



ATLANTO-AXIAL MALFORMATION AND SUBLUXATION IN A CZECHOSLOVAKIAN WOLF DOG WITH PITUITARY DWARFISM

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Pituitary dwarfism in dogs is due to an autosomal, recessively inherited disorder, caused by a genetic defect in LHX3⁽¹⁾. It is encountered most often in German shepherd dogs, but has been mentioned to occur in the Czechoslovakian wolf dog as well. The disorder is characterized by underdevelopment of the pituitary gland and a combined pituitary hormone deficiency. Pituitary dwarfism can lead to a wide range of clinical manifestations and not all dwarfs display the same signs. The most common clinical manifestations are marked growth retardation, retention of secondary hairs (puppy hair coat) with concurrent lack of primary hairs, and bilateral symmetrical alopecia⁽²⁾.

A 7-month-old female intact Czechoslovakian wolf dog, suffering from pituitary dwarfism, was presented with progressive ataxia, sopor and intermittent star gazing. Neurological exam revealed ataxia of her limbs, trunk and head, paraparesis, impaired proprioception and a hypermetric gait of all four limbs. History and clinical signs suggested a combination of a cerebellar and a spinal problem. Radiography, computed tomography and magnetic resonance imaging were performed of the neck and skull. Imaging revealed an abnormal dens and incomplete ossification between the three bony elements of the atlas with concurrent atlanto-axial subluxation and dynamic compression of the spinal cord by the dens. In addition, the calvaria caused pressure on the cerebellum. The malformations and aberrant motion at C1-C2 were confirmed at necropsy.

In human patients that suffer from dwarfism caused by a mutation in LHX3, anatomical abnormalities in the occipito-atlantoaxial joint in combination with a basilar impression of the dens axis can be found⁽³⁾. This is the first report of similar abnormalities in a dog with pituitary dwarfism.

References:

1. Voorbij AMWY, van Steenbeek FG, Vos-Loohuis M, Martens EECF, Hanson-Nilsson JM, van Oost BA, Kooistra HS, Leegwater PA. A contracted DNA repeat in LHX3 intron 5 is associated with aberrant splicing and pituitary dwarfism in German shepherd dogs. PLoS ONE 2011;6: e27940. doi:10.1371/journal.pone.0027940
2. Voorbij AMWY, Leegwater PAJ, Kooistra HS. Hypofysaire dwerggroei bij Duitse herdershonden, Saarloos wolfhonden en Tsjechooslowaakse wolfhonden. Beschikbaarheid van een genetische test. TvD 2010;135:950-4
3. Krström B, Zdunek AM, Rydh A, Jonsson H, Sehlin P, Escher SA. A novel mutation in the LIM homeobox 3 gene is responsible for combined pituitary hormone deficiency, hearing impairment, and vertebral malformations. J Clin Endocrinol Metab 2009;94:1154-61