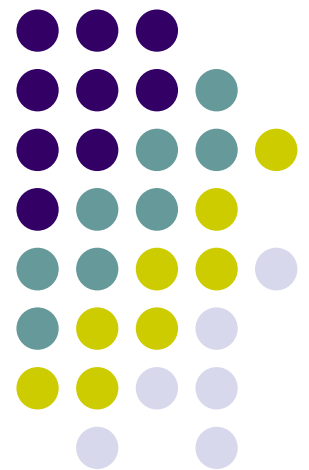


Age of Identification of Delayed Onset Hearing Loss in Infants and Young Children

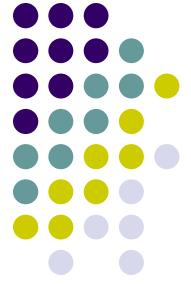
Angelique Boerst, MA

Marc Thorne, MD

University of Michigan Medical Center

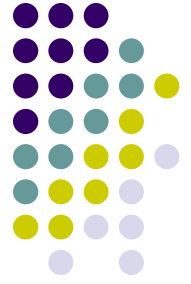


History of Risk Indicators and Delayed Onset Hearing Loss:



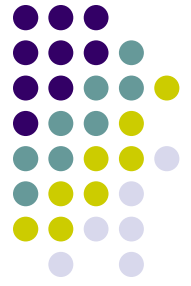
- First recommendations to monitor for delayed onset hearing loss made in 1982 by JCIH
- 3 criteria initially identified
 - Family History
 - Degenerative disease
 - Intrauterine infection
- No recommendations for frequency of monitoring

History of Risk Indicators and Delayed Onset Hearing Loss:



- 1990 indicators added:
 - Meningitis
 - Chronic lung disease
 - PPHN
 - Ototoxic medication
- 1994 suggested monitoring every six months until three years of age. Additional factors added:
 - NFII and neurodegenerative disorders
 - Conductive hearing loss including chronic OM, anatomic deformities, neurodegenerative disorders

JCIH 2000 Position Statement

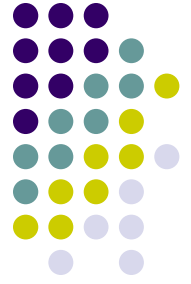


- Statement introduced 8 Principles for Effective EHDI Systems
- Principle 4: “Infants with indicators associated with late-onset, progressive or fluctuating hearing loss as well as auditory neural conduction disorders and/or brainstem auditory pathway dysfunction should be monitored.”



JCIH 2000

- Continue recommendation to monitor every 6 months until 3 years of age
- Indicators include
 - Parental caregiver concern
 - Stigmata associated with syndrome
 - Postnatal infections
 - In-utero infections
 - Hyperbili requiring transfusion
 - PPHN
 - Mechanical ventilation
 - ECMO
 - Syndromes associated with progressive hearing loss
 - Neurodegenerative disorders
 - Head trauma
 - Recurrent/persistent otitis media



JCIH 2007 Position Statement

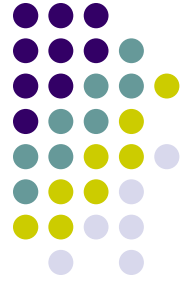
- Risk indicator categories collapsed due “to significant overlap among those indicators associated with congenital/neonatal hearing loss and those associated with delayed-onset/acquired or progressive HL”
- Indicators specific to delayed onset noted

Monitoring for LOHL

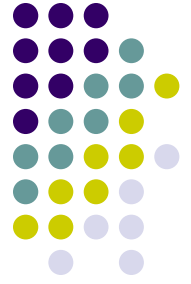


“Infants who pass the neonatal screening but have a risk factor should have at least one diagnostic audiology assessment by 24-30 months of age. Early and more frequent assessment may be indicated for children with CMV, syndromes associated with SNHL; for children who have received ECMO or chemotherapy; and when there is a caregiver concern or family history of hearing loss.”

