

MLYCD gene

malonyl-CoA decarboxylase

Normal Function

The *MLYCD* gene provides instructions for making an enzyme called malonyl-CoA decarboxylase. This enzyme helps regulate the formation and breakdown of a group of fats called fatty acids. Many tissues, including heart (cardiac) muscle, use fatty acids as a major source of energy. The body also uses fatty acids to build cell membranes, produce hormones, and carry out many other important processes.

Malonyl-CoA decarboxylase is responsible for the chemical reaction that converts a molecule called malonyl-CoA to a molecule called acetyl-CoA. This reaction is an important step in the breakdown of fatty acids. Acetyl-CoA is then used to make new fatty acids and can also be used to produce energy.

Malonyl-CoA decarboxylase is most active in cardiac muscle and in muscles used for movement (skeletal muscles). It is also found in other organs and tissues, including the brain, small intestine, liver, kidney, and pancreas. This enzyme probably functions in several parts of the cell, including mitochondria, which are cells' energy-producing centers, and peroxisomes, which are small sacs that process fatty acids and other molecules. Malonyl-CoA decarboxylase also functions in the fluid that surrounds these cell structures (the cytoplasm).

Health Conditions Related to Genetic Changes

Malonyl-CoA decarboxylase deficiency

More than 20 mutations in the *MLYCD* gene have been identified in people with malonyl-CoA decarboxylase deficiency. Some of these mutations lead to the production of an abnormally short, nonfunctional version of malonyl-CoA decarboxylase or prevent the gene from producing any of this enzyme. Other mutations change the structure of the enzyme so it cannot be delivered to the parts of the cell where it is needed (such as mitochondria and peroxisomes).

A lack of malonyl-CoA decarboxylase disrupts the normal balance of fatty acid formation and breakdown in the body. As a result, fatty acids cannot be converted to energy, which leads to characteristic features of this disorder including low blood glucose (hypoglycemia) and a heart condition called cardiomyopathy. Byproducts of fatty acid

processing build up in tissues, which also contributes to the signs and symptoms of malonyl-CoA decarboxylase deficiency.

Other Names for This Gene

- DCMC_HUMAN
- hMCD
- malonyl coenzyme A decarboxylase
- MCD

Additional Information & Resources

Tests Listed in the Genetic Testing Registry

- Tests of MLYCD ([https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=23417\[geneid\]](https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=23417[geneid]))

Scientific Articles on PubMed

- PubMed (<https://pubmed.ncbi.nlm.nih.gov/?term=%28%28MLYCD%5BTIAB%5D%29+OR+%28malonyl-CoA+decarboxylase%5BTIAB%5D%29%29+OR+%28malonyl+coenzyme+A+decarboxylase%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+3600+days%22%5Bdp%5D>)

Catalog of Genes and Diseases from OMIM

- MALONYL-CoA DECARBOXYLASE; MLYCD (<https://omim.org/entry/606761>)

Gene and Variant Databases

- NCBI Gene (<https://www.ncbi.nlm.nih.gov/gene/23417>)
- ClinVar ([https://www.ncbi.nlm.nih.gov/clinvar?term=MLYCD\[gene\]](https://www.ncbi.nlm.nih.gov/clinvar?term=MLYCD[gene]))

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65)

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Genomic Location

The *MLYCD* gene is found on chromosome 16 (<https://medlineplus.gov/genetics/chromosome/16/>).

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