

Figure S1

**a**

PATIENT	SEX	AGE (At Biopsy)		LOCALIZATION
1	♂	14w	Fetus	QUADRICEPS
2	♂	22w	Fetus	QUADRICEPS
3	♂	22w	Fetus	QUADRICEPS
4	♂	37w	Fetus	QUADRICEPS
5	♂	37w	Fetus	QUADRICEPS
6	♂	17	Teenager	LEFT DELTOID
7	♂	17	Teenager	QUADRICEPS
8	♂	18	Teenager	LEFT DELTOID
9	♂	18	Teenager	QUADRICEPS
10	♂	19	Teenager	LEFT DELTOID
11	♂	24	Adult	LEFT DELTOID
12	♂	27	Adult	LEFT DELTOID
13	♂	35	Adult	QUADRICEPS
14	♂	60	Elder	RIGHT BICEPS
15	♂	65	Elder	LEFT DELTOID
16	♂	68	Elder	QUADRICEPS
17	♂	69	Elder	LEFT BICEPS
18	♂	71	Elder	QUADRICEPS
19	♂	71	Elder	QUADRICEPS
20	♂	72	Elder	QUADRICEPS

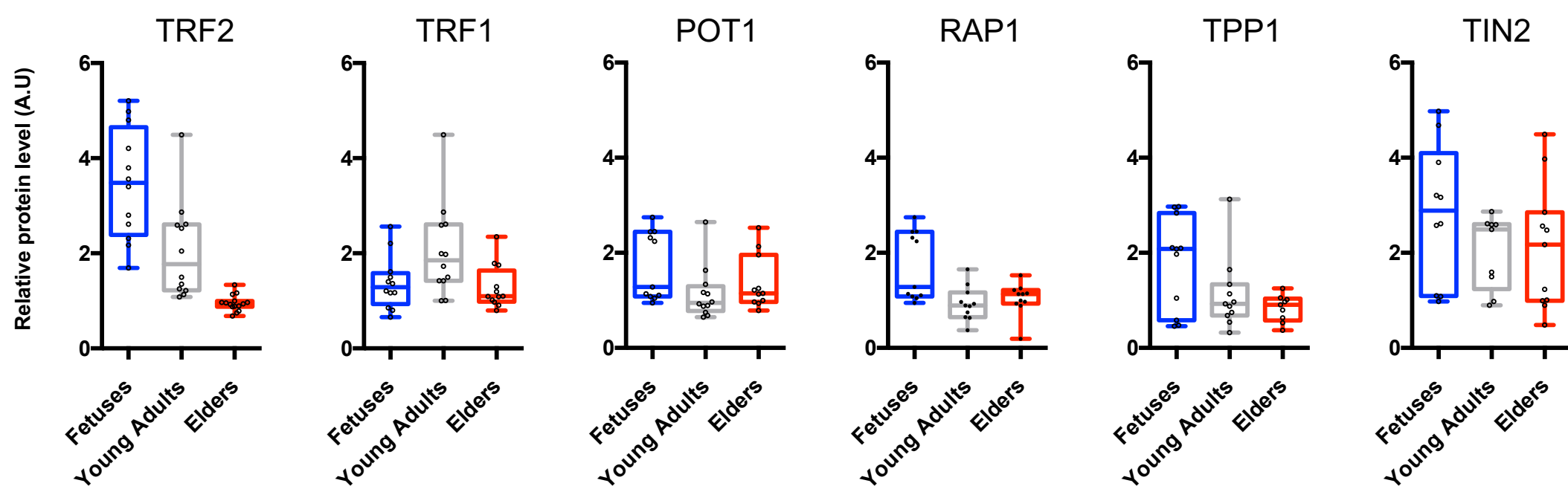
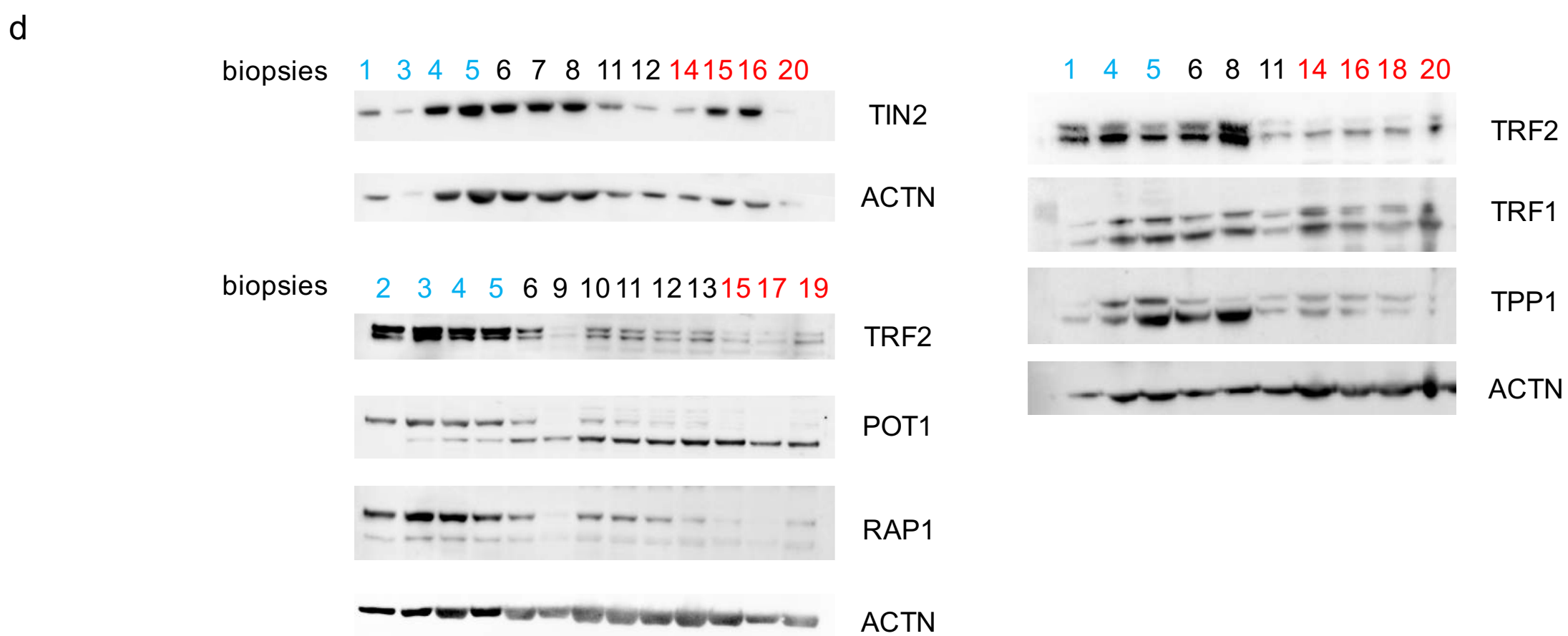
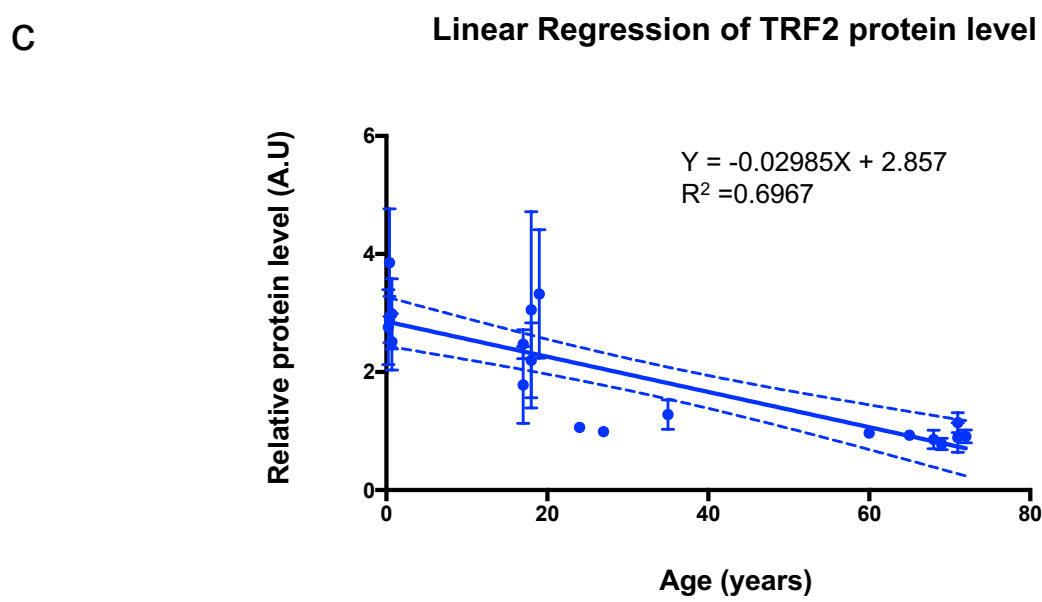
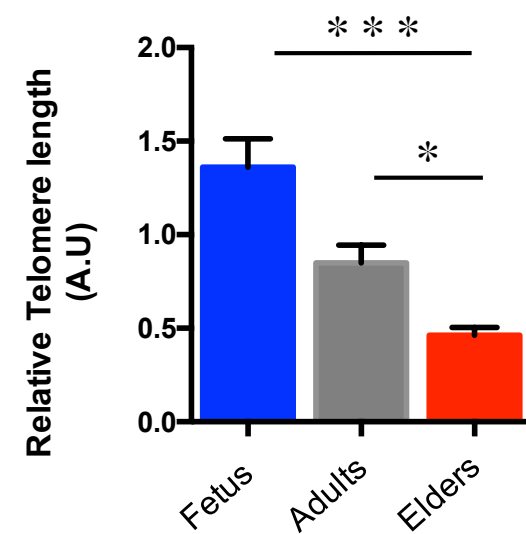
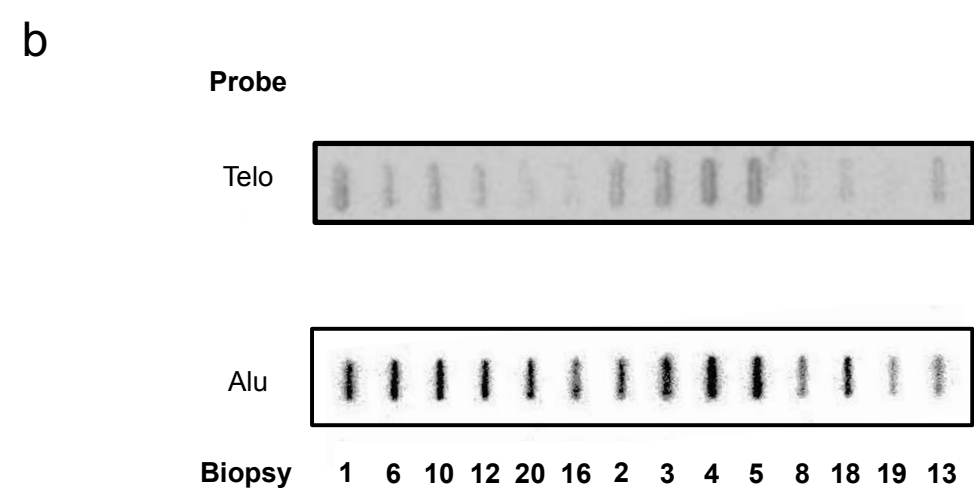
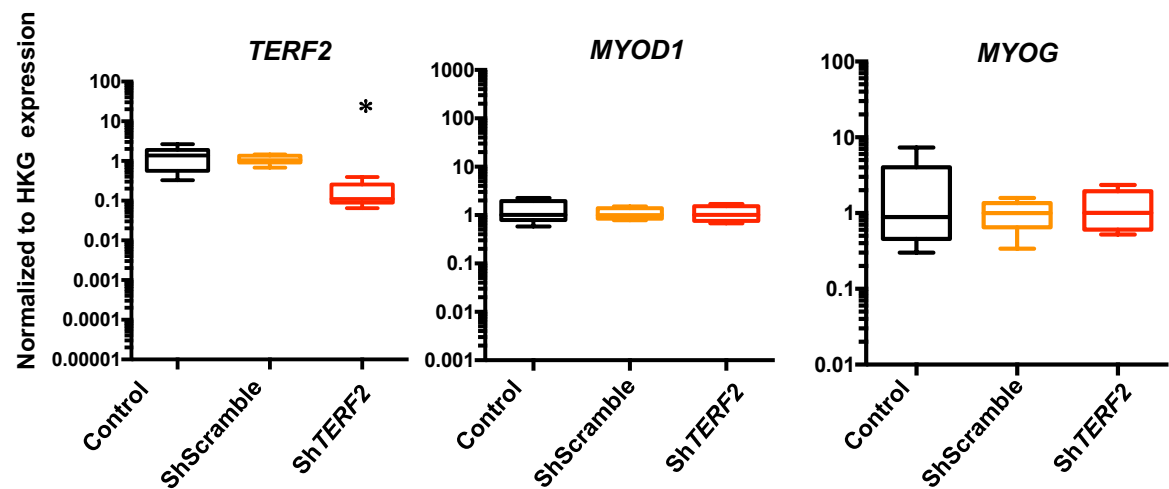


