Il documento di consenso degli esperti su sincope e cardiomiopatia ipertrofica

Firenze 7-8 luglio 2022

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

European Heart Journal

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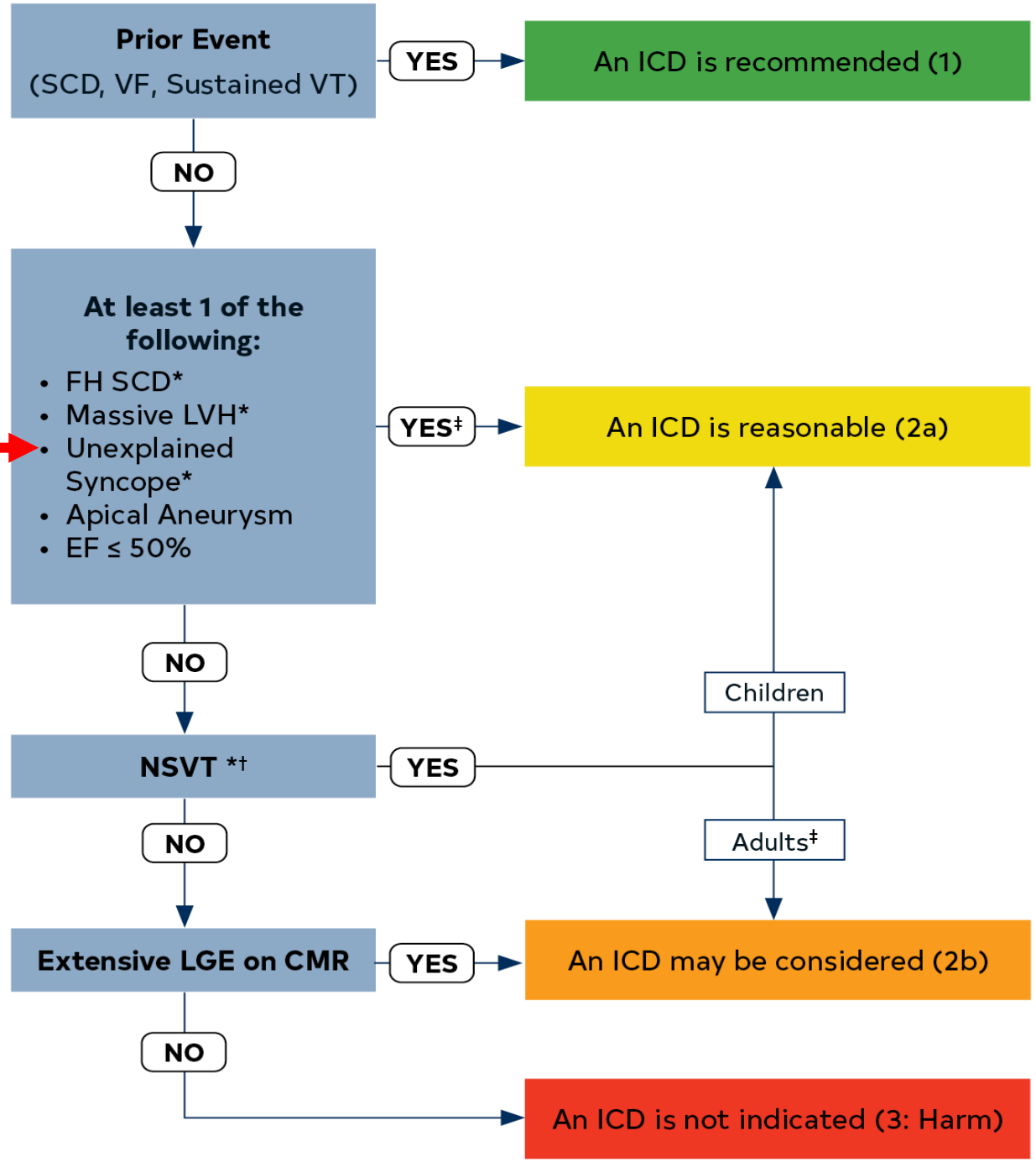
Risk of SCD at 5 years (%):

