

# NON COMPACTION VG

## LVNC LEFT VENTRICULE NON COMPACTION

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Cardiologie                      CHU Poitiers

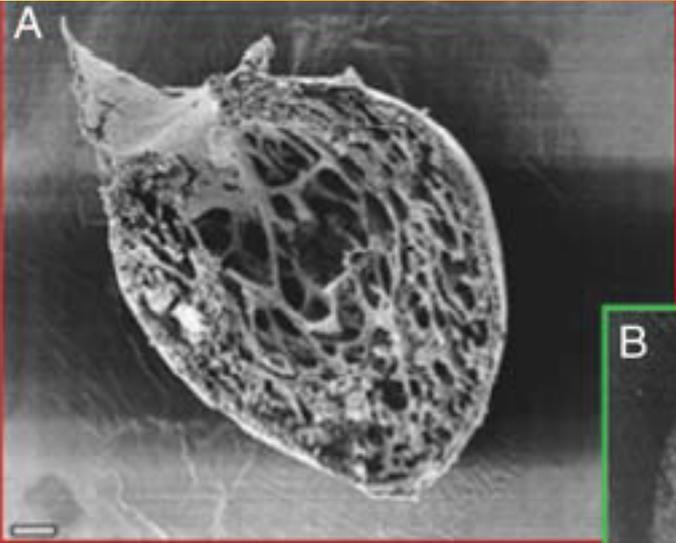
[l.christiaens@chu-poitiers.fr](mailto:l.christiaens@chu-poitiers.fr)

Conflit d'intérêt = 0

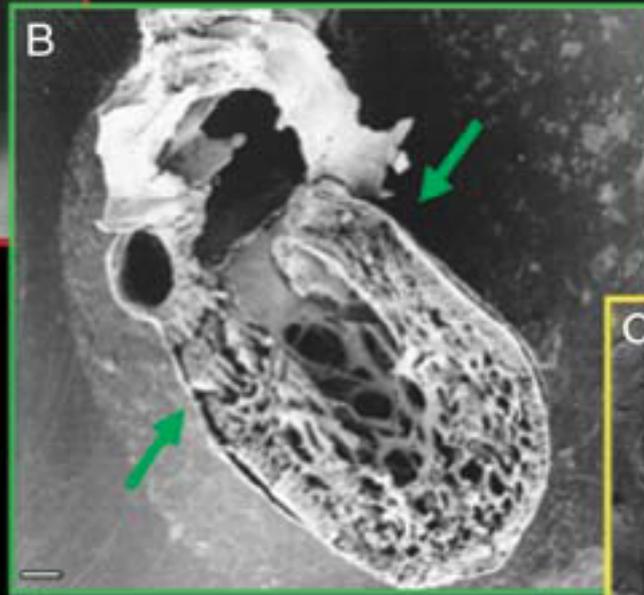


# NON COMPACTION / OBJECTIFS

- Connaître les définitions et les critères diagnostiques en fonction du type d'imagerie
- Connaître les complications secondaires éventuelles



At 6 weeks:  
Abundant fine trabeculations



At 8 weeks:  
Trabeculae start to solidify



Early foetal period:  
Compaction almost completed

The process of compaction

# NON COMPACTION VG

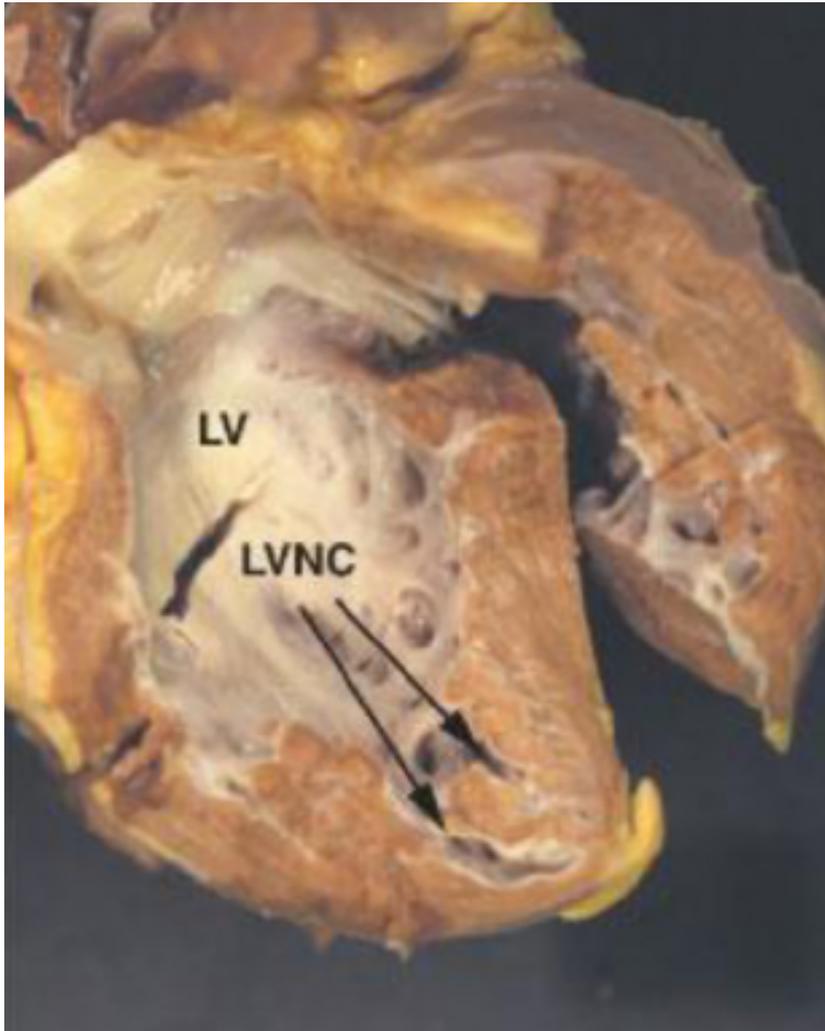
- Compaction progressive normale des trabéculations VG:
  - de la base vers l'apex
  - de l'épicarde vers l'endocarde
  - du septum vers la paroi latérale
- 70% des cœurs normaux présentent des zones non compactées à l'autopsie

Petersen SE. JACC 2005

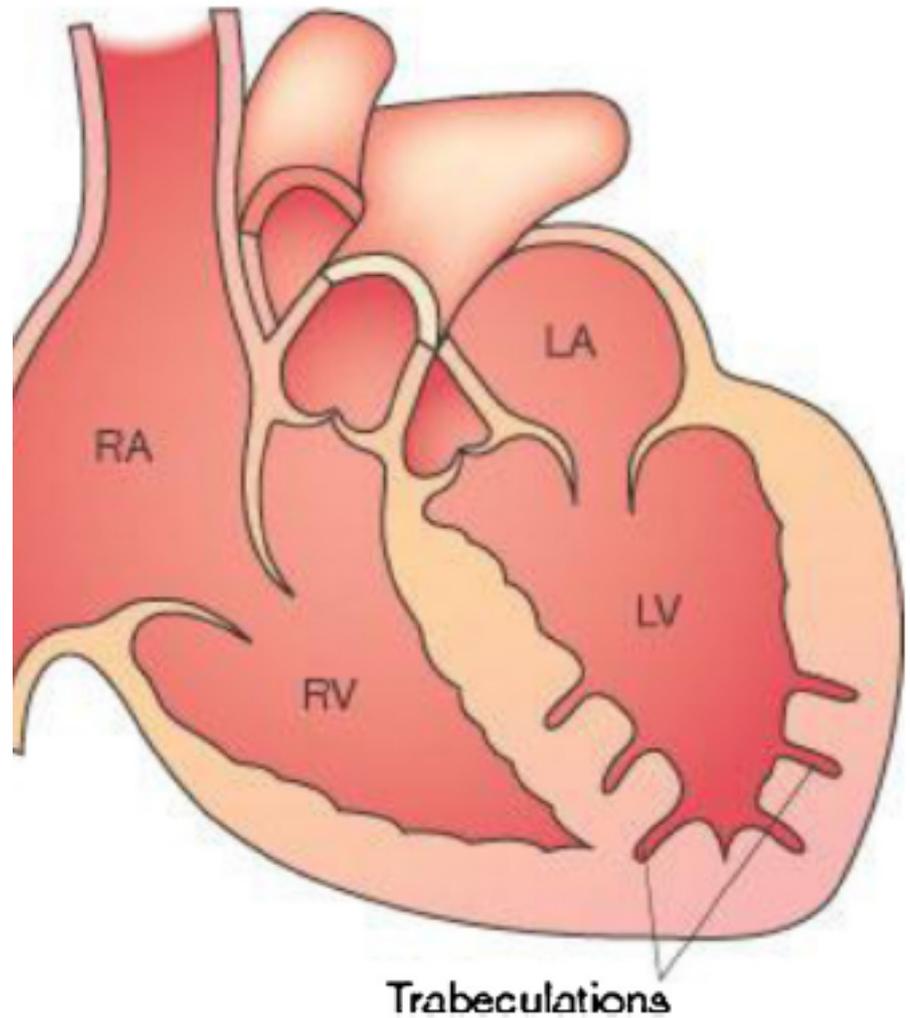
# NON COMPACTION VG (DÉFINITION ?)

