

## The Genetics of Hearing Loss in the Palestinian Population

Hereditary Research Laboratory

### Why Hearing Loss?

### Hearing Loss in the Palestinian Population\*

Gender	Population	Affected Individuals	Percentage
Females	1,495,055	2,410	0.16
Males	1,524,649	2,885	0.18

\*Does not include those parts of Jerusalem annexed by Israel in 1967.  
Palestinian Central Bureau of Statistics, 1999, Population, Housing and Establishment Census 1997, Statistical Brief, (Summary of Census Results) Ramallah -Palestine.

### Hearing loss affecting the Palestinians is severe to profound bilateral congenital sensorineural in nature

Sample audiogram of an affected individual from Family K

Classification of Hearing Loss

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- There is no policy yet that addresses hearing loss as a public health threat.
  - Limited number of specialized schools for the deaf
  - Education is limited to up to the age of 16
  - Chances for rehabilitation are minimum and limited to vocational training
  - Lack of database
- The HRL is interested in the basic science that addresses relevant problems and ties it to the new advancement in the field of genetics

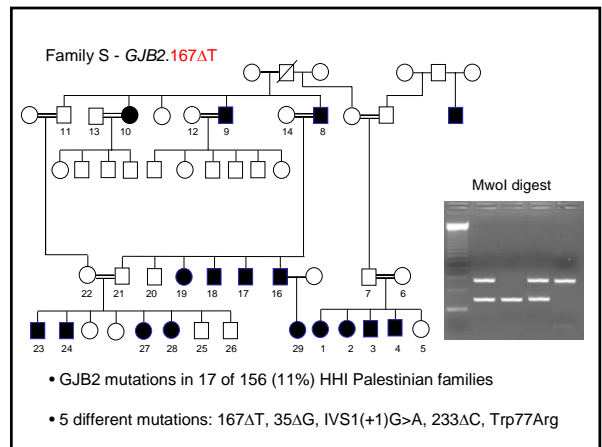
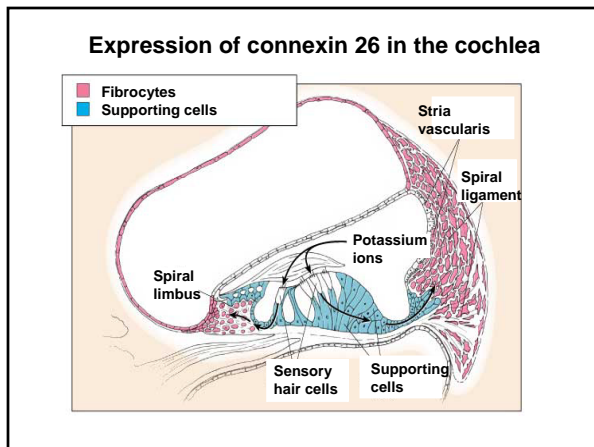
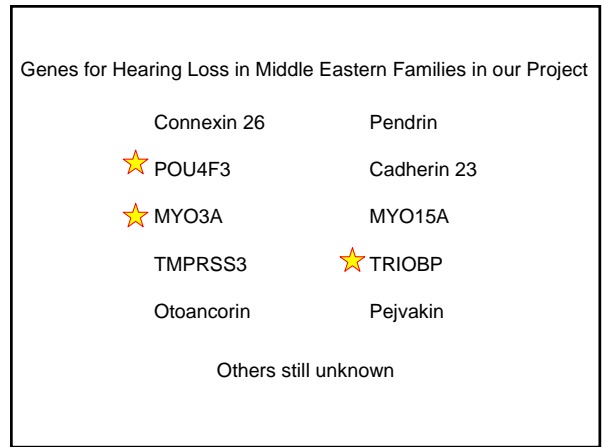
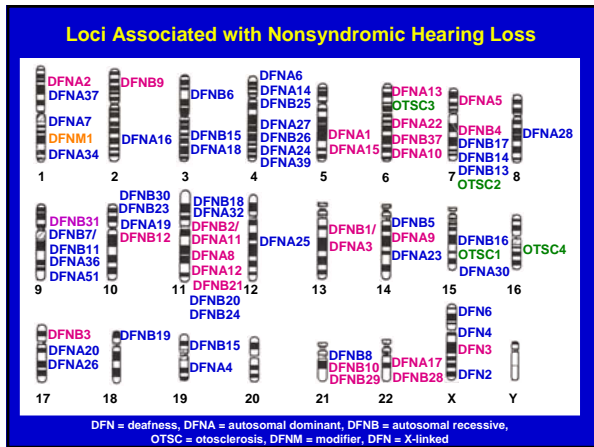
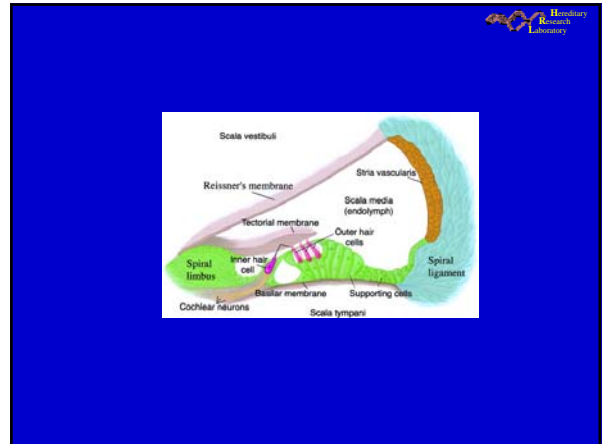
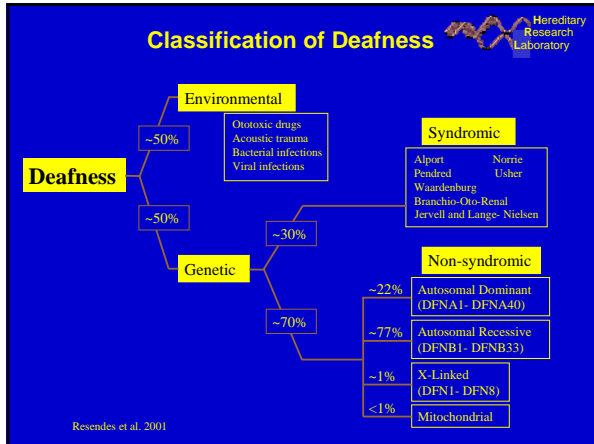
## The Inner Ear

