



# Les recommandations IC et Cardiomyopathies 2023

Emmanuelle Berthelot

CHU Bicêtre

# 2023 Focused Update of the 2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure



## Updated the following sections:

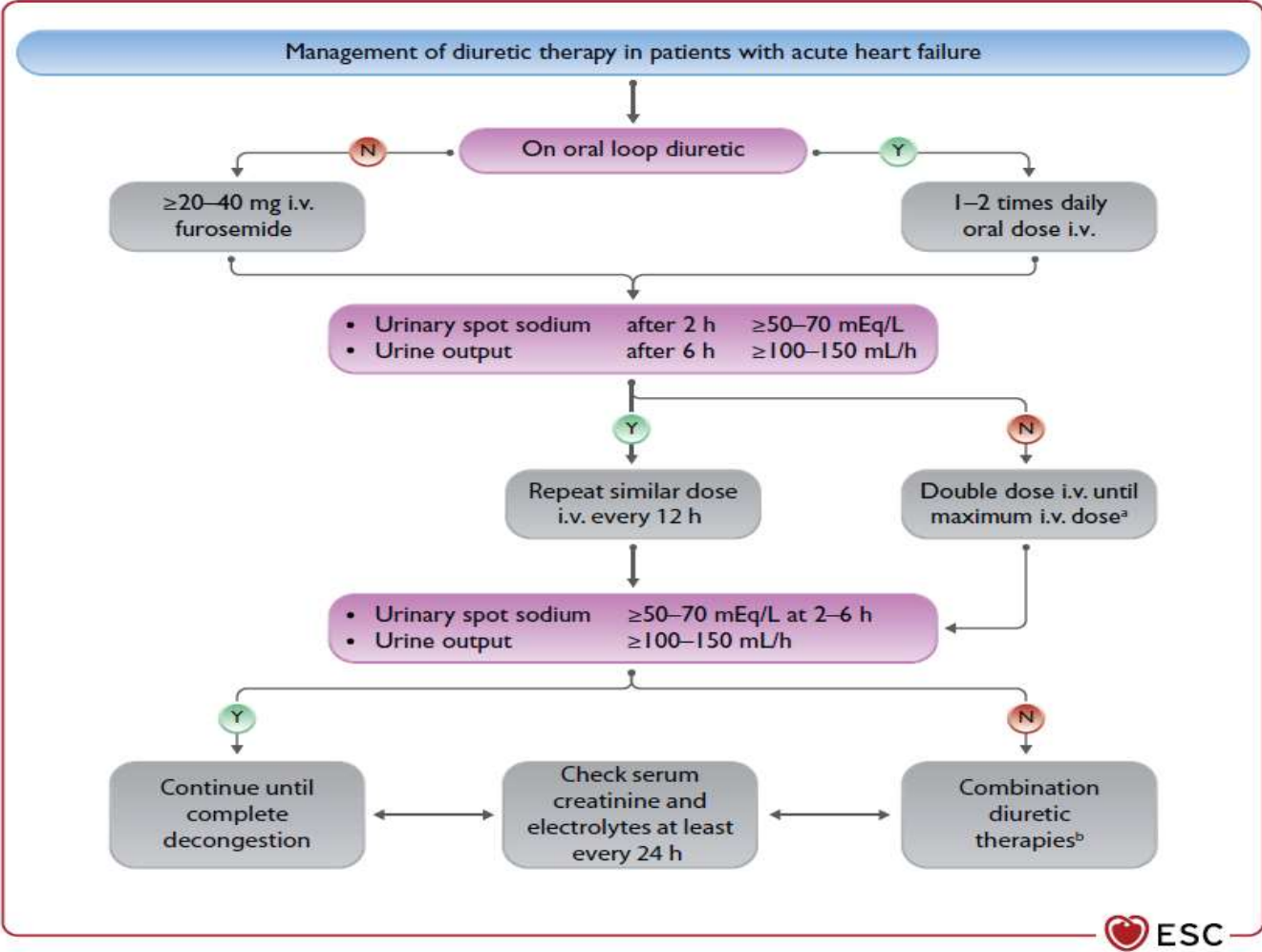
### Chronic HF

- HF with mildly reduced ejection fraction (HFmrEF)
- HF with preserved ejection fraction (HFpEF)

### Acute HF

### Comorbidities and prevention of HF

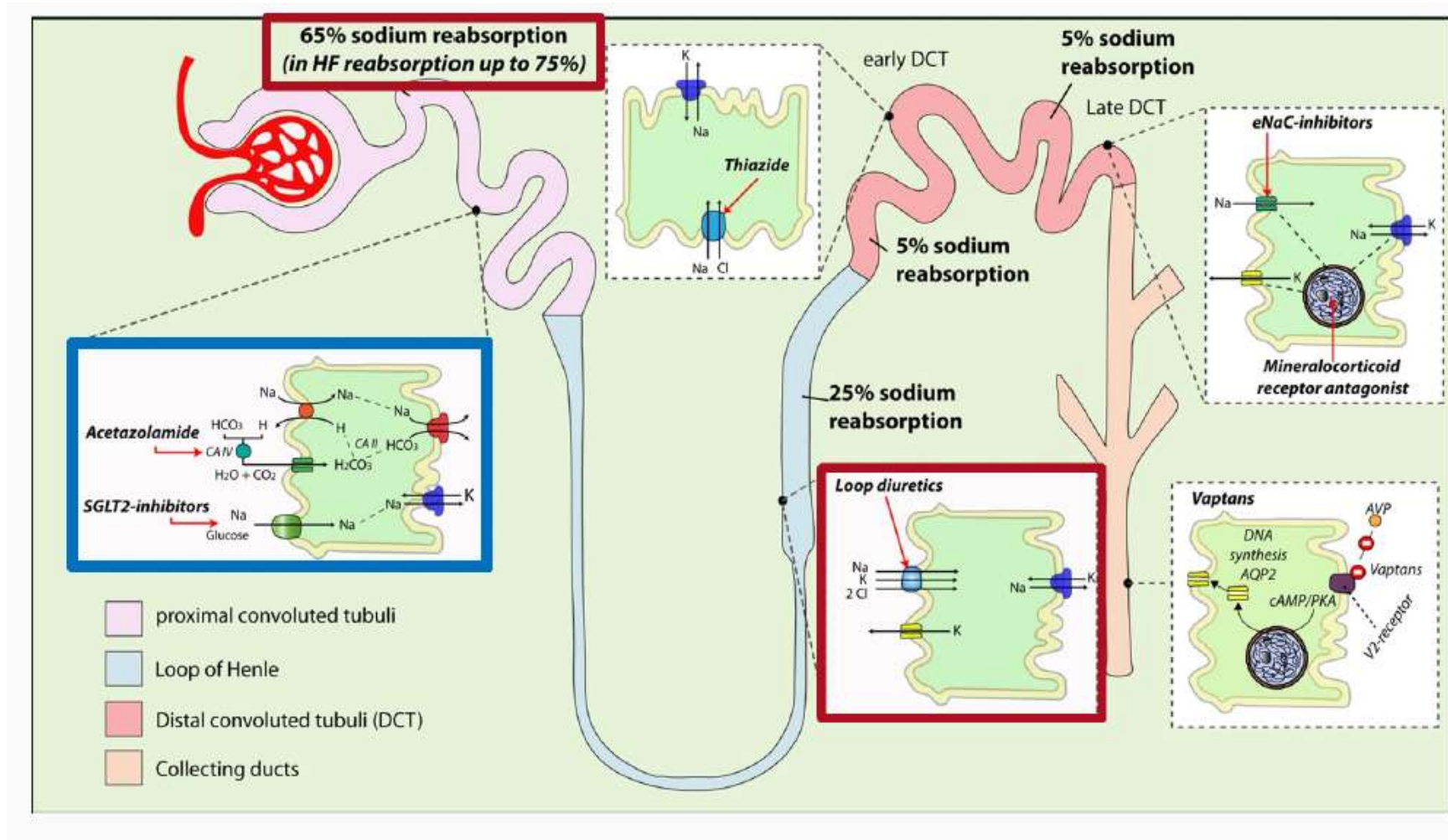
# Comment utiliser les diurétiques dans l'insuffisance cardiaque aiguë ?



Dose maximale de furosémide IV : 400 à 600 mg, jusqu'à 1000 mg si IR sévère

McDonagh T, et al. Eur Heart J 2021;42(36):3599-3726

# Réabsorption sodée au niveau du néphron et localisation de l'action des différents diurétiques





# Acute heart failure

## Acetazolamide in Acute Decompensated Heart Failure with Volume Overload

Wilfried Mullens, M.D., Ph.D., Jeroen Dauw, M.D., Pieter Martens, M.D., Ph.D., Frederik H. Verbrugge, M.D., Ph.D., F  
Tartaglia, M.Sc., Fabien Chenot, M.D., Samer Moubayed, M.D., Riet Dierckx, M.D., Ph.D., Philippe Blouard, M.D., P  
Group\*

Article Figures/Media

24 References 108 Citing Articles Letters

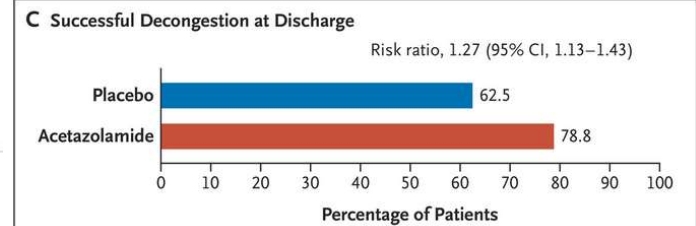
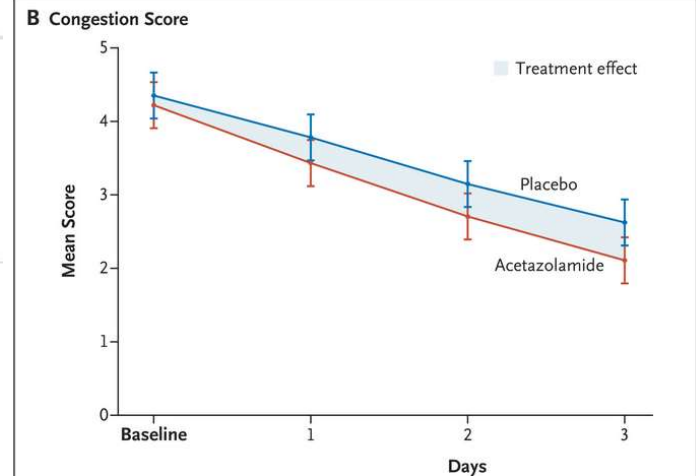
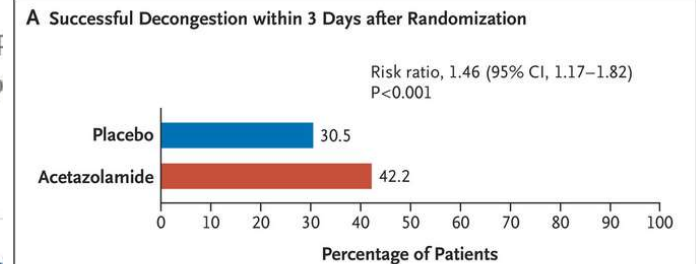
### Abstract

#### BACKGROUND

Whether acetazolamide, a carbonic anhydrase inhibitor that reduces proximal tubular sodium reabsorption, can improve the efficiency of loop diuretics, potentially leading to more and faster decongestion in patients with acute decompensated heart failure with volume overload, is unclear.

