

## **Author contribution statement**

JB, MJW, AAG and MH conceptualized the mouse model and performed the initial characterization. VK and ER assisted in the initial histological characterization of the model. ER also provided tissue sections of humans with myositis. JN continued with the characterization, i.e. performed in situ hybridizations of transgenic mice, analyzed chemo- and cytokine expression in human cases of myositis, determined weight and fiber size distribution in muscle of transgenic mice as well as qPCR of HSA-LT mice at different time points. CL performed the qPCRs of HSA-LT compared to HSA-LT,CreAtg mice under supervision of KPH. PT did the p62 and pTDP43 immunostaining. TB did the hanging wire and forelimb grip strength test. MP provided the infrastructure for this and for processing epon blocks for electron microscopy. The resulting sections were analyzed by CE and HZ for mitochondrial morphology (Fig. 4H) and further by JB and JW (Fig. 6). KKZ and TP performed and analyzed the Western blots. RR performed and analyzed the MRI experiments. UR managed the mouse colony in Heidelberg. MH, KPH, JS and JZ conceptualized the drug treatment. JS provided the infrastructure for the drug treatment and the continuous behavioral monitoring during treatment. JS and JZ supervised the drug treatments. LAF, ASMS, LW, JJR performed the drug treatments. LAF, ASMS, LW, JJR, JS, JZ, DL and FO analyzed the outcome, especially the behavioral outcome of drug-treated mice. CL and ALE performed the qPCRs of drug-treated mice, JB analyzed these qPCRs. AI determined the number of lymphocytes in liver samples of drug-treated mice. FK performed and analyzed the RNAseq and the mitochondrial DNA sequencing. CCK analyzed the published human IBM RNAseq data. JB analyzed and provided human histological images, mouse immune and enzyme histochemistry, human in situ hybridizations, assembled all the final figures. JB wrote the initial draft of the manuscript and of the revised version. JB, JW, MH, ER, JS, MJW, FK and JZ revised the manuscript. All authors approved the manuscript.