Vestibular Anatomy and Dysfunction

Chris Adams, BVetMed, MRCVS
Neurology Resident
Southeast Veterinary Neurology of Boynton Beach

Anatomy of the inner ear

- Bony labyrinth
    - Contained within the petrous portion of the temporal bone
    - Consists of three continuous fluid-filled portions
        - Vestibule, three semicircular canals and cochlea
        - Dilation in one end of each of the bony semicircular canals is the ampulla
        - All three bony components contain perilymph, a fluid similar to cerebrospinal fluid
    - There are two openings within the bony labyrinth the vestibular and cochlear windows
        - Each opening is covered by a membrane and the stapes is inserted in the membrane that covers the vestibular window
- Membranous labyrinth
    - Contained within the bony labyrinth
    - Consists of four fluid-filled compartments - all of which communicate
        - Saccule and utriculus within the bony vestibule, the three semicircular ducts within the bony semicircular canals and a cochlear duct within the bony cochlea
    - Endolymph is continued within these compartments and is thought to be derived from the blood vessels along one wall of the cochlear duct and is absorbed back into blood through the blood vessels surrounding the endolymphatic sac
    - The three semicircular ducts are the anterior (vertical), posterior (vertical), and lateral (horizontal)
        - Oriented at right angles to each other
        - Rotation of the head around any plane causes endolymph to flow within one or more of the ducts
        - Each semicircular duct connects at both ends with the utriculus, which, in turn, connects with the saccule by way of the intervening endolymphatic duct and sac.
        - The saccule connects with the cochlea duct by the small ductus reuniens
- Crista Ampullaris
    - At one end of each membranous semicircular duct there is a dilation called the ampulla
        - On one side of the membranous ampulla there is a proliferation of connective tissue that forms a ridge called the crista lined on the internal surface by columnar neuroepithelial cells
        - On the surface a gelatinous structure that is composed of a protein-polysaccharide material called the cupula is present
            - The cupula extends across the lumen of the ampulla

    - The neuroepithelium is composed of two basic cell types: hair cells and supporting cells
        - The dendritic zones of the neurons of the vestibular portion of the vestibulocochlear nerve are in synaptic contact with the base of the hair cells
        - The hair cells have 40-80 hairs on their luminal surface
            - They are modified microvilli (stereocilia)
            - A single modified cilium (kinocilium)
            - They project into the lumen of the cupula
    - Movement of the fluid in the semicircular ducts causes deflection of the cupula
        - Bending of the stereocilia is the source of stimulus to the dendritic zone of the vestibular neuron that is in synaptic relationship with the plasmalemma of the hair cell
        - At one end of the semicircular duct is one membranous ampulla with its crista ampullaris
        - Since they are at right angles to each other movement of the head in any plane or angular rotation affects a crista ampullaris and stimulates vestibular neurons
    - Each semicircular duct on one side is paired with the semicircular duct on the opposite side
        - Left and right lateral ducts, the left anterior and right posterior ducts, and the left posterior and right anterior ducts
        - When movement in the direction of one of these three planes stimulates the vestibular neurons of the crista of one duct, they are inhibited in the opposite duct of the synergistic pair
    - The anatomic orientation of the stereocilia relative to the kinocilium on the surface of the crista is responsible for the difference in activity relative to the direction of the cupula deflection
        - Deviation towards increases the vestibular neuronal activity
    - These cristae function in dynamic equilibrium
        - Not affected by constant velocity of movement but respond to acceleration or deceleration, especially when the head is rotated
    - Vestibular neurons are tonically active and their activity is excited or inhibited by deflection of the cupula in different directions
- Macula
    - Receptor found in each utriculus and saccule within the bony vestibule
    - The macula is on one surface of each of these saclike structures
    - Oval-shaped plaque in which the membranous labyrinth has proliferated and columnar epithelial cells
    - Covering the neuroepithelium is a gelatinous material, the statoconiorum (otolithic) membrane
        - On the surface of this membrane are calcareous crystalline bodies known as statoconia (otoliths)

    - Macula hair cells have projections of their luminal cell membranes, stereocilia and kinocilia, into the overlying statoconiorum membrane
    - Macula in the saccule is oriented in a vertical direction (sagittal plane), whereas the macula of the utriculus is in a horizontal direction (dorsal plane)
        - Gravitational forces continually affect the position of the statoconia relative to the hair cells
    - Function in static equilibrium
        - Responsible for sensation of the static position of the head and linear acceleration or deceleration
        - The macula of the utriculus may be more important as a receptor for sensing changes in head posture, whereas the macula of the saccule may be more sensitive to vibrational stimuli and loud sounds

