Early Hearing Detection and Intervention (EHDI): A Primer for Residents

A PRESENTATION FROM THE AMERICAN ACADEMY OF PEDIATRICS & IOWA EARLY HEARING DETECTION & INTERVENTION PROGRAM

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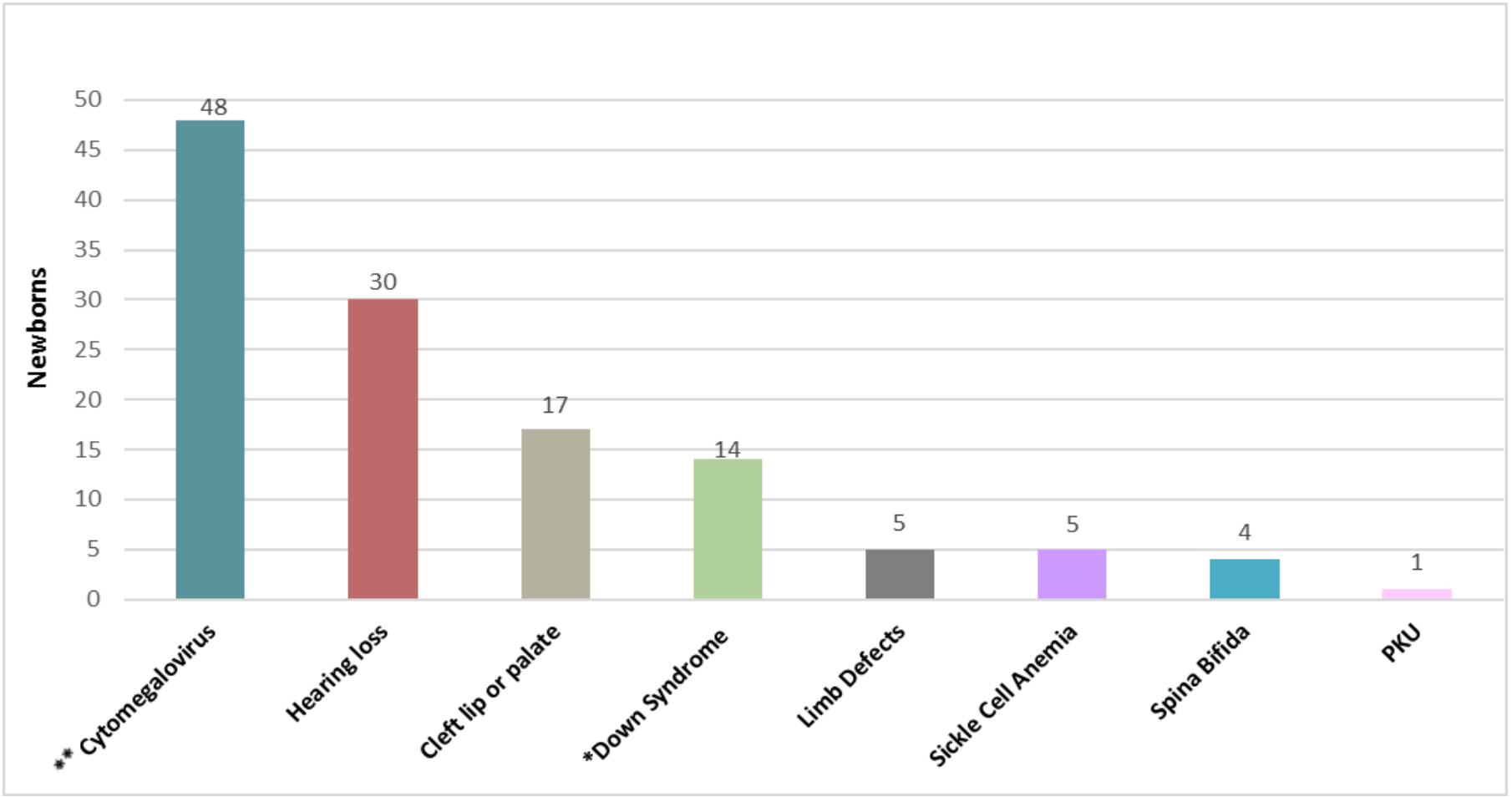
Hearing Facts

Early identification and intervention of a child who is Deaf or Hard of Hearing (D/HH) will support the development of good communication, language, and social skills.

Delayed Early Intervention can be associated with speech and language delays and inability to reach each child's full potential.

Comparison of Select Congenital Conditions

Incidence per 10,000 of Congenital Conditions



*In mother's age 25 and younger **CMV is the most common non-genetic cause of permanent childhood hearing loss

Spoken Language Development in Late vs Early Identification of Child who is Deaf or Hard of Hearing (D/HH)