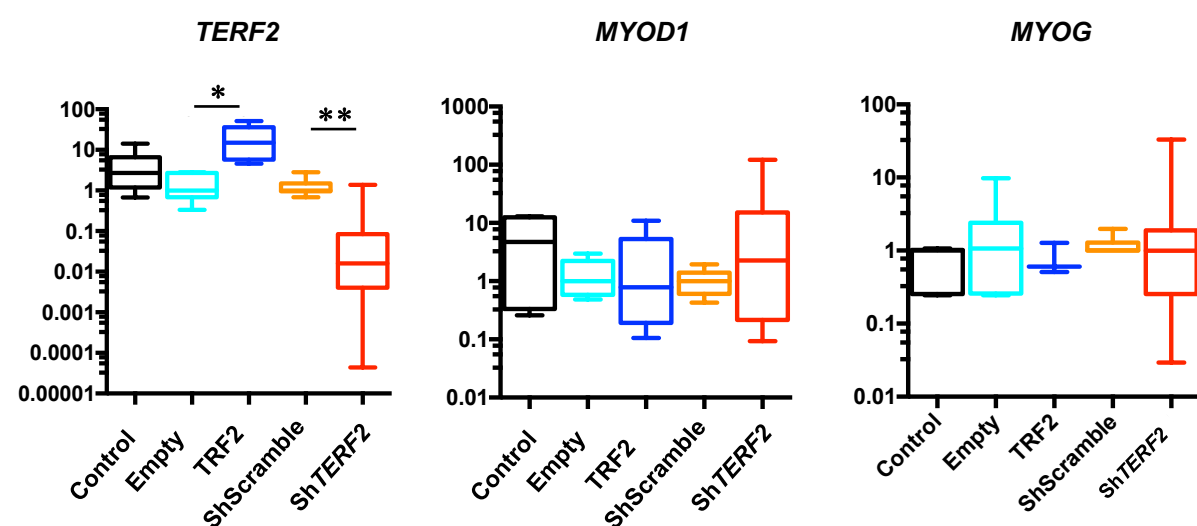
Figure S2

a

MYOBLASTS

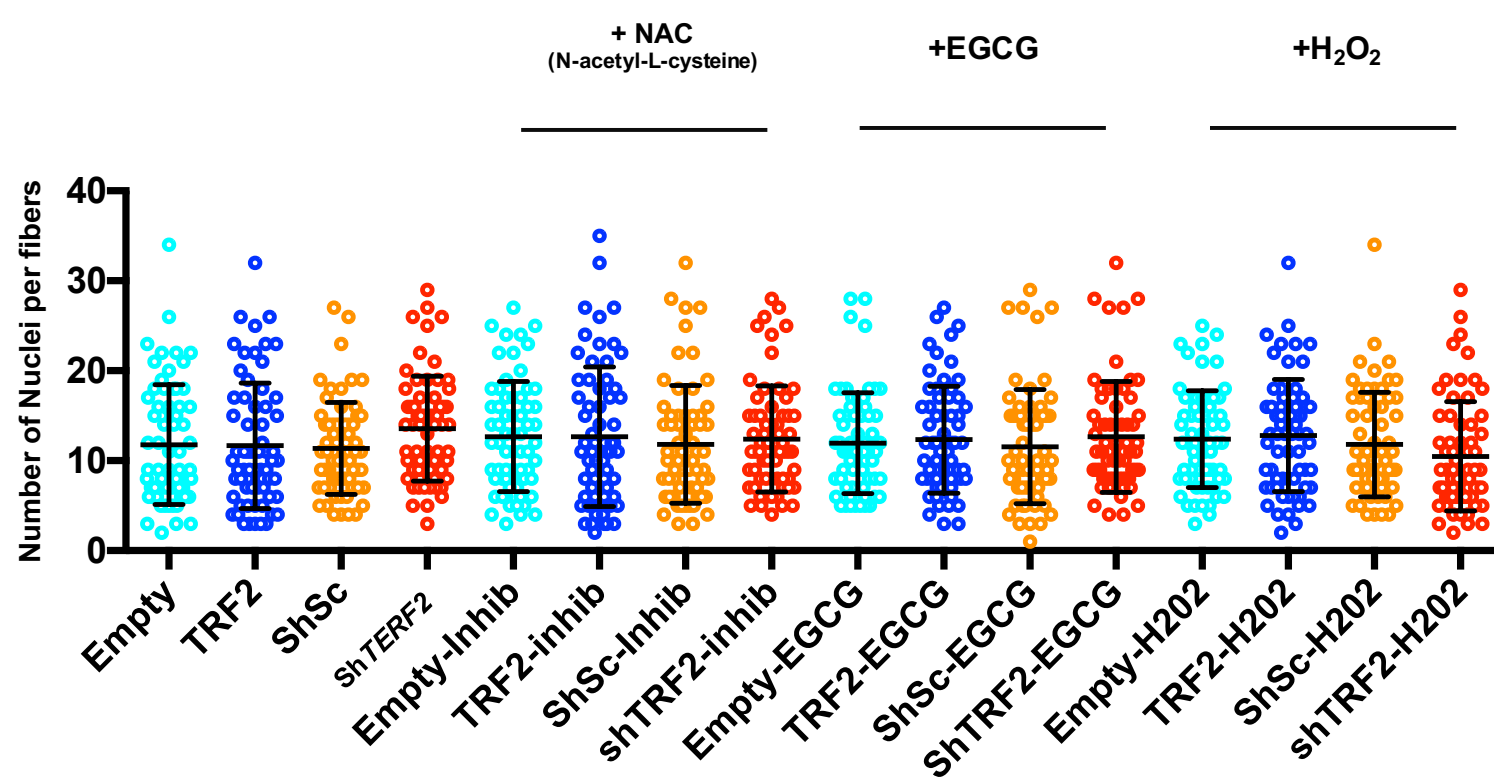


MYOTUBES



b

Fusion Index



c

MYOTUBES

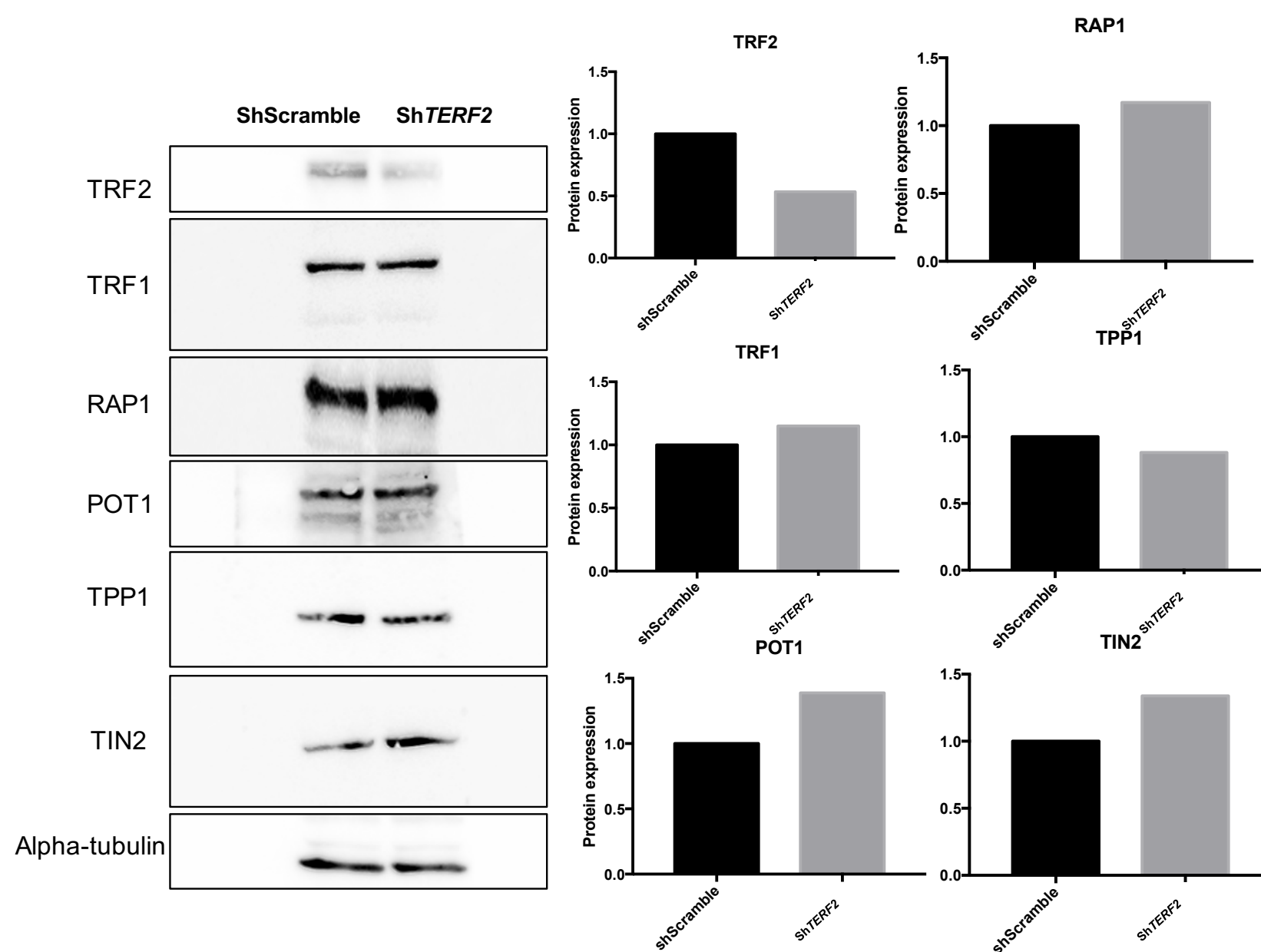
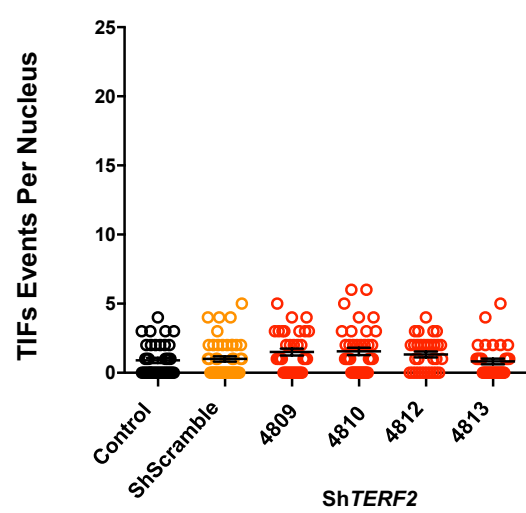
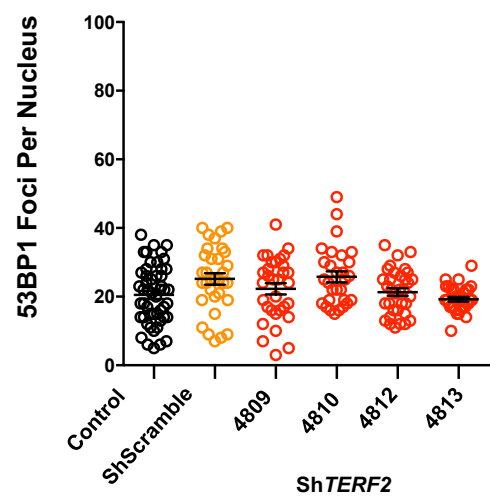
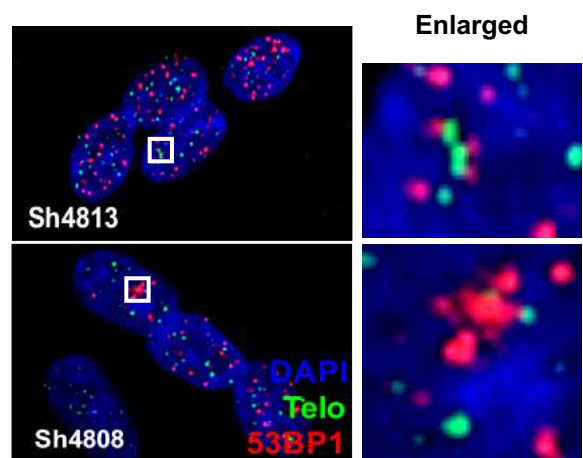
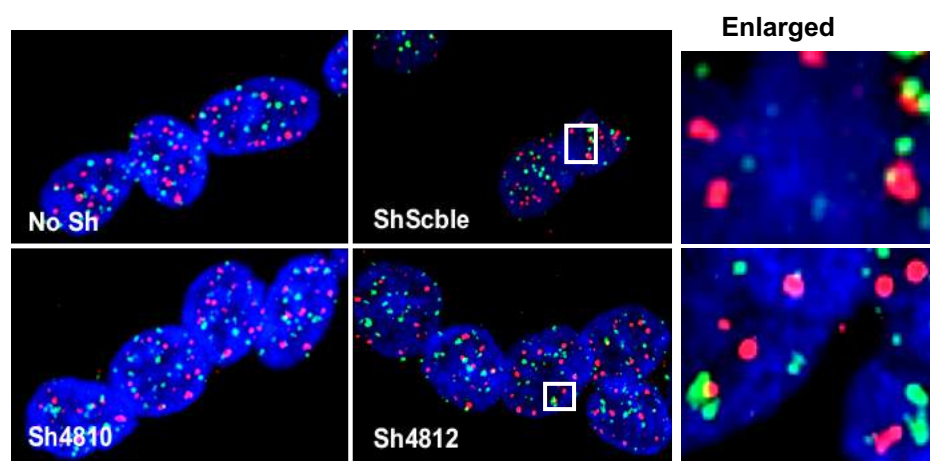
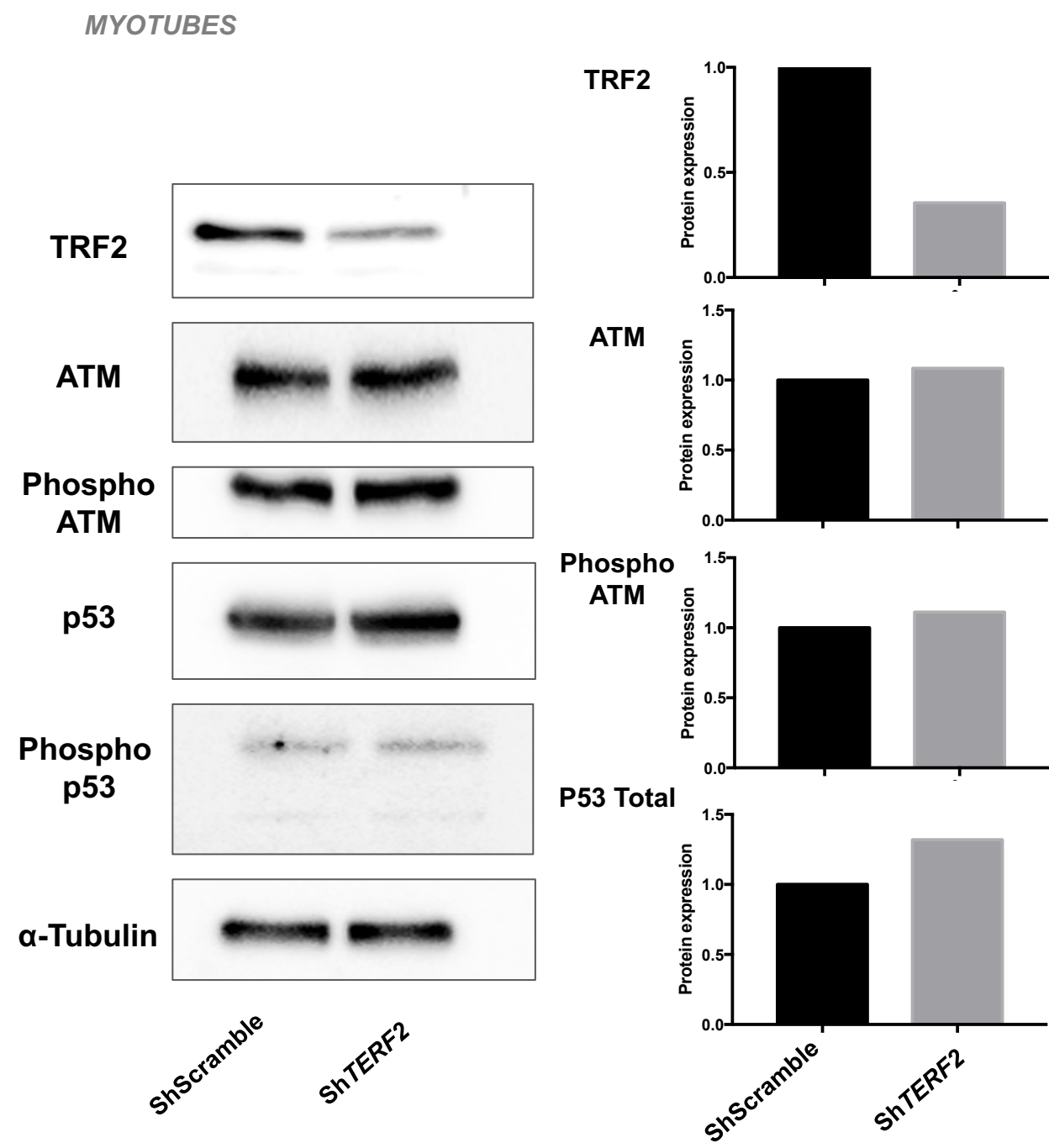


