

# **MIOCARDIOPATIA HIPERTROFICA**

**Dr. Luis Aguinaga**

**Corrientes 2010**

# Miocardiopatías: Clasificación

**PRIMARIAS**

**GENETICA**

**M.HIPERTROFICA**

DISPLASIA ARR. DEL VD  
VI NO COMPACTADO  
ALTER.CONDUCCION  
CANALOPATIAS

**ADQUIRIDA**

**MIXTA**

**SECUNDARIAS**

# DEFINICION

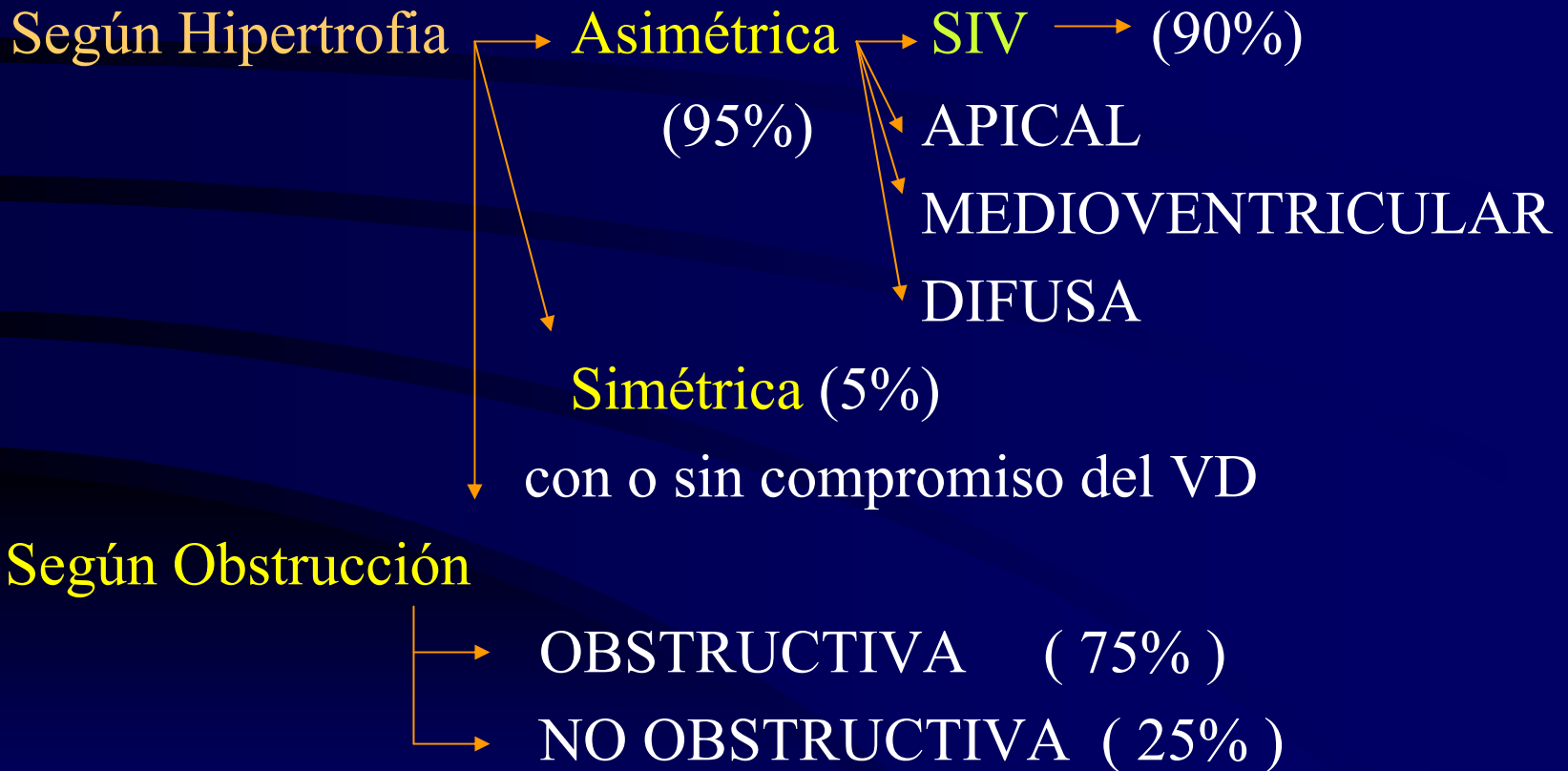
- Es una enfermedad con un **Factor de transmisión autosómica dominante**, de **expresión variable**, gran heterogenicidad morfológica, funcional y clínica, caracterizada por **Hipertrofia miocárdica del VI**, **desorden miofibrilar** y **riesgo de muerte súbita**.

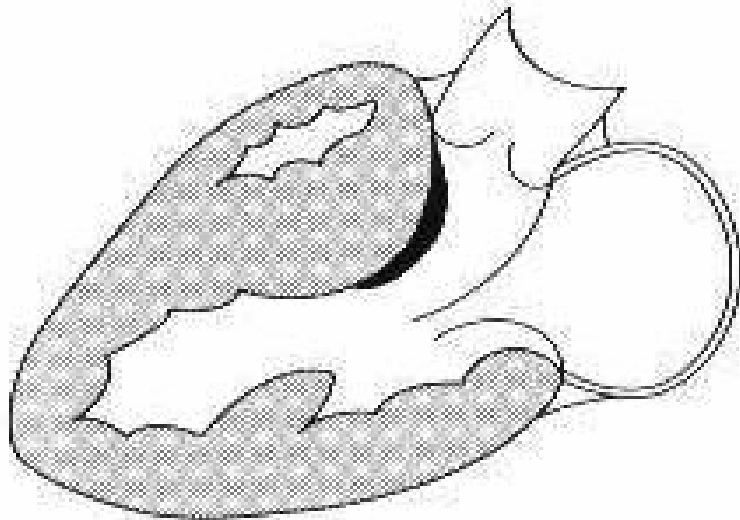
# ASPECTOS GENÉTICOS

- Mutaciones en 11 genes con 400 mutaciones identificadas
- B myosin cadena pesada, Myosin Binding protein C, Troponin T, representan mas del 50%
- Las expresiones fenotípicas depende del genotipo mas factores ambientales

