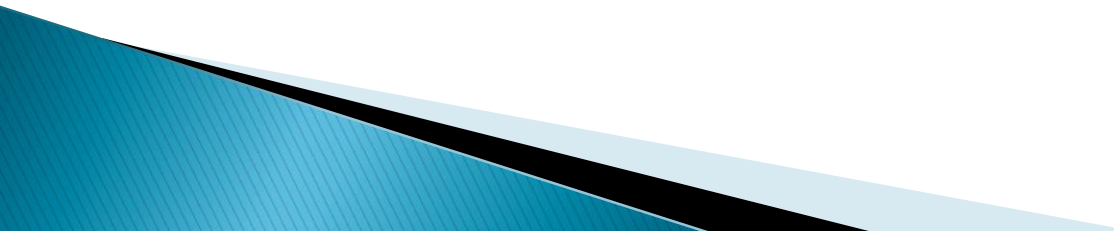


Why Children with Down Syndrome have an Excuse not to Listen - Hearing Problems

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Scope of Problem

- 38 - 78% incidence of hearing loss in Down Syndrome
 - Usually "mild"
 - Worse in young children
 - Can influence speech acquisition
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Mild hearing loss affects articulation


- Vocabulary
- Education (IQ)
- Emotional development (frustration)

Types of Hearing Loss

▶ Conductive
Sound energy
getting to nervous
system: 78%

▶ Sensorineural
Problems with inner
ear or nerves: 22%

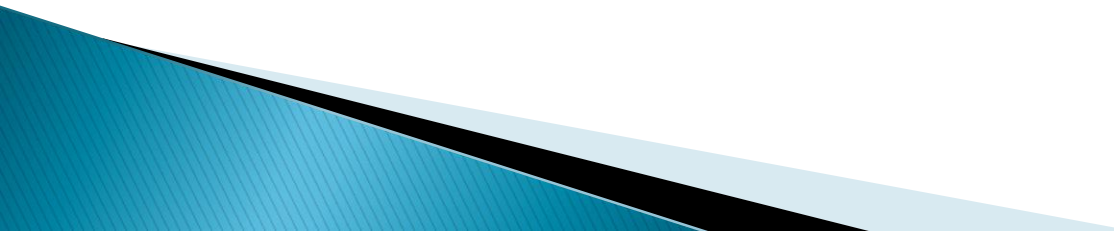
Conduction Steps

- ▶ Sound enters outer ear and ear canal
 - ▶ Sound hits eardrum and causes vibration
 - ▶ Eardrum movement causes little ear bones (in middle ear) to move
 - ▶ Innermost bone causes fluid wave in inner ear
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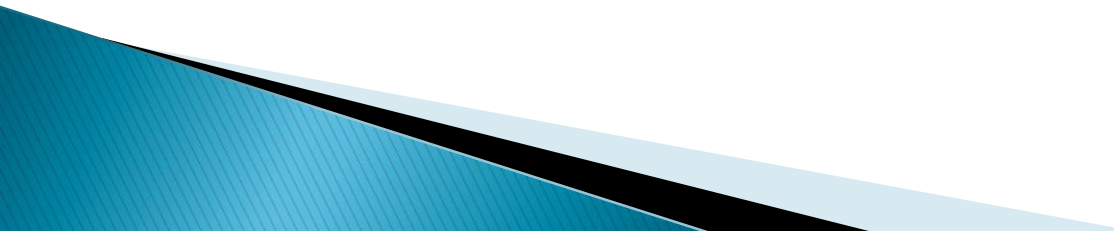
For System to work, ear canal must be open, and air must be present on both sides of eardrum.

Childhood "ear infections" cause fluid in middle ear which can persist after infection is "cured".

Let us begin in outer ear canal

- 40 -50% of newborns with DS have very small ear canals
 - If walls collapse or was plugs canal sound cannot pass
 - Canals grow with age: easier to examine by 3rd birthday
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Problems

- Wax may require removal
 - Primary care MD may not have equipment needed to examine such small ears
 - Primary care MD may "miss" seeing eardrum and middle ear fluid
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Recommendation

- ▶ Ear canals can be cleaned under office microscope to ensure exam/accurate diagnosis
- ▶ Until age 2-3, ENT exam might be necessary every 3-4 months

Let us move to middle ear
increased incidence of chronic
disease in DS

One study:

Only 5% of children had NO infections

Only 15% "avoided" ear tubes of patients
with tubes, 45% required > one set,
12.5% > 2 sets

