Supplementary material

Supplemental Table 2. Search strategies

Supplemental Table 3. Risk of bias table

Supplemental Table 4. Characteristics of included studies

Supplemental Table 5. Result of multiple linear regression analysis coefficients for logarithmic-

transformed prevalence of thromboembolism

Supplemental Table 6. Comparison of clinical characteristics between patients with and without TE

Supplemental Table 7. Comparison of clinical characteristics between adult patients with and without TE

Supplemental Figure 1. Summaries of forest plots of the mean prevalence of arrhythmia and LVEF in patients with LVNC

Supplemental Figure 2. Funnel plot of the assessment of publication bias

Supplemental Figure 3. Forest plot of the mean prevalence of thromboembolism in the patients with LVNC according to subgroup analysis

Supplemental Figure 4. Forest plot of the mean incidence of thromboembolism in the patients with LVNC according to subgroup analysis

Supplemental Figure 5. Forest plot of the mean prevalence and incidence of thromboembolism in the patients with LVNC excluding studies in which antithrombotic treatment in more than 30% of patient's population

Supplemental Figure 6. Forest plot of the mean mortality rates in the patients with LVNC according to subgroup analysis

Supplemental Figure 7. Forest plot of the mean mortality and heart plantation rates in the patients with LVNC according to subgroup analysis

References.

Supplemental Table 2. Search strategies

Search strategies using MEDLINE (Supplemental Table 1a), Embase (Supplemental Table 1b), and Cochrane Cochrane Central Register of Controlled Trial (Supplemental Table 1c) are shown below.

No.	Query	Hit retrieved
	(('isolated noncompaction of the ventricular myocardium'/exp) OR hypertrabeculation OR LVNC OR (left ventricular trabeculation) OR (left ventricular noncompaction) OR (left ventricular non-compaction) OR (noncompacted cardiomyopathy) OR (noncompacted myocardium) OR (noncompaction cardiomyopathy) OR (noncompaction myocardium))	6,800
S 6	((non-compacted cardiomyopathy) OR (non-compacted myocardium) OR (non- compaction cardiomyopathy) OR (non-compaction myocardium) OR (noncompaction of left ventricular myocardium) OR (spongy myocardium))	2,770
S 8	(warfarin OR anticoagulants OR aspirin OR apixaban OR edoxaban OR rivaroxaban OR (antithrombotic therapy) OR dabigatran)	433,156
S 9	(thromboembolism OR thrombosis OR embolism)	1,034,463
S10	S6 OR S5	7227

Supplemental Table 2a. Search strategy using MEDLINE

S 11	S10 AND S8	219
S12	S10 AND S9	738
S13	S12 OR S11	833
S14	(S12 OR S11) and (subt.exact("human"))	744

Supplemental Table 2b. Search strategy using Embase

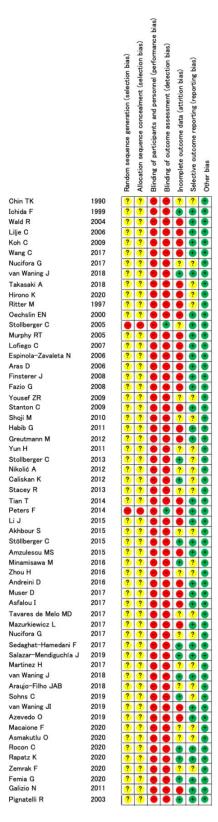
No.	Query	Hit retrieved
S 1	(('isolated noncompaction of the ventricular myocardium'/exp) OR hypertrabeculation OR LVNC OR (left ventricular trabeculation) OR (left ventricular noncompaction) OR (left ventricular non-compaction) OR (noncompacted cardiomyopathy) OR (noncompacted myocardium) OR (noncompaction cardiomyopathy) OR (noncompaction myocardium)) and (human(yes))	3,754
S 3	((non-compacted cardiomyopathy) OR (non-compacted myocardium) OR (non- compaction cardiomyopathy) OR (non-compaction myocardium) OR (noncompaction of left ventricular myocardium) OR (spongy myocardium)) and (human(yes))	1,842
84	('warfarin'/exp OR 'anticoagulants'/exp OR 'aspirin'/exp OR apixaban OR edoxaban OR rivaroxaban OR (antithrombotic therapy) OR 'dabigatran'/exp) and (human(yes))	36,963
\$5	(warfarin OR anticoagulants OR aspirin OR apixaban OR edoxaban OR rivaroxaban OR (antithrombotic therapy) OR dabigatran) and (human(yes))	208,200
S 6	('thromboembolism'/exp OR 'thrombosis'/exp OR 'embolism'/exp) and (human(yes))	1
S 7	(thromboembolism OR thrombosis OR embolism) and (human(yes))	509,254
S 8	\$3 OR \$1	3,889
S9	\$8 AND \$5	184
S 10	\$8 AND \$7	650
S 11	\$10 OR \$9	731
S15	(S10 OR S9) and (rtype.exact("Article") AND subt.exact("human"))	339

Supplemental Table 2c. Search strategy using Cochrane Central Register of Controlled Trial

No.	Query	Hit retrieved
#1	MeSH descriptor: [Isolated Noncompaction of the Ventricular Myocardium]	0
	explode all trees	
#2	hypertrabeculation	0
#3	LVNC	1
#4	left ventricular trabeculation	3
#5	left ventricular noncompaction	6
#6	left ventricular non-compaction	5
#7	noncompacted cardiomyopathy	2
#8	noncompacted myocardium	1
#9	noncompaction cardiomyopathy	7
#10	noncompaction myocardium	4

#11	non-compacted cardiomyopathy	2
#12	non-compacted myocardium	1
#13	non-compaction cardiomyopathy	6
#14	non-compaction myocardium	4
#15	noncompaction of left ventricular myocardium	4
#16	spongy myocardium	0
#17	#1 OR #2 OR #3 OR #4 OR #5 OR #6 OR #7 OR #7 OR #8 OR #9 OR #10 OR #11	448,627
	OR #12 OR #13 OR #14 OR 15 OR 16	
#18	MeSH descriptor: [Warfarin] explode all trees	1,700
#19	MeSH descriptor: [Anticoagulants] explode all trees	4,663
#20	MeSH descriptor: [Aspirin] explode all trees	5,935
#21	apixaban	978
#22	MeSH descriptor: [Dabigatran] explode all trees	312
#23	edoxaban	594
#24	rivaroxaban	1,738
#25	antithrombotic therapy	2,119
#26	#18 OR #19 OR #20 OR #21 OR #22 OR #23 OR #24 OR #25	14,025
#27	MeSH descriptor: [Thromboembolism] explode all trees	2,112
#28	MeSH descriptor: [Thrombosis] explode all trees	4,841
#29	MeSH descriptor: [Embolism] explode all trees	1,427
#30	#27 OR #28 OR #29	7,272
#31	#17 AND #26	1
#32	#17 AND #30	0
#33	#31 OR #32	1

Supplemental Table 3. Risk of bias table



According to the Cochrane Hand book for Systematic Reviews of Interventions, version 5.1.0. +: low risk of bias; -: high risk of bias; ?: unclear risk of bias.

