

Recommended Protocols for Diagnostic Audiological Assessment Follow-up to Newborn Hearing Screening in Ohio

The Early Hearing Detection and Intervention (EHDI) Program at the Ohio Department of Health (ODH) and the Coalition of Ohio Audiologists and Children's Hospitals (COACH) formed a taskforce in 2015 to establish standardized diagnostic evaluation measures for infants who did not pass their newborn hearing screening. Members of this taskforce included audiologists at Children's Hospitals and in other settings who provide follow-up for newborn hearing screening, pediatric otolaryngologists, and the Ohio Department of Health EHDI program. A series of statewide stakeholder meetings identified the need for standardized follow-up audiologic protocols. The task force members have widespread knowledge and skills with pediatric and infant audiologic testing. Collaboration among the members provided many contributions to the development of this standardized diagnostic evaluation process. This diagnostic protocol focuses on the first 3 months after birth, in which the diagnostic process should be completed. The protocol is designed to assist audiologists to comply with best practice for optimal universal hearing loss detection and intervention (EHDI) goals.

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I. Introduction

The Joint Committee on Infant Hearing (JCIH 2007 position statement), Ohio Department of Health and COACH all recommend that infants referred from newborn hearing screening have a diagnostic audiological assessment as soon as possible, with the goal of complete diagnostic testing before three months of age.

The target population for this protocol is infants referred for follow-up from newborn hearing screening from birth to 6 months of age. The process in this document provides specific guidance for physiological testing to identify ear specific information, including type, degree and configuration of hearing loss. It is important to note that prenatal history, medical history and hospital screening results including risk factors are an integral part of the evaluation. The scope of this protocol is the initial diagnostic assessment, and thus it does not discuss screening procedures at the hospital, nor does it cover intervention services.

The overarching goal of a statewide Early Hearing Detection and Intervention program (EHDI) is to identify hearing loss very early in life to ensure intervention before 6 months of age, promoting optimal brain development and allowing for subsequent development of language and communication skills. Behavioral audiometry provides valuable information for infants with a developmental age of six months or greater, and will be the subject of a future protocol.

The JCIH 1-3-6 goals state that screening be performed before 1 month of age, however, Ohio's legislation requires newborn hearing screening to be done at birth before hospital discharge. Babies who do not pass the initial screening receive a secondary screening on both ears before discharge. JCIH states that babies who did not pass the hospital screening are referred for audiological follow up, and that diagnostic assessment be

completed before 3 months. Babies who return for follow-up shortly after birth usually do not need sedation and may be tested in natural sleep. JCIH recommends that infants in whom permanent hearing loss is diagnosed should be fitted with an amplification device within 1 month of diagnosis for families who elect amplification and enrollment in early intervention should occur as soon as possible after diagnosis, before 6 months of age.

In addition, infants who pass the newborn hearing screening but have one or more higher-risk factors for late onset or progressive hearing loss should have a diagnostic ABR by 3 months of age. Otherwise, infants with at least one risk factor should have at least one diagnostic audiological assessment by 12 months of age. The frequency of diagnostic follow-up may depend on risk factors or caregiver concern. More information about types of risk factors is provided in the Appendix.

II. Abbreviations

AC: Air conduction

ABI: Auditory Behavior Index

ABR: Auditory Brain Response

ASSR: Auditory Steady State Response

ANSD: Auditory Neuropathy Spectrum Disorder

BBN: Broad band noise

BC: Bone conduction

dBA: Decibels weighted according to the A scale

dB nHL: Decibels referenced to normal behavioral thresholds for ABR stimuli

DPOAE: Distortion Product Otoacoustic Emissions

eHL: Estimated hearing level in dB

ECMO: Extracorporeal Membrane Oxygenation

EHDI: Early Hearing Detection and Intervention

JCIH: Joint Committee on Infant Hearing

OM: Otitis media

TEOAE: Transient Evoked Otoacoustic Emission

UNHS: Universal Newborn Hearing Screening

VRA: Visual Reinforcement Audiometry

III. Qualified Personnel

1. Licensed audiologist who is proficient in providing audiology services to infants and children
2. Assistance to the audiologist may be provided by the following:

- a. Audiology Interns under direct supervision by a licensed audiologist
- b. Audiologist assistants under direct supervision by a licensed audiologist

IV. Safety and Health Precautions

1. All procedures ensure the safety of the patient and clinician and adhere to standard health precautions (e.g., prevention of bodily injury and transmission of infectious disease).
2. Decontamination, cleaning, disinfection, and sterilization of multiple-use equipment before reuse are carried out according to facility-specific infection control policies and procedures and according to manufacturer's instructions.
3. Disposable probe tips, otoscope tips and electrodes are recommended to reduce infectious disease transmission.

V. Optimal Test Environment

1. Infant Preparation: Refer to preparation instruction sheet in the Appendix, which should be provided to caregivers before appointment. The purpose of preparation instructions is to help ensure that infants arrive for testing hungry, awake and ready to feed and go to sleep.
2. The test environment must be quiet and infants should ideally be sleeping or at least quiet and comfortable for adequate results. A sound booth is desirable, but not necessary in a quiet environment (less than 50 dBA unoccupied) *if* the probe and earphone fit snugly.
3. A sink with warm water and soap for handwashing and clean up.
4. Supplies including diapers, wipes, and drinking water.
5. Dimmable lights, privacy and provisions for nursing (comfortable chair with arm support)
6. A crib or rocking chair for feeding and to aid and maintain sleep. Rocking chairs and cribs should be covered with a fresh sheet that is laundered after each use.
7. Best results are obtained for most infants if they go to sleep on their back so that both ears are available. This is also recommended as the safest position for the infant to sleep.
8. Some infants will sleep well in a crib or infant carrier after going to sleep, while others will sleep better in their caregiver's arms. Note that infants who are held may be more restless, and the electrodes and probes are harder to keep in place.
9. Infants with cardiac or pulmonary issues, stridor, or tracheostomy may be noisier and have difficulty remaining asleep when on their backs, so the caregiver should be asked about the best sleep position for these special needs infants.

