

A new model for studying sporadic CJD

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Difficulties in studying sporadic CJD

The causes of sporadic CJD are unknown

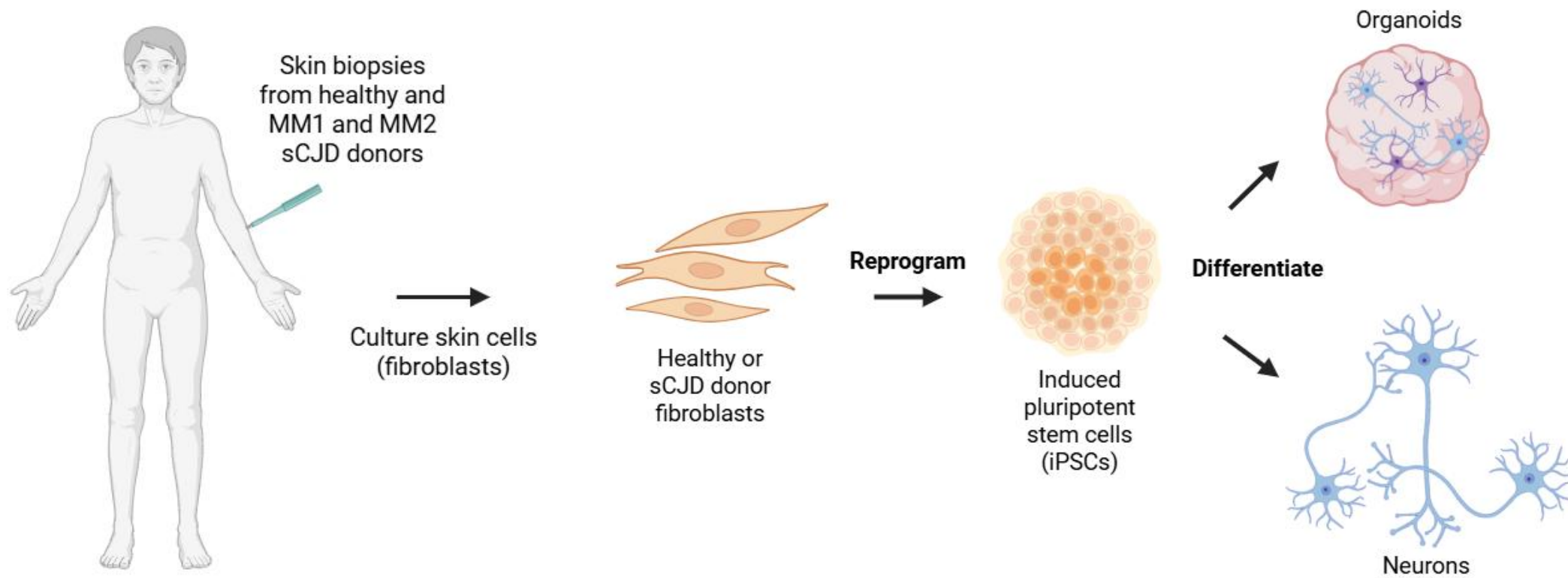
People don't know they have sCJD until after symptoms appear, which is after irreversible neuronal loss has occurred

Mouse models and cell culture models do not develop CJD spontaneously, they need to be infected

Mice do not always recapitulate what is seen in humans

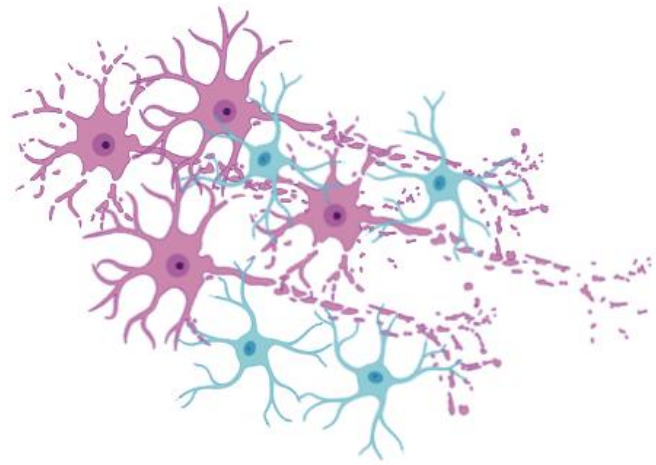
Can we develop a new research model for studying sporadic CJD?

Neurons and organoids derived from sCJD donor cells

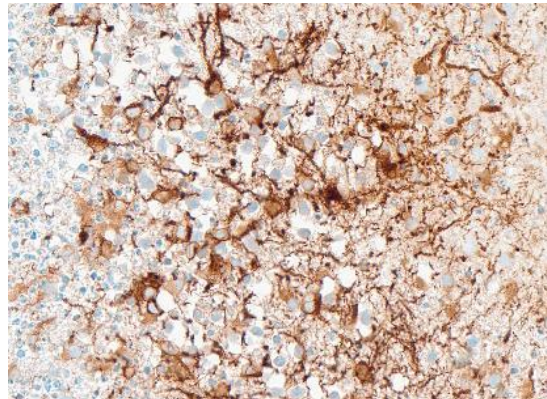


Skin cells kindly donated by Dr. Wenquan Zhou

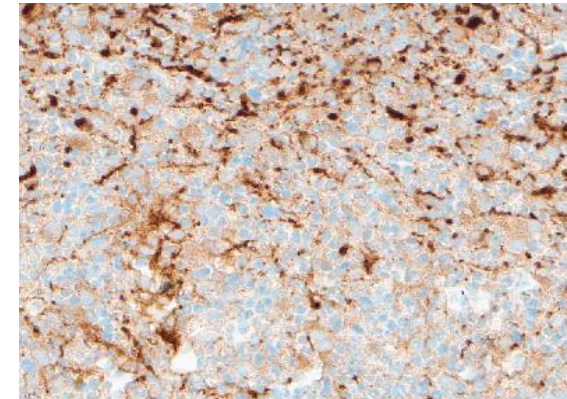
sCJD organoids show significant neuronal loss as they age



