Background:
Since 1997, the number of newborns screened for hearing loss in Michigan has improved from 5% to 98%. As part of the “1-3-6” goals, the Joint Committee on Infant Hearing (JCIH) recommends that infants have a complete diagnostic evaluation no later than three months of age. In 2012, only 54% of Michigan-born infants were diagnosed with hearing loss by this benchmark. In an effort to improve the age of identification of hearing loss, this “Best Practice Guideline” was created. The purpose of this document is to provide direction from the time of referred hearing screen until diagnosis of hearing loss is confirmed with the goal of ensuring consistent, quality care across the state.

Infant Re-screen Facility
If an infant refers on a newborn hearing screen (in either one or two ears), it is recommended that follow-up testing be done as soon as possible or no later than one month of age to meet the 1-3-6 National EHDI goals. The next step in testing might be done as either an outpatient rescreen or as a comprehensive assessment through a designated pediatric diagnostic center*1. Similarly, a child who does not receive a newborn hearing screening should follow the same procedures.

For purposes of this document, an EHDI Rescreen Facility is defined as one which offers a hearing screen following a nonpass (refer/missed) result at a birth hospital. A rescreen facility may or may not offer diagnostic evaluations and/or be staffed by an audiologist. A rescreen facility plays a specific role within the EHDI process by offering timely screening within a reasonable distance to families. As such, the minimal equipment requirements and level of professional staff training are expected to be different from that of a diagnostic facility. The role of a rescreen facility is identical to the EHDI screen – to determine if full diagnostic evaluation is warranted.

Auditory Brainstem Response (ABR) is the preferred method for screening whenever feasible. A disorder such as Auditory Neuropathy Spectrum Disorder (ANSD) is a type of hearing loss in which the auditory pathway is impaired, but Otoacoustic emission (OAE) may be present. Infants with a Neonatal Intensive Care Unit (NICU) stay are at greater risk for ANSD. Therefore, all NICU graduates should have hearing screened using ABR. Depending upon the expertise of the person completing a rescreen, the screen may be done using an automated ABR (A-ABR) or through standard (non-automated) testing.

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*1 This list is available at www.michigan.gov/documents/mdch/PedAudFeb2008_223684_7.pdf.

Updated 6/2014
Facilities meeting the criteria below would be eligible for inclusion on the Michigan EHDI re-screen center list.

Minimal Facility Requirements:
- Objective hearing screening equipment, i.e., Otoacoustic emission (OAE), Auditory Brainstem Response (ABR) or Automated ABR (A-ABR).
- Routine calibration of hearing screening equipment per manufacturer’s recommendations.

Minimal Staff Requirements:
- Professional staff with experience using equipment. An appropriate level of experience includes knowledge of appropriate test set up, ability to verify equipment is functioning and basic troubleshooting. As part of training, hands-on supervised screening of at least 5 infants is recommended.
- Well-versed in counseling families regarding results, whether pass or refer, and the importance for further evaluation.

Suggested Rescreen Procedure:
If an outpatient rescreen is completed, the following is considered best practice:
- Rescreen may use OAE, ABR (automated or standard) or combination of both, although ABR technology is preferred. ABR must be completed in the following situations:
  - When the initial hearing screen is completed with ABR (automated or standard).
  - Infant with a NICU stay greater than 5 days.
- **Multiple screens/rescreens are not recommended.** In some cases, middle ear effusion may prevent an infant with normal hearing from passing a hearing screen. Diagnostic evaluation should not be delayed in these cases. Diagnostic evaluation including bone conduction ABR testing can provide further information regarding the degree and nature of hearing loss.
- Regardless of initial screening results, both ears must be rescreened.
- If the baby passes the rescreen in both ears, determine if the infant is at risk for late onset hearing loss (Appendix A) and recommend monitoring audiograms as appropriate. If no risk factors are identified, infant can be released.
- Immediate referral for a diagnostic audiologic evaluation is warranted for any of the following outcomes:
  - Rescreen result is a “refer” in one or both ears.
  - Unilateral referral in which the ear that refers changes from the first to the second rescreen (i.e., initially refer right/pass left, followed by pass right/refer left).
  - Upon completion of an OAE and ABR and the infant “refers” on ABR (referral should be made regardless of OAE result.)

Diagnostic audiologic evaluation should be scheduled as soon as possible and no later than 3 months of age. There is a short window in which natural sleep diagnostic evaluations can be easily accomplished, continual rescreens delay diagnosis and may necessitate sedation which could be avoided. It is strongly recommended that infants be referred to a designated infant diagnostic center. As mandated by law, rescreen results must be reported to the Michigan EHDI Program by faxing to 517-335-8036.

