HEARING LOSS IN CHILDREN: THE EHDI PROGRAM AND BEYOND

Alaska Nurse Practitioner's Meeting
September 18, 2014
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DISCLOSURE OF COMMERCIAL INTERESTS

Neither I nor any members of my immediate family have a financial interest (currently or within the past 12 months) with any proprietary entity producing health care goods or services related to the content of this CME activity.

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31st Annual Alaska Nurse Practitioner's Conference

LEARNING OBJECTIVES

1. Learn about the principles of newborn screening in general and the EHDI program in specific.
2. Understand the PCPs responsibilities (in the EHDI program) to their patients.
3. Congenital versus Delayed/Progressive Forms of Hearing Loss
4. Cytomegalovirus- Its role in childhood hearing loss
5. Mild and Unilateral Hearing Loss – "It's Not Nothing."

- "Deafness separates people from people"
  - Helen Keller

AAP TASK FORCE ON NEWBORN INFANT HEARING

- Endorses implementation of universal newborn hearing screening
- Defines standards for:
  - Screening
  - Tracking & Follow-up
  - Identification & Intervention
  - Program Evaluation
- Encourages AAP Chapters to provide leadership in developing statewide programs

PREVIOUS HIGH RISK SCREENING:

- 50% of newborn hearing loss went undetected (many full term infants with no risk factors)
- Until recently, average age of identification of hearing loss was 2.5 years; even later for mild and moderate hearing loss

PREREQUISITES FOR A POPULATION SCREENING PROGRAM

Condition sufficiently frequent in screened population - YES

Condition serious or fatal without intervention-YES

Condition must be treatable or preventable-YES

Effective follow-up program possible-YES

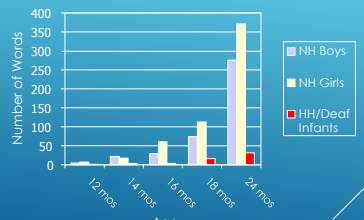
INCIDENCE OF ROUTINELY SCREENED NEWBORN CONDITIONS

Per 10000	Condition
	Metabolic Screening (45)
2.4	Amino Acid Disorders (11)
0.7*	Fatty Acid and Organic Acid Disorders (10/15)
4/0.6	Endocrine (2 – CHT, CAH)
*	Hemoglobinopathies (4)
3-4	Cystic Fibrosis
0.25	Other (galactosemia, biotinidase deficiency)
0.3	Cyanotic Heart Disease
30	Congenital Hearing Loss

WHY IS EARLY IDENTIFICATION OF HEARING LOSS IMPORTANT?

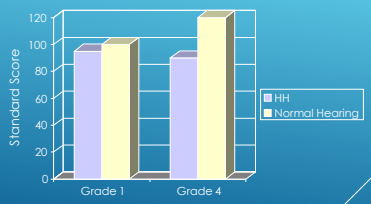
- Hearing loss is the most common birth defect
- Previous methods for detecting hearing loss have been ineffective
- Undetected hearing loss can delay speech, language, social & academic development

VOCABULARY DEVELOPMENT IN INFANTS



Mayne, et al, 2000 & MCDI norms

READING COMPREHENSION IN CHILDREN WITH MILD-MODERATE LOSS



Blair, et al, 1988

DEGREES OF HEARING LOSS

Profound (over 90 dB)

- No response to speech.
- Must use visual cues to communicate.
- May respond to vibration from sound

Severe (71dB-90dB)

- Speech heard only when loud and at close range
- Difficulty hearing and understanding always
- May show awareness, not recognition to sound (clap)

DEGREES OF HEARING LOSS

Moderately severe (56-70dB)
- Misses nearly all speech signal necessary to develop normal speech and language
- May respond to loud environmental sounds

Moderate (41-55dB)
- Misses 50-75% of speech signal
- May do well close up and face to face but have difficulties across the room
- Difficulty discriminating consonants

DEGREES OF HEARING LOSS

Mild (26-40dB)
- Misses 25-40% of speech signal
- Trouble understanding (Hears sound, but not all info)
- More trouble at distance or in noisy environment

Slight (16-25dB)
- Difficulty with quiet speech or at distance
- Background noise interferes
- May miss 10% of signal at more than 3 feet

WHAT DOES IT SOUND LIKE TO HAVE A HEARING LOSS?

Severe hearing loss

Moderate hearing loss

Mild hearing loss

Normal hearing

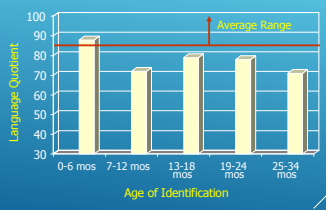
WHY IS EARLY IDENTIFICATION OF HEARING LOSS IMPORTANT?

