

Nonsyndromic Hearing Loss, *GJB6*-Related

What Your Results Mean

Test results indicate that you are a carrier of nonsyndromic hearing loss, *GJB6*-related. Carriers are not expected to show symptoms. You and your partner would both have to be carriers of nonsyndromic hearing loss, *GJB6*-related and/or nonsyndromic hearing loss, *GJB2*-related for there to be an increased chance to have a child with symptoms; this is known as autosomal recessive inheritance. Carrier testing of your partner or donor is recommended in addition to consultation with a genetic counselor for a more detailed risk assessment. Since this is an inherited gene change, this information may be helpful to share with family members as it may impact their family planning.



Recommended Next Steps

Carrier testing of your partner or donor is recommended in addition to consultation with a genetic counselor for a more detailed risk assessment. If both you and your partner are carriers for nonsyndromic hearing loss, *GJB2*- or *GJB3*-related, each of your children has a 1 in 4 (25%) chance to have the condition.

Nonsyndromic Hearing Loss, *GJB6*-Related Deficiency Explained

What is Nonsyndromic Hearing Loss, *GJB6*-Related?

Nonsyndromic hearing loss, *GJB6*-related is an inherited condition that affects a part of the inner ear called the cochlea, preventing auditory information from being transmitted to the brain. The condition causes mild-to-severe sensorineural hearing loss that is typically present from birth and not progressive. Other variants in the *GJB6* gene cause a skin disorder known as Clouston syndrome, which is characterized by fingernail abnormalities, hair loss, and thickened skin on the palms of the hands and soles of the feet.



Prognosis

Prognosis is generally favorable, as the condition does not affect life expectancy.

Treatment

Management may consist of hearing aids and enrollment in appropriate educational interventions. Cochlear implantation may be considered for individuals with profound deafness.



Resources

National Association of the Deaf

<https://www.nad.org/>

National Society of Genetic Counselors

<https://www.nsgc.org/>