

Figure 1a

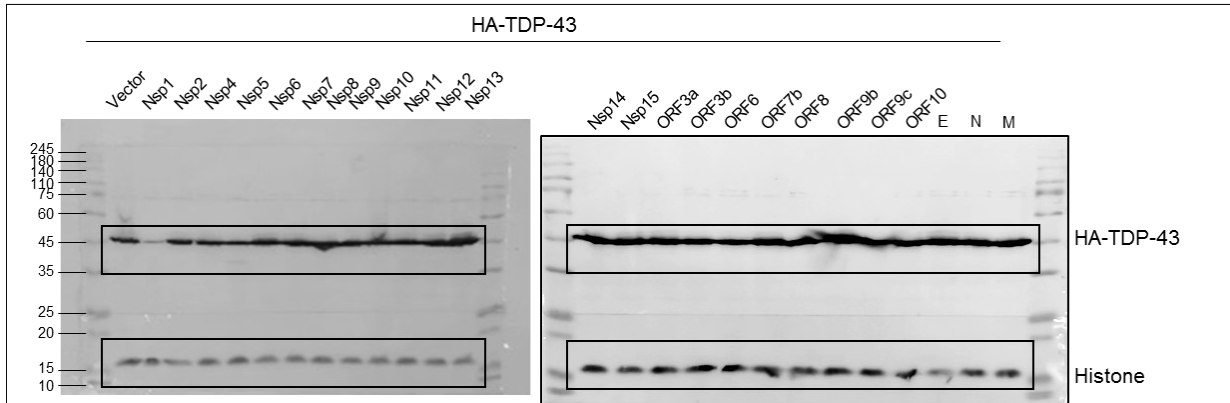


Figure 1b

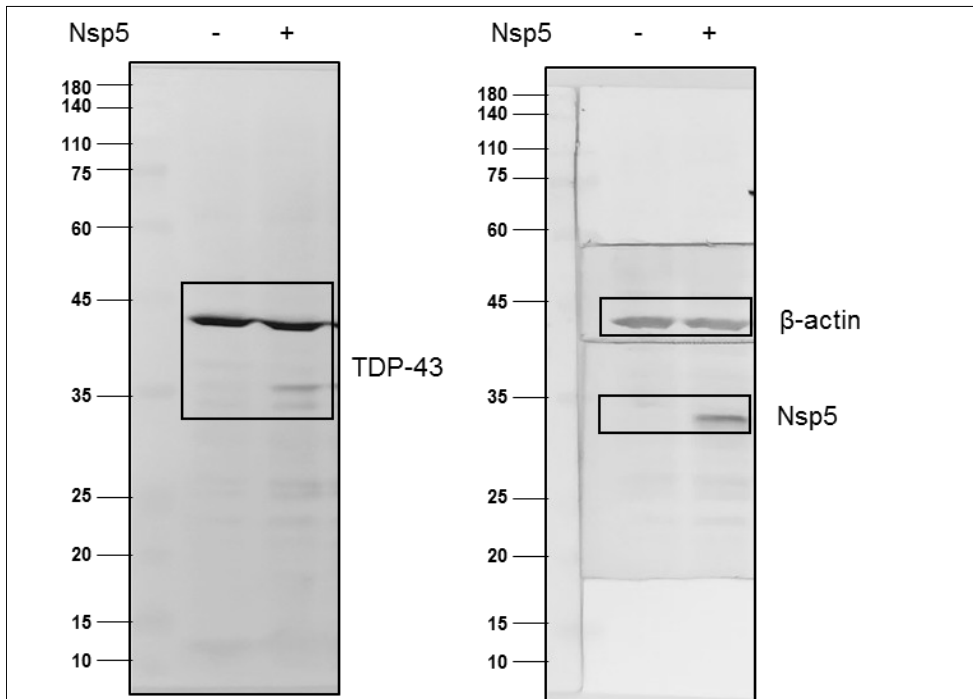


Figure 1c

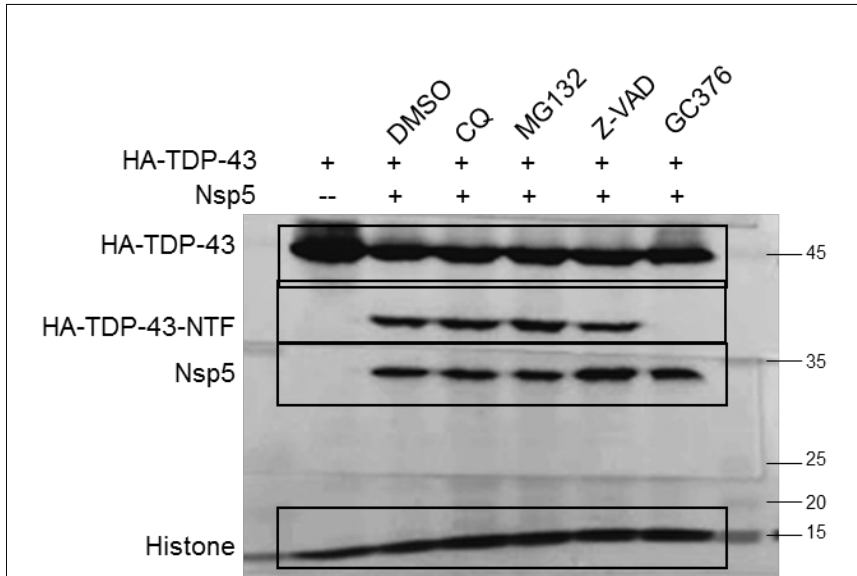


Figure 1d

