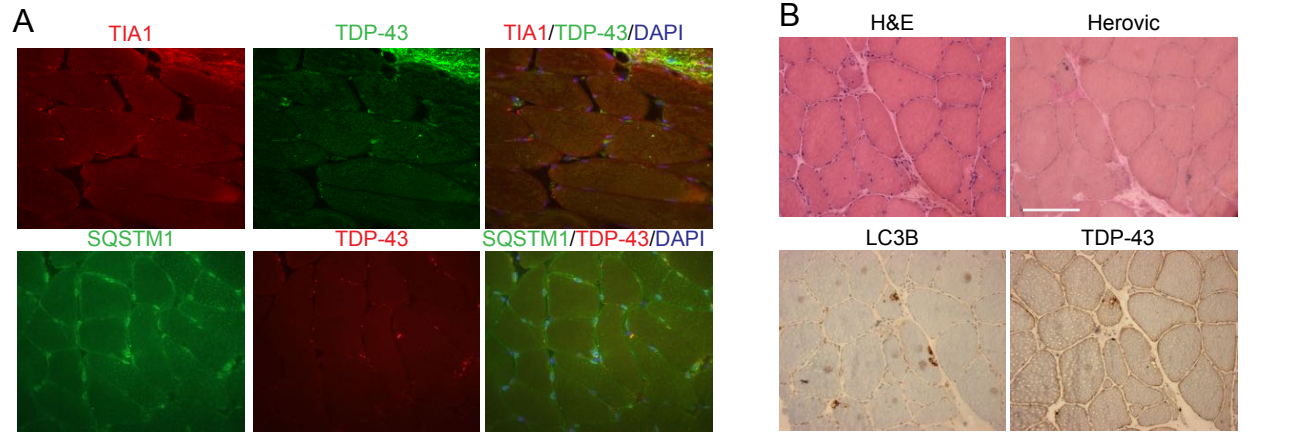
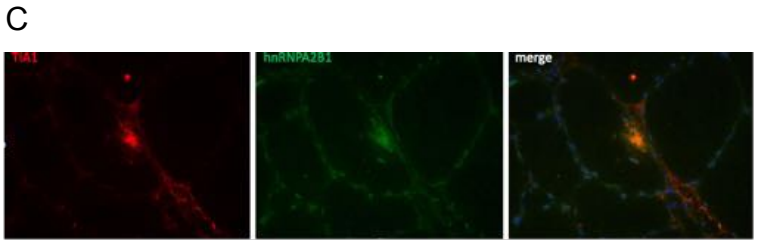
Supplemental Figure 1: Histopathology of SQSTM1-TIA1 patients and controls: A) Immunofluorescent staining of TIA1 (red in upper panel) or TDP-43 (green) and SQSTM1 (green in lower panel). B) Serial sections from patient muscle biopsy histochemically stained with hematoxylin and eosin (H&E) and Herovici staining demonstrating myopathy with rimmed vacuoles. Immunohistochemistry of similar sections stained with antibodies against autophagic marker LC3B and TDP-43. C) Co-immunofluorescent staining of patient muscle with antibodies to TIA1 (red) and HNRNPA2B1 (green). D) Patient and control muscle immunoblot for TIA1 and SQSTM1. Myosin is shown as a loading control.

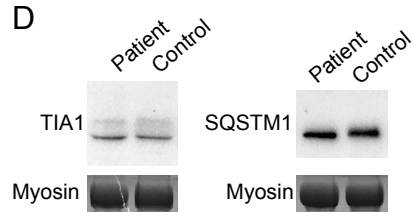
Supplemental Figure 2: Immunoblot images of GFP-TIA1 and live cell images following photobleaching. A) Immunoblot from control MEFs expressing GFP-TIA1-Empty, WT, NS, or EK for GFP and SQSTM1. GAPDH is shown as a loading control. B) Representative fluorescence images of GFP-TIA1-WT, EK, or NS stress granules from live MEFs cells expressing GFP-TIA1, EK, or NS in (Figure 2E). The circle indicates the photobleached area. Scale is 1μm.

Supplemental Figure 3: Immunostaining of stress granules accumulating ubiquitin conjugates and bar graph of cells treated with proteasomal inhibitor (MG132) or lysosomal autophagic inhibitor (BafilomycinA). A) Immunofluorescent images of control or p62-/- MEFs for TIA1 and ubiquitin (FK2) before HS, after incubation at 42°C for 1hr and following 30 min HS recovery. Representative data were pooled from three independent experiments (n=150~200). Blue color indicates DAPI staining. B) Bar graph of the percentage of MEFs cells containing TIA1/G3BP1 positive SGs. Control of p62-/- MEFs were incubated at 42°C for 1hr (HS). They were subsequently returned to 37°C and incubated with DMSO, MG132, or BafilomycinA for 1hr. All error bars are mean ± SEM. * denotes p value <0.05.

