

SCRAPIE

by

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CLINICAL CASE

A two-year-old Suffolk ram with clinical signs of scrapie was admitted to the Iowa State Veterinary Clinic on August 19, 1964. The ram came from a flock of Suffolk sheep on a farm near Clemons, Iowa. He had been purchased from the Iowa Falls area. The owner first noticed symptoms around July 1, 1964. He said the ram "seemed to have a fever and did a lot of scratching." The ram was the only one in the flock affected. He was isolated immediately and the farmer was instructed by a veterinarian to wash the ram with an aqueous solution of Ivory Flakes every few days. The flock was quarantined on July 28th for a period of six months or until final diagnosis was completed. Further regulations will depend on the final diagnosis.

INTRODUCTION

Scrapie is a chronic, slowly progressive, non-febrile, degenerative disease of the central nervous system. It is characterized by pruritus, locomotor disturbances, hypersensitivity or marked dullness, and a long incubation period (1, 13, 15, 17).

The disease has been known in Europe for over 200 years. It is enzootic in Britain and Europe with smaller outbreaks in Australia, New Zealand, Canada, and the United States. It was first introduced into Canada in 1938, the United States in 1947, New Zealand in 1952, and just recently, India. The outbreaks were usually traced to imports from countries

where the disease was enzootic. This has led to embargoes by several countries against sheep from these areas (1, 13, 16, 17).

The morbidity ranges up to 20%, but cases of 40% have been reported. The mortality can generally be considered to be 100% (1).

The following chart will show the situation in the United States in 1962 and 1963.

ETIOLOGY

The cause of scrapie is not yet fully understood but recent work indicates that it is a hardy virus that is transmitted by contact to susceptible sheep. The susceptibility or resistance of individual sheep is controlled by heredity (1, 5, 17, 20).

Due to complications in isolating and identifying the specific virus and the high percentage of occurrence in the Suffolk breed, much work has been done in the last few years (9). At first it was hypothesized that "scrapie is controlled by an autosomal recessive gene" (7, 16). Parry stated that "scrapie is an example of balanced polymorphism, unconsciously favored by current methods of selection which allow the frequency of the gene to reach very high levels. It arises by a dual mechanism of gene and provirus. Scrapie falls into a special category of genetic disorder, in which the physiological action of the gene is mediated by a specific particle with independent powers of self-replication and pathogenicity" (16). Practically all of the experimental work to determine the hereditary significance was carried out with mice (3, 6, 12, 19).

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Sheep showing cutaneous lesions associated with scrapie.

Latest work indicates a virus which is probably contagious and does not support the simple recessive gene hypothesis (11, 20). Hourrigan states that the heritable factor is probably related to variable resistance and susceptibility (11), while Dickinson states in very recent work that "the concept of absolute resistance is questionable" (6).

TRANSMISSION

Except for one case in a goat under natural conditions, scrapie is a disease exclusively of sheep (1). For experimental purposes, scrapie has been transmitted to mice, hamsters, rats, and goats (3, 4, 18).

The virus is probably transmitted directly by contact with infected sheep or premises and indirectly by ingestion of contaminated water (2). Accidental transmission through Louping-ill vaccine has been exhibited (1).

Draper suggests that the heredity component is paramount but not dependent entirely on either parent, and that the environmental or infectious component is non-existent or very small (7).

Chandler favors a combination of ideas by saying that the infection is not likely

to be due purely to genetic factors. The ewe is the most important factor in the transmission (2).

Presently, experiments are being performed to determine genetic and congenital transmission, the presence of neutralizing antibodies, and immune tolerance (14).

PATHOLOGY

The most spectacular lesion noted is the vacuolation of neurons in the spinal cord, medulla, pons, and mid-brain (20). This lesion was also noted in laboratory animals (4, 17, 20). Muscular lesions were often noticed but no direct correlation was seen (13). The neurological lesions produced three forms of the disease in mice. They are: a) hyperexcitability, b) obesity, and c) lethargy (21). The course is very prolonged and always exceeds 30 days with death occurring in 2-6 months (13).

CLINICAL FINDINGS

The incubation period is long, usually over 18 months (13). The first signs are transient nervous phenomena occurring under periods of stress. This is manifested

by loss of wool due to rubbing, scratching, biting and gnawing, with sudden collapse and behavioral changes seen (1, 13). Loss of wool is bilateral and most apparent over the shoulders, thighs and base of tail (1).

Other clinical findings are hyperexcitability, incoordination, exaggerated flexion, abnormal gait with the hind legs moving together and the front legs at a trot, slight tremors of the head and neck, rapid muscular tremors of the thighs and flanks, loss of weight, dry lusterless wool, change in voice, loss of bleat, blindness, and lagging behind the rest of the flock. Abnormal posture or position of the head is common, especially when responding to the "scratch reflex". This is initiated by scratching the back and results in a nibbling motion of the lips, rapid viper-like movements of the tongue, and wiggling of the tail. Anorexia is not evident until the latter stages when extreme emaciation and fatigue are noted. Sternal and lateral recumbency follow. Pyrexia is never evident (1, 12, 15).

