Validation of the 2014 ESC Guidelines Risk Prediction Model for the Primary Prevention of Sudden Cardiac Death in Hypertrophic Cardiomyopathy

Journal club, May 20th, 2015

HCMにおけるICD植え込みの適応（JCS2012）

Class I

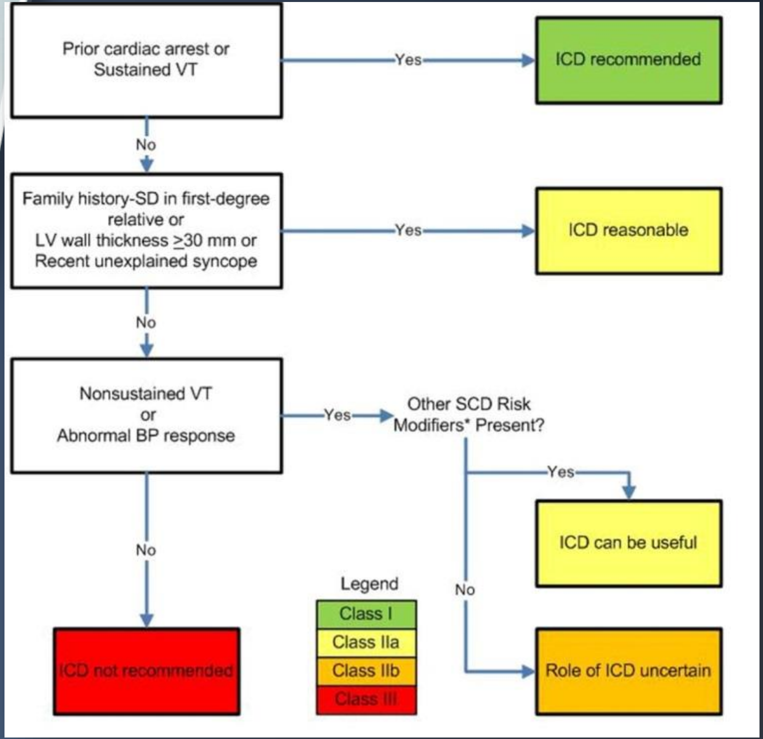
過去に持続性心室頻拍，心室細動，心停止の既往を有する場合

Class IIa

非持続性心室頻拍
突然死の家族歴
失神
左室壁厚30mm以上
運動時の血圧反応異常
のいずれかを認める場合に考慮する

日本循環器学会　肥大型心筋症の診療に関するガイドライン（2012年改訂版）

HCMにおけるICD植え込みの適応（ACC/AHA2011）



Prior cardiac arrest or Sustained VT

Yes

ICD recommended

No

Family history-SD in first-degree relative or LV wall thickness ≥30 mm or Recent unexplained syncope

Yes

ICD reasonable

No

Nonsustained VT or Abnormal BP response

Yes

Other SCD Risk Modifiers* Present?

Yes

ICD can be useful

No

Role of ICD uncertain

No

ICD not recommended

Legend

🟩 Class I
🟨 Class IIa
🟧 Class IIb
🟥 Class III

Class I
心停止、VF、持続性VTに対する二次予防目的
（Level B）

Class IIa
1)　1親等以内の突然死家族歴
壁厚≥30mm
最近の失神　　のいずれかを認める場合

2)　NSVT or 血圧反応異常
かつ他のリスクあり
（LVOT obstruction、LGE on CMR、
LV apical aneurysm、Malignant mutation(?)）
（Level C）

Class IIb
NSVTのみ or 血圧反応異常のみ
（Level C）

2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy
J Am Coll Cardiol. 2011 Dec 13;58(25):e212-60.

HCMにおけるICD植え込みの適応（ESC2014）

SECONDARY PREVENTION

- Cardiac arrest due to VT or VF
- spontaneous sustained VT causing syncope or haemodynamic compromise

Life expectancy >1 year

ICD recommended

Class I (Level B)

PRIMARY PREVENTION

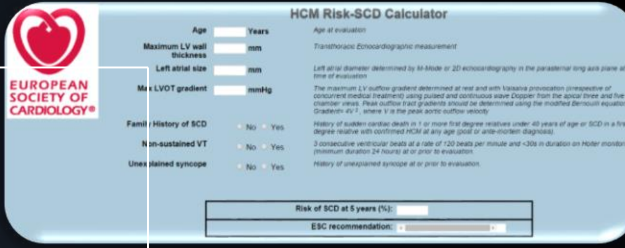
Recommended assessment:

History
2D/Doppler echocardiogram
48–hour ambulatory ECG

HCM Risk-SCD variables:

- Age
- Family history of sudden cardiac death
- Unexplained syncope
- Left ventricular outflow gradient[a]
- Maximum left ventricular wall thickness[a]
- Left atrial diameter[a]
- NSVT

HCM Risk-SCD Score



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HCM Risk-SCD Calculator

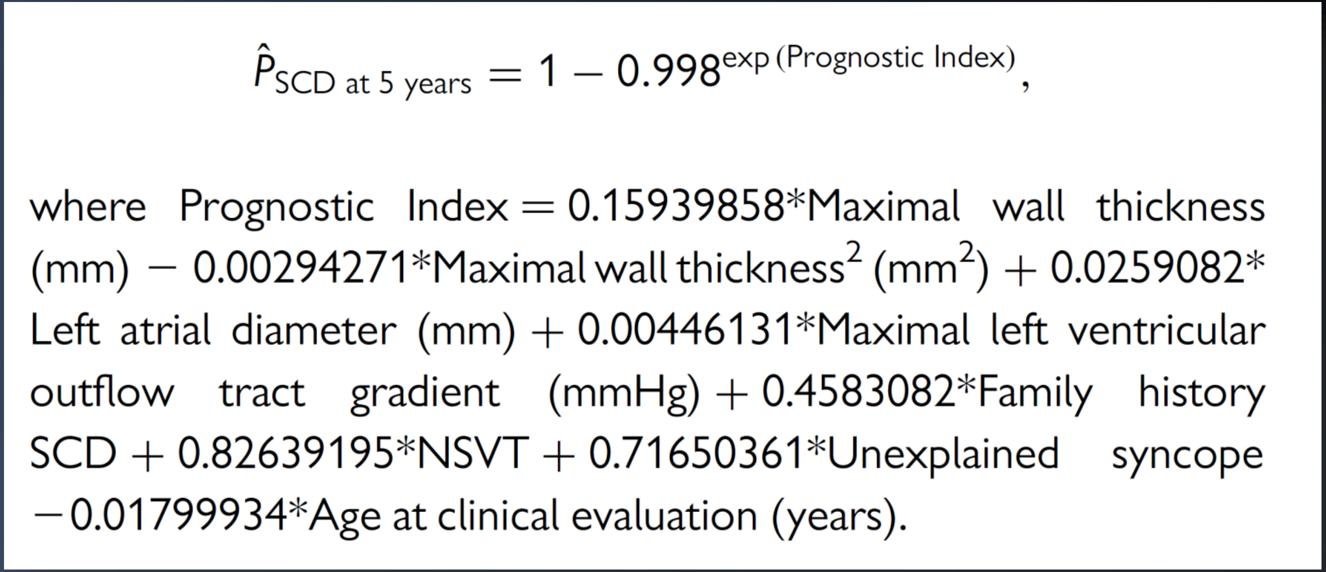
Age	Years	*Age at evaluation*
Maximum LV wall thickness	mm	*Transthoracic Echocardiographic measurement*
Left atrial size	mm	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	mmHg	*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernoulli equation: Gradient= 4V², where V is the peak aortic outflow velocity*
Family History of SCD	○ No ○ Yes	*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis)*
Non-sustained VT	○ No ○ Yes	*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation*
Unexplained syncope	○ No ○ Yes	*History of unexplained syncope at or prior to evaluation*

Risk of SCD at 5 years (%):
ESC recommendation:

LOW RISK 5–year risk <4%	INTERMEDIATE RISK 5–year risk ≥4-<6%	HIGH RISK 5–year risk ≥6%
ICD generally not indicated[b]	ICD may be considered	ICD should be considered
Class IIb or III (Level B)	Class IIb (Level B)	Class IIa (Level B)

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy: the Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC)
Eur Heart J. 2014 Oct 14;35(39):2733-79.