Addendum to 2007 statement



- “Early and more frequent can be interpreted as every six months, or more, depending upon the clinical findings and concerns.”



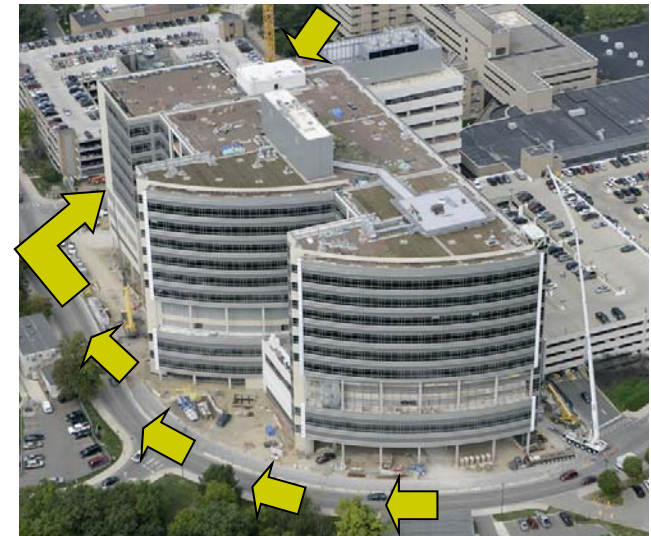
Clinical Concerns

- How will recommendation be interpreted?
Will children with delayed onset hearing loss be identified later if first monitoring audiogram doesn't take place until 24-30 months?
 - Age of identification
 - Types of risk indicators
 - Parental concern as a function of age of identification

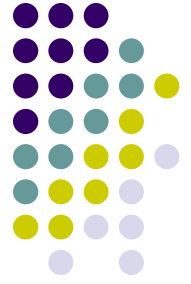


Population

- Retrospective review
- 21 children identified with delayed onset bilateral SNHL at University of Michigan prior to 48 months of age
- Born across 21 year span (1988-2009)
- Excluded children with
 - Auditory neuropathy
 - Conductive hearing loss
 - Meningitis
 - Chemotherapy



Newborn Hearing Screening at the University of Michigan



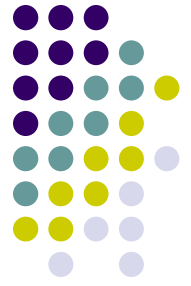
- Tertiary Care Hospital with Level III NICU
- Program started in 1987
- Screened infants at high risk for hearing loss
- All NICU charts reviewed by audiologists to determine which infants should be screened
- All screening done by audiologist using Nicolet Spirit, Nicolet Compact 4 or Biologic Navigator/Traveler system