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 Bernouilli equation: Gradient= $4V^2$, 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.*

AHA-ACC HCM Guidelines 2020
SCD Risk Assessment & ICD Recommendations



Causes of syncope in HCM patients

1. Hypovolaemia
2. Complete AV block
3. Sinus node dysfunction
4. Sustained fast VT
5. LVOTO and abnormal vascular reflexes
6. Occasionally atrial arrhythmias with fast ventricular response

   (in individuals with preserved atrial function and high filling pressures)
7. Comorbidities (Epilepsy, Hypoglicaemia in diabetic pts)

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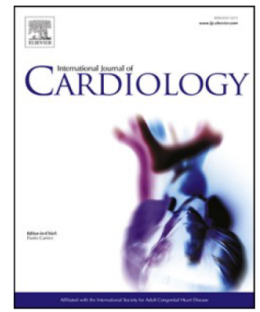

Review

Syncope in hypertrophic cardiomyopathy (part I): An updated systematic review and meta-analysis

https://doi.org/10.1016/j.ijcard.2022.03.028

Giuseppe Mascia [a], Lia Crotti [b], Antonella Groppelli [b], Marco Canepa [a,b,c], Carlo Merlo Andrea [a,b,c], Stefano Benenati [a,b,c], Paolo Di Donna [a], Roberta Della Bona [a], Davide Soranna [b], Antonella Zambon [b], Italo Porto [a,b,c], Olivotto Iacopo [d], Gianfranco Parati [b], Michele Brignole [b,1], Franco Cecchi [b,1,*]

