Identification of hearing impairment in pediatrics - the role of pediatricians and primary care providers

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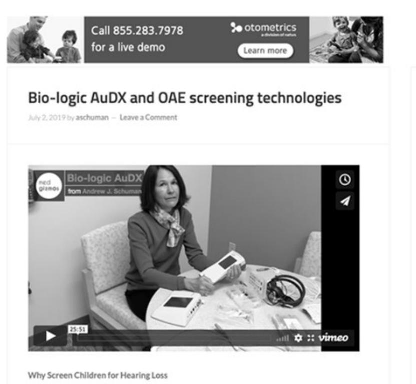
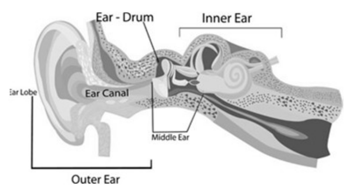
Disclosures

- This presentation is sponsored by Natus Medical/Otometrics
- Otometrics advertises on the Medgizmos web site



Learning Outcomes

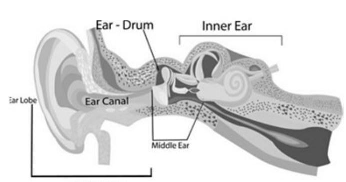
- After this course learners will be able to describe the incidence of hearing impairments in infants and children.
- After this course learners will be able to discuss how existing (and potentially future) technologies can assist with pediatric referrals for timely diagnostic evaluations leading to early intervention in cases of hearing impairments in infants and children.
- After this course learners will be able to list ways in which primary care providers can work with the audiologist and ENT specialists to assure timely diagnosis and treatment.



Identification of hearing impairment in pediatrics - the role of pediatricians and primary care providers

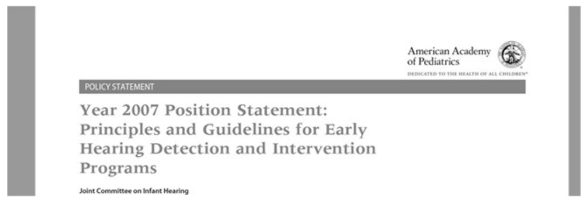
Goals and Objectives

- **Review the incidence/causes of hearing impairment in infants and children**
- **Review how physicians utilize technologies to identify pediatric patients with hearing impairment**
- **Review forthcoming technologies that may facilitate identification of hearing impairment/ear pathology**
- **Discuss how audiologists and physicians can form alliances to better identify and care for the hearing-impaired child**



Importance of detection of hearing loss in childhood

- Early Hearing Detection and Intervention (EHDI) goal is to maximize linguistic competence and literacy for children with hearing impairment.
- Without intervention affected child will fall behind in communication, cognition, reading and social-emotional development.
- Hearing off all newborns tested by 1 month, referred infants should be assessed by 3 months of age and intervention begun by 6 months of age.
- Early identification allows children with hearing loss to receive help they need during the first two years of life, a critical period for the development of speech and language skills.
- All infants should receive ongoing surveillance of communicative development/hearing throughout childhood.



American Academy of Pediatrics
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POLICY STATEMENT

Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs

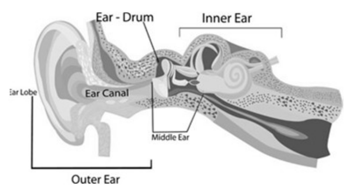
Joint Committee on Infant Hearing

Childhood hearing loss – Categories Causes

- **Congenital hearing loss – non genetic and genetic**
  - Non-genetic - Infections, prematurity, CNS injury, drug or alcohol use by mothers
  - Genetic (roughly 50%) of congenital hearing loss) – autosomal recessive, and autosomal dominant
- **Acquired hearing loss**
  - A perforated eardrum
  - Otosclerosis
  - Infections including meningitis, measles, mumps or pertussis
  - Ototoxic medications
  - Head injury
  - Exposure to loud noises
  - Untreated or frequent otitis media