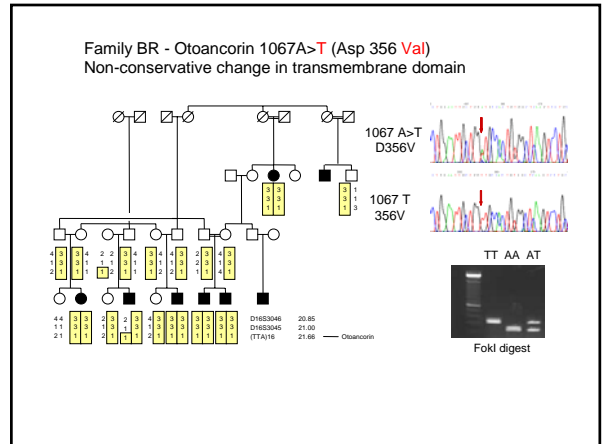
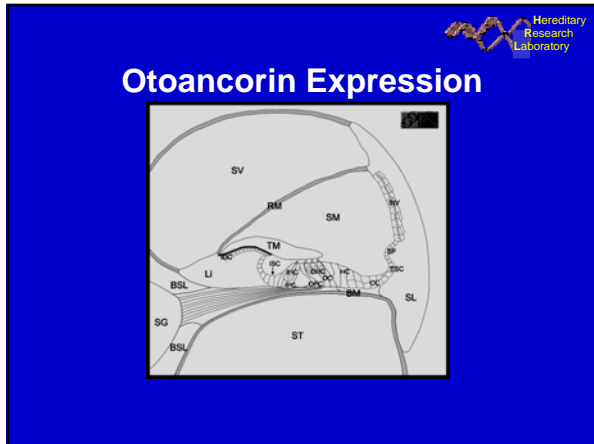
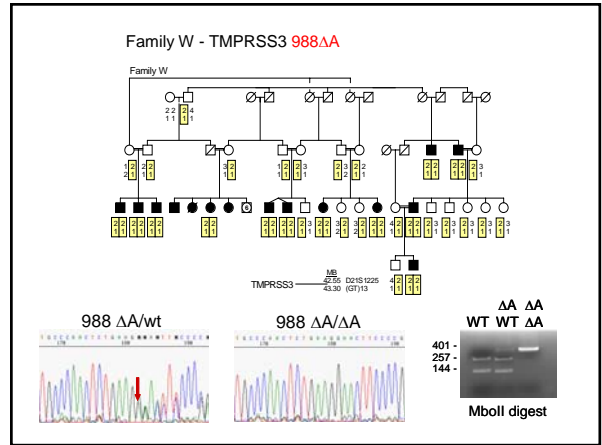
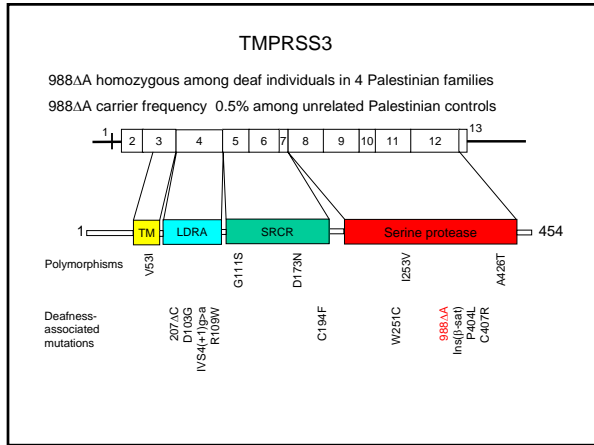
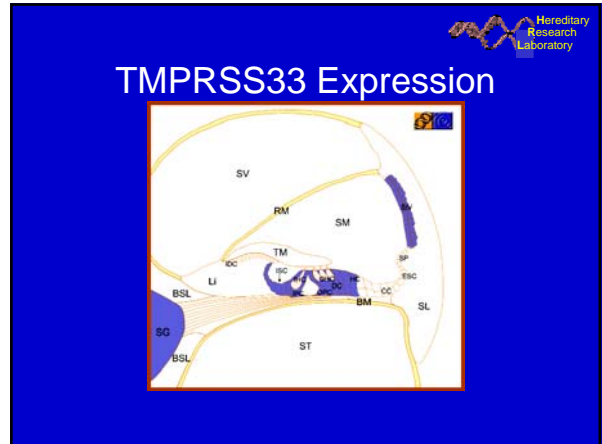
