

# **Effect of small molecule eRF3 degraders on premature termination codon readthrough**

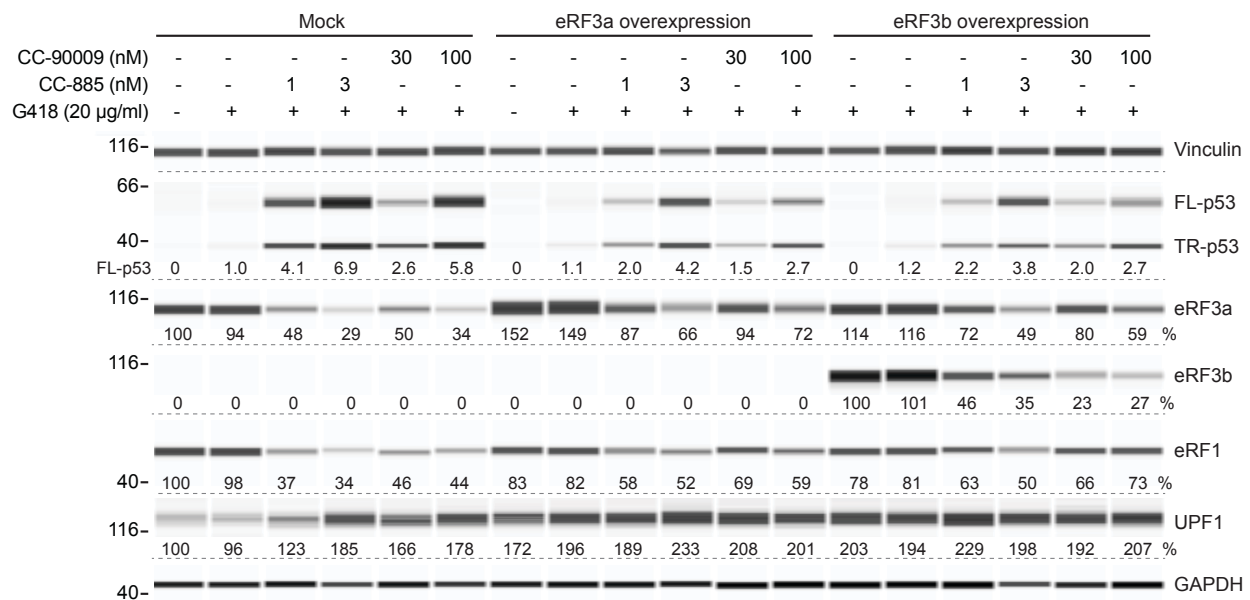
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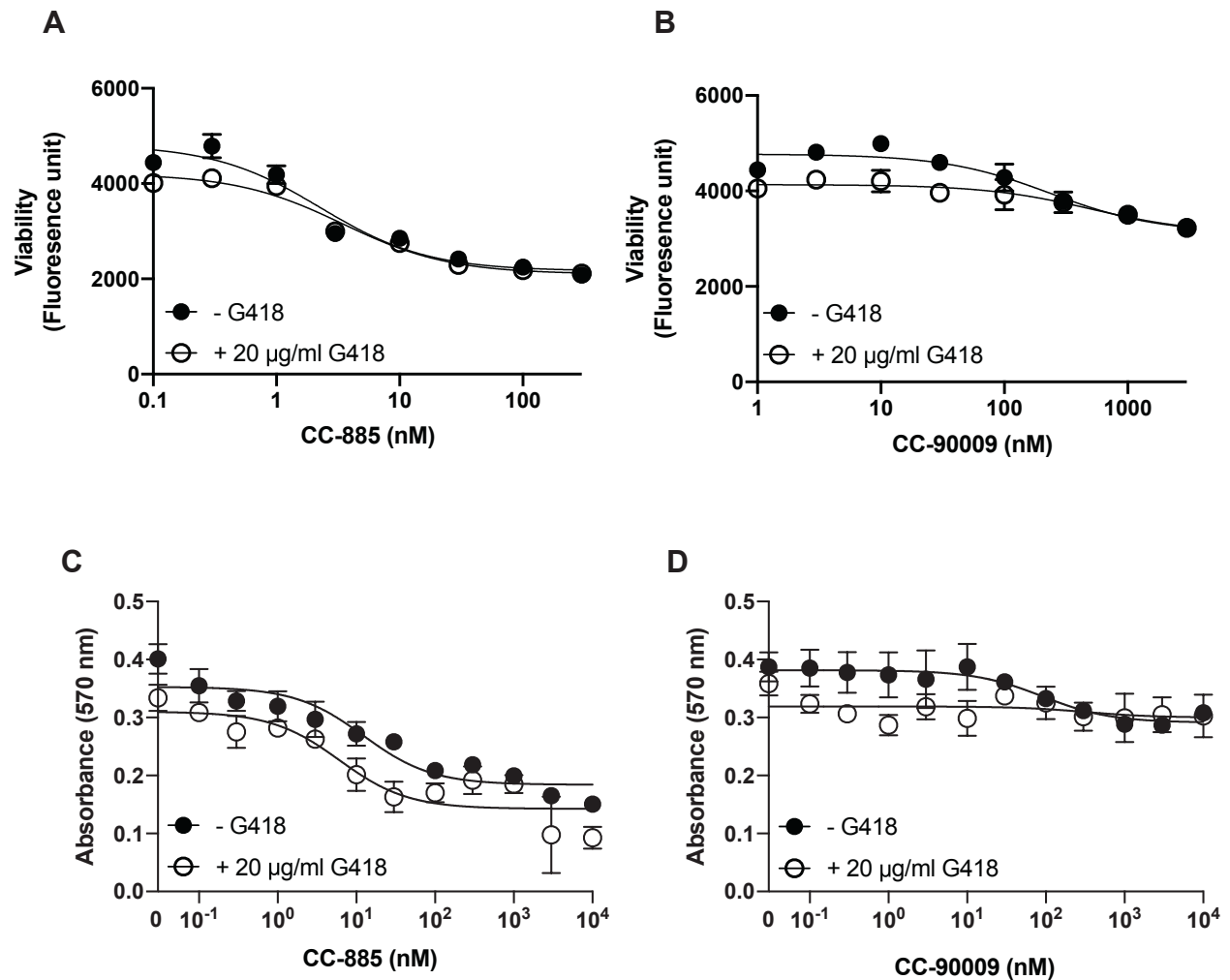
**Supplementary Figure S1.** Partial rescue of the effects of CC-885 and CC-90009. HDQ-P1

