Neurology Survival Kit

Mini Series

Session 2:

Droops, Tilts & The Head 'Stuff'

RCVS & European Specialist in Veterinary Neurology
Dr Laurent Garosi  DVM, Dip ECVN, MRCVS
DROPPED JAW, LOCK JAW AND DROOPY FACES

The trigeminal nerve (CN V) provides motor innervation of the masticatory muscles (temporalis, masseter, medial and lateral pterygoid and rostral part of the digastric muscles) and sensory innervation of the face (including cornea, mucosa of the nasal septum and mucosa of the oral cavity). It consists of three branches: ophthalmic, maxillary and mandibular. The mandibular branch serves both a motor and sensory function while the other two have only sensory function. The motor function of CN V is assessed by evaluating the size and symmetry of the masticatory muscles and testing the resistance of the jaw to opening the mouth.

DROPPED JAW

An inability to close the mouth (also described as ‘dropped jaw’) is a commonly encountered presentation in dogs. Affected animals frequently present as well with difficulty eating and drinking, and hypersalivation. The most common cause for this presenting sign is idiopathic trigeminal neuropathy (also called trigeminal neuritis, trigeminal neurapraxia or trigeminal nerve palsy). Other neurological causes for bilateral paralysis of the mandibular branch of the trigeminal nerve include multicentric lymphosarcoma (neoplastic lymphoid cell infiltration), myelomonocytic leukaemia, idiopathic hypertrophic chronic pachymeningitis, disseminated, non-suppurative ganglionicradiculoneuritis and rabies. Non-neurological causes are usually responsible for mechanical obstruction and include bilateral luxation of the temporomandibular joints, fracture of the mandible or oral foreign body.

Idiopathic trigeminal neuropathy

Idiopathic trigeminal neuropathy is the most common cause of ‘dropped jaw’. Onset is usually acute. Horner’s syndrome, some degree of sensory loss in the sensory distribution of the trigeminal nerve and facial nerve paralysis can occasionally be associated with the ‘drop jaw’. It is a diagnosis of exclusion and cannot be confirmed by any antemortem test. The only tests that showed abnormal results in most dogs are electromyography (EMG) and CSF. EMG often reveals positive sharp wave and/or fibrillation potentials in the masticatory muscles unless the dog is tested to soon after the onset of mandibular paralysis (up to 7 days after the onset). CSF
analysis can reveal a mild mononuclear pleocytosis, often with normal or mildly elevated protein content or can be normal. The etiology remains unknown. A non-suppurative inflammatory neuritis in motor branches of the trigeminal nerve and ganglion has been confirmed in some cases, however it is unknown whether this inflammatory process occur in all cases. Some report postulated a link with dogs carrying heavy object which was hypothesize to stretch the mandibular nerve from hyperextension of the jaw. Treatment is mainly supportive, helping the animal to eat and drink. Corticosteroid administration appears not to affect the clinical course of the disease. Use of tape muzzles has been recommended to improve ingestion of food as these dogs are unable to grab food but can swallow normally. Mean time for recovery ranges from 2 to 10 weeks. Dogs with longer recovery frequently show marked atrophy of the masticatory muscle caused by prolonged denervation.

**Chronic polyradiculoneuritis**

Although rare, unilateral atrophy or bilateral paralysis of muscles of mastication can be caused by chronic polyradiculoneuritis in which neural involvement is largely focused in the trigeminal nerves. Syndromes that may be classified as chronic polyradiculoneuritis include chronic relapsing polyradiculoneuritis, hypertrophic neuropathy, polyradiculoneuritis, and chronic polyneuritis. The etiology and pathogenesis of these syndromes are largely undertermined. Very little is known about the treatment and prognosis of these conditions.

**Idiopathic hypertrophic chronic pachymeningitis**

Idiopathic hypertrophic chronic pachymeningitis is a recently recognised cause of multiple cranial nerve deficits. ‘Dropped jaw’ is the most common complain for this condition, however, most dogs also show associated other cranial nerve deficits. No long tract signs (limb weakness, postural reaction deficit, ataxia) are usually observed as this condition mainly affect the meninges without brain parenchymal involvement. Lurcher and Greyhound seem to be predisposed. This condition is characterised by diffuse thickening of the dura mater caused by fibrosing inflammatory process that involves the dura mater. The etiology is unknown. The CSF in most cases showed inflammatory changes but can be normal. Neuroimaging studies revealed diffuse or localized thickening of the dura. MR imaging is key to diagnosis of this disorder with increased meningeal thickening and diffuse or localized hyperintensity in T2W and FLAIR images and severe contrast enhancement. Treatment consists in using immunosuppressive doses of corticosteroids (prednisolone 1 to 2 mg/kg every 12 hours orally until remission then at decreasing dosage). The addition of other immunosuppressant such as cytosine arabinoside seems to help getting a better control of the disease. Although most dogs get into clinical remission, cure is more difficult to obtain.
Neoplastic cell infiltration of the trigeminal nerve

