Alkoholová ablace septa u HOCM – jen symptomatickým jedincům

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2014 ESC Guidelines for the Diagnosis & Management of Hypertrophic Cardiomyopathy

The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC)

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www.escardio.org/guidelines European Heart Journal (2014);35:2733–2779 - doi:10.1093/eurheartj/ehu284

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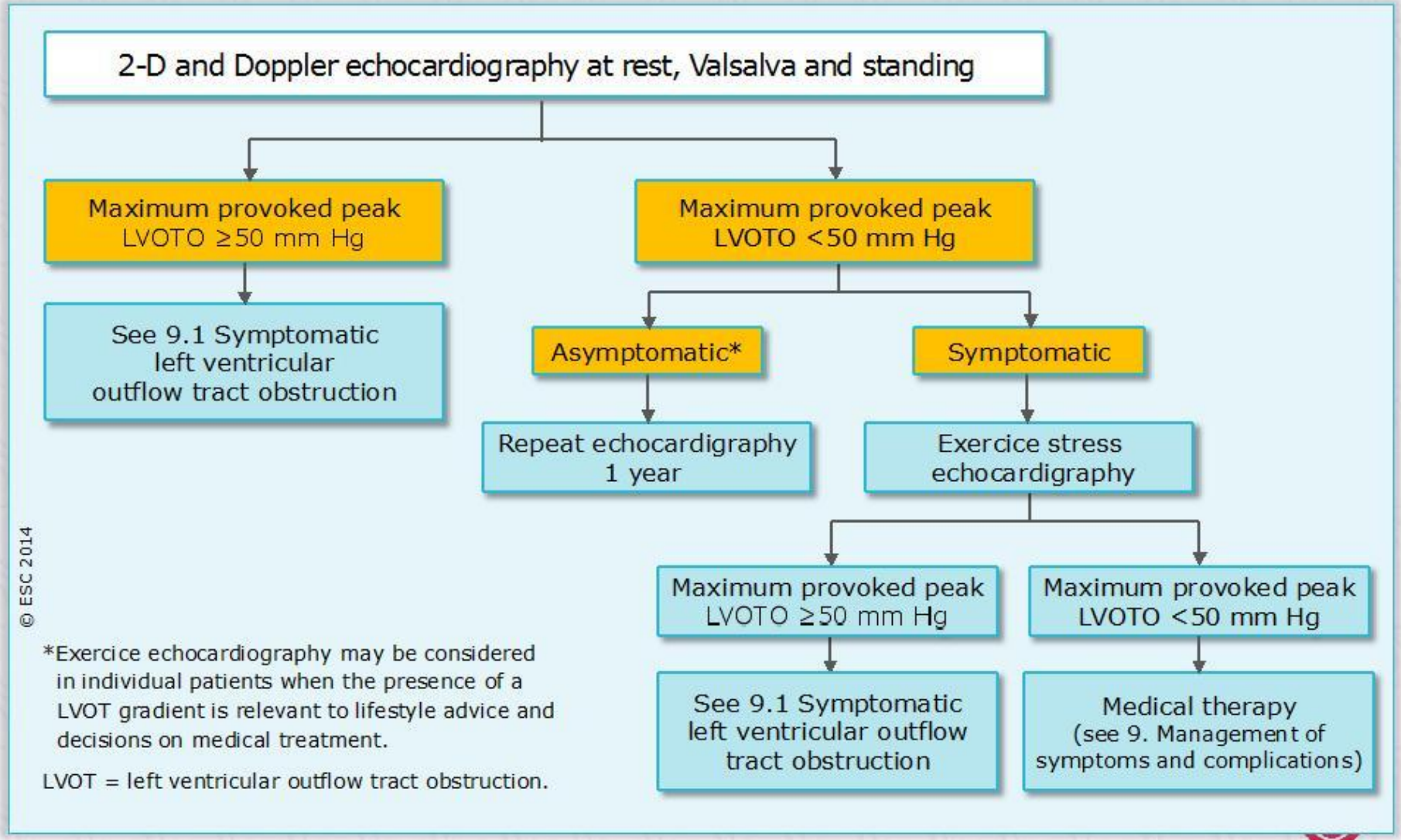
Protocol for the assessment and treatment of left ventricular outflow tract obstruction



*Exercice echocardiography may be considered in individual patients when the presence of a LVOT gradient is relevant to lifestyle advice and decisions on medical treatment.