#### METHODS

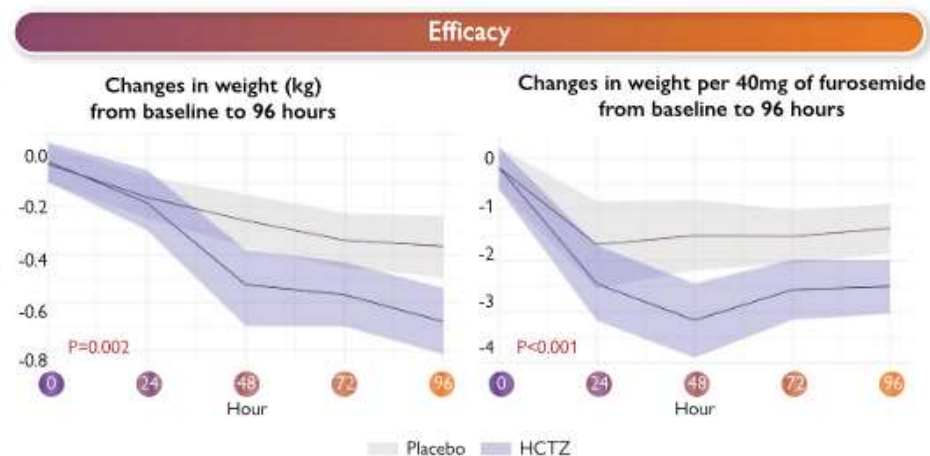
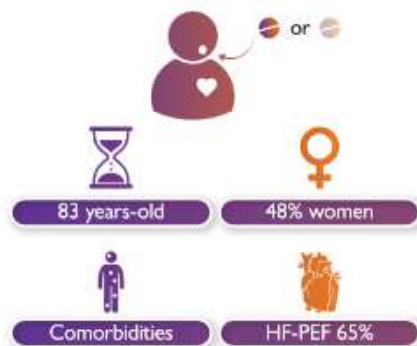
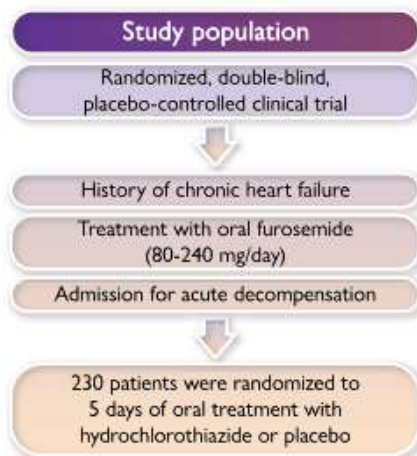
In this multicenter, parallel-group, double-blind, randomized, placebo-controlled trial, we assigned



# Acute heart failure

## Combining loop with thiazide diuretics for decompensated heart failure: the CLOROTIC trial

Joan Carles Trullàs<sup>1,2\*</sup>, José Luis Morales-Rull<sup>3</sup>, Jesús Casado<sup>4</sup>, Margarita Carrera-Izquierdo<sup>5</sup>, Marta Sánchez-Marteles<sup>6</sup>, Alicia Conde



Safety	Placebo	HCTZ	p-value
All-cause mortality at 90 days	19 (16.4%)	23 (20.2%)	0.566
All-cause rehospitalizations at 90 days	40 (34.5%)	43 (37.7%)	0.709
Impaired renal function (serum creatinine and eGFR)	20 (17.2%)	53 (46.5%)	<0.001
Hyponatraemia (Na <sup>+</sup> ≤ 130 mmol/L) - (Na <sup>+</sup> ≤ 125 mmol/L)	6 (5.2%) - 2 (1.7%)	10 (8.8%) - 3 (2.6%)	0.416 - 0.682
Hypokalaemia (K <sup>+</sup> ≤ 3.0 mmol/L) - (K <sup>+</sup> ≤ 2.5 mmol/L)	18 (16.1%) - 0 (0.0%)	43 (40.6%) - 2 (1.8%)	<0.001 - 0.245
Serious adverse events	27 (23.3%)	26 (22.8%)	0.93

# Acute heart failure

## ARTICLES

<https://doi.org/10.1038/s41591-021-01659-1>

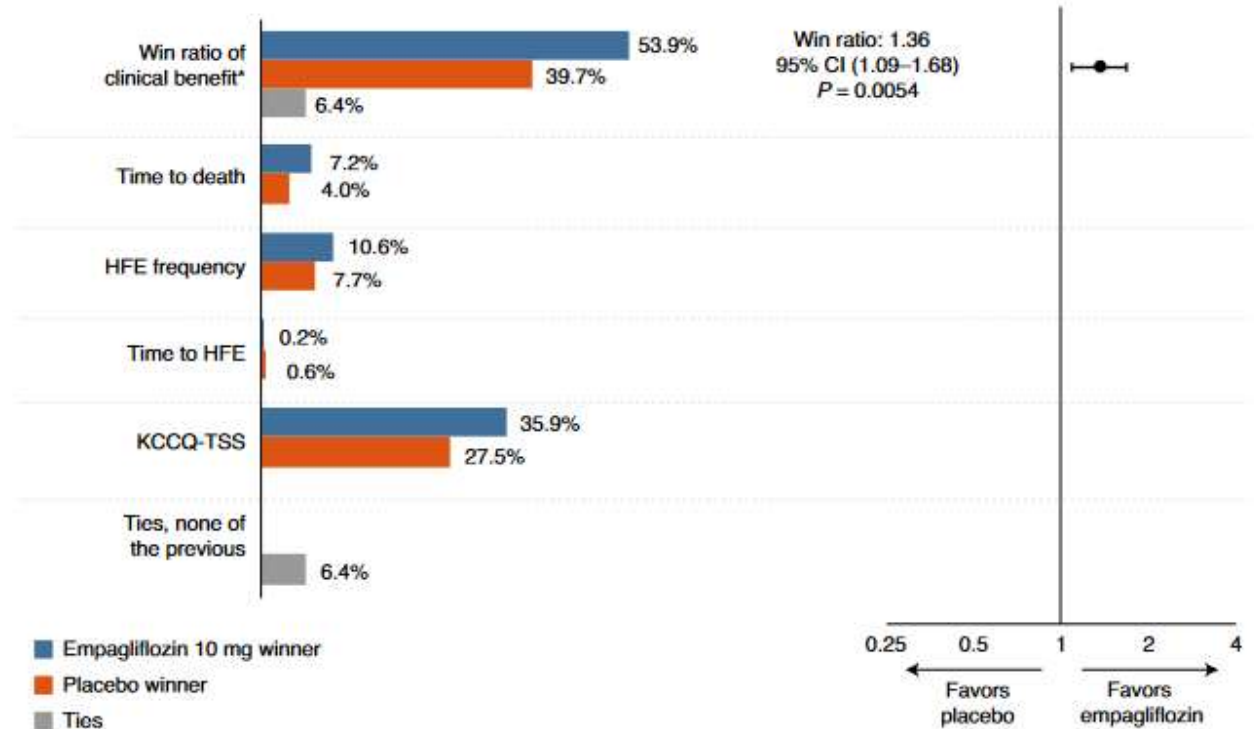
nature  
medicine

Check for updates

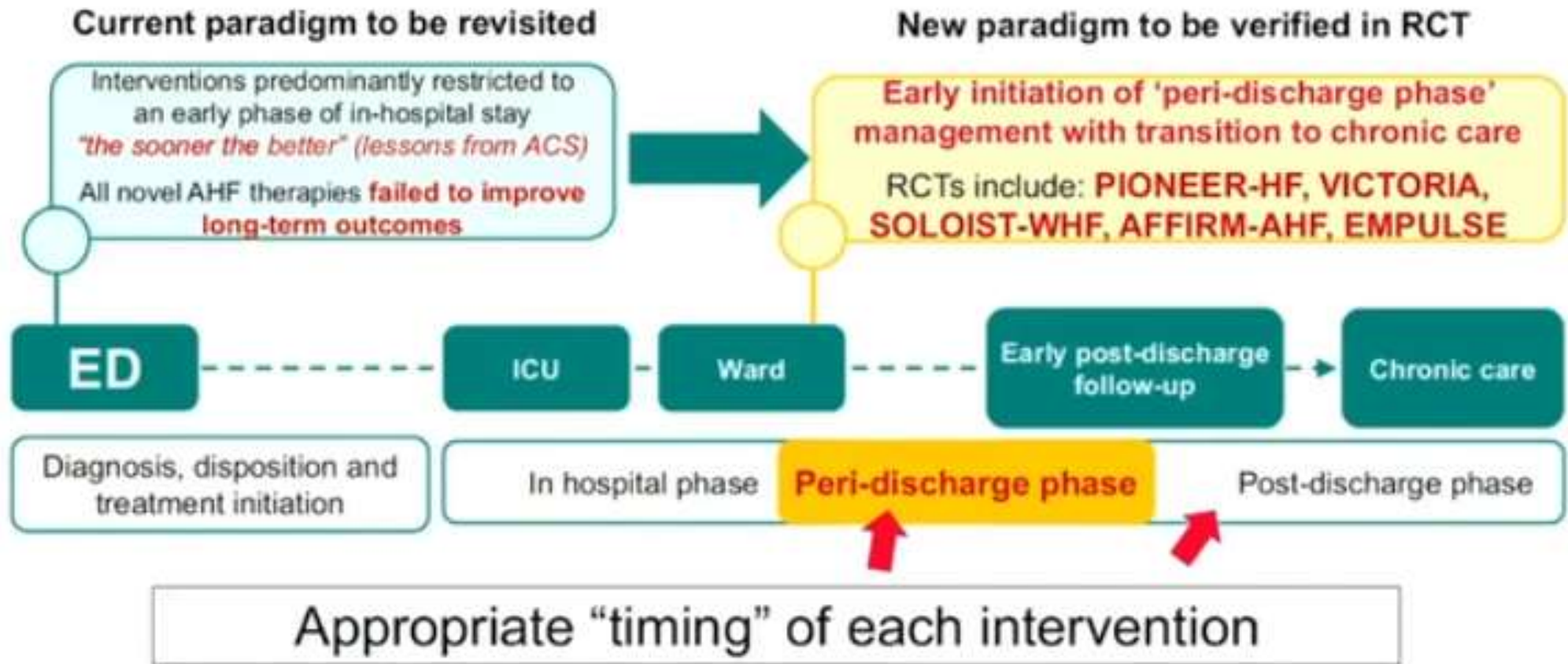
OPEN

## The SGLT2 inhibitor empagliflozin in patients hospitalized for acute heart failure: a multinational randomized trial

The sodium–glucose cotransporter 2 inhibitor empagliflozin reduces the risk of cardiovascular death or heart failure hospitalization in patients with chronic heart failure, but whether empagliflozin also improves clinical outcomes when initiated in patients who are hospitalized for acute heart failure is unknown. In this double-blind trial (EMPULSE; NCT04157751), 530 patients with a primary diagnosis of acute de novo or decompensated chronic heart failure regardless of left ventricular ejection fraction were randomly assigned to receive empagliflozin 10 mg once daily or placebo.



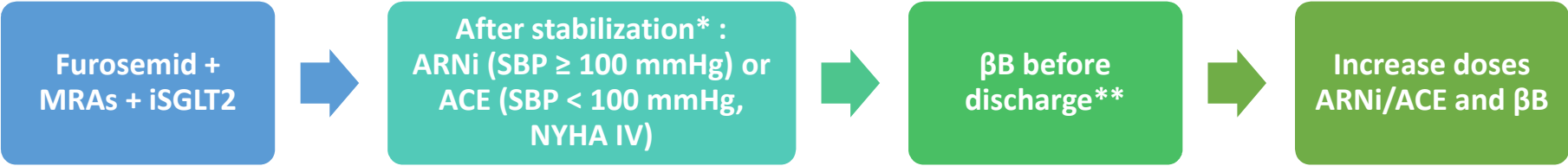
# Acute heart failure





# Comment introduire les différentes classes pharmacologiques essentielles chez un patient présentant une ICFer hospitalisé pour décompensation ?

Diuretic effects of MRAs and SGTL2i participate to decongestion induced by furosemid



\* Meets stabilization criteria :

All of the following criteria must apply for inclusion

- 1** Systolic BP  $\geq 100$  mmHg and no symptoms of hypotension in the preceding **6 hours**
- 2** No increase in IV diuretic dose for **6 hours** prior to randomization
- 3** No IV vasodilators including nitrates within the last **6 hours** prior to randomization
- 4** No IV inotropic drugs for **24 hours** prior to randomization


BP, blood pressure; IV, intravenous.

