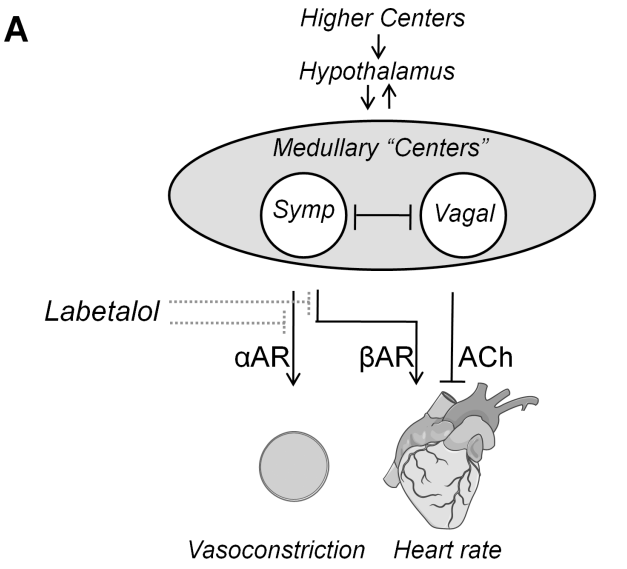
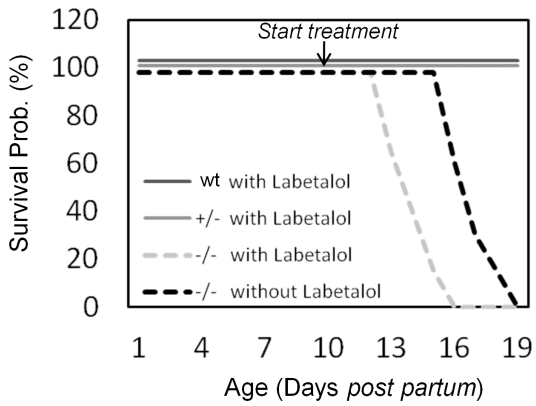




B

Age	Parameter Measured	wt		+/-		-/-	
		No Labetalol	*Labetalol*	*No Labetalol*	*Labetalol*	*No Labetalol*	*Labetalol*
PP13	P time (ms)	23.8 ± 3.3	25.3 ± 1.8	26.1 ± 2.7	25.2 ± 2.6	23.7 ± 2.6	29.8 ± 3.2*
	QRS time (ms)	14.0 ± 2.4	17.4 ± 0.9*	13.7 ± 0.8	16.5 ± 0.3*	14.4 ± 1.7	17.7 ± 1.2*
	QT time (ms)	37.2 ± 9.6	32.6 ± 2.4	33.7 ± 3.1	32.2 ± 2.9	33.2 ± 5.9	30.9 ± 3.1
	Heart Rate (bpm)	345 ± 30	383 ± 57	422 ± 58	399 ± 55	396 ±51	232 ±20*

Figure S1. Cardiac effects of α- and β-adrenergic receptor antagonist treatment in *LMNA*^{GT} mice. (A) Labetalol inhibits sympatic activity by antagonizing α- and β-adrenergic receptors, thereby suppressing sympatic stimulation of heart rate and vasoconstriction. Arrows and T-shaped ends indicate respectively stimulatory and inhibitory actions. **(B)** ECG analysis indicating heart rate ,P time, QRS duration and QT time at the age of PP13, 3 days after the start (PP10) of subcutaneous labetalol or control injections in all genotypes (See Materials & Methods). Asterisks in this figure indicate significant (P<0.05; N=5) differences between labetalol and control injections within the same genotype. **(C)** Survival curve for *LMNA*$^{GT-/-}$ mice treated from PP10 onward with labetalol or control injections (dashed lines) versus survival curves for labetalol treated *LMNA*$^{GT+/-}$ and WT siblings (straight lines) (N=5 each condition).

