Prion Disease: A protein gone rogue

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Background

'Creutzfeldt-Jakob Disease' or CJD is the most common form of human prion diseases, which is a **rare brain disease.** It has a **100% death rate**, with **no treatments** where **survival is weeks to months** after symptoms start.

Causes : The misfolding (& clumping) of the prion protein (happen by chance, or a mutation, or exposure to misfolded prion protein





Proteins are like origami, if folding is **wrong = clumpy and misfolded =** other copies to also misfold, like a domino effect.

How is it diagnosed?

Through a test called 'RT-QUIC' which detects misfolded prion protein in a spinal fluid sample by an increase in fluorescence.

<u>The problem</u>: Diagnosis of this disease **is difficult** because of its rarity and the **current test (RT-QUIC) takes too long** (120 hours) for this **rapid** disease. This can be because of the **overall low amount of small misfolded prion** protein in spinal fluid relative to other proteins.

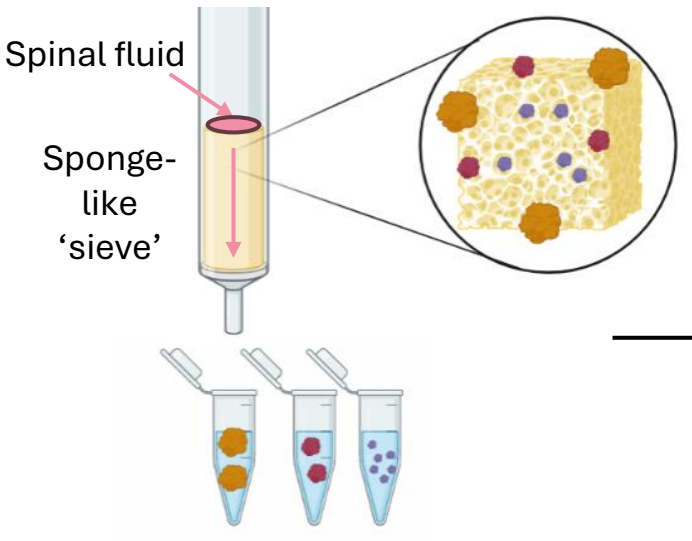
Aim

To make the diagnosis faster by processing spinal fluid samples using a 'sieve' method.

Hypothesis

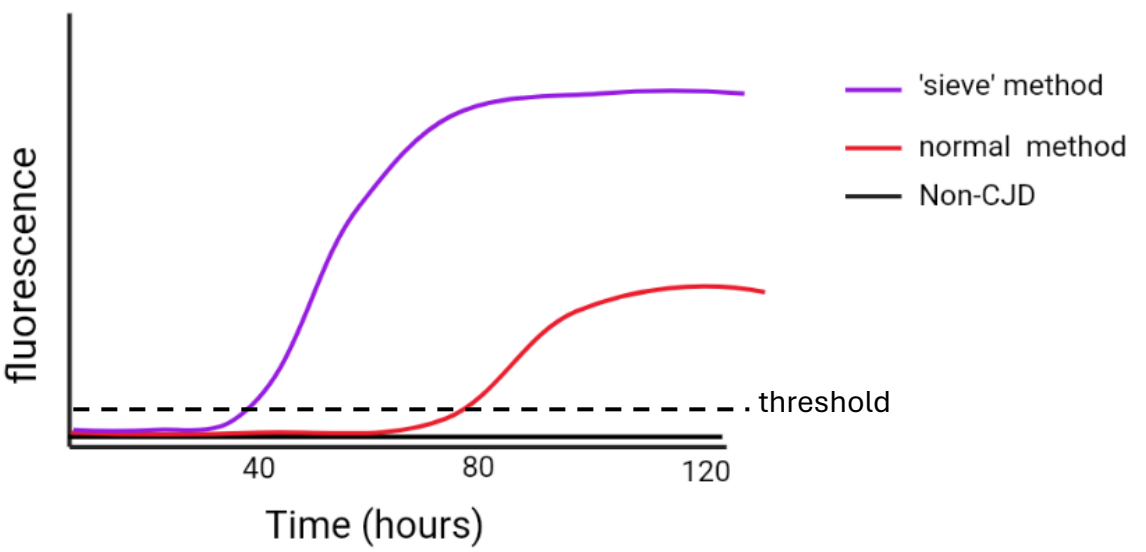
We hypothesise that using the 'sieve' method to process samples before the RT-QUIC will separate out 'junk' to distinguish the smaller, misfolded prion protein.

Method



Larger proteins come out first

Results



Key Takeaways

- Method can be adapted for earlier detection for Parkinson's and Alzheimer's Disease
- Education about Prion diseases is necessary for all medical and dental personnel
- Standardise sterilisation to kill Prion at 141C.
- Earlier detection = earlier treatment = more decision-making time for patients, loved ones & clinicians

  = more time to find a possible cure