Figure S3

a



b



c

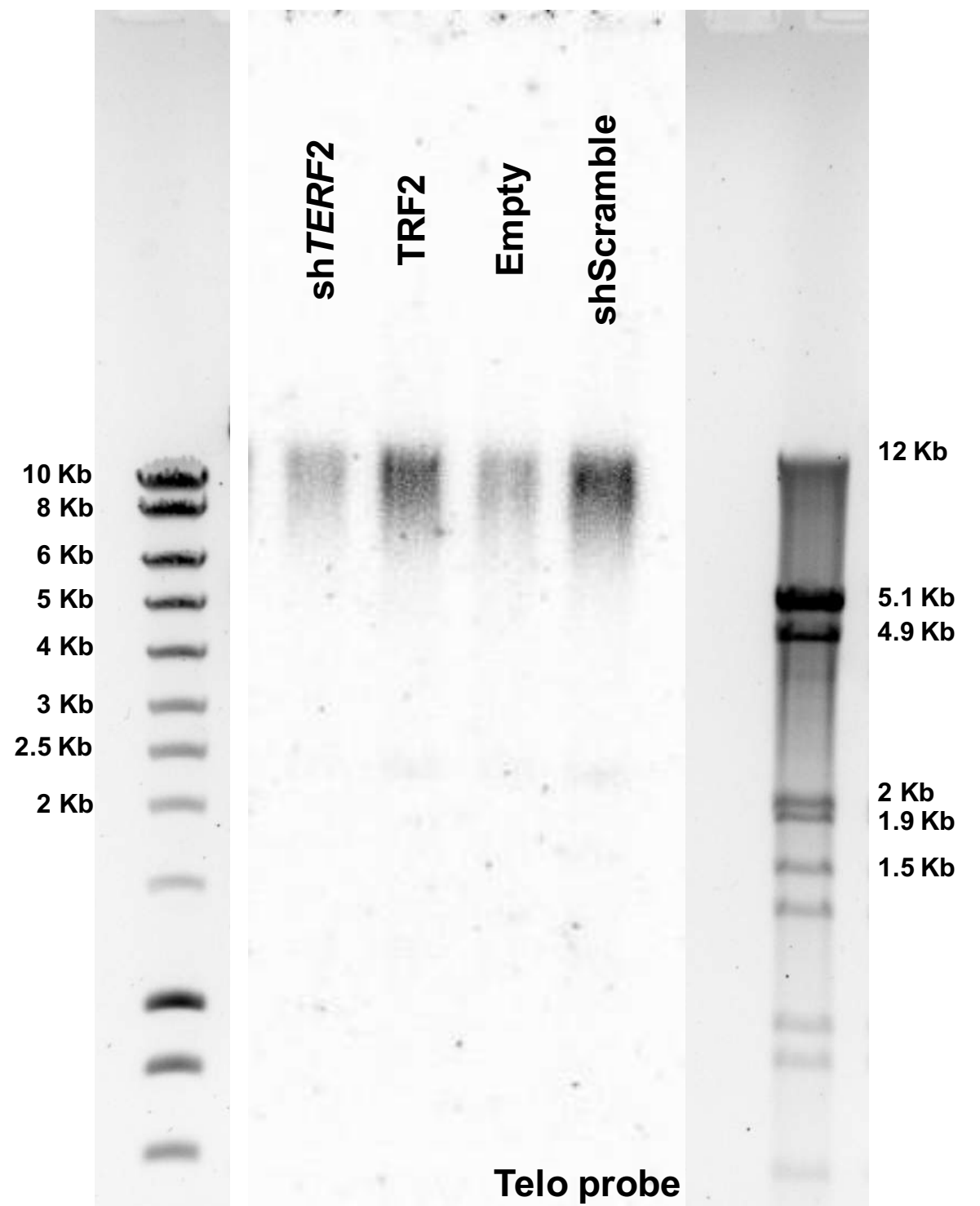




Figure S4

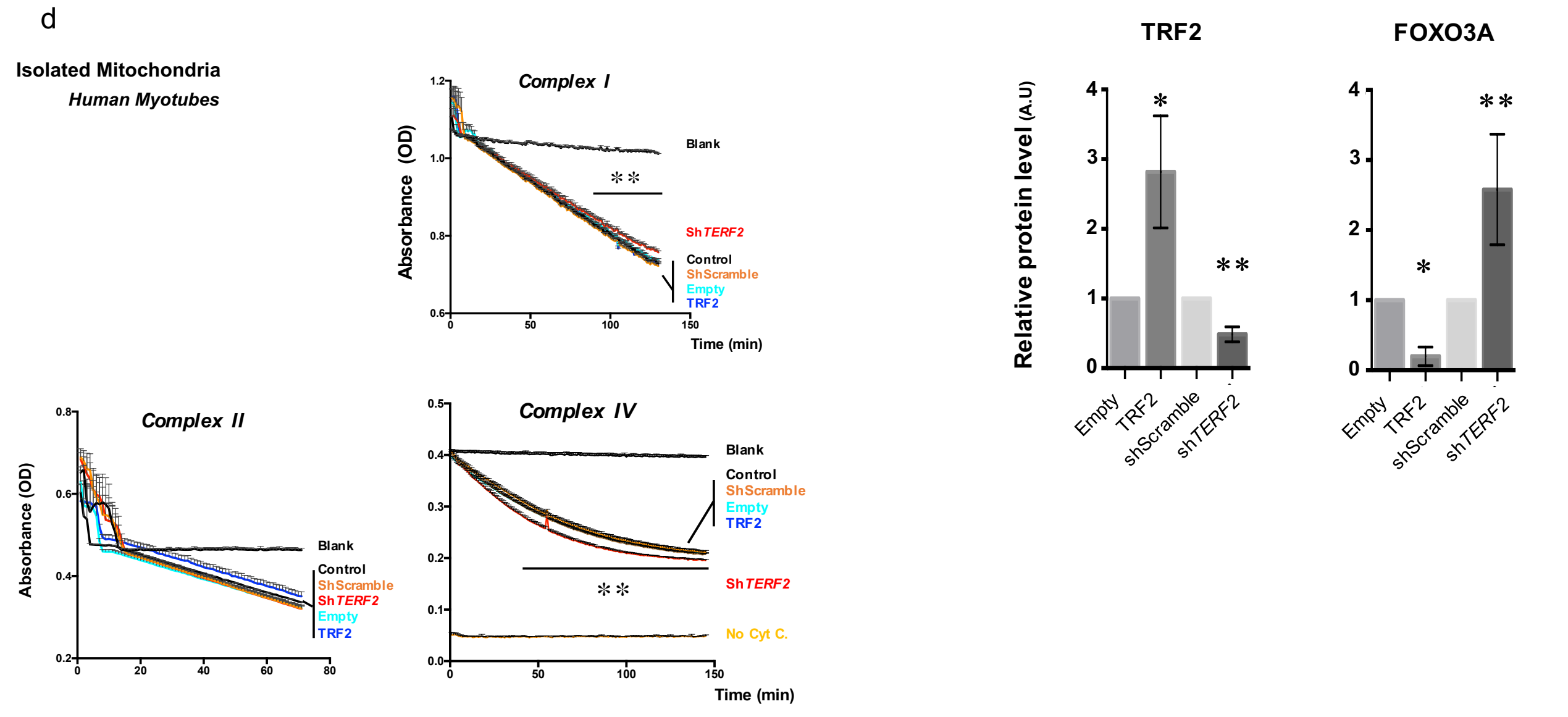
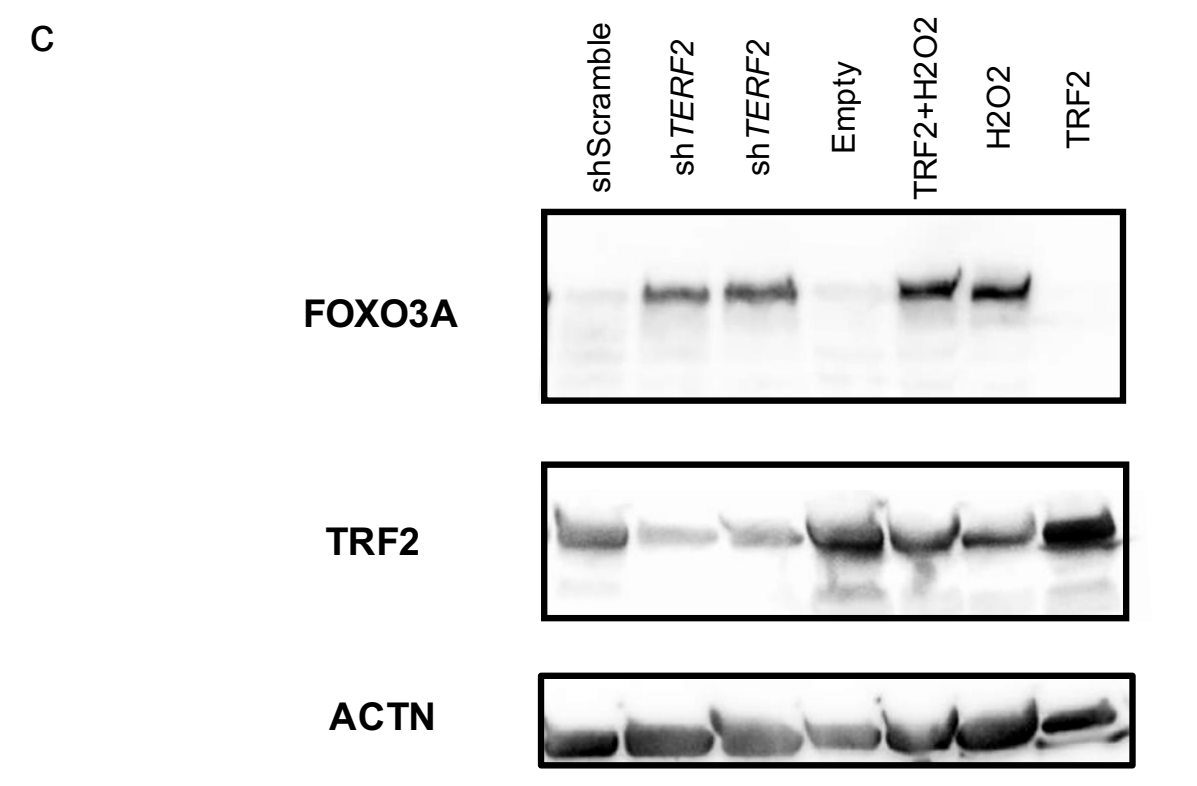