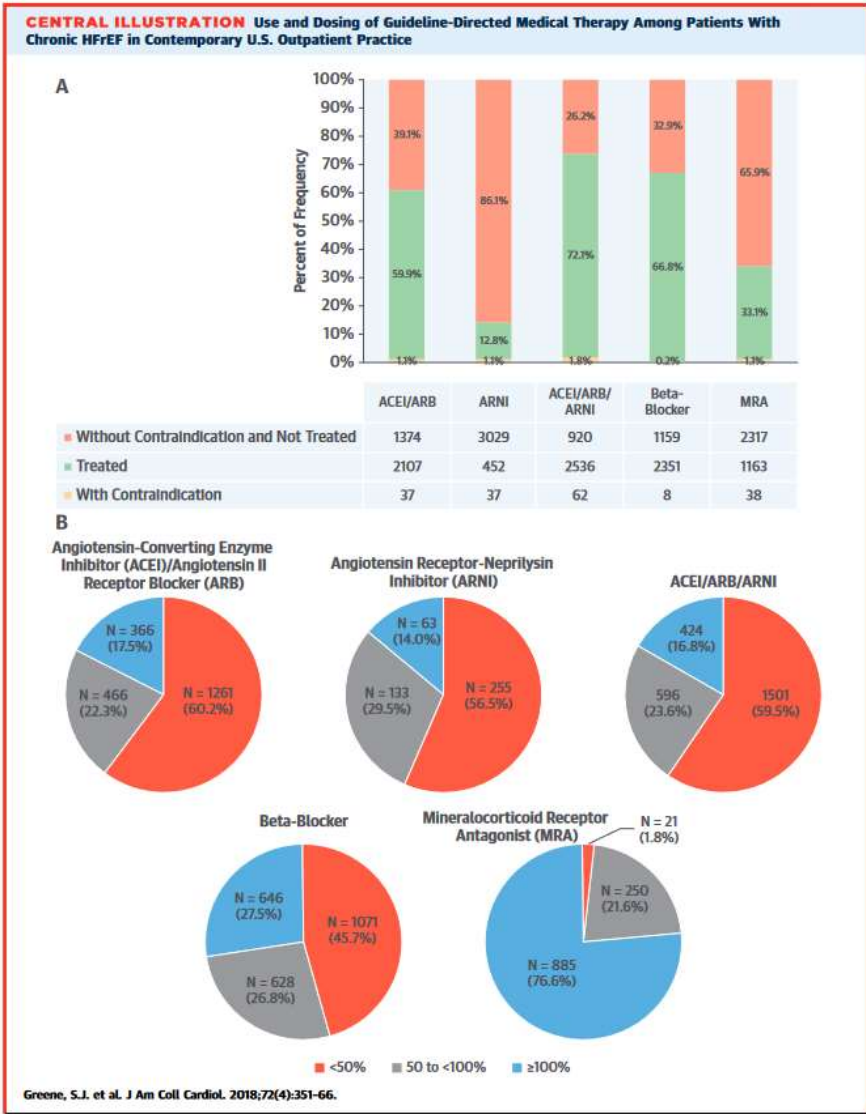
\*\* Introduce early  $\beta$ B if arrhythmic events

**Starting all 4 classes in a short-time period ( $\leq 1$  week) is feasible during hospitalization**

Tromp J, et al. Eur J Heart Fail 2021;23(5):826-834

# Management strategies

  
**Multiple real-life registries show significant gaps in the optimal use and dosing of GDMT, both in outpatients and in patients hospitalized for heart failure<sup>1-11</sup>**



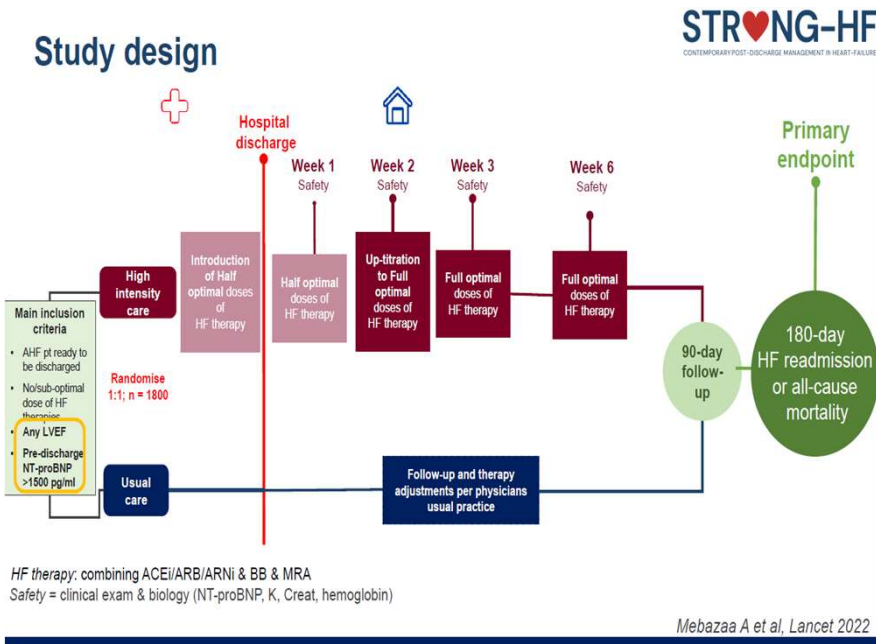
CHAMP registry J Am Coll Cardiol 2018;72:351-66

*Continued on the next page*

# Management strategies

Safety, tolerability, and efficacy of up-titration of guideline-directed medical therapies for acute heart failure (STRONG-HF): a multinational, open-label, randomised, trial

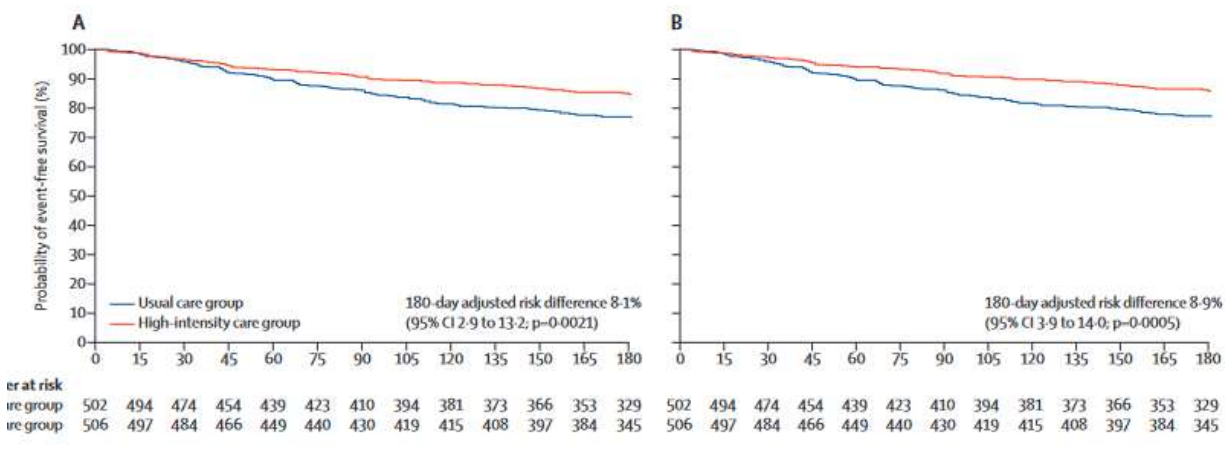
Alexandre Mebazaa, Beth Davison, Ovidiu Chioncel, Alain Cohen-Solal, Rafael Diaz, Gerasimos Filippatos, Marco Metra, Piotr Ponikowski, Karen Sliwa, Adriaan A Voors, Christopher Edwards, Koji Takagi, Albertino Damasceno, Hadiza Saidu, Etienne Gayat, Peter S Pang, Jelena Celutkiene, Gad Cotter



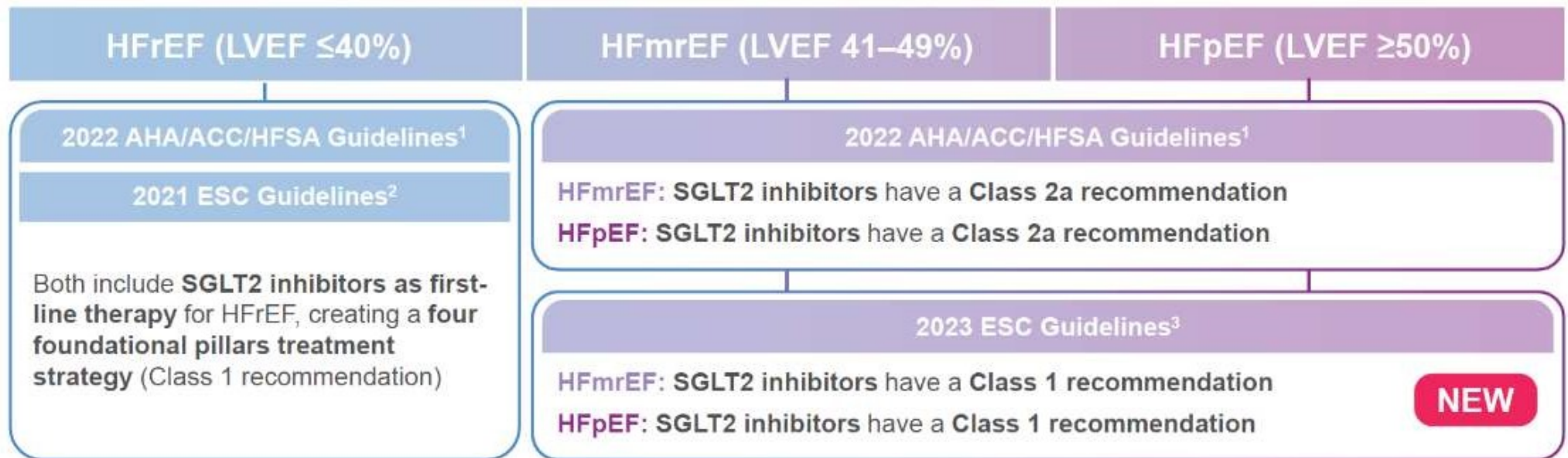
## Recommendation Table 3 — Recommendation for pre-discharge and early post-discharge follow-up of patients hospitalized for acute heart failure

Recommendation	Class <sup>a</sup>	Level <sup>b</sup>
An intensive strategy of initiation and rapid up-titration of evidence-based treatment before discharge and during frequent and careful follow-up visits in the first 6 weeks following a HF hospitalization is recommended to reduce the risk of HF rehospitalization or death. <sup>c,d,e 16</sup>	I	B

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# International guidelines support the use of SGLT2 inhibitors for patients with heart failure regardless of LVEF, including in the hospital setting



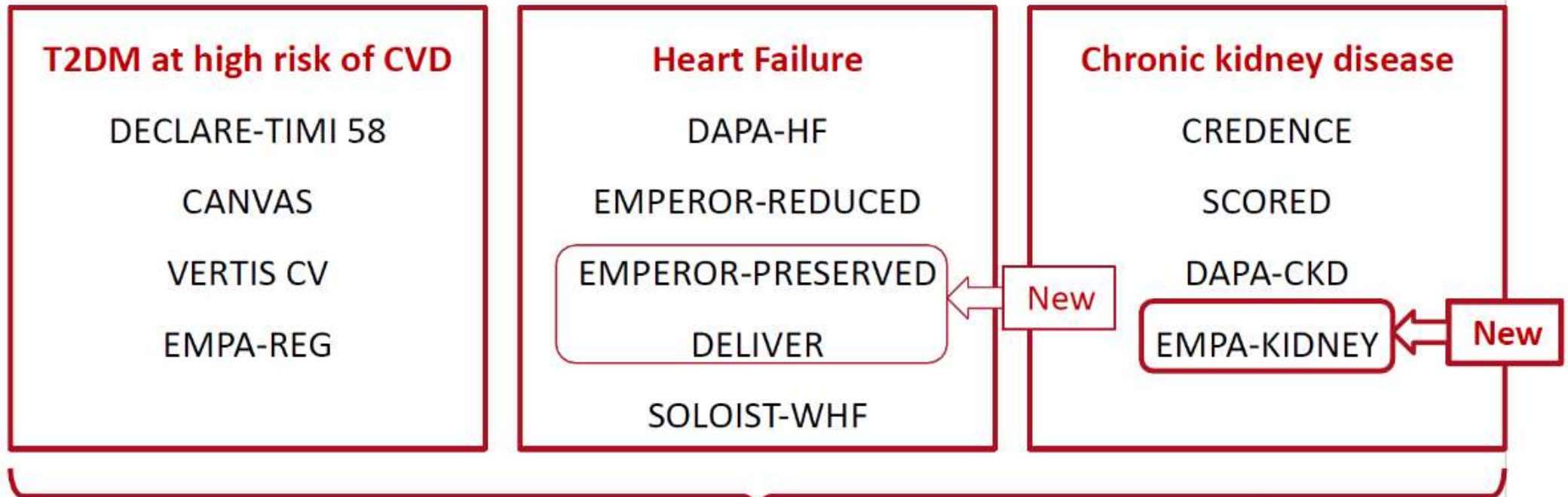
ACC, American College of Cardiology; AHA, American Heart Association; ESC, European Society of Cardiology; HFmrEF, heart failure with mildly reduced ejection fraction; HFpEF, heart failure with preserved ejection fraction; HFrEF, heart failure with reduced ejection fraction; HFSA, Heart Failure Society of America; LVEF, left ventricular ejection fraction; SGLT2, sodium-glucose co-transporter-2.

