Clinical Policy Title: Brainstem auditory evoked response

Clinical Policy Number: 09.01.06

Effective Date: October 1, 2014
Initial Review Date: May 21, 2014
Most Recent Review Date: May 1, 2018
Next Review Date: May 2019

Policy contains:
- Auditory neuropathy.
- Auditory brainstem response.
- Hearing loss.
- Evoked responses.

Related policies:

CP# 17.02.02 Altered auditory feedback devices for speech dysfluency (stuttering)
CP# 10.02.03 Non-pharmacologic treatments for chronic vertigo
CP# 02.01.18 Genomic tests in sensorineural hearing loss
CP# 15.02.05 Speech generating devices

ABOUT THIS POLICY: Select Health of South Carolina has developed clinical policies to assist with making coverage determinations. Select Health of South Carolina’s clinical policies are based on guidelines from established industry sources, such as the Centers for Medicare & Medicaid Services (CMS), state regulatory agencies, the American Medical Association (AMA), medical specialty professional societies, and peer-reviewed professional literature. These clinical policies, along with other sources, such as plan benefits and state and federal laws and regulatory requirements, including any state- or plan-specific definition of “medically necessary,” and the specific facts of the particular situation are considered by Select Health of South Carolina when making coverage determinations. In the event of conflict between this clinical policy and plan benefits and/or state or federal laws and/or regulatory requirements, the plan benefits and/or state and federal laws and/or regulatory requirements shall control. Select Health of South Carolina’s clinical policies are for informational purposes only and not intended as medical advice or to direct treatment. Physicians and other health care providers are solely responsible for the treatment decisions for their patients. Select Health of South Carolina’s clinical policies are reflective of evidence-based medicine at the time of review. As medical science evolves, Select Health of South Carolina will update its clinical policies as necessary. Select Health of South Carolina’s clinical policies are not guarantees of payment.

Coverage policy

Select Health of South Carolina considers the use of brainstem auditory evoked response to be clinically proven and, therefore, medically necessary as a primary (first-line) test when any of the following criteria are met (American Academy of Family Practice [AAFP], 2013; Ptok, 2011; Harlor, 2009; American Academy of Pediatrics [AAP], 2007; New York State Department of Health [NYSDH] Early Intervention Program, 2007):

- For initial screening for hearing loss in newborns (using limited auditory evoked potentials only).
- To assess infants and children under age 5 years for hearing loss, for one of the following conditions:
  - When pure tone screening is not developmentally appropriate (ability levels less than 36 months).
  - When the member passed neonatal hearing screening, but is at risk of having sensorineural hearing loss.
- When the member did not pass the initial hearing screening and requires diagnostic confirmation of a hearing disorder.

- To assess infants and children suspected of having a hearing disorder when either:
  - Behavioral audiometry is not reliable.
  - Ear-specific thresholds cannot be obtained.
  - Results from other tests are inconclusive regarding the type, degree, or configuration of hearing levels.

- To assess suspected hearing disorders in individuals of any age who are unable to cooperate in other methods of hearing testing (e.g., behavioral audiometry).

- To assess acoustic neuroma in patients for whom magnetic resonance imaging is contraindicated or results are equivocal (Stachler, 2012; American College of Radiology [ACR], 1996).

- To determine the degree and configuration of hearing deficiency in each ear for the fitting of amplification devices using frequency-specific brainstem auditory evoked response testing in persons with a permanent hearing deficit.

- To assess the auditory system through the level of the brainstem when its neurological integrity is in question.

Select Health of South Carolina considers the use of brainstem auditory evoked response to be clinically proven and, therefore, medically necessary when primary (standard) testing fails to provide a diagnosis for any of the following clinical conditions (American Society of Neurophysiological Monitoring, 2014; AAP, 2007; American Speech-Language-Hearing Association [ASHA], 2005):

- Cerebellopontine angle lesions (acoustic neuromas).
- Demyelinating disease, such as multiple sclerosis.
- Chiari malformation and syringomyelia.
- Asymmetric hearing loss.
- Unilateral tinnitus.
- Sudden hearing loss.
- Functional hearing loss.
- Ototoxic drug therapy monitoring, including chemotherapy or antibiotics.
- Auditory neuropathy.
- Preoperative baseline: posterior fossa surgery or cochlear implant.
- Postoperative testing for cochlear implant.

**Limitations:**

All other uses of brainstem auditory evoked response are not medically necessary.

**Alternative covered services:**

- Acoustic immittance measures.
- Conventional and high-frequency audiometry.
• Electrocochleography.
• Electroencephalography.
• Electromyography.
• Gadolinium-enhanced magnetic resonance imaging.
• Motor-evoked potentials.
• Otoacoustic emissions testing.
• Somatosensory-evoked potentials.
• Speech recognition tests.
• Tympanometry.
• Visual-evoked potentials.