VI. Procedures

The standardized comprehensive test battery includes a specific series of procedures designed to efficiently obtain ear-specific information. Guidance for each of the below procedures is provided in an overall flow chart (Figure 1). The below order of tests is recommended to provide ear-specific results that assess type, degree and configuration of hearing status for both well-baby and high risk newborn hearing screening referrals.

The audiologist should plan the assessment to obtain the most important information based on the case and the infant's state. Thus, while some flexibility is encouraged in the steps outlined, following the standard protocol will provide the necessary information to either clear or diagnose an infant with hearing loss, determine need for ongoing monitoring, or to plan for intervention if a hearing loss is diagnosed.

The case history and otoscopy can be accomplished while the infant is feeding. Otoacoustic emissions and prepping for ABR can be accomplished in a quiet awake state, while obtaining ABR threshold responses generally requires the infant to be quiet or sleeping.

1. Case History: A sample case history is provided in the appendix. The purpose of the case history is to obtain a thorough infant and family history, determination of congenital and neonatal risk factors for hearing loss, including caregiver/family report of the infant's responses to sound.
2. Otoscopic Examination: To ensure that the infant has an open ear canal and to assist with selection and accurate placement of appropriately sized probe tips and earphones.
3. DPOAE or TEOAE: Recommended as part of an abbreviated diagnostic protocol and as a cross-check for ABR results.
4. ABR: Can be done as a limited or complete diagnostic protocol, depending on risk factors and OAE results.
5. Immittance: Tympanometry and acoustic reflexes are recommended if OAE or ABR is abnormal to determine if middle ear function is normal and acoustic reflexes are present as a test for neural function.
6. Interpretation, counseling, appropriate referrals and resources.
7. Completion of required paperwork, timely and appropriate referral for habilitation services if needed.

VII. Equipment

Multiple pieces of specialized and calibrated equipment are needed to provide a comprehensive evaluation. Equipment should be versatile to meet the unique needs of each child. At a minimum, an otoscope, 1000-Hz middle ear acoustic immittance, acoustic reflexes, DPOAE or TEOAE, and tone burst air and bone-conduction ABR equipment are necessary for diagnostic evaluation. Auditory steady state responses (ASSR) and chirp stimuli are helpful optional procedures.

1. Equipment Calibration and daily checks: Equipment will be maintained and used per manufacturer guidelines. Instruments must be calibrated at least annually according to ANSI standards where available and daily calibrations for tympanometry and OAEs should be performed using a calibration cavity supplied by the manufacturer. Daily biologic (listening) checks are recommended ABR equipment.
2. Otoscope: Infant size disposable specula should be available.
3. Acoustic immittance: For immittance and acoustic reflex threshold measurement. Equipment with multifrequency or wideband probe tones (minimum of 226-Hz and 1000-Hz) is recommended. Calibration procedures are specified in ANSI S 3.1 (1987).
4. OAE: Equipment can be distortion product (DPOAE) or transient-evoked (TEOAE). It should offer varying test parameters and the ability to change stimulus levels.
5. ABR: Equipment should be able to produce many stimulus types such as clicks, chirps and tone bursts at different levels. Insert earphones and bone conduction oscillator should be available. Chirps may be used in place of clicks if desired.

6. Auditory Steady-State Responses (ASSR): ASSR may be used as an adjunct to ABR testing, and may be preferred for test time and multiple frequency responses. Detailed ASSR testing is not covered in this initial protocol, but may be added later.
7. Audiometer for Auditory Behavioral Index (ABI) and Visual-Reinforcement Audiometry (VRA): ABI and VRA are not covered in this protocol, but are important to complete as a validation of physiologic results, and to monitor hearing and intervention outcomes.

VIII. Important Considerations

1. Re-screening does not provide a diagnosis of normal hearing versus hearing loss, only an indication that further testing is necessary. In order to prevent delayed diagnosis, if the infant has already had two screenings (hospital and outpatient, or two screening tests within the hospital), additional rescreening is **not recommended**. A comprehensive test battery of otoscopy, immittance, OAE and ABR is recommended for adequate follow-up.
2. Both ears should be evaluated at the follow-up, even if only one ear did not pass the screening.
3. Only audiologists with experience in pediatric assessment and counseling should provide comprehensive follow up evaluation. ODH provides a referral directory of audiologists who can provide screening and diagnostic services.
4. Otoacoustic emissions (OAE) and middle ear assessments are important cross-check measures to diagnose type and location of problems in the middle ear and cochlea, but cannot provide diagnosis without ABR threshold testing.
5. Auditory brainstem response (ABR) is the fundamental test for accurate, frequency-specific and ear-specific pure tone threshold estimates, and is also able to assess neural problems such as Auditory Neuropathy Spectrum Disorder (ANSI).

IX. Case History (Form in Appendix A)

1. Expected Outcome: To gain knowledge on infant's birth history, risk factors and other pertinent medical background as well as build rapport with family
2. Obtain case history information by caregiver interview.
3. Ask about the reason for referral and the family's goals for the assessment.
4. Ask about hospital hearing screening results and risk factors.
5. Ask about infant's behavioral responses to sound.
6. Ask how the infant is best able to achieve sleep to provide appropriate support in the test environment.

X. Otoscopic and Outer Ear Examination

1. Expected Outcome: Assess the status of the outer ear canal prior to testing.

2. Otoscopy is performed to ensure that there are no contraindications to placing an earphone or probe in the ear canal.
3. Visual inspection for obvious structural abnormalities (e.g., ear pits, ear tags, atresia, stenosis and low set ears) of the pinna and/or ear canal should be completed and documented.
4. Newborn ear canal size and anatomy may make it difficult to identify the tympanic membrane or any landmarks, and to detect presence of fluid.
5. To diagnose ear drum or middle ear problems, referral to an otolaryngologist experienced in newborns is recommended.