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2 In the case of premature birth (infants born at less than 36 weeks gestational age), corrected chronological age should be used.
Infant Diagnostic Facility
Best Practice Guidelines

As stated by the Joint Committee on Infant Hearing (JCIH), comprehensive audiological evaluation of infants who fail newborn hearing screen should be performed by audiologists experienced in pediatric hearing assessment who utilize an evidence-based diagnostic protocol. Audiologists working with pediatric populations should be skilled at communicating with families and have thorough knowledge of support systems available for families of children with hearing loss. In addition to the requirements listed below, the following factors are also noted to be of importance:

- Desire to work with infants and their families.
- Staffed trained in counseling families.
- Familiarity with state resources (i.e. Children's Special Health Care Services (CSHCS), Guide By Your Side, Early On).

Facilities meeting these criteria would be eligible for inclusion on the Michigan EHDI Infant Diagnostic Center List.

1. Equipment Requirements:
To complete a comprehensive diagnostic evaluation on an infant less than six months of age, the following equipment is recommended:

- Diagnostic Auditory Brainstem Response (ABR) equipment capable of click, tone burst and bone conduction testing.
- Diagnostic Otoacoustic Emission (OAE) (Distortion Product or Transient Evoked OAE).
- High Frequency Tympanometer.
- Process to obtain ABR with sedation if needed. If facility cannot provide testing with sedation, documentation of appropriate referral source is needed to ensure this service is provided in a timely fashion.

2. Recommended Diagnostic Evaluation Battery:
The importance of using a battery of tests to assess an infant's hearing status is widely recognized. The following are considered to be minimal components of an infant diagnostic evaluation whenever hearing loss is suspected. As appropriate, the following elements should be obtained for each ear whenever possible. It is recognized that in some instances, this testing may not be able to be completed (i.e., air conduction thresholds on atretic ear). These elements were selected as each is needed to appropriately fit an infant with hearing aids.

- Case History to include family and medical history.
- Otoscopy.
- Diagnostic OAE.
- ABR Thresholds
  - Click and Tone Burst stimuli which represent at least one low frequency and one high frequency.
  - Additional tone burst thresholds and /or Auditory Steady-State Response (ASSR) information as time allows.
- Bone Conduction thresholds by ABR (preferred) and/or high frequency tympanometry (for infants less than 6 months of age) to rule out conductive and mixed hearing loss.
- Recording of cochlear microphonic (preferred) and/or acoustic reflex testing to rule out neural hearing loss (i.e., auditory neuropathy spectrum disorder, VIII nerve aplasia).
With regards to ABR testing, it is essential for each clinic to establish their own normative data. As a guide, the EHDI advisory committee recommends that ABR thresholds greater than 25 dBnHL be considered outside the normal range of hearing.

3. Minimal Staff Requirements:
Specialized skills and experience are important to accurate diagnosis of infants with hearing loss. The following criteria have been established in an attempt to quantify the level of experience and skills needed to provide expert and timely diagnosis of hearing loss in infants.

Facilities meeting these criteria would be included on the Michigan EHDI diagnostic center list.
- Michigan audiology licensure.
- At least one staff member has two years of experience working with children. Mentorship of staff members with less experience is encouraged.
- Experience and expertise in assessment of hearing in infants, defined as (must meet at least 3 criteria):
  - > 20% of client population is younger than 24 months.
  - On average each week, > 3 patients under the age of 24 months.
  - At least two diagnostic threshold ABRs completed each month.
  - Identification of hearing loss in children less than 12 months of age should be commensurate with area birth rates (i.e., not less than 1-3 babies with hearing loss per 1,000 births) *Refer to Appendix B for further information.

Further recommendations:
- Recommend that 30% of continuing education hours are pediatric focused.

4. Recommended Procedures Following Diagnosis of Hearing Loss:
Once hearing loss has been confirmed, the role of the audiologists does not end. In order to reach the final goal of the EHDI program (intervention no later than 6 months), the following is recommended:
- Provide families with unbiased written and/or electronic resources regarding hearing loss.
- Notify Pediatrician/Primary Care Physician of results.
- Refer to Otolaryngologist with pediatric experience.
- Schedule monitoring appointments to rule out progressive hearing loss (minimally every six months).
- As mandated by Michigan law, audiometric results are required to be reported to Michigan EHDI at 517-335-8036. Report form found at: http://www.michigan.gov/documents/FORMAUDMED_53429_7.pdf.
- Referral for early intervention services (Early On / Special Education).
- As needed:
  - CSHCS application.
  - Hearing aid evaluation with audiologist experienced in fitting infants.

