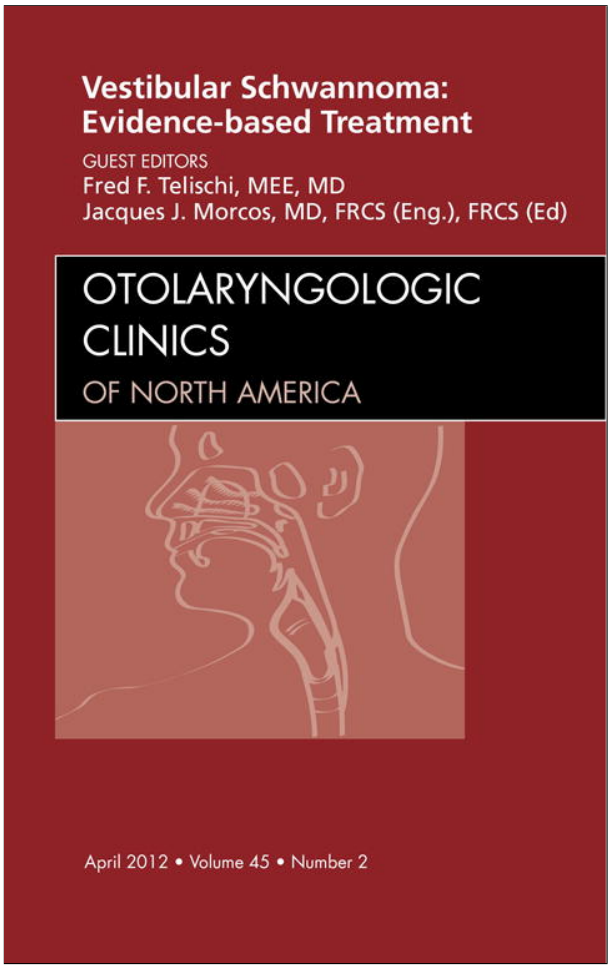

Acoustic Neuroma Neurophysiologic Correlates: Vestibular-Preoperative, Intraoperative, and Postoperative

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KEYWORDS

• Vestibular schwannoma • Acoustic neuroma • Vestibular loss • Dizziness
• Dysequilibrium • Vestibular testing • Electronystagmography • Rotational chair
• Vestibular evoked myogenic potentials • Posturography

Key Abbreviations: Acoustic Neuroma Neurophysiologic Correlates

AN	Acoustic neuroma
BPPV	Benign paroxysmal positional vertigo
CDP	Computerized dynamic posturography
CPA	Cerebellopontine angle
ENG	Electronystagmography
IAC	Internal auditory canal
IVN	Inferior vestibular nerve
SVN	Superior divisions of the vestibular nerve
SOT	Sensory organization test
VEMP	Vestibular evoked myogenic potentials
oVEMP	Ocular vestibular evoked myogenic potentials

Acoustic neuroma (AN) is a misnomer for the benign tumor discussed in this article, because it does not originate from the acoustic nerve and is not a neuroma. AN is a benign tumor arising from the Schwann cells of the vestibular nerve, more appropriately called a vestibular schwannoma. Consequently, the vestibular nerve is always involved with AN and there is virtually always some vestibular system pathology secondary to AN development. Although by far the most common presenting complaints in AN patients are hearing loss and tinnitus, this disorder is primarily one of the vestibular system, specifically the vestibular nerve. It is important to keep this in mind even though most patients do not complain of vestibular symptoms before intervention. However, vestibular complaints are common among patients after intervention.[1]

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Because intervention for AN by nature results in loss of vestibular function beyond the damage already inflicted by the tumor, it behooves the surgeon to have some knowledge of the preoperative status of the vestibular system because this can, on occasion, affect the treatment mode. Intraoperatively, the origin of the AN from either the inferior (IVN) or superior divisions of the vestibular nerve (SVN) can have some prognostic importance regarding hearing preservation and possible residual vestibular function. Postoperatively, in terms of morbidity from AN treatment, balance dysfunction is one of the most common consequences.[1]