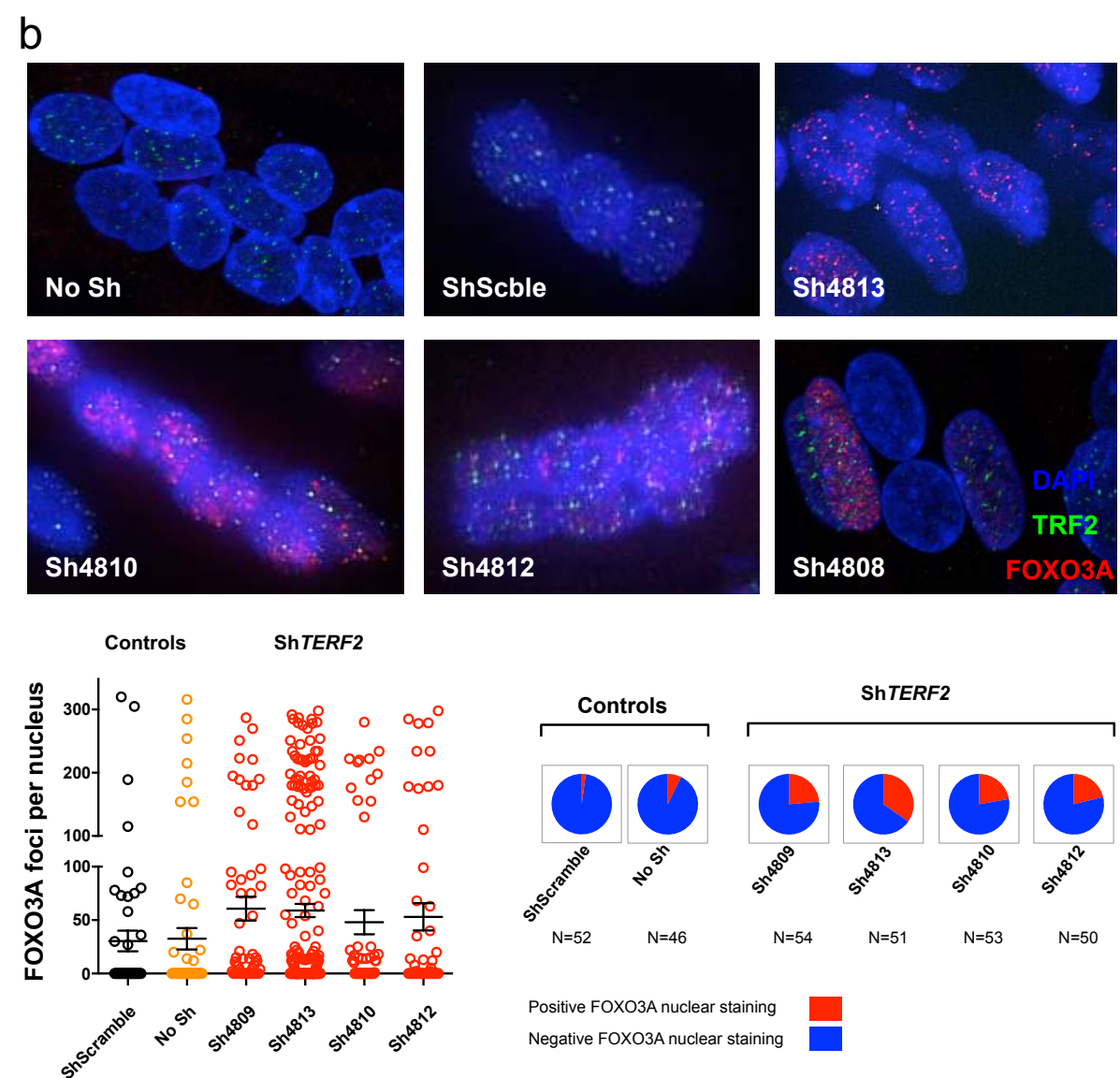
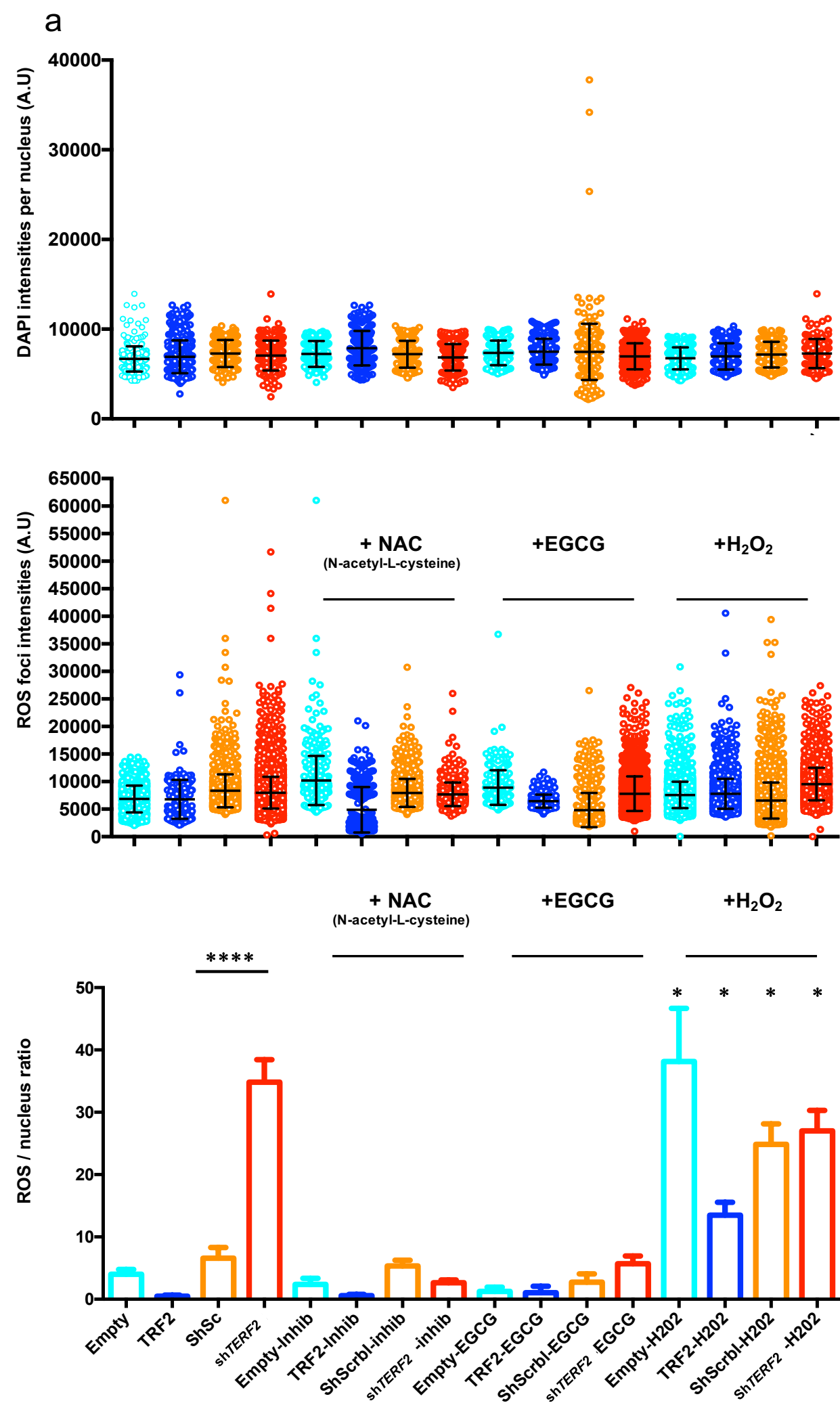
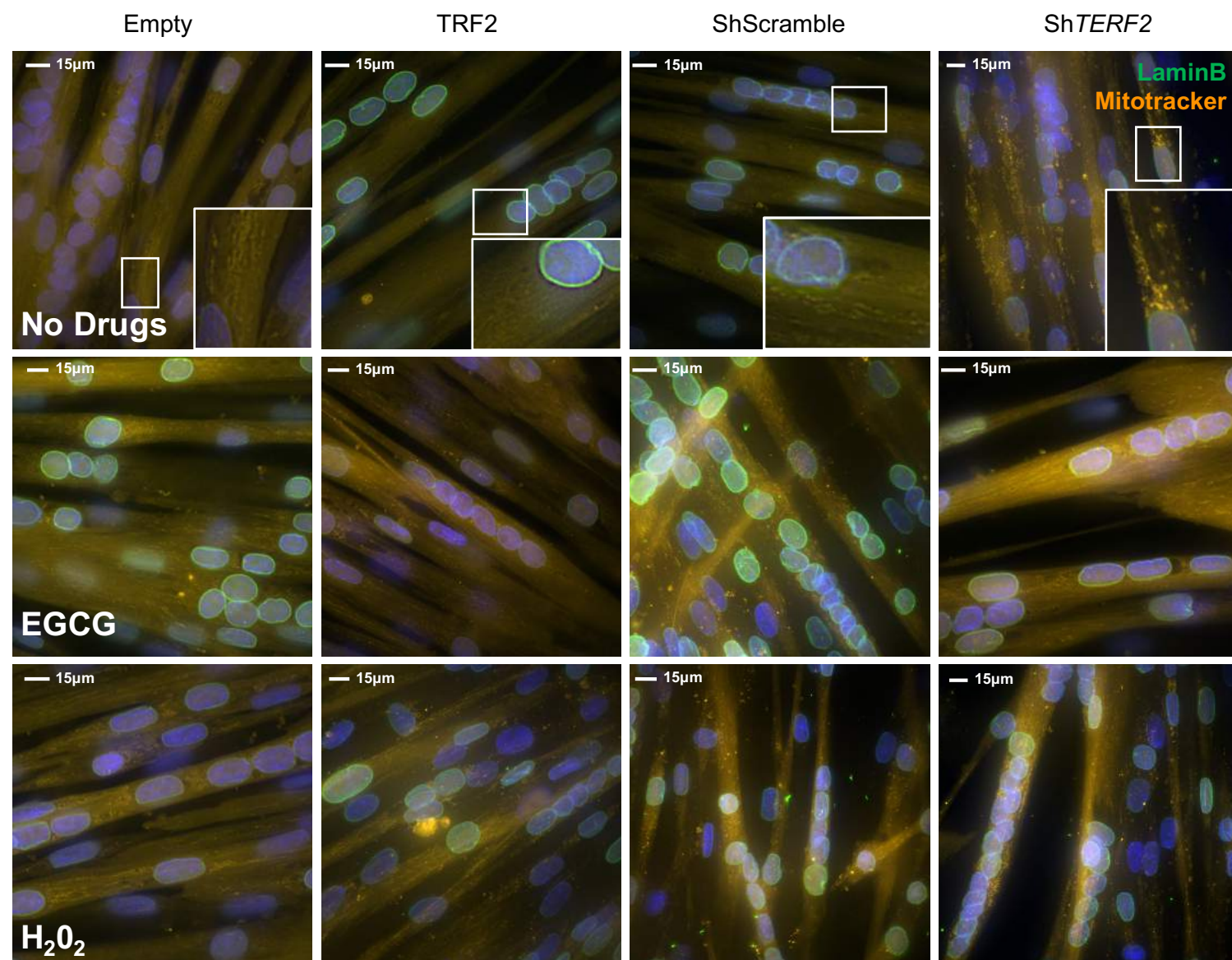