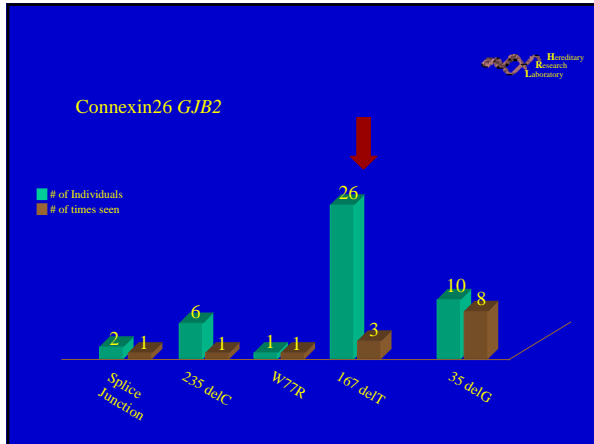
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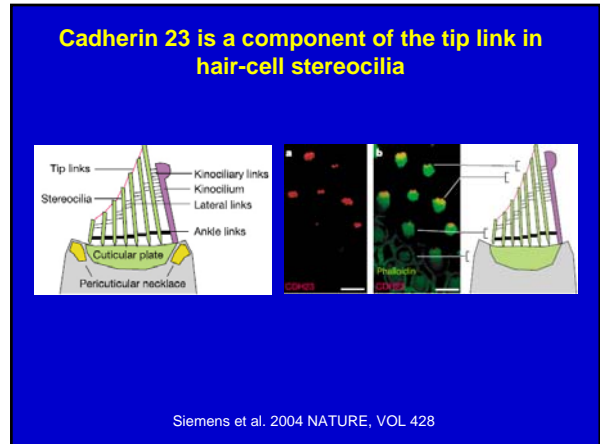
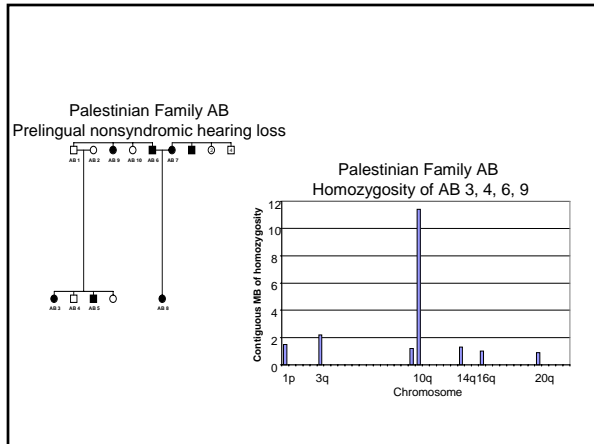