Current EHDI Program

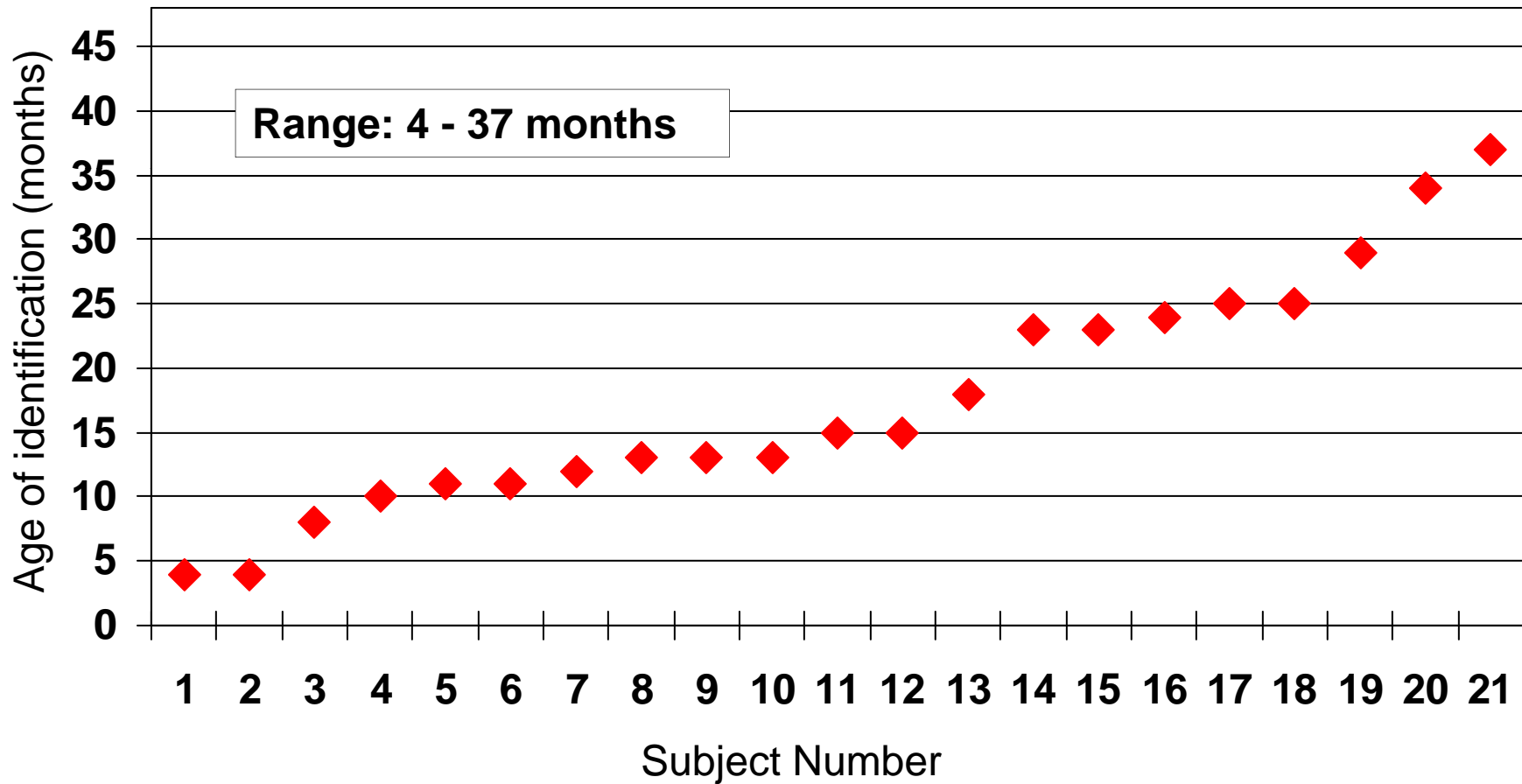
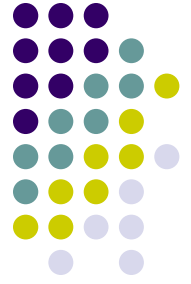
- Started in 2001
- Screenings done by trained audiology technicians using Biologic ABAER
- Program supervised by audiologist
- Technicians determine infants at risk for delayed onset HL through case review and obtaining family history information from parent interview

Population: Screening Results



- All with passed bilateral hearing screen
- 19 screened at University of Michigan
- Screening results for remaining two confirmed before included
- Screening completed using:
 - OAE (1)
 - ABR (12)
 - AABR (8)

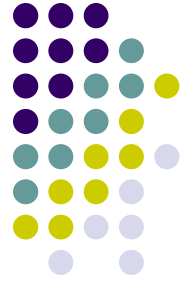
Population: Age of Identification



How does frequency of monitoring impact age of identification?