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SYNCOPE IN HCM PTS

22279 pts from 60 studies

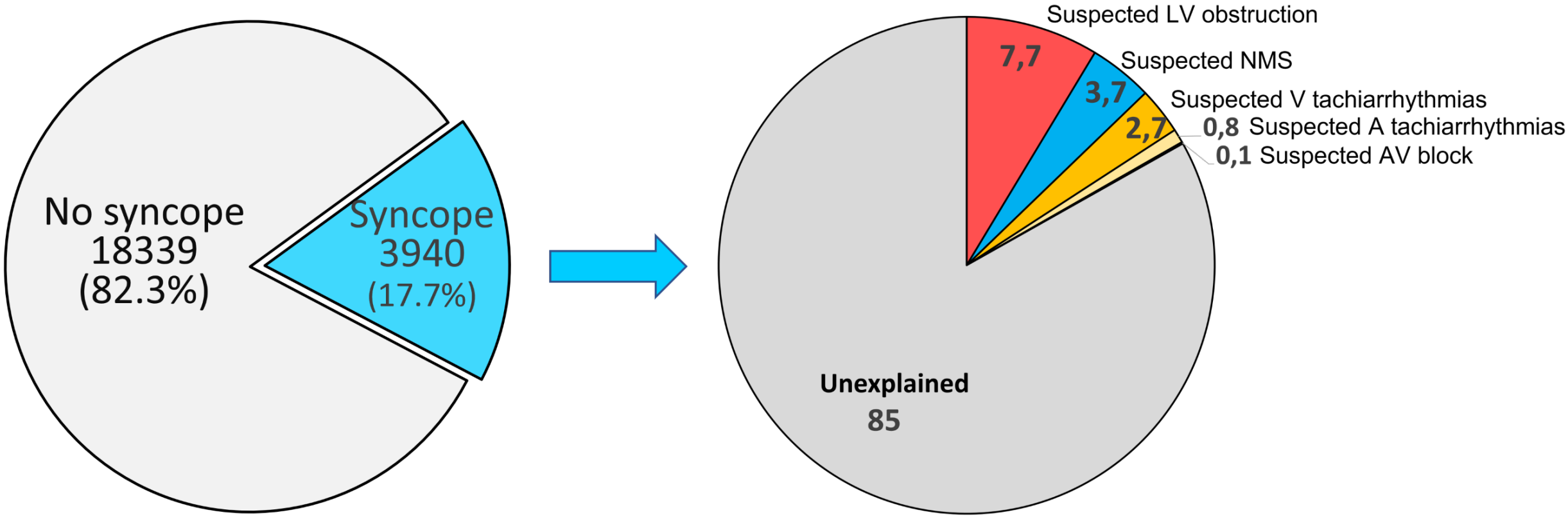

Table 1. Raw data of life-threatening arrhythmic events in 14 HCM studies that reported data for patients with and without a history of syncope

Reference	Pts	FU (years)	Mean age	No syncope		Syncope	
				Patients	Events	Patients	Events
Rowin et al, 2020	146	5,8	16	129	9	17	1
O'Mahony et al, 2014	3703	5,9	52	3229	56	474	17
Adler et al, 2017	168	1,3	59	154	0	14	0
O'Mahony et al, 2018	3675	5,7	48	3168	146	507	52
Finocchiaro et al, 2012	84	8,5	43	50	6	34	3
Furushima et al, 2010	66	0,8	55	55	14	11	0
Efthimiadis et al, 2009	123	2,9	52	106	0	17	2
Spirito et al, 2009	1511	5,6	46	1306	61	205	13
D'Andrea et al, 2006	123	4	39	99	10	24	2
Barletta et al,2004	28	0,75	44	20	0	8	2
Kawasaki et al, 2003	73	2,3	52	63	0	10	0
Isobe et al, 2003	58	10,4	51	50	3	8	2
Zhu et al, 1998	53	3,9	42	40	0	13	3
Dilsizian et al, 1993	23	0,4	17	8	0	15	8
Total	9834	5,6	49	8477	305 (3.6%)	1357	105 (7.7%)

