Cardiomyopathie Hypertrophique 2011

Erwan DONAL

Cardiologie – CHU Rennes

erwan.donal@chu-rennes.fr

UNIVERSITÉ DE RENNES 1

LTSI

Instituts thématiques

Inserm

Institut national de la santé et de la recherche médicale

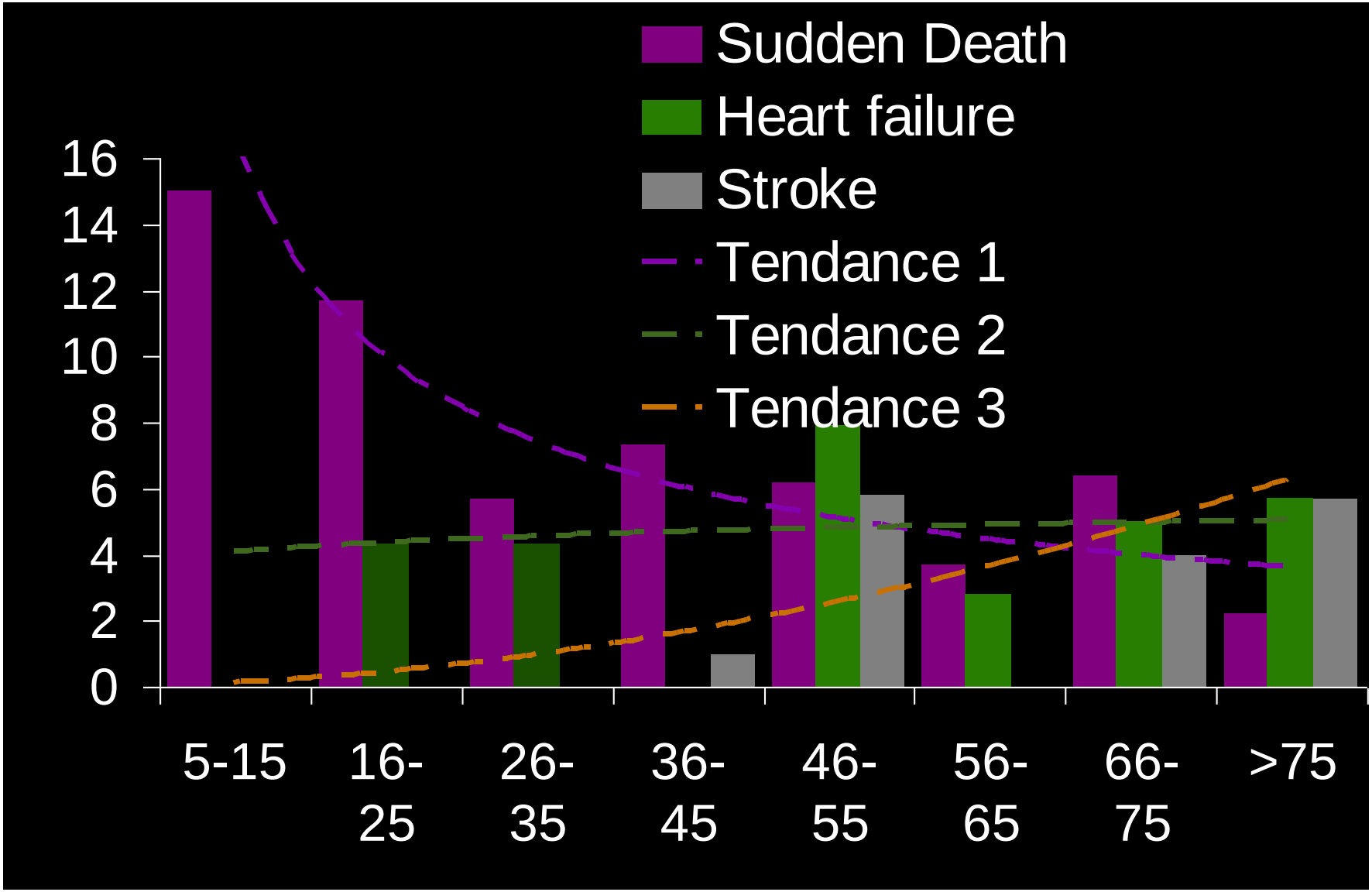
CIC Rennes IT

CHU Rennes

Epidémiologie

- 1ere cause de mort subite cardiaque chez les jeunes
- 1personne / 500
- Transmission autosomique dominante
- Conséquences variables

CMH Sévère: 3 Complications Majeures



Maron et al. Circulation. 2000

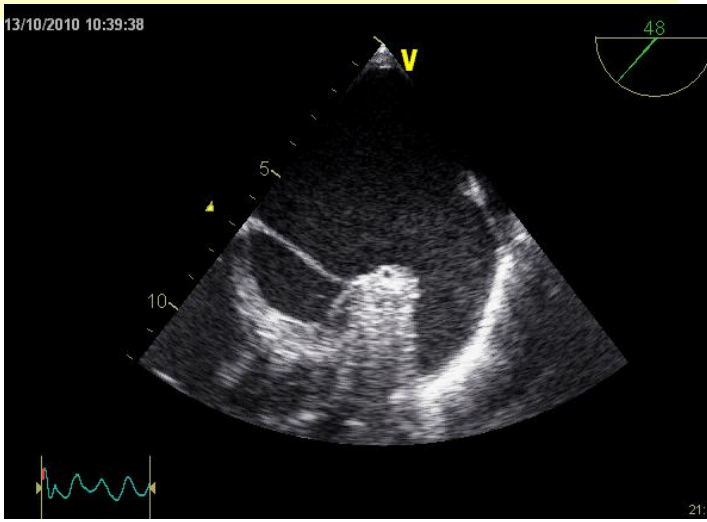
Who is At Risk ?
Major Risk Factors to Sudden Cardiac Death in HCM

- **History of cardiac arrest (VF)**
- **Spontaneous sustained VT**

- **Family history of premature HCM-related SCD**
- **High-risk mutation (closely linked to family history)**
- **Unexplained syncope**
- **LV thickness ≥ 30mm**
- *Abnormal exercise response*
- *Non-sustained VT (Holter): 20-30% of HCM patients*

Maron BJ, McKenna WJ et al JACC 2003

CMH
Maladie Hétérogène



Stabilité
Evolution Bénigne

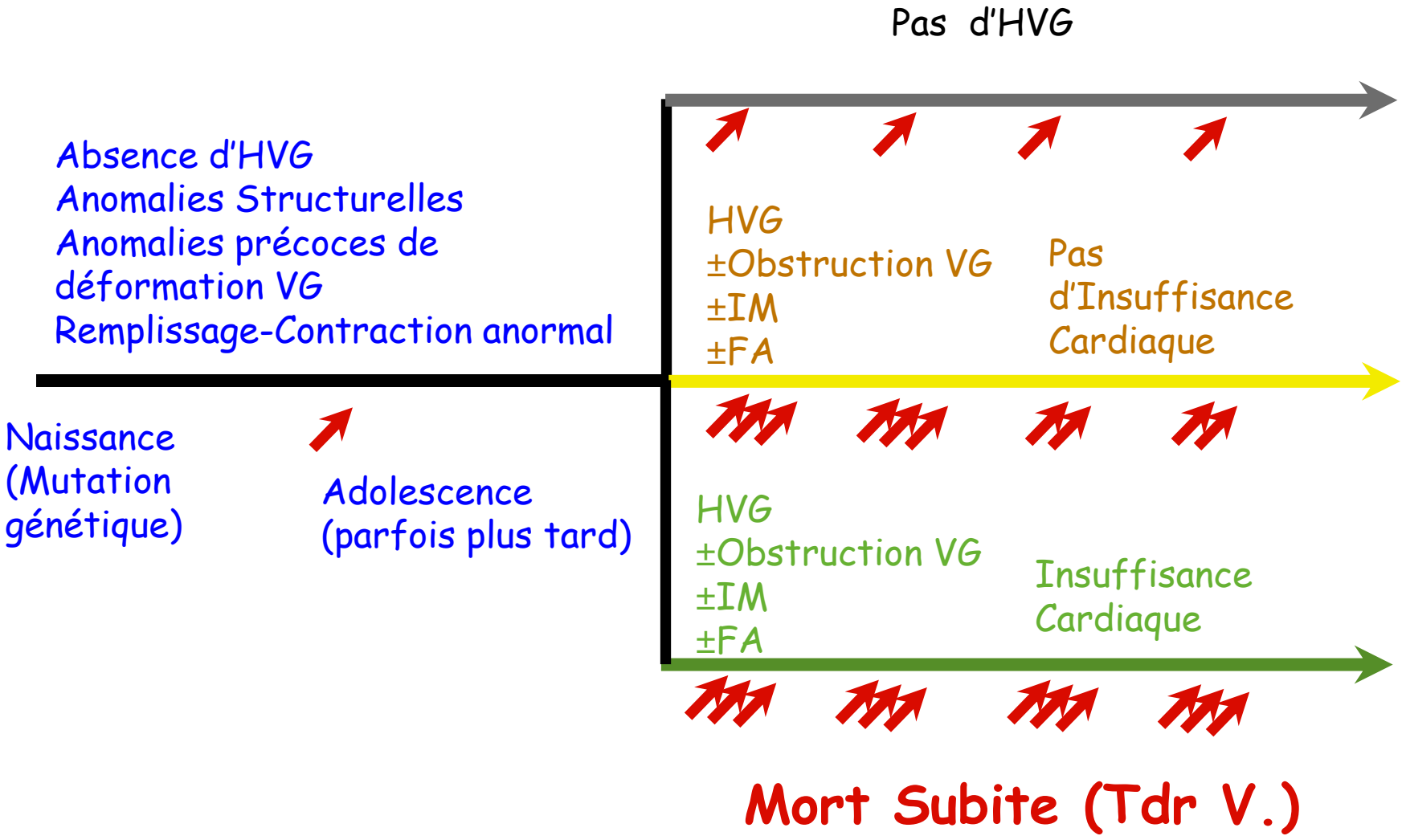
Profils Pronostiques des CMH

Ins.Cardiaque

Mort Subite

FA-AVC

Histoire Naturelle Hétérogène



Reconnaître une CMH Sévère

Apport de L'Echo et de l'IRM

Hypertrophie VG souvent asymétrique, sans dilatation ventriculaire et sans cause décelable qui ferait évoquer une HVG secondaire (HTA, RAo, amylose,…)

Epaisseur pariétale ≥15mm

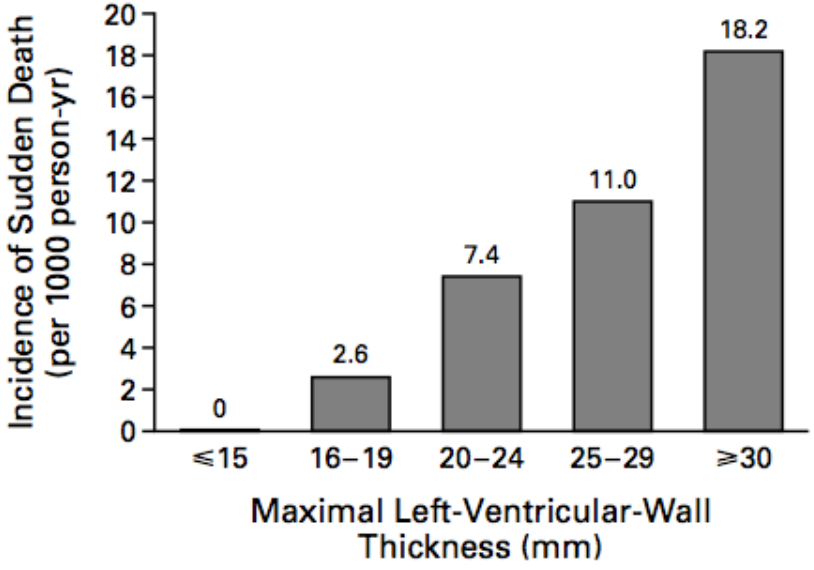
Mais toutes les épaisseurs peuvent être compatibles avec la présence d'un gène mutant de CMH

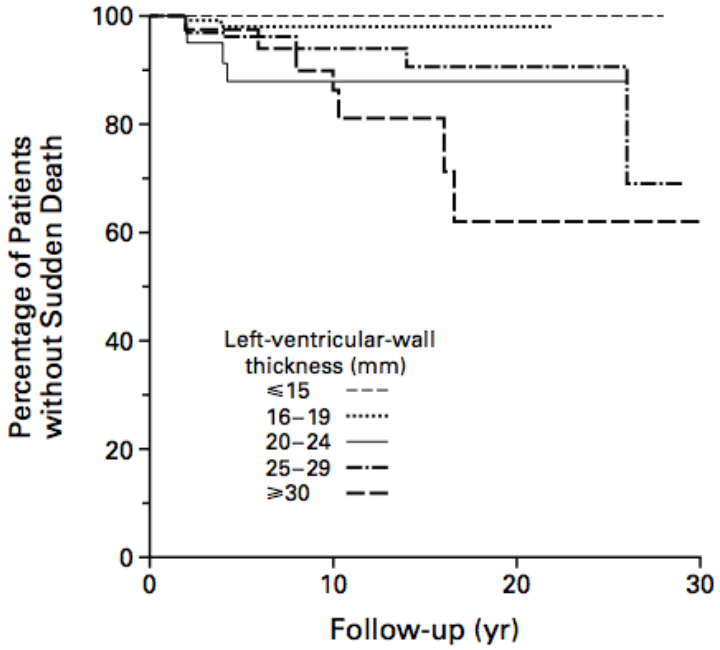
Donc **Epaisseur ≥13 mm** suffit pour les **apparentés du 1er degré**

Reconnaître une CMH Sévère
Apport de L'Echo

et de l'IRM

HVG: Critère Pronostique Majeur *(≥30mm)*





Spirito et al. New Engl J Med. 2000

Reconnaître une CMH Sévère
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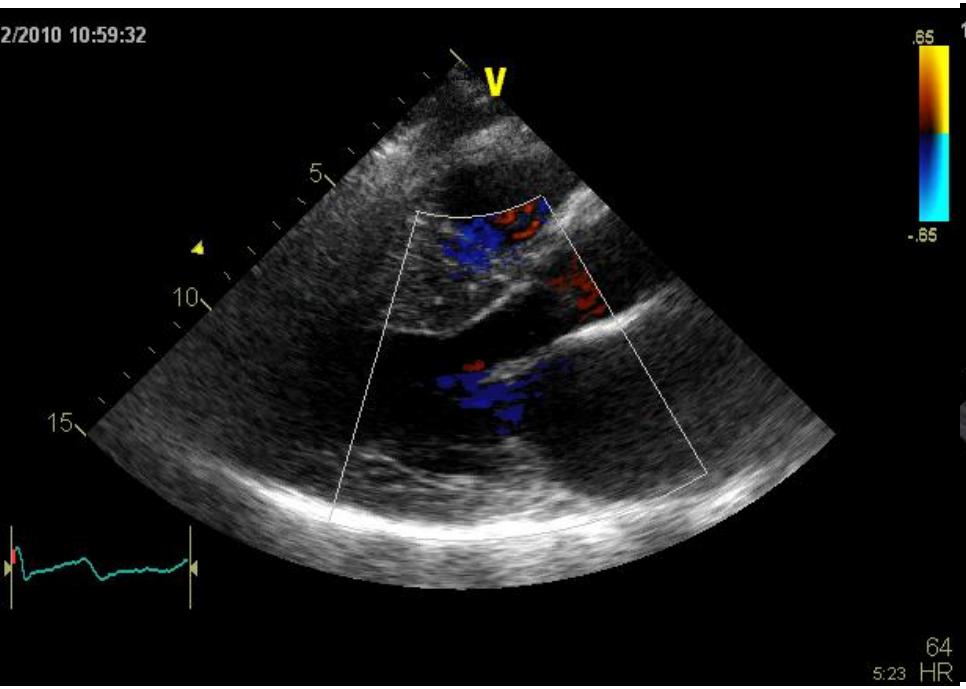
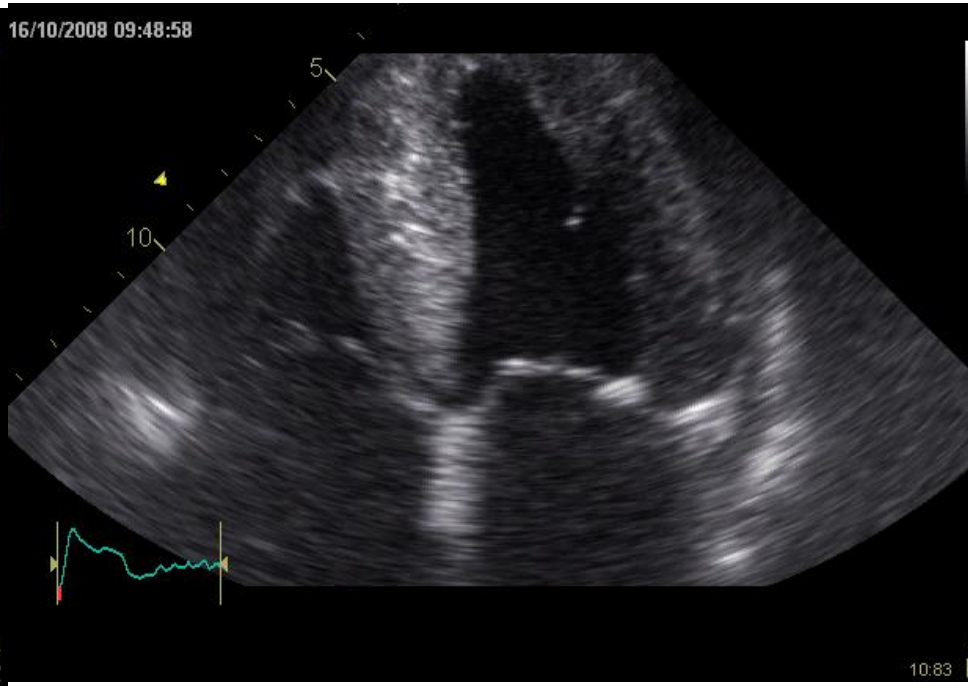




