

## Supplemental Figure Legends

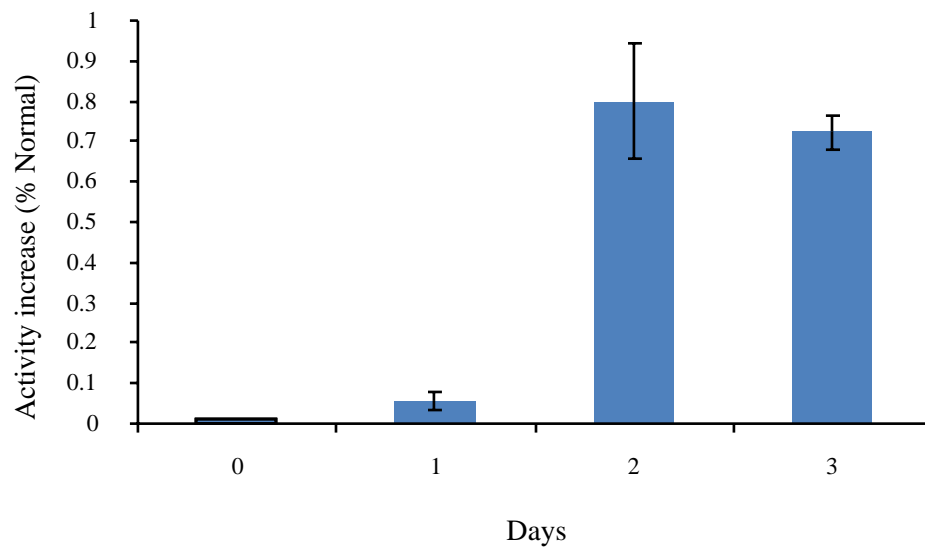
**Supplemental Figure 1. Time dependent effect of PTC124 on PPT1 enzyme activity in INCL cells.** Cells were treated with PTC124 (5  $\mu\text{g/ml}$ ) for varying length of time (0-3 days). Following PTC124 treatment PPT1 activity was measured and expressed as the mean  $\pm$  SD.

**Supplemental Figure 2. PPT1 activity induction by PTC124 in INCL patient cells with nonsense mutation.** Increase in PPT1 enzyme activity in PTC124 treated fibroblast cells (**A**) isolated from 3 different patients (P1, P2 and P3) and lymphoblasts (**B**) isolated from 3 different patients (P4, P5 and P6) was determined and is expressed as percentage of normal.

**Supplemental Figure 3. Effect of PTC124 on PPT1 enzyme assay.** To evaluate whether PTC124 interferes with PPT1 assay PTC124 (5  $\mu\text{g/ml}$ ) or DMSO were added to the reaction mixture for PPT1 assay. No significant alterations in PPT1 activity were noted due to the addition of PTC124.

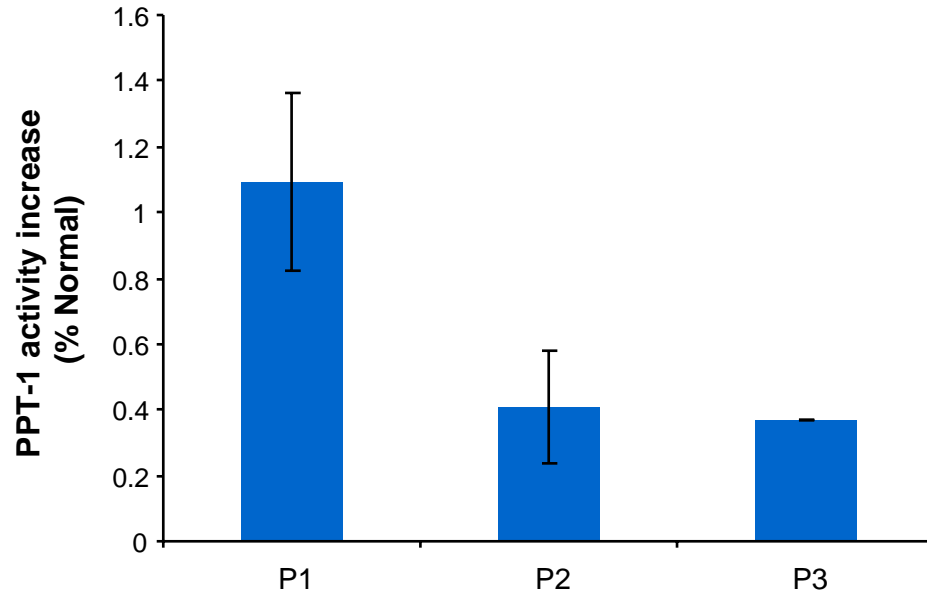
**Supplemental Figure 4.** Reduction in cell death in INCL cells after PTC124 treatment. Viability of INCL cells treated with or without PTC124 (5  $\mu\text{g/ml}$ ) for 1 week has been determined by trypan blue dye exclusion method. The results are presented as the mean of at least 3 independent experiments  $\pm$  SD.