Dogs with ‘dropped jaw’ that present with multiple cranial nerve or other neurological deficits are more at risk of having malignant disease and further diagnostic test to rule-out neoplastic or infectious disease should be considered. Both multicentric lymphosarcoma and myelomonocytic leukaemia have been identified as possible causes of ‘dropped jaw’ in dogs primarily via neoplastic infiltration of the trigeminal nerve. Lymphosarcoma in dogs can take a number of different forms and can involve the CNS, the peripheral nervous system (PNS), or both. Dogs with CNS involvement frequently present immature lymphoid pleocytosis on CSF analysis while dogs with PNS involvement may have normal CSF. Treatment options are limited and include the use of corticosteroids, lomustine (CCNU), carmustine (BCNU), cytosine arabinoside and radiation therapy. Regardless of treatment modality used, the long-term survival for dogs with multicentric lymphoma is guarded to poor with most patient showing only a partial or brief remission of clinical signs.

LOCK JAW (TRISMUS)

Trismus is defined as difficulty opening the jaw. Neurological conditions responsible for trismus include masticatory muscle myositis, muscular dystrophy, polymyositis, extraocular myositis (referred jaw pain) and tetanus. Non-neurological causes comprise of craniomandibular osteopathy, retrobulbar abscess and temporomandibular joint disease including luxation/subluxation. Complete physical and neurological examination is important to try distinguishing there different conditions. Patients should be closely examined for evidence of trauma that could have resulted in temporomandibular joint luxation/subluxation. Thorough oral and ophthalmic examination should be performed. Retrobulbar masses often cause a visible swelling or drainage behind the carnassial teeth. Animal with trismus caused by tetanus often show a characteristic facial expression (‘risus sardonicus’) resulting from an increase in facial muscle tone.

Masticatory muscle myositis

Masticatory muscle myositis (MMM) is an auto-immune, focal inflammatory myopathy with clinical signs restricted to the muscles of mastication (masseter, temporalis, pterygoid and rostral digastricus) which are innervated by the mandibular branch of the trigeminal nerve. Masticatory muscles contain a unique muscle fiber type (type 2M) that differs both histochemically and biochemically from fiber type present in limb muscles (types 1A and 2A). Biopsies of dogs with MMM are characterised by intense multifocal lymphocytic and plasmacytic perivascular infiltration, occasional eosinophils, necrosis and phagocytosis of type 2M myofibers. Circulating auto-antibodies against masticatory muscle type 2M fibers (fiber type-specific auto-antibodies)
can be detected in more than 80% of dogs with MMM and are the basis of serology testing for this condition. Despite many hypotheses proposed to explain the formation of auto-antibodies directed specifically against type 2M fibers including molecular mimicry, the primary initiating factor to this auto-immune disorder is unknown.

The most common clinical signs associated with MMM are inability to open the jaw, jaw pain, and masticatory muscle atrophy. Some dogs may present with pyrexia, mandibular lymphadenopathy, trismus, swollen and painful masticatory muscles, and bilateral exophthalmos from swelling of the pterygoid muscles during the acute phase of the condition. Many owners however do not recognise a problem until the chronic phase when marked muscle atrophy and enophthalmos because of atrophied pterygoid muscles are present.

MMM can be seen in any breed of dog with no apparent gender predilection. The average age of onset is 3 years, although dogs as young as 4 months of age with MMM have been reported. Diagnosis can be confirmed by detection of significant levels of anti-type 2M muscle fiber antibodies in the serum of suspected dogs. False negative results may occur if corticosteroids have been administered before sampling. Serum creatinine kinase (CK) levels are modestly elevated in some dogs in the acute phase of MMM. EMG can help to confirm the selective involvement of masticatory muscles and differentiate MMM from polymyositis. However, EMG may be normal in dogs with end-stage disease because of severe fibrosis and myofiber depletion. Evaluation of muscle biopsy taken from the masticatory muscles can also provide diagnostic confirmation of the disease as well as prognostic information by determining the stage of the disease.

