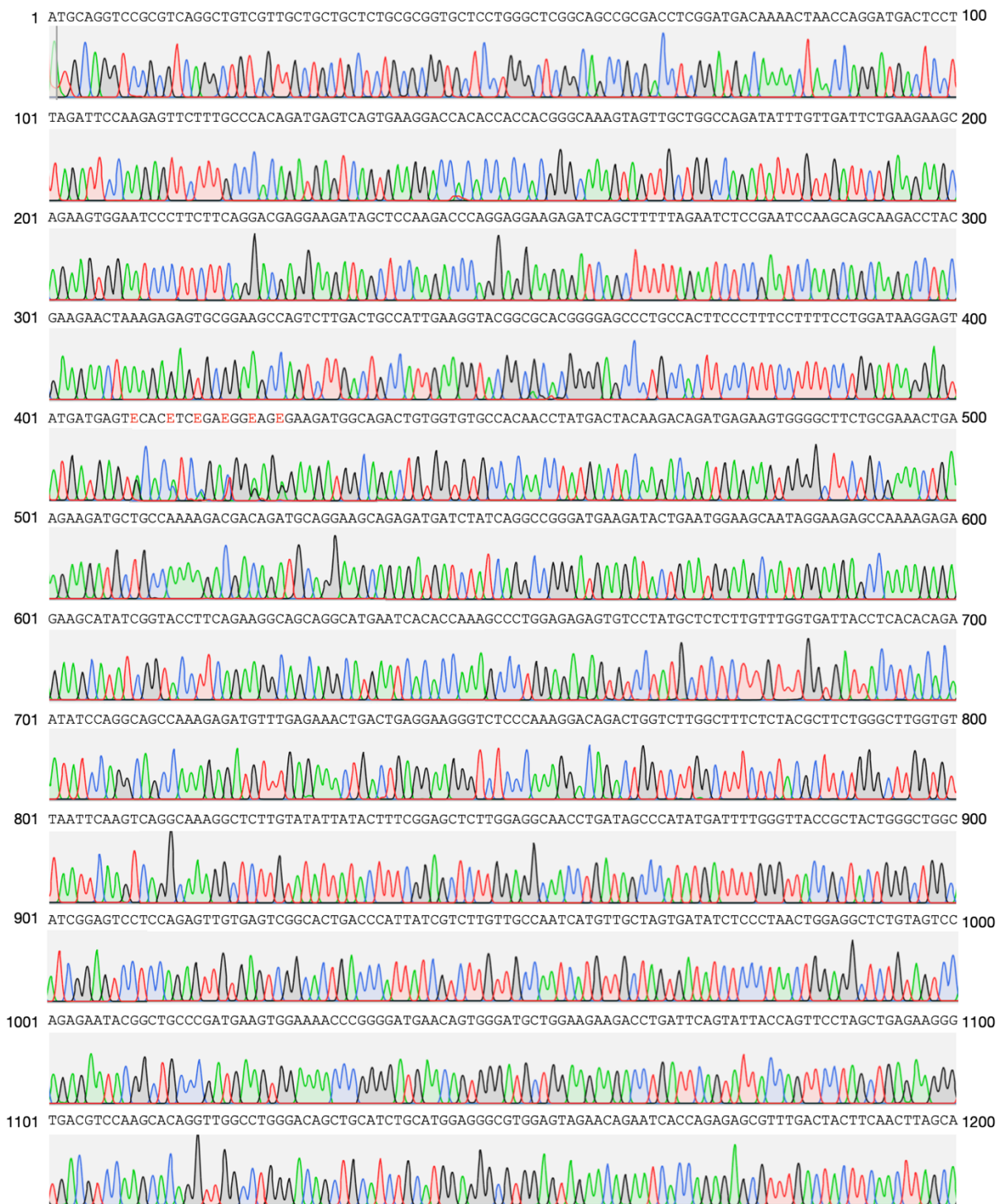


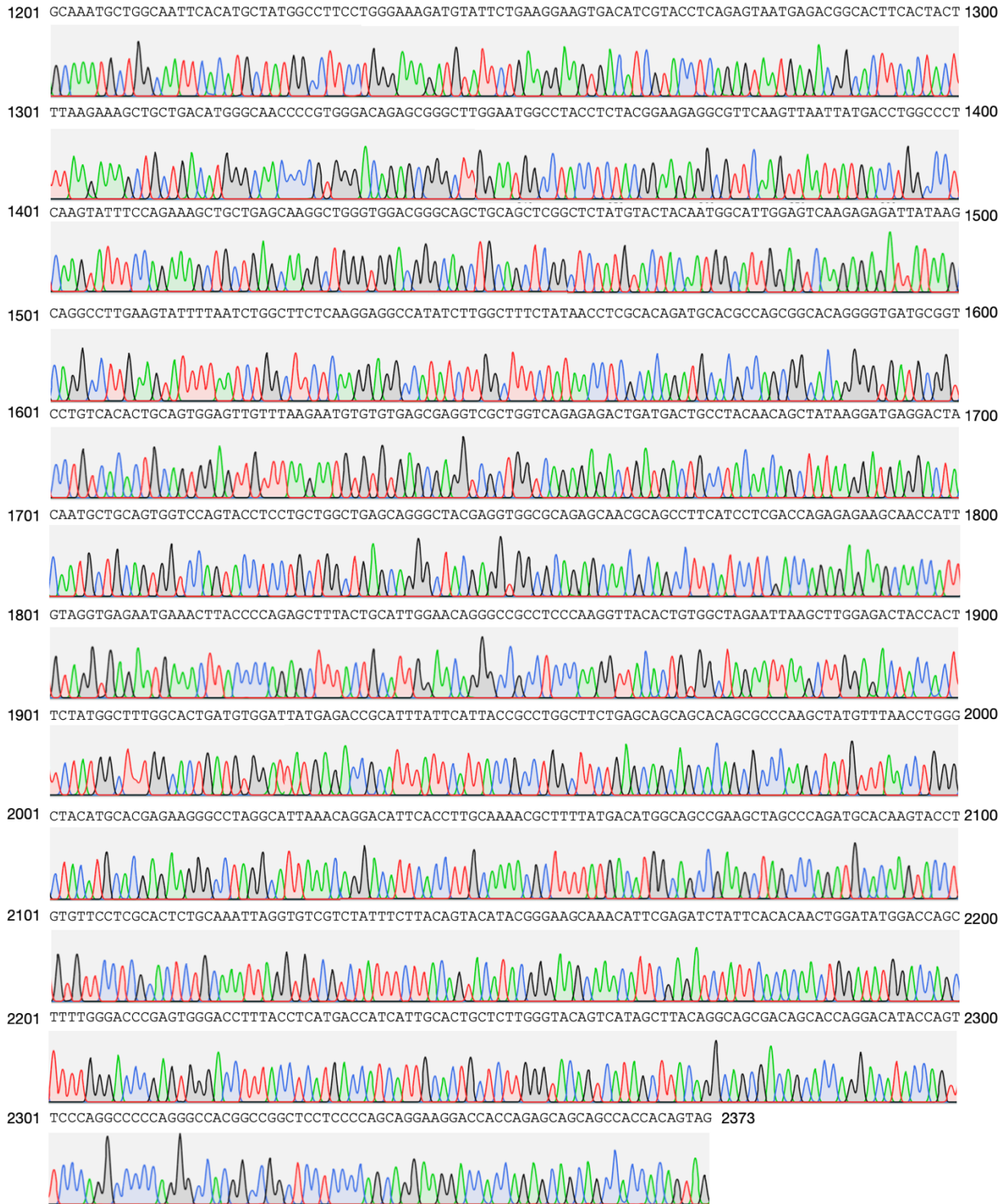
Appendix for

**Functional rescue of a disease-linked ERAD pathway mutation via
alternative splicing**

Appendix Figure S1

Appendix Table S1





Appendix Figure S1. The Sanger sequencing confirmation of *Sei1L* C141Y Heterozygous mouse coding DNA sequence. Sanger sequencing confirmation of the *Sei1L* C141Y heterozygous coding DNA sequence in mouse liver, obtained from multiple Sanger sequencing fragments. E, heterozygous allele.

