Auditory Neuropathy (ANSD) In Children

Auditory Neuropathy (ANSD) In Children

Auditory neuropathy spectrum disorder (ANSD) is a rare condition that can affect a person's ability to hear. Although sounds enter the inner ear normally, signals from the inner ear to the brain are not transmitted properly. As a result, the condition may be associated with mild to severe hearing loss and poor speech-perception abilities (difficulty understanding speech clearly). ANSD can be associated with other neurological disorders such as Friedrich ataxia, Stevens-Johnson syndrome, Ehlers-Danlos syndrome, and Charcot-Marie-Tooth syndrome. The exact underlying cause of ANSD is not completely understood; however, researchers have proposed many possible explanations including damage to the hair cells of the inner ears, faulty connections between the hair cells and the auditory nerve (the nerve connecting the inner ear to the brain), damage to the auditory nerve, and/or a combination of these abnormalities. In many cases, ANSD occurs sporadically in people with no family history of the condition; however, the condition does run in some families suggesting that genetic factors may play a role in some cases. Treatment varies based on the severity of the condition but aims to improve hearing (i.e. hearing aids and cochlear implants) and communication skills.

FINAL EXAM – PEDIATRICS

MAY 14TH, 2019 – DEVELOPMENTAL, COGNITIVE, SPEECH AND LANGUAGE MILESTONES
Four main categories that we create milestone tools around are All of these are going to be interacting with each other. Cognitive and social emotions are hard to separate.

- This will happen due to a child's trial and error. A child will observe->theorize predict experiment observe results
- Most important part of our invention is letting parent know milestones
- MOTOR DEVELOPMENT
    - Starts out as then becomes voluntary and controlled

        - Limited movement preliminary neck control more control of torso rolling over sit up
        - Can be broken down further into gross motor, fine motor.
            - There are also red flags (if they haven't done it, if there are different things they're doing, or if there is a persistence)
            - Examples mentioned by Dr. Dunckley:
                - suckling reflex
                - froglegs
                - dominance of a hand way too early
            - SOCIAL COGNITION
                - Cognition is anything related to a mental activity: Thinking, Reasoning, Perceiving the world, Subsections: memory, attention
                - Hierarchy of cognitive development:
                    - Sense stimuli (our concern for auditory comes in here)
                    - Perceive stimuli (our concern for auditory comes in here)
- Make a decision
- Take an action
- Problem Solving is a subset of this.
- Red flags
    - Failure to alert to environmental stimuli. We ask them "Does your kid respond to their name?"
    - Failure to reach for objects
- Absence of babbling
- Absence of localization
- No consonant sounds? Might not be hearing it.
- Lack of imitation

	0-3 months	4- 9 months *TOYS*	9-18 In which age range does the concept of object permanence typically develop?

Active Learning	Forming Pathways Poor motor control; must rely on others for much of their exposure to the environment	-Refining motor skills -Realizes she/he can be an agent -Awareness of cause and effect	Object permanence (if I hide something under a blanket, it doesn't disappear forever) ; means to desired ends
Differentiated actions		Preference for responsive objects (e.g., a rattle) Coordinated actions on objects (e.g., mouthing, throwing)	
Development of differential action schemes			Causality; Symbolic play (pretending something is a phone)
Seek out joint focus	Tracking of objects	Continuous visual contact with adults in the environment	
Communication			Prelinguistic intentional communicative; discovery of intentional communication; development of communicative gestures
Description	□ Early socializing □ Rewarding adults with smiles and coos □ Punishing adults with squirms and cries		

<table>
<tr>
<td>PIAGET'S 4 STAGES
more about the hardwired nature qualities</td>
<td>VYGOTSKY
Much more nurture guy. What is the culture surrounding communication and how does that lead to shared knowledge?</td>
</tr>
<tr>
<td>In the Piagettian tradition, child development is driven by the child. They contain a level of previous knowledge (Assimilated Knowledge) and they fit knew knowledge (Accommodated Knowledge) into their schemata

Sensorimotor Stage
● Random and reflex actions
Preoperational Stage
● Egocentric: "emotional terrorist"
Concrete Operational Stage
● Talk about talking and thinking
● Socialized speech
● Development of logical thought
● Reversibility (turn taking, if this then that and vice versa)
Equilibration = present in all stages
● Children want to learn but things that are not too far outside of their comfort zone
● It is important to maintain a balance between applying previous knowledge (assimilation) and changing behavior to account for new knowledge (accommodation).</td>
<td>Inner Speech – what's going on inside
● Children's speech parallels activity/play
● Becomes a tool for planning, conceptualization
● Transitions to cognitive tool, language becomes though
● Babies can't develop in isolation
o Adults act as a scaffold for learning and development to occur. The rate of things happening depends on adults. (Zone of Proximal Development)

The "distance between the actual developmental level and the level for potential development with scaffolding" is described by what model: Zone of Proximal Development</td>
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THEORY OF MIND

– More recent Rudimentary workings of theory of mind can be seen around 4 years of age. Even from a young age, because of attention, children are developing their own internal world.

- Mental states can cause, explain, and predict others' behaviors.
    - Thoughts, Desires, Intentions
- Infants who do not show attention by 7-9 months of age may be at risk for poor ToM (Baron-Cohen, 1991)
- False-belief tasks
    - Change in Location: Person A thinks Object is in Location A. Person B moves Object to Location B without Person A's knowledge. Question: Where does Person A believe the object to be?
    - Unexpected Contents: Question: What is in the candy box? (Child answers "candy.") Experimenter shows the child there are rocks in the box. Question: What would another person think is in the box?
- Deficits
    - Difficulty determining the intentions of others
    - Lack understanding of how their behavior affects others
    - Difficult time with social reciprocity
    - Less empathic
- Hearing Vs. Deaf Children
    - If you get some kind of language, you're okay. But if you don't have access to language, you're more likely to have deficits.
- SOCIAL DEVELOPMENT

JOINT ATTENTION

- Coordinating attention to an event or object with another individual, sharing interest and social engagement, and showing an understanding that the partner is sharing the same focus
    - 6 months – Shifting attention
    - 8 months – Gaze monitoring
    - 10-12 months – Follow pointing
    - 12-14 months – Protoimperative pointing (request)
    - 14-16 months- Protodeclarative pointing (comment)
- Research suggests that pediatrician's appraisal of a child's developmental status is often inaccurate without the use of standardized screening tools.

DEVELOPMENTAL SCREENING AND SURVEILLANCE:

assess issues as soon as possible

- Developmental and behavioral problems are estimated to be present in 12%-16% of US children. [development and behavioral will have parent interviews. Parent knows the child best]
- Only 20-30% of children with developmental or behavioral problems are detected prior to school age.

- Early Intervention has shown to be effective : Improving long-term educational and vocational outcomes for children who have developmental or behavioral disabilities, Preventing teen pregnancy, preventing criminality
    - Screening is a brief assessment procedure designed to identify children who should receive more intensive diagnosis or assessment.
    - A variety of validated, parent-report, developmental screening tools for the early identification of developmental delays are now available
    - Increase in referrals, low return of ASQ questionnaires, physician referrals 42

FINAL EXAM – PEDIATRICS

MAY 21ST, 2018 – LANGUAGE DEVELOPMENT RECEPTIVE AND EXPRESSIVE LANGUAGE DEVELOPMENT

A child's ability to parse words into sound units, or combine phonemes into words are dependent upon the child's: phonological awareness

- LANGUAGE DOMAINS

PHONETICS	PHONOLOGY	MORPHOLOGY	SYNTAX	SEMANTICS

the physical properties of speech sounds (phones), and their physiological production Phoneme: smallest segmental unit of sound employed to form meaningful contrasts All phonemes can be described in terms of: Place Manner Voice	the way sounds function in languages, including phonemes, syllable structure, stress, accent, intonation The way sounds are distinctive units within a language	the identification, analysis and description of the structure of words Morpheme: the smallest linguistic unit that has semantic meaning	the principles and rules for constructing sentences in natural languages	how language conveys meaning

PRAGMATICS: Society's rules for language
1. Pay attention to another person's language utterance. 2. Try best to understand what the speaker's intentions or desires are. 3. Do your best to respond to those intentions cooperatively and constructively.

