Neurologic evaluation of the ear

Laurie B. Cook, DVM

Animal Neurological Clinic, 352 Warren Avenue, Portland, ME 04103, USA

The ear is a highly complex sensory organ responsible for the sense of hearing and for vestibular control of posture and eye movements. Diseases of the inner ear are common in dogs and cats and are often associated with neurologic dysfunction, including peripheral vestibular syndrome, deafness, facial paralysis, and Horner’s syndrome. In some cases, differentiating neurologic manifestations of inner ear disease from central disease can be difficult. In addition, extension of inner ear disease into the central nervous system (CNS) can lead to signs of brain stem dysfunction. Sorting through these signs early in the course of disease can facilitate accurate diagnosis, prognosis, and treatment to prevent progression of neurologic signs.

Neuroanatomy and physiology

Receptors for the vestibular system lie within the inner ear and connect with the central components of the vestibular system to control posture and eye movements. The boney labyrinth lies within the petrous temporal bone of the inner ear and includes the vestibule, three semicircular canals, and cochlea. Within the boney labyrinth lies the membranous labyrinth, including three semicircular ducts: the utricle and saccule, which contain the receptors for the vestibular system, and the cochlear duct. These structures are filled with a fluid called endolymph [1–4].

These three semicircular ducts are oriented at 90° angles to each other, representing all three planes in space, and they are sensitive to rotational acceleration of the head. At one end of each semicircular duct is a dilation called the ampulla. Within the ampulla lies the crista ampullaris. The crista ampullaris is the receptor organ within the semicircular ducts and is lined with specialized columnar epithelial cells called hair cells. These hair cells synapse with the vestibular portion of the vestibulocochlear nerve at their basilar surface and have a constant level of neuronal discharge at rest. Cilia project...
from the hair cells into a gelatinous mass composed of a protein-polysaccharide material called the cupula. When the head begins to rotate, the inertia of the fluid within the ducts allows it to remain stationary while the semicircular ducts move with the head. The flow of endolymph deflects the cupula and cilia. Sodium channels in the cilia open, allowing flow of ions into the hair cells, which leads to their depolarization and propagation of nerve impulses to the vestibular nerve. Movement in one plane stimulates the impulses in one duct and inhibits impulses in the same duct on the opposite side, allowing bilateral sensation of head movements. When acceleration ceases and velocity is constant, the ducts are no longer stimulated, allowing for appropriate sensation of movement of the head [1–6].

The utricle and saccule are responsible for the sensation of the static position of the head and linear acceleration. The saccule is oriented in a sagittal plane (vertical plane), and the utricle is oriented in a dorsal plane (horizontal plane). On the floor of these structures is a thickening called the macula. Like the crista ampullaris, it is also lined by hair cells that give rise to cilia. On their basilar surface, these cilia synapse with the vestibular nerve. The cilia extend into a gelatinous structure called the otolithic membrane. Within this membrane lie many calcium carbonate crystals called otoliths or statoconia. Gravitational forces continually affect the position of these otoliths, relaying the sense of static equilibrium. When the body accelerates, the otoliths, which have greater inertia than the surrounding fluid, fall backward, deflecting the hair cells that stimulate the vestibular nerve [2–6].

Vestibular impulses are relayed through the vestibular nuclei that lie within the brain stem lateral to the fourth ventricle and communicate with many tracts through the brain and spinal cord. The vestibulospinal tract descends through the spinal cord to facilitate ipsilateral extensor muscle tone (antigravity muscles) and inhibit contralateral extensor tone. Through this pathway, head movements can be coordinated with limb and trunk activity to maintain normal posture. For example, when the head is at rest, vestibular impulses to both sides are equal. If the head tilts to the left, receptors on the left side are stimulated, whereas those on the right are inhibited. The left vestibulospinal tracts support extensor tone on the left side, preventing the animal from falling to the left and returning its posture to normal. There are also connections between the vestibular nuclei and the cerebellum that help to maintain equilibrium during movement [1–3,6,7].

The vestibular system is crucial to the coordination of eye movements. There are tracts that pass through the medial longitudinal fasciculus (MLF) to the nuclei of cranial nerves III, IV, and VI supplying the extraocular muscles. These tracts allow eye movements to be coordinated with changes in position of the head. This way, the visual system can compensate for movements while keeping images stable in the retina.

The vestibular nuclei project some axons to areas of the reticular formation, including the vomiting center and reticular activating system, the
brain stem region that maintains the awake state by activating the cerebral cortex [1,2,6,7]. Finally, there are poorly defined projections to the temporal lobe of the cerebral cortex through the thalamus for conscious perception of changes in posture [1,4,7].