XI. Diagnostic OAE Evaluation

1. Expected Outcome:
 - a. To assess cochlear function at the level of the outer hair cells.
 - b. OAEs are not a direct measure of hearing, but provide a valuable physiologic cross-check for ABR testing.
 - c. Using the below guidelines, OAE levels are sensitive to hearing losses of 30 dB HL and greater, although a small percentage of borderline and mild losses may have normal OAE levels (Gorga et al., 2000; Gorga et al., 2005; Norton et al., 2000).
 - d. It is not possible to predict degree of hearing loss from OAE measures alone.
 - e. Middle ear assessment is necessary to interpret abnormal OAE responses, since the middle ear both conducts the sound stimulus and the cochlear response.
2. Equipment:
 - a. Either Distortion Product (DPOAE) or Transient Evoked Otoacoustic Emissions (TEOAE) methods can be used.
 - b. Equipment should have multiple test parameters including the ability to adjust stimulus levels.
 - c. DPOAEs are generally recommended when high frequency regions are important to assess (2-8 kHz)
 - d. TEOAEs are more sensitive in the 1-2 kHz region (Norton et al., 2000).
3. Testing Conditions: Environment is quiet and patient is sleeping or quiet. A bottle or pacifier may be used to encourage sleep, but noise levels will increase if the infant is actively feeding or sucking. A sound booth is desirable, but not necessary in a quiet environment (below 45 dBA) and most importantly, with a tightly fitted probe.
4. Frequency range:
 - a. DPOAE: Assess five to six frequencies from 2000-8000 Hz. Noise levels are higher below 2000 Hz in infants, so frequencies between 2-8 kHz and above are most sensitive and specific to hearing loss (Gorga et al., 2000; 2005).
 - b. TEOAE: Assess above 1000 Hz for the same reasons of noise floor and better sensitivity (Norton et al., 2000).

5. Recommended stimulus levels:

- a. DPOAE: Recommended levels are L1=65 dB SPL and L2=50 - 55 dB SPL. The stimulus level should be within 3 dB of these target levels when verified in the ear.
- b. TEOAE recommended levels are 80 dB peSPL (± 3 dB) (AAA Guidelines, 2012).

6. Classification of Results:

- a. Age-specific reference data are recommended to determine the presence or absence of an emission at each frequency evaluated.
- b. DPOAE normal criterion at each frequency: The emission is present if the DP-NF (Distortion product minus noise floor, or SNR) value is 6dB or greater at each frequency and the overall response is consistent with age appropriate norms.
- c. DPOAE normal overall criteria: DP level and SNR criteria are in normal range, consistent with age appropriate norms.
- d. TEOAE normal criterion at each frequency band: The SNR must be >6 dB and reproducibility must be $>70\%$ for each frequency bands assessed, and the response amplitude is consistent with age appropriate norms.
- e. TEOAE normal overall criteria: Response is obtained at a SNR of greater than or equal to >6 dB at $>50\%$ of frequency bands and the overall response reproducibility is $>50\%$.
- f. If results are not normal, ensure that they were not impacted by noise in the room or by the patient, a poor probe fit or debris in the probe assembly. Repeating the test after the infant is sleeping is the best way to ensure quality recordings.

7. Interpretation of Results:

- a. An overall normal result is consistent with normal peripheral function through the level of the outer hair cells at all frequencies where OAEs were present.
- b. Borderline levels may be consistent with mild hearing loss or middle ear dysfunction and should be repeated, particularly if there are any other abnormal test results or caregiver concern for hearing.
- c. Responses that are present (meeting SNR criteria), but have reduced amplitude could indicate sub-clinical outer hair cell damage or a mild hearing loss.
- d. If middle ear function is normal, the absence of responses is generally consistent with a problem with the outer hair cells and a sensory hearing loss of at least 30 dB HL.
- e. If middle ear function and OAE results are not normal, refer for medical assessment. Repeat OAE testing after medical treatment.
- f. If ABR is markedly abnormal or absent, complete acoustic reflexes to distinguish between middle ear and neural pathology. Infants who pass ABR, but do not pass OAE should be referred for medical treatment for external or middle ear pathology. Repeat audiologic evaluation after medical treatment.

Normative data for newborns – 6 months for DP Level and for SNR.

From Blankenship, Hunter, Keefe et al, 2016. The SNR and the DP level should both meet these criteria at $>50\%$ of frequencies tested.

DPOAE using 65/55 primary tone levels for newborns to 6 months.

Frequency (kHz)	Cut-off Point to judge responses normal (greater than or equal to these levels)	
	DPOAE Level (dB)	SNR (dB)
2	6	6
3	4	5
4	5	12
5.5	2	10
8	-9	6

XII. Diagnostic Threshold Auditory Brainstem Response (ABR) Protocol

1. Expected Outcome(s):

- a. To assess status of the peripheral and central auditory system;
- b. To determine thresholds for ABR presence and estimated hearing levels;
- c. To determine need for audiologic habilitation in accordance with JCIH guidelines.

2. Clinical Indications:

- a. Patients who did not pass newborn hearing screening;
- b. Older patients who are difficult to test and/or who cannot provide accurate behavioral test information to rule out hearing loss;
- c. Any time objective information is needed to determine auditory sensitivity and/or neural pathway status.

3. Patient Preparation/Electrode Placement

- a. Either one or two-channel recordings are acceptable. Two channel recordings are useful for determining side of bone conduction response, and for assessing neurologic integrity (Wave I will be present in the ipsilateral recording at higher intensities, while Wave V should be present in both channels).
- b. For one-channel recordings: Three electrode sites will be prepared in the below locations. This montage can be used for 2-channel recordings, and also for systems that provide automatic electrode switching in a one-channel recording. The ipsilateral and contralateral electrodes can also be manually switched in systems that do not provide electrode switching.
 - i) High Forehead (FZ)/Vertex (CZ) (positive electrode)
 - ii) Ipsilateral mastoid or earlobe (negative electrode)
 - iii) Contralateral mastoid/earlobe (common or ground)
- c. For two-channel recordings: Four electrode sites will be prepared in the below locations. This montage can be used for 2-channel recordings, and also for systems that provide automatic electrode switching in a one-channel recording. The ipsilateral and contralateral electrodes can also be manually switched in systems that do not provide electrode switching.

