



Fig. S2. Elimination of keratin aggregates is reduced by a proteasome inhibitor. To evaluate whether aggregated keratin is degraded through the proteasome-mediated degradation, differentiated epidermolytic ichthyosis (EI) cells were pre-incubated with 400 nM MG132 (a proteasome inhibitor) prior to heat stress. Treatment with MG132 increased keratin aggregate-containing cells to 35%, in comparison with 25% in vehicle-treated cells. Data are expressed as aggregate-containing cells/keratin 10+ cells (mean \pm standard deviation, $n=3$), * $p<0.05$, ** $p<0.01$ and *** $p<0.001$.