Aetna considers any of the following surgical procedures medically necessary for the treatment of chronic refractory Meniere's disease (see selection criteria in the Appendix of the background section):

Surgical procedures non-destructive to hearing:

- Endolymphatic mastoid shunt
- Endolymphatic sac decompression/endolymphatic sac drainage
- Intra-tympanic corticosteroids injections/perfuisions
- Lateral semi-circular canal plugging
- Perilymphatic fistula patching
- Sacculotomy
- Tympanostomy tube insertion
- Vestibular nerve decompression
- Vestibular neurectomy (nerve section) or neurotomy (including middle fossa or retrosigmoid vestibular neurotomy).

*Please see amendment for Pennsylvania Medicaid at the end of this CPB.*
Surgical procedures destructive to hearing

- Cochleosacculotomy
- Intra-tympanic gentamicin
- Labyrinthectomy
- Translabyrinthine vestibular neurectomy
- Vestibulocochlear neurectomy

**Note:** For bilateral Meniere's disease, ablative treatments are relatively contraindicated due to the risks of bilateral vestibular and cochlear hypofunction.

Aetna considers surgical intervention (e.g., middle fossa craniotomy and trans-mastoid canal plugging) medically necessary for the treatment of superior semicircular canal dehiscence if diagnosis is confirmed by audiometry and computed tomography.

Aetna considers any of the following surgical procedures experimental and investigational for the treatment of chronic refractory Meniere's disease because their effectiveness has not been established:

- Cochleostomy with neurovascular transplant
- Intra-tympanic injection of dexamethasone thermo-sensitive gel
- Simultaneous labyrinthectomy with cochlear implantation (for bilateral Meniere's disease)
- Tenotomy of the stapedius and tensor tympani muscles
- Triple semi-circular canal plugging.

**Background**

Meniere's disease (MD) is a pathological condition of the inner ear characterized by vertigo, tinnitus and a progressive loss of hearing. Delayed MD can develop in an ear that was
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damaged years earlier, usually by viral or bacterial infection. The majority of patients with MD also experience a sense of fullness and pressure in the area of the affected ear. This disorder affects male and female equally; it may occur in children, but has a peak onset between 20 and 50 years of age. The incidence of disease affecting both ears increases to over 40% with long-term follow-up. Moreover, it has been reported that a high percentage of patients (57%) had complete resolution of symptoms in 2 years.

The key pathological finding for patients with MD is an increase in the volume of endolymph, in conjunction with distention of the whole endolymphatic system (known as endolymphatic hydrops). However, the exact cause of MD is still unclear, and no treatment has prospectively modified the clinical course of the condition and thereby prevented the progressive hearing loss. Conservative management of patients with MD may include dietary salt restriction (1 to 2 g sodium daily) with or without diuretic (e.g., hydrochlorothiazide, dyazide, furosemide, amiloride, acetazolamide, and methazolamide), avoidance of caffeine, alcohol and nicotine. For acute attacks, vestibular suppressants (e.g., benzodiazepines and diazepam) or anti-emetics (e.g., anti-cholinergics such as glycopyrrolate, anti-dopaminergics such as droperidol, prochlorperazine, and anti-histamines such as dimenhydrinate, diphenhydramine, meclizine, and promethazine) have been used. Medical ablation of the inner ear with systemic administration of ototoxic aminoglycosides, such as streptomycin and gentamicin, has been useful in advanced bilateral MD when poor but aidable hearing precludes surgical intervention. Moreover, the indications for this approach are limited, especially with the advent of intra-tympanic placement of gentamicin.

For MD patients whose vertiginous symptoms are disabling and refractory to dietary and medical treatments, surgery may be the last resort in achieving relief. In general, surgical procedures for MD can be categorized as non-destructive or
destructive regarding to hearing. The former includes endolymphatic sac surgery and vestibular nerve section (vestibular neurectomy), while the latter includes labyrinthectomy (extirpation of the labyrinth) and cochleosacculotomy.

Endolymphatic sac surgery is the most frequently employed conservative surgical approach for patients with MD when hearing is still serviceable. It has been reported to achieve complete or substantial control of vertigo in 81 % of patients, with significant improvement in hearing in about 20 % of patients. Although it has shown to be less likely to completely eliminate vertigo than vestibular nerve section, endolymphatic sac surgery has been reported to be a low morbidity procedure. Endolymphatic sac decompression and sacculotomy are two common types of endolymphatic sac surgery. In the former procedure, some of the bone surrounding the inner ear is removed. In some cases, endolymphatic sac decompression is coupled with the placement of an endolymphatic shunt. Sacculotomy entails the implantation of a permanent device that allows endolymph to drain out of the inner ear whenever pressure builds up.

Brinson et al (2007) compared the effectiveness of endolymphatic mastoid shunt (EMS, n = 88) versus endolymphatic sac decompression (ESD, n = 108) without sac incision for the treatment of MD. The American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) guidelines for the diagnosis and evaluation of therapy in MD were used to retrospectively identify suitable candidates for the study. All patients who failed medical management and underwent either EMS or ESD were selected for review using the AAO-HNS guidelines. The study was carried out at a tertiary care neurotology private practice. EMS and ESD were equally effective in reducing the incidence and severity of vertigo attacks with significant improvement in 67 % and 66 % of patients, respectively. The authors concluded that both
EMS and ESD are effective, non-destructive alternatives for patients who have failed medical management of MD with similar long-term hearing outcomes.

Available medical literature suggests that vestibular nerve section can be performed by exposing the middle fossa or the posterior fossa. The posterior fossa vestibular neurectomy can be further divided into the retrolabyrinthine approach, the retrosigmoid-internal auditory canal approach, and the combined retrolabyrinthine-retrosigmoid approach. Vestibular nerve section has been shown to be generally very effective in the control of vertigo. According to available literature, vestibular nerve section is contraindicated in persons with vertigo arising from the only hearing ear (i.e., the other ear is deaf); central nervous system disease; persons in poor medical condition; persons with ataxia; and in most cases of bilateral MD.