- Late diagnosis of hearing status:

https://www.youtube.com/watch?v=5yK4Z7n5NsI#t=454s

- Early diagnosis of hearing status:

https://www.youtube.com/watch?v=5yK4Z7n5NsI#t=526s

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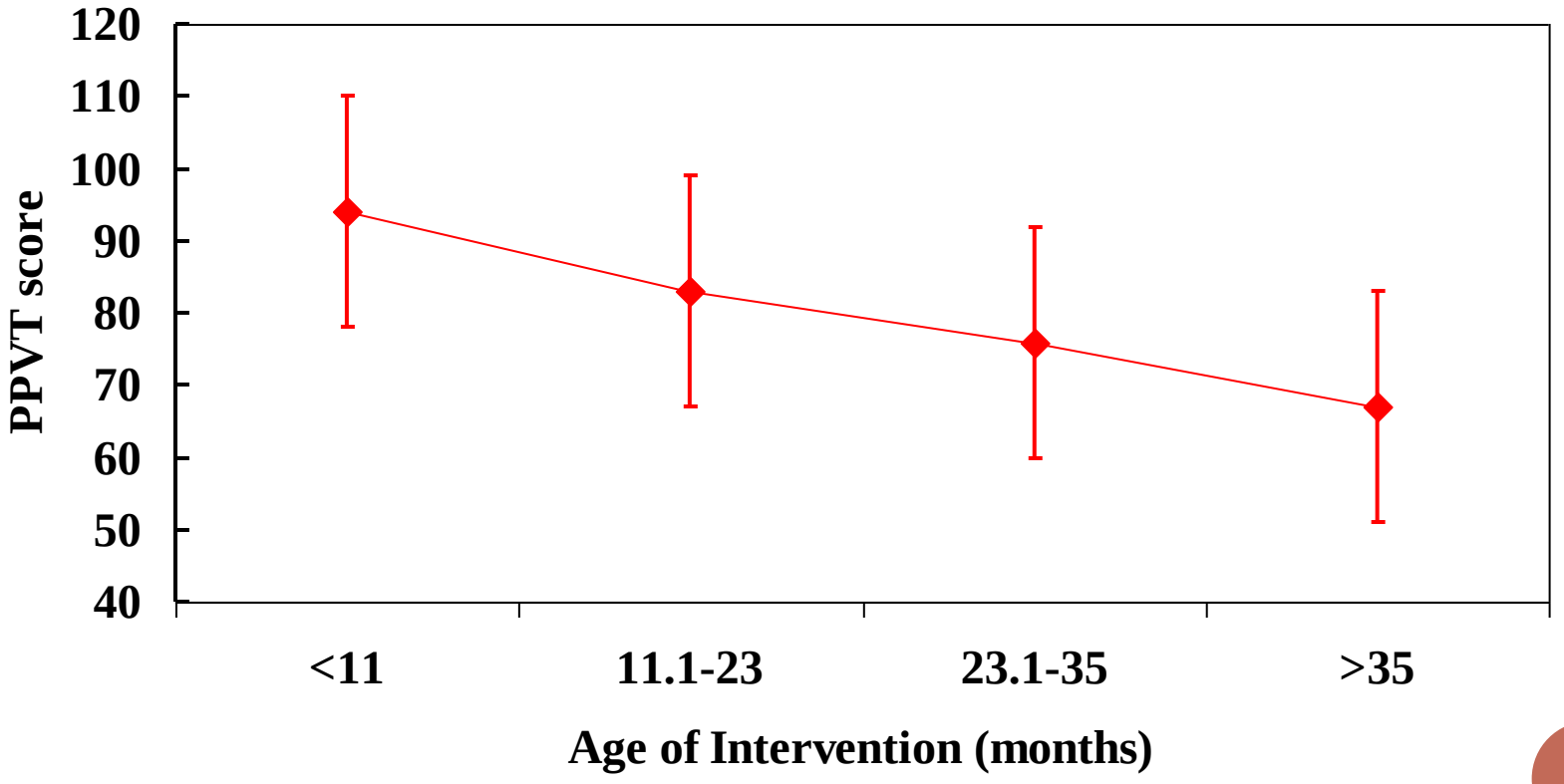
Moeller(2000): Receptive vocabulary scores at age 5 years as a function of age of enrollment in intervention (Children who are deaf or hard-of-hearing)