1. Heidenreich PA *et al.* *J Am Coll Cardiol.* 2022;79:e263; 2. McDonagh T *et al.* *Eur Heart J.* 2021;42:3599; 3. McDonagh T *et al.* *Eur Heart J.* 2023: doi.org/10.1093/eurheartj/ehad195.





# SGLT2 inhibitors trials



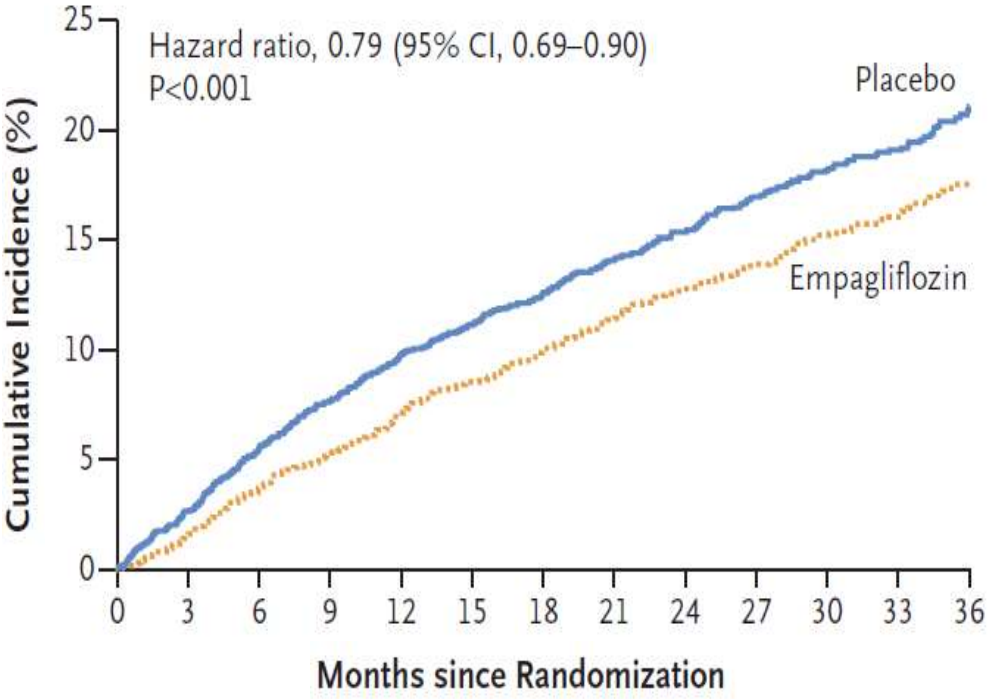
Impact of diabetes on the effects of sodium glucose co-transporter-2 inhibitors on kidney outcomes: collaborative meta-analysis of large placebo-controlled trials



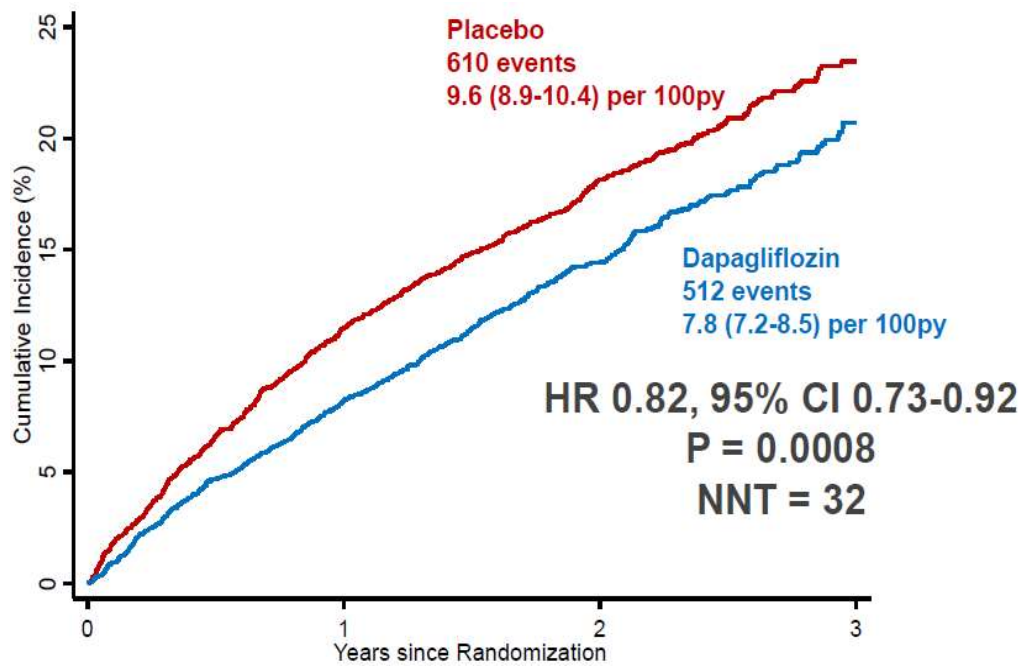
The Nuffield Department of Population Health Renal Studies Group\* and the SGLT2 inhibitor Meta-Analysis Cardio-Renal Trialists' Consortium\*

# Effets des iSGLT2 dans l'ICFEp

### Etude EMPEROR-Preserved Décès cxvx ou hospitalisations pour IC



### Etude DELIVER Décès cxvx ou aggravation IC



Anker SD, et al. N Engl J Med 2021;385(16):1451-1461

Solomon SD, et al. N Engl J Med 2022;387:1089-1098

## IC et comorbidités : diabète, insuffisance rénale, carence martiale

Principales comorbidités dans les essais sur l'ICFep

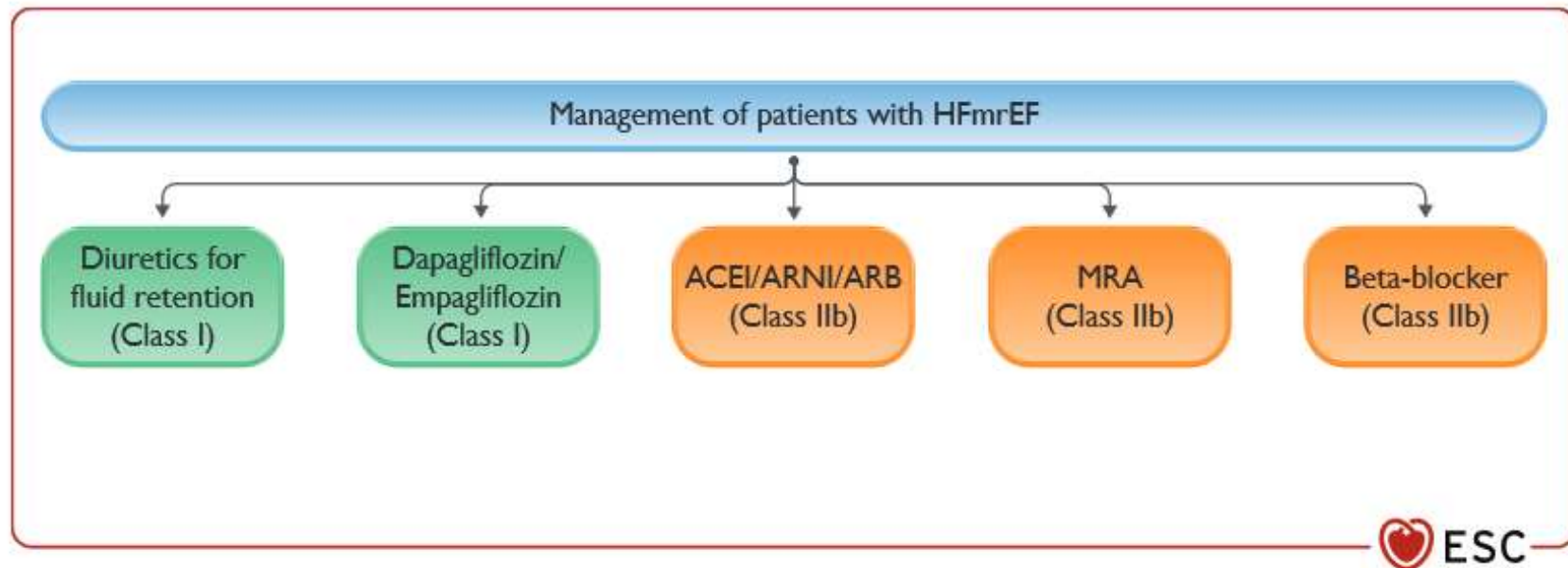
	PARAGON-HF	EMPEROR-Preserved	DELIVER-HF
Age	73 ans	72 ans	72 ans
Sexe féminin	52 %	45 %	44 %
HTA	96 %	90 %	89 %
Diabète	43 %	49 %	45 %
Obésité	49 %	36 %	
FA	32 %	51 %	42 %
Insuffisance rénale	47 %	50 %	49 %

Les comorbidités sont à l'origine de la majorité des décès au cours de l'ICFep en dehors des cardiomyopathies restrictives

# Chronic HF : HFmREF

**Recommendation Table 1** — Recommendation for the treatment of patients with symptomatic heart failure with mildly reduced ejection fraction

Recommendation	Class <sup>a</sup>	Level <sup>b</sup>
An SGLT2 inhibitor (dapagliflozin or empagliflozin) is recommended in patients with HFmREF to reduce the risk of HF hospitalization or CV death. <sup>c 6,8</sup>	I	A



**Figure 1** Management of patients with heart failure with mildly reduced ejection fraction. ACE-I, angiotensin-converting enzyme inhibitor; ARB, angiotensin receptor blocker; ARNI, angiotensin receptor–neprilysin inhibitor; HFmREF, heart failure with mildly reduced ejection fraction; MRA, mineralocorticoid receptor antagonist.



