Respiratory Chain

The respiratory chain, also called the electron transport chain (ETC), helps a cell create energy (in the form of ATP). Mitochondrial respiratory chain disorders (MRCDs) are a group of conditions that happen when one of the five major parts, or complexes, of the respiratory chain doesn't work as well as it should.

Each of the five complexes in the ETC is formed from a group, or complex, of proteins. When the body breaks down food, tiny bits of energy called electrons (which have a negative charge) are made. Molecules called NADH and FADH2 carry the electrons to the ETC. The ETC takes these electrons and uses them to make ATP, the type of energy that is used by cells.

Steps in the ETC

Step 1: Complex I (or NADH oxidoreductase), takes an electron (which has a negative charge) and moves it into the next step of the chain while also pumping a proton (which has a positive charge) into the space outside of the mitochondria.

Step 2: Complex II (or succinate oxidoreductase) takes an electron from FADH2.

Step 3: Complex III (or ubiquinol c oxidoreductase) receives the electrons and pumps more protons outside of the mitochondria.

Step 4: Complex IV (or cytochrome c oxidase) receives the electrons and uses them to form water by combining oxygen and the protons.

Step 5: The protons that have been pumped into the space out of the mitochondria move back into the mitochondria through complex V (or ATP synthase). As the protons move through complex V it spins, which helps change a molecule called ADP into ATP.

Step 6: The electrons that had moved through the ETC combine with oxygen and protons to make water.

Related Conditions

- Complex I Deficiency
- Complex II Deficiency
- Complex III Deficiency
- Complex IV Deficiency
- Complex V Deficiency
- Primary CoQ10 Deficiency

Frequency

Around 1 in 4300 people have an MRCD.

Clinical Trials

For specific details on other clinical trials, visit the MitoAction Clinical Trials page or www.clinicaltrials.gov.

Resources

Connecting with others who are impacted by a rare disease allows for important information to be shared about day-to day life, prevents isolation, and gives hope. Please contact MitoAction for peer support opportunities at 888-MITO-411 or email mito411@mitoaction.org. Other resources we recommend are:

- New Patient Kit for Mitochondrial Conditions
- Planning and Preparation
- Monthly Expert Series
- Energy in Action Podcast

MitoAction does not provide medical advice, diagnosis, treatment, or legal advice. It is essential that all those living with or caring for someone with a Mitochondrial or FAOD disease have an emergency protocol letter. These letters, which are written and signed by a doctor, share details about prescribed treatment during crises and in emergency room settings. Always check with your doctor if you or your child has concerns as everyone may present with symptoms differently. Before beginning any treatment or therapy, please consult with your physician.