Figure S5



**NAD<sup>+</sup> / NADH enzymatic staining of transduced myotubes  
(NADH-TR)**

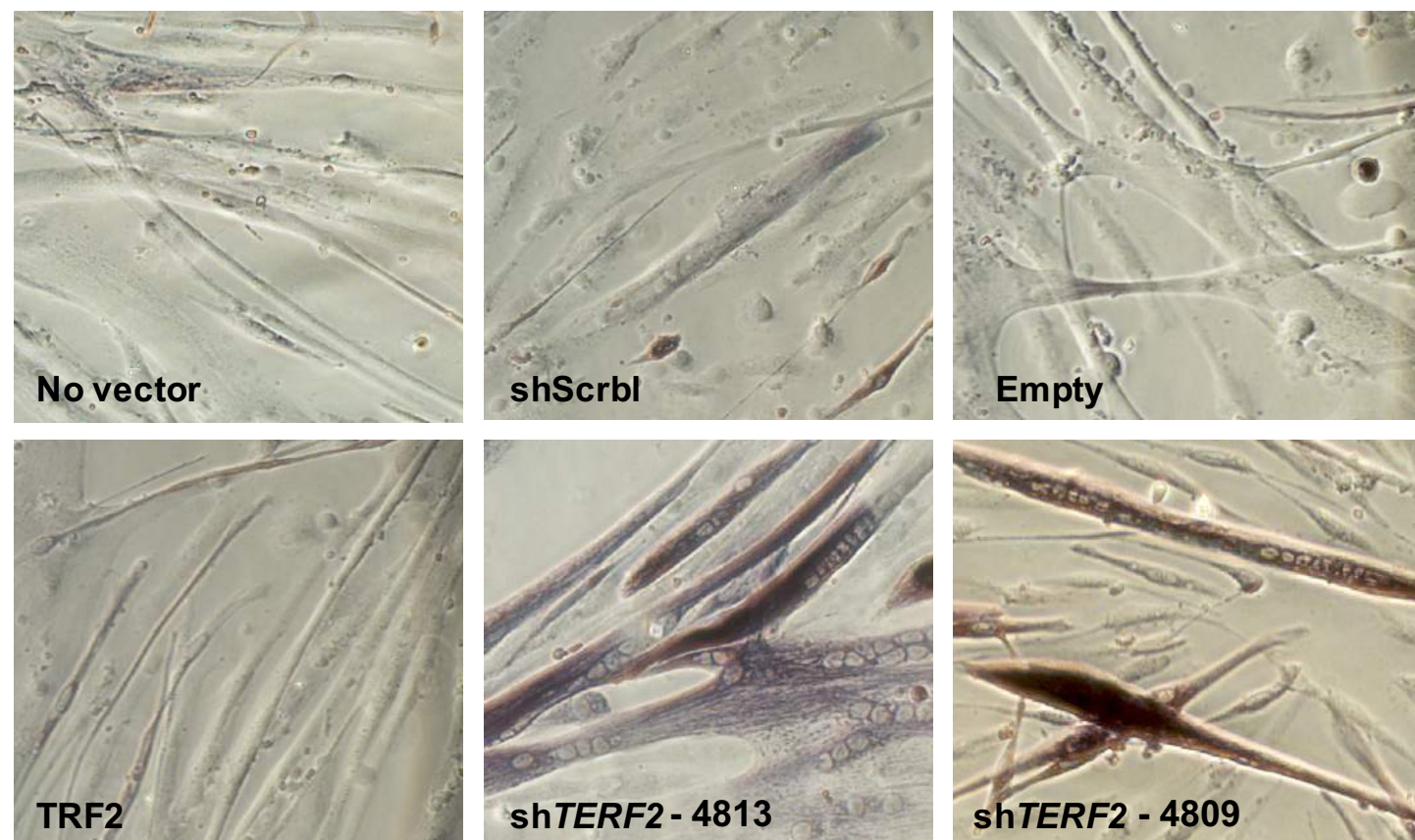
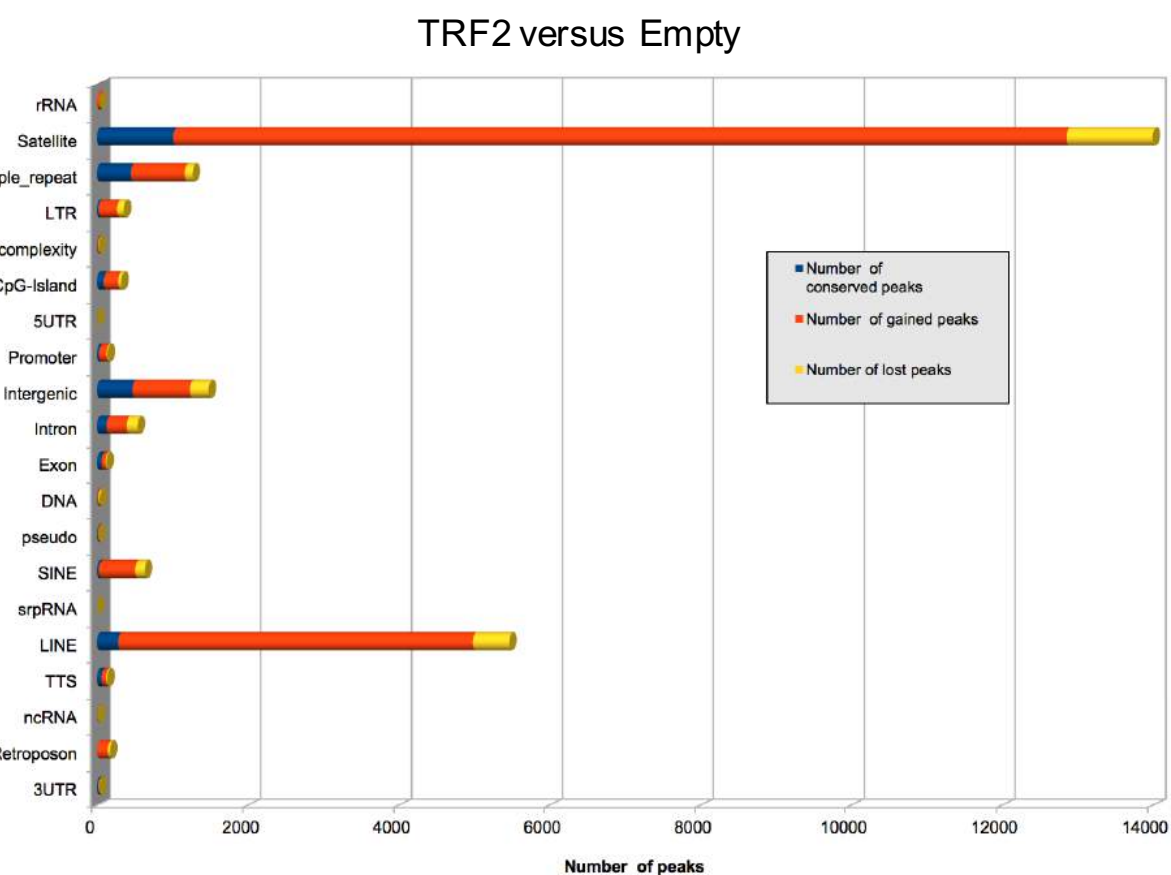
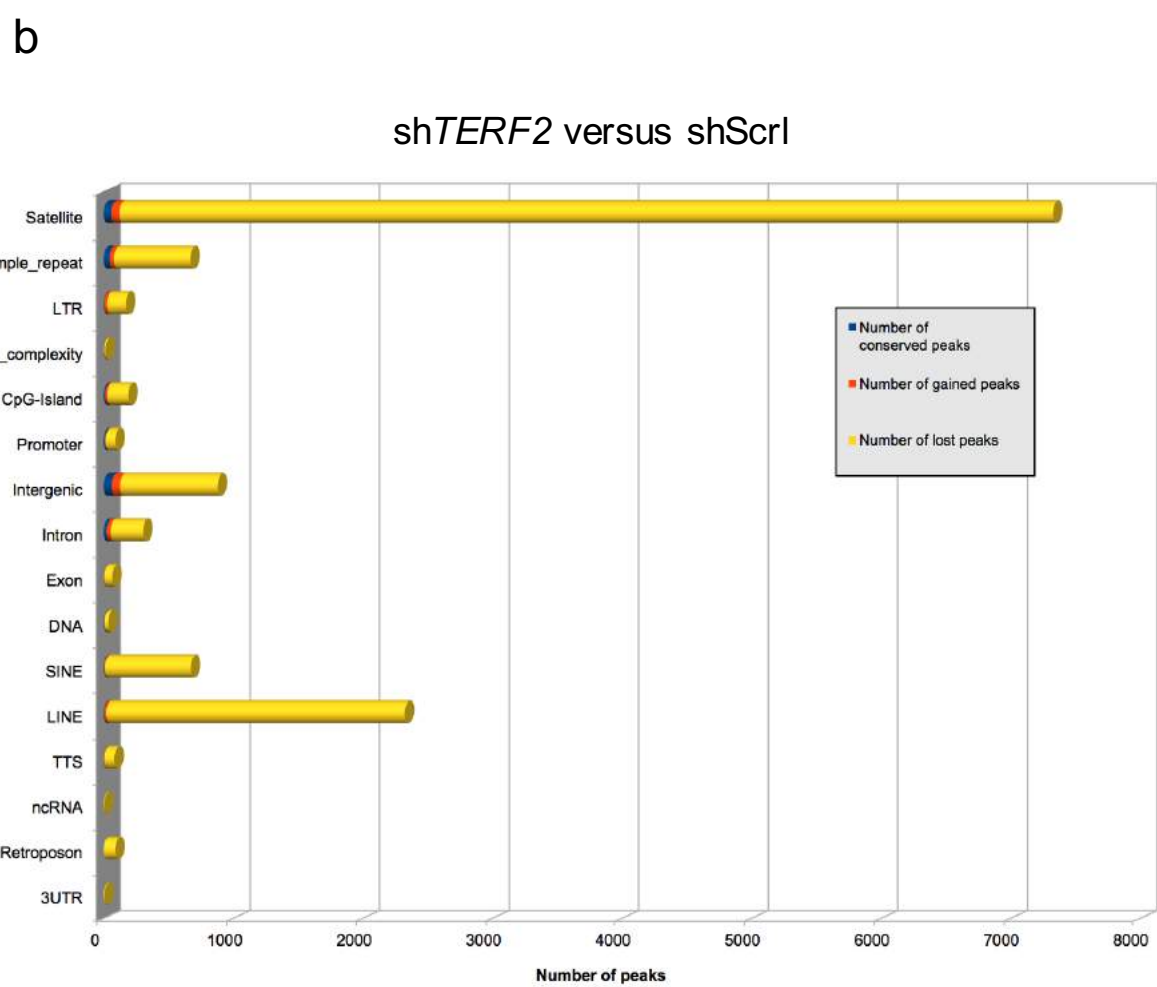
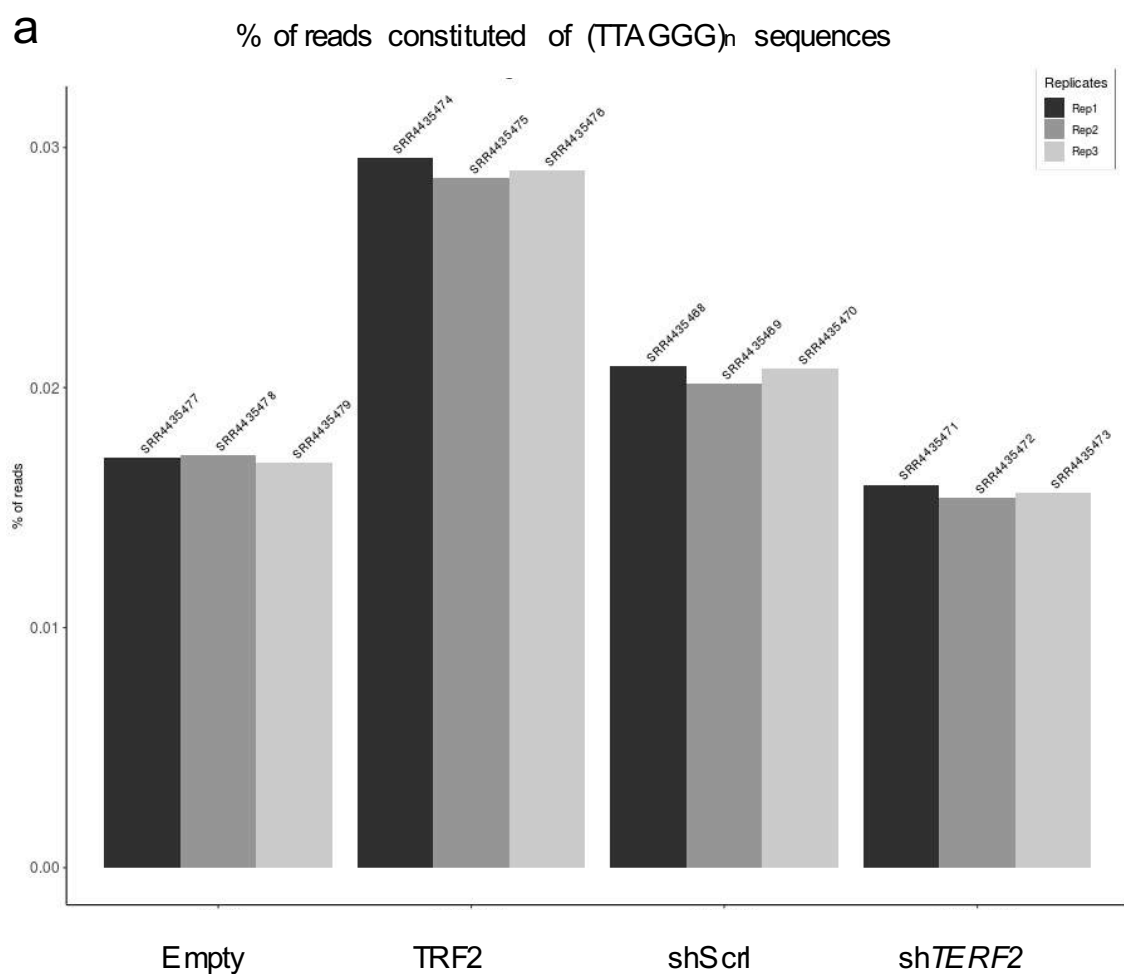




Figure S6



**C** Genes loss

Top Canonical Pathways

Name	p-value	Overlap
G-Protein Coupled Receptor Signaling	1,57E-04	13,5 % 38/281
CREB Signaling in Neurons	5,09E-04	13,8 % 30/217
GP6 Signaling Pathway	5,45E-04	15,9 % 21/132
Netrin Signaling	6,98E-04	20,0 % 13/65
cAMP-mediated signaling	8,63E-04	13,4 % 30/224

	GO term	Description	P-value
Biological process	GO:0007186	G protein-coupled receptor signaling pathway	5.69E-9
	GO:0007187	G protein-coupled recep sign path, coupled to cyclic nt second messenger	2.23E-6
	GO:0046777	protein autophosphorylation	5.92E-5
	GO:0007165	signal transduction	8.98E-5
	GO:0007193	adenylate cyclase-inhibiting G protein-coupled recep signaling pathway	2.14E-4
	GO:0007188	adenylate cyclase-modulating G protein-coupled recep signaling pathway	2.32E-4
	GO:0065007	biological regulation	2.91E-4
	GO:0065008	regulation of biological quality	5.24E-4
	GO:0006796	phosphate-containing compound metabolic process	7.14E-4
	GO:0006793	phosphorus metabolic process	7.14E-4
Molecular function	GO:1901564	organonitrogen compound metabolic process	9.26E-4
	GO:0044248	cellular catabolic process	9.84E-4
	GO:0004930	G protein-coupled receptor activity	1.43E-6
Cellular component	GO:0099528	G protein-coupled neurotransmitter receptor activity	5.04E-5
	GO:0003824	catalytic activity	1.13E-4
	GO:0044425	membrane part	1.66E-4
	GO:0005887	integral component of plasma membrane	6.18E-4

Genes gain

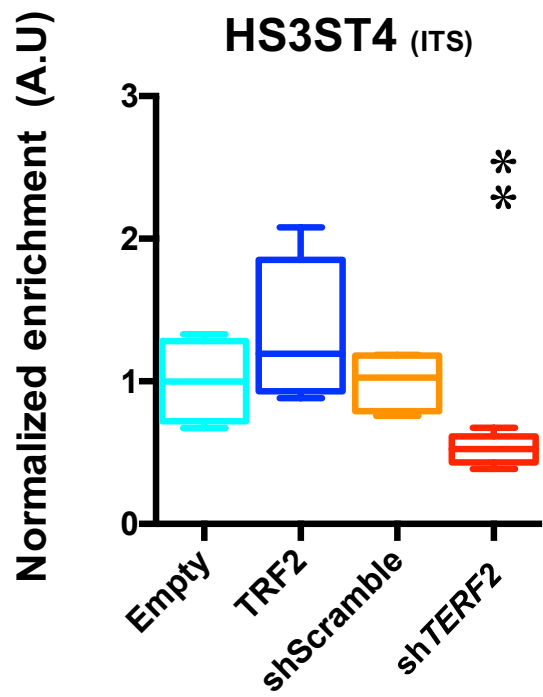
Top Canonical Pathways

Name	p-value	Overlap
Leukocyte Extravasation Signaling	3,64E-03	11,1 % 23/208
Netrin Signaling	5,27E-03	15,4 % 10/65
Choline Biosynthesis III	6,04E-03	30,8 % 4/13
Protein Kinase A Signaling	9,42E-03	9,1 % 35/383
Iron homeostasis signaling pathway	1,24E-02	11,5 % 15/131

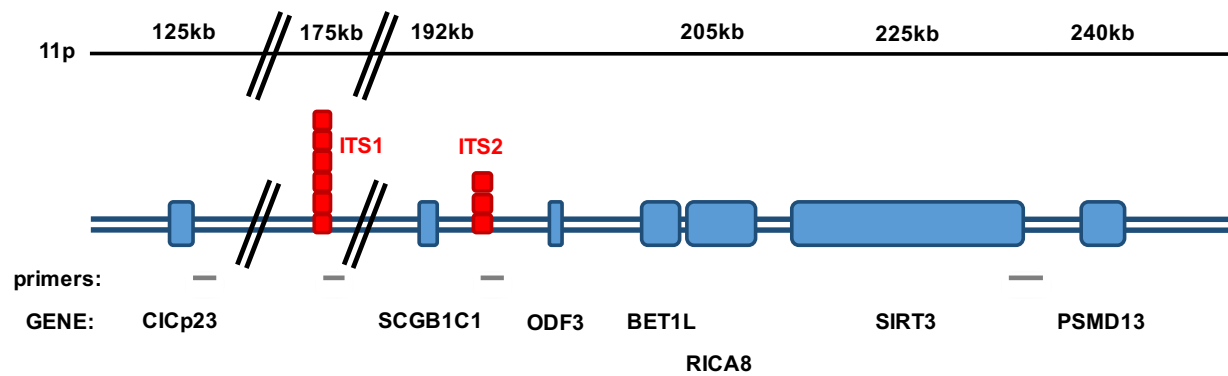
	GO term	Description	P-value
Biological process	GO:0022610	biological adhesion	4.41E-5
	GO:0007155	cell adhesion	4.41E-5
	GO:0098609	cell-cell adhesion	9.05E-5
	GO:0032879	regulation of localization	1.16E-4
	GO:0030198	extracellular matrix organization	1.41E-4
	GO:0043062	extracellular structure organization	1.41E-4
	GO:0071840	cellular component organization or biogenesis	1.43E-4
	GO:0016043	cellular component organization	1.43E-4
	GO:0033554	cellular response to stress	4.42E-4
	GO:0040012	regulation of locomotion	6.51E-4
	GO:2000145	regulation of cell motility	6.51E-4
	GO:0051894	positive regulation of focal adhesion assembly	7.02E-4
	GO:0010811	positive regulation of cell-substrate adhesion	7.02E-4
	GO:0001954	positive regulation of cell-matrix adhesion	7.02E-4
	GO:1901890	positive regulation of cell junction assembly	7.02E-4
	GO:1903393	positive regulation of adherens junction organization	7.02E-4
	GO:0002376	immune system process	7.67E-4
	GO:0034613	cellular protein localization	7.85E-4
	GO:0070727	cellular macromolecule localization	7.85E-4
	GO:0051270	regulation of cellular component movement	7.85E-4
Molecular function	GO:0048523	negative regulation of cellular process	8.88E-4
	GO:0050789	regulation of biological process	9.42E-4
Cellular component	GO:0031267	small GTPase binding	3.69E-4
	GO:0017016	Ras GTPase binding	3.69E-4
	GO:0051020	GTPase binding	3.69E-4
	GO:0017048	Rho GTPase binding	3.69E-4
	GO:0030054	cell junction	1.94E-5
	GO:0005911	cell-cell junction	9.19E-5
	GO:0043229	intracellular organelle	8.44E-4
	GO:0070161	anchoring junction	8.78E-4
	GO:0005886	plasma membrane	9.49E-4