As already mentioned, the AN arises from the Schwann cells surrounding the vestibular division of the eighth cranial nerve. Specifically, it has been proposed that ANs arise at the transition zone of the central myelin and peripheral myelin sheaths, known as the Obersteiner-Redlich zone. This location is also in the same vicinity as the vestibular (Scarpa) ganglion. It should be noted that although this region appears to be the most common site of AN origin, the site of this zone is variable and there are many examples of AN arising distal to this region as well. In any event, the vast majority of ANs appear to arise from within the medial portion of the internal auditory canal (IAC), with only a small minority originating in the lateral end of the IAC. Even fewer appear to originate at the porus acusticus or in the cerebellopontine angle (CPA) cistern itself. There is a small proportion that even arises within the vestibular end organ. AN can arise from either the IVN or SVN, giving somewhat differing functional vestibular pathology.

AN growth results in compression of the surrounding structures. Presumably the resulting compression of the associated vestibular nerve, and/or its blood supply, results in primary neural dysfunction; as already mentioned, it is rare for direct end-organ involvement. However, the degree of compression, or whether compression of the associated nerve occurs at all, is dependent on the size of the tumor and its location. Medially based AN, especially if there is minimal or no IAC component, will tend to have less impact on function, whereas laterally based tumors will tend to have more profound effects on vestibular function. Such effects are likely attributable to the limited degree of expansion for IAC tumors because of their bony confines, compared with the relatively larger area for expansion within the CPA for a medially based AN. This difference explains the incongruity of a large medial tumor with no significant vestibular loss in contrast to a small IAC tumor that may demonstrate near total loss of vestibular function.

Of course, at its outset a small tumor may cause no neural compression and cause no dysfunction, and the patient will be asymptomatic. As the tumor grows and causes surrounding compression, dysfunction occurs as the associated structures are compressed. The 4 anatomic stages of AN growth are:

1. Intracanalicular
2. Cisternal
3. Brainstem compressive
4. Hydrocephalic

In direct opposition to what one may expect, the worst vestibular symptoms are typically seen at the intracanalicular stage. During this stage, as the vestibular nerve is being progressively destroyed the patients may be asymptomatic or may have episodic vertigo suggestive of a peripheral vestibular disorder. As further growth occurs, the vertigo diminishes and the vestibular symptoms usually subside. Later, as the cisternal phase progresses and the brainstem compressive phase begins, onset of dysequilibrium occurs and signs of central vestibular dysfunction arise. The dysequilibrium worsens as the hydrocephalic stage of growth takes place.

Although the primary means of vestibular dysfunction caused by AN appears to be neural compression, distinct end-organ effects have been demonstrated. Specifically, histopathology of the inner ear in some AN patients has demonstrated endolymphatic hydrops, and eosinophilic proteinaceous precipitate in the perilymphatic and endolymphatic spaces. In addition, there is degeneration of other inner ear structures, including hair cells, the stria vascularis, and the spiral ligament.[2] This process may possibly explain how a minority of patients with small AN present with episodic vertigo.

PREOPERATIVE VESTIBULAR TESTING

Vestibular testing has been used in the past as a screening procedure for AN. Although vestibular testing will frequently be abnormal in AN patients, there is a lack of sensitivity or specificity for the purpose of diagnostic screening tests. For vestibular testing to demonstrate an abnormality in an AN case, the tumor must be of adequate size to compromise either the blood supply (to the vestibular nerve or end organ) or the nerve itself to a sufficient degree so as to reduce the vestibular function on the side affected. In addition, the degree of vestibular loss must be sufficient to appear significant on vestibular testing; it must be borne in mind that the state of vestibular testing is such that minor differences in vestibular test responses are often considered insignificant. Consequently, small tumors may not have attained adequate size to result in vestibular loss, thus resulting in false-negative outcomes for vestibular testing as a screening device. By contrast, although vestibular test abnormalities are often the rule among AN patients, abnormalities on vestibular studies are a common finding among a host of other vestibular disorders, resulting in a relatively low degree of specificity in a large screening population. However, this does not mean that vestibular testing in AN patients is without merit.

