prevents cells from entering apoptosis – cell self-destruction – unless they are extremely damaged.

Amino Acids:	239
Mutations:	72
Publications:	69,473
Chromosome:	18
MBU Rating:	23

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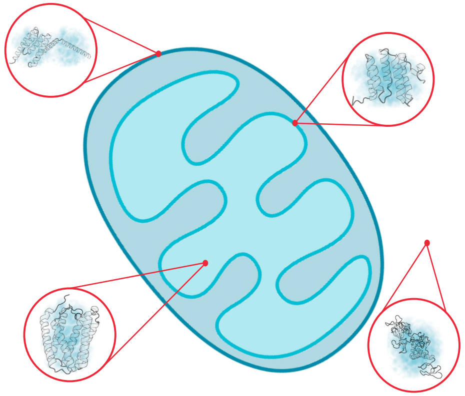

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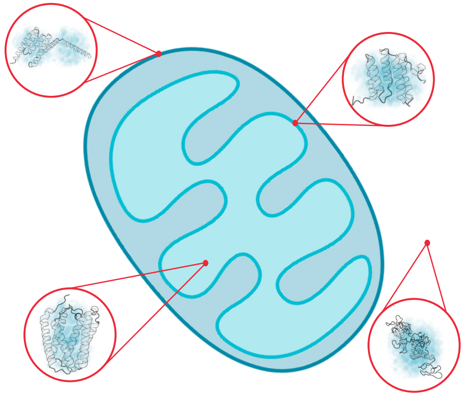

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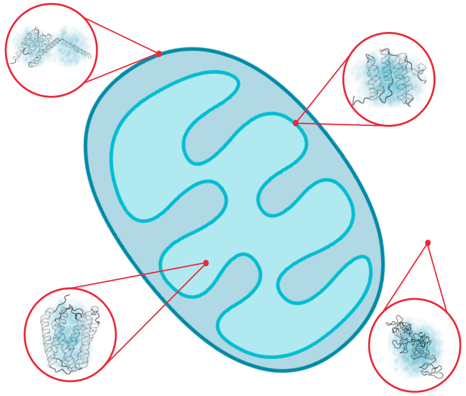

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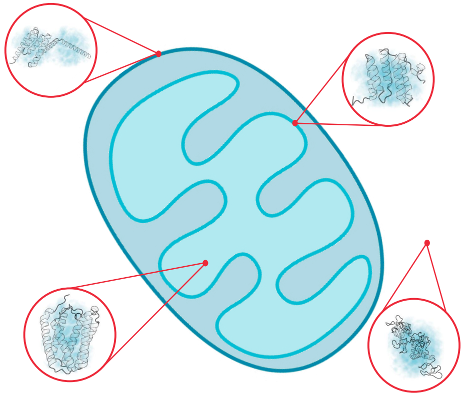

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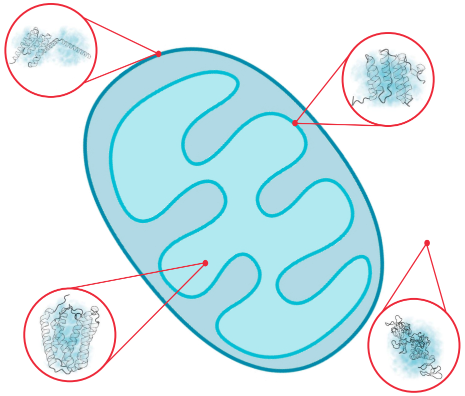

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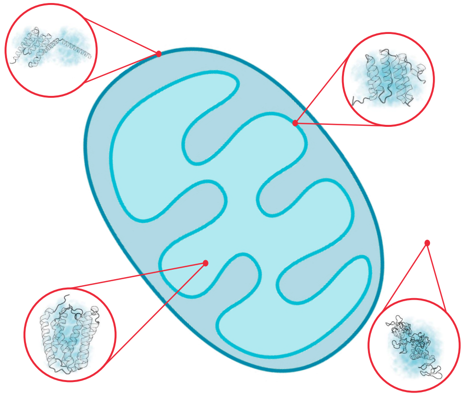