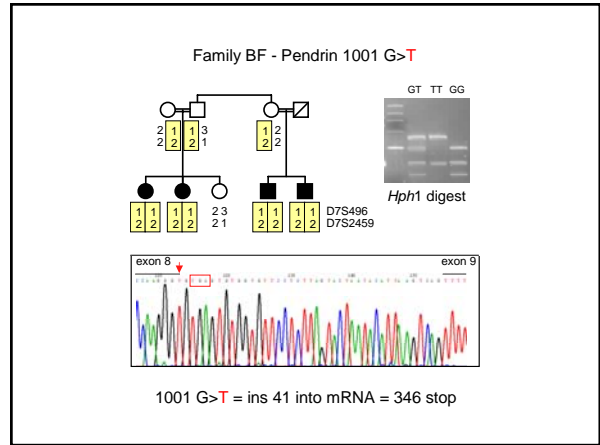
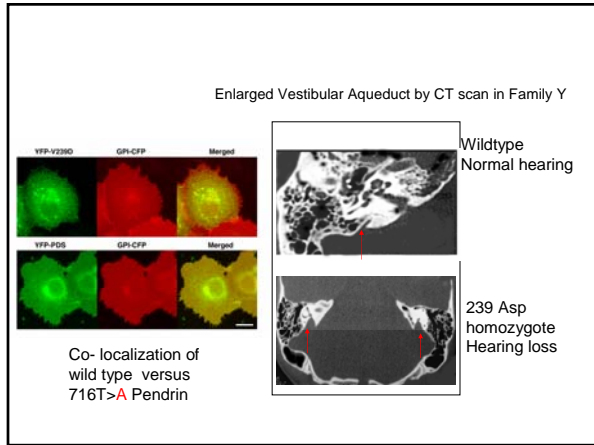
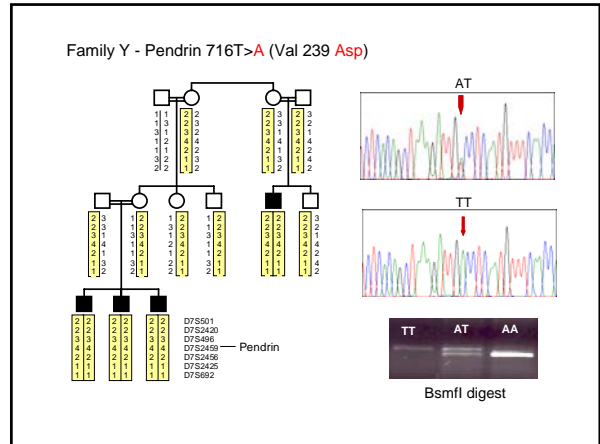
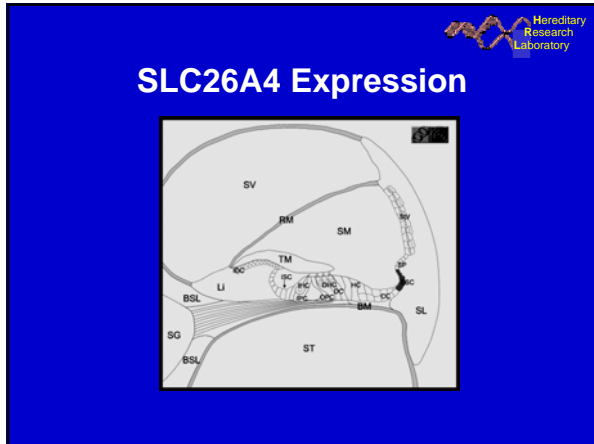
## Hair Cells – Function

