**Feline Spongiform Encephalopathy**

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**Importance**

Feline spongiform encephalopathy (FSE) is a neurodegenerative disease, caused by a prion, that affects members of the cat family. Once the symptoms appear, this disease is invariably fatal. FSE is caused by the same agent that is responsible for bovine spongiform encephalopathy (BSE) in cattle. BSE was first reported in the 1980s, when it caused an explosive epidemic among U.K. cattle. This disease eventually spread to many other countries. FSE was first reported in 1990, and was apparently transmitted to individual cats in BSE-contaminated food. As the BSE epidemic has declined, and controls have been placed on feeding high-risk bovine tissues to animals, FSE has become increasingly rare. However, this disease has a long incubation period and occasional cases continue to be reported in housecats and zoo animals.

**Etiology**

FSE is a member of the transmissible spongiform encephalopathies (TSEs), a group of neurodegenerative disorders caused by unconventional disease agents. These agents are resistant to the treatments that ordinarily destroy bacteria, spores, viruses and fungi. They are generally thought to be prions, although a minority opinion suggests that TSEs may be caused by virinos or retroviruses. Prions are infectious proteins that appear to replicate by converting a normal cellular protein into copies of the prion. The cellular protein, which is called PrP\(^c\), is found on the surface of neurons. Pathogenic isoforms of PrP\(^c\) are designated PrP\(^\text{Sc}\), PrP\(^\text{TSE}\) or PrP\(^\text{FSE}\). Prions that cause different diseases (e.g. FSE or scrapie) are considered to be different strains of PrP\(^c\). FSE is caused by the same agent that is responsible for BSE in cattle (see the BSE factsheet for additional details on this disease).

One TSE in a housecat, reported in 1998, was caused by a prion that was distinct from BSE. The authors suggested that this may have been a new type of FSE. No other infections with this prion have been reported in cats.

**Species Affected**

FSE has been found in domesticated cats (housecats) and captive wild cats including cheetahs, pumas, ocelots, tigers, lions and Asian golden cats.

**Geographic Distribution**

FSE has been found in countries where BSE occurs, and in animals imported from these countries. Most cases have been seen in the United Kingdom. In addition, a few infected housecats have been found in Norway, Switzerland, Northern Ireland and Liechtenstein, and infected zoo cats have been reported from Australia, Ireland, France and Germany. Most of the cases in zoo animals occurred in cats that had lived in the U.K., but one cheetah had been born in France, and another is thought to have been infected in the Netherlands.

FSE has not been documented in the U.S., where only three cases of BSE have been reported in cattle as of 2007.

**Transmission**

The BSE prion is thought to be transmitted to cats when they ingest contaminated bovine tissues. Cooking or rendering does not destroy this agent. Horizontal transmission has not been reported between cats.

The origins of BSE are unknown, but the cattle epidemic occurred when prions were amplified by recycling tissues from infected cattle into ruminant feed supplements. Banning ruminant tissues from ruminant feed significantly reduced the number of new cases of BSE, and has controlled the epidemic in cattle. In addition, tissues that have a high risk of transmitting BSE have been banned from pet foods in the U.K. since 1990, and are no longer fed to zoo cats. The vast majority of FSE cases have occurred in cats born before this ban; however, some cases have been seen in housecats born after the ban.
Clinical Signs

In housecats with FSE, the symptoms develop gradually. The first signs are usually behavioral changes such as uncharacteristic aggression, or unusual timidity and hiding. Gait abnormalities and ataxia are also characteristic; these defects initially affect the hindlegs. Affected cats often display poor judgment of distance. Some cats develop a rapid, crouching, hypermetric gait. Hyperesthesia is common, particularly when cats are stimulated by sound or touch. Some cats may have an abnormal head tilt, develop tremors, stare vacantly or circle. Excessive salivation, decreased grooming, polyphagia, polydypsia and dilated pupils have also been reported. In the late stages of the disease, somnolence is common and convulsions may occur. Similar symptoms have been reported in zoo cats. Once the symptoms of FSE appear, this disease is relentlessly progressive and fatal. Death occurs after 3 to 8 weeks in housecats, and 8 to 10 weeks in cheetahs.

Post Mortem Lesions

No gross lesions are found in cats with FSE. The typical histopathologic lesions are confined to the central nervous system. Neuronal vacuolation and non-inflammatory spongiform changes in the gray matter are pathognomonic.

Morbidity and Mortality

The number of FSE cases has paralleled the BSE epidemic, and declined as this epidemic has been controlled. The BSE epidemic peaked in the U.K. in 1992, but the peak of the epidemic curve occurred later in countries where feed bans were established more recently. As of September 2007, nearly a hundred cases of FSE have been diagnosed in housecats worldwide. Most of these cats were four to nine years old, but cats as young as two years have been affected. Eighty-nine cases of FSE were diagnosed in housecats in the UK, and five cases have been reported outside the U.K. The two most recent cases occurred in a cat from the U.K. in 2001, and a Swiss cat in 2003. If FSE was underdiagnosed or underreported, the true incidence may have been higher. Some sources estimate an annual incidence, at the height of the U.K. epidemic, of 10-15 cases per million cats. However, in a recent survey of clinically suspect cases of FSE (mainly from the U.K.), none of 192 cats had histopathological evidence of this disease, and prions were found in only one of 173 cases examined for these proteins. A similar retrospective study revealed no evidence of FSE in 286 cats that died of neurological disorders before 1990.

