

FAQ

Atypical Bovine Spongiform Encephalopathy (BSE)



What is atypical BSE?

In addition to classical BSE, which belongs to the transmissible spongiform encephalopathies (TSE) and causes fatal disease characterized by sponge-like changes in the brain of cattle, cases of atypical BSE are seen. While classical BSE is caused by feeding of insufficiently heated ruminant fats and proteins containing the pathogenic prion protein to cattle, rarely spontaneous cases of atypical BSE are seen in older animals.

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There are two types (H and L) of atypical BSE, which differ from each other and from classical BSE with regard to their biological properties and the biochemical characteristics of the pathogenic prion protein. So far, cases have only been observed in animals aged eight years or older.

In the H-type the molecular mass of degradation products of the prion protein is slightly higher than in classical BSE, thus the designation “H” for “high”, while in contrast the L-type is characterized by lower molecular masses (“L” for “low”).

Where does atypical BSE come from?

The worldwide distribution of atypical BSE cases, even in countries where previously no cases of classical BSE have been reported, and the fact that the disease only occurs in older animals support the assumption that this extremely rare disease develops spontaneously. This is similar to the spontaneously occurring cases of Creutzfeldt-Jakob disease in humans, which is also caused by prions. Therefore, it must be expected that single cases of atypical BSE in cattle will continue to occur in the future, which are not associated with feeding of infectious animal products.

Is atypical BSE infectious?

Although it is assumed that atypical BSE occurs spontaneously, it can be transmitted in animal experiments, and model animals for the human species barrier could also be infected. However, a possible etiological association between classical BSE and atypical BSE remains unclear.

In which countries has atypical BSE occurred so far?

So far (last updated February 2023) 145 cases of atypical BSE (H- and L-type) have been detected worldwide, 125 of these in fourteen member states of the European Union, with France and Poland being the most affected. In addition to the countries of the European Union, the United States of America, Canada, and Japan were also affected. Both types were predominantly detected in animals older than eight years.

In Germany, atypical BSE so far has been detected in five animals. H-type BSE was observed in 2004 in a 13-year-old and in 2014 in an 11-year-old beef cattle. The three L-type cases were detected in 2002 in a 15-year-old beef cattle, in 2014 in a 10-year-old beef cattle, and in 2021 in a 14-year-old beef cattle. In addition, in 2012, an H type case was detected in a cattle born in Germany and subsequently exported to Switzerland.

Which cattle are tested for BSE?

In the EU, BSE testing is still carried out as part of *active surveillance*. This includes samples from cattle older than 48 months that show a BSE risk. These can be animals that have been emergency slaughtered due to injury, animals with a pronounced suspicion of BSE, and cattle that have died or been killed due to other causes and are disposed of in a rendering facility.