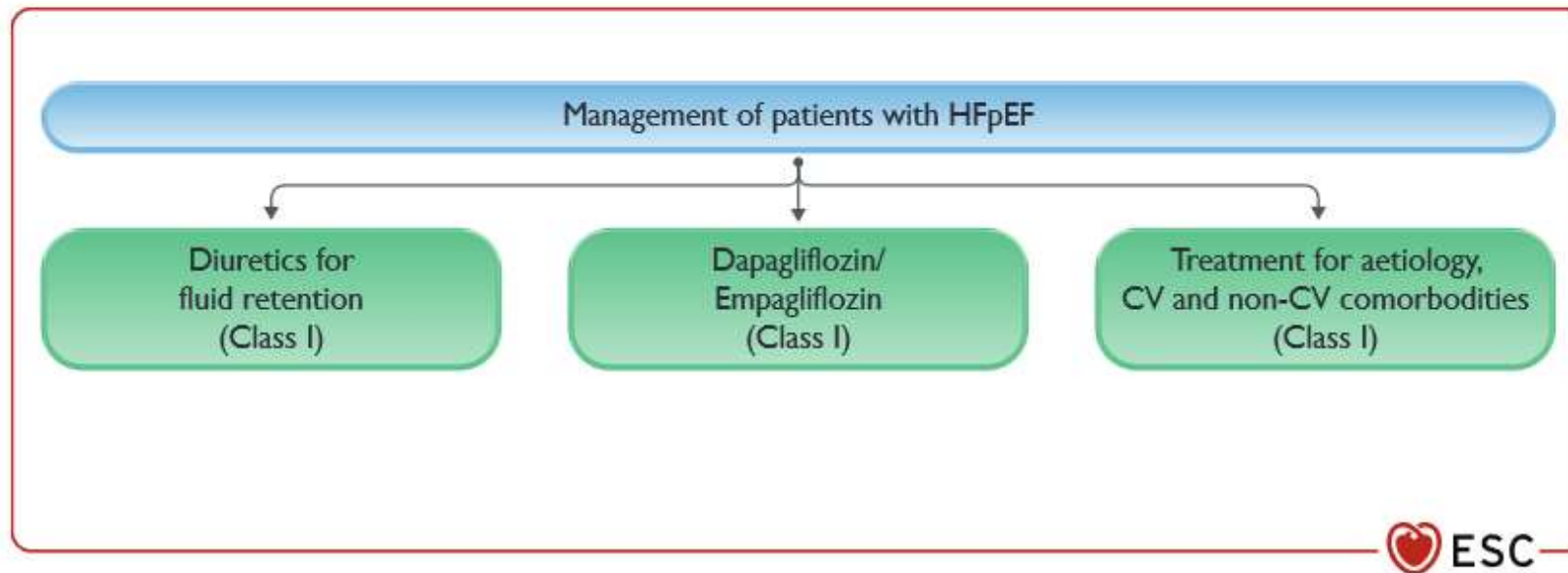
# Chronic HF : HFpEF

## Recommendation Table 2 — Recommendation for the treatment of patients with symptomatic heart failure with preserved ejection fraction

Recommendation	Class <sup>a</sup>	Level <sup>b</sup>
An SGLT2 inhibitor (dapagliflozin or empagliflozin) is recommended in patients with HFpEF to reduce the risk of HF hospitalization or CV death. <sup>c 6,8</sup>	I	A

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CV, cardiovascular; HF, heart failure; HFpEF, heart failure with preserved ejection fraction; SGLT2, sodium–glucose co-transporter 2.  
<sup>a</sup>Class of recommendation.



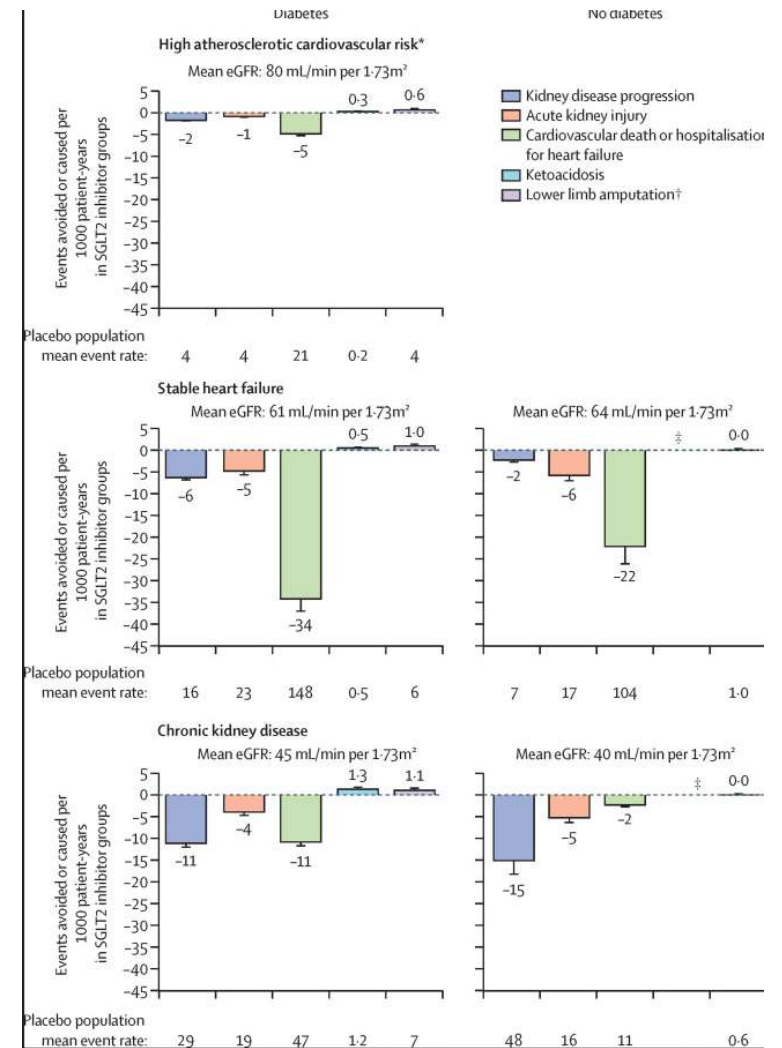
**Figure 2** Management of patients with heart failure with preserved ejection fraction. CV, cardiovascular; HFpEF, heart failure with preserved ejection fraction.

# Comorbidities : Diabetes and HF

## 5.1. Chronic kidney disease and type 2 diabetes mellitus

The 2021 ESC HF Guidelines gave recommendations for the prevention of HF in patients with diabetes. This update provides new recommendations for prevention of HF in patients with chronic kidney disease (CKD) and T2DM.<sup>5,7,10,11,34,35</sup>

Previous trials have shown the effects of ARB in preventing HF events in patients with diabetic nephropathy.<sup>36,37</sup> Both the Kidney Disease: Improving Global Outcomes (KDIGO) and the 2022 American Diabetes Association Standards of Medical Care in Diabetes and KDIGO recommendations indicate treatment with an ACE-I or ARB for patients with CKD, diabetes, and hypertension or albuminuria.<sup>38,39</sup>



Nuffield et al. Lancet 2022;400:1788–801.

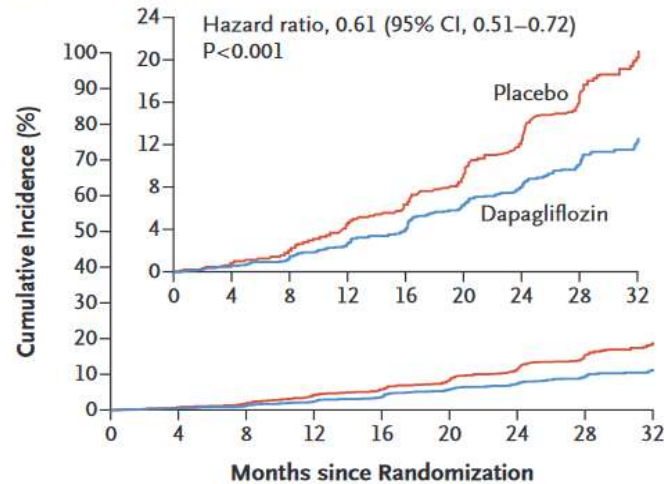
# Les iSGLT2 au cours des maladies rénales chroniques

Progression de la maladie rénale ou décès cardiovasculaires

## DAPA-CKD (46% diabétiques)

Déclin de la fonction rénale ( $\downarrow \geq 50\%$  DFG), maladie rénale terminale, décès de causes rénales ou cardiovasculaires

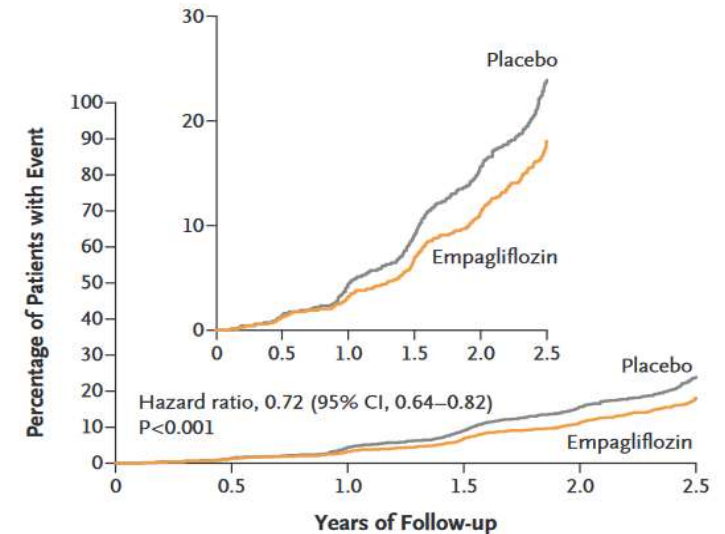
**A Primary Composite Outcome**



No. at Risk		0	4	8	12	16	20	24	28	32
Placebo		2152	1993	1936	1858	1791	1664	1232	774	270
Dapagliflozin		2152	2001	1955	1898	1841	1701	1288	831	309

## EMPA-KIDNEY (67% diabétiques)

Progression maladie rénale ( $\downarrow \geq 40\%$  DFG, DFG < 10 mL/min), décès de causes rénales ou cardiovasculaires



No. at Risk		0	0.5	1.0	1.5	2.0	2.5
Placebo		3305	3250	3129	2243	1496	592
Empagliflozin		3304	3252	3163	2275	1538	624

**Chez les patients atteints de MRC, diabétiques ou non, la dapagliflozine et l'empagliflozine à 10 mg améliorent le pronostic rénal et cardiovasculaire**

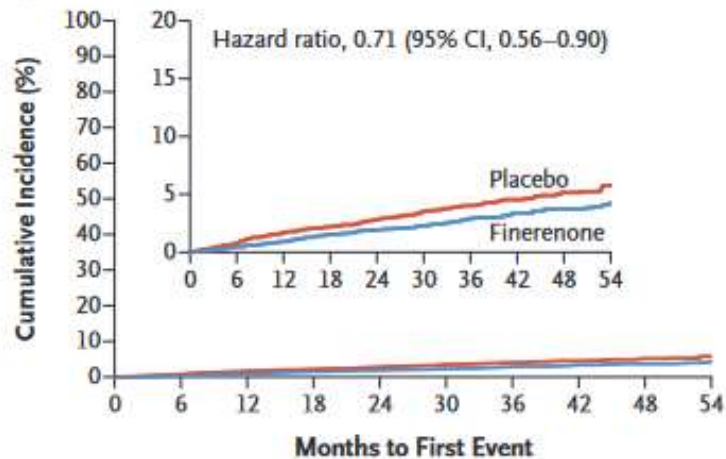
# Comorbidities : Diab and CKD

ORIGINAL ARTICLE

## Cardiovascular Events with Finerenone in Kidney Disease and Type 2 Diabetes

B. Pitt, G. Filippatos, R. Agarwal, S.D. Anker, G.L. Bakris, P. Rossing, A. Joseph, P. Kolkhof, C. Nowack, P. Schloemer, and L.M. Ruilope, for the FIGARO-DKD Investigators\*

### E Hospitalization for Heart Failure



#### No. at Risk

Placebo	3666	3610	3538	3471	3376	2849	2239	1751	1134	619
Finerenone	3686	3640	3581	3515	3429	2887	2284	1790	1142	629

### Recommendation Table 4 — Recommendations for the prevention of heart failure in patients with type 2 diabetes mellitus and chronic kidney disease

Recommendations	Class <sup>a</sup>	Level <sup>b</sup>
In patients with T2DM and CKD, <sup>c</sup> SGLT2 inhibitors are recommended to reduce the risk of HF hospitalization or CV death. <sup>35</sup>	I	A
In patients with T2DM and CKD, <sup>c</sup> finerenone is recommended to reduce the risk of HF hospitalization. <sup>10,11,34,40</sup>	I	A



ESC 2023 : Traitement de la carence martiale chez les patients avec ICFEr ou mr (FE < 50 %)

Recommendations	Class <sup>a</sup>	Level <sup>b</sup>
Intravenous iron supplementation is recommended in symptomatic patients with HFrEF and HFmrEF, and iron deficiency, to alleviate HF symptoms and improve quality of life. <sup>c 12,41,47–49</sup>	<b>I</b>	<b>A</b>
Intravenous iron supplementation with ferric carboxymaltose or ferric derisomaltose should be considered in symptomatic patients with HFrEF and HFmrEF, and iron deficiency, to reduce the risk of HF hospitalization. <sup>c 12,41,43–46</sup>	<b>IIa</b>	<b>A</b>