**Background**

Hearing loss is a major public health issue and is the third most common such affliction after arthritis and heart disease. According to the National Institute on Deafness and Other Communication Disorders (NIDCD), an estimated two to three of every 1,000 children in the United States are born deaf or hard-of-hearing, and approximately 15 percent (37.5 million) of American adults report some degree of hearing loss, with the prevalence of reported hearing loss increasing with age (NIDCD, 2016).

The prevalence of newborns with congenital hearing loss in the United States varies between one and six per 1,000 newborns (Cunningham, 2003). Most children with congenital hearing loss have hearing impairment at birth and are potentially identifiable by newborn and infant hearing screening. However, some congenital hearing loss may not become evident until later in childhood (Cunningham, 2003).

The hearing process can be divided into sound conduction, transformation of sound waves into bioelectrical signals, and neural processing. Hearing impairments can be classified as one or a combination of these functions as follows (Ptok, 2011):

- Impairments of conduction (defective transport of sound waves from the external environment to the inner ear).
- Sensory impairment (defective sensation and transformation of stimuli between the base of the stapes and the first neuron of the auditory nerves).
- Retrocochlear and central hearing impairment and auditory perception disorders (defective transmission, processing and perception of stimuli).
- Combined hearing impairments.

A variety of tests can be used to identify and diagnose a hearing loss. The method used depends in part on the age and competency of the individual and clinical indication. Behavioral pure tone audiometry is the standard for hearing evaluation (Cunningham, 2003). Other tests include:

- Speech testing.
- Tests of the middle ear.
- Otoacoustic emissions.
- Brainstem auditory evoked response.
Brainstem auditory evoked response:

Brainstem auditory evoked response measures auditory nerve and auditory pathway structural integrity in the brainstem (Evans, 2014). Its measurements reflect the status of the auditory (cranial) nerve and pathways and peripheral auditory system, not for identifying central hearing deficits. Infants with risk factors for central hearing deficits may pass newborn hearing screens but develop hearing loss in early infancy. Brainstem auditory evoked response is also referred to as auditory brainstem response, auditory evoked response, auditory evoked potential, evoked auditory potential, brainstem auditory evoked potential, and brainstem evoked response audiometry.

The test involves placing electrodes on the scalp and earlobes. Auditory stimuli, such as tones or clicking noises, are delivered to one ear. The sound stimulation moves through the outer ear (canal), through the middle ear (tympanic membrane and ossicles) to the inner ear (cochlea), through the vestibular and eighth cranial nerve to the brain. The electrodes sense an electrical response from the brainstem (Evans, 2014).

The rationales for using brainstem auditory evoked response over standard auditory testing are (Evans, 2014):

- Its resistance to alteration by anything other than structural pathology in the brainstem auditory tracts (e.g., systemic metabolic abnormalities, medications, or pronounced changes in the state of consciousness of the patient).
- The close association of waveform abnormalities to underlying structural pathology. The short-latency test is used generally for clinical purposes and can be performed with the patient under either sedation or general anesthesia. However, testing accuracy may be compromised if the child is not sleeping or quiet, or by the presence of middle-ear effusion or debris in the external canal.

Searches

Select Health of South Carolina searched PubMed and the databases of:

- UK National Health Services Centre for Reviews and Dissemination.
- The Centers for Medicare & Medicaid Services (CMS).

Searches were conducted on March 19, 2018. Search terms were: “brainstem auditory evoked response,” “brainstem auditory evoked potentials,” “auditory brainstem response,” and "evoked potentials, auditory" (MeSH). These terms were crossed with free-text terms for “screening,” “monitoring,” “retrocochlear,” “vestibular schwannoma,” “acoustic neuroma,” “Chiari malformation,” and “coma.”

We included:
• **Systematic reviews**, which pool results from multiple studies to achieve larger sample sizes and greater precision of effect estimation than in smaller primary studies. Systematic reviews use predetermined transparent methods to minimize bias, effectively treating the review as a scientific endeavor, and are thus rated highest in evidence grading hierarchies.

• **Guidelines based on systematic reviews.**

• **Economic analyses**, such as cost-effectiveness, and benefit or utility studies (but not simple cost studies), reporting both costs and outcomes — sometimes referred to as efficiency studies — which also rank near the top of evidence hierarchies.

**Findings**

There is a great deal of evidence regarding brainstem auditory evoked response, adding weight to decisions relative to clinical support and use of this technology. Seven systematic reviews and one cost-effectiveness analysis were identified for this policy, covering its use in several clinically important domains:

• Early childhood screening and diagnosis for hearing impairment (three systematic reviews and one cost-effectiveness analysis).