Labyrinthectomy is indicated for patients with symptoms who have poor or non-serviceable hearing. There are several approaches to the labyrinth -- transcanal labyrinthectomy with section of the posterior ampullary nerve, transmastoid labyrinthectomy, and transmeatal cochloevestibular neurectomy. For the older patients (greater than 60 years of age), available medical literature suggests that a transmastoid labyrinthectomy may be preferable to a transcanal labyrinthectomy because the incidence of permanent post-surgical imbalance is less with the former approach. Labyrinthectomy has been reported to produce similar or better results than vestibular neurectomy.

Cochleosacculotomy creates a permanent fistula in the cochlear duct by passing a small pick through the round window. It is mainly used for elderly patients with poor hearing who are unable to tolerate a prolonged operation.

Lacombe (2009) stated that spontaneous recovery or central compensation makes surgical procedures rare in patients with vertigo. The main target in treating MD is to promote

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vestibular compensation, which is possible only with a non-progressive and stable deficit leading to readjustment of vestibular reflexes. Surgical procedures can be classified as non-destructive (endolymphatic sac decompression, vestibular nerve decompression, patching of perilymphatic fistulas), selectively destructive (middle fossa or retrosigmoid vestibular neurotomy, lateral semi-circular canal plugging) and destructive (labyrinthectomy). Surgical indications essentially concern incapacitating vertigo and depend mainly on hearing status. In MD, vestibular neurotomy can be regarded as the gold standard considering its good results on vertiginous episodes; however, scoring with functional and quality-of-life scales bring out residual deficiency in some cases.

In a prospective, follow-up study, Charpiot and colleagues (2010) assessed the safety and effectiveness lateral semi-circular canal plugging to control vertigo in severe MD. A total of 28 MD patients with refractory vertigo and severe disability (functional scale 5 or 6) were included in this study. Lateral semi-circular canal plugging was performed in the pathological ear for each patient. The evaluation of therapy followed the guidelines for diagnosis and evaluation of therapy in MD. Hearing, frequency of vertigo, and functional disability were assessed in the early follow-up (6 months) for all the patients and in the late follow-up (2 years) for 16 patients. In addition, canal paresis was evaluated by the caloric test. No vital complication occurred. The hearing was preserved in 82 % of cases. Lateral semi-circular canal plugging induced in all cases canal paresis that was persistent after 2 years. After 2 years (n = 16), the control of vertigo was complete or substantial in 75 % of cases (restoration of a normal life = 62.5 %; no functional restriction =12.5 %). The authors concluded that lateral semi-circular canal plugging is a safe procedure that induces canal paresis and allows a good control of vertigo. In view of these results, lateral semi-circular canal plugging should be a therapeutic option for controlling rotatory vertigo in severe MD.
In a review on bilateral MD, Nabi and Parnes (2009) stated that for bilateral MD, ablative treatments are relatively contraindicated due to the risks of bilateral vestibular and cochlear hypofunction.

In a Cochrane review, Pullens et al (2011) evaluated the effectiveness of intra-tympanic gentamicin in the treatment of vertigo in MD. These investigators searched the Cochrane Ear, Nose and Throat Disorders Group Trials Register; the Cochrane Central Register of Controlled Trials (CENTRAL); PubMed; EMBASE; CINAHL; Web of Science; BIOSIS Previews; Cambridge Scientific Abstracts; ISRCTN and additional sources for published and unpublished trials. The date of the most recent search was June 30, 2010. All randomized or quasi-randomized controlled trials of intra-tympanic gentamicin versus placebo, or versus another treatment for MD were selected for analysis. Two review authors independently assessed trial quality and extracted data. They contacted study authors for further information. These investigators identified 2 trials, involving 50 participants, which fulfilled the inclusion criteria. Both of these trials are prospective, double-blind, placebo-controlled randomized clinical trials on the effect of intra-tympanic gentamicin on vertigo complaints. Both of these trials found a significant reduction in vertigo complaints in the gentamicin group when compared to the placebo group. Due to clinical heterogeneity these researchers could not perform a meta-analysis. The authors concluded that based on the results of the 2 included studies, intra-tympanic gentamicin seems to be an effective treatment for vertigo complaints in MD, but carries a risk of hearing loss.

Gawecki et al (2012) estimated the results of treatment of MD with intra-tympanic injections of gentamicin. A total of 37 patients with defined, pharmacological treatment resistant MD were injected intra-tympanic with 0.3 ml (12 mg) of gentamicin once or few times with 7 days or longer breaks and a number of injections depended on the reaction of the inner ear. These
investigators estimated the patients' subjective feelings and results of equilibrium and hearing organ examination in early (3 months) and late (2 years) period after treatment. Complete control of vertigo (class A) was achieved in 84.6 %, and complete and essential control (class A and B) in 96.1 %. Hearing deterioration usually mild or moderate was observed directly after treatment in 16.2 % and after 2 years in 23 % patients. The results of pure tone audiometry showed deterioration of hearing in 16.2 % (early) and 26.9 % (late). In 1 patient hearing deterioration was essential. The authors concluded that intra-tympanic injections of gentamicin are effective and not troublesome method of treatment of pharmacological treatment resistant MD. In most of patients hearing can be preserved, but they should be always informed about possible risk of hearing deterioration. The number of injections and breaks between them depends on the effect of therapy and of expectations of patients.

