# Supplementary Figure 1- Uncropped scans.

## **Main Figures**



Actin  $\frac{55}{35}$ 



35 - -

# Figure 4b











## **Extended Data**

#### Ext Data Figure 1b





#### Ext Data Figure 1c



















Ext Data Figure 3e





Ext Data Figure 3f







### Ext Data Figure 7c







## Ext Data Figure 8c



Ext Data Figure 8e











#### Ext Data Figure 9d (continue)



# IP K63 Ub 250-130-K63 Ub 250-



K48 Ub 250-130polyQ



polyQ





#### Ext Data Figure 9d

Input

### Ext Data Figure 10a



	Input	IP
<b>AR</b>	,	
100-	17	
AR		
130— -		(*******
100- *		li
Beclin 1		
70		
70		
55 — -		
55 — · 35 — ·	[]	
55 - ' - 35 IgG → <sup>55</sup>	[]	
$10 - 1$ $55 - 35 - 35 - 35$ $IgG \rightarrow 55 - 55$ Actin 35 - 35		
10 - 1 $55 - 35 - 35 - 35 - 35 - 35 - 35 - 35 -$		

Actin  $55 \rightarrow$ 55 — Actin 35 — Ext Data Figure 10b



35 —

#### Ext Data Figure 10c



## Ext Data Figure 10b (continue)



Table S1.	<b>Full statistical</b>	analysis of	the data from	Figure 1	and Figure 4.

Figure	P value for test	P value for post-test
Figure 1a	Two-way ANOVA (column factor shRNA *** P<0.001, row factor BafA1 *** P<0.001, interaction *	Bonferroni's post-test (* P<0.05, *** P<0.001, N.S. not significant).
	P<0.05).	
Figure 1b	Two-way ANOVA (column factor shRNA ** P<0.01, row factor BafA1 *** P<0.001, interaction P value N.S).	Bonferroni's post-test (** P<0.01, *** P<0.001, N.S).
Figure 1c	One-way ANOVA (** P<0.01).	Post-hoc Tukey's test (* P<0.05, ** P<0.01).
Figure 1d	Two-way ANOVA (column factor siRNA ** P<0.01, row factor fasting ** P<0.01, interaction P value N.S).	Bonferroni's post-test (* P<0.05, ** P<0.01, N.S).
Figure 1e	Two-way ANOVA (column factor siRNA * P<0.05, row factor fasting ** P<0.01, interaction P value N.S).	Bonferroni's post-test (* P<0.05, ** P<0.01, N.S).
Figure 4 c	Two-way ANOVA (column factor HD ** P<0.01, row factor fasting N.S, interaction P value N.S).	Bonferroni's post-test (** P<0.01, N.S).
Figure 4 d	Two-way ANOVA (column factor HD *** P<0.001, row factor fasting * P<0.05, interaction * P<0.05).	Bonferroni's post-test (** P<0.01, *** P<0.001, N.S).

 Table S2. List of patient-derived fibroblasts analysed in this study.

Disease	Catalogue number	Ref in the paper
HD	GM04285	HD1
	GM04287	HD 2
	GM04476	HD 3
	GM04867	HD 4
	HD 940-01	HD 5
	HD960-01	HD 6
	HD305-01	HD 7
DRPLA	AT2140102	DRPLA 1
	GM13716	DRPLA 2
	GM13717	DRPLA 3
SCA3	GM06151	SCA3 1
	GM06153	SCA3 2
SCA7	GM03561	SCA7 1