

Supplementary data

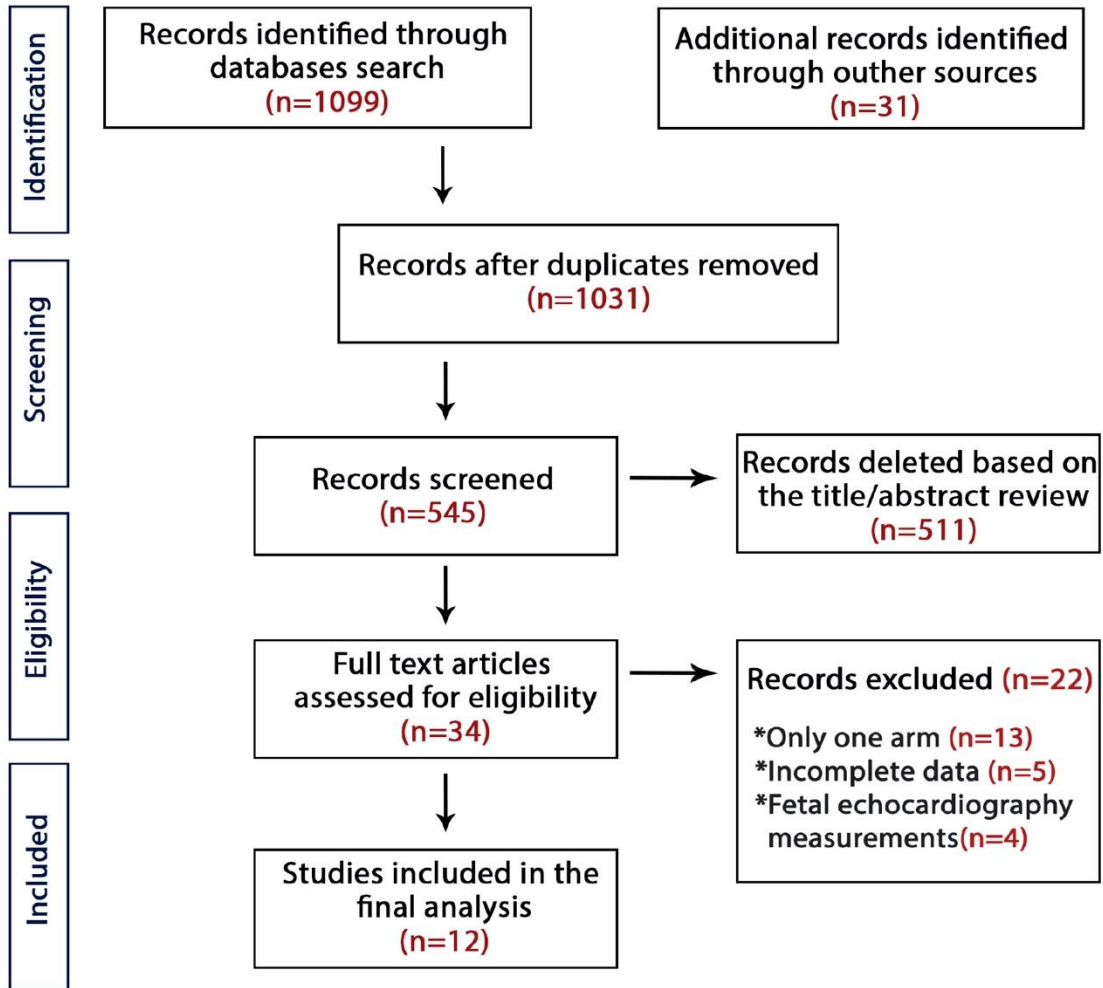
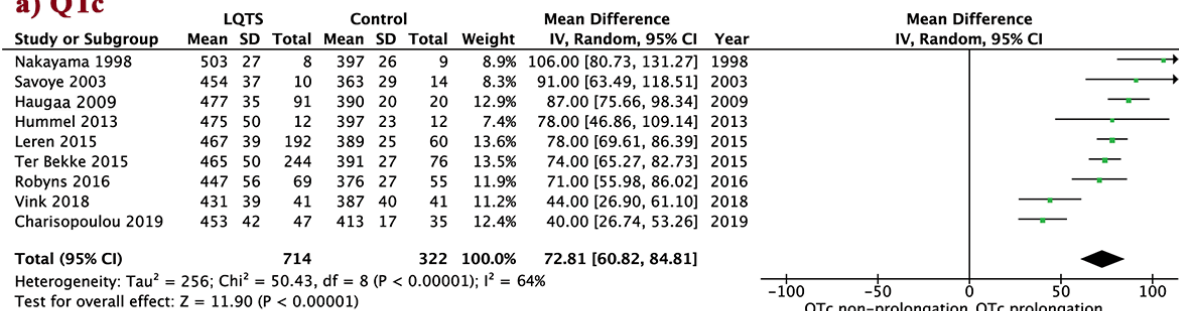
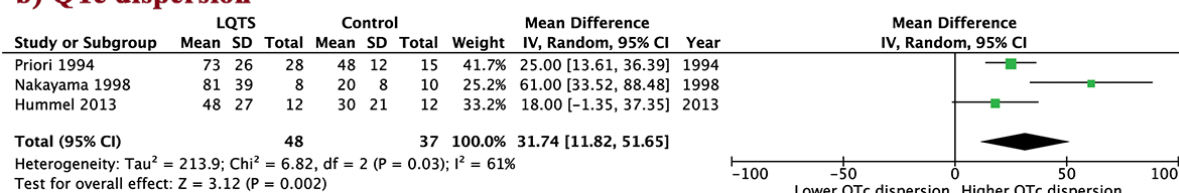


