

Supplemental Figure 1. Schematic representation of the exon structure of the two NDUFAF7 isoforms labeled a and b. Exons are shown in grey and black. The short isoform is missing exon 3, part of exon 4 and exon 5. The position of the DUF185 domain is indicated, as is Motif I of the methyltransferase domain, the target sequence for the siRNA used in this study, and the peptide epitope used to raise a polyclonal rabbit antibody.

Supplemental Figure 2. Immunoblot analysis of NDUFAF7 in human fibroblasts following extraction with alkaline sodium carbonate showing the expression of the long (49kDa) and short (38k Da) isoforms and the predominantly soluble nature of the protein.

Supplemental Figure 3. Steady state levels of mitochondrial-encoded mRNAs in NDUFAF7 knock down. Northern blot analysis of control, Alexa (siRNA transfection control) and NDUFAF7-siRNA-2 cell lines. Specific probes were hybridized for detection of the mRNA subunits of complex I: ND1, ND5 and ND6, complex IV: COX1 and complex V: ATP6/8. The 16S mitochondrial ribosomal RNA was used as a loading control

Supplemental Figure 4. Identification of the dimethylated site of NDUFS2 by mass spectrometry. MS/MS spectrum of the dimethylated NDUFS2 tryptic peptide
KDPHIGLLHR(me₂)GTEK

Supplemental Table 1. Mass spectrometry analysis of the ND1 immunoprecipitate. Scaffold scores for PGS1 (control) and PGS1+NDUFAF7-siRNA-2 (methyl transferase mutant) cell lines were obtained using the total spectrum count. Complex I structural subunits and assembly factors are highlighted in blue.