An UpToDate review on “Meniere disease” (Dinces and Rauch, 2013) states that “Destructive procedures for the treatment of Meniere disease include intratympanic gentamicin injection, surgical labyrinthectomy, and vestibular nerve section. In general, destructive techniques are better suited to patients who have failed medical therapy and who have unilateral disease”.

The National Coverage Determination (NCD) for “Cochleostomy with Neurovascular Transplant for Meniere's Disease” stated that “While there are 2 recognized surgical procedures used in treating MD (decompression of the endolymphatic hydrops and labyrinthectomy), there is no scientific evidence supporting the safety and effectiveness of cochleostomy with neurovascular transplant in treatment of Meniere's syndrome. Accordingly, Medicare does not cover cochleostomy with neurovascular transplant for treatment of MD”.
Furthermore, UpToDate reviews on “Meniere disease” (Dinces and Rauch, 2013) and “Treatment of vertigo” (Furman and Barton, 2013) do not mention the use of cochleostomy and tenotomty as therapeutic options.

Mick et al (2014) stated that MD that results in bilateral severe to profound sensori-neural hearing loss (SNHL) is a rare indication for cochlear implantation (CI); only a few studies had documented performance in these patients. These researchers compared the difference in pre-operative to 12-month post-operative speech perception scores among subjects with MD and controls. Group-wise comparisons of secondary post-operative outcomes (Tinnitus Handicap Inventory [THI] scores, 36-Item Short Form [SF-36] scores, and post-operative dizziness) were also performed. A retrospective cohort study was conducted. Subjects with MD and controls matched by age, device manufacturer and model, pre-operative sentence score, and sentence test used for pre-implantation and post-implantation performance assessments were identified from 1,130 patients in the prospectively maintained CI database at the authors’ center. Speech perception, THI, and SF-36 scores and demographic variables were obtained from the database. Vestibular outcomes were obtained by retrospective chart review. Statistical comparisons were performed to compare pre-operative to post-operative change between groups. A total of 20 patients with MD were identified. At 1 year after CI, improvements in sentence and word understanding did not differ in magnitude from the controls. Tinnitus was reduced significantly in patients with MD, whereas there was a trend for improvement in the controls. Quality of life as measured by the SF-36 improved in both groups. Patients with MD had significant improvements in 1 domain compared with 5 domains for the controls. Subjects with MD had significantly more chronic dizziness in the post-operative period than did controls. The authors concluded that patients with MD who have bilateral severe to profound SNHL benefited significantly from CI; ongoing dizziness in some patients with MD may result in
quality of life improvements that were slightly less than seen for the average adult patient with CI. Moreover, they stated that larger studies are needed to corroborate the results.

Mackeith et al (2014) noted that the development of second-side MD in the only remaining serviceable ear is difficult to treat. These researchers described an intervention to control disabling disease combining labyrinthectomy and CI to restore hearing. Following a thorough pre-operative assessment and consenting process, 2 patients underwent labyrinthectomy of the affected ear with simultaneous CI. Both patients achieved control of Meniere's attacks with improved hearing rehabilitation. Oscillospia was noted by both patients; both patients were pleased to have undergone the treatment. The authors proposed this as a potential management strategy in this rare but complex group of patients in whom all less destructive measures have failed. These preliminary findings need to be validated by well-designed studies.

Endolymphatic Sac Drainage

Yu et al (2007) investigated the effectiveness of endolymphatic sac drainage for MD (n = 26). Of the 26 patients, 18 were followed-up for more than 2 years post-surgery. According to Chinese Meniere disease's diagnosis and curative effect standard evaluation criteria published in 1996, for vertigo symptom of these 18 patients, there were 9 cases (50 %) with grade A (completely controlled), 8 cases (44.4 %) with grade B (fundamentally controlled) and 1 case (5.6 %) with grade D (not controlled). The vertigo fully controlled rate was only 50 %, but the vertigo completely or fundamentally controlled rate reached 94.4 %. Tinnitus of the patients after operation disappeared in 2 cases (11.1 %), reduced in 9 cases (50 %) and unchanged in 7 cases (38.9 %). Hearing post-surgery was improved in 6 cases (33.3 %), unchanged in 4 cases (22.2 %) and decreased in 8 cases (44.5 %). The authors
concluded that endolymphatic sac drainage was a safe and effective management as well as with less complication for intractable MD patients with residual hearing before operation.

Kitahara et al (2013) examined endolymphatic sac drainage with intra-endolymphatic sac application of large doses of steroids for intractable MD patients and observed long-term results from 2 years to over a decade until 13 years. Between 1998 and 2009, these researchers enrolled and assigned 286 intractable MD patients to 2 groups: group-I (G-I) included patients who underwent endolymphatic sac drainage with steroid instillation and group-II (G-II) included those who declined endolymphatic sac drainage. Definitive spells and hearing improvement in these 2 groups were determined for 2 to 13 years after treatment. According to the established criteria, vertigo was completely controlled in 88 % of patients in G-I in the 2nd year, in 73 % in the 12th year and in 70 % in the 13th year. These results in G-I were significantly better than those in G-II for 13 years after treatment. Hearing was improved in 49 % of patients in G-I in the 2nd year, in 27 % in the 12th year and in 25 % in the 13th year. These results in G-I were significantly better than those in G-II for 12 years after treatment, but this was not significant in the 13th year. The authors concluded that endolymphatic sac drainage with intra-endolymphatic sac application of large doses of steroids could improve long-term follow-up results of hearing as well as vertigo control. This meant that the drainage with local steroids could also improve patients’ long-term quality in the prime of life.

