

# Hereditary Hearing Loss: An Audiologist's Perspective

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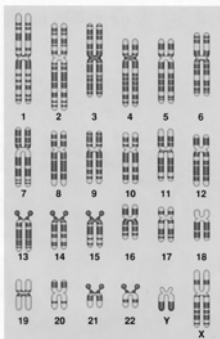
With special thanks to:  
Bronya Keats, PhD – LSUHSC, now Australia  
Kathleen Arnos PhD – Gallaudet University  
Walter Nance MD – Medical College of Virginia  
Arti Pandya MD – Georgetown University



# Genetics and Hearing Loss

- Congenital hearing loss occurs in approximately 1 to 2 of 1,000 births; higher in some populations and regions of the world.
- Genes are responsible for no less than 60% of cases of severe hearing impairment, and an underlying genetic component may contribute to almost all cases of hearing loss.
- A genetic etiology should be considered for every patient with a hearing problem. If no obvious environmental insult can be determined, the cause is most likely genetic.
- A family history of hearing loss is not necessary for the cause to be a genetic defect.
- Hereditary hearing loss can also have a later onset.

# Chromosomes



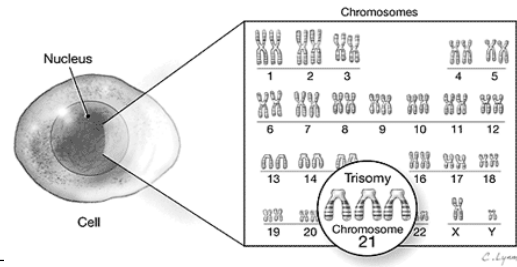
- 23 pairs for a total of 46 in the nucleus.
- X and Y and 22 paired autosomal chromosomes. Everybody has 2 copies of every gene o. The two copies are termed alleles.
- Down syndrome is the most common chromosomal defect (Trisomy 21).

Fig. 2.18 A karyogram of the human chromosome set

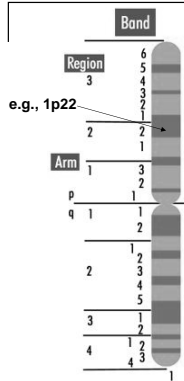
Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

# Karyotype of Down syndrome (trisomy 21)

Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

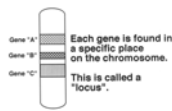


[http://www.meden.com/Medem/images/ama/da\\_AMA\\_Genetics\\_UnderstandingGenetics\\_Lev20\\_DownSyndrome\\_JPP\\_01.gif](http://www.meden.com/Medem/images/ama/da_AMA_Genetics_UnderstandingGenetics_Lev20_DownSyndrome_JPP_01.gif)



# Incidence of Chromosome Disorders

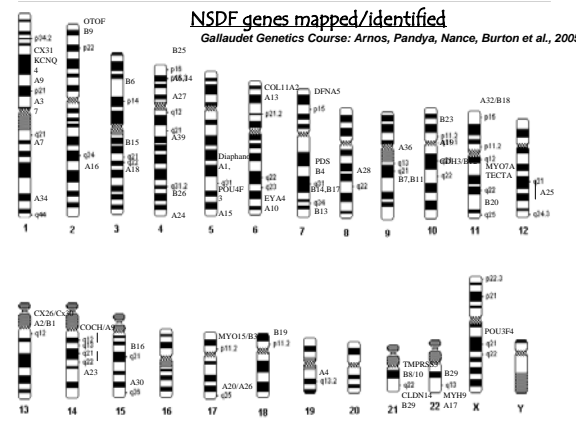
- Found in >7% of human conceptions
- Responsible for >50% miscarriages
- 1/200 live born with chromosome disorder



Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

# NSDF genes mapped/identified

Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005



## Inheritance Patterns

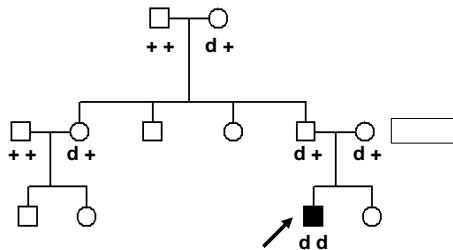
- **Autosomal<sup>1</sup> recessive**
  - 70 to 80% of non-syndromic hearing losses
- **Autosomal<sup>1</sup> dominant**
  - 15 to 20% of non-syndromic hearing losses
- **X-linked (2-3%) and mitochondrial (<1%)**
  - X-linked: 7 loci reserved, 1 gene identified
  - Mitochondrial: 7 nonsyndromic and 8 syndromic mutations identified
  - Y-linked: 1 locus reserved

<sup>1</sup>Autosomal: pertaining to chromosomes other than the sex chromosomes

## Method of Recessive Inheritance

- Each parent carries one normal and one abnormal gene.
- To display a recessive trait, two abnormal genes are inherited, one from each parent.
- Often no history of hearing loss in either parent's family.

## Method of Recessive Inheritance



From R. Morell, 2000

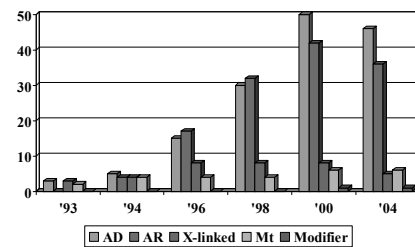
## Method of Recessive Inheritance

- The probability of inheriting two abnormal genes and being affected is 25%.
- The probability of being a carrier is 50%.
- The probability of inheriting two normal genes is 25%.

## Autosomal Recessive Inheritance

- Largest subgroup of hereditary hearing loss.
- Hearing loss is usually congenital.
- Males and females are both affected.
- Loci: DFNB1, DFNB2, etc.
- Currently:
  - 77 loci for non-syndromic recessive hearing loss reserved
  - 26 genes related to recessive hearing loss identified

## Non-Syndromic Deafness Loci

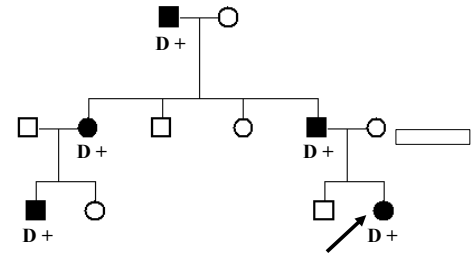


Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

## Method of Dominant Inheritance

- Inheriting a single copy of an abnormal gene can result in hearing loss.
- An affected parent has a 50% chance of passing that gene to their child.
- Occurs in each generation.

## Method of Dominant Inheritance



## Autosomal Dominant Inheritance

- Hearing loss has a tendency to be later in onset and progressive.
- More often part of a syndrome (e.g., Waardenburg syndrome)
- Males and females are both affected.
- Loci: DFNA1, DFNA2, etc.
- Currently:
  - 57 loci for non-syndromic dominant hearing loss reserved
  - 21 genes related to dominant hearing loss identified

## Genes for hereditary hearing loss

- Modifier genes:
  - 2 loci reserved: DFNM1, DFNM2
  - Pakastani family with 7 members homogeneous for DFNB26, but unaffected.
  - A dominant modifier gene was found that suppressed the deafness.

## Genes and the Ear

- Development
- Homeostasis
- Energy
- Structure

## Genes and the Ear

- Development
  - External/middle ear, branchial arches
    - Auricle, ossicles, TM
  - Inner ear
    - Labyrinths, organ of Corti
      - Hair cells, hair cell bundle, stereocilia, stria vascularis, tectorial membrane, supporting cells, etc.
    - Spiral ganglion, auditory nerve
    - Etc.