Three Key Components of Early Hearing Detection & Intervention Programs

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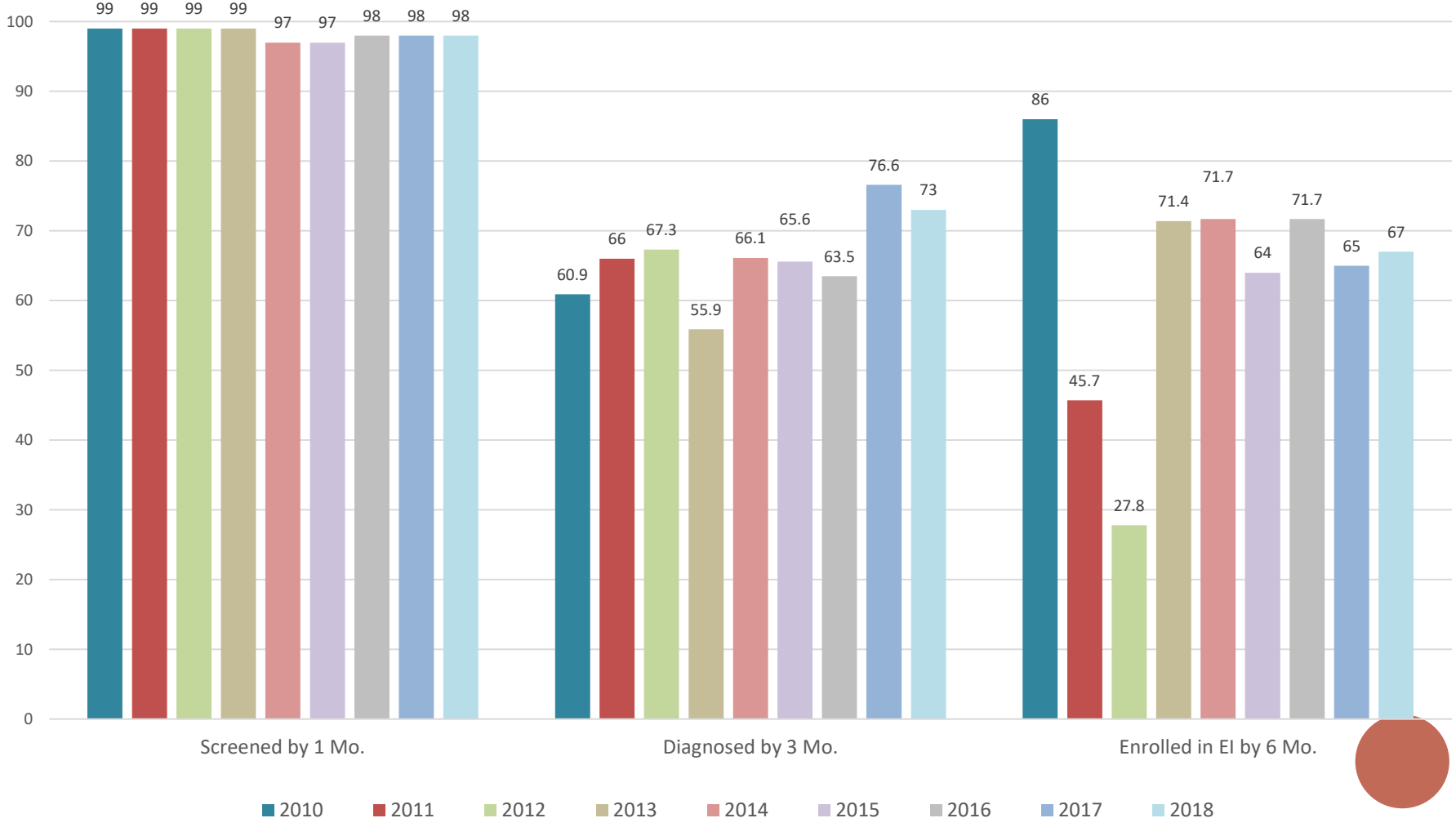
Early Hearing Detection & Intervention (EHDI) Program

National EHDI Goals

- All infants will receive a hearing screening no later than 1 month of age. In Iowa, they must also have a hearing re-screen no later than 1 month
- Infants not passing the hearing re-screen will receive an appropriate audiologic and medical evaluation to determine diagnosis no later than 3 months of age
- All infants identified as D/HH be enrolled in early intervention no later than 6 months of age

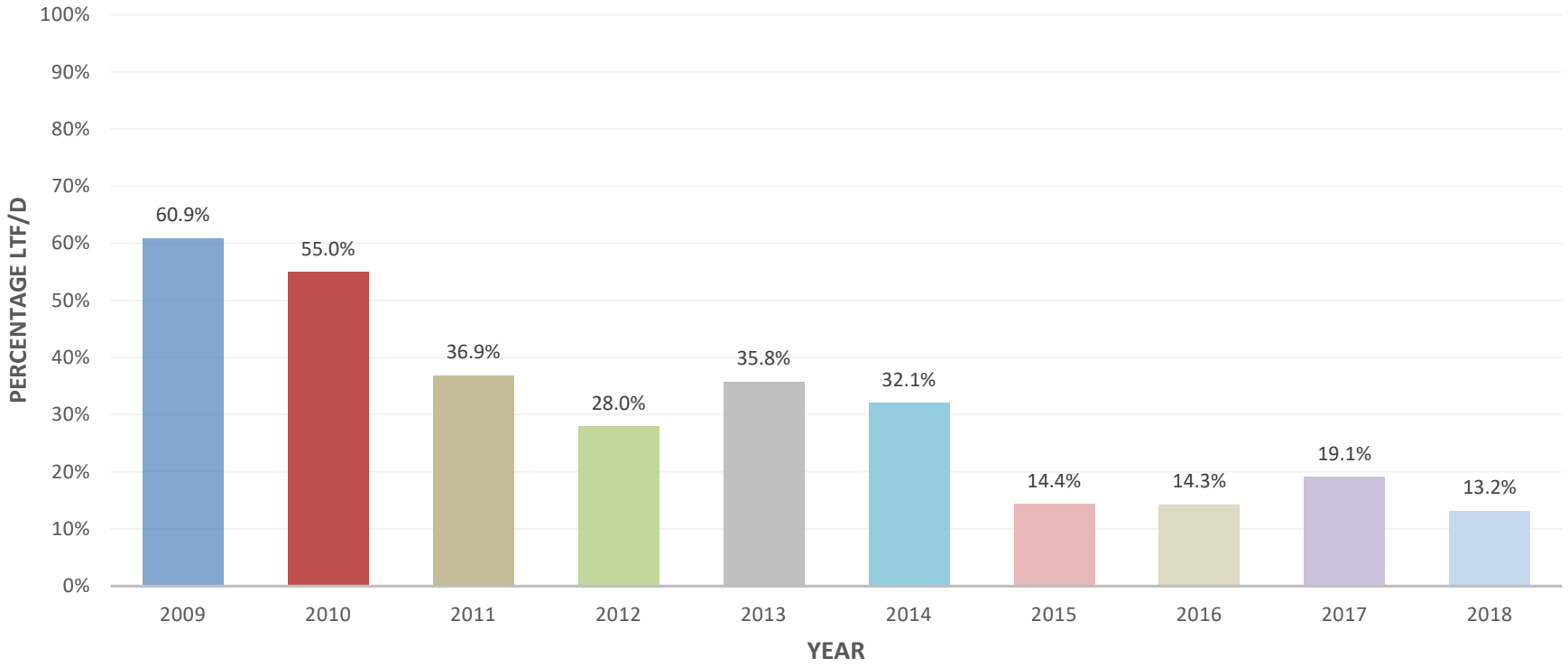
1-3-6

How Is Iowa Doing in Meeting National 1-3-6 Goals?