- THEORIES OF LANGUAGE DEVELOPMENT

Principles and Parameters Theory	Connectionist – kids are constantly figuring out what the rules are

● Innate capability for language ● Through exposure, child learns which language model to follow [exposure is key] ● Different cognitive parameters are set and become fixed	● Child derives rules of language through repeated exposure to similar constructions ● Rules can be derived out of these multiple exposures

ROLE OF PARENTS

- People talk differently to babies – an innate sense that things need to be positive and simple.
    - CHILD-DIRECTED SPEECH (aka “motherease”) when mapped, it seems to be a lot clearer
    - Responsive to needs – “Aw baby, why are you crying? Are you wet?”
    - Language is learned against the backdrop of human interaction beginning with two basic behaviors:
    - In turn, Communication with infants is driven to support interaction. Adults try to establish and engage in joint attention and turn taking.
        - JOINT ATTENTION
            - By 9 months of age, Adults put selves or other objects in the baby's line-of-sight to be the object of attention or adults attend to objects currently in the baby's line-of-sight.
            - Repeated interactions offering intensive language learning opportunities
        - Peek-a-boo , Chasing, Toys/games, Naptime/bedtime routines, Mealtime routines, Early receptive vocabulary

Safety words No-no! Hot! Dirty!	Politeness words Please. Thank you.	Survival Words
Substantive words Milk. Mommy. More	Expressive words Oops! Uh-oh! Where!	

- RECIPROCITY/TURN TAKING – very core of communication
    - Adults: Do something Wait
    - Children learn: to fill turn to allow partner’s turn
- BABY’S COMMUNICATION PROGRESS

- Baby's intentional communicative acts increase
- Parents respond to the communicative intent
- Parents use more words during interaction. Parents "up the ante" – parents who are talked to more, have higher literacy rates.
    - Naming things, actions, and states that are meaningful to the interaction
    - Improvements of children's utterances
        - Accuracy
        - Complexity
        - Appropriateness
- COMMUNICATIVE INTENT
- Before 9 months, babies are reactive and interactive, but not intentional.
- 9-12 months of age, children realize their actions cause an adult's reaction.
- Not necessarily linguistic but leads to communicative gesture
    - Pointing, Reaching, Open-hand request
- Gesture
- Contact gestures: giving, showing
- Distal gestures: pointing (very significant predictor of language), waving
- Order: giving, pushing away, raising arms, showing, reaching, waving, pointing, shaking head "no", nodding "yes"

Communication functions	Communication means
⚫ Attention (to self, others, objects) ⚫ Request (objects, actions, information) ⚫ Greets ⚫ Protesting/rejecting ⚫ Responding/acknowledging ⚫ Commenting/informing	⚫ Gestural ⚫ Vocal ⚫ Verbal ⚫ Combinations Rate: 2.5/ min; whether vocal, gestural, verbal, and combinations

- SPOKEN LANGUAGE – What age do we expect true words? 12 months
- Around 10-12 months parents "feed" children words: Picture books, Constant naming of things, actions, & states
- Hart and Risley (1995)
    - Children in lower SES heard far fewer words in at-home interactions than children in higher SES.
- REPAIR TECHNIQUES
- Amending: correcting a child's utterance. "I gots two pennies." "No, you've got three pennies. You try. Say, 'I've got three pennies.'"

- Expanding: modeling an expanded form, higher level of grammar. “Horsey run.” “Yes, the horse is running. The horse is running quickly in the field.”
- ROLE OF CHILDREN
- Babies’ innate behavior allows them to learn from people and things in their environment.
- Knowledge of the here and now precedes comprehension or production of the language used to refer to the here and now.

Specific strategies of babies	Specific strategies of toddlers
• Paying attention to the subject of discussion • Listening selectively to what adults say • Providing feedback that indicates whether or not the message was understood	• Selective imitation of what others say • Using words to learn more words

- MASTERY MOTIVATION
- Children seem to have an innate desire to master their environments and will work hard to do so.
- PRENATAL LANGUAGE EXPOSURE
- Babies have had linguistic exposure from in-utero. At birth, they already recognize their mother’s voice.
- STATISTICAL LEARNING: after massive hours of exposure, some things are more common than others.
- Phonotactic probability
    - Certain phonetic sequences are more common than others within a language.
        - g., more likely to hear /…gz…/ than /…l zg…/
        - “dogs” vs “daddy’s girl”
    - Infants as young as 8 months are sensitive to these statistical particularities of the native language (Saffran, 1996)
    - Multiple perceptual modalities
    - Synchronicity of perceptual events creates stronger connections between those particular events
- CHILD’S GENERAL LEARNING STRATEGIES
- Attend to and act on the people and objects in the environment
- Observe, listen, and learn from other people
- Imitation- 7m old: children can imitate without seeing selves
- Seeking and taking instruction= Adults may scaffold, set up a situation so child only has to do a small action to achieve success

- Explore and experiment via play

LANGUAGE COMPREHENSION STRATEGIES

- Attend to the common focus of an interaction
- Listen selectively to adults' language input
- Provide feedback to adults regarding their messages
- LANGUAGE & SPEECH: PHONOLOGY
  - Adds word shapes (no longer just CV)
  - Adds consonants that can be used to "keep words separate" (e.g., sun, fun, done, run, ton, pun, nun, bun, gun)
  - "Sheds" common simplification processes
- SPEECH/PHONOLOGY
  - Know:
    - Intelligibility levels (with unfamiliar listener)
      - 24 months ≅ 50% intelligible
      - 36 months ≅ 75% intelligible
      - 48 months ≅ 100% intelligible
    - Consonant place/manner growth patterns
    - Age when common phonological processes are "gone" (suppressed)
- NOT ALL EARLY WORDS ARE CREATED EQUAL WHEN IT COMES TO PHONOLOGY
  - Level 1 words: words with NO true consonants (i.e., Vs only, or Cs limited to glides and glottals)
  - Level 2 words: words with ONE true consonant
  - Level 3 words: words with TWO true consonants that differ by place and/or manner
- LANGUAGE PRODUCTION STRATEGIES
- Imitate adult language selectively – Another marker for language development
- Use language to learn language
- Wh- Questions
- Hypothesis testing
- Evocative utterances – a lot of emotional attachment
- DEVELOPMENTAL STAGES – some things will improve. Some things will drop off. We can use these milestones to lead to development of specific scales.

What is the order of speech milestones? (4) (1) cooing (2) vocal play (3) canonical babble (4) jargon

- BABBLING STAGE
  - Phonation/reflexive babble

0-2 months	Cry and fussing	Vegetative sounds

STAGE 1: Cooing/laughter

- 2-4 months
- Usually velar, separated by glottal stops

STAGE 2: Vocal play

Addition of trills, raspberries	Changes in intensity, pitch	4.-7. months

STAGE 3: Canonical babbling – more repetitive with some easy consonants ; Failure to enter canonical babble stage indicative of possible language delay

- 7-10 months
- Reduplicative (mama, gaga)
- Non Reduplicative
- “contoids” [d,b,m,t,g,w] and “vocoids”
- CV sequences with rapid formant transitions between C & V (120 msec)

Variegated babble – much more repetition and more prosody ● 10-12 months ● Simple forms (CV, V, VCV) ● Low prosody	STAGE 4 Jargon – all prosody with no content ● 12-14 months ● Highly inflected, but lacking content or grammar

- Assessment
  - Observe infant-caregiver interaction for: affect, responsiveness, reciprocity, appropriateness, encouragement of joint attention, language stimulation
  - Broad developmental scales: Batelle, Bayley, Denver (Pediatric practices),
  - Communication-specific scales:
    - Rossetti (0-36 mo)
    - PLS4 (0-6;11)
    - MCDI
- ONE-WORD STAGE
- Substantive form. Labels for objects or actions “Mama,” “bang”
- Functional/relational form
  - Relationship or state of object/action
  - “mine,” “bye-bye”
- TWO-WORD STAGE
- ~24 months of age children begin putting two words together.
- How does Word 1 relate to Word 2?