## Development – Hair Cell Bundle

- **Cadherin & Protocadherin**
  - Cell-cell adhesion molecules, stereocilia
- **Ptprq**
  - Receptor like lipid phosphatase, stereocilia
- **Vlgr1**
  - G-protein couple receptors, hair cell development
- **Usherin**
  - Localized in stereocilia and synaptic region
- **Myosin**
  - Molecular motors, hair cells and stereocilia
- **PDZ**
  - Whirlin and harmonin, stereocilia

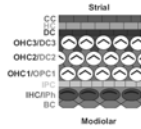


Fig. 2 Schematic diagram of cellular partitioning in the organ of Corti. Inner hair cells (IHC) are arranged in a single row located on the modiolar side. Outer hair cells are arranged in three rows (OHC1, OHC2, and OHC3) on the stria vascularis side. On the stria vascularis side, the outer hair cells are arranged in three rows (OHC1, OHC2, and OHC3) and the inner hair cells are arranged in a single row (IHC). The diagram shows the arrangement of cells in the organ of Corti, including the stria vascularis, the modiolus, and the inner and outer hair cells.

## Genes and the Ear

- **Homeostasis**
  - **Cochlear endolymph is uniquely characterized by high potassium concentration**
    - Influx of K into inner hair cells is needed to convert hair cell stimulation into nerve impulses
  - **Several forms of genetic deafness are related to defects in potassium maintenance**
    - Jervell and Lange-Nielsen syndrome
    - Connexins (Cx26 and Cx31) - role in high K maintenance in the cochlea
    - DFN3: bony defect allowing mixing of endolymph and perilymph; exceeds capacity of stria vascularis to maintain high K levels

## Genes and the Ear

- **Energy**
  - Ear needs high energy to maintain its sensitivity
  - Stria vascularis, responsible for maintaining the endolymphatic potential (store of energy for the ear)
  - Energy loss is connected to mitochondrial defects that can underlie some forms of deafness as part of a syndrome; also related to diabetes and ototoxicity

## Genes and the Ear

- **Structure**
  - Genes that code for forms of collagen
    - Deficient in Alport syndrome; Stickler syndrome
  - Expressed in structures of stria vascularis
  - Also has been related to the tectorial membrane

## Non-syndromic hereditary hearing loss

## Hearing Loss related to *GJB2* (connexin 26) mutations

- The gene *GJB2* encodes a protein called connexin 26 (Cx26).
- Cx26 proteins form gap junctions that facilitate exchange of electrolytes, second messengers, and metabolites.
- Cx26 is essential to maintenance of high potassium in the scala media of the inner ear.
  - Problems in K recycling, cell death, organ of Corti degeneration
- *GJB2* locus maps to 13q11.12

Denoyelle et al., 1997; Zelante et al., 1997

## Connexin 26 (GJB2)

- High percentage of recessive pre-lingual deafness
- Single mutation (35delG) most common
- Also associated with dominant deafness
  
- Single copy mutation of GJB2 (Cx26) in combination with GJB6 (Cx30)
  - Proximity on chromosome 13
  - Effect on transcription, regulatory elements
  - Digenic?

Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

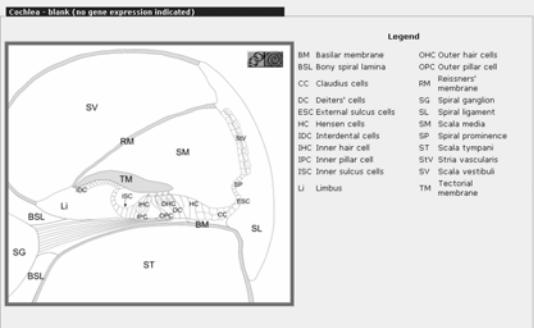
## Mutational Spectrum in the GJB2 gene

### More than 80 mutations

- |                        |                   |
|------------------------|-------------------|
| ■ 35delG               | European descent  |
| ■ 167delT              | Ashkenazi Jewish  |
| ■ 235delC              | Asian descent     |
| ■ W44C (G to C at 132) | dominant mutation |
| ■ G59A /D66H / R75W    | dominant          |
| ■ M34T (T to C at 101) | mild, pathogenic? |

