

4. DISEASE MODELS

PROJECT:

Evaluation of a novel inducible muscle-specific TDP-43 mouse model of MND

It is not yet certain where in the body MND begins. Although a potential location is the body's muscles, only a small amount of research has so far examined how muscle tissue may be involved in triggering MND. This project aims to overcome this obstacle by producing a mouse model in which TDP-43, a protein that is dysfunctional in the majority of people with MND, is designed to misbehave only in muscle. Investigators will use this new model to study specific roles for muscle in the onset and progression of MND. The model will also provide a valuable tool for testing how effective muscle targeting strategies are for treating MND.

KEY HIGHLIGHTS:

This project is developing a 'world-first' model of MND, in which the onset of the disease occurs in muscle.

AMOUNT INVESTED BY FIGHTMND IN THIS RESEARCH PROJECT:

\$249,994

Q&A:

What problem are you trying to solve with this project?
Skeletal muscle appears to play a key role in MND disease progression and potentially in disease onset. This project will allow us to determine if causing a protein (TDP-43) to aggregate specifically in skeletal muscle cells causes the onset of MND-like symptoms.

“The successful development and validation of our mouse model will provide a valuable tool to investigate the potential molecules inside muscle that impact MND.” – Professor Aaron Russell

PROJECT LEAD:

Professor Aaron Russell
Deakin University, VIC



Above: Professor Aaron Russell | Below: Russell lab at Deakin University (L-R): Miss Anuska Podar (PhD student), Dr Paul Della Gatta (Research Fellow), Dr Felicity Dunlop (Research Fellow) and Professor Aaron Russell