TYPICAL SYNTAX

- Mean Length of Utterance (MLU): A “global” measure of syntactic growth

2 years -2-word sentences -Noun-verb combinations -Introduction of morphological markers (tense, number, possession)	3 years -Three word sentences -Verbs begin to predominate ~75% intelligible -Understands most simple questions dealing with his environment and activities “what do you do when you are sleepy, hungry, or thirsty?“ -Morphosyntactic system relatively figured out -Can make more complex sentences, 2-clauses	4 years -Demonstrates understanding of over and under -Understands such concepts as longer, larger, when a contrast is presented	5 years -Can use many descriptive words spontaneously-both adjectives and adverbs -Should use fairly long sentences and should use some compound and some complex sentences -Speech on the whole should be grammatically correct
8 years -Complex & compound sentences used easily with few lapses in grammatical constructions-tense, pronouns, plurals			

- Telegraphic stage

PRAGMATIC DEVELOPMENT 12 months old

⚫ Use of gestures, vocalizations, and a few “true” words to: ⚫ Request objects/actions ⚫ Refuse/protest ⚫ Comment ⚫ Play communicative games	Primitive speech acts ⚫ Request action ⚫ Request attention ⚫ Answer ⚫ Request answer ⚫ Protest ⚫ Greet ⚫ Call ⚫ Label ⚫ Repeat ⚫ Practice

PRAGMATIC DEVELOPMENT 36 months old

⚫ Use of language to: ⚫ Replace gestures of 12 month old stage ⚫ Requesting information ⚫ Answering questions ⚫ Acknowledging ⚫ Asking permission ⚫ Requesting indirectly	⚫ Talks beyond the “here-and-now” ⚫ Tells little stories ⚫ Has a conversation ⚫ Reciprocity, turn taking ⚫ Says more than one utterance per turn ⚫ Rate of intentions increases

- Awareness of social distance
  - To friend: “Turn on the TV”
  - To my friend's mom: “May we please watch TV?”
- Developing request strategies
  - Telegraphic directive [1;3-2;0]: “Gimme that”
  - Oblique strategy [4;0+]: “We haven’t had candy in a long time”

PLAY DEVELOPMENT

• ⚫ 12 months old • ⚫ Presymbolic Schemes • ⚫ Conventional use of a few objects; no pretending • ⚫ picks up a brush, touches to hair, then drops it; • ⚫ picks up toy phone, puts to ear, then sets aside)	• ⚫ 36 months old • ⚫ Hierarchical pretend schemes • ⚫ “Feeds” baby doll, kisses, puts to bed and covers with cloth • ⚫ Carries out a tea party with dolls, stuffed animals • ⚫ Sets up and carries out story “scenarios” with characters and props

LANGUAGE DISORDERS- The concept of “normalization”

- Later = delay
- Outside norm = disorder
  - We need to know the difference because it will tell us how to approach intervention.

PREVALENCE OF CHILD LANGUAGE AND RELATED DISORDERS: HIGH INCIDENCE

- SLI: 7.4 % of all kindergarten children
- Learning Disabilities: 5 -10 % (+)
- Mental retardation: 2%
- Autistic Spectrum Disorder: 1 in 110
- Reading: Approximately 25% of the nation’s secondary students read and/or write below a basic level

- Reading in children with language disorder: 50% read below a basic level

LATE TALKERS

- Toddlers whose deficits appear to be primarily confined to language
- Criteria for LT
  - < 10th % on McArthur-Bates Communicative Development Inventory (MCDI)
  - at age 24 mo, < 50 words and no 2-wd combinations in spontaneous speech
  - can be expressive only or both receptive and expressive
  - normal range performance on sensory, motor, and nonverbal cognitive measures
  - goes hand-in-hand with decreased speech sound development
- Prevalence of LT: 15% (10% – 18% depending on source)

CLINICAL PRESENTATION

- “(my toddler) is not talking very much”
- Sizeable # of EI referrals
- No-man's land for parents in terms of physicians, internet
- Caregiver questions:
  - Why isn’t my child talking?
  - What should I do to help my child?
  - Will my child grow out of this

LATE TALKER OR LATE BLOOMER ?

- ~50% of LT move into normal range for language by 36-42 mo, becoming “late bloomers” (50% normalization rate)
- ~75% of LT “normalize” for speech by age 6
- After vocabulary moves into normal range, children have a higher risk for production delays in phonology, morphology, syntax, and narrative abilities.
- Compared to the control group, they are “in the normal range” but still significantly below age peers.
- How do we manage these children in this period between 24 – 48 mo?

PRELINGUISTIC/RISK/PREMATURITY

⚫ Prematurity: ⚫ <37 weeks GA ⚫ 12% of all babies ⚫ In 70's a baby < 3 lbs had a 50% chance of surviving; today 90% ⚫ Low Birth Weight: ⚫ 1500 – 2500 grams ⚫ Very Low Birth Weight: ⚫ 1000 – 1500 grams ⚫ Extremely Low Birth Weight: ⚫ < 1000 grams ⚫ 1 lb = grams	⚫ Why do VLBW and ELBW have neurodevelopmental problems? ⚫ What is a neurodevelopmental problem and how is one determined by physicians? ⚫ One recent study compared ELBW and term children at age 3: found differences in most language domains of 1 SD

BILINGUALISM

Common "Errors" in ESL children

- Prepositions
- Multipurpose verbs
- Grammar and Syntax
- Vocabulary and Semantics

A disorder is considered present if errors outside of expected patterns occur, and only if they occur in BOTH languages

CLINICAL MANAGEMENT: CHOICES THAT THE AUDIOLOGIST NEEDS TO KNOW ABOUT

- wait and see
- watch and see
- intervention (perhaps guided by risk factors)
- As an audiologist, what would you Rx and why?

SEMANTICS: Vocabulary DEVELOPMENT/ Types of words

- Qualitative shift in types of words learned:
    - Low frequency words

Abstract words (e.g., can't point to them)

Derived words (derivational suffixes and prefixes)

- A "literate" lexicon

Polysemous words (multiple meanings)

Adverbs of likelihood and magnitude

Cognitive verbs Connectives (adverbial conjuncts) Curriculum-specific words

Metalinguistic and metacognitive verbs

- Metalinguistic development
- First grade: students asked to identify words in a sentence that sound the same but look different, or identify the word in a sentence that tells how an agent performed some action.

        - “He saw the man’s son standing in the sun”
        - “The pig walked away slowly”
    - By second grade, children laugh at jokes whose humor stems from phonological, lexical, or syntactic ambiguity.
    - Vocabulary growth
    - Age 7 years, children begin to use dictionary
    - Increased number of abstract notions added to vocabulary…
        - prophetic, circumstantial, officialdom
    - Mirrors increased abstractions in cognition
        - Santa Claus changes from a real man to a symbol of the Christmas spirit
    - Seventh grade: students asked to determine meanings of unfamiliar compound words.
        - Yachtsman, landfall, etc.
    - Development of word definition ability
        - More words can be defined
        - Accuracy of definition increases
        - For 9-14 year olds, most accurate definitions are synonyms
        - Increased placement of objects into superordinate categories (e.g., “A ruby is a type of gemstone”)
        - Increased awareness of boundaries between semantically-related categories (e.g., cup – glass)

MULTIPLE MEANINGS ASCRIBED TO SAME WORD. ABSTRACT ONES LEARNED LATER. “THE BOY IS BRIGHT.”

- Understanding of increasingly complex verb forms develops through college age, and may never be mastered by some adults.
- Literate verbs (hypothesize, concede) – used in discussions of spoken and written language interpretation
- Factive (know, forgot) – presupposes truth of complement (“He forgot I live in Chicago”)
- Non Factive (think, believe) – cannot be sure the complement is true (“I think the car has gas”)
- Reading development: Stages
- Learning to read (K – 3rd grade):
    - Decoding skill determines reading skill
    - With practice, letter patterns (orthography) become automatically recognized (e.g., light, right, right)
- Reading to learn (3rd grade +)
    - Reading is “good enough” that it is now a tool for learning
    - Reading skill is determined by decoding fluency (accuracy and rate) AND general comprehension abilities
- Phonological awareness

- Norms: Kindergarteners (5-6 y)
    - Know the names and sounds of most letters (names are slightly earlier)
    - Detect and generate rhyme
    - Tell whether two words “start” with the same sound (pig, house, pan, sun)
    - Write their names, and write other short words and notes
- How do we test Phonological Awareness?
    - Remove a phoneme from a word
    - Blend phonemes said separately and slowly back together to make a word
    - Say phonemes of a word separately
    - Match words on the basis of phonemes
    - Switch/reverse phonemes (say it backwards)
    - Make phoneme “substitutions”
- Can use felt squares or cards to represent parts of a word on the table.
- Literacy development
- Phonology
    - Steeper growth at earlier grades
- Orthography
    - Steeper growth at earlier grades
- Morphology
    - Steeper growth at earlier grades
- Birth to 3 literacy accomplishments
- Recognizes specific books by cover
- Pretends to read books
- Understands books are handled in particular ways
- Enters into book-sharing routine with primary caregivers & listens to stories
- Birth to 3 literacy accomplishments
- Begins to attend to print (e.g., letters in names)
- Purposeful scribbling – may produce some letter-like forms
- Labels objects in books
- Comments on characters in books
- Components of reading (Whitehurst & Lonigan)
- Outside-in: sight words
    - Sources from outside the printed word that directly support understanding of the meaning
        - Vocabulary, conceptual knowledge, story schema
    - Inside-out: sounding out
        - Sources of information within the printed word that support translation between print and sounds

