

LZTR1 gene

leucine zipper like post translational regulator 1

Normal Function

The *LZTR1* gene provides instructions for making a protein whose exact function is unknown. The LZTR1 protein is made in cells throughout the body. Within cells, it is found in the Golgi apparatus, which is a structure in which newly produced proteins are modified. Studies suggest that the LZTR1 protein may help stabilize this structure. Researchers suspect that this protein may also be associated with the CUL3 ubiquitin ligase complex, which is part of the cell machinery that breaks down (degrades) unneeded proteins.

Based on its role in several tumor types, the LZTR1 protein is thought to act as a tumor suppressor. Tumor suppressors are proteins that keep cells from growing and dividing too rapidly or in an uncontrolled way.

Health Conditions Related to Genetic Changes

Schwannomatosis

More than 50 different mutations in the *LZTR1* gene have been found in people with schwannomatosis, a disorder characterized by multiple noncancerous (benign) tumors called schwannomas that grow on nerves. This type of tumor arises from Schwann cells, which are specialized cells that normally form an insulating layer around the nerve.

LZTR1 gene mutations associated with schwannomatosis lead to production of an altered LZTR1 protein that is less able to control cell growth and division, which allows tumors to develop. It is unknown why these gene mutations are predominantly associated with schwannomas, instead of other types of tumor, in people with schwannomatosis.

It appears that mutations in the *LZTR1* gene alone are not enough to trigger the development of schwannomas. Additional genetic changes (somatic mutations) that are acquired during a person's lifetime and are present only in certain cells may also be required for schwannomas to form.

Some people who have a mutation in the *LZTR1* gene never develop tumors, which is a situation known as reduced penetrance.

Noonan syndrome

MedlinePlus Genetics provides information about Noonan syndrome

Cancers

Mutations in the *LZTR1* gene have also been found in a type of cancerous (malignant) brain tumor called glioblastoma. These mutations are classified as somatic and are present only in the cells that give rise to the tumor. Scientists believe that somatic changes in the *LZTR1* gene, or a loss of this gene, are among the factors that allow certain brain cells to grow and divide uncontrollably to form a tumor.

Other Names for This Gene

- BTBD29
- leucine-zipper-like transcriptional regulator 1
- LZTR-1
- NS10
- SWNTS2

Additional Information & Resources

Tests Listed in the Genetic Testing Registry

- Tests of LZTR1 ([https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=8216\[geneid\]](https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=8216[geneid]))

Scientific Articles on PubMed

- PubMed (<https://pubmed.ncbi.nlm.nih.gov/?term=%28%28LZTR1%5BTIAB%5D%29+OR+%28leucine+zipper+like+transcription+regulator+1%5BTIAB%5D%29%29+OR+%28LZTR-1%5BTIAB%5D%29+OR+%28SWNTS2%5BTIAB%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D%29>)

Catalog of Genes and Diseases from OMIM

- GLIOMA SUSCEPTIBILITY 1; GLM1 (<https://omim.org/entry/137800>)
- LEUCINE ZIPPER-LIKE TRANSCRIPTIONAL REGULATOR 1; LZTR1 (<https://omim.org/entry/600574>)

Gene and Variant Databases

- NCBI Gene (<https://www.ncbi.nlm.nih.gov/gene/8216>)
- ClinVar ([https://www.ncbi.nlm.nih.gov/clinvar?term=LZTR1\[gene\]](https://www.ncbi.nlm.nih.gov/clinvar?term=LZTR1[gene]))

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Genomic Location

The *LZTR1* gene is found on chromosome 22 (<https://medlineplus.gov/genetics/chromosome/22/>).

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