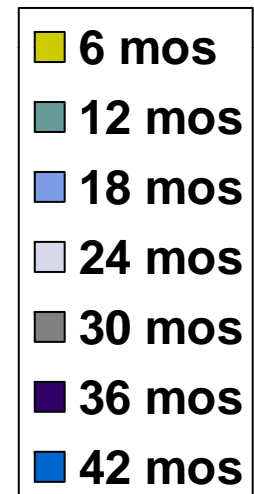
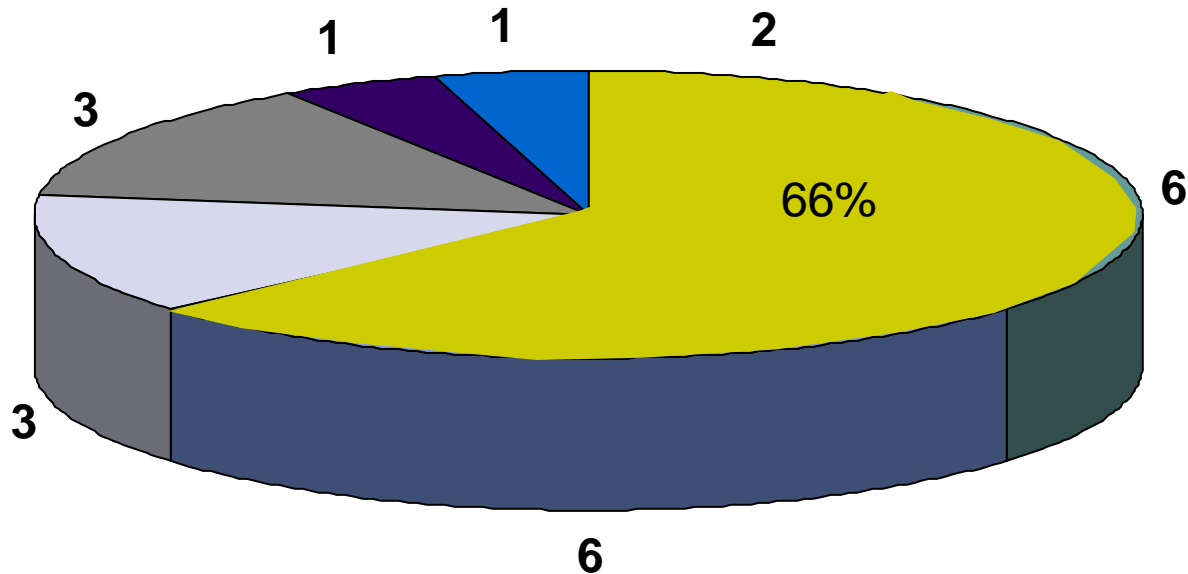
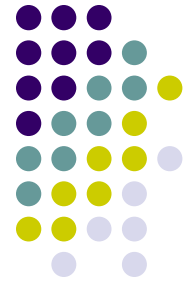
- Once child is identified as at risk for delayed onset hearing loss, how often should monitoring occur?



Protocols

- University of Michigan protocol is every 6 months until preschool age (48 months)
- JCIH at least once before 24-30 months of age.

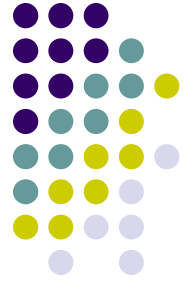
Children identified using six month monitoring intervals