Reasons

Higher #s of respiratory infections

- ▶ possible relationship to immature immune function

Mid face structure

- ▶ Small area behind nose where air enters ear through Eustachian tube easily obstructed
- ▶ Role of adenoids
- ▶ E tube not oriented normally so muscles which "pop" ears during swallowing fail to do so
- ▶ E tube muscles suffer from poor muscle tone seen in DS
- ▶ E tube cartilage weaker structurally

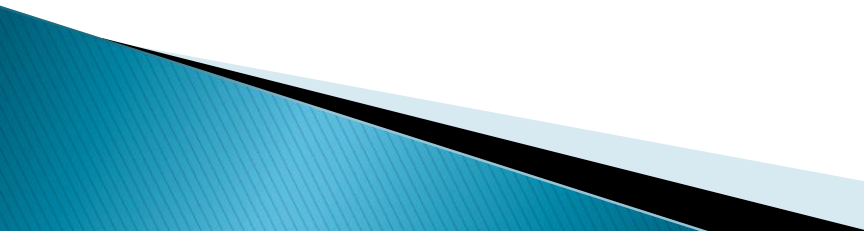
Treatment

- ▶ Antibiotics for acute infections
- ▶ Surveillance for persistent fluid
- ▶ Ear tubes

Surveillance for proper tube function

- ▶ disease returns after tubes extrude
- ▶ Evaluation and treatment of nasal disease, especially adenoids

Hearing tests

- ▶ All newborns are screened
 - ▶ DS children should have ABR (special test of hearing ability of brain) @ 3 and 6 months
 - ▶ Hearing tests every 6 months if even mild hearing loss present
 - ▶ Hearing test annually if hearing is normal
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Hearing tests 3.5 times more likely to be normal in DS children with ear tubes than age-matched children without **FUNCTIONAL** ear tubes.

Results are even better when tubes are not delayed.

Early use of hearing aids for even mild hearing loss is helpful

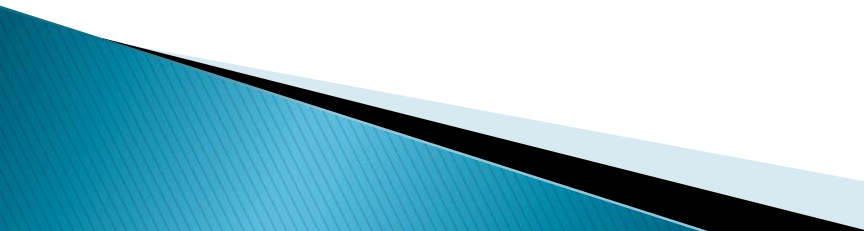
Advantages:

- ▶ Children get "used to" wearing them
- ▶ Present during critical age for language development

Disadvantages:

- ▶ Hard to fit into very small ear canals
- ▶ Tend to increase chance of wax blockage

Secondary problems of chronic ear disease

- ▶ Eardrum perforations (holes)
 - ▶ Middle ear cholesteatoma (skin cysts)
 - ▶ Middle ear bone abnormality
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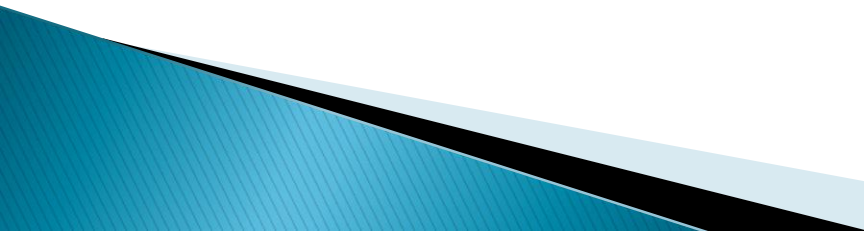
Chronic secondary problems amenable to surgery, BUT

- ▶ Higher failure rates reported.
- ▶ Even if successful in closing holes, hearing may not improve!
- ▶ Unless surgery necessary, hearing aid might be best.

Problems with hearing aids (wax or ear drainage)

- » Consider implanted hearing aid (BAHA)

Last: the inner ear

- ▶ A problem in 4-20% of DS patients
 - ▶ Some related to changes to inner ear caused by unrecognized middle ear disease
 - ▶ Not known incidence of true inner ear isolated disease
 - ▶ Genetic "deafness" incidence similar to general population, but acquired deafness higher
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Treatment

- ▶ Hearing aids
- ▶ Cochlear implants

Conclusion

- ▶ Careful follow-up
 - ▶ Prompt treatment
 - ▶ Check therapy for effectiveness
 - ▶ Problem solving
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