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SUPPLEMENTARY DATA

Supplementary Table 1. Antibodies used for immunofluorescence analysis

Usage	Antibodies	Manufacturer	Catalogue number	Dilution
Western Blotting	Lamin A/C (N-18)	Santa Cruz Biotechnology, CA	Sc-6215	1:500
	Beta-Actin	Santa Cruz Biotechnology, CA	Sc-47778	1:500
IPS characterization	OCT4	Stemgent, Cambridge, MA	09-0023	1:100
	SSEA-4		09-0006	
	Tra1-60		09-0010	
	Nanog		09-0020	
CM staining	Alpha-actinin	Sigma-Aldrich, St. Louis, MO	A7811	1:200
EC staining	Lectin	Sigma-Aldrich, St. Louis, MO	L9006	1:100
	vWF	Millipore	AB7568	1:100
Fibroblast staining	Fibronectin	Santa Cruz Biotechnology, CA	Sc-69777	1:100
	Vimentin		Sc-6260	1:100
Nuclear blebbing analysis	Lamin A/C (N-18)	Santa Cruz Biotechnology, CA	Sc-6215	1:100

Supplementary Table 2. PCR primers and conditions for reprogramming transgene silencing analysis

Gene	Accession no.	Forward/reverse (5'→3')	Annealing temperature (˚C)	Product (bp)	Cycles
Endo-OCT4	NM_001159542.1	5'- GACAACAATGAAAATCTTCAGGAGA -3'	57	223	30
		5' - TTCTGGCGCCGGTTACAGAACCA -3'			
Endo-NANOG	NM_024865.2	5'-AAGACAAGGTCCCGGTCAAG	57	583	30
		5'- CCTAGTGGTCTGCTGTATTAC			
*Exo-OCT4	NM_001159542.1	5'-TCAAGCCTCAGACAGTGGTTC3'	57	236	30
		5'-GGCCCGATTCCTGGCCCTCA3'			
*Exo-NANOG	NM_024865	5'-TCAAGCCTCAGACAGTGGTTC-3'	57	296	30
		5'-CTTCAAAGCAAGGCAAGCTT-3'			
GAPDH	NM_011406	5'- AGCCACATCGCTCAGACACC -3'	60	157	30
		5'- GTACTCAGCGGCCAGCATCG -3'			

Abbreviation: OCT4 : octamer-binding transcription factor 4 ; GAPDH: Glyceraldehyde 3-phosphate dehydrogenase.
*Forward primer was probed on EF1-alpha coding region, which is upstream of the OCT-4/NANOG cDNA sequence in the lentiviral reprogramming vector.