Follow-up Echo/MRI LVEF (%) AF VT Anticoagulant Antiplatelet An Author Year Patients Age (yeras) Male Diagnostic criteria (%) (%) (n) (%)period (years) Children Chin[1] 1990 8.9 (0.9-22.5) 63.0 N/A Chin Echo N/A 0 62.5 N/A N/A 8 6.0 (0-17.0) Ichida[2] 1999 27 5.0 (0-15.0) 55.6 Ichida Echo 61.5 ± 11.6 3.7 N/A 0 N/A Pignatelli[3] 2003 36 0.3 (0-17) 55.6 3.2 (0.5-12.0) Jenni Echo 25 (18.0-32.0) 0 2.8 N/A N/A Wald[4] 2004 22 3.9 (0-16.0) 40.9 3.0 (0.1-16) Jenni Echo 34.0 (6.0-67) 0 9.1 N/A N/A Lilje[5] 2006 66 4.0 (0-21.0) 48.5 1.0 (0-4.3) Chin, Jenni Echo N/A N/A N/A N/A N/A Koh[6] 2009 10 2.0 (0-12.0) 70.0 2.0 (0.2-3.0) Echo N/A N/A N/A N/A N/A Jenni 53.7 4.6 (0-22.0) 2017 205 3.6 (0-15.0) Echo 49.9 ± 2.1 N/A N/A Wang[7] Jenni N/A N/A Nucifora[8] 2017 12 15.0 ± 3.0 75.0 N/A Petersen, Jacquire MRI 64.0 ± 8.0 N/A 8.0 N/A N/A van Waning[9] 2018 52 7.0 (0-14.0) 52.0 5.0 (1.5-9.4) Jenni, Petersen UCG/MRI N/A 10.0 6.0 N/A N/A Takasaki[10] 2018 82 0.8 (0-35.0) 56.1 4.8 (0-22.0) Paterick Echo 42.4 ± 3.5 N/A N/A N/A N/A Hirono[11] 2020 206 0.3 (0-16.0) 57.8 2.0 (0-33.0) Paterick Echo 47.1 ± 19.1 0 11.2 N/A N/A Adults Ritter[12] 1997 17 41.6 (18.0-71.0) 82.3 2.5 ± 2.3 Stollberger Echo N/A 29.4 11.8 N/A N/A 74.0 Oechslin[13] 2000 34 42.0 ± 17.0 3.7 ± 3.3 Echo 33.0 ± 13.0 26.0 41.0 N/A N/A Jenni Stollberger[14] 2005 62 53.0 86.0 N/A Stollberger Echo FS 25.1 8.1 N/A N/A yes 2.7 (0.5-14.9) Murphy[15] 2005 45 37.0 ± 17.0 62.2 Chin, Jenni Echo FS 21.0 ± 9.0 6.5 22.0 N/A yes Lofiego[16] 2007 65 42.0 ± 17.0 N/A 3.8 (0.5-16.1) Jenni Echo 31.0 ± 11.0 1.5 6.0 yes N/A 47.2 Espinola-Zavaleta[17] 2006 53 43.2 ± 14.8 0.6 ± 0.4 Jenni Echo 39.0 ± 18.5 5.7 7.5 N/A N/A Aras[18] 2006 67 41.0 ± 18.0 66.0 3.0 ± 1.1 Jenni Echo 43.5 ± 14.4 12.0 36.0 N/A yes Finsterer[19] 2008 104 14.0-94.0 71.0 12.0 Stollberger Echo N/A N/A N/A N/A N/A Stollberger Fazio[20] 2008 238 41.5 (1.0-92.0) 58.0 7.3 (1.0-12.0) Echo 24.6 N/A N/A N/A N/A Yousef[21] 2009 42 48.7 ± 2.3 59.5 N/A Jenni Echo 41.1 ± 14.8 28.6N/A N/A N/A Stanton[22] 2009 30 39.0 ± 19.5 60.0 2.5 ± 1.2 Echo 41.0 ± 15.0 10.0 N/A Jenni 16.6 yes 187 65.8 Shoji[23] 2010 41.3 ± 16.8 N/A Jenni Echo 62.2 4.8 1.1 N/A N/A Habib[24] 2011 105 45.0 ± 17.0 66.0 2.3 ± 1.5 Echo 46.0 ± 18.0 7.0 7.0 N/A Jenni yes Greutmann[25] 2012 132 41.0 ± 17.0 35.0 2.7 (0.1-19.4) Ienni Echo 41.0 ± 18.0 24.0 7.0 yes yes Yun[26] 2011 11 35.0 (15.0-58.0) 81.8 N/A MRI 40.0 (23.0-70.0) 0 0 N/A N/A Petersen Stollberger[27] Stollberger 2013 169 52.9 ± 16.0 70.0 16.0 Echo FS 24.2 ± 11.0 N/A N/A 16.6 N/A 58.3 Nikolić[28] 2012 12 45.0 ± 15.0 N/A Jenni Echo 49.8 ± 8.0 33.3 25.0 N/A N/A Caliskan[29] 2012 84 40.0 (17.0-77.0) 47.6 5.3 (1.5-9.6) Jenni Echo FS 20.8 ± 8.2 9.5 19.0 N/A N/A Stacey[30] 122 57.0 ± 17.5 59.0 N/A MRI 44.0 ± 16.0 4.9 N/A 2013 Jacquier N/A N/A Tian[31] 2014 106 46.0 ± 17.0 78.0 2.9 ± 2.1 Jenni Echo 39.0 ± 14.0 13.0 2.0 N/A yes Peters[32] 2014 55 42.2 ± 11.5 38.2 1.4 ± 0.5 Jenni Echo 29.6 ± 11.8 1.8 1.8 N/A yes

Supplemental Table 4. Characteristics of included studies

nticoagulant/	Hx of	TE
Antiplatelet	TE (%)	events
(%)		(%)
N/A	38.0	N/A
N/A	N/A	0
N/A	2.8	0
N/A	N/A	0
N/A	N/A	13.9
N/A	N/A	0
N/A	N/A	4.9
N/A	0	N/A
N/A	2.0	4.0
N/A	3.7	N/A
N/A	2.4	N/A
N/A	5.9	24.0
N/A	N/A	24.0
6.5	1.6	10.0
60.0	N/A	4.0
52.3	N/A	5.0
N/A	N/A	5.7
28.3	6.0	9.0
N/A	9.0	15.0
N/A	14.0	2.1
N/A	14.0	N/A
30.0	0	0
N/A	0.5	N/A
29.2	4.0	8.5
36.0	3.0	4.0
N/A	9.1	N/A
N/A	15.0	N/A
N/A	0	N/A
N/A	10.7	N/A
N/A	8.2	N/A
9.0	6.0	4.0
29.1	16.3	1.8

Author	Year	Patients (n)	Age (yeras)	Male (%)	Follow-up period (years)	Diagnostic criteria	Echo/MRI	LVEF (%)	AF (%)	VT (%)	Anticoagulant	Antiplatelet	Anticoagulant/ Antiplatelet (%)	Hx of TE (%)	TE events (%)
Li[33]	2015	20	42.5 (36.3–52.5)	60.0	1.1±0.5	Stollberger	Echo	32.0 (22.0-40.0)	5.0	5.0	yes	yes	60.0	N/A	0
Akhbour[34]	2015	24	42.7 ± 13.1	66.7	N/A	Petersen	MRI	37.8 ± 13.9	16.7	20.8	N/A	N/A	N/A	4.2	N/A
Stöllberger[35]	2015	232	52.0 ± 17.0	69.0	6.0 ± 5.2	Stollberger	Echo	FS 25.0 ± 11.0	16.0	N/A	N/A	N/A	N/A	N/A	4.3
Amzulescu[36]	2015	59	52.0 ± 13.0	57.6	3.4 (1.5 - 6.3)	Petersen	MRI	24.1 ± 8.3	3.0	N/A	yes	yes	22.0	12.0	6.8
Minamisawa[37]	2016	23	54.9 ± 9.1	69.6	5.1	Jenni	Echo	27.4 ± 8.5	43.5	N/A	yes	N/A	43.4	8.7	N/A
Zhou[38]	2016	31	40.8 ± 4.8	61.3	N/A	Jenni	Echo	42.0 ± 5.9	N/A	N/A	N/A	N/A	N/A	6.5	N/A
Andreini[39]	2016	113	44.0 ± 17.0	62.0	4 ± 2	Jenni, Petersen	Echo /MRI	42.8 ± 16.2	N/A	N/A	yes	yes	5.0	N/A	4.4
Muser[40]	2017	9	42.0 ± 15.0	55.6	4.0 (1.0 - 11.0)	Jenni	Echo	40.0 ± 12.1	N/A	44.4	yes	N/A	66.6	N/A	11.1
Asfalou[41]	2017	23	47.0 ± 13.0	65.2	2.0	Jenni	Echo	27.0 ± 8.0	21.7	16.6	yes	N/A	28.6	N/A	4.3
Tavares de Melo[42]	2017	30	41.0 ± 12.0	53.0	N/A	Petersen	MRI	39.0 ± 14.0	N/A	33.0	yes	N/A	77.0	13.0	N/A
Mazurkiewicz[43]	2017	127	33.1 ± 8.8	61.4	2.2 ± 1.0	Grothoff	MRI	27.7 ± 6.8	11.8	N/A	N/A	N/A	N/A	8.7	1.6
Nucifora[8]	2017	20	35.0 ± 7.0	75.0	N/A	Petersen, Jacquire	MRI	55.0 ± 14.0	N/A	10.0	N/A	N/A	N/A	10.0	N/A
Sedaghat-	2017	95	41.0 ± 14.4	70.6	5.1	Jenni, Stollberger	Echo	38.0 ± 15.3	29.4	35.3	N/A	N/A	N/A	N/A	10.3
Hamedani[44]															
Salazar-	2019	75	50.4 ± 14.8	68.0	5.0 (2.4 - 6.7)	Jenni	Echo	32.0 (29.0 - 34.0)	38.7	4.0	yes	yes	53.3	1.3	5.3
Mendiguchía[45]															
Martinez[46]	2017	13	N/A	N/A	N/A	Jenni	Echo	N/A	N/A	N/A	N/A	yes	N/A	8.0	N/A
van Waning[9]	2018	275	45.0 (33.0 - 56.0)	54.0	2.1 (0.3 - 4.8)	Jenni, Petersen	Echo /MRI	N/A	16.0	6.0	N/A	N/A	N/A	3.0	13.0
Araujo-Filho[47]	2018	36	41.0 ± 13.0	61.0	N/A	Petersen	MRI	42.0 ± 6.0	N/A	30.6	yes	N/A	55.6	8.3	N/A
Sohns[48]	2019	18	43.5 ± 18.0	66.7	5.2 ± 3.5	Jenni	Echo	35.0 ± 15.0	11.1	66.7	yes	N/A	77.8	27.8	N/A
van Waning[49]	2019	216	38.0 (23.0 - 52.0)	54.0	3.7 (0.8 - 7.8)	Jenni, Petersen, Grothoff	Echo /MRI	N/A	11.6	11.6	N/A	N/A	N/A	N/A	4.6
Azevedo[50]	2019	78	47.0 ± 17.0	62.8	4.4 ± 2.8	Jenni, Stollberger, Chin, Petersen Jacquier	Echo /MRI	47.2 ± 15.2	13.5	21.6	N/A	N/A	N/A	5.1	11.5
Macaione[51]	2020	27	47.6 ± 22.6	77.8	N/A	Jenni, Petersen, Grothoff	Echo /MRI	48.5 ± 17.6	N/A	11.1	N/A	N/A	N/A	3.7	N/A
Asmakutlu[52]	2020	42	32.4 (18.0 - 63.0)	52.3	N/A	Stollberger, Petersen	Echo /MRI	50.3 (22.0 - 67.0)	4.7	11.9	N/A	N/A	N/A	14.2	N/A
Rocon[53]	2020	108	38.3 ± 15.5	48.1	5.8 ± 3.9	Chin, Jenni, Stollberger, Petersen	Echo /MRI	45.3 ± 14.6	N/A	61.7	N/A	N/A	N/A	N/A	17.0
Rapatz[54]	2020	113	57.0 (45.0 - 69.0)	76.0	6.1 ± 5.3	Stollberger	Echo	FS 20.0 (15.0 – 30.0)	20.0	N/A	N/A	N/A	N/A	12.0	6.0
Zemrak[55]	2020	142	N/A	N/A	N/A	Petersen	MRI	N/A	N/A	N/A	N/A	N/A	N/A	1.3	N/A
Femia[56]	2020	98	47.0 (31.0 - 65.0)	56.7	6.5 (5.5 - 7.5)	Petersen	MRI	81.2 ± 7.2	1.0	16.3	N/A	N/A	N/A	7.1	4.1
Galizio[57]	2011	80	41.0 ± 17.0	66.3	1.1 (0.5 – 2.1)	Chin, Jenni, Stollberger, Petersen	Echo /MRI	38.6 ± 11.4	N/A	18.8	N/A	N/A	N/A	N/A	3.75