The values of preoperative vestibular testing are mainly to help distinguish the nerve of origin for the AN, to establish a baseline function in not just the tumor ear but also the contralateral ear, and as a means to help prognosticate about functional postoperative vestibular status. The vestibular-nerve origin has more than just academic significance. SVN tumors have a better prognosis for hearing preservation than IVN tumors. Differentiation between IVN-based and SVN-based tumors can help determine whether a surgical approach to hearing preservation may or may not be warranted. Establishing normal vestibular function in the non-AN ear is important for postoperative rehabilitation. Severely diminished or absent vestibular function in the non-AN ear may warrant a more conservative approach to maintaining vestibular function in the AN ear.

Electronystagmography

Electronystagmography (ENG) has been the most studied vestibular test battery among AN patients, and its most useful subtest is the bithermal caloric test. On caloric testing, the vast majority of AN patients will demonstrate reduced or absent response on the affected side.[3] Because the caloric test is a test of the horizontal semicircular canal function and the SVN, tumors arising from the SVN have a higher rate of reduced caloric response than tumors arising from the IVN. Of course, once tumors of the IVN attain sufficient size they could compromise the adjacent SVN. In addition, the larger the tumor is the more likely that the caloric response will be diminished. As alluded to earlier, this is not a good screening test for AN, but from a practical standpoint a large AN without a corresponding hypoactive caloric response would lead one to suspect that the pathology may be something else, such as a meningioma.

As one would expect, spontaneous nystagmus frequently accompanies AN. Most typically, it is a nystagmus of the paretic type from the loss of unilateral vestibular function. However, other types of nystagmus can also be seen. As the AN grows and causes central compression, signs of brainstem and cerebellar dysfunction may manifest with Bruns nystagmus.[4] Bruns nystagmus is a combination of a gaze-evoked nystagmus from cerebellar compression and a paretic nystagmus from unilateral deafferentation. The resulting nystagmus is an asymmetric one with slow, large-amplitude beats of nystagmus looking to the side of the AN and fast, small-amplitude nystagmus when gaze is directed away from the side of the AN. As the tumor continues to grow, causing bilateral cerebellar compression, a more pure gaze-evoked nystagmus may predominate. Alternatively, with brainstem compression and increased intracranial pressure, vertical down-beat or up-beat nystagmus may be present.

Other abnormalities may be seen on ENG, including positional nystagmus, which can be seen with tumors of all sizes, and central findings that are virtually exclusive to the larger tumors presenting with compressive features. Among the central findings seen with larger tumors are impaired fixation suppression, optokinetic and smooth-pursuit abnormalities, and the aforementioned gaze-evoked nystagmus and vertical nystagmus.

Rotational Studies

Similar to ENG, rotational studies lack both specificity and sensitivity as a means for making the diagnosis of AN[5]; however, rotational chair studies are frequently abnormal, particularly when the tumor has grown in sufficient size to compromise the SVN.[6] Tumors arising from the IVN that are small and do not compromise the SVN should have no identifiable abnormality on rotational chair testing, because this is a test of the horizontal semicircular canal and the corresponding SVN. There is some evidence to suggest that high-frequency, vestibular autorotation testing in the vertical plane may identify vestibular abnormalities in this subgroup of AN patients.[7] When abnormalities are identified, the findings typically reflect evidence of a unilateral vestibular deficit with varying evidence of central compensation. In general, gain reduction is seen, especially if there is a concomitant caloric hypofunction. However, in some of the large tumors an increase in gain may be seen, representing central vestibular dysfunction, which is the inability to suppress vestibular eye movements. Asymmetry tending to the side of the AN is most characteristically seen, but eye asymmetry may occasionally go to the non-AN side.