LVOT = left ventricular outflow tract obstruction.

European Heart Journal (2014);35:2733–2779 - doi:10.1093/eurheartj/ehu284

Medical treatment of left ventricular outflow tract obstruction

Recommendations	Class	Level
Non-vasodilating ß-blockers, titrated to maximum tolerated dose, are recommended as first-line therapy to improve symptoms in symptomatic patients with resting or provoked LVOTO.	I	B
Verapamil, titrated to maximum tolerated dose, is recommended to improve symptoms in symptomatic patients with resting or provoked[a] LVOTO, who are intolerant or have contra-indications to ß-blockers.	I	B
Disopyramide, titrated to maximum tolerated dose[b], is recommended in addition to a ß-blocker (or, if this is not possible, with verapamil) to improve symptoms patients with resting or provoked[a] LVOTO.	I	B
Disopyramide, titrated to maximum tolerated dose[b], may be considered as monotherapy to improve symptoms in symptomatic patients with resting or provoked[a] LVOTO (exercise or Valsalva manoeuvre) taking caution in patients with–or prone to–AF, in whom it can increase ventricular rate response.	IIb	C
ß-Blockers or verapamil may be considered in children and *asymptomatic* adults with resting or provoked[a] LVOTO, to reduce left ventricular pressures.	IIb	C

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Septal reduction therapy

Recommendations	Class	Level
It is recommended that septal reduction therapies be performed by experienced operators, working as part of a multidisciplinary team expert in the management of HCM.	I	C
Septal reduction therapy to improve symptoms is recommended in patients with a resting or maximum provoked LVOT gradient of ≥50 mm Hg, who are in NYHA functional Class III–IV despite maximum tolerated medical therapy.	I	B
Septal reduction therapy should be considered in patients with recurrent exertional syncope caused by a resting or maximum provoked LVOTO gradient ≥50 mm Hg despite optimal medical therapy.	IIa	C
Septal myectomy, rather than SAA, is recommended in patients with an indication for septal reduction therapy and other lesions requiring surgical intervention (e.g. mitral valve repair/replacement, papillary muscle intervention).	I	C
Mitral valve repair or replacement should be considered in symptomatic patients with a resting or maximum provoked LVOTO gradient ≥50 mm Hg and moderate-to-severe mitral regurgitation not caused by SAM of the mitral valve alone.	IIa	C
Mitral valve repair or replacement may be considered in patients with a resting or maximum provoked LVOTO gradient ≥50 mm Hg and a maximum septal thickness ≤16 mm at the point of the mitral leaflet–septal contact or when there is moderate-to-severe mitral regurgitation following isolated myectomy.	IIb	C

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Pre-assessment check list for patients being considered for invasive septal reduction therapies

Are there alternative/additional explanations for symptoms?

- Obesity
- Respiratory Disease
- Coronary artery disease
- Anaemia
- Thyroid disease
- Arrhythmia (e.g. AF)
- Drug side-effects
- Systemic disease (e.g. amyloid)
- RVOT obstruction

What is the mechanism of obstruction?

- SAM-related
- Mid-cavity
- Sub-aortic membrane
- Aortic stenosis
- Anomalous papillary muscle insertion
- Accessory mitral valve tissue

