TECTA Gene

Subjects: Genetics & Heredity
Contributor: Rui Liu

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

Keywords: 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

References

1. Balciuniene J, Dahl N, Jalonen P, Verhoeven K, Van Camp G, Borg E, Pettersson U, Jazin EE. Alpha-tectorin involvement in hearing disabilities: one gene--twophenotypes. Hum Genet. 1999 Sep;105(3):211-6.
2. Hildebrand MS, Morín M, Meyer NC, Mayo F, Modamio-Hoybjor S, Mencía A,Olavarrieta L, Morales-Angulo C, Nishimura CJ, Workman H, DeLuca AP, del CastilloI, Taylor KR, Tompkins B, Goodman CW, Schrauwen I, Wesemael MV, Lachlan K,Shearer AE, Braun TA, Huygen PL, Kremer H, Van Camp G, Moreno F, Casavant TL,Smith RJ, Moreno-Pelayo MA. DFNA8/12 caused by TECTA mutations is the mostidentified subtype of nonsyndromic autosomal dominant hearing loss. Hum Mutat.2011 Jul;32(7):825-34. doi: 10.1002/humu.21512.
3. Meyer NC, Alasti F, Nishimura CJ, Imanirad P, Kahrizi K, Riazalhosseini Y,Malekpour M, Kochakian N, Jamali P, Van Camp G, Smith RJ, Najmabadi H.Identification of three novel TECTA mutations in Iranian families with autosomal recessive nonsyndromic hearing impairment at the DFNB21 locus. Am J Med Genet A. 2007 Jul 15;143A(14):1623-9.
4. Moreno-Pelayo MA, del Castillo I, Villamar M, Romero L, Hernández-Calvín FJ,Herraiz C, Barberá R, Navas C, Moreno F. A cysteine substitution in the zonapellucida domain of alpha-tectorin results in autosomal dominant, postlingual,progressive, mid frequency hearing loss in a Spanish family. J Med Genet. 2001May;38(5):E13.
5. Mustapha M, Weil D, Chardenoux S, Elias S, El-Zir E, Beckmann JS, Loiselet J, Petit C. An alpha-tectorin gene defect causes a newly identified autosomalrecessive form of sensorineural pre-lingual non-syndromic deafness, DFNB21. HumMol Genet. 1999 Mar;8(3):409-12.
6. Naz S, Alasti F, Mowjoodi A, Riazuddin S, Sanati MH, Friedman TB, Griffith AJ,Wilcox ER, Riazuddin S. Distinctive audiometric profile associated with DFNB21alleles of TECTA. J Med Genet. 2003 May;40(5):360-3.
7. Pfister M, Thiele H, Van Camp G, Fransen E, Apaydin F, Aydin O,Leistenschneider P, Devoto M, Zenner HP, Blin N, Nürnberg P, Ozkarakas H, KupkaS. A genotype-phenotype correlation with gender-effect for hearing impairmentcaused by TECTA mutations. Cell Physiol Biochem. 2004;14(4-6):369-76.
8. Verhoeven K, Van Laer L, Kirschhofer K, Legan PK, Hughes DC, Schatteman I,Verstreken M, Van Hauwe P, Coucke P, Chen A, Smith RJ, Somers T, Offeciers FE,Van de Heyning P, Richardson GP, Wachtler F, Kimberling WJ, Willems PJ, Govaerts PJ, Van Camp G. Mutations in the human alpha-tectorin gene cause autosomaldominant non-syndromic hearing impairment. Nat Genet. 1998 May;19(1):60-2.Erratum in: Nat Genet 1999 Apr;21(4):449.
