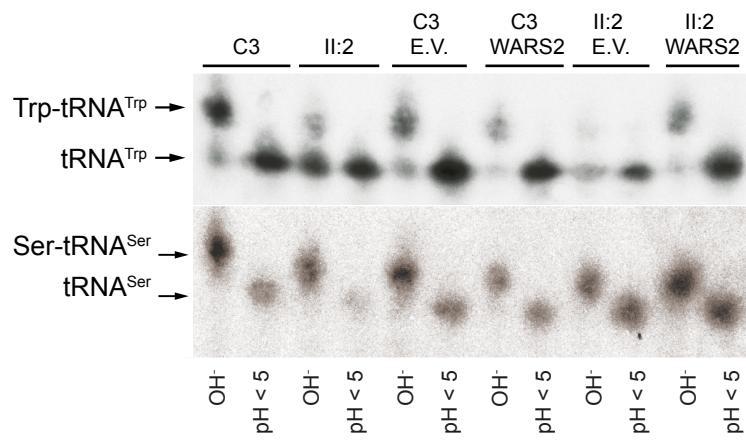
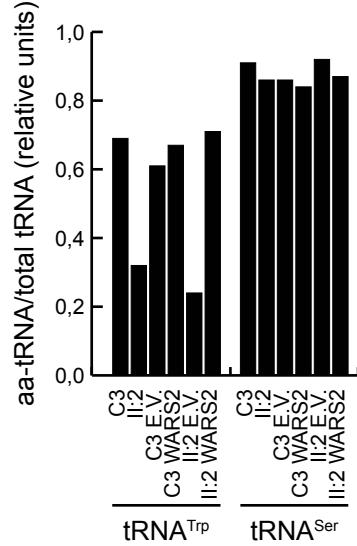


A)



aa-tRNA/total tRNA (relative units)



B)

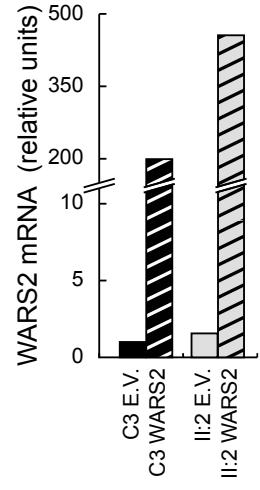


Figure S3. Rescue of the aminoacylation defect in NES cells from subject II:2. (A) Left: Representative acidic-urea PAGE on control (C3), and subject II:2 NES cells transfected with empty pIRES-eGFP (E.V.) or overexpressing WARS2 (WARS2), n=3 independent experiments.

Aminoacylated tRNAs (Trp-tRNA^{Trp}, Ser-tRNA^{Ser}), migrate slower, when isolated under acidic (pH < 5) conditions. Uncharged tRNAs (tRNA^{Trp}, tRNA^{Ser}) migrate as seen under basic (OH⁻). Right: Quantification of (A) under acidic (pH < 5) conditions. (B) qRT-PCR analysis of WARS2 expression in control (C3) and subject II:2 NES cell lines transfected with either empty pIRES-eGFP (E.V.) or overexpressing WARS2.

Maffezzini et al. Figure S3