BlueCross BlueShield Nebraska

GENETIC TESTING: HEARING LOSS (REQUIRES PREAUTHORIZATION)

V.35

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DESCRIPTION

Hereditary hearing loss can be classified as syndromic or nonsyndromic. Syndromic hearing loss refers to hearing loss associated with other medical or physical findings, including visible abnormalities of the external ear. Because syndromic hearing loss occurs as part of a syndrome of multiple clinical manifestations, it is often recognized more readily as hereditary. Nonsyndromic hearing loss is defined as hearing loss not associated with other physical signs or symptoms. Nonsyndromic hearing loss accounts for 70% to 80% of genetically determined deafness, and it is more difficult to determine whether the etiology is hereditary or acquired.

This policy primarily focuses on the use of genetic testing to identify a cause of suspected hereditary hearing loss. The diagnosis of syndromic hearing loss can be made on the basis of associated clinical findings. However, at the time of hearing loss presentation, associated clinical findings may not be apparent; furthermore, variants in certain genetic loci may cause both syndromic and nonsyndromic hearing loss. Given this overlap, the policy focuses on genetic testing for hereditary hearing loss more generally.

If there is not a high suspicion for a specific hearing loss etiology, ideally the evaluation should occur in a stepwise fashion. About 50% of individuals with autosomal recessive hereditary hearing loss have pathogenic variants in the *GJB2* gene, in the other 50% of patients with apparent autosomal recessive hereditary hearing loss; numerous other genes are implicated. There is no single identifiable gene responsible for most cases of autosomal dominant hereditary hearing loss.

hearing loss with a multigene panel would be efficient. An alternative strategy for suspected autosomal recessive or autosomal dominant hearing loss would be to obtain a multigene panel that includes GJB2 and GJB6 as a first step.

Given the extreme heterogeneity in genetic causes of hearing loss, these 2 strategies may be considered reasonably equivalent.

Dates

Original Effective
07-21-2016
Last Review
08-07-2024
Next Review
08-11-2025

RELATED POLICIES

This policy document provides coverage criteria for genetic testing for hereditary hearing loss. Please refer to:

Genetic Testing: Prenatal and Preconception Carrier Screening for coverage criteria related to carrier screening for hereditary hearing loss.

Genetic Testing: Multisystem Inherited Disorders, Intellectual Disability, and Developmental Delay for coverage criteria related to genetic disorders that affect multiple organ systems.

Genetic Testing: General Approach to Genetic and Molecular Testing for coverage criteria related to genetic testing for hearing loss that is not specifically discussed in this or another non-general policy.

REFERENCE TABLE

The tests, associated laboratories, CPT codes, and ICD codes contained within this document serve only as examples to help users navigate claims and corresponding coverage criteria; as such, they are not comprehensive and are not a guarantee of coverage or non-coverage. Please see the Concert Platform for a comprehensive list of registered tests.

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GJB2 and *GJB6* Sequencing and/or Deletion/ Duplication Analysis or Multigene Panel Analysis	*GJB2* Gene Sequencing (GeneDx)	81252	H90-H90.8, H90.A-90.A3, H91.3-H91.93	1, 2
	GJB2 Deletion/Duplication Analysis (GeneDx)	81479		
	GJB6 Common Variant Analysis	81254		
	GJB6 Sequencing Analysis	81479		
	Hearing Loss Panel (GeneDx)	81430, 81431		
	Comprehensive Hearing Loss NGS Panel (Sequencing & Deletion/Duplication) (Fulgent Genetics)			
	Hereditary Hearing Loss and Deafness Panel (PreventionGenetics, part of Exact Sciences)			

POLICY

HEREDITARY HEARING LOSS

GJB2 and *GJB6* Sequencing and/or Deletion/Duplication Analysis or Multigene Panel Analysis

I. *GJB2* sequencing and/or deletion/duplication (81252, 81479) and/or *GJB6* sequencing and/or deletion/duplication analysis (81479) or multigene panel analysis (81430, 81431, 81254) to establish a diagnosis of hereditary hearing loss is considered **medically necessary** when:
   A. The member has hearing loss, **AND**
   B. There is no known acquired cause of the hearing loss (i.e., TORCH infections [*Toxoplasma gondii*, other agents, rubella, cytomegalovirus, and herpes simplex virus], bacterial infection, age-related or noise-related hearing loss).

II. *GJB2* sequencing and/or deletion/duplication (81252, 81479) and/or *GJB6* sequencing and/or deletion/duplication analysis (81479) or multigene panel analysis (81430, 81431, 81254) to

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BACKGROUND

GJB2 and _GJB6_ Sequencing and/or Deletion/Duplication Analysis or Multigene Panel Analysis

American College of Medical Genetics and Genomics (ACMG)

The American College of Medical Genetics and Genomics (ACMG) published guidelines in 2022 to guide genetics evaluation for individuals with hearing loss: “For individuals lacking physical findings suggestive of a known syndrome, a tiered diagnostic approach should be implemented. Unless clinical and/or family history suggests a specific genetic etiology, comprehensive HL gene panel testing should be initiated.” (p. 9)

The guidelines also state the following: “Although nonsyndromic HL [hearing loss] demonstrates high genetic heterogeneity, the DFNB1 locus, which includes the *GJB2* gene encoding the gap junction protein connexin 26 and the *GJB6* gene encoding the gap junction protein connexin 30, accounts for an estimated 50% of all autosomal recessive nonsyndromic HL and 15% to 40% of all deaf individuals in a variety of populations.” (p. 3)

GeneReviews: Hereditary Hearing Loss and Deafness Overview

GeneReviews is an expert-authored review of current literature on a genetic disease, and goes through a rigorous editing and peer review process before being published online.

In the section that discusses possible differential diagnosis for hereditary hearing loss and deafness, it is stated that, In developed countries approximately 65% of prelingual hearing loss is due to genetic causes. The remainder of cases are due to environmental (acquired) causes, which should be differentiated from genetic causes to inform the evaluation and required ancillary testing. Acquired hearing loss in children commonly results from prenatal infections from ‘TORCH’ organisms…or postnatal infections (such as CMV). Acquired hearing loss in adults is most often attributed to environmental factors.

GeneReviews also states that molecular genetic testing includes the use of multigene hearing loss panels and/or genomic testing. Single-gene testing (sequence analysis of a given gene, followed by gene-targeted deletion/duplication analysis) is rarely useful and typically NOT recommended.

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Procedure

Please type a procedure code

Enter at least the first 3 characters of the code

Diagnosis

Please type a diagnosis code

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CODES

\+ **CPT4**

REFERENCES

2020

Shearer AE, Hildebrand MS, Smith RJH. Hereditary Hearing Loss and Deafness Overview. 1999 Feb 14 [Updated 2017 Jul 27]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2020. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1434/

2022

Li MM, Tayoun AA, DiStefano M, et al. Clinical evaluation and etiologic diagnosis of hearing loss: A clinical practice resource of the American College of Medical Genetics and Genomics (ACMG). *Genet Med*. 2022;24(7):1392-1406

REVISIONS

01-01-2024
Minor updates to Background information.

09-28-2023
Changed "may be considered medically necessary" to "may be considered scientifically validated"

Added: "investigational when the above criteria is not met and for all other indications"

06-05-2023
Updated reference and formatting

12-06-2022
new references and formatting for 01/01/2023

06-14-2022
Added criteria for known familial variant analysis effective 07/01/2022

01-04-2022
Review of policy with no changes.

04-20-2021
Updated policy - no code changes

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