Sakagami et al (2015) noted that patients with MD sometimes face recurrent problems years after endolymphatic sac drainage with steroid instillation surgery (EDSS) due to endolymphatic sac closure and/or disease progression. These investigators examined the effects of EDSS with posterior tympanotomy with steroids at the round window (EDRW) on vertigo and hearing after revision surgery for intractable relapsed MD. A total of 16 patients with MD had revision
surgery due to intractable recurrence of disease, and were followed-up regularly at least for 2 years. As revision surgery, EDSS was performed in 8 cases and EDRW was performed in the other 8. There were no significant differences between the patients' backgrounds in the 2 groups. Periods of hearing recovery to the pre-operative level were 11.5 ± 4.4 months after the first EDSS, although it took 16.4 ± 2.6 months longer after revision surgery with the second EDSS ($p = 0.038 < 0.05$: first EDSS versus second EDSS) and was 10.0 ± 3.3 months shorter after revision surgery with EDRW ($p = 0.010 < 0.05$: second EDSS versus EDRW). The authors concluded that at the second post-operative year, there were no significant differences between results for vertigo and hearing after EDSS and EDRW. In particular, as regards hearing recovery to the pre-operative level, the periods after EDRW were shorter than those after the second EDSS.

Tenotomy of the Stapedius and Tensor Tympani Muscles

In an interventional cohort study, Loader et al (2012) compared the unique long-term results of tenotomy of the stapedius and tensor tympani muscles in definite MD refractory to medical treatment and presented a hypothesis on why tenotomy seems effective. The study sample comprised 30 patients (15 males, 15 females; average age of 57 +/- 13.1 years) with definite MD (AAO-HNS criteria, 1995). Patients were evaluated pre- and post-operatively using pure tone audiometry, AAO-HNS questionnaires regarding vertigo attacks, functional level scores, and tinnitus, and were followed-up for 2 to 9 years. Post-operative values were calculated for the patient collective as a whole and consequently divided into 3 equal post-operative terms of 3 years each. A statistically significant improvement of inner ear hearing levels post-operatively ($p = 0.041$) and a major reduction in vertigo attacks in all groups ($p < 0.001$) with complete absence of attacks in 26/30 patients was noted. Results remained constant up to 9 years post-operatively. Although tinnitus persisted, the intensity was lower overall ($p =
0.013). Based on the immediate and persistent reduction of vertigo and a clear improvement in hearing function and functional scales, the authors concluded that tenotomy is effective in unilateral, definite MD. They stated that the findings of this study laid the foundation for future prospective, randomized controlled trials.

Albu et al (2015) compared the outcomes of patients with MD submitted to either endolymphatic mastoid shunt (EMS) or tenotomy of the stapedius and tensor tympani muscles (TSTM). This was a retrospective chart review of patients treated with EMS or TSTM between 2000 and 2010 and followed-up for at least 12 months. The main outcomes were represented by: (i) vertigo class, hearing stage and functional level according to the AAO-HNS criteria; (ii) adjustment of dizziness handicap inventory (DHI), and (iii) complete and substantial vertigo control using the Kaplan-Meier survival method. A total of 63 patients met the inclusion criteria: 34 underwent EMS and 29 TSTM. The baseline demographic characteristics, the hearing stage, the functional level, the DHI and hearing levels were not different between the 2 groups. No significant difference in vertigo class was demonstrated: 66% of TSTM patients attained class A compared to 44% in the EMS group (p = 0.14). Kaplan-Meier survival curves specific to class A showed significant differences, favoring TSTM (log-rank test, p = 0.022). Patients who underwent TSTM demonstrated significantly improved functional level (p = 0.0004) and improved DHI scores (p = 0.001); 8 EMS patients (25%) demanded a 2nd surgical attempt compared to none in the TSTM. Aural fullness was significantly improved in TSTM group (p = 0.01), while the difference in tinnitus improvement was non-significant. Hearing preservation was significantly better in TSTM group (p = 0.001). The authors concluded that TSTM is a safe surgical procedure, with significant vertigo control rates, and important hearing preservation rates. Moreover, they stated that more patients and longer follow-up are needed to support these preliminary findings.
Reichmayr and colleagues (2019) noted that TSTM is a relatively unknown therapeutic procedure in MD. Widespread approaches include pharmacotherapies with betahistine or diuretics as well as interventional procedures, such as intra-tympanic gentamicin or glucocorticoid injection, vestibular neurectomy, labyrinthectomy or endolymphatic sac surgery. The exact pathomechanism of this approach is not fully known. It is assumed that by cutting the tendons of both middle ear muscles in cases of endolymphatic hydrops the stapes is not additionally actively pushed against the oval window but can deviate laterally and thereby does not augment the inner ear pressure even further. Studies have shown that this method does not only improve vestibular symptoms but also, in contrast to most other strategies, increases the hearing level. The formation of scar tissue and the resulting reduction of ossicular chain mobility, especially due to post-operative infections, may limit the success of tenotomy and should be considered as a possible factor in cases of limited post-operative vertigo control.

Furthermore, an UpToDate review on “Meniere disease” (Moskowitz and Dinces, 2019) does not mention tenotomy of the middle ear muscles as a therapeutic option.

**Intra-Tympanic Corticosteroids Injections / Perfusions**

In a retrospective study, She and colleagues (2015) examined the long-term effectiveness of intra-tympanic methylprednisolone perfusion for the treatment of patients with intractable MD (n = 17). Treatment effectiveness was evaluated according to the AAO-HNS criteria. Short- and long-term control or improvement rates were calculated after 6 and 24 months, respectively; 16 patients were followed for more than 2 years. Short- and long-term vertigo control rates were 94 % and 81 %, respectively; short- and long-term functional activity improvements were 94 % and 88 %, respectively. The pure tone average was 53 ± 14 dB before treatment, and 50 ± 16 dB at 6 months and 52 ± 20 dB at 24 months after intra-
typanic methylprednisolone perfusion. Tinnitus was controlled or improved in 5 patients over the 2-year follow-up period. The authors concluded that intra-tympanic methylprednisolone perfusion could effectively control vertigo and improve functional activity in intractable MD patients with good hearing preservation. They stated that this approach may therefore be a viable alternative treatment for intractable MD. The main drawbacks of this study were its retrospective design and its small sample size (n = 17). These findings need to be validated by well-designed studies.

