

MITOCHONDRIAL DNA DEPLETION SYNDROME

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ENERGY PRODUCTION BY MITOCHONDRIA

- most cells (except red blood cells) contain mitochondria, which convert energy in food to ATP (adenosine triphosphate) that can be used by the cell for it to function
- the process that generates energy in the mitochondria is oxidative phosphorylation
- the oxidative part of oxidative phosphorylation involves oxygen consumption by a pathway called the respiratory chain, which generates energy by pumping protons
- the phosphorylation part of oxidative phosphorylation involves the formation of ATP using the protons pumped by the respiratory chain
- besides oxygen, the respiratory chain needs a source of electrons, primarily NADH

MITOCHONDRIAL DNA

- the proteins needed in the respiratory chain depend on mitochondrial DNA to provide the message for their synthesis
- the four components needed to make DNA are deoxythymidine triphosphate (dTTP), deoxyguanosine triphosphate (dGTP), deoxyadenosine triphosphate (dATP), and deoxycytosine triphosphate (dCTP)
- the enzyme thymidine kinase (TK) converts thymidine to thymidine monophosphate (TMP), which is then further processed to TTP
- the enzyme deoxyguanosine kinase converts deoxyguanosine to deoxyguanosine monophosphate (dGMP), which is then further processed to dGTP