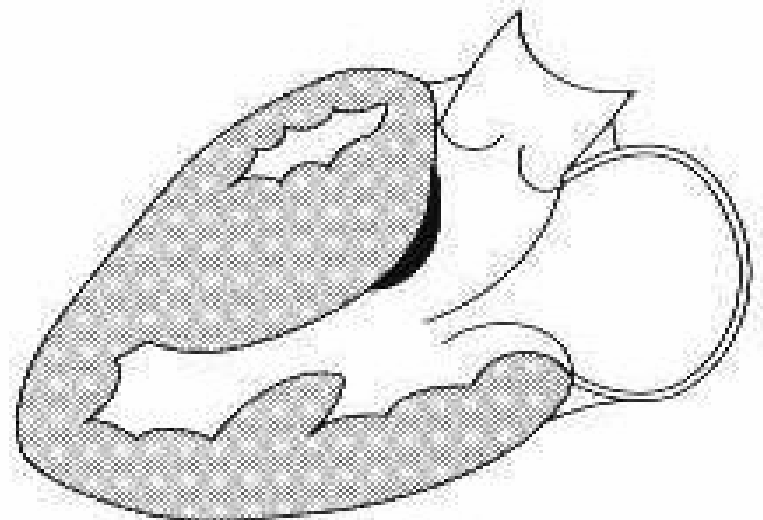


# M.H.: Formas de Presentación

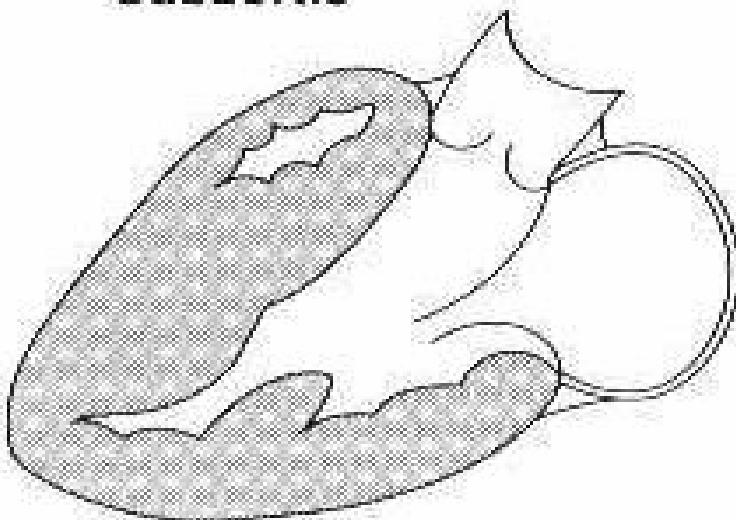




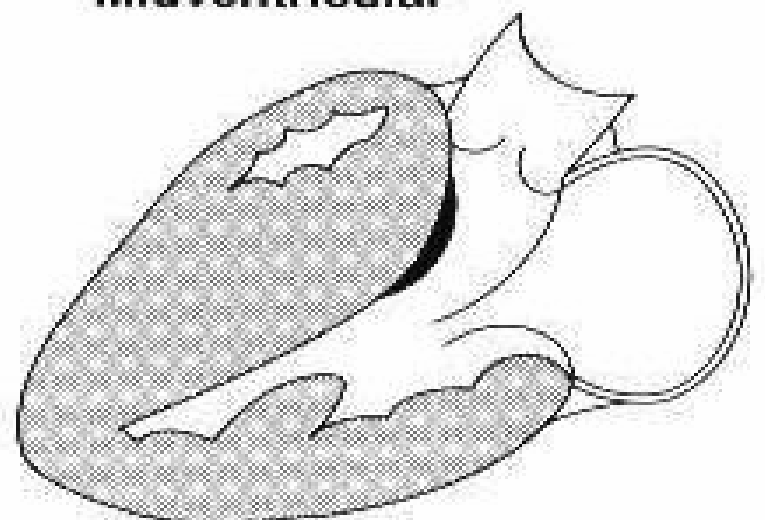
**Subaortic**



**Midventricular**



**Apical**



**Diffuse**

# MH: Fisiopatología Molecular

- Mutación genética



- Alteran integridad estructural y disposición estequiométrica del sarcómero

