Electrocochleogram and Perilymphatic Pressure Measurement

Number: 0564

**POLICY**

*Please see amendment for Pennsylvania Medicaid at the end of this CPB.*

Aetna considers electrocochleography (ECOG) medically necessary for evaluation of members with symptoms of episodic dizziness (vertigo, imbalance) or tinnitus, to rule out endolymphatic hydrops (Meniere’s disease) and perilymphatic fistula.

Aetna considers ECOG medically necessary when performed with auditory brainstem response (ABR) testing of members with profound hearing loss. *(Note): Profound hearing loss is defined as having a pure tone average (PTA) of 90 dB HL or greater at 500 Hz, 1,000 Hz, 2,000 Hz and 4,000 Hz).*

Aetna considers ECOG experimental and investigational for routine screening of hearing impairment, and for all other indications (e.g., during superior semicircular canal dehiscence repair and canal resurfacing) because of insufficient evidence of its clinical value for these indications.

Aetna considers the use of intra-operative ECOG for evaluation of cochlear trauma during cochlear implantation and prediction of preservation of residual acoustic hearing after cochlear implantation experimental and investigational because of insufficient evidence.
Aetna considers measurement of perilymphatic pressure experimental and investigational because its value in the management of individuals with Meniere’s disease or idiopathic sudden sensorineural hearing loss has not been established.

BACKGROUND

Meniere's Syndrome/Endolymphatic Hydrops

Meniere’s disease or Meniere’s syndrome is a potentially disabling condition involving varying degrees of fluctuating hearing loss, fluctuating tinnitus, episodic vertigo, and aural fullness (a feeling of fullness, pressure and discomfort in the ear). The syndrome may be idiopathic, in which case it is called Meniere’s disease, or secondary to various processes that interfere with the normal resorption of endolymph (e.g., neurosyphilis, viral infections, trauma, congenital anomalies, etc.). The disease appears to strike most commonly persons between 30 and 60 years of age, with men and women affected equally. Incidence of the disease is approximately 250 per million populations. Patients with Meniere’s disease have a progressive distention of the endolymphatic space of the inner ear, caused by fluid build-up of the endolymphatic space (endolymphatic hydrops), caused either by overproduction or reduced adsorption. The increased pressure exposes cochlear hair cells responsible for sensing movement and balance to progressive damage and paralysis, resulting in attacks of dizziness, often with nausea and vomiting.

Early in the course of disease, these attacks are usually brief (lasting 1 hour or so), as the damage to the cochlear hair cells is temporary and the hair cells resume normal function when the hydrops resolves. Chronic repetitive attacks may lead to irreversible damage to the hair cells, and hearing loss can become permanent. The hearing loss and tinnitus are usually unilateral, although up to a quarter of patients may go on to develop a severe bilateral disorder.
Trans-tympanic electrocochleography (ECOG) can be used to confirm cochlear involvement in hearing loss, and is an objective test for endolymphatic hydrops. Electrocochleography measures the ratio of the summating potential (SP) and the action potential (AP) on the most peripheral portion of the auditory system in response to auditory stimuli. The AP is the summed or averaged activity of the APs of the auditory nerve, which are elicited by acoustic stimulation. The SP is generated by the hair cells of the cochlea in response to acoustic stimulation. Surface electrodes, such as those used in auditory brainstem response, can not record these potentials; electrodes must be placed on or through the tympanic membrane. In ECOG, a fine needle is passed through an anesthetized tympanic membrane and placed in contact with the cochlear hair cells of the inner ear in order to record electrical activity from these cells. The ear is exposed to a train of about 1,000 click or tonal stimuli, and APs from auditory neurons are recorded for 10 milliseconds after each click. This information is recorded and summated by computer. Patients with endolymphatic hydrops have abnormal waveforms (widening of the waveform with multiple peaks). Endolymphatic hydrops is suggested when the ratio of the summating potential to the AP is greater than 35 %.

Electrocochleography allows the diagnosis of Meniere's disease to be confirmed or refuted so that appropriate prognostic advice can be given together with medical or surgical treatments if indicated.

In all patients who have unilateral persistent otological symptoms, a MRI is required to exclude acoustic neuroma, which can mimic the presentation of Meniere's disease. Meniere's is confirmed with an electrocochleogram so that appropriate effective treatments can be applied.

Acute attacks of Meniere's syndrome are treated with anti-emetics and sedatives. Long-term treatment is usually medical, including rigid salt restriction and diuretics. Occasionally chemoablation (intra-tympanic gentamycin) or surgical ablation (labyrinthectomy when hearing is already lost, vestibular nerve section when it is not) is necessary for refractory disease.
Perilymphatic Fistula

Electrocochleography has also been used to determine the presence of perilymphatic fistula, based on the SP/AP amplitude ratio. A perilymph fistula (perilymphatic fistula, labyrinthine fistula) is an abnormal communication between the fluid-filled perilymphatic space of the inner ear and the air-filled middle ear cavity, usually through the round or oval windows. This results in sensori-neural hearing loss and/or vestibular symptoms.

Most commonly, a tear in the round or oval window leads to loss of perilymph into the middle ear. This may be the result of stapes prosthesis surgery, trauma, barotrauma, bony erosion due to infection or neoplasm, or it may be idiopathic. In children, it is associated with congenital anomalies of the middle or inner ear.

Symptoms of perilymphatic fistula are similar to Meniere's disease, and include sensori-neural hearing loss, which may be sudden or fluctuating; aural fullness; and vestibular symptoms (vertigo (with or without head position changes), dysequilibrium, motion intolerance, nausea and vomiting, disorganization of memory and concentration, and perceptual disorganization in complex surroundings (such as crowds or traffic)). Tinnitus occurs in some cases, and can be roaring. In the absence of prior surgery or definite traumatic event, it may be difficult to distinguish a perilymph fistula from Meniere's syndrome.

In addition to ECOG, other tests that may be used by otologists for the diagnosis of perilymph fistula include audiograms to detect hearing loss and fistula tests. The subjective fistula test is performed by applying positive and negative pressure to the intact eardrum using a pneumatic otoscope. Positive results include the elicitation of nystagmus or onset of dysequilibrium with the sensation of motion or nausea. Some otologists administer the test with electronystagmography or using a specialized platform. Rigid or flexible endoscopy is performed to look for visible tears or fluid in the middle ear. The final diagnosis is made by direct inspection at the time of surgery, with visualization of perilymph fluid in the middle ear cavity.

