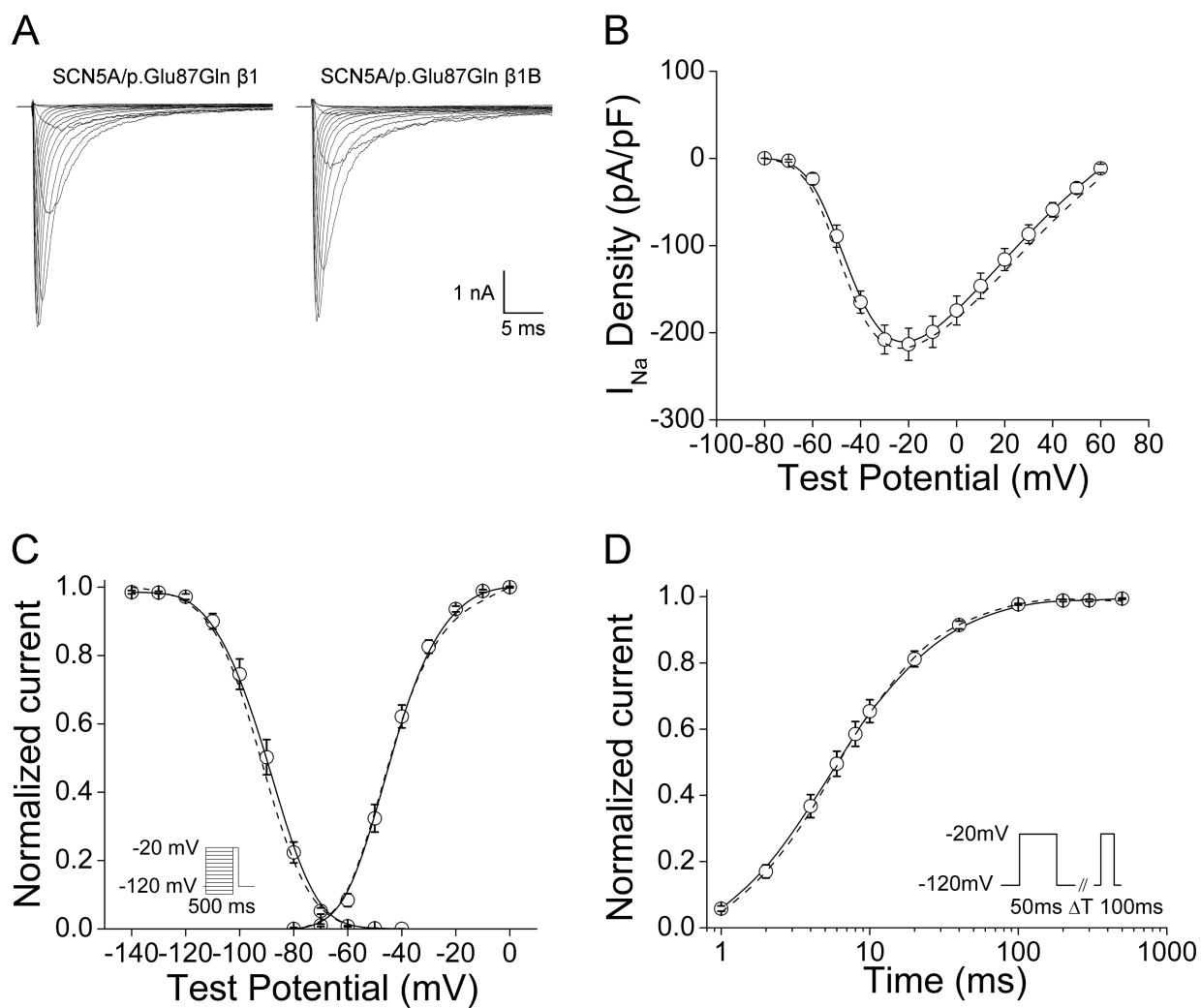


Supplemental Figure 1



Supplemental Figure 1 Comparison of p.Glu87Gln mutant function in $\beta 1$ - and $\beta 1B$ -subunits. (A) Representative traces of sodium current. (B) Current-voltage relationships for the p.Glu87Gln $\beta 1B$ -subunit (open circles, $n = 15$). The dotted lines in panels B-D indicate data for the Glu87Gln $\beta 1$ -subunit shown in Figure 5. (C) Voltage dependence of activation and inactivation. (D) Recovery from inactivation. Biophysical properties are provided in Supplemental Table 4.