Childhood hearing loss – Categories Physiology/Anatomy

- **Conductive** – problem between the external canal and cochlea. Includes vernix in newborns, cerumen in children, otitis media, aural atresia
- **Sensory** – Problem in the cochlea. Causes include ototoxic medications, infections, and genetic causes
- **Neural** – failure of the neural part of auditory pathway. Causes include structural problems (tumors), bleeding, infections, auditory neuropathy
- **Mixed** – any combination of the above



Hearing impairment in childhood - by the numbers

- 2 to 3/1000 newborns in well nursery are born with a hearing loss
- 2 to 4/1000 newborns in NICUs are born with a hearing loss
- 95% of Newborns born in the USA are screened for congenital hearing loss
- 1/75 newborns with risk factors and 1/811 without risk factors are born with a hearing -loss
- 5 to 7% of newborns are referred for follow up testing following newborn hearing screening
- 39% of babies referred by Newborn Hearing Screening Programs are lost to follow up
- In school age population as many of 10% of children have a hearing loss which may impact school performance
- The American Academy of Pediatrics recommends that children be screened for hearing loss at 4, 5, 6, 8, and 10 years of age and whenever risk factors are identified

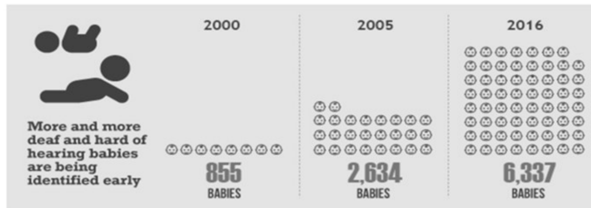
Take away: **Between the newborn period and 4 years of age, primary care providers *do not* routinely screen for hearing loss in children!**

However: **Young infants are screened for global assessment including speech-language competence at 9, 18, and 24 months!**

Annual Data Early Hearing Detection and Intervention (EHDI) Program

If you have any questions about this data please email the CDC EHDI program at: ehdi@cdc.gov.

Progress in Action



View text version of infographic

Congenital Cytomegalovirus (cCMV)

- 1/200 infants (25,000 yearly) in the USA are born with cCMV infections
- Toddlers bring home infections from daycare and infect pregnant mother
- 90% of CMV infections are asymptomatic
- Symptomatic newborns may have growth retardation, microcephaly, jaundice, seizures, rashes, petechiae
- Approximately 22% to 65% of children with symptomatic cCMV disease at birth and 6% to 23% of children with asymptomatic cCMV infection will have hearing impairment

Congenital CMV infection is diagnosed by detection of **CMV** DNA in the urine, saliva (preferred specimens), or blood, within three weeks after birth.

Connecticut, Iowa, New York, Utah, and Virginia require each newborn that fails the newborn hearing screening to be tested for congenital CMV. Illinois requires that a CMV test be offered to the parents of every child who fails the newborn hearing screening.

NATIONAL CMV FOUNDATION

March 27, 2019

DR. JOSEPH A. BOCCHINNI, JR., M.D., CHAIRMAN

Advisory Committee on Heritable Disorders in Newborns and Children
5600 Fishers Lane, Room 18W68
Rockville, MD 20857

Dear Dr. Bocchini:

The National CMV Foundation (NCMVF) is honored and excited to submit this nomination for congenital cytomegalovirus (CMV) to be considered for inclusion on the recommended universal screening panel (RUSP). We submit this nomination on behalf of all children infected with congenital CMV, their families, and a multi-disciplinary team of experts representing CMV research, clinical practice, public health, industry, and advocacy.

National Health Examination Surveys

The National Health and Nutrition Examination Survey (NHANES) is a program of studies designed to assess the health and nutritional status of adults and children in the United States. NHANES is a major program of the National Center for Health Statistics (NCHS). NCHS is part of the Centers for Disease Control and Prevention (CDC) and has the responsibility for producing vital and health statistics for the Nation.

