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**Editor's Viewpoint**

# Atypical BSE in Brazil and Worldwide

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In December 6th 2012 the MAPA - Ministério da Agricultura, Pecuária e Abastecimento of Brazil- announced the existence of a case of cattle with positive prionic immunohistochemistry (2). The cow, 13-year-old, of the Nelore breed, fed on pasture system had a sudden death. No signs described for BSE were noted before. Histopathological examination showed no spongiform or other type of lesions in the Central Nervous System (CNS). Exam for rabies was also negative. On the other hand, immunohistochemistry (IHC) was positive por $PrP^{res}$. This case is compatible with atypical BSE, a variant phenotype of the BSE as reported by authors in Europe and the USA (1, 3, 4).

Atypical BSE was first described in 2004 in Italy (4) as a novel molecular phenotype in cattle, the L type, in contrast with the C (classic) type of the typical BSE. During the same year a second atypical BSE variant, the H-type, was reported in France (1). Based on Western Blot analysis L-type demonstrates faster electrophoretic mobility of $PrP^{Sc}$ unglycosylated moiety. This type is also called bovine amyloidotic spongiform encephalopathy (BASE) because the presence of amyloid plaques. The H-type cases were characterized by a higher molecular weight of the unglycosylated fraction of $PrP^{Sc}$ than C-type BSE. It is acceptable that both types of atypical cases are not originated from the BSE prion that caused the epidemic outbreak in the UK in the decades of 1980 and 1990. The occurrence of atypical cases is supposed to be sporadic (6).

Intensive active surveillance has uncovered atypical cases in Germany in 2006 (3) and in the USA (7, 8), all H-type. The US BSE surveillance program focus on high-risk cattle defined as animals displaying clinical signs consistent with BSE or fallen stock. US surveillance has detected three BSE cases: one C-type and two H-type. (8) A molecular study performed with cases of BSE diagnosed in Poland from 2002 to 2006 revealed six L-type BSE and one H-type. All animals were above 9-year-old. None of these cases was reported to have clinical signs.

Three cases of H-type were reported in the UK (9). Canada, Denmark, Ireland, Netherland, Sweden and Switzerland have registered atypical cases (9). On the other hand, in Belgium out of 133 cases of BSE diagnosed in cattle aging seven years and older, no atypical case was found (5). At present, approximately 70 atypical BSE cases have been detected worldwide.

Four points are important in the atypical BSE: most of the animals show no clinical signs; most of the animals have no vacuolation or other microscopic lesion; the $PrP^{res}$ is more widespread in the CNS; and finally, the age of the animals is eight years and older, mostly 11 to 13-year-old. The number of animals diagnosed with atypical BSE is relatively low. Furthermore, the material collected usually is poor, because of the absence of clinical signs. IHC usually is made to attend surveillance programs. The focus is the typical BSE, that means, the priority are animals aged 2,5 to 5-year-old. Research on interspecies transmission of the atypical BSE is still incipient.

The case reported in Brazil (2) attended all the characteristics of the atypical BSE reported in the literature , i.e. no clinical signs, no histological lesions and old animal. Furthermore, no cases of typical BSE were noticed although a robust program of surveillance was implanted by the government authorities (2). The cow was maintained in pasture feed. All these facts point to the occurrence of an atypical form and also it is very likely that the disease is indeed spontaneous. Therefore, countries with significant cattle herds should have the incidence of the disease in extremely low ratio and therefore not likely to be diagnosed.

**Suggested Reading:**

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