        - Phonemic awareness, letter knowledge
- READING = Decoding + General comprehension
- Decoding/ Word recognition
- Using the visual representation of a word to access the word's pronunciation (and meaning)
- Requires CONSCIOUS PHONEMIC AWARENESS: The awareness that words can be decomposed into more basic elements – namely sounds (phonemes). If you can't do this, you have nothing to "map" letters onto.
- Hard for 20% of all children
- READING = Decoding + General comprehension
- Comprehension
- Once the word (meaning) has been accessed (via it's pronunciation) the same higher order comprehension processes "kick in"
- Literacy development
- Phonological processing skills are used to decode the written word
    - Sensitivity: ability to detect and manipulate sound structure of oral language
    - Memory: short-term memory for sound-based information
    - Naming: efficiency of retrieval of phonological information from permanent memory
- Literacy development
- Print awareness/letter knowledge
    - Letter discrimination, Letter recognition, Letter-sound relationship
- Emergent writing
- Oral language skills
    - Young readers with larger vocabularies have greater phonological sensitivity
    - In older children, better language supports reading which supports language development
    - Sentence length: Developmental benchmarks (Loban, 1976)
- Encouraging emergent literacy
- Shared reading
    - Vocabulary development
    - Print exposure
- Spoken conversations (e.g., family dinners)
    - Decontextualized language development
- Interventions for emergent literacy
- Dialogic reading
    - Child assumes role of storyteller

- Adult scaffolds child's role using questions
    - What color is that? Have you seen a duck like that before?
- Phonological sensitivity training
    - Exercise for phonemic analysis
        - Categorize words by first letter, final letter, etc.
    - Morphology and literacy
    - Relationship between phonological awareness and literacy well established, especially in younger children.
    - Morphological awareness is important for literacy in older children.

As children advance through school they are exposed to more complex vocabulary – words constructed from multiple morphemes.

- Morphological awareness most strongly related to vocabulary at 4th – 5th
- Morphological awareness most strongly related to spelling at 6th – 7th

FINAL EXAM – PEDIATRICS : MAY 28th , 2019 – CASE HISTORY

- Why NOT to skip:
    - Opportunity to build rapport with family, and observe family dynamics
    - Gives child time to adjust to surroundings
    - Guides focus of assessment
    - Gives picture of cognitive/developmental status of child
    - Foundation for counseling following evaluation
- First Question: Why have you brought your child here today?
    - Do the answers match up with the information gleaned from the chart (if available)?
- Best case scenario for testing : objective, subjective, and ear specific.
- What children would not be able to pass their NBHS? those w/ congenital aural atresia
- According to the American Academy of Pediatrics, what is an appropriate refer rate for a newborn hearing screening program? Less than 4%

MAY 28th , 2019 – IMMITTANCE

Infants younger than 7 months of age should be tested using a 1000 Hz probe tone because their middle ear system is: mass-dominated
Middle ear acoustic reflex testing may be improved in infants by using a _____ _____ rather than a probe tone wideband signal

COMPONENTS OF CASE HISTORY PAGE 50-51 OF MADELL/FLEXER CHAPTER

<table>
<tr>
<td colspan="2">Prenatal and birth history
● Complications, Low birth weight, NICU stay, Jaundice, Mother's age
Maternal illness – TORCH
⚫ Toxoplasmosis: 10-20% have SNHL
⚫ Others: Syphilis (3% have SNHL); Varicella-Zoster Virus (few have SNHL)
⚫ Rubella: >65% have hearing loss when rubella acquired in first 4 months of pregnancy
⚫ Cytomegalovirus: 10-20% have progressive SNHL
⚫ Herpes simplex: 25-90% depending on degree of virus dissemination
Rh Incompatibility
ABO Blood incompatibility
⚫ Apgar Scores
⚫ Apgar scores and SNHL
Appearance, Pulse, Grimace, Activity, Respiration
⚫ Utah criteria: (Eichwald & Mahoney, 1993)
⚫ 0-4 at 1 minute
⚫ 0-6 at 5 minutes
⚫ Represents greater risk for SNHL
Possible relation to perinatal hypoxia
Perinatal Period: Jaundice , NICU length, infections, syndromes, feeding issues, respiratory diseases</td>
<td>Developmental history
⚫ Psychosocial
⚫ Developmental
⚫ Evaluations:
physical therapy
occupational therapy
speech-language pathology</td>
</tr>
<tr>
<td>Educational history (special services)

⚫ Will results of today's test affect child's current placement?
⚫ Is child receiving special services in school and/or outside of school?</td>
<td>Health history
⚫ Genetic evaluation/referral
⚫ Surgeries?
⚫ Head injuries/falls?
⚫ Specific Diagnoses</td>
<td>Communication history
⚫ How would you describe your child's hearing?
⚫ Expressive language
⚫ How does your child communicate his needs?
⚫ Verbal (I'm hungry)
⚫ Gesture (pointing)
⚫ Other (taking parent by hand and dragging to fridge)
⚫ Receptive language
⚫ How well does your child understand you?
⚫ Simple commands (Say "thank you"; Go get your shoes)
⚫ Relative to other children the same age</td>
</tr>
</table>

Social History -Do they play with other children? -What kind of play do they do? -Do they have siblings? Are they developing similarly?		Hearing Health History -Family history of hearing loss -Family history of congenital malformations -How are they doing in school? -Noise exposure? -Behavioral changes -ear infections? Sick a lot?	

CLASS PRESENTATIONS				
VESTIBULAR IMPAIRMENT IN CHILDREN WITH COCHLEAR IMPLANTS (MALINASKY AND KOCON)				
-50-70% of children with hearing loss have VI -VI correlates to etiology, not severity of hearing loss	3 main etiologies cause both hearing loss and vestibular impairment ○ Congenital Cytomegalovirus ○ Inner ear malformation ○ Syndromic hearing loss		Classified vestibular functioning into 3 grades: 1: Normal 2: Partial dysfunction 3: severe dysfunction and areflexia (absence of calorics	66% of patients with Usher syndrome, meningitis, CMV, and inner ear malformations had abnormal cVEMPs1 • 58.3% of implanted children showed a decrease in cVEMPs2

Results: Depending on the etiology, children had worse vestibular functioning post-implantation when tested with calorics ○ 50% of the participants had an unknown etiology ○ 50% had either Usher Syndrome, inner ear malformations or Connexin 26	inner ear malformations lead to a higher risk of VI post-implantation			Best practice is to test calorics, posturography and VEMPs pre- and post-operatively in children – Can give insight into post-implant functioning – Can allow for better rehabilitation post-implantation
VESTIBULAR DYSFUNCTION AND DYSLEXIA (BARRY AND COPPOLA) – Auditory Neuropathy (ANSD) in Children				

Could central vestibular dysfunction be an underlying mechanism leading to learning disabilities such as dyslexia?	RESULTS FROM ARTICLES Significant symptomatic overlap within subject ● Learning disabilities are likely all linked to cerebellar vestibular dysfunction ○ One underlying mechanism for all learning disabilities Learning disabled patients had significantly different OPKs from the control group ● Learning disabilities and dyslexia appear to have the same underlying CV mechanism that leads to ADD ● Concentration plays a vital role in compensating for CV dysfunction -Patients with dyslexia employed concentration strategies to maintain ocular fixation ● Patients with dyslexia reported background blurring during testing, whereas control group did not Selective interaction between vestibular exercises, vestibular dysfunction and spatial perception ● Control group: ○ Improved in speed of both visual and spatial perceptual processing ● Abnormal VOR/Aerobic exercise: ○ Improved only in speed of general visual perceptual processing ● Abnormal VOR/Vestibular exercise: ○ Made significant improvements in spatial perceptual processing		CONCLUSIONS Cerebellar-vestibular dysfunction can result in a vast array of symptoms, including learning disabilities like dyslexia ● Vestibular tests are an important part of the test battery in identifying and treating learning disabilities ● Vestibular compensation exercises can help patients deal with the symptoms of the CV dysfunction, including dyslexia and other learning disabilities
HIV – (HAGER AND HUGHES)			□