Medical therapy is rarely reported. There are some reports of spontaneous healing with bedrest, head elevation to 30 degrees, and avoidance of lifting or middle ear pressure-increasing activities. Surgical treatment is available if conservative therapy fails.
Severe Sensori-Neural Deafness

Another clinical application of ECOG is identification of wave I of the auditory brainstem response (ABR) during combined ECOG-ABR testing, as wave I is frequently difficult to detect in patients with profound hearing loss when ECOG is not performed in conjunction with ABR testing. Auditory brainstem response testing involves the measurement of responses along the auditory pathway from cranial nerve VIII to the lateral lemniscus of the auditory brainstem. Five distinct electric waveforms generated in the 8th nerve, brainstem, and other regions in response to acoustic stimulation are examined. Wave I is generated at the distal part of the auditory nerve.

Screening for Hearing Impairment

According to the U.S. Preventive Services Task Force, ECOG is not an appropriate test for routine screening for hearing impairment.

Electrocochleography is available in virtually all otolaryngology departments, takes only 20 mins or so and requires an otolaryngologist and usually an audiologist.

Perilymphatic Pressure Measurement

Assessment of perilymphatic pressure has also been used to diagnose Meniere's disease. However, published reports do not support a diagnostic role for this approach. Rosingh and colleagues (1996) did not find any significant differences in perilymphatic pressure measurements between patients with Meniere's disease and young normal hearing subjects. This is in accordance with the findings of Ayache and associates (2000) who concluded that assessment of perilymphatic pressure does not seem to be useful in Meniere's disease. Furthermore, Rosingh and co-workers (2000) reported that perilymphatic pressure measured in the affected ear of patients with Meniere's disease or idiopathic sudden sensori-neural hearing loss did not differ significantly from the pressure in the non-affected and normal hearing ear. In a follow-up study by Ayache et al (2002), the authors concluded that perilymphatic
pressure measurements by means of the Tympanic Displacement Analyzer are not useful in the evaluation of patients with Meniere’s disease.

Intra-Operative Electrocochleography for Evaluation of Cochlear Trauma During and After Cochlear Implantation

Calloway and colleagues (2014) noted that electrophysiological responses to acoustic stimuli are present in nearly all recipients of cochlear implantation (CI) when measured at the round window (RW). Intra-cochlear recording sites might provide an even larger signal and improve the sensitivity and the potential clinical utility of ECOG. These researchers compared RW to intra-cochlear recording sites and examined if such recordings can be used to monitor cochlear function during CI. Intra-operative ECOG recordings were obtained in subjects receiving CI from the RW and from just inside scala tympani (n = 26). Stimuli were tones at high levels (80 to 100 dB hearing level [HL]). Further recordings were obtained during insertions of a temporary lateral cochlear wall electrode (n = 8). Response magnitudes were determined as the sum of the 1st and 2nd harmonics amplitudes. All subjects had measurable extra-cochlear responses at the RW; 20 cases (78%) showed a larger intra-cochlear response, compared with 3 (11%) that had a smaller response and 3 that were unchanged. On average, signal amplitudes increased with increasing electrode insertion depths, with the largest increase between 15 and 20 mm from the RW. The authors concluded that ECOG to acoustic stimuli via an intra-cochlear electrode is feasible in standard recipients of CI. The increased signal can improve the speed and efficiency of data collection. The growth of response magnitudes with deeper intra-scalar electrode positions could be explained by closer proximity or favorable geometry with respect to residual apical signal generators. These investigators stated that reductions in magnitude may represent unfavorable geometry or cochlear trauma.

Campbell and co-workers (2015) recorded ECOG directly from a cochlear implant in awake recipients with residual hearing, using an adaptation of Neural Response Telemetry (NRT) that achieves a 10-ms recording window. Subjects were adults with CI422 cochlear implants who retained audiometric thresholds between 75 and 90 dB HL at 500 Hz in their
implanted ear. The cochlear implant was interfaced to a laptop via a Freedom speech processor connected by USB. Calibrated acoustic stimuli (clicks and tone bursts between 500 and 1,500 Hz) were presented via insert tube phones to the implanted ear. Responses were acquired through the adapted NRT system. Recordings were made from apical, mid-array, and basal electrodes; ECOG responses were compared with audiometric thresholds. Electrocochleography could be recorded from all 5 subjects. The compound action potential, cochlear microphonic, and summating potentials were identified. Good quality recordings were most reliably attained from apical electrodes using 40 to 100 repetitions. Audiometric thresholds were similar to compound action potential thresholds. The authors concluded that intra-cochlear responses to acoustic stimulation can be recorded directly from the CI in awake recipients with residual hearing; they stated that this may prove useful for monitoring post-operative hearing and for device fitting.