Computerized Dynamic Posturography

Within the standard vestibular test battery, computerized dynamic posturography (CDP) probably is the least sensitive to identification of AN. However, it is not unusual to see reduced scores among AN patients on CDP. There seems to be a general correlation between the size of the AN and outcomes on sensory organization test condition 5 (SOT 5), with larger tumors resulting in poorer performance.[8] It has also been suggested that CDP may be a means to help identify AN of IVN origin as opposed to SVN origin.[9] CDP is a measure of the vestibulospinal reflex arc, but is more sensitive to end-organ components innervated by the IVN and is relatively less sensitive to the SVN-innervated components. Consequently, one would expect to see more frequent CDP abnormalities (SOT 5) among patients with IVN-based tumors than among those with SVN-based tumors. However, in practice this is not so straightforward and has not yet been definitively shown in study. This finding may possibly be explained by the slow growth of AN and concomitant central compensation/adaptation, resulting

in somewhat better performance for some patients with IVN tumors and the eventual compromise of the IVN by tumors arising from the SVN.

Vestibular Evoked Myogenic Potentials

Vestibular evoked myogenic potentials (VEMP) is a testing technique that has recently made the transition from the research laboratory to the clinical laboratory, due to its popularity in the evaluation of patients with superior semicircular canal dehiscence. VEMP is a means of measuring otolithic function. Delivery of sound stimuli to the ear results in stimulation of the otolithic organ, which initiates reflex muscular contraction that can be measured and quantified for sensitivity, latency, and amplitude. The 2 most common methods are the cVEMP (cervical) with measurement of the sternocleidomastoid muscle and the oVEMP (ocular), which measures extraocular muscle contraction.

The generally accepted view at present is that the saccule may be more amenable to air-conducted sound stimuli, whereas the utricle is more sensitive to bone-conducted sound stimuli.[10] Because of this differential sensitivity to stimuli, the otolithic organs can be assessed separately for responsiveness. Consequently, the use of bone-conducted VEMP and air-conducted VEMP may be useful for distinguishing between IVN and SVN tumor origins. Air-conducted VEMP would be expected to more frequently demonstrate a deficit in IVN-based tumors, whereas bone-conducted VEMP would be more likely to demonstrate abnormalities in SVN-based tumors. Of course, the issue found with other distinguishing vestibular tests arises here as well. Once a tumor is of sufficient size, whether it arises from the SVN or the IVN, it has a tendency to compromise both divisions of the vestibular nerve. Therefore, the differential utility of this test is most useful with the smaller tumors.

INTRAOPERATIVE

There is no commonly used means or rationale for intraoperative vestibular nerve monitoring. A few studies, comparing total vestibular nerve resection with preservation of one branch, have demonstrated no significant difference in vestibular/balance outcomes. However, the distinction of AN arising from the IVN or the SVN has importance with regard to hearing preservation. The IVN lies in the inferior compartment of the IAC, adjacent to the cochlear nerve. Consequently, IVN tumors have a higher probability than SVN tumors of cochlear nerve invasion. Similarly, because of the IVN location, surgical dissection of an IVN tumor would more likely disrupt cochlear nerve blood supply to SVN tumors than would dissection of an SVN tumor. Because of these anatomic differences, AN patients who present for possible hearing-preservation surgery generally have a more favorable prognosis when vestibular studies suggest SVN origin for the tumor (absent/reduced caloric response, reduced gain on rotational studies, present air-conducted VEMP, and normal SOT 5 on CDP) than when vestibular studies suggest an IVN origin for the AN (normal caloric response, normal gain on rotational studies, present bone-conducted VEMP, and abnormal SOT 5 on CDP).

POSTOPERATIVE

Vestibular function is virtually always altered postoperatively. The surgical disruption of the vestibular nerve changes the patient's vestibular situation dramatically. Preoperatively, ANs grow slowly and cause a very slow decline in vestibular function. Consequently, central compensation likely occurs as the tumor grows, and most patients do not therefore encounter any significant vestibular symptoms. Postoperatively, the loss of residual vestibular function is sudden. In the case of a large AN that has destroyed

most or all of vestibular function preoperatively, there is little or no change in vestibular function after surgical resection. In the case of the small AN that has had little decline in vestibular function preoperatively, surgical resection results in a dramatic change in vestibular function. This process explains the paradoxic phenomenon of patients with a large AN having generally less vertigo immediately after surgery than those with a small AN.