Antiretroviral therapy (ART) is the standard treatment for HIV-infection for the pediatric population. ● Highly active antiretroviral therapy (HAART) is simply a combination of ART drugs for multiple viral targets.	Hearing loss in perinatally HIV-infected and HIV-exposed but uninfected children and adolescents. – Children exposed to HIV are more likely to qualify for audiometric screenings ○ Children exposed to antiretroviral in utero are more likely to qualify for an audiometric evaluation ○ Children with CDC Class C diagnosis are more likely to have hearing loss ○ PTA is higher in children with HIV “Hearing loss in HIV- infected children in Lilongwe, Malawi.” ○ 24% of children with HIV had CHL ○ 5.5% of children required hearing aids ○ There were higher instances of parental perception of hearing loss and documentation of hearing problems for those in WHO stage 3 or 4 Audiologic and vestibular findings in a sample of Human Immunodeficiency Virus type-1-infected Mexican children under highly active antiretroviral therapy Consistent abnormal ABR results between adults and children with HIV ○ HIV increases the chance of otitis media ○ Children with HIV had a lower amplitude and prolonged I-V interwave latency in ABR testing ○ HAART therapy did not play a role in hearing loss ○ Hearing loss could still present in asymptomatic children with HIV	CONCLUSIONS Children in the more severe stages of HIV-infection are more likely to have hearing loss ● Of those children who had a hearing loss, conductive losses were the most prevalent ● PTA is likely to be higher in children with HIV ○ Due to more cases of hearing loss ● 5.5% of children required hearing aids (Hrapcak, et al.) ○ (Where we can help!) ● Abnormal ABR results consistently found with children with HIV ○ Lower amplitudes and prolonged I-V interwave latencies ● Increased likelihood of recurrent otitis media ● Hearing loss could possibly be present in asymptomatic children with HIV
CARDIAC SURGERY IN INFANCY AS A RD (MOTT & SCHNEIDER)		

Is cardiac surgery the risk factor, or is it some aspect of undergoing cardiac surgery?	Article 1: Prevalence of 6% is significantly high ○ When accounting for syndromes, prevalence of 4% is still significantly high ○ Remember: no consideration of ototoxic drug us Article 2: Increased risk for hearing loss in children with CHD ○ Children who undergo cardiac surgery at 6 months of age or younger should have at least one audiologic evaluation by 24-30 months to evaluate hearing status Article 3: Unexpected association of PHL with hyposix and bolus administration of furosemide ○ Close follow-up is necessary to identify outcomes and seek modifiable predictive variables ○ Changes in the mode of furosemide administration may prevent this complication		This is an important start to understanding a complicated topic!! ● 3 articles with 3 approaches ○ All showed a correlation between cardiac surgery and hearing loss ● Evaluate all children who undergo cardiac surgery in infancy ● More research is necessary	
LANGUAGE ACQUISITION FOR PRELINGUAL				5
Spoken language development is negatively affected by delaying access to linguistic input until auditory input is initiated through hearing aids and/or CIs ● Bilingual-bimodal approach emphasizes the use of ASL and acquiring spoken language through the use of assistive technology			Conclusions: Improvements in identification of hearing loss and assistive technology leaves a need to improve and update the model of instruction and acquisition of language ● Even with assistive technology, language deprivation is still a problem leaving ASL and manual communication necessary for robust language development	

		● Further research is necessary to develop a formal methodology for the implementation of a Bi-Bi program	
HYPERBILIRUBINEMIA (PLUARD AND KIM) Affects ~60% of infants			
Risk for hyperbilirubinemia when levels >18 mg/dl. Excessive levels of bilirubin in the blood Lesion in brainstem = hearing loss Lesion in oculomotor = upward gaze Unconjugated bilirubin (UCB) is a breakdown product of red blood cells (hemoglobin) that is toxic to the brain in its free form UCB processed by liver into conjugated bilirubin and pooped out	Hyperbilirubinemia is a major risk factor for ANSD • Initiates cell apoptosis and causes damage to auditory structures including cochlear nuclei in the brainstem, the auditory nerve, and higher order neurons • ANSD is a neuropathy caused by dys-synchronous firing of auditory nerve fibers where sounds enter the inner ear normally, but signals from the inner ear to the brain are transmitted abnormally. Auditory Neuropathy (ANSD) in Children	Those diagnosed with ANSD will have predicatable audiometric results which include:: • Degree of hearing loss can vary from normal to severe • Reflexes will usually be absent • Present OAEs • Abnormal ABR results • Speech recognition scores will be worse than predicted by pure-tones	
	TREATMENT Exchange transfusion – Albumin infusion – Blue light phototherapy – CI may offer synchronous firing of auditory nerve fibers Visual Language Learning – Cued speech, ASL, SEE – Low gain amplification – Low expectations, but not without benefit – If OAE/ CM present, monitor closely – Should still consider a trial! – Cochlear Implant – If patient has very poor WR – Unsuccessful trial with amplification	Conclusions: Language acquisition is key in neonates Some will have sound awareness, but complex speech sounds will be distorted & indiscriminate Early diagnosis & intervention! – Parents will often be fooled that their child has normal hearing	
PEDIATRIC HEARING LOSS: refer siblings. They could be a carrier or it may be recessive. If the child has risk factors, we want to keep them on our assessment schedule.			

⚫ Risk Indicators for permanent congenital, delayed onset or progressive hearing loss: Caregiver concerns about hearing, speech, language, development ⚫ Family history* of permanent childhood hearing loss ⚫ NICU stay > 5 days or any of following (regardless of length of stay): ECMO assisted ventilation* ⚫ Ototoxic medications (gentimycin, tobramycin) ⚫ Loop diuretics (furosemide, Lasix) ⚫ Hyperbilirubinemia reguiring exchange transfusion ⚫ In Utero infections (cmv*, herpes, rubella, syphillis, toxoplasmosis ⚫ Craniofacial anomalies ⚫ Physical findings (e.g. white forelock)	⚫ Syndromes* involving hearing loss ⚫ Neurofibromatosis, osteopetrosis, Usher, Waardenburg, Alport, Pendred, Jervell & Lange-Nielson ⚫ Neurodegenerative disorders ⚫ Hunter syndrome ⚫ Sensory motor neuropathies (Frieidrich ataxia, Charcot-Marie-Tooth) ⚫ Culture positive postnatal infections associated with HL* ⚫ Herpes, varicella, meningitis ⚫ Head trauma (basal skull, temporal bone)* ⚫ Chemotherapy*

PIERRE ROBIN ⚫ Micrognathia ⚫ Glossoptosis ⚫ Upper airway obstruction ⚫ Cleft palate (U-shaped) ⚫ Gene ⚫ 17q24.3 – q25.1 ⚫ Associated with ⚫ Stickler (STL1; 12q13.11)	TREACHER COLLINS [Dominant] ⚫ Minor clinical features ⚫ Preauricular hair displacement (26%) ⚫ Atresia or stenosis of EAC (36%) ⚫ Conductive hearing loss (40-50%) ⚫ Ankylosis, hypoplasia or aplasia of ossicles ⚫ Ophthalmologic defects ⚫ Cleft palate ⚫ Airway abnormalities ⚫ Speech and motor development delays		PERILYMPHATIC FISTULA SNHL (sudden, progressive) Presence of cochlear malformation	ENGLARGED LARGE VESTIBULAR AQUEDUCT May not cause hearing loss but is probably comorbid with hearing loss.

MENINGITIS – Viral, bacterial infection of CSF – Cochlear implants soon	NF2 Most common are vestibular schwannoma”Balance disorders, Hearing loss, Tinnitus, Facial paralysis	WAARDENBURG SYNDROME Type II associated with deafness Partial albinism	USHER SYNDROME 3 types, different degrees of vision and hearing loss. CI helps
CONDUCTIVE ⚫ Apert Syndrome; Crouzon Syndrome; Pfeiffer syndrome ⚫ Craniosynostosis ⚫ Velocardiofacial (VCF) Syndrome ⚫ Hemifacial microsomia ⚫ Asymmetric skull development ⚫ Nager syndrome ⚫ Cleft palate, coloboma, absent eyelashes, dysmorphic ears, short forearms, clinodactyly/syndactyly	1.Screen by 1 month of age. 2. If they don’t pass, identify by 3 months of age. 3. Confirmed hearing loss? Intervention (fit) by 6 months. Auditory Neuropathy (ANSD) in Children		

Auditory Neuropathy (ANSD) In Children

IMMITTANCE			
Admittance = Y (mmho). The vector addition of susceptance and conductance. The middle ear is the impedance matching system between the outer and the inner ear.			
Infants less than 6 months of age do not have a lot of stiffness in their system (think of the cartilage, lack of ossification)	B and G curve Separate the two vectors Adults = there are distinct patterns. They refer to the number of peaks identified. Different tones = different patterns. As we move up in frequency, 70% are 3B1G When we look at babies, it’s a lot more variable.		