**Auditory system**

The external ear canal serves to direct sound vibrations toward the middle ear. The middle ear is air filled and consists of the tympanic membrane and bony ossicles (malleus, incus, and stapes) along with the ligaments and muscles that coordinate their function. The sound vibrations from the external ear canal are transmitted through the tympanic membrane through the ossicles, which articulate to amplify sound and transmit vibrations to the fluid-filled inner ear. The foot plate of the stapes connects through the oval window with the cochlear ducts. The cochlea is the sensory organ for hearing and is encased within the bony labyrinth of the petrous temporal bone. It includes the scala vestibuli and scala tympani, which are canals filled with perilymph, a fluid that communicates directly with cerebrospinal fluid of the subarachnoid space. The cochlear duct is a coiled portion of the membranous labyrinth that lies within the cochlea and is filled with endolymph. The scala media is part of the membranous labyrinth and is filled with endolymph. The basilar membrane separates the scala vestibule and scala media. The organ of Corti, the sensory receptor for hearing, lies on the basilar membrane. On the surface of the organ of Corti lie inner and outer hair cells that synapse with the spiral ganglion for transmission to the cochlear nerve. These hair cells project stereocilia into the gelatinous tectorial membrane of the scala media. There is a vascular bundle on the outer wall of the scala media called the stria vascularis, which is responsible for the production of endolymph. Endolymph contains a high concentration of potassium and a low concentration of sodium, which is exactly the opposite of perilymph. This difference is responsible for the endocochlear potential, which is an electrical potential of approximately +80 mV that exists between endolymph and perilymph and sensitizes the receptor. As sound vibrations pass through the middle ear and the stapes pushes the oval window inward, the perilymph within the scala vestibuli is compressed and the basilar membrane is deflected. Deflection of the basilar membrane causes the stereocilia embedded in the tectorial membrane to bend. Bending of the stereocilia depolarizes the hair cells, which excites the nerve fibers of the spiral ganglion that synapse at their bases. From the spiral ganglion, impulses are propagated through the cochlear nerve into the brain [2,6,7]. Impulses from the cochlear nerve are transmitted to the cochlear nuclei within the medulla oblongata. Tracts from the cochlear nuclei cross to the opposite side and pass through the lateral lemniscus of the brain stem to the medial geniculate nucleus of the thalamus. From the thalamus, there are relays to the cerebral cortex for conscious recognition of sound [1,7].
are tracts that communicate with the reticular formation of the brain stem and are responsible for arousal in response to sound stimuli. There are also tracts connecting the cochlear nuclei with descending motor tracts to the neck and limbs that coordinate reflex movements (ie, rapid turning of the head in response to sudden noises) [7].

**Neurologic dysfunction of inner ear disease**

**Peripheral vestibular disease**

Diseases of the inner ear can cause injury to the vestibular receptors of the membranous labyrinth. Peripheral vestibular syndrome is characterized by ataxia and loss of balance, with preservation of strength. Despite the ataxic gait and tendency to fall, strength and proprioception are preserved with peripheral lesions and can be assessed with placing, hopping, and hemiwalking responses. It is important to provide a surface with good traction when evaluating postural responses. Severely affected animals may be difficult to assess because of falling, rolling, and failure of the righting response [1,3,4,6,8–10].

Animals with peripheral vestibular disease are typically alert with normal mentation but may be disoriented. Nausea or vomiting may be present and can be severe in some cases because of the connection of the vestibular system to the vomiting center [1–4,6–9].

A head tilt toward the affected side is often present with peripheral vestibular disease. Loss of the normal vestibular tone on the affected side results in a relative increased stimulation on the normal side. Facilitation of extensor tone to the neck and limbs is lost on the affected side and increased on the opposite side. This imbalance causes the head to fall or “tilt” toward the affected side. Likewise the loss of extensor tone on the affected side and increased extensor tone on the opposite side may cause the animal to fall or roll toward the lesion. On flexion of the limbs, affected animals may have decreased extensor tone noted on the side of the lesion and increased extensor tone opposite the side of the lesion [1,3,4,6,8–10].