The risk of SCD in 5 years



$$\hat{P}_{\text{SCD at 5 years}} = 1 - 0.998^{\exp\,(\text{Prognostic Index})},$$

where Prognostic Index $= 0.15939858$*Maximal wall thickness (mm) $-$ 0.00294271*Maximal wall thickness^2 (mm^2) $+$ 0.0259082* Left atrial diameter (mm) $+$ 0.00446131*Maximal left ventricular outflow tract gradient (mmHg) $+$ 0.4583082*Family history SCD $+$ 0.82639195*NSVT $+$ 0.71650361*Unexplained syncope -0.01799934*Age at clinical evaluation (years).

NRI: net reclassification improvement

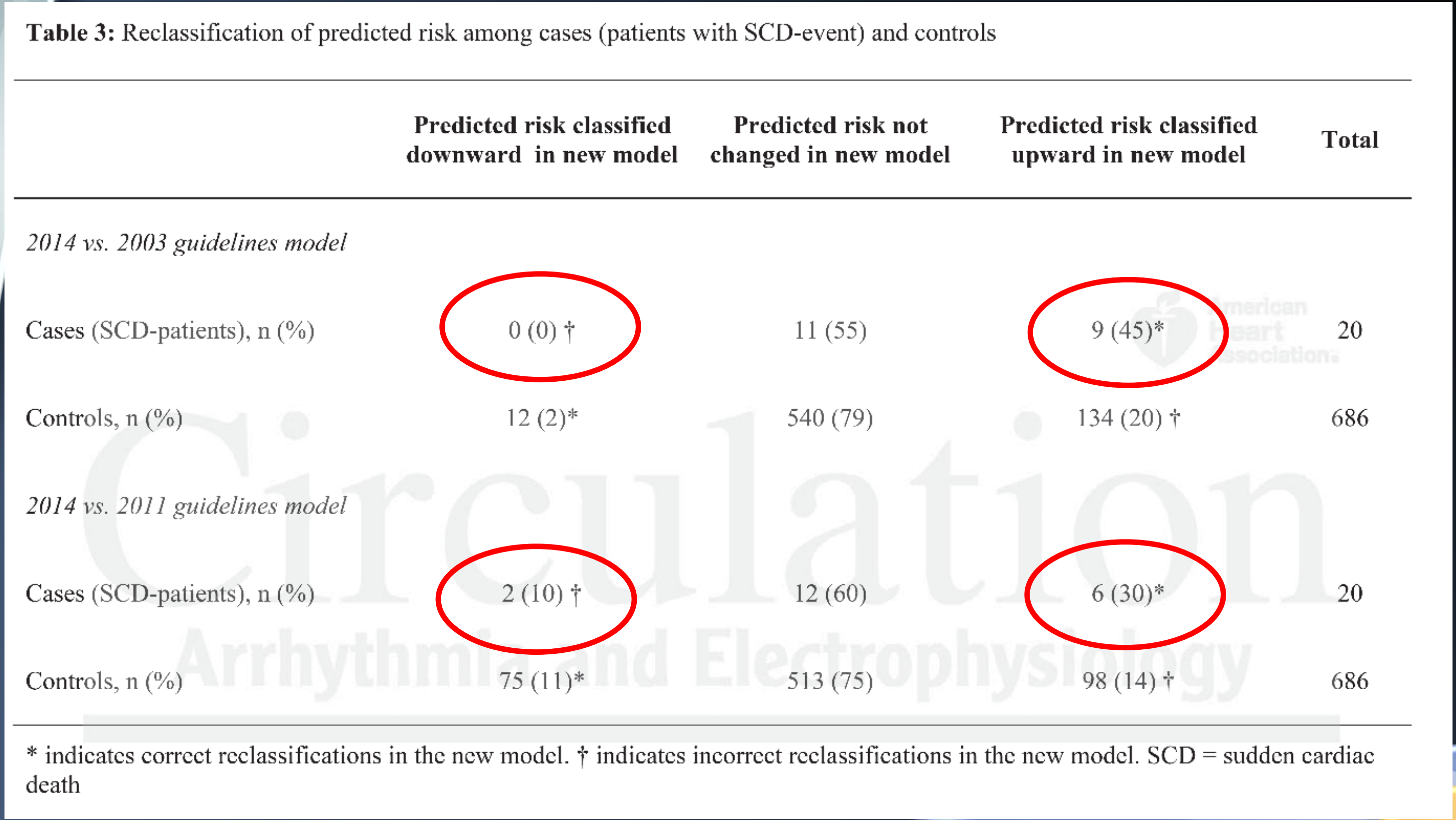


Table 3: Reclassification of predicted risk among cases (patients with SCD-event) and controls