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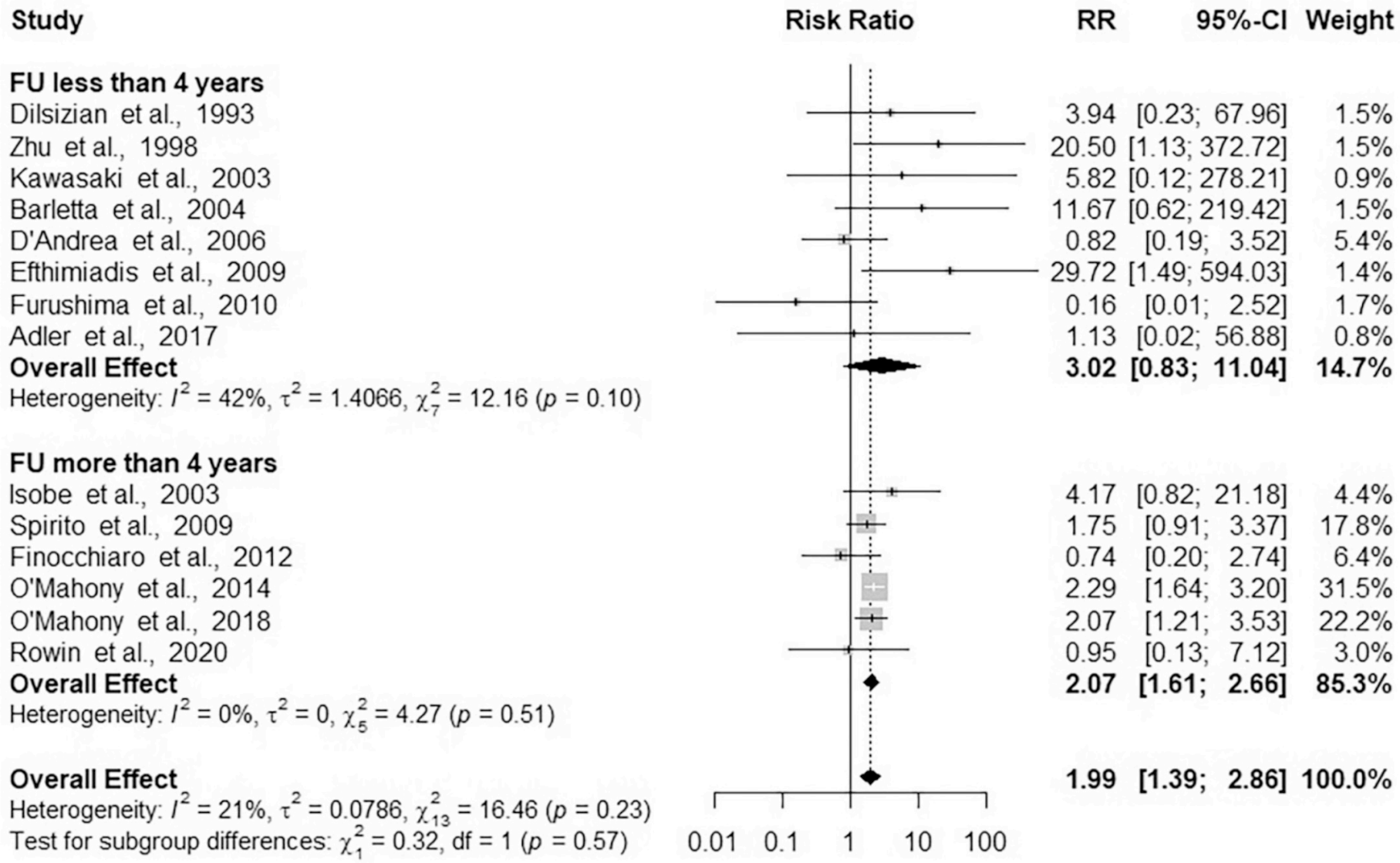