Figure S7

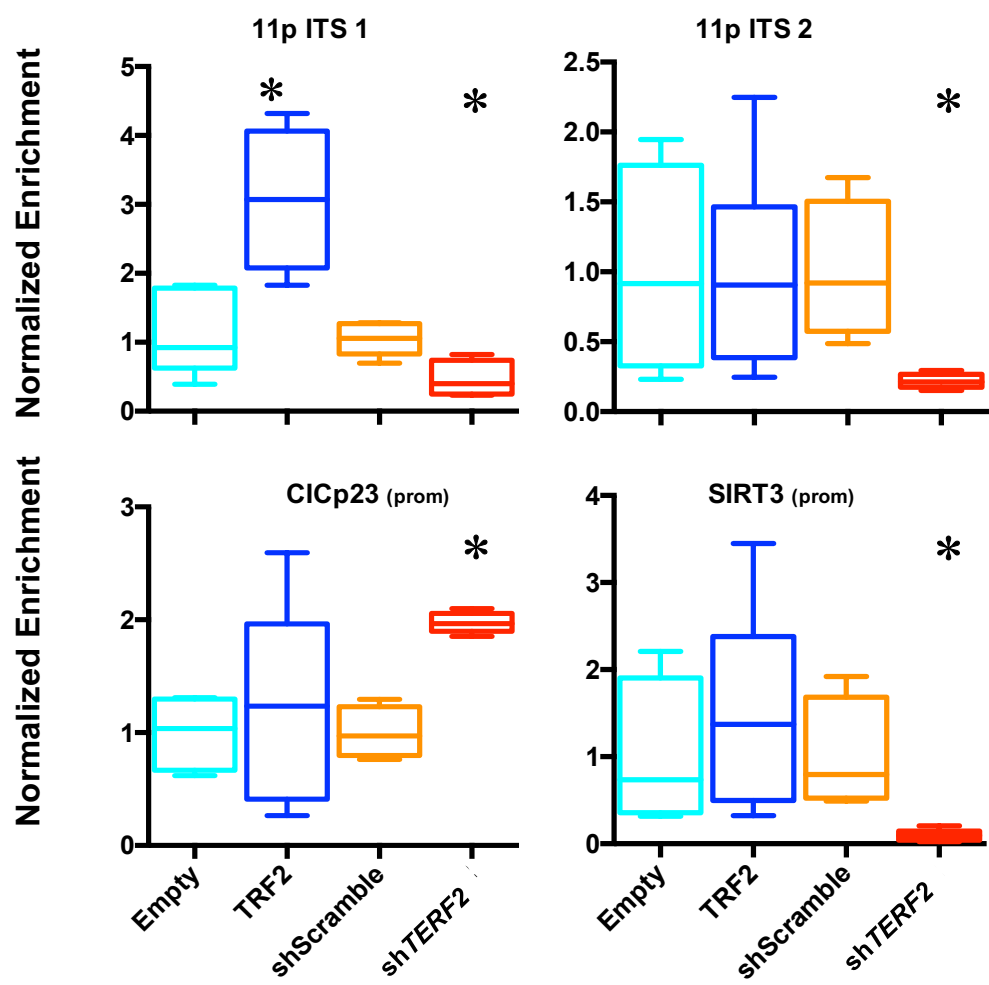
a



b

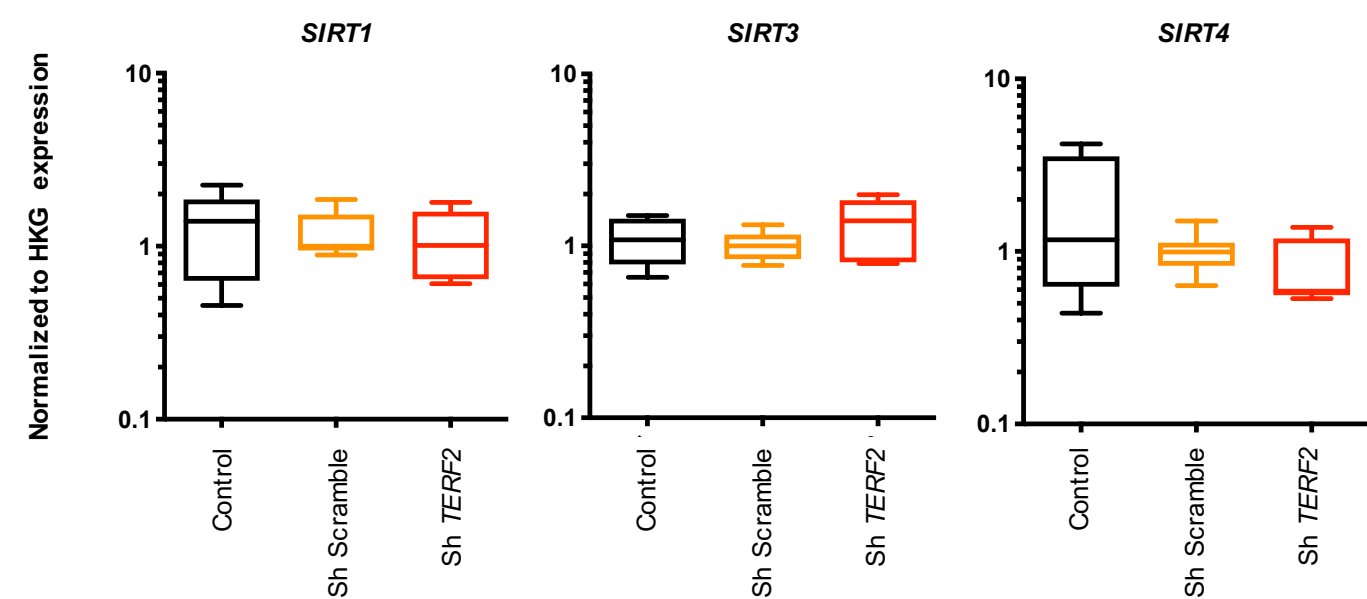


c



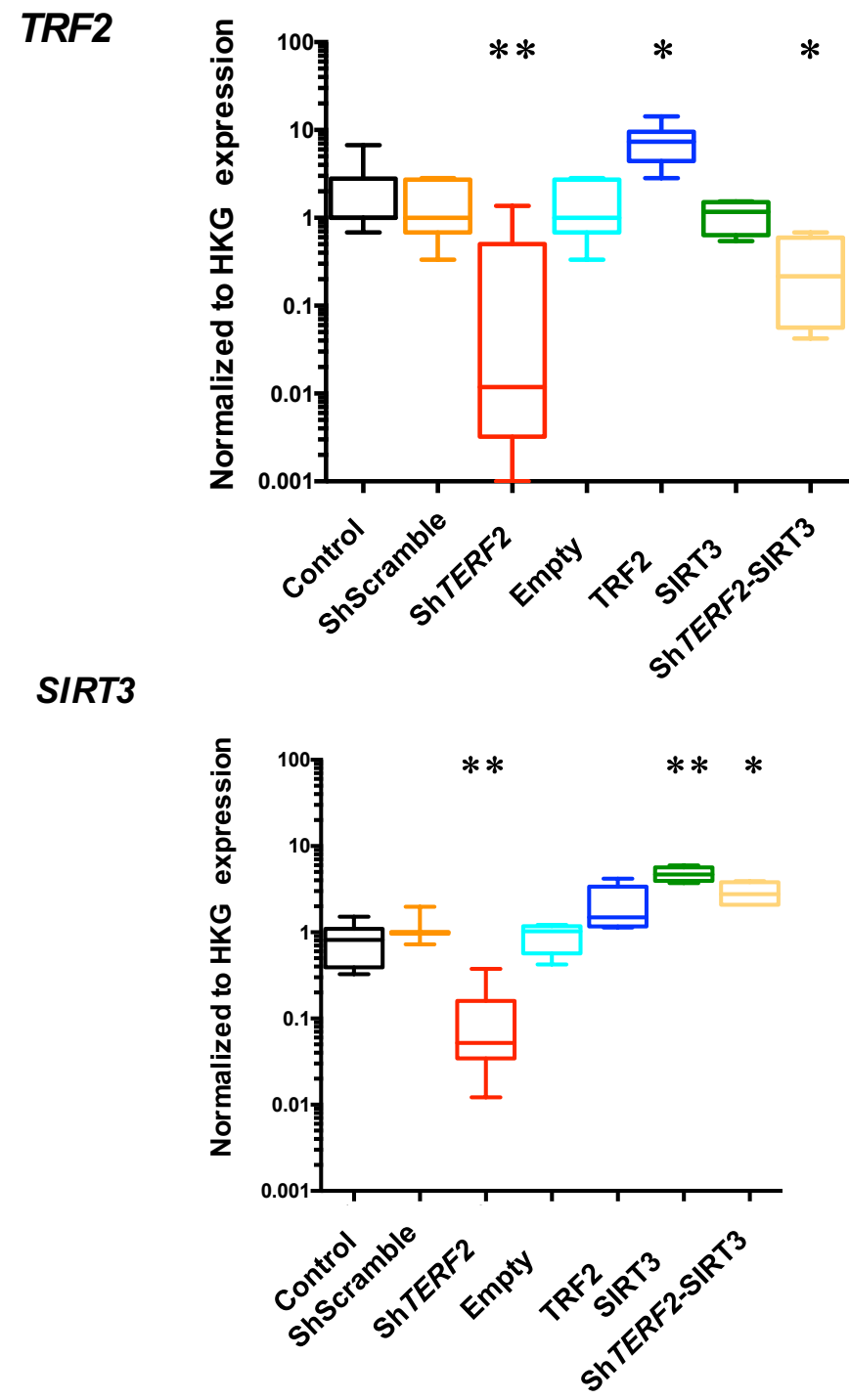
d

**MYOBLASTS**



e

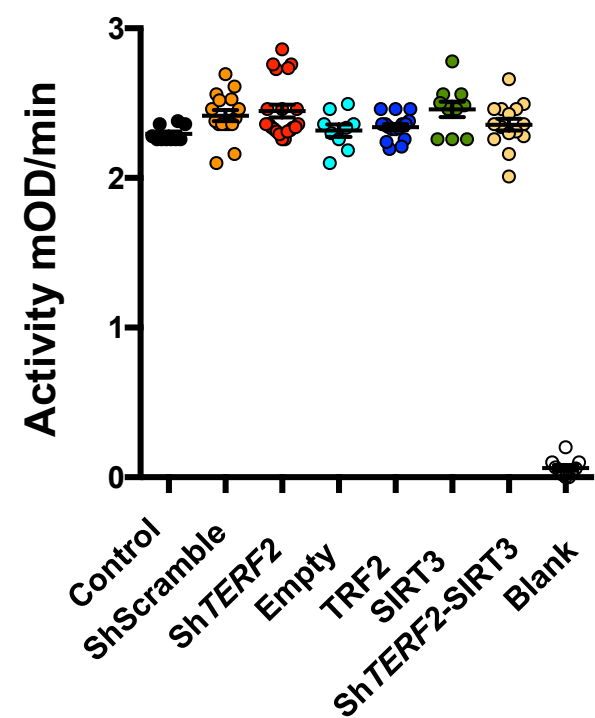
**MYOTUBES**



f

**Isolated Mitochondria  
Human Myotubes**

**Complex II**



**Complex IV**

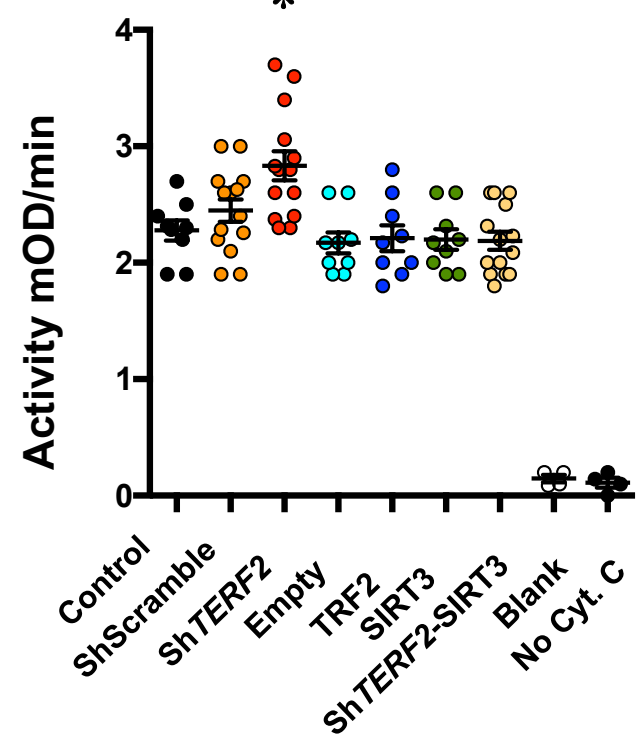
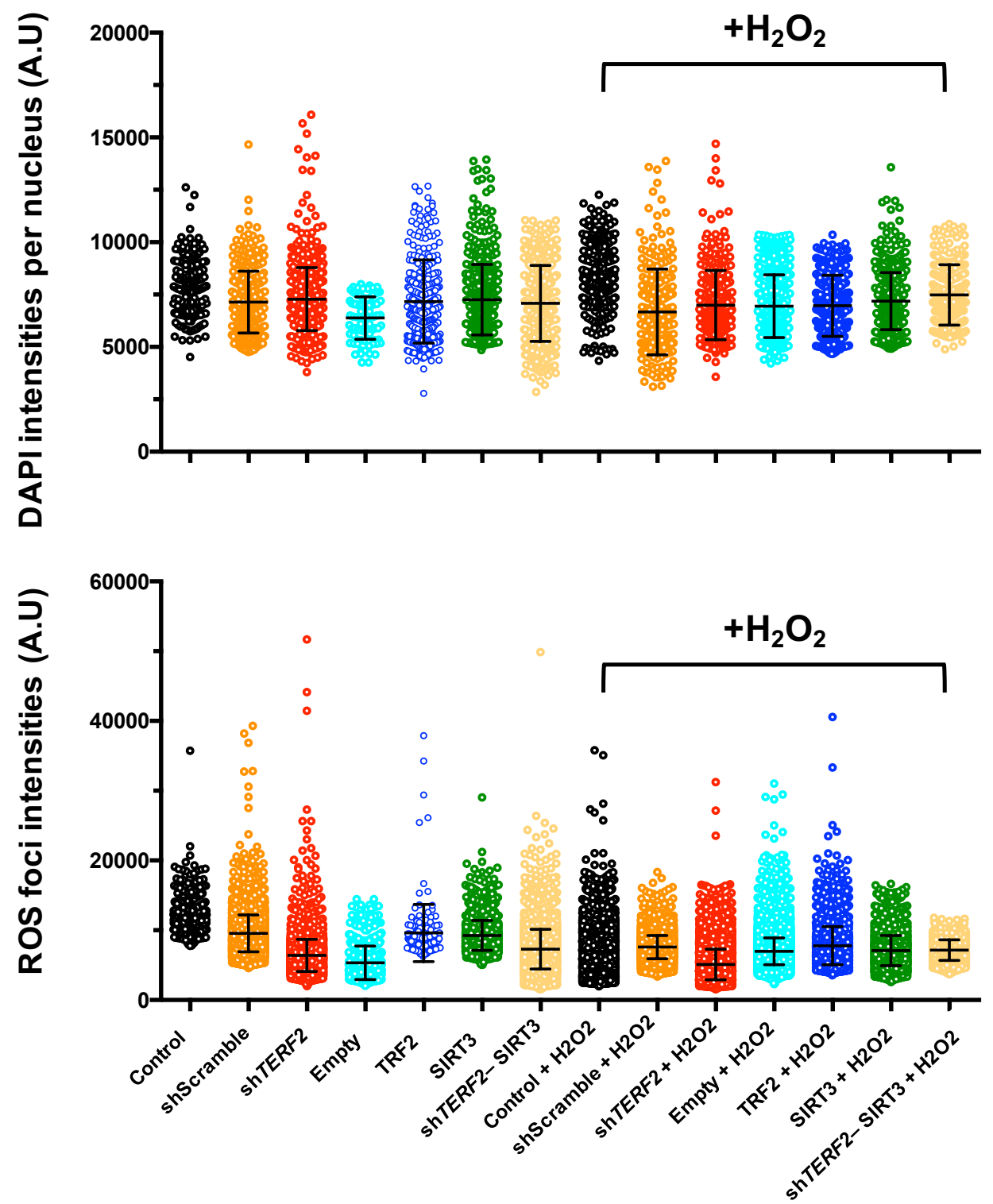