- Early identification and intervention can make a difference

YOSHINAGA-ITANO LANDMARK STUDIES

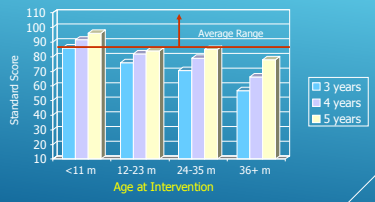
- **Children who are diagnosed with hearing loss before 6 month *and* receive appropriate intervention demonstrate:**
  - **Better receptive and expressive language**
  - **Better expressive vocabulary**
  - **Better personal-social skills**
  - ***and***
  - ***Maintain those skills within normal limits***

EFFECTS OF AGE OF IDENTIFICATION ON LANGUAGE DEVELOPMENT



Yoshinaga-Itano, et al, 1998

RECEPTIVE LANGUAGE OVER TIME BY AGE OF INTERVENTION



Moeller, 2001

3 YEAR OLD WITH MILD-MODERATE HEARING LOSS – DIAGNOSED AT AGE 3

3 YEAR OLD WITH MODERATE-SEVERE LOSS: DIAGNOSED AT BIRTH

5 YEAR OLD – MILD-MODERATE LOSS DIAGNOSED AGE 3 POST-INTERVENTION

SCREENING TECHNIQUES

- Otoacoustic emissions (OAE)
- Auditory brainstem response (ABR)
- Two stage screening (OAE + ABR)

OTOACOUSTIC EMISSIONS

- Sounds are presented to the ear canal and a small microphone measures the response in the ear canal
- Average test time is 5-15 minutes/baby

AUTOMATED AUDITORY BRAINSTEM RESPONSE (AABR)

- Sounds are presented and surface electrodes measure brainstem activity
- Average test time 20 min/baby

AUTOMATED AUDITORY BRAINSTEM RESPONSE

WHAT IF A BABY FAILS UNHS?

- Failure rates range from 1.5-5.0% in good screening programs
- The incidence of hearing loss is 3/1000 babies. Most babies who fail the initial screening will actually have normal hearing
- Babies should have re-screen within 2 weeks of birth to expedite referral for diagnostic testing.

COUNSELING AFTER NHS REFER

DIAGNOSTIC TESTING

- Referral for follow-up testing

  After second refer (fail), patient should be considered as having a *developmental emergency*.

  Early referral for diagnostic testing should be the rule.

  Diagnostic BAERs are labor intensive for audiologist and experienced pediatric-trained audiologist need to be availed ASAP. Referral at 3 months or later usually require sedation, but younger infants can often get non-sedated BAERs which lower cost/risk.

  Delays also increase risk of middle ear infections/fluid that can complicate diagnosis

COUNSELING AFTER DIAGNOSIS

WHAT IS A MEDICAL HOME?

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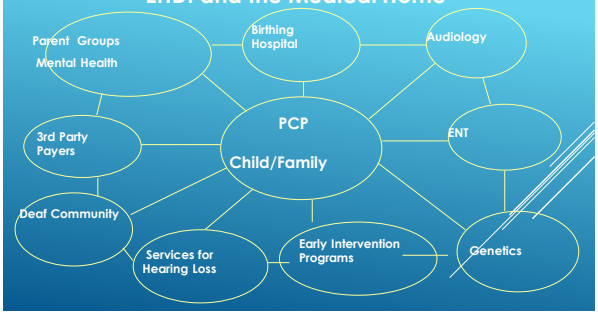
A philosophy of care emphasizing the primary care practitioner which is:

- Accessible
- Family-centered
- Comprehensive
- Continuous
- Coordinated
- Compassionate
- Culturally effective

PCP'S ROLE AFTER DIAGNOSIS

- History – prenatal, neonatal, family
- ENT referral – CT/MRI, ?middle ear disease
- Ophthalmology referral
- Evaluate for possible syndrome. Genetics referral
- Referral to Early Infant Program
- Provide support via other parents of children with hearing loss (ie Stone Soup group)

EHDI and the Medical Home



ROLES OF THE MEDICAL HOME

- Understand testing results at screening and diagnostic phases and their implications for follow up
- Assure follow-up screening; refer for diagnostic and medical specialty evaluations
- Support family in understanding severity & type of hearing loss
- Discuss Communication Options

ROLES OF THE MEDICAL HOME

- Offer partnership with parents to identify and develop a plan of health and habilitative care
- Provide ongoing surveillance, especially for infants with risk indicators for progressive or late onset hearing loss, and for infants with unilateral hearing loss

