## **Supplemental Material**

## Disease associated mutations in mitochondrial precursor tRNAs affect binding, m1R9 methylation and tRNA processing by mtRNase P

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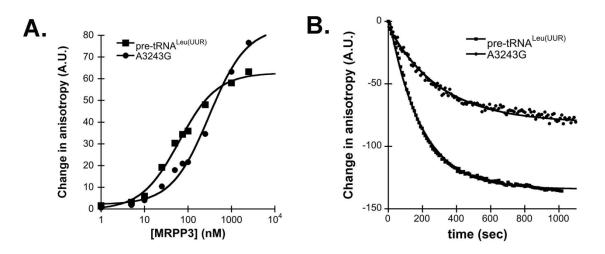
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**Supplemental Table 1.** Melting temperature values for first transitions for wild type (mt)pre-tRNAs and (mt)pre-tRNA<sup>Ile</sup> mutants.

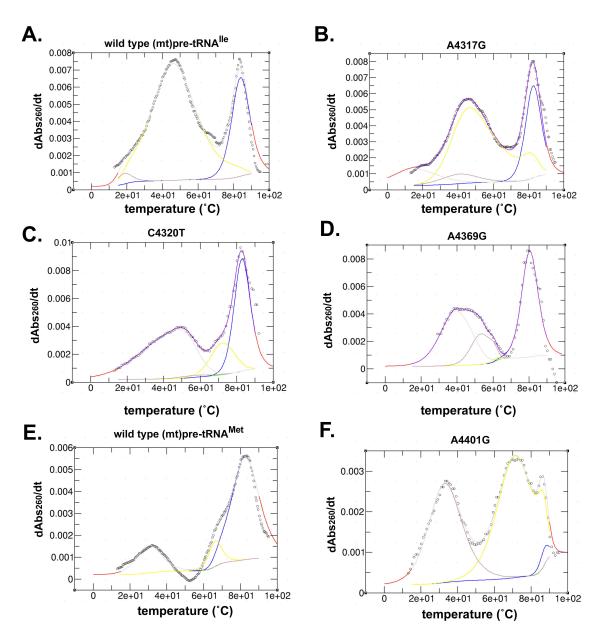
human mitochondrial pre- tRNA	T <sub>m</sub> 1 (°C)	T <sub>m</sub> 2 (°C)
(mt)pre-tRNA <sup>Leu(UUR)</sup>	28.7 ±0.1	-
(mt)pre-tRNA <sup>Met</sup>	32.6 ± 1.1	
(mt)pre-tRNA <sup>lle</sup>	47.7 ± 1.6	
A4269G	$40.4 \pm 5.4$	49.3 ± 1.0
A4317G	$42.5 \pm 0.9$	$56.2 \pm 0.5$
С4320Т	27.7 ± 3.7	$47.5 \pm 0.4$

Supplemental Figure 1.



Supplemental Figure 1. A. Example of a wild type and a mutant (mt)pre-tRNA binding in the function of MRPP3 concentrations when 150 nM MRPP1/2 is present using standard binding assay conditions. B. Example of the single turnover cleavage of a wild type and a mutant (mt)pre-tRNA using standard cleavage assay conditions.

Supplemental Figure 2.



Supplemental Figure 2. Examples of fitting data from UV melting experiments by fitUVData.py and Global Melt Fit [33, 34] for different (mt)pre-tRNAs.