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Neuromuscular Disorders Fact Sheet

An overview of the neuromuscular system in animals and common disorders affecting it.

What is the neuromuscular system?

The nervous system is divided into:

- The central nervous system (CNS) – composed of the brain and spinal cord
- Peripheral nervous system – also called the neuromuscular system and composed of nerves and muscles.

The neuromuscular system consists of the nerves leaving the back of the brain to innervate the muscles and glands of the head (cranial nerves) and peripheral nerves leaving the spinal cord to control, in particular, the muscles of the limbs. The junction between the peripheral nerves and the muscles is called the neuromuscular junction.

What is the role of the neuromuscular system?

The neuromuscular system is an essential part of any motor activity (standing up, walking, running, taking food, chewing or swallowing). The brain initiates any voluntary motor activity by sending command signals down the spinal cord to stimulate a peripheral nerve.

Each muscle of the body is controlled by its own peripheral nerve. Signals travel along the nerve as an electrical current. When the electrical nerve impulse reaches the end of the nerve, the signal must be conveyed across the gap between the nerve and the muscle (neuromuscular junction). A chemical messenger called acetylcholine bridges this gap. This messenger is released from the end of the nerve, flows across the gap and fixes itself to a receptor on the muscle causing muscle contraction and, by this way, motor activity.

What are the signs of a neuromuscular disorder?

Dogs or cats with a neuromuscular disorder present with varying degree of muscle weakness and muscle wastage. This muscle weakness may affect the limbs so that animals are unable to stand or exercise normally but also can affect other muscles in the body. The muscles of the pharynx or the oesophagus (the pipe carrying food from the mouth to the stomach) are weak in some types of neuromuscular disorder and this means that affected animals may have problems swallowing and will often bring back food after eating.

This weakness could be caused by a failure of the electrical nerve impulse to be conveyed by the nerve (disease of the nerve or neuropathy), failure of the chemical messenger to bridge the gap between the nerve and the muscle (disease of the neuromuscular junction or junctionopathy), or failure of the muscle to contract (disease of the muscle or myopathy).

In its mildest form, the muscle weakness may only be intermittent and triggered by exercise. This weakness might affect all four legs or only affect the back legs. It is frequently preceded by a short stride stiff gait with muscle tremors. In the most severe forms, neuromuscular disorders can cause the animal to be completely paralysed, unable to support its own weight, hold its head up and have breathing difficulty due to involvement of the muscles of the chest wall and diaphragm.

What diseases can affect the neuromuscular system?

A large number of diseases can affect the neuromuscular system. They can be divided into diseases directly affecting the various components of the neuromuscular system (i.e. peripheral nerves, neuromuscular junction or muscle) and diseases somewhere else in the body having an indirect effect on the function of the neuromuscular system.

The latter comprises hormonal diseases (also called endocrine diseases such as an under-active thyroid gland, diabetes, Cushing's disease, Addison's disease), cancer (causing low sugar level or the release of chemical toxic to the neuromuscular system), kidney disease (causing abnormal salt balance essential for proper muscle function), heart disease and lung disease.

Diseases which directly affect the various components of the neuromuscular system include:

- infectious diseases (particularly toxoplasmosis, neosporosis, botulism or tetanus)
- immune-diseases causing inflammation of the muscle (Myositis), inflammation of the nerve (neuritis or polyradiculoneuritis) or destruction of the acetylcholine receptor on the muscle (Myasthenia Gravis)
- toxic diseases
- inherited diseases
- and degenerative diseases.

How can neuromuscular diseases be investigated?

The first stage in the investigation of neuromuscular diseases involves eliminating diseases somewhere else in the body which may have an indirect effect on the function of the neuromuscular system. This may include doing a complete blood profile (including blood tests for hormonal and infectious diseases), X-rays of the chest and abdomen, as well as an abdominal ultrasound. If it's suspected that an animal may have a disease which affects the junction between the nerves and muscles, such as Myasthenia Gravis, a blood test which looks for antibodies directed toward the acetylcholine receptor (anti-AChR antibody titre) may be considered.

Other tests to re-enforce the suspicion of Myasthenia Gravis include a 'Tensilon test'. In this test a short-acting antidote to Myasthenia Gravis (Tensilon) is injected into a vein. In affected animals there will be a dramatic increase in muscle strength immediately after injection and collapsed animals may get up and run about. However, the effects will wear off after a few minutes.

A spinal tap is often considered at this stage of the investigation to look for inflammation of the root of the nerve (polyradiculoneuritis).

Finally, another test that can be used, not to identify the cause of a neuromuscular disorder but to confirm that we are looking at the right part of the nervous system, is an electromyogram (EMG). An EMG machine can be used to deliver a small electrical stimulation to an individual nerve or muscle in an anaesthetised animal. Using an EMG machine a vet can evaluate how well the muscles respond to stimulation from the nerves.

If the above preliminary tests fail to identify a cause, the next step is to take a biopsy of the nerve and muscle to determine if the problem is an inflammation or a degeneration of the nerve or muscle. This degeneration can be transient and of unknown origin (such as distal denervating disease or chronic relapsing demyelinating neuropathy), while others may be permanent and slowly progressive (inherited neuropathy or myopathy such as muscular dystrophy).

Can neuromuscular disorders be treated and what are their prognoses?

Treatment and prognosis of neuromuscular disorders directly depends on their primary cause. Some conditions carry a poor prognosis, such as neuromuscular disorders caused by cancer, inherited neuropathy or inherited myopathy. Other conditions have a good prognosis with adequate treatment (myositis, myasthenia gravis, distal denervating disease, under-active

thyroid gland and most forms of polyradiculoneuritis). It is important to be aware that, in the treatment of neuromuscular disorders, a quick fix is usually unlikely and some conditions (although carrying a good prognosis) can take weeks to improve.

Related Fact Sheets

Neuro-Diagnostic Tests Fact Sheet

If you are concerned about the health of your pet you should contact your veterinary surgeon.

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