Figure S1. Flow chart of study section

a) QTc



b) QTc dispersion



c) RR interval

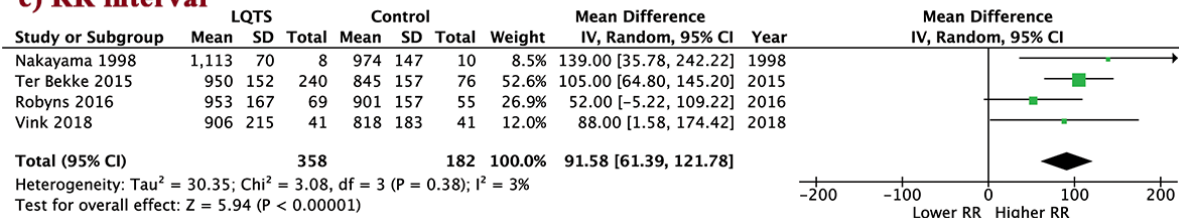
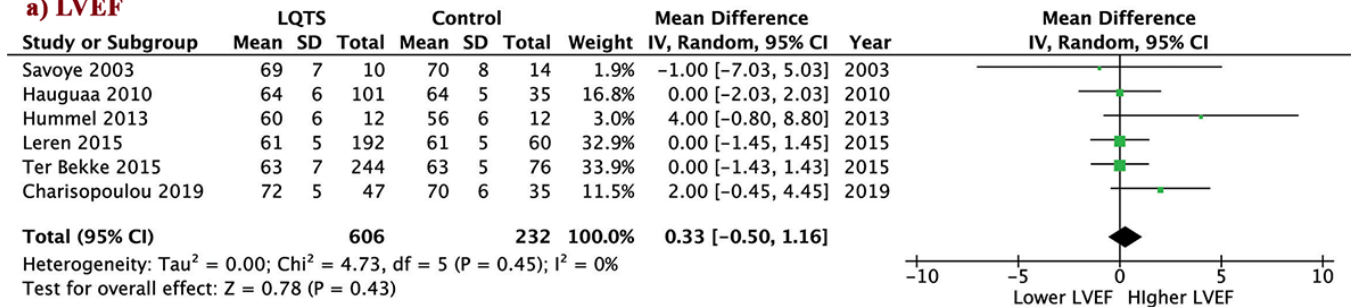


