SCOTTISH TERRIER CLUB OF AMERICA

Hyperkinetic Episodes In Scottish Terrier Dogs

By K. M. Meyers, Ph.D.; J. E. Lund, D.V.M., Ph.D.; G. Padgett, D.V.M.; W. M. Dickson, D.V.M., Ph.D.
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From the Department of Physiology and Pharmocology and the Department of Pathology, School Of Veterinary Medicine, Washington State University, Pullman, Wash. 99163. Dr. Lund's present address is Animal Care Facility, Stanford University School of Medicine, Palo Alto, Calif. 94304.

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SUMMARY
"Scottie cramp" can be elicited by excitement, exercise, and amphetamine sulfate* (0.5 to 2.0 mg/kg). Chlorpromazine** (1.0 to 1.75 mg/kg), acepromazine+ (0.075 to 0.1 mg/kg), and diazepam# (0.50 to 1.50 mg/kg) when injected intramuscularly were effective in suppressing the signs of the disease.

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SCOTTIE CRAMP IS a descriptive term for a hyperkinetic disorder which has been observed for some years in Scottish Terriers by kennel owners and veterinarians. Recent studies have demonstrated the disease to be a central nervous system (CNS) disorder which results in profound locomotion and postural signs. The purpose of this report is to describe the disease, criteria for its diagnosis, and methods of treatment.

Materials and Methods

Ten Scottish Terriers considered to have scottie cramp by their owners were sent to Washington State University from various locations in the United States and Canada. The tentative diagnosis was confirmed by clinical, physiologic, and pharmacologic means.
The electromyogram (EMG) was obtained by photographing the telemetered bioelectric activity of the biceps femoris muscle of the right hindlimb while the dog was walking.

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*Amphetasul, Pitman-Moore Company, Indianapolis, Ind.
**Thorazine, supplied by Smith, Kline & French Laboratories, Philadelphia, Pa.
+Acepromazine, Ayerst Laboratories, Incorporated, New York, N.Y.

#Valium, supplied by Hoffmann-LaRoche, Incorporated, Nutley, N.J.

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Clinical Description

Hyperkinetic episodes could be elicited by normal physical activity or by psychic changes. The dogs appeared normal when at rest or when first exercised, but with continued exercise the signs of the disease became apparent. The time interval and intensity of exercise required to elicit signs of the disease were variable. We have observed signs after walking the dog as little as 10 yd. (9.1 m.). According to one report, the distance varied from 100 yd. (91.4 m.) to 3/4 of a mile (0.69 km.).[9] The psychic state of the dog was found to be more important than the amount or intensity of exercise. Excitement and fear facilitated the hyperkinetic episodes, whereas anxiety and apprehension were often inhibitory.

Amphetamine sulfate (0.5 to 2.0 mg./kg.) elicited a hyperkinetic episode within 15 minutes when injected intramuscularly. The clinical signs were indistinguishable from those observed after exercise or excitement.

During exercise, the onset of a hyperkinetic episode was usually indicated by a slight abduction of the front legs, resulting in an arclike motion of the limbs while extending. The back then became arched in the lumbar region, and the hind legs were quickly overflexed and then swiftly returned to the ground. This motion has been appropriately described as a "stringhalt" gait.[8] The front legs became increasingly stiff, and while walking were quickly extended then flexed. In rare cases where the back was not arched, the dog walked with a "goose step" gait. Forward movement was usually hindered, and in severe cases was completely absent, resulting in the dog walking in place. Facial muscles did not appear to be affected at this time; the dogs had free jaw movement and were often seen panting, with the tongue hanging freely. The head tended to be extended, with the nose pointed downward; at times, the nose would be pulled toward the ground. The tail was often flexed. occasionally, when the younger dogs were running, the hindquarters would suddenly and strongly become elevated, often to such a degree that the dog somersaulted. If the inducing stimulus was continued, the hind legs became increasingly resistant to flexion, which finally resulted in a pillarlike stance, with the dog unable to walk. If the dog fell down, it would curl into a ball with its head, limbs, and tail tucked in; breathing would appear to cease. The severe seizure would last approximately 15 seconds, after which the dog appeared relaxed and panting. During or just preceding a severe episode, the facial muscles were often affected, and the dog was unable to open its jaws. None of the dogs lost consciousness during an episode, nor did they appear to be in pain. A short period of rest would alleviate the hyperkinetic episode in most dogs, but the signs would quickly reappear if the inducing factors were not eliminated.

Considerable variation was found in the frequency and the severity of the hyperkinetic episodes in the same or in different dogs. In some dogs, signs were hardly noticeable; others were nearly incapacitated. A perplexing feature of the disease was the variation in severity of signs in a given dog. The dog would have severe signs after a short period of exercise or excitement for a period of weeks or months, then suddenly, or over a period of days, it would become much more difficult to elicit signs of the disease. The signs, when manifested, were usually milder and more transient than those observed during the period when the threshold for eliciting the hyperkinetic episode was lower. We have not observed complete and permanent recovery in any dog we have studied. Environmental temperature may have an inverse relationship with the signs of the disease, but if so, it is certainly not the only factor involved. Except for the variations mentioned, the severity of the manifestations appeared to remain relatively constant in dogs personally observed. However, 2 Scottish Terrier breeders told us that they observed a steady increase in the severity of the sings as their dogs aged.