MEDICAL HOMES AND FAMILY FUNCTIONING

With Medical Homes, families report less difficulties with :

- Parental Coping
- Parental Aggravation
- Child care/Workplace
- Missed school days

Arauz Bourdreau et al, Academic Pediatrics, 2012

AMPLIFICATION

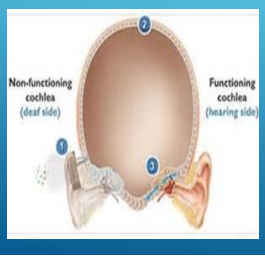
- Hearing aids can be fitted as young as 1 month of age

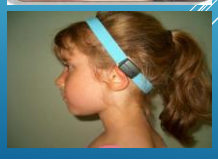
Management of Infant Hearing Loss: Cochlear Implants

CI CANDIDACY CRITERIA

- 3-6 month trial with hearing aids; lack of benefit
- Age of implantation: 12 mos. for profound loss (90dB+); >18 mos. for severe to profound loss (+70dB)
- No medical contraindications
- Education focusing on auditory-oral skills
- Family factors (motivation, expectations)

BONE ANCHORED HEARING APPARATUS (BAHA)





CHOICES IN COMMUNICATION

- It is about COMMUNICATION, not methodology
- Choosing methods or devices is a process that is flexible, ongoing & changeable
- Early ID provides a window of time for exploration
- "Goodness of Fit", family goals, family resource allocation, potential "risks" & effectiveness matter

COMMUNICATION OPTIONS

Bi-lingual/Bi-cultural – ASL/English
- No speech skills, amplification not used

Aural/Oral – Cued speech, Speech-reading
- Stresses speech, listening, technology

Total Communication – Signing Exact English-SEE
- Pidgen Signed English-PSE
- Simultaneous Communication-SimCom
- Interacts with Deaf and Hearing world

ALASKA'S EHDI PROGRAM

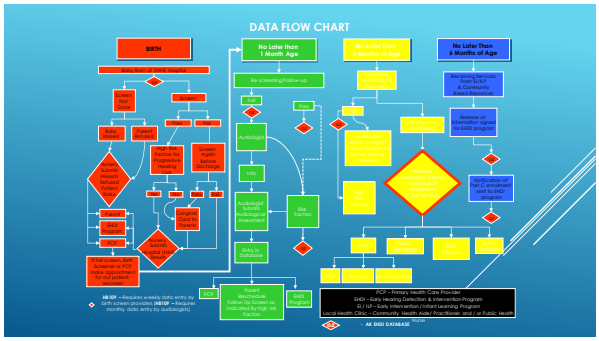
SOA, Division of Public Health,
Section of Women's Children's & Family Health

2000 – Voluntary program – paper reporting

2005 – Database goes "live"

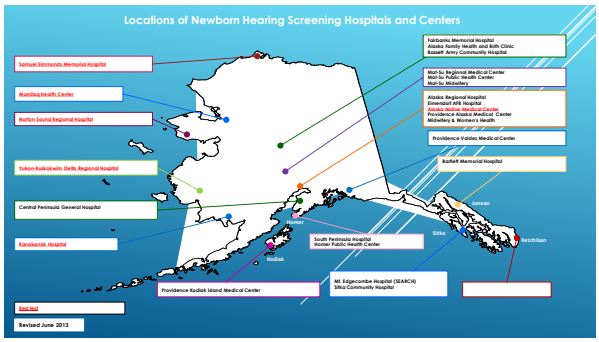
2006 – State law is passed mandating Newborn Hearing Screening, Tracking and Intervention Program

January 2008 – law and regulations take effect



CURRENT STATUS OF SCREENING

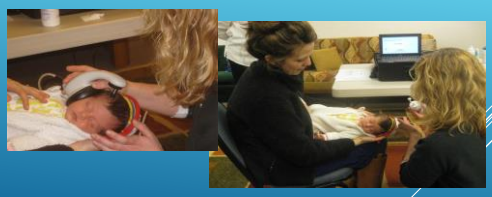
- Screening equipment in all **20 birthing hospitals** in Alaska
- Screening equipment currently in 3 **Public Health Centers and 4 midwifery centers**
- **2012 data – 96%** of all Alaskan newborns were screened –
  - 99% of all infants born in hospitals screened!