- Between 2007-2008 and the 2009-20010 the National Health and Nutrition Examination Surveys indicate that the incidence of hearing loss > 15 dB dropped from 17.5% to 12.8%
- NHANES provide compelling evidence that hearing loss ≥25 dB affects 3% to 5% of adolescents and hearing loss >15 dB affects 15% to 20% of adolescents
- NHANES data from 1994 to 2010, indicate that hearing loss among adolescents in the United States is not increasing.

Risk factors for childhood hearing loss

- Caregiver/Pediatrician concern regarding hearing, speech, language, or developmental delay.
- Family history of permanent childhood hearing loss.
- Neonatal intensive care of longer than 5 days or any of the following, regardless of length of stay: extracorporeal membrane oxygenation; assisted ventilation; exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix); and hyperbilirubinemia that requires exchange transfusion.
- In utero infections (eg, cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis).
- Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
- Syndromes associated with hearing loss or progressive or late-onset hearing loss: neurofibromatosis; osteopetrosis; Usher syndrome; Waardenburg syndrome; Alport syndrome; Pendred syndrome; Jervell and Lange-Nielson syndrome.
- Neurodegenerative disorders (eg, Hunter syndrome) or sensory motor neuropathies (eg, Friedreich ataxia, Charcot-Marie-Tooth disease).
- Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (eg, herpes viruses, varicella) meningitis.
- Head trauma, especially basal skull/temporal bone fracture that requires hospitalization.
- Chemotherapy.
- Recurrent or persistent otitis media for at least 3 months.

Bottom Line…..

- There are many newborns who refer following newborn hearing screening **who are lost to follow up!**
- OAE testing, will not identify the 7-10% of hearing impaired newborns born with auditory neuropathy.
- A significant number of children will develop hearing impairment from otitis media.
- A significant number of children will develop hearing impairment in childhood unrelated to otitis media.
- A significant number of children with speech delay and or developmental delay will have associated hearing impairment.
- Incidence of childhood hearing impairment is 6/1000 by Kindergarten.

Bottom Line…..

Pediatricians/Primary care providers need to identify children with suspected hearing loss

AND

Refer children to Audiologists for diagnosis and treatment!

Index of Suspicion - ? Need for testing and/or audiology follow up

- Delayed Speech
- Developmental delay (Development screen at 9 months, Autism screen at 18 months, 2 years)
- Syndrome associated with hearing impairment
- Presence of ear malformations
- History of prematurity, ototoxic medication exposure, hyperbilirubinemia, etc.
- Parental concern
- Family history of hearing loss

Impediments to Hearing Screening in Pediatrics/Primary Care

- Physician burnout – overwhelmed by paperwork, EHRs
- Pediatricians see fewer patients per day than ever before – 20 per day, due to non-clinical burdens
- Need to get a lot done in short visit
- Technology can be expensive and time consuming
- Poor communication between Audiologists and Primary Care physicians

Technology for Hearing Screening in Pediatrics/Primary Care

- OAE
- Pure Tone Audiometry
- Tympanometry

Late 1990s

2019

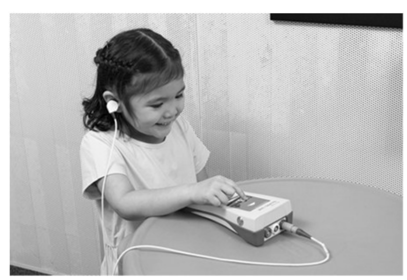
Technology for examining ears

- Otoscope (Macroview or forthcoming Wispr) – should be used with insufflation, but most pediatricians do not use
- Acoustic Otoscope – Once very popular but unavailable for well over a decade (but coming back next year!)
- TomiScope - experimental and not widely available
- Otocam video otoscope