Nystagmus is an involuntary rhythmic movement of the eyes that can occur in any direction and is typically characterized by a slow phase and a fast phase. By convention, nystagmus is described by the direction of the fast phase (ie, a left nystagmus means the fast phase of the nystagmus is to the left). Normal or physiologic nystagmus is the movement that the eyes make as they track in the direction that the head turns, allowing the animal to maintain visual fixation on a stationary point. This is called the vestibulo-ocular reflex. As the head turns to one side, hair cells of the vestibular receptors on that side are stimulated, causing the eyes move slowly away from the direction of the head turn. When elicited, this is referred to as the “doll’s eye” maneuver. This slow phase is followed by a quick movement in the direction of the head turn. The fast phase occurs when tension in the
extraocular muscles reaches its threshold, causing a corrective movement of
the eye back in the direction of the head turn. The fast phase is under
control of higher brain stem centers. Delayed physiologic nystagmus may be
seen with peripheral vestibular dysfunction. Caloric nystagmus is a type
of physiologic nystagmus that can be induced by irrigating the ear canal with
ice-cold water (0°C) or warm water (44°C) for 3 to 5 minutes and watching
for induced nystagmus. The flow of water causes flow of endolymph within
the ducts. Absence of response or asymmetry between sides may indicate
vestibular dysfunction. This test is often unreliable and is not used widely in
clinical settings [1,3,6].

The presence of spontaneous nystagmus at rest indicates vestibular
dysfunction. Animals with peripheral vestibular disease show horizontal or
rotary nystagmus that always occurs away from the side of the lesion. Damage
to the semicircular canals impedes the resting baseline activity on the affected
side. The opposite side continues to emit baseline activity that is interpreted as
head rotation to the normal side. Therefore, the nystagmus occurs with the
fast phase away from the damaged ear and with the slow phase toward the
damaged ear. Some animals with chronic peripheral vestibular disease, such as
dogs with chronic otitis media/interna, may show positional nystagmus when
the head is extended or the animal is placed in lateral or dorsal recumbency.
Early in the course of disease, these animals may show spontaneous
nystagmus that resolves within a few days to weeks. The damaged vestibular
system can compensate over time with central preprogramming of eye
movements and postural responses as well as reliance on visual and other
sensory input that replaces lost vestibular input. In these cases, no nystagmus
is present at rest, but it can be induced by changing the animal’s position,
which decompensates the altered vestibular system [3,6,7,9,10].

Animals with peripheral vestibular disease may also show a positional or
resting ventral or ventrolateral strabismus on the side of the lesion. In
a normal animal, when the head and neck are extended, the eye should
remain centered within the palpebral fissure. This response fails when there
are disturbances of the vestibular receptors in the membranous labyrinth
[1,3,4,6,8–10].

Animals with bilateral otic disease may present with bilateral peripheral
vestibular disease. These animals present with symmetric ataxia and loss of
balance, with preserved strength and postural reactions. There is no head
tilt, but the animal often crouches close to the ground with side-to-side head
movements [1,6]. Spontaneous nystagmus is not present, and normal
physiologic nystagmus cannot be elicited, because input from the vestibular
receptors is absent [1,3,4,6].

Deafness

Otic disease may also cause hearing loss through conductive or
sensorineural impairment. Conductive deafness occurs when there is
impedance to transmission of sound waves to the inner ear and CNS. Sensorineural deafness results from congenital or acquired abnormalities in the inner ear structures, cochlear nerve, or auditory nervous system [8,11,12]. Acquired conductive hearing loss may be caused by stenosis or obstruction of the ear canal from chronic otitis externa, foreign bodies, ceruminoliths, or aural neoplasms [8,13]. One study showed that accumulation of debris within the ear canal significantly affected hearing ability [13]. Dogs in this study had significantly lower hearing thresholds detected by brain stem auditory evoked response (BAER) testing after removing the obstructive debris by ear cleaning [13]. Other mechanisms of conductive hearing loss include rupture of the tympanic membrane, damage to the ossicular chain, or fluid accumulation within the middle ear. Damage to the tympanic membrane from trauma or otitis has been shown to cause conductive hearing impairment as well. Dogs with perforated tympanic membranes had significantly elevated hearing thresholds assessed by BAER testing [8,14]. Changes in hearing thresholds returned to baseline by 3 to 4 weeks after tympanic membrane perforation [8,14]. Acquired sensorineural deafness has also been associated with ototoxicity from drugs or chemicals that damage hair cells, the stria vascularis, the organ of Corti, or cochlear neurons [8,15,16].

Presbycusis is hearing impairment that accompanies aging in dogs and cats. Affected dogs have loss of spiral ganglion cells, atrophy of the organ of Corti, atrophy of the stria vascularis, thickening of the basilar membrane, lipofuscin accumulation within cochlear hair cells, and nerve cell loss and gliosis within the cochlear nuclei. These degenerative changes are hypothesized to be the result of aging changes as well as exposure to ototoxic agents [17].