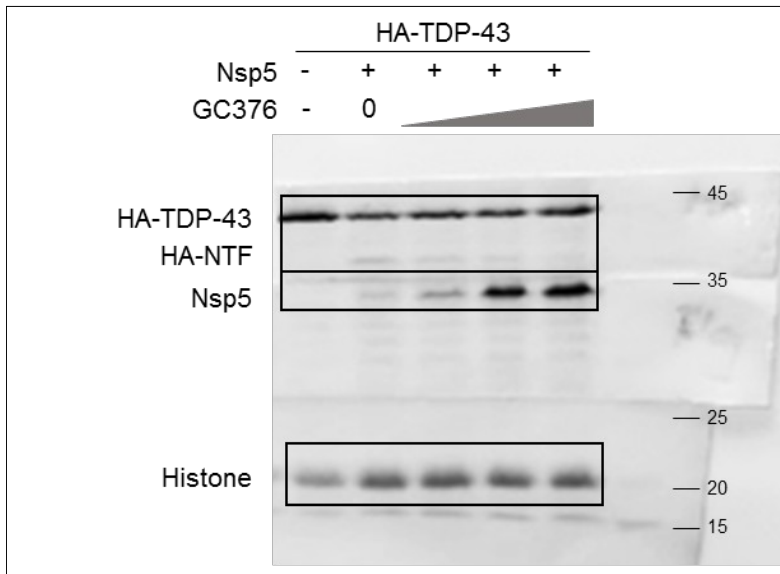


Figure 1e

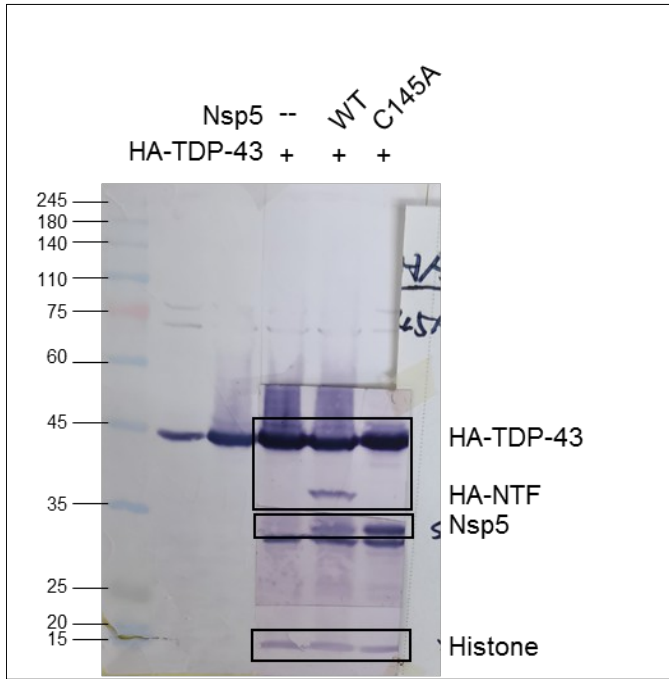


Figure 1f

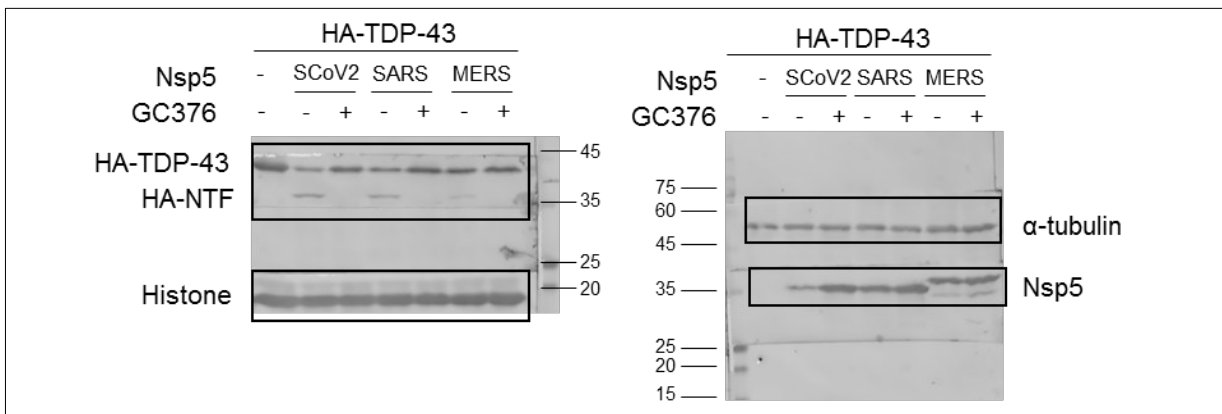


Figure 1i

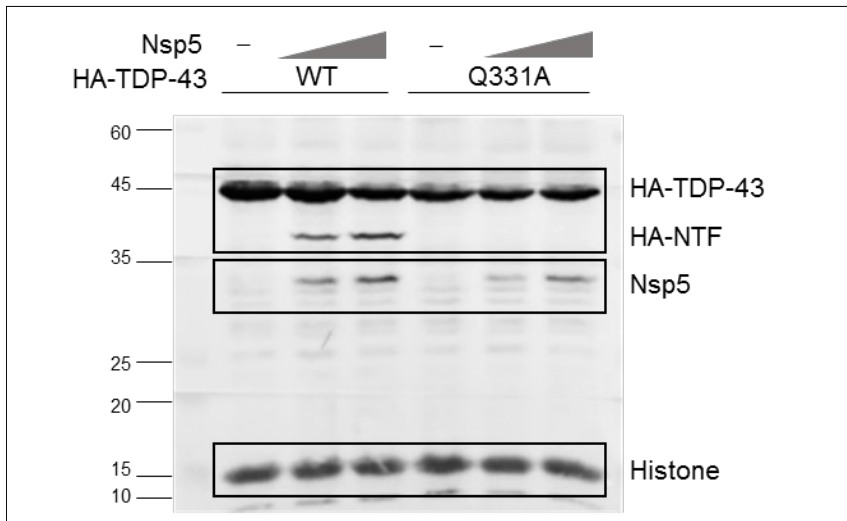


