## *Dissecting neuronal susceptibility to mitochondrial disease*

Mitochondria are the powerhouses of the cell. Mutations that render mitochondria unable to generate energy cause a group of rare and usually fatal pathologies collectively known as mitochondrial disease. It has been estimated that 1 in 5000 children in the US will develop a mitochondrial disease. Currently, there is no cure for mitochondrial disease and the treatments available are mostly ineffective. Energy-demanding cells such as neurons are especially sensitive to mitochondrial disease, and they account for most of the clinical signs and symptoms observed in humans, such as hypotonia, ataxia, seizures and early death. However, even if every single cell in the body carries the mutation, only specific brain areas seem to be affected by the deficiency. **The Quintana lab** current research focuses on identifying the neuronal populations susceptible to mitochondrial disease and which mechanisms are making these neurons die. This knowledge is essential to understand and fight these incurable diseases. The Quintana lab uses a wide array of approaches, combining molecular biology, stereotaxic surgery, mouse genetics and behaviour, biochemistry, histology, optogenetics and in vivo electrophysiology to reveal novel pathways and mechanisms in neuronal function and pathology and open new and unexplored lines of research and therapeutic targets to treat mitochondrial disease encephalopathy.

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