

SUPPLEMENTAL MATERIAL

Supplemental Table 1: LQT1 and LQT2 Mutations by Location and Amino Acid Coding

Group	Location and Coding	Exon	Number of Patients
LQT1	Pre-Pore		
	Ile 235 Asn	exon 5	2
	Gly 168 Arg	exon 3	5
	Ala 46 Thr	exon 1	2
	Arg 174 Leu	exon 3	1
	Subtotal		10
	Pore		
	Val 280 Glu	exon 6	1
	Thr 322 Met	exon 7	3
	Ala 344 Ala	exon 7	1
	Ser 349 Ter	exon 8	5
	Gly 325 Glu	exon 7	1
	Subtotal		11
	Post-Pore		
	P400 fs/62X	exon 9	1
	Arg 594 Gln	exon 15	1
	Ser 546 Leu	exon 13	1
	Arg 360 Met	exon 8	1
	Gln 530 Ter	exon 12	1
	Gly 635 Arg	exon 16	1
	Gln 359 Ter (X)	exon 8	1
	Subtotal		7
	Other	intron 12	1
LQT2	Pore		
	Ile 593 Lys	exon 7	1
	Leu 552 Ser	exon 7	5
	His 562 Arg	exon 7	1
	Thr 613 Met	exon 7	1
	Gly 628 Ser	exon 7	1
	Pro 605 Leu	exon 7	1

	Thr 613 Met	exon 7	1
	Subtotal		11
	Non-pore		
	His 70 Arg	exon 2	1
	Pro 123 fs +15x	exon 2	1
	Arg 176 Trp	exon 4	3
	Thr 74 Arg	exon 2	1
	Pro 347 Ser	exon5	1
	Val 94 Met	exon 2	1
	Pro 393 Leu	exon 6	2
	Subtotal		10

Supplemental Table 2: QTc Intervals (ms) During EST in LQTS Patients with and without Beta-Blockers.

	Rest	Peak exercise	1 min recovery	3 min recovery	5 min recovery	7 min recovery	9 min recovery
LQT1							
Beta-Blocker (n=17)	450±23	479±31	491±42	485±30	484±20	480±24	480±25
No Beta-Blocker (n=12)	454±39	495±37	474±29	495±44	489±39	487±33	491±36
<i>P</i> -value	0.75	0.22	0.24	0.48	0.67	0.5	0.36
LQT2							
Beta-Blocker (n=10)	491±38	429±16	425±16	457±36	475±20	474±18	482±20
No Beta-Blocker (n=11)	460±25	447±39	441±32	464±33	462±23	482±20	474±25
<i>P</i> -value	0.05	0.18	0.18	0.64	0.17	0.32	0.83

Supplemental Table 3: QTc Intervals (ms) During EST in Concealed (cLQT) Patients.

	cLQT1	cLQT2	<i>P</i> -value
Rest	428±25	440±14	0.25
Peak exercise	467±26	441±24	0.04
1 min recovery	471±39	428±28	0.01
3 min recovery	477±42	456±34	0.68

5 min recovery	474±27	466±23	0.49
7 min recovery	475±34	472±17	0.78
9 min recovery	472±34	471±19	0.95

Supplemental Table 4: QTc intervals during EST Based on LQT1 and LQT2 Gene Location

	Peak Exercise	1 minute	3 minutes	5 minutes	7 minutes	9 minutes
LQT1						
Pre-Pore	456±40	458±37	472±41	482±25	481±21	482±30
Pore	471±30	456±37	470±30	470±23	480±27	475±26
Post-Pore	482±60	461±45	490±40	484±23	490±30	473±25
LQT2						
Non-Pore	471±34	458±28	472±32	471±30	476±27	479±24
Pore	472±51	475±52	488±53	489±42	485±27	489±37

P-value NS for comparison of LQT1 pre-pore vs. pore vs post pore

P-value NS for comparison of LQT2 non-pore vs. pore