A Pediatrician’s Guide to Healthy Ears

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• No financial relationships to disclose
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Objectives

• Review challenges in diagnosis and timely intervention of pediatric congenital hearing loss
• Discuss the current practice guidelines of management of infections of the ear
• Outline the risk factors of ear disease in young adults

Infancy:

CONGENITAL HEARING LOSS
Congenital Hearing Loss: A Common Problem with Profound Effects

• 1/1000 live births
• Without hearing, there is NO SPEECH…
  – Affects communication, psychosoc., and educational progress
• Lifetime costs for people with hearing loss born in 2000 will be $2.1 billion
  – medical costs, education, lost wages, unemployment

Congenital Hearing Loss: A Common Problem with Profound Effects

• Mandatory newborn screening occurs in all 50 states but THE SCREENING TEST CANNOT DIAGNOSE THE CONDITION
• Follow-up diagnostic testing by 1 month of age
• Diagnose no later than 3 months of age
• Hearing aids/therapy enrollment by 6 months
• Cochlear implantation at 12 months if no progress

2007 Position Statement: Joint Committee on Infant Hearing
Challenges Facing Kentucky

- Rural State
- Socioeconomic Barriers
- Healthcare shortage
- Educational limitations
- Geographic extremes
- Cultural diversity

Is congenital hearing loss common in KY and is diagnosis delayed?

Summary of Statewide Data

<table>
<thead>
<tr>
<th>Region of Birth</th>
<th>Appalachia</th>
<th>Non-Appalachia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Live births</td>
<td>43,036</td>
<td>110,615</td>
</tr>
<tr>
<td>Failed Newborn Screens</td>
<td>1,788</td>
<td>5,182</td>
</tr>
<tr>
<td>Permanent childhood hearing loss</td>
<td>56</td>
<td>223</td>
</tr>
<tr>
<td>Severe sensorineural hearing loss</td>
<td>28</td>
<td>93</td>
</tr>
<tr>
<td>Percentage obtaining diagnostic testing (after unilateral or bilateral failed screening test)</td>
<td>76.1%</td>
<td>62.7%</td>
</tr>
<tr>
<td>Percentage obtaining diagnostic testing (following bilateral failed screening test)</td>
<td>73.4%</td>
<td>84.6%</td>
</tr>
<tr>
<td>Children with hearing loss enrolled in early intervention program</td>
<td>51.8%</td>
<td>52%</td>
</tr>
</tbody>
</table>

1 out of 585 births
25% lost to follow-up
Age of first follow-up after birth

Weeks after birth to first ABR

- Non-Appalachian KY: 10 weeks
- Appalachian KY: 13 weeks

p = 0.33

Delayed Diagnosis in Kentucky
(Worse in Appalachia)

Weeks after birth to final diagnosis

- Non-Appalachian KY: 22 weeks
- Appalachian KY: 31 weeks

p = 0.05
Age of Diagnosis

Poor Appalachian Compliance

Average number of "no-shows"

p=0.3
Correlation of Distance and Diagnosis Timing

(\(r=0.4, p=0.002\))

Bridging the Gaps in Infant Hearing Loss

- High rate of congenital hearing loss in Kentucky
- Poor follow-up and intervention after screening
- Delay in diagnosis of hearing loss
- Distance may play a role in the delay
- Development of programs to address this
  - Telemedicine = bring care to the patients
  - Patient navigators = bringing patients to timely care
Cochlear Implants: The Bionic Ear

Surgically implanted device that electrically stimulates the cochlea to restore hearing

Cochlear Implant Components

- **External**
  - Microphone
  - Speech processor
  - Transmitter
- **Internal**
  - Receiver/Stimulator
  - Electrode array
Cochlear Implant Candidacy

Pediatric
- Severe-profound SNHL bilaterally
- Limited benefit from proper amplification
- Family commitment and realistic expectations
- No medical contraindication
- Intact auditory nerve and “acoustic areas of the CNS”

Cochlear Implant Candidates
- Short period of deafness to implantation
- Good speech and language skills
- Prelingually-deaf children (between 1-2.5)
- Pre-lingually deaf adults have the most variable and unreliable results
The Process of Implantation