Vestibulocochlear Nerve: Cranial Nerve VIII - vestibular division

- The dendritic zones are in a synaptic relationship with the hair cells of each of the crista ampullaris and the macula utriculi and sacculi
- Axons course through the internal acoustic meatus with those of the cochlear division
- Bipolar-type sensory neurons are inserted along the course of the axons within the petrous portion of the temporal bone, where they form the vestibular ganglion
- After leaving the internal acoustic meatus with the cochlear division of the vestibulocochlear nerve, the vestibular nerve axons pass to the lateral surface of the rostral medulla at the cerebellomedullary angle, which occurs at the level of the trapezoid body and the attachment of the caudal cerebellar peduncle to the cerebellum
- Axons then enter the medulla between the caudal cerebellar peduncle and the spinal tract of the trigeminal nerve and terminate in telodendria at one of the two sites
    - Majority terminate in the vestibular nuclei in the medulla and pons
    - A few course directly into the cerebellum by way of the caudal peduncle and terminate in the fastigial nucleus in the cerebellar medulla and the cortex of the flocculonodular lobe
        - These form the direct vestibulocerebellar tract

Vestibular Nuclei

- Either side of the dorsal part of the pons and medulla adjacent to the lateral wall of the fourth ventricle are four vestibular nuclei forming a continuous column on each side
- From the level of the rostral and middle cerebellar peduncles, they extend caudally to the level of the caudal portion of the fourth ventricle
- The four vestibular nuclei are the rostral, medial, lateral and caudal
- Receive afferents from the vestibular division of the vestibulocochlear nerve
- From the vestibular nuclei are numerous projections, which may be grouped into spinal cord, brainstem and cerebellar pathways
- *Spinal Cord*
    - The lateral vestibulospinal tract courses caudally in the ipsilateral ventral funiculus through the entire spinal cord and terminates in all of the spinal cord segments on interneurons in the ventral gray columns

        - The interneurons
            - Facilitatory to ipsilateral alpha- and gamma-motor neurons to extensor muscles
            - Inhibitory to the ipsilateral alpha-motor neurons to flexor muscles
            - Some interneurons cross to the opposite ventral gray column, where they are inhibitory to the contralateral alpha- and gamma-motor neurons to extensor muscles
        - Most of the cell bodies are located in the lateral vestibular nucleus
    - The medial vestibulospinal tract course caudally in the ipsilateral ventral funiculus of the cervical and cranial thoracic spinal cord segments and terminate on interneurons in the ventral gray columns
        - Activate the alpha- and gamma-motor neurons that innervate primarily neck muscles
        - Also projects axons into the medial longitudinal fasciculus, which courses caudally in the dorsal portion of the ventral funiculus through the cervical and cranial thoracic spinal cord segment
        - Cell bodies located within the rostral, medial and caudal vestibular nuclei
    - Through these spinal cord pathways the position and activity of the limbs, neck and trunk can be coordinated with movements of the head
- *Brainstem*
    - Three general terminations in the brainstem
        - Motor nuclei of cranial nerves VI, IV and III
            - Axons course rostrally in the medial medial longitudinal fasciculus to terminate on the motor nuclei of abducens (VI), trochlear (IV) and oculomotor (III) nerves
            - Purpose is to provide coordinated movements associated with changes in the position of the head
            - Can be elicited by moving the head from one side to the other and observing the jerk nystagmus that is produced
                - When the brainstem is severely contused by a head injury, these pathways may be disrupted, and eyeball movements cannot be elicited by changing the position of the head
        - Reticular formation
            - Some of these axons provide afferents to the vomiting center located in the reticular formation
            - Involved with motion sickness
        - Conscious perception
            - Involves a relay through a thalamic nucleus
            - This pathway is not well defined for the vestibular system
            - Axons of neuronal cell bodies in vestibular nuclei course rostrally through the midbrain to terminate in the contralateral medial geniculate nucleus of the thalamus or some other thalamic nucleus where synapse occurs before project by way of the internal capsule to the cerebral cortex

- *Cerebellum*
    - Axons of neuronal cell bodies in the vestibular nuclei, in addition to some in the vestibular ganglia, project into the cerebellum through the caudal cerebellar peduncle
    - Terminate mostly in the cortex of the flocculus of the hemisphere and the nodulus of the vermis (flocculonodular lobe)
    - Collaterals synapse in the fastigial nucleus, which is the most medial of the three nuclei in the cerebellar medulla
- Through these systems the vestibular system functions to coordinate the position of the eyes, neck, trunk and limbs with the position and movements of the head; maintain equilibrium during active and passive movements and when the head is at rest