PROBE TONE Why doe we use 226 Hz? It's easier to interpret one peak. Real reason = if we use 226Hz, then it's easy to come up with your ear canal volume. G becomes 0 so Y and B are the same. 226 Hz probe tone – most humans have 1B1G pattern. Some infants have more complicated patterns 1000 Hz probe tone – most adults have 3B1G pattern. Children have various patterns. To get something close to 1B1G, it's better to use 1000 Hz. 7 months is the cut off. Anything below that, you should use 1000 Hz. □ Higher frequency probe tone more sensitive to ME effusion		Post Natal Changes to Ear □ Increased bony portion, decreased cartilaginous portion □ Increased size of ear canal, ME space □ Decreased tympanic membrane tilt □ Fusion of tympanic ring □ Decreased ossicular density □ Increased stiffening of ossicular joints □ Closer coupling of stapes to oval window □ Mesenchymal absorption
WIDEBAND REFLECTANCE		
Measure admittance of middle ear system of all frequencies. You use a chirp as your measurement.		
		□

Auditory Neuropathy (ANSD) In Children : UNHS & OAEs – JUNE 4th

- Otoacoustic emissions are thought to arise primarily from the somatic motility of the outer hair cell. What other mechanical response of the OHCs is hypothesized to contribute to the cochlear amplifier? Stereocilia bundle motor
- Two criterion used clinically to identify a present/absent TEOAE (1) TE-noise floor (SNR ≥6 dB) (2) reproducibility (70%)
- Two criterion used clinically to identify a present/absent DPOAE. (1) SNR ≥6 dB (2) Absolute amplitude: >-5 dB SPL
- What kind of stimuli is better for identifying hearing loss using DPOAE? mid-level (65 dB, high level is 75 dB)
- 3 factors when comparing OAE to published norms: Above 10th, below 90th

- Why do TEOAEs and DPOAEs?
- OAES won't rule out auditory neuropathy.
- OAEs are generally larger in newborns, but there are higher noise levels.
- "Present but abnormal" doesn't work at Rush.
- Why 6 dB for noise ratio?
    - Boys Town research
    - If it's greater than 6 dB, you are more likely to have a threshold that is 20 dB HL or better.
    - If your hearing is worse than 20 dB HL, most likely your TEOAE/Noise is < 6 dB
- DPOAEs and TEOAEs are coming from multiple contributions. They can cancel each other out or add together.
    - Because of fine structure, you can't say a person doesn't have OAEs based on one frequency. Presence of fine structure means that a Pass/Refer decision should NEVER be based on a single frequency
    - If even one is absent, you can still say they have present OAEs.
- When we calibrate, we are looking for a reference. Why are we looking for this? Because the system is not alive. Auditory Neuropathy (ANSD) in Children
- Distortion Product: 2f1-f2 is largest distortion product otoacoustic emission in humans
- DPOAEs are more place specific. Where are we stimulating in the cochlea along the basilar membrane.
- If you play two tones close enough in frequency, most people perceive a third tone. That's because of intermodulation distortion along the basilar membrane. Those two tones have to be played to the same ear.
- Discovered around 1978.
- 2f1-f2 is what we are measuring, but we vary the tones we are playing.
- As each pair of tones is played, we average . Plotted of f2
- We measure the size of the OAE at the distortion product—at a frequency slightly lower than the two tones. Lower in frequency and much smaller in amplitude.

OAES ARE TINY AND SOFT. -OAE = healthy ear. -You can have 30 dB HL loss and still have OAES (mild), so they can't rule out hearing loss. -Are strongly affected by middle ear pathology (even negative pressure) and/or external ear pathology the signal is traveling back through the ear canal. -Are not recorded if there is too much noise in the environment.	WHAT'S AN OAE? -An acoustic signal measured with a microphone in the sealed ear canal. [like tympanometry] As opposed to an electrical signal.		MEASUREMENT – DPOAE 2f1-f2 RUSH – Absolute amplitude: > -5 dB SPL ≥6 dB Signal to Noise Ratio	CONDUCTIVE LOSS Not only will conductive loss decrease the effective level of the stimulus at the cochlea, it will attenuate the emission. TEOAEs will be absent or reduced in amplitude – dependent on severity and type of conductive pathology. Fluid – OAES typically absent. Perfs/PE tubes – whether or not you record OAEs depends on size/position of perforation. Not always recordable. Negative middle ear pressure – OAE amplitude decreased. Presence or absence depends on magnitude of ABG and the amount of negative pressure. Auditory Neuropathy (ANSD) in Children

WHAT IS AN OTOACOUSTIC EMISSION? Assessment of inner ear. Present at birth. USE -Assess OHC function (test tube shape and Prestin electromotility) -Site of lesion battery -Screening for sensory hearing loss -Monitoring cochlear status [if we suspect something is changing the outer hair cells.] -Pseudohypocusis	DPOAEs have two speakers because two signals are delivered		MEASUREMENT – TEOAE Transient-evoked OAEs: (1500 Hz-3000 Hz) Reproducibility (looking at two bins and averaging) TE – NF (how much larger is the OAE than the noise floor in a band) -If you meet the two criteria, the system will say “PASS” and finish. If it doesn’t get it, it will keep averaging. If it fails either, it’s a FAIL. -Frequency reported is the center frequency of narrowband RUSH – 70% Reprod. 0 dB Amplitude ≥6 dB Signal to Noise Ratio	SNHL: OAEs are… NOT sensitive to retrocochlear hearing loss

-An acoustic signal measured with a microphone in the sealed ear canal. [like tympanometry] As opposed to an electrical signal.	HOW DO WE CHANGE FROM TIME DOMAIN TO FREQUENCY DOMAIN? Fourier transform. We are looking for how much energy is occurring in different frequency bands? 500 Hz – 4000 Hz. (largest response from cochlea) Time Window = 20 ms		Newborn -smaller ear canal, larger ear pressure -higher noise level -higher rejection level	Adult	

Auditory Neuropathy (ANSD) In Children

PE Tubes – JUNE 11th

- Most common surgical procedure on kids
- What does AAA mean when they say that children with otitis media may be doomed to mediocrity?
    - -deprivation of a stable auditory base
    - -fall behind peers
- List 4 groups that AAA identifies as at-risk for otitis media with effusion
    - (1) Infants w/ OM before 6 mos
    - (2) multi-child daycare setting
    - (3) structural abnormalities (cleft)
    - (4) other risk factors (DS/Native American populations)

NBHS

- According to the American Academy of Pediatrics, what is an appropriate refer rate for a newborn hearing screening program? Less than 4%
- You must pass both ears on the same day to be a full pass
- Pediatrics uses automated ABR. Only present at 35 dB NHL. They don't have a hard and fast rule about how many sweeps.
- Sent to the Illinois Department of Public Health

- If you must recreen, it must be the same test that they failed.
- You can have a pass, a fail, or a pass with at risk monitoring.
    - If they have a high risk, we still have to keep an eye on them. The state also wants to know this.
    - “Yes it was a pass, but we still want to see you in 3-6 months.”

ABR / ANSD

- What does Stapells recommend using for pediatric ABR protocols? Screening 2kHz in each ear before obtaining thresholds
- According to Stapells, what information would constitute a complete tone-ABR evaluation? AC: 500 & 2000 Hz both ears, BC: 2000 Hz if elevated
- Using a vertex electrode (rather than forehead) will enhance which feature of the ABR waveforms? Amplitude
- Masking is never necessary when measuring ABR in infants False
- You have recorded condensation and rarefaction runs to loud clicks. You are not sure if the early waveform you see is a cochlear microphonic or some other abnormal response. You add the waveforms and subtract the waveforms to yield two new waveforms. How do you make your interpretation? If the subtracted wave has early components greater in amplitude than the added wave, then it’s probably the cochlear microphonic
- Which of the following would be absent in a patient with ANSD? Wave V
- Which syndrome is most closely associated with auditory neuropathy? Charcot-Marie-Tooth
- Children with which diagnosis are likely to show some improvement in hearing despite an early diagnosis of auditory neuropathy? HBR
- Air conduction measures should be done with insert earphones
    - Headphones can affect latency of waveform HOW?. Diaphragm of the insert is further away.
- Bone conduction measures are needed to rule out conductive loss or find conductive component.
    - Use B-70 bone vibrator (max output ~50-55 dB HL)
    - Use mastoid placement
- British Columbia:
    - Is an ear’s AC threshold normal or elevated? Is the other ear’s AC threshold normal or elevated?
    - If elevated, is the elevation conductive in nature or is there a sensorineural component?
    - If elevated, what are the specific thresholds (AC and/or BC). Auditory Neuropathy (ANSD) in Children

