GANGLIOSIDOSIS IN SHEEP CAUSED BY A DEFECTIVE $\beta$ GALACTOSIDASE GENE

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ABSTRACT

Gangliosidosis is a genetic disease that occurs in a variety of animals including sheep and humans. It is similar to Tay Sachs disease and Morquio B syndrome in humans. We are using this disease in sheep as preliminary research for developing tests and treatment for similar human genetic diseases. Gangliosidosis is caused by an inactive $\beta$-galactosidase gene. The normal $\beta$-galactosidase gene cleaves the terminal sugar from oligosaccharide chains allowing for their degradation. A buildup of these sugar chains accumulates in neurons and visceral organs of affected animals causing disease symptoms and eventual death. The symptoms in affected lambs are progressive neurological degeneration and loss of physical fitness occurring at 4-6 months with death occurring at 6-8 months. To identify mutated $\beta$-galactosidase genes, we are isolating the normal and mutated genes from tissues and blood of normal and affected lambs respectively. Once sequenced, the two genes can be compared to identify the mutations that cause the inactivation of the mutated gene. Similar diseases in other animals have shown that mutations are mainly base pair inserts and point mutations. The discrepancies between the two genes will be utilized to develop a rapid test for in-utero identification of Gangliosidosis.