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# Comorbidities : iron deficiency

Intravenous ferric derisomaltose in patients with heart failure and iron deficiency in the UK (IRONMAN): an investigator-initiated, prospective, randomised, open-label, blinded-endpoint trial

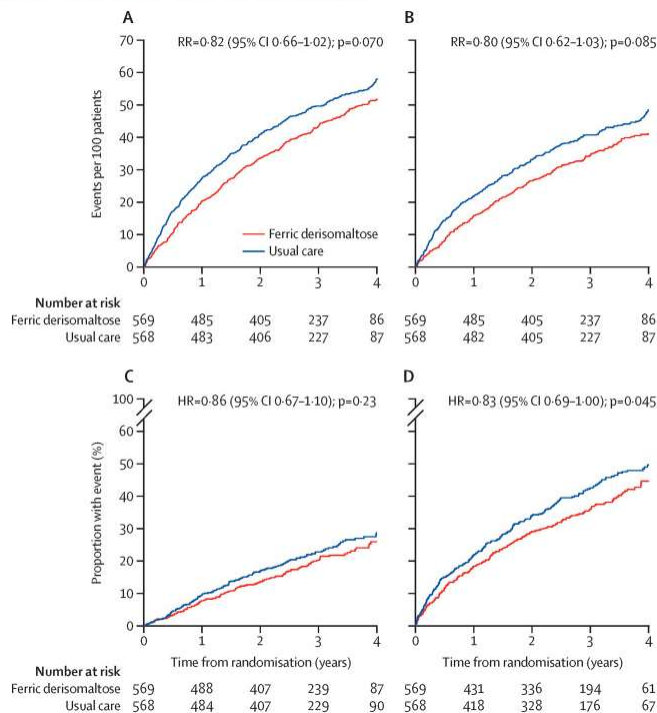
Paul R Kalra, John G F Cleland, Mark C Petrie, Elizabeth A Thomson, Philip A Kalra, Iain B Squire, Fozia Z Ahmed, Abdallah Al-Mohammad, Peter J Cowburn, Paul W X Foley, Fraser J Graham, Alan G Japp, Rebecca E Lane, Ninian N Lang, Andrew J Ludman, Iain C Macdougall, Pierpaolo Pellicori, Robin Ray, Michele Robertson, Alison Seed, Ian Ford, for the IRONMAN Study Group\*

## Summary

**Background** For patients with heart failure, reduced left ventricular ejection fraction and iron deficiency, intravenous ferric carboxymaltose administration improves quality of life and exercise capacity in the short-term and reduces hospital admissions for heart failure up to 1 year. We aimed to evaluate the longer-term effects of intravenous ferric derisomaltose on cardiovascular events in patients with heart failure.



Lancet 2022; 400: 2199–209  
Published Online  
November 5, 2022  
[https://doi.org/10.1016/S0140-6736\(22\)02083-9](https://doi.org/10.1016/S0140-6736(22)02083-9)



## Recommendation Table 5 — Recommendations for the management of iron deficiency in patients with heart failure

Recommendations	Class <sup>a</sup>	Level <sup>b</sup>
Intravenous iron supplementation is recommended in symptomatic patients with HFrEF and HFmrEF, and iron deficiency, to alleviate HF symptoms and improve quality of life. <sup>c</sup> 12,41,47–49	I	A
Intravenous iron supplementation with ferric carboxymaltose or ferric derisomaltose should be considered in symptomatic patients with HFrEF and HFmrEF, and iron deficiency, to reduce the risk of HF hospitalization. <sup>c</sup> 12,41,43–46	IIa	A

HF, heart failure; HFmrEF, heart failure with mildly reduced ejection fraction; HFrEF, heart failure with reduced ejection fraction.

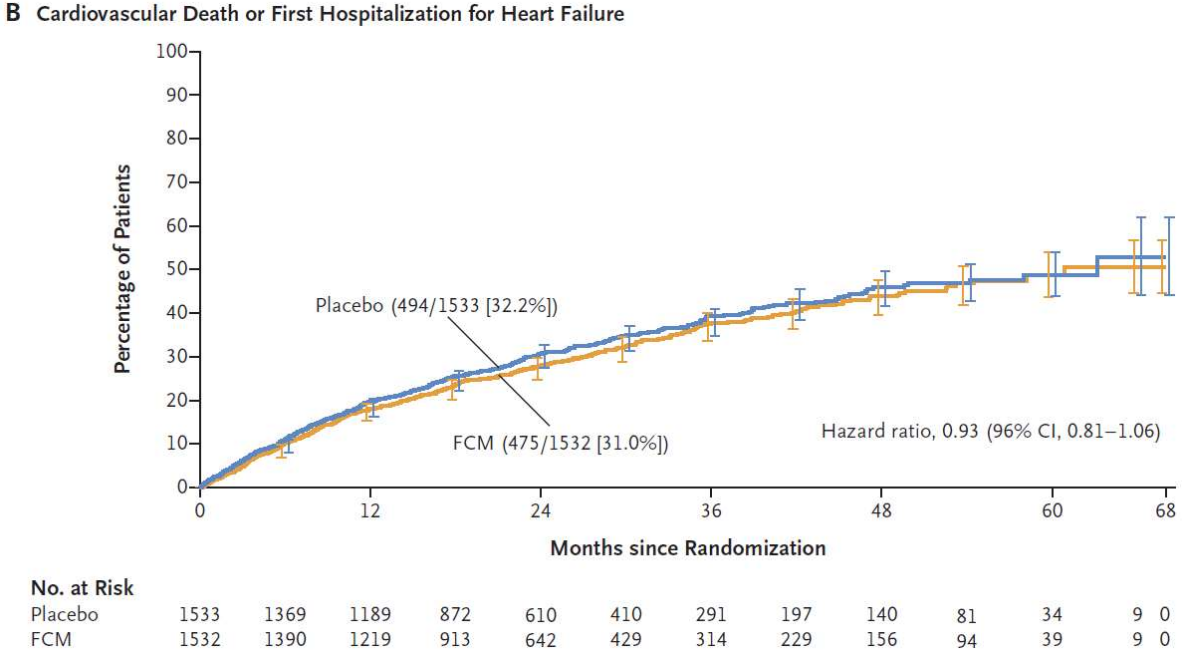
<sup>a</sup>Class of recommendation.

<sup>b</sup>Level of evidence.

<sup>c</sup>Most of the evidence refers to patients with left ventricular ejection fraction  $\leq 45\%$ .

# Etude HEART-FID : effets du carboxymaltose ferrique IV sur les hospitalisations pour insuffisance cardiaque et les décès cardiovasculaires chez les patients avec ICFEr chronique (FE ≤ 40 %) carencés en fer

Etude randomisée, en double aveugle, de 3065 pts traités par fer carboxymaltose tous les 6 mois si nécessaire, vs placebo, suivis 12 mois



La correction de la carence martiale par voie IV n’a diminué ni l’incidence des hospitalisations pour insuffisance cardiaque, ni celle des décès cardio-vasculaires dans l’ICFER chronique stable

Mentz RJ, et al. N Engl J Med 2023;August 26th. <https://doi.org/10.1056/NEJMoa2304968>

# Méta-analyse des données individuelles des patients traités par fer carboxymaltose IV pour une carence martiale au cours ICFer et mr dans les essais CONFIRM-HF, AFFIRM-AHF, HEART-FID sur les hospitalisations et la mortalité

4501 pts, suivi : 52 semaines

	RR	IC 50 %	P
Critères primaires			
Hospitalisations cxvx et décès cxvx	0,86	0,75-0,98	0,029
Hospitalisations pour IC et décès cxvx	0,87	0,75-1,01	0,076
Critères secondaires			
Hospitalisations cxvx	0,83	0,73-0,96	0,009
Hospitalisations pour IC	0,84	0,71-0,98	0,025
Décès cxvx	0,97	0,80-1,17	0,72

La correction de la carence martiale dans l'ICFer et mr par du fer carboxymaltose semble diminuer le risque d'hospitalisation cxvx et pour IC mais pas la mortalité cxvx, avec des effets d'autant plus marqués que le CST (< 15 %) et l'hémoglobine sont bas.





**ESC**

European Society  
of Cardiology


European Heart Journal (2023) **44**, 3503–3626

<https://doi.org/10.1093/eurheartj/ehad194>

**ESC GUIDELINES**

# **2023 ESC Guidelines for the management of cardiomyopathies**

**Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC)**

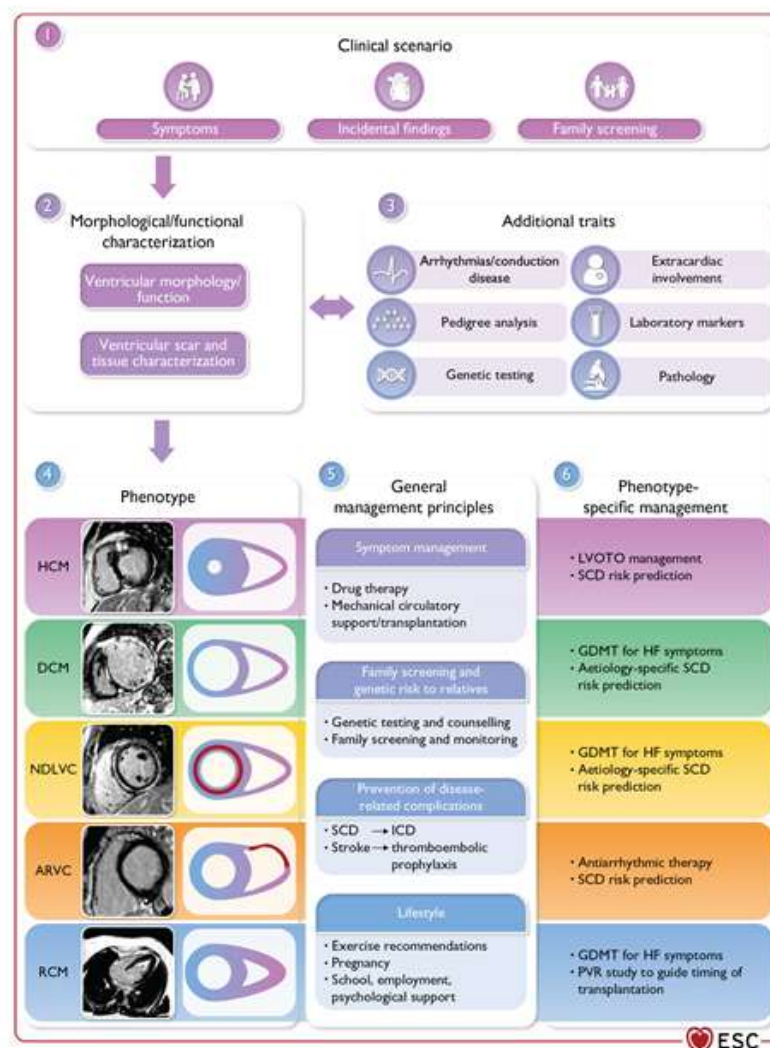
**Authors/Task Force Members: Elena Arbelo  \*<sup>†</sup>, (Chairperson) (Spain), Alexandros Protonotarios  <sup>‡</sup>, (Task Force Co-ordinator) (United Kingdom), Juan R. Gimeno  <sup>‡</sup>, (Task Force Co-ordinator) (Spain), Eloisa Arbustini  (Italy), Roberto Barriales-Villa  (Spain), Cristina Basso  (Italy), Connie R. Bezzina  (Netherlands), Elena Biagini  (Italy), Nico A. Blom<sup>1</sup> (Netherlands),**

## Definitions



A **cardiomyopathy** is defined as *'a myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of coronary artery disease (CAD), hypertension, valvular disease, and congenital heart disease (CHD) sufficient to cause the observed myocardial abnormality'*.