Age of identification

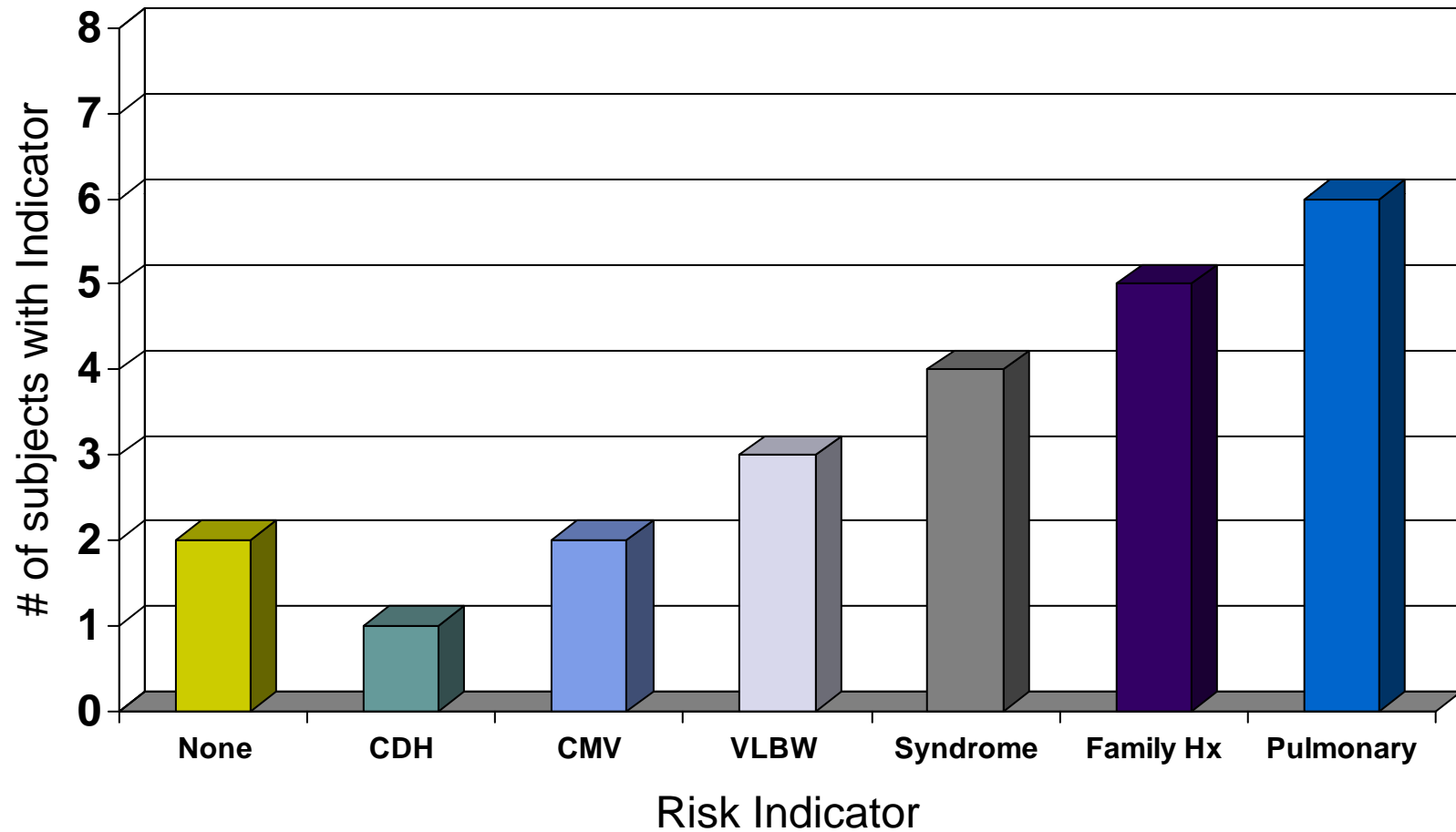
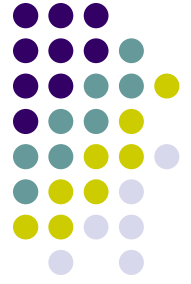
- Over half of the infants were identified with hearing loss before 24 months
- Identification is likely to over estimate date of onset. Unable to determine if onset of remaining children occurred before 24 months.