	Predicted risk classified downward in new model	**Predicted risk not changed in new model**	**Predicted risk classified upward in new model**	**Total**
2014 vs. 2003 guidelines model				
Cases (SCD-patients), n (%)	0 (0) †	11 (55)	9 (45)*	20
Controls, n (%)	12 (2)*	540 (79)	134 (20) †	686
2014 vs. 2011 guidelines model				
Cases (SCD-patients), n (%)	2 (10) †	12 (60)	6 (30)*	20
Controls, n (%)	75 (11)*	513 (75)	98 (14) †	686

* indicates correct reclassifications in the new model. † indicates incorrect reclassifications in the new model. SCD = sudden cardiac death

American Heart Association
Circulation
Arrhythmia and Electrophysiology

NRI

0.27
(95%CI -0.02-0.57,
p=0.07)

0.16
(95%CI -0.17-0.45,
p=0.2)

NNT: number needed to treat

1人のSCDを予防するのに何人ICDを植え込めばよいか

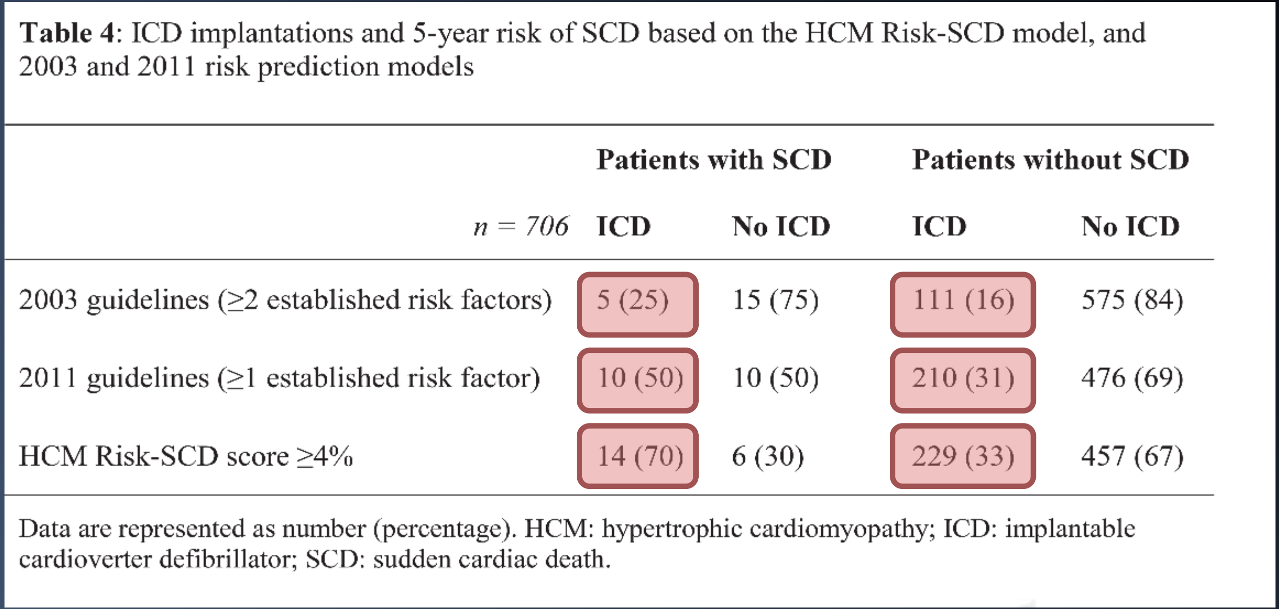


Table 4: ICD implantations and 5-year risk of SCD based on the HCM Risk-SCD model, and 2003 and 2011 risk prediction models

	Patients with SCD		Patients without SCD	
n = 706	**ICD**	**No ICD**	**ICD**	**No ICD**
2003 guidelines (≥2 established risk factors)	5 (25)	15 (75)	111 (16)	575 (84)
2011 guidelines (≥1 established risk factor)	10 (50)	10 (50)	210 (31)	476 (69)
HCM Risk-SCD score ≥4%	14 (70)	6 (30)	229 (33)	457 (67)

Data are represented as number (percentage). HCM: hypertrophic cardiomyopathy; ICD: implantable cardioverter defibrillator; SCD: sudden cardiac death.

NNT

22

20

17