Echo, echocardiography; MRI magnetic resonance imaging; LVEF, left ventricular ejection fraction; AF; atrial fibrillation; VT, ventricular tachycardia; Hx, history; TE, thromboembolism; N/A, not applicable; FS, fractional shortening.

	Unstandardized coefficients				Standardized coefficients	_		
Model	β	Standard error	Lower CI	Upper CI	β	Т	P value	Variance Inflation Factor
Constant	3.0518441	0.672898	1.5296435	4.5740446	0	4.54	0.0014	
Follow-up period (y)	0.386106	0.164966	0.0129269	0.7592851	0.730529	2.34	0.0440	1.8558142
LVEF (%)	-0.046194	0.017301	-0.085332	-0.007057	-0.83592	-2.67	0.0256	1.8671364
AF (%)	-0.053726	0.018199	-0.094894	-0.012558	-0.90918	-2.95	0.0162	1.8067165

Supplemental Table 5. Result of multiple linear regression analysis coefficients for logarithmic-transformed prevalence of thromboembolism

CI, confidence interval; LVEF, left ventricular ejection fraction; AF; atrial fibrillation.

	TE negative (n	TE positive (n	P value
	= 281)	= 6)	
Male	142	4	1.0000
FH	72	2	1.0000
CHD	37	0	1.0000
HF at diagnosis	154	5	0.2303
LVEF (%)	46.9 ± 18.9	30.3 ± 19.9	0.0648
LVEF<40%	89	4	0.0608
AF	2	0	1.0000
VT	29	1	0.4876
Anticoagulant	3	1	0.0298
Antiplatelet	7	0	1.0000

Supplemental Table 6. Comparison of clinical characteristics between pediatric patients with and without TE

FX, family history; CHD, congenital heart disease; HF, heart failure; LVEF, left ventricular ejection fraction; AF, atrial fibrillation; VT, ventricular tachycardia

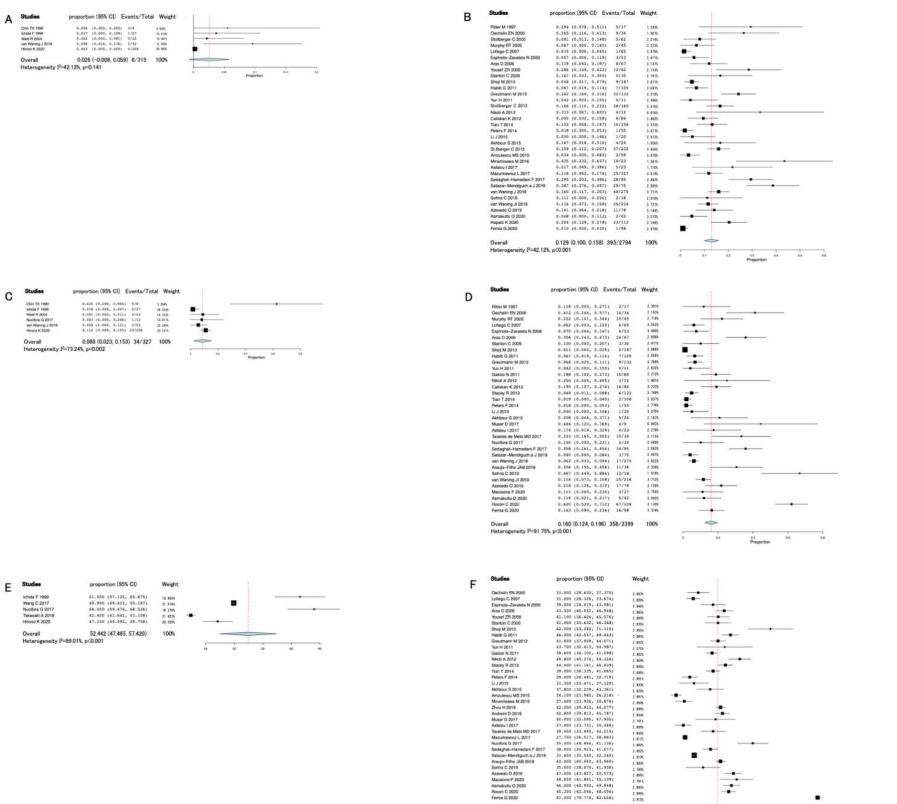
	TE negative (n	TE positive (n	P value
	= 21)	= 5)	
Male	14	5	0.2782
FH	2	1	0.3333
CHD	0	0	1.0000
HF at diagnosis	7	3	0.3402
LVEF (%)	39.8 ± 13.8	40	1.0000
LVEF<40%	3	0	1.0000
AF	4	1	1.0000
VT	9	1	0.6169

Supplemental Table 7. Comparison of clinical characteristics between adult patients with and without TE

FX, family history; CHD, congenital heart disease; HF, heart failure; LVEF, left ventricular ejection fraction; AF, atrial fibrillation; VT, ventricular tachycardia

Supplemental Figure 1. Summaries of forest plots of the mean prevalence of arrhythmia and LVEF in patients with LVNC

Mean prevalence of atrial fibrillation (AF) in pediatric (A) and adult patients (B). Mean prevalence of ventricular tachycardia (VT) in pediatric (C) and adult patients (D). Mean LVEF in pediatric (E) and adult patients (F).



Overall

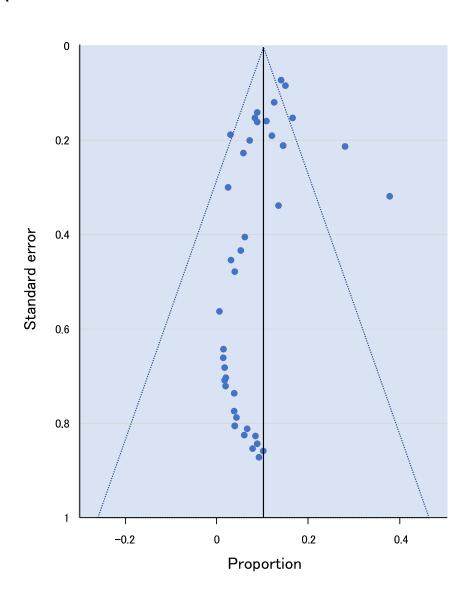
40.852 (35.585, 46.118) neity P=99.26% p<0.001 100%

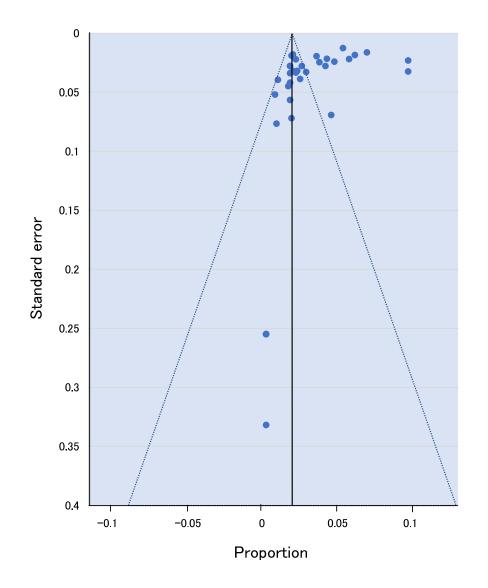
В

Supplemental Figure 2. Funnel plot of the assessment of publication bias for studies

Funnel plot of the assessment of publication bias for studies regarding the prevalence (A) and incidence (B) of TE in the patients with LVNC

Α





Supplemental Figure 3. Forest plot of the mean prevalence of thromboembolism in the patients with LVNC according to subgroup analysis

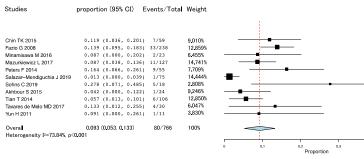
Mean prevalence of thromboembolism in studies with average LVEF <40% (A) and >40% (B).