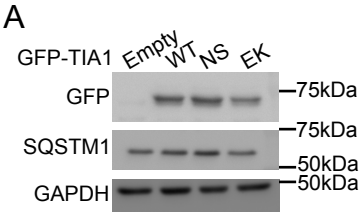
Supplemental Figure1.

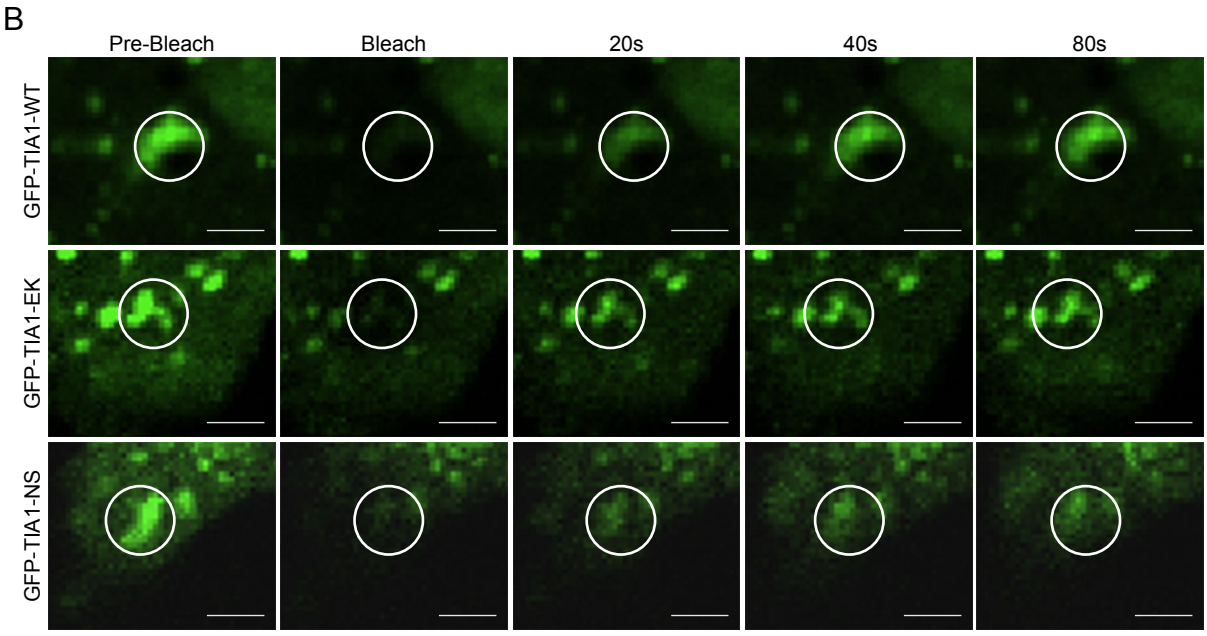




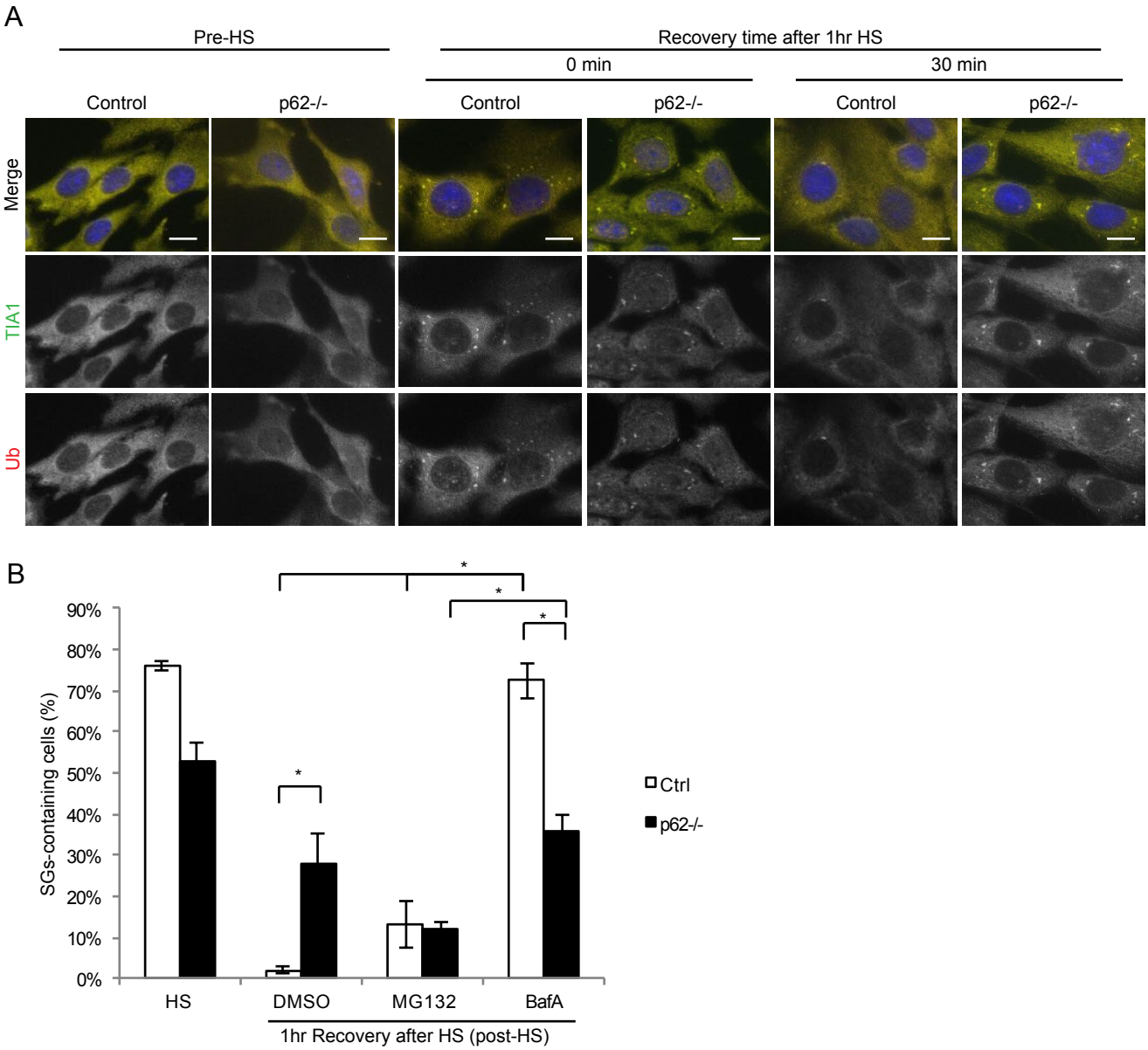


Supplemental Fig 2.





Supplemental Fig 3.



Patient	Gender	Age	Onset	*SQSTM1*	*TIA1* SNP
1	F	67	67	P392L/WT	WT/WT
2	F	71	56	G425R/WT	WT/WT
3	M	66	53	P392L/WT	WT/WT
4	F	83	63	P392L/WT	WT/WT
5	M	68	60	P392L/WT	WT/WT
6	M	59	56	G425R/I424S	WT/WT
7	F	69	55	G425R/WT	WT/WT
8	F	71	55	M404V/WT	WT/WT
9	F	74	47	P392L/WT	WT/WT
10	F	78	69	P392L/WT	WT/WT
11	F	81	49	P392L/WT	WT/WT
12	M	74	45	P392L/WT	WT/WT
13	M	74	64	P392L/WT	WT/WT
14	M	71	66	P392L/WT	WT/WT
15	F	69	50	M404T/WT	WT/WT
16	M	91	54	P392L/WT	WT/WT
17	M	72	58	P392L/WT	WT/WT
18	M	80	78	P392L/WT	WT/WT
19	F	62	54	G425R/WT	WT/WT
20	M	64	54	P392L/WT	WT/WT
21	F	67	48	P392L/WT	WT/WT