cells were transiently transfected with transfection reagents only (Mock), *eRF3a* or *eRF3b*

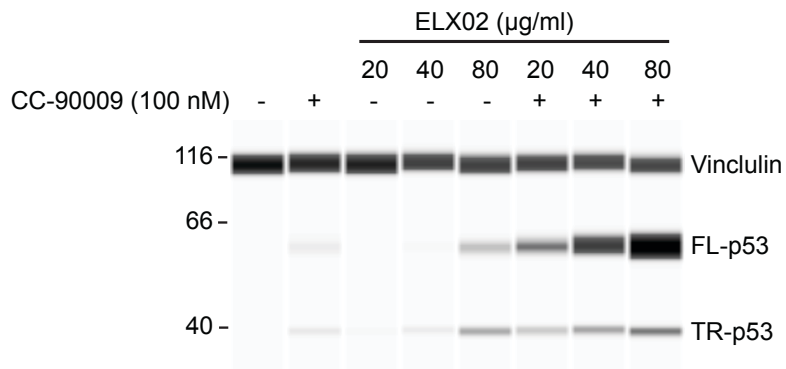
constructs and exposed to the indicated concentrations of CC-885 or CC-90009 in combination

with 20 µg/ml G418 for 48 h and p53, eRF3a, eRF3b, eRF1 and UPF1 were measured. Vinculin

and GAPDH were used as loading controls.



**Supplementary Figure S2.** Measurement of cell viability. Fibroblasts derived from an unaffected individual (**A**, **B**) or MPS I-H patient (**C**, **D**) were exposed to various concentrations of CC-885 (**A**, **C**) or CC-90009 (**B**, **D**) without or with 20  $\mu$ g/ml G418 for 48 h and cell viability was measured using the Promega ApoLive-Glo Multiplex Assay kit (**A**, **B**) or the MTT assay (**C**, **D**) in triplicate samples ( $\pm$  S.D.).



**Supplementary Figure S3.** Effect of CC-90009 on PTC readthrough by ELX-02.

HDQ-P1 cells were exposed to the indicated concentrations of ELX-02 with or without 100 nM CC-90009 for 48 h and p53 levels (full-length, FL-p53; truncated, TR-p53) were determined using automated capillary electrophoresis western analysis. Vinculin was used as loading control.