5. Required Protocols/Tracking:
Each facility should have a written best practice protocol. The purpose of the protocol is to ensure evidence-based practice that is consistent across clinicians within a single practice. The exercise of completing a protocol provides an opportunity to discuss possible scenarios to improve clinical decision making. As with any protocol, situations will arise in which deviations from the standard protocol are required.
Michigan EHDI recognizes the unique challenges involved in the diagnostic evaluation of infants and views a written protocol as a necessary component of a quality diagnostic facility. In order for a facility to be included on the Infant Diagnostic Center List, a copy of the protocol must be provided to Michigan EHDI. The protocol should include the following:

- Outline of steps from time of referral on EHDI screen to final diagnosis with timeframe consistent with National EHDI goals.
- Protocol that lists collection parameters for diagnostic ABR.
- Pass/Refer criteria for OAE measurements.
- Established routine for reporting to Michigan EHDI.
- Established routine for making referrals as listed in #4.
- List of risk indicators for delayed onset hearing loss and monitoring schedule.
- Routine monitoring and tracking of infants seen in facility to ensure timelines set forth in protocol are consistently being met.
- Schedule for equipment calibration used for diagnostic evaluations.
Appendix A

Centers are encouraged to use a defined set of risk indicators to identify those infants at risk for delayed onset or progressive hearing loss. Risk indicators may be based upon evidence-based practice or published recommendations such as those listed below.

Risk Indicators Associated with Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss in Childhood:

Excerpt from: Joint Committee on Infant Hearing Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs.

Risk indicators that are marked with an *** are of greater concern for delayed-onset hearing loss.

- Caregiver concern* regarding hearing, speech, language, or developmental delay.
- Family history* of permanent childhood hearing loss.
- Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO*, assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.
- In utero infections, such as CMV*, herpes, rubella, syphilis, and toxoplasmosis. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
- Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss. 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.
- Neurodegenerative disorders*, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
- Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
- Head trauma*, especially basal skull/temporal bone fracture that requires hospitalization.
- Chemotherapy*.
Appendix B

Calculation of expected number of hearing loss cases in your specific area (per live birth rate).

Rationale:
The incidence of permanent hearing loss is 1-3 infants per 1,000 live births. For this reason, the actual number of infants diagnosed with hearing loss will vary across facilities based upon the number of live births in an area. To determine if the number of infants diagnosed with hearing loss at your facility reflects prevalence of hearing loss in Michigan, the following steps are recommended:

1. Determine the number of births in your area by referring to the “Vital Statistics” website located at: http://www.mdch.state.mi.us/pha/osr/Chi/Births/frame.asp. Depending upon the location of your facility, select the county/countyies served by your facility. If appropriate, add the births across counties.

   Example:
   Ingham County 2008 live births= 3,538

2. Complete the following calculations:
   Multiply:
   \[(\text{Total Births}) \times .001 = A\]  Example: \(3,538 \times .001 = 3.5\) infants
   \[(\text{Total Births}) \times .003 = B\]  Example: \(3,538 \times .003 = 10.5\) infants

3. The expected number of infants with hearing loss for your area, in the specified time period (i.e. 2008) will be a range from A to B.

   Example:
   The expected number of infants with hearing loss for Ingham County in 2008 is 3-11 infants.
To be included on the *EHDI Infant Diagnostic Facility* List, please complete this application. Applications and supporting documents can be submitted to Michelle Garcia’s attention by email or fax. EHDI will contact you with questions or approval within 2 weeks upon receipt of application.

If approved as an Infant Diagnostic Center, your facility will also be included on the *EHDI Rescreen Facility* listing.

Please refer to the *EHDI Infant Diagnostic Facility Best Practice Guidelines* for additional information. Inclusion on the list is voluntary.