• Diagnosing retrocochlear disease, specifically acoustic neuroma (two systematic reviews).

• Managing patients with Chiari malformation (one systematic review).

• Predicting outcomes in coma (one systematic review).

While the rationales for using brainstem auditory evoked response make it a potentially attractive option for many clinical indications, no systematic reviews were identified that would support other clinical uses. For other popular indications, namely, ototoxicity monitoring and intraoperative neuromonitoring, the evidence is confined to case series and anecdotes that suggest the feasibility of using brainstem auditory evoked response but are insufficient to determine diagnostic efficacy or impact on treatment management or patient outcomes.

**Universal early childhood screening for hearing impairment:**

Because approximately half of the children with hearing loss have no identifiable risk factors, several professional societies, including the AAFP and AAP, recommend universal screening (as opposed to targeted screening) in newborns prior to hospital discharge and in young children to avoid delaying diagnosis beyond the age of language acquisition, which may result in life-long psychological and cognitive handicaps (AAFP, 2013; Harlor, 2009; AAP, 2007).

Neonatal screening methods include a limited brainstem auditory evoked response test using a significantly low intensity level (35 to 40 dB) and otoacoustic emissions testing either alone or sequentially if the first test fails (Ptok, 2011). Infants who fail the first screening test are rescreened, and those who fail rescreening are referred for additional outpatient testing and diagnostic evaluation. Diagnostic evaluation may consist of repeat otoacoustic emissions screenings, comprehensive diagnostic brainstem auditory evoked response testing, behavioral audiometry at an appropriate age, tympanometry, and otoscopy (Ptok, 2011).
Brainstem auditory evoked response and otoacoustic emissions screening tests have comparable sensitivities and specificities when employed individually or in combination as part of a screening protocol. Limited evidence suggests early intervention using either test is associated with positive developmental outcomes (Wolff, 2010; NYSDH Early Intervention Program, 2007; Hayes, 2005). One cost-effectiveness analysis found otoacoustic emissions testing at birth followed by repeat testing at follow-up demonstrated the lowest cost ($13 per infant) and had the lowest cost-effectiveness ratio — $5,100 per infant with hearing loss identified (Kezirian, 2001). Screening brainstem auditory evoked response test at birth with no screening test at follow-up showed greater effectiveness, but was associated with higher costs ($25 per infant) and higher cost-effectiveness ratio ($9,500 per infant with hearing loss identified).

There is a lack of consensus among professional societies regarding the frequency of screening, the most appropriate tests for different age groups, and when brainstem auditory evoked response or otoacoustic emissions testing is appropriate outside of the screening setting (Ptok, 2011). The AAP Joint Committee on Infant Hearing recommends automated brainstem auditory evoked response technology as the only appropriate technique for screening infants in the neonatal intensive care unit (AAP, 2007). The AAP recommends otoacoustic emissions for children of any developmental age, and automated brainstem auditory evoked response testing for infants with a developmental age between birth and 9 months (Harlor, 2009). The American Academy of Audiology (AAA) (2011) recommends otoacoustic emissions for preschool- and school-age children for whom pure tone screening is not developmentally appropriate (ability levels less than 3 years).

Either brainstem auditory evoked response or otoacoustic emissions testing is appropriate for making a confirmatory diagnosis of hearing disorders in infants and children (developmental age of birth to 36 months) who did not pass the initial screening test (AAP, 2007). When a permanent hearing deficit is detected, frequency-specific brainstem auditory evoked response testing is appropriate to determine the degree and configuration of hearing deficiency in each ear for fitting of amplification devices. When there are risk indicators for neural hearing disorders, click-evoked brainstem auditory evoked response testing using both condensation and rarefaction single-polarity stimulus are needed to determine if a cochlear microphonic is present (AAP, 2007). Brainstem auditory evoked response is an appropriate test for children suspected of hearing loss with risk factors for hearing loss or who are being evaluated for amplification and are developmentally delayed or too young (under 5 months) for reliable conditioned behavioral testing procedures (NYSDH, 2007).

**Retrocochlear pathology:**

Retrocochlear diseases may involve the vestibulocochlear nerve, brainstem, or central nervous system. Among the most common pathologies affecting the vestibulocochlear nerve are vestibular schwannoma (also called acoustic neuromas, acoustic schwannoma, acoustic neuromas, and vestibular neurilemoma). Vestibular schwannoma is a group of benign Schwann cell-derived tumors that commonly arise from the vestibular portion of the eighth cranial nerve. As a schwannoma grows, it presses against the nerves affecting hearing and balance. There may be no symptoms or mild symptoms at first, progressing to loss of hearing on one side, ringing in the ears, dizziness and balance problems. The tumor can eventually cause
numbness or paralysis of the face and, if large enough, press against the brain, becoming life-threatening (National Library of Medicine [NLM], 2014).

