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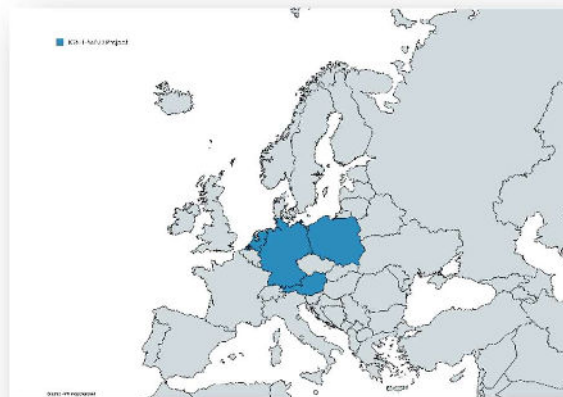
IgLON5 disease: NeuroInflammatory Tauopathy and Encephalopathy: the Model for Inflammation towards early treatment of NeuroDegeneration

Tauopathies are a group of dementia disorders, in which tau proteins cluster in the brain and brain cells die. Frontotemporal dementias are a subgroup of these affecting planning, executive functioning and behaviour, more than the typical memory problems seen with Alzheimer's Disease. IgLON5 disease is also a tauopathy, that was only discovered in 2014. In this disease the immune system attacks the own brain by creating antibodies against IgLON5. How this exactly leads to clustering of tau proteins and loss of brain cells is unknown. But, it is known that the IgLON5 antibodies – and the immune system – are very important. IgLON5 disease can serve as a model for how the immune system can create dementia. In neurodegenerative dementias, (hereditary) errors in tau proteins are found, but why and how this leads to dementia is also unknown. It is very likely that the immune system plays a role in why, when and how quick patients become demented. In this study, we will look at the role of the immune system in IgLON5 disease in patients at all stages of the disease: when they just became ill, later in disease and in brains of patients who died of this disease. We will study blood, brain fluid and brain tissue with several techniques. These techniques are the most novel and up-to-date methods we can use nowadays. We will also investigate if we can mimic the disease in cultured brain cells: we will use IgLON5 antibodies from patients and add these to brain cultures. We will check if and how the tau proteins change and cluster. This last step would be the missing link between antibodies and dementia. In anti-IgLON5 disease we can and will compare the role of the different parts of the immune systems and determine the role of the immune system in dementia. Understanding the role of the immune system in the clustering of tau proteins and loss of brain cells will have a major impact on potential treatments. This role can be direct, (causing dementia) or indirect (accelerating the decay in dementia). Either way, the immune system can be changed by medication and this might open up new ways of treating dementia. These can also be considered in other dementia types, like frontotemporal dementias. It might also provide markers that better predict how quickly a patient will become ill or deteriorate.

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