Figure S2. Electrical abnormalities in LQTS patients vs. control

LV systolic function

a) LVEF



b) LV GLS

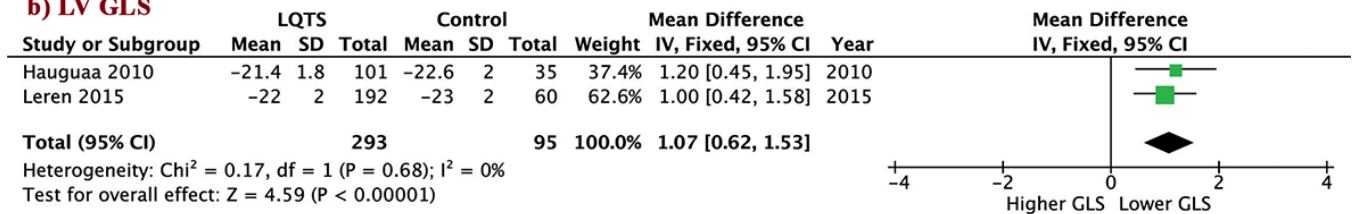
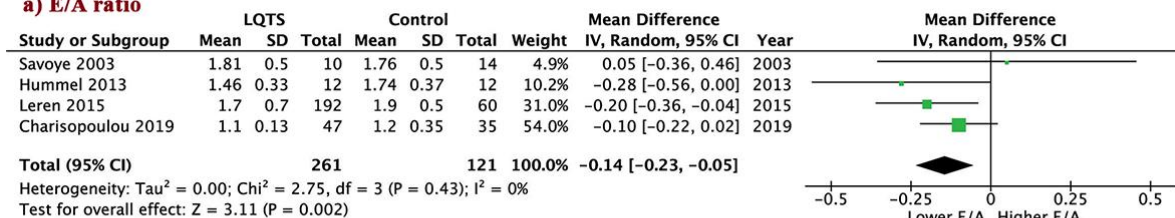


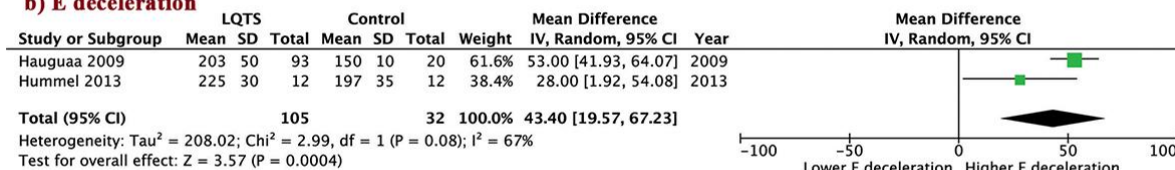
Figure S3. LV systolic function difference: comparison between LQTS vs. Control