Brainstem auditory evoked response demonstrates high sensitivity and specificity for screening clinically suspected moderate to large vestibular schwannoma, but significantly lower values in patients with a low clinical suspicion for disease, particularly those with tumors less than 1 cm (Koors, 2013; Fortnum, 2009). Brainstem auditory evoked response failed to provide clinically useful results in patients with severe to profound hearing impairment — typically a hearing threshold greater than 70 dBHL at 4 kHz (Fortnum, 2009).

Magnetic resonance imaging is the method of choice for confirming diagnosis of retrocochlear pathology. However, brainstem auditory evoked response may have a role when magnetic resonance imaging is contraindicated or its results are equivocal. Brainstem auditory evoked response and gadolinium-enhanced magnetic resonance imaging are used to discriminate among idiopathic, viral, and other causes of sensorineural hearing loss (ACR, 1996). The American Academy of Otolaryngology supports magnetic resonance imaging, brainstem auditory evoked response, or audiometric follow-up to evaluate adult patients with sudden sensorineural hearing loss for retrocochlear pathology, based on observational studies with a preponderance of benefit over harm, but identifying this pathology may not influence outcomes in all cases (Stachler, 2012). Therefore, brainstem auditory evoked response and follow-up audiometry would be acceptable alternatives for the initial follow-up of sudden sensorineural hearing loss in adults, as long as there is appropriate counseling about the limitations of these modalities (Stachler, 2012).

**Chiari malformation s:**

Chiari malformation s — also called Arnold-Chiari malformations — are structural defects in the cerebellum, which can block the flow of cerebrospinal fluid and cause a range of symptoms, including dizziness, muscle weakness, numbness, vision problems, headaches, and problems with balance and coordination (National Institute of Neurological Disorders and Stroke, 2014). Brainstem auditory evoked response has been proposed in patients with Chiari malformation or myelomeningocele to assess the degree of damage to the brainstem and predict which infants may go on to develop symptoms.

There is insufficient evidence from low-quality case series to support the role of brainstem auditory evoked response for predicting infants who would develop Chiari malformation -related brainstem symptoms within a 2-year to 2.5-year period, for identifying neurologic abnormalities in older patients with documented symptomatic Chiari malformation, or for detecting risk of neurological injury intraoperatively (Hayes, 2010). No studies demonstrated any positive effects of brainstem auditory evoked response testing on patient outcomes (Hayes, 2010). No evidence-based guidelines were identified on this topic.

**Predicting outcome in comatose patients:**

The American Academy of Neurology (AAN) found insufficient evidence to support brainstem auditory and visual-evoked tests and event-related potential tests for prognosis in anoxic-ischemic encephalopathy
Findings from a recent large case series of more than 100 subjects suggest brainstem auditory evoked response may be best suited to patients with massive hemispheric infarction to predict poor outcome (Zhang, 2011). Unfavorable electroencephalography patterns showed the highest sensitivity (96.3 percent, 95 percent confidence interval [CI] 86.2 percent to 99.4 percent), while bilateral absence of somatosensory evoked potentials (N20 component) and wave V showed the highest specificity (100 percent, 95 percent CI 85.9 percent to 100 percent) and positive predictive value (100 percent, 95 percent CI 80.8 percent to 100 percent), but these results require further confirmation.

**Intraoperative neuromonitoring:**

Intraoperative neuromonitoring is performed to minimize neurological damage during surgery and to identify important neural structures in the operative field with the goal of avoiding and/or limiting significant postoperative impairments. The evidence is insufficient to support the clinical role of brainstem auditory evoked response in assessing hearing preservation during excision of vestibular schwannoma. The maintenance of waves I and V corresponds to the peripheral cochlear nerve and the inferior colliculus, respectively. While some evidence from surgical case series suggests preservation of waves I and V correlates with better postoperative hearing preservation rates, others have found poor hearing outcomes despite wave preservation. When actual changes are seen on brainstem auditory evoked response, the severity or presence of postoperative deficits cannot be predicted reliably. While such brainstem auditory evoked response waveform irregularities may alert the surgeon to potential cranial nerve damage, the evidence for affecting surgical decisions and patient outcomes is anecdotal (Oh, 2012).

