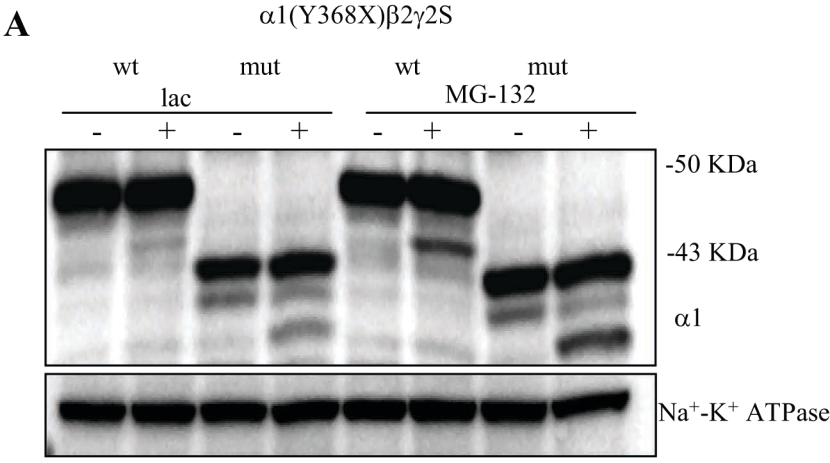

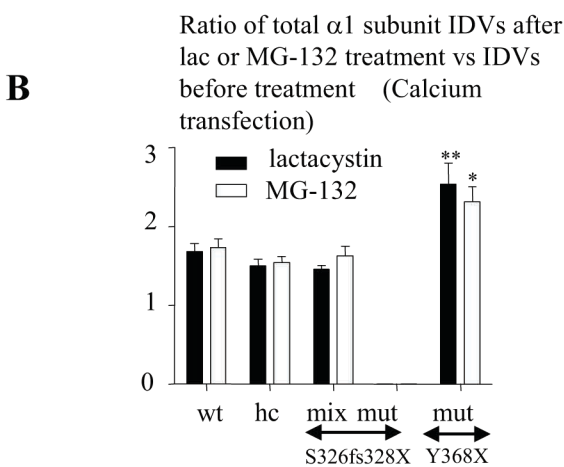

Supplementary Figure 3. ***The mutant α1(Y368X) protein, the NMD-insensitve α1 minigene subunit protein was increased with proteasome inhibition.*** **(A)** HEK 293T cells were cotransfected with β2 and γ2S subunit cDNAs and wild-type α1 subunit minigene (ratio of 1:1:1, (wt) or mut α1(Y368X) subunit minigene (ratio of 1:1:1 (mut). The cells were incubated with or without lactacystin (10 μM) (left) or MG-132 (3μM) (right) for 6 hr before harvest. The total cell lysates (15 μg/lane) were analyzed by 10% SDS-PAGE. With proteasomal inhibition, there was an increase in the α1(Y368X) subunit main band (43 KDa) and appearance of a third band that was likely an unglycosylated form of the mutant band or a degraded fragment. **(B)** The histogram displays the ratio of increase of protein with lactacystin or MG-132 treatment over the protein without treatment. The treatment with either lactacystin or MG-132 increased the total wild-type a1 subunit expression in wt, hc and mixed conditions and mutant α1(Y368X) subunit protein expressions. There was no mutant α1(S326fs328X) protein detected by Western blot either before or after lactacystin or MG-132 treatments (* $p<0.05$,** $p<0.01$ vs wild-type n = 4).