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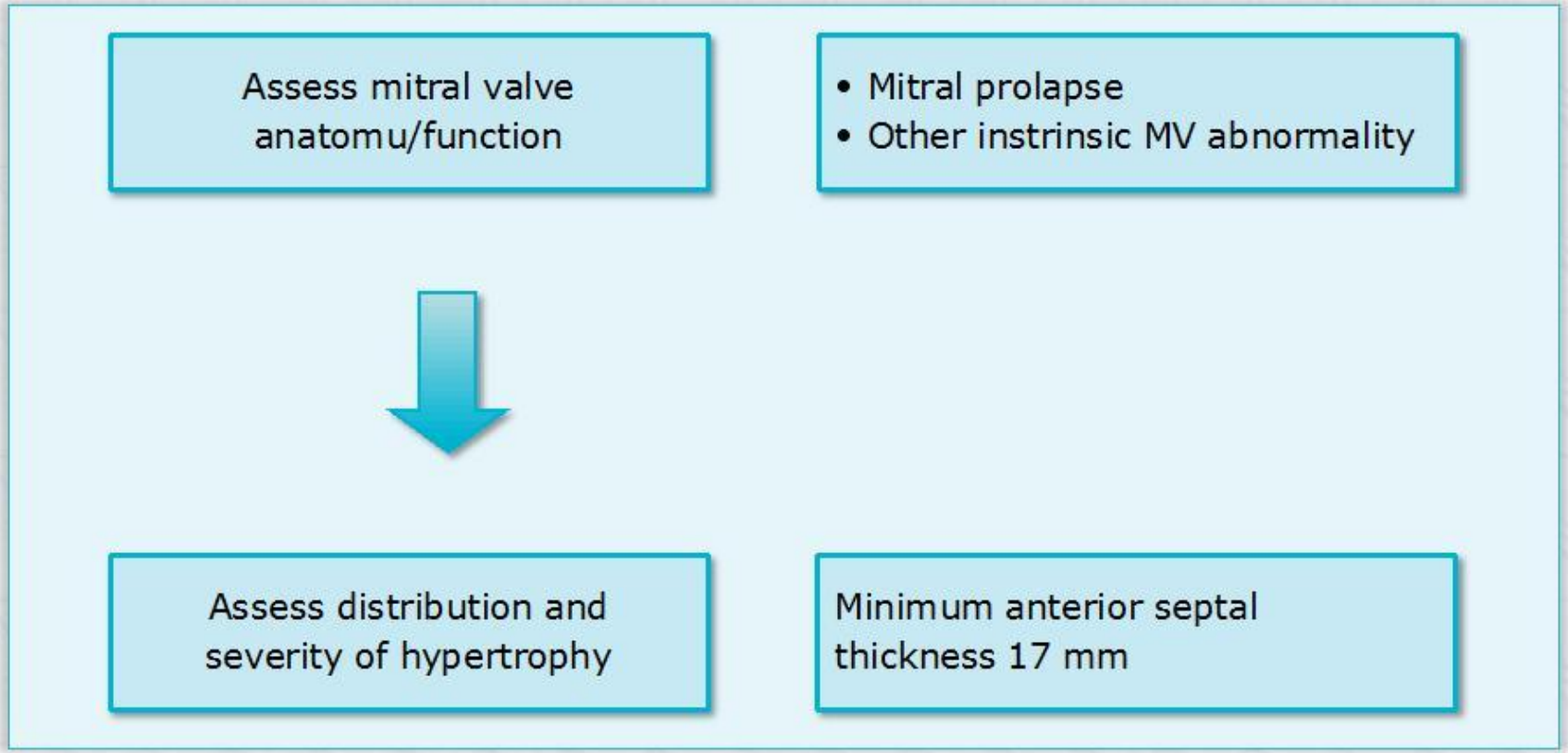
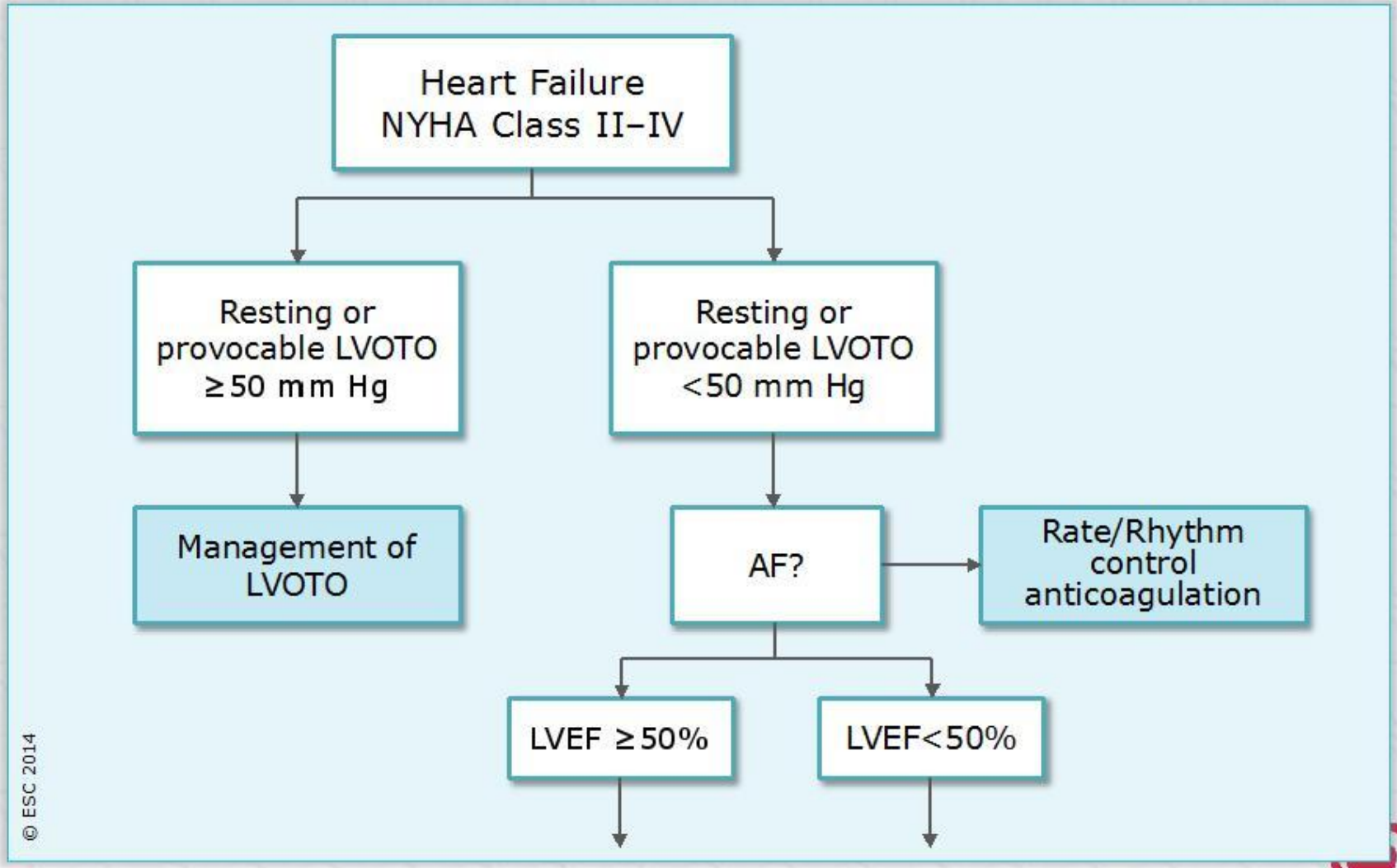
Pre-assessment check list for patients being considered for invasive septal reduction therapies *(Cont.)*



