

Effect of small molecule eRF3 degraders on premature termination codon readthrough

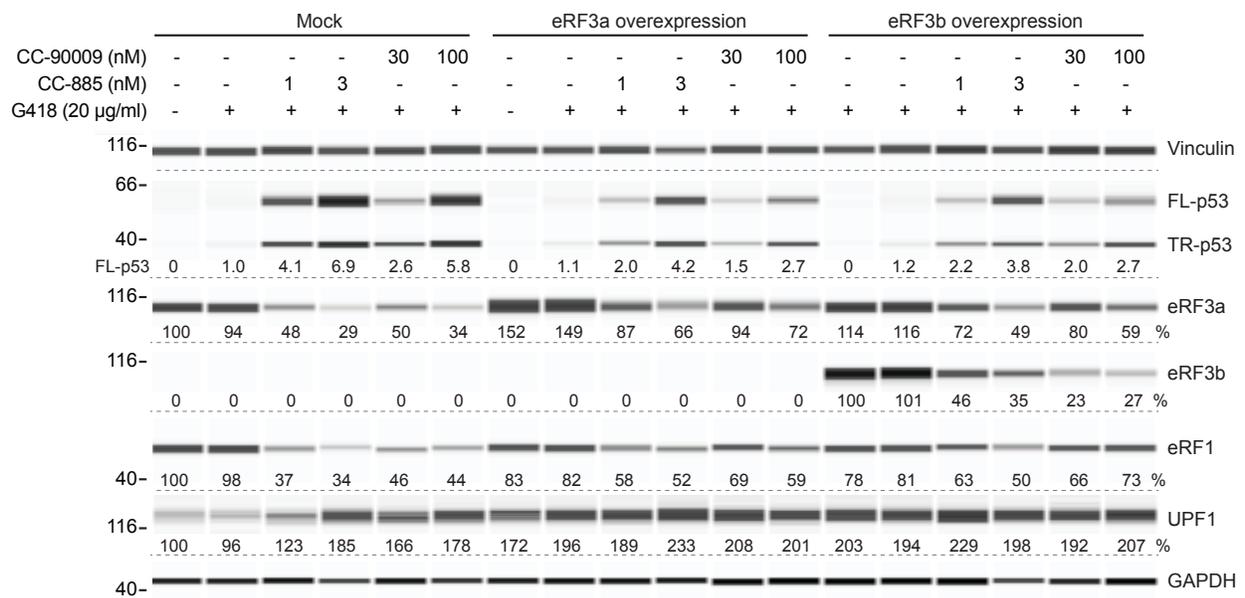
Alireza Baradaran-Heravi¹, Aruna D. Balgi¹, Sara Hosseini-Farahabadi¹, Kunho Choi¹, Cristina Has² and Michel Roberge¹

¹Department of Biochemistry and Molecular Biology, University of British Columbia, Vancouver, BC, Canada V6T 1Z3