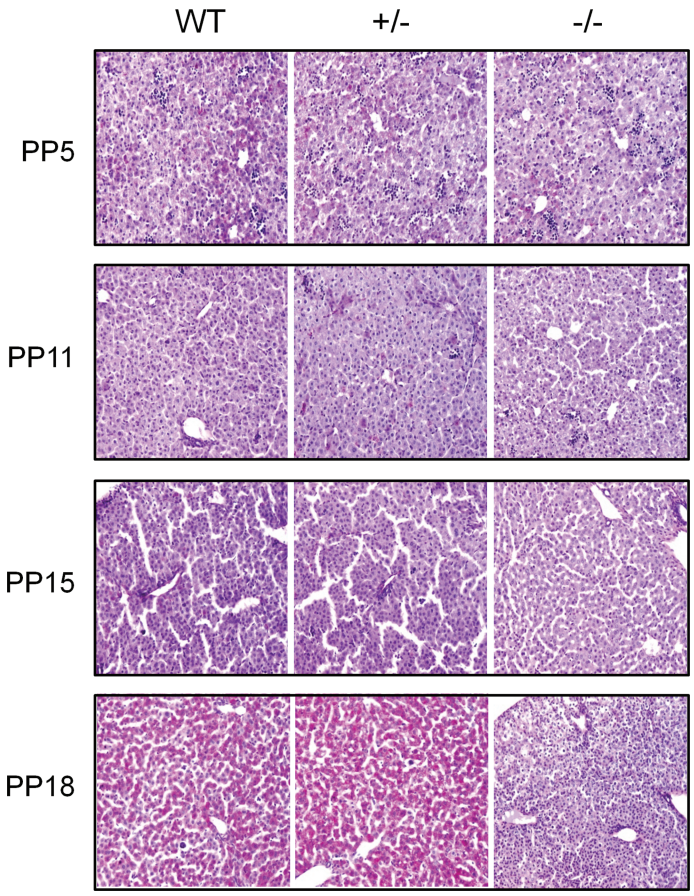
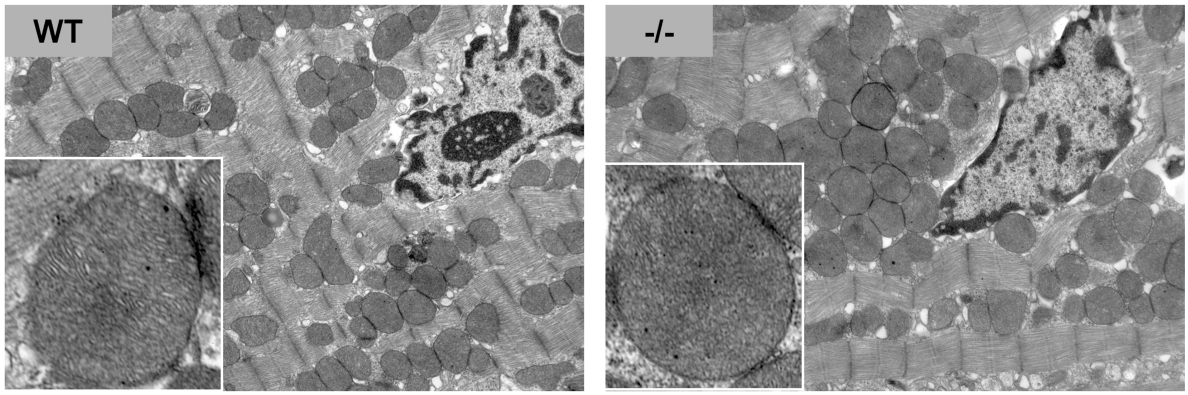


Figure S2. Post-natal hepatic glycogen storage in *LMNA*$^{GT-/-}$ mice. Formalin fixed liver sections were Periodic Acid Shiff (PAS) stained to visualize glycogen deposits in a light purple/pink color. *LMNA*$^{GT-/-}$ mice show absence of glycogen deposits at all ages investigated, whereas in WT and *LMNA*$^{GT+/-}$ livers glycogen is detectable at day PP18.

A



B

Age	Parameter Measured	N=	wt	-/-
	Mitochondrial DNA copy number:			
	Heart Left Ventricle	4	6.93 ± 2.18	8.38 ± 2.36
	Skeletal Muscle	4	4.08 ± 1.24	4.18 ± 0.42
PP15	Mitochondrial Complex Activity:			
	Citrate Synthase (CS)	5	1508 ± 118	1712 ± 307*
	Complex I/CS	5	0.182 ± 0.013	0.178 ± 0.011
	Complex II/CS	5	0.122 ± 0.007	0.114 ± 0.0024*
	Complex III/CS	5	0.406 ± 0.046	0.345 ± 0.122
	Complex IV/CS	5	0.408 ± 0.05	0.372 ± 0.06
	Complex V/CS	5	0.461 ± 0.028	0.525 ± 0.061

Figure S3. Mitochondrial morphology and functioning in *LMNA*$^{GT-/-}$ mice. (A) Transmission Electron Microscopy pictures of left ventricle cardiac tissue at day PP15 WT and *LMNA*$^{GT-/-}$ mice, showing cardiac muscle fiber and Z-disc orientation, and (in close-up) a typical example of a mitochondrion. **(B)** Mitochondrial DNA copy numbers (N=4) and complex activities (N=5) in both quadriceps skeletal and cardiac muscle as determined at PP15 (See material and methods). Asterisks indicate significant differences for WT and *LMNA*$^{GT-/-}$values ($P < 0.05$).