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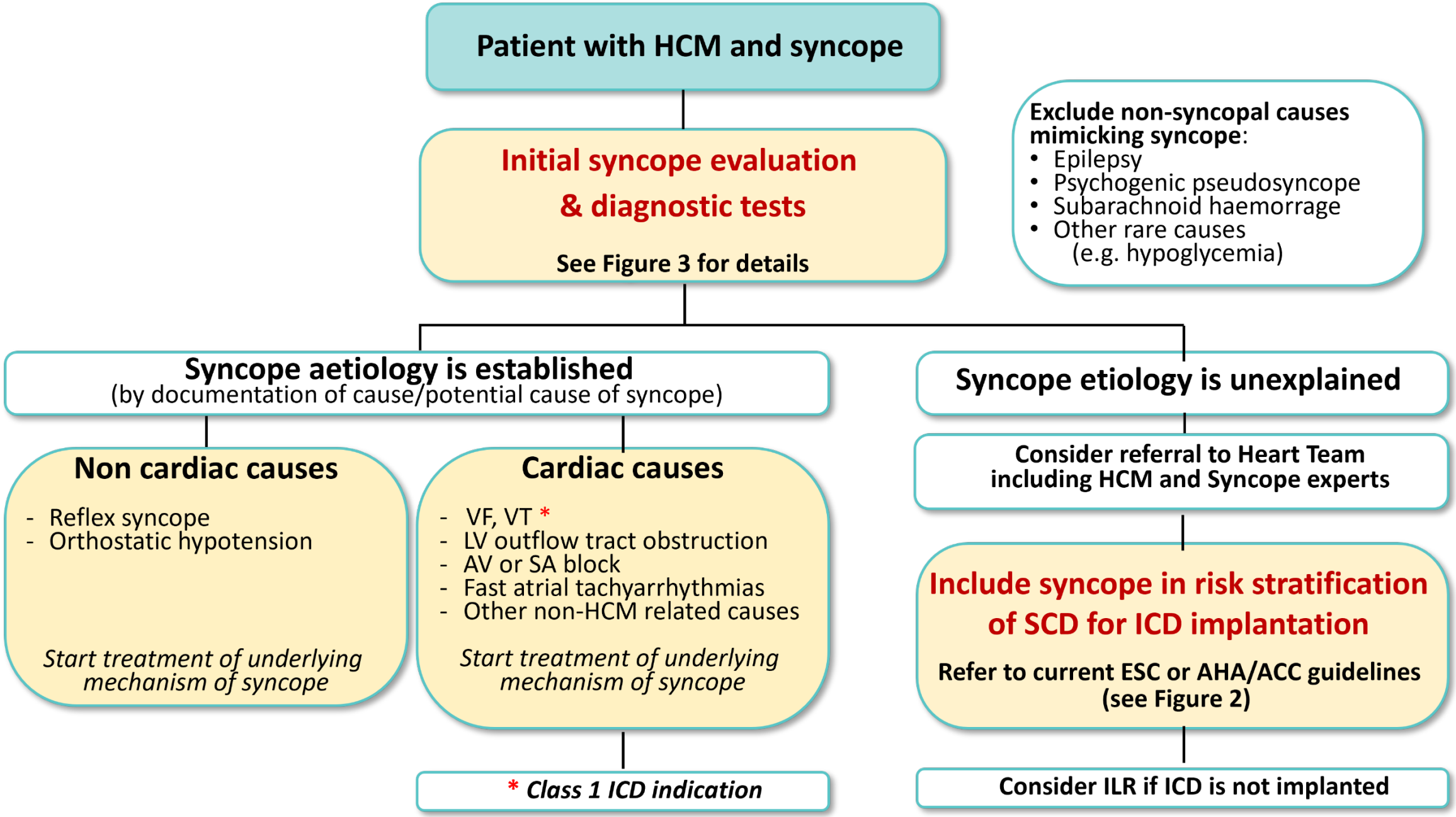
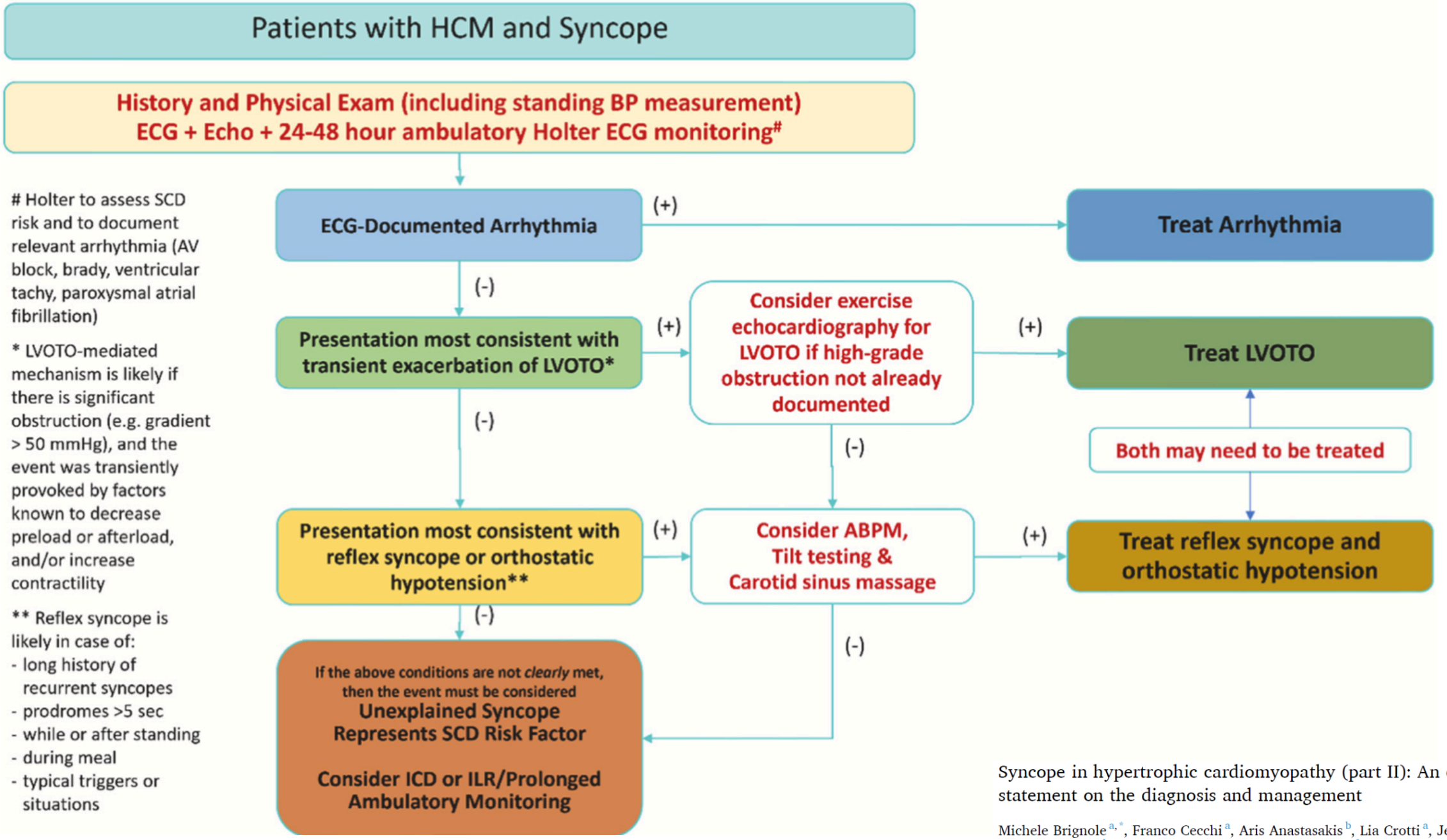
Review