In a prospective, cohort study, Adunka and colleagues (2016) stated that previous reports have documented the feasibility of utilizing ECOG responses to acoustic signals to assess trauma caused during CI. The hypothesis is that intra-operative RW ECOG before and after electrode insertion will help predict post-operative hearing preservation outcomes in recipients of CI. Intra-operative RW ECOG responses were collected from 31 CI recipients (14 children and 17 adults) immediately before and just after electrode insertion. Hearing preservation was determined by post-operative changes in behavioral thresholds. On average, the post-insertion response was smaller than the pre-insertion response by an average of 4 dB across frequencies. However, in some cases (12 of 31) the response increased after insertion. The subsequent hearing loss was greater than the acute loss in the ECOG, averaging 22 dB across the same frequency range (250 to 1,000 Hz). There was no correlation between the change in the ECOG response and the corresponding change in audiometric threshold. The authors concluded that intra-operative ECOG is a sensitive method for detecting electrophysiological changes during CI; but had limited prognostic value regarding hearing preservation in the current conventional CI patient population where hearing preservation was not intended.
Dalbert and associates (2016) evaluated cochlear trauma during CI by ECOG and cone beam computed tomography (CBCT) and correlated intra-operative cochlear trauma with post-operative loss of residual hearing. Electrocochleography recordings to tone bursts at 250, 500, 750, and 1,000 Hz and click stimuli were recorded before and after insertion of the cochlear implant electrode array, using an extra-cochlear recording electrode. Cone beam CTs were conducted within 6 weeks after surgery. Changes of intra-operative ECOG recordings and CBCT findings were correlated with post-operative threshold shifts in pure-tone audiograms (PTAs). A total of 14 subjects were included. In 3 subjects a decrease of low-frequency ECOG responses at 250, 500, 750, and 1,000 Hz occurred after insertion of the electrode array. This was associated with no or minimal residual hearing 4 weeks after surgery. Electrocochleography responses to click stimuli were present in 6 subjects and showed a decrease after insertion of the electrode array in 3. This was associated with a mean hearing loss of 21 dB in post-operative PTAs. Scalar dislocation of the electrode array was assumed in 1 subject because of CBCT findings and correlated with a decrease of low-frequency ECOG responses and a complete loss of residual hearing. The authors concluded that hearing loss of less than or equal to 11 dB was not associated with detectable decrease in ECOG recordings during CI. However, in a majority of patients with threshold shifts of greater than 11 dB or complete hearing loss, an intra-operative decrease of high- or low-frequency ECOG signals occurred, suggesting acute cochlear trauma.

Harris and colleagues (2017) noted that intra-operative, intra-cochlear electrocochleography (ECOG or ECochG) could provide a means to monitor cochlear hair cell and neural response during CI electrode insertion. Distinct patterns in the insertion track can be characterized. Intra-cochlear ECochG was performed in 17 patients. During electrode insertion, a 50-ms tone burst acoustic stimulus was delivered with a frequency of 500 Hz at 110 dB SPL. The ECochG response was monitored from the apical-most electrode. The amplitude of the 1st harmonic was plotted and monitored in near real time by the audiologist-surgeon team during CI electrode insertion. Three distinct patterns in 1st harmonic amplitude change were observed across subjects during insertion: Type A (52 %), overall increase in amplitude from the beginning of insertion until completion; Type B (11 %), a maximum amplitude at the beginning of insertion, with a decrease in amplitude as insertion
progressed to completion; and Type C (35%), comparable amplitudes at the beginning and completion of the insertion with the maximum amplitude mid-insertion. The authors concluded that 3 ECochG patterns were observed during electrode advancement into the cochlea. Moreover, they stated that ongoing and future work will broaden the scope of knowledge regarding the relationship among these patterns, the presence of cochlear trauma, and functional outcomes related to hearing preservation.

Riggs and associates (2017) stated that auditory neuropathy spectrum disorder (ANSD) is characterized by an apparent discrepancy between measures of cochlear and neural function based on auditory brainstem response (ABR) testing. Clinical indicators of ANSD are a present cochlear microphonic (CM) with small or absent wave V. Many identified ANSD patients have speech impairment severe enough that CI is indicated. To better understand the cochleae identified with ANSD that lead to a CI, these researchers performed intra-operative round window ECochG to tone bursts in children (n = 167) and adults (n = 163). Magnitudes of the responses to tones of different frequencies were summed to measure the "total response" (ECochG-TR), a metric often dominated by hair cell activity, and auditory nerve activity was estimated visually from the compound action potential (CAP) and auditory nerve neurophonic (ANN) as a ranked "Nerve Score". Subjects identified as ANSD (45 ears in children, 3 in adults) had higher values of ECochG-TR than adult and pediatric subjects also receiving CIs not identified as ANSD. However, nerve scores of the ANSD group were similar to the other cohorts, although dominated by the ANN to low frequencies more than in the non-ANSD groups. To high frequencies, the common morphology of ANSD cases was a large CM and summating potential, and small or absent CAP. Common morphologies in other groups were either only a CM, or a combination of CM and CAP. These results indicated that responses to high frequencies, derived primarily from hair cells, were the main source of the CM used to evaluate ANSD in the clinical setting. However, the clinical tests did not capture the wide range of neural activity seen to low frequency sounds. The authors concluded that the difference between ANSD and non-ANSD subjects lied primarily in the high frequency regions of the cochlea. These regions produced a larger CM and SP, and were less likely to produce a CAP, compared to non-ANSD subjects. These features were consistent with a large hair cell
response combined with a limited neural response expected for ANSD. In contrast, for responses to low frequencies the neural components, primarily in the form of the ANN, were similar between ANSD and non-ANSD subjects, and varied from no evidence of neural contributions to clear evidence of CAP and/or ANN. Thus, responses from low frequency parts of the cochlea produced a similarly wide distribution of evidence for neural activity between ANSD and non-ANSD subjects. These researchers stated that it remains to be determined if the levels of neural activity observed using acoustic stimuli by ECochG are important in speech perception outcomes with the CIs.

O’Connell and co-workers (2017) determined the relationship between ECochG (measured from the CI electrode array during and after CI), and post-operative audiometric thresholds. These investigators also determined the relationship between ECochG amplitude and electrode scalar location determined by CT; and examined if changes in CM amplitude during electrode insertion were associated with post-operative hearing. A total of 18 subjects undergoing CI with an Advanced Bionics Mid-Scala device were prospectively studied. ECochG responses were recorded using the implant coupled to a custom signal recording unit. ECochG amplitude collected intra-operatively concurrent with CI insertion and at activation was compared with audiometric thresholds post-operatively; 16 patients also underwent post-operative CT to determine scalar location and the relationship to ECochG measures and residual hearing. Mean low-frequency pure tone average (LFPTA) increased following surgery by an average of 28 dB (range of 8 to 50). Threshold elevation was significantly greater for electrodes with scalar dislocation. No correlation was found between intra-operative ECochG and post-operative behavioral thresholds collapsed across frequency; however, mean differences (MDs) in thresholds measured by intra-operative ECochG and post-operative audiometry were significantly smaller for electrodes inserted completely within scala tympani (ST) versus those translocating from ST to scala vestibuli. A significant correlation was observed between post-operative ECochG thresholds and behavioral thresholds obtained at activation. The authors concluded that post-operative audiometry currently serves as a marker for intra-cochlear trauma though thresholds were not obtained until device activation or later. When measured at the same time-point post-operatively, low-frequency ECochG thresholds correlated with behavioral thresholds.
Intra-operative ECochG thresholds, however, did not correlate significantly with post-operative behavioral thresholds suggesting that changes in cochlear physiology occurred between electrode insertion and activation. These investigators stated that ECochG may hold clinical utility providing surgeons with feedback regarding insertion trauma due to scalar translocation, which may be predictive of post-operative hearing preservation. They stated that CI insertion trauma is generally not evident until post-operative audiometry when loss of residual hearing is confirmed; ECochG has the potential to provide estimates of trauma during insertion as well as reliable information regarding degree of hearing preservation.