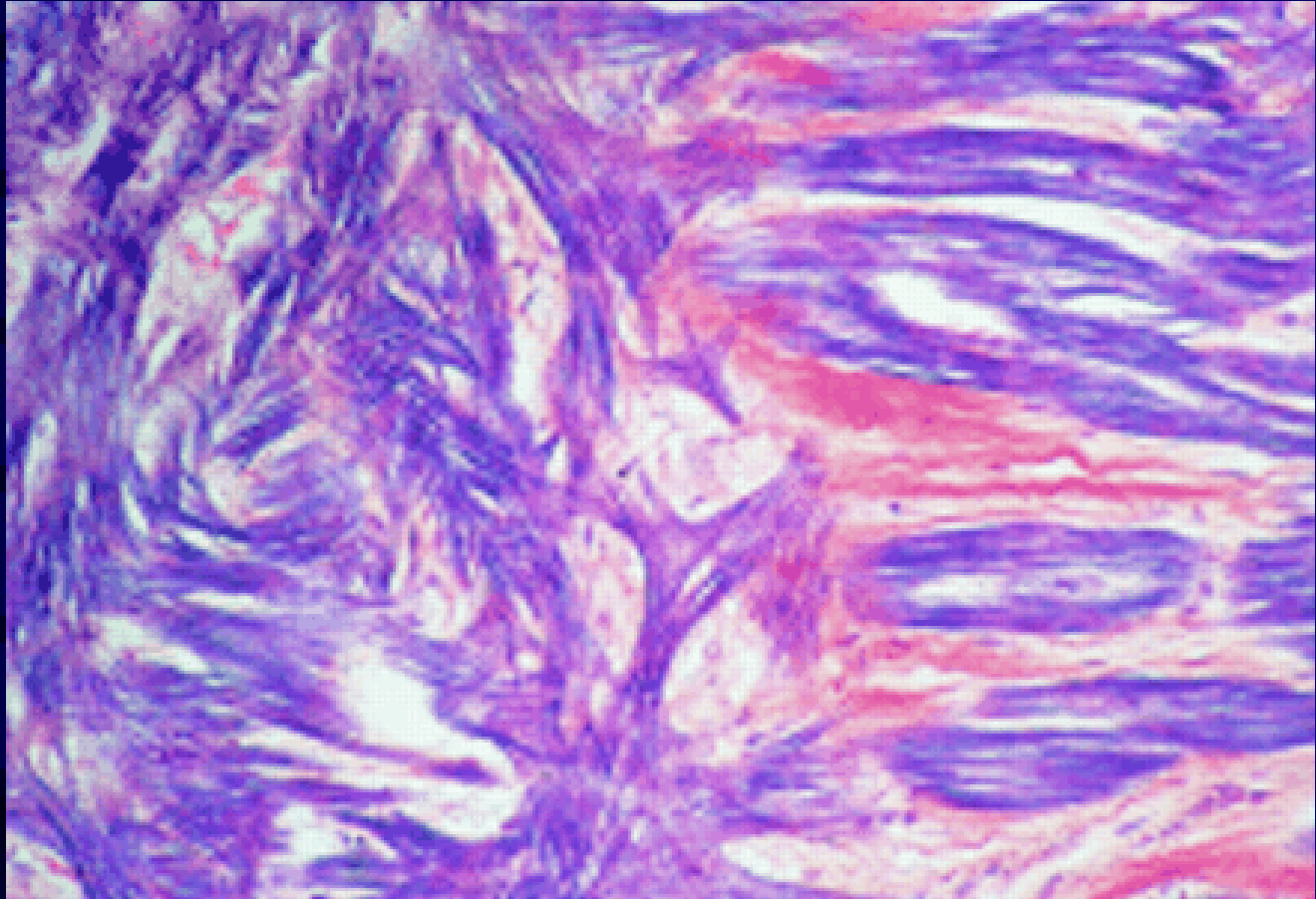


- < Contractilidad a nivel molecular



- Hipertrofia → Compensa para mantener función sistólica

# Pathogenesis



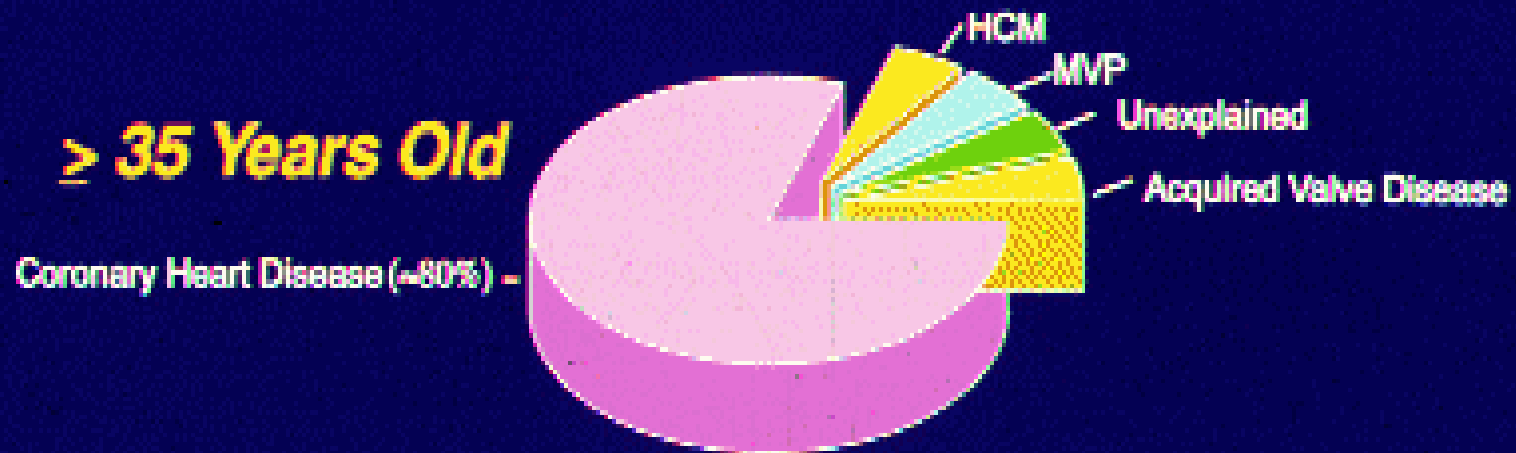
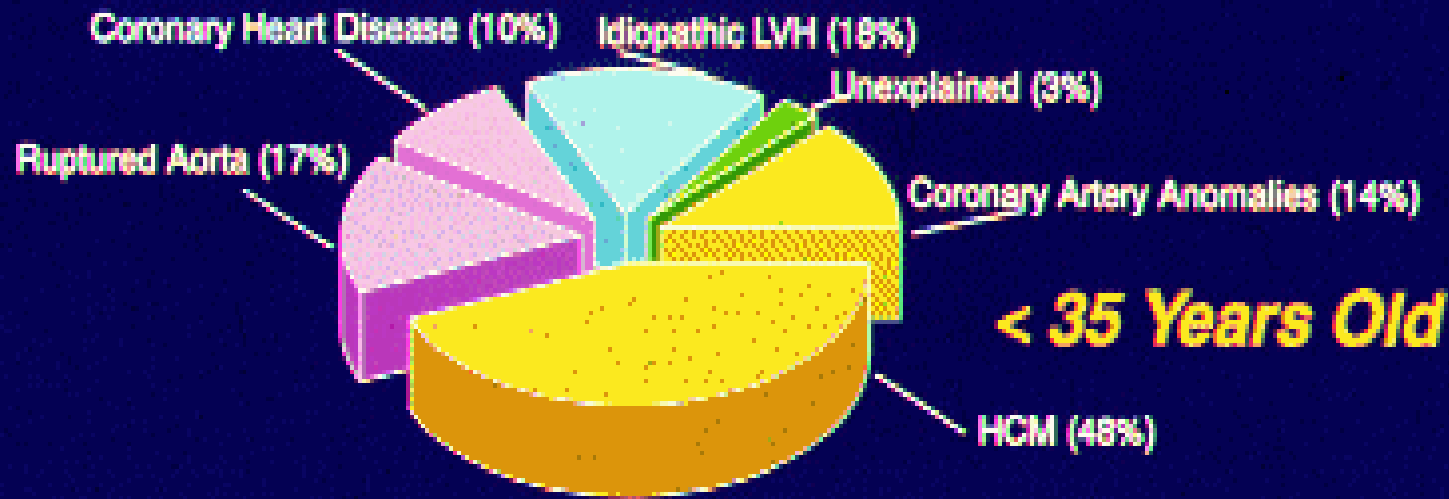
Focal distribution of myocyte disarray (to the left) adjacent to normal parallel alignment of myocytes;