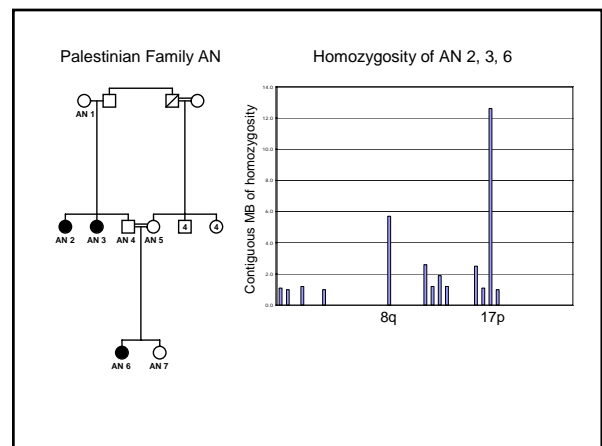
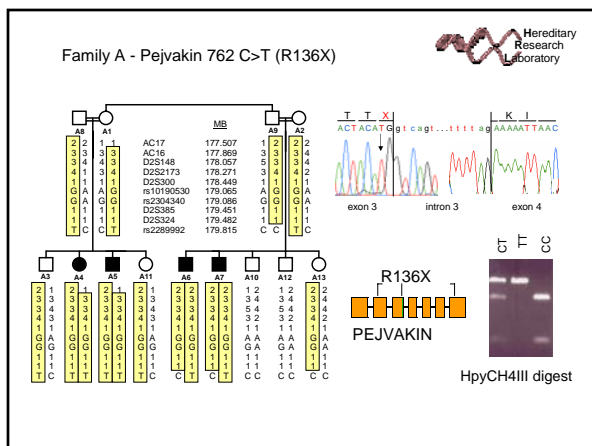
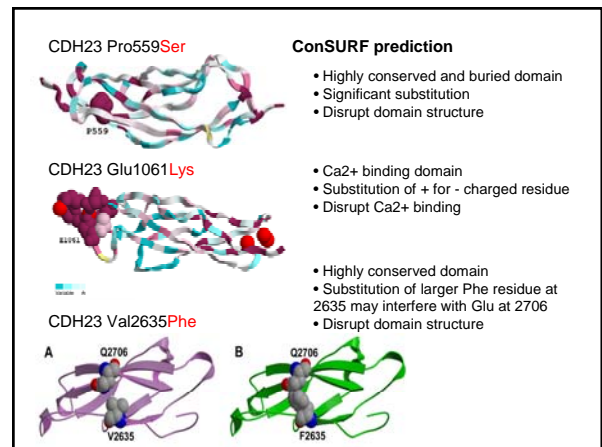
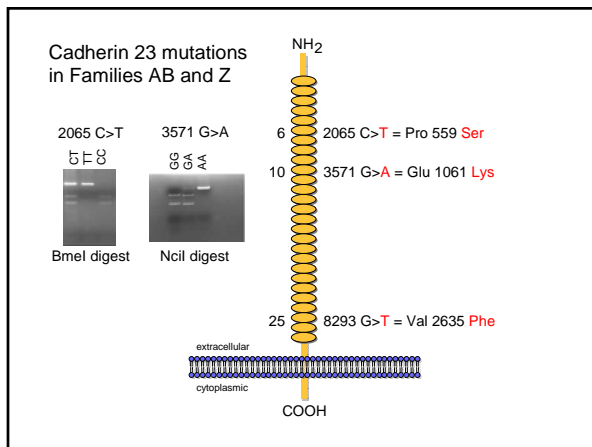
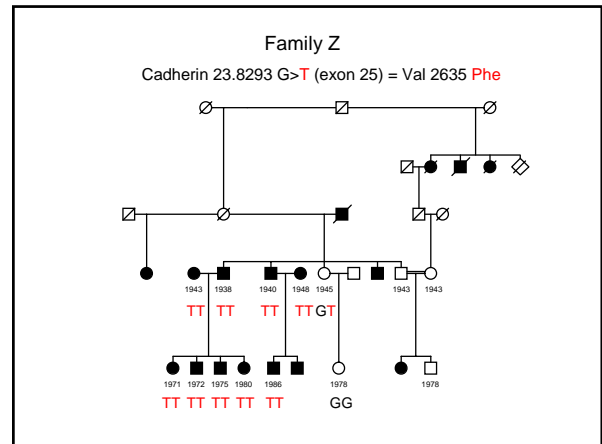
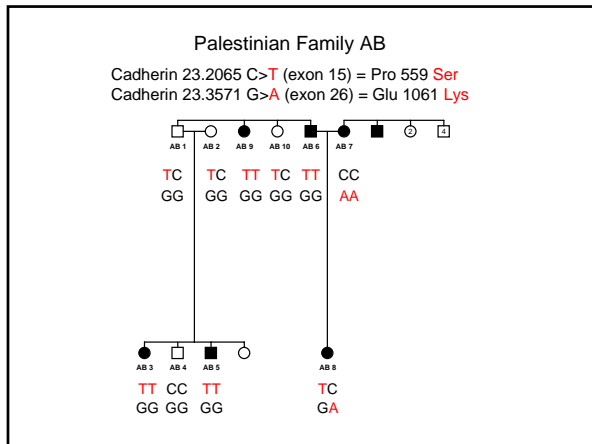
