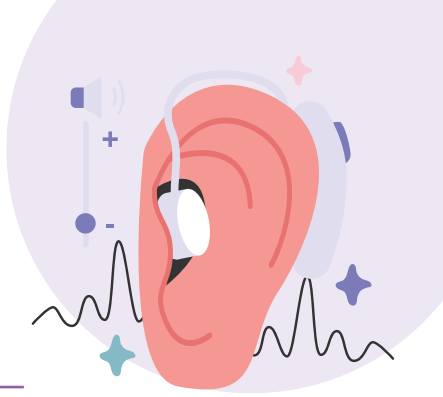
PRIME PATH LAB

HEREDITARY HEARING LOSS GENOMIC PANEL



When the Audiogram Tells You “How Much,” the Gene Tells You “Why.”

About half of childhood hearing loss is genetic, yet the audiogram the test physicians reach for first measures severity, never cause. This panel reads the molecular basis of sensorineural hearing loss across the common nonsyndromic genes and the syndromic forms that hide systemic disease behind the ear. The result tells you whether the loss will progress, how a cochlear implant is likely to perform, and which organ to watch next.

Did you know?

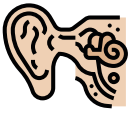
In 2026 the FDA approved the first gene therapy for genetic hearing loss a one-time cochlear infusion for OTOF (otoferlin)-related deafness. In the pivotal trial, about 80% of treated children gained hearing and many reached normal levels. A confirmed OTOF result is no longer just a diagnosis; it can be the doorway to treatment and the window is in early childhood.

Which genes carry the most weight on this panel?

A few account for a large share of real-world diagnoses: The full panel spans more than 120 genes; these five anchor the most actionable findings.

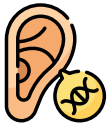
GJB2
the single most common cause of inherited hearing loss

OTOF
now a gene-therapy target



SLC26A4
and enlarged vestibular aqueduct

KCNQ1/KCNE1
Jervell and Lange-Nielsen syndrome



USH2A
the leading cause of Usher syndrome

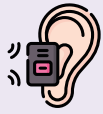
My patient does fine with a hearing aid — why sequence?

Because the gene reveals what an audiogram cannot. KCNQ1/KCNE1 pairs congenital deafness with long QT and a genuine risk of sudden cardiac death — a finding that turns an audiology visit into a life-saving cardiology referral. SLC26A4 flags thyroid and inner-ear disease; USH2A predicts future vision loss; and a positive aminoglycoside-ototoxicity result identifies a maternally inherited, entirely preventable cause where a single dose of an aminoglycoside can cause permanent deafness across an entire maternal line.

Which of my patients qualify?

- Congenital, prelingual, or early-childhood bilateral sensorineural hearing loss, or a failed newborn hearing screen.

- Progressive or adult-onset sensorineural hearing loss with an autosomal-dominant family history.



- Auditory neuropathy spectrum disorder (present OAE / cochlear microphonic with absent or abnormal ABR).

- Aminoglycoside exposure, or a maternal/family history of aminoglycoside-induced deafness.

What changes when the result comes back?

Management moves. An OTOF result flags gene-therapy candidacy and sharpens implant planning; a syndromic result starts the right surveillance — cardiac, thyroid, renal, or ophthalmic; an aminoglycoside-risk result rewrites the antibiotic plan for the patient and the maternal line. Every positive result enables cascade testing of relatives and informed reproductive counseling.

Why Prime Path Lab?

Hearing loss is the rare genetic diagnosis with a closing window: gene therapy, implant timing, and language development all reward early action. Identifying the cause before speech and schooling are affected and before a cardiac or renal complication declares itself — converts a single test into protection for the patient and the whole pedigree.

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