- i) High Forehead (FZ)/Vertex (CZ) – positive electrode, with jumper cable
- ii) Low Forehead or nape of neck – common or ground
- iii) ipsilateral mastoid or earlobe – Channel 1 negative electrode
- iv) contralateral mastoid/earlobe– Channel 2 negative electrode
- d. Skin preparation: Clean the skin with electrode preparation liquid on a gauze pad, taking care to wipe away all excess liquid before applying electrodes.
- e. Disposable electrodes are recommended for infection control, consistent impedance and ease of clean up.
- f. Insert earphones with silicone tips or infant foam tips should be inserted as fully as possible. Trimming of foam tips may be necessary for newborns.
- g. Bone conduction (BC) should be done whenever air conduction (AC) ABR is abnormal. Reliable results can be obtained with the BC transducer placed at the temporal bone just above the pinna. Hand-held with a fingertip or using a headband has been shown to be equivalent (Small, Hatton and Stapells, 2007). Measurement of adequate force with a strain gauge is recommended.
- h. Effort will be made to ensure appropriate impedance for the electrode array. Impedance values should be < 5 kOhm for each electrode, and balanced with no more than 3 kOhm differences.

3. Clinical Process:

- a. The limited diagnostic protocol is the minimum suggested battery for otherwise healthy infants. The purpose of the limited battery is to balance sensitivity with cost and time needed. The audiologist or site may elect to do full diagnostic batteries at their discretion.
- b. The limited diagnostic protocol is intended for initial assessment of well infants who do not have risk factors for delayed/progressive hearing loss, and do not have caregiver concerns for hearing or a family history of hearing loss.
- c. For the limited diagnostic protocol, the decision to proceed to full diagnostic testing is based on any abnormality of OAE and click or toneburst ABR results.
- d. For either the limited or full test batteries, start in the ear that referred, or either ear if both referred.
- e. Always test both ears even if refer was unilateral since screening documentation errors can occur, and hearing loss may occur in a previously normal ear after birth.
- f. See tables below for suggested order of test stimuli/transducer and initial stimulus levels.
- g. The cutoff for estimated hearing level (eHL) is 20 dB HL or less to consider hearing status as normal. The below cutoff criteria were determined with this goal in mind.

Limited and Full Diagnostic ABR	
For infants with Normal OAEs and no risk factors, including no caregiver concerns for hearing	<p>Limited diagnostic protocol:</p> <ol style="list-style-type: none"> 1) Alternating split-sweep clicks or chirps at 60 and 25 dB nHL 2) If no response present at 60 dB, increase in 20 dB steps to determine overall starting level for toneburst stimuli and proceed to full diagnostic protocol. 3) If the click or chirp ABR is present in both ears at 25 dB nHL with normal OAEs, the assessment is considered normal. 4) Always test both ears even if only one referred.
For infants with Abnormal OAE or Risk factors present	<p>Full diagnostic protocol:</p> <ol style="list-style-type: none"> 1) Alternating split-sweep clicks or chirps at 60 and 25 dB nHL* 2) If no response present at 60 dB, increase in 20 dB steps to determine overall starting level for toneburst stimuli and proceed to full diagnostic protocol. 3) Continue with AC tone bursts starting at 10-20 dB above click threshold. <ul style="list-style-type: none"> • 4000 Hz • 1000 Hz 4) Decrease or increase as needed in 10-20 dB steps to threshold. Obtain a no response recording 5-10 dB below lowest repeatable response to determine threshold. 5) Switch ears and repeat. If above abnormal and if infant sleep state permits: <ul style="list-style-type: none"> • 500 Hz • 2000 Hz 6) Continue with BC tone bursts at below starting levels if AC thresholds were abnormal. Decrease or increase in 10-20 dB steps to threshold. Obtain a no response recording 5-10 dB below lowest repeatable response to determine threshold. <ul style="list-style-type: none"> • 4000 Hz at 30 dB nHL • 1000 Hz at 30 dB nHL 7) If two-channel recordings are performed, the latency value and amplitude can be used to infer side of BC response, since ipsilateral recordings are better than contralateral (Hatton et al., 2012). 8) Note that at less than 12 weeks of age and bone conduction levels of less than 15 dB nHL, there is no cross over so masking is not necessary.

5. Correction Factors for estimated behavioral hearing level (dB eHL; McCreery et al., 2014):

The degree of hearing loss influences the ABR-behavioral threshold differences for all frequencies. Of particular importance is the observation that ABR thresholds underestimate behavioral thresholds in cases of greater than moderate hearing loss. This underestimation has the potential to result in under-amplification when ABR is used as the basis for hearing aid gain prescriptions. Thus, correction factors should differ for different ABR threshold levels.

Negative numbers indicate that the correction factor (dB) should be subtracted from the ABR threshold to estimate behavioral hearing level, and positive numbers mean that the correction factor (dB) should be added to the ABR threshold to estimate behavioral hearing level.

Toneburst Freq.	20 dB nHL	40 dB nHL	60 dB nHL	80 dB nHL
500 Hz	-5	3	7	12
1000 Hz	-5	-3	0	2
2000	-5	-2	1	4
4000 Hz	-6	-3	0	3

These correction factors were obtained using calibration for peak equivalent SPL (peSPL) for 0 dB nHL reference for 250, 1000, 2000, and 4000 Hz tone burst and the click are 43, 24, 28, 32, and 35 dB peSPL, respectively (Gorga et al. 2006).

5. General Test Parameters:

- a. Window latency – 20 ms for click or chirp and 1000-4000 Hz toneburst; 25 ms for 500 Hz tone bursts.
- b. Toneburst ramp: Blackman window, rise fall times recommended (McCreery et al., 2014)
 - i) 500 Hz : 2-0-2 msec
 - ii) 1000 Hz: 2-0-2 msec
 - iii) 2000 Hz: 1.5-0-1.5 msec
 - iv) 4000 Hz: 1-0-1 msec
- c. Stimulus Rate – range between 27.1 and 37.9. Use odd numbers to minimize 60 Hz harmonic components. A slower rate, such as 13.1 or 11.1 may be needed for neurologic immaturity (premature or neurological diagnosis).
- d. Filter range – High pass filter set in range of 30 to 100 and low pass filter 1500 to 3000 Hz.
- e. Notch filter - Not recommended since infant responses are dominant in low frequencies.
- f. Waveform smoothing– if used, should be no higher than 5.
- g. Minimize/Maximize tracings – is not recommended to be used unless it is applied to all of the tracings.
- h. Stopping rules: Recommended rules for acceptable waveform quality are available for most instruments based on residual noise and correlation values, and are highly recommended for consistency. If the residual noise is sufficiently low, and correlation values are acceptable, the response may be stopped and the next level or frequency started.
- i. Sweeps: If stopping rules are not available, a recommended approach is to set the sweeps to a very large number such as 5000, and monitor the recording until waveforms are quiet, stable and the response is clear in the expected latency range. Waveform replication is recommended only to establish threshold.