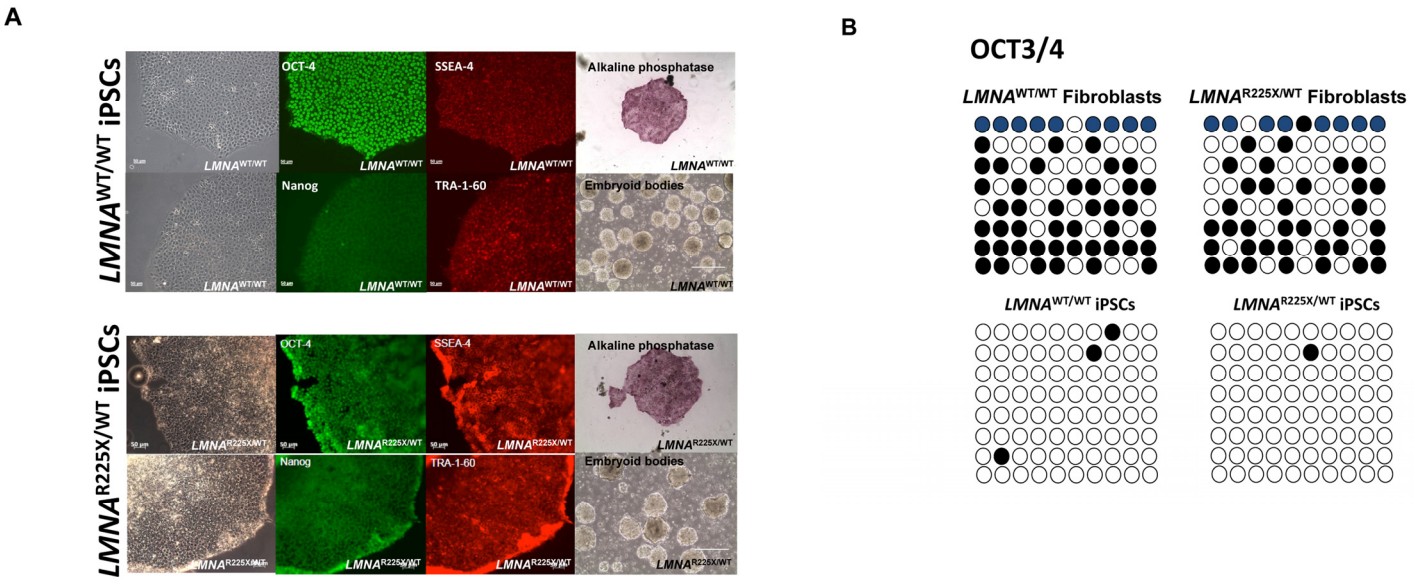

Figure 1. Generation of patient-specific iPSC lines. (A) Immunofluorescence analysis of pluripotent markers OCT4 (Green), SSEA4 (Red), NANONG (Green), and TRA-1-60; the expression of alkaline phosphatase; and embryoid body formation in representative iPSC clones derived from the proband (II:7) and the healthy control, **(B)** Oct-4 promoter methylation analysis with bisulfate pyro-sequencing in two parent fibroblast lines ($LMNA^{R225X/WT}$ and $LMNA^{WT/WT}$), and their iPSC lines.

