Potential new treatments for Huntington's disease

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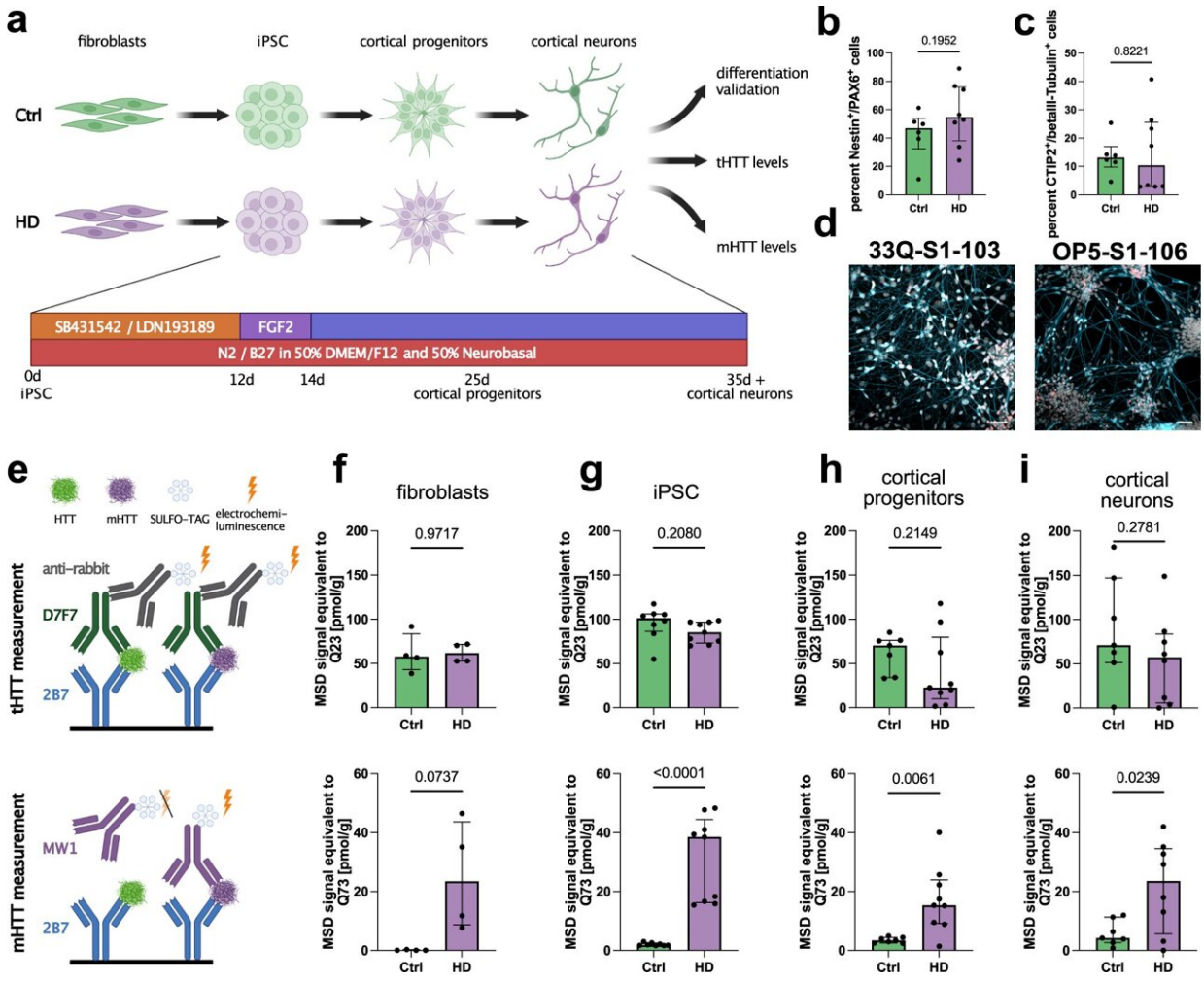

Mutant HTT is increased in HD patient-derived cells using an MSD assay. a Paradigm illustrating the HD patient-based disease model (fibroblasts, iPSC, cortical progenitors (25d old), and cortical neurons (35d old)) and readouts. Created with BioRender.com. b Bar plot depicting FACS quantification of

NESTIN/PAX6 double-positive cells. Statistics: Welch's test. Bars: median ± IQR. c Bar plot illustrating FACS quantification of bIII-Tubulin/CTIP2 double-positive cells. Statistics: Welch's test. Bars: median ± IQR. d Representative pictures of cortical neurons. Scale bar 50 µm. e Illustration depicting the MSD HTT quantification assay, where the added protein samples bind to 2B7 antibody, used for coating the plates. The SULFO-TAG coupled antibodies D7F7 and MW1 are added for quantification of total HTT and mutant HTT, respectively. Note: numeric values from 2B7/D7F7 assay (total HTT) cannot be directly set in relation to numeric values from 2B7/MW1 assay (mutant HTT). Created with BioRender.com. f Bar plots quantifying total (tHTT, top) and mutant (mHTT, bottom) levels in fibroblasts (4 Ctrl lines, 4 HD lines) with 2B7/D7F7 and 2B7/MW1 MSD assays, respectively. Statistics: tHTT: Welch's test (P value = 0.9717); mHTT Welch's test (P value = 0.0737). Bars: median ± IQR. g Bar plots quantifying total (tHTT, top) and mutant (mHTT, bottom) levels in iPSC (8 Ctrl lines, 9 HD lines) with 2B7/D7F7 and 2B7/MW1 MSD assays, respectively. Statistics: tHTT: Welch's test (P value = 0.2080); mHTT Mann–Whitney test (P value