Répartition Variable
de l'Hypertrophie

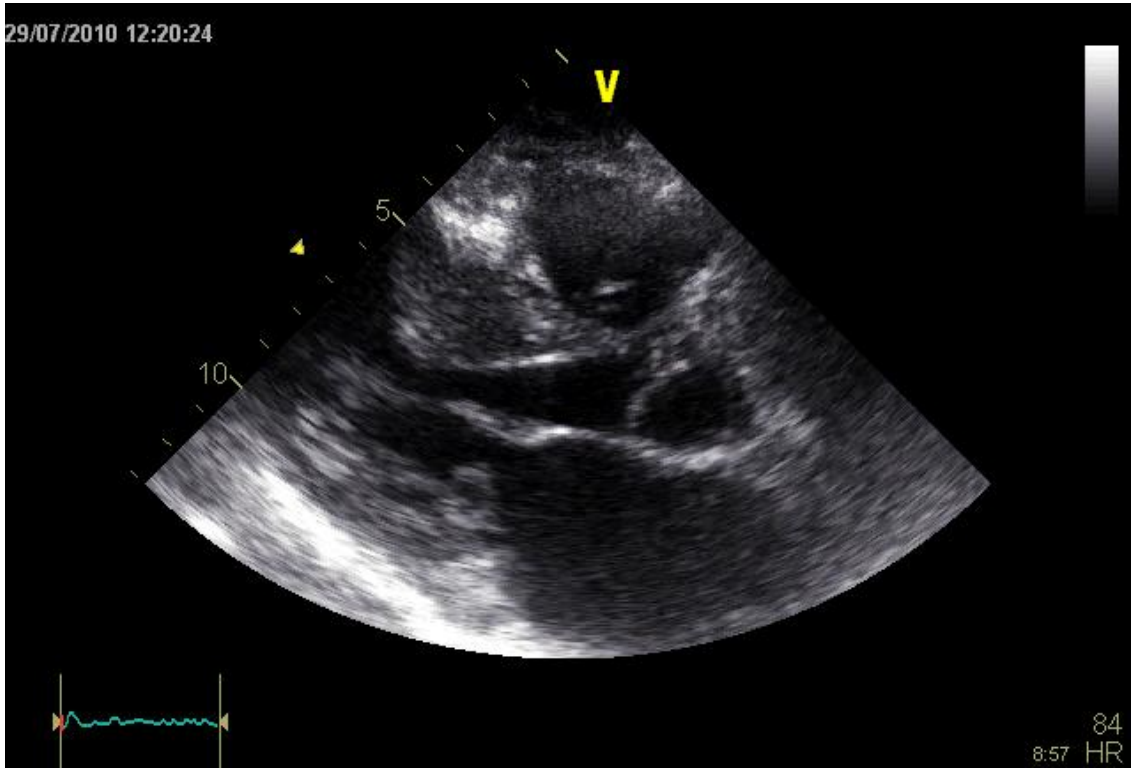
Reconnaître une CMH Sévère
Apport de L'Echo et de l'IRM

HVG: Critère Pronostique Majeur *(≥30mm)*

Pièges de Mesures

Masse VG mode TM non valable car HVG asymétrique
Coupe pariétale oblique

Piège du myocarde VD



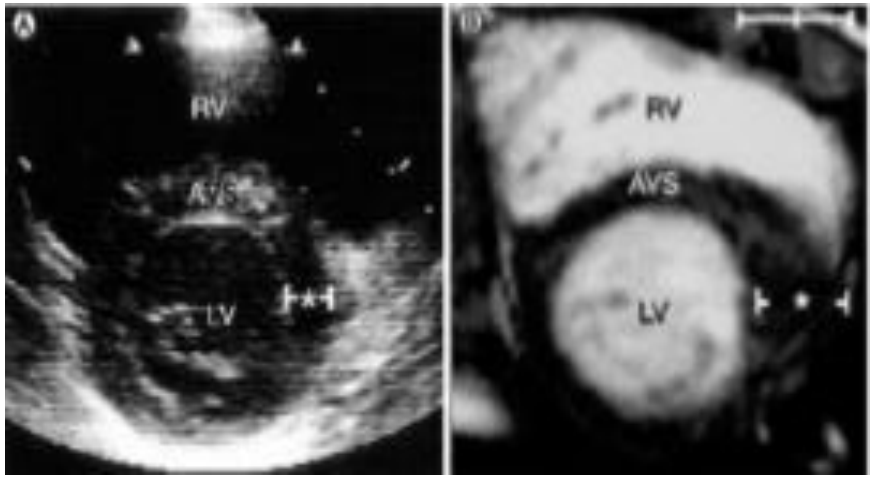
Interêt non exeptionnel d'une injection de Sonovue

Reconnaître une CMH Sévère
Apport de L'Echo et de l'IRM

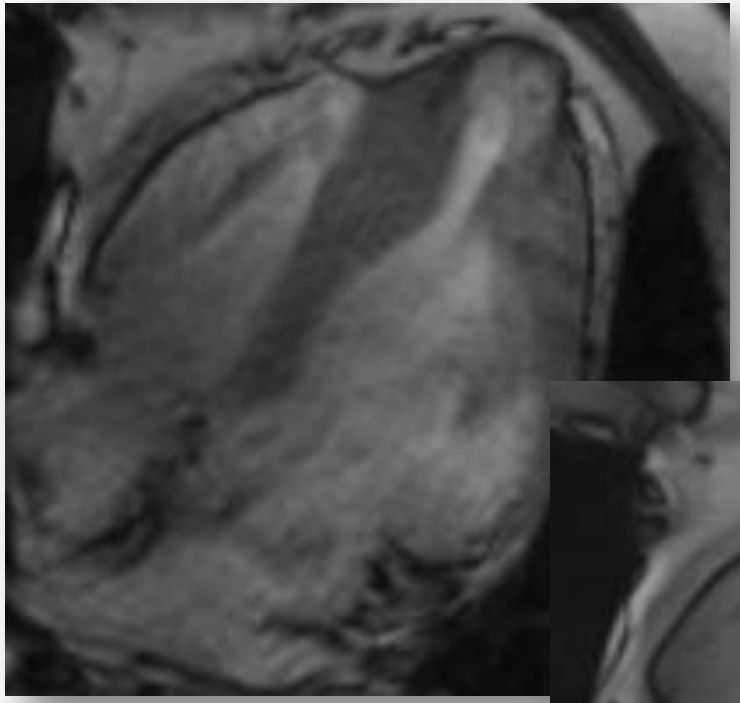
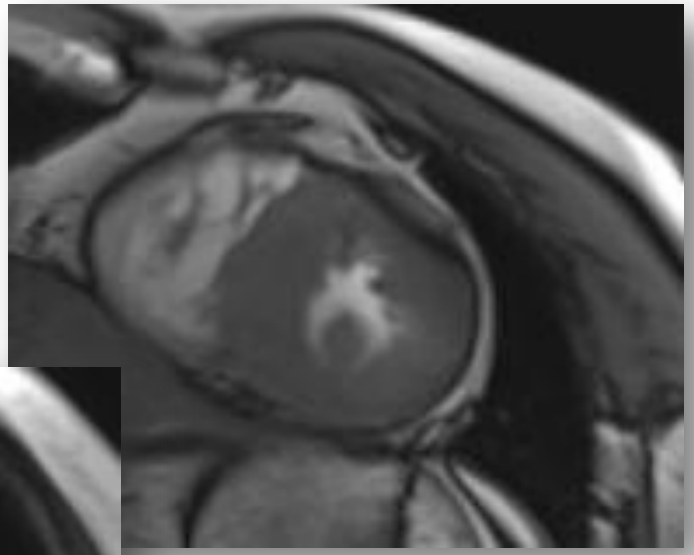
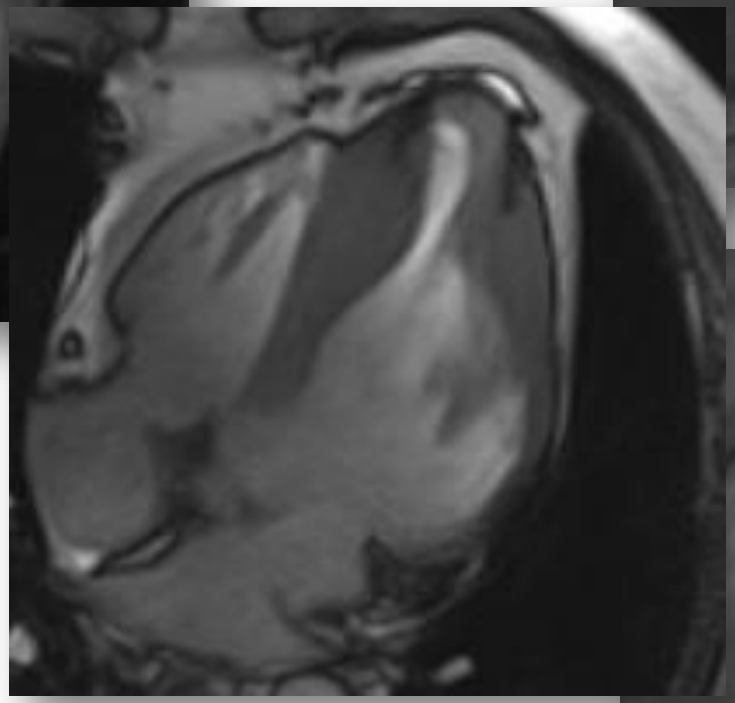
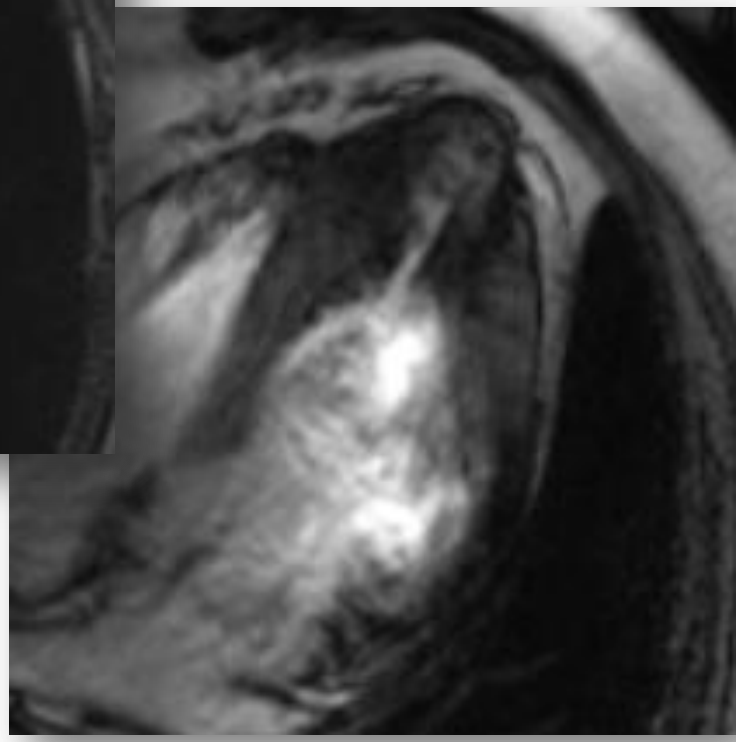
HVG: Critère Pronostique Majeur *(≥30mm)*

Formes Localisées

- Echo - vs. IRM +: 6% de cas
- Sous-estimation echo >3cm: 10% de cas
- Limites de la résolution spatiale de l'echo dans les formes antéro-latérales



Rickers et al. Circulation 2005;112:855.

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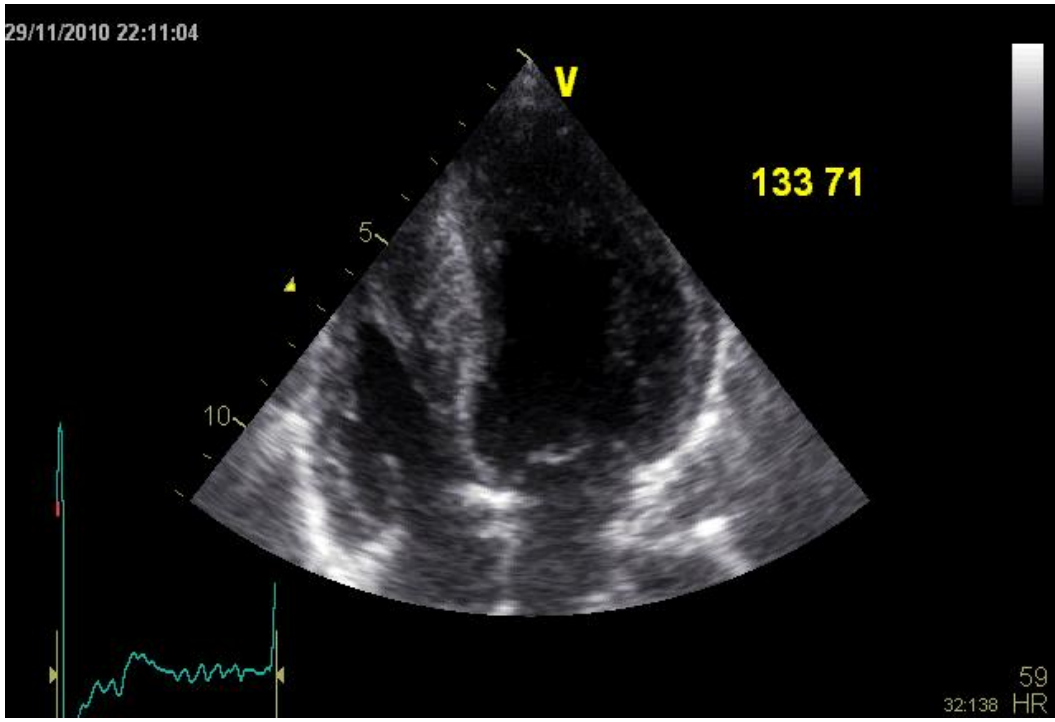
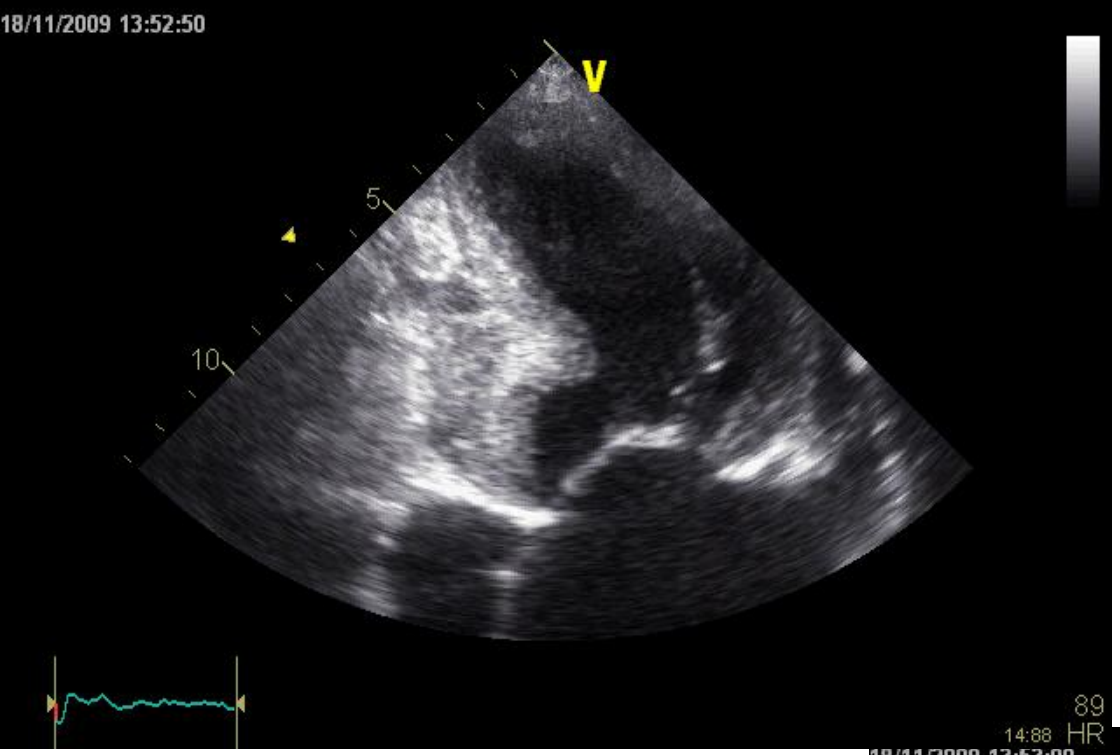
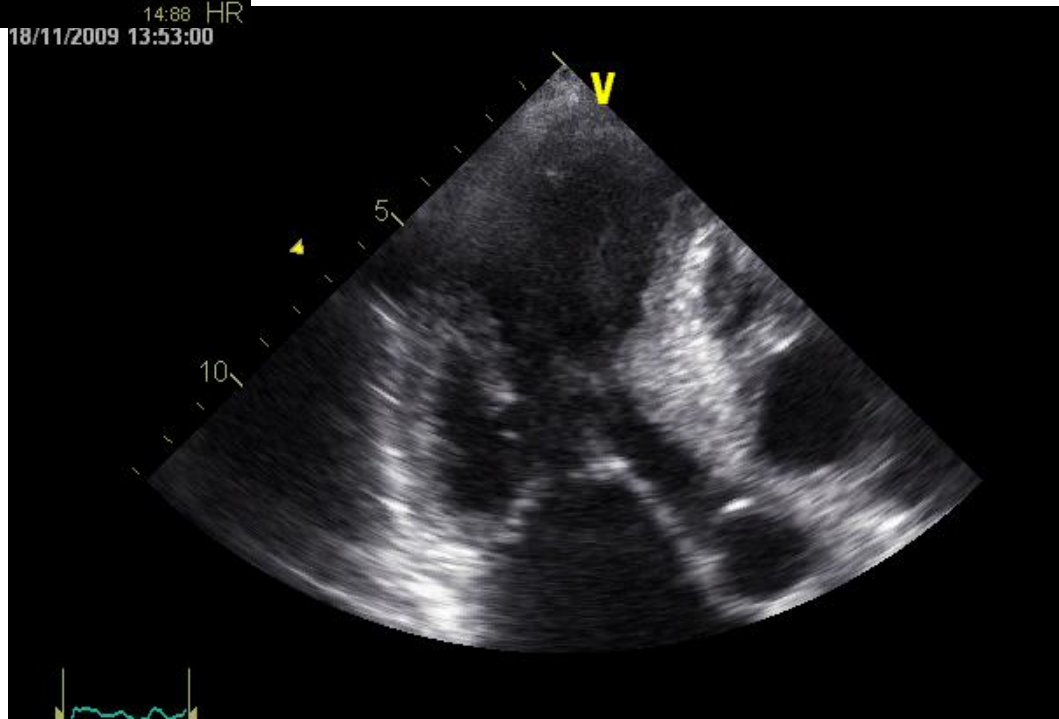