Lost to Follow-Up/Documentation

Percentage of Infants Needing Follow-up That Did Not Complete Recommendations



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Iowa Early Hearing Detection and Intervention Legislation

- January 1, 2004 – mandatory screening of all Iowa newborns with provision for active refusal
- Mandatory reporting of hearing screen results to parents (before discharge), infant's primary care provider (PCP) and Iowa Department of Public Health (IDPH)
- Outpatient re-screen of infants who do not pass the birth hearing screen at 2-4 weeks of age (hospitals, AEAs, healthcare provider) – mandatory reporting of results to IDPH w/in six days of screen
- Mandatory reporting of diagnostic audiological evaluation results on children under 3 by all licensed audiologists to IDPH and infant's PCP w/in six days of evaluation
- Mandatory reporting of risk factors (2009) for hearing loss

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Challenges

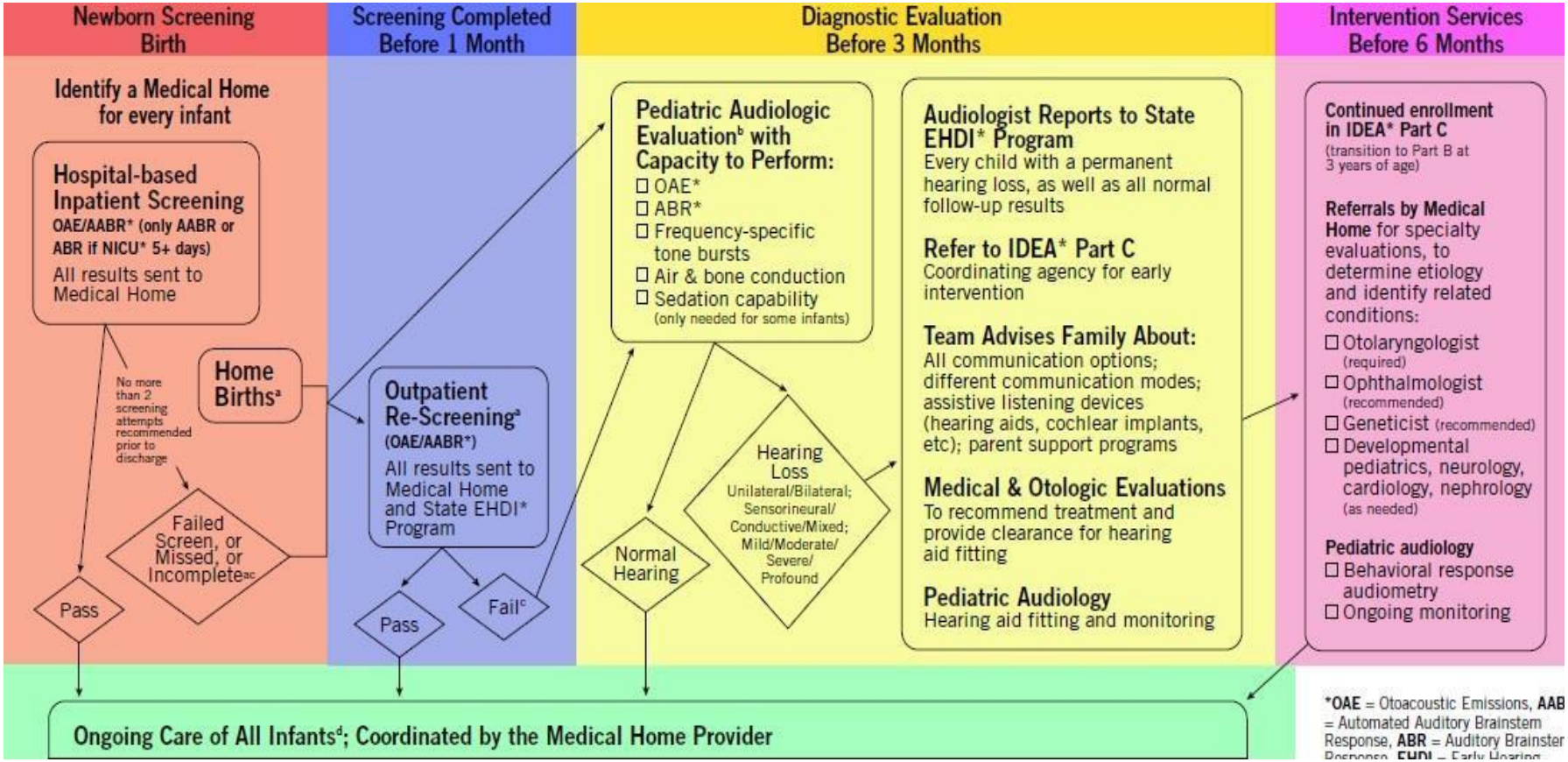
- Unfunded mandate
- Re-screening infants over and over rather than referring to pediatric audiologist for diagnostic assessment following failed hearing re-screen
- Hand off between some practitioners not organized causing delays or unnecessary follow-up
- Families unclear on next steps and importance of timely follow-up
- Few pediatric diagnostic audiology centers
- Infant and families not being referred to Early ACCESS (Part C, IDEA) upon diagnosis of hearing loss
- Minimal funding for family support and deaf mentorship, but no sustainable funding
- Hearing aids are often not covered by insurance which may cause a hardship for some families