European Heart Journal (2014);35:2733–2779 - doi:10.1093/eurheartj/ehu284

Algorithm for the treatment of heart failure in hypertrophic cardiomyopathy



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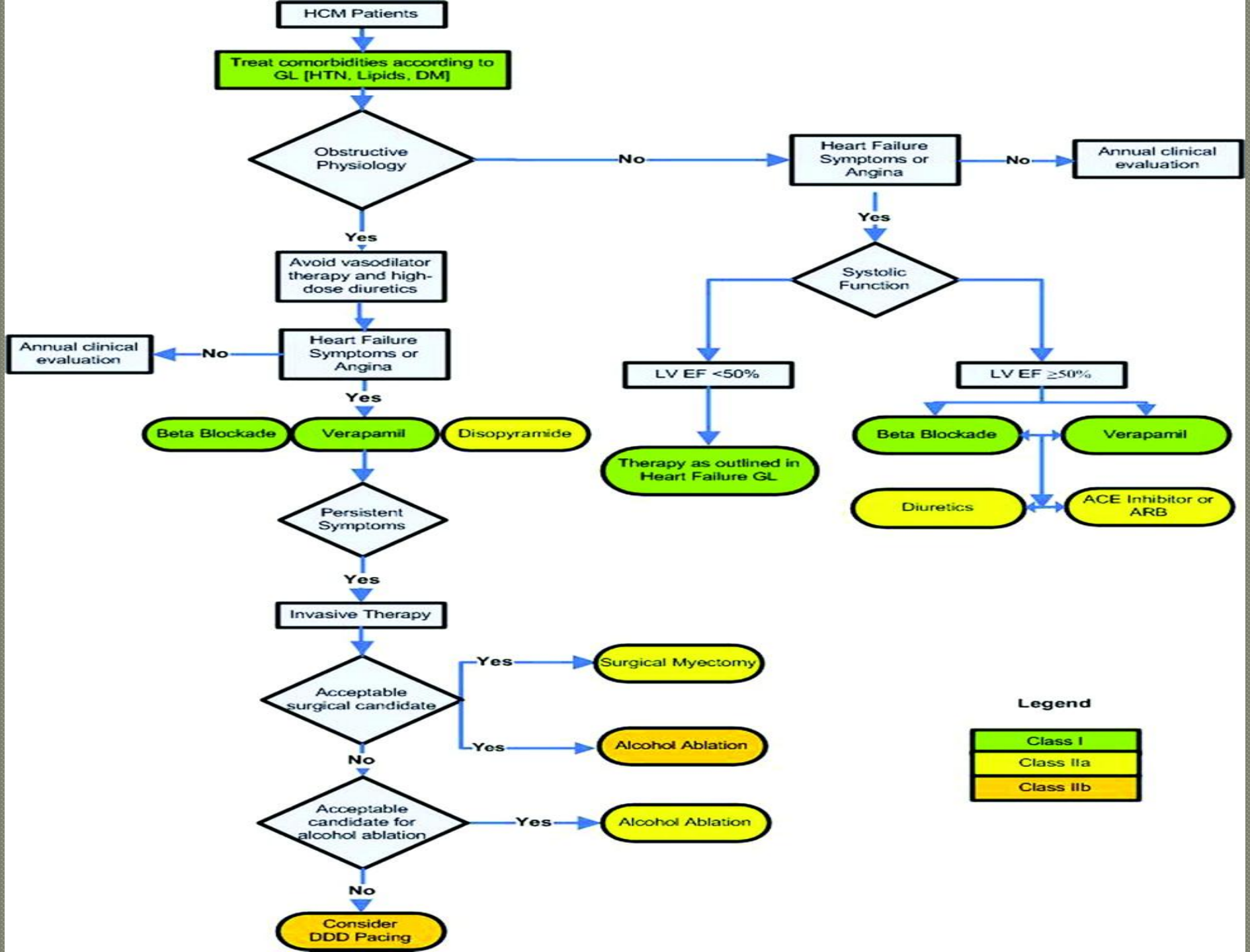

HCM Patients
Treat comorbidities according to GL [HTN, Lipids, DM]
Obstructive Physiology
No
Heart Failure Symptoms or Angina
No
Annual clinical evaluation
Yes
Avoid vasodilator therapy and high-dose diuretics
Yes
Systolic Function
LV EF <50%
LV EF ≥50%
Therapy as outlined in Heart Failure GL
Beta Blockade
Verapamil
Diuretics
ACE Inhibitor or ARB
Annual clinical evaluation
No
Heart Failure Symptoms or Angina
Yes
Beta Blockade
Verapamil
Disopyramide
Persistent Symptoms
Yes
Invasive Therapy
Acceptable surgical candidate
Yes
Surgical Myectomy
Yes
Alcohol Ablation
No
Acceptable candidate for alcohol ablation
Yes
Alcohol Ablation
No
Consider DDD Pacing
Legend
Class I
Class IIa
Class IIb

2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy
Asymptomatic Patients—Recommendations

- **Septal reduction therapy should not be performed for asymptomatic adult and pediatric patients with HCM with normal effort tolerance regardless of the severity of obstruction.**[9,10]
- **In patients with HCM with resting or provocable outflow tract obstruction, regardless of symptom status, pure vasodilators and high-dose diuretics are potentially harmful.**[3,9]

JACC:Cardiovascular Interventions. Volume9, Issue 5, 14March 2016, Pages 463–469

Max Liebregts et al. Long-Term Outcome of Alcohol Septal Ablation for Obstructive Hypertrophic Cardiomyopathy in the Young and the Elderly.