Immunosuppressive doses of corticosteroids (prednisolone 1 to 2 mg/kg every 12 hours orally) comprises the cornerstone of treatment of MMM. This dose should be maintained until jaw function and serum CK level (when initially elevated) have both returned to normal. Dosage of prednisolone is then slowly decreased over a few months to the lowest every-other day dose that keeps the clinical signs at bay. Other immuno-suppressive agents such as azathioprine (1 to 2 mg/kg every 24 hours orally) are indicated in dogs that failed to respond to corticosteroids treatment or that relapse when the dose is tapered. Short-term prognosis is usually good however many dogs that are treated for insufficient period of time will experience relapses. Life-long treatment is occasionally necessary. The prognosis of dogs in the more chronic phase of the disease (gradual replacement of myofibers by fibrous tissue) is guarded. Persistent muscle atrophy is a common manifestation of the disease.

MASTICATORY MUSCLE ATROPHY

Masticatory muscle atrophy can occur unilaterally or bilaterally. It can result from impaired innervation due to lesions of the motor branch of the trigeminal nerve, lesions affecting the
masticatory muscle themselves or systemic disorders. The later is not a cause of unilateral muscle atrophy.

**Unilateral masticatory muscle atrophy**

Unilateral masticatory muscle atrophy is uncommon with myositis, and when present, a trigeminal nerve disorder should be suspected. Unilateral involvement of the motor part of CN V causes ipsilateral masticatory muscle atrophy secondary to neurogenic atrophy and decreased jaw tone. Enophthalmia and protrusion of the third eyelid can be observed in the ipsilateral eye (passive retraction of the eyeball secondary to loss of temporalsis and digastric muscle mass). Decreased or complete loss of facial sensation may be seen with associated involvement of trigeminal sensory nerve branches. Involvement of the ophthalmic branch of CN V can also produce decreased tear secretion and neurotropic keratitis secondary to the loss of afferent stimulation to the lacrimal reflex. Other neurological signs (e.g., hemiparesis, facial nerve paralysis, circling, mydriasis…) may be seen as the result of expansion of the mass and subsequent damage to adjacent brain structures. Underlying causes for this unilateral masticatory muscle atrophy come down to trigeminal nerve sheath tumors and neuritis. In comparison to nerve sheath tumors, inflammatory diseases restricted to cranial nerves are uncommon in dogs. Possible causes of cranial neuritis include autoimmune or infectious processes. MR imaging allows earlier diagnosis of these trigeminal nerve lesion.

**Bilateral masticatory muscle atrophy**

Bilateral masticatory muscle atrophy can be caused by the following:

- Bilateral involvement of the motor branches of CN V (see section on dropped jaw) which is usually associated with reduced jaw tone, a drop jaw and/or inability to close the mouth voluntarily
- Systemic disorder (cachexia, hyperadrenocorticism or exogenous steroid administration)
- Chronic masticatory muscle myopathy. In case of chronic masticatory myositis, the atrophy is caused by destruction of myofibres and scarring. It is usually associated with reduced ability to open the jaw (see section on trismus)

As with unilateral atrophy, bilateral masticatory muscle atrophy may cause enophthalmia and protrusion of the third eyelid.

**DROOPY FACES: FACIAL NERVE PARALYSIS**

The facial nerve (CN VII) is motor to the muscle of facial expression and sensory (providing the sense of taste) to the rostral two thirds of the tongue and palate. Its parasympathetic component innervates the lacrymal gland, the mandibular and sublingual salivary glands. Neurons
innervating the muscles of facial expression are located in the facial nucleus in the rostral medulla oblongata. The axons pass in the internal acoustic meatus of the petrosal bone on the dorsal surface of the vestibulocochlear nerve and leave the skull through the stylomastoid foramen. The facial nerve courses through the middle ear before branches are distributed to the muscles of facial expression (ear, eyelids, nose, cheeks, lips) as well as caudal portion of the digastricus muscle.

The motor function of CN VII is primarily assessed by observation of the face for symmetry (position of the ears and lip commissure on each side within the same plane, symmetry of the palpebral fissure), spontaneous blinking and movement of the nostrils. It also is the motor response (efferent part) of following tests: palpebral reflex, corneal reflex, menace response, pinching of the face. The parasympathetic supply of the lacrimal gland associated with CN VII can be evaluated by the Schirmer tear test strips. This later test quantitatively assesses the tear flow by measuring the amount of wetting on a filter paper inserted in the lower conjunctival fornix at the outer half of the palpebral fissure. In normal dog and cats, the wetting of the tear test paper ranges from 10 to 25 mm in one minute.

Salivation can be subjectively assessed by examining the mouth for a moist mucosa.

