Supplemental Material

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

Agnes Karasik¹, Carol A. Fierke², Markos Koutmos^{3*}

¹Department of Biochemistry and Molecular Biology, Uniformed Services University of the Health Sciences, Bethesda, MD, 20814, USA

²Department of Chemistry, Department of Biological Chemistry, University of Michigan,

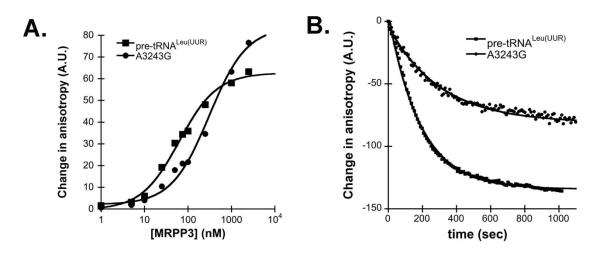
Ann Arbor, MI 48109, USA

³Department of Chemistry, Program in Biophysics, University of Michigan, Ann Arbor, MI, 48109, USA

Supplemental Table 1. Melting temperature values for first transitions for wild type (mt)pre-tRNAs and (mt)pre-tRNA^{Ile} mutants.

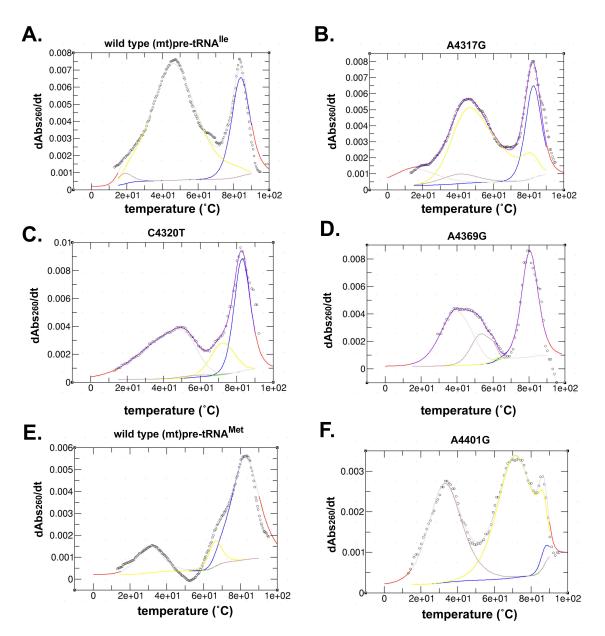
| human mitochondrial pre- tRNA | T _m 1 (°C) | T _m 2 (°C) |
|----------------------------------|-----------------------|-----------------------|
| (mt)pre-tRNA ^{Leu(UUR)} | 28.7 ±0.1 | - |
| (mt)pre-tRNA ^{Met} | 32.6 ± 1.1 | |
| (mt)pre-tRNA ^{lle} | 47.7 ± 1.6 | |
| A4269G | 40.4 ± 5.4 | 49.3 ± 1.0 |
| A4317G | 42.5 ± 0.9 | 56.2 ± 0.5 |
| С4320Т | 27.7 ± 3.7 | 47.5 ± 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.