The Role of the Medical Home in Follow-up for Infants that Do NOT Pass Their Hearing Screen at Birth

If there is any suspicion an infant may be D/HH

- **Do listen to parents concerns and refer immediately for full audiology evaluation for any parental worries about hearing or language development**
- Encourage prompt follow-up with re-screen and diagnostic evaluations
- Make sure diagnostic evaluations are done by a pediatric audiologist with appropriate equipment
- Set up electronic medical record (EMR) system to include results of hearing screens
- Flag all patient charts for children that require follow-up hearing screens or diagnostic evaluations
- Flag all patient charts for children that are at risk for late onset hearing loss

EHDI Guidelines for Pediatric Medical Home Providers



Guidelines and Referral checklist for providers available at
http://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Early-Hearing-Detection-and-Intervention.aspx

The Role of the Medical Home for Infants Identified as D/HH

- Address the family's concerns
- Ensure the family is seeing an experienced pediatric audiologist
- Refer the family to appropriate specialists
  - Otolaryngology, Genetics, Ophthalmology
- Help the family obtain early intervention services and family support
- Monitor developmental milestones and ear infections

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Specialty Referrals

Otolaryngology

- Assess integrity of ear canal and middle ear
- Order appropriate diagnostic screening such as temporal bone CT, MRI, etc.
- Discuss possible surgical interventions
- Counsel family and follow for success of intervention

Genetics

- Evaluate for possible genetic causes of hearing change
- Counsel family and patient

Ophthalmology

- Assess integrity of visual system
- Evaluate for visual changes known to be associated with hearing changes

Case Study 1: Baby James

- Baby James comes to your office for a well child visit at 2 weeks of age
- Full term, 3300 g, uncomplicated pregnancy and delivery
- Parents are concerned because he did not pass his newborn hearing screen in the hospital and they wonder what to do now
- How do you counsel them?

OAE: Otoacoustic Emissions

Picture of OAE Screening. [Photograph]. Retrieved from http://www.medicalexpo.com/prod/otometrics/product-70796-473134.html

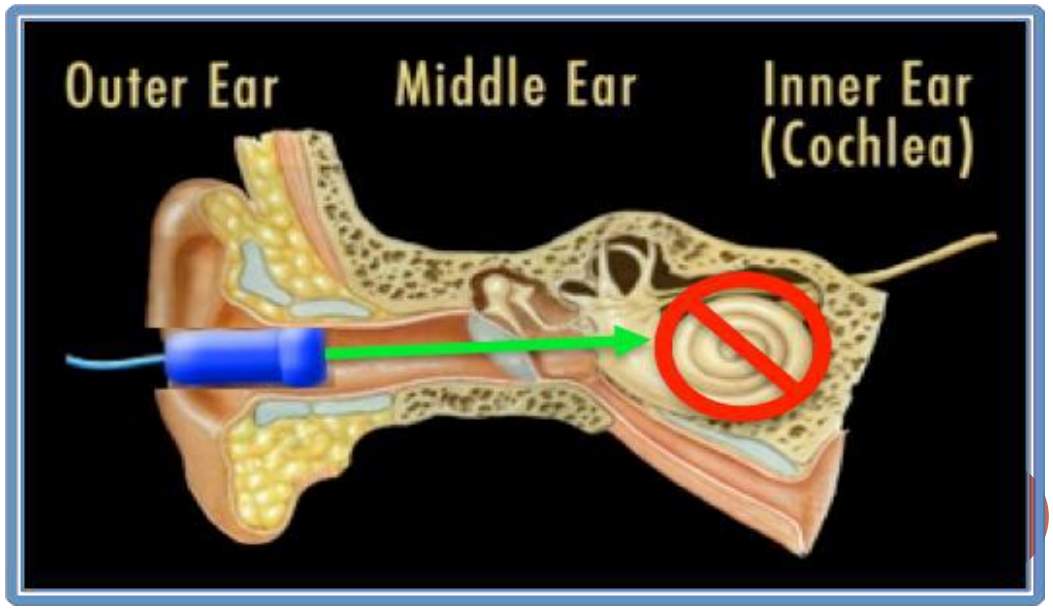

Diagram of infant ear. Retrieved from http://archive.constantcontact.com/fs154/1102782899287/archive/1115854843507.html

AABR:

Automated Auditory Brainstem Response

Picture of AABR Screening. [Photograph] Retrieved from http://infanthearing.vihsp.org.au/home