In a systematic review, Lavigne and colleagues (2016) examined the evidence of intra-tympanic steroids injections (ITSI) for effectiveness in the management of the following inner ear diseases: MD, tinnitus, noise-induced hearing loss (NIHL) and idiopathic sudden sensorineural hearing loss (ISSNHL). The data sources were literature review from 1946 to December 2014, PubMed and Medline. A systematic review of the existing literature was performed. Databases were searched for all human prospective randomized clinical trials using ITSI in at least 1 treatment group. The authors identified 29 prospective randomized clinical trials investigating the benefits of an intra-tympanic delivery of steroids. A total of 6 articles on MD were identified, of which 1 favored ITSI over placebo in vertigo control. Of the 5 randomized clinical trials on tinnitus therapy, 1 study found better tinnitus control with ITSI. The only available trial on NIHL showed significant hearing recovery with combination therapy (ITSI and oral steroids therapy). A total of 17 studies were identified on ISSNHL, of which 10 investigated ITSI as a first-line therapy and 7 as a salvage therapy. Studies analysis found benefits in hearing recovery in both settings. Due to heterogeneity in treatment protocols and follow-up, a meta-analysis was not performed. The authors concluded that given the low adverse effects rates of ITSI therapy and good patient tolerability, local delivery should be considered as an interesting adjunct to the therapy of the ISSNHL and NIHL. Moreover, they stated that only 1 article over 6 where ITSI therapy offered potential
benefits to patients with MD in the control of tinnitus and vertigo was found; and ITSI did not appear to be effective in the treatment of tinnitus.

An UpToDate review on “Meniere disease” (Dinces, 2016) states that “Intratympanic glucocorticoids may improve vertigo symptoms, but await controlled trials”.

Patel and colleagues (2016) stated that MD is characterized by severe vertigo attacks and hearing loss. Intra-tympanic gentamicin, the standard treatment for refractory MD, reduces vertigo, but damages vestibular function and can worsen hearing. In a double-blind, comparative effectiveness clinical trial, these researchers examined if intra-tympanic administration of the corticosteroid methylprednisolone (non-destructive to hearing) reduces vertigo compared with gentamicin. Patients aged 18-70 years with refractory unilateral MD were enrolled at Charing Cross Hospital (London, UK) and Leicester Royal Infirmary (Leicester, UK). Patients were randomly assigned (1:1) by a block design to two intra-tympanic methylprednisolone (62.5 mg/ml) or gentamicin (40 mg/ml) injections given 2 weeks apart, and were followed-up for 2 years. All investigators and patients were masked to treatment allocation. The primary outcome was vertigo frequency over the final 6 months (18 to 24 months after injection) compared with the 6 months before the first injection. Analyses were done in the intention-to-treat population, and then per protocol. Between June 19, 2009, and April 15, 2013, a total of 256 patients with MD were screened, 60 of whom were enrolled and randomly assigned: 30 to gentamicin and 30 to methylprednisolone. In the intention-to-treat analysis (i.e., all 60 patients), the mean number of vertigo attacks in the final 6 months compared with the 6 months before the first injection (primary outcome) decreased from 19.9 (SD 16.7) to 2.5 (5.8) in the gentamicin group (87 % reduction) and from 16.4 (12.5) to 1.6 (3.4) in the methylprednisolone group (90 % reduction; mean difference -0.9, 95 % confidence interval: -3.4 to 1.6). Patients whose
vertigo did not improve after injection (i.e., non-responders) after being assessed by an unmasked clinician were eligible for additional injections given by a masked clinician (8 patients in the gentamicin group versus 15 in the methylprednisolone group); 2 non-responders switched from methylprednisolone to gentamicin. Both drugs were well-tolerated with no safety concerns; 6 patients reported 1 adverse event (AE) each: 3 in the gentamicin group and 3 in the methylprednisolone group. The most common AE was minor ear infections, which was experienced by 1 patient in the gentamicin group and 2 in the methylprednisolone group. The authors concluded that methylprednisolone injections are a non-ablative, effective treatment for refractory MD; the choice between methylprednisolone and gentamicin should be made based on clinical knowledge and patient circumstances.

Vestibular Neurotomy

In a prospective study, Miyazaki and colleagues (2017) evaluated the state of dizziness and anxiety of patients with incapacitating MD and its improvement through minimally invasive vestibular neurotomy (MIVN). A total of 118 patients with incapacitating MD who underwent MIVN in France and Japan were evaluated. The DHI, SAST (Short Anxiety Screening Test), and STAI (State Trait Anxiety Index) questionnaires were used to evaluate disequilibrium and anxiety. The authors concluded that the MIVN method appeared safe and effective for patients with incapacitating MD. Pre-operative assessment results by DHI and SAST were significantly related to each other, and were influenced by lifestyle and profession.