Gibson (2017) noted that recently ECOG has been used to monitor cochlear implant insertion and to record residual hearing using an electrode on the cochlear implant array as the non-inverting (active) electrode; it has the potential to indicate adverse changes during surgery.

Dalbert and associates (2018) examined the correlation between electrophysiological changes during CI and post-operative hearing loss, and detected the time-points that electrophysiological changes occur during CI. Extra- and intra-cochlear ECoG were used to detect electrophysiological changes during CI. Extra-cochlear ECoG recordings were conducted through a needle electrode placed on the promontory; for intra-cochlear ECoG recordings, the most apical contact of the cochlear implant electrode itself was used as the recording electrode. Tone bursts at 250-, 500-, 750-, and 1,000-Hz were used as low-frequency acoustic stimuli and clicks as high-frequency acoustic stimuli. Changes of extra-cochlear ECoG recordings following full insertion of the cochlear implant electrode were correlated with pure-tone audiometric findings 4 weeks after surgery. Changes in extra-cochlear ECoG recordings correlated with post-operative hearing change ($r = -0.44$, $p = 0.055$, $n = 20$). Mean hearing loss in subjects without decrease or loss of extra-cochlear ECoG signals was 12 dB, compared to a mean hearing loss of 22 dB in subjects with a detectable decrease or a loss of ECoG signals ($p = 0.0058$, $n = 51$). In extra-cochlear ECoG recordings, a mean increase of the ECoG signal of 4.4 dB occurred after opening the cochlea. If a decrease of ECoG signals occurred during insertion of the cochlear implant electrode, the decrease was detectable during the 2nd half of the insertion. The authors concluded that ECoG recordings allowed detection of
Electrophysiological changes in the cochlea during CI. Decrease of extracocharl ECoG recordings during surgery had a significant correlation with hearing loss 4 weeks following surgery. Trauma to cochlear structures appeared to occur during the final phase of the cochlear implant electrode insertion. Baseline recordings for extra-cochlear ECoG recordings should be conducted after opening the cochlea; ECoG responses can be recorded from an intra-cochlear site using the cochlear implant electrode as recording electrode. This technique may prove useful for monitoring cochlear trauma intra-operatively in the future; however, further studies are needed to elucidate the implications of intra-operative findings.

Kim and co-workers (2018) noted that shorter electrode arrays and soft surgical techniques allow for preservation of acoustic hearing in many cochlear implant users. Recently, these researchers developed a method of using the Neural Response Telemetry (NRT) system built in Custom Sound EP clinical software to record acoustically evoked ECoG responses from an intra-cochlear electrode in Nucleus Hybrid CI users. They recorded responses dominated by the hair cells (cochlear microphonic, CM/DIF) and the auditory nerve (auditory nerve neurophonic, ANN/SUM). Unfortunately, the recording procedure was time consuming, limiting potential clinical applications. This report described a modified method to record the ECoG response more efficiently. These investigators referred to this modified technique as the "short window" method, while their previous technique was referred as the "long window" method. In this study, these researchers examined the feasibility of the short window method to record the CM/DIF and ANN/SUM responses, characterized the reliability and sensitivity of the measures recorded using the short window method, and evaluated the relationship between the CM/DIF and ANN/SUM measures recorded using the modified method and audiometric thresholds. A total of 34 post-lingually deafened adult Hybrid CI users participated in this study.

Acoustic tone bursts were presented at four frequencies (250-, 500-, 750-, and 1,000-Hz) at various stimulation levels via an insert earphone in both condensation and rarefaction polarities. Acoustically evoked ECoG responses were recorded from the most apical electrode in the intra-cochlear array. These 2 responses were subtracted to emphasize the CM/DIF responses and added to emphasize the ANN/SUM responses. Response thresholds were determined based on visual inspection of time
waveforms, and trough-to-peak analysis technique was used to quantify response amplitudes. Within-subject comparison of responses measured using both short and long window methods were obtained from 7 subjects. These investigators also assessed the reliability and sensitivity of the short window method by comparing repeated measures from 19 subjects at different times. Correlations between CM/DIF and ANN/SUM measures using the short window recording method and audiometric thresholds were also assessed. Regardless of the recording method, CM/DIF responses were larger than ANN/SUM responses. Responses obtained using the short window method were positively correlated to those obtained using the conventional long window method. Subjects who had stable acoustic hearing at 2 different time-points had similar ECoG responses at those points, confirming high test-retest reliability of the short window method. Subjects who lost hearing between 2 different time points showed increases in ECoG thresholds, suggesting that physiologic ECoG responses were sensitive to audiometric changes. Correlations between CM/DIF and ANN/SUM thresholds and audiometric thresholds at all tested frequencies were significant. The authors concluded that this study compared 2 different recording methods. Intra-cochlear ECoG measures recorded using the short window technique were efficient, reliable, and repeatable. They were able to collect more frequency specific data with the short window method, and observed similar results between the long window and short window methods. Correlations between physiological thresholds and audiometric thresholds were similar to those reported previously using the long window method. This was an important finding because it demonstrated that clinically-available software could be used to measure frequency-specific ECoG responses with enhanced efficiency, increasing the odds that this technique might move from the laboratory into clinical practice.