Adapted from: Varnavaa AM et al., Heart 2000;84:476-482

# M.Hipertrófica:Prevalencia

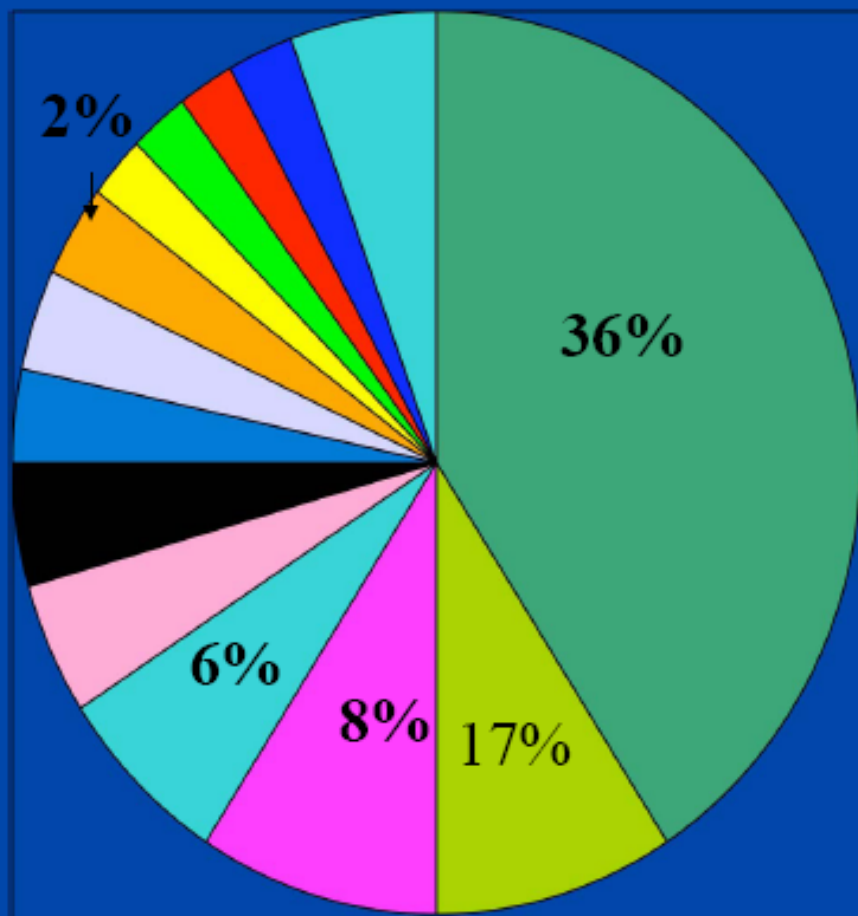
- \* Alrededor 0.17 % ( 1 c / 500 sujetos) en población de 25-35 a
- \* 3 / 1 a favor de **sexo masculino**
- \* Doble en **raza negra**
- \* Suele expresarse en 3° ó 4° decada aunque puede manifestarse en cualquier momento de la vida.

# SUDDEN DEATH IN ATHLETES

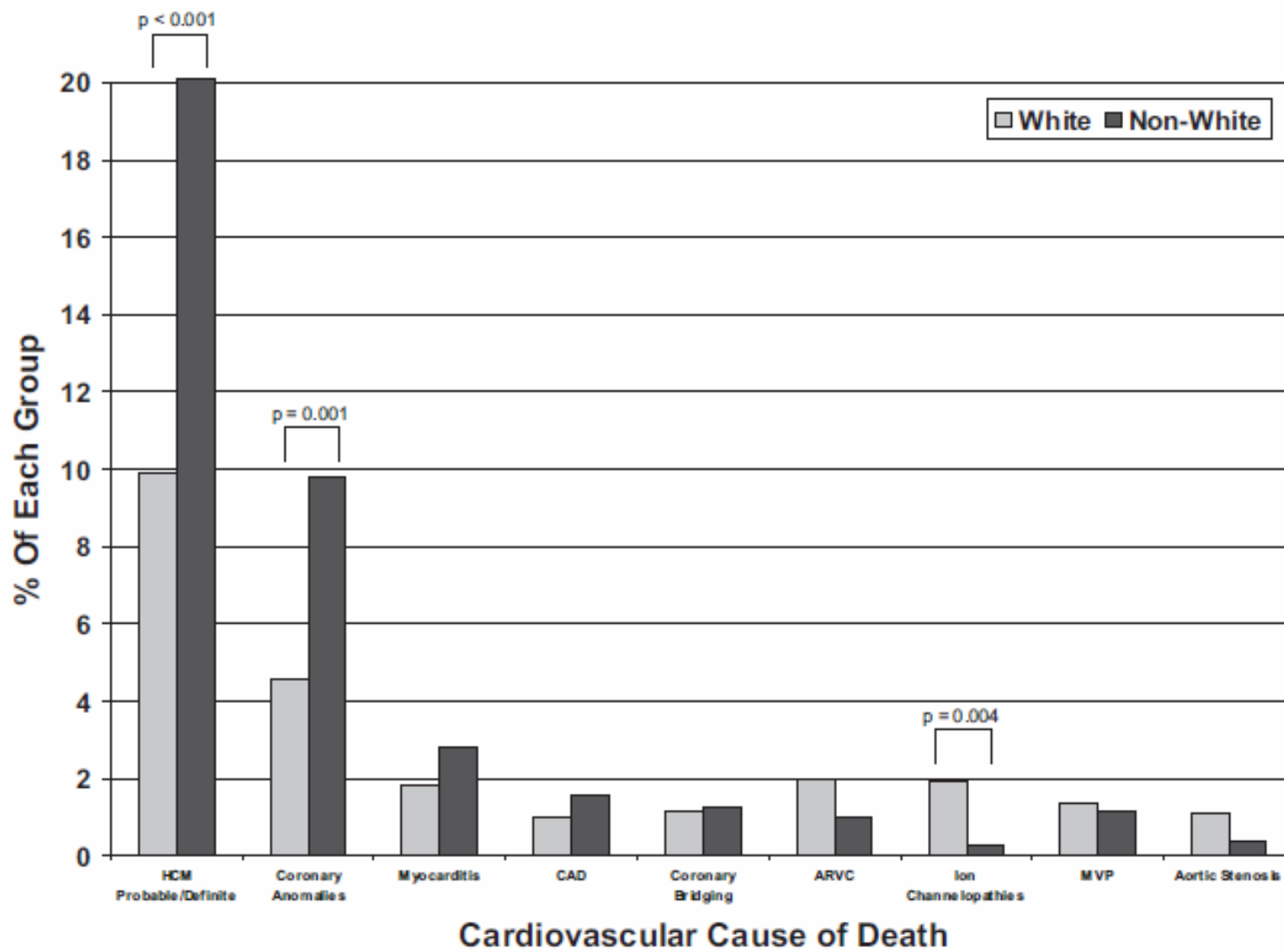


Maron, B.J., Epstein, S., Roberts, W.C. Causes of Sudden Death in Competitive Athletes. JACC:1986:204-14