Risk Indicators

- JCIH 2007 states the need for early and frequent monitoring for CMV, syndromes associated with SNHL, family history, ECMO and caregiver concern
- Does this sample agree with these recommendations?
- Of note, pulmonary factors include PPHN and vent > 14 days (not 5 days)

Risk Indicators for Delayed Onset Hearing Loss



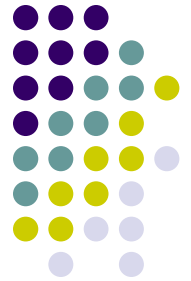


Odds Ratio for Risk Factors

Risk Factor	Relative Risk	95% Confidence Interval	P value (Fisher's exact test)
FHHL	33.0	(12.2-89.5)	<0.0001
Pulmonary	48.3	(20.5-114.0)	<0.0001
CDH	25.3	(3.5-185.0)	0.04
Perinatal Infection	173.3	(43.9-683.4)	0.0001
Syndrome	37.4	(11.2-125.1)	0.0001
VLBW	82.7	(25.1-272.5)	<0.0001

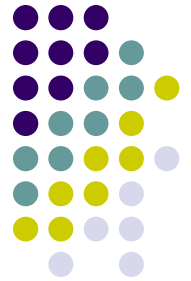
Values >1 indicate increase odds (or risk)

Multiple Logistic Regression



Risk Factor	Odds Ratio	95% Confidence Interval	P value
Perinatal Infection	642.1	121.9 - 3383.1	<0.001
Family History	49.7	15.7 - 157.4	<0.001
Pulmonary	31.4	9.9 - 99.9	<0.001
Syndrome	31.1	7.0 - 137.8	<0.001
CDH	14.4	1.6 - 127.0	0.016
VLBW	<1		

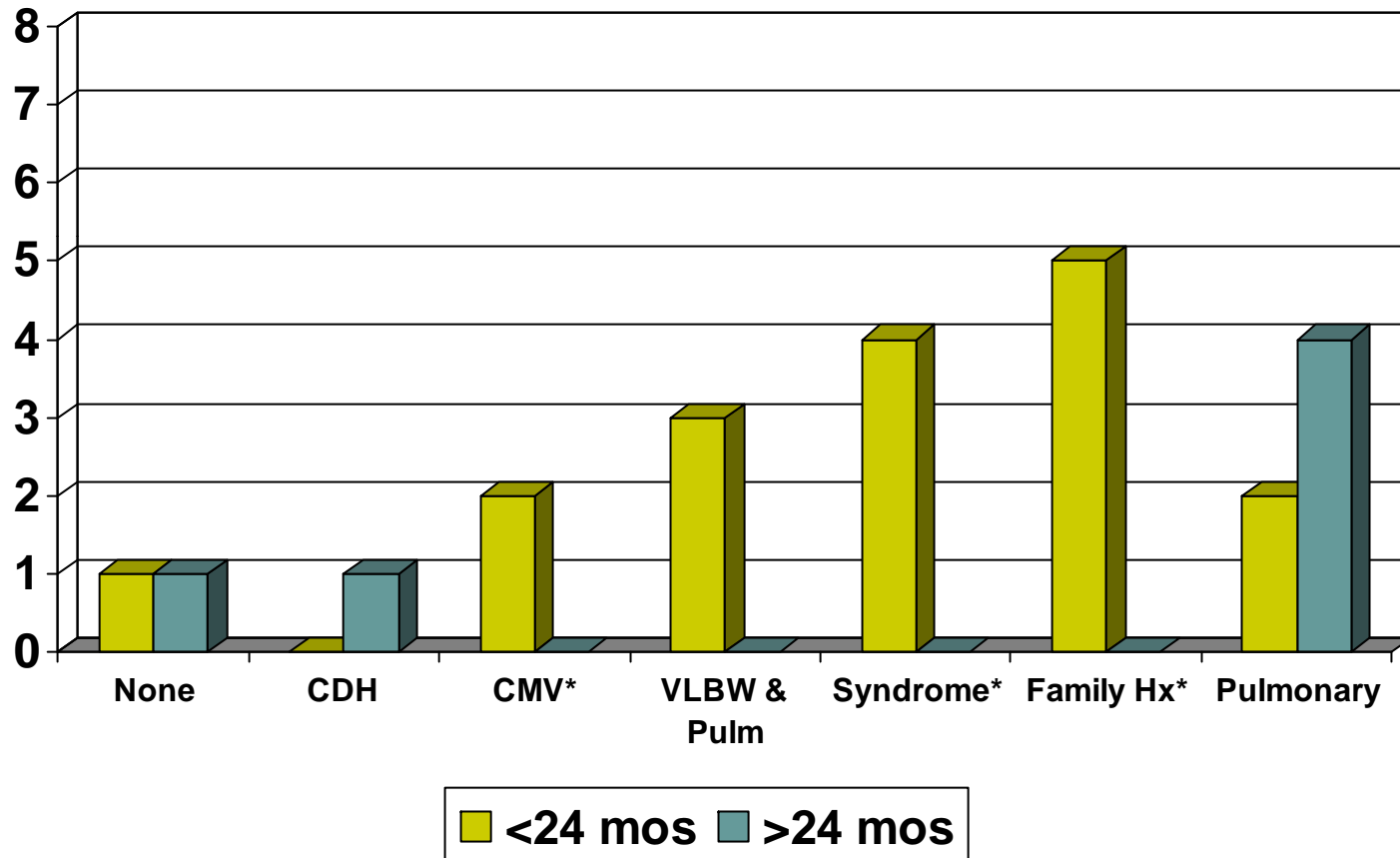
Risk Indicators & Delayed Onset Hearing Loss