There is no consensus on the exact alarm criteria of intraoperative brainstem auditory evoked response changes for intraoperative neural damage and subsequent postoperative hearing loss. Significant time delay inherent in signal averaging, the high prevalence of false-positive results, and the dependence on the individual’s baseline results further limit the clinical utility of brainstem auditory evoked response. The auditory preservation rates of combined techniques that incorporate brainstem auditory evoked response do not yet approximate those of facial nerve preservation. Further efforts and investigations are needed to study and incorporate adjunctive intraoperative neuromonitoring techniques such as brainstem auditory evoked response in an attempt to improve preservation of auditory function (Kim, 2013; Oh, 2012).

No evidence-based guidelines were identified that addressed the clinical use of brainstem auditory evoked response for intraoperative neuromonitoring. Brainstem auditory evoked response provides direct evidence of a change in function along the auditory pathway that may warrant the immediate attention of the surgical team (American Society of Neurophysiological Monitoring, 2014).

**Ototoxicity monitoring:**

Common drugs such as aminoglycosides, chemotherapeutic agents, and heavy metals are known for their ototoxic potential. The goal of monitoring for ototoxicity is to identify cochlear dysfunction early in an effort to reduce or prevent further auditory damage. Two overviews noted conventional brainstem auditory evoked response uses clicks to stimulate middle (1 – 4 kHz) rather than higher frequencies where the cochlea is affected by ototoxic drug exposure, and responses to high-frequency tone bursts require
considerable recording time (Jacob, 2006; Mitchell, 2004). Evidence from early publications of case reports and case series suggest conventional and high-frequency brainstem auditory evoked response is feasible for the early detection of drug-induced hearing loss, but studies comparing it to alternative tests are conflicting.

No evidence-based guidelines were identified. There is little consensus on the optimal protocol for monitoring ototoxicity using objective measures, but the potential for brainstem auditory evoked response in this context is an active area of investigation (AAA, 2009; ASHA, 2005). ASHA (2005) supports otoacoustic emissions or brainstem auditory evoked response to monitor for ototoxicity in children with limited attention spans and in patients who are unable to provide reliable behavioral data; brainstem auditory evoked response may be more appropriate than otoacoustic emissions for patients with abnormal middle ear function and baseline hearing loss greater than about 40 dB HL.

**Policy updates:**

A contemporary cohort study found no association between the wave component of brainstem auditory evoked response and cumulative lead values in 130 children with a history of low blood lead levels, suggesting that brainstem auditory evoked response may not be the most sensitive method in this population (Alvarenga, 2015). A narrative review noted that hypoacusis is the most prevalent sensory disability in the world and is amenable to effective hearing screening tests using electroencephalography technologies (Paulraj, 2015). Electroencephalography-based hearing threshold level determination is most suitable for persons who lack verbal communication and behavioral response to sound stimulation, while brainstem auditory evoked response reflects the auditory ability level of an individual. Systematic evaluation of electroencephalography hearing perception level may predict hearing loss in newborns, infants, and children.

To improve identification of patients at high risk of vestibular schwannoma, a new systematic review and meta-analysis assessed the diagnostic accuracy of non-imaging screening protocols for patients presenting with asymmetrical sensorineural hearing loss and/or unilateral audiovestibular dysfunction (Hentschel, 2017). While more than 95 percent of magnetic resonance imagings are negative, non-imaging protocols, including those with brainstem auditory evoked response, were less accurate and would offer no improvement in patient selection.

Results of one case series of 46 patients at a single institution found that Intraoperative neuromonitoring and post-operative auditory brainstem response monitoring in patients who undergo vestibular schwannoma excision suggest ongoing changes of auditory brainstem response quality and hearing function during and after surgery (Hummel, 2016a and 2016b). Both tests may be predictive of postoperative course and hearing outcome, and monitoring immediately after surgery may be able to identify patients at risk of a secondary hearing deterioration. It is unclear whether these results would affect intraoperative or post-operative decision-making, and these findings should be replicated in studies at other institutions before widespread use.

In 2018, we added no new information to the policy. No policy changes are warranted.
### Summary of clinical evidence:

<table>
<thead>
<tr>
<th>Citation</th>
<th>Content, Methods, Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hentschel (2017)</strong></td>
<td><strong>Key points:</strong></td>
</tr>
</tbody>
</table>
| Diagnostic accuracy of screening protocols for detecting vestibular schwannoma | • Systematic review and meta-analysis of 12 crossover studies (4,969 total patients) comparing non-imaging screening to magnetic resonance imaging in patients with asymmetrical hearing loss and/or unilateral audiovestibular dysfunction.  
  • Overall quality: low to moderate, high heterogeneity in all studies except for studies using pure tone audiometry protocols.  
  • None of the currently available non-imaging screening protocols appear to be accurate as magnetic resonance imaging in detecting vestibular schwannoma. |
| **Hummel (2016a and 2016b)** | **Key points:**                    |
| Intraoperative neuromonitoring and postoperative monitoring for vestibular schwannoma excision | • From 2010 to 2012, 46 patients underwent surgery and 4-hour post-operative monitoring. Hearing function was correlated with auditory brainstem response monitoring.  
  • Strong correlation found between different types of auditory brainstem response development and postoperative hearing (P < 0.001); auditory brainstem response quality after 60% tumor removal was independently significant for hearing outcome.  
  • Post-operative course strongly correlated with the intraoperative development (P < 0.001) and with hearing outcome (P = 0.003). |
| **Alvarenga (2015)**      | **Key points:**                    |
| Brainstem auditory evoked potential in children with low level cumulative lead exposure | • A contemporary cross-sectional cohort examined tympanometry, pure tone audiometry and speech audiometry, transient evoked otoacoustic emissions and brainstem auditory evoked potentials, with blood lead monitoring over 35.5 months.  
  • The study included 130 children, with ages ranging from 18 months to 14 years, 5 months (mean age 6 years, 8 months ±3 years, 2 months). The mean time-integrated cumulative blood lead index was 12 g/dL (SD ± 5.7, range: 2.433).  
  • No association was found between the absolute latencies of waves I, III, and V; the interpeak latencies I – III, III – V and I – V; and the cumulative lead values.  
  • No evidence of toxic effects from chronic low lead exposures was observed in the auditory function of children living in a lead-contaminated area. |
| **Koors (2013)**          | **Key points:**                    |
| Diagnosing vestibular schwannoma with brainstem auditory evoked response | • Systematic review and meta-analysis of 43 studies (3,314 total patients).  
  • Overall quality: not systematically assessed, but high risk of bias in most studies.  
  • Sensitivity (Se) and specificity (Sp) of brainstem auditory evoked response for screening clinically suspected vestibular schwannoma:  
    - Pooled Se = 93.4% (95% CI 92.6 to 94.3, P = 0.0000).  
    - For tumors < 1 cm Se = 85.8% (95% CI 80.6 to 90.1, P = 0.0116, eight studies, 176 patients). |
<table>
<thead>
<tr>
<th>Citation</th>
<th>Content, Methods, Recommendations</th>
</tr>
</thead>
</table>
| Hayes (2010) | **Key points:**  
- Systematic review of studies to assess damage to the brainstem or to predict which infants may go on to develop symptoms.  
- Overall quality: low, case series.  
- Brainstem auditory evoked response accuracy = 70% to 84% for identifying infants who would develop Chiari malformation-related brainstem symptoms within a 2-year to 2.5-year period (two studies), but less accurate (39%) for identifying neurologic abnormalities in older patients with documented symptomatic Chiari malformation (several case series).  
- Value of intraoperative brainstem auditory evoked response for guiding surgery or detecting risk of neurological injury is unclear (two studies).  
- Effects of brainstem auditory evoked response testing on patient outcomes and overall clinical role is unclear. |
| Wolff (2010) | **Key points:**  
- Systematic review of 16 studies.  
- Overall quality: generally poor, highly heterogeneous with high risk of bias.  
- Brainstem auditory evoked response results: Se increased with tumor size:  
  - Size ≤ 1.0 cm: Se 79% (95% CI 72% to 85%).  
  - Size 1.0 to 2.0 cm: Se 95% (95% CI 91% to 97%).  
  - Size > 2.0 cm: Se 98% (95% to 99%).  
- Brainstem auditory evoked response fails to provide clinically useful results in patients with severe to profound hearing impairment (typically a hearing threshold greater than 70 dBHL at 4 kHz).  
- Magnetic resonance imaging is still considered the gold standard, but brainstem auditory evoked response may have a role in patients for whom magnetic resonance imaging is contraindicated. |
| Fortnum (2009) | **Key points:**  
- Systematic review of 16 studies.  
- Overall quality: generally poor, highly heterogeneous with high risk of bias.  
- Brainstem auditory evoked response results: Se increased with tumor size:  
  - Size ≤ 1.0 cm: Se 79% (95% CI 72% to 85%).  
  - Size 1.0 to 2.0 cm: Se 95% (95% CI 91% to 97%).  
  - Size > 2.0 cm: Se 98% (95% to 99%).  
- Brainstem auditory evoked response fails to provide clinically useful results in patients with severe to profound hearing impairment (typically a hearing threshold greater than 70 dBHL at 4 kHz).  
- Magnetic resonance imaging is still considered the gold standard, but brainstem auditory evoked response may have a role in patients for whom magnetic resonance imaging is contraindicated. |
<table>
<thead>
<tr>
<th>Citation</th>
<th>Content, Methods, Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>NYSDOH (2007)</td>
<td>Clinical practice guideline: hearing loss, assessment, and intervention for young children (ages 0 – 3 years)</td>
</tr>
</tbody>
</table>
| **Key points:** | - Systematic review of studies evaluating aspects of newborn screening: newborn screening (five studies); screening infants and young children (five studies).  
- Overall quality: variable.  
- Otoacoustic emissions and brainstem auditory evoked response can identify moderate-to-profound sensorineural hearing loss, brainstem auditory evoked response more effective than otoacoustic emissions for moderate hearing loss at 1 kHz; both have a low Se for mild hearing loss (moderate).  
- Recommend universal neonatal hearing screening using a physiologic test prior to discharge (moderate quality), or by one month of age for those with no access to hospital screening or not born in a hospital (consensus).  
- Recommend otoacoustic emissions and brainstem auditory evoked response screening in infants and young children and in children in whom behavioral testing would be unreliable or unattainable (consensus).  
- Recommend repeat testing for failed initial screening, either using same technology as the initial test (consensus).  
- Recommend brainstem auditory evoked response to detect suspected conditions of the auditory nerve or brainstem (consensus).  
- Brainstem auditory evoked response can identify conductive and sensorineural hearing loss and provide some diagnostic information about type of loss through algorithms (consensus). Recommended for:  
  - Detection of sensorineural hearing loss with air-conducted and bone-conducted click brainstem auditory evoked responses (moderate).  
  - For children suspected of hearing loss, or with risk factors for hearing loss, who are developmentally delayed or are too young (under age 5 months) for reliable conditioned behavioral testing procedures (consensus).  
  - When two attempts at behavioral audiometry by a pediatric audiologist are not successful in testing the hearing status of a child within a two-month period (consensus).  
  - To assess provision of amplification if behavioral audiologic assessment cannot be performed accurately because of the infant's age or developmental level (consensus). |