Diagnosed Hearing Loss

- Alaska births – approximately 11,300
  - Incidence of diagnosed hearing loss
    1-3 per 1,000 diagnosed through newborn hearing screening (National prevalence 2011: 1.5 per 1,000)
- Infant diagnosed with hearing loss:
  2012-- 22 infants
  2011 – 18 infants (prevalence 1.6)
  2010 – 20 infants
  2009 – 19 infants

OUT OF HOSPITAL SCREENING



INFANTS BORN OUT OF HOSPITAL

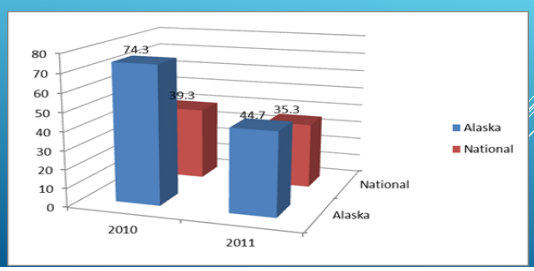
- 6 percent of all Alaskan newborns are born out-of-hospital (OOH)
- Statute addresses notification by Bureau of Vital Statistics (monthly) to EHDI Program of infants born out of hospital.
- EHDI Program to notify parents of the merits of newborn hearing screening.
- Names checked against database and letters generated.
- Letter sent to Parents – screening location card and milestone checklist

Screening rate for OOH: 38% in 2010
72% in 2013

FOLLOW-UP FOR INFANTS WITH A MISSED OR FAILED SCREENING

- Reports generated from EHDI database of infants with a missed or failed hearing screening – name, birth date, screening result, parents name
- Monthly "fax back" system to communicate with birth screen providers as to status of follow-up screens
- Parent Contacts
- Referrals to Audiology
- Communication with Infant's Medical Home

IMPROVEMENT IN LTF/LTD



Challenges/Opportunities

YEAR	TOTAL	MILITARY	ANMC	Remote Regional Hospitals	PRIVATE	OTHER
2010	150	76 - (51%)	18 - (12%)	18 - (11%)	32 - 21%)	5 - (2%)
2011	72	12 - (17%)	12 - (17%)	18 - (25%)	27 - (38%)	3 - (4%)

Parent Involvement

Parent Involvement

Parent Navigation

- Stone Soup Group Partnership
  - Resources
  - Parent to Parent

Resources

- Communicate with Your Child

http://www.communicatewithyourchild.org/brochures.html

Alaska specific insert

- Decision Guide to Communication Choices

http://www.cdc.gov/ncbddd/hearingloss/freematerials/Decision-Guide.pdf

- Hearing Aid Loaner Program

New EHDI Resources

- Coming Soon from CDC:
  - **"What Else Checklist"** – Considerations for audiologists before, during and after audiology appointment. Developed by the Parent to Parent Committee from the parent perspective
  - EHDI Packet for Ear, Nose & Throat Doctors (Otolaryngologists)
  - **"Just in Time"** materials (revised) for primary care providers

HEARING LOSS: INCIDENCE

- **Congenital: 3/1000 live births**
- **Late onset/acquired: Less studied-estimated 12-19/1000 by age 19**

RISK FACTORS FOR HEARING LOSS

Family History of Hearing Loss *	Caregiver Concerns *
Intra-Uterine Infections *	Syndromes assoc. with HL * Physical findings assoc. with above
Neurodegenerative Diseases *	Hyperbilirubinemia with transfusion
Post-natal Infections *	ECMO *
Assisted Ventilation	Head Trauma *
Oto-toxic meds, loop diuretics	NICU>5 days
Cranio-facial anomalies	Chemotherapy *
	* *Great concern of late onset/progressive hearing loss*

AT RISK FOR CONGENITAL HEARING LOSS

- More than 48 hours in NICU
- Family history of congenital or delayed hearing loss
- Stigmata associated with SNHL or conductive HL
- Cranio-facial anomalies especially with external ear anomalies
- Intra-uterine/gestational infection (CMV, Rubella, Syphilis, Herpes, Toxoplasmosis)

GENETIC CAUSES

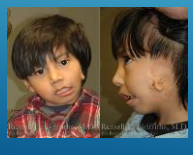
Single gene — Connexin 26, 70genes

Gene + *environment* (Epigenetics) — Mitochondrial + ototoxic

Multiple genes

PHYSICAL MARKERS FOR SYNDROMES ASSOCIATED WITH HEARING LOSS - HEAD

Waardenburg syndrome
- assymmetric facies
- eye abnormalities
- ear abnormalities
- Vertebral changes

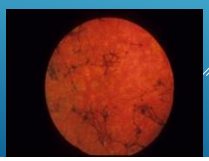


Goldenhar syndrome
- **white forelock**
- **Iris heterochromia**
- **midface anomalies**