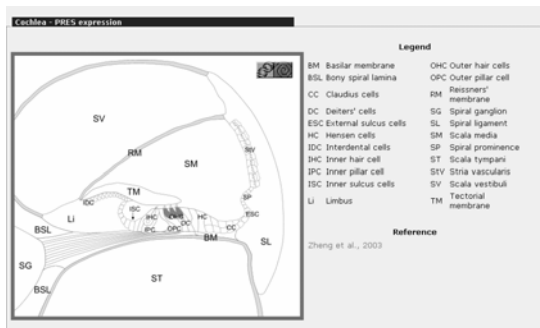
Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

## Cochlear Structures



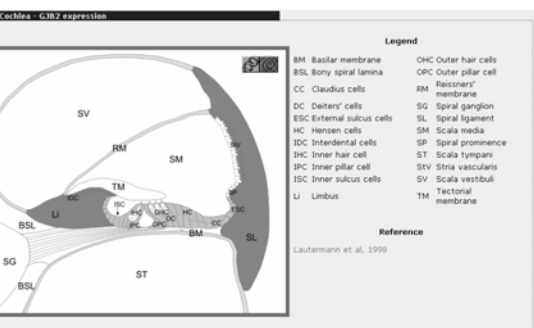
Hereditary Hearing Loss Homepage – Van Camp and Smith

## PRES (prestin) expression



Hereditary Hearing Loss Homepage – Van Camp and Smith

## GJB2 Expression



Hereditary Hearing Loss Homepage – Van Camp and Smith

## Severity of Hearing Loss as a function of type of Cx 26 mutation

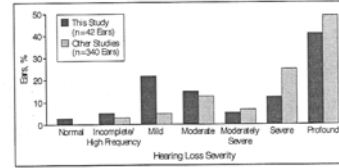
(from Cohn et al., 1999)

Degree of Loss	35delG/ 35 del G	167delT/ 167delT	Compound Heterozygote
Profound	11	2	4
Severe-Profound	9	3	1
Severe	1	0	1
Moderate-severe	4	1	3
Mild-severe	1	0	0
Moderate	1	0	0
Mild-Moderate	4	0	0

**Progression of Hearing Loss as a function of type of Cx 26 mutation**  
(from Cohn et al., 1999)

Degree of Loss	35delG/ 35 del G	167delT/ 167delT	Compound Heterozygote
Progression	5	2	3
Stable	14	2	1
Fluctuation	3	0	0

**Audiometric phenotype in Cx26 patients**



Hearing loss severity in patients with bilateral connexin 26 mutations in this study and others<sup>11-12</sup> in this study, severity of hearing loss was scored separately in each ear. Data were not always available in this format in the other studies, so the combined reported severity was assumed to be present in both ears.

From Kenna et al., 2001: Connexin 26 studies in patients with SNHL

**Mutations: changes in the genetic code**

Normal AGT ATA GGA CTT TTG  
 Mutant AGT ATA AGA CTT TTG

**Deletion or insertion**

Normal AGT ATA CGA CTT TTG ...  
 Serine Isoleucine Arginine Leucine Leucine  
 Mutant AGT ATA CGA TCT TTT ...  
 Serine Isoleucine Arginine Serine Phenylalanine

From D Emanuel, Towson State University

**Substitution of single base**

Normal AGT ATA CGA CTT TTG  
 Serine Isoleucine Arginine Leucine Leucine  
 Mutant AGT ATA TGA CTT TTG  
 Serine Isoleucine Stop

From D Emanuel, Towson State University

**Types of Mutations – A sentence comprised of 3-letter words can be used as an analogy to the effect of mutations on a gene's sequence.**