Intra-Tympanic Injection of Dexamethasone Thermo-Sensitive Gel

In a randomized, double-blind, placebo-controlled, phase IIb clinical trial, Lambert and colleagues (2016) evaluated the safety and effectiveness of a single intra-tympanic injection of
OTO-104, sustained-exposure dexamethasone, in patients with unilateral MD. A total of 154 patients (77 per group) aged 18 to 85 years from 52 academic and community otolaryngology centers were included in this study. Participants received single intra-tympanic injection of OTO-104 (12 mg dexamethasone) or placebo. Main outcome measures were efficacy (vertigo) and safety (adverse events [AE], otoscopy, audiometry, and tympanometry). Primary endpoint (change from baseline in vertigo rate at Month 3) was not statistically significant (placebo [-43 %], OTO-104 [-61 %], p=0.067). Improvements with OTO-104 were observed in prospectively defined secondary end-points number of days with definitive vertigo, (Month 2 [p =0.035], Month 3 [p=0.030]), vertigo severity (Months 2 to 3, p=0.046) and daily vertigo counts (Month 2, p=0.042), and in some Short Form-36 (SF-36) subscales (Month 2 bodily pain p=0.039, vitality p=0.025). No difference in tinnitus loudness or THI-25 was observed; OTO-104 was well-tolerated; no negative impact on safety compared with placebo. Persistent tympanic membrane perforation was observed in 2 OTO-104 treated patients at the end of the trial. The authors concluded that OTO-104 was well-tolerated, did not significantly affect change from baseline in vertigo rate, but reduced number definitive vertigo days, vertigo severity, and average daily vertigo count compared with placebo during Month 3. They stated that these findings provided insight into analyzing for a vertigo treatment effect and support advancing OTO-104 into phase III clinical trials for the treatment of MD symptoms.

Triple Semi-Circular Canal Plugging

Zhang and colleagues (2016) examined the long-term effectiveness of triple semi-circular canal plugging (TSCP) in the treatment of intractable MD. A total of 79 patients diagnosed with unilateral MD referred to a vertigo clinic of the hospital between December 2010 and December 2013 were included in this study for retrospective analysis; TSCP was
performed in the affected ear for each patient. Vertigo control and auditory function were measured. Pure tone audiometry, caloric test, and cervical vestibular evoked myogenic potential (cVEMP) were performed in 2-year follow-up; 36 MD patients, who accepted ESD operation were selected as a comparison group. The total control rate of vertigo in the TSCP group was 98.7% in the 2-year follow-up, with a complete control rate of 81.0% and substantial control rate of 17.7%. The rate of hearing preservation was 70.9%. The total control rate of vertigo in the ESD operation group was 72.2%. The vertigo control rate of TSCP was significantly higher than that of ESD operation; 24 months after treatment, canal paresis was found in the operation side of all patients of TSCP by means of caloric test. These preliminary findings need to be validated by well-designed studies.

Furthermore, an UpToDate review on “Meniere disease” (Moskowitz and Dinces, 2017) does not mention triple semi-circular canal plugging as a therapeutic option.

Superior Semicircular Canal Dehiscence Surgery

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 (Furman, 2020).

Welgampola et al (2008) stated that diagnosis of the superior canal dehiscence syndrome (SCDS) relies on symptoms such as sound- or pressure-induced vertigo or oscillopsia, demonstration of sound or pressure-evoked vertical/torsional eye movements, and the presence of a defect in the bony roof overlying the superior semicircular canal. Lowered thresholds for eliciting vestibular evoked myogenic potentials (VEMPs) provide additional confirmation. These researchers examined
VEMP characteristics before and after canal plugging for SCDS. VEMPs evoked by air- and bone-conducted tones were measured from the sternocleidomastoid muscles (cVEMP) and peri-ocular sites (oVEMP) of 20 normal volunteers, 10 newly diagnosed subjects with SCDS, and 12 subjects who underwent successful superior canal plugging. In all SCDS ears, thresholds for evoking VEMP using air-conducted tones were pathologically lowered, with average values of 83.85 +/- 1.40 dB sound pressure level (SPL) for cVEMP and 85.38 +/- 1.32 dB SPL for oVEMP, 20 to 30 dB below those of controls. Successful canal plugging resulted in normal reflex thresholds. For bone vibration, average thresholds in SCDS ears were 114.62 +/- 1.54 dB FL (force level) for cVEMP and 116.0 +/- 1.52 dB FL for oVEMP, 10 to 20 dB below controls, yet 3 SCDS ears had normal thresholds. The authors concluded that ocular and cVEMPs evoked by air-conducted sound were equally useful in the diagnosis and follow-up of SCDS. Stimulus thresholds were consistently lowered upon presentation and normalized after corrective surgery. Thresholds for bone vibration, in contrast, have a lower diagnostic yield.

In a retrospective review, Niesten et al (2013) examined the utility of cervical VEMP thresholds in the surgical management of bilateral SCDS. These investigators identified patients who underwent surgical treatment for SCDS from their database of 147 patients diagnosed with SCD between 2000 and 2011 at their institution. The diagnosis of SCDS was based on clinical signs and symptoms, audiometric and cervical VEMP testing, and high-resolution computed tomography (CT). These researchers identified 38 patients who underwent SCD surgery in 40 ears (2 bilateral). In 7 patients with bilateral SCD, the more symptomatic ear had lower cervical VEMP thresholds, a larger air bone gap and a lateralizing tuning fork. In 13 patients with peri-operative cervical VEMP testing, thresholds increased in 12 patients following primary repair, and no threshold shift was observed in 1 patient with persistence of symptoms after revision surgery. Audiometric data showed a
significant mean decrease of the low-frequency air-bone gap and a mild (high-frequency) bone conduction loss after surgical repair. The authors found that pre-operative cervical VEMP thresholds, the magnitude of the air-bone gap and tuning-fork testing were important to confirm the worse ear in patients with bilateral SCD; elevation of cervical VEMP thresholds following surgery correlated with improvement of symptoms and underscored the importance of post-operative testing in patients with bilateral disease or recurrence of symptoms; and SCD plugging was associated with a partial closure of the air-bone gap and a mild (high-frequency) SNHL.

