

Supplementary Material

for

“Actin Cytoskeleton Polymerization and Focal Adhesion as Important Factors in the Pathomechanism and Potential Targets for Mucopolysaccharidosis Treatment”

Figure S1. F-actin abundance in cells incubated in the presence of genistein or appropriate enzyme. Investigated lines of fibroblasts were incubated for 24 h and stained with CellMask™ Green Actin Tracking Stain. Panel A demonstrate representative micrographs of treated control lines and MPS cells. Panels B and C show quantitative analysis from 100 randomly chosen cells (10,000 events). Error bars represent standard deviation of three independent repetitions of a given experiment. Statistical analysis was performed using one-way ANOVA test. Differences were considered statistically significant relative to the control cell line (*) or untreated MPS I or MPS II (#) when $p < 0.05$.

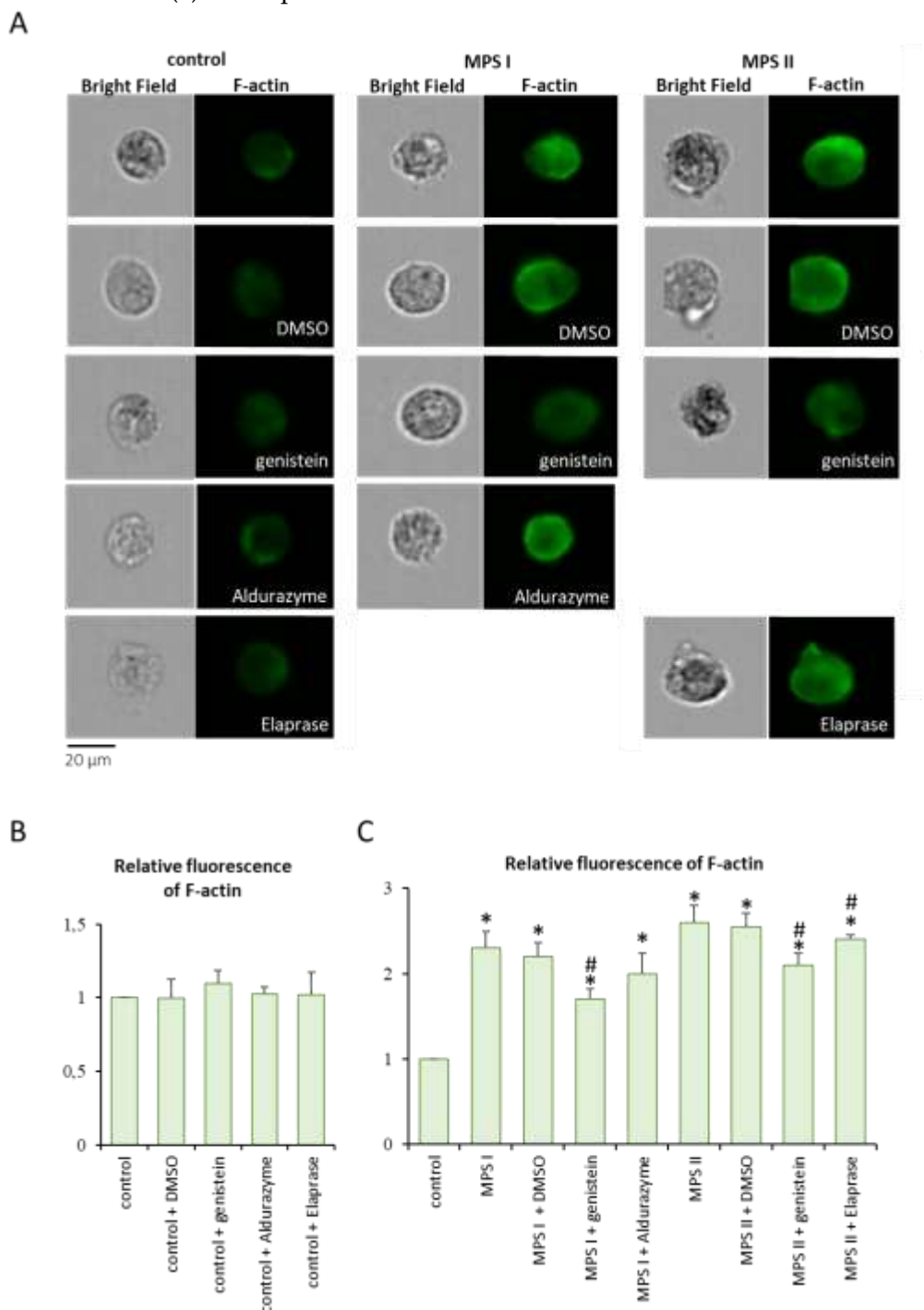


Table S1. Characteristics of MPS patients’ derived fibroblast used for transcriptomic analysis.

Cell Line	Sex *	Age *#	Mutated Gene and Its Locus *	Mutation *			
				Allele 1		Allele 2	
				DNA	Protein	DNA	Protein
MPS I	Female	1	<i>IDUA</i> , 4p16.3	G1293A	Trp402X	G1293A	Trp402X
MPS II	Male	3	<i>IDS</i> , Xp28	208insC	His70ProfsX29	none	none
MPS IIIA	Female	3	<i>SGSH</i> , 17q25.3	G1351A	Glu447Lys	G746A	Arg245His
MPS IIIB	Male	7	<i>NAGLU</i> , 17q21	C1876T	Arg626X	C1876T	Arg626X
MPS IIIC	Male	8	<i>HGSNAT</i> , 8p11.1	ND	ND	ND	ND
MPS IIID	Male	7	<i>GNS</i> , 12q14	C1063T	Arg355X	C1063T	Arg355X
MPS IVA	Female	7	<i>GALNS</i> , 16q24.3	ND	ND	ND	ND
MPS IVB	Female	4	<i>GLB1</i> , 3p22.3	TG851- 852CT	Trp273Leu	G1561T	Trp509Cys
MPS VI	Female	3	<i>ARSB</i> , 4q14.1	ND	ND	ND	ND
MPS VII	Male	3	<i>GUSB</i> , 7q21.11	G1881T	Trp627Cys	G1068A	Arg356X
MPS IX	Female	14	<i>HYAL1</i> , 3p.21.3	ND	ND	ND	ND

* According to cell line description in Coriell Institute; #age (years) at the time of cell collection; ND = not determined.

Table S2.Number of up-and down-regulated transcripts (at FDR<0.1;*p*<0.1) of gene included into Actin filament-based process (GO:0030029) term in cells of different MPS type relative to control cells (HDFa).

No. of transcripts with altered expression in particular MPS type vs HDFa line											
MPS type:	I	II	IIIA	IIIB	IIIC	IIID	IVA	IVB	VI	VII	IX
up-regulated	23	13	27	29	21	18	17	22	12	34	18
down-regulated	7	6	13	13	13	3	5	18	4	8	12
total	30	19	40	42	34	21	22	48	16	42	30