Wild Type THE ONE BIG FLY HAD ONE RED EYE  
 Missense THQ ONE BIG FLY HAD ONE RED EYE  
 Nonsense THE ONE BIG  
 Frameshift THE ONE QBI GFL YHA DON ERE DEY  
 Deletion THE ONE BIG HAD ONE RED EYE  
 Duplication THE ONE BIG FLY FLY HAD ONE RED EYE  
 Insertion THE ONE BIG WET FLY HAD ONE RED EYE

## Genotype and Phenotype

- **35delG/35delG homozygote**
  - Most common genotype
    - Truncating mutation
  - Severe-profound deafness in 82-90% of patients
- **35delG heterozygote**
  - With other *GJB2* mutation (e.g., 35delG/167delT)
  - Severe-profound deafness in about 60% of patients
- **Two missense mutations**
  - Substitution of one amino acid for another
  - Severe-to-profound deafness rarely observed

Azaiez, van Camp, Smith, 2006 Sem Hearing

Genotype \ Phenotype	Phenotype			
	Mild	Moderate	Severe	Profound
35delG/35delG				
Truncating/ Truncating				
Truncating/ Nontruncating				
Nontruncating/ Nontruncating				

□ Percentage of patients < 1%  
 □ Percentage of patients < 15%  
 □ Percentage of patients < 25%  
 □ Percentage of patients < 45%  
 □ Percentage of patients

Figure 4. Phenotype-genotype correlations. Truncating mutations are nonsense mutations, insertions, and deletions that disrupt open reading frame (nonsense mutations). Nontruncating mutations are missense mutations, deletions, or insertions of three or six nucleotides. Based on data from (Pittard et al., 2002; Azaiez et al., 2006; and Crisic et al., 2006).

Azaiez, van Camp, Smith, 2006 Sem Hearing

## Genetics and AN/AD

- Affected siblings or other family members with auditory neuropathy/dys-synchrony suggests a genetic form of AN/AD.
- Recessive, dominant, and mitochondrial inheritance patterns are associated with AN/AD.
- AN/AD can be part of a syndrome or non-syndromic.

Candidate Genes (gene name, loci or protein product):

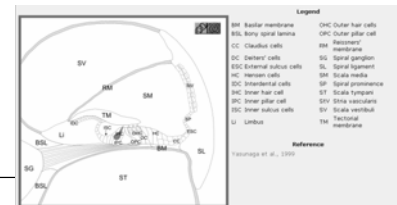
- |  |                             |
|--|-----------------------------|
| • <i>OTOF</i>  | • <i>PJVK</i>               |
| • 13q14-21 ( <i>AUNA1</i> )  | • Xq23-27.3 ( <i>AUNX</i> ) |
| • 12SrRNA (mtDNA)  | • <i>SLC19A2</i>            |
| • <i>FXN</i>   | • <i>ERG2</i>               |
| • <i>PMP22</i>   | • <i>MPZ</i>                |
| • <i>Cx26</i> ( <i>GJB2</i> ), <i>Cx29</i> , <i>Cx30</i> ( <i>GJB6</i> ), <i>Cx31</i> ( <i>GJB3</i> ), <i>Cx32</i> ( <i>GJB1</i> ) |                             |

## Genetics and AN/AD

- Non-syndromic recessive AN is associated with abnormalities in *OTOF* – *otofelin* (Varga et al., 2003).
  - *Otofelin* is expressed in the inner hair cells, possible roles in membrane trafficking and/or IHC synaptic vesicle fusion
  - In mice, *otofelin* has been localized to IHC associated synaptic vesicles

**OTOF expression**

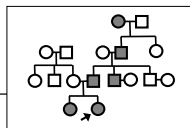
From: Hereditary Hearing Loss Homepage  
(Smith and Van de Camp)



## Genetics and AN/AD

AN/AD occurs as part of a syndrome with various inheritance patterns

- Accompanying other hereditary motor sensory neuropathies - HMSN (e.g., Butinar et al., 1999; Starr et al., 2004)
- Charcot-Marie-Tooth disease
- Friedreich's ataxia
- AN and optic nerve abnormalities
- Case reported with cri-du-chat
  - 5p del syndrome; Swanepoel (2007)