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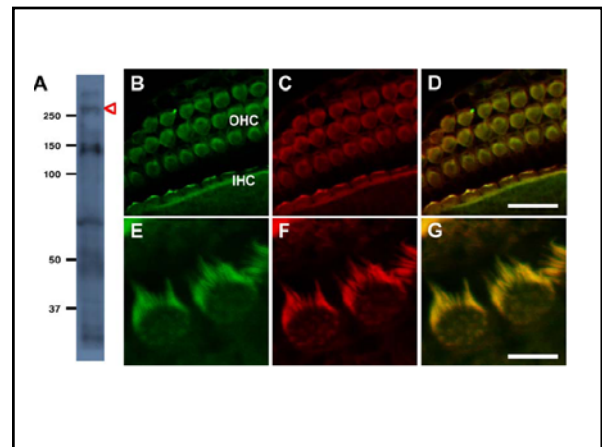
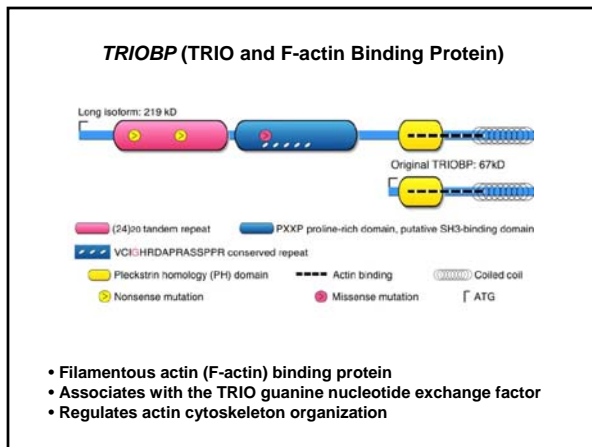
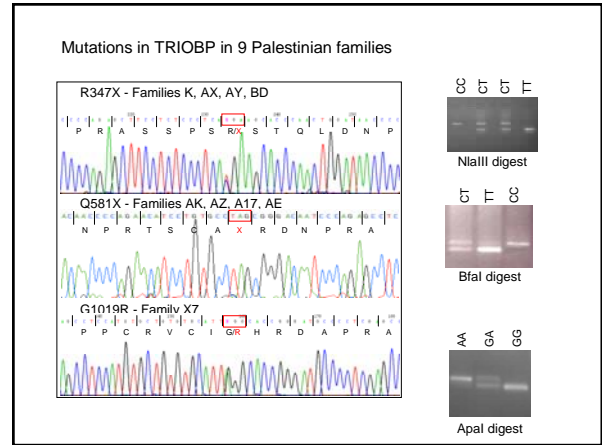
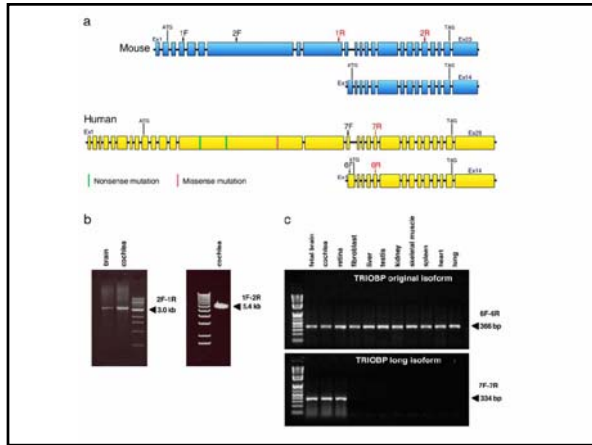
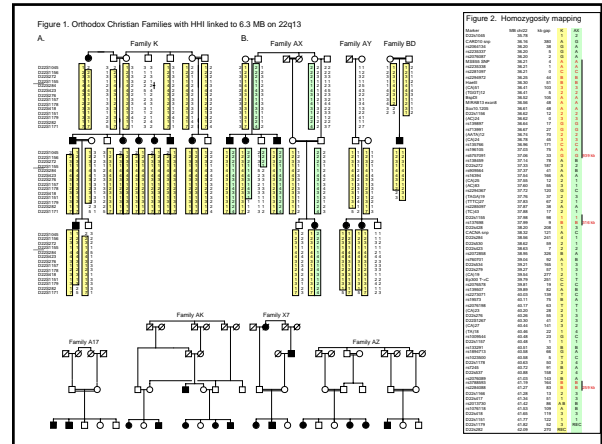
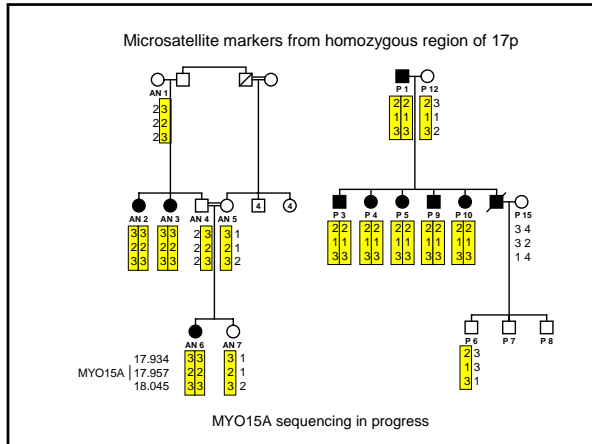
Auditory/Vestibular stimulus →  
 Stereocilia deflection →  
 Opening of ion channels & entrance of Ca<sup>++</sup> and K<sup>+</sup> ions →  
 Hair cell depolarization →  
 Neurotransmitter release at synapse