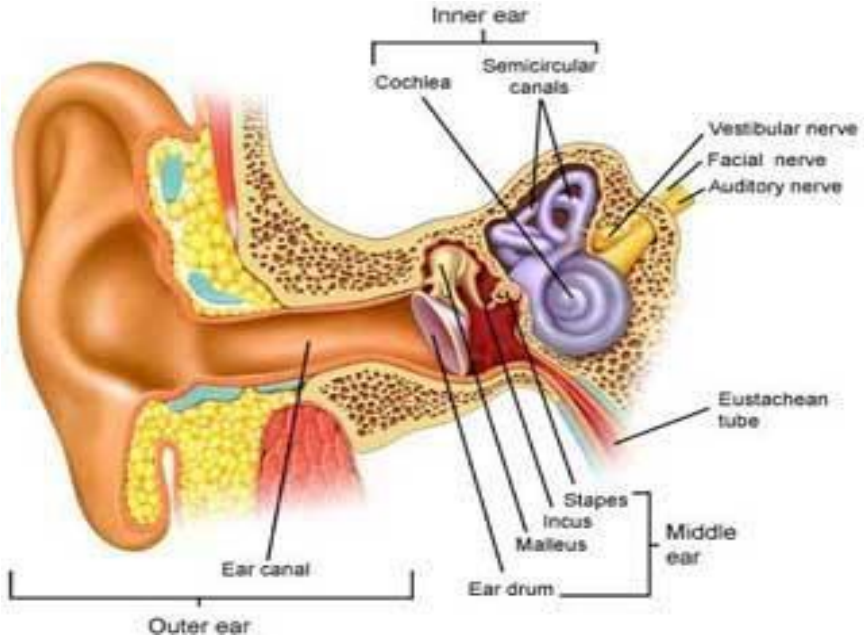

Diagram of infant ear. Retrieved from: https://www.medicalhomeportal.org/image/65

Newborn Hearing Screening

Otoacoustic Emissions vs. Automated Auditory Brainstem Response

	Otoacoustic Emissions (OAE)	Automated Auditory Brainstem Response (AABR)
Technique	Probe with microphone placed in the ear canal. Acoustic stimuli presented.	Earphone placed in the ear canal, electrodes placed on baby's scalp. Acoustic stimuli presented.
Measurement	OAEs are measured in the ear canal. With outer/middle ear and/or cochlear problems, no OAEs are detected.	Neural activity of cochlea, auditory nerve and brainstem is measured. Problems with peripheral auditory and/or auditory nerve and/or brainstem result in abnormal or absent measurements of auditory neural activity.
Advantage	Is easier and quicker.	Can indicate auditory nerve or auditory brainstem pathway dysfunction.
Disadvantage	Will NOT identify auditory nerve or auditory brainstem dysfunction.	May require sedation after 4 months of age.

