BETHLEHEM UNIVERSITY

Hereditary Research Laboratory

HEARING AWARENESS WEEK

Better hearing for persons of all nations is an achievable, important goal, given that a disabling hearing impairment affects about 4.2 percent of the world's population, with two thirds of such persons living in developing countries

Why Hearing Loss?

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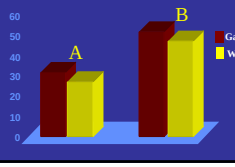
Hearing loss occurs at a rate almost double the rate world wide. It affects 2/1000 live births.

Hearing Loss in the Palestinian Population*

Gender	Population Number	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.



A: First cousin marriages
B: Marriages within extended family

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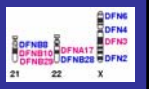
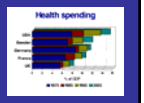
Three Important Developments

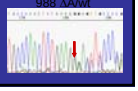
1-The first was the application of objective noninvasive physiological tests for hearing loss

2-The second that tests could be administered by nonprofessional personnel.

3-The third was the demonstration that the early detection influences the educational outcome of affected infants.







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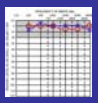
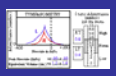
Techniques In Hearing Evaluation:

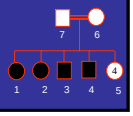
1-The automated auditory brain-stem response measures average neural response to a large number of repeated sound signals of the same pitch and intensity,



2-Whereas measurement of spontaneous or sound-induced otoacoustic emissions detects sound produced by movements of outer hair cells of the cochlea.



Both methods have acceptable sensitivities and specificities and are often used together in two-stage screening protocols.



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Ramifications

1-The average age at which hearing loss is confirmed has dropped from 24 to 30 months to 2 to 3 months

2-Infants in whom remediation is begun within six months are able to maintain language and social and emotional development that is commensurate with their physical development, in striking contrast to those whose hearing loss is first detected after six months of age

3-Would also permit prompt initiation of genetic evaluation, counseling, and testing and would serve as model programs for the delivery of these services.

4-provide valuable epidemiologic data on secular trends in the incidence of genetic and environmental causes of hearing loss, as well as variation in specific forms of hearing loss between populations.

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Limitation Of Existing Screening Program

1-Existing universal screening programs to identify hearing defects in newborns still do not enjoy the extraordinarily high follow up rates for positive test results that characterize most metabolic screening programs for newborns.

2-Another limitation is that some forms of early-onset hearing loss are not apparent at birth.

3-Testing protocols are not standardized to permit the comparison and aggregation of data from different sites and to avert the failure to identify infants with specific forms of hearing loss, such as auditory neuropathy.

4-Finally, most screening programs have lacked an etiologic focus, which may compromise meaningful interpretation of the results of early intervention.

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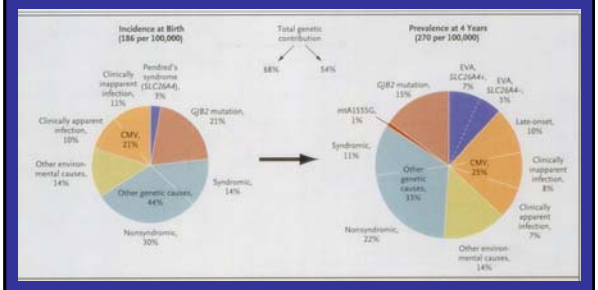

Cynthia; et al. NEJM.2006

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Table 1. Risk Indicators for Audiologic Monitoring for Progressive or Delayed-Onset Sensorineural Hearing Loss, Conductive Hearing Loss, or Both, in Infants (29 Days through 2 Years of Age) with Normal Hearing on Newborn Screening.*

Parental or caregiver concern regarding child's hearing, speech, language, or developmental delay
Family history of permanent hearing loss in childhood
Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian-tube dysfunction
Postnatal infections associated with sensorineural hearing loss, including bacterial meningitis
In utero infections such as cytomegalovirus infection, herpes, rubella, syphilis, and toxoplasmosis
Neonatal indicators such as hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal-membrane oxygenation
Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and some forms of Usher's syndrome
Neurodegenerative disorders such as Hunter's syndrome or sensory neuropathies such as Friedreich's ataxia and Charcot–Marie–Tooth syndrome
Head trauma
Recurrent or persistent otitis media with effusion for at least 3 months

* Information is from the Joint Committee on Infant Hearing.[21]

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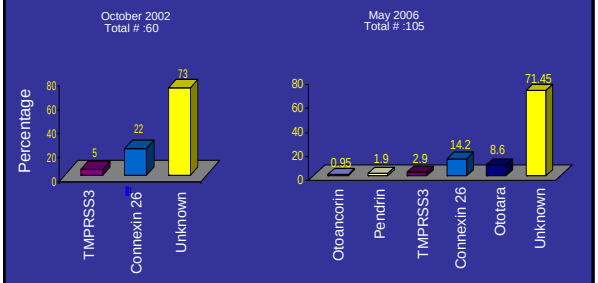
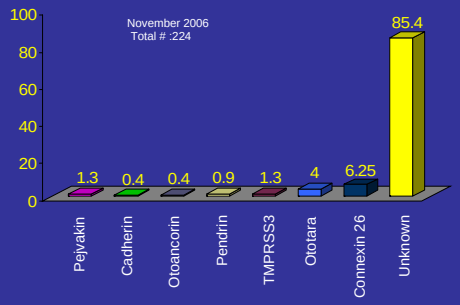
Palestinian Families With Deafness



Palestinian Families With Deafness …continue



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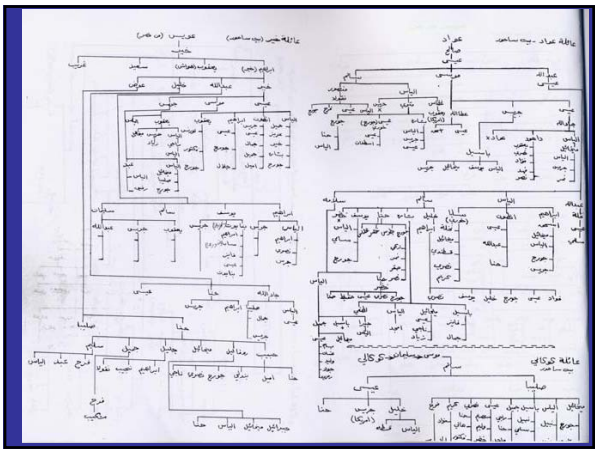
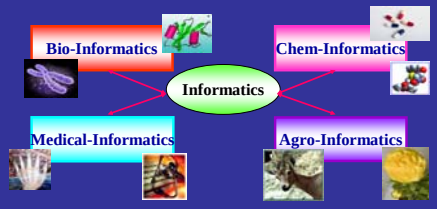


Bioinformatics



Application of IT in biology

- IT - acquire, store and manage data
- Bio/IT - develop software and databases
- Bio – gain biological understanding