Motor involvement of CN VII produces the following signs: drooping and inability to move the ear and lip, drooling, widened palpebral fissure, absent spontaneous and provoked blinking, absent abduction of the nostril during inspiration, deviation of the nose toward to the normal side due to the unopposed muscle tone on the unaffected side. With chronic denervation, the lips are retracted further than normal and the nostril is deviated to the affected side as a result of muscle fibrosis. Unilateral involvement can be seen in the asymmetry of the ears, eyelids, lips and nose. Lesion of the individual branches of the facial nerve long their course produces paresis or paralysis to the specific muscle they innervate. Involvement of the parasympathetic supply of the lacrimal gland produces keratoconjunctivitis sicca.

Differential diagnosis of facial nerve paralysis using the mnemonic mean VITAMIN D should include:

- **I**nfectious and inflammatory disorder: middle ear infection, viral/bacterial/protozoal and immune-mediated meningoencephalitis and inflammatory/immune-mediated neuromuscular disorders such as polyradiculoneuritis, myasthenia gravis
- **T**raumatic disorder: iatrogenic injury to the peripheral facial nerve, head trauma
- **M**etabolic: hypothyroid polyneuropathy
- **I**diopathic: acute unilateral or bilateral paralysis may be seen
- **N**eoplasm: CNS neoplasm located to the caudal fossa, middle ear neoplasm

Compared to with middle/inner ear disease where facial nerve paralysis may be associated with Horner’s syndrome and peripheral vestibular disorder, diseases of the facial nerve nucleus in the medulla oblongata are usually associated with brainstem signs such as ipsilateral paresis and
postural reaction deficit, decreased mentation, central vestibular signs and other cranial nerve deficits. With generalised neuromuscular disorders, any combination of cranial nerve deficit as well as limb signs of neuromuscular weakness (intermittent or permanent weakness, decreased segmental spinal reflexes and muscle tone associated or not with muscle atrophy).
VESTIBULAR DISORDER

The vestibular system is essential in maintaining balance and preventing the animal falling over by keeping and adapting the position of the eyes, head and body with respect to gravity. It is therefore not surprising that disease of the vestibular system results in some of the most dramatic and distressing neurological signs. Head tilt, falling, rolling, leaning, circling, abnormal nystagmus and ataxia commonly result. Clinical signs of vestibular disease may be a result of lesions involving either the receptor organs in the inner ear or the vestibular portion of the eighth cranial nerve (ie, peripheral vestibular disease) or lesions involving the brainstem vestibular nuclei or vestibular centers in the cerebellum (ie, central vestibular disease). This article reviews the physiology of the vestibular system and discusses the clinical approach of an animal with vestibular disorder.

THE VESTIBULAR SYSTEM

The vestibular system is a sensory system. Its main function is to control balance and usually prevents the animal from falling over by maintaining and adapting the position of the eyes, head and body with respect to gravity. The vestibular system consists of a receptor organ within the petrous temporal bone (inner ear), the vestibular nerve, and a balance control centre at the back of the brain (four brainstem nuclei located in the rostral medulla oblongata on each side of the fourth ventricle). The receptor organ (saccule, utricule and semi-circular canals) detects the position and movement of the head in space while the animal is standing at rest or when it is moving. The information on the position of the head is converted into electrical signals which are sent via the vestibular nerve to the brain. The balance control centre in the brainstem processes this information and sends messages to the rest of the body to keep the animal upright (facilitatory effect on the ipsilateral extensor muscles of the limb via the vestibulospinal tract). Messages are also sent to the muscles controlling movement of the eyes (via medial longitudinal fasciculus and cranial nerves III, IV, VI) to change the position of the eyes according to the position of the head. Finally, the brainstem vestibular nuclei receive some influences from higher vestibular centers in the thalamus and from vestibular centers in the cerebellum (flocculonodular
lobe and fastigial nuclei). The latter have inhibitory effect mainly on the brainstem vestibular nuclei.

Through these pathways, the vestibular system controls the position of the eyes, trunk and limbs depending on the position and movement of the head.

VESTIBULAR DISORDERS: CLINICAL SIGNS

Vestibular disorders (VD) are common in dogs and cats and may result in any or all of the following clinical signs: head tilt, falling, rolling, leaning, circling, abnormal nystagmus, positional strabismus, and ataxia.

- **Head tilt** often indicates a VD. This abnormal head posture is characterized by a rotation of the median plane of the head (one ear is held lower than the other). It occurs in VD as a result of the loss of antigravity muscle tone on one side of the neck. It must be differentiated from a head turn where the median plane of the head remains perpendicular to the ground but the nose is turned to one side. Such head turn is usually associated with a body turn. A head turn does not indicate a vestibular disorder and is usually toward the side of a forebrain lesion.