Gioacchini et al (2016) noted that superior semicircular canal dehiscence (SSCD) represents a rare condition that may be associated to some particular symptoms as vertigo, autophony, and Tullio phenomenon. In those patients who present severe symptoms surgical treatment is needed. Middle fossa craniotomy and trans-mastoid approaches were both described. Concerning repairing techniques, plugging and/or resurfacing are typically used to close the defect. These investigators analyzed the overall outcomes and complications of this surgery; and compared the different surgical modalities to examine the eventual advantages and disadvantages. They carried out a search through Ovid Medline in January 2015 to include all eligible articles. A statistical analysis of the obtained data was performed. A total of 20 studies comprising a total of 150 procedures were included; 4 modalities of canal repair were described (plugging, capping, resurfacing, plugging with resurfacing). The overall rate of success resulted in 94 % (95 % confidence interval [CI]: 87 % to 97 %). No statistically significant differences were observed among the different modalities of canal repair concerning both success rate and surgical complications. Differences observed between the middle fossa approach and trans-mastoid approach in terms of outcome were not statistically significant. The authors concluded that the findings of this review showed that surgical treatment for SSCD represented a safe option for those
patients with severe symptoms of this condition. Differences observed in terms of success rate or complications were analyzed between the different surgical techniques with results that were not statistically significant.

In a case-series study with chart review, Banakis Hartl and Cass (2018) examined changes in subjective symptoms in patients following trans-mastoid canal plugging for SSCD syndrome; and quantified changes in hearing in patients who have undergone trans-mastoid canal plugging for SSCD syndrome. These researchers retrospectively reviewed patients with SSCD who underwent repair with canal plugging via a trans-mastoid approach between January 2012 and January 2017. Symptom severity was assessed prospectively (autophony, sound/pressure-induced vertigo, disequilibrium, aural fullness, and pulsatile tinnitus) and after surgery. Pure-tone and speech audiometry were measured before and after surgery; 2-sided Wilcoxon rank-sum tests were used to evaluate changes in subjective symptoms and audiometric outcomes. A total of 17 patients (19 ears) met inclusion criteria. The superior canal was successfully plugged via the trans-mastoid approach in all cases. Patients reported a statistically significant improvement in autophony, vertigo, aural fullness, and pulsatile tinnitus (p < 0.01), without significant improvement in disequilibrium rating (p = 0.06). There were no changes noted in pure-tone average or word recognition score; however, there was a statistically significant improvement in air-bone gap at 250-Hz of 10.9 dB (p = 0.04) with 12.9-dB improvement in air conduction thresholds (p = 0.02) and no difference (0.9 dB, p = 0.9) in bone conduction thresholds. The authors concluded that in this study, patients with SSCD demonstrated excellent hearing outcomes and resolution of most otologic symptoms after surgical repair; and trans-mastoid canal plugging, which has been described to-date only in smaller case series, was a safe and effective alternative to the traditional middle cranial fossa approach.
In a retrospective, multi-center, comparative cohort study, Schwartz et al (2019) compared outcomes for patients undergoing a trans-mastoid approach versus a middle fossa craniotomy approach with plugging and/or resurfacing for repair of SSCD. Outcome measures include symptom resolution, hearing, operative time, hospital stay, complications, and revision rates. Adult patients undergoing repair for SSCD between 2006 and 2017 at 3 neurotology centers were included. Demographics and otologic history collected by chart review. Imaging, audiometric data, and VEMP measurements were also collected for analysis. A total of 68 patients (74 ears) were included in the study; 21 patients underwent middle fossa craniotomy repair (mean age of 47.9 years), and 47 underwent trans-mastoid repair (mean age of 48.0 years). There were no significant differences in age or sex distribution between the groups. The trans-mastoid group experienced a significantly shorter duration of hospitalization and lower recurrence rate as compared with the middle fossa craniotomy group (3.8 % versus 33 %). Both groups experienced improvement in noise-induced vertigo, autophony, pulsatile tinnitus, and non-specific vertigo. There was no significant difference among symptom resolution between groups. Additionally, there was no significant difference in audiometric outcomes between the groups. The authors concluded that both the trans-mastoid approach and the middle fossa craniotomy approach for repair of SSCD offered symptom resolution with minimal risk. The trans-mastoid approach was associated with shorter hospital stays and lower recurrence rate as compared with the middle fossa craniotomy approach.

Nguyen et al (2019) stated that SSCD is caused by a fistula in the arcuate eminence, creating vestibular and auditory disturbances. These researchers examined the effects of gender and age on symptom prevalence and resolution in patients with SSCD. A Boolean search was conducted through 4 separate scientific data-bases. Full-text English articles for SSCD patients, who underwent surgery were
included. Demographics and outcomes were extracted. A total of 198 patients were identified, and available for quantitative analysis. Between genders, there were no differences in the prevalence of pre- or post-operative symptomology. Both genders had statistically significant improvement in symptomatology with females experiencing significantly high rates of hearing loss improvement compared to male patients. Compared to their younger cohort, patients over 65 years of age had similar symptom frequencies before and after surgery with similar rates of symptom resolution. The authors concluded that SSCD repair was safe and effective for resolving auditory and vestibular symptoms. Gender and age may not be strong predictors of patient presentation or symptom resolution; and gender and age-associated factors may not influence patient outcomes.

Steenerson et al (2020) noted that SCDS is a vestibular disorder caused by a pathologic 3rd window into the labyrinth that could present with autophony, sound- or pressure-induced vertigo, and chronic disequilibrium among other vestibulocochlear symptoms. Careful history taking and examination in conjunction with appropriate diagnostic testing could accurately diagnose the syndrome. Key examination techniques include fixation-suppressed ocular motor examination investigating for sound- or pressure-induced eye movements in the plane of the semicircular canal. Audiometry, VEMP s, and CT confirm the diagnosis. Corrective surgical techniques can be curative, but many patients find their symptoms are not severe enough to undergo surgery. Although a primarily peripheral vestibular disorder, as 1st-line consultants for most dizziness complaints, neurologists will serve their patients well by understanding SCDS and its role in the differential diagnosis of vestibular disorders.