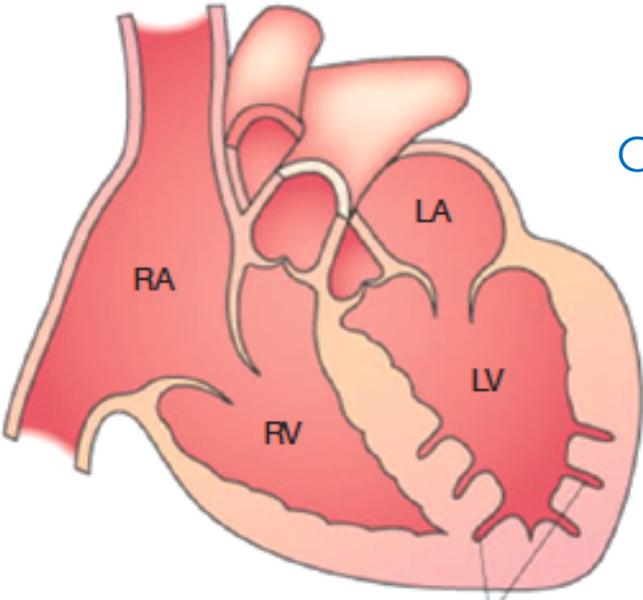
- Cardiopathie (congénitale / acquise ?)
  - Association de trabéculations ventriculaires myocardiques nombreuses et profondes en général localisées au niveau de l'apex du ventricule gauche, séparées par des espaces intertrabéculaires ou récessus communiquant avec la cavité VG
- Pouvant se compliquer:
  - Risque d'insuffisance cardiaque (Dysfonction systolique VG fréquente)
  - Risque thrombo-embolique (stase dans les récessus)
  - Risque d'arythmies graves
- Diagnostic à tout âge, variabilité clinique +++
- Formes familiales possibles (autosomique dominant) ou sporadiques
- Cardiomyopathie non classée pour l'ESC

# LVNC (LEFT VENTRICULAR NON-COMPACTION)



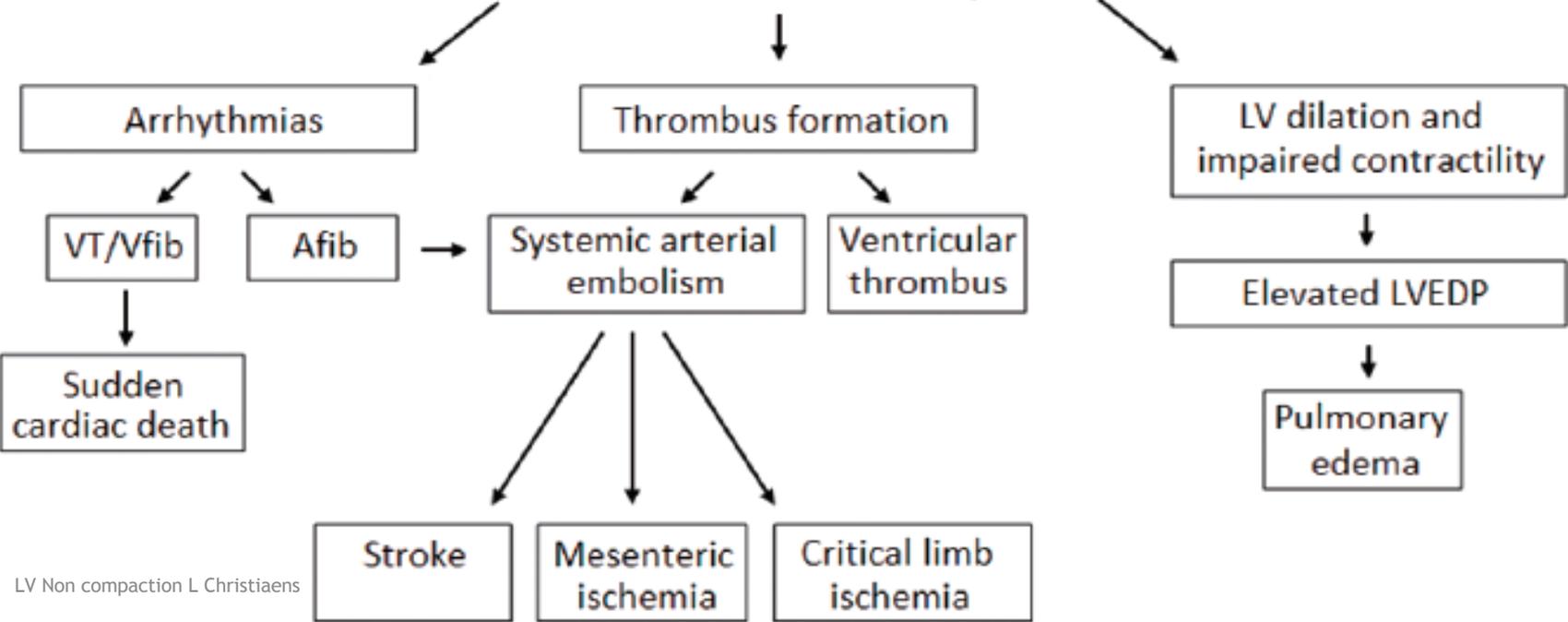
LV Non compaction L Christiaens





Trabeculations

Isolated Ventricular Noncompaction



# NON COMPACTION VG

## Difficultés diagnostiques

- Pas de critères diagnostiques anapathologiques
- Critères diagnostiques variables / méthode
- Pas de “Gold standard”

It is important to emphasize here that currently there is no diagnostic tool, neither genetic nor imaging, that can, in all patients, reveal the underlying truth of whether or not somebody is affected by LVNC.

**a likely diagnosis of LVNC and not a definitive diagnosis**

Petersen SE, JACC Cvi 2013

# NON COMPACTION VG

- Hypokinésie fréquente dans les zones de NC
- Anomalies de la microcirculation coronaire responsables d'ischémie et de fibrose sous-endocardique, considérées comme cause possible de la dysfonction systolique
- Nombreuses mutations génétiques rapportées

Shemisa Kamal, Cardiovasc Diagn Ther 2013

Ikeda U, J Cardiol 2014

## LVNC: INCERTITUDES

The natural history of LVNC with normal ejection fraction compared to normally functioning and normally trabeculated hearts remains unknown

The natural history of LVNC with impaired LV systolic function compared to that in matched dilated cardiomyopathy patients is unclear.

Should patients with heavily trabeculated hearts and normal function be treated to reduce the likelihood of adverse remodelling or thromboembolic events?

Should LVNC patients with impaired systolic function be managed differently from those with dilated cardiomyopathy?

# ECHO ET LVNC

- ETT : examen de dépistage
- 0.05% population (?)
- rapport couche NC / couche C  $> 2$  en systole
- Avec au moins 3 sillons profonds avec signal couleur +
- Prédominance apex et paroi latérale

**Table 1** Frequency of zones of noncompaction

Location	Frequency
Apex	91%
Mid-cavity	78%
Base	21%

Two-layered structure of the myocardium (epicardial compacted, endocardial non-compacted layer)

Determination of the X-to-Y ratio ( $\leq 0.5$ )

X—Distance between the epicardial surface and through of intertrabecular recess

Y—Distance between epicardial surface and peak of trabeculation

*Acquisition of the images:* parasternal short-axis view, measurements of the X-to-Y ratio at end-diastole

Thickened myocardium with a two-layered structure consisting of a thin compacted epicardial layer/band (C) and a much thicker, non-compacted endocardial layer (N) or trabecular meshwork with deep endomyocardial spaces; N/C ratio  $> 2.0$

Predominant location of the pathology: mid-lateral, mid-inferior, and apex

Colour Doppler evidence of deep intertrabecular recesses filled with blood from the left ventricular cavity

Absence of coexisting cardiac abnormalities (in the presence of isolated LVNC)

*Acquisition of the images:* short-axis views, measurements of the N/C ratio at end-systole

More than three trabeculations protruding from the left ventricular wall, located apically to the papillary muscles and visible in one image plane

Trabeculations with the same echogenicity as the myocardium and synchronous movement with ventricular contractions

Perfusion of the intertrabecular spaces from the left ventricular cavity

Ratio of non-compacted to compacted segment  $> 2.0$  at end-diastole (this criterion was introduced later)

*Acquisition of the images:* apical four chamber view; angulation of the transducer and acquisition of pictures in atypical views to obtain the technically best picture quality for differentiation between false chords/aberrant bands and trabeculations

Diagnostic criteria have changed during the last years

30% seulement des LVNC répondent aux 3 définitions.

Anomalies plus fréquentes chez les africains et les asiatiques

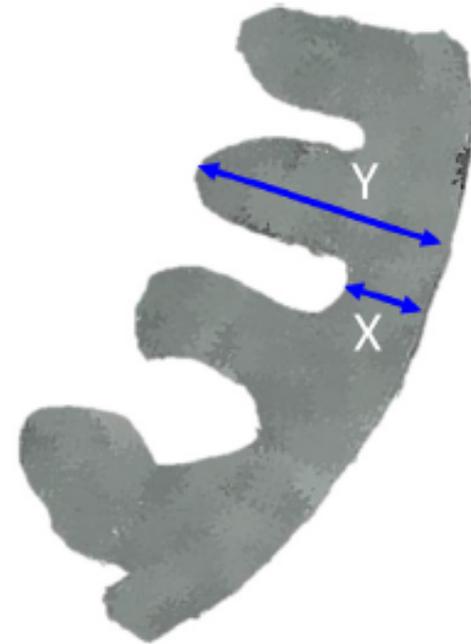
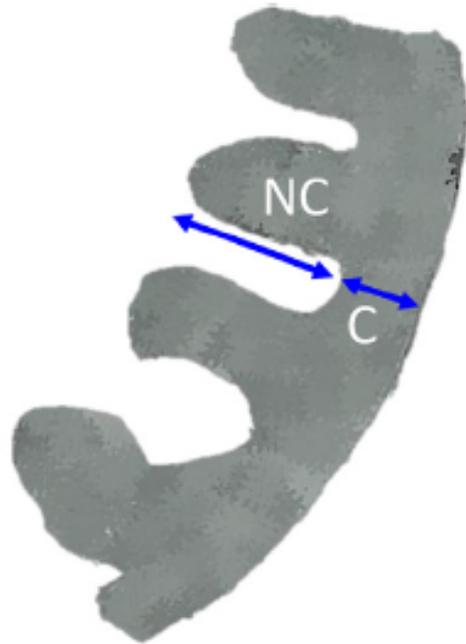
## DIASTOLE

Définitions

écho ?????

## SYSTOLE

# LVNC ECHO



## The criteria by Jenii et al.[20]

- 1) A two-layer structure with a thin, compacted layer (C) and a thickened non-compacted layer (NC) at end-systole. ←
- 2) A ratio of  $NC/C > 2$ .
- 3) Intertrabecular spaces are filled by blood flow from the ventricular cavity on color Doppler.
- 4) Other cardiac abnormalities do not exist.

## The criteria by Chin et al.[3]

- 1) Distance from the epicardial surface to the trough of the trabecular recess (X) and distance from the epicardial surface to peak of trabeculation (Y) at end-diastole. ←
- 2) A ratio of  $X/Y \leq 0.5$ .
- 3) Focuses on trabeculations at the LV apex.

## ECHO ET LVNC

LVNC is considered a distinct, primary cardiomyopathy despite variable diagnostic criteria and limited validation of such criteria. Our study demonstrates that the qualitative diagnosis and quantitative measurements to fill diagnostic criteria are poorly reproducible between observers. Severe outcomes in our cohort of patients were associated with poor LV function and not morphologic findings. We call into question whether LV hypertabeculation represents a primary cardiomyopathy or a secondary morphologic trait of variable significance.

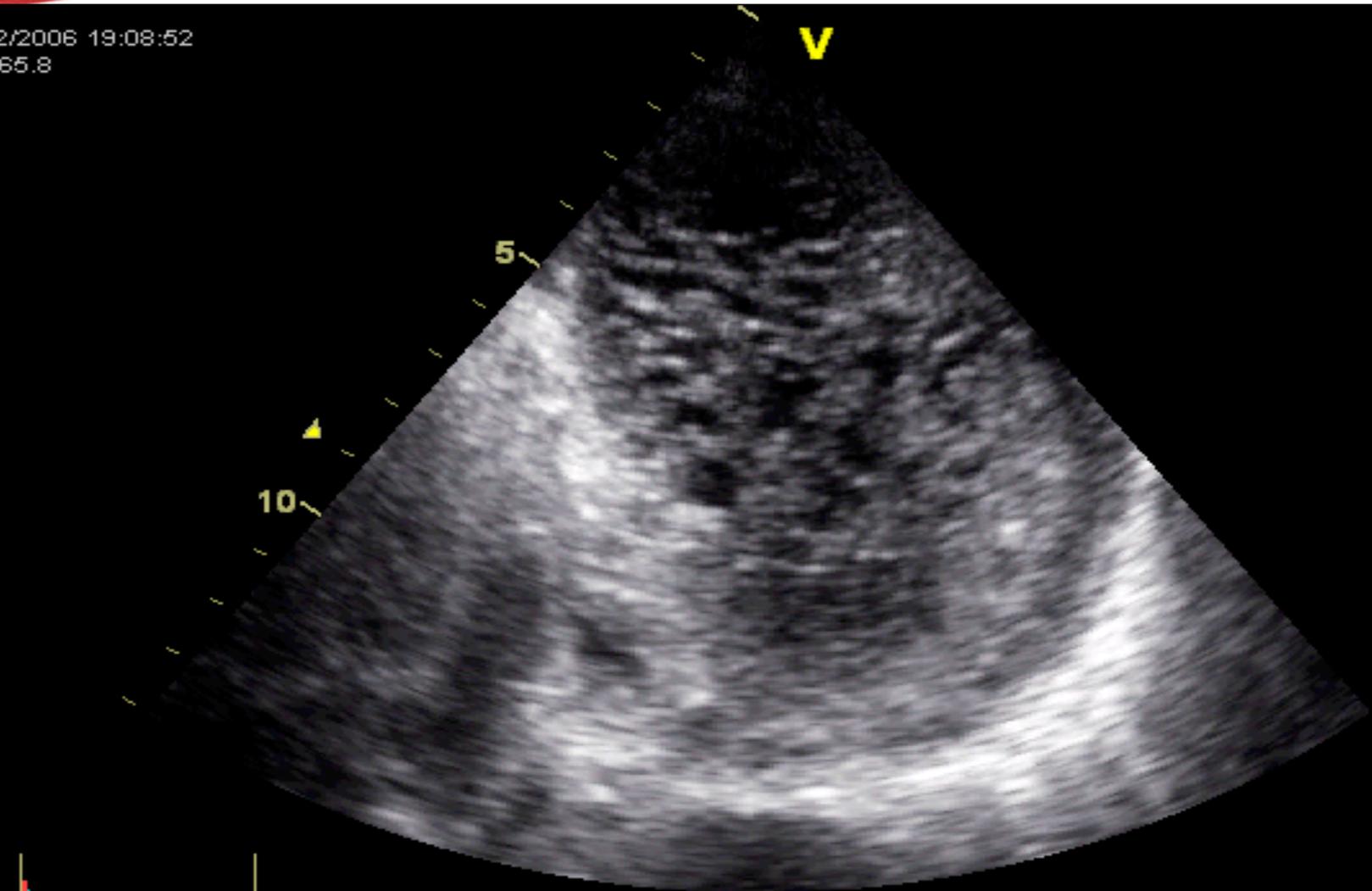
**Conclusions:** The reproducibility of making measurements to diagnose LVNC by accepted criteria is poor. Heart transplantation and death are associated with significant ventricular dysfunction and not with increased trabeculations or NC/C ratios. (J Am Soc Echocardiogr 2012;25:194-202.)

# NORMAL OU LVNC ??

- Volume Trabéculations / m<sup>2</sup> SC
  - hommes > femmes
  - diminue avec l'age
  - Inversement % à FEVG mais rôle de VTDVG ++
- Même chez des volontaires sains 26 à 70% ont des critères de LVNC (NC/C ratio en diastole > 2.3), % age et le sexe
  - Plus de faux + chez la femme
- Attention à l'exclusion des muscles papillaires !!

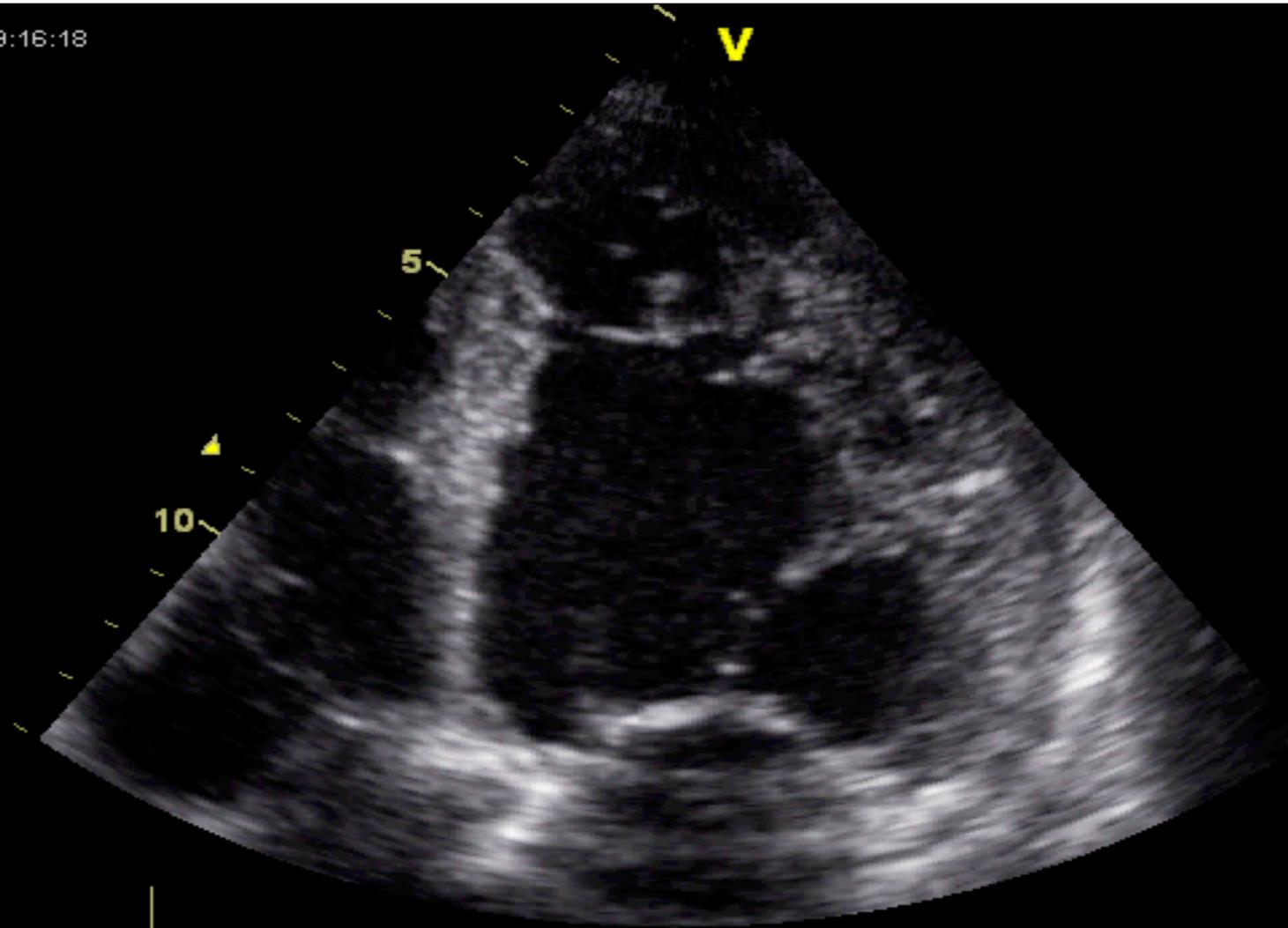
# ECHO ET LVNC

04/12/2006 19:08:52  
IPS: 65.8



88  
2:47 HR

04/12/2006 19:16:18  
IPS: 65.8

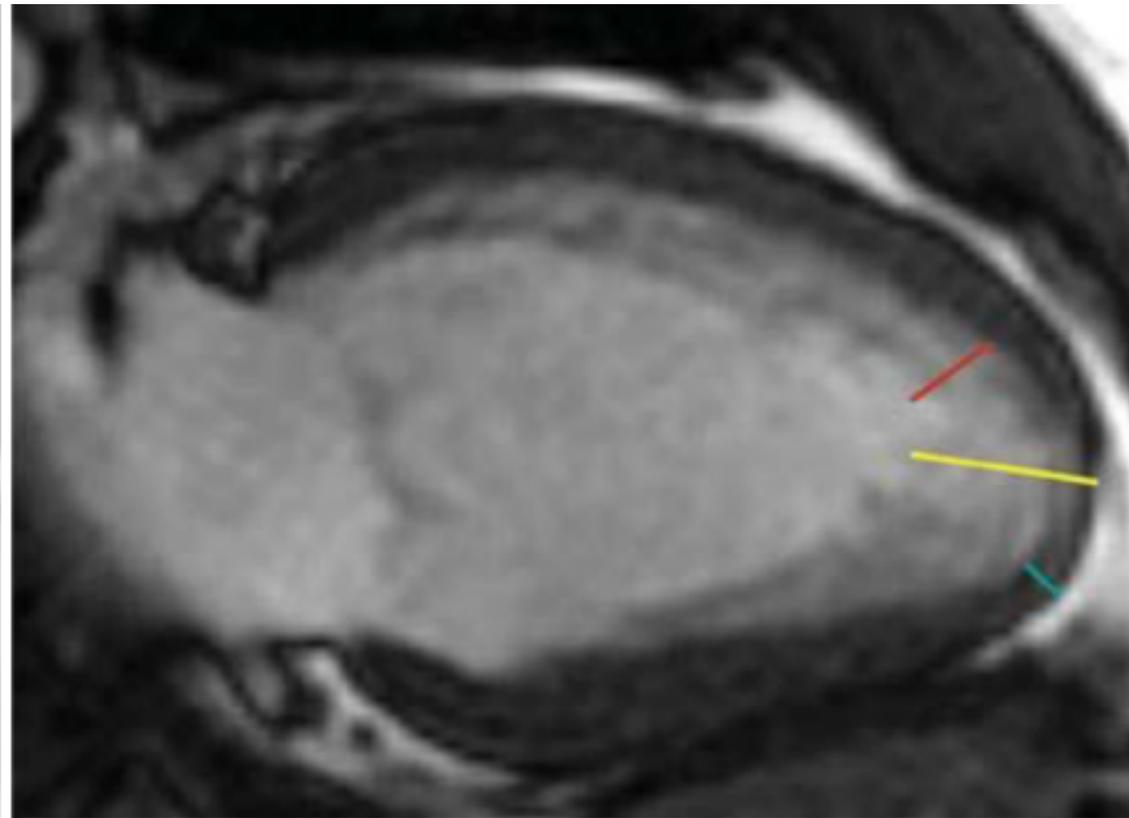
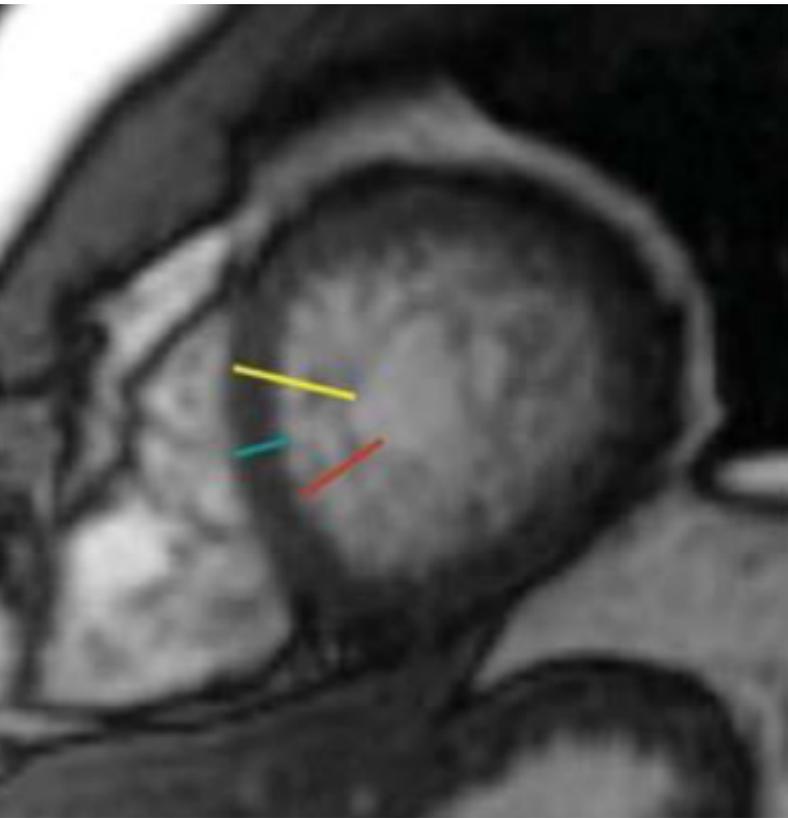


89  
46:90 HR

# NON COMPACTION VG / **IRM**

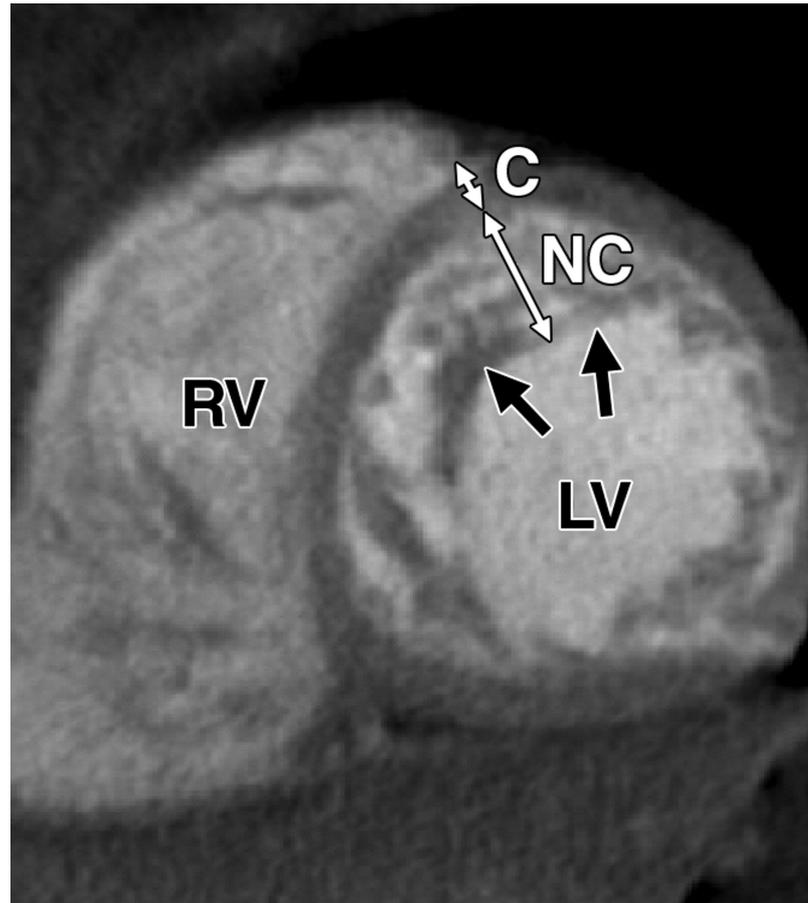
- 2005: mesures en diastole de NC/C car qualité des images en systole insuffisante
- Mesures en diastole: plus sensible mais moins spécifiques
- Mesures en petit axe et non en long axe
- **NC/C en systole: > 2 pour le diagnostic de LVNC**
- **NC/C en diastole: > 2.3**
- **Agrément entre les critères diagnostiques: 77%**

# NON COMPACTION VG / **IRM**



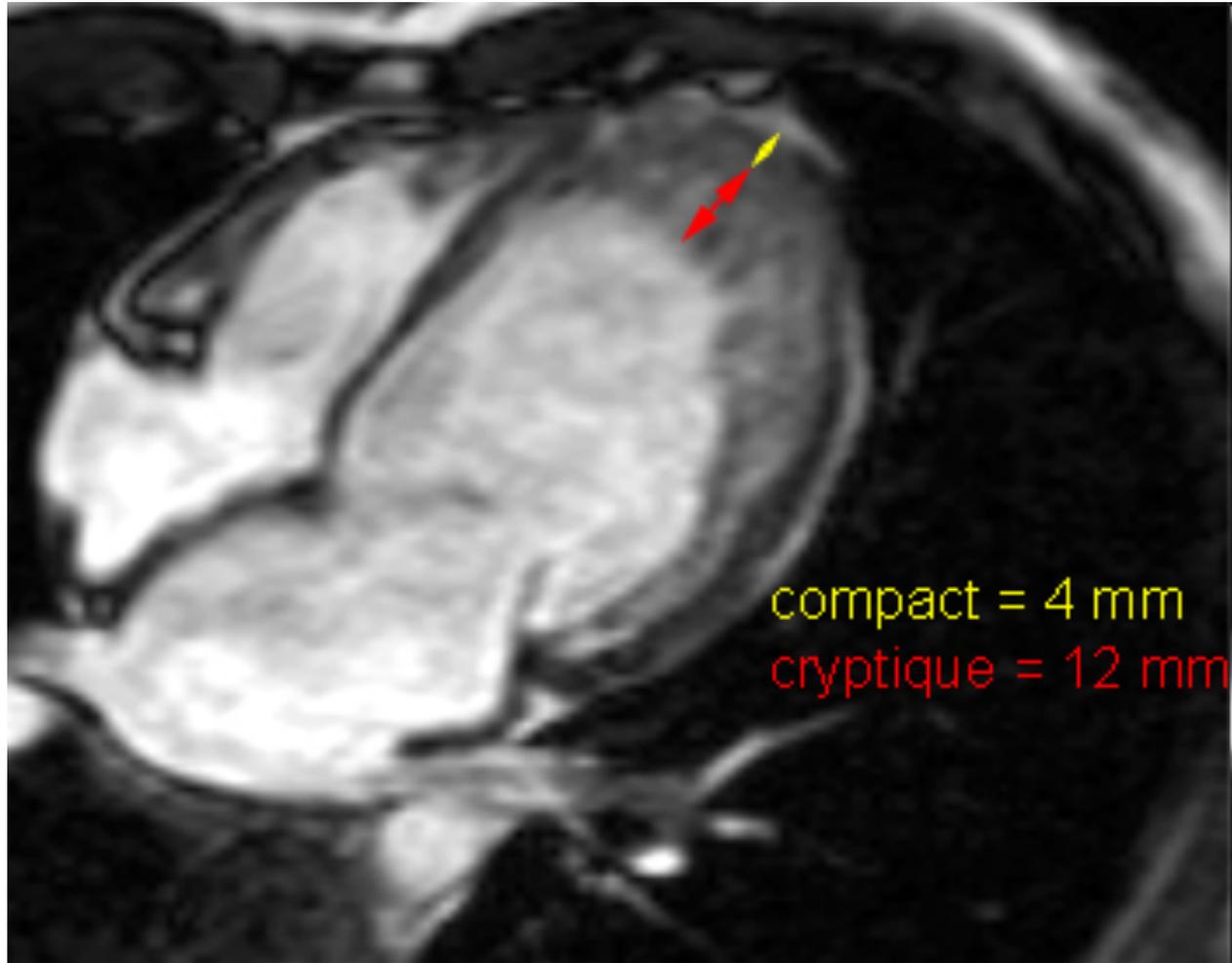
Télédiastole. Jaune: épaisseur totale de la paroi, Rouge: épaisseur NC, Bleu: épaisseur C  
Maximum NC/C = 4

# NON COMPACTION VG / **IRM**

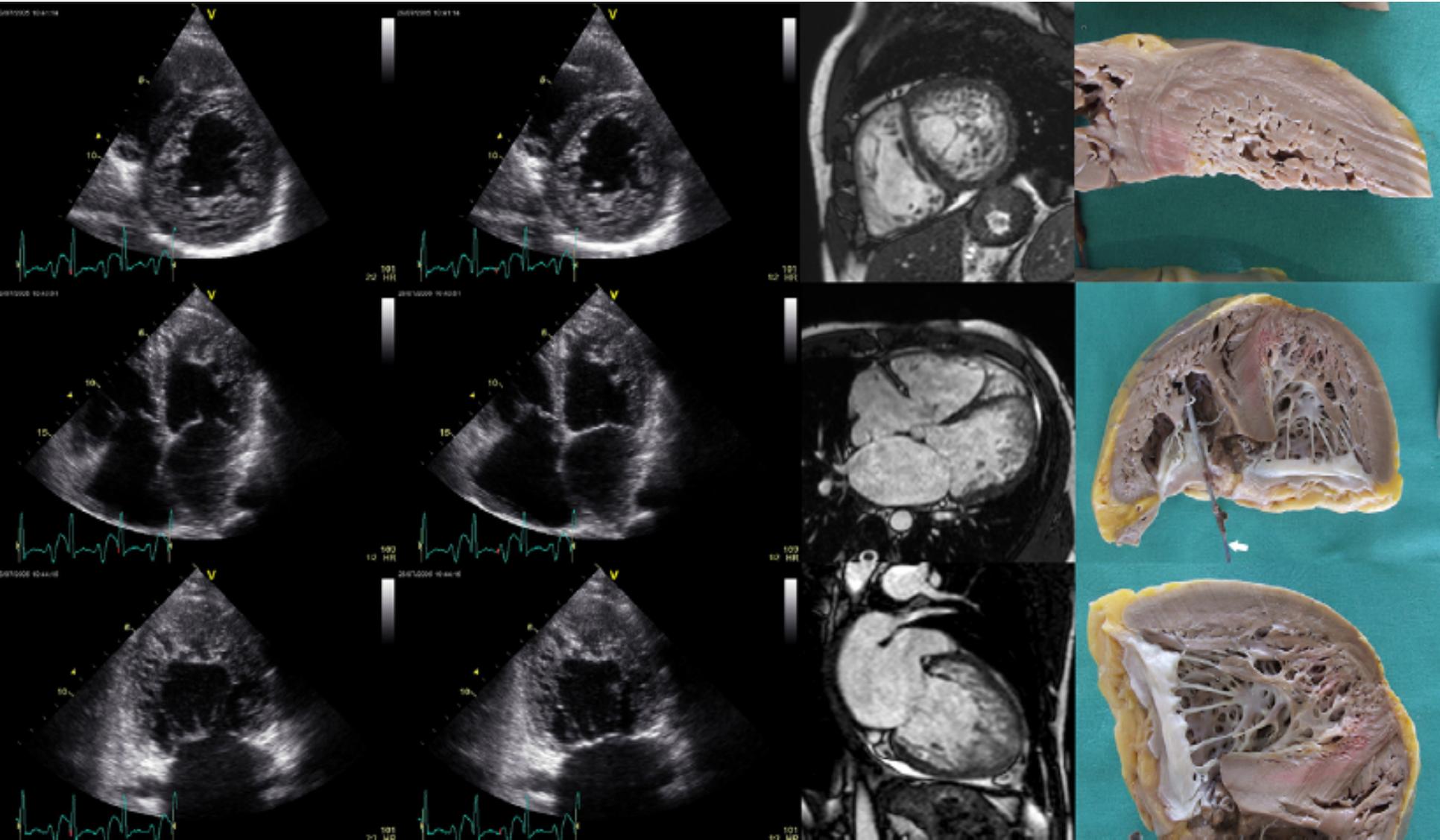


Lee, T. et al. Am. J. Roentgenol. 2007

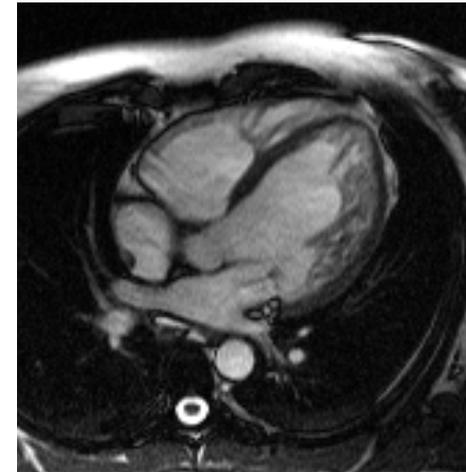
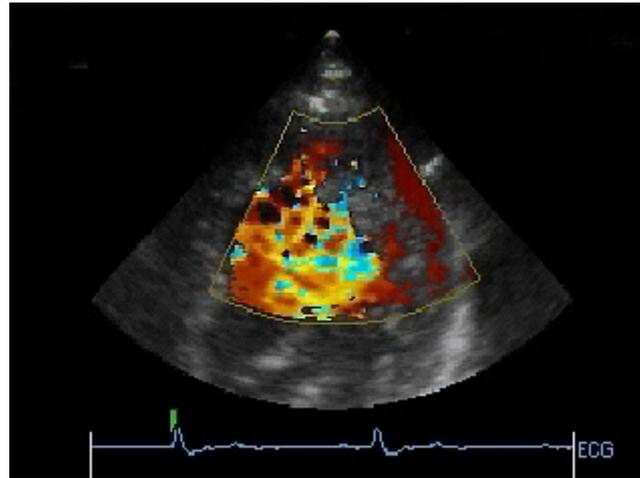
# NON COMPACTION VG / **IRM**



Thuny F, 2010 Arch Mal Coeur Vx  
Distinction des 2 couches plus facile en IRM  
IRM mieux que écho

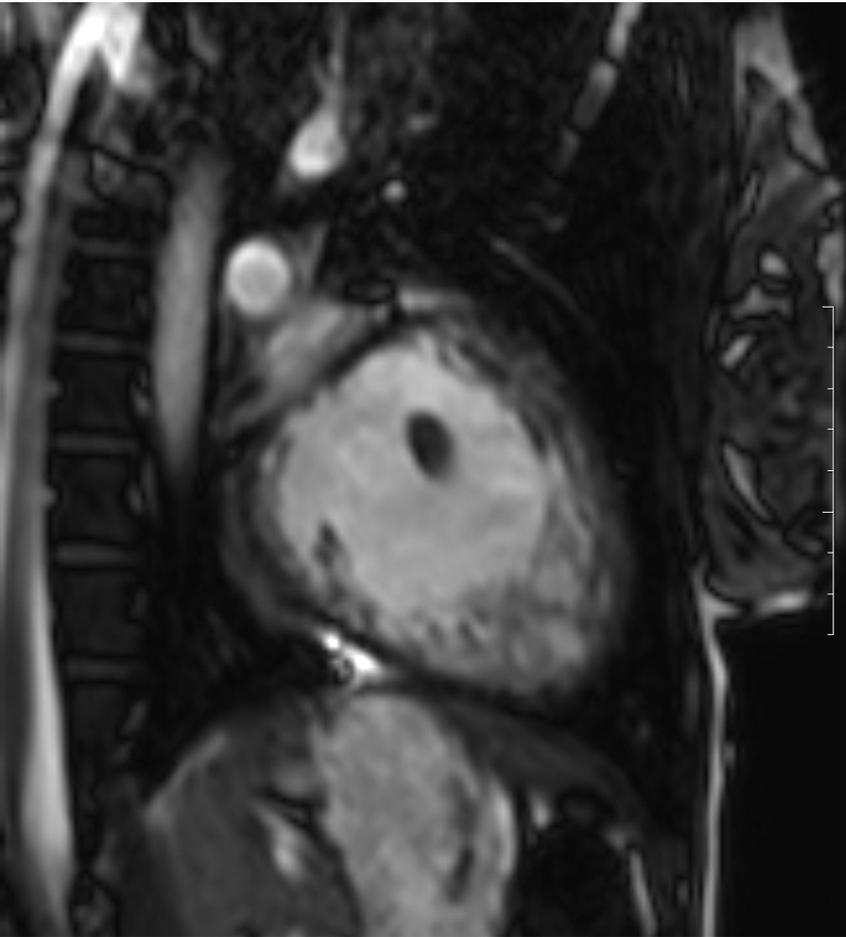


# NON COMPACTION VG

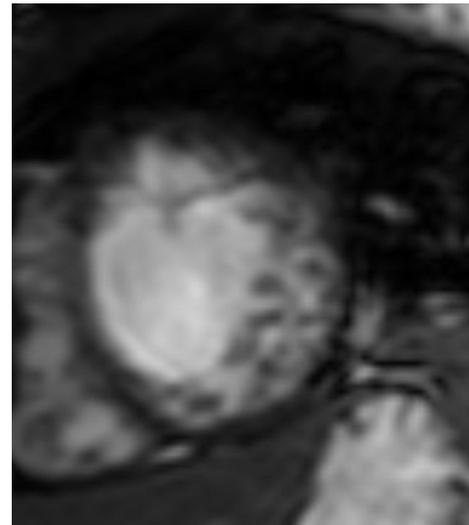


Baumhackel M. et al. *Circulation* 2002;106:e16-e17  
Vidéo accessible en ligne

# NON COMPACTION VG / IRM

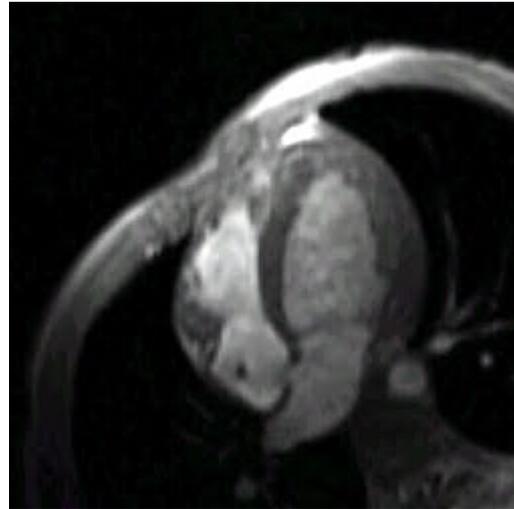
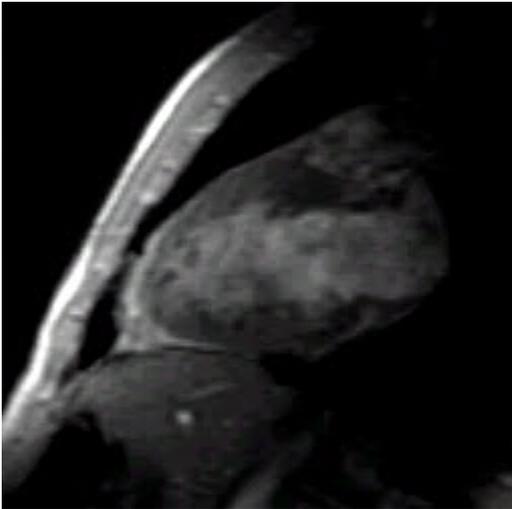


- Coupe long axe et petit axe
- couche compactée
- couche non compactée
- Récessus profonds entre les trabéculations mieux vus en diastole



# NON COMPACTION VG / IRM

## Piège diagnostique ??

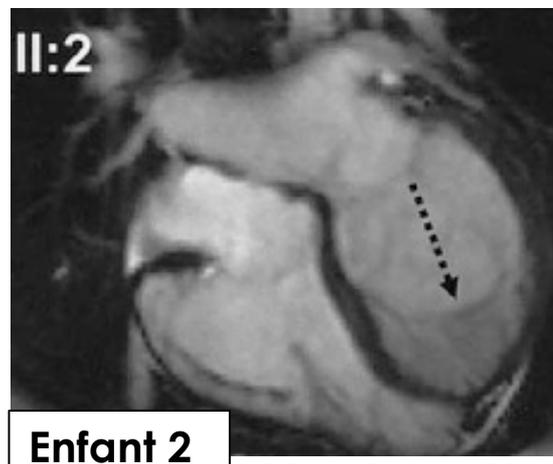
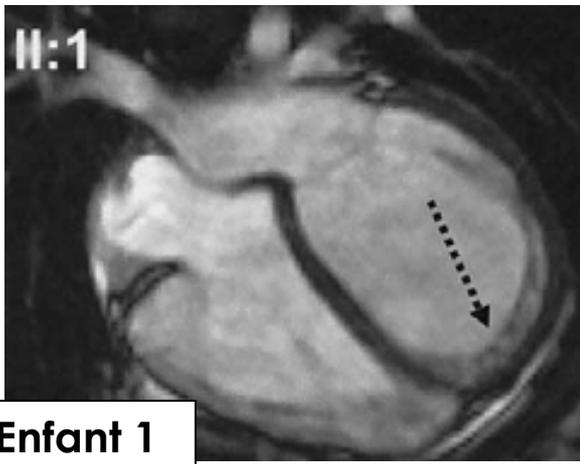
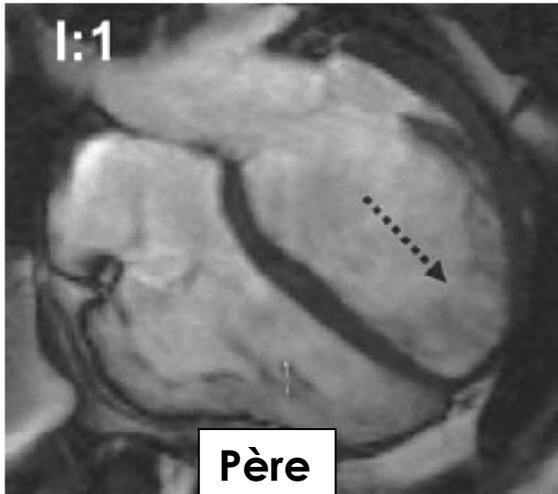


Borreguero, L. J.J. et al. *Circulation* 2002;105:e177-e178

Vidéo en ligne

MR gradient echo images are very sensitive to slow flow. The presence of deep apical recesses filled with blood that communicate with the ventricle (bright areas within the myocardium) is diagnostic of noncompacted myocardium. In contrast to diastolic images, systolic images do not show flowing blood within the myocardial wall because the trabeculations are very close to each other, and recesses are collapsed.

# NON COMPACTION VG / IRM



Forme autosomique dominante

Petersen SE  
JACC 2005

# NON COMPACTION VG / IRM

## Critère diagnostique en IRM

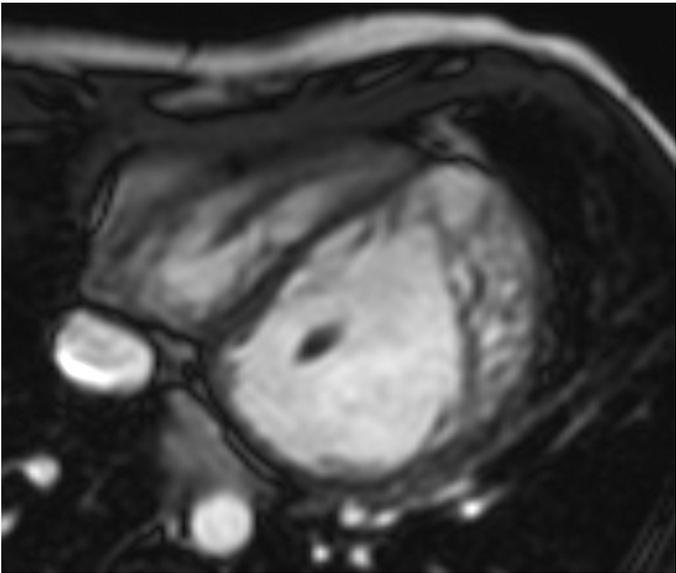
épaisseur couche non compactée / couche compactée >  
2.3 en télédiastole

Sensibilité	86%
Spécificité	99%
VPP	75%
<b>VPN</b>	<b>99%</b>

(En excluant le segment « apex » dans la classification en 17 segments)

Petersen SE  
JACC 2005

# IRM / NON COMPACTION VG



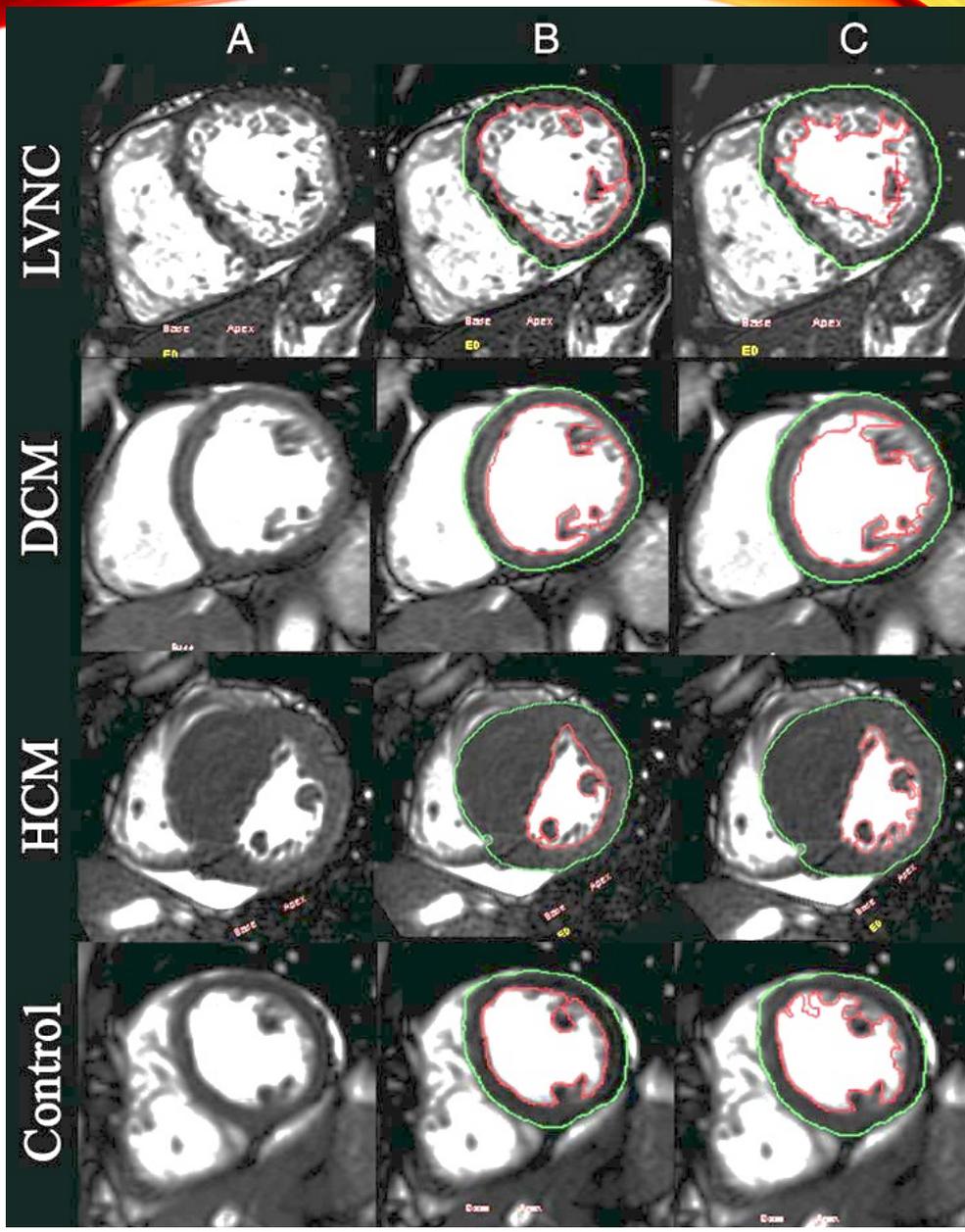
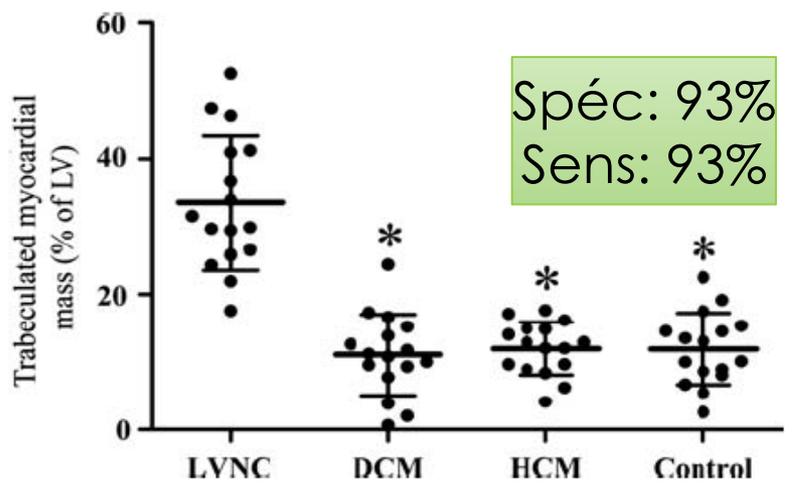
- IRM en écho de gradient, sang blanc
- Coupe 4 cavités montrant des trabéculations importantes de l'apex du ventricule gauche et de la paroi latérale.
- couche compactée
- couche non compactée
- Ratio  $> 2.3$  (en diastole)

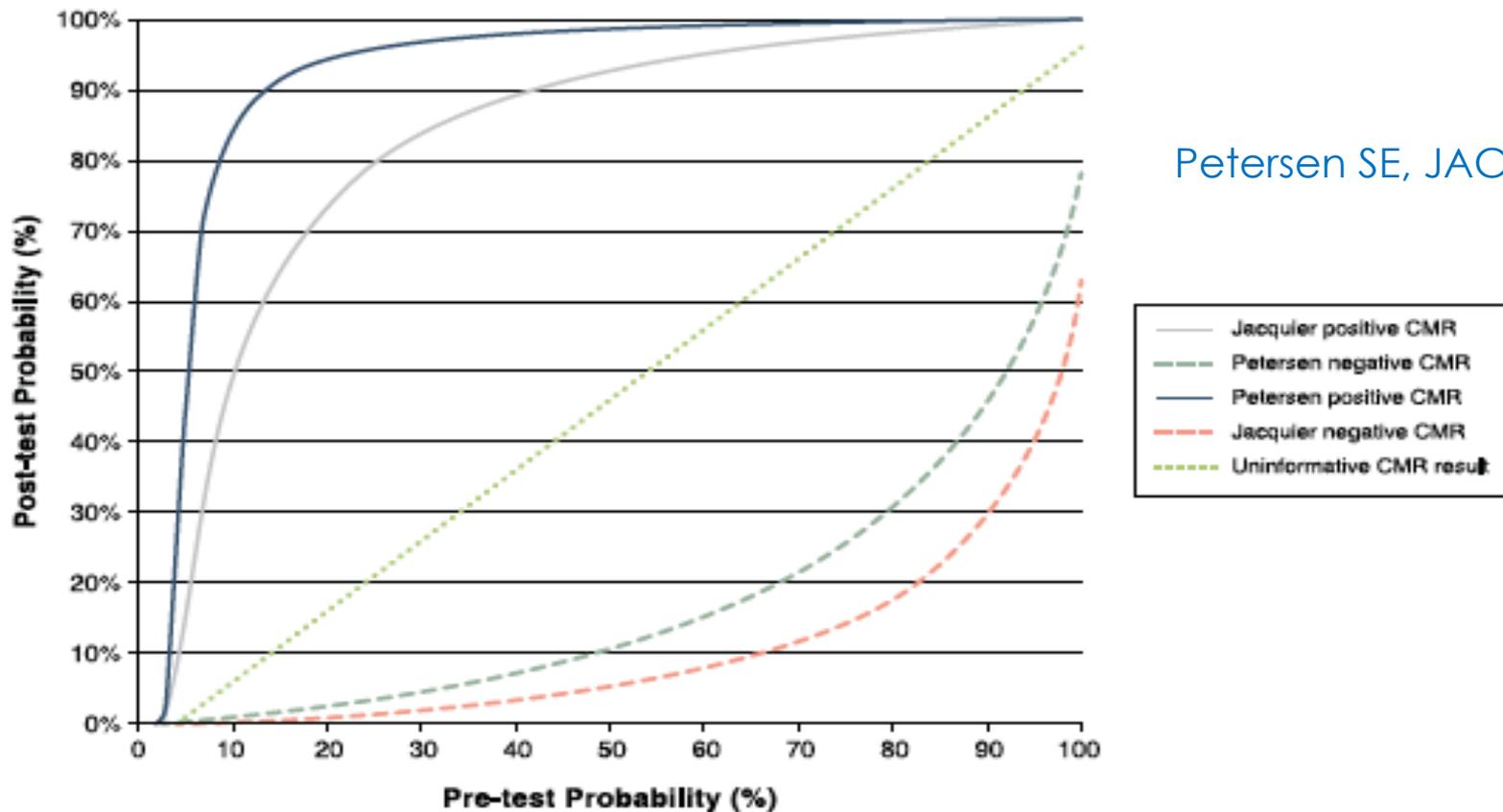
# CRITÈRES IRM / NON COMPACTION VG

- Trabéculatation ++ apex et paroi libre (partie moyenne et apicale) du VG
  - Au moins 3 trabéculations / plan de coupe
  - Entre l'apex et l'insertion des muscles papillaires
- Multiples recessus intra-trabéculaires
- Structure en 2 couches du myocarde (compactée et non compactée)
- **Rapport NC / C > 2.3** (ou 2.5 suivant les auteurs) en diastole mais faible spécificité (1)
- **Masse des trabéculations / masse du VG > 20%** en télédiastole (2)
- Masse des trabéculations / masse du VG > 40% en systole avec inclusion des piliers (3)

1. Attenhofer Jost CH and M.Connolly H. EHJ 2012
2. Jacquier A, et al. EHJ 2010
3. Stacey BR, et al. JACC Cvi 2013

- Masse des trabéculations / VG > 20%
- En télédiastole





Petersen SE, JACC Cvi 2013

**Figure 1. Relationship Between Pre- and Post-Test Probability for Published Sensitivities and Specificities to Diagnose LVNC**

Petersen criteria: sensitivity: 86%; specificity: 99%. Jacquier criteria: sensitivity: 93.7%; specificity: 93.7%. An uninformative test has a sensitivity and specificity of 50%. In very low pre-test probabilities consistent with the reported left ventricular noncompaction (LVNC) prevalence of 0.014% to 0.5%, neither cardiac magnetic resonance (CMR) criteria are very informative. In low and intermediate pre-test probabilities both CMR criteria are informative, one slightly better at ruling out disease and one better at ruling in disease.

# NON COMPACTION VG / IRM

**Séquence SSFP**

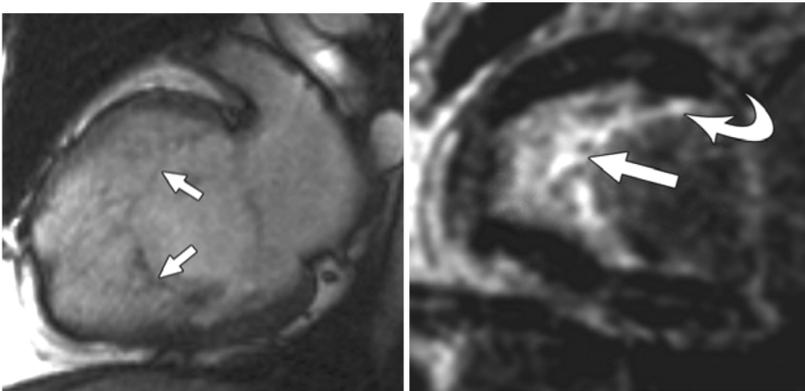
**Non compaction VG  
hypokinésie globale  
OG dilatée  
Auricule G ectasique**

**Dodd J. 2007 AJR  
Vidéo en ligne**

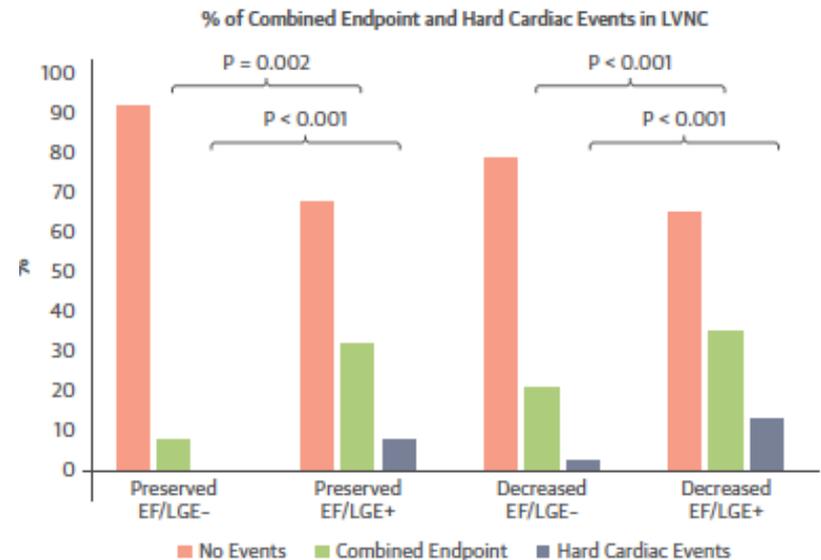


# LVNC ET RÉHAUSSEMENT TARDIF

- Réhaussement tardif % fibroélastose des couches non compactées
- % altération FEVG
- % au risque de TV ou mort subite
- **Très bon pronostic si LVNC avec absence de LGE et FEVG conservée.**



Mavrogeni S . Int J Cardiol 2012



## Report

- 1. Dimensions, mass (corrected for BSA) and function**
  - LV: EDV, ESV , SV, EF
  - Mass of non-compacted and compacted layer
- 2. Regional wall motion abnormalities**
- 3. Location and extent of segments with increased non-compacted to compacted (NC/C) myocardial ratio**

## Key Points

### 1. Current diagnostic criteria:

- **NC/C  $\geq$  2.3 : 1** on **end-diastolic** image\*

**Note:** NC/C 2:1 on end-systolic echo images

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- **Non-compacted LV mass above 20%** of the global mass

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### 2. **LGE** may represent **severe or late forms** of LVNC

### 3. **Diagnosis may not be based on imaging criteria alone**

- Often over-diagnosed, particular in DCM (thin compacted myocardium) and in patients of Afro-American descent
- Current diagnostic criteria may overdiagnose LVNC and new guidance is anticipated

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# NON COMPACTION VG: POINTS CLES

- Dépistage par écho cœur
- Intérêt de l'IRM ++ pour analyser l'ensemble des zones ventriculaires
- Intérêt de l'analyse familiale
- Attention aux critères diagnostiques
- Attention aux formes frontières
- Intérêt de la FEVG et du LGE et de la recherche de thrombus VG