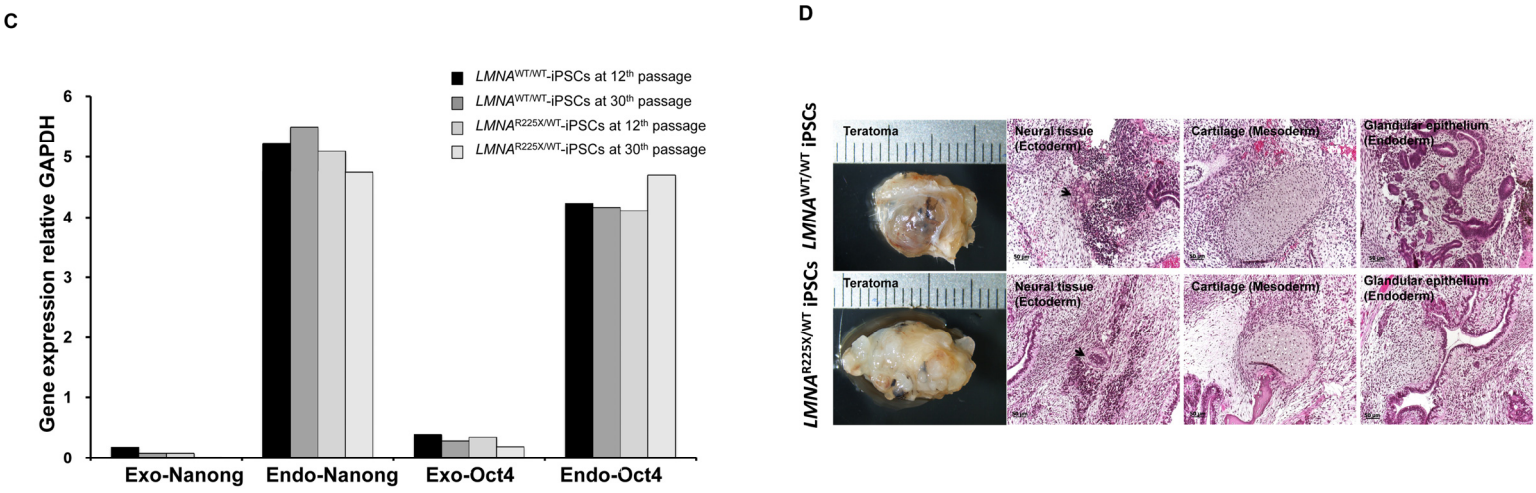

Figure 1. Generation of patient-specific iPSC lines. (C) RT-PCR analyses of the endogenous and exogenous level of OCT4 and Nanog of LMNA$^{R225X/WT}$ and LMNA$^{WT/WT}$ iPSC lines at 12th and 30th passages, **(D)** Teratoma formation and the histological section of teratoma formed 4-6 weeks after subcutaneous injection of *LMNA*$^{R225X/WT}$ and *LMNA*$^{WT/WT}$ iPSC lines into NOD/SCID mice.

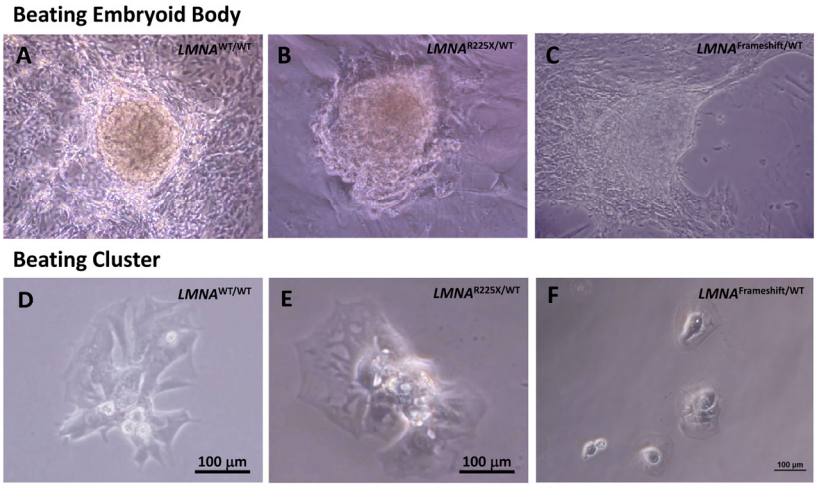

Figure 2. (A, B, & C) Beating embryoid bodies derived from *LMNA*$^{R225X/WT}$, *LMNA*$^{Frameshift/WT}$, and *LMNA*$^{WT/WT}$ iPSCs; **(D, E, & F)** Spontaneously beating cell clusters after dissociation. Videos of these beating embryoid bodies and clusters were available in supplemental materials.