Figure 1j

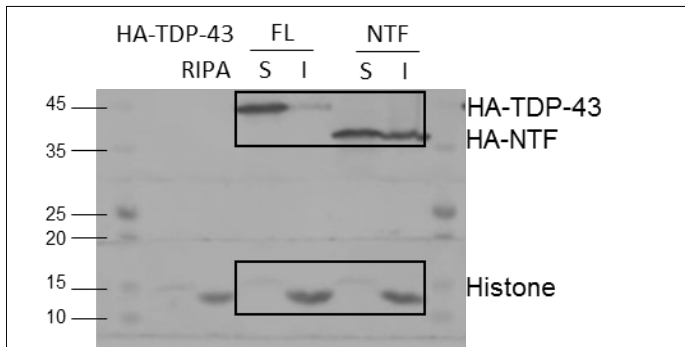


Figure 1k

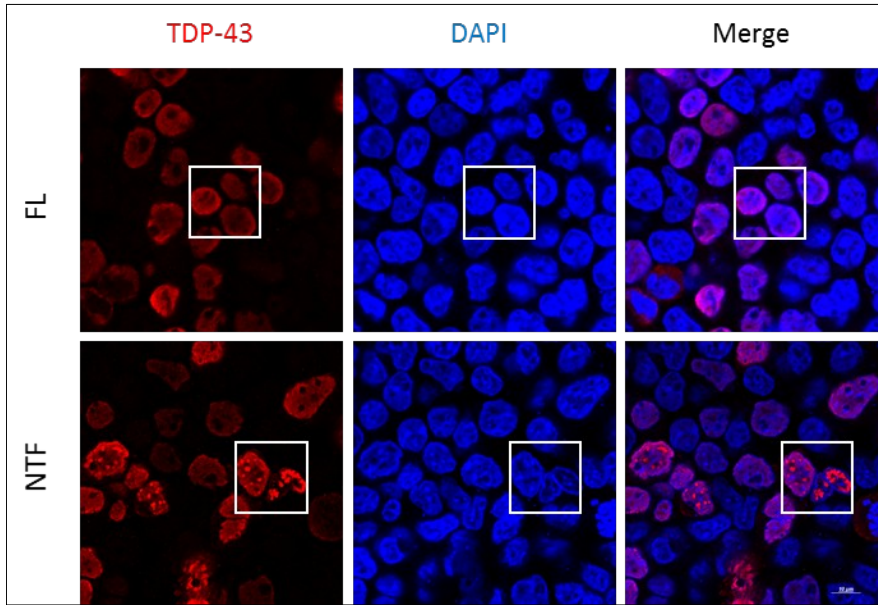


Figure 1m

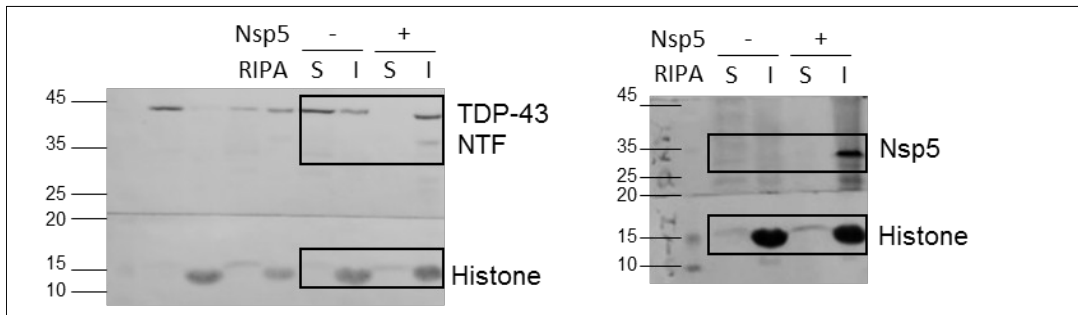


Figure 1n

SH-SY5Y

TDP-43	-	WT			Q331A		
Nsp5(ng)	0	0	100	200	0	100	200
	0.372	0.471	0.517	0.585	0.515	0.521	0.563
	0.403	0.457	0.547	0.607	0.548	0.536	0.591
	0.404	0.469	0.546	0.604	0.521	0.506	0.58