- **Circling** may occur in conjunction with VD as well as an asymmetrical or focal lesion in the forebrain. Tight circles are usually but not exclusively associated with a VD, while wide circles are often associated with a forebrain lesion.

- **Nystagmus** is an involuntary rhythmic movement of the eyeballs. Physiologic nystagmus is nystagmus that occurs in normal animals while pathologic nystagmus reflects an underlying VD. The direction of the nystagmus is typically defined by the direction of the fast phase. Physiologic nystagmus can be induced in the normal animal by rotating the head from side to side (oculo-vestibular reflex). It is characterized by a slow phase in the opposite direction of the head movement and a fast compensatory phase in the same direction as the head rotation. Physiologic nystagmus can be depressed in animal with unilateral VD or absent in animal with bilateral VD. Pathologic nystagmus can be either spontaneous (observed when the head is in a normal position at rest) and/or positional (that which occurs, or is altered in character, intensity or direction, with alteration in the position of the head, for example by placing the animal upside down on its back). Nystagmus is usually classified on the basis of its direction and may be horizontal, vertical or rotatory and may change in direction on changing position of the head. The fast phase of pathologic nystagmus is typically directed away from the side of the vestibular lesion.
- **Strabismus** refers to an abnormal position of the globes. Strabismus can be seen in VD when the head is placed in an abnormal position (extended dorsally or the animal placed upside down on its back). VD often causes a ventral or ventrolateral positional strabismus in the eye on the same side as the vestibular lesion.

- **Ataxia** is defined as an uncoordinated gait and can be caused by a vestibular disorder (vestibular ataxia), a cerebellar disorder (cerebellar ataxia) or a peripheral nerve, spinal cord or brainstem disorder (proprioceptive or sensory ataxia) (Table 1). Vestibular lesions often cause ataxia characterized by swaying of the trunk and head, base-wide stance, leaning, falling and rolling to one side with unilateral lesion. With bilateral VD, affected animals tend to fall to either side and often show wide excursion of the head from side to side.

<table>
<thead>
<tr>
<th>Type of ataxia</th>
<th>Anatomic diagnosis</th>
<th>Neurologic signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proprioceptive</td>
<td>General proprioceptive pathways</td>
<td>Abnormal postural reactions. Limb paresis may also be evident as spinal cord and peripheral nerve lesions usually concurrently affect the motor pathways</td>
</tr>
<tr>
<td></td>
<td>- peripheral nerve</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- dorsal root</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- spinal cord</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- brainstem</td>
<td></td>
</tr>
<tr>
<td>Vestibular</td>
<td>Vestibular apparatus</td>
<td>Head tilt, leaning, falling or rolling to one side, abnormal nystagmus, strabismus, normal (peripheral) or abnormal (central) postural reactions in case of unilateral dysfunction</td>
</tr>
<tr>
<td></td>
<td>- vestibular nuclei (central)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- vestibular portion of CN VIII or vestibular receptors (peripheral)</td>
<td>Crouched posture, reluctance to move and wide head excursion in case of bilateral dysfunction</td>
</tr>
</tbody>
</table>
| Cerebellar     | Cerebellum         | Broad-based stance, swaying of the trunk, intention tremors of the head, loss of balance on both sides as well as forward and backward, dysmetric gait, pendular nystagmus, delayed and then exaggerated response to postural reactions testing, ipsilateral menace deficit with normal
Laurent S Garosi  DVM, Dip ECVN, MRCVS

| vision, absence of limb paresis and normal mentation (pure cerebellar disease) |

Table 1. Classification and criteria for clinical differentiation of ataxia

Animal with acute vestibular disease may additionally display vomiting associated with disequilibrium.

VESTIBULAR DISORDERS: LOCALIZING THE LESION

Clinical signs of VD may be a result of lesions involving the receptor organs in the inner ear or the vestibular portion of the eighth cranial nerve running in the petrous part of the temporal bone (ie, peripheral VD) or lesions involving the brainstem vestibular nuclei (ie, central VD).

- **Peripheral or central?**

Most lesions affect a region, rather than a specific nerve or nucleus, so accompanying neurologic abnormalities can often be used to localize the lesion to the peripheral or central vestibular system (Table 2).