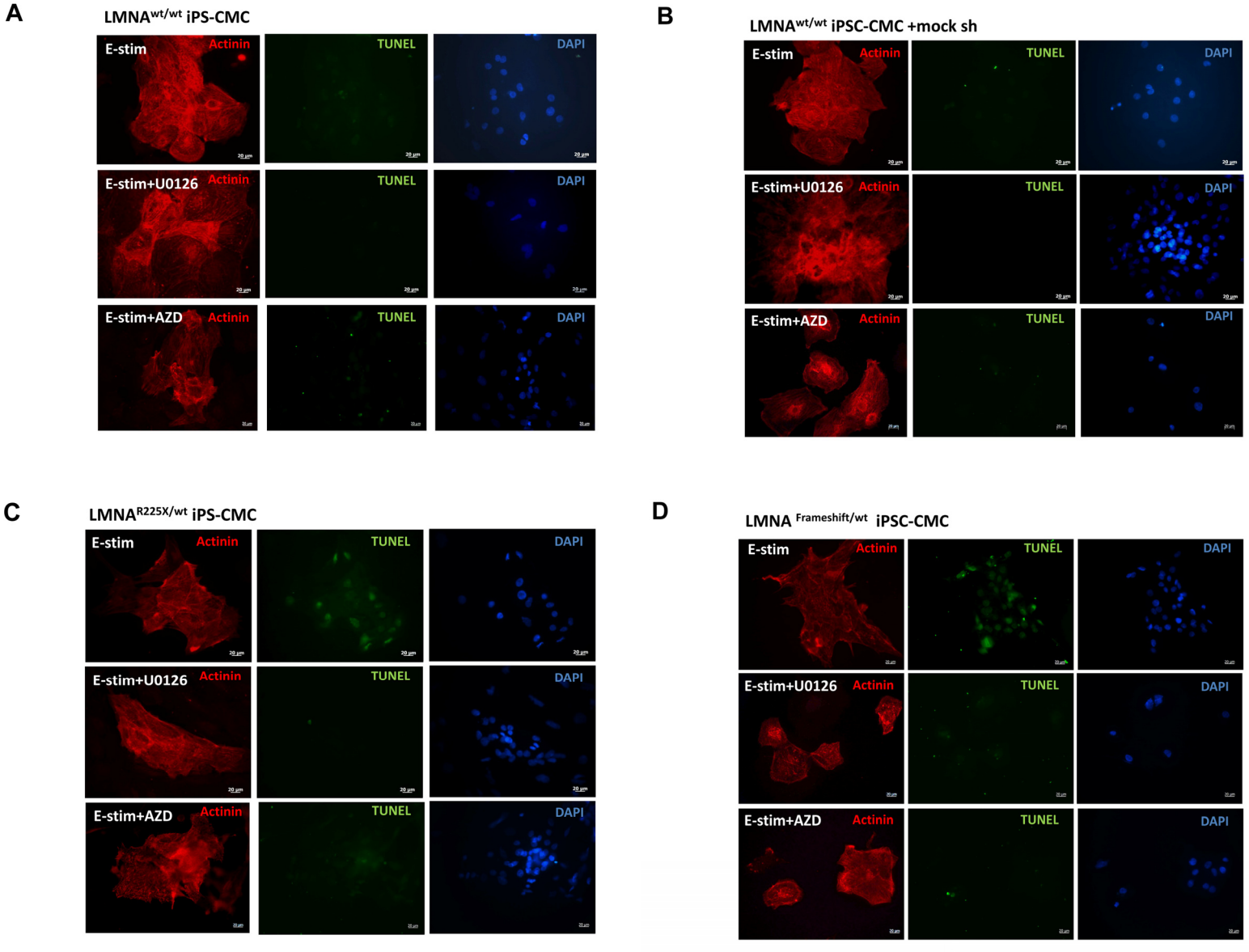

Figure 3. Electrical stimulation inducing apoptosis in cardiomyocytes derived from *LMNA*$^{R225X/WT}$ & *LMNA*$^{Frameshift/WT}$ iPSCs. Representative TUNEL assay and co-immunofluorescence staining of alpha-actinin in cardiomyocytes derived from **(A)** *LMNA*$^{WT/WT}$ iPSCs, **(B)** *LMNA*$^{R225X/WT}$ iPSCs, **(C)** *LMNA*$^{Frameshift/WT}$ iPSCs, **(D)** *LMNA*$^{WT/WT}$ iPSCs treated with mock shRNA.