Clinical Signs of Vestibular System Disease

- Unilateral peripheral vestibular disease
    - Normally there is a constant flow of impulses occurring between the receptors of the inner ear to the components in the pons, medulla and cerebellum
        - When disruption of this constant signal occurs clinical signs develop due to loss of impulses
    - Posture
        - Loss of coordination between head and neck, trunk and limbs is seen in a head tilt
        - More ventral ear is directed toward the side of the vestibular lesion
        - The neck and trunk will lean, fall or even roll toward the side of the lesion
        - The neck and trunk may be flexed laterally with the concavity directed toward the side of the lesion
        - May tend to circle, in small diameter circles, toward the side of the lesion
            - Different than prosencephalic lesions which tend to be wide circles
        - Cats with vestibular disease will often adopt a crouched position and be reluctant to move
        - May be possible to elicit mild hypertonia in the limbs on the side of the body opposite to the side of the vestibular system lesion
    - Ataxia
        - Asymmetry may be explained by the loss of tonic activity in the vestibulospinal tract on the side of the lesion
            - Removes facilitation of ipsilateral extensor muscles
            - The unopposed activity of the contralateral vestibulospinal tract causes the neck and trunk to be forced toward the side of the lesion by excessive unopposed extensor muscle tonus
        - In some cases, you can blindfold the patient and this will make the vestibular ataxia worse due to the loss of visual feedback
    - With peripheral vestibular disease the rolling is usually limited to the first 24 to 48 hours after a peracute onset of clinical signs
    - Nystagmus
        - Involuntary rhythmic oscillation of the eyes

- Eye movements that are unequal, with a slow movement in one direction and a fast return of the eye to its starting position, indicate a jerk nystagmus
- The direction of the nystagmus by convention is ascribed to the direction of the quick of fast phase
- *No normal or abnormal nystagmus may occur with bilateral loss of function in the peripheral vestibular system, its central components the medial longitudinal fasciculus or the GSE motor nuclei of the abducent, trochlear and oculomotor nerves*
- Normal
    - Jerk nystagmus is a normal response to any rapid movement of the head
    - Stand over the patient and observe the horizontal jerk nystagmus as you move their head from side to side
        - As you move to the right both eyes will repeatedly jerk quickly to the right and slowly return to the left and vice versa for movement in the left direction
        - This is known as a normal vestibular or physiologic nystagmus
        - Tests not only vestibular system but also the medial longitudinal fasciculus in the brainstem and the abducent nerve innervation of the lateral rectus muscle that abducts the eye and the oculomotor nerve innervation of the medial rectus muscle that adducts the eye
    - If you flex and extend the animal's neck so that the head moves up and down you will see the same movements in the vertical direction
    - A normal reflex in which the slow component is initiated by way of the vestibular receptors in the membranous labyrinth and the quick component involves a brainstem center related to the vestibular system
    - Requires normal function of the vestibular system components, normal medial longitudinal fasciculus bilaterally and normal general somatic efferent neurons in the motor nuclei of the abducent, trochlear and oculomotor nerves
- Abnormal
    - May be observed when the head is held in its normal extended position and would be termed resting or spontaneous nystagmus
    - If induced only by holding the head fixed in lateral flexion or full extension it is called positional nystagmus
        - Important to remember a normal nystagmus will be present when moving the head; assess once movements have stopped

        - With peripheral vestibular diseases the imbalance represents a loss
        - In disorders of the peripheral vestibular system, the abnormal resting or positional nystagmus is directed in a horizontal-dorsal plane or its rotatory but is always directed away from the side of the lesion
        - We no longer us vertical nystagmus alone to distinguish peripheral from central vestibular system disease
            - Some patients will have an almost vertical nystagmus but with careful examination, often will still have a rotary component to the nystagmus
        - Resting nystagmus is more common in acute disorders of the peripheral components of the vestibular system
    - Postrotatory
        - If an animal is rotated rapidly, with acceleration, the labyrinth moves around the endolymph, which deflects the cupula of the crista ampullaris, stimulating the vestibular nerve and thus eliciting eye movements
        - At a constant velocity the cupulae are not deflected but when the rotation is stopped the endolymph continues to flow for a short time which deflects the cupulae, this causes a jerk nystagmus with a fast phase in the OPPOSITE direction of the rotation
    - Caloric
        - Unreliable and not practical in veterinary medicine
        - Using cold water this test will normally produce a jerk nystagmus to the side opposite to the ear being stimulated
        - Irrigation of the external ear canal for 3-5 minutes will cause endolymph to flow in the semicircular ducts
    - Congenital
        - Congenital pendular resting nystagmus occurs in humans as an inherited abnormality or secondary congenital lesions in the visual system
        - Typically pendular in that the eye movements are equal in velocity in both directions and it is very rapid
        - Does not interfere with vision
        - In cattle, a congenital rapid fine pendular nystagmus is observed in many breeds and usually persists for the lifespans of these animals
            - Sporadic in occurrence
        - Congenital rapid fine pendular nystagmus is most often observed in cats with a varying degree of ocular albinism
            - An abnormality in the retinogeniculate projections and the neuronal organization of the lateral geniculate nucleus has