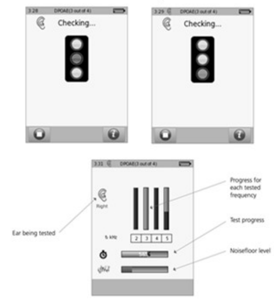
OAE

- Many pediatricians have adopted OAE
- Quick and easy
- Cartoon OAE devices entertain while testing
- Reimbursed reasonably well - $25 per test
- Good return on investment
- Some also include tympanometry and pure tone audiometry

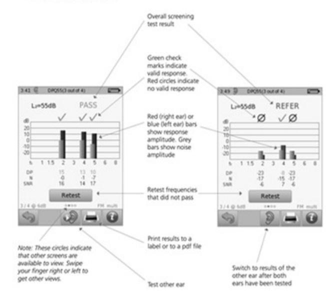
OAE/Audiometry



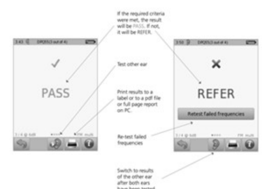
OAE/Audiometry



- Test results (bar graphs)



- Start menu

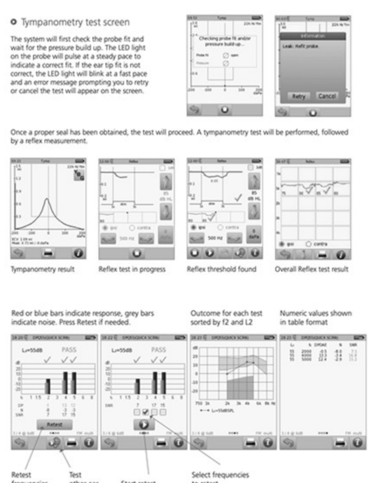


OAE/Tympanometry/Audiometry

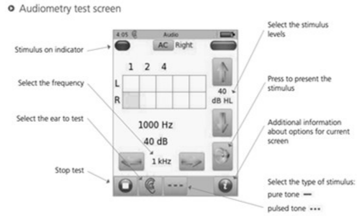
- Tympanometry test screen

The system will first check the probe fit and wait for the pressure build up. The LED light on the probe will pulse at a steady pace to indicate a correct fit. If the ear tip fit is not correct, the LED light will blink at a fast pace and an error message prompting you to retry or cancel the test will appear on the screen.

Once a proper seal has been obtained, the test will proceed. A tympanometry test will be performed, followed by a reflex measurement.

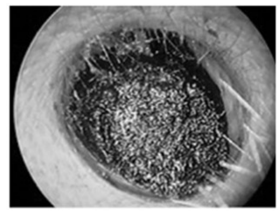


- Audiometry test screen

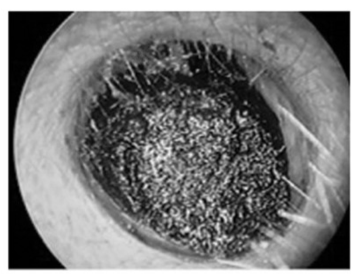


When should primary care providers perform OAE?

- Babies who refer on Newborn Hearing Screen who are "lost to follow up"
- Routinely screened for hearing loss at 4, 5, 6, 8, and 10 years of age and whenever risk factors are identified
- Prolonged/Recurrent otitis media
- Speech or developmental delay
- Poor school performance
- Parental request
- History of impacted cerumen



Cerumen Impaction

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- ☑Please note: Ear tips only, no bottle included.

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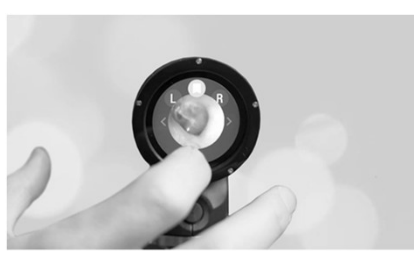
When should primary care providers perform pure tone audiometry?