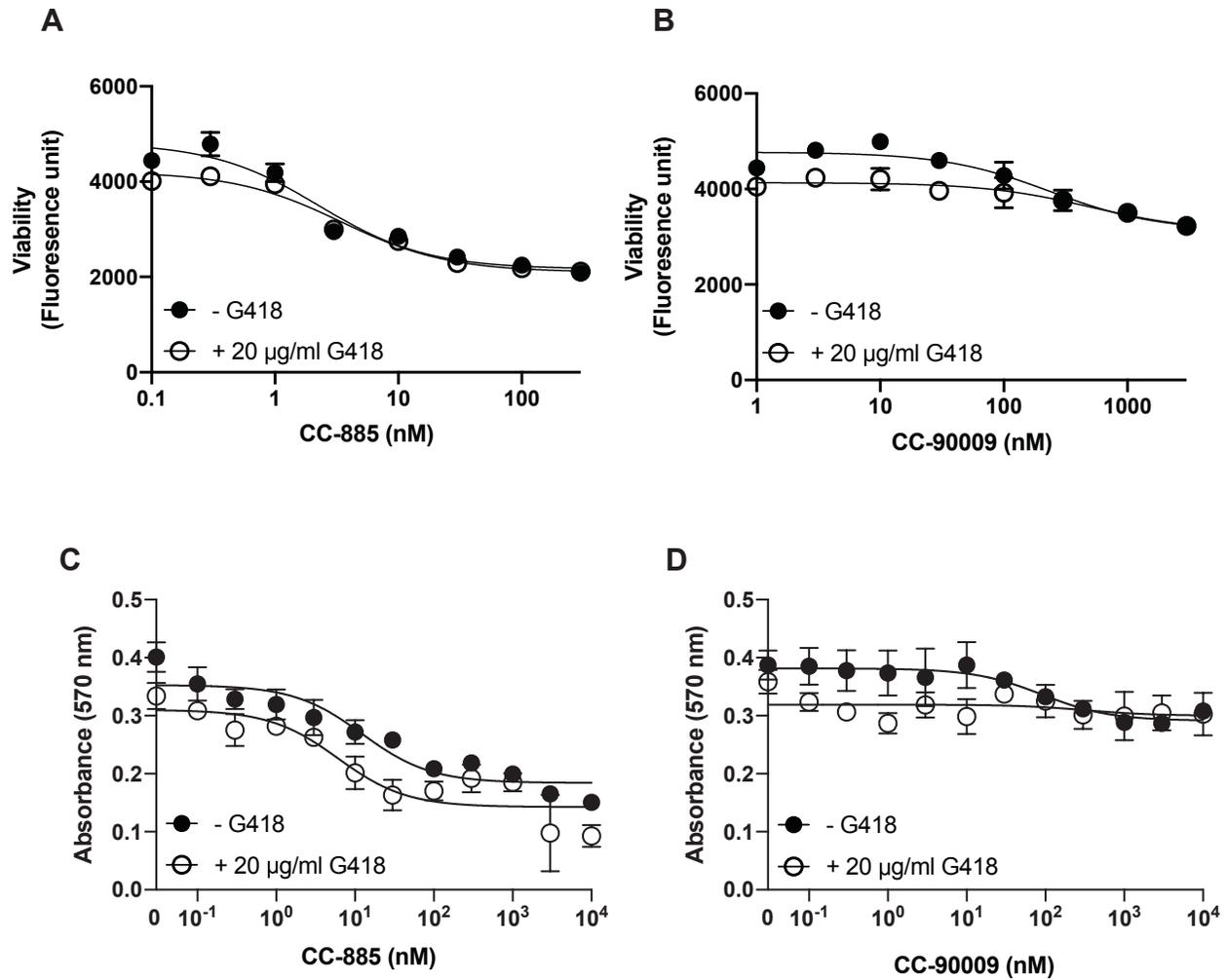
²Department of Dermatology, Medical Center-University of Freiburg, Faculty of Medicine, Freiburg, Germany

Corresponding author:

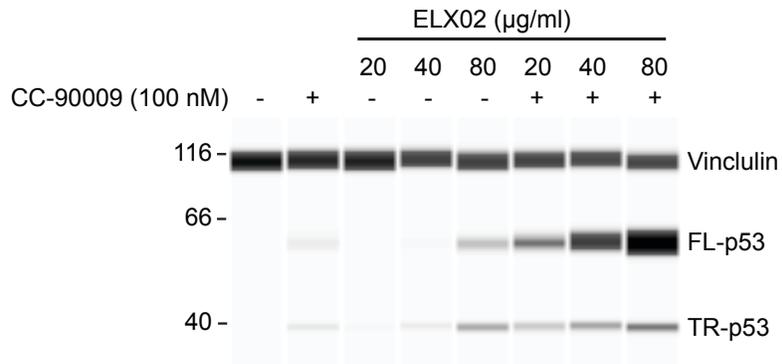
Michel Roberge
Dept Biochemistry & Molecular Biology
University of British Columbia
2350 Health Sciences Mall
Vancouver, BC, Canada, V6T1Z3
T. 604-652-9997
michelr@mail.ubc.ca



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 $\mu\text{g/ml}$ 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.