Congenital sensorineural deafness is common in many breeds of dogs and cats with a predilection for white coat colors, and there is a strong association of deafness with blue irises. The most commonly affected canine breeds include Dalmatians, English Setters, Australian Shepherds, Border Collies, and Shetland Sheepdogs, although there are many others [8,11,12]. The deafness results from degeneration of the stria vascularis apparently as a result of an absence of melanocytes. Subsequently, there is collapse of Reissner’s membrane and the cochlear duct and degeneration of the hair cells of the organ of Corti [8,11].

**Horner’s syndrome**

Horner’s syndrome may be seen in animals with diseases of the middle ear [1,3,18,19] or after surgery in patients that undergo total ear canal ablation or other surgeries of the ear canal [20–22]. Sympathetic innervation to the eye arises from the C8 to T7 spinal cord segments. Preganglionic sympathetic nerve fibers pass through the thorax and neck with the vagosympathetic trunk and synapse in the cranial cervical ganglion deep to the tympanic bulla. Postganglionic fibers pass with the internal carotid artery into the middle ear cavity via the tympano-occipital fissure and then run along the internal
carotid foramen to join the ophthalmic nerve at the trigeminal ganglion. Sympathetic fibers go to the smooth muscle that lifts the upper eyelid, bands of smooth muscle that tonically pull the eye rostrally, the retractor of the third eyelid, and the dilator of the pupil [18,23]. Damage to postganglionic fibers passing through the middle ear often produces Horner’s syndrome, which is characterized by miosis, enophthalmos, ptosis (drooping of the upper eyelid), and protrusion of the third eyelid [1,3,18,19,24].

Sympathetic hyperirritability has been reported in early otitis media. Irritation of the postganglionic sympathetic fibers in the middle ear can result in dilation of the pupil [3,7,18].

**Facial paralysis**

Facial paralysis commonly accompanies middle and inner ear disease. The facial nerve exits the cranial vault through the internal acoustic meatus accompanied by the vestibular and cochlear nerves. From the meatus, the facial nerve runs through the facial canal of the petrous temporal bone and through the middle ear on its way out of the skull. The motor portion of the facial nerve supplies the superficial muscles of the head and face. The facial nerve also supplies preganglionic parasympathetic fibers to the lacrimal gland and salivary glands [23]. Diseases of the inner ear may damage the facial nerve, resulting in ipsilateral facial paralysis. Affected patients may have an obvious ear, lip, and eye droop on the affected side. Owners may report that the patient drools excessively or drops food out of the affected side of the mouth. Palpebral, menace, and corneal responses are reduced or absent because of an inability to close the eyelid [1,3,6,25]. Neurogenic keratoconjunctivitis sicca may accompany facial nerve paralysis associated with middle ear disease. Damage to the parasympathetic supply to the lacrimal gland interferes with normal lacrimation, resulting in xeromycteria or dry eye [1,6]. Affected animals may have recurrent corneal ulceration, mucous discharge from the eye, and chronic keratitis.

**Hemifacial spasm**

Hemifacial spasm may be seen early in the course of middle ear diseases. Inflammation of the facial nerve as it runs through the facial canal of the petrous temporal bone may cause the facial muscles on the affected side to become hypertonic, causing the face and nose to be pulled caudally. There may also be a narrowed palpebral fissure caused by partial closure of the eyelids, elevation of the ear, and wrinkling of the face. In some cases, hemifacial spasm may precede facial paralysis [26,27].

**Specific conditions**

The neurologic syndromes described here may be seen with many diseases of the inner ear. They are most commonly reported with otitis media/interna
in dogs and cats [5,10,28,29]. Other causes include traumatic injury to the inner ear [3,4,6,10], nasopharyngeal polyps [30,31], aural neoplasms [32–34], cholesteatomas [33], otolithiasis [35], lymphocytic labyrinthitis [36], and others. Facial paralysis or Horner’s syndrome often follows total ear canal ablation and other surgeries of the ear canal [20–22]. Damage to the peripheral nervous structures may be transient, and clinical signs may resolve if the underlying condition is treated early, particularly in the case of otitis media/interna or after surgeries of the ear canal [5,10,20–22,28]. Chronic conditions or trauma to the inner ear structures may result in permanent damage.