6. Adjustment of ABR Parameters:

Suggested stimulus and recording parameters can be adjusted within recommended ranges as needed to obtain the best test recording. For example, if low frequency noise is contaminating the tracing, the high pass filter can be adjusted slightly higher. If high frequency noise is apparent, the low pass filter can be moved lower. More averaging may help, or waiting until the infant is quiet. Checking and improving impedance, turning off unnecessary equipment and unneeded monitors or cellphones may help in cases of electrical noise.

7. Diagnostic Assessment (Full Protocol):

A minimum of clicks or chirps, normal thresholds at 1000 Hz and 4000 Hz, along with normal DPOAE data should be obtained in both ears. It is then up to the judgment of the audiologist to determine if enough data was obtained to confirm normal hearing sensitivity or if further testing is necessary. When possible, results should be reviewed by another audiologist for confirmation.

If results were abnormal, immittance testing is recommended as a cross-check of hearing loss type (conductive, sensory, neural). Testing may be done prior to or after ABR testing, as the audiologist's discretion.

XIII. Immittance (tympanometry and acoustic reflex)

1. Expected Outcome(s):

- a. To assess middle ear function and auditory pathway integrity
- b. To evaluate for middle ear abnormalities

2. Select appropriate probe stimulus:

- a. Birth to 6 months: 1000 Hz or wideband (click) stimulus
- b. Older than 6 months: 226 Hz
- c. Choose a probe tip of sufficient size to achieve a hermetic seal and place tightly in ear canal.

3. Normal ranges for 1000-Hz probe tone (birth to age 6 months):

- a. Normal: ≥ 0.6 peak static admittance, relative to the positive baseline
- b. Abnormal: < 0.6 peak static admittance (Margolis et al., 2003)

4. Normal ranges for 226-Hz probe tone (6-36 months):

<i>Ear Canal Volume (mL or cc)¹</i>	Blocked: 0.0 – 0.09 ml Normal: 0.1 – 1.0 ml Possible perforation: ≥ 1.1 ml
<i>Peak Pressure (daPa)</i>	Negative: < -150 daPa Normal: -150 to $+100$ daPa Positive ² : > 100 daPa
<i>Admittance (mmho)</i>	Reduced: < 0.2 mmho Normal: 0.2^3 – 1.2 mmho Increased: > 1.2

<i>Tympanometric Width (daPa)</i> ⁴	Infant/Child: > 250 daPa
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¹ Volumes are guidelines and must be interpreted cautiously such as for patients with abnormally sized ears. Comparison to opposite ear is recommended (should be similar except in cases of unilateral ear canal stenosis)

² Positive pressure may be an indicator of acute OM.

³ Combine tympanometric width with admittance for better sensitivity for 226-Hz. For 1000-Hz, admittance alone is recommended since notching and collapsing ear canals at this frequency is often present, and can make width measurements inaccurate.

⁴ Tympanometric width and admittance are based on gold standard of myringotomy (Nozza et al., 1994)

5. Tympanometry interpretation:

- a. Normal: Identifiable peak is observed at or near atmospheric pressure and admittance and tympanometric width values are typical for the patient's age (see norms).
- b. Abnormal: No identifiable pressure peak
 - i. With normal volume: Consistent with middle ear fluid
 - ii. With abnormally large volume: Consistent with patent tympanostomy tube or tympanic membrane perforation
- c. Abnormal: Peak is observed, but static admittance values indicate reduced or increased mobility (see norms).
 - i. Abnormally low admittance: Consistent with reduced middle ear mobility (such as middle ear fluid, ossicular fixation, or other abnormalities of middle ear function).
 - ii. Abnormally high admittance: Consistent with increased middle ear mobility (such as ossicular anomaly or abnormalities of the tympanic membrane).
- d. Peak is observed, but tympanometric width is abnormally increased or gradient is abnormally reduced (see norms).
 - i. Abnormally broad width: Consistent with reduced middle ear mobility (may be due to OM, ossicular fixation, or other abnormalities of middle ear function).
- e. Peak is observed at a pressure outside of the normal range (see norms)
 - i. Consistent with abnormally negative or positive middle ear pressure.

6. Acoustic Reflexes: Acoustic reflexes may be helpful in cases of suspected ANSD, neurologic history, or when ABR is markedly abnormal, as the acoustic reflex is nearly always absent or elevated in confirmed cases of ANSD (Berlin et al., 2005).

- a. Select Probe Tone: Children Birth to 6 months: 1000-Hz; Children > 6 months: 226-Hz.
- b. Select ear: Ipsilateral or Contralateral.
- c. Place appropriately sized tip into ear canal.
- d. Start test and when pressure is equalized (i.e. at peak admittance) and begin stimulation.

- e. A response is present with clear deflection (0.02 mL or greater) from the baseline, continued contraction with stimulation and return to baseline when stimulus is discontinued.

7. Acoustic Reflex Interpretation:

- a. Normal response: The average acoustic reflex threshold for infants with normal hearing is between 65-80 dB between 500-4000 Hz, and is 60 dB for broad band noise (Kei, 2012).
- b. Abnormal response: The upper limit for the acoustic reflex threshold is >95 dB HL for 500 Hz, 85 dB at 3000 Hz, 80 dB at 4000 Hz and 75 dB for broad band noise (Kei et al., 2012).
- c. Note: maximum stimulus level should not exceed 95 dB HL in infants due to the possibility of noise induced hearing loss caused by the reflex stimulus (Hunter et al., 1999).

XIV. Counseling and follow-up

1. Informational counseling:

Provide the family with informational counseling regarding hearing test results, even if incomplete, so that they understand the current status and next steps. The family should be informed that specific findings and recommendations will be conveyed through a written report following complete analysis. If hearing loss is confirmed, provide unbiased family-centered counseling. Refer to Appendix on effective counseling techniques for family-centered adjustment counseling and initiating Early Intervention.