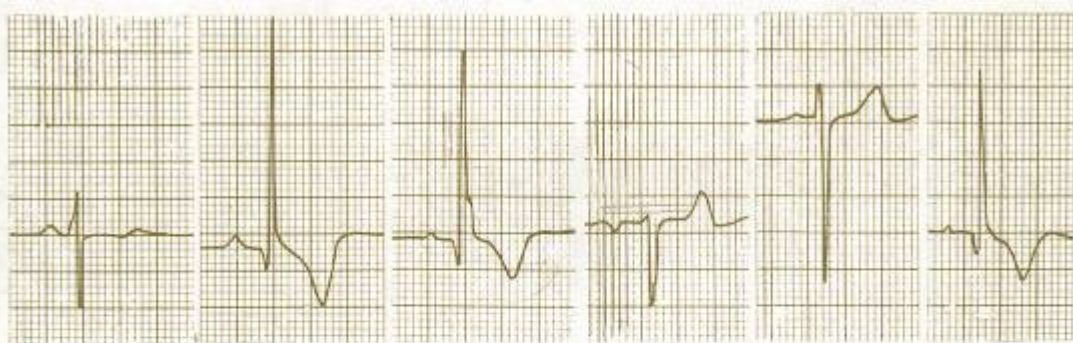
# Cardiovascular Causes of Sudden Cardiac Death in 690 Young Athletes



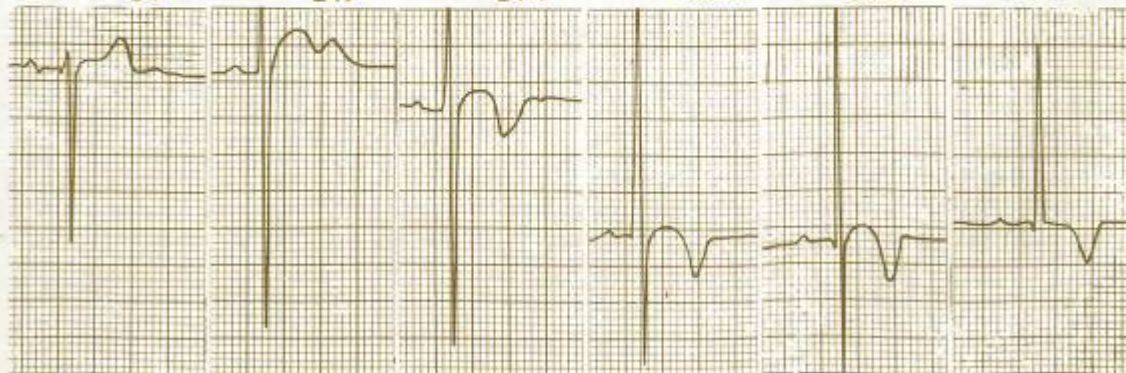
- HCM
- Coronary anomalies
- LVH/?HCM
- myocarditis
- ARVC
- Ion Channel Dx
- MVP
- LAD bridge
- CAD
- Aortic Rupture
- Aortic Stenosis
- DCM
- WPW
- other



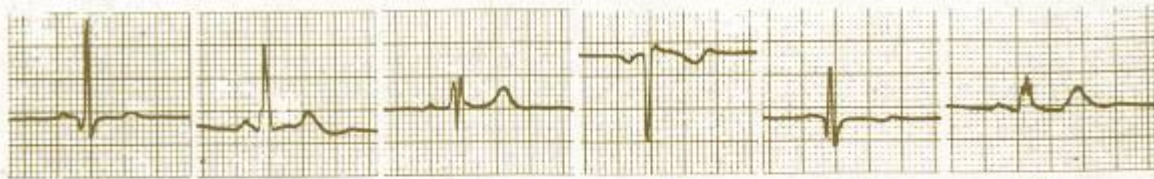
# METODOS COMPLEMENTARIOS DE DIAGNOSTICO