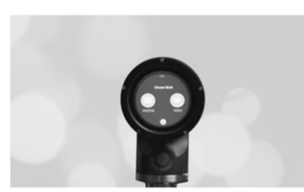
- When a child refers on OAE to establish a baseline/threshold for hearing
- A child passes OAE but one suspects auditory neuropathy

Approximately 20% of children 3- 5 years of age are unable to complete pure Tone audiometry.

Otoscope

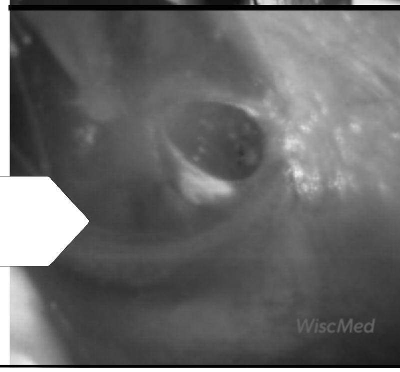
- Pneumatic otoscopy – rarely done by primary care providers
- Wispr (coming soon) provides video/image capture (lacks option for pneumatic otoscopy)



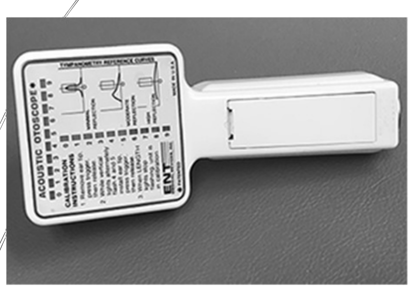


Wispr Video Otoscope

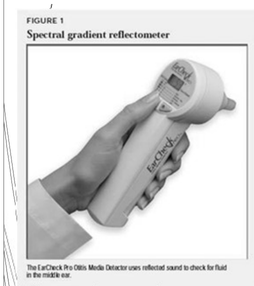
(Available November 2019)

Acoustic Otoscope



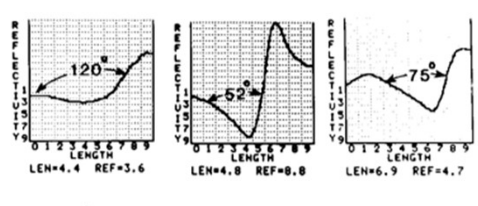
Spectral Gradient Acoustic Reflectometry (Acoustic Otoscope)



- Handheld probe containing an acoustic speaker that emits sound bursts of 44 different frequencies at 80 dB sound level.
- Using a microphone and microprocessor the device analyzes thre frequency spectra of the reflected sound and presents the output as a spectral gradient angle
- The spectral gradient angle corresponds to the probability of effusion
- No seal is required, and measurement takes seconds

Spectral gradient level	Spectral gradient angle	Risk of effusion
5	<49 degrees	High
4	49-50 degrees	Moderate to High
3	60-69 degrees	Moderate
2	70- 95 degrees	Low to Moderate
1	>95 degrees	Low

Spectral Gradient Acoustic Reflectometry (Acoustic Otoscope)



Nurses comments re: SGAR measurements vs Tympanometry

- The SGAR is easier to use because of how quickly a readout is obtained.
- If a child is crying or moving, they can still get a readout.
- You don't have to change the tip of the SGAR for the size of the external ear canal.
- The SGAR is easier to read than the tympanometer.
- The SGAR is easier to interpret for the parents.
- You don't have to get a seal with the ear canal with SGAR, as you do with a tympanometer.
- The SGAR uses a disposable tip.