| Wijdicks (2006) for the AAN | Practice parameter: prediction of outcome in comatose survivors after cardiopulmonary resuscitation |
| **Key points:** | - Systematic review identified one study for brainstem auditory evoked response (13 total patients).  
- Overall quality: low, small, retrospective.  
- Middle latency brainstem auditory evoked response was absent in all patients who died or remained in a persistent vegetative state (Se = 34%, 95% CI 19% to 49%, false positive rate = 0%).  
- Insufficient evidence. |
### Citations

<table>
<thead>
<tr>
<th>Citation</th>
<th>Content, Methods, Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hayes (2005)</td>
<td>Neonatal Hearing Screening Key points:</td>
</tr>
<tr>
<td></td>
<td>• Systematic review of 17 studies published from 1998 – 2005 evaluating newborn hearing screening programs (updates a previous review).</td>
</tr>
<tr>
<td></td>
<td>• Overall quality: variable.</td>
</tr>
<tr>
<td></td>
<td>• These programs are feasible and effective.</td>
</tr>
<tr>
<td></td>
<td>• Limited evidence suggests early intervention is associated with positive developmental outcomes.</td>
</tr>
<tr>
<td></td>
<td>• Detection rates were higher for targeted screening compared with universal screening, but approximately half of all hearing-impaired infants had no known risk factors and would have been missed by screening limited to high-risk infants.</td>
</tr>
<tr>
<td></td>
<td>• Sufficient evidence supports universal neonatal hearing screening programs using either brainstem auditory evoked response or otoacoustic emissions when the screening program includes a protocol for rescreening infants who fail the initial screen, referral for age-appropriate diagnostic testing for infants who fail both newborn screens, parent and community education, and a support system to ensure diagnostic testing is performed and effective intervention provided when indicated.</td>
</tr>
<tr>
<td>Kezirian (2001)</td>
<td>Cost and cost-effectiveness of universal screening for hearing loss in newborns Key points:</td>
</tr>
<tr>
<td></td>
<td>• Cost and cost-effectiveness analysis of universal newborn hearing screening programs based on published data.</td>
</tr>
<tr>
<td></td>
<td>• Decision analysis model using the hospital perspective; compared four distinct protocols for screening a fixed and defined hypothetical cohort of newborn infants.</td>
</tr>
<tr>
<td></td>
<td>• The prevalence of hearing loss per 1,000 infants = 3.5 (range: 2.3 – 4.6).</td>
</tr>
<tr>
<td></td>
<td>• Results: otoacoustic emissions testing at birth followed by repeat testing at follow-up demonstrated the lowest cost ($13 per infant) and had the lowest cost-effectiveness ratio ($5,100 per infant with hearing loss identified). Screening brainstem auditory evoked response testing at birth with no screening test at follow-up had greater effectiveness, but also had the highest cost ($25 per infant) and highest cost-effectiveness ratio ($9,500 per infant with hearing loss identified).</td>
</tr>
<tr>
<td></td>
<td>• Results were robust regardless of prevalence of hearing loss or the fraction of infants returning for follow-up testing.</td>
</tr>
</tbody>
</table>