**Figure 1**  
**Central illustration.**  
**Key aspects in the**  
**evaluation and**  
**management of**  
**cardiomyopathies**





# Morphological and functional traits used to describe cardiomyopathy phenotypes



## Morphological traits

Ventricular hypertrophy: left and/or right











Ventricular dilatation: left and/or right

Non-ischæmic ventricular scar and other myocardial tissue characterization features on cardiac magnetic resonance

## Functional traits

Ventricular systolic dysfunction (global, regional)

Ventricular diastolic dysfunction (restrictive physiology)

Phenotype	General management principles	Phenotype-specific management
<b>HCM</b>  	<b>Symptom management</b> <ul style="list-style-type: none"> <li>• Drug therapy</li> <li>• Mechanical circulatory support/transplantation</li> </ul>	<ul style="list-style-type: none"> <li>• LVOTO management</li> <li>• SCD risk prediction</li> </ul>
<b>DCM</b>  	<b>Family screening and genetic risk to relatives</b> <ul style="list-style-type: none"> <li>• Genetic testing and counselling</li> <li>• Family screening and monitoring</li> </ul>	<ul style="list-style-type: none"> <li>• GDMT for HF symptoms</li> <li>• Aetiology-specific SCD risk prediction</li> </ul>
<b>NDLVC</b>  	<b>Prevention of disease-related complications</b> <ul style="list-style-type: none"> <li>• SCD → ICD</li> <li>• Stroke → thromboembolic prophylaxis</li> </ul>	<ul style="list-style-type: none"> <li>• GDMT for HF symptoms</li> <li>• Aetiology-specific SCD risk prediction</li> </ul>
<b>ARVC</b>  	<b>Lifestyle</b> <ul style="list-style-type: none"> <li>• Exercise recommendations</li> <li>• Pregnancy</li> <li>• School, employment, psychological support</li> </ul>	<ul style="list-style-type: none"> <li>• Antiarrhythmic therapy</li> <li>• SCD risk prediction</li> </ul>
<b>RCM</b>  	<ul style="list-style-type: none"> <li>• Exercise recommendations</li> <li>• Pregnancy</li> <li>• School, employment, psychological support</li> </ul>	<ul style="list-style-type: none"> <li>• GDMT for HF symptoms</li> <li>• PVR study to guide timing of transplantation</li> </ul>



## Key epidemiological metrics in adults and children for the different cardiomyopathy phenotypes

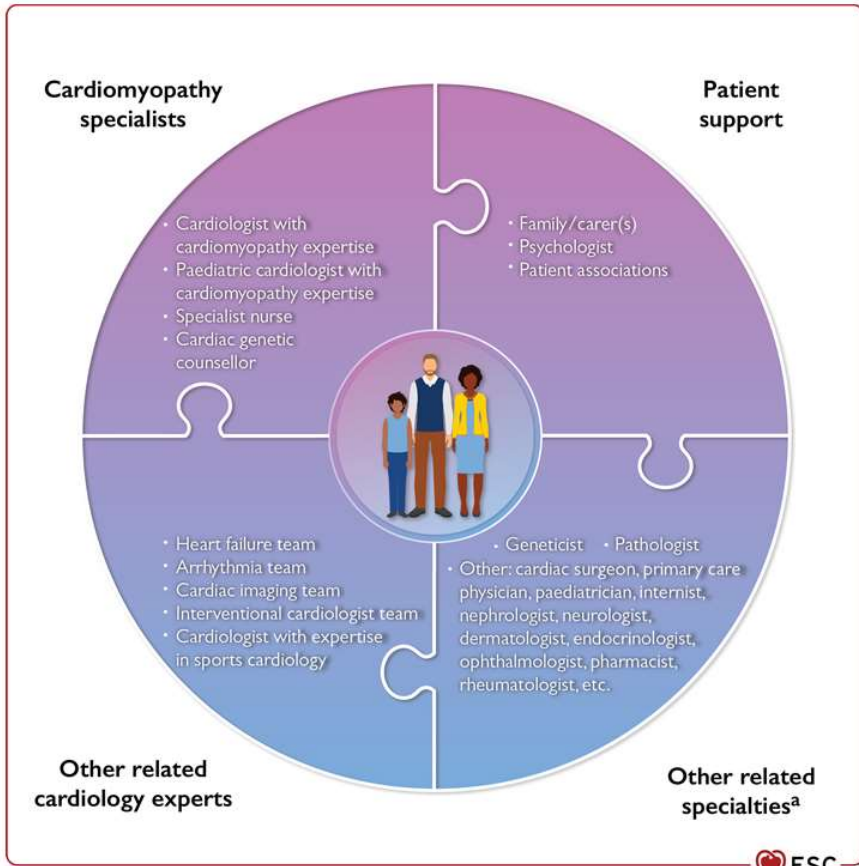


Cardiomyopathy phenotype	Adults	Children
HCM	Prevalence: 0.2%	Childhood incidence: 0.002–0.005% Childhood prevalence: 0.029%
DCM	Prevalence: 0.036–0.400%	Childhood incidence: 0.003–0.006% Childhood prevalence: 0.026% Infantile incidence: 0.038–0.046%
NDLVC	To be determined	To be determined
ARVC	Prevalence: 0.078%	Very rare in infancy and early childhood; to be determined in older children and adolescents
RCM	Rare	Childhood incidence: 0.0003%

## Special populations

- Alcoholic cardiomyopathy
- Cancer therapy-induced cardiomyopathy
- Péripartum cardiomyopathy
- Acute myocarditis
- Anderson Fabry disease in LVH and HCM
- Amyloidosis in severe aortic cardiomyopathy, HFpEF
- Left ventricular hypertrabeculation (left ventricular non-compaction)
- Takotsubo syndrome

# Multidisciplinary care of cardiomyopathies

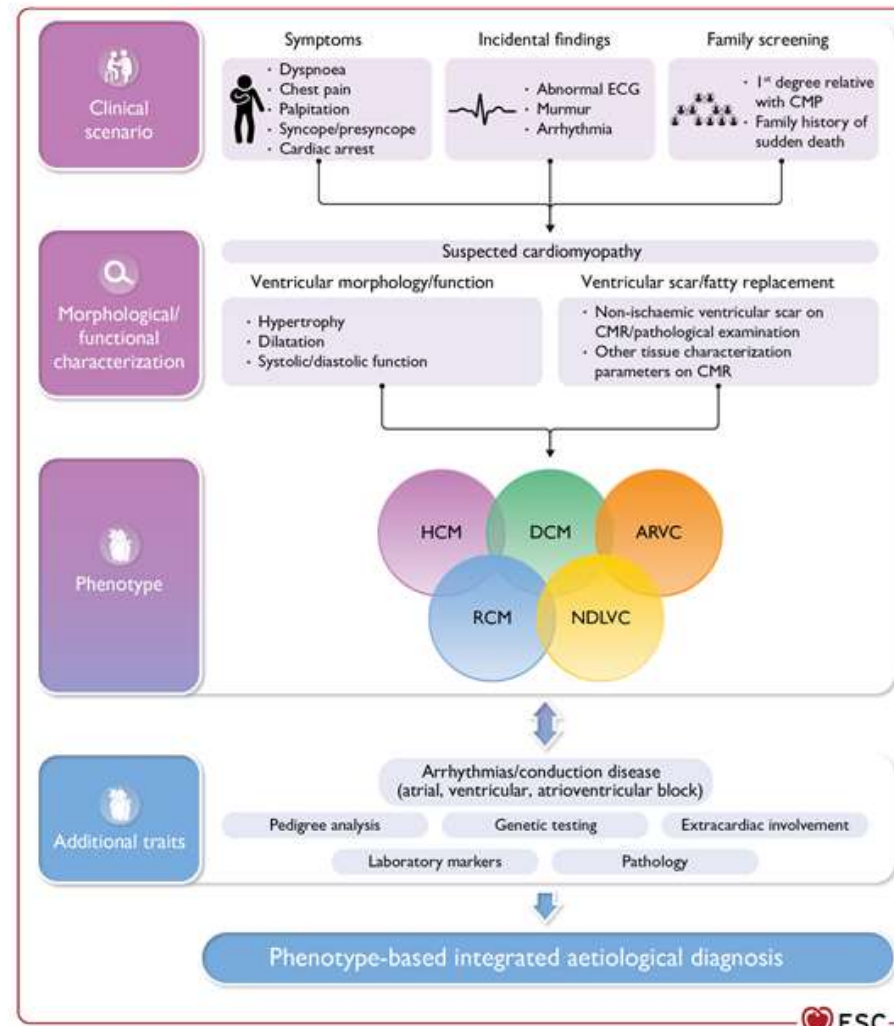


Recommendations	Class	Level
It is recommended that all patients with cardiomyopathy and their relatives have access to <b>multidisciplinary teams</b> with expertise in the diagnosis and management of cardiomyopathies.	I	C
Timely and adequate preparation for <b>transition of care from paediatric to adult services</b> , including joint consultations, is recommended in all adolescents with cardiomyopathy.	I	C

**A shared and coordinated care approach between cardiomyopathy specialists and general adult and paediatric cardiology centres is strongly recommended**

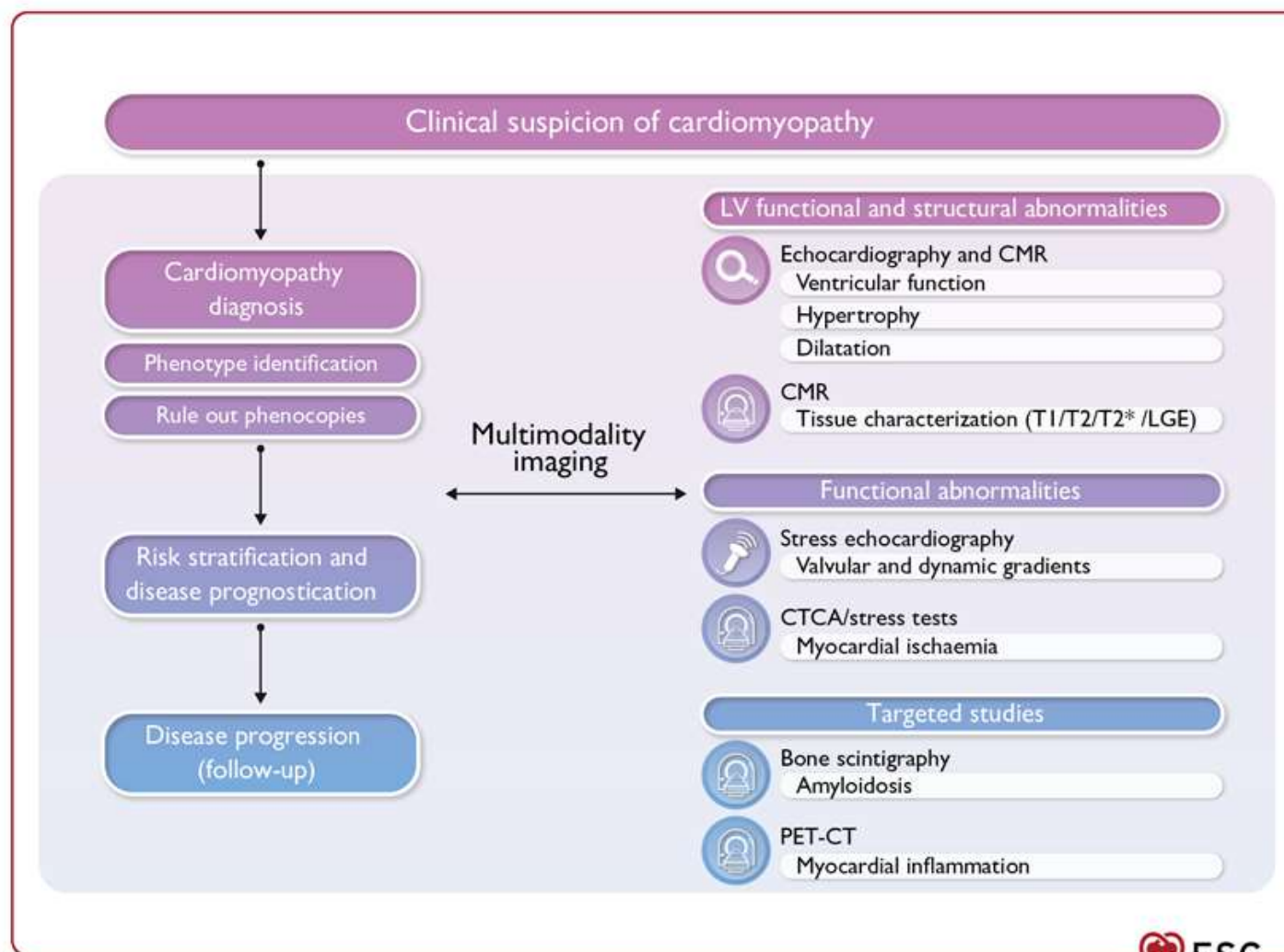
**Figure 2**  
**Clinical diagnostic workflow of cardiomyopathy**