- Risk indicators identified by JCIH 2007 for delayed onset hearing loss demonstrate a statistically significant increased risk.
- Are any risk factors more likely to be associated with hearing loss prior to 24 months and more likely to benefit from “early and frequent” monitoring?

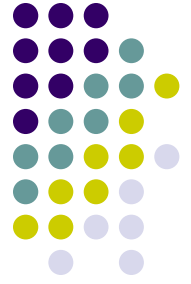


Risk Indicators vs Age of ID



*JCIH 2007 recommended "early and more frequent monitoring"

Risk Factors and Age of Onset



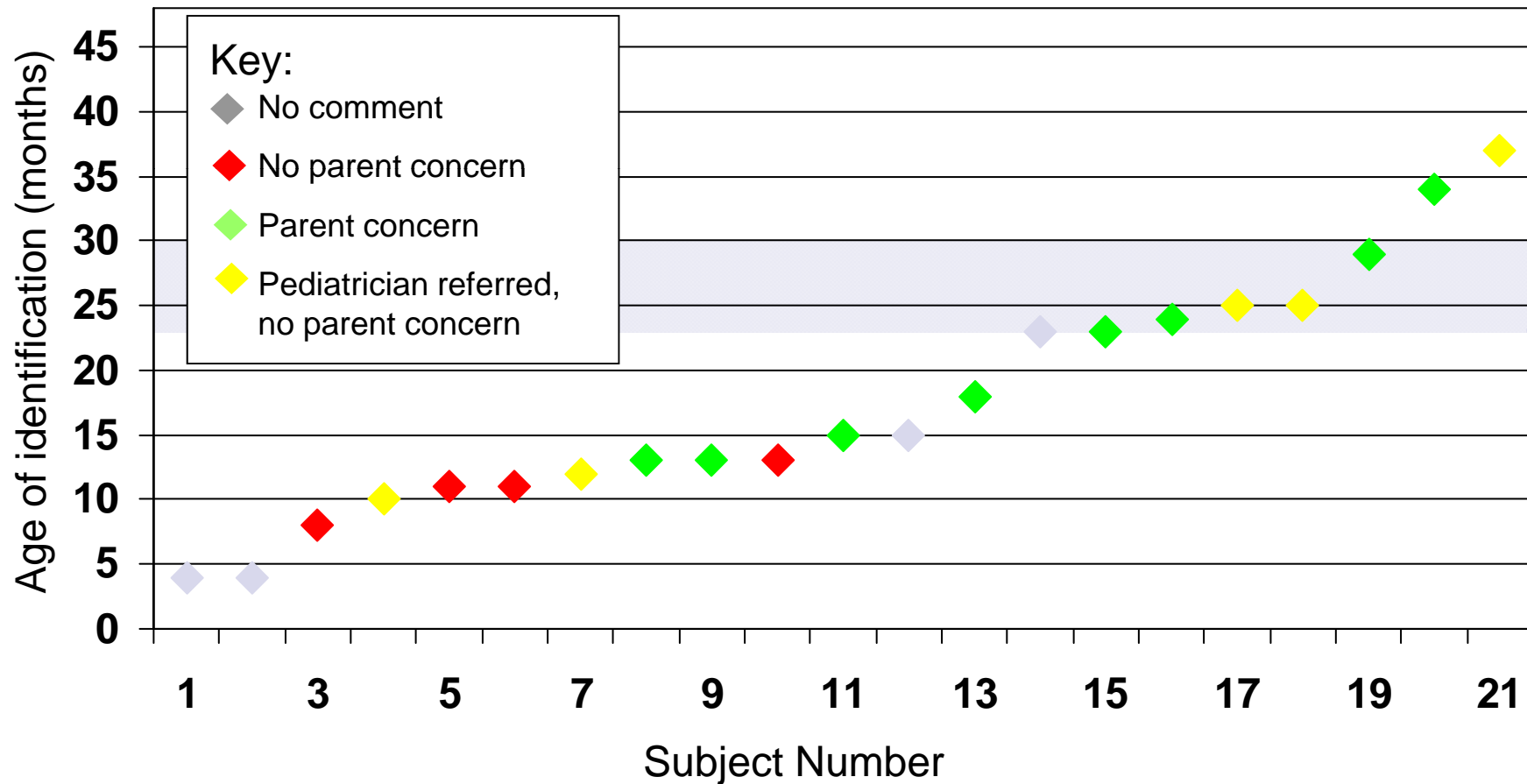
- This sample was in good agreement with JCIH 2007 recommendations.
- In this sample, 3 infants with VLBW and pulmonary indicators had onset of hearing loss of less 24 months. Multiple Logistic Regression did not demonstrate a statistical significance in the combination of these factors.