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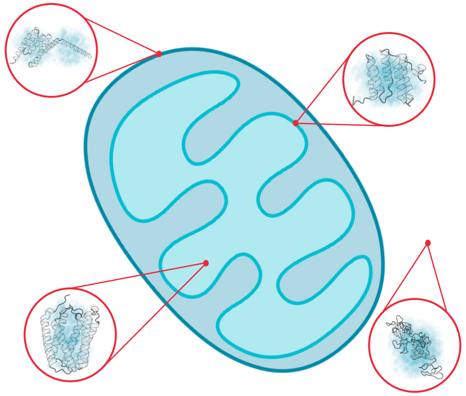

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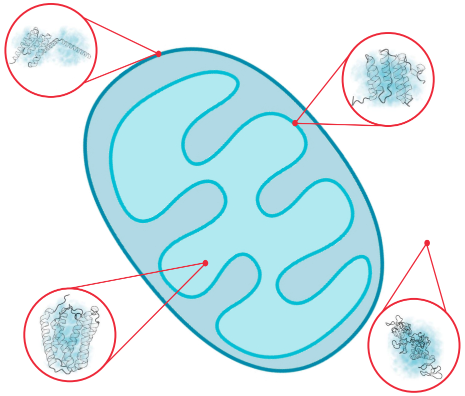

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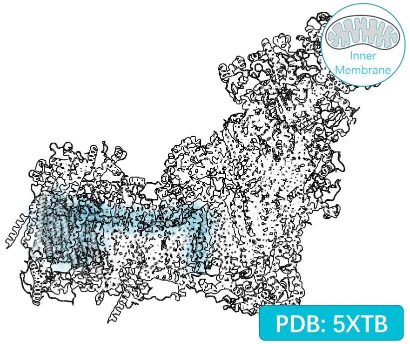
ND5
NADH-Ubiquinone Oxidoreductase, Complex I

Inner Membrane



PDB: 5XTB

ND5 is one of the 44 complex I subunits! It sits in the inner membrane.

Amino Acids:	603
Mutations:	421
Publications:	12,932
Chromosome:	mtDNA
MBU Rating:	30

NDUFV1
NADH-Ubiquinone Oxidoreductase, Complex I

Inner Membrane

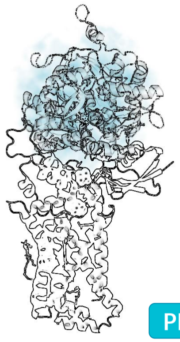
PDB: 5XTB

One of 44 complex I subunits! NDUFV1 has the flavin which accepts the electron from NADH.

Amino Acids:	434
Mutations:	315
Publications:	12,932
Chromosome:	11
MBU Rating:	19

SDHA
Succinate Dehydrogenase, Complex II

Inner Membrane



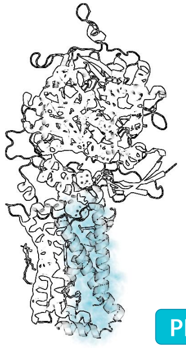
PDB: 1ZOY

SDHA is one of four complex II subunits. Complex II is the second part of the electron transport chain.

Amino Acids:	664
Mutations:	400
Publications:	20,646
Chromosome:	5
MBU Rating:	17

SDHC
Succinate Dehydrogenase, Complex II

Inner Membrane



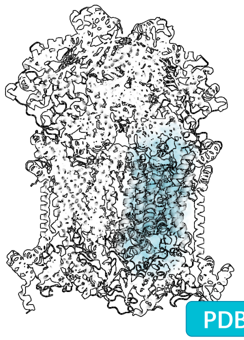
PDB: 1ZOY

SDHC is one of the four complex II subunits. Complex II's full name is succinate dehydrogenase!

Amino Acids:	169
Mutations:	118
Publications:	20,646
Chromosome:	1
MBU Rating:	13

Cyt B
Coenzyme Q-Cytochrome c Reductase, Complex III

Inner Membrane



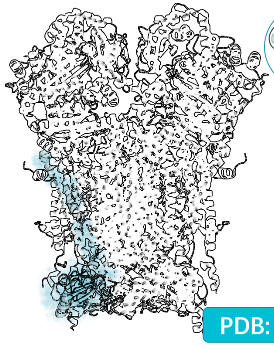
PDB: 5XTE

Cytochome B is part of mitochondrial complex III. It binds two heme groups!