Mean prevalence of thromboembolism in studies with mean follow-up period <4 years (C) and >4 years (D).

Mean prevalence of thromboembolism in studies with mean prevalence of AF <10% (E) and prevalence of AF >10% (F).

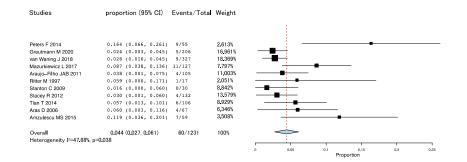
CI, confidence interval.



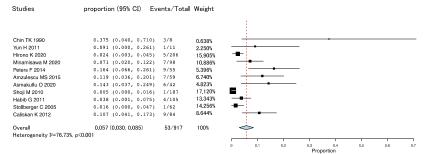


В			
Studies	proportion (95% CI) Events/Tota	al Weight	
Stanton C 2009	0.016 (0.000, 0.060) 0/30	6.969%	_
Greutmann M 2012	0.030 (0.001, 0.060) 4/132	9.763%	_
Yousef ZR 2009	0.143 (0.037, 0.249) 6/42	2.063%	
Zhou H 2016	0.065 (0.000, 0.151) 2/31	2.880%	
Araujo-Filho JAB 2018	0.083 (0.000, 0.174) 3/36	2.687%	
Takasaki A 2018	0.037 (0.000, 0.077) 3/82	7.584%	
Aras D 2006	0.060 (0.003, 0.116) 4/67	5.269%	e
Stacey R 2013	0.082 (0.033, 0.131) 10/122	6.313%	_
Habib G 2011	0.038 (0.001, 0.075) 4/105	8.308%	
Hirono K 2020	0.024 (0.003, 0.045) 5/206	11.476%	
Azevedo O 2019	0.051 (0.002, 0.100) 4/78	6.274%	
Macaione F 2020	0.037 (0.000, 0.108) 1/27	3.870%	
Nikolic A 2012	0.038 (0.000, 0.143) 0/12	2.106%	
Asmakutlu O 2020	0.143 (0.037, 0.249) 6/42	2.063%	
Nucifora G 2017	0.062 (0.000, 0.146) 2/32	3.023%	
Shoji M 2010	0.005 (0.000, 0.016) 1/187	13.360%	
Femia G 2020	0.071 (0.020, 0.122) 7/98	5.991%	
Overall	0.043 (0.027, 0.060) 62/1329	100%	\sim
Heterogeneity I2=57.66%, p	=0.002		· · · · · · · · · · · · · · · · · · ·
			0 0.05 0.1 0.15 0.2
			Proportion

С



Е



D

Studies	proportion (95% CI)	Events/Total	Weight					
					1.0			
Azevedo O 2019	0.051 (0.002, 0.100)	4/78	10.322%		-			
Takasaki A 2018	0.037 (0.000, 0.077)	3/82	11.013%		-			
Salazar-Mendiguchia J 2019	0.013 (0.000, 0.039)	1/75	12.074%	-	1.1			
Minamisawa M 2016	0.087 (0.000, 0.202)	2/23	5.388%		+			
Sohns C 2019	0.278 (0.071, 0.485)	5/18	2.343%				•	
Caliskan K 2012	0.107 (0.041, 0.173)	9/84	8.846%			-		
Rapatz K 2020	0.124 (0.063, 0.185)	14/113	9.307%					
Femia G 2020	0.071 (0.020, 0.122)	7/98	10.147%					
Fazio G 2008	0.139 (0.095, 0.183)	33/238	10.746%					
Finsterer J 2008	0.087 (0.033, 0.141)	9/104	9.885%					
Stollberger C 1 2013	0.148 (0.094, 0.201)	25/169	9.929%					
Overa	0.089 (0.054, 0.124)	112/1080	100%	~	\sim			
Heterogeneity I2=78.90%, p<0	.001							
				0	0.1	0.2	0.3	0.4

F

Studies proportion (95% CI) Events/Total Weight

Sohns C 2019	0.278 (0.071, 0.485)	5/18	1.015%		
Mazurkiewicz L 2017	0.087 (0.038, 0.136)	11/127	7.506%		_
Aras D 2006	0.060 (0.003, 0.116)	4/67	6.638%		
Tian T 2014	0.057 (0.013, 0.101)	6/106	8.092%		
Azevedo O 2019	0.051 (0.002, 0.100)	4/78	7.502%		
van Waning J 2018	0.028 (0.010, 0.045)	9/327	11,186%	-8-	
Stanton C 2009	0.016 (0.000, 0.060)	0/30	8.049%		
Stollberger C 1 2013	0.148 (0.094, 0.201)	25/169	6.984%		
Akhbour S 2015	0.042 (0.000, 0.122)	1/24	4.597%		-
Rapatz K 2020	0.124 (0.063, 0.185)	14/113	6.227%		-
Greutmann M 2012	0.030 (0.001, 0.060)	4/132	9.920%		
Yousef ZR 2009	0.143 (0.037, 0.249)	6/42	3.135%		•
Ritter M 1997	0.059 (0.000, 0.171)	1/17	2.885%		
Nikolic A 2012	0.038 (0.000, 0.143)	0/12	3.193%		
Salazar-Mendiguchia J 2019	0.013 (0.000, 0.039)	1/75	10.312%		
Minamisawa M 2016	0.087 (0.000, 0.202)	2/23	2.759%		
Overall	0.060 (0.038, 0.082)	93/1360	100%	-	
Heterogeneity I2=66.28%, p<0.	001				

0.1 0.2 0.3 0.4 Proportion

0.2 Proportion

Supplemental Figure 4. Forest plot of the mean incidence of thromboembolism in the patients with LVNC according to subgroup analysis

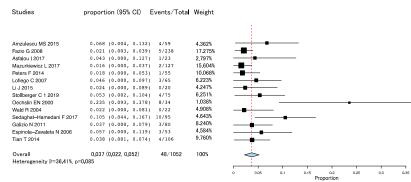
Mean incidence of thromboembolism in studies with average LVEF <40% (A) and >40% (B).

Mean incidence of thromboembolism in studies with mean follow-up period <4 years (C) and >4 years (D).

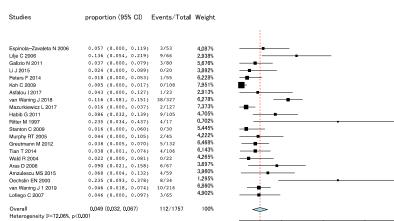
Mean incidence of thromboembolism in studies with mean prevalence of AF <10% (E) and prevalence of AF >10% (F).

CI, confidence interval.









100%

	_		
•			
•		•	
-	 		
_		•	

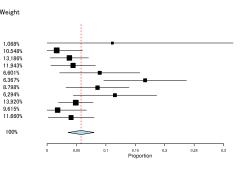
0.3

0.2 Proportion

ser D 2017	0.111	(0.000,	0.316)	1/9	
nton C 2009	0.016	(0.000,	0.060)	0/30	
utmann M 2012	0.038	(0.005,	0.070)	5/132	
freini D 2016	0.044	(0.006,	0.082)	5/113	
s D 2006	0.090	(0.021,	0.158)	6/67	
on C 2020	0.167	(0.096,	0.237)	18/108	