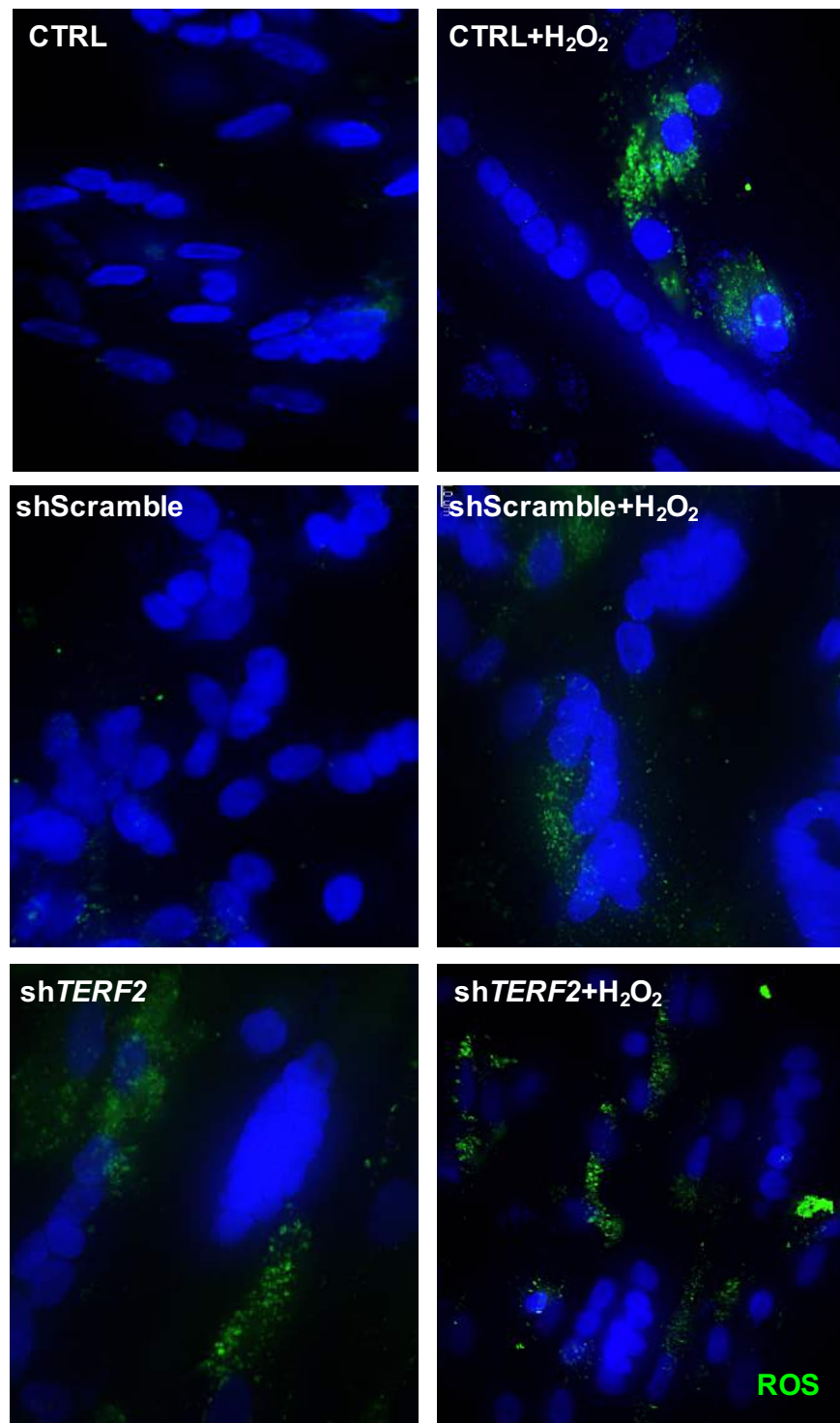


Figure S8

a



b

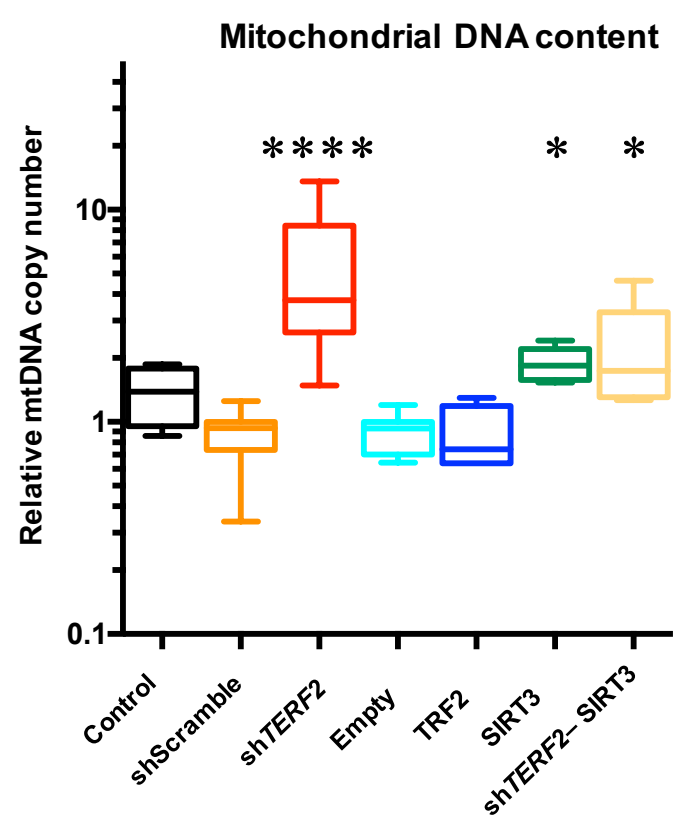
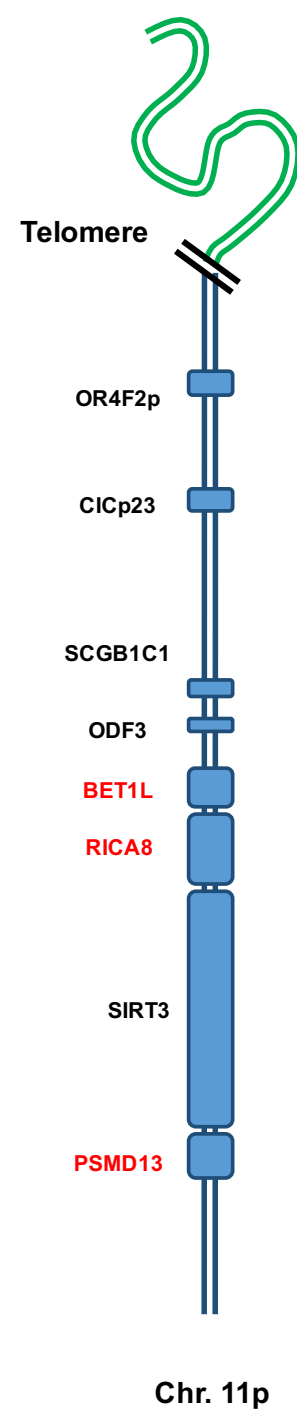
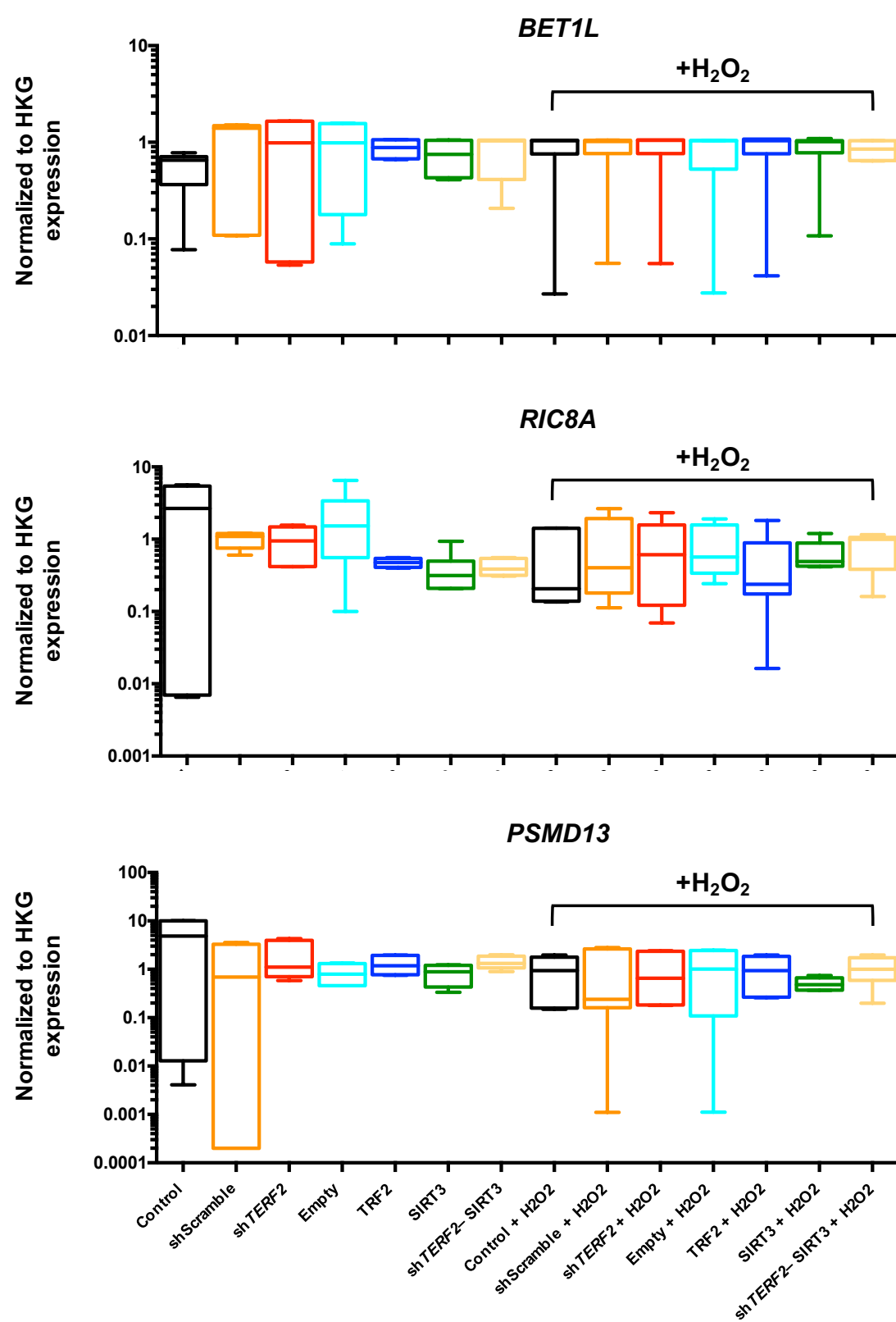


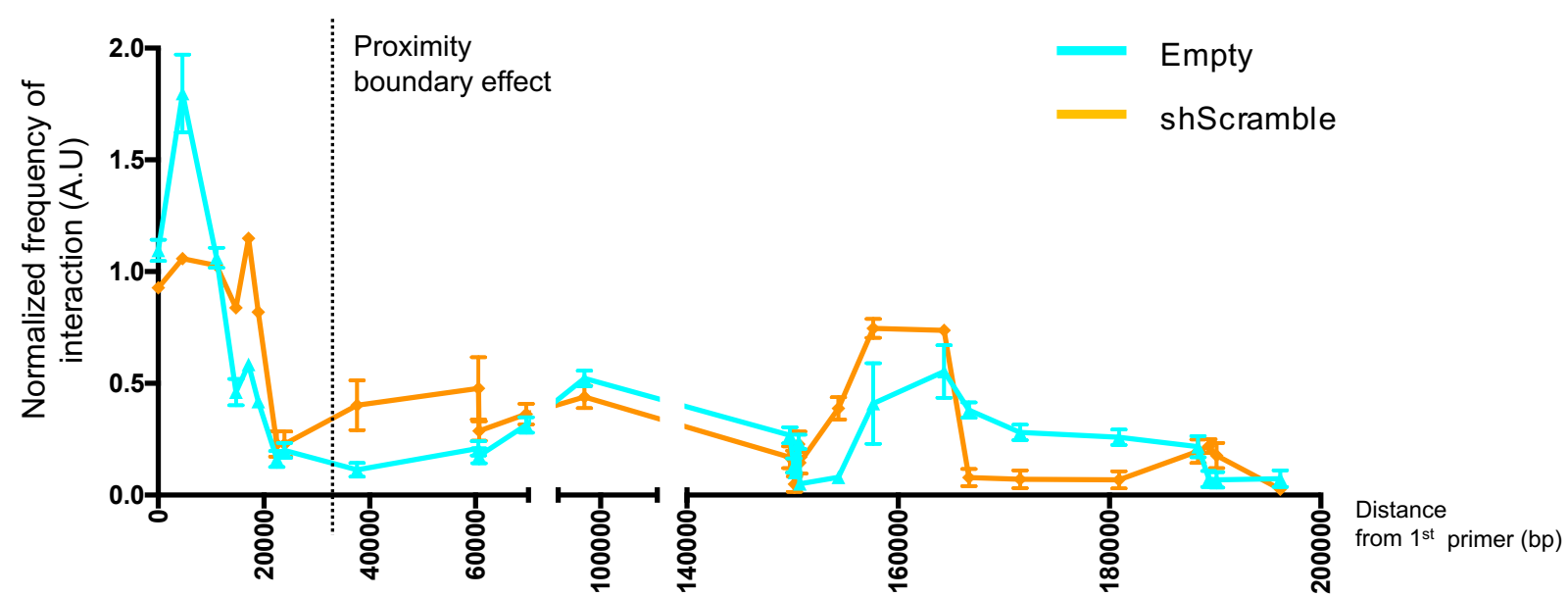


Figure S9

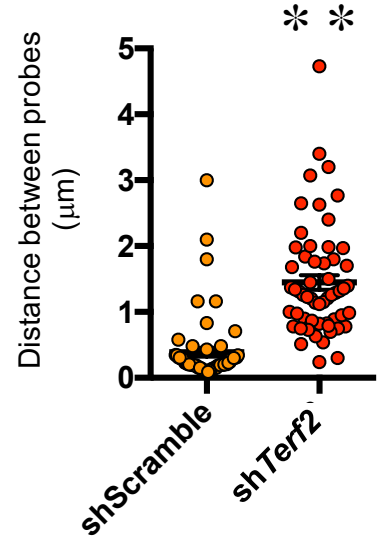
a



b



c



c *Mouse Embryonic Fibroblasts* (Mitotically arrested)