SCIENCE TRANSLATIONAL MEDICINE | RESEARCH ARTICLE

EAR INFECTION

Detecting middle ear fluid using smartphones

Justin Chan[1*†], Sharat Raju[2,3*†], Rajalakshmi Nandakumar[1], Randall Bly[2,3], Shyamnath Gollakota[1†]

The presence of middle ear fluid is a key diagnostic marker for two of the most common pediatric ear diseases: acute otitis media and otitis media with effusion. We present an accessible solution that uses speakers and microphones within existing smartphones to detect middle ear fluid by assessing eardrum mobility. We conducted a clinical study on 98 patient ears at a pediatric surgical center. Using leave-one-out cross-validation to estimate performance on unseen data, we obtained an area under the curve (AUC) of 0.898 for the smartphone-based machine learning algorithm. In comparison, commercial acoustic reflectometry, which requires custom hardware, achieved an AUC of 0.776. Furthermore, we achieved 85% sensitivity and 82% specificity, comparable to published performance measures for tympanometry and pneumatic otoscopy. Similar results were obtained when testing across multiple smartphone platforms. Parents of pediatric patients ($n = 25$ ears) demonstrated similar performance to trained clinicians when using the smartphone-based system. These results demonstrate the potential for a smartphone to be a low-barrier and effective screening tool for detecting the presence of middle ear fluid.

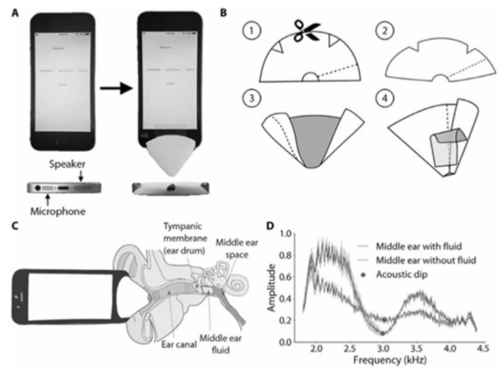
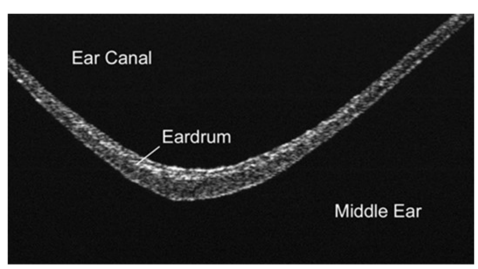


Fig. 1. Using a smartphone to detect middle ear fluid. (**A**) Location of speaker and microphone on the bottom of an iPhone 5s, without and with paper funnel attached. (**B**) Process of assembling smartphone funnel. (**C**) Proper placement of smartphone and funnel at ear canal entrance. (**D**) Raw acoustic waveform obtained when chirps are played into an ear with middle ear fluid (red) and without fluid (blue). The SD (gray) is computed across 10 chirp instances on a patient's ear.

Tomiscope

- Optical Coherence Tomography is an established non-invasive imaging technology similar to ultrasound, except that it uses near-infrared light waves instead of sound waves to provide 3-D views inside living tissue.
- Cross-sectional images of the middle ear are shown on the system's screen, alongside video of the surface of the eardrum. The physician can evaluate the revealing OCT visual images of the middle ear without giving up the more familiar otoscopic view. Images can be saved for later analysis with the click of a button.

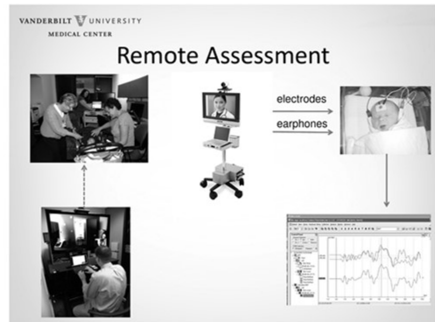


Reimbursement & Licensing

(National Telehealth Policy Resource Center, a program of the Center for Connected Health Policy; Feb. 2013)

- 44 states have some form of reimbursement for telehealth in their public program
  - Those that do not : CT, DC, IA, MA, NH, NJ, RI
- Proposed The Telehealth Promotion Act of 2012 (H.R. 6719) would require reimbursement of telehealth for any service covered face-to-face

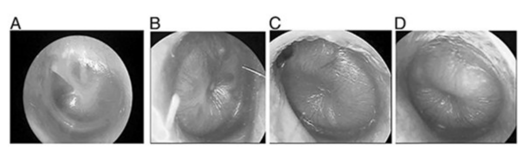
NCHAM National Center for Hearing Assessment and Management Utah State University™



Tele-audiology!

