MYASTHENIA GRAVIS

BASICS

OVERVIEW
- A disorder of signal transmission between nerves and muscles (known as "neuromuscular transmission"), characterized by muscular weakness and excessive fatigue

GENETICS
- Congenital (present at birth) familial (runs in families or lines) forms—Jack Russell terriers, English springer spaniels, smooth fox terriers; smooth-haired miniature dachshunds, autosomal recessive mode of inheritance
- Acquired (not inherited, present later in life/after birth)—as with other autoimmune diseases, requires appropriate genetic background for disease to occur; multiple factors involved, including environmental, infectious, and hormonal influences
- Familial (runs in families or lines) forms of acquired myasthenia gravis occur in the Newfoundland and Great Dane

SIGNALMENT/DESCRIPTION of ANIMAL

Species
- Dogs and cats

Breed Predilections
- Congenital (present at birth)—Jack Russell terriers; English springer spaniels; smooth fox terriers, smooth-haired miniature dachshunds
- Acquired (not inherited, present later in life/after birth)—several dog breeds: golden retrievers; German shepherd dogs; Labrador retrievers; dachshunds; Scottish terriers; Akitas and cat breeds: Abyssinian and Somali

Mean Age and Range
- Congenital (present at birth)—6 to 8 weeks of age
- Acquired (not inherited, present later in life/after birth)—bimodal age of onset; dogs: 1 to 4 years of age and 9 to 13 years of age

Predominant Sex
- Congenital (present at birth)—none
- Acquired (not inherited, present later in life/after birth)—may be a slight susceptibility for females in the young age group; none in the old age group (as described in ***Mean Age and Range***)

SIGNS/OBSERVED CHANGES in the ANIMAL
- Acquired (not inherited, present later in life/after birth)—may have several clinical presentations ranging from localized involvement of the muscles of the esophagus (the tube running from the throat to the stomach), muscles of the throat (pharynx), and muscles adjacent to the eye (known as "extraocular muscles") to acute generalized collapse
- Any dog with acquired megaesophagus (enlargement of the esophagus), loss of normal reflexes (known as "lower motor neuron weakness"), or a mass in the front central area of the chest (known as a "cranial mediastinal mass") should be evaluated for myasthenia gravis
- Regurgitation (return of food or other contents from the esophagus or stomach back up through the mouth)—common; important to differentiate between vomiting (forceful ejection of stomach contents up through the esophagus and mouth) and regurgitation
- Voice change
- Exercise-related weakness
- Acute collapse
- Progressive weakness
- Sleep with eyes open
- May look normal when at rest
- Excessive drooling, repeated attempts at swallowing
- Loss of muscle mass (known as "muscle atrophy")—usually not found
- Difficulty breathing (known as "dyspnea")—with aspiration pneumonia
- Fatigue or cramping—with mild exercise
- Subtle nervous system findings findings: decreased or absent blink reflex (known as "palpebral reflex"); may note a poor or absent gag reflex; spinal reflexes usually normal but may fatigue (rarely absent and dog unable to support its weight)
- Abnormal position of the neck (known as "ventroflexion")—cats, uncommon in dogs

CAUSES
- Congenital (present at birth)
- Immune-mediated disease
- Secondary to cancer (known as "paraneoplastic")

RISK FACTORS

- Appropriate genetic background
- Tumor or cancer—particularly thymus tumor (known as "thymoma")
- Methimazole treatment (cats)—may result in reversible disease
- Vaccination can exacerbate active myasthenia gravis
- Intact female

TREATMENT

HEALTH CARE

- Inpatient—until adequate dosages of drugs that inhibit acetylcholinesterase, an enzyme in the central nervous system (drugs are known as "anticholinesterase drugs"), are achieved
- Aspiration pneumonia—may require intensive care
- Feeding tube—may be required if patient is unable to eat or drink without significant regurgitation (return of food or other contents from the esophagus or stomach back up through the mouth)
- Oxygen therapy, intensive antibiotic therapy, intravenous fluid therapy, and supportive care—generally required for aspiration pneumonia
- Nutritional maintenance with a feeding tube—multiple feedings of a high-caloric diet; good hygiene care
- Elevation of food and water bowls

ACTIVITY

- Self-limited, owing to the severity of muscle weakness and extent of aspiration pneumonia

DIET

- May try different consistencies of food—gruel; hard food; soft food; evaluate what is best tolerated

SURGERY

- Cranial mediastinal mass—thymus tumor (thymoma)
- Before attempting surgical removal, stabilize patient with drugs (anticholinesterase drugs) that inhibit acetylcholinesterase, an enzyme in the central nervous system, and treat aspiration pneumonia
- Weakness may not be clinically evident initially
- Suspected thymus tumor (thymoma)—test all patients for acquired myasthenia gravis before surgery

MEDICATIONS

Medications presented in this section are intended to provide general information about possible treatment. The treatment for a particular condition may evolve as medical advances are made; therefore, the medications should not be considered as all inclusive.

- Drugs that inhibit acetylcholinesterase, an enzyme in the central nervous system (anticholinesterase drugs)—prolong the action of acetylcholine (a chemical that transmits messages between nerves and muscles) at the neuromuscular junction; pyridostigmine bromide syrup (Mestinon® syrup)
- Steroids—initiated if poor response to pyridostigmine or if no response to edrophonium chloride challenge
- Azathioprine, a chemotherapeutic drug used to decrease the immune response

FOLLOW-UP CARE

PATIENT MONITORING

- Return of muscle strength should be evident
- Chest X-rays—evaluated every 4 to 6 weeks for resolution of enlarged esophagus (megaesophagus)
- Acetylcholine receptor (AchR) antibody titers—evaluated every 6 to 8 weeks; decrease to the normal range with clinical remission

POSSIBLE COMPLICATIONS

- Aspiration pneumonia
- Breathing may stop (known as "respiratory arrest")

EXPECTED COURSE AND PROGNOSIS

- No severe aspiration pneumonia or weakness of the throat (pharynx)—good prognosis for complete recovery; resolution usually within 6 to 8 months.
- Thymus tumor (thymoma) present—guarded prognosis, unless complete surgical removal and control of myasthenic signs are achieved

KEY POINTS

- Although the disease is treatable, most patients require months of special feeding and medication
- A dedicated owner is important to a favorable outcome for acquired myasthenia gravis