Both peripheral and central VD can cause a head tilt, horizontal or rotatory nystagmus, and ataxia. Facial paralysis and Horner’s syndrome can be seen with peripheral VD due to the proximity of cranial nerve VII (facial nerve) and the sympathetic nerve supply to the eye to the vestibular nerve in the region of the petrous temporal bone. Correctly identifying central VD requires identification of clinical signs that cannot be attributed to diseases of the peripheral vestibular system. Lesions that affect the central vestibular system typically have additional clinical signs suggestive of brainstem involvement. Such lesions often involve the reticular formation as well as ascending and descending motor and sensory pathways to the ipsilateral limbs. Therefore, abnormal mental status, ipsilateral paresis, and conscious proprioceptive deficits are commonly associated with central VD. Deficits of cranial nerves V through XII can also be associated with central VD. The presence of spontaneous or positional jerk nystagmus indicates vestibular dysfunction but does not further localize the lesion to the peripheral or central vestibular system. However, vertical nystagmus and nystagmus that changes in direction on changing position of the head are a feature of central vestibular lesions. Rate of nystagmus (number of beats per minute with the head in a neutral position as well as with the animal in dorsal recumbency) can further assist with differentiation between central VD from peripheral VD. Median rate of resting and positional nystagmus appears to be significantly faster for dogs with
peripheral VD with a resting nystagmus ≥ 66 beats per minute providing the highest combined sensitivity and specificity in diagnosing peripheral VD. With peripheral and central VD, the head is usually tilted in the direction of the lesion. With paradoxical VD, however, the head is usually tilted opposite to the direction of the lesion.

<table>
<thead>
<tr>
<th></th>
<th>Peripheral</th>
<th>Central</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head tilt</td>
<td>Ipsilateral</td>
<td>Ipsilateral (or contralateral in case of paradoxical VD)</td>
</tr>
<tr>
<td>Asymmetrical ataxia</td>
<td>Ipsilateral</td>
<td>Ipsilateral</td>
</tr>
<tr>
<td>Nystagmus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Horizontal</td>
<td>Ipsilateral</td>
<td>Ipsilateral or contralateral</td>
</tr>
<tr>
<td>• Rotatory</td>
<td>Ipsilateral</td>
<td>Ipsilateral or contralateral</td>
</tr>
<tr>
<td>• Vertical</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>• Positional</td>
<td>No (except during the early phase of recovery)</td>
<td>Yes</td>
</tr>
<tr>
<td>Postural reaction deficits</td>
<td>No</td>
<td>Yes (always ipsilateral to the lesion)</td>
</tr>
<tr>
<td>Abnormal mental status</td>
<td>No</td>
<td>Possible</td>
</tr>
<tr>
<td>Circling</td>
<td>Ipsilateral</td>
<td>Ipsilateral or contralateral</td>
</tr>
</tbody>
</table>

Table 2. Clinical findings associated with peripheral and central vestibular disease

Occasionally, intracranial lesions can result in signs suggestive of a peripheral lesion. Although animals with a peripheral VD have a normal level of consciousness and no evidence of weakness or postural reaction deficits, the absence of these signs does not rule-out the presence of a central VD. If in doubt about the localization of the lesion, the clinician should evaluate the animal for central VD as well as peripheral VD.

In very rare cases, VD may be part of a diffuse polyneuropathy or cranial polyneuropathy. Other cranial nerve dysfunction such as dysphagia, tongue weakness, jaw weakness and/or facial paralysis as well as limb weakness with depressed segmental spinal reflexes may be seen. Note also that lesions of the thalamus and/or extrapyramidal basal nuclei may also cause abnormal head posture and signs of central VD.
• **Ipsilateral, contralateral or bilateral lesion?**

With both central and peripheral VD, the head tilt, circling and nystagmus typically occur ipsilateral to the side of the lesion. Less frequently, lesions affecting the caudal cerebellar peduncle, the fastigial nucleus, or the flocculonodular lobes of the cerebellum can cause central VD with a resulting paradoxical head tilt. This syndrome is called paradoxical because the head tilt and circling occur contralateral to the side of the lesion. Bilateral VD is characterized by head sway from side to side, loss of balance on both sides and symmetrical ataxia with a wide-based stance. A physiological nystagmus usually cannot be elicited and a head tilt is not observed.

**DIFFERENTIAL DIAGNOSIS OF VESTIBULAR DISORDER**

A lesion must be localized to a particular section of the vestibular apparatus before an appropriate differential diagnosis can be established and further test conducted. The formation of a differential diagnosis list is essential in choosing and interpreting any diagnostic test. Diseases affecting the nervous system are classically classified in disease processes using the mnemonic **VITAMIN D**.