Otitis media

- Most common infection in childhood
- Accounts for most ED/physician visits in the USA
- Criteria for diagnosis has changed over the years
- Responsible for conductive hearing loss and speech delay
- Complications include mastoiditis, perforated ear drum, cholesteatoma



Otitis Media

Epidemiology of Acute Otitis Media in the Postpneumococcal Conjugate Vaccine Era

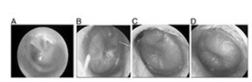
Ravinder Kaur, PhD, Matthew Morris, PhD, Michael E. Pichichero, MD

- By 1 year of age, 23% of the children experienced ≥1 episode of AOM; by 3 years of age, 60% had ≥1 episodes of AOM, and 24% had ≥3 episodes.
- increased risk of AOM associated with male sex, non-Hispanic white race, family history of recurrent AOM, day care attendance, and early occurrence of AOM

American Academy of Pediatric 2004 Policy Re: AOM diagnosis

1. Recent, usually abrupt, onset of signs and symptoms of middle-ear inflammation

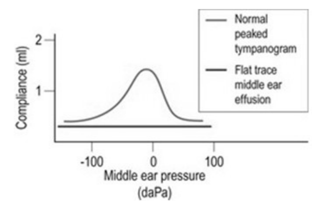
2. The presence of middle ear effusion that is indicated by any of the following:
a. Bulging of the tympanic membrane
b. Limited or absent mobility of the tympanic membrane
c. Air-fluid level behind the tympanic membrane
d. Otorrhea



3. Signs or symptoms of middle-ear inflammation as indicated by either
a. Distinct erythema of the tympanic membrane or
b. Distinct otalgia (discomfort clearly referable to the ear[s] that results in interference with or precludes normal activity or sleep)

AAP Middle Ear Effusion Policy Statement 2004

- Middle Ear Effusion diagnosed by
- Pneumatic Otoscopy
- Tympanometry



AAP AOM 2004 Treatment guidelines

TABLE 4. Criteria for Initial Antibacterial-Agent Treatment or Observation in Children With AOM

Age	Certain Diagnosis	Uncertain Diagnosis
<6 mo	Antibacterial therapy	Antibacterial therapy
6 mo to 2 y	Antibacterial therapy	Antibacterial therapy if severe illness; observation option* if nonsevere illness
≥2 y	Antibacterial therapy if severe illness; observation option* if nonsevere illness	Observation option*

AAP AOM Policy Statement 2013

- Diagnosis of AOM should be made in children who present with moderate to severe bulging of the tympanic membrane or new onset otorrhea not due to acute otitis externa
- Diagnosis of AOM may be made in children who present with mild bulging of the TM and recent onset of ear pain, or intense erythema of the TM
- Diagnosis of AOM should not be made in children who do not have middle ear effusion based on pneumatic otoscopy or tympanometry

AAP AOM Policy Statement 2013

TABLE 4 Recommendations for Initial Management for Uncomplicated AOM[a]

Age	Otorrhea With AOM[a]	Unilateral or Bilateral AOM[a] With Severe Symptoms[b]	Bilateral AOM[a] Without Otorrhea	Unilateral AOM[a] Without Otorrhea
6 mo to 2 y	Antibiotic therapy	Antibiotic therapy	Antibiotic therapy	Antibiotic therapy or additional observation
≥2 y	Antibiotic therapy	Antibiotic therapy	Antibiotic therapy or additional observation	Antibiotic therapy or additional observation[c]

[a] Applies only to children with well-documented AOM with high certainty of diagnosis (see Diagnosis section).
[b] A toxic-appearing child, persistent otalgia more than 48 h, temperature ≥39°C (102.2°F) in the past 48 h, or if there is uncertain access to follow-up after the visit.
[c] This plan of initial management provides an opportunity for shared decision-making with the child's family for those categories appropriate for additional observation. If observation is offered, a mechanism must be in place to ensure follow-up and begin antibiotics if the child worsens or fails to improve within 48 to 72 h of AOM onset.

