Mitochondria AREN'T ALWAYS MODEL EMPLOYEES

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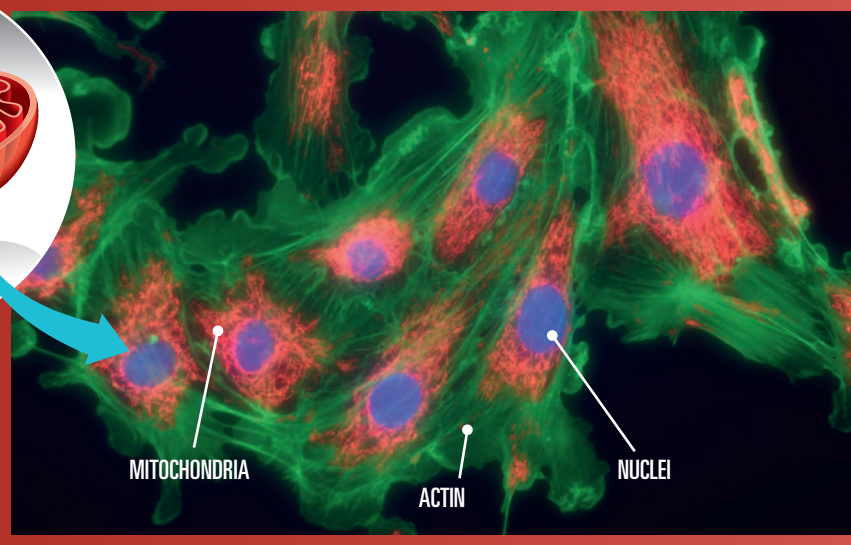
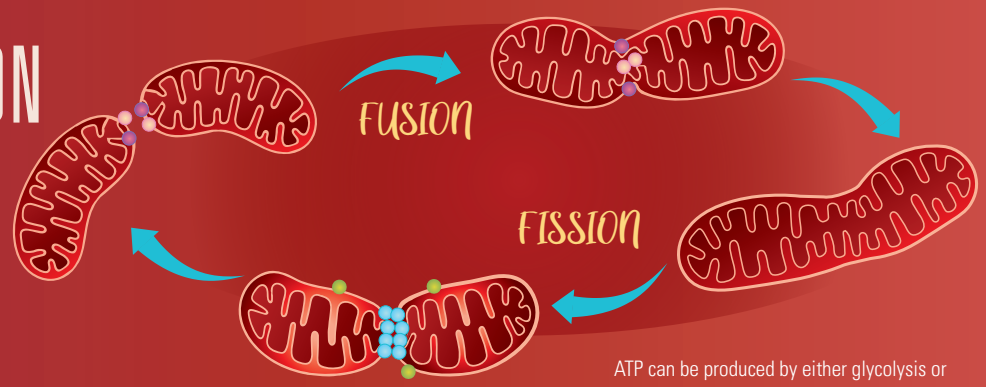
Mitochondria have an important job in the cell: produce most of the energy powering cellular function. When they get lazy and slack on the job, they can cause mitochondrial disorders such as Leigh syndrome, a severe, often lethal, neurological disorder (1). But an overzealous mitochondrion is just as problematic. When mitochondria work overtime, they tinker on the edge of "biological burnout." Overenergetic mitochondria can't handle stress, and their dysfunction contributes to autism, a spectrum of behavioral neurological disorders marked by challenges with social skills, speech, and nonverbal communication (2-3).

Mitochondria are often depicted as simplistic beans filled with folded membranes, but they are much more dynamic. They form large networks in a cell, giving them a long, filamentous appearance under a microscope.



FISSION & FUSION

Mitochondria undergo fission and fusion in cells to accommodate the energetic needs of the cell. The main energetic currency cells work in is ATP, which is produced via the electron transport chain.