Ichida F 1999 Femia G 2020 Overall	0.041	(0.000, (0.002,	0.080)	0/27 4/98 67/9		9.615% 11.660 100%
			,			
Ichida F 1999	0.018	(0.000.	0.067)	0/27		9.615%
Wang C 2017	0.049	(0.019,	0.078)	10/205		13.920
Azevedo O 2019	0.115	(0.044,	0.186)	9/78		6.294%
Habib G 2011	0.086	(0.032,	0.139)	9/105		8.798%
Rocon C 2020	0.167	(0.096,	0.237)	18/108		6.367%
Aras D 2006	0.090	(0.021,	0.158)	6/67		6.601%
Andreini D 2016	0.044	(0.006,	0.082)	5/113		11.943
Greutmann M 2012	0.038	(0.005,	0.070)	5/132		13,186
Stanton C 2009	0.016	(0.000,	0.060)	0/30	•	10.548
Muser D 2017	0.111	(0.000,	0.316)	1/9	•	1.068%
	Stanton C 2009 Greutmann M 2012 Andreini D 2016 Aras D 2006 Rocon C 2020 Habib G 2011	Stanton C 2009 0.016 Greutmann M 2012 0.038 Andreini D 2016 0.044 Aras D 2006 0.090 Rocon C 2020 0.167 Habib G 2011 0.086	Stanton C 2009 0.016 (0.000, Greutman M 2012 0.038 (0.063, Andreini D 2016 0.044 (0.006, Aras D 2006 0.090 (0.021, Rocon C 2020 0.167 (0.086, G 0.032, Abbi G 2011 0.086 (0.032, G 0.032, G 0.032,	Stanton C 2009 0.016 (0.000, 0.060) Greutmann M 2012 0.038 (0.005, 0.070) Andreinin D 2016 0.044 (0.006, 0.082) Aras D 2006 0.090 (0.021, 0.158) Rocon C 2020 0.167 (0.096, 0.237) Habib G 2011 0.086 (0.032, 0.138)	Stanton C 2009 0.016 (0.000, 0.060) 0/30 Greutmann M 2012 0.038 (0.055, 0.070) 5/132 Andreini D 2016 0.044 (0.060, 0.082) 5/131 Aras D 2006 0.090 (0.021, 0.158) 6/67 Rocon C 2020 0.167 (0.096, 0.237) 18/108 Habib G 2011 0.066 (0.032, 0.139) 9/105	Stanton C 2009 0.016 (0.000, 0.060) 0/30 . Greutmann M 2012 0.038 (0.005, 0.070) 5/132 Andreini D 2016 0.044 (0.060, 0.082) 5/133 Aras D 2006 0.090 (0.021, 0.158) 6/67 Rocon C 2020 0.167 (0.096, 0.0237, 0.139) 9/105

proportion (95% CI) Events/Total Weight

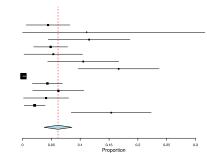


D

В

Studies

Studies	proportion (95% CI)	Events/Total	Weight
Andreini D 2016	0.044 (0.006, 0.082)	5/113	8.848%
Muser D 2017	0.111 (0.000, 0.316)	1/9	1.242%
Azevedo O 2019	0.115 (0.044, 0.186)	9/78	5.747%
Wang C 2017	0.049 (0.019, 0.078)	10/205	9.675%
Salazar-Mendiguchia J 2019	0.053 (0.002, 0.104)	4/75	7.545%
Sedaghat–Hamedani F 2017	0.105 (0.044, 0.167)	10/95	6.523%
Rocon C 2020	0.167 (0.096, 0.237)	18/108	5.796%
Ichida F 1999	0.002 (0.000, 0.006)	0/327	11.250%
Stollberger C 1 2015	0.043 (0.017, 0.069)	10/232	9.981%
Rapatz K 2020	0.062 (0.018, 0.106)	7/113	8.186%
Femia G 2020	0.041 (0.002, 0.080)	4/98	8.721%
Fazio G 2008	0.021 (0.003, 0.039)	5/238	10.613%
Finsterer J 2008	0.154 (0.085, 0.223)	16/104	5.872%
Overal Heterogeneity I2=87.51%, p<0.0	0.062 (0.037, 0.086) 01	99/1795	100%

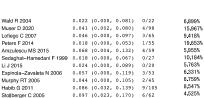


F

Studies	proportion (95% CI) Ev	vents/Total	Weight					
van Waning J 1 2019	0.400 (0.096, 0.704)	4/10	4.783%	-		•		-
Mazurkiewicz L 2017	0.041 (0.002, 0.080)	4/98	11.690%					
Aras D 2006	0.333 (0.000, 0.711)	2/6	3.606%			•		-
Tian T 2014	0.500 (0.010, 0.990)	2/4	2.435%					
Azevedo O 2019	0.444 (0.120, 0.769)	4/9	4.404%			•		
Stollberger C 1 2015	0.400 (0.096, 0.704)	4/10	4,783%	-		•		-
van Waning J 2018	0.105 (0.008, 0.203)	4/38	10.371%					
Stanton C 2009	0.094 (0.016, 0.173)	5/53	10.883%		- :			
Rapatz K 2020	0.571 (0.205, 0.938)	4/7	3.751%	_			•	
Asfalou 2017	0.010 (0.000, 0.028)	1/105	11.916%					
Greutmann M 2012	0.065 (0.003, 0.126)	4/62	11,293%		-			
Oechslin EN 2000	0.500 (0.154, 0.846)	4/8	4.048%					
Ritter M 1997	0.900 (0.637, 1.000)	4/4	5.628%		1			
Sedaghat–Hamedani F 2017	0.400 (0.096, 0.704)	4/10	4.783%	-				-
Salazar-Mendiguchia J 2019	0.900 (0.637, 1.000)	4/4	5.628%					
Overall	0.279 (0.193, 0.364)	54/428	100%		~	>		
Heterogeneity I2=89.95%, p<	(0.001							
				0	0.2	0.4 Prop	0.6 ortion	0.8

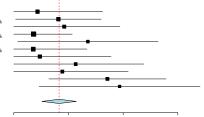
Е Studies

Amzulescu MS 201



proportion (95% CI) Events/Total Weight

Sedaghat–Hamed Li J 2015 Espinola-Zavaleta N 2006 Murphy RT 2005 Habib G 2011 Stollberger C 2005 0.041 (0.026, 0.057) 32/611 Overall Heterogeneity I2=0%, p=0.535



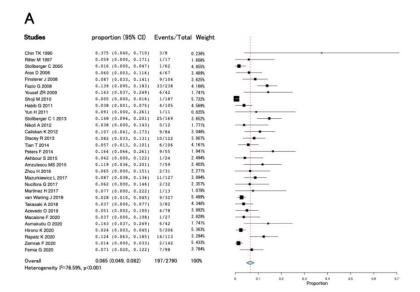
Proportion

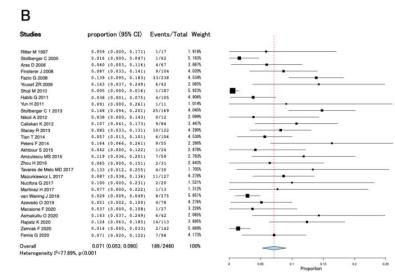
Supplemental Figure 5. Forest plot of the mean prevalence and incidence of thromboembolism in the patients with LVNC excluding studies in which antithrombotic treatment in more than 30% of patient's population

Mean prevalence of thromboembolism in all patients (A) and adult patients (B) with LVNC.

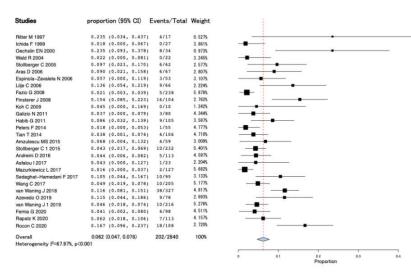
Mean incidence of thromboembolism in the whole aged (C) and adult patients (D) with LVNC.

CI, confidence interval.







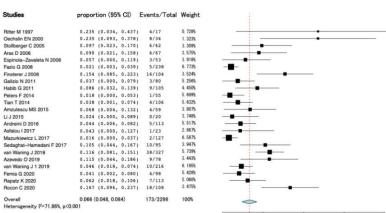


D

Studies

Ritter M 199

Oechslin EN 20 Stollberger C 2 Aras D 2006 Espinola-Zava Fazio G 2008





0.2 Proportion

В

D

Studies

Supplemental Figure 6. Forest plot of the mean mortality rates in the patients with LVNC according to subgroup analysis

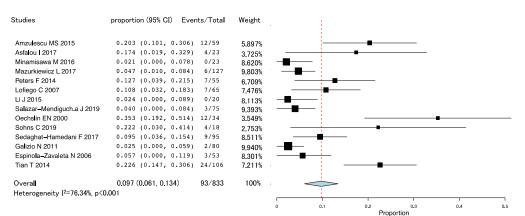
The mean mortality rates in studies with average LVEF $\leq 40\%$ (A) and $\geq 40\%$ (B).

The mean mortality rates in studies with mean follow-up period <4 years (C) and >4 years (D).

The mean mortality rates in studies with mean prevalence of AF <10% (E) and prevalence of AF >10% (F).

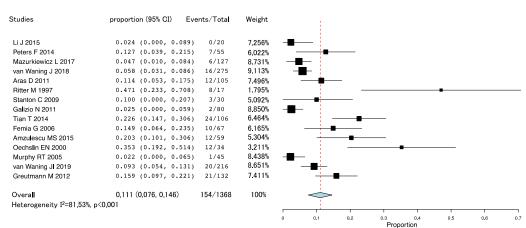
CI, confidence interval.