The postoperative vestibular affliction is a total unilateral loss of function in most cases. In the vast majority of cases, central compensation proceeds to the point of essentially normal daily function.[11] Of course, when a patient has only one functioning vestibular end organ there will always be some situations that provoke symptoms of dysequilibrium. These situations are typically novel ones whereby central compensation has not occurred, and with high-velocity movement toward the lesioned ear. In addition, patients who have attained complete central compensation can have recurrent symptoms of dysequilibrium when central decompensation occurs. This decompensation is often brought about during times of extreme fatigue, severe physical stress, and severe emotional stress.

For a minority of patients, central compensation does not occur and postoperative dysequilibrium will persist. The cause for this uncompensated vestibulopathy usually falls into 1 of 3 categories:

1. Central vestibular abnormality
2. Incomplete deafferentation with fluctuating vestibular function in the surgical ear
3. Unrecognized fluctuating vestibulopathy in the nonsurgical ear

Of course, it is certainly possible for more than one of these conditions to exist as well.

Central Vestibular Abnormalities

Central vestibular abnormalities are typically seen with the large AN that has progressed to the point of brainstem or cerebellar compression. Postoperatively, these patients will demonstrate central vestibular test abnormalities in addition to unilateral vestibular deafferentation. The existence of central vestibular abnormalities is the differentiating test finding for this group of patients. Vertical nystagmus, gaze nystagmus, impaired fixation suppression, and optokinetic and smooth-pursuit abnormalities can be seen on ENG. Rotational studies typically demonstrate reduced gain (peripheral loss finding), but may demonstrate an increased gain (central finding) in the rare case where the SVN is preserved. CDP does not help in differentiating these patients from the others, but does help in identifying multisystem balance dysfunction, which can complicate the postoperative AN dysequilibrium picture.

Incomplete Deafferentation

The scenario of incomplete deafferentation with fluctuating vestibular function in the surgical ear requires preservation of one branch of the vestibular nerve and preservation of the vestibular end organ. This situation is seen in hearing-preservation cases using either a middle fossa or retrosigmoid approach for hearing preservation in the typically smaller tumors. There is usually an absence of central vestibular test findings for these patients. The ENG and rotational studies show the picture of unilateral loss of vestibular function with poor compensation, exhibiting paretic spontaneous nystagmus, and unilateral absent caloric response on ENG, with reduced gain and asymmetry on rotational testing. If there has been preservation of the IVN, posterior canal benign paroxysmal positional vertigo (BPPV) may be present, which may cause the

continued fluctuation of vestibular stimulation. Similarly, an atypical BPPV (from the horizontal or anterior semicircular canal) may be present with preservation of the SVN. In either of the aforementioned situations, the Dix-Hallpike test likely would be abnormal.

Unrecognized Fluctuating Vestibulopathy

The third scenario, of the unrecognized fluctuating vestibulopathy in the nonsurgical ear, would typically be easiest to recognize when there has been total deafferentation of the AN ear from surgical extirpation in a small or medium AN that had no compression of the brainstem or cerebellum. In this case, as in most cases of dysequilibrium, BPPV must be suspect because of its ubiquity. For absent concomitant BPPV, the test findings one would expect include unilateral absent caloric response in the surgical ear and a hypoactive caloric response in the nonsurgical ear. Spontaneous nystagmus would be present, and rotational studies would demonstrate reduced gain suggestive of bilateral loss of vestibular function.

SUMMARY

AN and vestibular schwannoma are benign tumors that grow from the Schwann cells surrounding the vestibular division of the eighth cranial nerve. Because of their origin, there is virtually always a concomitant vestibular lesion. Depending on the size and location of the AN, there will be a loss of peripheral vestibular function and/or central vestibular dysfunction. Although vestibular testing is frequently abnormal in AN patients, it lacks the sensitivity and specificity for use as a diagnostic screening device. However, vestibular testing is very useful in defining the vestibular abnormality associated with the AN preoperatively and postoperatively, may be helpful in planning surgery for hearing preservation, and is helpful in the diagnosis of postoperative dysequilibrium.

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