Syncope in hypertrophic cardiomyopathy (part II): An expert consensus statement on the diagnosis and management

Michele Brignole [a,*], Franco Cecchi [a], Aris Anastasakis [b], Lia Crotti [a,p], Jean Claude Deharo [c,o], Perry M. Elliott [d], Artur Fedorowski [e], Juan Pablo Kaski [f], Giuseppe Limongelli [g], Martin S. Maron [h], Iacopo Olivotto [i], Steve R. Ommen [j], Gianfranco Parati [k], Win Shen [l], Andrea Ungar [m], Arthur Wilde [n]

Name	Remark
Anastasakis Aris	Member, 2014 ESC HCM guidelines
Brignole Michele	Chair, 2018 ESC Syncope guidelines
Cecchi Franco	Member, 2014 ESC HCM guidelines
Crotti Lia	Member of ERN and EHRA)/HRS/APHRS/LAHRS Expert Consensus Statement on the State of Genetic Testing for Cardiac Diseases
Deharo Jean Claude	Member, 2018 ESC Syncope Guidelines
Elliott Perry	Chair, 2014 ESC HCM guidelines Member, 2020 AHA/ACC HCM guidelines
Fedorowski Artur	Member, 2018 ESC Syncope Guidelines
Kaski Juan Pablo	Chair, 2023 ESC Cardiomyopathy guidelines
Limongelli Giuseppe	Member of ERN; Member, 2014 ESC HCM guidelines
Maron Martin S	Member, 2020 AHA/ACC HCM guidelines
Olivotto Iacopo	Member, 2019 EAPC Recommendations for sports participation in athletes with cardiomyopathies, myocarditis and pericarditis.
Ommen Steve R	Chair, 2020 AHA/ACC HCM guidelines
Parati Gianfranco	Member, 2018 ESC/ESH Hypertension guidelines
Shen Win	Chair, 2017 AHA/ACC Syncope guidelines
Ungar Andrea	Member, 2018 ESC Syncope Guidelines
Wilde Arthur	Member, ERN 2020 APHRS/HRS expert consensus statement on the investigation of decedents with sudden unexplained