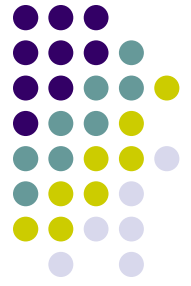
Role of Caregiver

- Caregiver concern has been cited as another reason to complete diagnostic testing
- What is the impact of caregiver concern on returning for testing following passed screen?
- Reviewed audiology report at time of initial identification for statement related to parental/pediatrician concerns.

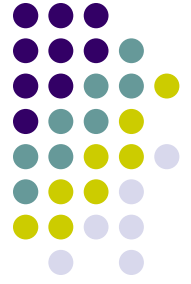
Report of Parent Concern & Age of Identification



Parental & Physician Concern



- As children become older, parents and physicians are more likely to have concerns about changes in hearing as speech and language skills are impacted.



Thoughts...Pros

- Current JCIH statement reduces the number of monitoring appointments recommended. This in turn may improve follow up with recommendations.
- Global recommendation of at least one hearing test between 24-30 months of age for all children with risk indicator may lead to less confusion regarding recommended protocol.

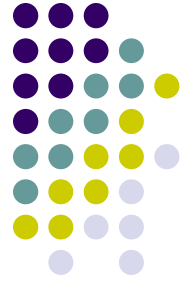


Thoughts...Cons

- If recommendation for early and frequent monitoring is not made, identification may be delayed.
- If monitoring isn't recommended until 24 months, parents may view as "not important," a test that "can wait" or forget monitoring was recommended.
- Current recommendation of 24-30 months occurs in tandem with parent and pediatrician concerns about lack of speech and language development secondary to hearing loss.

Thoughts...

Possible Directions



- If recommended window for screening were changed to 12-18 months, this data suggests that a majority of delayed onset hearing loss would be identified within this time frame.
- Hearing loss with onset after 18 months likely to be suspected due to its impact on speech and language.