Amino Acids:	380
Mutations:	410
Publications:	12,276
Chromosome:	mtDNA
MBU Rating:	30

Rieske
Coenzyme Q-Cytochrome c Reductase, Complex III

Inner Membrane



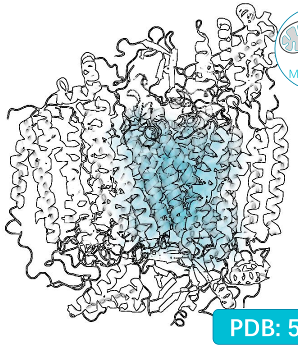
PDB: 5XTE

The Rieske protein is the part of complex III which transfers one electron to cytochrome c.

Amino Acids:	274
Mutations:	89
Publications:	44
Chromosome:	19
MBU Rating:	17

COX1
Cytochrome C Oxidase, Complex IV

Inner Membrane



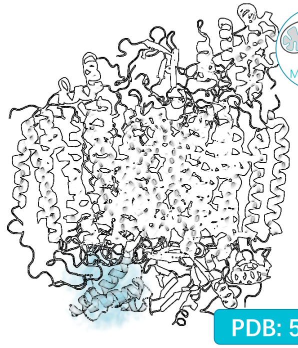
PDB: 5Z62

COX1 is a subunit of mitochondrial complex IV, the last complex in the electron transport chain.

Amino Acids:	513
Mutations:	294
Publications:	10,039
Chromosome:	mtDNA
MBU Rating:	21

COX6B1
Cytochrome C Oxidase, Complex IV

Inner Membrane



PDB: 5Z62

COX6B1 is a subunit of mitochondrial complex IV. The full name of complex IV is cytochrome C oxidase!

Amino Acids:	86
Mutations:	43
Publications:	26,663
Chromosome:	19
MBU Rating:	14

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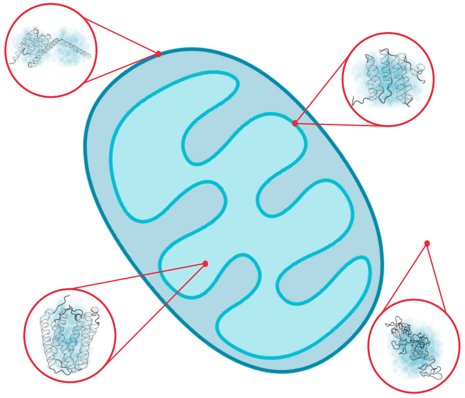

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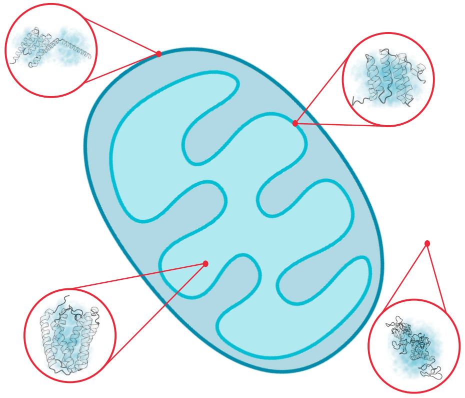

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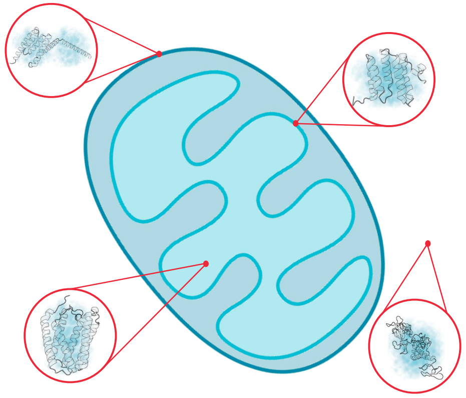

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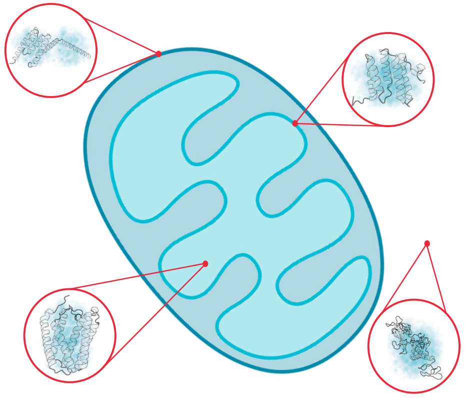