А



Studies	proportion (95% CI) Events	/Total Weight	
Muser D 2017		/9 4.039%	
Stanton C 2009	0.100 (0.000, 0.207) 3	/30 5.533%	•
Greutmann M 2012	0.159 (0.097, 0.221) 21	/132 9.538%	
Andreini D 2016	0.044 (0.006, 0.082) 5	/113 12.435%	5%
Aras D 2006	0.149 (0.064, 0.235) 10	/67 7.219%	
Stacey R 2013	0.049 (0.011, 0.088) 6	/122 12.382%	2%
Rocon C 2020	0.056 (0.012, 0.099) 6	/108 11.809%)% :
Habib G 2011	0.114 (0.053, 0.175) 12	/105 9.714%	á · · · · · · · · · · · · · · · · · · ·
Azevedo O 2019	0.006 (0.000, 0.024) 0	/78 14.459%	3% -
Femia G 2020	0.031 (0.000, 0.065) 3	/98 12.872%	2%
Overal	0.068 (0.036, 0.099) 6	6/862 100%	
Heterogeneity I ² =78.60%	i. p<0.001		
			0 0.05 0.1 0.15 0.2 Proportion

С

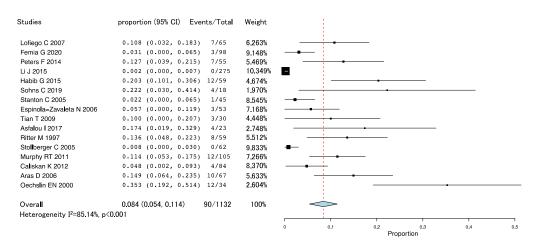


0.033 (0.092, 0.014) 0.022 (0.000, 0.065) 0.093 (0.054, 0.131) 0.159 (0.097, 0.221) 0.111 (0.076, 0.146)	1/45 20/216 21/132 154/1368	3.211% 8.438% 8.651% 7.411% 100%		■				
0.022 (0.000, 0.065) 0.093 (0.054, 0.131) 0.159 (0.097, 0.221) 0.111 (0.076, 0.146)	1/45 20/216 21/132	8.438% 8.651% 7.411%	•	■				
0.022 (0.000, 0.065) 0.093 (0.054, 0.131)	1/45 20/216	8.438% 8.651%	- B					
0.022 (0.000, 0.065)	1/45	8.438%	- B					
		8.438%						
0.555 (0.192, 0.514)	12/34							
0.353 (0.192, 0.514)	12/34	3.211%						
0.203 (0.101, 0.306)	12/59	5.304%		-				
0.149 (0.064, 0.235)	10/67	6.165%		I				
0.226 (0.147, 0.306)	24/106	6.464%	-					
0.025 (0.000, 0.059)	2/80	8.850%	-8-					
0.100 (0.000, 0.207)	3/30	5.092%						
0.471 (0.233, 0.708)	8/17	1.795%				•		
0.114 (0.053, 0.175)	12/105	7.496%	_	_				
0.058 (0.031, 0.086)	16/275	9.113%						
0.047 (0.010, 0.084)	6/127	8.731%	— — —					
0.127 (0.039, 0.215)	7/55	6.022%						
0.024 (0.000, 0.089)	0/20	7.256%						
	0.127 (0.039, 0.215) 0.047 (0.010, 0.084) 0.058 (0.031, 0.086) 0.114 (0.053, 0.175) 0.471 (0.233, 0.708) 0.100 (0.000, 0.207) 0.225 (0.000, 0.059) 0.226 (0.147, 0.306) 0.149 (0.064, 0.235) 0.203 (0.101, 0.306)	$\begin{array}{ccccccc} 0.127 & (0.039, \ 0.215) & 7/55 \\ 0.047 & (0.010, \ 0.084) & 6/127 \\ 0.058 & (0.031, \ 0.086) & 16/275 \\ 0.114 & (0.053, \ 0.175) & 12/105 \\ 0.471 & (0.233, \ 0.708) & 8/17 \\ 0.100 & (0.000, \ 0.207) & 3/30 \\ 0.025 & (0.000, \ 0.059) & 2/80 \\ 0.226 & (0.147, \ 0.306) & 24/106 \\ 0.149 & (0.064, \ 0.235) & 10/67 \\ 0.203 & (0.101, \ 0.306) & 12/59 \\ \end{array}$	$\begin{array}{cccccccccccccccccccccccccccccccccccc$	0.127 (0.039, 0.215) 7/55 6.022% 0.047 (0.010, 0.084) 6/127 8.731% 0.058 (0.031, 0.086) 16/275 9.113% 0.114 (0.053, 0.175) 12/105 7.496% 0.471 (0.233, 0.708) 8/17 1.795% 0.100 (0.000, 0.207) 3/30 5.092% 0.025 (0.000, 0.059) 2/80 8.850% 0.226 (0.147, 0.306) 24/106 6.464% 0.149 (0.064, 0.235) 10/67 6.165% 0.203 (0.101, 0.306) 12/59 5.304%	0.127 (0.039, 0.215) 7/55 6.022% 0.047 (0.010, 0.084) 6/127 8.731% 0.058 (0.031, 0.086) 16/275 9.113% 0.114 (0.053, 0.175) 12/105 7.496% 0.471 (0.233, 0.708) 8/17 1.795% 0.100 (0.000, 0.207) 3/30 5.092% 0.025 (0.000, 0.059) 2/80 8.850% 0.226 (0.147, 0.306) 24/106 6.464% 0.149 (0.064, 0.235) 10/67 6.165% 0.203 (0.101, 0.306) 12/59 5.304%	0.127 (0.039, 0.215) 7/55 6.022% 0.047 (0.010, 0.084) 6/127 8.731% 0.058 (0.031, 0.086) 16/275 9.113% 0.114 (0.053, 0.175) 12/105 7.496% 0.471 (0.233, 0.708) 8/17 1.795% 0.100 (0.000, 0.207) 3/30 5.092% 0.025 (0.000, 0.059) 2/80 8.850% 0.226 (0.147, 0.306) 24/106 6.464% 0.249 (0.644, 0.235) 10/67 6.165% 0.203 (0.101, 0.306) 12/59 5.304%	0.127 (0.039, 0.215) 7/55 6.022% 0.047 (0.010, 0.084) 6/127 8.731% 0.058 (0.031, 0.086) 16/275 9.113% 0.114 (0.053, 0.175) 12/105 7.496% 0.471 (0.233, 0.708) 8/17 1.795% 0.100 (0.000, 0.207) 3/30 5.092% 0.025 (0.000, 0.059) 2/80 8.850% 0.226 (0.147, 0.306) 24/106 6.464% 0.149 (0.064, 0.235) 10/67 6.165% 0.203 (0.101, 0.306) 12/59 5.304%	0.127 (0.039, 0.215) 7/55 6.022% 0.047 (0.010, 0.084) 6/127 8.731% 0.058 (0.031, 0.086) 16/275 9.113% 0.114 (0.053, 0.175) 12/105 7.496% 0.471 (0.233, 0.708) 8/17 1.795% 0.100 (0.000, 0.207) 3/30 5.092% 0.025 (0.000, 0.059) 2/80 8.850% 0.226 (0.147, 0.306) 24/106 6.464% 0.249 (0.644, 0.235) 10/67 6.165% 0.203 (0.101, 0.306) 12/59 5.304%

Weight

proportion (95% CI) Events/Total

Е



F

Studies	proportion (95% CI) Eve	nts/Total	Weight					
Minamisawa M 2016	0.021 (0.000, 0.078)	0/23	8.063%					
Azevedo O 2019	0.006 (0.000, 0.024)	0/78	9.415%	-				
Peters F 2014	0.226 (0.147, 0.306)	24/106	7.014%					
Mazurkiewicz L 2017	0.047 (0.010, 0.084)	6/127	8.884%					
Rapatz K 2020	0.336 (0.249, 0.423)	38/113	6.666%		1.1	_		
van Waning JI 2019	0.093 (0.054, 0.131)	20/216	8.821%		-			
Sedaghat–Hamedani F 2017	0.095 (0.036, 0.154)	9/95	7.985%			_		
Espinola-Zavaleta N 2013	0.003 (0.000, 0.011)	0/169	9.545%					
Salazar-Mendiguchia J 2019	0.040 (0.000, 0.084)	3/75	8.606%					
Greutmann M 2012	0.159 (0.097, 0.221)	21/132	7.825%					
Sollberger C 2015	0.293 (0.235, 0.352)	68/232	7.999%				•	_
van Waning J 2018	0.058 (0.031, 0.086)	16/275	9.176%		•			
Overal	0.105 (0.063, 0.146)	205/1641	100%		\sim	-		
Heterogeneity I ² =95.17%, p<0								
installagenergy i boilting poi				0	0.1	0.2	0.3	04
				-		Proportion		

В

D

Supplemental Figure 7. Forest plot of the mean mortality and heart plantation rates in the patients with LVNC according to subgroup analysis

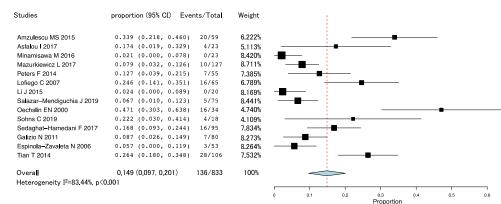
The mean mortality and heart plantation rates in studies with average LVEF <40% (A) and >40% (B).