been observed in the Siamese cat and the white Persian tiger
                - More retinal ganglion neurons project their axons contralaterally in Siamese cats than the normally pigmented feline breeds
            - This congenital pendular nystagmus occurs in some cats and cattle with the Chediak-Higashi syndrome, in which pigmentation and melanin granules are abnormal
        - Some aberration of the architecture of the visual pathway mauy be the common factor in these patients
            - The albino cat exhibits excessive contralateral projection of optic nerve axons, and the Belgian Shepherds shows complete lack of any contralateral projection
    - Strabismus
        - Abnormal eye position relative to the orbit or palpebral fissure
        - In the normal animal, then the neck and head are extended the eyes should elevate and remain in the center of the palpebral fissure
            - With disorders of ANY component of the vestibular system this effect may not occur on the side of the lesion, resulting in a dropped or ventrally deviated eye that exposes the sclera dorsally
        - Occasionally a ventral or ventrolateral strabismus is observed without head and neck extension but disappears when the head position is changed
            - Often will mimic an oculomotor nerve strabismus but normal physiologic nystagmus is present indicating that cranial nerves III and VI are not impaired
        - In farm animals it is normal for their eyes to not elevate completely when the head and neck are extended; therefore you expect to see some sclera dorsal to the cornea in these species, but it should be equal on both sides
- Bilateral peripheral vestibular system disease
    - When both the peripheral components of the vestibular system are dysfunctional bilaterally, no postural asymmetry is noted
    - Balance is lost to both sides which results, typically, in a crouched posture closer to the ground
    - The most characteristic clinical sign is wide head excursions
    - No normal or abnormal nystagmus can be seen
    - If you support their head and the support is then suddenly withdrawn the head may rapidly descend ventrally beyond the normal neutral position and is termed *head rebound phenomenon*
        - Typically seen with cerebellar dysfunction but may also be seen with bilateral vestibular dysfunction
- Central vestibular system disease
    - A nystagmus that changes direction when the position of the head is changed or a horizontal or rotary nystagmus directed toward the side of the head tilt and

body deviation will occur with central vestibular system disease BUT NOT peripheral vestibular system disease
    - If the nystagmus is absolutely vertical, then this likely suggests central vestibular system dysfunction
    - The most reliable clinical signs that determines that a lesion exists in the pons or medulla that is affecting the vestibular nuclei is ipsilateral proprioceptive deficits or a recongnizable spastic hemiparesis and ataxia from involvement of the UMN and GP systems adjacent to these nuclei in the caudal brainstem
    - Clinical signs related to cerebellar dysfunction or any cranial nerve other than facial or vestibulocochlear nerves implicate a central vestibular system disease
        - Remember that facial nerve paralysis and Horner syndrome may occur along with clinical signs of vestibular nerve dysfunction with diseases of the middle and inner ear in small animals and just facial paralysis in horse and farm animals
    - Resting nystagmus is more common in acute disorders of the peripheral components of the vestibular system, and the rate or either resting or positional nystagmus tends to be more rapid in peripheral dysfunction than central dysfunction
    - Lesions that involve solely the vestibular nuclei on one side cause ipsilateral clinical signs similar to all the lesions that affect the peripheral components of the vestibular system with the patients head tilt and loss of balance directed toward the side of the lesion
- Paradoxical vestibular system disease
    - Unique syndrome in which the head tilt and loss of balance are directed toward the side *opposite* to the central lesion
    - A small population of Purkinje neurons, most of which are located in the cortex of the folia of the flocculus in the hemisphere and the nodulus in the vermis
        - Have axons that leave the cerebellum directly as a component of the caudal cerebellar peduncle where they terminate in the vestibular nuclei
        - They are inhibitory to the activation of these neuronal cell bodies
    - A lesion in the caudal cerebellar peduncle interferes with this inhibition, resulting in excessive discharge of vestibular system neurons on that side
        - Head tilt and loss of balance to the side opposite to this lesion the direction of the head tilt and balance loss will be toward the side with the <u>least</u> activity of the vestibular system
    - In practice the side of this unilateral lesion will be determined on your neurological examination by the side of the postural reaction deficit
- The peripheral vestibular system is the ONLY system involved with movement of the animal that when deficient does not interfere with the performance of the postural reactions
    - The ability for the animal to right itself from lateral recumbency may be altered
- Vomiting as a continuous event is an uncommon clinical sign of vestibular system dysfunction in domestic animals

- In approximately 25% of animals presented with an acute onset of vestibular system dysfunction, the owners will report observing an episode of vomiting at the onset of clinical signs

Reference:

1. De Lahunta, A., Glass, E.N. and Kent, M., 2015. *Veterinary Neuroanatomy and Clinical Neurology, Fourth Edition*. Elsevier Health Sciences.