The age at which the disease first became apparent to the dog's owner was variable, but the clinical signs were usually noticed before the dog was 18 months old. In the dogs observed in our laboratory, signs were noticed from 6 weeks to 18 months of age. According to one report, an affected dog was 3 years old before the disease was first noticed.[20] The variation might be explained by the owner's unfamiliarity with the signs of the disorder. The degree of affliction and frequency of episodes are certainly factors which would alter the age at which the disease would first be recognized. A review of the kennel records of several Scottish

Terrier breeders did not reveal a high death loss during puppyhood or a decreased longevity in the dogs with the disease. The clinical signs observed in our dogs were similar to those described previously.[8,9,20]

Treatment

Since it has been demonstrated that the hyperkinesis was of CNS origin, various drugs which act upon the CNS were evaluated for their therapeutic value.

Chlorpromazine had been shown to decrease classical Sherrington decerebrate rigidity and to antagonize some of the actions of amphetamine.[5,7,11] Because of the appearance of the affected dogs and their response to amphetamine, chlorpromazine was given. Five dogs were given chlorpromazine, 1.0 to 1.75 mg./kg. of body weight, intramuscularly during a hyperkinetic episode. A complete remission of signs of the disease was observed within 15 minutes after the injection. Some sedation was apparent at this dose level. Acepromazine maleate, a phenothiazine derivative and a commonly used tranquilizer, effectively eliminated signs at a dose of 0.1 to 0.75 mg./kg.

Diazepam, a benzodiazepine derivative used extensively as an antianxiety drug, has certain CNS muscle-relaxant properties23 that suggested it might be beneficial.

While 6 dogs were in a hyperkinetic state, induced either by exercise, excitement, or amphetamine, diazepam, 0.50 to 1.5 mg./kg. of body weight, was injected intramuscularly. Prompt cessation of signs occurred, and a normal-appearing EMG resulted. Hyperkinesis could not be induced by any means when the affected dogs were given an oral dose of diazepam, 0.5 mg./kg. t.i.d. When treatment was reduced to twice daily (mornings and evenings), mild signs occurred before the evening administration. We have had dogs on daily treatment for 1 month without apparent side effects. The psychological state of the dogs was not affected. The increased activity induced by amphetamine administration was not hindered. The dogs were not depressed, and their aggressiveness toward rodents was not modified.

Vitamin E as alphatocopherol acetate has been reported to have therapeutic value in high doses.8 A Scottish Terrier breeder reported that large daily doses of vitamin B complex produced improvement within 2 weeks of treatment. In this laboratory, an 8 month old male dog that was mildly affected (i.e., the episodes were easily precipitated, but their duration was extremely short) was given vitamins once a day for 2 weeks without improvement. Treatment consisted of vitamin A, 5,500 IU/lb.; vitamin D2, 883 IU/lb.; vitamin E, 0.55 IU/lb.; thiamine, 0.33 mg./lb.; riboflavin, 0.33 mg./lb.; pyridoxine HCl, 0.138 mg./lb.; vitamin B12, 0.111 ug./lb.; calcium pantothenate, 0.555 mg./lb.; desiccated liver, 16167 mg./lb.; and brewer's yeast, 8.33 mg./lb.

Discussion

There are many diseases in man and animals which affect the musculature and produce locomotion and postural problems. The diseases in man result from a wide variety of defects. McArdle's disease[12,16,19] results from a deficiency in myophosphorylase. The affected persons have electrically silent muscle cramping after exercise. Persons with myotonia congenita have painless tonic muscle contractions which may be produced by voluntary movement. The muscle spasms appear to be the result of an abnormal myomembrane and are not affected by curare or nerve block.[24,10,11] A disease in 2 siblings was characterized by painful muscle spasms induced by attempted movement.[18]The spasms became milder and less frequent with continued muscular activity. The stiffman syndrome[14,15,21,22] is a neurologic disorder wherein muscle spasms are precipitated by various sensory inputs and are characterized by constant electrical activity even

at rest. The disease was reported to be effectively controlled by diazepam.[6] A familial myopathy has been characterized by mild proximal muscle weakness and painless muscle cramp following exercise.1 In the center of 75% of the skeletal muscle fibers from biopsy specimens, a zone of abnormal myofibrils was found which stained blue with Mallory's aniline blue and orange G stain. The outer fibrils stained a normal orangered.

Transient hyperkinesis in Scottish Terriers does not appear to have a direct human counterpart. The muscular hypertonicity appears to be the result of a CNS rather than a muscle defect. The muscle contractions are inhibited by curare and nerve block.[13] There were no histopathologic lesions in biopsy and postmortem muscle sample we have examined. The condition resembles most closely the condition in man of painful muscle spasms18 and the stiffman syndrome. It differs from the former in that the muscular hypertonicity does not decrease with continued exercise and, unlike the latter, the electrical activity is transient rather than continuous.

A spastic syndrome in cattle[17] has many clinical signs similar to the Scottish Terrier disease. It is a CNS disorder characterized by spastic muscle contractions, particularly noticeable in the muscles of the back and hindquarters when the animal tries to stand or move.

A positive diagnosis of scottie cramp in a Scottish Terrier required the fulfillment of 3 criteria: (1) The clinical history should indicate an abnormality in gait or seizures during excitement; (2) an injection of amphetamine should induce transient hyperkinetic episodes; and (3) the administration of diazepam or promazine derivative during a natural or induced seizure should produce prompt remission. Diazepam has qualifications for a practical suppressant. It is fast acting, produces complete cessation of clinical signs, does not appear to have adverse side effects, does not depress the animal, and can be administered orally. The only disadvantage is the relative short duration of action. The dosage of either diazepam or phenothiazine derivatives depends upon the severity and frequency of seizures and should be separately established in each case. The dosages given in this report ar for fairly severe cases and are intended to serve as a basis for establishing individual treatment.
If amphetamine is to be administered, the induced psychic stimulation should be counteracted by one of the phenothiazine derivatives.

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