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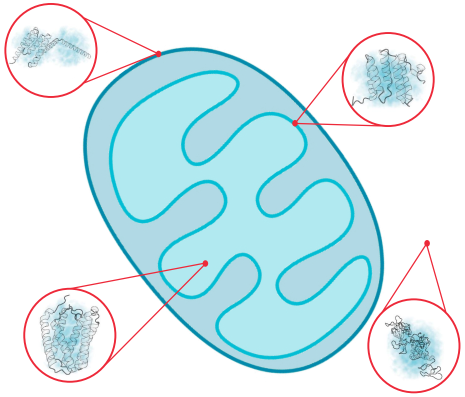

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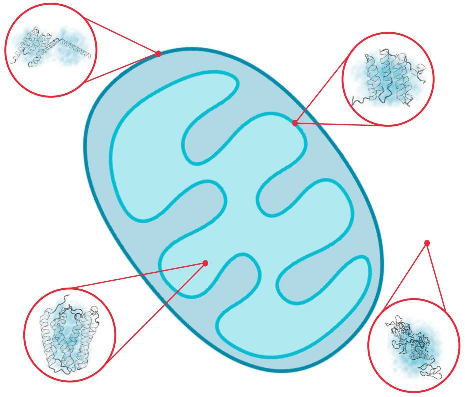

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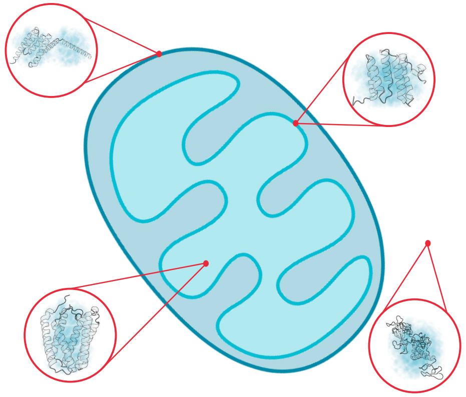

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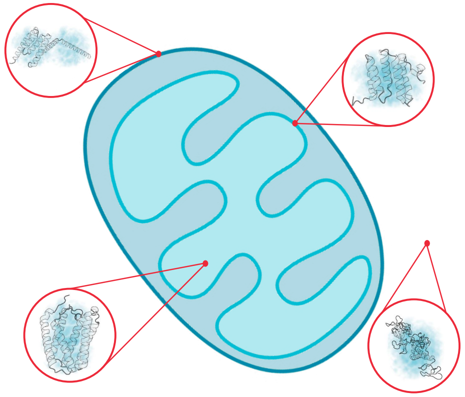

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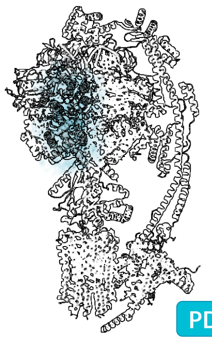
ATP Synthase α

Inner Membrane



PDB: 6ZQN

Each ATP synthase has three α subunits which bind ADP, the nucleotide needed to make ATP.

Amino Acids:	553
Mutations:	217
Publications:	6,968
Chromosome:	18
MBU Rating:	45

ATP Synthase C

Inner Membrane

PDB: 6ZQN

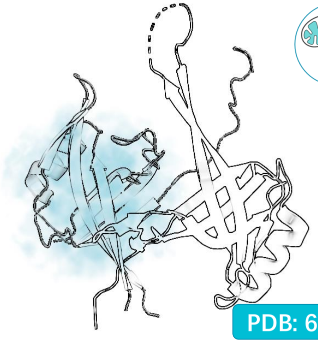
The c subunits of ATP synthase form the membrane half of the complex, known as Fo.

Amino Acids:	136
Mutations:	57
Publications:	6,968
Chromosome:	17
MBU Rating:	37

SSBP1

Single Stranded DNA
Binding Protein

Matrix



PDB: 6RUP

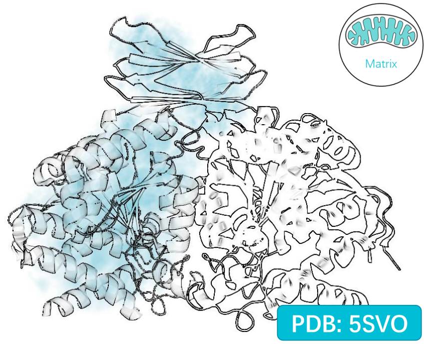
SSBP1 binds single stranded mtDNA to protect it during DNA replication.