<table>
<thead>
<tr>
<th>Disease mechanism</th>
<th>Peripheral vestibular disease</th>
<th>Central vestibular disease</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Vascular</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Brain infarct</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Brain hemorrhage</td>
</tr>
<tr>
<td><strong>Inflammatory/Infectious</strong></td>
<td>Otitis media/interna</td>
<td>Infectious encephalitis</td>
</tr>
<tr>
<td></td>
<td>Nasopharyngeal polyps</td>
<td>(Distemper, Toxoplasma,</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Neospora, Fungus, Bacterial, FIP)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Meningo-encephalitis of unknown etiology</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(GME, necrotizing, idiopathic)</td>
</tr>
<tr>
<td><strong>Trauma</strong></td>
<td>Head trauma</td>
<td>Head trauma</td>
</tr>
<tr>
<td><strong>Toxic</strong></td>
<td>Aminoglycosides, topical chlorhexidine</td>
<td>Metronidazole</td>
</tr>
<tr>
<td><strong>Anomalous</strong></td>
<td>Congenital vestibular disease</td>
<td>Intracranial intra-arachnoid cyst, dermoid/epidermoid cyst, Dandy-Walker syndrome, Chiari-like</td>
</tr>
</tbody>
</table>
Idiopathic vestibular syndrome and otitis media/interna are the two most common causes of peripheral VD.

**Idiopathic vestibular syndrome** is common in adult cats and dogs (often geriatric). Clinical signs are usually peracute and initially severe with affected animal appearing extremely disable in the first 48 to 72 hours. If facial nerve paralysis or Horner’s syndrome (miosis, enophtalmia, protrusion of third eyelid, ptosis of upper eyelid) is also present then other differentials should be considered. Diagnosis is based on the presence of compatible history and exclusion of other causes of peripheral VD. Most animals tend to improve over 1 to 3 weeks period and often return to normal. However, some animal may be left with a permanent head tilt or episodic ataxia. No treatment has proved beneficial and recurrence is possible.

**Otitis media/interna** can be secondary to otitis externa, oropharyngeal infection (spreading via the auditory tube) or hematogenous spreading. Therefore, the absence of sign of otitis externa does not rule-out the presence of otitis media/interna. Clinical course can be acute or progressive. Signs of VD caused by otitis media/interna are often associated with ipsilateral facial nerve paralysis and/or Horner’s syndrome. Diagnosis is made by otoscopic examination and imaging studies (bulla radiographs, CT or MR scan) and/or exclusion of other causes of peripheral VD. If fluid is visualized within the middle ear then attempt should be made to obtain a sample via myringotomy for cytology and bacterial culture. Treatment of otitis media/interna consists of systemic antibiotic for a minimum of four to six weeks (oral amoxicillin/clavulanate, fluoroquinolone or cephalosporin if no culture can be obtained following myringotomy). Surgical drainage and debridement via bulla osteotomy should be considered in case of failure of medical treatment. Prognosis is guarded to fair as some animal may be left with permanent head tilt and/or facial paralysis.

Tumours of the caudal fossa and meningo-encephalitis of unknown etiology (MUE) such as granulomatous meningo-encephalitis (GME) are the two most common causes of central VD. Common types of tumours found in the caudal fossa include meningioma and choroid plexus tumour both which have a tendency to arise at the level of the emergence of the vestibulo-cochlear nerve at the cerebellomedullary angle. Less common tumours include glioma, ependymoma or medulloblastoma. Signs of VD associated with these tumours are often slowly
progressive. Presumptive diagnosis is made by advance imaging (CT or MRI) but the exact type of tumour can only be confirmed histologically (either by surgical tissue biopsy or post-mortem). Prognosis is fair for surgically accessible cerebellomedullary angle meningioma. Prognosis is more guarded for other tumors.

Meningo-encephalitis of unknown etiology (MUE) is often attributed to GME, the diagnosis of which can only be confirmed on histopathology. Clinical signs can be acute or progressive in onset. Neurolocalisation often suggests multifocal involvement but can occasionally be focal. A presumptive diagnosis can be made based on a consistent history, clinical signs, signalment (frequently young to middle-aged female terrier breeds), multifocal, contrast-enhancing lesions on MRI, CSF analysis (pure mononuclear pleocytosis or a mixed cell population) and exclusion of infectious aetiologies on serological or PCR tests (mostly Distemper, Toxoplasma and Neospora). Immunosuppressive doses of corticosteroids have been the mainstay of treatment for presumptive GME. Other immunomodulatory drugs such as azathioprine, procarbazine, cytosine arabinoside and cyclosporine as sole agent or as an adjunctive treatment with prednisone have been reported to be effective in some dogs. Overall, the prognosis is guarded but survival times range from weeks to years.