Audiogram

Normal: 0-15 dB

Mild: 15-40 dB

Moderate: 40-55 dB

Moderate to Severe: 55-70 dB

Severe: 70-90 dB

Profound: 90-110 dB

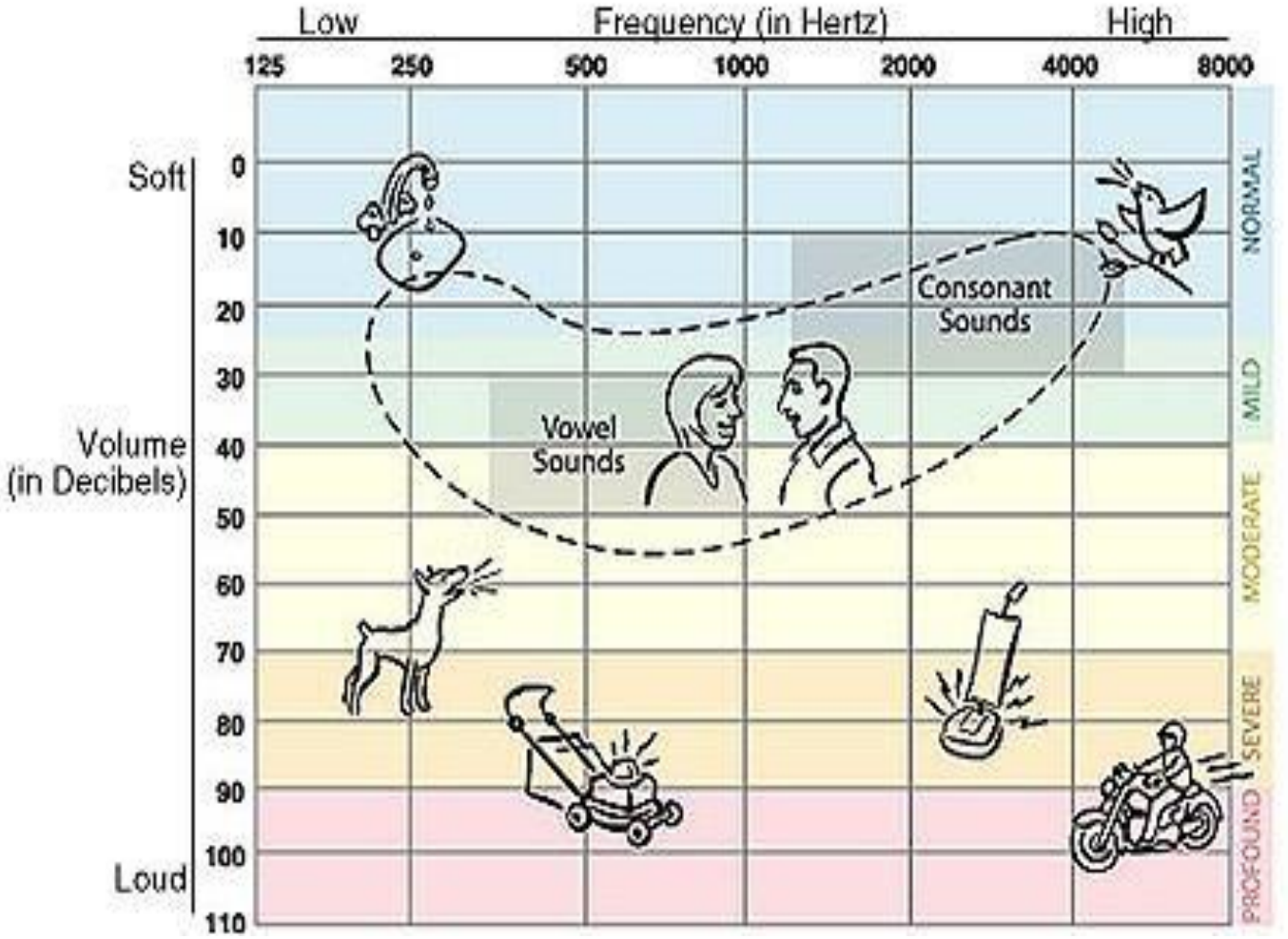

Image of an audiogram. Retrieved from: http://www.fairviewebenezer.org/HealthLibrary/Article/83640

Case Study 1: James at 2 months of age

- James returns for his 2 month well child visit
- Parents report that after the 2 week visit, he returned to the birth hospital for a repeat OAE, and because he did not pass this, had an diagnostic ABR with a pediatric audiologist
- ABR revealed that he had bilateral moderate to severe sensorineural hearing loss. The audiologist recommended return visit for hearing aid fitting
- Parents are doubtful of the results because James is cooing and startles to loud noises
- What other referrals need to be placed at this time?
- What needs to happen by 6 months?

Case Study 2: James' Brother

- The same mother now presents with the child's two and a half year old brother, Nathan, at his well child check up

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Case Study 2: James' Brother

- She is concerned he is not responding to her consistently, and doesn't appear to have as much spoken language as the other children in his daycare
- She confides to you that she thinks she is just being paranoid since the recent identification of his brother's hearing status

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Case Study 2: James' Brother

What is the appropriate next step?

A. Use developmental assessment tool (eg, ASQ, PEDs) to evaluate language development – if normal, reassure mother and re-evaluate child in 6 months

B. Stand behind child and whisper "what is your name?" to see if they answer correctly

C. Use pure tone hearing screening in office

D. Refer directly to audiology

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Ongoing Care of All Infants[d]; Coordinated by the Medical Home Provider

- Refer promptly for audiology evaluation when there is any parental concern‡ regarding hearing, speech, or language development

- Refer for audiology evaluation (at least once before age 30 months) infants who have any risk indicators for later-onset hearing loss:

- Family history of permanent childhood hearing loss‡
- Neonatal intensive care unit stay of more than 5 days duration, or any of the following (regardless of length of stay):
  ECMO‡, mechanically-assisted ventilation, ototoxic medications or loop diuretics, exchange transfusion for hyperbiliruinemia
- In utero infections such as cytomegalovirus‡, herpes, rubella, syphilis, and toxoplasmosis
- Postnatal infections associated with hearing loss‡, including bacterial and viral meningitis
- Craniofacial anomalies, particularly those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
- Findings suggestive of a syndrome associated with hearing loss (Waardenburg, Alport, Jervell and Lange-Nielsen, Pendred)
- Syndromes associated with progressive or delayed-onset hearing loss‡ (neurofibromatosis, osteopetrosis, Usher Syndrome)
- Neurodegenerative disorders‡ (such as Hunter Syndrome) or sensory motor neuropathies (such as Friedreich's ataxia and Charcot Marie Tooth disease)
- Head trauma, especially basal skull/temporal bone fracture that requires hospitalization
- Chemotherapy‡

‡Denotes risk indicators of greater concern. Earlier and/or more frequent referral should be considered

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Risk Factors for Hearing Loss

Three Month Evaluation

- Bacterial and viral meningitis (especially herpes viruses and varicella) or encephalitis
- Congenital Cytomegalovirus (CMV) confirmed in infant
- Extra-corporeal membrane oxygenation (ECMO)
- Family history of hearing loss (permanent, sensorineural hearing loss since childhood)
- Head injury (especially basal skull/temporal bone fracture requiring hospitalization)
- Chemotherapy

Risk Factors for Hearing Loss

Nine Month Evaluation

- Family history of childhood hearing loss
- NICU stay longer than five days
- Cranio-facial anomalies (cleft lip or palate, microtia, atresia, white forelock, microphthalmia, congenital microcephaly, congenital or acquired hydrocephalus, temporal bone abnormalities)
- Hyperbilirubinemia with exchange transfusion regardless of length of stay
- Asphyxia or Hypoxic Ischemic Encephalopathy
- Aminoglycosides administration for more than 5 days (e.g. Vancomycin, Kanamycin, Streptomycin, Tobramycin)
- In utero infections such as herpes, rubella or syphilis or toxoplasmosis confirmed in infant
- Syndromes (e.g. Trisomy 21-Down Syndrome, Goldenhar, Pierre Robin,

  CHARGE Syndrome, etc.)

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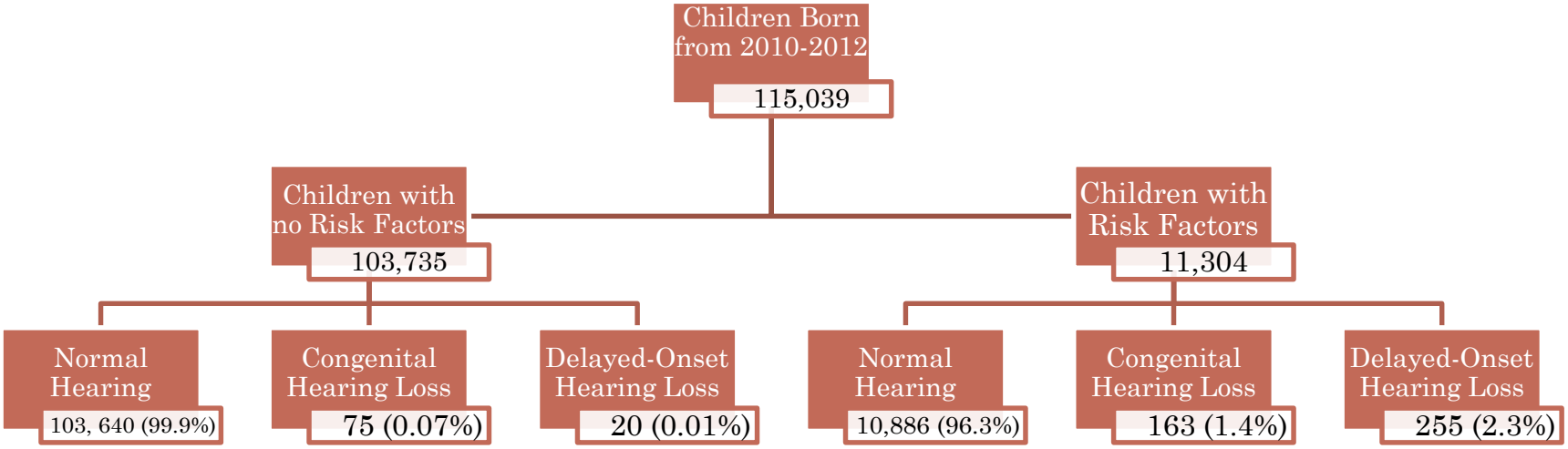
3 YEAR REVIEW OF CHILDREN WITH RISK FACTORS IN IOWA (DUMANCH ET AL., 2017)

- What percent of children with risk factors (CWRF) have hearing loss? Normal hearing?



2 per 1000 born with hearing loss, additional 2.4 per 1000 develop hearing loss before age 3 (Most have risk factors)

Odds ratios of delayed onset hearing loss associated with selected risk factors (Dumanch et al., 2017)

- NICU stay greater than 5 days: OR = 24.9
- This means a baby with a NICU stay of more than 5 days is at least 24 times more likely to develop permanent hearing loss than a baby without that risk factor
- Family history of hearing loss alone: OR = 8
- Ototoxic medications alone: OR = 9
- Risk factors in combination increase the odds ratios

CMV screening for infants

- CMV is the most common non-genetic cause of congenital hearing loss
- Physicians are required by law to provide CMV education and screening of infant upon a failed second hearing screen
- When an infant does not pass the initial hearing screen, the repeat screen should occur within 10 days & no later than 21
- If infant does not pass repeat hearing screen, a CMV screen should be ordered immediately and occur no later than 21 days of age
- Diagnostic assessment should be ordered upon failed second screen
- Antiviral treatment needs to start by 30 days of age in infants with hearing loss and confirmed CMV

Take Home Points

- When an infant does not pass the initial hearing screen, the repeat screen should occur no later than 1 month of age
- If infant does not pass repeat hearing screen, diagnostic ABR should be completed by pediatric audiologist no later than 3 months of age
- Appropriate hearing testing is required to identify hearing status. You must keep them moving through the point of diagnosis
- Early intervention referral should be completed immediately following diagnosis and no later than 6 months of age, along with subspecialty referrals

Take Home Points

- If infant does not pass repeat hearing screen, a CMV screen should be ordered immediately and occur no later than 21 days of age
- If infants does not pass repeat hearing screen, a diagnostic assessment should be ordered immediately
- **Parent concern is a very sensitive indicator of hearing changes, and warrants an immediate referral for audiology evaluation**

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Take Home Points

- It is important to develop practice parameters for tracking high risk registry infants/children for late onset hearing changes
  - For example, include "at risk for late-onset hearing loss" in the running problem list (ICD-10 code Z91.89, "Other specified personal risk factors, not elsewhere specified")
- Any child with risk factors for late-onset hearing should be evaluated by audiology either at 3 or 9 months of age
- Even if language development is on track and there are no concerns, any child with risk factors should still be evaluated

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Useful Web Sites

- American Academy of Pediatrics (AAP) EHDI page
  http://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Early- Hearing-Detection-and-Intervention.aspx
- Iowa Early Hearing Detection & Intervention Program
  http://idph.iowa.gov/ehdi
- Joint Committee on Infant Hearing (JCIH)
  http://www.jcih.org/
- Boys Town National Research Hospital
  http://www.boystownhospital.org/

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Acknowledgement

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