PE TUBES Most common surgical procedure for kids.	Reason Reduce otitis media with effusion
BENEFITS -Reduced prevalence of OME -Improved hearing -Improved quality of life	⚫ Adverse Effects: ⚫ Otorrhea ⚫ Persistent TM perforation ⚫ Pathological abnormalities of the TM
Research – Follow up 25 years later. -doesn't affect your hearing loss later	

NBHS	First screening ● Baby passes both ears -> DONE ● Baby refers one or both ears -> RESCREEN Rescreen ● Baby passes referred ear -> DONE ● Baby refers again -> OUTPATIENT SCREEN Outpatient screen ● Baby passes both ears -> DONE ● Baby refers again -> DIAGNOSTIC EVALUATION	1970s – risk was assessed at 2 or 3 years. NOW: 1 Screen all infants before one month of age 3 complete diagnostic evaluations on all failed screenings by 3 months 6 Medical, educational and audiologic intervention before 6 months of age for diagnosed hearing loss	Maternal Anxiety ● Mothers would rather know Cost/Benefit ● Better to test and have some false positives than spend money on special education

Started in Rhode Island – Can a newborn screening program be effectively implemented? -99% of babies born in Rhode Island were screened using transient otoacoustic emissions (TEOAE) -Of the 53,121 babies screened, 111 had permanent hearing loss: about 0.2%			

Risk Factors for hearing loss □ Parent concern □ Family history of hearing loss □ Perinatal asphyxia □ Craniofacial anomalies □ TORCH infections -Bacterial meningitis □ Haemophilus influenzae B □ Vaccinations may reduce incidence □ Hyperbilirubinemia -Low birthweight □ <1500 g	Pulse, Grimace, Activity, Respiration Utah criteria: (Eichwald & Mahoney, 1993) □ 0-4 at 1 minute □ 0-6 at 5 minutes Represents greater risk for SNHL -Possible relation to perinatal hypoxia		Distribution of causes for profound hearing loss in infants Genetic – 50% Environmental – 50% Nonsyndromic – 70%	Babies identified with hearing loss should be evaluated by a team □ Audiologist □ Physician (Peds/ENT) □ SLP □ Others who may contribute to IFSP Outpatient rescreen □ Audiologist or Audiology Assistant □ Screening ABR □ Outcome report sent to IDPH

Auditory Neuropathy (ANSD) in Children

ABR, BAER, AEP	Pediatric Sedation for ABR □ 4 months to 5 years □ conscious sedative, mild general anesthesia □ Monitoring: administered and managed by nurse □ monitor O2, HR and BP □ crash cart and suction available □ Negative outcomes associated with: □ overdoses, drug interactions □ non-trained personnel □ injuries on the way to facility (administered at home) □ drugs with long half-lives (chloral hydrate, pentobarbital)
Summed neural response (microvolt). All the nerve fibers are synchronously firing.	

ADVANTAGES -Estimates normal hearing thresholds -Ear-specific BC finding Diagnosis of AN	DISADVANTAGES -Can't estimate profound HL -Skilled analysis required -Limited BC intensity levels	□ Air conduction measures should be done with insert earphones □ Headphones can affect latency of waveform HOW? □ Bone conduction measures are needed to rule out conductive loss or find conductive component. □ Use B-70 bone vibrator (max output ~50-55 dB HL) □ Use mastoid placement		
ELECTROCOCHLEOGRAPHY – Cochlear microphonic (depolarization repolarization of stereocilia) – SP – AP (wave I) All three of these break down the cochlear potential.	ASSR – way to extract different information from the cochlea using modulated signals, so you're using one signal to get responses from multiple locations. -done with ABR clicks	ANSD □ Diagnosed based on a pattern of audiologic results =Degree of loss: Normal to severe, bilateral 95% -OAEs: present 75% -Acoustic reflexes: absent 90%, elevated 10% =ABR: abnormal =Cochlear microphonic: present HBR -Genetic (charcot-marie-tooth, freidrich's	Management of ANSD □ Amplification – Limited benefit – Addition of FM technology helps in face of low SNR □ Implantation -May improve speech perception -Varied results -What would be a good management strategy for a 7 month old child you identified with ANSD?	

USE: NEUROLOGICAL ASSESSMENTS – High intensity – Interpeak intervals (I-V, I-III, III-V) – Interaural differences – Screening Threshold estimations Absence or presence of wave V and latency of wave V as a function of stimulus level		FREQUENCY SPECIFIC Tone bursts -Provides information for narrower frequency regions -Better relates to pure tone audiogram -Bone-Conducted stimuli -Should get when either the click or 500-Hz tone bursts responses are not present at expected normal levels. -Why/when to perform BC ABR?		ataxia) -unknown -Decreased speech recognition -Worse than predicted by pure tone thresholds -poor frequency discrimination (worse in LF than HF) -CI? -FM helps w/ SNR			

Auditory Neuropathy (ANSD) In Children

Toddler/Preschool Hearing Assessment – JUNE 18th

Fresh Noise/Pediatric Noise – Frequency Specific Hearing Noise. Warble tones are boring. Narrowband noise was mixed up to keep it interesting for children. Can't be used for responses—it's not an accurate signal. Can be used to get the child's attention again.

Madell VIDEO – "Prior to beginning testing, it's important to determine child's cognitive age" critical to choosing a test

- Instead of BOA why not just do an ABR? Limitations of the time, limitations of sleep expectations, doesn't really give us information about functional component.
- Headphones are useful because of ear specificity
- What makes habituation more likely? 100% reinforcement
- What patient history/status piece of information is most important to consider when choosing a behavioral testing technique? -chronological age, -cognitive age/birth history
- Age ranges for behavioral testing [dependent on cognitive age]
    - BOA: younger than 6 mos
    - VRA: 6 months-3 years
    - CPA: 2.5-5 years
- Speech testing by age:
    - -under 2: EPS
    - -2-5: NU-CHIPS
    - -4-6: WIPI
    - -5-8: PBK
    - ->8: NU 6
- Normal progression of frequencies behavioral testing? SNHL -2000, 500, 4000, 1000 // CHL -500, 2000, 250, 1000 (also prioritize BC)

Age	Warble tones (dB HL)	Speech (dB HL)	Response
0-6 wks	75	40-60	Arousal, Eye widens
6 wks – 4 mos	70	45	Rudimentary turn
4-7 mos	50	20	Lateral head turn
7-9 mos	45	15	Direct to side
9-13 mos	40	10	Direct to side, below

13-16 mos	30	5	Direct localization
16-24 mos	25	5	Direct localization

Auditory Neuropathy (ANSD) in Children

Toddler/Preschool Hearing Assessment Can present in sound field, bone oscillator, speakers, headphones of some sort.			
WHY DON'T YOU USE NARROWBAND? The bandwidth is quite wide. If a child had a sloping/rising hearing loss, there could be some off frequency listening going on and responses that were not indicative of that loss.	BOA: younger than 6 mos // VRA: 6 months-3 years [developmental age] // CPA: 2.5-5 years	HEARING TESTING □ Ultimately you want to get ear specific information. □ Frequency presentation varies from Hughson-Westlake □ Based on frequency band importance and ability to interpolate □ 2000 500 4000 1000 Time is of the essence	

Visual Reinforcement Audiometry VRA (6 months – 3 years) Children older than 6 months -Muscle control [looking for head turn] -Cognitive development equal to 5 ½ months of age on Bayley Scales of Infant Development	Detectable stimulus presented child turns toward stimulus reinforcer activated □ Begins by building association between the stimulus □ Once relationship is established the reinforce is used to reward the headturn □ the reinforcer can be a lighted, animated toy or an animation on a screen	Conditioning Orienting Response –Reinforcer only presented when child localizes correctly -Adds layer of cognitive complexity that may be too much for younger children -Requires two reinforcers, one for each ear.	CONDITIONING Response shaping – Sound presented at level that child can hear – When child looks to toy, tester lights up toy – If child doesn't look, assistant can prompt child to lookat the toy Conditioning criteria – Three consecutive responses at same level as response shaping with no prompting. Auditory Neuropathy (ANSD) in Children	

OPERANT BEHAVIOR -Willful or purposeful behavior – rather than a reflex -Increased or decreased in frequency by the changes it brings about in the environment – Conditioning shapes the desired behavior by applying positive or negative reinforcement			-If the child doesn't condition well to tones, and hearing loss is suspected, switch to bone conduction -Condition child to the vibrotactile input. When the child feels the vibration, the toy will light up! -Move from hand to mastoid. □ If BC thresholds can be □ obtained, move back t □ o AC.	