### References

**Professional society guidelines/other:**


Peer-reviewed references:


**CMS National Coverage Determinations (NCDs):**


**Local Coverage Determinations (LCDs):**


**Commonly submitted codes**

Below are the most commonly submitted codes for the service(s)/item(s) subject to this policy. This is not an exhaustive list of codes. Providers are expected to consult the appropriate coding manuals and bill accordingly.

<table>
<thead>
<tr>
<th>CPT Code</th>
<th>Description</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>92585</td>
<td>Auditory evoked potentials for evoked response audiometry and/or testing of the central nervous system; comprehensive.</td>
<td></td>
</tr>
<tr>
<td>92586</td>
<td>Auditory evoked potentials for evoked response audiometry and/or testing of the central nervous system; limited.</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>ICD-10 Code</th>
<th>Description</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>D33.3</td>
<td>Acoustic neuroma</td>
<td></td>
</tr>
<tr>
<td>G35</td>
<td>Multiple sclerosis</td>
<td></td>
</tr>
<tr>
<td>H91.01</td>
<td>Otoxic hearing loss, right ear</td>
<td></td>
</tr>
<tr>
<td>H91.02</td>
<td>Otoxic hearing loss, left ear</td>
<td></td>
</tr>
<tr>
<td>H91.03</td>
<td>Otoxic hearing loss, bilateral</td>
<td></td>
</tr>
<tr>
<td>H91.09</td>
<td>Otoxic hearing loss, specified</td>
<td></td>
</tr>
<tr>
<td>H91.20</td>
<td>Sudden hearing loss, unspecified</td>
<td></td>
</tr>
<tr>
<td>H91.21</td>
<td>Sudden hearing loss, right</td>
<td></td>
</tr>
<tr>
<td>H91.22</td>
<td>Sudden hearing loss, left</td>
<td></td>
</tr>
<tr>
<td>H91.23</td>
<td>Sudden hearing loss, bilateral</td>
<td></td>
</tr>
<tr>
<td>H91.8X1</td>
<td>Other specified hearing loss, unspecified ear (asymmetrical)</td>
<td></td>
</tr>
<tr>
<td>H91.8X2</td>
<td>Other specified hearing loss, right ear</td>
<td></td>
</tr>
<tr>
<td>H91.8X3</td>
<td>Other specified hearing loss, left ear</td>
<td></td>
</tr>
<tr>
<td>H93.11</td>
<td>Tinnitus, right ear</td>
<td></td>
</tr>
<tr>
<td>H93.12</td>
<td>Tinnitus, left ear</td>
<td></td>
</tr>
<tr>
<td>H93.13</td>
<td>Tinnitus, bilateral</td>
<td></td>
</tr>
<tr>
<td>H93.19</td>
<td>Tinnitus, unspecified</td>
<td></td>
</tr>
<tr>
<td>H93.3X1</td>
<td>Disorder of right acoustic nerve</td>
<td></td>
</tr>
<tr>
<td>H93.3X2</td>
<td>Disorder of left acoustic nerve</td>
<td></td>
</tr>
<tr>
<td>H93.3X3</td>
<td>Disorder of bilateral acoustic nerve</td>
<td></td>
</tr>
<tr>
<td>ICD-10 Code</td>
<td>Description</td>
<td>Comment</td>
</tr>
<tr>
<td>------------</td>
<td>--------------------------------------------------</td>
<td>------------------</td>
</tr>
<tr>
<td>H93.3X9</td>
<td>Disorder of unspecified acoustic nerve</td>
<td></td>
</tr>
<tr>
<td>Z01.10</td>
<td>Encounter for exam of ears and hearing without abnormal findings</td>
<td></td>
</tr>
<tr>
<td>Z01.110</td>
<td>Encounter for hearing examination following failed hearing screening</td>
<td></td>
</tr>
<tr>
<td>Z01.118</td>
<td>Encounter for examination of ears and hearing with other abnormal findings</td>
<td></td>
</tr>
<tr>
<td>Z01.818</td>
<td>Preop exam for cochlear implant</td>
<td></td>
</tr>
<tr>
<td>Z13.5</td>
<td>Encounter for screening for ear and eye disorders</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>HCPCS Level II Code</th>
<th>Description</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>N/A</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>