18/11/2009 13:52:50
V
5
10
89
14:88 HR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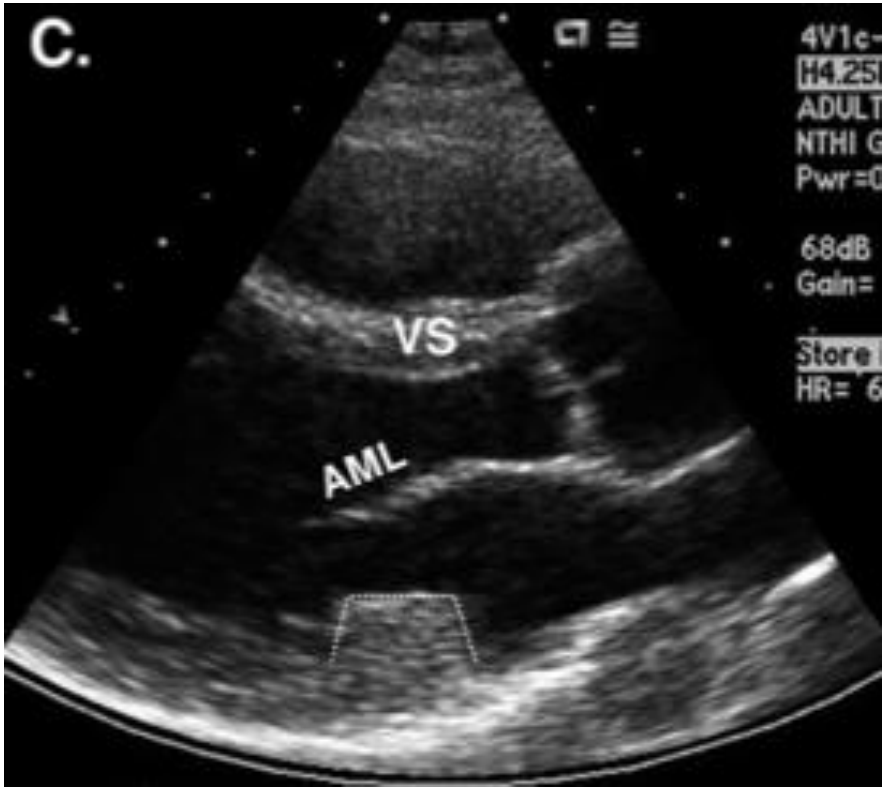
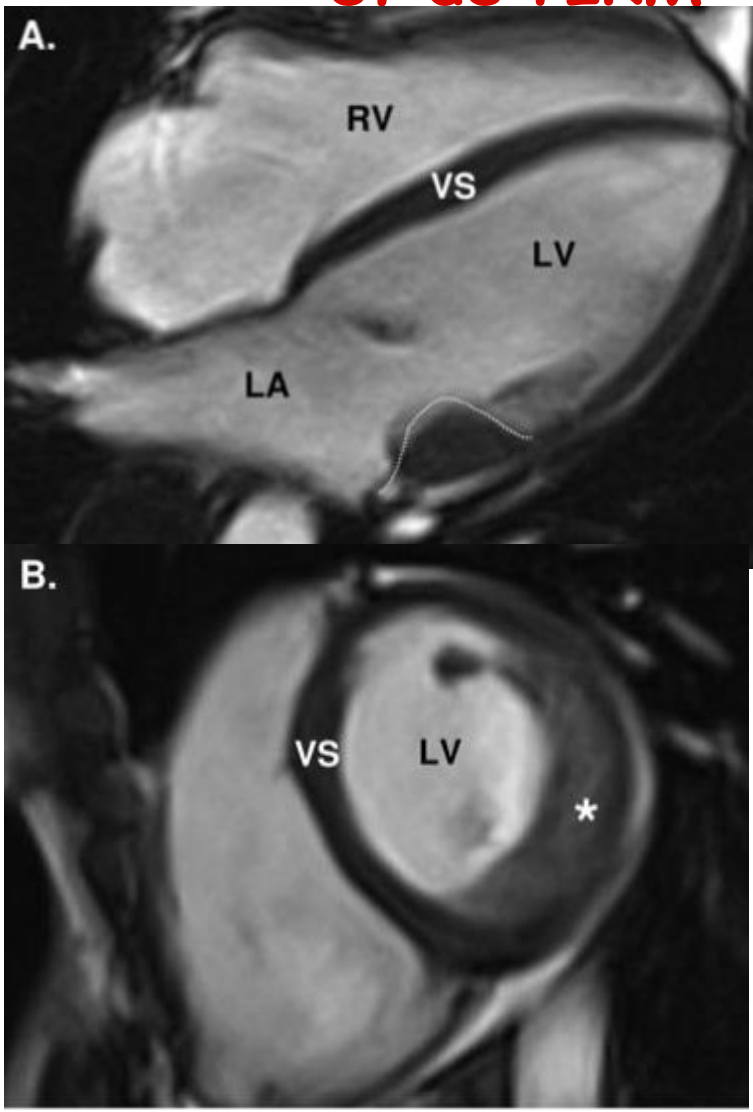
10

Reconnaître une CMH Sévère
Apport de L'Echo

et de l'IRM



Formes Postéro-basales

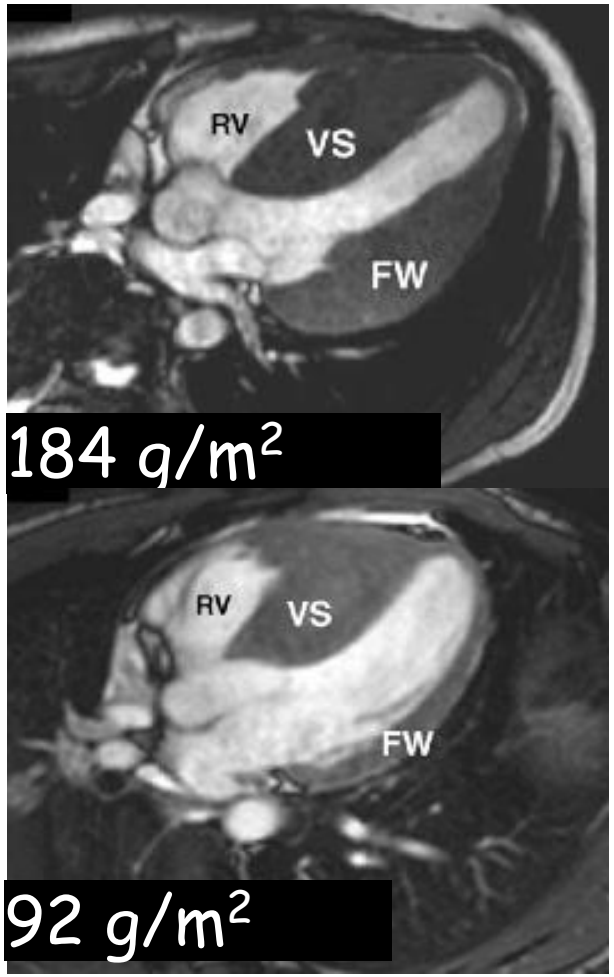


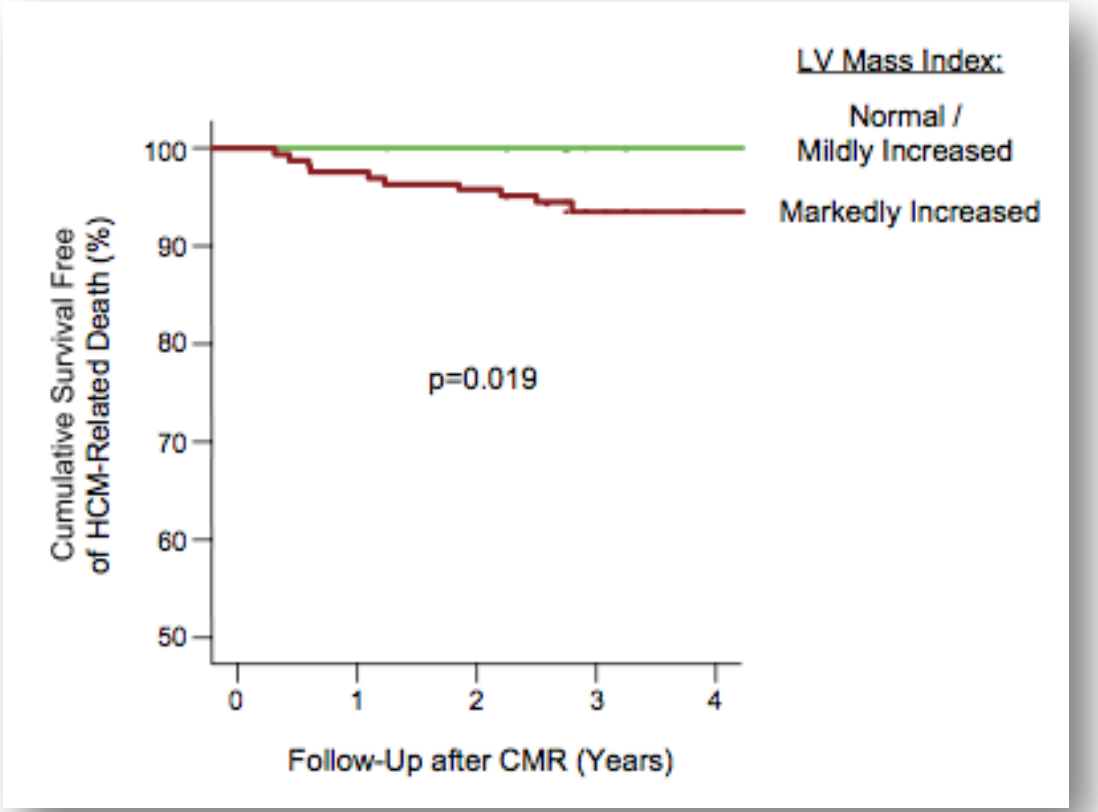
Maron BJ et al. Am J Cardiol 2010

La Masse

- Normale: 20%
- Non valable en Echo TM
- Echo 3D plus appropriée
- IRM: valeur pronostique

(moins spécifique que l'épaisseur)



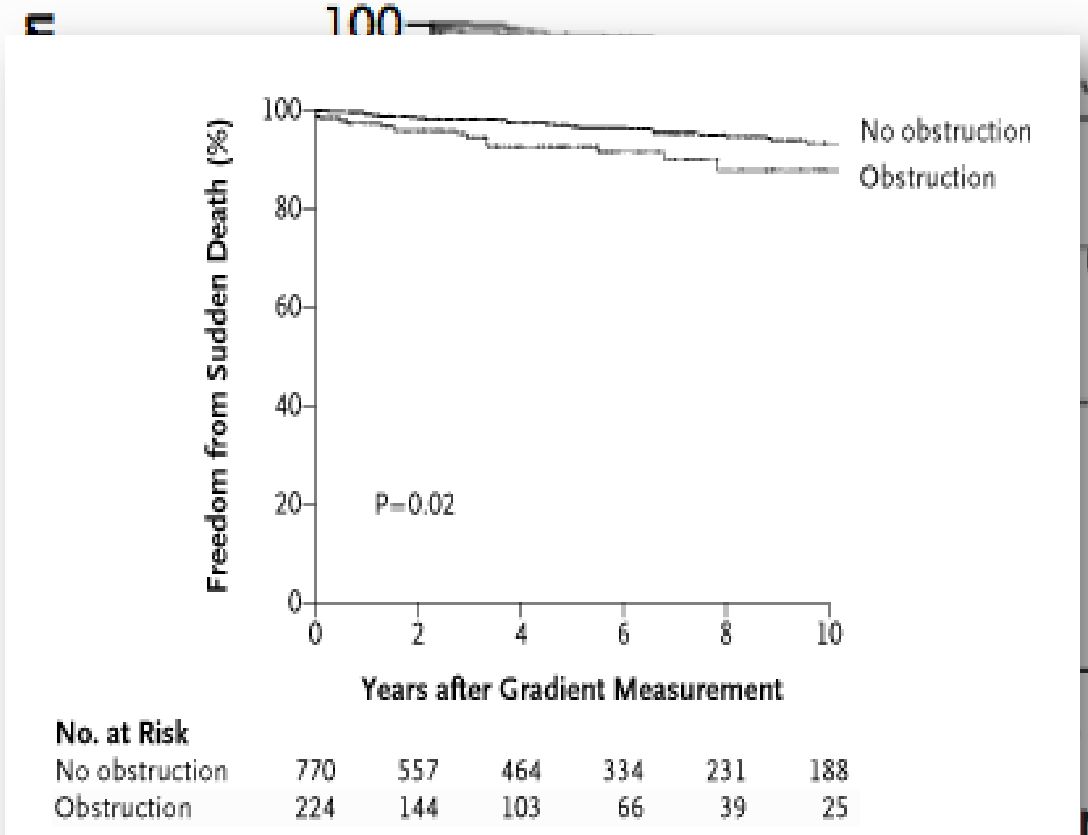


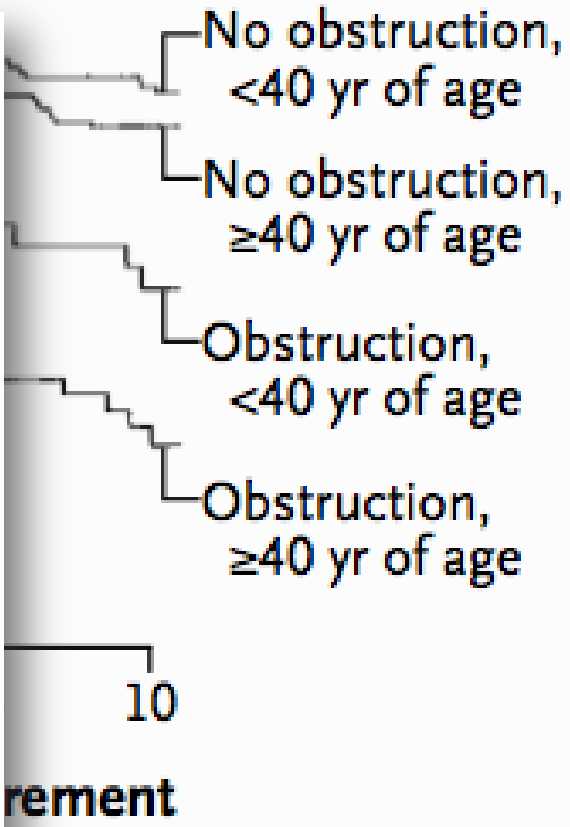
Olivotto et al. J Am Coll Cardiol 2008

Obstruction VG

Critère Pronostique Mineur de Mort Subite

Critère Majeur d'Ins.Cardiaque et AVC



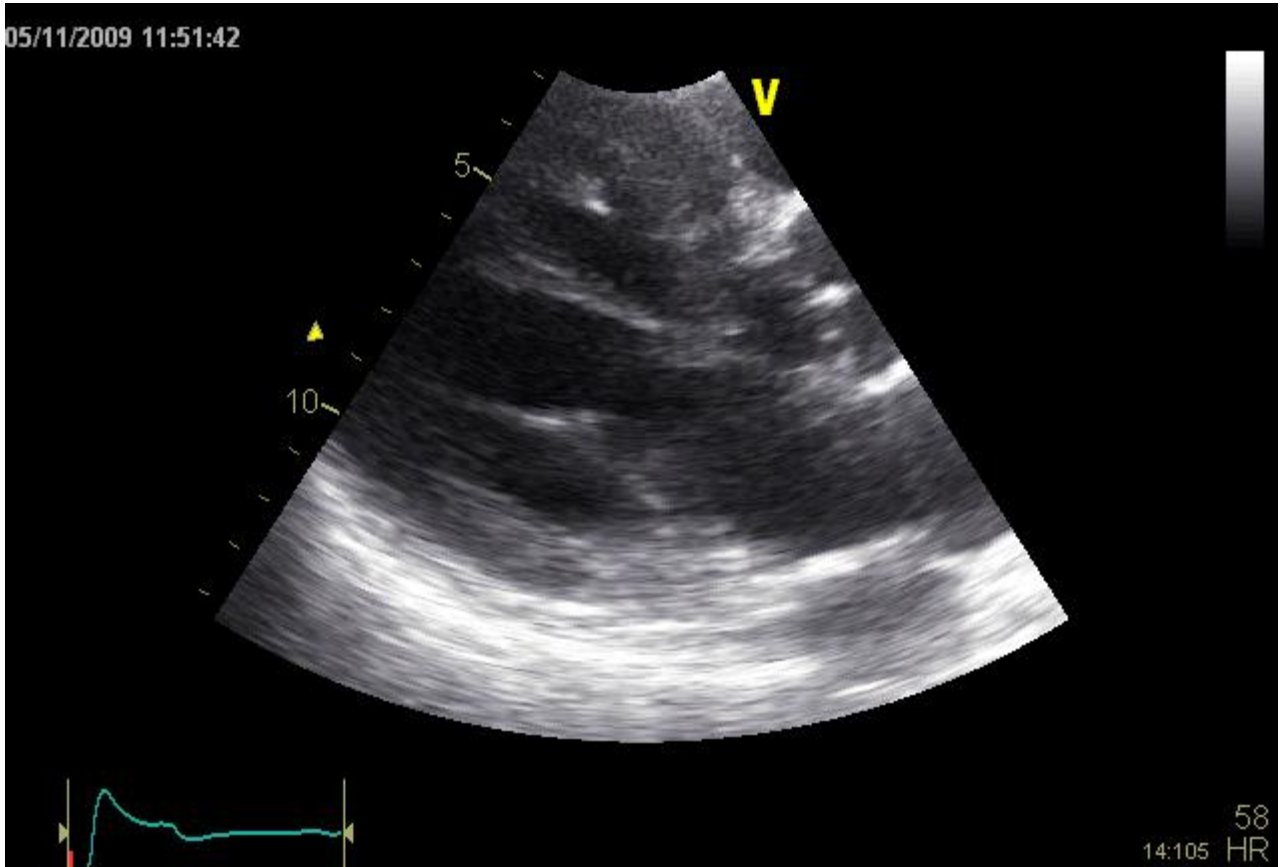


Maron et al. New Engl J Med 2003

Obstruction VG

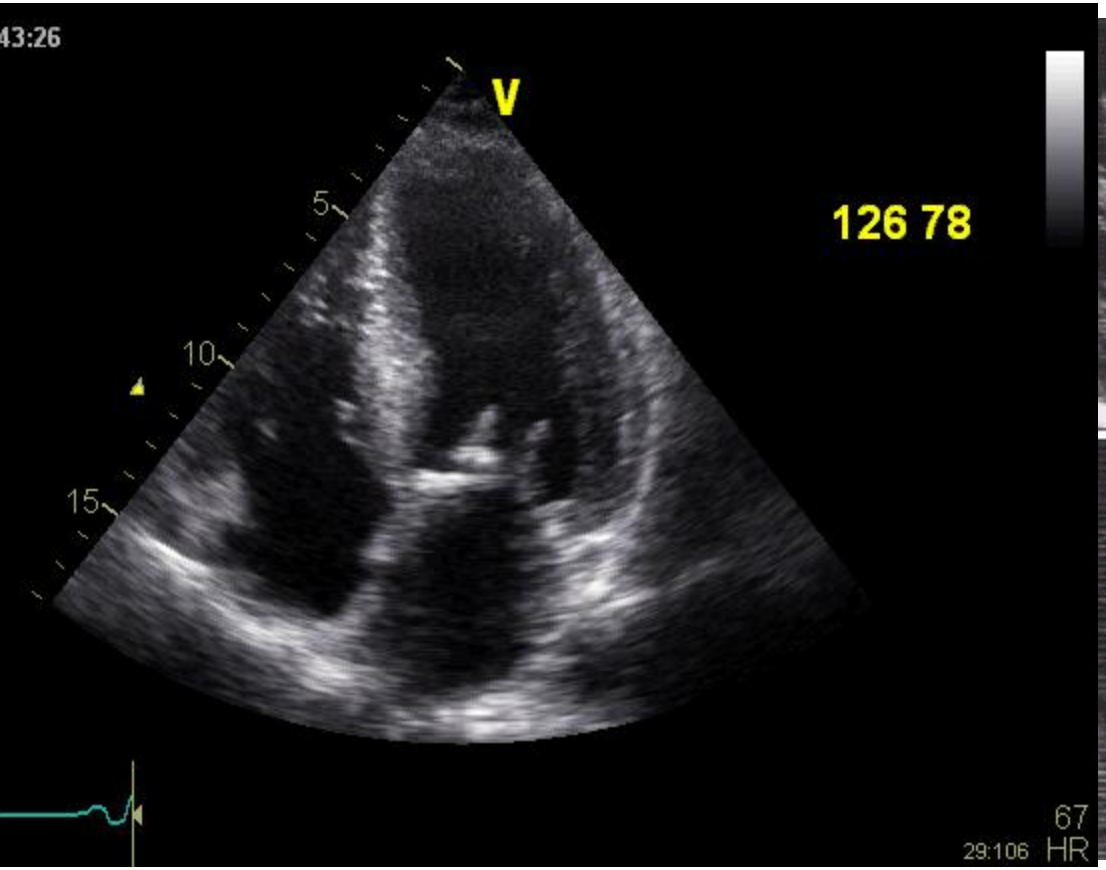
Mécanismes

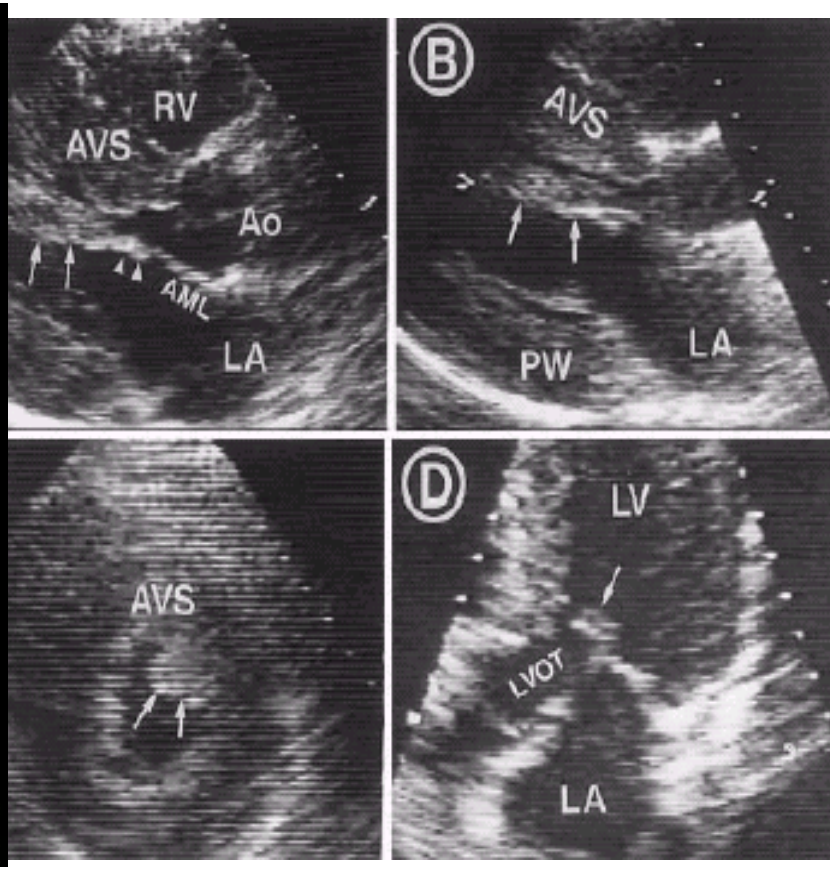
Le plus fréquent: *Hypertrophie du septum basal + Mouvement Systolique antérieur de la valve mitrale (SAM)*



Autres Mécanismes: OBSTRUCTION MEDIO-VENTRICULAIRE

Autres Mécanismes: INSERTION DU PILIER SUR FEUILLET MITRAL





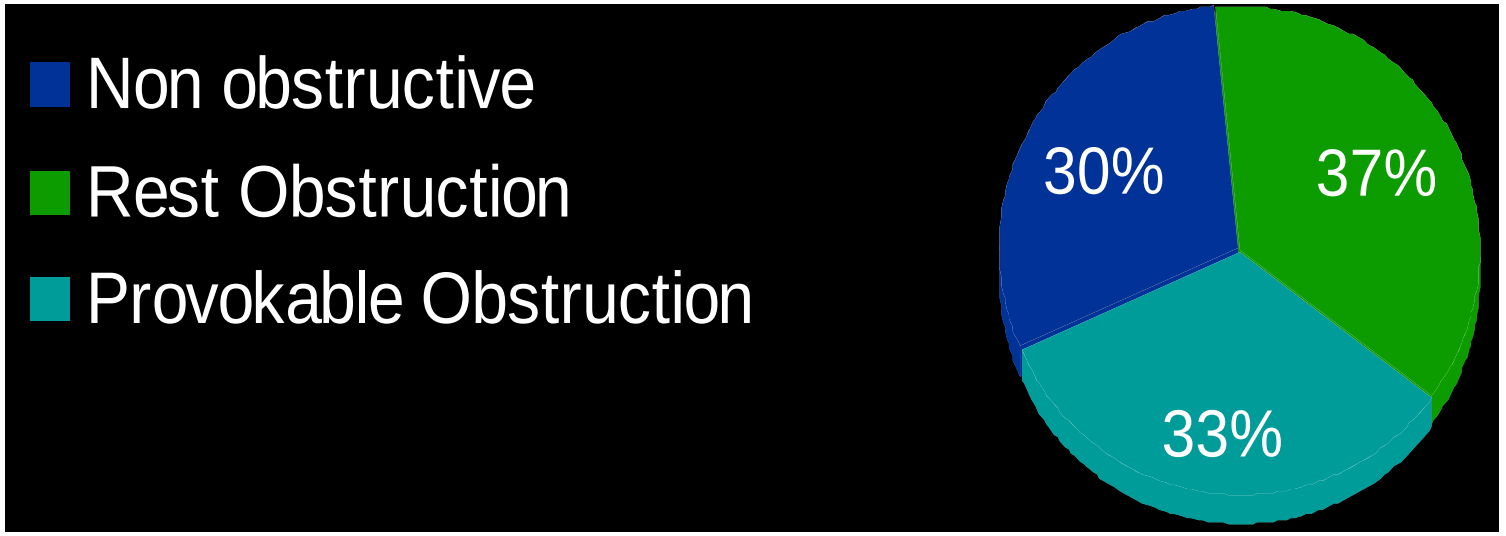
Maron et al. Circulation 1998

ECHO D'EFFORT

- L'obstruction est un phénomène dynamique
- Echo d'effort chez le patient symptomatique on considère

30 mmHg au repos et Gmax > 50mmHg à l'effort

- 70% des patients ont une obstruction de repos ou d'effort



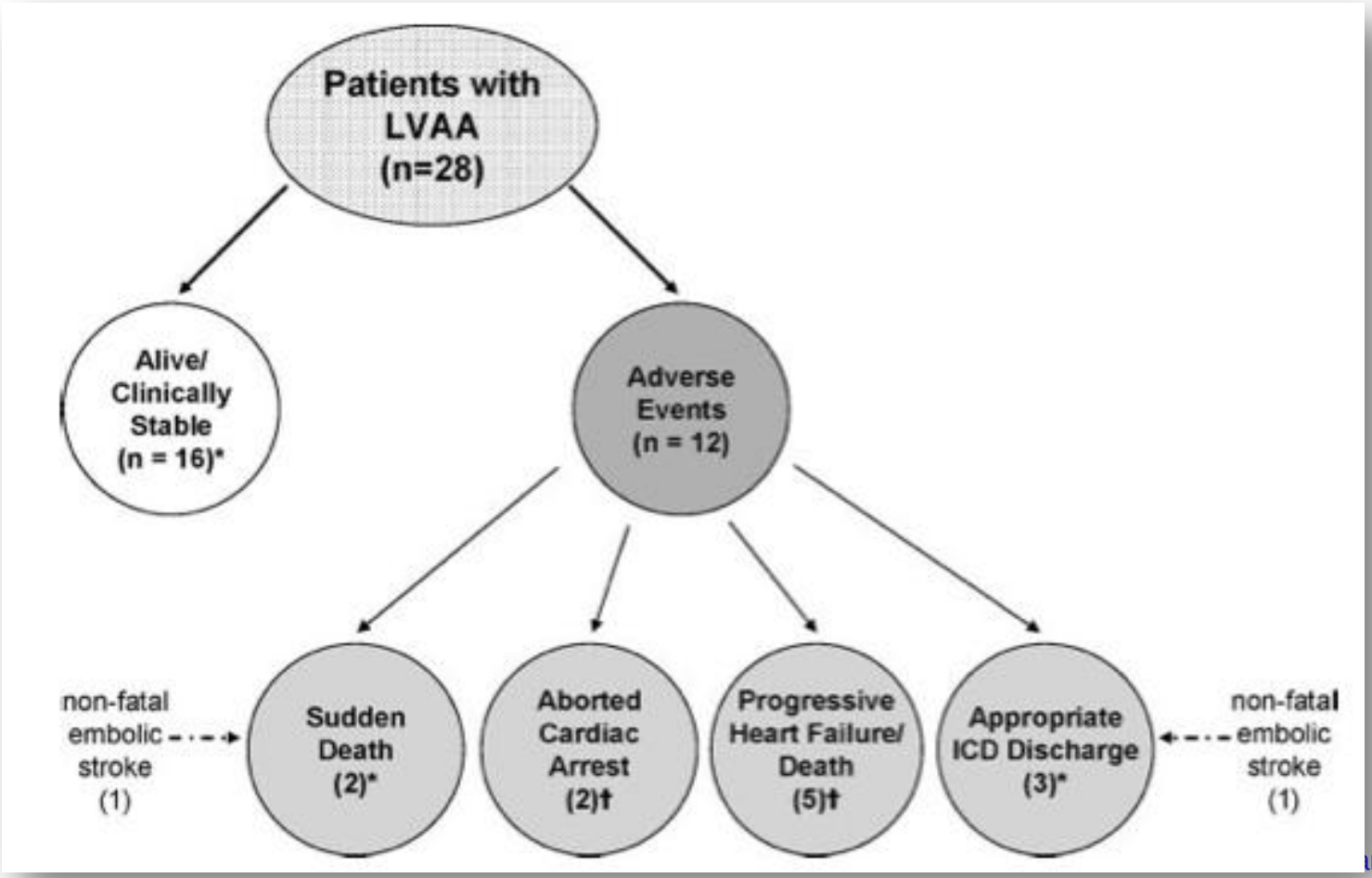
70%

Maron et al. Circulation 2006

Anévrysme VG des Formes Médio-Ventriculaires

Mauvais Pronostic
Ev.nts: 10.5%/ an

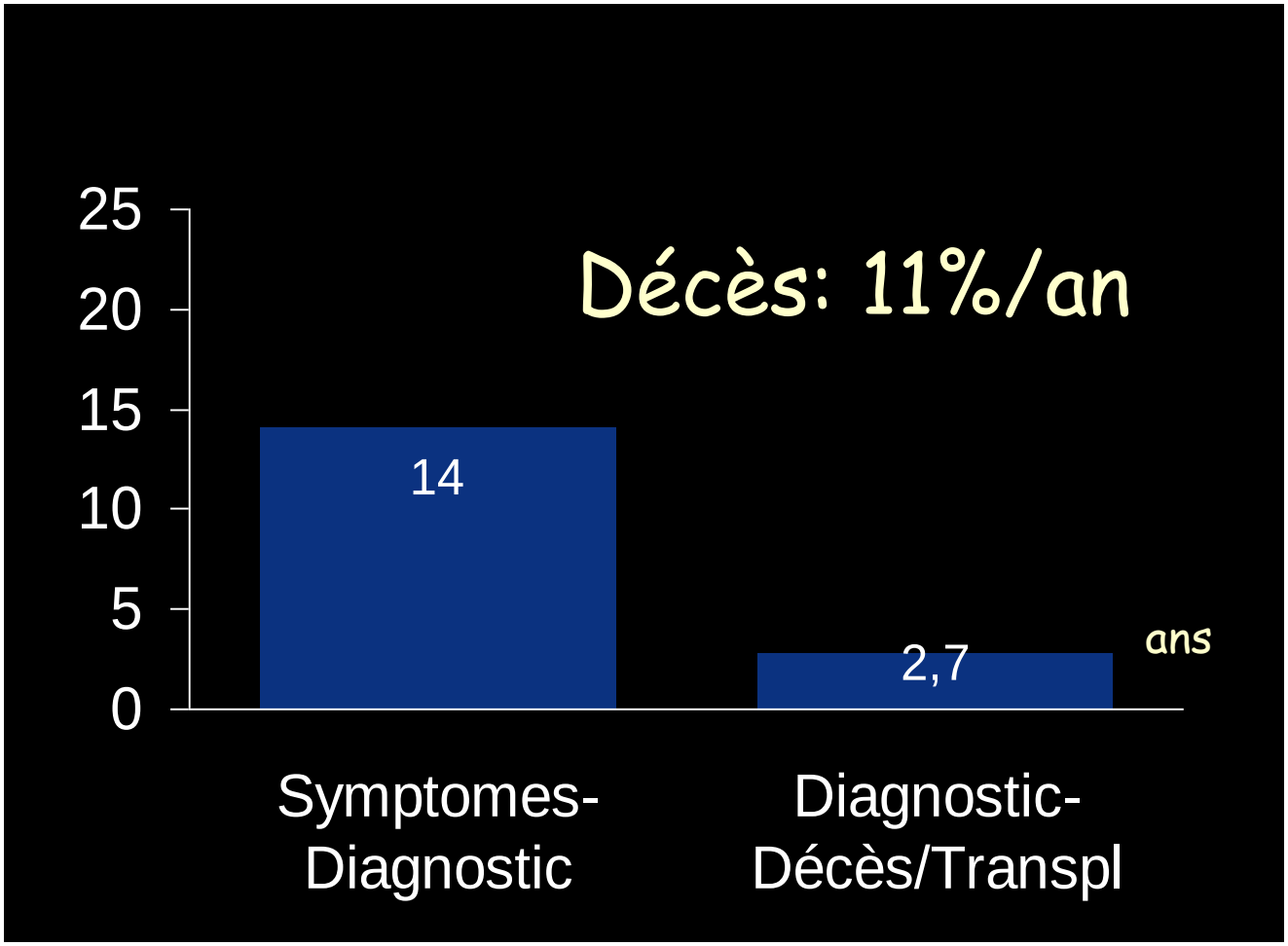
Echo ne voit pas 43% des Anévrysmes: IRM



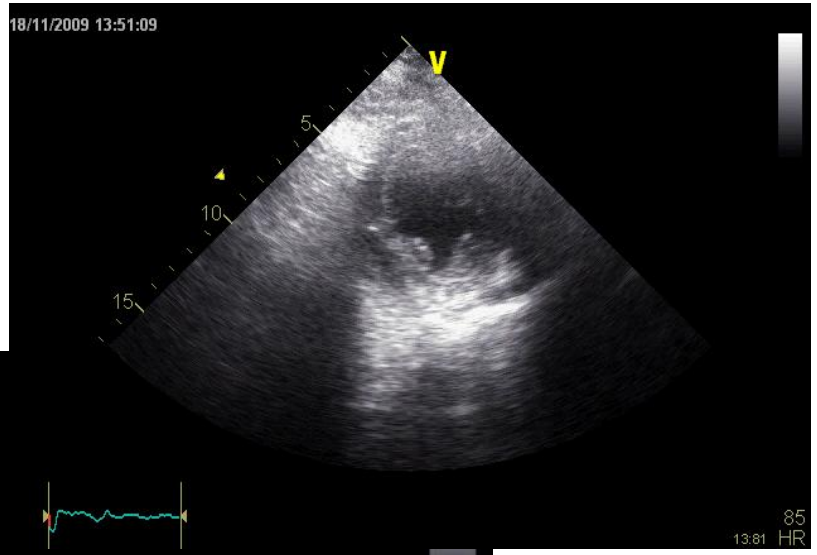
aron et al. Circulation 2008

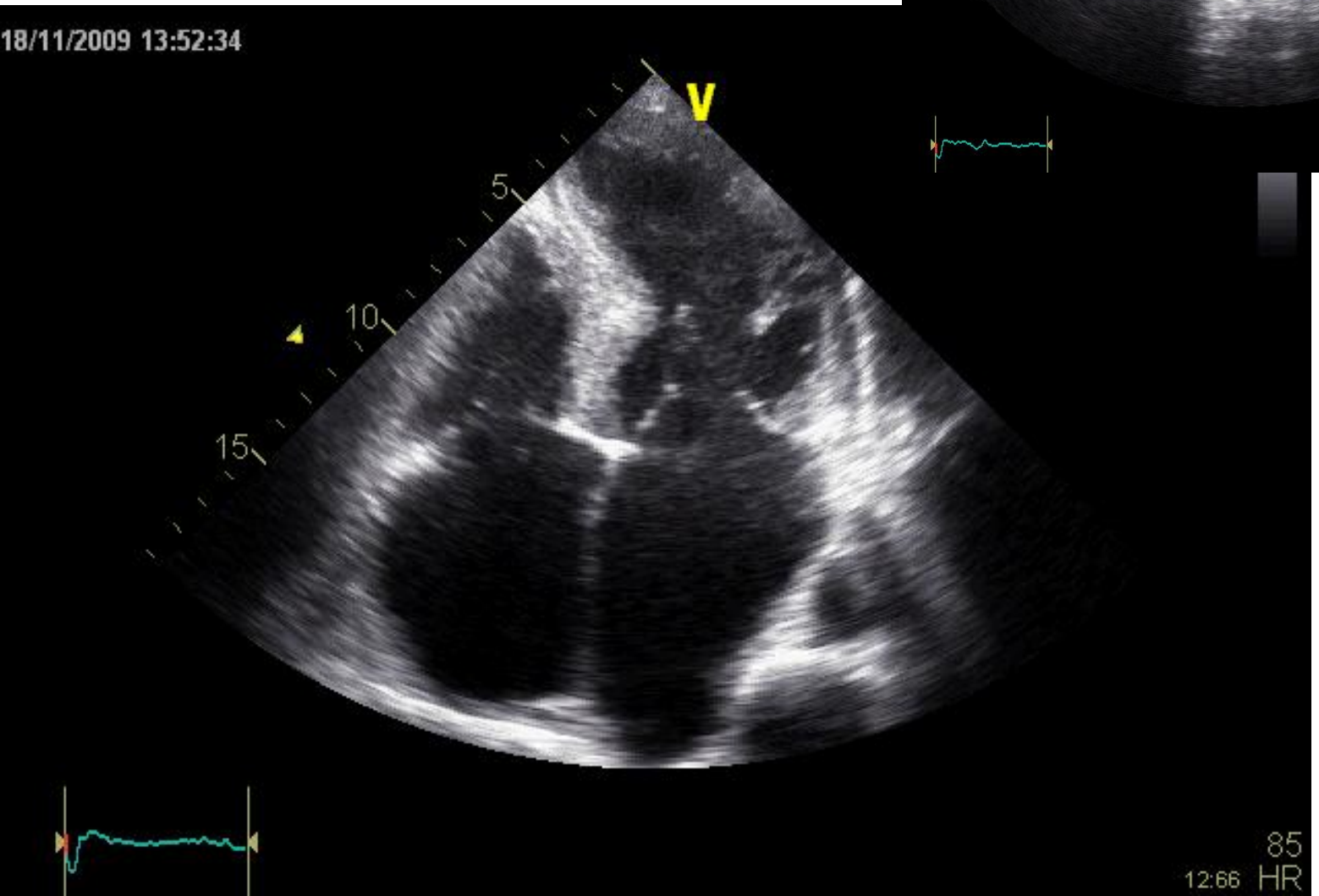
«La Forme Terminale» CMH avec FEVG <50%

- Fréquence: 3.5% des CMH
- Formes familiales
- Formes sans dilatation ou amincis
57%



Harris et al. Circulation 200

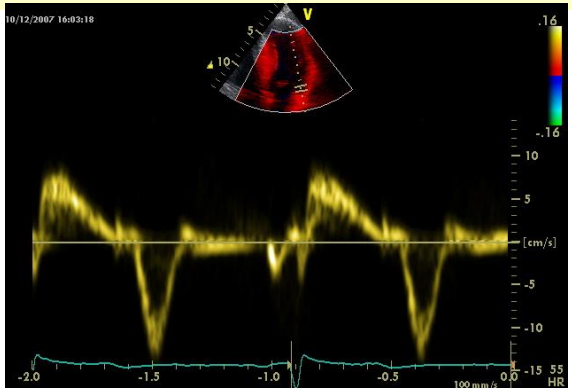

18/11/2009 13:51:09
V
5
10
15
85
13:81 HR


18/11/2009 13:52:34
V
5
10
15
85
12:66 HR

Dysfonction Diastolique

- Anomalies de Relaxation et Compliance
- Rigidité VG
- Diminution de «l'Effet Suction» du VG

Valeur Pronostique du DTI à l'Anneau Mitral (e')



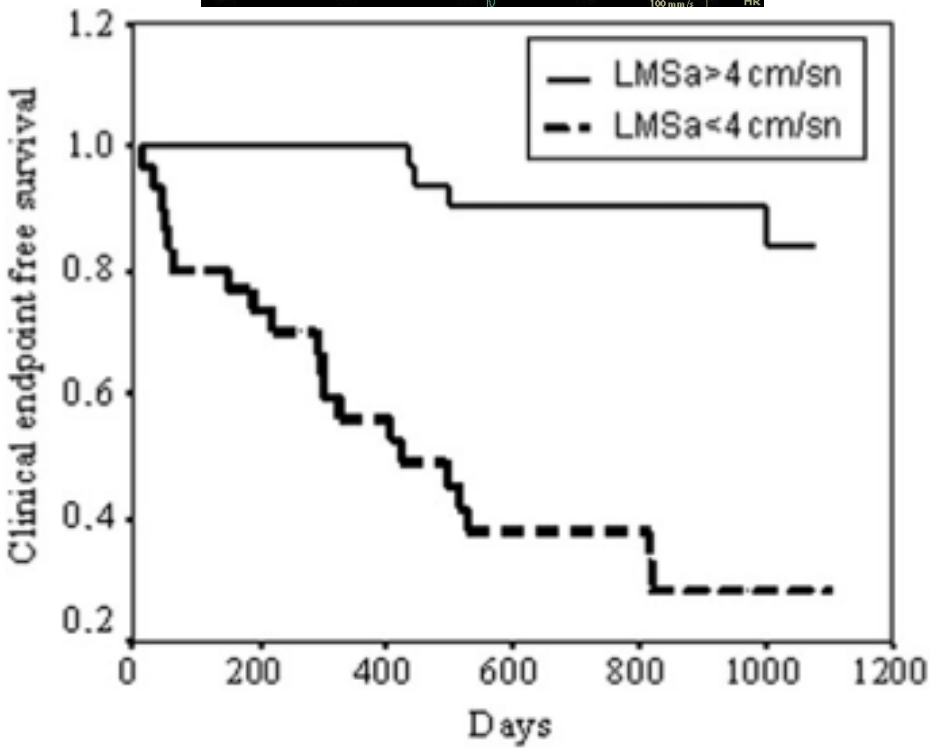


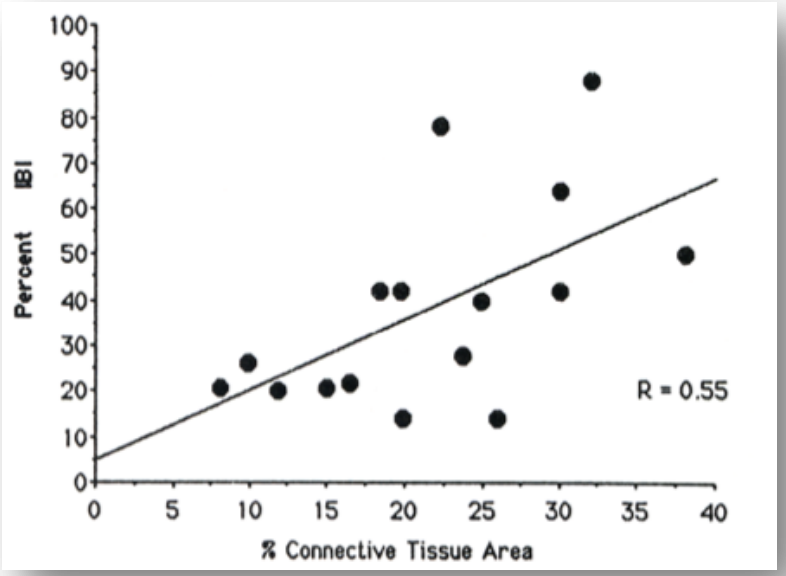
Table 3 Univariate and multivariable relations for prediction of clinical endpoints

	Univariate		Multivariable		
	F-value	P-value	R^2	Wald	P-value
LV outflow obstruction	5.0	0.02	0.29	4.6	0.03
Left atrium diameter	6.2	0.04	–	–	NS
Maximum wall thickness	5.0	0.02	–	–	NS
E/Ea (septal)	8.5	0.004	–	–	NS
E/Ea (lateral mitral)	9.3	0.003	–	–	NS
Lateral mitral Ea (cm/s)	16	0.0001	–	–	NS
Lateral mitral Aa (cm/s)	19	0.0001	–	–	NS
Lateral mitral Sa (cm/s)	27	0.0001	0.25	15.8	0.0001
Septal mitral Ea (cm/s)	10	0.002	–	–	NS
Septal mitral Aa (cm/s)	7.9	0.006	–	–	NS
Septal mitral Sa (cm/s)	5.5	0.02	–	–	NS

Bayrak F et al. Eur J Echocardiogr 2008.

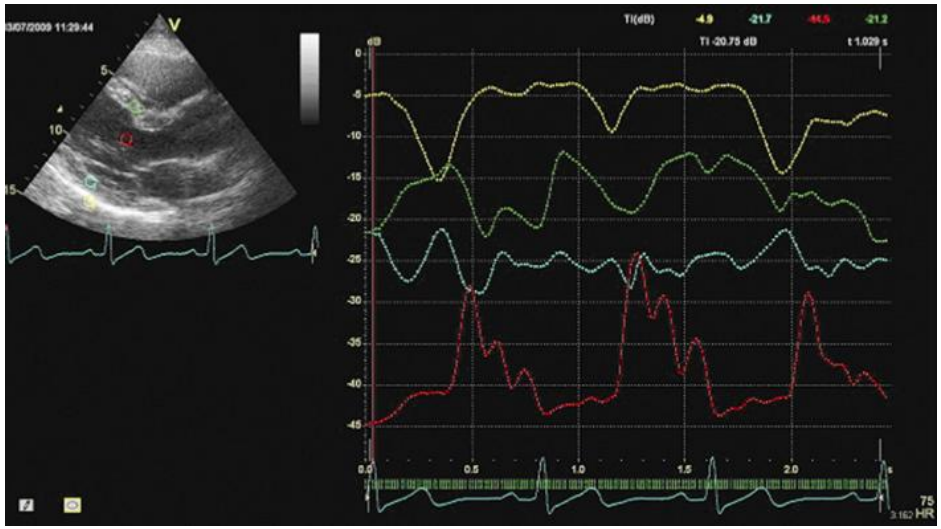
Fibrose Myocardique

Echocardiographie «Réflectivité des Tissus»



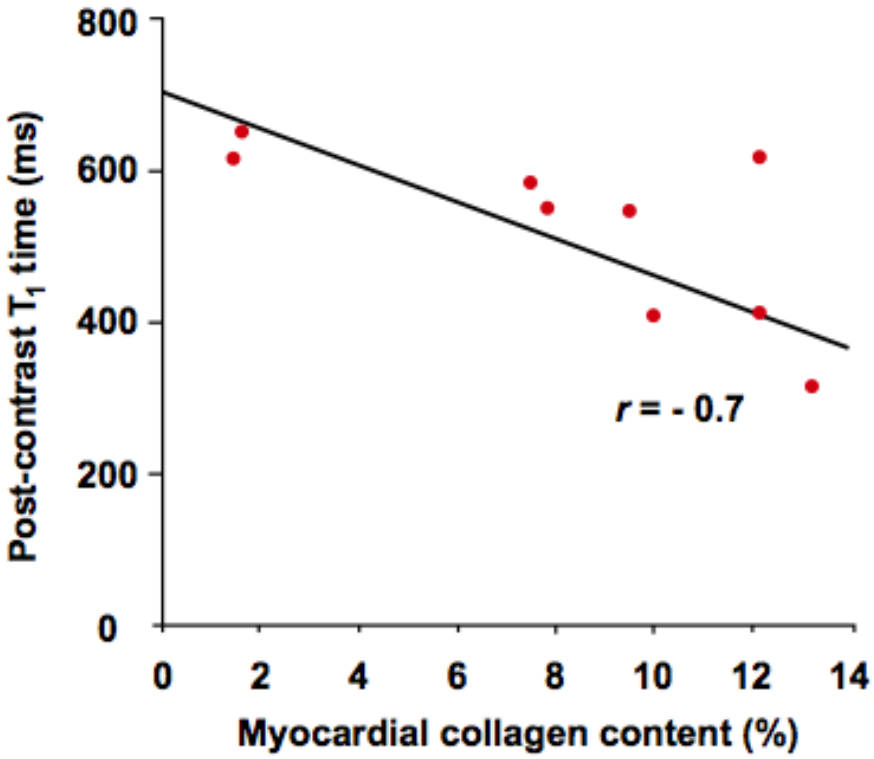
Péricarde = référence

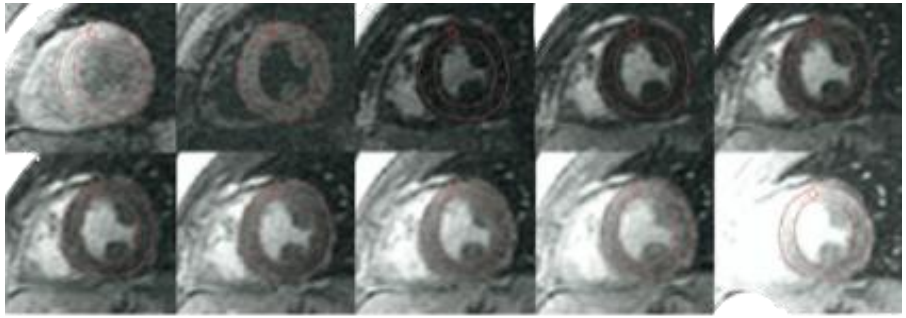
Mesure de la rétrodiffusion pour chaque zone d'intérêt en fonction de celle du péricarde (en % ou dB)

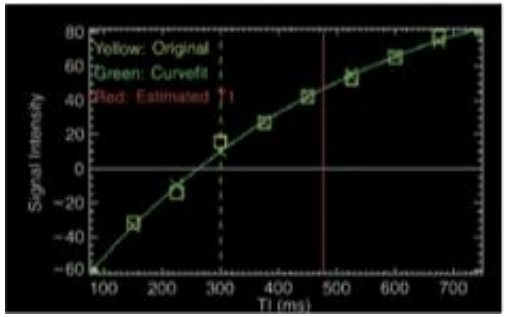


Picano E. *Circulation.* 1990;81:58-64

Quantification IRM de la Fibrose Interstitielle «T1 Mapping»







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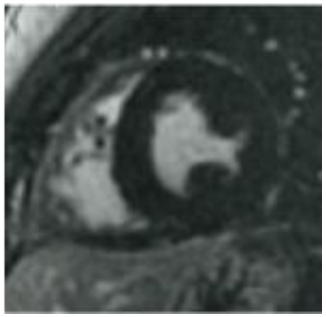
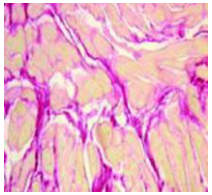
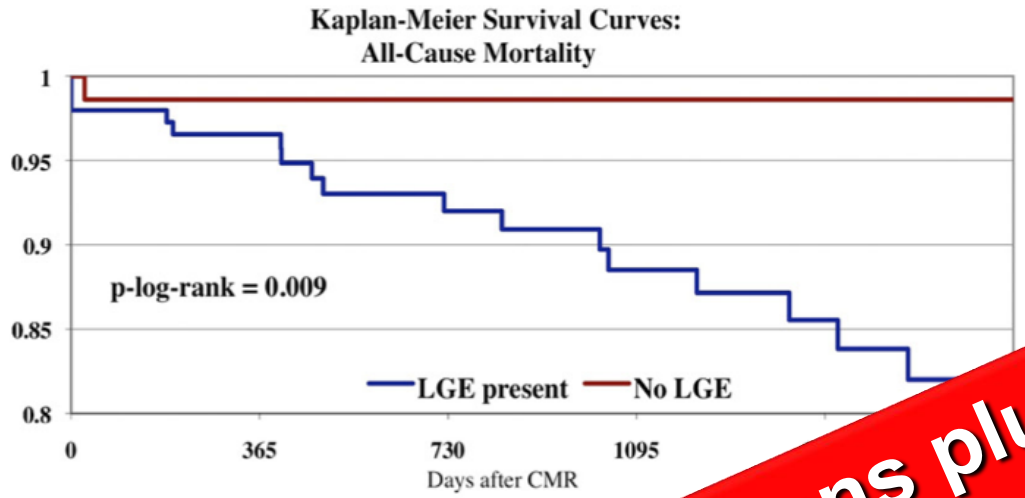
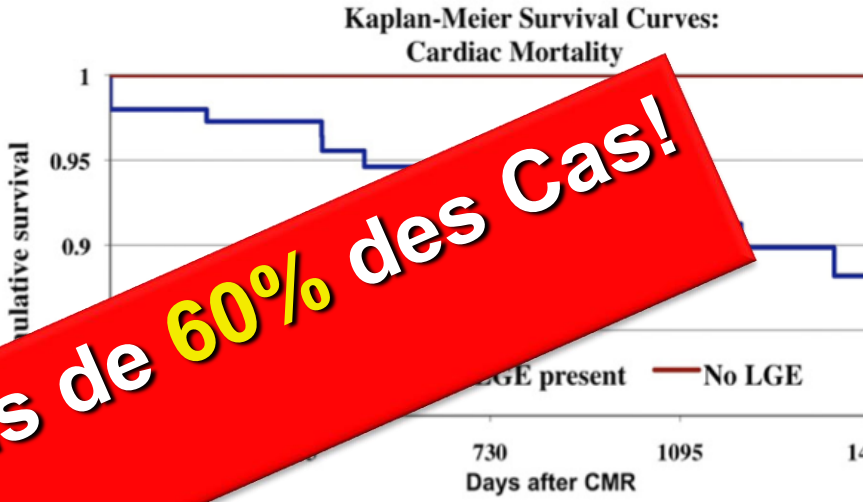


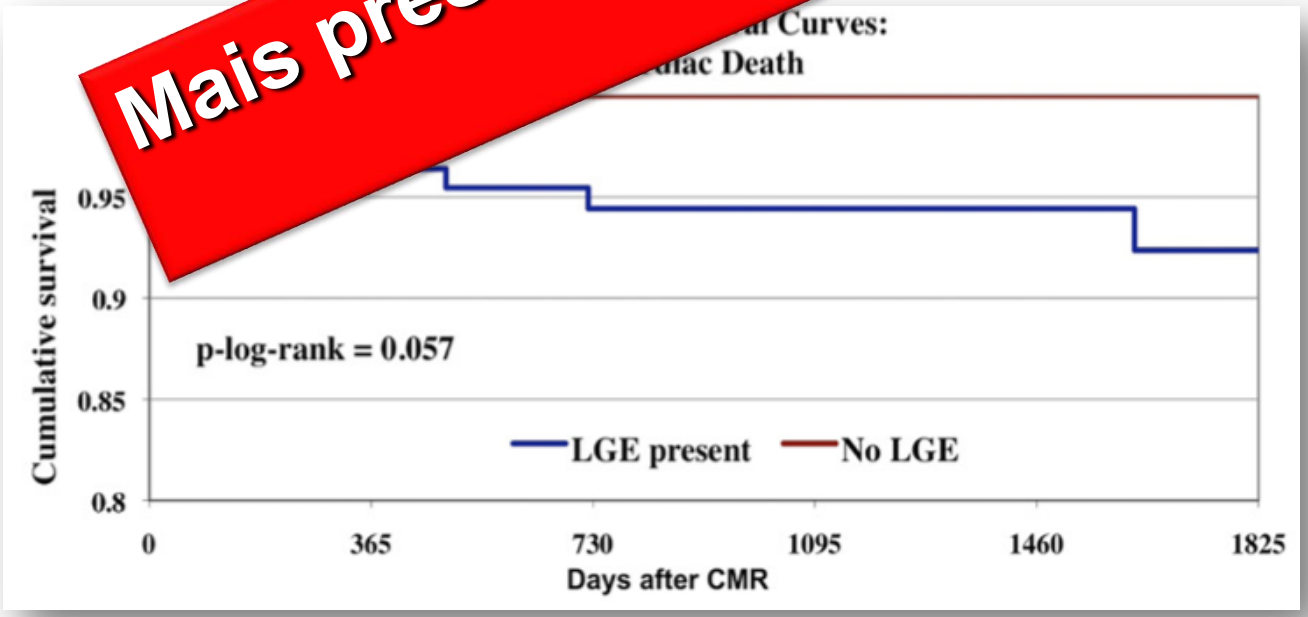


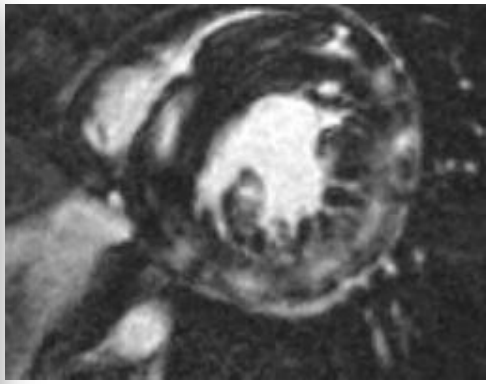
Iles L. *J Am Coll Cardiol.* 2008;52:1574-80





Mais présente dans plus de 60% des Cas!





Bruder O et al. J Am Coll Cardiol 2010

Journal of the American College of Cardiology
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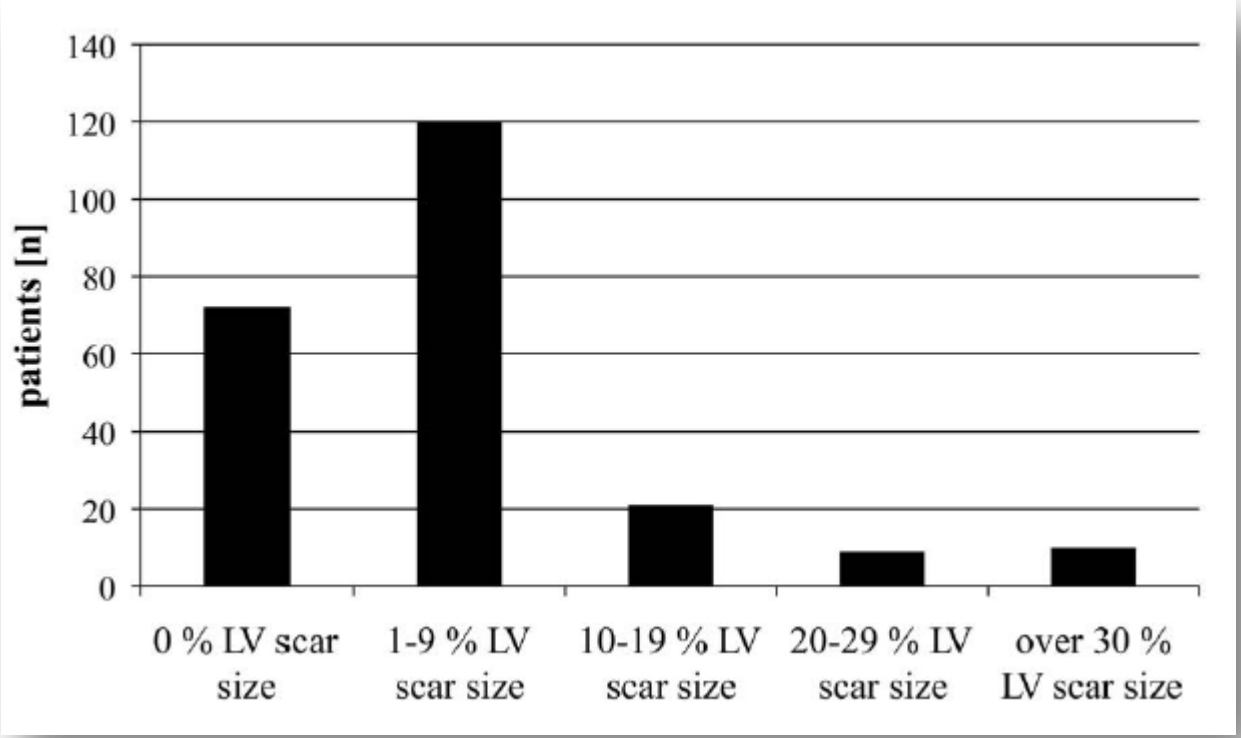
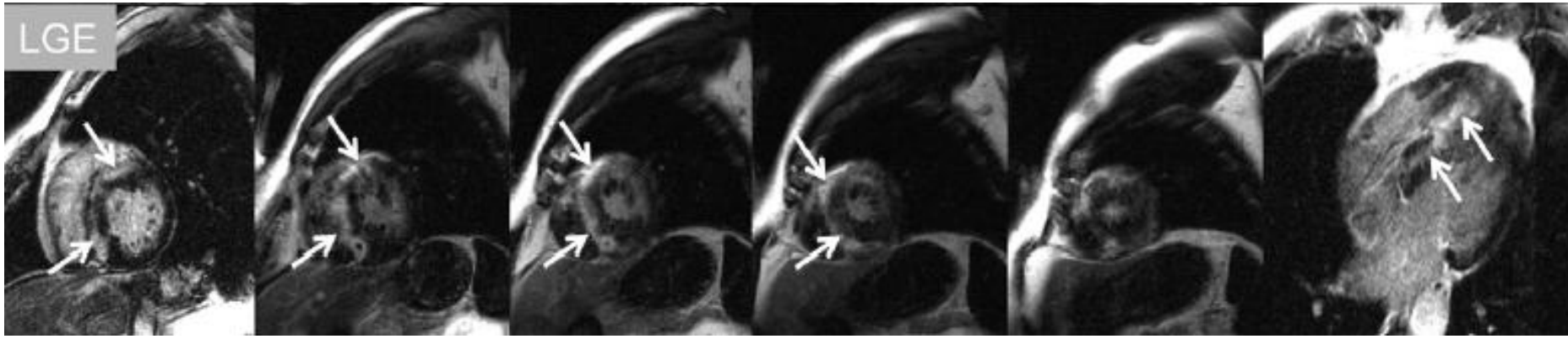
Vol. 56, No. 11, 2010
ISSN 0735-1097/$36.00
doi:10.1016/j.jacc.2010.05.007

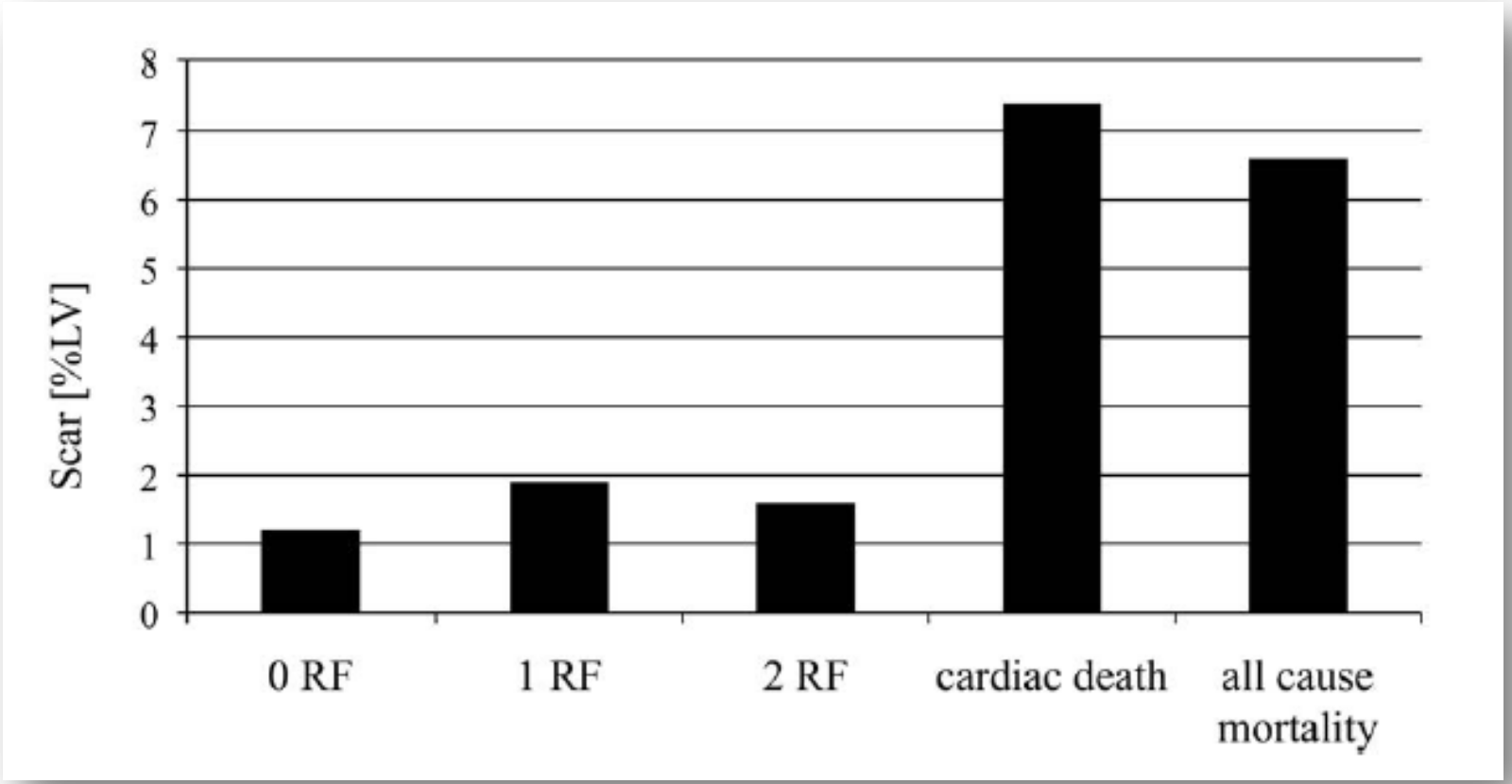
Myocardial Scar Visualized by Cardiovascular Magnetic Resonance Imaging Predicts Major Adverse Events in Patients With Hypertrophic Cardiomyopathy

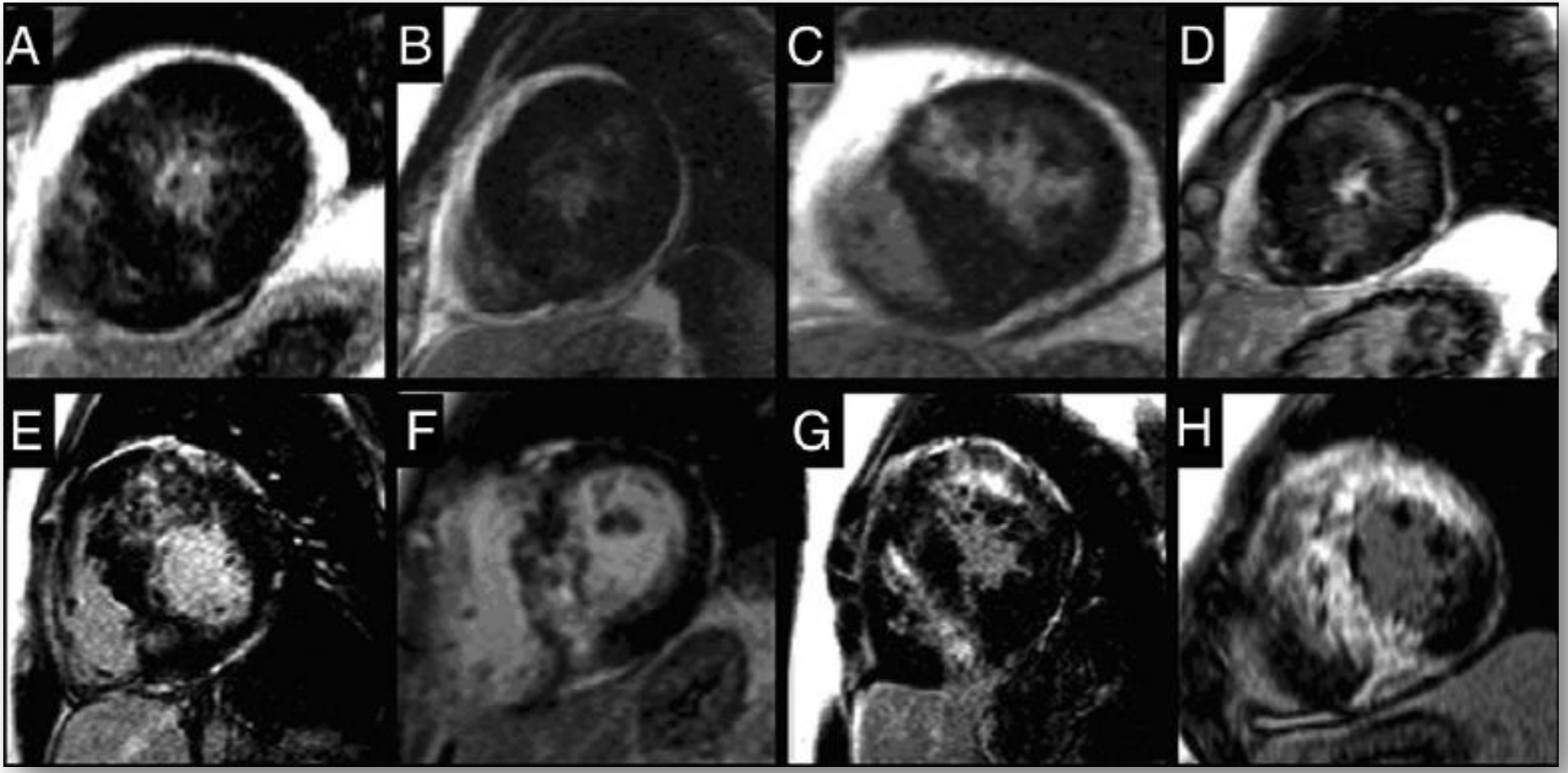
Oliver Bruder, MD,* Anja Wagner, MD,† Christoph J. Jensen, MD,* Steffen Schneider, PHD,‡ Peter Ong, MD,§ Eva-Maria Kispert, RN,§ Kai Nassenstein, MD,‖ Thomas Schlosser, MD,‖ Georg V. Sabin, MD,* Udo Sechtem, MD,§ Heiko Mahrholdt, MD§

Essen, Ludwigshafen, and Stuttgart, Germany; and Philadelphia, Pennsylvania

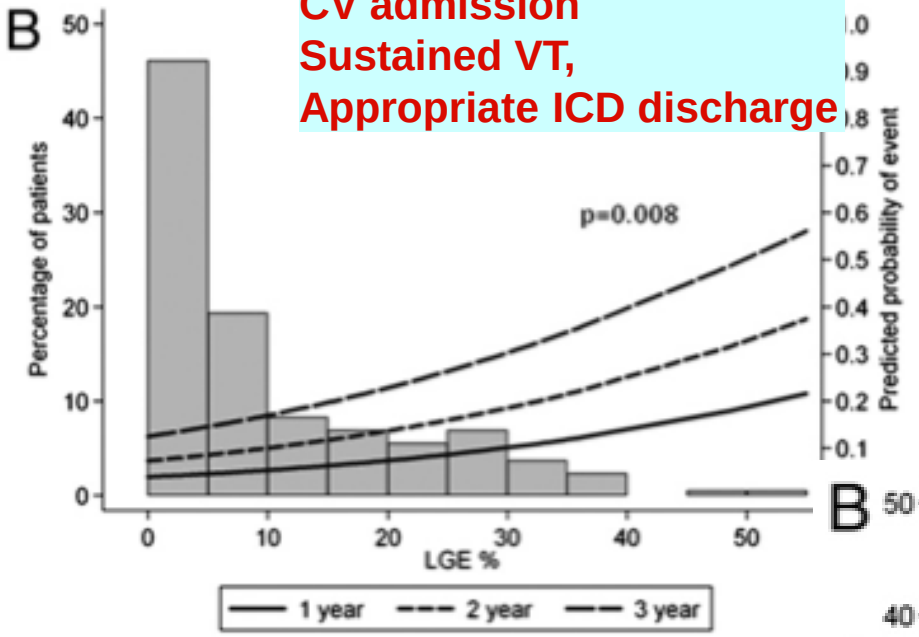



LGE

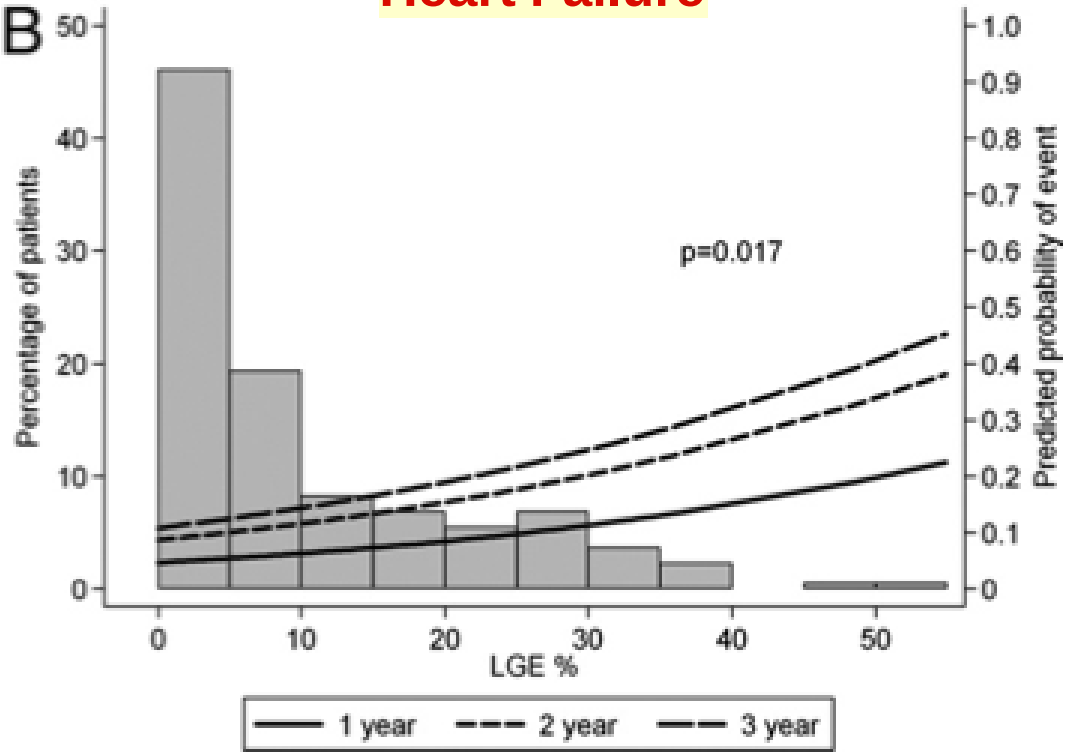

Scar [%LV]
8
7
6
5
4
3
2
1
0
0 RF
1 RF
2 RF
cardiac death
all cause mortality


A
B
C
D
E
F
G
H

CV death,
CV admission
Sustained VT,
Appropriate ICD discharge

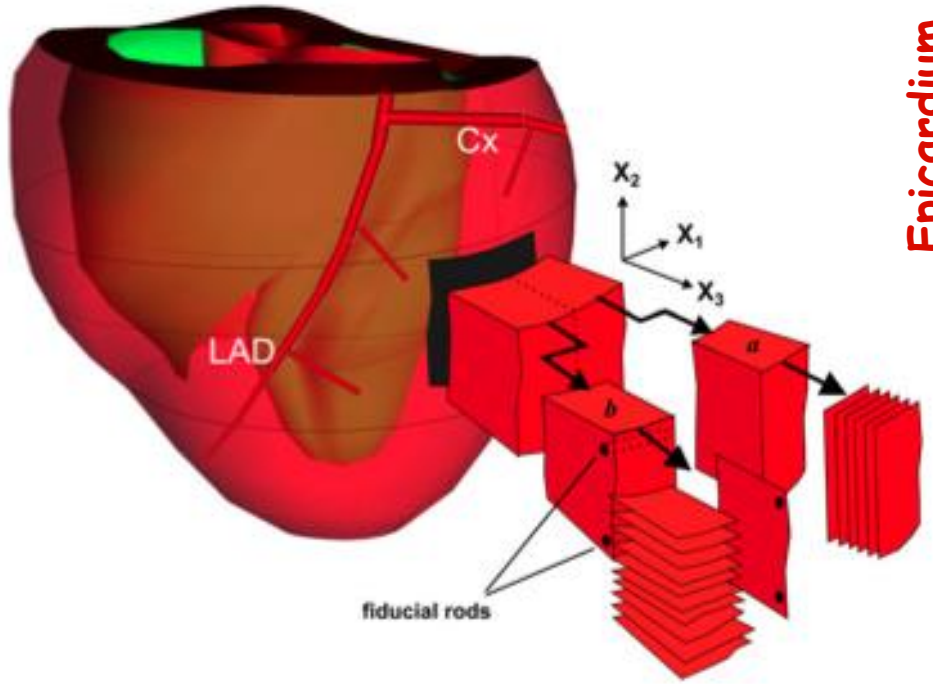


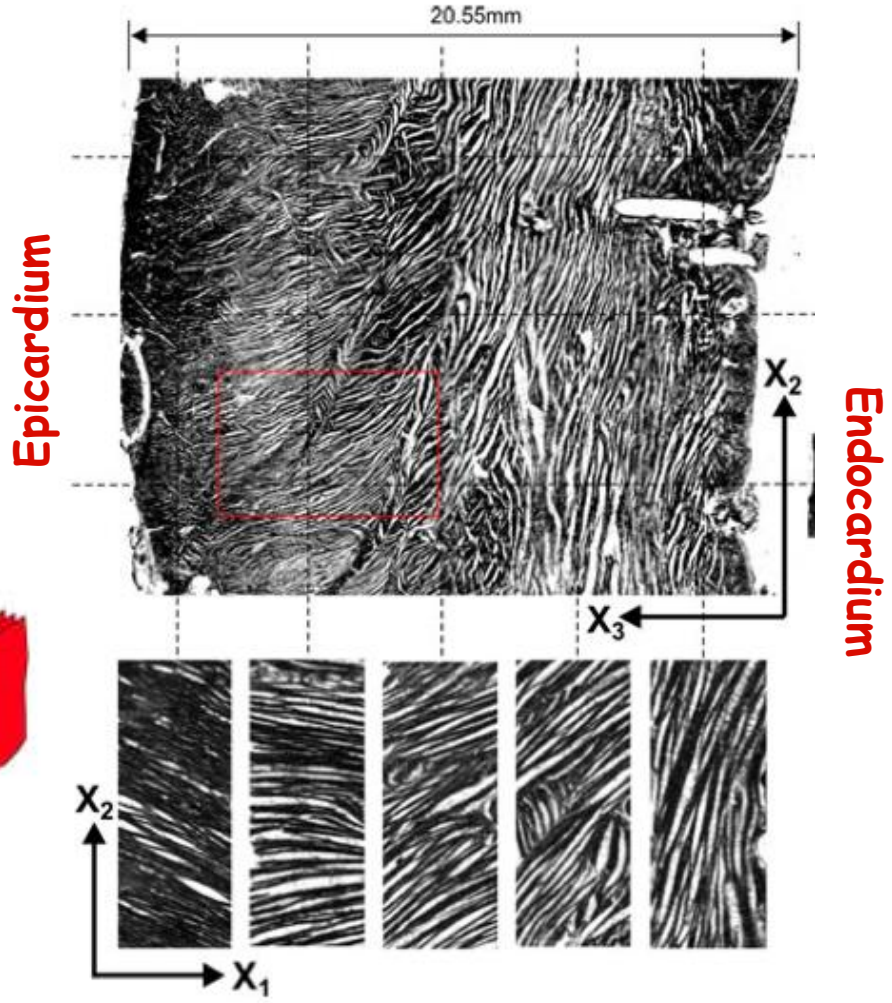
Heart Failure



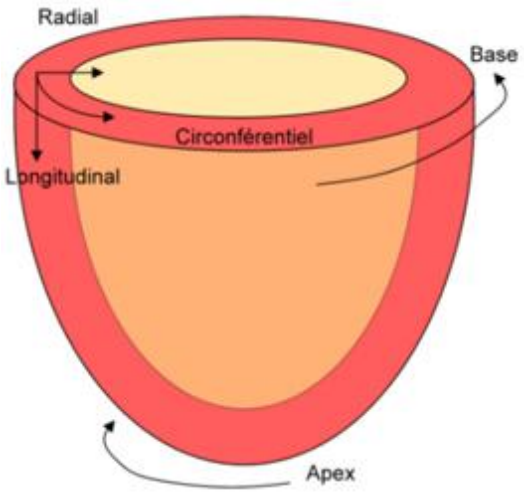
O'Hanlon R et al. J Am Coll Cardiol 2010

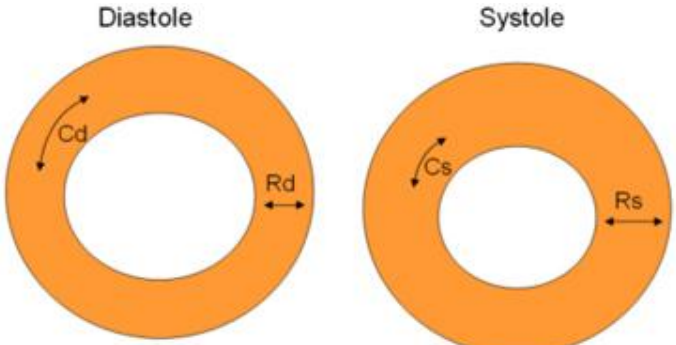
Imagerie de Déformation Myocardique



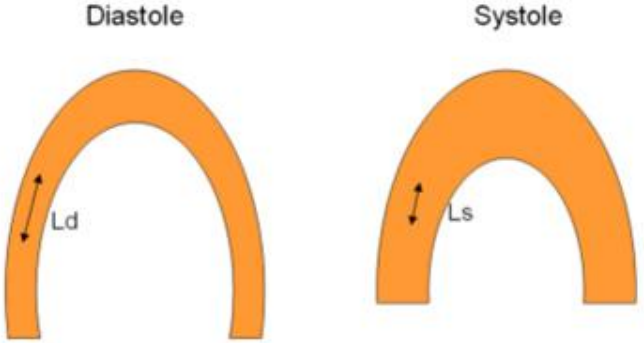


Imagerie de Déformation Myocardique

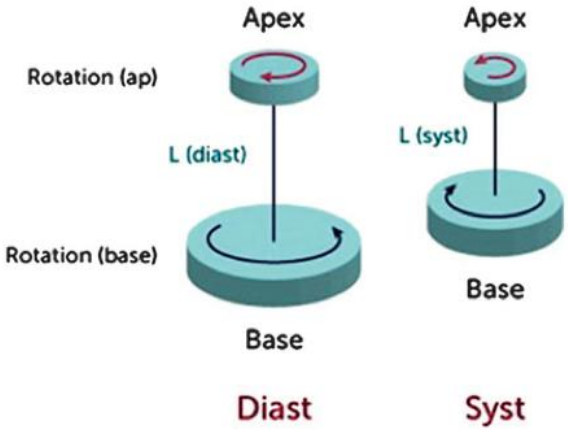




Strain circonférentiel = (Cs-Cd)/Cd, de l'ordre de - 20%
Strain radial = (Rs-Rd)/Rd, de l'ordre de +40%

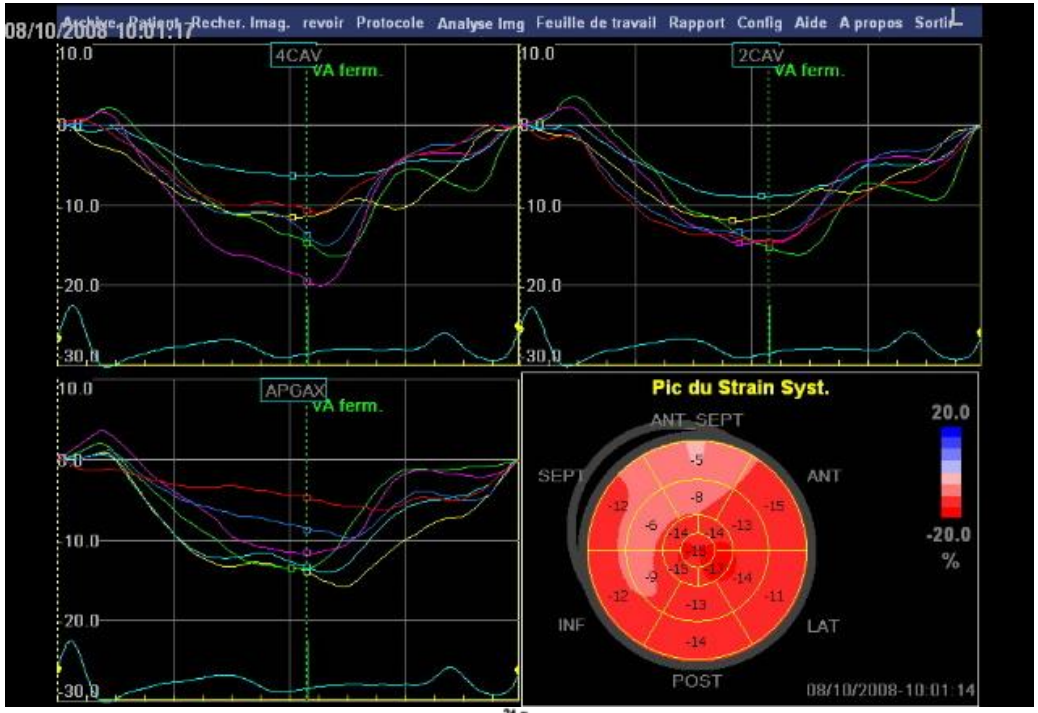


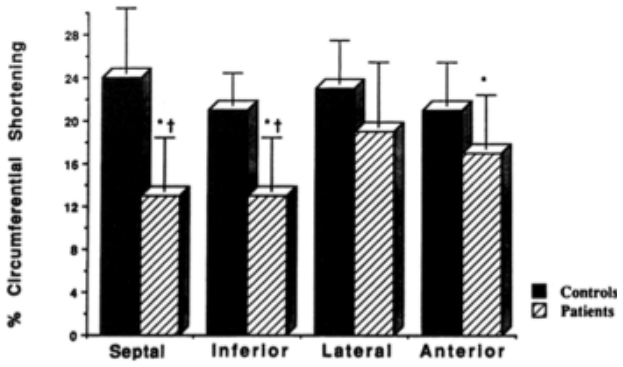
Strain longitudinal = (Ls-Ld)/Ld, de l'ordre de - 20%