Figure 1o

SH-SY5Y

	Vector	Nsp5	Nsp5+GC376
	0.169	0.205	0.19
	0.189	0.213	0.197
	0.186	0.212	0.192

Figure 1p

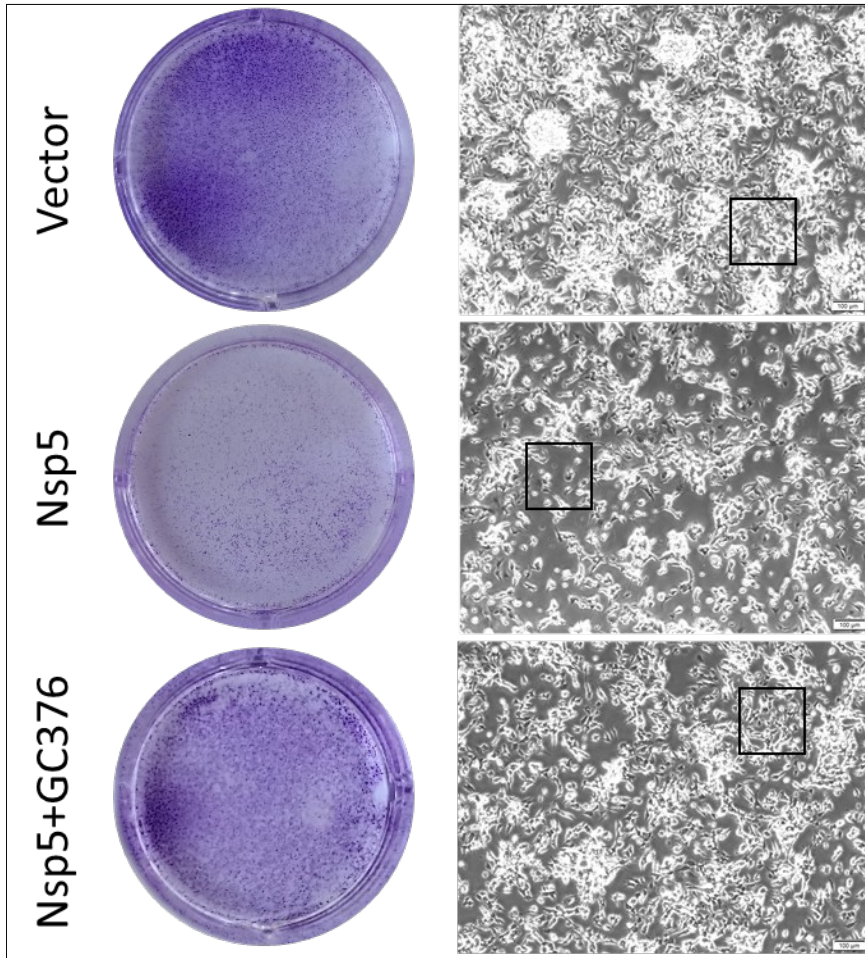


Figure S1b

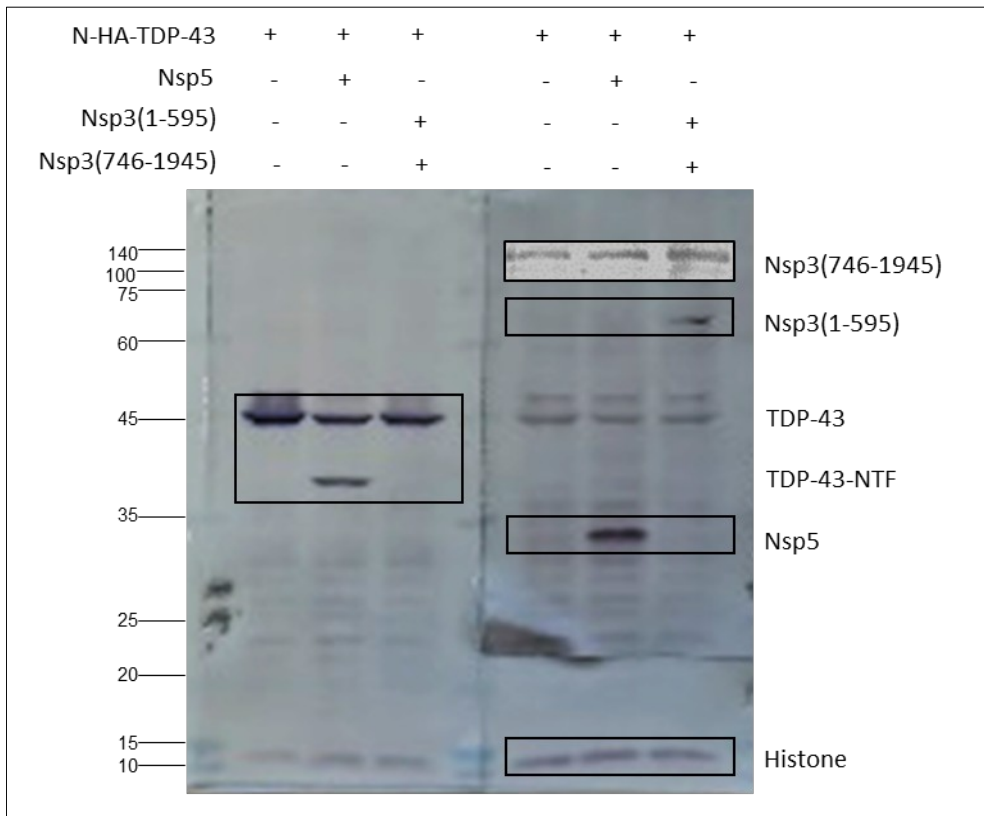


Figure S1c