Supplemental Table 1 Biophysical parameters of p.Glu87Gln β 1B

	Voltage dependence of activation			Voltage dependence of inactivation			Recovery from inactivation		
	$V_{1/2}$, mV	k , mV	N	$V_{1/2}$, mV	k , mV	N	τ_f , ms (%)	τ_s , ms (%)	N
Nav1.5 ^C	-46.2±1.0	7.1±0.4	29	-83.8±1.8	7.6±0.2	17	7.7±1.1 (87.2±1.1)	56.4±9.8 (11.6±1.0)	12
Nav1.5/WT β 1B ^C	-50.6±0.7 ^A	6.3±0.3	28	-94.2±1.3 ^A	7.6±0.2	14	7.4±1.0 (86.5±1.2)	43.3±8.6 (13.1±1.1)	14
Nav1.5/p.Glu87Gln β 1B	-44.2±0.6 ^B	8.2±0.4	15	-90.6±0.8 ^A	6.7±0.2	12	7.4±1.0 (87.6±1.1)	48.9±4.1 (11.5±1.1)	12
Nav1.5/p.Glu87Gln β 1 ^{C,D}	-44.9±1.4	7.7±0.4	18	-92.5±1.7 ^A	6.8±0.2	12	7.7±1.1 (89.1±1.1)	52.0±9.5 (10.4±1.1)	10

Values are shown as mean $\pm$ SEM. ^A P < 0.01 when comparing versus Nav1.5 alone; ^B P < 0.01 when comparing versus Nav1.5 / WT β 1B. ^CValues of Nav1.5, Nav1.5/WT β 1B, and Nav1.5/p.Glu87Gln β 1 are also shown in Tables 1 and 2.

^DValues of Nav1.5/p.Glu87Gln β 1 were not compared with those of Nav1.5/WT β 1B.

Supplemental Table 2 Sequence of oligonucleotide primers for *SCN1B* screening

Exon	Forward primer	Reverse primer
1	CCTCTCGCGCCGCTATTAATACC	CGTGTCCCGAGCGCCACTC
2	GTGAGGGGTCTGGCATTGCT	TCCAGGTCAGCAATCACAGCAT
3	GGGTCAGGTAAGGGAAGAGAGG	TGTCCCTCCATCTGGCTCTC
4	CCCAGGGAGGTTGAGCCACT	CCTCTCTGGCAAGTGTGACGG
5	ACCGCTACCTTCCCCCATCC	GACTTCGGCCCCTCCCAATC
3A	GTCTACCGCCTGCTCTTCTTCG	CAGGCGTGTGCCTGCAGCTG

Supplemental Table 3 Characteristics of
non-diseased heart donors

Subject No.	Gender	Age (years)	Tissues
1	Female	65	LV, RV
2	Female	49	LV, RV
3	Female	41	LV, RV
4	Female	35	LV, RV
5	Male	43	LV
6	Male	42	LV
7	Male	38	RV
8	Female	18	RV
9	Male	18	Purkinje
10	Male	20	Purkinje
11	Female	46	Purkinje
12	Female	49	Purkinje
13	Female	28	Purkinje
14	Female	65	Purkinje

LV, left ventricle; RV, right ventricle.

Supplemental Table 4 Sequence of oligonucleotide primers for quantitative PCR

Gene isoform	GenBank accession No.	Target Exons	Forward primer	Reverse primer	Probe
<i>SCN1B</i> (transcript isoform β 1)	NM 001037	3-4	TaqMan® Gene Expression Assay Hs00168897_m1		
<i>SCN1B</i> (transcript isoform β 1B)	NM 199037	3A	CCACCGGAGAGCCAGAT	GCCTGTCCTGTCCACTGC	CTGCCATCTGTCCCTCC
<i>HPRT1</i>	NM 000194	6-7	TaqMan® Gene Expression Assay Hs99999909_m1		

HPRT1, hypoxanthine phosphoribosyltransferase 1.