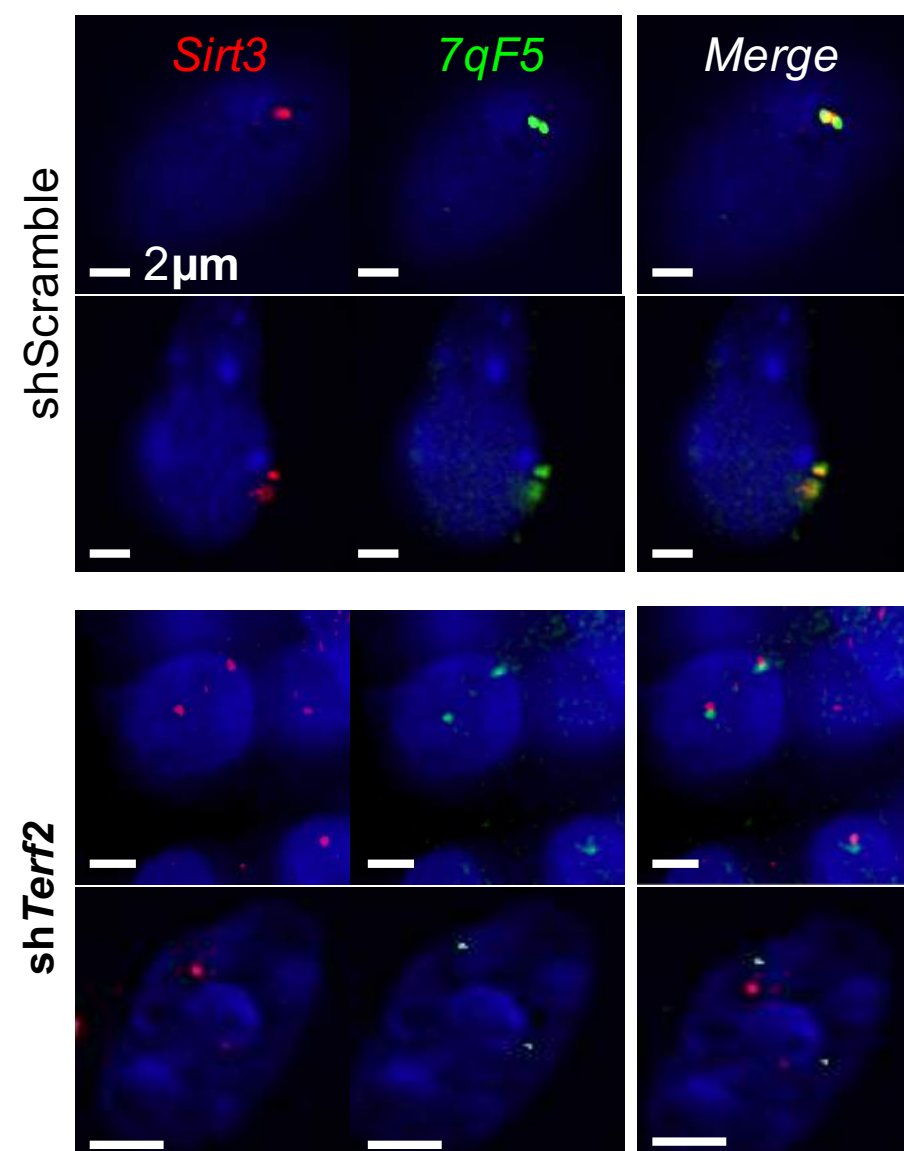
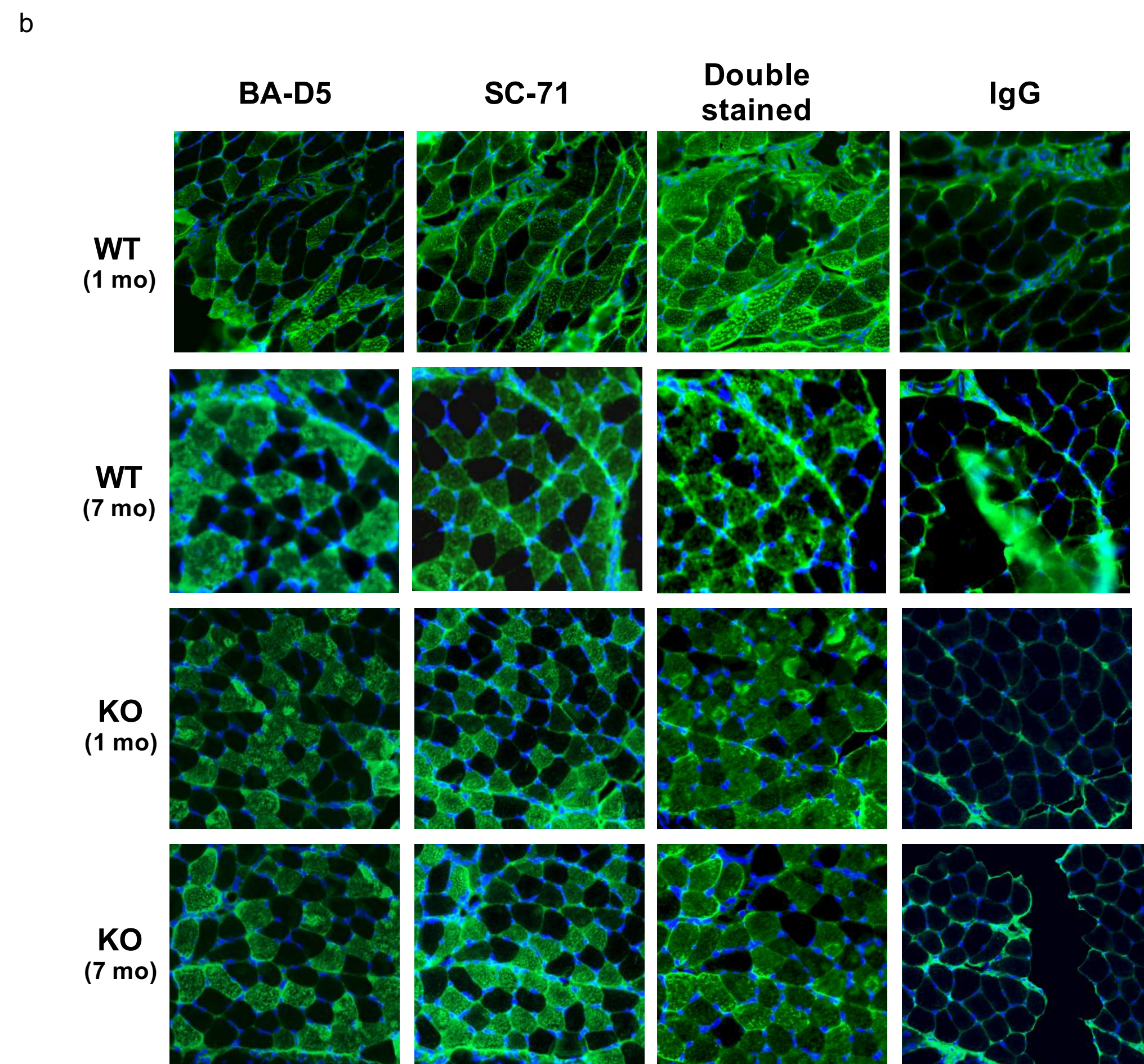
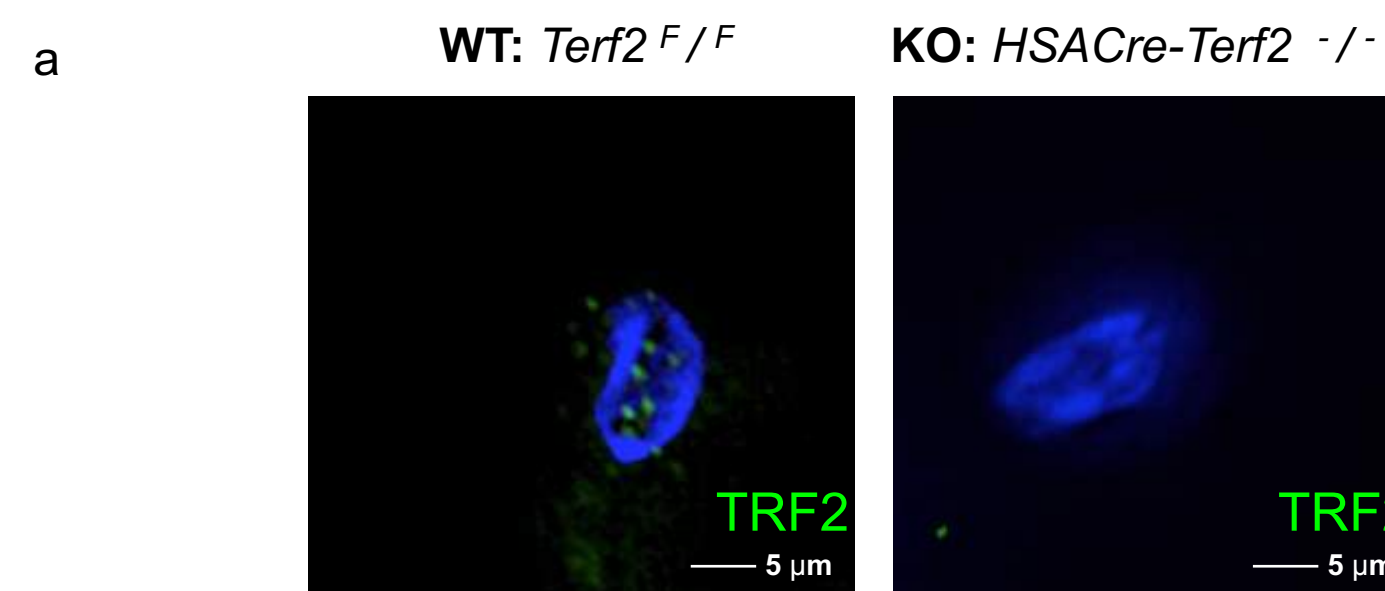




Figure S10



**BA-D5 staining** : fiber Type I determination

**SC-71 staining** : fiber Type IIa determination

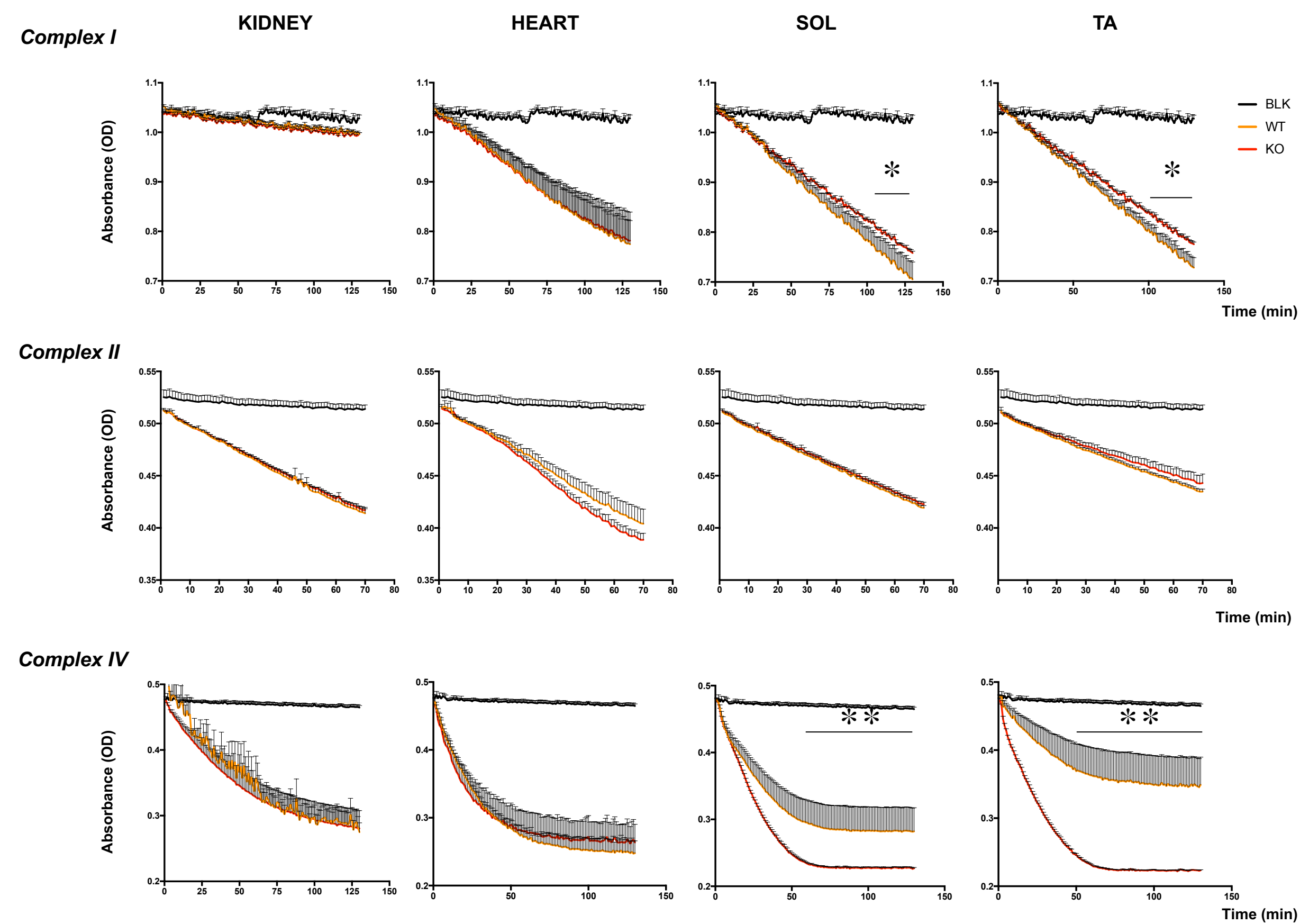
**Double staining** : unidentified fiber Type (as not stained fibers)

**IgG** : negative control



Figure S11

a



b

**Tibia Anteralis**

**Control mice: *HSACre***

