Pediatric Sudden Hearing Loss

David H. Chi, MD
April 28, 2020

Collaborative Multi-institutional Otolaryngology Residency Education Program

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World that is Completely Flipped

Chicago Midway Airport
at noon, April 17, 2020

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Case Presentation

- 8 year old boy, healthy, born full term, passed NBHS, no otologic history
- He presents with right hearing loss suspected after school screen 4 months ago
- Physical exam is normal
  - Tympanic membranes intact, no effusion

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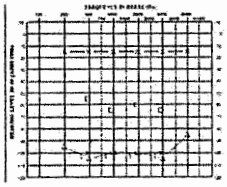
Case Presentation



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Clinical Practice Guideline: Sudden Hearing Loss (Update)

Sujana S. Chandrasekhar, MD[1,2,*], Betty S. Tsai Do, MD[3], Seth R. Schwartz, MD, MPH[4], Laura J. Bontempo, MD, MEd[5], Erynne A. Faucett, MD[1], Sandra A. Finestone, PsyD[6], Deena B. Hollingsworth, MSN, FNP-BC[7], David M. Kelley, MD[8], Steven T. Kmucha, MD, JD[9], Gul Moonis, MD[10], Gayla L. Poling, PhD, CCC-A[11], J. Kirk Roberts, MD[12], Robert J. Stachler, MD[13], Daniel M. Zeitler, MD[3], Maureen D. Corrigan[14], Lorraine C. Nnacheta, MPH, DrPH[15], and Lisa Satterfield, MS, MPH[11]

Clinical Practice Guideline 2019

- First paragraph: Guideline "focuses on SSNHL in adult patients aged > 18 years and primarily on those with idiopathic sudden sensorineural hearing loss."

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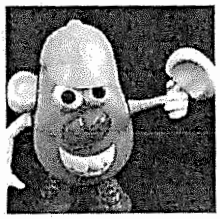
Pediatric SSNHL

- How do kids present?
- What is the evidence for treatment?
- What are unique etiologies of pediatric SSNHL?
- What are the challenges in the management of pediatric SSNHL?



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Sudden SNHL

- 30 dB hearing loss over three continuous frequencies over 3 days
- Annual incidence is 5 to 20 cases per 100,000
  - –3.5-10% occur in <18 years old
- Bilateral in only 1-2% of cases
- 32-65% recovery rate

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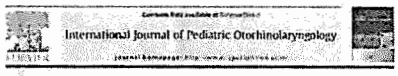
International Journal of Pediatric Otorhinolaryngology

Pediatric sudden sensorineural hearing loss: Etiology, diagnosis and treatment in 20 children*

Kavita Dedhia *, David H. Chi *

- 20 pts from 2000 to 2013
- Average age was 11 years, 3 months
- Patients presented an average of 122 days from onset
- Tinnitus was the most common associated symptom 55%
  - Otalgia 25%
  - Vertigo 20%
- 40% had bilateral hearing loss

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Pediatric sudden sensorineural hearing loss: Etiology, diagnosis and treatment in 20 children*

Kavita Dedhia *, David H. Chi *

- No patients had routine laboratory tests
- 90% had radiographic imaging
  - –Scans were abnormal in 40% (8 pts)
    - Enlarged vestibular aqueduct in 4 pts
- Eight pts (40%) were treated with oral steroids
  - 50% showed improvement on postop audiogram

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Pediatric sudden sensorineural hearing loss: Etiology, diagnosis and treatment in 20 children*

Kavita Dedhia *, David H. Chi *

Punchline:

- Presentation often delayed
  - –70% presented beyond 2 weeks
- Steroids, when used, were primarily systemic for kids
- Pediatric SSNHL has different etiologies than adults

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Multivariate Analysis of Prognostic Factors for Idiopathic Sudden Sensorineural Hearing Loss in Children

Jae Ho Chung, MD; Seok Hyun Cho, MD; Jin Hyeok Jeong, MD; Chul Won Park, MD; Seung Hwan Lee, MD