- If patients with obstructive hypertrophic cardiomyopathy remain severely symptomatic despite optimal medical therapy, septal reduction therapy should be considered. This can be done either by surgical myectomy or alcohol septal ablation…

Příčiny tlakového gradientu u HOKMP

- Komplexnější mechanismus
  - Hypertrofie septa, zvl. bazálního
  - Abnormality mitrálního aparátu a chlopně (pozice vpředu papilárních svalů, šlašinek, elongace a redundance jednoho nebo více skallopů cípů a šlašinek, kalcifikace v prstenci)
  - Aktuální hemodynamické faktory (preload, afterload)

Způsoby řešení obstrukce (a MR) u HOKMP

- **Katetrizační**
  - - ASA
  - - plikace MCH *(Sorajja 2016)*
  - - mitraClip *(Shäfer 2014)*, katetrizační implantace MV *(Deharo 2016)*
  - - radiofrekvenční ablace septa *(Cooper 2015, Crossen 2016, Shelke 2016)*
- **Kardiochirurgické**
  - - (náhrada MCH), transaortální myektomie septa, resekce a plikace cípů + chord *(Colley 1991…, Ferrazi 2015, Bulgerov 2016)*
- **Kardiostimulace**
  - - dvoudutinová *(Krejčí 2013…Roman 2016, Albano 2017)*, biventrikulární *(Giraldeau 2016)*

Vždy pouze u symptomatických.

Eur.J.Cardiothoracic Surgery. Volume 50, Issue 1, 1 July 2016, Article number ezv473, Pages 61-65

- **Hypertrophic obstructive cardiomyopathy: The mitral valve could be the key**
- Dulguerov F., Marcacci, Alexandrescu, C.,
- Chan K.M.J., Dreyfus G.D.

Zákroky u HOKMP a přežití : kardiochirurgie

- Steve R. Ommen, Barry J. Maron et al. J Am Coll Cardiol 2005;46:470.
- **Surgical myectomy** performed to relieve outflow obstruction and **severe symptoms** in HCM was associated with long-term survival equivalent to that of the general population, and superior to obstructive HCM without operation. In this retrospective study, septal myectomy seems to reduce mortality risk **in severely symptomatic patients** with obstructive HCM.

Zlepšení prognózy **symptomatických** s HOKMP

- **Po kardiochirurgických výkonech** *(Gersh 2011, Liebregts 2015, 2016, Maron 2013, Ommen 2005, Shaff 2012)*
- **Po ASA** *(Aksu 2016, Ball 2011, Jensen 2013, Liebregts 2015, 2016, Maron 2016, Quiao 2016, Singh 2016, Sorajja 2009, 2012, Veselka 2014, 2016, Yang 2016)*
- Vždy šlo o symptomatické pacienty