PHYSICAL MARKERS FOR SYNDROMES ASSOCIATED WITH HEARING LOSS - HEAD

Stickler syndrome
- Flattened facial profile
- Cleft palate
- Ocular disorders
- conductive and high frequency, progressive HL

Usher syndrome – 3 types
- Retinitis pigmentosa
- Moderate-profound CHL
- Vestibular problems



PHYSICAL MARKERS FOR SYNDROMES ASSOCIATED WITH HEARING LOSS

Thyroid
- Pendred syndrome (5% of congenital HL)
  - Thyroid goiter (euthyroid) – may be undetected
  - Enlarged vestibular aqueduct (Mondini malf.)
    - Trauma may cause deafness
  - Variable hearing loss

Skeletal
- Osteogenesis imperfecta
  - Susceptibility to bone fracture
  - Blue sclerae
  - Wide variability of expression

PHYSICAL MARKERS FOR SYNDROMES ASSOCIATED WITH HEARING LOSS-HEART

CHARGE syndrome –
- coloboma,
- Heart abnormalities,
- atresia choanae,
- retarded growth and devel., genital hypoplasia/undecended testicles,
- ear anomalies/deafness

Jervell and Lange-Nielson syndrome

Prolonged QT syndrome (SIDS)

Profound SNHL

PHYSICAL MARKERS FOR SYNDROMES ASSOCIATED WITH HEARING LOSS - KIDNEY

Most Common Syndrome Association
- Timing of prenatal development similar
  - Infections/toxins may affect both
- Similarities of structure and function
  - Membranes maintain chemical balance
  - Molecular structures damaged by drugs

PHYSICAL MARKERS FOR SYNDROMES ASSOCIATED WITH HEARING LOSS - KIDNEY

Branchio-oto-renal syndrome
- Branchial cleft fistulas/cysts
- Auricular anomalies, pre-auricular pits
- Varying degrees of renal anomalies

Alport syndrome
- Nephritis (progressive), Hematuria
- Progressive SNHL
- x-linked

PHYSICAL MARKERS FOR SYNDROMES ASSOCIATED WITH HEARING LOSS - KIDNEY

Muckle - Wells Syndrome
- Urticaria
- Recurrent fever, joint pain
- Amyloidosis of kidney
- Progressive SNHL

Renal Tubular Acidosis –autosomal recessive
- 2 types – early and late onset
- Renal stones
- Early profound or progressive SNHL

PHYSICAL MARKERS FOR SYNDROMES ASSOCIATED WITH HEARING LOSS - KIDNEY

Charcot-Marie-Tooth syndrome
- Progressive neuropathy - lower extremities
- Nephritis
- Progressive SNHL
- Presents in adolescence

Epstein syndrome
- Macrothrombocytopenia
- Nephritis
- Deafness

Auditory and Visual Defects Resulting from Symptomatic and Subclinical Congenital Cytomegaloviral and *Toxoplasma* Infections

Sergio Stagno, M.D., David W. Reynolds, M.D., Catherine S. Amos, O.D., Arthur J. Dahle, Ph.D., Faye P. McCollister, M.A., Indra Mohindra, O.D., Rufino Ermocilla, M.D., and Charles A. Alford, M.D.

From the Divisions of Infectious Disease, Speech and Hearing, and Vision Function (Center for Developmental and Learning Disorders), Departments of Pediatrics and Pathology, Medical School, University of Alabama in Birmingham, and The Children's Hospital, Birmingham, Alabama

ABSTRACT. Sensorineural hearing loss was present in ten of 59 (17%) patients with congenital cytomegalovirus (CMV) infection (three of eight born with symptomatic and seven of 51 born with subclinical infection). The defect was bilateral in eight, moderate to profound in eight, and of progressive nature in two. Hearing loss did not occur in 21 patients with natal CMV infection nor in seven of 12 patients with congenital toxoplasmosis. Histopathologic and immunofluorescent studies of the inner ear in two of three neonates who died with severe infection revealed that viral antigens were widely distributed in cochlear structures.

Eye pathology was associated only with congenital *Toxo-*

common abnormality resulting from congenitally acquired rubella,[1] and it is the one most easily overlooked in infancy.[2-4] In Eichenwald's follow-up studies,[4] auditory impairment occurred in approximately 15% (15 of 101) of children born with congenital toxoplasmosis, otherwise symptomatic; and 20% (four of 20) of those with congenital cytomegaloviral (CMV) infection had varying degrees of hearing loss in the McCracken et al. series.[5]

PATIENT WITH HEARING LOSS AND CMV

- J.O. – DOB 10/10 at ANMC(?)
- Referred NHS
- BAER – Bilateral profound SNHL
- Urine positive for CMV
- No apparent signs of Symptomatic Congenital CMV
- No intra-cranial calcifications
- Ht and Wt 70-80%, HC 8%

