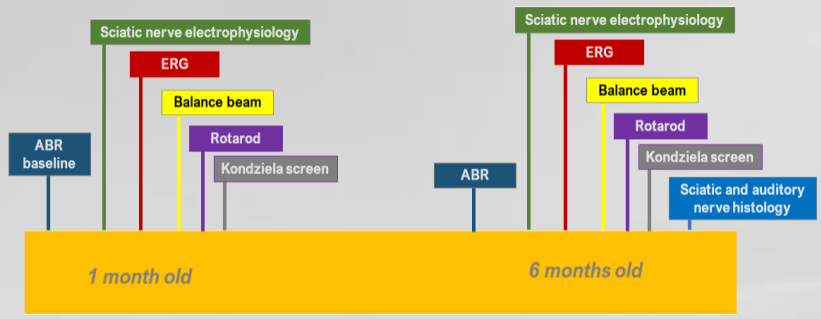
Charcot–Marie–Tooth neuropathy

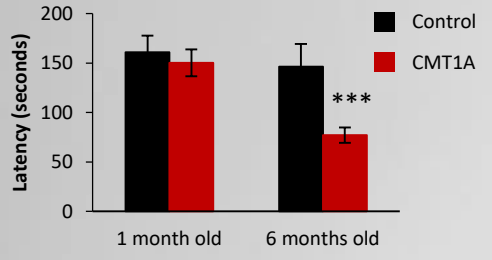
Charcot–Marie–Tooth disease (CMT) is one of the hereditary motor and sensory neuropathies of the peripheral nervous system characterized by progressive loss of muscle tissue and touch sensation across various parts of the body. Currently incurable, this disease is the most commonly inherited neurological disorder, and affects about one in 2,500 people.

The most prevalent type of CMT is the CMT1A characterized by a duplication of the PMP22 gene leading to an accumulation of the pmp22 protein in the Schwann cell and a progressive demyelination.
The C3-PMP22 mice express 3 copies of a wild-type human PMP22 gene making these mice a robust and reproducible model to analyze the sensory neuropathy and neuromuscular disorders observed in CMT 1A disease. This model allows to determine the efficacy of new pharmacological candidates targeting demyelinating diseases such as CMT1A disorder.

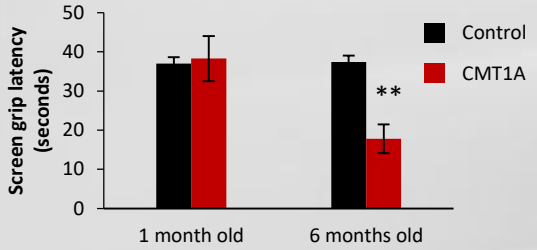


Neuromotor system

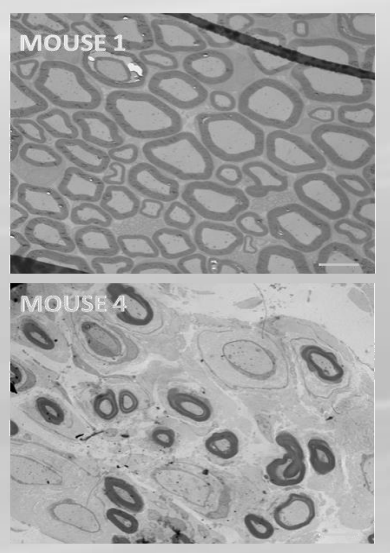
Rotarod (motricity and equilibrium)



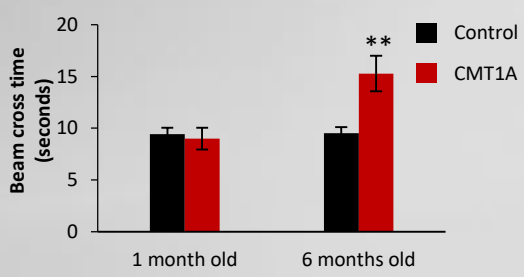
Kondziela' screen (muscular strength)



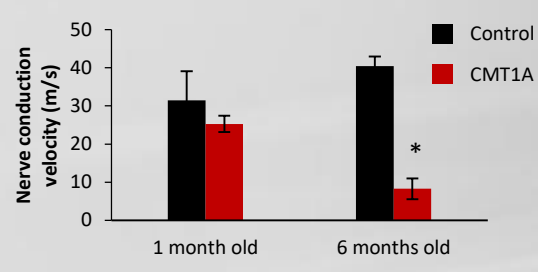
Sciatic nerve myelin



Balance beam cross time (walking performances)

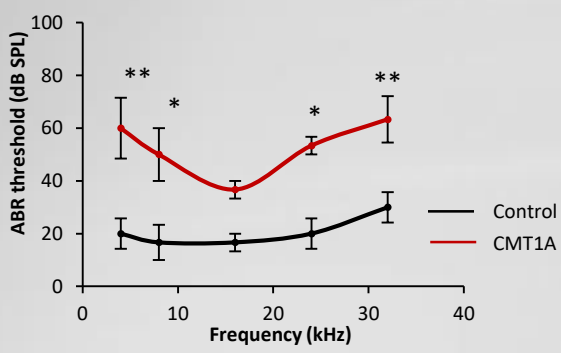


Sciatic nerve conduction velocity

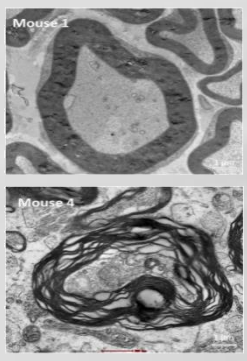


Hearing system

ABR

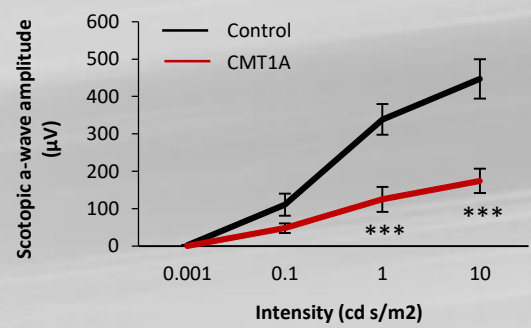


Auditory nerve histology (TEM)



Visual system

Electroretinogram



Decrease of sciatic nerve action potential velocity, neuromuscular disorders, sciatic nerve and auditory nerve demyelination, hearing loss and decrease of scotopic a-wave amplitude (visual acuity) observed in CMT1A mice at 6 months old.