

**'CJDSGN Memorial Award 2019 in memory of Jennifer Duckworth' - \$25,000**  
**– awarded in October 2019**

**'Treatment of medically relevant prion disease' during 2019/2020**

*A major challenge to the management of medically relevant prion disease is the uncertainty of the effect of strain variation on the detection, prevention and treatment of disease. We have developed mouse adapted strains of sporadic Creutzfeldt Jakob Disease and familial prion disease that reflect the major molecular strain types identified in human prion diseases. We will infect cells with these prion strains and use them to screen a panel of therapeutic molecules that have been previously shown to be effective in sheep scrapie models of prion disease and novel therapeutic compounds identified by our collaborator Ina Vorberg to inhibit prion formation. This unprecedented study will involve screening five medically relevant forms of human prion disease with ten therapeutic compounds and ensure the clinical and translational relevance of prion disease research in Australia and offer hope to patients suffering from prion diseases and their families.*

Ms Zoe Revill was recruited as an Honours student in 2019 to investigate the effect of strain variation in medically relevant prion disease. In this very ambitious project, Zoe was able to screen existing (2) and novel (3) compounds for their efficacy in a cell culture (in vitro) model using two prion strains derived from sporadic and familial forms of prion disease. Although not as extensive as the proposed project a key outcome was the demonstration that these two strains responded differently to treatment with some but not all drugs tested.

That is for example, PPS was able to efficiently reduce the prion load in cells infected with M1000 prions but was not as effective against MU-02 prions, whereas congo red efficiently reduced prions from cells infected with both prion strains. Importantly we identified a novel compound that appeared to work effectively against both strains tested. Future studies will be required to establish the significance of this observation.