The mean mortality and heart plantation rates in studies with mean follow-up period <4 years (C) and >4 years (D).

The mean mortality and heart plantation rates in studies with mean prevalence of AF <10% (E) and prevalence of AF >10% (F).

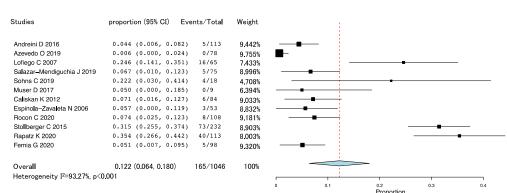
CI, confidence interval.

А



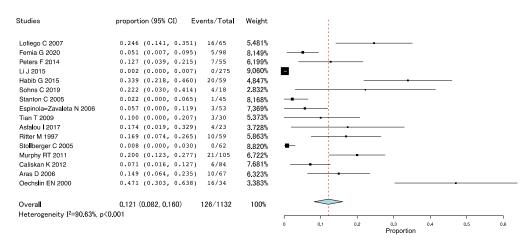
Studies	proportion (95% CI) Ever	nts/Total	Weight						
Muser D 2017	0.008 (0.000, 0.031)	0/59	12.449%		_				
Stanton C 2009	0.100 (0.000, 0.207)	3/30	6.003%			•			
Greutmann M 2012	0.205 (0.136, 0.273)	27/132	8.824%					-	
Andreini D 2016	0.044 (0.006, 0.082)	5/113	11.434%		-	÷			
Aras D 2006	0.149 (0.064, 0.235)	10/67	7.502%			1			
Stacey R 2013	0.049 (0.011, 0.088)	6/122	11.399%		-				
Rocon C 2020	0.074 (0.025, 0.123)	8/108	10.491%	-					
Habib G 2011	0.200 (0.123, 0.277)	21/105	8.190%			1		-	
Azevedo O 2019	0.006 (0.000, 0.024)	0/78	12.727%	-					
Femia G 2020	0.051 (0.007, 0.095)	5/98	10.981%		-				
Overal	0.078 (0.042, 0.113)	85/912	100%						
Heterogeneity I ² =86.86%,	p<0.001					1.00			
				0	0.05	0.1 F	0.15 Proportion	0.2	0.25

С



proportion (95% CI) Events/Total Studies Weight Li J 2015 0.024 (0.000, 0.089) 0/20 7.305% Peters F 2014 0.127 (0.039, 0.215) 7/55 6.472% 0.079 (0.032, 0.126) 10/127 0.080 (0.048, 0.112) 22/275 7.902% 8.285% Mazurkiewicz L 2017 van Waning J 2018 Habib G 2011 Ritter M 1997 0.200 (0.123, 0.277) 21/105 0.588 (0.354, 0.822) 10/17 6.900% 2.561% 21/105 Stanton C 2009 Galizio N 2011 0.100 (0.000, 0.207) 0.087 (0.026, 0.149) 3/30 5.766% 7.419% 7/80 Tian T 2014 0.264 (0.180, 0.348) 28/106 6.626% Aras D 2006 0.043 (0.017, 0.069) 10/232 8.406% Amzulescu MS 2015 0.339 (0.218, 0.460) 20/59 5.296% Oechslin EN 2000 0.471 (0.303, 0.638) 16/34 3.892% 0.022 (0.000, 0.065) 8.010% Murphy RT 2005 1/45 0.125 (0.081, 0.169) 27/216 0.205 (0.136, 0.273) 27/132 van Waning J**I** 2019 7.981% Greutmann M 2012 7.178% Overa 0.148 (0.103, 0.192) 209/1533 100% Heterogeneity I2=88,16%, p<0.001

Е



F

Studies	proportion (95% CI) Ev	ents/Total	Weight					
Minamisawa M 2016	0.021 (0.000, 0.078)	0/23	8.315%	_				
Azevedo O 2019	0.006 (0.000, 0.024)	0/78	9.277%	-	-			
Peters F 2014	0.264 (0.180, 0.348)	28/106	7.342%				•	
Mazurkiewicz L 2017	0.079 (0.032, 0.126)	10/127	8.639%					
Rapatz K 2020	0.354 (0.266, 0.442)	40/113	7.180%					
van Waning JI 2019	0.125 (0.081, 0.169)	27/216	8.718%					
Sedaghat-Hamedani F 2017	0.168 (0.093, 0.244)	16/95	7.670%					
Espinola-Zavaleta N 2013	0.003 (0.000, 0.011)	0/169	9.365%					
Salazar-Mendiguchia J 2019	0.067 (0.010, 0.123)	5/75	8.338%		e			
Greutmann M 2012	0.205 (0.136, 0.273)	27/132	7.907%				_	
Sollberger C 2015	0.315 (0.255, 0.374)	73/232	8.227%			-		
van Waning J 2018	0.080 (0.048, 0.112)	22/275	9.022%					
Overa	0.133 (0.084, 0.182)	248/1641	100%	L	_			
Heterogeneity I ² =96.15%, p<0				_				
·····, 1 · ····, 9 ·				0	0.1	0.2 Proportion	0.3	0,4

REFERENCES

1 Chin TK, Perloff JK, Williams RG, et al. Isolated noncompaction of left ventricular myocardium. A study of eight cases. *Circulation* 1990;**82**:507-13.

2 Ichida F, Hamamichi Y, Miyawaki T, et al. Clinical features of isolated noncompaction of the ventricular myocardium: long-term clinical course, hemodynamic properties, and genetic background. *J Am Coll Cardiol* 1999;**34**:233-40.

3 Pignatelli RH, McMahon CJ, Dreyer WJ, et al. Clinical characterization of left ventricular noncompaction in children: a relatively common form of cardiomyopathy. *Circulation* 2003;**108**:2672-8.

4 Wald R, Veldtman G, Golding F, et al. Determinants of outcome in isolated ventricular noncompaction in childhood. *Am J Cardiol* 2004;**94**:1581-4.

5 Lilje C, Razek V, Joyce JJ, et al. Complications of non-compaction of the left ventricular myocardium in a paediatric population: a prospective study. *Eur Heart J* 2006;**27**:1855-60.

Koh C, Lee PW, Yung TC, et al. Left ventricular noncompaction in children. *Congenit Heart Dis* 2009;**4**:288-94.

7 Wang C, Takasaki A, Watanabe Ozawa S, et al. Long-Term Prognosis of Patients With Left Ventricular Noncompaction- Comparison Between Infantile and Juvenile Types. *Circ J* 2017;**81**:694-700.

8 Nucifora G, Sree Raman K, Muser D, et al. Cardiac magnetic resonance evaluation of left ventricular functional, morphological, and structural features in children and adolescents vs. young adults with isolated left ventricular noncompaction. *Int J Cardiol* 2017;**246**:68-73.

9 van Waning JI, Caliskan K, Hoedemaekers YM, et al. Genetics, Clinical Features, and Long-Term Outcome of Noncompaction Cardiomyopathy. *J Am Coll Cardiol* 2018;**71**:711-22.

10 Takasaki A, Hirono K, Hata Y, et al. Sarcomere gene variants act as a genetic trigger underlying the development of left ventricular noncompaction. *Pediatr Res* 2018;**84**:733-42.

11 Hirono K, Hata Y, Miyao N, et al. Increased Burden of Ion Channel Gene Variants Is Related to Distinct Phenotypes in Pediatric Patients With Left Ventricular Noncompaction. *Circ Genom Precis Med* 2020;**13**:e002940.

12 Ritter M, Oechslin E, Sutsch G, et al. Isolated noncompaction of the myocardium in adults. *Mayo Clin Proc* 1997;**72**:26-31.

13 Oechslin EN, Attenhofer Jost CH, Rojas JR, et al. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol* 2000;**36**:493-500.

14 Stollberger C, Finsterer J. Left ventricular hypertrabeculation/noncompaction and stroke or embolism. *Cardiology* 2005;**103**:68-72.

15 Murphy RT, Thaman R, Blanes JG, et al. Natural history and familial characteristics of isolated left ventricular non-compaction. *Eur Heart J* 2005;**26**:187-92.

16 Lofiego C, Biagini E, Pasquale F, et al. Wide spectrum of presentation and variable outcomes of isolated left ventricular non-compaction. *Heart* 2007;**93**:65-71.

17 Espinola-Zavaleta N, Soto ME, Castellanos LM, et al. Non-compacted cardiomyopathy: clinicalechocardiographic study. *Cardiovasc Ultrasound* 2006;**4**:35.

Aras D, Tufekcioglu O, Ergun K, et al. Clinical features of isolated ventricular noncompaction in adults long-term clinical course, echocardiographic properties, and predictors of left ventricular failure. *J Card Fail* 2006;12:726-33.
Finsterer J, Stollberger C, Molzer G, et al. Cerebrovascular events in left ventricular hypertrabeculation/noncompaction with and without myopathy. *Int J Cardiol* 2008;130:344-8.