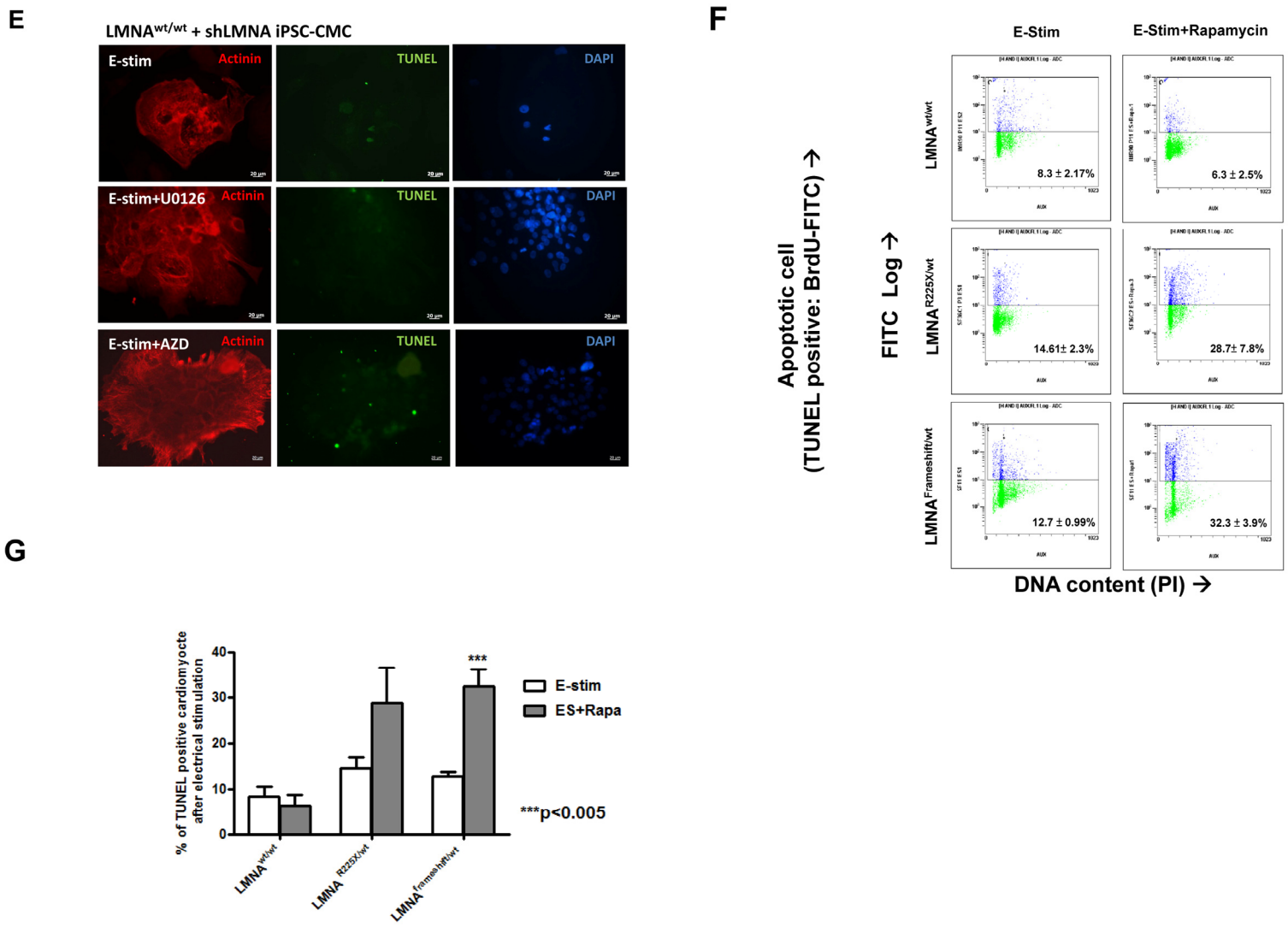

Figure 3. Electrical stimulation inducing apoptosis in cardiomyocytes derived from *LMNA*$^{R225X/WT}$ & *LMNA*$^{Frameshift/WT}$ iPSCs. **(E)** *LMNA*$^{WT/WT}$ iPSCs treated with sh*LMNA*, **(F and G)** Quantification of apoptotic cardiac differentiated iPSCs in presence of rapamycin by APO-BrdU TUNEL assay at baseline and after electrical stimulation. The percentage of cardiomyocytes with apoptosis was determined by FACS analysis by FL-1 positive gating. Unpaired t-test was performed between treatment and baseline n=3.