LV diastolic function

a) E/A ratio



b) E deceleration



c) IVRT

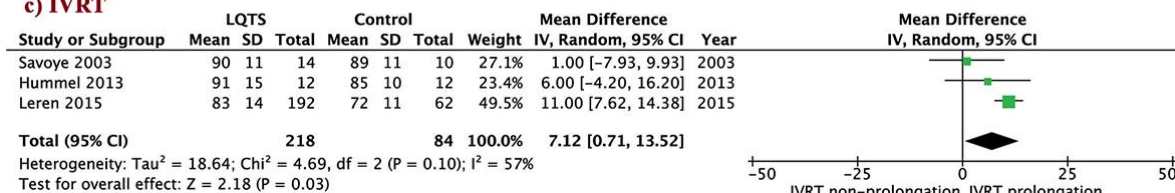


Figure S4. LV diastolic function difference: comparison between LQTS vs. Control

Electrical abnormalities in LQTS

a) QTc

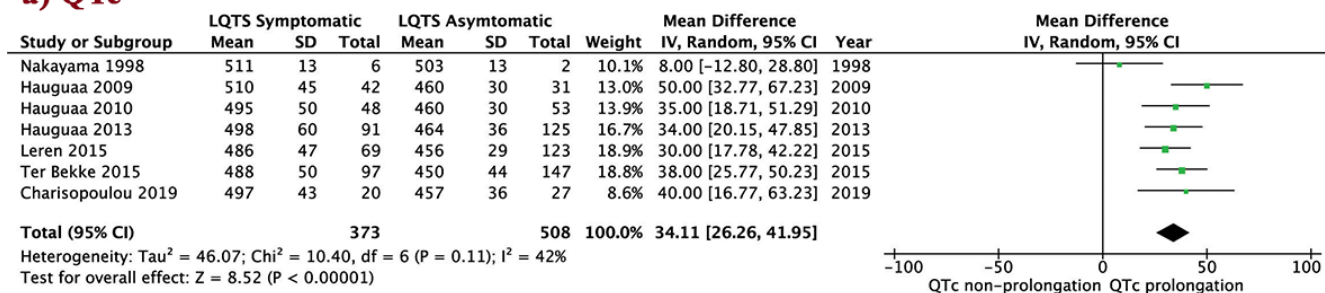


Figure S5. Electrical abnormalities in LQTS: comparison between symptomatic vs. asymptomatic

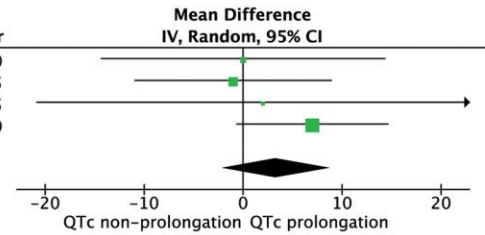
a) QTc

Study or Subgroup	LQT1			LQT2			Weight	Mean Difference IV, Random, 95% CI	Year
	Mean	SD	Total	Mean	SD	Total			
Hauguaa 2010	470	35	64	470	30	26	14.3%	0.00 [-14.37, 14.37]	2010
Leren 2015	462	34	130	463	30	53	29.8%	-1.00 [-10.97, 8.97]	2015
Ter Bekke 2015	491	54	38	489	50	42	5.7%	2.00 [-20.88, 24.88]	2015
Charisopoulou 2019	458	15	36	451	10	11	50.2%	7.00 [-0.68, 14.68]	2019

Total (95% CI) 268 132 100.0% 3.33 [-2.11, 8.77]

Heterogeneity: $\tau^2 = 0.00$; $\chi^2 = 1.82$, $df = 3$ ($P = 0.61$); $I^2 = 0\%$

Test for overall effect: $Z = 1.20$ ($P = 0.23$)



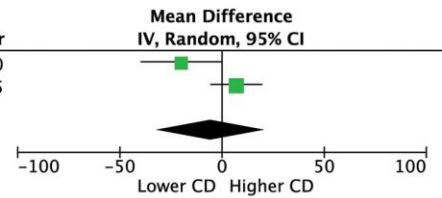
b) Contraction duration

Study or Subgroup	LQT1			LQT2			Weight	Mean Difference IV, Random, 95% CI	Year
	Mean	SD	Total	Mean	SD	Total			
Hauguaa 2010	441	63	130	461	62	53	45.8%	-20.00 [-39.90, -0.10]	2010
Leren 2015	425	37	130	418	41	53	54.2%	7.00 [-5.74, 19.74]	2015

Total (95% CI) 260 106 100.0% -5.37 [-31.74, 20.99]

Heterogeneity: $\tau^2 = 291.85$; $\chi^2 = 5.02$, $df = 1$ ($P = 0.03$); $I^2 = 70\%$

Test for overall effect: $Z = 0.40$ ($P = 0.69$)