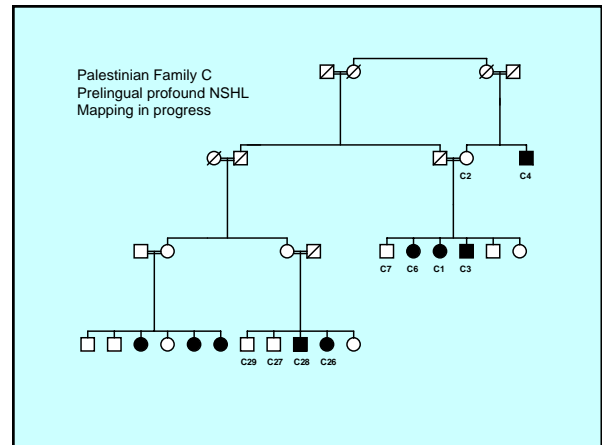
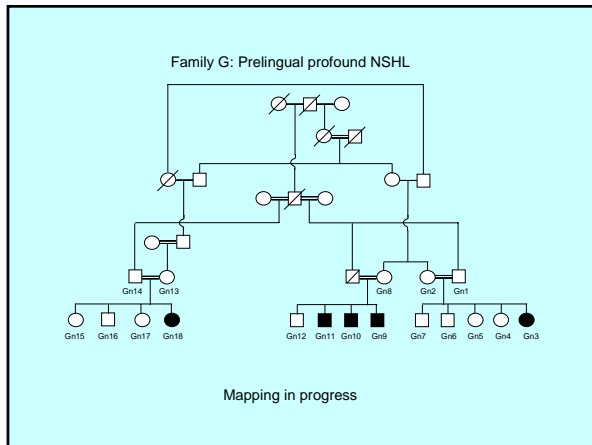
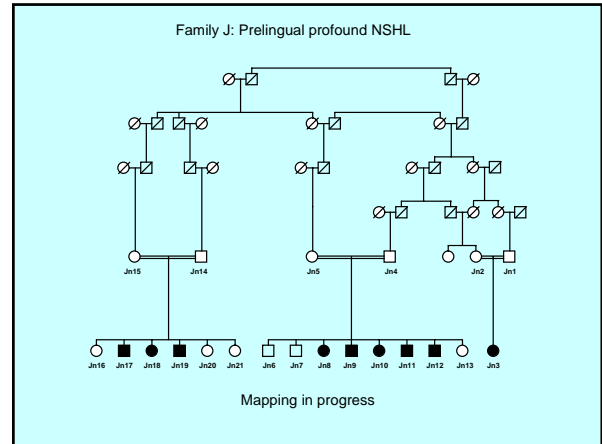
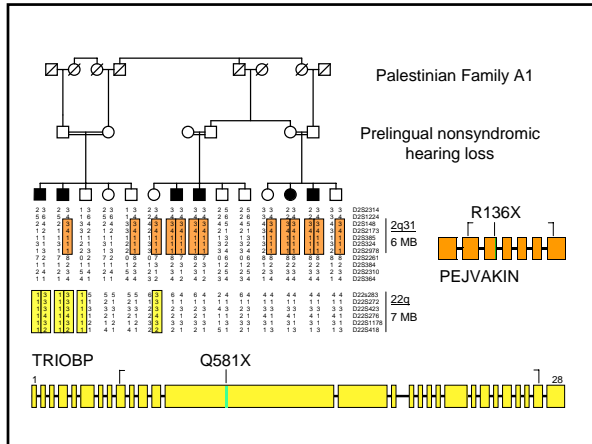












## Conclusions

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- Inherited hearing loss in the Palestinian population is highly heterogeneous, both in the number of genes involved and in the number of alleles at each gene.
- Alleles responsible for hearing loss in this population are individually very rare.
- Families with hearing loss who are wildtype for *GJB2* are likely to harbor novel alleles, either of known hearing-related genes (as in these three families) or of previously unknown genes.
- We have identified and cloned a new gene (*TRIOBP*) that underlines hearing loss in nine Palestinian families.
- Studies of multiple Palestinian kindreds are in progress to identify and characterize yet unknown genes.

## Genetics of Hearing Loss in Middle Eastern Families



### Bethlehem

Hashem Shahin  
Judeh Abu Sa'ed  
Amal Abu Rayan  
Wa'el Salhab  
Moien Kanaan



### Tel Aviv

Tama Sobe  
Zippora Brownstein  
Moshe Frydman  
Koret Hirschberg  
Karen B. Avraham



### Seattle

Tom Walsh  
Vanessa Walsh  
Lara Shamieh  
Ming K. Lee  
Mary-Claire King

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