Mitochondrial Deafness in a National Hereditary Deafness DNA Repository
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Purpose: To describe the largest cohort of patients with mitochondrial deafness, and to determine the prevalence of mitochondrial deafness in the US population.

Methods: Retrospective review of subjects with mitochondrial DNA mutations in a national repository.

Results: Of 86 subjects, 72.2% had hearing loss before 3 years of age, and 62.1% had hearing loss before 6 months of age. The most common mutation was 961delT+C(n), which was present in 38.1% of subjects.

Discussion: This study confirms that mitochondrial deafness is a common cause of childhood hearing loss, and that early intervention is critical.

Conclusion: Early diagnosis and intervention are crucial for improving outcomes in children with mitochondrial deafness.

REFERENCES