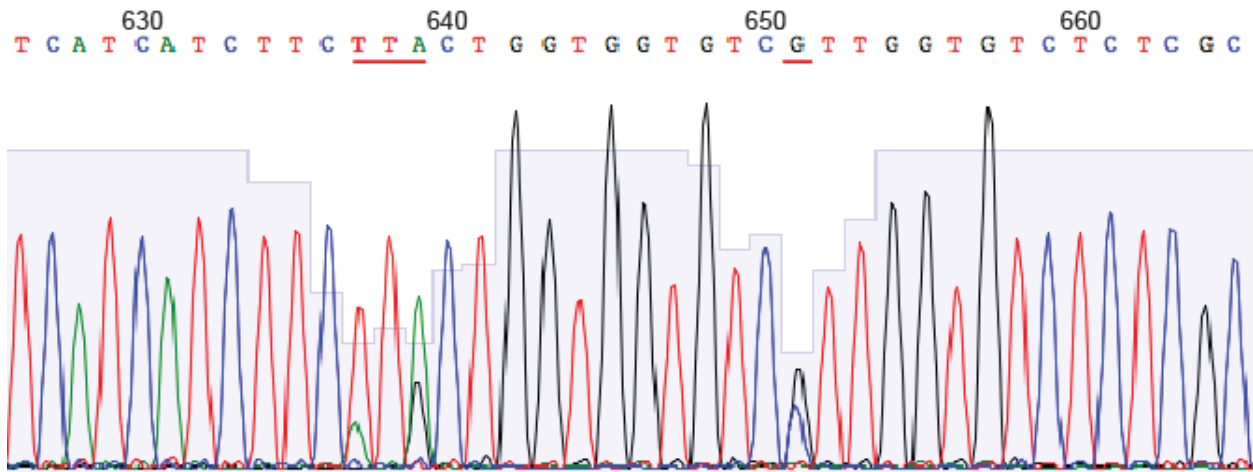


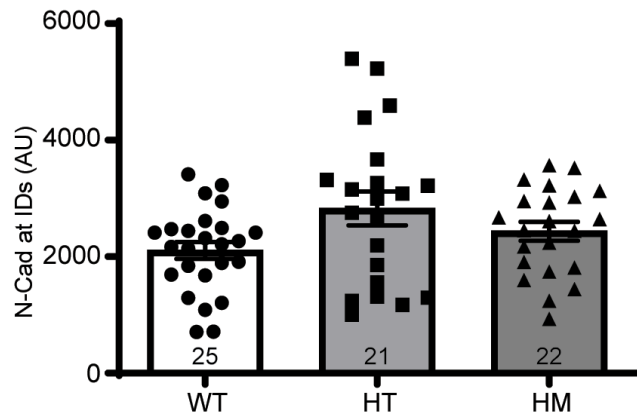
## **Supplemental Data**

### **GJA1-20k Deficiency Leads to Sudden Deaths via Limiting Connexin43 Trafficking**

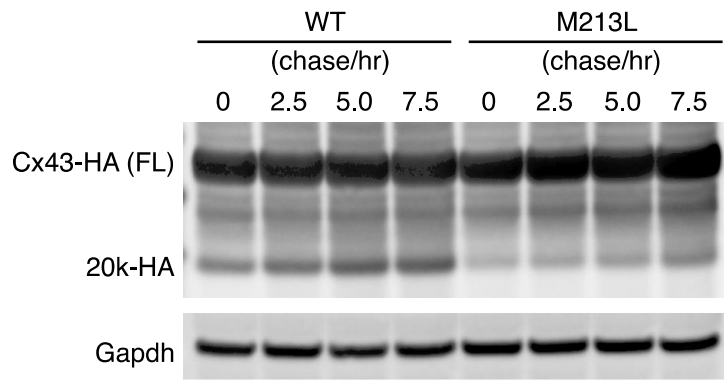
Shaohua Xiao, Daisuke Shimura, Rachel Baum, Diana M Hernandez, Sosse Agvanian, Yoshiko Nagaoka, Makoto Katsumata, Paul D Lampe, André G Kléber, TingTing Hong, Robin M Shaw



**Supplemental Figure 1. Genomic sequence flanking the targeted ATG to TTA mutation in the founder of a *GJAI*<sup>M213L</sup> mouse clone used in this study.** The sequence confirms the successful mutation of ATG to TTA, which results in the M213L mutation in Cx43, and TCC to TCG, a silent mutation to disrupt a nearby PAM. The mutations are underlined.



**Supplemental Figure 2. The quantification of N-cadherin at the intercalated discs of the heart sections of 2-3 weeks old mice.** There was no significant difference in N-Cad expression at intercalated discs among genotypes. Kruskal-Wallis test, followed by Dunn's multiple comparisons, with n=25, 21, and 22 images (WT, HT, and HM, respectively) from 7 (WT) or 6 (HT and HM) mice. Data represents mean  $\pm$  SEM.



**Supplemental Figure 3. Expression of GJA1-20k in HEK293 cells transfected with WT and M213L mutated mouse Cx43.** A representative western blot of exogenously expressed, HA-tagged full length Cx43 (FL) and GJA1-20k protein. Gapdh was used as a loading control. n = 4 experiments.

**Table S2. Echocardiographic parameters of adult M213L mice (9-10 weeks old)**

	WT (n=8)		HT(n=5)		<i>P</i> value
LVAWd, mm	0.73	± 0.04	0.66	± 0.03	0.2082
LVAWs, mm	1.11	± 0.06	1.02	± 0.06	0.4126
LVIDd, mm	3.89	± 0.07	3.66	± 0.13	0.1826
LVIDs, mm	2.70	± 0.09	2.52	± 0.17	0.5237
LVPWd mm	0.67	± 0.06	0.74	± 0.06	0.6216
LVPWs mm	0.96	± 0.06	1.02	± 0.09	0.4561
HR, bpm	518.30	± 10.29	474.80	± 34.83	0.3543
LVmass, mg	75.58	± 4.63	68.54	± 3.62	0.3543
EF(LAX), %	58.48	± 1.68	60.65	± 1.81	0.2844
FS(LAX), %	21.44	± 0.84	19.70	± 1.30	0.1709
LVvol,d, μL	55.52	± 3.65	43.61	± 2.26*	0.0295
LVvol,s, μL	23.28	± 2.11	17.19	± 1.17	0.0932
E/A	2.07	± 0.48	1.55	± 0.15	0.8329
E/e'	18.44	± 2.66	18.44	± 1.69	0.9433

<sup>A</sup> \* *P* < 0.05 vs. WT, Mann-Whitney test with two-tailed *P* value. Data represents mean ± SEM.