

TECTA Gene

Subjects: **Genetics & Heredity**

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TECTA: Tectorin alpha. The TECTA gene provides instructions for making a protein called alpha-tectorin.

genes

1. 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.

2. Health Conditions Related to Genetic Changes

2.1. 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.

3. Other Names for This Gene

- DFNA12
- DFNA8
- DFNB21
- *TECTA_HUMAN*

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