An UpToDate review on “Causes of vertigo” (Furman, 2020) states that “Semicircular canal dehiscence syndrome -- While this condition is increasingly recognized, it may still be
underdiagnosed. The diagnosis can be established with high-resolution CT of the temporal bone. Some patients benefit by surgical repair of their anatomic deficit”.

Appendix

Selection criteria for surgery for Meniere’s disease

1. Member has disabling vertigo; and
2. Member has exhibited symptoms (hearing loss, tinnitus, vertigo) for at least 2 years; and
3. Member has failed conservative management, including dietary restrictions (avoidance of caffeine, alcohol and nicotine; low sodium), and medical therapy (anti-emetic, diuretic, and vestibular suppressants); and
4. Member has unilateral Meniere’s disease and hearing loss is severe to profound in the involved ear (for labyrinthectomy only).

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>
</tr>
</thead>
<tbody>
<tr>
<td>61590</td>
<td>Infratemporal pre-auricular approach to middle cranial fossa (parapharyngeal space, infratemporal and midline skull base, nasopharynx), with or without disarticulation of the mandible, including parotidectomy, craniotomy, decompression and/or mobilization of the facial nerve and/or petrous carotid artery</td>
</tr>
<tr>
<td>Code</td>
<td>Code Description</td>
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<tr>
<td>---------</td>
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<tr>
<td>61591</td>
<td>Infratemporal post-auricular approach to middle cranial fossa (internal auditory meatus, petrous apex, tentorium, cavernous sinus, parasellar area, infratemporal fossa) including mastoidectomy, resection of sigmoid sinus, with or without decompression and/or mobilization of contents of auditory canal or petrous carotid artery</td>
</tr>
<tr>
<td>61592</td>
<td>Orbitocranial zygomatic approach to middle cranial fossa (cavernous sinus and carotid artery, clivus, basilar artery or petrous apex) including osteotomy of zygoma, craniotomy, extra- or intradural elevation of temporal lobe</td>
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<tr>
<td>64716</td>
<td>Neuroplasty and/or transposition; cranial nerve (specify) [vestibular (non-destructive to hearing)]</td>
</tr>
<tr>
<td>69433</td>
<td>Tympanostomy (requiring insertion of ventilating tube), local or topical anesthesia</td>
</tr>
<tr>
<td>69436</td>
<td>Tympanostomy (requiring insertion of ventilating tube), general anesthesia</td>
</tr>
<tr>
<td>69676</td>
<td>Tympanic neurectomy</td>
</tr>
<tr>
<td>69801</td>
<td>Labyrinthotomy, with perfusion of vestibuloactive drug(s); transcanal [intra-tympmanic gentamicin]</td>
</tr>
<tr>
<td>69805</td>
<td>Endolympathic sac operation; without shunt</td>
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<tr>
<td>69806</td>
<td>with shunt</td>
</tr>
<tr>
<td>69905</td>
<td>Labyrinthectomy; transcranial [not covered with simultaneous cochlear implantation]</td>
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<tr>
<td>69910</td>
<td>with mastoidectomy [not covered with simultaneous cochlear implantation]</td>
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<tr>
<td>Code</td>
<td>Code Description</td>
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<td>--------</td>
<td>----------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>69915</td>
<td>Vestibular nerve section, translabyrinthine approach</td>
</tr>
<tr>
<td>69950</td>
<td>Vestibular nerve section, transcranial approach</td>
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<td><strong>CPT codes not covered for indications listed in the CPB:</strong></td>
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<tr>
<td>69930</td>
<td>Cochlear device implantation, with or without mastoidectomy</td>
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<tr>
<td>92601-</td>
<td>Diagnostic analysis of cochlear implant</td>
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<tr>
<td>92604</td>
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<td><strong>HCPCS codes not covered for indications listed in the CPB:</strong></td>
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<tr>
<td>L8614-</td>
<td>Cochlear implant and other components</td>
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<td>L8629</td>
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<td><strong>Dexamethasone thermos-sensitive gel, tenotomy of the stapedius and tensor tympani muscles, semi-circular canal plugging - no specific code:</strong></td>
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<td><strong>ICD-10 codes covered if selection criteria are met:</strong></td>
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<td>H81.01-</td>
<td>Meniere's disease [active]</td>
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<td>H81.09</td>
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<tr>
<td>H83.8x1-</td>
<td>Other specified diseases of inner ear [superior semicircular canal dehiscence]</td>
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<tr>
<td>H83.8x9</td>
<td></td>
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</tbody>
</table>

The above policy is based on the following references:

2. Albu S, Babighian G, Amadori M, Trabalzini F. Endolymphatic sac surgery versus tenotomy of the stapedius and tensor tympani muscles in the


11. Charpiot A, Rohmer D, Gentine A. Lateral semicircular canal plugging in severe Ménière's disease: A clinical


18. Furman JM, Barton JJS. Treatment of vertigo. UpToDate [online serial]. Waltham, MA: UpToDate; reviewed April, 2013.


40. Moskowitz HS, Dinces EA. Meniere disease. UpToDate [online serial]. Waltham, MA: UpToDate; reviewed March 2017. (Updated March 2019).


52. Sajjadi H, Paparella MM, Williams T. Endolymphatic sac enhancement surgery in elderly patients with


AETNA BETTER HEALTH® OF PENNSYLVANIA

Amendment to
Aetna Clinical Policy Bulletin Number: 0514 Meniere's Disease and Related Disorders: Surgery

There are no amendments for Medicaid.

www.aetnabetterhealth.com/pennsylvania revised 07/09/2020