Scrapie is seen in sheep 1¼ to 11 years old (4½-mean). It is found in variable incidence in many improved breeds (16), but has also been artificially transmitted to grade lambs (9). Male and female are affected equally.

NECROPSY

Although both neural and muscular lesions have been seen, significance is placed only on the neural lesion; however, a thorough examination of the entire carcass should be undertaken and recorded. Gloves should be used and they, along with the instruments, should be heat sterilized following necropsy.

If a brain specimen is sent to Diagnostic Service Section of NADL, address it as follows: (13)

Chief Veterinarian
Diagnostic Service—NADL
P. O. Box 70
Ames, Iowa

TREATMENT AND CONTROL

There is no known treatment. Flocks into which sheep have been moved from

infected flocks within a 42 month period are quarantined. Sheep from the infected flock and their progeny are slaughtered. The remainder are observed for 42 months and inspected every six months (15).

DIAGNOSIS

A positive diagnosis can only be made on the basis of positive necropsy findings or transmission experiments (1).

A tentative diagnosis can be made by pruritis and locomotor disturbances. The veterinarian should examine the skin microscopically by a skin scraping to rule out cutaneous acariasis.

Scrapie should be differentiated from listeriosis, pregnancy disease, pseudorabies, rabies and parasites (13). Kuru is a human disease giving similar cellular changes (10). History or the presence of other significant lesions may refute a scrapie diagnosis. Edema is usually the only microscopic lesion in the brain tissue of parasitized animals that die (13).

Disturbances in locomotion and coordination are detected and evaluated by making a small group, including suspected cases, walk backwards, sideways, over obstacles, and around sharp turns. The "scratch reflex" may be absent in scrapie or present in other diseases.

If scrapie is suspected, contact state or federal veterinary regulatory officials. If the animal recovers, it is not scrapie but must be inspected from time to time for 6 months due to the possibility of remission. When possible, refer them to the State Diagnostic Laboratory or College of Veterinary Medicine for observation and necropsy (13).

Changes in the brain stem consist of vacuolated neurons, progressive degeneration of neurons, astrocytosis, and vacuolation of the neuroparenchyma imparting a spongy appearance.

Vacuolated neurons are characteristic if not pathognomonic for scrapie. These appear as large holes in the cytoplasm. The Nissl substance, nuclei and other intracellular components are often pushed to one side of the cell body. These vacuoles are often found in clumps and may have eosinophilic round bodies in them (50 vacuoles per section).

In addition, neurological degeneration is characterized by pyknosis and chromatolysis (tigrolysis).

Confirmation of clinical diagnosis of scrapie is possible when clinical manifestations and duration indicate scrapie, and when vacuolation and degeneration of neurons in the brain stem is 30 or more per section, and a total for 54 sections of the medulla is over 400.

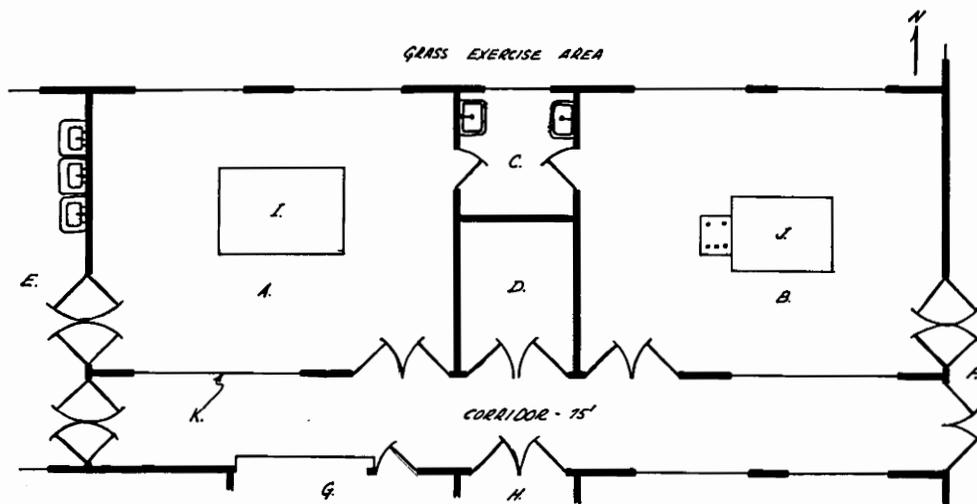
The work of Zlolnik is an excellent guideline for the neurological examination (13).

SUMMARY

Scrapie is a chronic debilitating disease of the central nervous system of sheep. It is probably caused by a virus transmitted to hereditarily susceptible sheep. Long incubation and duration are characteristic, as are the slowly progressing nervous symptoms. Being a fatal disease with no treatment, the infected flock and their progeny are slaughtered. A positive diagnosis can only be made by positive microscopic findings or transmission experiments.

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LARGE ANIMAL SURGERY AREA. A. Equine surgery room, B. Bovine surgery room, C. Surgical scrub room, D. Tack room, E. Equine treatment area, F. Corridor to Bovine stalls, G. Dispensary, H. Case record hall, I. Hydraulic surgery table (recessed), J. Hydraulic tilting surgery table, K. Observation window.