As of September 2007, 22 cases of FSE had been confirmed in zoo cats. The most recent case in a cheetah was reported in 2007.

FSE is always fatal once the symptoms appear.

Diagnosis

Clinical

FSE may be a consideration in cats that develop a progressive, fatal neurologic disease. Behavioral changes and ataxia are the most common symptoms.
Differential diagnosis

Other neurologic diseases must be ruled out. The differential diagnosis includes neoplasia, inflammatory disorders such as feline infectious peritonitis, congenital lesions, trauma, metabolic diseases and toxins.

Laboratory tests

FSE is usually diagnosed by detecting prion proteins (PrP\textsuperscript{res}) in the CNS by immunoblotting or immunohistochemistry. The diagnosis can also be confirmed by finding characteristic prion fibrils (called scrapie-associated fibrils) with electron microscopy in brain extracts. Histological examination of the brain is also very helpful, but some animals in early stages of the disease may have few or no spongiform changes. Serology is not useful for diagnosis, as antibodies are not made against prions.

Samples to collect

FSE is generally diagnosed from brain samples; the laboratory should be contacted for advice before collecting samples. No live animal test is currently available.

Recommended actions if feline spongiform encephalopathy is suspected

Notification of authorities

FSE is an exotic disease and should be reported promptly to state or federal officials.

Federal: Area Veterinarians in Charge (AVIC):
http://www.aphis.usda.gov/animal_health/area_offices/
State Veterinarians:

Control

FSE can be prevented by not feeding bovine tissues that may contain prions to cats. Complete avoidance is generally necessary, as cooking or rendering cannot completely inactivate these agents. Tissues that have a high risk of transmitting BSE, such as the brain and spinal cord, have been banned from pet foods in the U.K. since 1990. These tissues are no longer fed to zoo cats. Controlling BSE in cattle also reduces the risk to cats.

Although horizontal transmission of FSE has never been reported, other prions can be transmitted between some species iatrogenically. Human prion diseases have been transmitted in blood transfusions or by surgical instruments. Decontamination of prion-contaminated tissues, surfaces and environments is difficult. These agents are highly resistant to most disinfectants (including formalin), heat, ultraviolet radiation and ionizing radiation, particularly when they are protected in organic material or preserved with aldehyde fixatives, or when the prion titer is high. Prions can bind tightly to some surfaces, including stainless steel and plastic, without losing infectivity. Prions bound to metal seem to be highly resistant to decontamination. Few effective decontamination techniques have been published. A 1-2 N sodium hydroxide solution, or a sodium hypochlorite solution containing 2% available chlorine, has traditionally been recommended for equipment and surfaces. Surfaces should be treated for more than 1 hour at 20°C (68°F). Overnight disinfection is recommended for equipment. Cleaning before disinfection removes organic material that may protect prions. Recently, milder treatments including a phenolic disinfectant, an alkaline cleaner (KOH with detergents), and an enzymatic cleaner combined with vaporized hydrogen peroxide have been shown to inactivate scrapie prions. The alkaline cleaner and phenolic disinfectant were also effective against BSE prions. These disinfectants may be useful for items that cannot withstand harsher decontamination procedures. Physical inactivation of prions can be carried out by porous load autoclaving at 134-138°C (273-273°F) for 18 minutes at 30 lb/in2. Autoclaving items in water is more effective than autoclaving without immersion. Dry heat is less effective; hamster-adapted scrapie prions can survive dry heat at temperatures as high as 360°C (680°F) for an hour. A combination of chemical and physical decontamination can be more effective than either procedure alone; chemical disinfection should be carried out first, then the items should be rinsed and autoclaved. However, even the harshest combination of chemical and physical disinfection is not guaranteed to destroy all prions. For this reason, disposable instruments are often used during high-risk procedures (e.g. brain surgery) in humans with prion diseases.

Because prions may be able to survive in the environment for years and are difficult to disinfect, precautions should be taken to avoid contamination of surfaces and equipment during necropsies. Disposable plastic-coated paper sheets can be used to protect tables and other surfaces. Disposable instruments and work clothing can also be used.

Public Health

Although eating BSE-contaminated tissues can cause a fatal TSE (variant Creutzfeldt-Jakob disease) in people, there is no indication that humans have ever acquired this disease from cats. Nevertheless, precautions are advisable when conducting necropsies on FSE-suspects or handling tissues, particularly high-risk tissues such as the CNS. Standard general precautions for prion-related work include the use of protective clothing and the avoidance of penetrating injuries, contamination of abraded skin, and ingestion. A negative pressure laminar flow hood should be used for tissue manipulations whenever possible.

Spongiform encephalopathies were reported simultaneously in a cat and its owner in 1998; however, the man was found to have the sporadic form of Creutzfeldt-Jakob disease, rather than the BSE-associated form, and the disease in the cat differed clinically from FSE. The prions isolated from both man and cat appeared to be similar, but differed from the BSE prion. It is not known whether these prions might have been transmitted between the man and the
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cat, whether both contracted the disease from a common source, or if the incident was due to chance. No other infections with this prion have been reported in cats.

Internet Resources

U.K. Department for Environment Food and Rural Affairs. Other TSEs

World Organization for Animal Health (OIE)
http://www.oie.int

References


U.K. Department for Environment Food and Rural Affairs
[DEFRA] TSE- Statistics. DEFRA; 2007 Sept. Available at:


* Link defunct as of 2007