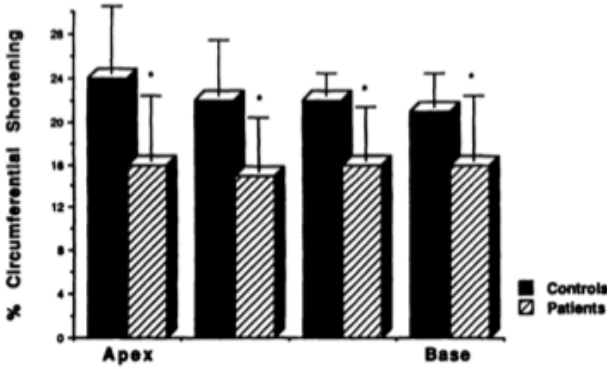


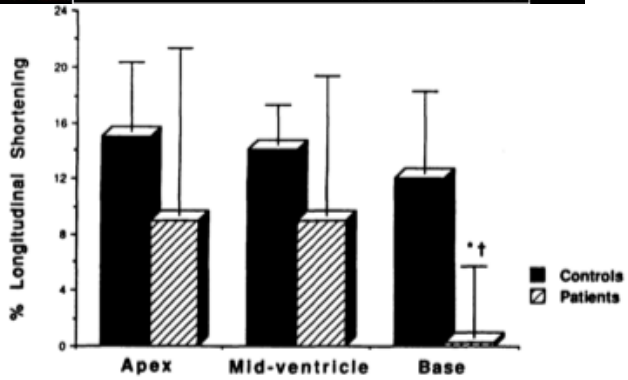
Torsion (Twist)=Rot ap-Rot bas

Hétérogénéité de Contraction Spatiale et Temporelle





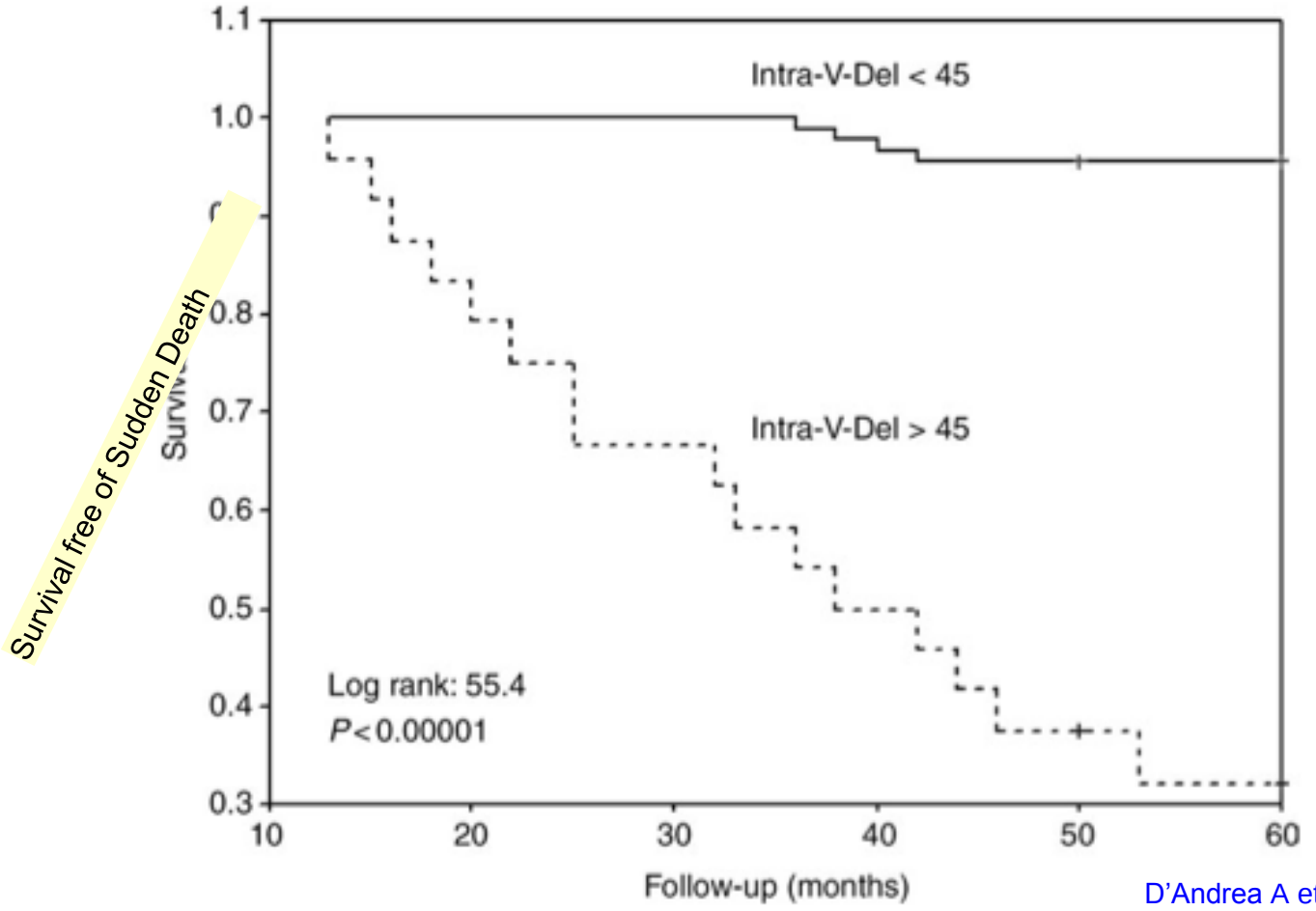




Kramer et al. Circulation 1994

Hétérogénéité de Contraction Spatiale et Temporelle

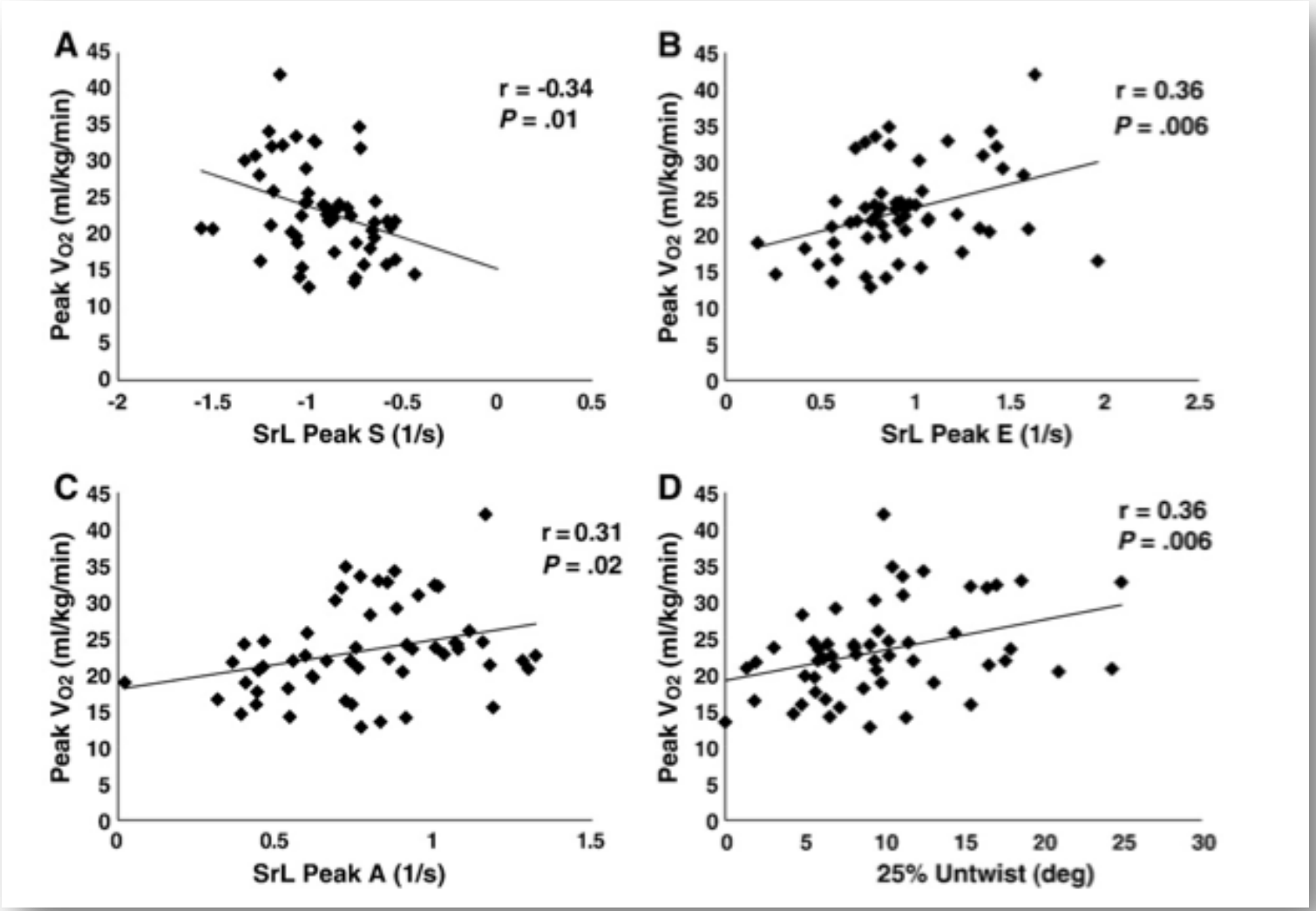
Asynchronisme Intra-VG Prédit la Mort Subite?



D'Andrea A et al. Eur Heart J 2006

Imagerie de Déformation Myocardique

Relation avec la Capacité d'Effort



Abozguia K. Am Heart J 2010

Reconnaître une CMH Sévère
Apport de L'Echo et de l'IRM

L' Imagerie Cardiaque a une place fondamentale dans la Stratification du Risque

Critères "Reconnus"

- HVG >30mm (majeur)
- Forme «terminale» (FEVG <50%)
- Anévrysme Apical
- Obstruction (>30mmHg)
- Obstruction effort
- OG >34mL/m^2 (>47mm)
- E' <4cm/s
- Forme «restrictive»
- Fibrose de remplacement à l'IRM (Rétention tardive de Gado)

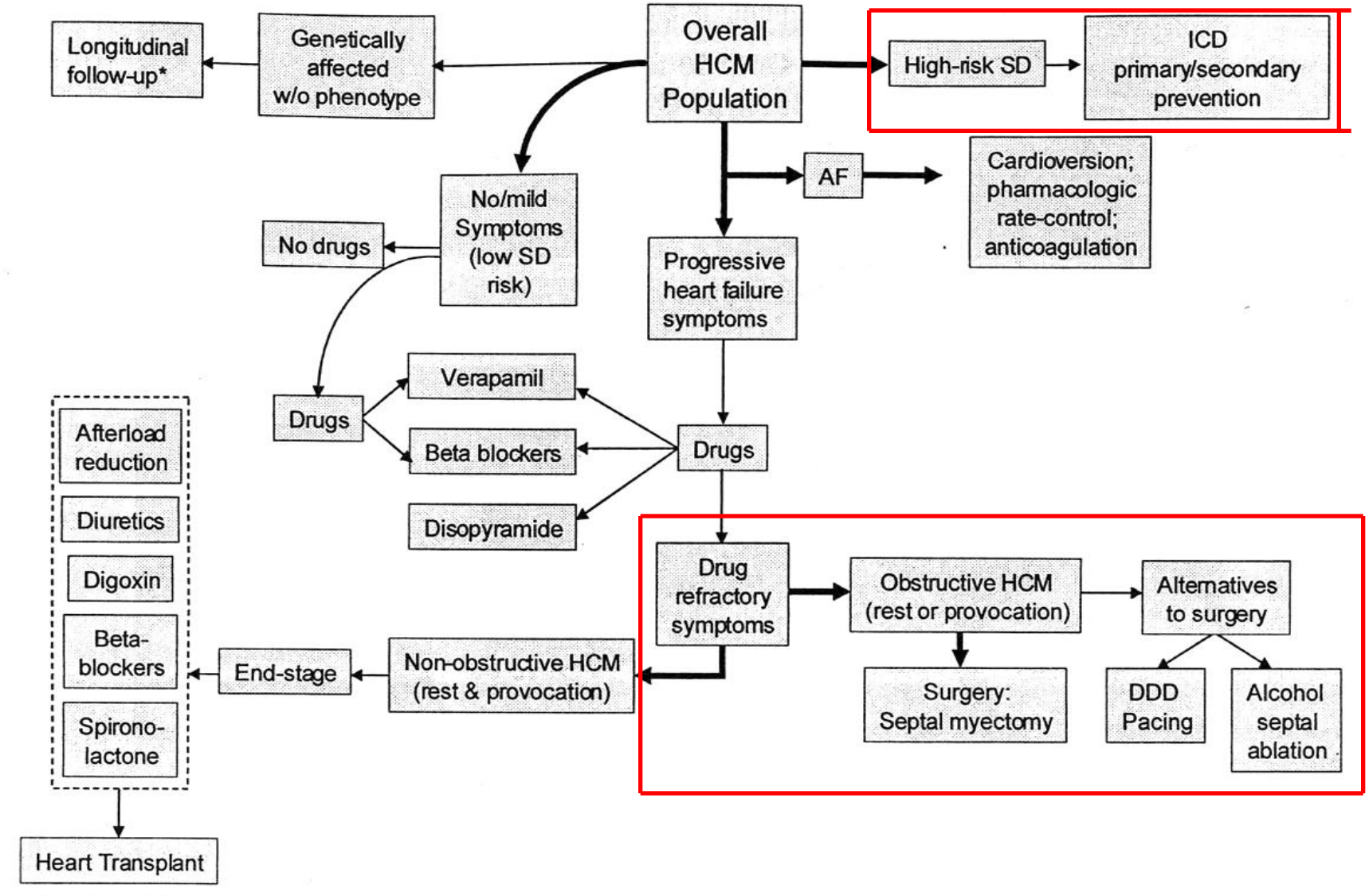
Critères Potentiels?

- Anomalies de déformation VG
- Anomalies de déformation OG
- Asynchronisme VG
- Anomalies de réserve coronaire
- Quantification de la Fibrose interstitielle (T1 mapping)

Objectives of treatment in Hypertrophic Cardiomyopathy

- ***Improve symptoms, when invalidating: NYHA Class > 2***
- ***Prevent sudden cardiac death***
- ***Prevent disease progression and progression to heart failure***

ACC-ESC Guidelines on Management of HCM



Maron BJ, Mc Kenna WJ et al JACC, 2003

Non-Pharmacological Therapies for Hypertrophic Cardiomyopathy

- ✓ **To prevent the risk of sudden death:**

  ICD in high risk patients

- ✓ *For hemodynamic support:*
  - Surgery: myectomy±mitral valvuloplasty
  - Transcatheter Alcohol Septal Ablation (TASH)
  - DDD pacing

Who is At Risk ?

Major Risk Factors to Sudden Cardiac Death in HCM

- **History of cardiac arrest (VF)**
- **Spontaneous sustained VT**

- **Family history of premature HCM-related SCD**
- **High-risk mutation (closely linked to family history)**
- **Unexplained syncope**
- **LV thickness ≥ 30mm**
- *Abnormal exercise response*
- *Non-sustained VT (Holter): 20-30% of HCM patients*

Maron BJ, McKenna WJ et al JACC 2003

Dépistage génétique

- 3 gênes = 85% des cas en Europe
  -Chaîne lourde de la béta-myosine (MYH7)
  -Protéine C (MYBPC3)
  -Troponine T (TNNT2)

- Grande hétérogénéité intragénique avec plus de 200 mutations identifiées dont certaines sont considérées comme à haut risque

Dépistage génétique

- **Mutations à haut risque:**

  -Chaîne lourde de la béta-myosine:

- **R403Q**
- **R453C**
- **G716R**
- **R719W**

  -Troponine T cardiaque:

- **R92W**

ICD for secondary or primary prevention in HCM

Multicenter retospective study *(Maron BJ et al N Engl J Med, 2000)*

- **3-year f/u**
- **Appropriate therapies (AT): 25% patients**
- **4:1 excess of ICD 's implanted to lives saved**
- **Secondary prevention: 11% AT per year**
- **Primary prevention: 5% per year**

« ICD is the most effective and reliable treatment when risk is judged unacceptably high and deserving of intervention »

ACC-AHA-NASPE Guidelines 2002:

- Secondary prevention: class I indication

- Primary prevention: class II b indication

Non-Pharmacological Therapies for Obstructive Hypertrophic Cardiomyopathy

✓ ***As hemodynamic support* in HCM patients with:**

. Marked LVOT obstruction: resting gradient >50 mmHg

. Severe limiting symptoms: NYHA class III-IV

. Failure to «optimal» drug treatment

✓ ***Technical solutions:***

- Myectomy ± mitral valvuloplasty: reference
- Transcatheter Alcohol Septal Ablation (TASH):

  Alternative to surgery ?
- **Pacing therapy: which place ?**

No proven efficacy to reduce the risk of sudden cardiac death

Merci beaucoup