

# Putative Lafora body disease in a cow in the Netherlands

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## Introduction

Lafora body disease has been previously reported in humans, in several breeds of dogs, a cat, a flying fox, a cockatiel and in cattle in the USA and in the UK. To our knowledge this is the first case described in a cow in continental Europe.

## Materials and methods

An 8-year old Holstein-Friesian breed cow with a ruminal fistula from a research facility showed progressive decrease in milk production, diminished appetite, severe loss of weight and progressive esophageal spasms over a few weeks time. The cow was euthanized and sent in for postmortem examination.

A complete necropsy was performed, with histopathology of brain, spinal cord, esophagus, small and large intestine, liver, kidney, vagosympathetic trunc, thoracic vagal nerve, and recurrent laryngeal nerve. Quantitative mineral and trace element content of liver tissue was analysed.

## Results

At necropsy the only abnormal finding was mild dilation of the esophagus. The animal was in moderate body condition. Histopathology of the brain stem at the transition of the caudal thalamus to the midbrain, at the level of the medial and lateral corpora geniculata showed many intraneuronal round to cauliflower shaped inclusion bodies. The inclusions measured 6-17 µm in diameter, were pale basophilic, ground-glass appearing in the HE stain (figures 1 and 2), with an occasional dense core, and located in the neuronal perikarya as well as scattered in the neuropil. The inclusions stained brightly eosinophilic both in the PAS stain (not shown) and the PAS-diastrase stain (figure 3), consistent with polyglucosan bodies. No inclusions were present in the other brain regions, the autonomic nerves and intramural autonomic plexi of esophagus and intestine as well as the liver and kidney. The esophageal wall showed few small inflammatory foci consisting of few lymphocytes and rare macrophages centered on few autonomic ganglia cross sections. The mineral and trace element contents of the liver were within the normal ranges.

## Discussion

Polyglucosan bodies are basophilic to amphophilic inclusions and occur in neurologic disease and as an incidental finding in aged animals. Polyglucosan bodies within the neuronal perikarya are called Lafora bodies and are suggestive of Lafora body disease, when present in high numbers and together with matching clinical signs (including myoclonus epilepsy). Scattered polyglucosan bodies in the neuropil only, called corpora amylacea, are more consistent with an incidental histological change in aging animals. Conclusions: The morphological and histochemical findings in this cow with progressive esophageal spasms are consistent with Lafora body disease.

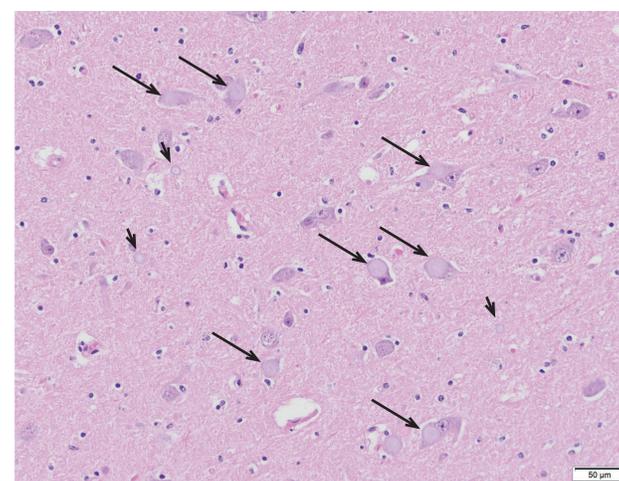


Figure 1. Brain stem, HE, 20x. Polyglucosan bodies within the perikarya (long arrows) and within the neuropil (short arrows)

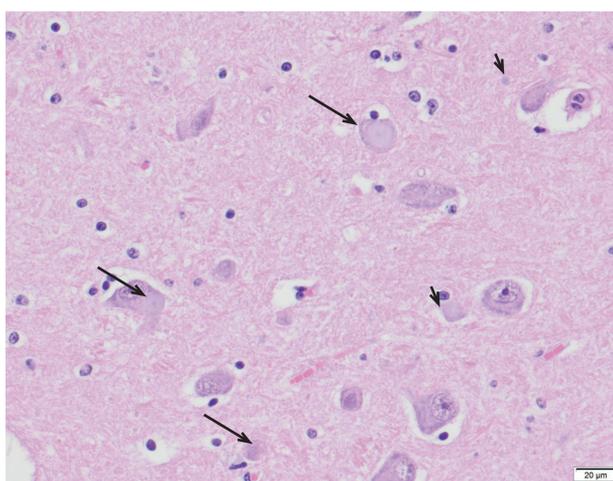


Figure 2. Brain stem, HE, 40x. Notice the pale grey aspect of the polyglucosan bodies, and the occasional dense core

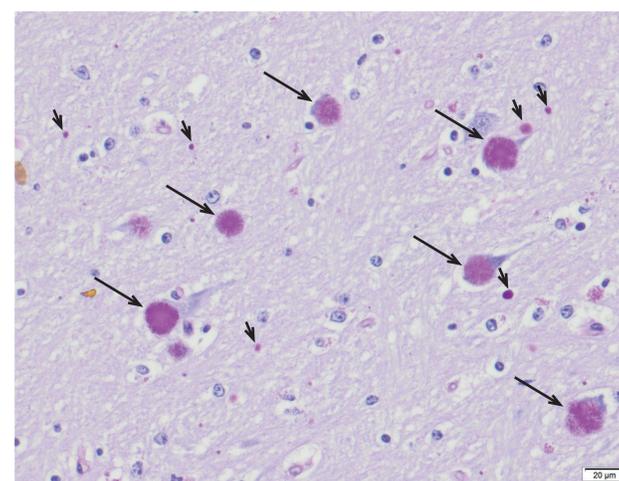


Figure 3. Brain stem, PAS-diastrase, 40 x. The polyglucosan bodies stain strongly magenta in the PAS stain after diastrase pretreatment, with radial staining pattern

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