20 Fazio G, Corrado G, Zachara E, et al. Anticoagulant drugs in noncompaction: a mandatory therapy? J

Cardiovasc Med (Hagerstown) 2008;9:1095-7.

21 Yousef ZR, Foley PW, Khadjooi K, et al. Left ventricular non-compaction: clinical features and cardiovascular magnetic resonance imaging. *BMC Cardiovasc Disord* 2009;**9**:37.

22 Stanton C, Bruce C, Connolly H, et al. Isolated left ventricular noncompaction syndrome. *Am J Cardiol* 2009;**104**:1135-8.

23 Shoji M, Yamashita T, Uejima T, et al. Electrocardiography characteristics of isolated non-compaction of ventricular myocardium in Japanese adult patients. *Circ J*2010;**74**:1431-5.

Habib G, Charron P, Eicher JC, et al. Isolated left ventricular non-compaction in adults: clinical and echocardiographic features in 105 patients. Results from a French registry. *Eur J Heart Fail* 2011;**13**:177-85.

Greutmann M, Mah ML, Silversides CK, et al. Predictors of adverse outcome in adolescents and adults with isolated left ventricular noncompaction. *Am J Cardiol* 2012;**109**:276-81.

26 Yun H, Zeng MS, Jin H, et al. Isolated noncompaction of ventricular myocardium: a magnetic resonance imaging study of 11 patients. *Korean J Radiol* 2011;**12**:686-92.

27 Stollberger C, Wegner C, Finsterer J. CHADS2- and CHA2DS2VASc scores and embolic risk in left ventricular hypertrabeculation/noncompaction. *J Stroke Cerebrovasc Dis* 2013;**22**:709-12.

28 Nikolic A, Jovovic L, Tomic S, et al. Left ventricular noncompaction: clinical-echocardiographic study. *Vojnosanit Pregl* 2012;**69**:32-6.

29 Caliskan K, Ujvari B, Bauernfeind T, et al. The prevalence of early repolarization in patients with noncompaction cardiomyopathy presenting with malignant ventricular arrhythmias. *J Cardiovasc Electrophysiol* 2012;**23**:938-44.

30 Stacey RB, Andersen MM, St Clair M, et al. Comparison of systolic and diastolic criteria for isolated LV noncompaction in CMR. *JACC Cardiovasc Imaging* 2013;**6**:931-40.

Tian T, Liu Y, Gao L, et al. Isolated left ventricular noncompaction: clinical profile and prognosis in 106 adult patients. *Heart Vessels* 2014;**29**:645-52.

32 Peters F, Khandheria BK, Botha F, et al. Clinical outcomes in patients with isolated left ventricular noncompaction and heart failure. *J Card Fail* 2014;**20**:709-15.

Li J, Franke J, Pribe-Wolferts R, et al. Effects of beta-blocker therapy on electrocardiographic and echocardiographic characteristics of left ventricular noncompaction. *Clin Res Cardiol* 2015;**104**:241-9.

Akhbour S, Fellat I, Fennich N, et al. Electrocardiographic findings in correlation to magnetic resonance imaging patterns in African patients with isolated ventricular noncompaction. *Anatol J Cardiol* 2015;**15**:550-5.

35 Stollberger C, Blazek G, Gessner M, et al. Age-dependency of cardiac and neuromuscular findings in adults with left ventricular hypertrabeculation/noncompaction. *Am J Cardiol* 2015;**115**:1287-92.

36 Amzulescu MS, Rousseau MF, Ahn SA, et al. Prognostic Impact of Hypertrabeculation and Noncompaction Phenotype in Dilated Cardiomyopathy: A CMR Study. *JACC Cardiovasc Imaging* 2015;**8**:934-46.

37 Minamisawa M, Koyama J, Kozuka A, et al. Regression of left ventricular hypertrabeculation is associated with improvement in systolic function and favorable prognosis in adult patients with non-ischemic cardiomyopathy. *J Cardiol* 2016;**68**:431-8.

38 Zhou H, Lin X, Fang L, et al. Characterization of Compacted Myocardial Abnormalities by Cardiac Magnetic Resonance With Native T1 Mapping in Left Ventricular Non-Compaction Patients- A Comparison With Late Gadolinium Enhancement. *Circ J*2016;**80**:1210-6.

39 Andreini D, Pontone G, Bogaert J, et al. Long-Term Prognostic Value of Cardiac Magnetic Resonance in Left Ventricle Noncompaction: A Prospective Multicenter Study. *J Am Coll Cardiol* 2016;**68**:2166-81. 40 Muser D, Liang JJ, Witschey WR, et al. Ventricular arrhythmias associated with left ventricular noncompaction: Electrophysiologic characteristics, mapping, and ablation. *Heart Rhythm* 2017;**14**:166-75.

41 Asfalou I, Boulaamayl S, Raissouni M, et al. Left ventricular noncompaction-A rare form of cardiomyopathy: Revelation modes and predictors of mortality in adults through 23 cases. *J Saudi Heart Assoc* 2017;**29**:102-9.

42 Tavares de Melo MD, Giorgi MCP, Assuncao AN, Jr., et al. Decreased glycolytic metabolism in noncompaction cardiomyopathy by 18F-fluoro-2-deoxyglucose positron emission tomography: new insights into pathophysiological mechanisms and clinical implications. *Eur Heart J Cardiovasc Imaging* 2017;**18**:915-21.

43 Mazurkiewicz L, Petryka J, Spiewak M, et al. Clinical and prognostic relevancy of left ventricular trabeculation assessed by cardiac magnetic resonance in patients with dilated cardiomyopathy. *Kardiol Pol* 2017;**75**:794-803.

44 Sedaghat-Hamedani F, Haas J, Zhu F, et al. Clinical genetics and outcome of left ventricular non-compaction cardiomyopathy. *Eur Heart J* 2017;**38**:3449-60.

45 Salazar-Mendiguchia J, Gonzalez-Costello J, Oliveras T, et al. Long-term Follow-up of Symptomatic Adult Patients With Noncompaction Cardiomyopathy. *Rev Esp Cardiol (Engl Ed)* 2019;**72**:169-71.

46 Martinez HR, Ware SM, Schamberger MS, et al. Noncompaction cardiomyopathy and heterotaxy syndrome. *Prog Pediatr Cardiol* 2017;**46**:23-7.

47 Araujo-Filho JAB, Assuncao AN, Jr., Tavares de Melo MD, et al. Myocardial T1 mapping and extracellular volume quantification in patients with left ventricular non-compaction cardiomyopathy. *Eur Heart J Cardiovasc Imaging* 2018;**19**:888-95.

48 Sohns C, Ouyang F, Volkmer M, et al. Therapy of ventricular arrhythmias in patients suffering from isolated left ventricular non-compaction cardiomyopathy. *Europace* 2019;**21**:961-9.

49 van Waning JI, Caliskan K, Michels M, et al. Cardiac Phenotypes, Genetics, and Risks in Familial Noncompaction Cardiomyopathy. *J Am Coll Cardiol* 2019;**73**:1601-11.

50 Azevedo O, Marques N, Craveiro N, et al. Screening for Fabry disease in patients with left ventricular noncompaction. *Rev Port Cardiol* 2019;**38**:709-16.

51 Macaione F, Meloni A, Positano V, et al. The planimetric Grothoff's criteria by cardiac magnetic resonance can improve the specificity of left ventricular non-compaction diagnosis in thalassemia intermedia. *Int J Cardiovasc Imaging* 2020;**36**:1105-12.

52 Asmakutlu O, Alis D, Topel C, et al. Late gadolinium enhancement on CMRI in patients with LV noncompaction: An overestimated phenomenon? *Clin Imaging* 2020;**66**:121-6.

Rocon C, Tabassian M, Tavares de Melo MD, et al. Biventricular imaging markers to predict outcomes in noncompaction cardiomyopathy: a machine learning study. *ESC Heart Fail* 2020;**7**:2431-9.

54 Rapatz K, Finsterer J, Voill-Glaninger A, et al. NT-pro-BNP in patients with left ventricular hypertrabeculation/non-compaction. *ESC Heart Fail* 2020.

55 Zemrak F, Raisi-Estabragh Z, Khanji MY, et al. Left Ventricular Hypertrabeculation Is Not Associated With Cardiovascular Morbity or Mortality: Insights From the Eurocmr Registry. *Front Cardiovasc Med* 2020;**7**:158.

56 Femia G, Zhu D, Choudhary P, et al. Long term clinical outcomes associated with CMR quantified isolated left ventricular non-compaction in adults. *Int J Cardiol* 2021;**328**:235-40.

57 Galizio NOG, José L.;Favaloro, Liliana E.;Diez, Mirta;Fernández, Adrián;Guevara, Eduardo;Palazzo, Alejandro A.;Robles, Federico;Casabé, José H.;Galizio, N. O. Non-compaction cardiomyopathy. Risk stratification of Sudden death for automatic cardioverter defibrillator implantation. *REVARGENT CARDIOL* 2011;**79**:7.