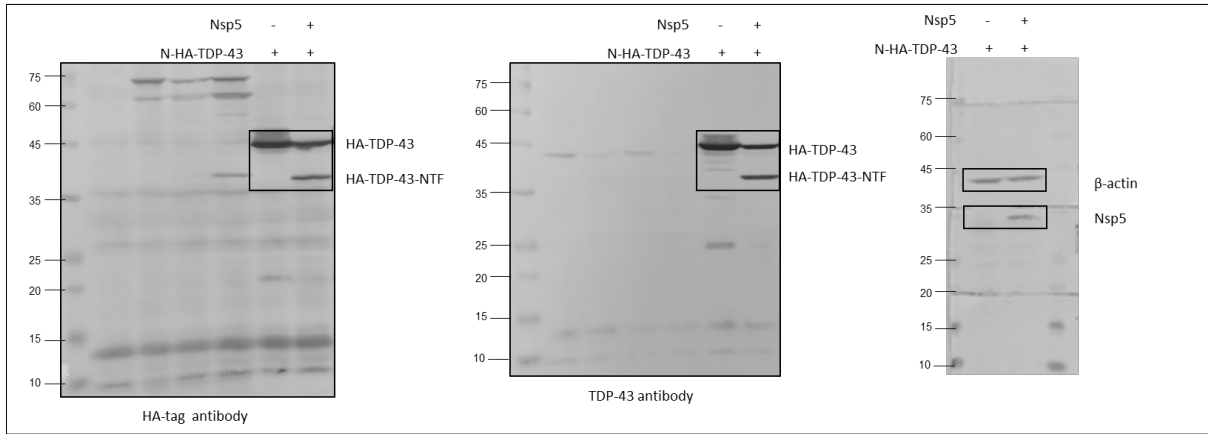


Figure S2b

GC376(μM)	Blank	0	0.001	0.01	0.1	1	10	100
OD450	0.337	1.931	1.959	2.041	1.823	1.946	1.897	1.734
	0.291	1.743	1.884	1.915	1.661	1.691	1.561	1.401
	0.309	1.725	1.876	1.676	1.851	1.614	1.675	1.299