Amino Acids:	148
Mutations:	66
Publications:	143
Chromosome:	7
MBU Rating:	25

IDH2

Isocitrate Dehydrogenase
2

Matrix



PDB: 5SVO

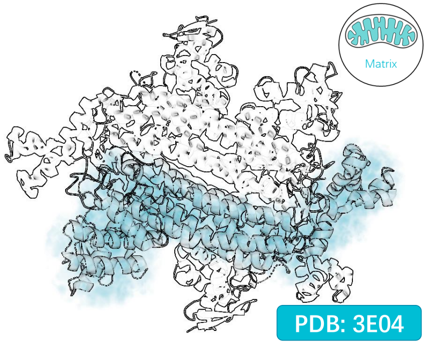
IDH2 is an enzyme that helps to break down glucose from food in order to power the electron transport chain.

Amino Acids:	452
Mutations:	209
Publications:	1,978
Chromosome:	15
MBU Rating:	19

Fumarase

Fumarate
Hydratase

Matrix



PDB: 3E04

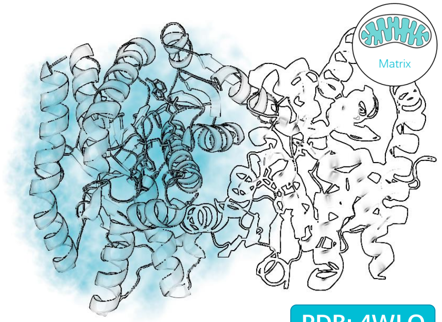
Fumarase is a key enzyme involved in the citric acid cycle, which occurs in the mitochondrial matrix.

Amino Acids:	510
Mutations:	256
Publications:	944
Chromosome:	1
MBU Rating:	34

MDH2

Malate
Dehydrogenase 2

Matrix



PDB: 4WLO

MDH2 is an enzyme in the citric acid cycle that converts malate into oxaloacetate.

Amino Acids:	338
Mutations:	221
Publications:	70
Chromosome:	7
MBU Rating:	12

MITOTrumps

Shuffle and deal the cards face down. Each player picks up their cards in a stack, so that they can see their **top card only**.

The player left of the dealer chooses and calls out their **best stat** (e.g. “Chromosome – 10”). The other players look to see if their cards beats this stat. If there is a tie between multiple people, the card with, the highest amino acid number wins.

The person with the **highest number** wins the top card from each player and adds them to the back of their stack. They then choose the category for the next card.

The **winner** is the person who **collects all the cards!**

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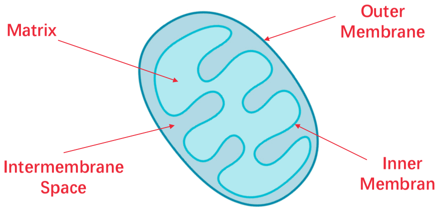
Proteins have many different structures and functions! The pictures on the cards are often the **real structures** for these proteins, and will have a ‘PDB’ number where they can be found! Some are **computer models** of that protein, and there will be a note saying it is a model.

Mitochondria have **proteins everywhere**, so if you look at the icon in the top right of your card you can see where inside a mitochondrion a protein can be found.



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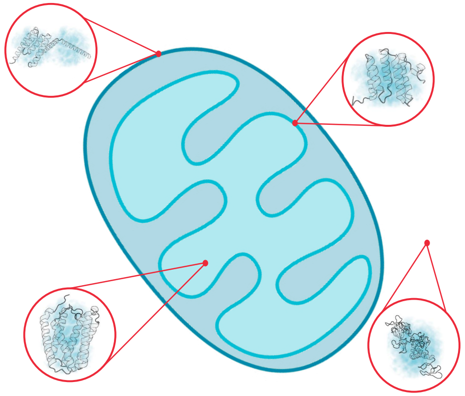