Central vestibular disease

Animals with disease of the medulla oblongata or cerebellum may have signs of central vestibular disease. Differentiating these signs from those of peripheral vestibular disease associated with inner ear disease is important for making an accurate diagnosis and prognosis (Table 1). Diseases that cause central vestibular syndrome generally carry a worse prognosis than many of the diseases that cause peripheral vestibular disease. In addition, some conditions that cause peripheral vestibular syndrome may extend into the CNS (ie, otitis media/interna), and early detection can expedite treatment, which may improve outcomes. The presence of proprioceptive deficits or weakness (paresis) is the most reliable sign used to differentiate central vestibular disease from peripheral vestibular disease [1,3]. Descending motor pathways and ascending proprioceptive pathways run through

<p>| Table 1 | Neurologic examination findings that differentiate peripheral vestibular syndrome from central vestibular disease |</p>
<table>
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<th>Clinical signs</th>
<th>Peripheral vestibular disease</th>
<th>Central vestibular disease</th>
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<td>Head tilt</td>
<td>Toward the lesion</td>
<td>Toward the lesion</td>
</tr>
<tr>
<td>Spontaneous nystagmus</td>
<td>Horizontal or rotary Fast phase away from the lesion</td>
<td>Horizontal, rotary, vertical, or positional Fast phase toward or away from the lesion</td>
</tr>
<tr>
<td>Paresis/proprioceptive deficits</td>
<td>None</td>
<td>Common ipsilateral to lesion</td>
</tr>
<tr>
<td>Mentation</td>
<td>Normal</td>
<td>Commonly stuporous, obtunded, or comatose</td>
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<tr>
<td>Cranial nerve deficits</td>
<td>Cranial nerve VII ipsilateral</td>
<td>Cranial nerves V, VII, IX, X, XII ipsilateral</td>
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<td>Horner’s syndrome</td>
<td>Common ipsilateral to lesion</td>
<td>Uncommon</td>
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the brain stem and are often damaged when the vestibular nuclei are involved. Hemiparesis or altered proprioseception is often present on the side of the lesion. Most commonly, a head tilt toward the side of the lesion is present. Animals with central vestibular disease may show spontaneous nystagmus in a horizontal, rotary, or vertical direction, or they may have positional nystagmus. Typically, the presence of vertical nystagmus or change in the direction of spontaneous nystagmus when the animal is turned in lateral or dorsal recumbency indicates central disease. Absence of oculocephalic reflexes usually suggests damage to the vestibular nuclei, which can occur with brain stem injury and typically carries a poor prognosis. Diseases causing central vestibular dysfunction may be accompanied by alteration of mentation. The reticular activating system of the brain stem facilitates the alert awake state in animals. Damage to this area may cause the animal to be disoriented, stuporous, or comatose [1,3,6].

Central vestibular syndrome may be accompanied by other cranial nerve dysfunction as well. Cranial nerves V, VI, VII, IX, X, and XII may be affected. Clinical signs suggesting involvement of these cranial nerves include ipsilateral facial hypalgesia, atrophy of masticatory muscles, reduced jaw tone, facial paralysis, tongue weakness, and loss of the swallow or gag reflex. Horner’s syndrome is uncommonly associated with central vestibular syndrome [1,3,6].

Paradoxic vestibular syndrome may be seen with central vestibular disease. Damage to the cerebellar medulla and peduncles may produce a head tilt directed away from the lesion. In these cases, paresis and proprioceptive deficits are still present on the affected side and are the most reliable signs for localizing the side of the lesion [1,3,4].

Severe brain stem involvement can interfere with respiratory and cardiovascular control centers within the pons and medulla oblongata. The pneumotaxic center within the pons regulates respirations per minute. In the medulla, the apneustic center stimulates respirations. There are a number of abnormal respiratory patterns that may be seen with brain stem disease. Cheyne Stokes respirations consist of waxing and waning depth of respiration. Caudal brain stem lesions may cause irregular or ataxic respirations [2,6]. Normal vasomotor tone is controlled at the level of the reticular formation in the pons and medulla oblongata. Ventricular premature contractions or other arrhythmias may be seen with brain stem injury. Bradycardia may be seen as a consequence of the Cushing response secondary to increased intracranial pressure [1,2,6].

Signs of central vestibular syndrome suggest damage to the brain stem and are not present in patients with inner ear disease unless there has been extension of the inner ear disease into the brain stem. Otitis media/interna is frequently associated with extension to the CNS in people [37] and has been reported in domestic animals [38,39]. Neoplasms of the inner ear may also progress to involve the brain stem. Affected patients may initially show signs of peripheral vestibular disease associated with inner ear disease. Progressive
alteration of mentation, paresis, proprioceptive deficits, or other cranial nerve deficits may suggest extension into the brain stem.

References