- Characterization of hearing loss by an Audiologist and Otolaryngologist
- Trial of hearing aids
- Cochlear Implant Evaluation
- CAT scan or MRI (based on medical Hx and ABR results)
- Genetic counseling
- Other testing: EKG, Ophtho, Renal ultrasound
- Implantation (2-3 hours surgery)
- Activation of Implant 3-6 weeks after surgery
- Rehabilitation with an Audiologist

Congenital Cochlear Malformations

- Mondini deformity
- Michel’s Aplasia
Technique for Cochlear Implantation

Sanna M, Khrais T, Falcioni M, Russo A, Taibah A.

Technique for Cochlear Implantation
Technique for Cochlear Implantation
Implantation Risks

- Inherent risks of general anesthesia
- Infection (0.03% chance of meningitis)
- Failure of the device (1% risk)
- Injury of facial nerve
- Permanent hearing loss in the implanted ear
- Scarring

Benefits

- Improved sensitivity
- Improved speech understanding
- Can use with hearing aid on better ear
- Can perform bilateral implantation
Spoken Language Development in Children Following Cochlear Implantation

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JAMA. 2010;303(15):1498-1506 www.jama.com

Language Comp. & Expression

[Graphs showing language development in children with normal hearing and cochlear implantation]
Early Childhood:

CONGENITAL MALFORMATIONS

Congenital Malformations

http://www.mypacs.net/cases/AURAL-ATRESIA-559512.html
Early Childhood:

FOREIGN BODIES

Foreign Bodies

- Beads, Food, Bugs, Batteries, Eraser tips
- Do not manipulate the ear
- No fluid or drops
- May require anesthesia to remove

http://www.entusa.com/external_ear_cana1.htm
Childhood and Young Adulthood:

OTITIS MEDIA

Otitis Media

- Otitis Media #1 diagnosis in children <15 yrs
- 25-35 million office visits yearly
- 25% of prescriptions
- Annual cost U.S. = $5 billion
  - $406 per patient episode
  - $2,174 per patient requiring tube insertion
Current Management Protocol

• Children younger than 6 months → Antibiotics
  – Amoxicillin 80-90mg/kg/d for 10 days, or severe illness then Amoxicillin/Clavulanate
  – If allergy: cefdinir, cefuroxime, cefpodoxime, azithromycin, if severe: ceftriaxone 1-3 days
  – Watchful waiting not recommended

• Children 6 mo – 12 months of age
  – Abx in Bilateral or Severe cases (ear pain that is moderate or severe, lasts for at least 48 hours, or is accompanied by a temperature of >102.2°F

• Children older than 12 months
  – Abx if severe disease
  – Pneumococcal conjugate vaccine and annual flu shots are recommended for all children

2013 AOM Update

• Recommendations don’t apply to those with tympanostomy tubes, anatomic abnormalities such as cleft palate or Down syndrome, immune deficiencies, or cochlear implants.
• RAOM = 3 in 6 months/ 4 in prior year
• Prophylactic antibiotics not prescribed to reduce recurrences. Instead, these children may be offered tympanostomy tubes.
2013 AOM Update

- The 2004 guidelines used a three-part definition for acute otitis media: acute onset of symptoms, acute middle ear inflammation, and middle ear effusion. The 2013 update also requires middle ear effusion for diagnosis, but it now has to be based on tympanometry or pneumatic otoscopy.
- Additional diagnostic criteria include: moderate to severe bulging of the tympanic membrane or new onset of discharge not due to an infected ear canal, and mild bulging of the ear drum and onset of ear pain within 48 hours, which could be indicated by holding, tugging, rubbing of the ear for nonverbal children, or intense redness of the tympanic membrane.

Otitis Media with Effusion
Medical Treatment of OME

• Observation
• Antibiotics
  – Meta-analysis shows beneficial short-term resolution of OME
  – Unclear long-term impact
• Audiogram at 3 months with persistent effusion
• Follow-up every 6 weeks

When to Punt the Ball?