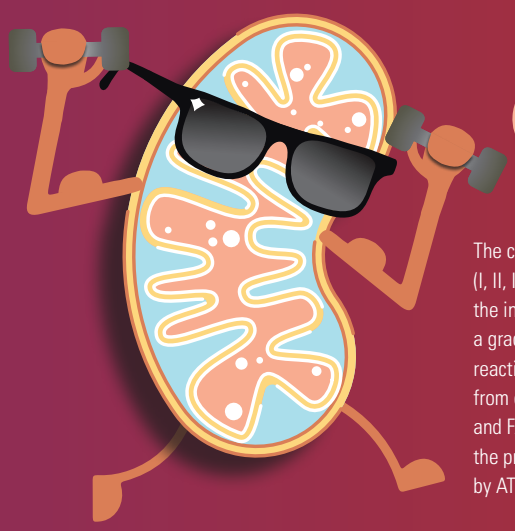
Key proteins mediate the fission and fusion of mitochondria. Mutations in some of these proteins cause conditions such as optic atrophy, the slow loss of vision due to optic nerve damage.

The connection between mitochondrial morphology and metabolic function holds potential as a type of diagnostic for diseases with known mitochondrial dysfunction such as autism spectrum disorder.

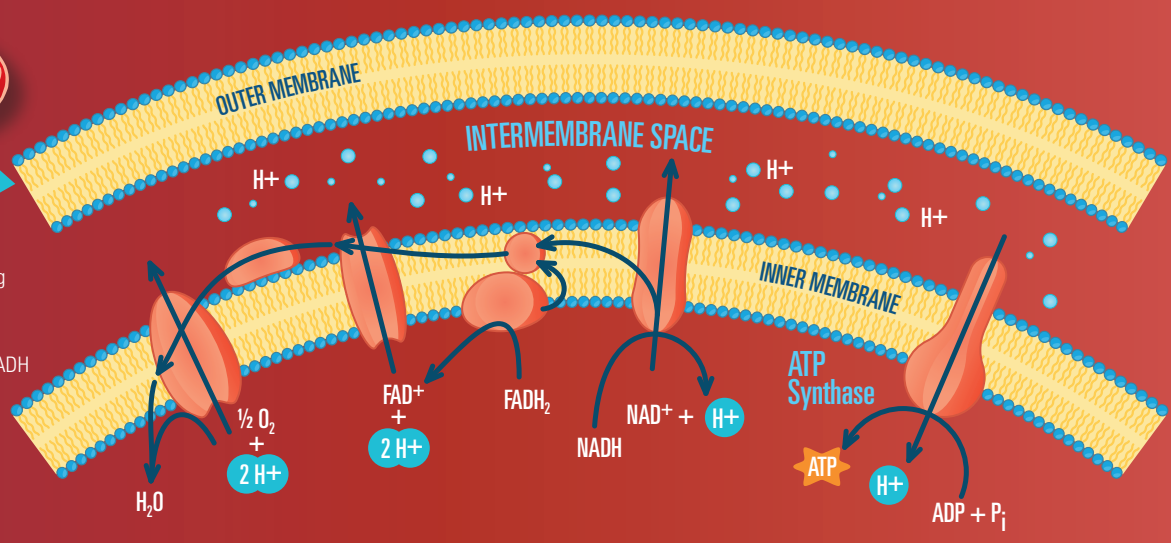
ATP can be produced by either glycolysis or mitochondrial respiration via the electron transport chain. When cells switch to glycolysis as their main energy producing source, mitochondria break apart, but when the cell relies on mitochondrial respiration for energy production, mitochondria fuse together and form large networks (4).

OXIDATIVE PHOSPHORYLATION

Mitochondria are composed of two membranes, an outer membrane and inner membrane. Key proteins found in the inner membrane make up the electron transport chain that moves electrons between forms of energy currency such as NADH, FADH, and ATP.



The cytochrome complex proteins (I, II, III, and IV) pump protons into the intermembrane space, creating a gradient that powers reduction reactions that remove electrons from energy currencies such as NADH and FADH, ultimately leading to the production of water and ATP by ATP synthase.



As the final step in the electron transport chain, ATP synthase phosphorylates ADP, producing the most valuable energetic currency in the cell: ATP. Rather than pump protons into the inner membrane space, the ATP synthase pumps protons into the mitochondrial matrix, powering the phosphorylation reaction.

Researchers recently reported that complexes I, III, and IV in the electron transport chain were dysfunctional in patients with autism spectrum disorder, although which complex and the level of dysfunction varied between individuals (5).

Altered metabolism in individuals with autism associated with changes in mitochondrial morphology. How fragmented or fused an individual's mitochondria were correlated with problems in the electron transport chain and symptom severity (5).

REFERENCES

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