Appendix Table S1. The primers and ASOs used in the study

Name	Sequence
Genotyping primer for <i>Sel1L</i> ^{C141Y} allele	F 5'- AGTACACATCCGATGGAAGAGAAG-3'; R 5'-GAAAATGCCTTCCAAATGCTGC-3';
Genotyping primer for <i>Sel1L</i> wildtype allele	F 5'- AGTGCACCTCAGACGGGAGGG-3'; R 5'- GAAAATGCCTTCCAAATGCTGC-3';
Sequencing primer for <i>mSel1L</i> genomic DNA Exon 4 and intron 4	F 5'- CTTAAGAACTCAAAGTCTACACTAAGTCT-3'; R 5'- CAGCTTGCCTCAAGGGTTTACAGAA-3';
Mutagenesis primers for alternative splicing donor site (Mutation 1)	F 5'-CCATTGAAGGCACGGCGCACGGGGA-3' R 5'-GCGCCGTGCCTTCAATGGCAGTCAAGA-3'
Mutagenesis primers for Exon 4 canonical splicing donor site mutation (Mutation 2)	F 5'-CTTCTGCGAAAGGCGTATTGTTCAAGTGGGG-3' R 5'-AACAATACGCCTTTTCGCAGAAGCCCCACTTCT-3'
Mutagenesis primers for Exon 4 C137Y mutation (Mutation 3)	F 5'- GTATGATGAGTACACCTCAGACG-3' R 5'- CGTCTGAGGTGTACTCATCATAC-3'
Mutagenesis primers for Exon 4 synonymous mutations (Mutation 4)	F 5'- GATGAGTGCACATCCGATGGAAGAGAAGATGG-CAGACTGTGG-3' R 5'- GCCATCTTCTCTTCCATCGGATGTGCACTCATCAT-ACTCC-3'
<i>mSEL1L</i> Full length (F1/R1)	F 5'- ATGCAGGTCCGCGTCAGGCTGTCGTTGCTGCT-3'; R 5'- CTACTGTGGTGGCTGCTGCTCTGG-3';
<i>hSEL1L</i> Full length (F1'/R1'):	F 5'- GCGGCTAGCATGCGGGTCCGGATAGGGCT-3'; R 5'- GCGAAGCTTTTACTGTGGTGGCTGCTGCTCTG-3';
<i>mSEL1L</i> Exon4 for acrylamide gel (F2/R2)	F 5'- AGCAAGACCTACGAAGAACT-3'; R 5'- GAAGGTACCGATATGCTTCTCTCT-3';
<i>mSEL1L</i> Exon4 for agarose gel (F3/R3)	F 5'- ATGCAGGTCCGCGTCAGGCTGTCGTTGCTGCT -3'; R 5'- CTACTGTGGTGGCTGCTGCTCTGG -3';
Minigene Exon4 (F4/R4)	F 5'- TGGTGAGCAAGGGCGAGG-3'; R 5'- CGTCCTTGAAGAAGATGGTGCG-3';
<i>hSEL1L</i> Exon4 (F5/R5)	F 5'- GGGGAAAGTGTACAGAAGATATCAG-3'; R 5'- GACACTCTCTCCAGGGCTTTG-3';

<i>mXbp1s</i>	F 5'- ACGAGGTTCCAGAGGTGGAG-3'; R 5'- AAGAGGCAACAGTGTCTCAGAG-3';
<i>hXBP1s</i>	F 5'- GAATGAAGTGAGGCCAGTGG-3'; R 5'- ACTGGGTCCTTCTGGGTAGA-3';
<i>mL32</i>	F 5'- GAGCAACAAGAAAACCAAGCA-3'; R 5'- TGCACACAAGCCATCTACTCA-3';
<i>hPPIA</i>	F 5'- GGCAAATGCTGGACCCAACACA-3'; R 5'- TGCTGGTCTTGCCATTCCTGGA-3'
Standard control oligos	CCTCTTACCTCAGTTACAATTTATA
ASO1 (+167bp - +192bp)	TGCCTCCTACTGAGCAATACTTACT
ASO2 (+137bp - +162bp)	AAAAGCCCCACTTTTCATCTGCTTT
ASO3 (+107bp - +132bp)	CATAGGTTGTAGCACACCACAGTCT
ASO4 (+77bp - +102bp)	CTTCCCTCCCATCTGATGTATATTC
ASO5 (+47bp - +86bp)	ACTCCTTATCTAGGAAAAGAAAAGG
ASO6 (+17bp - +42bp)	GGCAGGGCTCCCCATGTGCTGTGCC
ASO7 (-14bp - +11bp)	GGCGGTCAAAGCTGGAATGACAAGA