Tejani and associates (2019) stated that interest in ECoG has recently resurfaced as a potential tool to assess peripheral auditory function in cochlear implant users. ECoG recordings can be evoked using acoustic stimulation and recorded from an extra- or intra-cochlear electrode in cochlear implant users. Recordings reflect contributions from cochlear hair cells and the auditory nerve. These researchers recently demonstrated the feasibility of using Custom Sound EP (clinically available software) to record ECoG responses in Nucleus Hybrid CI users with preserved acoustic hearing in the implanted ear. While successful,
the recording procedures were time intensive, limiting clinical applications. The current report described how they improved data collection efficiency by writing custom software using Python programming language. The software interfaced with Nucleus Implant Communicator (NIC) routines to record responses from an intra-cochlear electrode. ECoG responses were recorded in 8 CI users with preserved acoustic hearing using Custom Sound EP and the Python-based software. Responses were similar across both recording systems, but the recording time decreased significantly using the Python-based software; 7 additional cochlear implant users underwent repeated testing using the Python-based software and showed high test-retest reliability. The authors concluded that the improved efficiency and high reliability increased the likelihood of translating intra-cochlear ECoG to clinical practice.

Haumann and colleagues (2019) noted that to preserve residual hearing during CI surgery, it is desirable to use intra-operative monitoring of inner ear function (cochlear monitoring), especially during electrode insertion; a promising method is ECochG. In this study, the relations between ongoing responses (ORs), recorded extra- and intra-cochlearly (EC and IC), and preservation of residual hearing were examined. Before, during, and after insertion of hearing preservation electrodes, intra-operative ECochG recordings were performed EC using a cotton wick electrode and after insertion also IC using the CI electrode (MED-EL) and a research software tool. The stimulation was delivered acoustically using low-frequency tone bursts. The recordings were conducted in 10 adult CI recipients. The amplitudes of IC ORs were detected to be larger than EC ORs. Intra-operative EC thresholds correlated highly to pre-operative audiometric thresholds at 1,000-Hz, IC thresholds highly at 250-Hz and 500-Hz. The correlations of both intra-operative ECochG recordings to post-operative pure tone thresholds were low. When measured post-operatively at the same appointments, IC OR thresholds correlated highly to audiometric pure tone thresholds. For all patients, it was possible to record ORs during or directly after electrode insertion. The authors concluded that they did not observe any cases with severe IC trauma; and delayed hearing loss could not be predicted with this method. Nevertheless, these researchers stated that intra-operative ECochG recordings are a promising tool to gain further insight into mechanisms impacting residual hearing; post-operatively recorded IC OR thresholds
appeared to be a reliable tool for frequency specific hearing threshold estimation.

**Intraoperative Electrocochleography for Prediction of Preservation of Residual Acoustic Hearing after Cochlear Implantation**

In a prospective, randomized study, Ramos-Macias and co-workers (2019) reported residual hearing preservation outcomes in patients with low-frequency hearing, after CI electrode insertion with 2 types of electrode arrays: one straight and other perimodiolar, when using intraoperative intra-cochlear ECochG during CI electrode insertion. A total of 15 patients ranging from 33 to 54 years of age (mean of 51.19 years). They had been diagnosed with a bilateral, profound sensorineural hearing loss (SNHL) and treated with a unilateral CI: 8 of them with the CI532 and 7 of them with the CI522 (Cochlear Ltd, Sydney, Australia). Pure-tone audiometry was conducted pre-operatively and at 1 and 6 months post-operatively. Inter-operatively, intra-cochlear ECochG was carried out using the apical-most electrode. The amplitude of the 1st harmonic was plotted and monitored in real time by the audiologist-surgeon team during their CI electrode insertion. The different ECochG patterns of the insertion track were recorded and analyzed. In 12 cases, ECochG responses were successfully recorded. In 3 cases, no ECochG responses could be recorded with no residual hearing observed post-operatively in 2 of them. With respect to the 1st harmonic amplitude changes, these investigators found: 4 cases with an overall increase in amplitude measured from the beginning of insertion until completion, all of them showed residual hearing (less than 15 dB HL) at 6 months post-operation; 3 cases with an increasing amplitude at the beginning of insertion, with a decrease in amplitude as insertion progressed to completion, in 2 cases dropping of residual hearing (15 to 30 dB HL) were observed after 6 months post-operation and, in 1 case, complete residual hearing was observed at 6 months post-operatively; 5 cases presented amplitudes at the start of insertion with modifications of amplitude during the insertion dynamic, with increasing and descending in amplitude range during the whole insertion, 2 of them showed residual hearing at 6 months post-operation and 3 cases a drop of residual hearing (15 to 30 dB HL) was observed after 6 months post-operation. No statistical
differences between CI532 and CI522 electrodes were found. Data of the
ECochG responses were also presented (p ≥ 0.05). The authors
concluded that ECochG was a useful tool to examine the residual hearing
in CI recipients with straight and perimodiolar cochlear implant.
Moreover, these researchers stated that further studies are needed to
fully understand the relationship between ECochG and the presence of
residual hearing, cochlear trauma, and functional outcomes.

Kim (2020) examined the use of ECochG as a tool for evaluating the
response of the peripheral auditory system and monitoring hearing
preservation in the growing population of CI recipients with preserved
hearing in the implanted ear. These investigators carried out a search in
PubMed and CINAHL databases up to August 2020 to locate articles
related to the ECochG measured during or after the CI surgery for
monitoring purposes. Non-English articles, animal studies, literature
reviews and editorials, case reports, and conference papers were
excluded. The quality of studies was evaluated using the National
Institute of Health (NIH) "Study Quality Assessment Tool for Case Series
Studies". A total 30 articles were included for the systematic review; 21
were intraoperative ECochG studies, while 7 were post-operative studies;
2 studies were conducted ECochG both during and after the surgery.
Intraoperative ECochG studies focused on monitoring changes in
ECochG response amplitudes during and/or after electrode insertion and
predicting the scalar location of the electrode array. Post-operative
ECochG studies focused on using the ECochG measurements to
estimate behavioral audiometric thresholds and monitor
pathophysiological changes related to delayed onset hearing loss post-
implant. The authors concluded that ECochG was feasible to provide
real-time feedback intraoperatively and has a potential clinical value to
monitor the status of hearing preservation post-operatively in CI recipients
with residual acoustic hearing. Moreover, these researchers stated that
further studies are needed to make this technique more clinically
accessible and understand how ECochG responses can be attributed to
considerable variabilities in CI outcomes.

