SUCLG1 gene

succinate-CoA ligase GDP/ADP-forming subunit alpha

Normal Function

The *SUCLG1* gene provides instructions for making one part (the alpha subunit) of an enzyme called succinyl-CoA ligase. The alpha subunit is used to make two slightly different versions of this enzyme. One version is most active in tissues such as the brain and muscle that require a large amount of energy, while the other version is most active in tissues that require less energy.

Both versions of succinyl-CoA ligase play an important role in mitochondria, which are the energy-producing centers inside the cell. These enzymes are involved in a series of chemical reactions known as the citric acid cycle (or Krebs cycle). These reactions allow cells to use oxygen and produce energy.

Mitochondria each contain a small amount of DNA, known as mitochondrial DNA or mtDNA. Studies suggest that succinyl-CoA ligase interacts with another enzyme, called nucleoside diphosphate kinase, to produce and maintain the molecules that make up mtDNA. Having an adequate amount of mtDNA is essential for normal energy production within cells.

Health Conditions Related to Genetic Changes

SUCLG1-related mitochondrial DNA depletion syndrome

Variants (also called mutations) in the *SUCLG1* gene cause *SUCLG1*-related mtDNA depletion syndrome, an inherited disorder that affects the early development of the brain.

Most *SUCLG1* gene variants change a single protein building block (amino acid) in the alpha subunit of succinyl-CoA ligase, which reduces but does not eliminate the activity of the enzyme. People with some enzyme activity typically have milder signs and symptoms than people with no enzyme activity.

A shortage (deficiency) of normal succinyl-CoA ligase leads to problems with the production and maintenance of mtDNA. A reduction in the amount of mtDNA (known as mtDNA depletion) impairs energy production in many of the body's cells and tissues and leads to the characteristic features of *SUCLG1*-related mtDNA depletion syndrome.

Leigh syndrome

MedlinePlus Genetics provides information about Leigh syndrome

Other Names for This Gene

- GALPHA
- succinate-CoA ligase, ADP-forming, alpha subunit
- succinate-CoA ligase, GDP-forming, alpha subunit
- succinyl-CoA synthetase, alpha subunit
- SUCLA1

Additional Information & Resources

Tests Listed in the Genetic Testing Registry

- Tests of SUCLG1 (https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=8802[geneid])

Scientific Articles on PubMed

- PubMed (https://pubmed.ncbi.nlm.nih.gov/?term=%28SUCLG1%5BALL%5D%29+OR+%28succinate-CoA+ligase%5BALL%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+3600+days%22%5Bdp%5D)

Catalog of Genes and Diseases from OMIM

- SUCCINATE-CoA LIGASE, GDP/ADP-FORMING, SUBUNIT ALPHA; SUCLG1 (https://omim.org/entry/611224)

Gene and Variant Databases

- NCBI Gene (https://www.ncbi.nlm.nih.gov/gene/8802)
- ClinVar (https://www.ncbi.nlm.nih.gov/clinvar?term=SUCLG1[gene])

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Genomic Location

The *SUCLG1* gene is found on chromosome 2 (https://medlineplus.gov/genetics/chromosome/2/).

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