NEURODIAGNOSTIC INVESTIGATIONS OF VESTIBULAR DISORDER

The choice of neurodiagnostic tests in patient with VD depend essentially on where the lesion is suspected on the basis of the neurological examination. If in doubt about the localization of the lesion, the animal should be evaluated for both peripheral and central VD.

- Peripheral VD

Diagnostic plan for patients with signs suggestive of peripheral VD include at least otoscopic and pharyngeal examination, imaging of the tympanic bullae with radiographs, computed tomography (CT) or magnetic resonance imaging (MRI) and thyroid function testing. Middle ear pathology should be suspected if the tympanic membrane is ruptured, bulging, cloudy or red in colour on otoscopy. If the tympanic membrane is ruptured, swabs for cytology and culture (aerobic, fungal and yeast) can be taken directly from the middle ear. If the tympanic membrane is intact but bulging or of an abnormal colour, a small hole (myringotomy) can be made in the tympanic membrane with a 20-gauge spinal needle to obtain samples for cytology and culture. Additionally, the middle ear cavity can be flushed by attaching a 10 to 20 cc syringe of warm saline to the spinal needle. Warm water is flushed into the tympanic cavity and gently suctioned. The resulting fluid can then be submitted for cytology and culture. Radiographic evaluation of the tympanic bullae requires general anesthesia to allow adequate positioning. Four radiographic
projections are classically used (dorsoventral, latero-lateral, latero 20° ventral-laterodorsal and rostro 30° ventral-caudodorsal open-mouth projection). Although positive radiographs can be seen as highly specific in the diagnosis of middle ear disease, negative radiographs do not rule out the presence of middle ear disease. CT and MRI are more sensitive than radiographs in detecting middle ear pathology (Figure 10). Brainstem auditory evoked response test may be abnormal if the cochlea, vestibulocochlear nerve or auditory brainstem pathways are involved and can sometime be used to differentiate central from peripheral VD. Finally, electromyography (EMG) and motor nerve conduction study are indicated in patients suspected of multiple cranial nerve neuropathy or of a more diffuse polyneuropathy.

- Central VD

Evaluation of patients suspected of central VD include in first instance the use of advance imaging (CT or MRI), cerebrospinal fluid (CSF) analysis (nucleated cell count and cytology, total protein concentration), serum and CSF titers (serology and/or PCR) for various infectious organisms (toxoplasma gondii, neospora caninum, canine distemper, coronavirus, fungal agents...) (Figures 11 to 13). Further investigations may be require in cases suspected of brain tumour (tissue biopsy by surgical or stereotactic biopsy, thoracic and abdominal imaging to investigate metastatic disease), thiamine deficiency (urinary organic acids excretion screening or transketolase activity in fresh erythrocytes) or cerebrovascular accident (routine hematology and serum biochemistry, clotting profile, evaluation of arterial blood pressure, thyroid, kidney, adrenal and heart function).
VESTIBULAR SIGNS?  
HEAD TILT, NYSTAGMUS, POSITIONAL STRABISMUS, ATAXIA

SIGNS THAT CANNOT BE ATTRIBUTED TO PERIPHERAL VDz?  
ABNORMAL MENTATION, POSTURAL REACTION DEFICITS, PARESIS, VERTICAL OR VARIABLE NYSTAGMUS, INVOLVEMENT OF OTHER CRANIAL NERVES THAN CN VII OR VIII CEREBELLAR OR FOREBRAIN SIGNS?

YES  NO

METRONIDAZOLE administered?  
AMINGLYCOSIDES or TOPICAL CHLOREXIDINE administered?

EVALUATE FOR CAUSES OF CENTRAL VDz  
NORMAL  EVALUATE FOR CAUSES OF PERIPHERAL VDz

ADVANCED IMAGING (CT, MRI)  
CSF EVALUATION  
INFECTION DISEASE TITRES  
+- BAER

NO OTHER NEUROLOGICAL SIGNS AND NON-PROGRESSIVE  
IDIOPATHIC

BRAIN INFARCT  
BRAIN HEMORRHAGE  
INFLAMMATORY CNS DISEASE  
INFECTION CNS DISEASE  
HEAD TRAUMA  
ANOMALOUS  
BRAIN TUMOUR  
THIAMINE DEFICIENCY

OTITIS MEDIA/INTERNA  
EAR FOREIGN BODY  
POLYPS  
MIDDLE EAR TUMOUR  
HYPOTHYROIDISM  
INFLAMMATORY OR INFECTION POLY-NEUROPATHY

OTOSCOPIC EXAMINATION  
SKULL RADIOGRAPHS  
CT or MRI FOR EVALUATION OF MIDDLE/INNER EAR  
THYROID PROFILE  
ELECTROPHYSIOLOGY

10