2. Initiation of referrals and intervention:

If the assessment is complete and indicates sensory hearing loss, intervention should proceed as soon as possible. If the assessment is incomplete, indicates a conductive component, neural involvement or there are inconsistent test results, repeat ABR should be scheduled as soon as possible for further information and confirmation, prior to determining an intervention plan. Appropriate medical referral and intervention should not be delayed if abnormal hearing is suspected, simply in order to obtain complete results. If results are insufficient to achieve audiologic diagnosis of type and degree of hearing loss after two unsedated ABRs, and the child is older than 3 months, referral for a sedated ABR is strongly recommended in order to prevent delay in diagnosis and intervention.

3. Follow-up recommendations:

- a. A referral to general or pediatric otolaryngology is recommended if loss is minimal in a well-baby or due to conductive hearing loss, and caregivers do not need additional support.
- b. A referral to pediatric otolaryngology is recommended if mild or greater hearing loss is diagnosed, especially if amplification is recommended. This will facilitate assessment of etiology and medical clearance.
- c. Referral to a multidisciplinary team referral is recommended if neural involvement is suspected, if there are craniofacial or other syndromic stigmata, or high risk factors (refer to high risk factor list below)
- d. An informational packet regarding hearing loss will be offered to caregivers, including handouts such as Early Intervention program information, Familiar sounds audiogram, Parent to parent support (Hands and Voices – www.handsandvoices.org), hearing loss brochures, etc.
- e. Discuss appropriate amplification options and explain the process to obtain amplification.

- f. Referrals should be made to the Early Intervention program for children under 3 years of age who are deaf or hard of hearing.
- g. Behavioral testing should be performed to confirm/monitor hearing loss as soon as the patient is developmentally able to participate in VRA testing (usually by 8 months of age).
- h. Refer to genetics and ophthalmology by age 2 years, consistent with JCIH, 2007 recommendations.

4. Follow-up recommendations for conductive hearing loss:

- a. When making a referral to ENT for conductive hearing loss due to suspected fluid, counsel the family that if a middle ear problem is found and even if tubes are placed, repeat audiologic testing will be needed to establish normal hearing after medical intervention.
- b. Schedule follow-up testing in 6-8 weeks regardless of ear status to document need for intervention.
- c. Clearly state the need for audiologic follow-up after treatment, including tube insertion, since underlying permanent hearing loss could be present.

5. Follow-up recommendations for normal ABR with risk factors (JCIH, 2007):

- a. Higher Risk Factors: Earlier and more frequent monitoring is recommended (eg. every 6 months).
 - i. Caregiver concern regarding hearing, speech, language, or developmental delay
 - ii. In-utero infections (Congenital cytomegalovirus (CMV) infection)
 - iii. Culture-positive postnatal infections associated with sensorineural hearing loss (Bacterial and viral meningitis); syndromes associated with progressive hearing loss
 - iv. Syndromes associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson
 - v. Neurodegenerative disorders
 - vi. Extracorporeal Membrane Oxygenation (ECMO)
 - vii. Head trauma, especially basal skull/temporal bone fracture that requires hospitalization
 - viii. Chemotherapy
- b. Other Risk Factors: Repeat audiologic testing is indicated at least once by 24-30 months of age. Testing can include behavioral and/or objective testing measures.
 - i. Family history of permanent childhood hearing loss.
 - ii. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix).
 - iii. Hyperbilirubinemia that requires phototherapy.

6. Documentation:

- a. Necessary documentation, including ODH paperwork, is completed by the audiologist no later than 7 days after the appointment.
- b. Corrected thresholds should be reported where indicated (dB eHL).
- c. No Response ABR: When a "no response" is obtained, indicate the level at which there was no response.
- d. The copies of the written report will be mailed and faxed to the primary care providers and mailed to the caregiver. Calling the PCP to answer questions and provide guidance is extremely helpful to reinforce need for quick action.

- e. The audiologist should be available to the family for counseling and education by phone following receipt of the written report so that questions can be answered. Scheduling a follow-up counseling session with behavioral validation is recommended.
- f. The Ohio Department of Health (ODH) electronic UNHS Follow-up Hearing Evaluation Reporting Form is completed on all infants screened in Ohio regardless of residency no later than 7 business days after the appointment.
- g. For Kentucky residents the UNHS Program Form for Commission for Children with Special Health Care Needs form is completed. The form is available at http://chfs.ky.gov/NR/ronlyres/EC55F3B1-4D3F-4548-80F8-26300F36347F/0/Hearing_Screen_AUF_Blank.pdf
- h. For Indiana residents, the Diagnostic Audiology Evaluation (DAE) form is completed and can be obtained at [http://www.in.gov/isdh/files/Diagnostic_Audiology_Form_\(DAE\).pdf](http://www.in.gov/isdh/files/Diagnostic_Audiology_Form_(DAE).pdf)
- i. For Pennsylvania residents, use the diagnostic audiology report template, Newborn Hearing Screening Program by telephone at 717-783-8143; or by email at nbhs@pa.gov
- j. For West Virginia residents, use the Audiological Evaluation Form, available at http://www.wvdhhr.org/nhs/provider/NHS_Referral_Diagnostic_Audio_Evaluation_Form.pdf

7. Confirmation of Hearing Loss

- a. A subsequent, confirmatory evaluation may be recommended in a timely manner to minimize stress and anxiety for the family and to ensure timely transition of the infant from the diagnostic to intervention phase, so that the 1, 3, 6 guidelines are met.
- b. It is recommended that all diagnostic testing be conducted by the same audiologist, when possible to enhance continuity of care and to encourage the development of rapport and trust, and minimize the likelihood of receiving different interpretations of the results from different audiologists.
- c. Collaboration among audiologists, such as peer review, is important.
- d. Both informational and adjustment counseling for caregivers of newly identified infants with hearing loss should be provided.
- e. Written documentation and unbiased informational literature should be provided to the caregivers/family.
- f. Caregivers should receive information regarding the need for medical evaluation and diagnosis.
- g. Caregivers should receive information regarding the availability and importance of parent to parent support.
- h. Caregivers should receive information and referral for funding assistance in all cases. Do not make any assumptions about family's ability to pay for hearing aids privately.
- i. With consultation with the infant's PCP, the infant should be referred to an otolaryngologist for medical assessment.
- j. If appropriate, discuss additional specialty evaluations (e.g., genetics, ophthalmology, developmental pediatrician) with caregivers and the infant's PCP.
- k. If appropriate, initiate the amplification process and ensure that medical clearance for amplification has been obtained.