Dalbert and associates (2020) examined the correlation between
intraoperative changes of ECochG responses and traumatic CI insertions
as well as post-operative hearing loss. ECochG, radiological, and
audiological data were collected prospectively in a CI recipient with
otosclerosis and assumed cochlear trauma during electrode insertion. These researchers also carried out a systematic review within PubMed-NCBI, Embase, and the Cochrane Library using the terms "Cochlear implant" and "Electrocochleography". Original studies that examined intraoperative ECochG responses and post-operative hearing loss were selected and analyzed. The case report revealed a drop of intra- and extra-cochlear ECochG signals during electrode insertion. The postoperative CT scan suggested a scalar dislocation. There was no measurable hearing 4 weeks after surgery. Within the database search, 9 articles met the inclusion criteria. All were case-series studies (ranging from 2 to 36 subjects) with a total of 173 subjects. Due to the heterogeneous data, a meta-analysis was unfeasible. The authors concluded that in concordance with some findings in the literature, the presented case report suggested that a drop of intra- and extra-cochlear ECochG signals during the insertion of the electrode array was associated with cochlear trauma and post-operative hearing loss in some cases; however, the literature is inconclusive regarding the correlation between intraoperative changes of the ECochG signals and postoperative hearing preservation. These investigators stated that more studies examining the correlation are needed to provide sufficient data.

Yin and colleagues (2021) examined the use of intraoperative ECochG as a predictive tool for preservation of residual acoustic hearing after CI. These researchers carried out a systematic review employing a multi-database search strategy (Ovid Medline, Embase, EBM Cochrane, and Scopus) from inception to August 1, 2019; only English language studies in humans were included. All articles were independently reviewed by 2 authors according to Preferred Reporting Items of Systematic Reviews and Meta-analysis (PRISMA) guidelines. Studies without intraoperative ECochG obtained during CI were excluded. Extracted variables included number of patients, ECochG recording technique, success rate of obtaining ECochG potentials, intraoperative changes in ECochG signal, and post-operative hearing preservation outcomes. Among 537 eligible articles, 22 met inclusion criteria encompassing 498 unique patients; 10 studies featured extra-cochlear measurements, 8 featured intra-cochlear measurements, and 4 featured both. Extra-cochlear ECochG had an average (SD) recording success rate of 94.9 % (12.7% ) while intra-cochlear ECochG had an average (SD) recording success rate of 91.8 % (9.8 %). A total of 145 unique patients from 6 studies had complete
intraoperative ECochG data with post-operative behavioral audiometry. After accounting for study-specific definitions of ECochG signal disturbance, worsening changes in intraoperative ECochG signal predicted post-operative hearing loss with limited sensitivity and specificity and notable heterogeneity across studies. The authors concluded that intraoperative ECochG recordings can be obtained in over 90% of patients; however, accuracy in predicting post-operative hearing loss remains limited. These researchers stated that standardization of intraoperative ECochG monitoring technique and data interpretation are needed to more robustly examine outcomes and refine technique.

Electrocochleography During Superior Semicircular Canal Dehiscence Repair and Canal Resurfacing

In a case-series study, Adams et al (2011) determined the electrocochleographic characteristics of ears with superior semicircular canal dehiscence (SSCD) and examined its use for intra-operative monitoring in canal occlusion procedures. A total of 33 patients (45 ears) had clinical and computed tomographic (CT) evidence of SSCD; 8 patients underwent intra-operative electrocochleography (ECoG) during superior canal occlusion; 9 patients underwent post-operative ECoG after SSCD occlusion. Interventions entailed diagnostic, intra-operative, and post-operative extra-tympanic ECoG; middle fossa or trans-mastoid occlusion of the superior semicircular canal. Main outcome measures included summating potential (SP) to action potential (AP) ratio, as measured by ECoG, and alterations in SP/AP during canal exposure and occlusion. Using CT as the standard, elevation of SP/AP on ECoG demonstrated 89% sensitivity and 70% specificity for SSCD. The mean SP/AP ratio among ears with SSCD was significantly higher than that among unaffected ears (0.62 versus 0.29, p < 0.0001). During occlusion procedures, SP/AP increased on exposure of the canal lumen (mean change ± standard deviation [SD], 0.48 ± 0.30). After occlusion, SP/AP dropped below the intra-operative baseline in most cases (mean change, -0.23 ± 0.52). All patients experienced symptomatic improvement. All patients who underwent post-operative ECoG 1 to 3 months after SSCD repair maintained SP/AP of 0.4 or lesser. The authors concluded that these findings expanded the differential diagnosis of abnormal ECoG.
conjunction with clinical findings, ECoG may support a clinical diagnosis of SSCD; and intra-operative ECoG facilitated dehiscence documentation and allowed the surgeon to confirm satisfactory canal occlusion.

Park et al (2015) examined the ECoG findings of patients with superior canal dehiscence (SCD) syndrome and determined their diagnostic values and relationships with audiometric parameters. A total of 13 symptomatic SCD patients (1 bilateral) confirmed by temporal bone CT (TBCT) and cervical vestibular evoked myogenic potentials (cVEMP) were recruited; SCD sizes were measured on reformatted images in the plane of the superior canal (SC). Results of audiologic tests (audiometry, cVEMP, ECoG) for 14 affected and 12 contralateral unaffected ears were evaluated. Relationships between SP to AP ratios, as measured by ECoG, and other audiometric parameters were evaluated. Sensitivity analysis of SP/AP ratios was performed by plotting receiver operating characteristic (ROC) curves for SCD syndrome patients and 19 age-matched healthy controls. Mean SP/AP ratio of SCD ears was significantly higher than that of unaffected ears (0.52 versus 0.25, p < 0.001) and SPs were significantly elevated in affected ears (p = 0.011), whereas APs were similar for affected and unaffected ears. SP/AP ratio showed a sensitivity of 92.3% and a specificity of 94.0% for distinguishing SCD syndrome patients given the inclusion criteria applied (symptoms, TBCT, cVEMP threshold) at a cut-off value of 0.34 (p < 0.001); SP/AP ratio was not correlated with SCD size or cVEMP threshold in affected ears. Negative absolute values of bone conduction at low frequency tended to increase with SP/AP ratio; 5 out of 13 patients underwent surgical repair experienced symptomatic improvement with normalization of SP/AP ratios. The authors concluded that ECoG appeared to be a valuable diagnostic adjunct for functional demonstration of the 3rd window in the otic capsule with high sensitivity and specificity, and thus, could support a clinical diagnosis of SCD when used in conjunction with clinical and radiological findings. This study did not address the intra-operative use of ECoG.