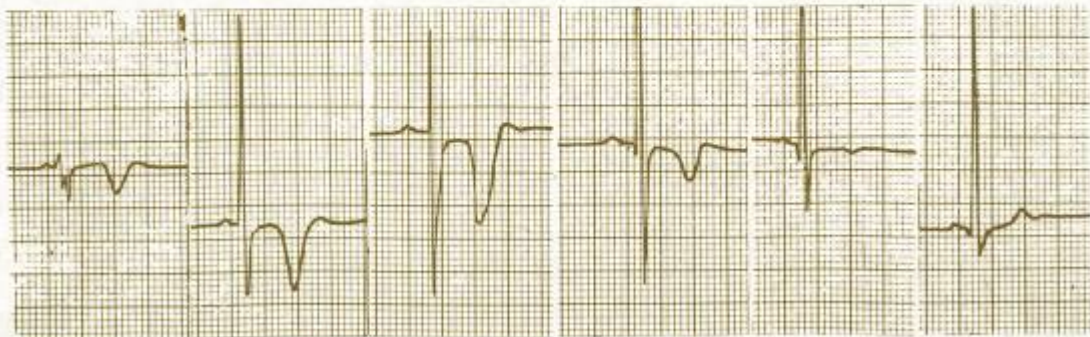
I II III aVR aVL aVF



V1 V2 V3 V4 V5 V6



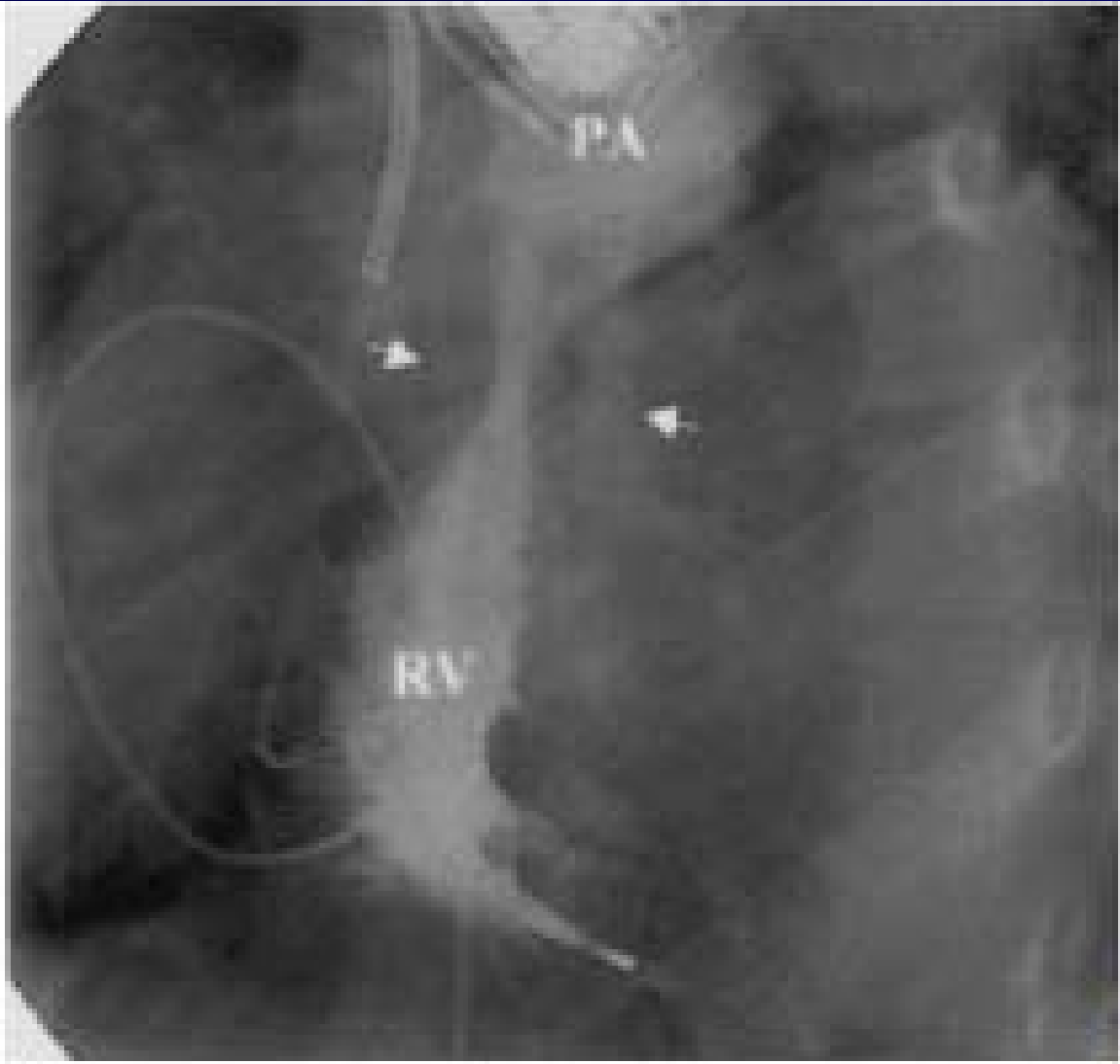
I II III aVR aVL aVF



V1 V2 V3 V4 V5 V6

# DIFERENTES PATRONES DE HIPERTROFIA





D