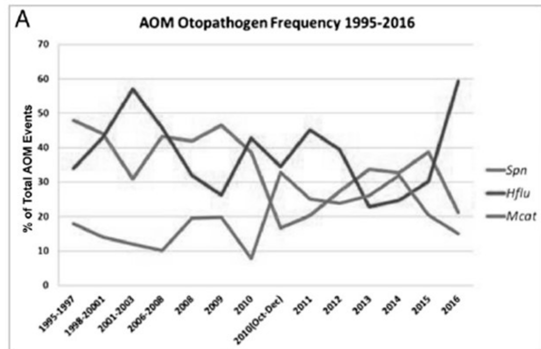
Causes/Consequences of OM

By using comprehensive and sensitive microbiologic testing, bacteria and/or viruses can be detected in the middle ear fluid in up to 96% of AOM cases (eg, 66% bacteria and viruses together, 27% bacteria alone, and 4% virus alone).

3 most common bacterial pathogens in AOM are S pneumoniae, nontypeable Haemophilus influenzae, and Moraxella catarrhalis.

Two weeks after successful antibiotic treatment of AOM, 60% to 70% of children have MEE, decreasing to 40% at 1 month and 10% to 25% at 3 months after successful antibiotic treatment.

Assurance that OME resolves is particularly important for parents of children with cognitive or developmental delays that may be affected adversely by transient hearing loss associated with MEE



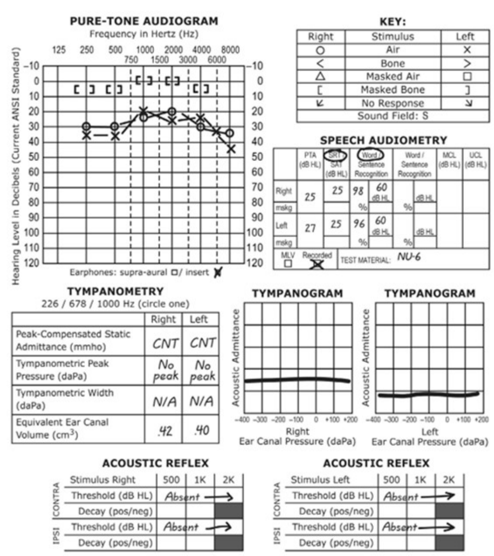
FebriDx – indicates if respiratory infection is caused by virus or bacteria

Indications for tympanostomy tubes

- Clinicians may offer tympanostomy tubes for recurrent AOM (3 episodes in 6 months or 4 episodes in 1 year, with 1 episode in the preceding 6 months).
- Long-term sequelae of tympanostomy tubes include TM structural changes including focal atrophy, tympanosclerosis, retraction pockets, and chronic perforation.

Barriers to Audiology Follow Up

- Education of Primary Care Providers re: importance of audiology evaluation and treatment
- Confusion re: Audiology reports
- Compliance on part of patients to follow up as recommended.
- Assumption that ENT referral will guarantee audiology evaluation and intervention.



Amblyopia is correctable vision loss in young children that occurs in 2.5% of the population!

20% of children referred by Dartmouth-Hitchcock Clinic to ophthalmologists for suspicion of amblyopia were evaluated and treated

Solution: Make appointment for follow up before patient leaves the primary physician office!!!!

Assuring Audiology Follow Up

- Effort on the part of audiologist to meet with and educate primary care providers
- Readable and meaningful reports
- Provision of "back-line" access to facilitate communication and appointments.

Schedule a meeting with primary care providers (and bring food!)

Questions?