8. Periodicity Schedule for Evaluation:

- a. After hearing loss is diagnosed, routine audiologic evaluation should occur starting at age 6-8 months until full audiograms are obtained, and at six month intervals through age three.
- b. Due to rapid growth for infants, new earmolds may need to be obtained frequently.
- c. Immediate re-evaluation should be completed if caregiver concern is expressed or if behavioral observation by caregiver, therapist or teacher suggests a change in hearing.
- d. More frequent evaluation is appropriate when middle ear disease is chronic or recurrent or when risk factors for progressive hearing loss are present.

9. Referrals:

- a. Infants and children in the state of Ohio (birth to three) identified with hearing loss are referred to Early Intervention services.
- b. Early Intervention provides service coordination, eligibility assessments, development of the Individualized Family Service Plan (IFSP) and subsequent transition planning.
- c. Other referrals may be necessary that include genetics, speech-language pathology, neurology, ophthalmology, developmental pediatrics or other services.
- d. Infants who have one or more risk factors, should be evaluated at least once before 24-30 months of age.

10. Sharing information with Families

- a. Always build rapport at the beginning of the appointment, engage the caregiver/family in the appointment, care and communication with their infant.
- b. Share why hearing is so important for every day, ongoing communication. Describe how we communicate and how hearing loss can impact communication acquisition.
- c. Understand and recognize the emotional impact that a non-pass hearing screening and/or diagnosis of hearing loss can have on a family.
- d. Provide results of the testing in caregiver friendly language and assess emotional readiness of caregivers to receive informational counseling. Remember to have sensitivity to the emotional status of the family/caregivers. Emotional counseling (Refer to Appendix for more information) may need to precede informational counseling.
- e. Provide family support resources via Hands and Voices national website.
- f. Emphasize the importance of communication and explain how sign language, amplification and/or cochlear implants can enhance language learning and communication development.
- g. Share the benefits of early identification paired with early intervention. Talk about brain development and how a language rich environment is needed to develop language and communication.
- h. Recognize that families may not be emotionally ready to discuss communication options, and schedule additional appointments as needed to discuss the variety of options.
- i. Before the initial appointment ends, remind caregivers/families that infants and children identified with hearing loss early can have excellent language and communication ability. Provide reassurance and emphasize the value of family-family support locally, if available, or through Hands and Voices national website.
- j. At future appointments, share unbiased information about communication options. Be sensitive to the family's needs and desires as well as, language and culture. Provide all communication options and discuss the benefits of each.
- k. Additional follow up appointments may include counseling visits, social work referrals, etc.
- l. Provide packet of educational materials on hearing loss, non-biased communication choices, and Early Intervention resources available to infants.

11. Diagnostic follow up reporting:

- a. UNHS Ohio Administrative Code (OAC; Rule 3701-40-08) requires follow up reports to be completed and send to ODH within 7 days of the completed evaluation.
- b. All follow up information should be complete and accurate and recommendations for follow up notes, including additional referrals.
- c. Reports are submitted electronically via a secure web based data system called HI*TRACK. Please follow the appropriate reporting for your site.
- d. Copies to caregiver, PCP, and other healthcare/educational provider as requested in writing by the caregivers.

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Appendix A

Sample Case History Form

Date: _____ Child's name: _____

Age: _____ Date of Birth: _____

Where was baby born? _____ Was baby transferred after birth? YES NO

If yes, Where? _____ Reason for transfer: _____

Parent(s) Name(s): _____

PCP name and Address: _____

Reason for referral/testing: _____

Newborn Hearing Screening results: Right: _____ Left: _____

Type of testing if known: OAE ABR OAE+ABR

Infant responses to sound noted? _____

Family History

Other children with hearing loss YES NO

If yes, who? _____ Age loss identified: _____

Family history of hearing loss in childhood YES NO

(parents, grandparents, aunts, uncles, cousins)

If yes, who? _____ Age loss identified: _____

Maternal/pregnancy History

Full-term pregnancy YES NO If, weeks premature: _____

Complications High Risk Gestational Diabetes Other _____

Cesarean section (C-section) YES NO

Medications/drugs taken during pregnancy (including street drugs) YES NO

If yes, list: _____

Neonatal abstinence (withdrawal) YES NO

Illnesses during pregnancy YES NO If yes, list: _____

Exposure during pregnancy: Chickenpox Measles Zika virus

Mumps Toxoplasmosis

During pregnancy, was mother diagnosed with: Syphilis Herpes

Influenza Cytomegalovirus (CMV)

HIV/AIDS

Birth history

Birth weight: _____ APGAR score if known: _____

Medications given (eg., gentamicin, Lasix, etc.) _____

Intensive care (NICU) YES NO If yes, how long: _____

Breathing problems at birth YES NO

Oxygen at birth or later YES NO If yes, specify type and how long: _____

Jaundice YES NO Phototherapy YES NO

Exchange Transfusion YES NO Congenital heart disease YES NO

Congenital CMV YES NO If yes, list problems _____

Defects of ear, head or neck YES NO (e.g., dysmorphic appearance, cleft palate, ear tags or pits)
If yes, explain: _____

Paralysis at birth YES NO Seizures at birth YES NO

Cerebral palsy YES NO Head/skull injury YES NO

Brain injury (eg. hemorrhage) YES NO Eye problems YES NO

Other diagnoses: _____

Illnesses since birth

Meningitis	Encephalitis	Measles	Influenza	Cytomegalovirus
(CMV) Chickenpox	Septicemia	Diabetes	Sickle Cell	Rubella
Ear infection	RSV	Failure to thrive	Reflux	High fever

List any hospitalizations or surgeries: _____

Doctors/other specialists

Please list other doctors and/or specialists that treat your child:

Appendix B: ABR (Hearing) Testing Instructions

Your baby/child has been scheduled for an Auditory Brainstem Response (ABR) evaluation. It is the most accurate way to measure hearing in infants who are too young for other tests.