CYTOMEGALOVIRUS

One of the Herpesviruses (HHV5)
- Betaherpesvirinae (like HHV 6, 7)

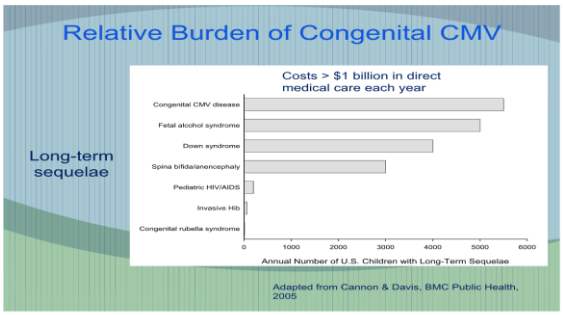
Sero-prevalence increases with age and varies widely due to location, socio-economic status, ethnicity

Excreted in all bodily fluids

Infections usually asymptomatic
- May cause mononucleosis in immunocompetent
- Serious disease in immunoincompetent

Cost of treatment of CMV and complications –
- $1-2 billion/year

Relative Burden of Congenital CMV



HERPESVIRIDAE - HUMAN

Type	Synonym	Sub family	Primary Target Cell	Site of Latency	Means of Spread
HHV-1	HSV1	alpha	mucoepithelial	neuron	Close contact (STD)
HHV-2	HSV2	alpha	mucoepithelial	neuron	Close contact (STD)
HHV-3	VZV	alpha	mucoepithelial	neuron	Resp and Close Contact (STD)
HHV-4	EBV	gamma	B cells Epithelial cells	B cell	Close contact congenital ,*
HHV-5	CMV	beta	Monos, lymphs, Epithelial cells	Mono, lymphs and ?	Saliva *
HHV-6	Roseola	beta	T cells and ?	T cells and ?	Resp. and close contact
HHV-7	Roseola	beta	T cells and ?	T cells and ?	?
HHV-8	KSHV	gamma	Lymphs and others	B cell	Close contact, Saliva, sexual

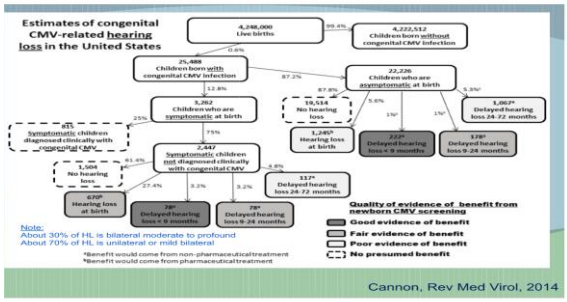
CHARACTERISTICS OF SYMPTOMATIC CONGENITAL CMV

- Hepatosplenomegaly
- Microcephaly
- Thrombocytopenia
- Petechiae (Blueberry muffin)
- Jaundice with conjugated hyperbilirubinemia
- Chorioretinitis

HEARING LOSS AND CMV (UAB)

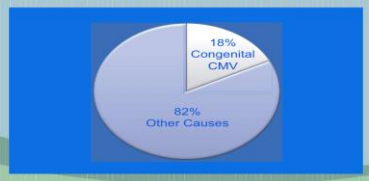
	Symptomatic	Asymptomatic
Subjects	209	651
Total Hearing Loss	85 (40.7%)	48 (7.4%)
Unilateral Hearing Loss	28 (32.9%)	25 (52.1%)
Bilateral Hearing Loss	57 (67.1%)	23 (47.9%)
Delayed Onset	23 (27.1%)	18 (37.5%)
Age Range	6-197 months	24-182 months
Progressive	46 (54.1%)	26 (54.2%)
Age Range	2-209 months	3-186 months
Sex, Male	59.1%	48.6%
Race, Black	50.0%	82.9%

Estimates of congenital CMV-related hearing loss in the United States



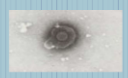
Cannon, Rev Med Virol, 2014

Bilateral Moderate to Profound Hearing Loss Attributable to Congenital CMV



Adapted from Grosse, J Clin Virol, 2008

CMV Natural History



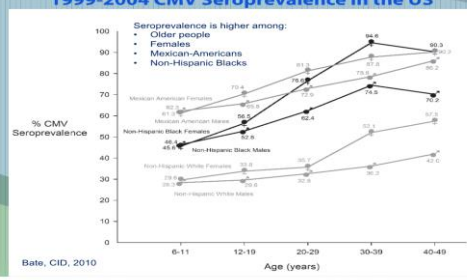
Primary infection → Latency → Reactivation → Recurrent or secondary infection

