Nonsyndromic Hearing Loss, *FOXI1*-Related

NxGen MDx

What Your Results Mean

Test results indicate that you are a carrier of nonsyndromic hearing loss, *FOXI1*-related. Carriers are not expected to show symptoms. You and your partner would both have to be carriers of nonsyndromic hearing loss, *FOXI1*-related and/or nonsyndromic hearing loss, *FOXI1*-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- *FOXI1* related or Pendred syndrome, each of your children would have a 25%, or 1 in 4, chance to develop symptoms.

Nonsyndromic Hearing Loss, *FOXI1*-Related Deficiency Explained

What is Nonsyndromic Hearing Loss, *FOXI1*-Related?

Nonsyndromic hearing loss, *FOXI1*-related is a rare form of inherited hearing loss. The hearing loss often develops shortly after birth and is often severe to profound, although mild-to-moderate progressive hearing impairment also occurs. The condition causes differences in the way the inner ear forms, including enlarged vestibular aqueduct, which can lead to problems with balance and coordination. Some people with nonsyndromic hearing loss, *FOXI1*-related may also have a kidney condition known as renal tubular acidosis. People with this condition have kidneys that do not remove enough acidic compounds from the body. As a result, they can have nausea, vomiting, dehydration, and problems with feeding and gaining weight. Short stature, kidney cysts and stones, weak bones, and muscle paralysis can also be seen. Renal tubular acidosis with deafness usually presents in infancy with failure to thrive. Hearing loss usually begins between childhood and young adulthood, and the hearing loss gradually gets worse over time.

Prognosis

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

Treatment

There is no cure for nonsyndromic hearing loss, *FOXI1*-related. Treatment options may include hearing aids, cochlear implants, speech therapy, and educational programs for the hearing impaired.

Resources

National Association of the Deaf
https://www.nad.org/

National Society of Genetic Counselors
https://www.nsgc.org/

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Revised: 4/2022