Please arrive 15 minutes prior to your appointment. If you are late for the appointment, we may not be able to complete testing and will need to reschedule. Your appointment date and time is:

_____. The appointment will last 1-2 hours.

IMPORTANT INSTRUCTIONS TO FOLLOW BEFORE ARRIVING:

- ***Please keep your baby awake on the way to the appointment, so she or he will be tired and able to sleep during testing. To obtain clear results, it is best if your baby is asleep during the test.***
- ***Tips to keep your baby awake: Skip a nap before the appointment, entertain your baby with toys, dab a cool washcloth on face, wiggle hands or feet and talk to your baby.***
- ***Delay nursing or feeding until you arrive for the test to assist your child in falling asleep. Be sure to bring diapers, favorite cuddly, pacifier if needed, and anything else that will make your baby comfortable.***
- ***Please do not bring other children to this appointment, since you will be busy holding your baby during the test.***

About the test and test procedures:

The ABR test is a procedure that measures hearing and nerve work in each ear. This test does not rely on a baby's/child's ability to respond to sound. It measures how the hearing nerves send sounds to the brain.

Small recording sensors or patches will be placed on your baby's forehead and behind each ear. A small, soft ear tip will be placed in the ear to deliver sounds. The audiologist will measure responses to different sounds to check hearing function for your baby.

After the test, the audiologist will share the results with you and will offer any recommendations. Sometimes, further testing by the audiology or ENT clinic may be recommended.

Please call the clinic with any questions or concerns about the tests or to reschedule if your baby is ill the day of the test.

Name

Phone

Appendix C: Effective Counseling of Newly-Identified Hearing Loss

There are two main forms of counseling that audiologists need to be trained and practiced in providing effectively: 1) Informational-educational and 2) Emotional-behavioral counseling. Patients will not be able to absorb and understand information unless they are emotionally ready to hear it. Audiologists are generally more comfortable and prepared to provide education and information about hearing loss than to provide emotional and behavioral counseling.

Sharing unwelcome news requires using a specific approach and a sensitive, professional demeanor. The SPIKES model is one such approach of emotional counseling that has been developed specifically for medical professionals. A realistic and proactive approach is most effective to assist families with a new diagnosis. Families are encouraged by working with a professional who conveys confidence and is able to earn their trust. Families will have a range of emotions, usually ranging from sadness to disbelief to shock. In some cases they may be relieved or even happy if they are culturally Deaf themselves. This reaction can be surprising to audiologists, so being prepared for different reactions is helpful.

Unwelcome news is defined as information that is likely to alter a patient's view of his or her future (Buckman, 1992). This type of news includes situations where there is either a feeling of decreased hope, a threat to one's mental or physical well-being, a risk of upsetting an established lifestyle, or where the message given conveys fewer choices in his or her life. Professionals may have reasons for fear and anxiety when they need to explain a new diagnosis to families:

- Concern for how the news will affect family
- Family's fears of social stigma and impact of disability and illness
- Fear of family's reaction to the news
- Uncertainty in dealing with intense emotional response
- Fear of being blamed
- Fear of how this affects you and expressing emotion
- Challenge of delivering the news appropriately and sensitively
- Not wanting to take away hope

Setting up the interview: Arrange for privacy, involve significant others, sit down, make a personal connection with the patient, avoid time constraints and interruptions.

Perception of the patient: "Before you tell, ask" e.g. "What have you been told about your baby's screening so far?" "What is your understanding of why we did the ABR?" Determine family's expectations, and assess their understanding of the reasons for assessment.

Invitation by the patient: "How would you like me to give you information about the test results?" If patient does not want to know details, involve a relative or friend. The majority of patients express a desire for full information. Discuss information disclosure (who else should receive results) while planning for referrals to other professionals (primary care, ENT, social work, intervention, genetics).

Knowledge of the patient: Start by providing an introduction e.g. "I have some results to go over with you". The results do show hearing loss (describe in one or both ears and say severity). Share that is a permanent loss. Provide reassurance that their baby will still grow and develop, but will need some early help to learn to communicate. Make this a positive statement. Practice certain lines or language. Start at level of

comprehension of the patient; use nontechnical words such as “inner ear or nerve” instead of “cochlea or brainstem”; avoid being blunt, but do be realistic and positive about benefits of early intervention *Wait to see reaction before giving any additional information.*

Emotions of the patient: Respond to patients’ emotions with empathy. Often they will respond with shock (silence, numb look), isolation (looking down, lack of eye contact), disbelief (questioning the diagnosis), grief (crying or appearing sad) or denial (challenging the diagnosis). Observe for these emotions on the patient’s part. Identify the emotion, by affirming what you observe (I can tell you might be feeling upset). Identify the reason for the emotion (It makes sense to feel his way, you may not have been expecting this today). Connect with the patient (Listen and wait for them to say something, or offer a tissue, pat on the shoulder, ask if they would like to take a little break and you can come back to talk some more. Empathic statements can be helpful, such as:

- I can tell this is not what you expected to hear
- I understand you may have been expecting different results

Techniques for talking through emotions: Use exploratory questions, such as: How do you mean? Tell me more about it; you said it frightens you; you said you were concerned about your child; tell me more; could you tell me what you are worried about?

Techniques for validating responses: I can understand how you felt that way; I guess anyone might have the same reaction; you are perfectly correct to think that way; your understanding of the reason for the tests is good; many other patients have had a similar experience.

Strategy and summary

- Ask if family is ready to hear more information
- Ask if they would like to discuss next steps
- Explore patients’ ideas, concerns and expectations
- Do not minimize results, be clear about findings
- Patients who have a plan for the future are less likely to feel anxious and uncertain
- Discuss goals family may have, like hearing their child’s voice, learning sign language, or learning to read
- Explain the positive outcomes of language development and communication when intervention starts early Set a follow-up time to talk over the phone or in person in the next 2-3 days

Written Information

Prepare a family-friendly discharge summary (these can be templates that are customized as needed).

It is enormously helpful to have a ready packet of easy to understand information that the family can take home.