Wenzel, et al (2015) noted that recent findings in patients with superior semicircular canal dehiscence (SCD) have shown an elevated ratio of summating potential (SP) to action potential (AP), as measured by electrocochleography (ECochG). Changes in this ratio can be seen during surgical intervention. The investigators sought to evaluate the utility of
intraoperative ECochG and auditory brainstem response (ABR) as predictive tools for postoperative hearing outcomes after surgical plugging via middle cranial fossa approach for SCD syndrome (SCDS). The investigators reported on a review of 34 cases (33 patients) in which reproducible intraoperative ECochG recordings were obtained during surgery. Diagnosis of SCDS was based on history, physical examination, vestibular function testing, and computed tomography imaging.

Simultaneous intraoperative ECochG and ABR were performed. Pure-tone audiometry was performed preoperatively and at least 1 month postoperatively, and air-bone gap (ABG) was calculated. Changes in SP/AP ratio, SP amplitude, and ABR wave I latency were compared with changes in pure-tone average and ABG before and after surgery. Median SP/AP ratio of affected ears was 0.62 (interquartile range [IQR], 0.45-0.74) and decreased immediately after surgical plugging of the affected canal to 0.42 (IQR, 0.29-0.52; p < 0.01). Contralateral SP/AP ratio before plugging was 0.33 (IQR, 0.25-0.42) and remained unchanged at the conclusion of surgery (0.30; IQR, 0.25-0.35; p = 0.32). Intraoperative changes in ABR wave I latency and SP amplitude did not predict changes in pure-tone average or ABG after surgery (p > 0.05). The investigators concluded that this study confirmed the presence of an elevated SP/AP ratio in ears with SCDS. The SP/AP ratio commonly decreases during plugging. However, an intraoperative decrease in SP/AP does not appear to be sensitive to either the beneficial decrease in ABGs or the mild high-frequency sensory loss that can occur in patients undergoing surgical plugging of the superior semicircular canal. The investigators stated that future work will determine the value of intraoperative ECochG in predicting changes in vestibular function.

Bi and colleagues (2017) noted that SSCD syndrome is an increasingly recognized cause of vestibular and/or auditory symptoms in both adults and children. These symptoms are believed to result from the presence of a pathological mobile "3rd window" into the labyrinth due to deficiency in the osseous shell, leading to inadvertent hydro-acoustic transmissions through the cochlea and labyrinth. The most common bony defect of the superior canal is found over the arcuate eminence, with rare cases involving the postero-medial limb of the superior canal associated with the superior petrosal sinus. Operative intervention is indicated for intractable or debilitating symptoms that persist despite conservative management and vestibular sedation. Surgical repair can be accomplished by
reconstruction or plugging of the bony defect or reinforcement of the round window through a variety of operative approaches. The authors reviewed the etiology, pathophysiology, presentation, diagnosis, surgical options, and outcomes in the treatment of this entity, with a focus on potential pitfalls that may be encountered during clinical management. These investigators stated that ECoG offers an alternative peri-operative testing modality to assess SSCD pathological features and repair. ECoG measurements reveal a significantly higher cochlear SP/AP ratio in ears affected by SSCD, with improvement observed following operative repair. The ECoG measurement is not specific to canal dehiscence because abnormal values are also observed in other inner-ear conditions such as Meniere's disease and perilymphatic fistula. Isolated observation of intra-operative ECoG has also noted normalization of the SP/AP ratio immediately following canal occlusion.

Ward et al (2017) superior canal dehiscence syndrome (SCDS) is a condition in which an abnormal communication between the superior semicircular canal and the middle cranial fossa causes patients to hear internal noises transmitted loudly to their affected ear as well as to experience vertigo with pressure changes or loud sounds. Patients with SCDS can have an elevated ratio of SP to AP as measured by ECoG. Changes in this ratio have been observed during surgical intervention to correct this abnormal communication. These researchers presented a case of SCDS along with history, physical examination, vestibular function testing, and CT imaging. Due to the disabling symptoms, the patient elected to undergo surgery for plugging of the superior semicircular canal by middle cranial fossa approach. Simultaneous intra-operative ECoG and auditory brainstem response (ABR) were performed. Changes in SP/AP ratio, SP amplitude, and ABR wave I latency were observed during surgery, with a large ECoG SP amplitude generating a new wave, identifiable on the ABR and preceding the traditional wave I. The patient's symptoms resolved after surgery, and no long-term detriment to hearing was observed. The authors concluded that this case demonstrated the intra-operative changes in ECoG during surgery for repair of a SCDS. The substantial intra-operative changes in the SP could create a novel wave on intra-operative ABR. These investigators stated that while the clinical consequences of a large spike in the SP is unknown, this finding likely reflects a change in inner ear physiology and may lead to new understandings of origin of the ECoG SP.
Moreover, the authors stated that ECoG has been historically a test used for the assessment of endolymphatic hydrops, associated with Meniere’s disease. Only recently has ECoG been studied in cases of SCDS. The SP/AP ratio is commonly elevated, and often decreases after surgical plugging of the superior semicircular canal. Other physiologic abnormalities associated with SCDS are corrected after surgical plugging, including the elevated low-frequency air-bone gaps identified on pure tone audiometry, decreased cervical VEMP thresholds, and elevated ocular VEMP amplitudes. Each of these findings is attributed to abnormal inner ear physiology reflecting the presence of a 3rd mobile window on the inner ear, in addition to the oval and round windows. The plugging of the extra or "3rd" mobile window, restores normal inner ear physiology, and is shown in the normalization of these tests. Elevations in the SP/AP ratio seen here and in other cases of SCDS likely also reflect abnormal physiology caused by the 3rd mobile window. How the presence of a dehiscence leads to elevations in SP is unclear. One potential explanation is that the opening into the middle cranial fossa creates a pressure differential between endolymphatic and peri-lymphatic compartments of the inner ear, such that a hydrops ex vacuo due to lower pressure peri-lymphatic compartment leads to biasing of the basilar membrane and increased SP. An alternative theory is that sound-sensitive hair cells in the vestibular system contribute to the summating potential, similar to the increased vestibular sensitivity to sound observed in the abnormal cervical and ocular VEMP responses in patients with SCDS. Finally, a recent theory has proposed that patients with SCDS may have endolymphatic hydrops, although the pathophysiology of this is unexplained. It is curious that the SP/AP ratio was noted to rise during surgery, prior to manipulating the labyrinth, as shown in the ECoG stack traces in this study. During exposure of the superior semicircular canal by middle cranial fossa approach, the dura is retracted away from the labyrinth, with more retraction required as the superior semicircular canal is approached. This finding could reflect elevations of intra-cranial pressure (ICP) transmitted to the labyrinth, as has been proposed by Buki et al in measuring oto-acoustic emissions, another measure that like ECoG reflects hair cell function. The use of ECoG in cases of SCDS may provide insight into the origin of the SP, leading to new applications of this electrophysiologic measure.
Furthermore, an UpToDate review on "Causes of vertigo" (Furman, 2019) states that "In semicircular canal dehiscence syndrome, the bone overlying the superior aspect of the superior semicircular canal becomes thin or even absent, thereby allowing pressure to be transmitted to the inner ear. Vertigo is provoked by coughing, sneezing, and Valsalva maneuver. Patients may experience nausea and instability during brief episodes of vertigo … While this condition is increasingly recognized, it may still be under-diagnosed. The diagnosis can be established with high-resolution CT of the temporal bone. Some patients benefit by surgical repair of their anatomic deficit". However, this review does not mention ECoG as a management tool.