Reinfection

The Laboratory Vocabulary

Measurement	Labelled	Detects	Test format
Ever been infected	Seropositive	Antibody	ELISA
At risk for transmission	Shedding or excreting	Virus or viral DNA	PCR or culture

1999-2004 CMV Seroprevalence in the US



Bate, CID, 2010

Summary CMV Annual Seroconversion

Risk group	Summary annual seroconversion rate (%)	95% confidence interval (%)
Pregnant women	2.2	2.1 - 2.4
Parents with child not shedding CMV	2.1	0.3 - 6.8
Healthcare workers	2.7	2.3 - 3.2
Day care providers	8.5	6.1 - 11.6
Women attending STD clinics	13	10 - 17
Parents with child shedding CMV*	24	18 - 30

*Annual infection rate of less 25% in this high risk group suggests that CMV is not easily transmitted.

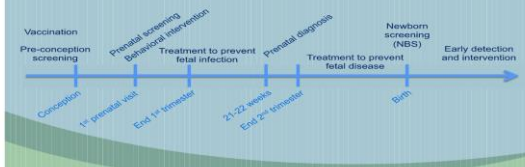
Adapted from Hyde, Rev Med Virol, 2010

Comparison of Models of Contagiousness

Study	Design	Force of infection (100 p-y)	Basic reproductive rate	Age of infection (years)
Measles Mumps Rubella	Review Ages 11-17	20 12 10		
Varicella	Convenience Ages ≥10	6		
CMV Griffiths (2001)	Hospital-based Ages 16-40	3.1 and 3.5	2.4 and 2.7	29 and 32 (median)
CMV Colugnati (2007)	Pop.-based Ages 12-49	1.8	1.7	28.7 (mean)
HSV-2	Pop.-based Ages ≥12	0.84		
Hepatitis A	Pop.-based Ages ≥10	0.2-1.0		
Hepatitis B	Pop.-based Ages 6-39	0.15		

Adapted from Colugnati, BMC Infect Dis, 2007

Potential Clinical and Public Health Interventions for Congenital CMV



Currently, none of these interventions is routine in the U.S.

Utility of Newborn CMV Screening

Probably satisfies	May not yet satisfy
• Important health problem	• Suitable test available
• Recognizable latent or early symptomatic stage	• Test acceptable to population
• Natural history adequately understood	• Agreed on policy on whom to treat
	• Facilities for diagnosis and treatment available
	• Cost-effective

Grosse, J Clin Virol, 2009

Laboratory Approaches to Newborn CMV Screening

Specimen	Method	Advantages	Disadvantages
Dried blood spot	PCR from DBS	NBS program already in place	CMV viral load lower in blood, less available specimen
Saliva	PCR from cheek swab	CMV viral load higher in saliva	Not part of existing NBS program
Urine	PCR from bagged urine or diaper insert	CMV viral load higher in urine	Not part of existing NBS program



Dollard, J Inherit Metabol Dis, 2010

TREATMENT-GANCICLOVIR

- **Kimberlin et al, JPeds, 2003**
- **42 patients with symptomatic CCMV had ABRs at base, 6, 12mo**
- **6 weeks IV ganciclovir**
- **Stable or improved hearing at 6mo (P=.06)**
- **21/25 (84%) treated patients 10/17 (59%) control patients**
- **Worsening hearing at 6mo**
- **0% of treated 41% of controls**
- **Stable or improved hearing at 12 mo (P=.08)**
- **79% of treated 39% control**
- **Worsening hearing at 12 mo (P=.002)**
- **21% of treated 61% of controls**

OTHER OUTCOMES WITH GANCICLOVIR

- **No sustained difference in the two groups between head circumference, thrombocytopenia, hepatomegaly, retinitis, weight gain, or hyperbilirubinemia**
- **Neutropenia in 63% if ganciclovir patients vs 21% of controls (P<.01)**
- ***Valganciclovir – oral pro-drug***
  JID 2008: 197 – similar drug levels, 38% neutropenia

PATIENT PRESENTATION - MD

1. MD : 3yo female, born in FL, military, noted to have GM delays and had some PT before coming to AK
2. After visit to our office, referred by PNP for evaluation and testing.
3. MRI showed white matter abnormalities.
4. Neuro said ?CP, ? Intrauterine viral infection, ? Leukodystrophy syndrome – refer to genetics
5. W/U: Neuro – VLDL fatty acids, Urine Organic acids, plasma amino acids, acyl carnitine
   Genetics- urine for polysaccharides, oligosaccharides, 13-enzyme lysosomal package
   If above normal – suggest chromosomal microarray and molecular study
6. Sedated ABR showed profound HL on left, normal on right at age 2, repeat recommended
7. Chromosomal array might cost parents $2000 out of pocket
8. Speculation ?? What if he was CMV+ as newborn??

