TECTA gene

Tectorin alpha

Normal Function

The TECTA gene provides instructions for making a protein called alpha-tectorin. This protein is found in the tectorial membrane, which is part of a snail-shaped structure called the cochlea in the inner ear. The cochlea converts sound waves into nerve impulses, which are then transmitted to the brain. This process is critical for normal hearing.

Alpha-tectorin is large protein with multiple regions (called domains) through which it interacts with other proteins. These interactions are critical for the normal formation of the tectorial membrane.

Health Conditions Related to Genetic Changes

Nonsyndromic hearing loss

Researchers have identified at least 40 TECTA gene mutations that can cause nonsyndromic hearing loss, which is loss of hearing that is not associated with other signs and symptoms. Mutations in this gene can cause two forms of nonsyndromic hearing loss: DFNA8/12 and DFNB21.

DFNA8/12 is inherited in an autosomal dominant pattern, which means one mutated copy of the TECTA gene in each cell is sufficient to cause the condition. This form of hearing loss can be present before a child learns to speak (prelingual) or begin after a child learns to speak (postlingual). In some cases the hearing loss is stable, while in others it becomes more severe over time.

The TECTA gene mutations that cause DFNA8/12 change single protein building blocks (amino acids) in alpha-tectorin. The characteristics of the hearing loss depend on the domain in which the mutation occurs. Mutations in one domain tend to affect the ability to hear mid-frequency sounds, while mutations in another generally affect the ability to hear high-frequency sounds. All of these mutations alter the structure of the tectorial membrane and disrupt the conversion of sound to nerve impulses. However, it is unclear why changes in different areas of the alpha-tectorin protein lead to different hearing loss characteristics.

DFNB21 is inherited in an autosomal recessive pattern, which means both copies of the TECTA gene are mutated in each cell. This form of hearing loss is usually severe to profound and is prelingual.

The TECTA gene mutations that cause DFNB21 mutations create a premature stop signal in the instructions for making the alpha-tectorin protein. These mutations lead
to the production of a nonfunctional version of alpha-tectorin or prevent cells from making any of this protein. A total loss of alpha-tectorin function alters the structure of the tectorial membrane in such a way that sound cannot be converted to nerve impulses.

**Chromosomal Location**

Cytogenetic Location: 11q23.3, which is the long (q) arm of chromosome 11 at position 23.3

Molecular Location: base pairs 121,101,243 to 121,191,490 on chromosome 11 (Homo sapiens Updated Annotation Release 109.20200522, GRCh38.p13) (NCBI)

Credit: Genome Decoration Page/NCBI

**Other Names for This Gene**

- DFNA8
- DFNA12
- DFNB21
- TECTA_HUMAN

**Additional Information & Resources**

**Educational Resources**

- Neuroscience (second edition, 2001): The Inner Ear
  https://www.ncbi.nlm.nih.gov/books/NBK10946/

**Clinical Information from GeneReviews**

- Hereditary Hearing Loss and Deafness Overview
  https://www.ncbi.nlm.nih.gov/books/NBK1434
Scientific Articles on PubMed

- PubMed
  https://www.ncbi.nlm.nih.gov/pubmed?term=%28%28TECTA%5BTIAB%5D%29+OR+%28tectorin+alpha%5BTIAB%5D%29+OR+%28DFNA12%5BTIAB%5D%29+OR+%28DFNA8%5BTIAB%5D%29+OR+%28DFNB21%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+1800+days%22%5Bdp%5D

Catalog of Genes and Diseases from OMIM

- TECTORIN, ALPHA
  http://omim.org/entry/602574

Research Resources

- Atlas of Genetics and Cytogenetics in Oncology and Haematology
  http://atlasgeneticsoncology.org/Genes/GC_TECTA.html

- ClinVar

- HGNC Gene Symbol Report

- Monarch Initiative
  https://monarchinitiative.org/gene/NCBIGene:7007

- NCBI Gene

- The Hereditary Hearing Loss Homepage
  https://hereditaryhearingloss.org/

- UniProt
  https://www.uniprot.org/uniprot/O75443

Sources for This Summary

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/10987647

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/21520338
  Free article on PubMed Central: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3326665/
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/17431902

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  http://omim.org/entry/602574

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/9590290

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