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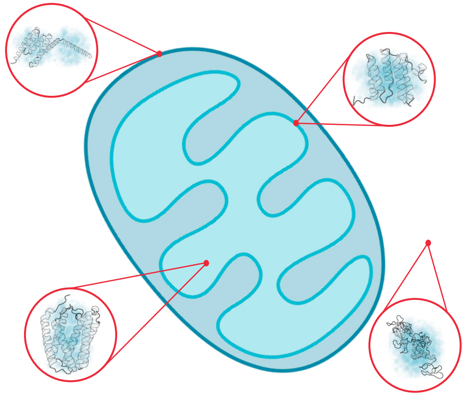

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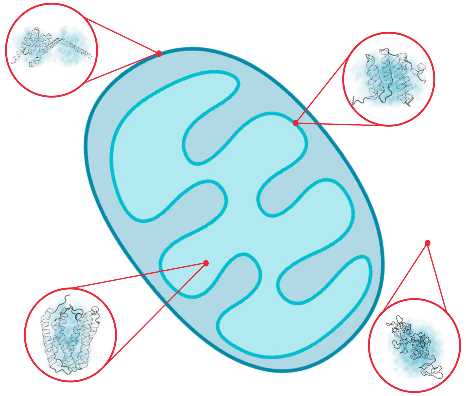

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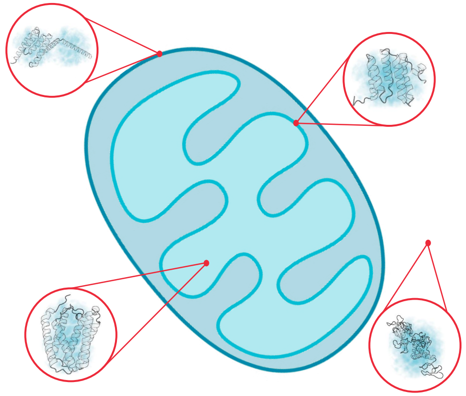

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The MRC Mitochondrial Biology Unit (MBU) is dedicated to the **study of mitochondria**, and the role they play in **disease**.

Due to the many essential functions of mitochondria, such as releasing energy from food, damaged mitochondria can cause disease. Many mitochondrial diseases are **life-long, difficult to diagnose**, and **incurable**. Mitochondria also play a role in diseases such as **cancer** and **dementia**. Through understanding how mitochondria work, we will be able to **better treat these diseases**.

*MitoTrumps was developed by **Alannah King.***
*Thank you to the **MBU Public Engagement Committee** and **the MBU** for their contributions, and to **Stewart Ashcroft-Quinn** and **Tyler Smyth** for the inspiration!*

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Amino Acid – The **building blocks** of proteins! This is the number of amino acids in the protein.

Mutations – This is the **number of different variants** of this protein across humans listed on gnomAD as of 10/01/21. Some of these cause diseases.

Publications – The **number of published scientific articles** on this protein, as of 10/01/22 on PubMed.

Chromosome – The **chromosome that the gene for this protein is on**. Some genes are on the mitochondrial DNA, mtDNA! For the ‘Chromosome’ stat, mtDNA proteins trump all others!

MBU Rating – We asked our scientists to **vote for their favourite proteins in this deck**! The MBU rating reflects the number of points a protein received.

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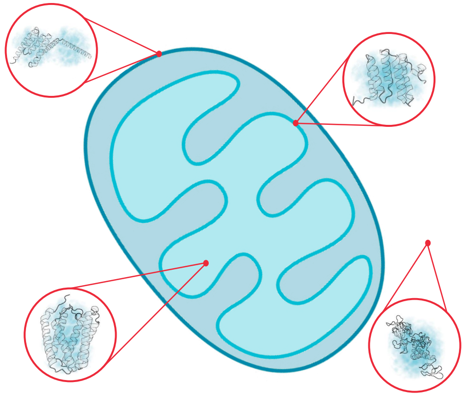

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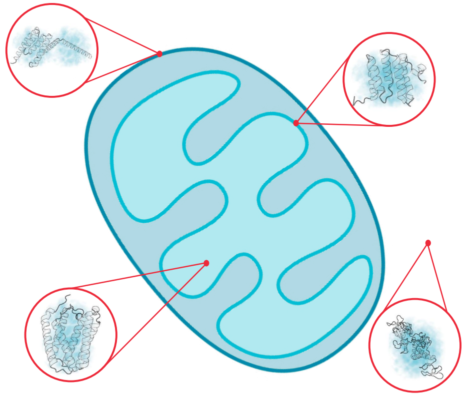

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