# MANIFESTACIONES CLINICAS

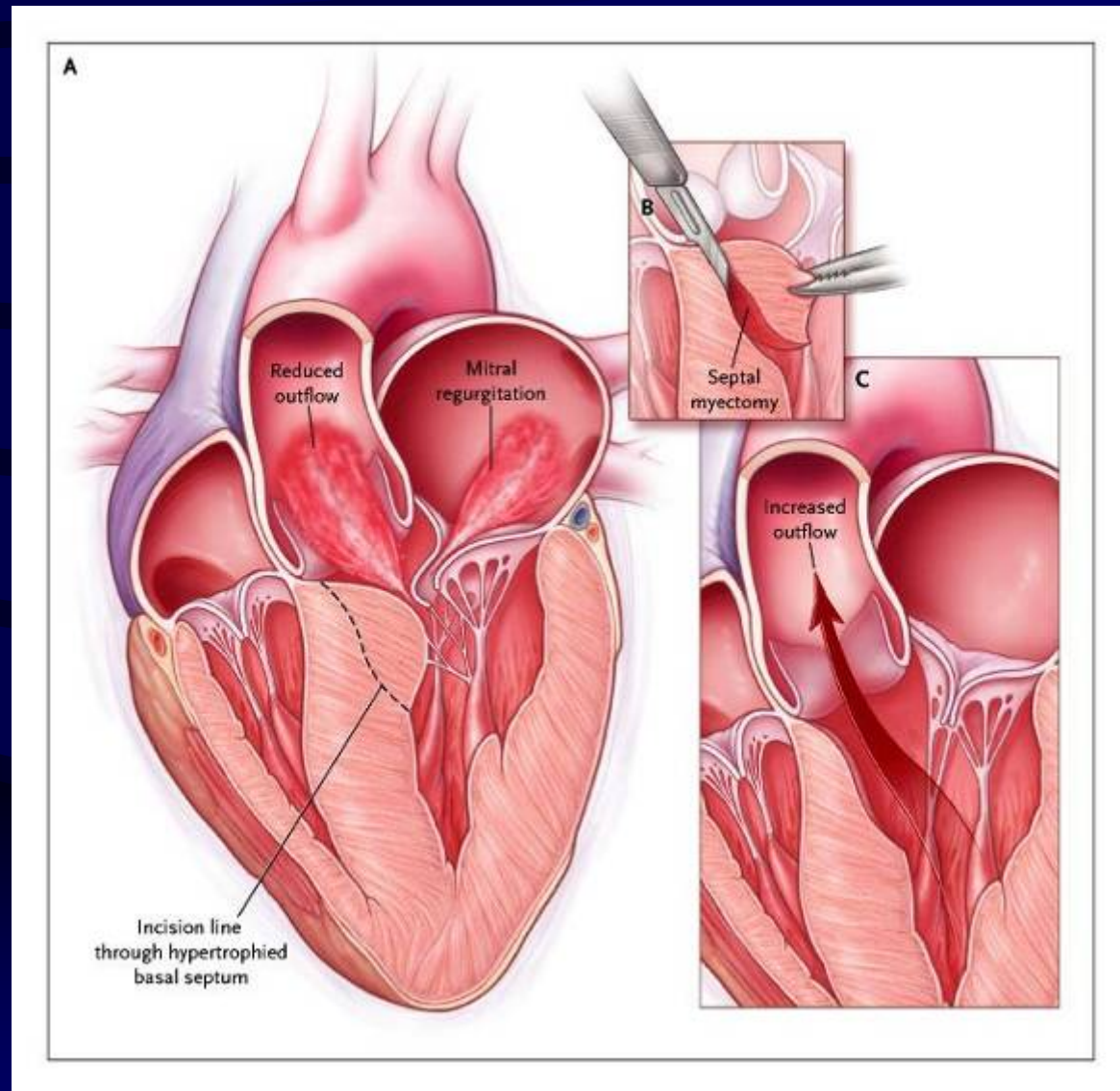
# Manifestaciones clínicas

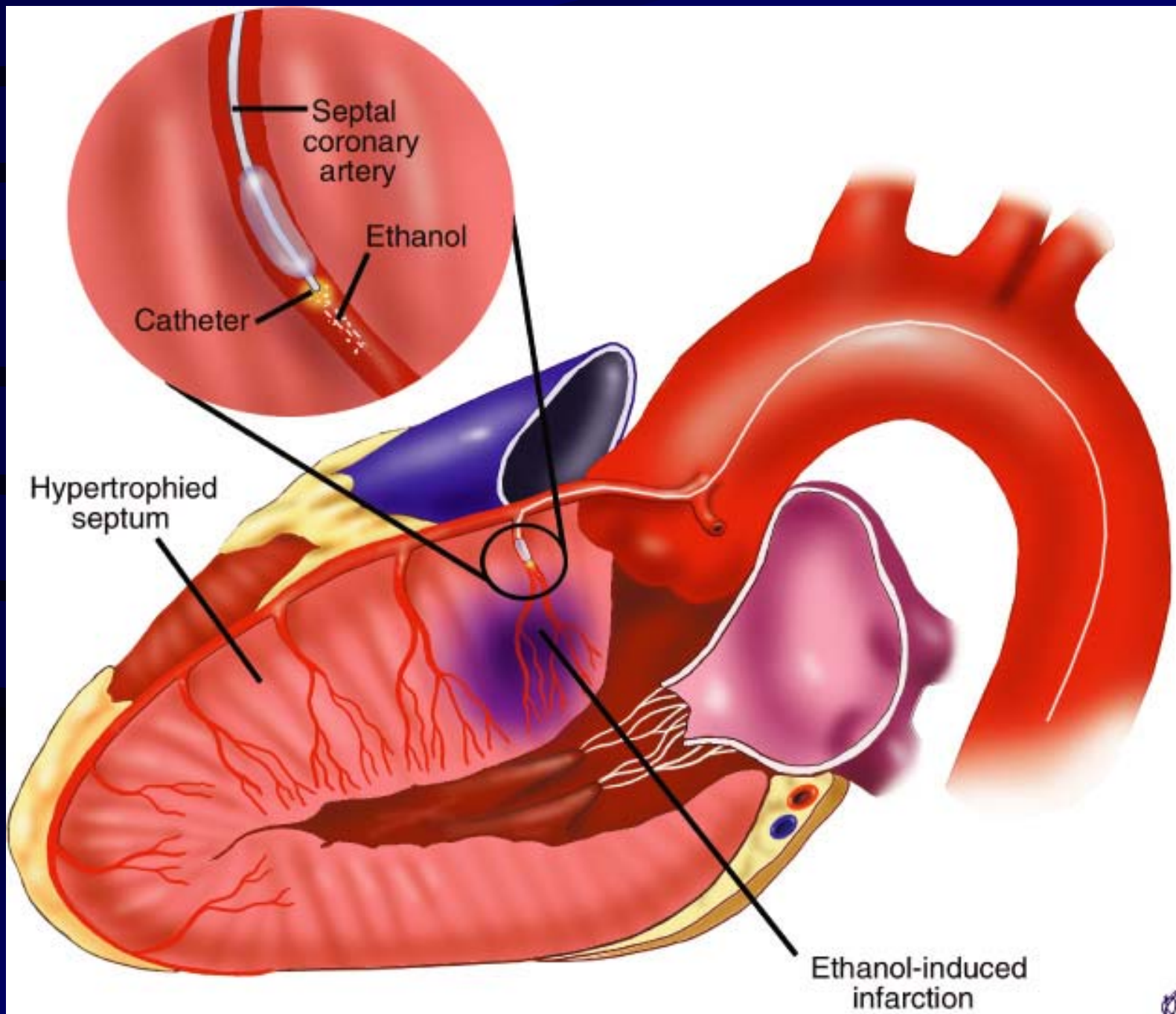
- ASINTOMATICO
- ANGOR
- SINCOPE
- DISNEA
- MUERTE SUBITA