- Retrospective review over 6 years
  - 37 children <18 years old; comparison group of 276 patient ≥19 year old
  - SSNHL with any causative agent were excluded
    - Viral infection with positive serology
    - Vestibular schwannoma
    - Congenital anomaly
  - Patients with delay >2 weeks prior to treatment were excluded
  - All patients were admitted for 6 days for oral prednisolone(1mg/kg/day) for 7 days then tapered for 7 days as well as IV dextran

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Multivariate Analysis of Prognostic Factors for Idiopathic Sudden Sensorineural Hearing Loss in Children

Jae Ho Chung, MD; Seok Hyun Cho, MD; Jin Hyeok Jeong, MD; Chul Won Park, MD; Seung Hwan Lee, MD

- Average age =14.3 years
  - –Average PTA 61.9 dB
- Pediatric ISSNHL represented 6.6% of the total cases
- Recovery
  - –Complete: in **46.6%** of peds, **30.8%** of adults
  - –Overall recovery (complete + partial) was 57.4% vs 47.2%

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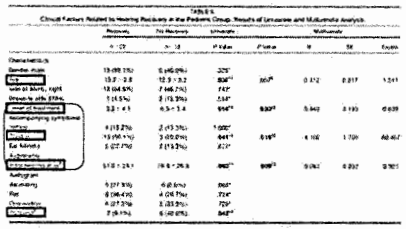

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Sudden Sensorineural Hearing Loss in Children, Management and Outcomes: A meta-analysis

- 13 studies provided a total of 605 patients (total 289 ears)
- 24.6% abnormal imaging
- Improvement:
  - 51.5% no improvement
  - 25% partial improvement
  - 24.5% complete resolution

Positive Prognostic Factors	Negative Prognostic Factors
Unilateral hearing loss	Profound hearing loss
Tinnitus	Delay > 6 days
Ascending audiogram	
Age > 12 years	

Wood, Shaffer, Kitsko. Chi. Presented, in part, at ASPO National Meeting, 2017

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Differential Diagnosis of "Sudden" loss in children

Apparent	Not apparent
Infection • Meningitis • Labyrinthitis • CMV- symptomatic • Lyme disease	**Infection** • CMV
Trauma • Temporal Bone fracture • Traumatic perilymphatic fistula	**Genetic** • Late onset or Progressive hearing loss
Ototoxins • Chemotherapy • Aminoglycoside	**Structural** • EVA • Mondini
Vascular • Sickle cell disease	**Idiopathic**

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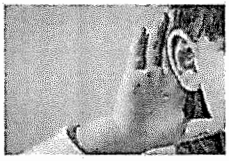
Pediatric Sudden Sensorineural Hearing Loss

not Sudden Onset, but Suddenly Identified



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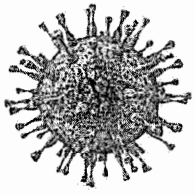
What is Cytomegalovirus?

- Another dreaded virus that starts with C
- A Herpesvirus
- 0.5 to 1% of all newborns
- 30,000 congenital CMV infections
- 140 deaths
- Most common intrauterine infection



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Transmission Mother to Fetus

- **Primary** CMV infection
  - Seronegative mothers who develop infection during pregnancy
    - 30-40% risk with primary infection
- **Secondary** CMV infection:
  - Can transmit in seropositive mothers via reactivation of latent virus or reinfection with new strain
    - 2% risk with 2ndary infection

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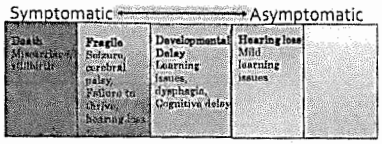
Two types of infections

- Symptomatic (evident at birth) — 5%-10%
  Sensorineural hearing loss (50%)
- Asymptomatic (silent at birth) — 90%-95%
  Sensorineural hearing loss (5 - 15%)



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CMV and Hearing Loss

- Onset between birth and ~5 years of age
- No predictable method of which children get hearing loss
- Unilateral or bilateral
- Mild to profound
- May fluctuate but usually progressive
- Approximately 33-50% of hearing loss due to congenital CMV may be late onset