• When to refer to Oto-HNS?
  – 3 bouts AOM in 6 months
  – 4 bouts AOM in 12 months
  – Chronic OME >3mos, hearing loss, speech delay
  – Complication
  – Failed prophylaxis
  – Earlier if anatomic or immune problem
Complications of Otitis Media

- Acute mastoiditis
- Cholesteatoma
- Labyrinthitis
- Facial paralysis
- Meningitis
- Epidural/subdural abscess
- Brain abscess
- Sigmoid sinus thrombosis
- Otitic Hydrocephalus

Acute Mastoiditis with Abscess
Coalescent Mastoiditis

• Mastoiditis
  – Fluid accumulation in mastoid normal w/ AOM
  – Osteitis can occur (coalescent)
  – Abscess formation (intracranial, neck)

Myringosclerosis (Tympanosclerosis)
Tympanic Membrane Perforations

Pars Flaccida Retraction (ETD)
Total TM Retraction

Complications of Otitis Media

• Chronic suppurative otitis media (CSOM)
  – Ear drum gets pulled in creating pockets
  – Skin and debris can get caught in the pockets (cholesteatoma)
  – The pocket can expand and slowly erode surrounding bone
Cholesteatoma

- Epithelial lined pocket of squamous debris that slowly erodes bone
- CT scans used frequently to evaluate atypical symptoms (vertigo) or preoperative planning
- Erosion of HSCC

Cholesteatoma Labyrinthine Erosion

Adolescence and Young Adulthood:

OTITIS EXTERNA
Otitis Externa (Swimmer’s Ear)

• Symptoms
  – Otalgia
  – Otorrhea
  – Pruritis (chronic)

• Signs
  – Auricular Tenderness
  – Erythema/Edema of Ear Canal
  – Tympanic Membrane may or may not be involved

Treatment

• Etiology
  – Pseudomonas
  – Staphylococcus
  – Fungi

• Otic Drops +/- irrigations +/- microscopic cleaning

• “Wick” if canal cannot accept drops

• Keep water out of the ear

• Consider oral antibiotics

• Caution in diabetic patient
  – suspect malignant otitis externa (osteomyelitis of the skull base)
Otologic Trauma

- Auricle/Pinna
  - Piercing of cartilage can be problematic
  - Infection can compromise cartilage
  - Trauma to the ear can lead to permanent damage

[Image of ear piercings]
[Image of cauliflower ear]

References:
http://www.ear-piercings.engineeringdreams.com/
Otologic Trauma

• 20% of skull fractures involve the temporal bone
• 70% in 2nd - 4th decades
• Men 3-4 times more susceptible than women
• Common causes: MVA, Assault=Falls, MCA, Bicycle, gunshot wounds
• Classification based on otic capsule (CT scan is key)

Otologic Trauma

• Otic Capsule Sparing
  – 97.5% of fractures
  – Trauma to temporo-parietal region
  – Facial nerve paralysis 20%
  – CHL - Perforation of TM, EAC injury, ossicular damage (IS joint most common disruption)

Temporal Bone Trauma

- **Otic Capsule Disrupting**
  - 2.5% of fractures
  - Severe frontal or occipital trauma (deadly force)
  - Otic capsule/IAC damage = SNHL and vertigo
  - Bilateral otic capsule fx can be tx with CI

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Otologic Trauma

- **Noise induced hearing loss (NIHL)**
  - 26 million Americans affected
  - Occupational
  - Recreational
  - Simple steps to protect hearing (lower volume, limit exposure, plug the ears)
Otologic Trauma

• Noise Induced Hearing Loss (NIHL)
  – Control the TV (content and volume)
  – Buy quiet toys
  – Check out noise ratings of appliances
  – Sound-proof house (Seal cracks, carpet floors)
  – Furnish house

Otologic Trauma

• Noise induced hearing loss (NIHL)
  – 180 million iPod users
  – 100% volume for 5 min
  – 80% volume for 90 min
  – 70% volume for 4.5 hr
  – Noise cancelling headphones may help

http://www.time.com/time/health/article/0,8599,1881130,00.html?cnn=yes
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Thank You!
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