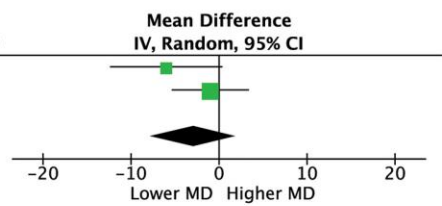
c) Mechanical dispersion

Study or Subgroup	LQT1			LQT2			Weight	Mean Difference IV, Random, 95% CI	Year
	Mean	SD	Total	Mean	SD	Total			
Hauguaa 2010	31	14	64	37	14	26	38.8%	-6.00 [-12.38, 0.38]	2010
Leren 2015	31	13	130	32	14	53	61.2%	-1.00 [-5.38, 3.38]	2015

Total (95% CI) 194 79 100.0% -2.94 [-7.71, 1.84]

Heterogeneity: $\tau^2 = 4.70$; $\chi^2 = 1.60$, $df = 1$ ($P = 0.21$); $I^2 = 38\%$

Test for overall effect: $Z = 1.21$ ($P = 0.23$)



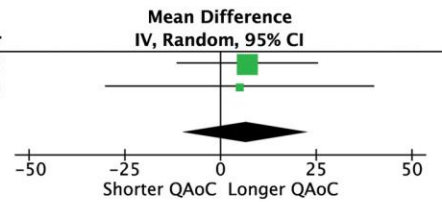
d) QAoC

Study or Subgroup	LQT1			LQT2			Weight	Mean Difference IV, Random, 95% CI	Year
	Mean	SD	Total	Mean	SD	Total			
Ter Bekke 2015	420	50	38	413	31	42	78.4%	7.00 [-11.46, 25.46]	2015
Charisopoulou 2019	390	45	36	385	54	11	21.6%	5.00 [-30.13, 40.13]	2019

Total (95% CI) 74 53 100.0% 6.57 [-9.77, 22.91]

Heterogeneity: $\tau^2 = 0.00$; $\chi^2 = 0.01$, $df = 1$ ($P = 0.92$); $I^2 = 0\%$

Test for overall effect: $Z = 0.79$ ($P = 0.43$)



e) EMW

Study or Subgroup	LQT1			LQT2			Weight	Mean Difference IV, Random, 95% CI	Year
	Mean	SD	Total	Mean	SD	Total			
Ter Bekke 2015	-68	37	38	-68	43	42	35.5%	0.00 [-17.54, 17.54]	2015
Charisopoulou 2019	-40	23	36	-48	18	11	64.5%	8.00 [-5.02, 21.02]	2019

Total (95% CI) 74 53 100.0% 5.16 [-5.30, 15.61]

Heterogeneity: $\tau^2 = 0.00$; $\chi^2 = 0.52$, $df = 1$ ($P = 0.47$); $I^2 = 0\%$

Test for overall effect: $Z = 0.97$ ($P = 0.33$)

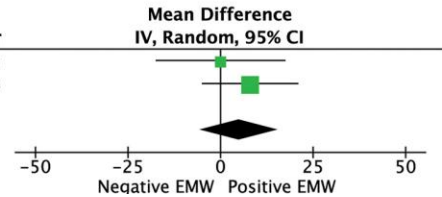


Figure S6. Electrical and mechanical abnormalities in LQT1 vs. LQT2

Table S1. Main characteristics of patients enrolled among trials included in the study