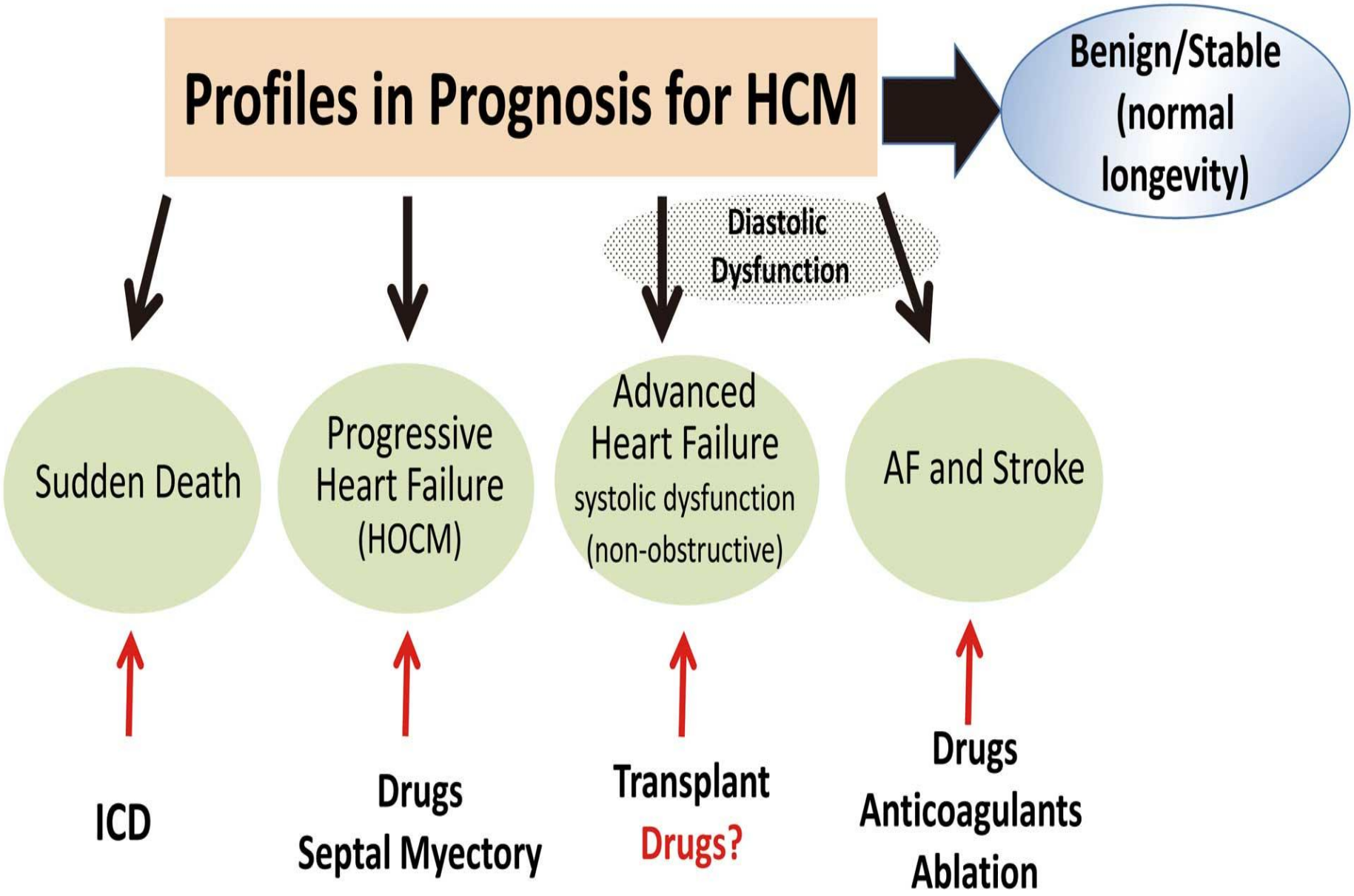
Indikační kriteria k operaci AS

(Vahanian, ESC, 2012)

- **Těžká symptomatická** - operovat vždy
- -AVA < 0,6 cm^2/m^2, PGmean > 40 mm Hg, PGmax > 60 mm Hg, Vmax > 4 m/s)
- -PGmean < 30 mm Hg při EF ≤ 35% (pro AS)
- **Těžká asymptomatická**
  - EF < 50% z důvodu AS
  - Současný CABG, jiná chlopeň vč. náhrady aorty
  - Objevení se symptomů při zátěžovém testu

    Pozitivní zátěžový test (↓TK, depresí ST > 2 mm, komorová tachykardie)
  - Velmi těžká AS (AVA < 0,6 cm^2, PG_{mean} > 60 mm Hg, V_{max} > 5 – 5,5 m/s při nízkém operačním riziku)
  - (Těžká hypertrofie ≥ 15 mm bez hypertenze, rychlá progrese vady - při nízkém operačním riziku)
- ***Důkazy u HKMP nejsou, inspirace při organizaci studie?***