## Genetic Susceptibility to Aminoglycoside Ototoxicity

- Genetic predisposition to developing deafness as a result of aminoglycoside sensitivity
- Common cause of irreversible hearing loss
- Mitochondrial inheritance
  - Characteristic matrilineal inheritance
- Point mutation (1555A>G)
- History of exposure to streptomycin
- Common in Spanish & Oriental populations

Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

## Classified by Association with Other System Disorders

- Eye disorders (Usher)
- Endocrine disorders (Pendred)
- Cardiac disorders (Jervell & Lange-Nielsen)
- Pigmentary disorders (Waardenburg)
- Renal disorders (BOR and Alport)
- Musculoskeletal disorders (Stickler)

*Percentage of hearing loss: About 15%*  
*Genes – predicted: About 400 (Gorlin et al.)*

## Syndromes

- Genes have been identified for:
  - Alport (2)
  - Branchial-oto-renal (BOR) (3)
  - Jervell and Lange-Nielsen (2)
  - Norrie (1)
  - Pendred (1)
  - Stickler (4)
  - Treacher-Collins (1)
  - Usher (9)
  - Waardenburg (7)

## Branchio-Oto-Renal Syndrome

- Autosomal Dominant
- Conductive, sensorineural or mixed loss, often progressive
- Pre-auricular pits, and/or malformation of ear with branchial fistulae or cysts
- Association with enlarged vestibular aqueduct (EVA, LVA)
- Renal anomalies
- Genes identified: EYA1 (8q13.3), SIX1 (14q21.3-q24.3)

## Jervell and Lange-Nielsen Syndrome

- Autosomal recessive
- Congenital sensorineural hearing impairment with electrocardiographic abnormalities, fainting spells, and sudden death
- 1 in every 100 infants with profound hearing impairment may have this syndrome
- Genes identified: KCNQ1 (11p15.5), KCNE1 (21q22.1-q22.2)
  - encode proteins that form potassium channels

## Pendred Syndrome

- Autosomal recessive
- Profound congenital sensorineural hearing impairment, goiter, enlarged thyroid
- 20% of children with severe to profound hearing loss may have Pendred syndrome
- Association with enlarged vestibular aqueduct (EVA, LVA)
- Locus name: PDS; chromosome location: 7q21-34; gene: SLC26A4
  - Mutations in PDS are also associated with non-syndromic hearing impairment (DFNB4)

## Usher Syndrome: Clinical Classification

Usher syndrome is characterized by hearing impairment and retinitis pigmentosa. Usher syndrome is classified into 3 types based on clinical findings.

Type	Hearing impairment	Vestibular responses	Onset of retinitis pigmentosa
Type I	Profound hearing loss congenital	Absent	Onset in first decade first decade
Type II	Sloping audiogram Congenital	Normal	Onset in first or second decade
Type III	Progressive loss	Variable	Variable

### Usher Syndrome: Molecular Classification

The three types of Usher syndrome are further divided into subtypes based on differences in underlying genetic location.

Locus Name	Location	Gene
USH1B	11q13.5	MYO7A
USH1C	11p15.1	USH1C
USH1D	10q22.1	CDH23
USH1E	21q21	unknown
USH1F	10q21-22	PCDH15
USH1G	17q24-25	SANS
USH2A	1q41	USH2A
USH2B	3p23-24.2	unknown
USH2C	5q14.3-q21.3	VLGR1
USH2D	9q32	WHRN
USH3	3q21-q25	USH3

### Waardenburg Syndrome

- Clinical description: dystopia canthorum; pigmentary abnormalities of hair, iris and skin; SNHL
- Hearing loss mild to profound, unilateral and bilateral
- Clinical classification
  - Type I – dystopia canthorum
  - Type II – no dystopia canthorum
  - Type III – Klein-Waardenburg syndrome
    - (Type I and upper limb abnormalities)
  - Type IV – Waardenburg-Shah syndrome
    - (Type II and Hirschsprung disease, AR)
- 7 molecular subtypes; 7 genes identified

### Role of Audiologists and Other Clinicians

- Obtain accurate phenotype information

### Defining Hearing Loss: Genotype and Phenotype

- Genotype describes an individual's genetic constitution.
  - The allele combinations in an individual that cause a particular trait or disorder.
- Phenotype relates to the physical characteristics of an individual.
  - Can include information from clinical studies (physiological, morphological, biochemical)

Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

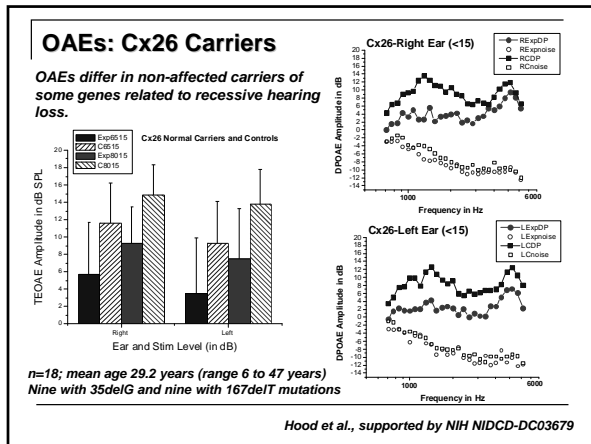
### Evaluation and classification of hearing loss

- Pure-tone audiogram
  - degree of loss, frequencies involved, configuration
  - type (conductive, sensorineural, mixed)
  - symmetry between ears
- OAEs and other auditory methods may show greater sensitivity and suggest altered cochlear or neural function where pure tone thresholds do not.
- Look for dysmorphic features and features that may suggest a syndrome.

### Distortion product otoacoustic emissions in patients with Waardenburg syndrome

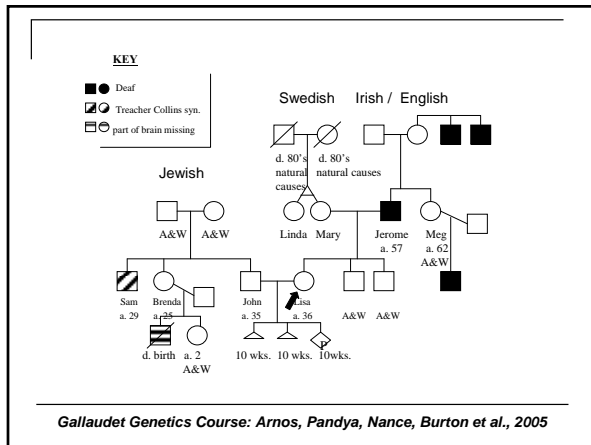
Lui and Newton, 1997

- Subjects: Eight Waardenburg syndrome patients (3 type I, 5 type II), no complaint of hearing loss, normal pure-tone audiogram
- 7 of 8 patients showed wide notches in DPOAEs in the 1000 to 3000 Hz range.
  - All type II patients had notches.
  - Two ears of type I patients had notches.
  - In one type I family, all four affected patients had notches, while two unaffected individuals had no notches.
  - Control subjects: 10% showed notches.



## Role of Audiologists and Other Clinicians

Obtain accurate family history information and construct a family pedigree



## Family History

- Couples with a previous child with a chromosome abnormality or genetic disorder
- Couples who have a genetic disorder, are known carriers of a disorder or have a family history of a genetic disorder
- Couples who have had more than 2 miscarriages
- Individuals of almost every ethnic group have an increased risks for particular genetic conditions

Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

## Role of Audiologists and Other Clinicians

Collaborate with geneticists and genetics counselors

## What to do if you suspect that a hearing loss is genetic?

- Refer the patient/family to a geneticist or a genetics counselor.
- Why?
  - Genetics and hereditary issues are not simple.
  - The questions and issues raised by gene testing can challenge family and other personal relationships.
  - Genetic counselors are trained to help persons as they consider genetic testing, when they receive results and in the weeks and months afterwards.

## Genetic Counseling

This process integrates:

- ▶ Collection and interpretation of family and medical histories to assess the chance of disease occurrence or recurrence
- ▶ Education about inheritance, testing, management, prevention, resources and research
- ▶ Counseling to promote informed choices and adaptation to the risk or condition.

Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

## The Human Genome Project: Science, Health, and Society

- The Human Genome Project and the resultant "new genetics" have great potential for:
  - Benefiting individuals
    - Preserving health
  - Harming individuals
    - Privacy issues
  - Changing health care
    - Individualizing care
  - Changing society
- Mapping the human genome is just the beginning...
- There is great need to decide how genetics information will be used and to prepare health professionals and society to use it effectively.

## Why is genetic information ethically complex?

- Includes family health data, parentage, reproductive options, future health risks
  - Who/What we are
- Non-medical uses of genetic information
  - Insurance, employment, criminal law, litigation, domestic relations, immigration, settle estates, paternity, kinship



Gallaudet Genetics Course: Arnos, Pandya, Nance, Burton et al., 2005

## Ethical, Legal and Social Issues (ELSI)

- Privacy of genetic information
- Safe and effective introduction of genetic information in the clinical setting
- Fairness in use of genetic information
- Professional and public education
- 3-5% of the Human Genome Project budget devoted to ELSI

## Contemporary Attitudes Towards Genetics Among the Deaf

- Attitudes towards:
  - Genetic testing for deafness in general
  - Prenatal diagnosis for deafness
  - The use of genetic testing for partner selection
  - Newborn hearing screening and newborn screening for deafness genes

## Attitudes Surveys

- 96 hearing parents of deaf children who had undergone genetic testing
  - The majority had a positive attitude towards genetic testing including prenatal diagnosis
  - None said they would use the information to terminate the pregnancy

Brunger et al. (2000). Parental attitudes toward genetic testing for pediatric deafness. *AJHG* 67:1621-1625.

## Attitude Surveys

- 644 deaf, 143 hard of hearing and 527 hearing individuals with either a deaf parent or a deaf child
  - 21% of deaf, 39% of hard of hearing and 49% of hearing said they would consider prenatal diagnosis for deafness
  - 2% of deaf participants said they would consider a therapeutic abortion of a hearing baby

Middleton, Hewison & Mueller (2001). Prenatal diagnosis for inherited deafness – what is the potential demand? *J Genet Counsel* 10:121-131.

## Attitude Surveys

- Stern, Arnos, Murrell, Welch, Nance, Pandya (2002). Attitudes of deaf and hard of hearing subjects towards genetic testing and prenatal diagnosis of hearing loss. *J Med Genet* 39:449-453.
- Taneja, Pandya, Foley, Nicely, Arnos (2004). Attitudes of deaf individuals towards genetic testing. *Am J Med Genet* 130A:17-21.

## Conclusions

- Most people support newborn screening, genetic testing, and new discoveries in genetics.
- Somewhat fewer support prenatal genetic testing.
- Attitudes vary amongst hearing, hard of hearing and deaf individuals.

## Websites

- Hereditary Hearing Loss Homepage
  - G. Van Camp and R. Smith
  - <http://webh01.ua.ac.be/hhh/>
- OMIM
  - Online Mendelian Inheritance in Man
  - <http://www.ncbi.nlm.nih.gov/omim/>

## OMIM

- OMIM is a comprehensive, authoritative, and timely compendium of human genes and genetic phenotypes.
- Full-text, referenced overviews contain information on all known Mendelian disorders and over 12,000 genes.
  - OMIM focuses on the relationship between phenotype and genotype.
  - It is updated daily, and the entries contain copious links to other genetics resources.
- This database was initiated in the early 1960s by Dr. Victor A. McKusick as a catalog of Mendelian traits and disorders.



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