Study (year)	Arms	Sample Size	Age (year)	Female (%)	QTc (ms)	HR (bpm)	LVEF (%)
Priori et al., 1994	P	28	17.3 ± 5.6	53.3	75	519 ± 41	NR
	C	15	17 ± 4	53.3	53	418 ± 22	NR
	S	18	17 ± 4.5	77.7	NR	NR	NR
	A	10	17 ± 8	80	NR	NR	NR
Nakayama et al., 1998	P	8	27 ± 17	50	509 ± 27	NR	NR
	C	10	23 ± 6	40	397 ± 26	NR	NR
	S	6	19.5 ± 7.48	50	512 ± 13	NR	NR
	A	2	29.5 ± 6.5	50	503 ± 3	NR	NR
Savoye et al., 2003	P	10	35.7 ± 12	70	454 ± 37	NR	69 ± 7
	C	14	33.6 ± 9	50	363 ± 29	NR	70 ± 8
	S	NR	NR	NR	NR	NR	NR
	A	NR	NR	NR	NR	NR	NR
Leren et al., 2015	P	192	36 ± 16	61	467 ± 39	64 ± 12	61 ± 5
	C	60	37 ± 10	53	389 ± 25	65 ± 10	61 ± 5
	S	69	32 ± 16	74	486 ± 47	62 ± 11	61 ± 6
	A	123	38 ± 16	54	456 ± 29	65 ± 12	60 ± 5
Haugaa et al., 2009	P	73	NR	NR	NR	NR	NR
	C	20	34 ± 11	55	390 ± 20	NR	67 ± 3
	S	42	32 ± 16	79	497 ± 42	NR	64 ± 6
	A	31	41 ± 14	68	460 ± 30	NR	64 ± 6
Haugaa et al., 2010	P	101	37 ± 16	70	NR	65 ± 13	64 ± 6
	C	35	34 ± 10	57	NR	69 ± 10	64 ± 5
	S	48	32 ± 16	81	495 ± 50	64 ± 13	64 ± 5
	A	53	41 ± 14	60	460 ± 30	67 ± 13	64 ± 6
Haugaa et al., 2013	P	216	23 ± 17	59	478 ± 50	70 ± 17	NR
	C	NR	NR	NR	NR	NR	NR

Hummel et al., 2013	S	91	24 ± 16	66	498 ± 60	65 ± 15	NR
	A	125	23 ± 17	50	464 ± 36	73 ± 17	NR
	P	12	35.7 ± 7.3	75	475 ± 50	64 ± 11	60 ± 6
	C	12	35.3 ± 6.2	58	397 ± 23	68 ± 11	56 ± 5
ter Bekke et al., 2015	S	NR	NR	NR	NR	NR	NR
	A	NR	NR	NR	NR	NR	NR
	P	244	38 ± 16	67	465 ± 50	NR	63 ± 5
	C	76	37 ± 12	67	391 ± 27	NR	63 ± 7
Robyns et al., 2017	S	97	37 ± 17	73	488 ± 50	NR	NR
	A	147	39 ± 16	63	450 ± 44	NR	NR
	P	69	34 ± 17	54	447 ± 56	NR	NR
	C	55	35 ± 16	44	376 ± 27	NR	NR
Vink et al., 2018	S	19	NR	NR	NR	NR	NR
	A	50	NR	NR	NR	NR	NR
	P	41	NR	37	431 ± 30	NR	NR
	C	41	NR	37	387 ± 17	NR	NR
Charisopoulou et al., 2019	S	5	NR	NR	NR	NR	NR
	A	36	NR	NR	NR	NR	NR
	P	47	45 ± 15	53	477.0 ± 40.0	86.3 ± 12.6	71.0 ± 4.11
	C	35	47 ± 13	54	405.7 ± 17.9	85.3 ± 11.5	73.3 ± 5.51
	S	20	NR	NR	495.9 ± 39.2	87.3 ± 13.0	NR
	A	27	NR	NR	468.3 ± 16.3	92.7 ± 16.5	NR

Abbreviations: P: Patients with LQTS; C: Control; S: Symptomatic; A: Asymptomatic; NR: Not rep

Table S2. Diagnostic accuracy of echocardiographic parameters in predicting CE in LQTS

	Sensitivity	Specificity	PPV	NPV	Accuracy	DOR
CD≥430ms	71 [40-94]	84 [67-94]	75 [62 – 84]	87 [72 – 92]	83 [73 – 90]	>19.5
EMW -59ms	82 [66-93]	56 [41-70]	56 [51 – 61]	83 [77 – 88]	67 [61 – 71]	>7.47
QTc≥460ms	53 [37-75]	73 [60-82]	59 [53 – 63]	75 [70 – 78]	68 [62 – 71]	>4.14

CD: Contraction duration; EMW: electromechanical window; QTc: QT corrected.