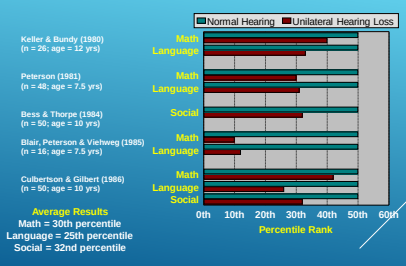
CMV-CONCLUSIONS

- Congenital CMV is a leading cause of hearing loss in children
- It is potentially preventable
  - -preconception and prenatal education
- Newborn screening is available
  - -need higher sensitivity, lower cost
- Prenatal and postnatal treatments are being tested in clinical trials
- Immunizations may be important future strategy

MILD AND UNILATERAL HEARING LOSS "IT'S NOT NOTHING"

- Minimal Hearing Loss (15-25dB) for children
  - Mild HL usually not picked up by UNHS
- Unilateral Hearing Loss-Normal hearing in one ear and a Permanent Hearing Loss in the other
  - 0.83/1000 overall (3.2/1000 NICU)(Prieve etal-2000)
- Eligibility for Services uncertain
  - Alaska – high probability of resulting in 50% DD

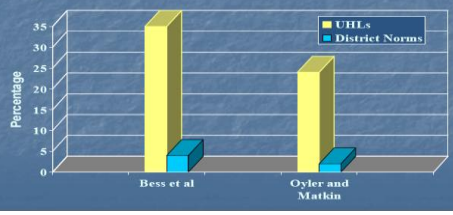
Effects of Unilateral Hearing Loss



DIFFICULTIES WHEN LISTENING WITH ONE EAR

- Localization
- Understanding in noise, at distance or if speech presented to ear with loss
- Loss of Binaural Summation- worth 3-8dB (Lieu)
- Problems magnified in children-difficulties "filling in the blanks" while developing language
- The more severe the UHL, the greater likelihood of academic failure
- Right UHL more problematic than Left UHL
- UHL may progress, develop into BHL (Cole,Flexer 2007)

Percent Failing at Least One Grade: UHL



Failure as a Function of Ear

MINIMAL HEARING LOSS IN SCHOOL AGED CHILDREN

BESS, ET AL

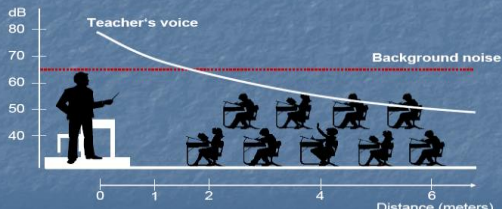
- 1218 Students – Grades 3, 6, 9
- MSNL vs NH controls (5.4% with MSNL)
- Evaluation Tools (CTBS, SIFTER, Behav.Checklist, Grade Retention, Functional Status)
- Results-CTBS lower at grade 3 (not 6 or 9)
  - 37% of MHLS retained 1 grade
  - Greater difficulties with behavior, energy, stress, social support and self-esteem

Failure Rates of Children with MSHL & with NH (Bess et al., 1998)

LISTENING FATIGUE AND SELF CONCEPT

- **"Because sound is audible, but not understandable (in mild/unil. HL), children may be inattentive, disinterested, or aloof…leading to problems in peer/social relations. …not able to hear conversations in groups, …may feel insecure, "left out" leading to difficulty with behavior and peer interactions"(Bess et al-1998)**
- **Problems accentuated if English is 2nd language**
- **Viewed as "giving up easily on new/difficult tasks**
- **Rated lower in areas of dependence and independence, attention to task, emotional ability, peer relations and social confidence.**
- **Teachers frequently unaware of problem (McKay2006)**
- **Between 33 – 50% of children with mild/unilat HL have academic, social, or behavioral difficulties**

Distance and background noise



How will I know if my child is having difficulty?

- Your child might: Get easily frustrated. Seem overly tired at the end of the day.
- Seem like he or she is not paying attention.
- Respond incorrectly to a question or request. "
- Act out" due to frustration.
- See more at: http://www.asha.org/public/hearing/Unilateral-Hearing-Loss-in-Children/#sthash.xgAUv0Vg.dpuf

EHDI WEBSITES

Alaska EHDI website:
http://dhss.alaska.gov/dph/wcfh/Pages/newborn/default.aspx

NCHAM website:
http://www.infanthearing.org/stateguidelines/index.php

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