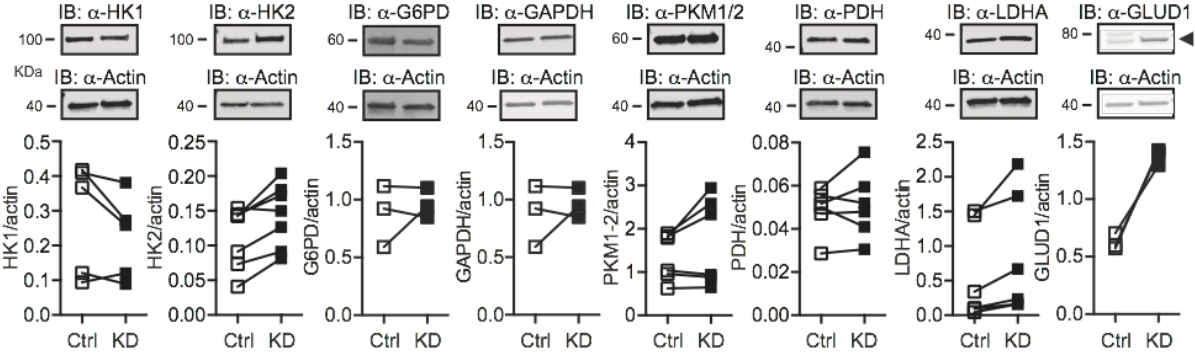


# **GDAP1 loss of function inhibits the mitochondrial pyruvate dehydrogenase complex by altering the actin cytoskeleton**

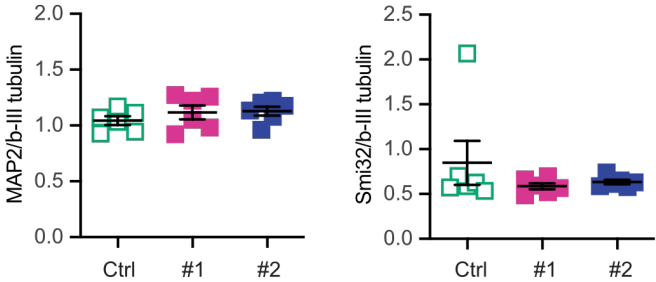
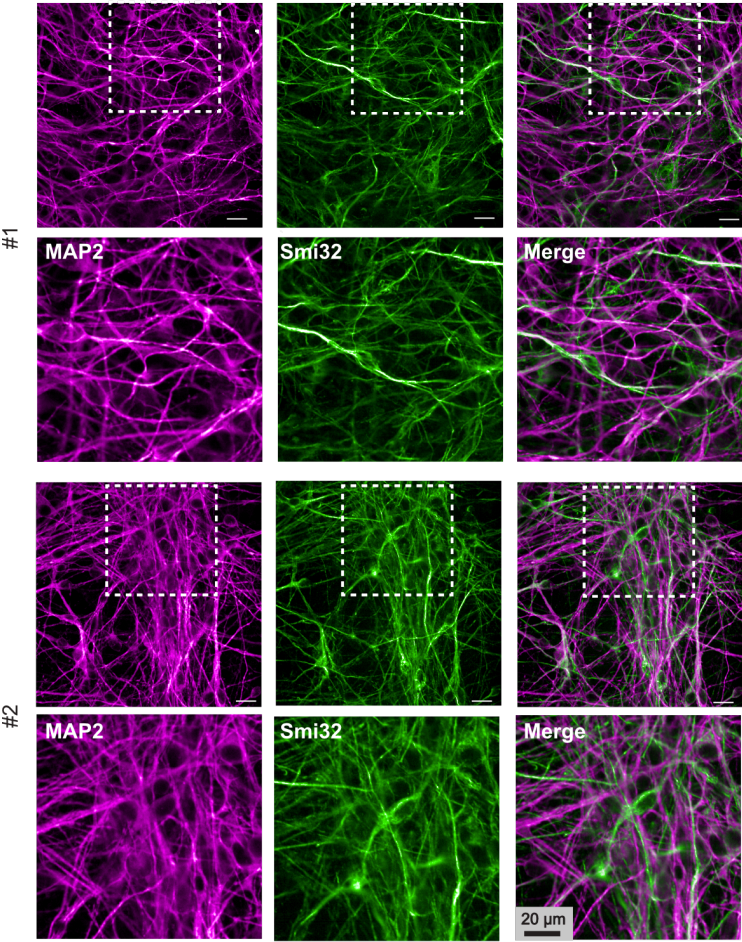
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**Supplemental figure 1**



**Supplemental Figure 1 Changes in protein abundance. (a)** Immunoblot of whole cell lysates from Ctrl and GDAP1 KD cells against Hexokinase 1 (HK1), HK2, Glucose-6-phosphate dehydrogenase (G6PD), Glyceraldehyde 3-phosphate dehydrogenase (GAPDH), Pyruvate kinase M1/2 (PKM1/2), Pyruvate dehydrogenase (PDH), Lactate Dehydrogenase A (LDHA), Glutamate dehydrogenase (GLUD1, arrowhead). Sizes are indicated and actin served as loading control. n=3-7

**Supplemental figure 2**



**Supplemental Figure 2 Patient-derived motoneurons express similar amounts of MAP2 and Smi32.** Immunocytochemical staining of patient-derived motoneurons with Smi32 and MAP2 normalized to the neuronal marker  $\beta$ -tubulin-III (not shown here). Batch analysis calculated the fluorescence intensity of each marker. Data are from three independent differentiation lines performed in triplicates. Statistical variation is shown as scatter plots with the indication of mean  $\pm$  SEM and significance calculated using the ordinary one-way ANOVA with Turkey’s multiple comparisons test.

