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Supplemental information

Aberrant cyclin C nuclear release induces mitochondrial fragmentation and dysfunction in *MED13L* syndrome fibroblasts

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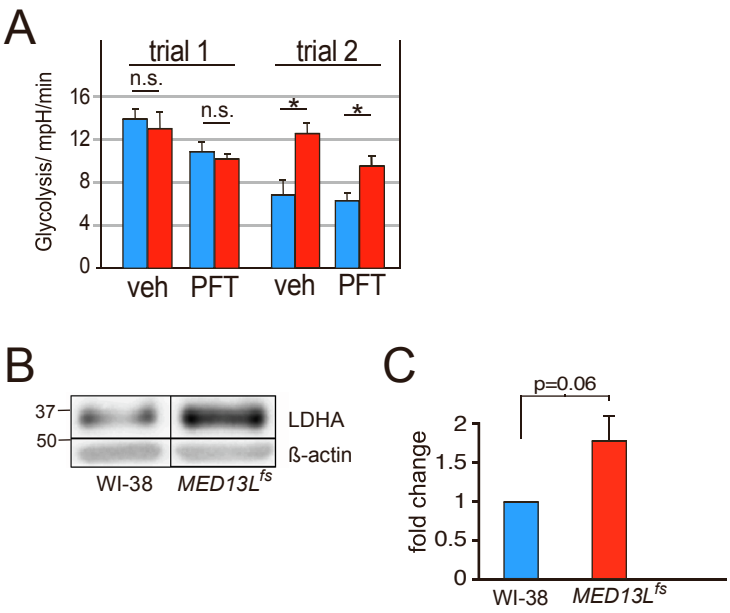
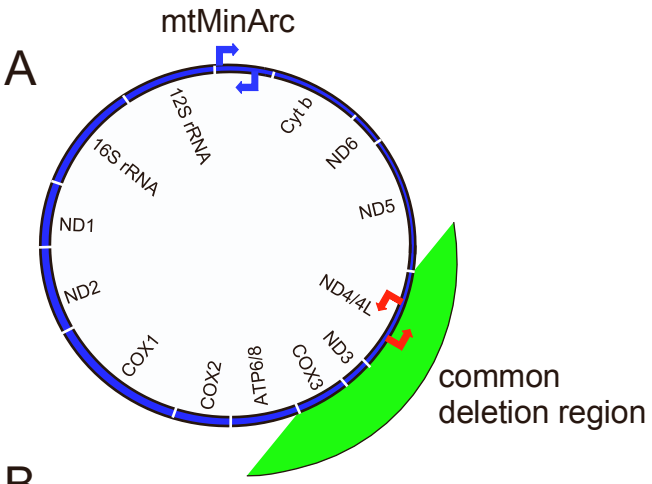


Figure S1. Glycolytic activity in *MED13L*$^{+/fs}$ cells. Related to Figure 1. (A) Seahorse analysis of lactate production to measure glycolysis in two trials with WI-38 and *MED13L*$^{+/fs}$ cells treated with PFT (1 μM, 96 h) or the vehicle control. * indicates p=0.05, n.s. indicates no statistical difference. (B) A representative Western blot analysis of lactate dehydrogenase A protein levels in WCE prepared from the growth matched cells as indicated. ß-actin served as a loading control. (C) LDHA signals relative to ß-actin were quantified using chemiluminescence imaging (n = 3 separate blots). The LDHA/ß-actin ratio for WI-38 was set at 1.

A



B

Annotation Cluster 1	Enrichment Score: 19.06	Count	P_Value	Benjamini
UP_KEYWORDS	Mitochondrion	222	1.5E-26	1.1E-24
GOTERM_CC_DIRECT	mitochondrion	327	1.7E-25	2.9E-23

Figure S2. Reduced mitochondrial DNA copy number and mitochondrial maintenance gene transcription in *MED13L*$^{+/fs}$ cells. Related to Figure 2. (A) Schematic of human mtDNA map with the qPCR primer locations indicated (adapted from Phillips et al., 2014). mtDNA locations of primers for the major arc (red arrows, ND4/4L locus) and minor arc (blue arrows at 0/360°). The vast majority of mtDNA mutations occur in the major arc (grey collar) with the most common deletions. (B) GO clusters enriched for genes involved in mitochondrial maintenance that require cyclin C for their full expression in MEF cultures. The number of genes identified in each cluster is given. These data sets contain overlapping genes. The P value and Benjamini value (a tool that decreases the false discovery rate) are given.