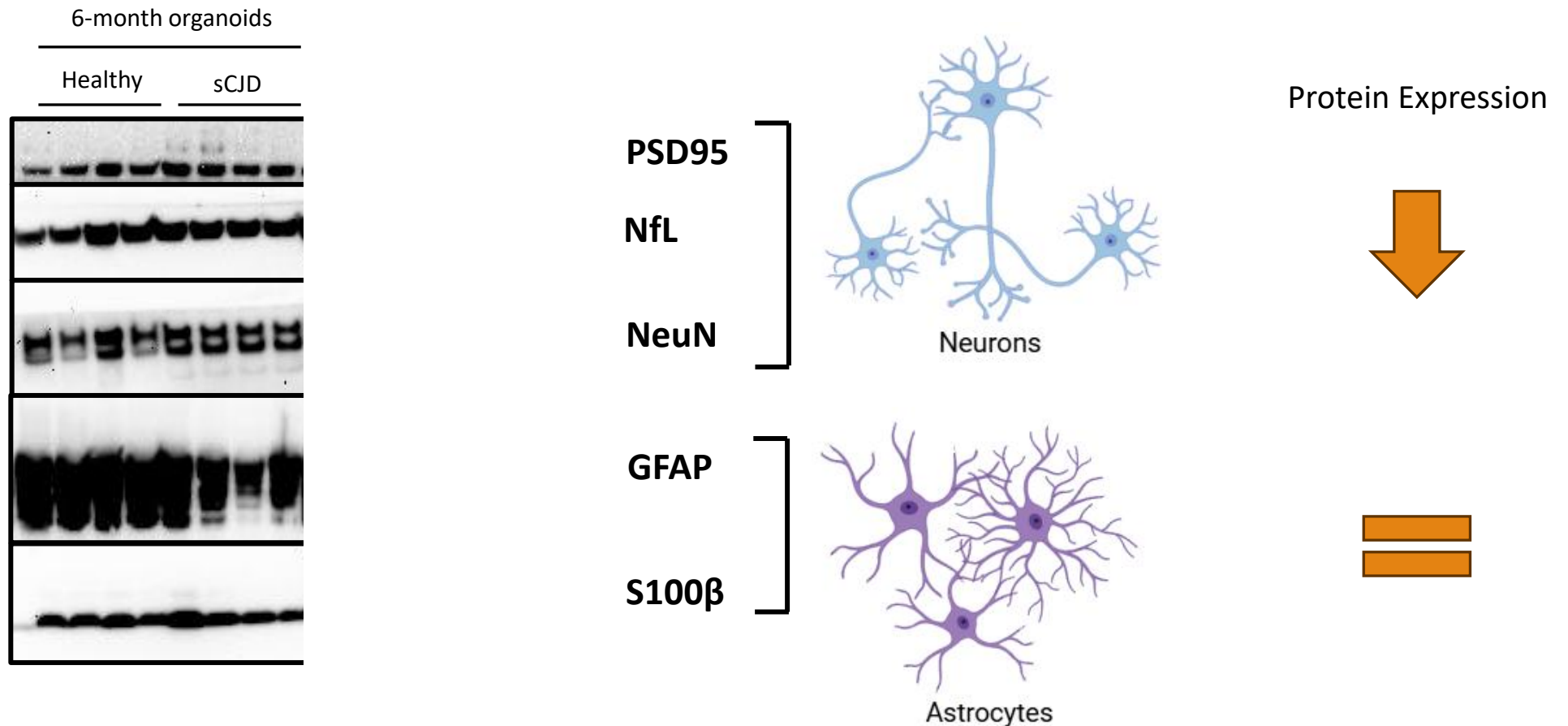
Healthy



sCJD



sCJD organoids show significant neuronal loss as they age



sCJD organoids and neurons can be used to study neurodegeneration

- We have successfully reprogrammed cells from a donor with MM1 sCJD and MM2 sCJD
- So far, none of these organoids or neurons have shown any prion seeding activity
- Organoids show neuronal loss as they age
- Neurons show functional abnormalities compared to healthy neurons

Future Directions

- Reprogram cells from other donors to study different types of sCJD
- Monitor neurons and organoids for prion seeding activity
- Test therapeutics to reverse neuronal dysfunction and loss

A huge thank you to the donors for
supporting this work!

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Summary

- We can model sporadic CJD by reprogramming cells from donors with sCJD and growing them into neurons and organoids
- sCJD neurons show dysfunction
- sCJD organoids show neurodegeneration as they age
- sCJD neurons and organoids can be used to further understand disease processes and test therapeutics to prevent neuronal loss