Patient with HCM and syncope
Initial syncope evaluation & diagnostic tests
See Figure 3 for details
Exclude non-syncopal causes mimicking syncope:
• Epilepsy
• Psychogenic pseudosyncope
• Subarachnoid haemorrage
• Other rare causes (e.g. hypoglycemia)
Syncope aetiology is established
(by documentation of cause/potential cause of syncope)
Syncope etiology is unexplained
Non cardiac causes
- Reflex syncope
- Orthostatic hypotension
Start treatment of underlying mechanism of syncope
Cardiac causes
- VF, VT *
- LV outflow tract obstruction
- AV or SA block
- Fast atrial tachyarrhythmias
- Other non-HCM related causes
Start treatment of underlying mechanism of syncope
* Class 1 ICD indication
Consider referral to Heart Team including HCM and Syncope experts
Include syncope in risk stratification of SCD for ICD implantation
Refer to current ESC or AHA/ACC guidelines (see Figure 2)
Consider ILR if ICD is not implanted



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Diagnostic criteria with initial evaluation (I)

Recommendations	Class	Level
Reflex syncope and OH		
1. VVS is highly probable if syncope is precipitated by pain or fear or standing, and is associated with typical progressive prodrome (pallor, sweating, nausea).	I	C
2. Situational reflex syncope is highly probable if syncope occurs during or immediately after specific triggers.	I	C
3. Syncope due to OH is confirmed when syncope occurs while standing and there is concomitant significant OH.	I	C
4. In the absence of the above criteria, reflex syncope and OH should be considered likely when the features that suggest reflex syncope or OH are present and the features that suggest cardiac syncope are absent.	IIa	C

Clinical and ECG features that suggest a reflex (neurally-mediated) syncope

- Long history of recurrent syncope, in particular occurring before the age of 40 years.
- After unpleasant sight, sound, smell, or pain.
- Prolonged standing.
- During meal.
- Being in crowded and/or hot places.
- Autonomic activation before syncope: pallor, sweating, and/or nausea/vomiting.
- With head rotation or pressure on carotid sinus (as in tumours, shaving, tight collars).
- Absence of heart disease.

2018 ESC Guidelines on Syncope – Michele Brignole & Angel Moya
EHJ Doi:10.1093/eurheartj/ehy037

Clinical and ECG features that suggest a syncope due to orthostatic hypotension

- While or after standing.
- Prolonged standing.
- Standing after exertion.
- Post-prandial hypotension.
- Temporal relationship with start or changes of dosage of vasodepressive drugs or diuretics leading to hypotension.
- Presence of autonomic neuropathy or Parkinsonism.

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Carotid sinus massage

Recommendations	Class	Level
Indication		
1. CSM is indicated in patients >40 years of age with syncope of unknown origin compatible with a reflex mechanism.	I	B
Diagnostic criteria		
2. CSS is confirmed if CSM causes bradycardia (asystole) and/or hypotension that reproduce spontaneous symptoms and patients have clinical features compatible with a reflex mechanism of syncope.	I	B

Tilt testing

Recommendations	Class	Level
Indications		
1. Tilt testing should be considered in patients with suspected reflex syncope, OH, POTS, or PPS.	IIa	B
2. Tilt testing may be considered to educate patients to recognize symptoms and learn physical manoeuvres.	IIb	B
Diagnostic criteria		
3. Reflex syncope, OH, POTS, or PPS should be considered likely if tilt testing reproduces symptoms along with the characteristic circulatory pattern of these conditions.	IIa	B

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