Revision log
Coding Implications

# CONCERT GENETIC TESTING: OTOLARYNGOLOGY

## OVERVIEW

**See Important Reminder at the end of this policy for important regulatory and legal information.**

This policy addresses the use of tests to identify a cause of hereditary hearing loss, or for other tests related to ear, nose, and throat disorders. Pre-test and post-test genetic counseling that facilitates informed decision-making, addresses the possibility of secondary or incidental findings, and a plan for returning results before testing occurs is strongly advised.

For additional information see the Rationale section.

The tests, CPT codes, and ICD codes referenced in this policy are not comprehensive, and their inclusion does not represent a guarantee of coverage or non-coverage. Please see the Concert Platform for additional registered tests.

## POLICY REFERENCE TABLE

**Coding Implications**

This clinical policy references Current Procedural Terminology (CPT®). CPT is a registered trademark of the American Medical Association. All CPT codes and descriptions are copyrighted 2024, American Medical Association. All rights reserved. CPT codes and CPT descriptions are from the current manuals and those included herein are not intended to be all-inclusive and are included for informational purposes only. Codes referenced in this clinical policy are for

informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

The tests, CPT codes, and ICD codes referenced in this policy are not comprehensive, and their inclusion does not represent a guarantee of coverage or non-coverage. Please see the Concert Platform for additional registered tests.

| CRITERIA SECTIONS | EXAMPLE TESTS (LABS) | COMMON BILLING CODES | REF |
|---|---|---|---|
| **Hereditary Hearing Loss** | | | |
| *GJB2* and *GJB6* Sequencing and/or Deletion/Duplication Analysis or Multigene Panel Analysis | *GJB2* Deletion/Duplication Analysis (GeneDx) | 81252, 81479, 81254, 81479, 81430, 81431, H90-H90.8, H90.A-90.A3, H91.3-H91.93 | 1, 2 |
| | *GJB6* Sequencing Analysis (Billing lab my vary) | | |
| | Hearing Loss Panel (GeneDx) | | |
| | Comprehensive Hearing Loss NGS Panel (Sequencing & Deletion/Duplication) (Fulgent Genetics) | | |
| | Hereditary Hearing Loss and Deafness Panel (PreventionGenetics, part of Exact Sciences) | | |
| | DFNB1 Autosomal Recessive Hearing Loss (*GJB2* sequencing and common | | |

| | | | |
|---|---|---|---|
| | *GJB6* deletions) (Billing lab may vary) | | |

# RELATED POLICIES

This policy document provides criteria for hereditary hearing loss, or for other tests related to ear, nose, and throat disorders. Please refer to:

- ***Reproductive Testing: Carrier Screening*** for criteria related to parental carrier screening for genetic disorders before or during pregnancy.
- ***Specialty Testing: Multisystem Genetic Conditions*** for criteria related to diagnostic tests for genetic disorders that affect multiple organ systems (e.g. whole exome and genome sequencing, chromosomal microarray, and multigene panels for broad phenotypes).
- ***General Approach to Laboratory Testing*** for criteria related to hearing loss, including known familial variant testing, that is not specifically discussed in this or another non-general policy.

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# CRITERIA

It is the policy of health plans affiliated with Centene Corporation® that the specific genetic testing noted below is **medically necessary** when meeting the related criteria:

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

I. *GJB2* sequencing and/or deletion/duplication and/or *GJB6* sequencing and/or deletion/duplication analysis or multigene panel analysis to establish a diagnosis of hereditary hearing loss is considered **medically necessary** when:

   A. The member/enrollee 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. Current evidence does not support *GJB2* sequencing and/or deletion/duplication and/or *GJB6* sequencing and/or deletion/duplication analysis or multigene panel analysis to establish a diagnosis of hereditary hearing loss for all other indications.

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## HEREDITARY HEARING LOSS

# RATIONALE

## *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 approximately 65% of prelingual hearing loss is due to genetic causes in developed countries. 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 recommends the use of multigene hearing loss panels and/or genomic testing.

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| Reviews, Revisions, and Approvals | Revision Date | Approval Date |
|---|---|---|
| Policy developed. | 03/23 | 03/23 |
| Semi-annual review. Updated title to reflect V1.2024 version. Overview, coding, reference-table, background and references updated. Throughout policy: replaced "coverage criteria" with "criteria. For Policy Reference Table: changed "Sequencing Analysis" to "Gene Sequencing"; added "GeneDx" to "GJB2 Deletion/Duplication Analysis; added "part of Exact Sciences" to "Hearing Loss and Deafness…". For Other Related Policies: added "and Molecular" to "Genetic Testing: General Approach…". For Hereditary Hearing Loss I.A. added "Targeted variant analysis for a known…". For Background and Rationale: replaced "inheritance patterns" to "genetic testing". For GJB2 and GJB6 Sequencing and/or Deletion Duplication Analysis or Multigene Panel Analysis: replaced "congenital" with "prelingual"; replaced "and the remainder" with "with 70% of prelingual…"; removed "Acquired cases"; added "which"; added statement "GeneReviews also states…". | 10/23 | 10/23 |
| Semi-annual review. Updated title to reflect V2.2024 version. In Known Familial Variant Analysis for Hereditary Hearing Loss criteria, moved criteria to policy "Genetic Testing: General Approach to Genetic and Molecular Testing" to consolidate criteria for known familial variant tests. Minor rewording for clarity throughout. Coding, reference-table, background and references updated. | 04/24 | 04/24 |
| Semi-annual review. Updated title to reflect V1.2025 version. GJB2 and GJB6 Sequencing and/or Deletion/Duplication Analysis or Multigene Panel Analysis: Streamlined portions of Background and Rationale section for brevity. | 11/24 | |
| Annual review. Policy title changed to Concert Genetic Testing: Otolaryngology from Concert Genetic Testing: Hearing Loss. Minor rewording without clinical significance. Changed "investigational" policy statement to note that "current | 11/25 | 12/25 |

| Reviews, Revisions, and Approvals | Revision Date | Approval Date |
|---|---|---|
| evidence does not support…" Coding table, rationale, background, and references updated. | | |

# REFERENCES

1. 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.
2. Shearer AE, Hildebrand MS, Smith RJH. Genetic Hearing Loss Overview. 1999 Feb 14 [Updated 2023 September 28]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2024. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1434/

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**Important Reminder**

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. "Health Plan" means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan's affiliates, as applicable.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions, and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment, or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care and are solely responsible for the medical advice and treatment of member/enrollees. This clinical policy is not intended to recommend treatment for member/enrollees. Member/enrollees should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom the Health Plan has no control or right of control. Providers are not agents or employees of the Health Plan.

 Providers, member/enrollees, and their representatives are bound to the terms and conditions expressed herein through the terms of their contracts. Where no such contract exists, providers, member/enrollees and their representatives agree to be bound by such terms and conditions by providing services to member/enrollees and/or submitting claims for payment for such services.

**Note: For Medicaid member/enrollees**, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

**Note: For Medicare member/enrollees,** to ensure consistency with the Medicare National Coverage Determinations (NCD) and Local Coverage Determinations (LCD), all applicable NCDs and LCDs and Medicare Coverage Articles should be reviewed prior to applying the criteria set forth in this clinical policy. Refer to the CMS website at http://www.cms.gov for additional information.