

## Opis choroby \*

### Definicja

A rare otorhinolaryngological malformation characterized by a hypoplastic or absent cochlear nerve, resulting in variable hearing loss or total deafness, depending on the quantity of nerve fibers present. The condition can be unilateral or bilateral, occur as an isolated malformation or in the context of a complex syndrome, and may be associated with a hypoplastic internal auditory or cochlear nerve canal.

### Dane

### Klasyfikacja

Wada morfologiczna

#### Kod ORPHA

502318

#### Kod OMIM

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#### Kod ICD10

H93.3

#### Kod ICD11

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### \*Źródło

orphanet