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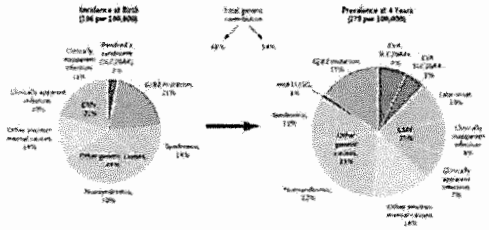
CMV: 20-25% of early childhood SNHL in children



Morton, NEJM, 2006

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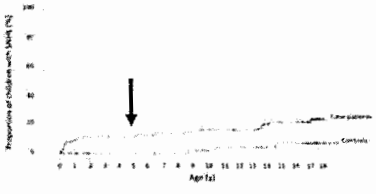
Late-onset Hearing loss in 15% of asymptomatic cCMV children

- At 5 years, 15% of children with asymptomatic cCMV had hearing loss
- Of these 2% had bilateral severe to profound SNHL



Lanzieri, *Pediatrics*, 2017

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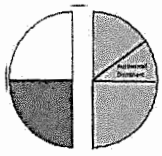
What about Genetics?



- Autosomal recessive 60-80%
- Autosomal dominant 15-20%
- X-linked 2-3%
- mitochondrial 0.5%

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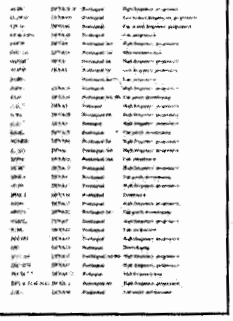
What about Genetics?

- **DFNA**: Autosomal dominant
  - Most cause post-lingual SNHL
- DNFA16
  - Fukushima, Smith RJH et al, 1999
  - Progressive loss
  - Responsive to steroids



Shearer AE, Hildebrand MS, Smith RJH. Gene Reviews, 2017

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Structural causes of pediatric sudden hearing loss

- EVA
  - >0.9mm at midpoint
  - >1.9mm at operculum
- Valvasorri and Clemis
  - LVA if greater than 1.5 mm at midpoint

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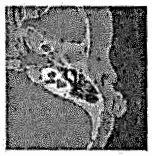
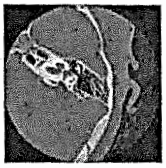
Incomplete Partition

- Type 1
  - Cochlea: cystic, absent modiolus
  - Vestibule: cystic
  - VA: not enlarged
- Type II:
  - Cochlea: Cystic apex
  - Vestibule: normal or dilated
  - Vestibular aqueduct: large





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Controversy about Head Trauma and Progressive SNHL with EVA

- Historically, there were concerns of head trauma and SNHL
- Recommend: avoiding contact sports, scuba diving and/or wear helmets

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Progressive Hearing Loss and Head Trauma in Enlarged Vestibular Aqueduct: A Systematic Review and Meta-analysis

A. Sean Alemi, MD[1], and Dylan K. Chan, MD, PhD[1,2]

- *Otolaryngol-Head Neck Surg*, 2015
- Systematic Review and Meta-analysis: 1115 EVAs (23 articles)
  - Overall, progression occurred in 40%
  - Only 12% had progression associated with trauma
  - Association with trauma not statistically significant

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Case Presentation

- No labs were obtained, based on lack of history and late presentation
- No steroid treatment due to late presentation
- Imaging was obtained

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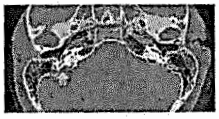
Case Presentation



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Summary

- Sudden onset vs. Sudden diagnosis
- Evaluate for etiology
  - genetic, CMV
  - May be associated with an anatomical-pericochlear cause more than retrocochlear mass
- Steroids has a role
- Challenges of Pediatric Sudden Hearing Loss
  - Children may not complain
  - Present later than 2 weeks
  - IT option not practical in kids

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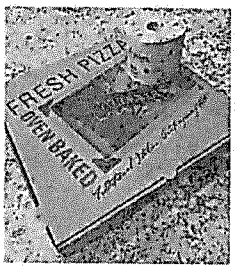
- Thank you everyone!
- Questions?



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