Recommendations	Class	Level
It is recommended that all patients with suspected or established cardiomyopathy undergo <b>systematic evaluation using a multiparametric approach</b> that includes clinical evaluation, pedigree analysis, ECG, Holter monitoring, laboratory tests, and multimodality imaging.	I	C
It is recommended that all patients with suspected cardiomyopathy undergo evaluation of <b>family history</b> and that a <b>three- to four-generation family tree</b> is created to aid in diagnosis, provide clues to underlying aetiology, determine inheritance pattern, and identify at-risk relatives.	I	C





**Figure 6**  
**Multimodality**  
**imaging process in**  
**cardiomyopathies**





## Recommendations for cardiac magnetic resonance indication in patients with cardiomyopathy

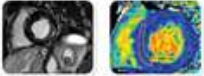
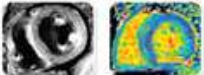












Recommendations	Class	Level
Contrast-enhanced CMR is recommended in patients with cardiomyopathy at initial evaluation.	I	B
Contrast-enhanced CMR should be considered in patients with cardiomyopathy during follow-up to monitor disease progression and aid risk stratification and management.	IIa	C
Contrast-enhanced CMR should be considered for the serial follow-up and assessment of therapeutic response in patients with cardiac amyloidosis, Anderson–Fabry disease, sarcoidosis, inflammatory cardiomyopathies, and haemochromatosis with cardiac involvement.	IIa	C
In families with cardiomyopathy in which a disease-causing variant has been identified, contrast-enhanced CMR should be considered in genotype-positive/phenotype-negative family members to aid diagnosis and detect early disease.	IIa	B
In cases of familial cardiomyopathy without a genetic diagnosis, contrast-enhanced CMR may be considered in phenotype-negative family members to aid diagnosis and detect early disease.	IIb	C

©ESC

**Figure 7**

Examples of cardiac magnetic resonance imaging tissue characterization features that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype

Cardiomyopathy phenotype	Finding	Cardiac CMR examples	Specific diseases to be considered
HCM	Posterolateral LGE and concentric LVH Low native T1		Anderson-Fabry disease
	Diffuse subendocardial LGE, high native T1		Amyloidosis
	Patchy mid-wall in hypertrophied areas		Sarcomeric HCM
DCM	Short T2*		Haemochromatosis
	Subepicardial LGE		Post-myocarditis
	Lateral wall epicardial LGE		Dystrophinopathy
	Subepicardial and midwall LGE at basal septum +/- extension into inferolateral wall and RV insertion points		Sarcoidosis
	Apical transmural LGE		Chagas disease
NDLVC	Ring-like and/or subepicardial LGE pattern		DSP variants FLNC variants DES variants
	Septal mid-wall LGE		Laminopathy
ARVC	Fat and LGE (transmural RV plus sub-epicardial-midmural LV free wall)		Desmosomal variants
RCM	Partial LV or RV apical obliteration + LGE at endocardial level		EMF/hypereosinophilia

## Recommendations for genetic counselling and testing in cardiomyopathies (1)

Recommendations	Class	Level
<b><i>Genetic counselling</i></b>		
Genetic counselling, provided by an appropriately trained healthcare professional and including genetic education to inform decision-making and psychosocial support, is recommended for families with an inherited or suspected inherited cardiomyopathy, regardless of whether genetic testing is being considered.	I	B
It is recommended that genetic testing for cardiomyopathy is performed with access to a multidisciplinary team, including those with expertise in genetic testing methodology, sequence variant interpretation, and clinical application of genetic testing, typically in a specialized cardiomyopathy service or in a network model with access to equivalent expertise.	I	B
Pre- and post-test genetic counselling is recommended in all individuals undergoing genetic testing for cardiomyopathy.	I	B



## Examples of inheritance patterns that should raise the suspicion of specific genetic aetiologies, grouped according to cardiomyopathy phenotype (1)

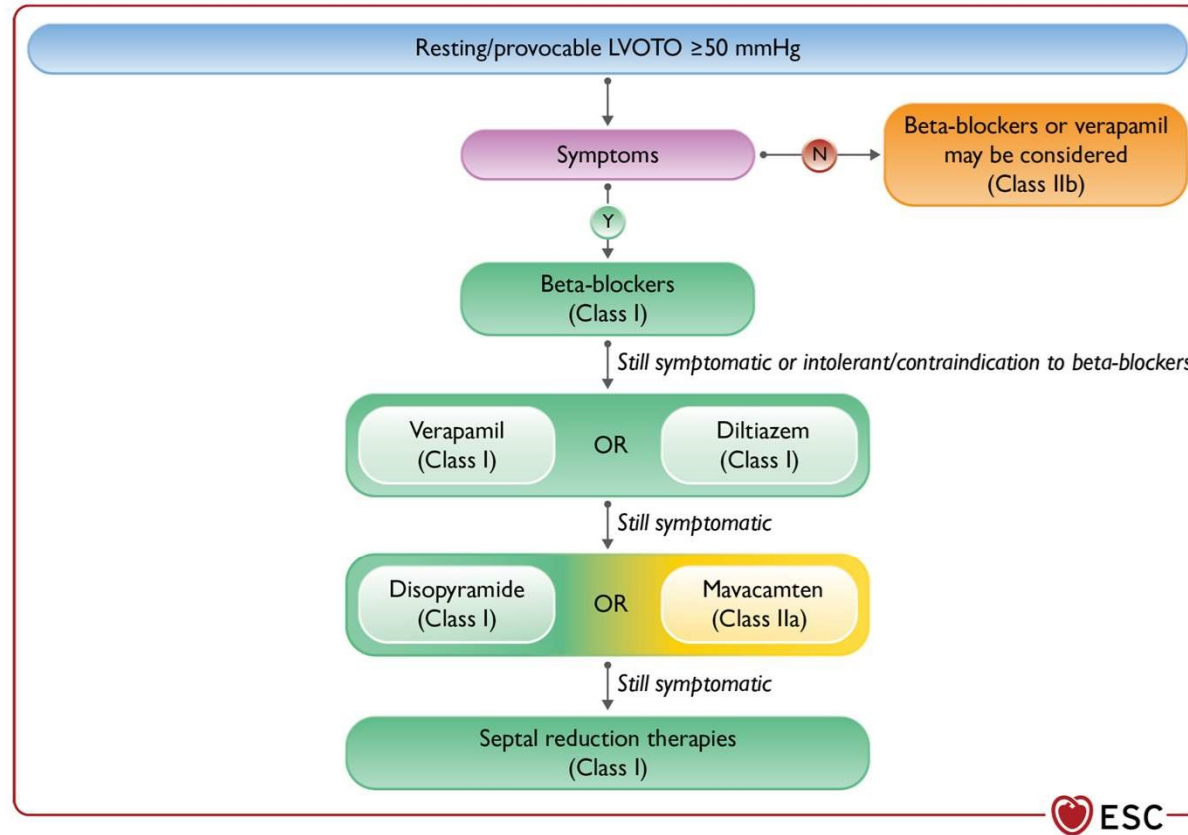
Cardiomyopathy phenotype		AD	AR	X-linked	Matrilineal
HCM	Sarcomeric	X			
	Anderson–Fabry			X	
	Danon			X	
	TTR amyloidosis	X			
	RASopathy	X	(X)		
	Friedreich ataxia		X		
	Mitochondrial				
	Mitochondrial DNA				X
	Nuclear DNA	X	X	X	



# Traitement des cardiomyopathies obstructives



## Flowchart on the management of left ventricular outflow tract obstruction



### Place des inhibiteurs de la myosine

- En association aux bêtabloquants ou aux ICA<sup>++</sup> : IIa, A
- En cas de CI ou d'intolérance aux bêtabloquants, aux ICA<sup>++</sup>, au disopyramide : IIa, B

# Effet de la diminution du gradient de pression intraventriculaire gauche au cours des CMH obstructives sous macavamten

## Etude EXPLORER-HCM (429 pts)

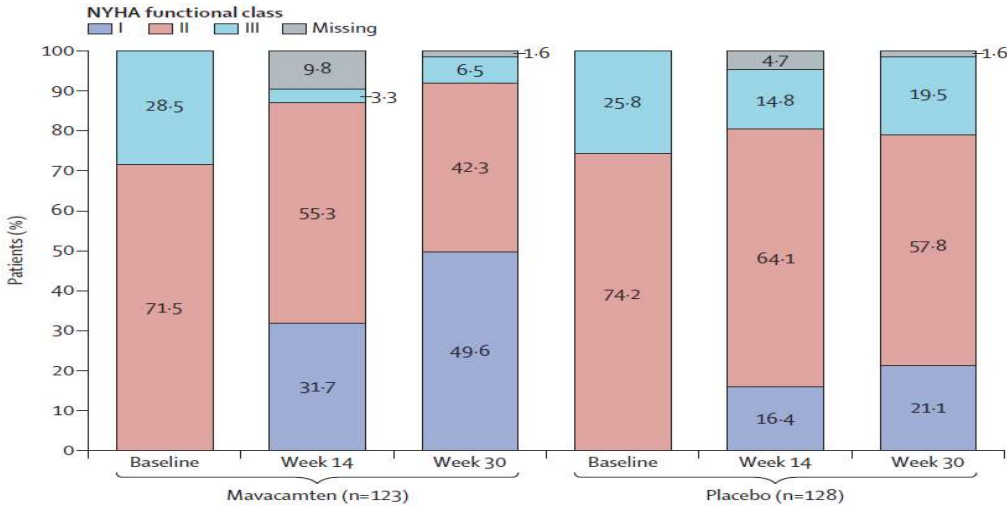
## Etude VALOR-HCM (112 pts)

Diminution moyenne du gradient intra VG en post-exercice

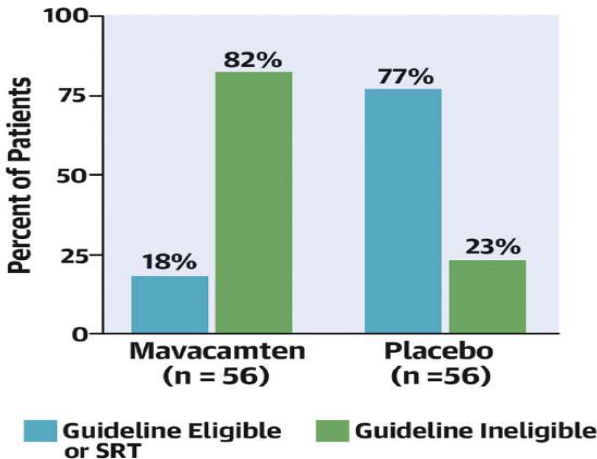
- 36 mmHg ( $p < 0,0001$  vs placebo)

- 37,2 mmHg

### Effets sur les symptômes (NYHA)



### Patients qui nécessitent ou restent selon les recommandations éligibles à un traitement de réduction septale



+ amélioration du pic VO<sub>2</sub> et KCCQ-CSS

## Actualités dans l'insuffisance cardiaque en 2023 : messages clefs

- IC aiguë : multiplier les sites d'action sur le néphron en cas de résistance au furosémide
- IC FEr : mettre en route les 4 classes thérapeutiques qui diminuent la mortalité, ARNi/IEC,  $\beta$ B, ARM, iSGLT2, le plus rapidement possible en augmentant les posologies aux doses maxima-tolérées en 6 semaines
- IC FEmr : même traitement que IC FEr
- IC FEp : iSGLT2 + diurétique de l'anse  $\pm$  ARM et traitement étiologique
- Co-morbidités :
  - Diabète : iSGLT2, finérénone
  - Carence martiale : fer IV à visée symptomatique
- Individualisation des cardiomyopathies ventriculaires gauches non dilatées
- Cardiomyopathies hypertrophiques obstructives : intérêt des inhibiteurs de la myosine