22	M	82	57	P392L/WT	WT/WT
23	M	76	72	P392L/WT	WT/WT
24	F	73	45	P392L/WT	WT/WT
25	F	72	67	P392L/WT	WT/WT
26	M	73	70	P392L/WT	WT/WT
27	M	89	60	P392L/WT	WT/WT
28	F	85	84	P392L/WT	WT/WT
29	M	83	54	E396X/WT	WT/WT

30	F	82	75	P392L/WT	WT/WT
31	M	72	26	I424S/WT	WT/WT
32	F	84	79	P392L/WT	WT/WT
33	F	74	64	M404V/WT	WT/WT
34	M	72	55	P392L/WT	WT/WT
35	F	86	76	P392L/WT	WT/WT
36	M	62	55	P392L/WT	WT/WT
37	M	75	63	P392L/WT	WT/WT
38	M	71	72	P392L/WT	WT/WT
39	F	80	70	P392L/WT	WT/WT
40	M	79	41	P392L/WT	WT/WT
41	F	70	70	P392L/WT	WT/WT
42	M	74	50	P392L/WT	WT/WT
43	F	67	54	P392L/WT	WT/WT
44	M	76	59	P392L/WT	WT/WT
45	F	59	51	P392L/WT	WT/WT
46	F	66	40	P392L/WT	WT/WT
47	F	78	52	P392L/WT	WT/WT
48	M	72	70	P392L/WT	WT/WT
49	F	71	66	P392L/WT	WT/WT
50	F	73	67	P392L/WT	WT/WT

Supplemental Table 1: Genotype of PDB patients