**Condition:** Hereditary deafness

**Inheritance:**
Genetically heterogeneous; most common single gene nonsyndromic cause due to mutation of *GJB2* encoding connexin26.

**Genetic etiology:**
Genetically heterogeneous.

**Frequency:**
Prelingual deafness affects approximately 1/1,000 children.

**Clinical features:**
Deafness is divided into sensorineural and conductive types. Approximately 50% of prelingual deafness is due to genetic causes, and approximately 70% of this consists of isolated deafness, referred to as “nonsyndromic.” Approximately half of this is DFNB1 due to *GJB2* mutation. Other forms are associated with additional features, such as thyroid disease (Pendred syndrome) or blindness (Usher syndrome).

**Management:**
Supportive care; cochlear implants are successful in some children with prelingual deafness.

**Genetic counseling:**
Based on specific genetic cause; molecular genetic testing available for many of the causative genes.