**CPT Codes/ HCPCS Codes/ICD-10 Codes**
Information in the [brackets] below has been added for clarification purposes. Codes requiring a 7th character are represented by "+"

<table>
<thead>
<tr>
<th>Code</th>
<th>Code Description</th>
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<tbody>
<tr>
<td>92584</td>
<td>Electrocochleography</td>
</tr>
<tr>
<td>69930</td>
<td>Cochlear device implantation, with or without mastoidectomy</td>
</tr>
<tr>
<td>70540</td>
<td>Magnetic resonance (e.g., proton) imaging, orbit, face, and/or neck; without contrast material(s)</td>
</tr>
<tr>
<td>70542</td>
<td>with contrast material(s)</td>
</tr>
<tr>
<td>92587</td>
<td>Evoked otoacoustic emissions; limited (single stimulus level, either transient or distortion products)</td>
</tr>
<tr>
<td>92588</td>
<td>comprehensive or diagnostic evaluation (comparison of transient and/or distortion product otoacoustic emissions at multiple levels and frequencies)</td>
</tr>
<tr>
<td>92650</td>
<td>Auditory evoked potentials; screening of auditory potential with broadband stimuli, automated analysis</td>
</tr>
<tr>
<td>92651</td>
<td>Auditory evoked potentials; for hearing status determination, broadband stimuli, with interpretation and report</td>
</tr>
<tr>
<td>Code</td>
<td>Code Description</td>
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<tr>
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<tr>
<td>92652</td>
<td>Auditory evoked potentials; for threshold estimation at multiple frequencies, with interpretation and report</td>
</tr>
<tr>
<td>92653</td>
<td>Auditory evoked potentials; neurodiagnostic, with interpretation and report</td>
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**CD-10 codes covered if selection criteria are met**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>H81.01 - H81.09</td>
<td>Meniere's disease</td>
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<tr>
<td>H81.10 - H81.13</td>
<td>Vertigo</td>
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<tr>
<td>H81.311 - H81.49</td>
<td>Labyrinthine fistula</td>
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<tr>
<td>H83.3X1 - H83.3X9</td>
<td>Noise effects on inner ear</td>
</tr>
<tr>
<td>H90.3</td>
<td>Sensorineural hearing loss, bilateral</td>
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<tr>
<td>H90.41 - H90.42</td>
<td>Sensorineural hearing loss, unilateral, with unrestricted hearing on the contralateral side</td>
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<tr>
<td>H90.5</td>
<td>Unspecified sensorineural hearing loss</td>
</tr>
<tr>
<td>H90.6 - H90.8</td>
<td>Mixed conductive and sensorineural hearing loss</td>
</tr>
<tr>
<td>H90.A11 - H90.A12</td>
<td>Conductive hearing loss, unilateral, with restricted hearing on the contralateral side</td>
</tr>
<tr>
<td>H91.20 - H91.23</td>
<td>Sudden idiopathic hearing loss</td>
</tr>
<tr>
<td>H91.8X1 - H91.8X9</td>
<td>Other specified hearing loss</td>
</tr>
<tr>
<td>H93.11 - H93.19</td>
<td>Tinnitus</td>
</tr>
<tr>
<td>R26.89</td>
<td>Other abnormalities of gait and mobility [imbalance]</td>
</tr>
<tr>
<td>R42</td>
<td>Dizziness and giddiness</td>
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</tbody>
</table>

**CD-10 codes not covered for indications listed in the CPB (not all-inclusive)**
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<table>
<thead>
<tr>
<th>Code</th>
<th>Code Description</th>
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<tbody>
<tr>
<td>S09.91xA</td>
<td>Unspecified injury of ear [cochlear trauma]</td>
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<tr>
<td>S09.91xS</td>
<td></td>
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<tr>
<td>Z01.10</td>
<td>Encounter for examination of ears and hearing without abnormal findings [routine screen without signs/symptoms]</td>
</tr>
<tr>
<td>Z01.110</td>
<td>Encounter for hearing examination following failed hearing screening [routine screen without signs/symptoms]</td>
</tr>
</tbody>
</table>

The above policy is based on the following references:

Electrocochleogram


22. Kim JS, Tejani VD, Abbas PJ, Brown CJ. Postoperative electrocochleography from hybrid cochlear implant users: An


Perilymphatic Pressure Measurement


AETNA BETTER HEALTH® OF PENNSYLVANIA

Amendment to
Aetna Clinical Policy Bulletin Number: 0564 Electrocochleogram and Perilymphatic Pressure Measurement

There are no amendments for Medicaid.

revision 08/10/2021