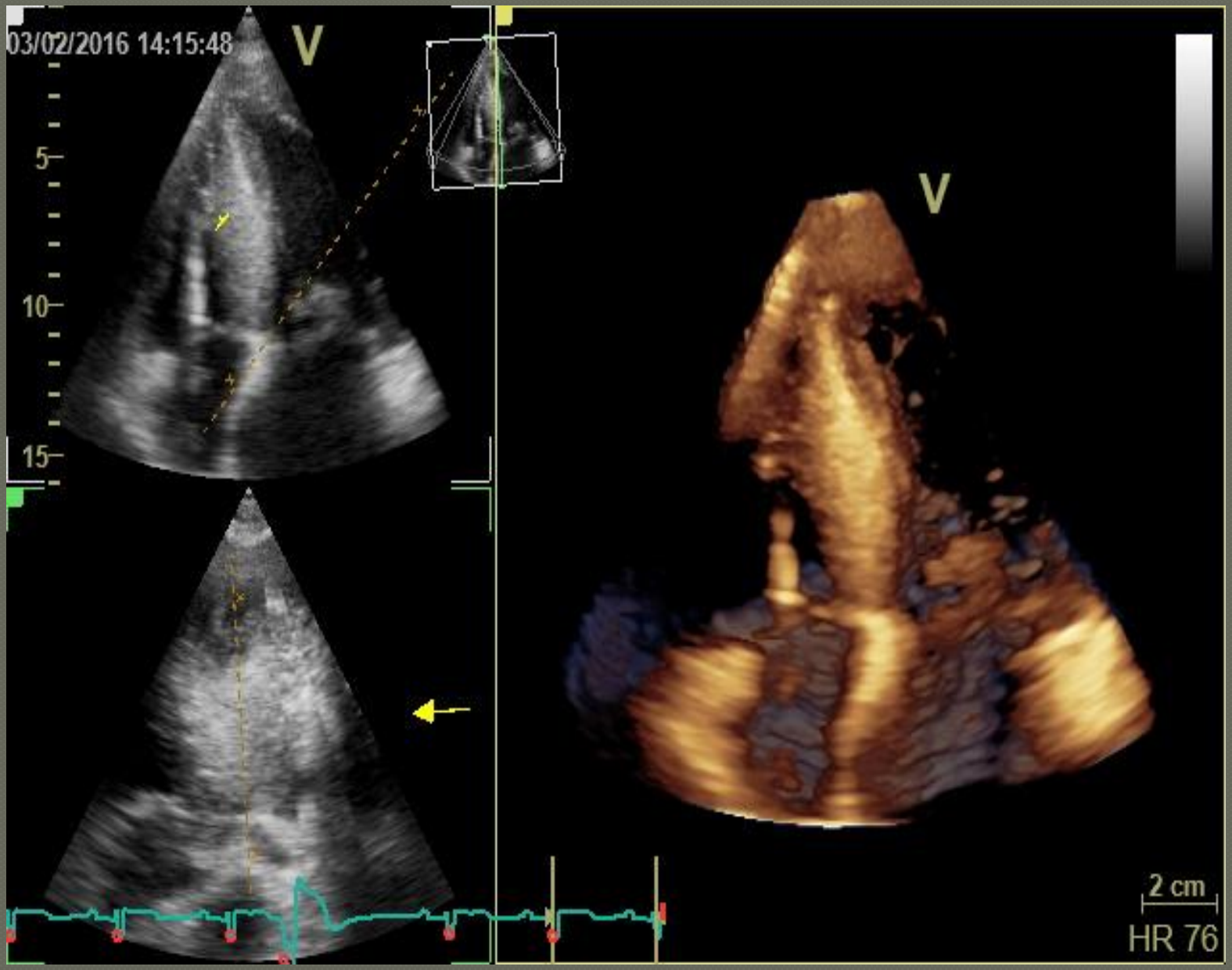
K.Yamauchi-Takihara, Circ.J, 2015 (modified from Maron et al., JACC, 2014)



Alkoholová ablace septa u HOCM i asymptomatickým?

- Jsou důkazy k tomu, abychom desítiletí trvající praxi změnili ? **Nejsou.**
- Přináší KCH i ASA naději na zlepšení životní prognózy i pro asymptomatické nemocné? **Nevíme.**

- **Prospektivní randomizovaná studie** *(Olivotto 2007)* event. širší konsenzus odborníků.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