The EHDI staff is available to make a site visit to facilities requesting assistance in developing protocols and applying for inclusion in the State of Michigan’s *Infant Diagnostic Facility* listing. If you would be interested in assistance, please contact:

Michelle Garcia, Au.D., CCC-A  
EHDI Follow-Up Consultant  
garciam@michigan.gov  
Ph: 517-335-8878  
Fax: 517-335-8036
EHDI Infant Diagnostic Facility
Application

**Equipment:** Please indicate whether the following equipment is available at your facility.

<table>
<thead>
<tr>
<th></th>
<th>Yes</th>
<th>No</th>
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<tbody>
<tr>
<td>Diagnostic Auditory Brainstem Response (ABR) with Click Stimuli.</td>
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<tr>
<td>Diagnostic ABR with Tone Burst Stimuli.</td>
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<tr>
<td>Diagnostic ABR with Bone Conduction Stimuli.</td>
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<td>Diagnostic Otoacoustic Emission (OAE) (Distortion Product or Transient Evoked OAE).</td>
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<tr>
<td>High Frequency Tympanometer.</td>
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**Sedation:**

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<th>Yes</th>
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<tr>
<td>Is sedation available if needed to complete ABR testing at your facility?</td>
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<td>If sedation is not available, is there a facility to which you refer for sedation?</td>
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<td>Please indicate facility: __________________________.</td>
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**Staff Requirements:** Please indicate whether the following statements accurately describe the staff at your facility.

<table>
<thead>
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<tr>
<td>Valid Michigan Audiology Licensure.</td>
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<td>One staff member with at least two years of experience working with children.</td>
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**Client Population:** Please indicate whether the following statements accurately describe the client population at your facility.

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<td>&gt; 20% of client population is younger than 24 months.</td>
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<td>On average each week, &gt; 3 patients under the age of 24 months.</td>
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<tr>
<td>At least two diagnostic threshold ABRs completed each month.</td>
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<tr>
<td>Identification of hearing loss in children less than 12 months of age is commensurate with area birth rates.</td>
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Please include a copy of your facility's written best practice protocol clinic protocol related to the diagnostic evaluation of infants.

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3 Rate of identification should not be less than 1-3 babies with hearing loss per 1,000 births. Refer to EHDI Infant Diagnostic Facility Best Practice Guidelines, Appendix B for further information.
To be included on the *EHDI Rescreen Facility List*, please complete this application. Applications and supporting documents can be submitted to Michelle Garcia’s attention by email or fax. EHDI will contact you with questions or approval within 2 weeks upon receipt of application.

Facilities which have completed an application as an Infant Diagnostic Center will automatically be included on the rescreen center listing. It is not necessary to complete this application if your facility has already completed an Infant Diagnostic Center application.

Please refer to the *EHDI Rescreen Facility Best Practice Guidelines* for additional information. Inclusion on the list is voluntary.

The EHDI staff is available to make a site visit to facilities requesting assistance in developing protocols and applying for inclusion in the State of Michigan's EHDI Rescreen Facility listing. If you would be interested in assistance, please contact:

Michelle Garcia, Au.D., CCC-A
EHDI Follow-Up Consultant
garciam@michigan.gov
Ph: 517-335-8878
Fax: 517-335-8036
EHDI Rescreen Facility
Application

**Equipment:** Please indicate whether the following equipment is available at your facility.

<table>
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</table>
Automated Auditory Brainstem Response (A-ABR).
Auditory Brainstem Response (ABR).
Otoacoustic Emission (OAE).
Calibration schedule for above equipment.

**Staff Requirements:** Please indicate whether the following statements accurately describe the staff at your facility.

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</table>
Professional staff with experience using equipment. An appropriate level of experience includes knowledge of appropriate test set up, ability to verify equipment is functioning and basic troubleshooting. As part of training, hands-on supervised screening of at least 5 infants is recommended.
Well-versed in counseling families regarding results, whether pass or refer, and the importance for further evaluation.

**Please indicate if the following are included in your facility’s rescreen procedures:**

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Regardless of initial screening results, both ears are rescreened.
A-ABR or ABR is used for rescreen whenever possible and always used if initial screening was completed using ABR.
Re-screen will be completed one time before referral for diagnostic evaluation.
In the case of a “pass” rescreen, risk factors for late onset hearing loss are reviewed.
Rescreen results are reported to Michigan EHDI.

**Please indicate which of the following warrant referral for full diagnostic evaluation:**

<table>
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</table>
Rescreen result is “refer” in one or both ears.
Unilateral referral in which the ear that refers changes from the first to the second rescreen (i.e., initially refer right/pass left, followed by pass right/refer left).
Upon completion of an OAE and ABR and the infant “refers” on ABR.

**Referral Facilities:**
If an infant is at risk for late onset hearing loss, please indicate referral facility:

If diagnostic evaluation is warranted, please indicate referral facility: