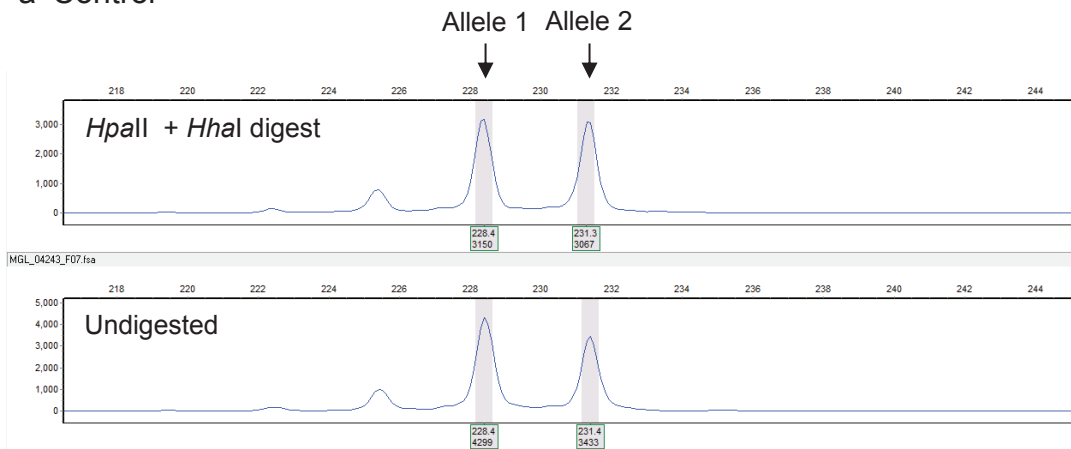
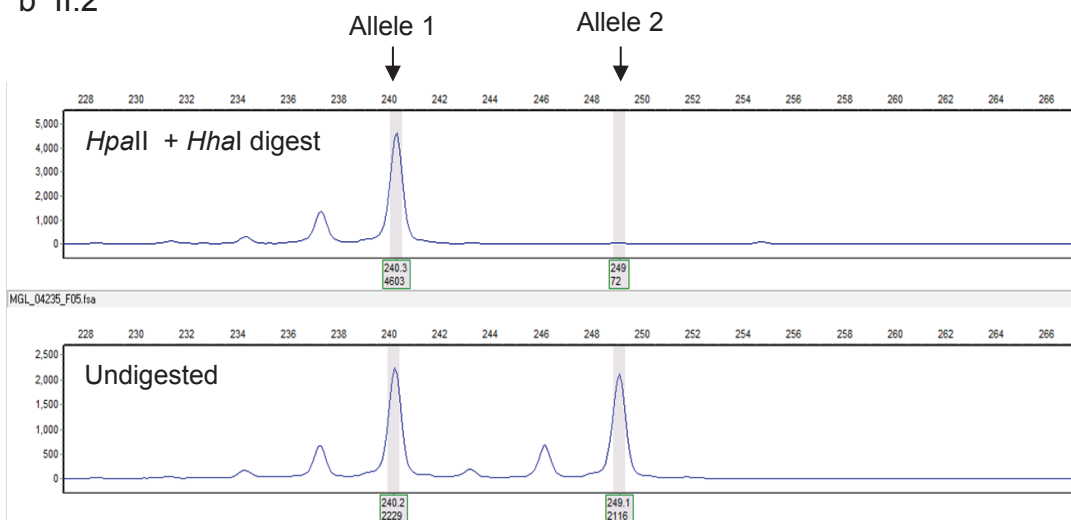


Supplementary Figure 1

a Control



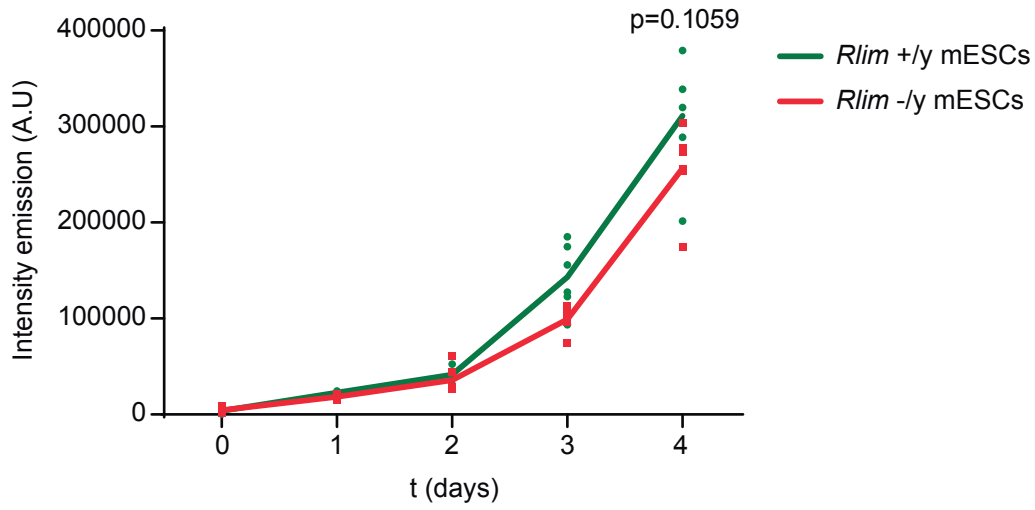
b II:2



Subject	Fragment length (bp) allele 1	Fragment length (bp) allele 2	Cells with allele 1 as active X-chromosome (%)	Cells with allele 2 as active X-chromosome (%)
Control	228	231	50	50
II:2	240	249	5	95

Supplementary Figure 1: Skewed X-chromosome inactivation in the proband's mother (II:2). DNA was subjected to methylation sensitive restriction enzyme digestion followed by PCR and fragment analysis. The inactive X-chromosome is not digested. Figure was generated using SoftGenetics GeneMarker v2.7 (<https://softgenetics.com/GeneMarker.php>).

Supplementary Figure 2



Supplementary Figure 2: Proliferation curves for indicated mESC lines cultured in LIF/FCS as determined by MTS assay. Data are represented as mean \pm standard error of the mean (n=3). Statistical significance was determined by student's t-test. Figure was generated using Graphpad Prism v8.0 (<https://www.graphpad.com/scientific-software/prism/>).

Supplementary Figure 3: Unprocessed gels and blots

Figure 2a

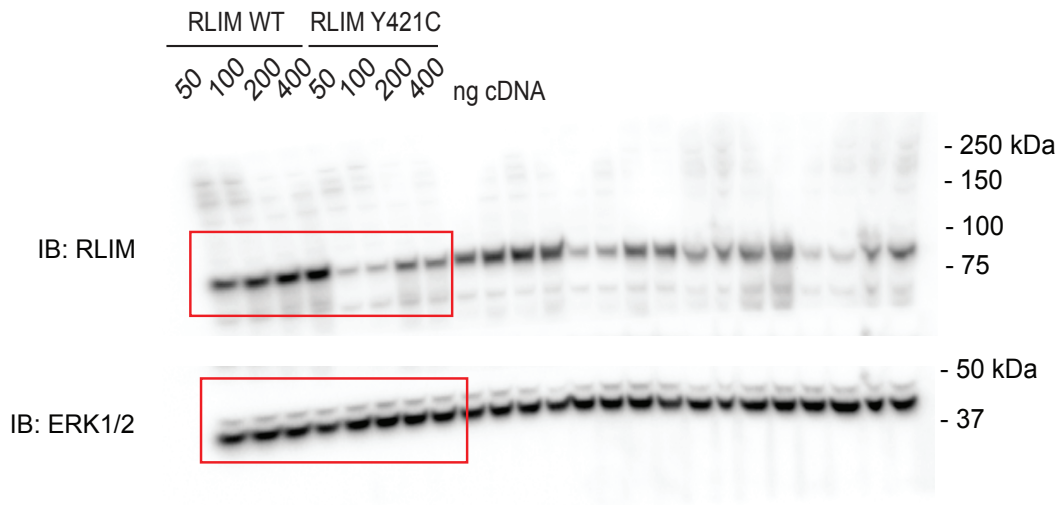


Figure 2c

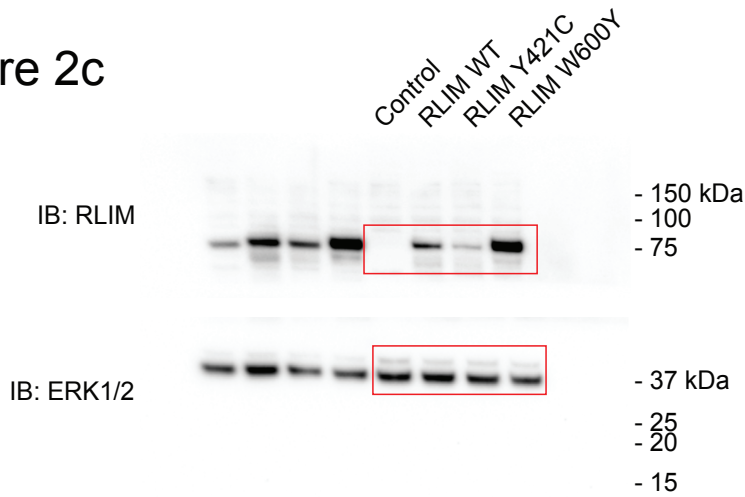


