

# 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/>