REINFORCEMENT Verbal praise, Clapping, A sticker	□ 100% reinforcement schedule [every time they look] □ If you drop, younger kids tend to be more interested. [Will I get it this time?] □ Rapid conditioning □ Rapid habituation □ Intermittent reinforcement schedule □ Slower conditioning □ Slower habituation □ Primus & Thompson, 1985 □ Found no difference in response of 2-year olds with either schedule □ If you are not sure the response was true, DO NOT REINFORCE □ Not reinforcing a true response won't habituate the child any sooner	
Conditioned Play Audiometry 2.5 5 years Children have left Piaget's Sensorimotor Stage and entered Preoperational Stage (Mastered turn-taking, Are able to make associations) □ They like to play simple games □ They have longer attention spans Establish rapport, elicit speech	SNHL -2000, 500, 4000, 1000 CHL -500, 2000, 250, 1000 (also prioritize BC)	

Procedure Instructions stimulus child takes play turn audiologist reinforces with praise	Young children need modeling - Audiologist can demonstrate the task - Parent can demonstrate the task - Physically manipulating the child, hand-over-hand, to do the task may be necessary the first couple times If fail: vibrotactile, revert to VRA	Test Order: [Select the order where you can get the most information.] Tympanometry, OAE, Pure tones, Soundfield, Phones Speech Soundfield, Phones	
Behavioral Observational Audiometry Younger Than 6 Mos Test Of Responsiveness, Not Sensitivity			
Madell: Baby is held Observing changes in sucking, startling, eyes widening, it is a reflex.	- You have to agree what your method will be and then stick to it. - Make sure it is repeatable and not just a random occurrence. - An extra set of eyes is extremely helpful. - Takes a lot of training to become reliable - If false alarm rate is greater than 25%, you can't make a just assessment.	Signal Noisemakers: Bells, percussion, etc. Broad or complex spectral characteristics, Hard to control intensity level Pure tones: Easy to control level, Infants less inclined to respond to tones and narrow bandwidth stimuli	

□ Probability of a response depends upon □ State of infant □ Nature of stimulus □ Ambient noise levels □ Agreement among observers □ Leads to many false-positives and false-negatives								

JULY 2ND – SPEECH TESTING

- Why do we have to use an average to describe speech? There will be fluctuations and differences in frequency content. When we take those two variables into account, we end up looking at a lot of fluctuation above and below that average. The peak can be hovering around 12 dB around the average.
- how to proceed with speech testing if it is too easy:
    - -start at 50 dB HL
      -if between 50-75%, administer at 35 dB HL and at 50 dB HL with +5 SNR
      -if >75%, administer a more difficult test at 50 dB HL (and do the same thing from there)
      -if <50%, go to an easier test
- Should you choose a speech perception test based on the child's degree of HL? No
- Speech Testing ages:
    - -under 2: EPS
      -2-5: NU-CHIPS
      -4-6: WIPI
      -5-8: PBK
      ->8: NU 6
- Body part identification is used for: SRT
- Advantages of phoneme scoring:

- -phoneme scoring can be used with any speech test
  -Can identify which parts of the auditory spectrum that are not being appropriately perceived by the listener.
  -Modifications to the frequency response of the child's hearing aids or cochlear implants, to make ear mold changes, and to make recommendations about auditory training goals can be applied.

- Why is SRT + 40 dB not necessarily an appropriate level for presenting stimuli for speech testing?
  - -want to see how each child is performing functionally in degraded environments
    -50 dB HL in quiet
    -35 dB HL in quiet (soft speech)
    -50 dB HL in 45 dB HL noise (+5 SNR)
    -50 dB HL in 50 dB HL noise (0 SNR)
    -35 dB HL in 35 dB HL noise (0 SNR)

As children develop language, they can complete more complicated speech tests with higher levels of vocabulary.
When performing speech testing, have in mind whether you are going to be monitoring development of this child and how you want to show changes. Auditory Neuropathy (ANSD) in Children

- How might it be different for a child with OME vs. a child with moderate SNHL?

Conductive loss :loss might come and go. Might do well at one appointment. Might be fine at conversational level, and then test at soft and they might do worse.
SNHL: can be pretty predictable.

SPEECH TESTING -JULY 2nd			
Average talker, average effort, 4 feet away ~60 dB SPL ~50 dB HL	Speech is a broadband signal From fundamental frequency ~100 Hz To fricated noise ~16,000 Hz Majority of speech is up to 8000 Hz. Our sensitivity for low frequencies is not as good.	Easiest sounds to see are the hardest to hear. Vowels are low frequency. /f/ is high frequency	Communication strategies – Reduce distance – Reduce noise – Sound-treat environments – Avoid poor acoustic environments. Technology – FM systems Looped systems

the summed level across all frequencies of the speech signal, averaged over time (the LTASS: long-term average speech spectrum).			

Speaker dependent – Vocal quality – Vocal amplitude – Clear speech (expanded vowel space, slower speech rate, word-final consonant release, more intense stop consonants)	Listener dependent – Hearing loss – Attention/fatigue (teach kids to look at you if they didn't understand) – Audiological intervention (hearing aids, cochlear implants) Medical/behavioral intervention (if it's attention/fatigue—which is outside our scope)	Environment dependent – Distance (reduced amplitude. 6 dB per doubled distance in freefield; reduced access to visual cues- children under six may not take advantage of lip-reading cues anyway.) – Noise (masking of speech signal, broadband noise less disruptive than speech noise. Children require greater signal to noise ratio for speech recognition equal to adults). Auditory Neuropathy (ANSD) in Children Reverberation (early reverberations enhance signal; late reverberations mask signal)	– Lower frequencies – more intensity – High frequencies – less intensity – So if you have a high frequency hearing loss, despite those fluctuations

Why do speech testing? Determine the extent of the hearing loss' effect on speech perception Identify childrn at risk for delayed speech/language development Monitor progress during treatment	Evaluating Speech -Threshold vs. suprathreshold -Open-set vs. Closed-set (Digits, alphabet) -Recorded vs. Live Voice -Presentation Level -Presentation Condition -Test Complexity -Scoring (total accuracy, phoneme scoring)	Speech Detection Threshold -Detection threshold: lowest level at which a stimulus is detected 50% of the time -Broadband detection: running speech -Narrowband detection: phonemes Ling-6 sounds /m/, /a/, /u/, /i/, /ʃ/, /s/ "Ling-3" sounds /ba/, /ʃ/, /s/ -Generally these sounds are presented via live voice -Adjust level with attenuator dial of audiometer -Keep an eye on the VUmeter	

Auditory skills hierarchy Detection (SRT) Discrimination Identification (also called recognition) Comprehension (replying to a question and answering. Don't tend to use comprehension in audiology)	Speech Reception/Recognition Threshold -Recognition threshold: lowest level at which a stimulus is identified 50% of the time -Spondee repetition -Body-part identification, Potato-Head task -Picture pointing	IT-MAIS/MAIS – Parent questionnaire "Does your child produce well-formed syllables and syllable-sequences that are recognized as speech?" Never – Always Can be used pre-post intervention Early Speech Perception (ESP) a closed-set test that examines pattern perception (Hop hop hop for a bunny. "Vroom" for a car)	Easier: WIPI (Word Intelligibility by Picture Identification) Harder: PB-K Words (Phonetically Balanced Kindergarten)
TESTS [brown – closed]			

PBK – open-set word list – monosyllabic -Four 50-word lists	NU-CHIPs -4 forced choice test -3-5 years	WIPI -6 forced choice (a little bit harder. That's why the age recommendation is older) -4-6 years	GASP – Sentences: Series of WH-Questions -Words: various syllabic	HINT-C (noise in testing) -Presented at 70 dB SPL in soundfield. -Recommended for children 5 years and older	Multisyllabic Lexical Neighborhood Word lists dependent on child's experience and their age Provide reliable information about the spoken word recognition abilities of children with profound

					hearing loss who use cochlear implants
SELECTING A PROTOCOL					
– A goal of pediatric speech testing is to find the limits of the child's auditory skills using a test appropriate for their cognitive and auditory abilities. –	Mendel □ Supports a battery that examines various domains of perception □ Phonetic segments □ Words □ Discourse	Madell □ Supports a battery that hones in on not-quite-mastered level of speech perception at environmentally valid levels □ Choose pick based on language age. □ Four year old that's cognitively on track, needed a hearing aid and just got it a few months ago you might need to pick something simpler. □ Should be tested in quiet			