Figure 2c

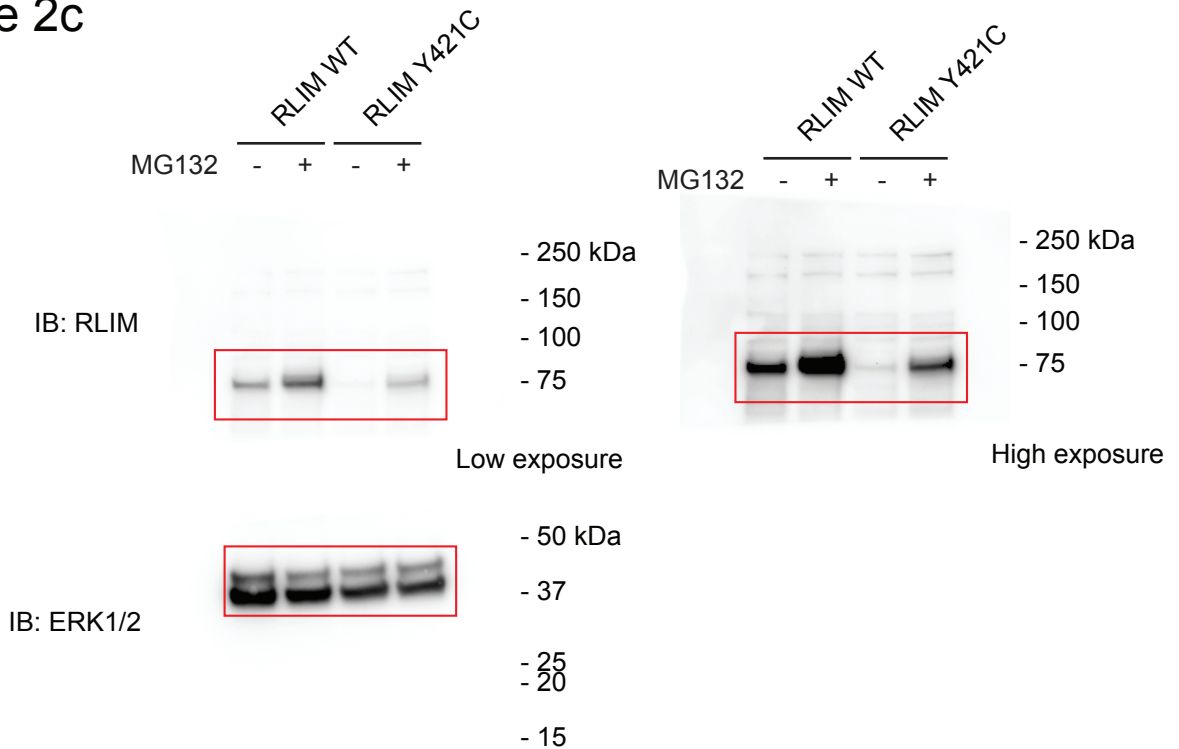


Figure 3a

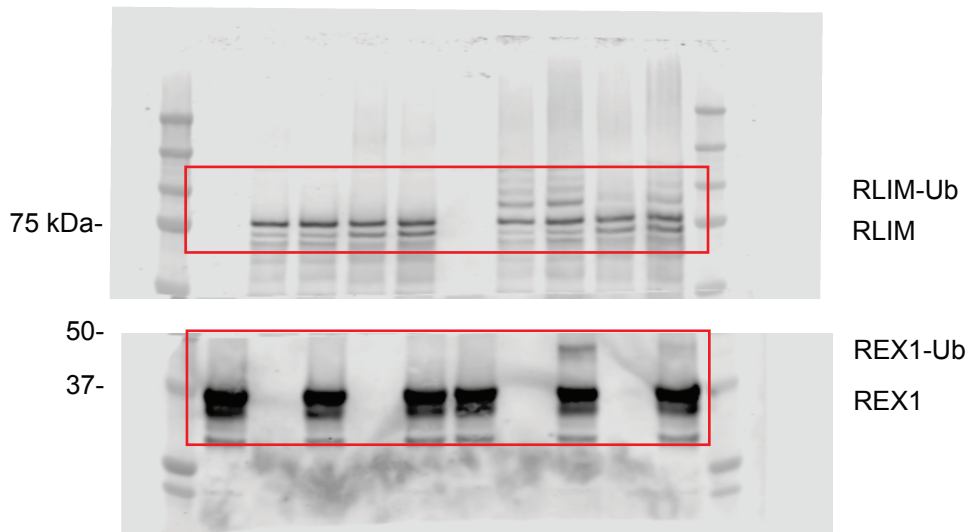
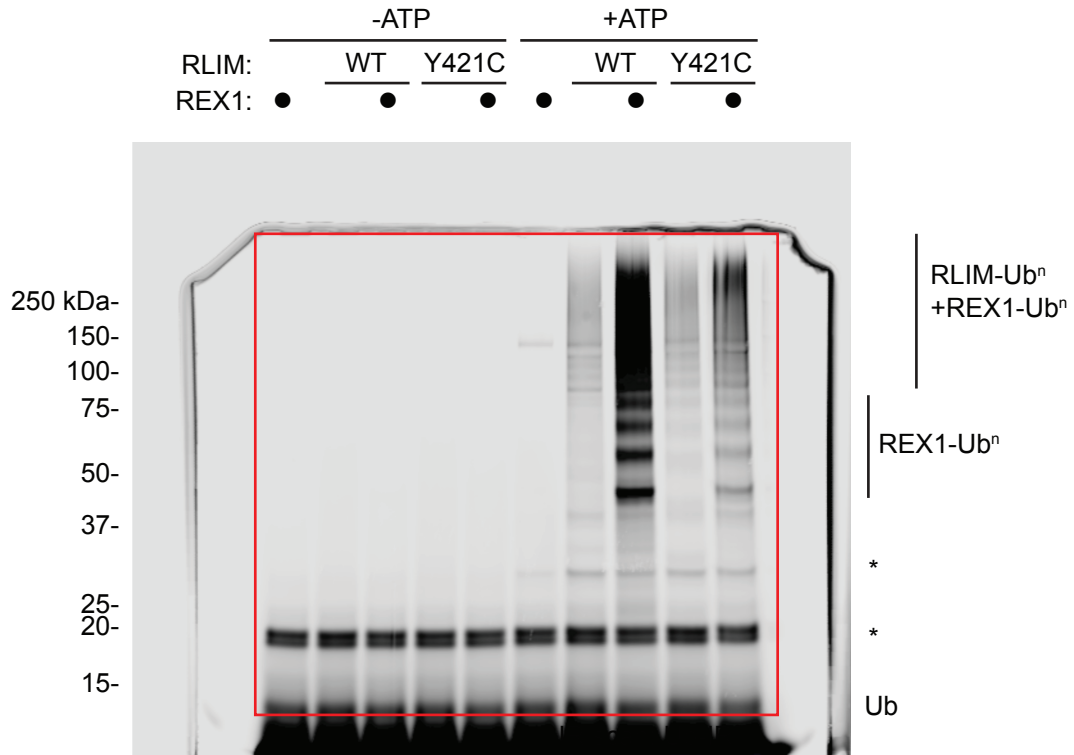


Figure 4b