# Supplemental Figure 1

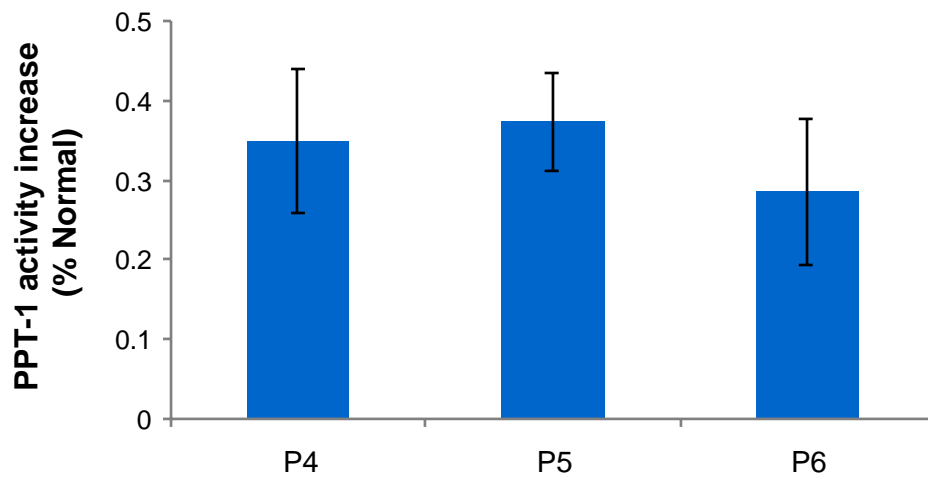


## Supplemental Figure 2

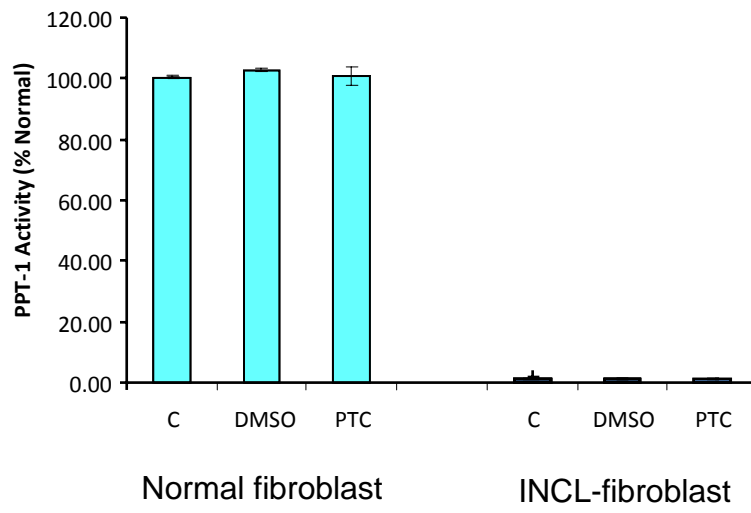
A



B



# Supplemental Figure 3



## Supplemental Figure 4