Figure S4

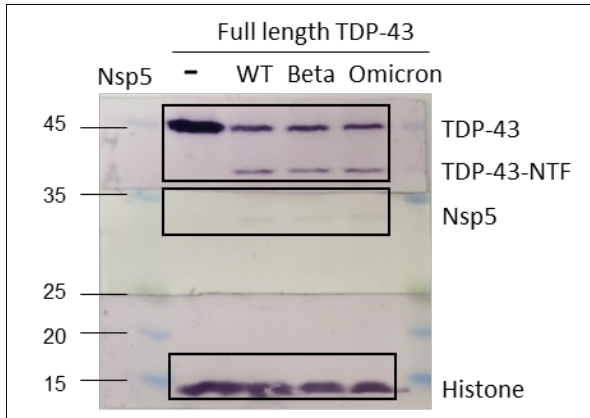


Figure S6

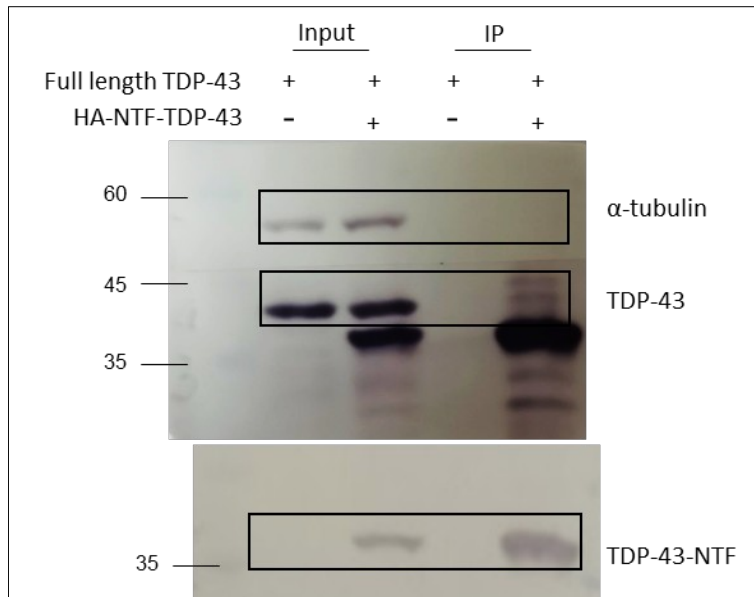


Figure S7

T98G

TDP-43	-	WT			Q331A		
Nsp5(ng)	0	0	100	200	0	100	200
	0.391	0.408	0.424	0.426	0.413	0.433	0.457
	0.375	0.386	0.415	0.428	0.428	0.437	0.47
	0.371	0.381	0.4	0.42	0.419	0.421	0.457

Figure S8

SH-SY5Y	Vector	TDP-43-Q331A	Nsp5	Nsp5+TDP-43-Q331A
	0.408	0.416	0.431	0.408
	0.413	0.421	0.442	0.421
	0.404	0.422	0.442	0.421

Figure S9

SH-SY5Y

GC376(μ M)	Blank	0	0.01	0.1	1	10	100
OD450	0.408	0.557	0.566	0.573	0.547	0.578	0.556
	0.415	0.563	0.575	0.564	0.594	0.547	0.595
	0.39	0.556	0.576	0.545	0.556	0.571	0.565