# MCH: DROGAS

- BETA-BLOQUEANTES
- VERAPAMILO
- DISOPIRAMIDA

# Surgical Septal Myectomy





# MARCAPASOS DDD EN MCH

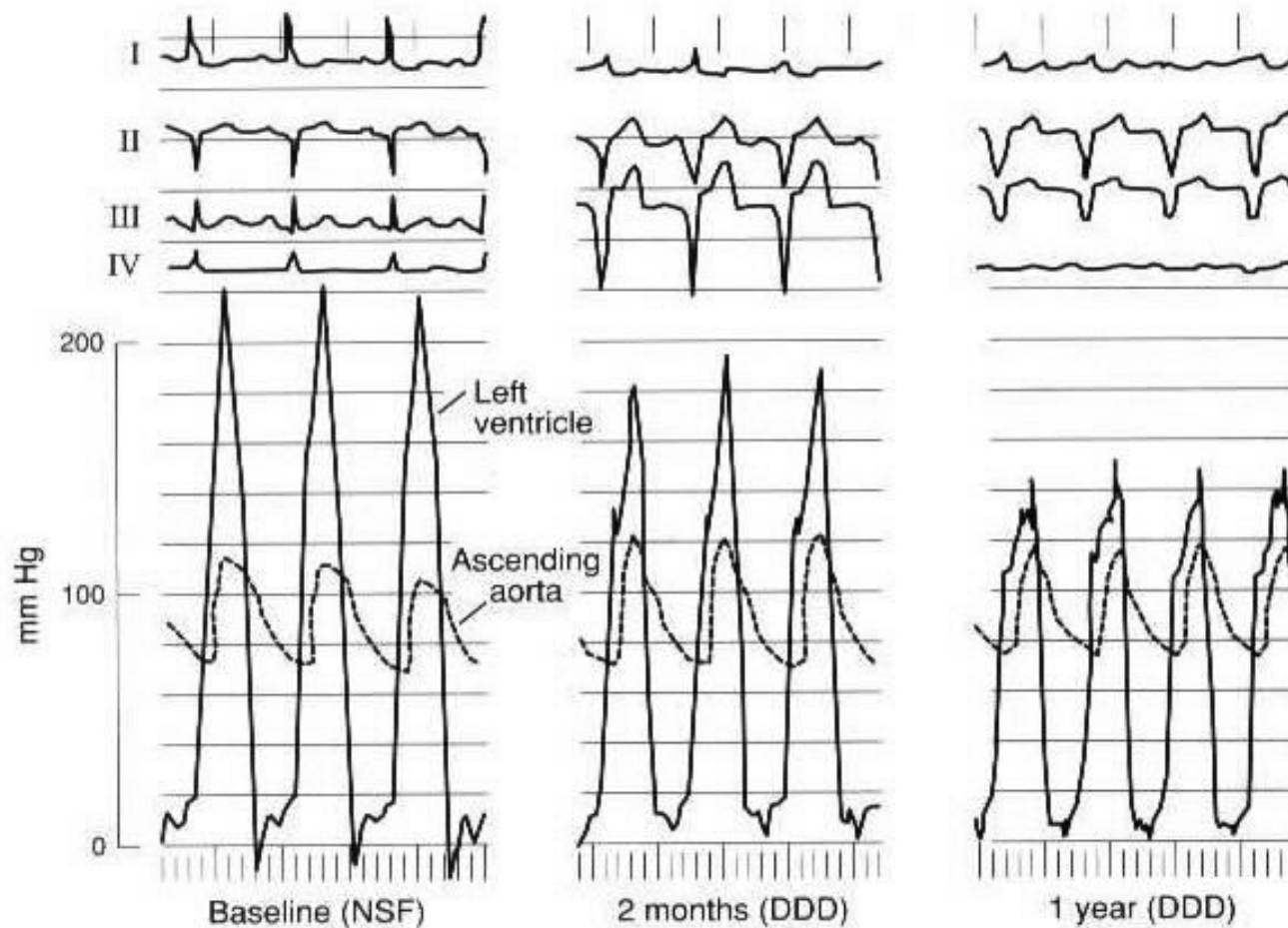


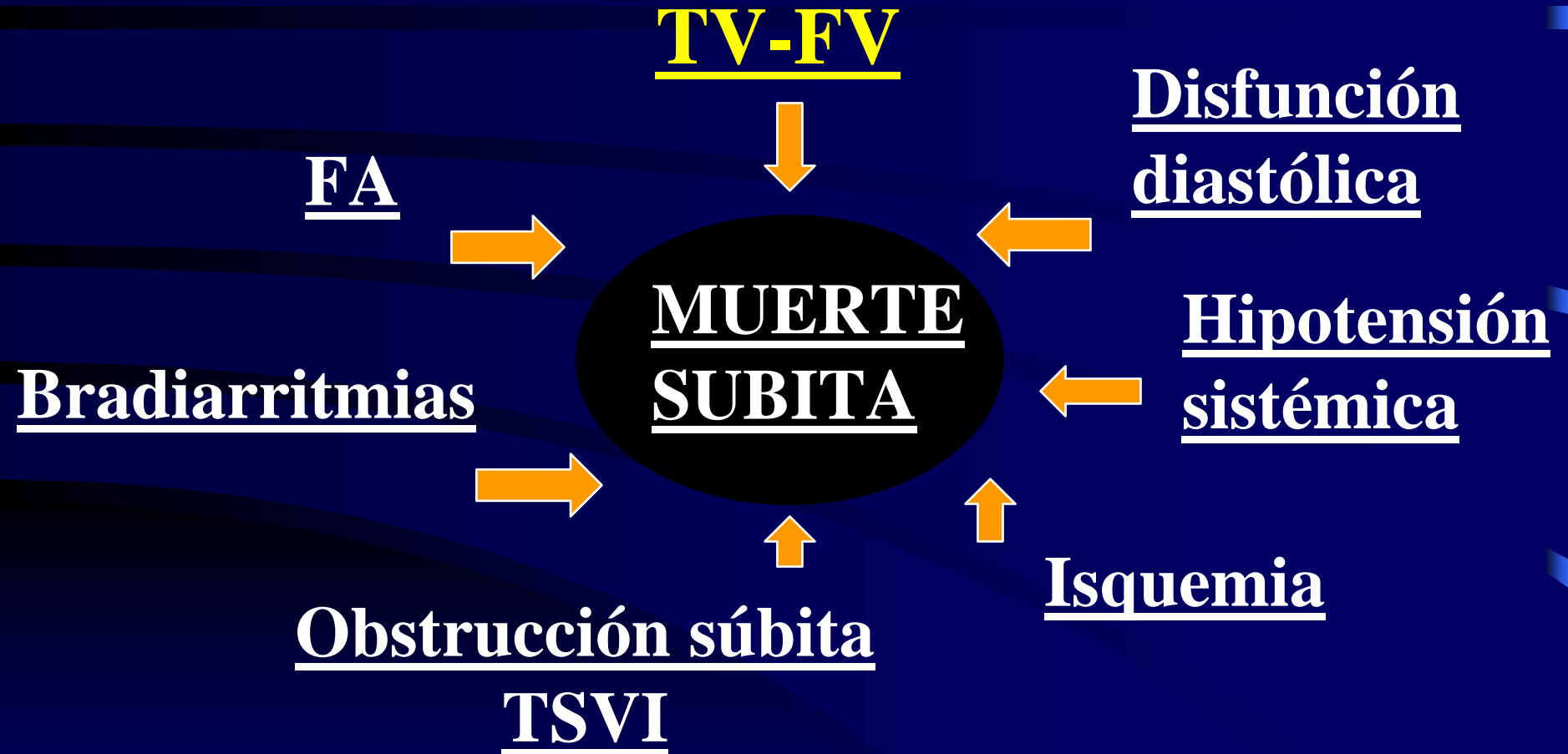
Fig. 15. Effect of dual-chamber pacing (DDD) on intraventricular gradient in hypertrophic cardiomyopathy at 2 months and 1 year. (From *Circulation* 90:2731-2742, 1994. By permission of the American Heart Association.)

# Manifestaciones clínicas

- ASINTOMATICO
- ANGOR
- SINCOPE
- DISNEA
- **MUERTE SUBITA**

# MUERTE SUBITA

# Desorganización fibrilar



# FA.....TV



# ESTRATIFICACION DE RIESGO DE MUERTE SÚBITA

## **FACTORES DE RIESGO MAYORES**

1. Historia familiar de MS
2. Síncope inexplicable
3. TVNS en holter de 24Hs
4. Comportamiento anormal de TA en esfuerzo
5. Severa HVI (mayor de 30mm)

## **POSIBLES FACTORES DE RIESGO**

1. Fibrilación auricular
2. Obstrucción TSVI
3. Mutación maligna



# Estratificación de Riesgo: Estudios de utilidad

- Historia clínica
- PEG
- Holter
- Ecocardiograma
- RMN
- Genético

# Estratificación de Riesgo: Estudios no útiles

- Q-t
- Señal Promediada
- VFC
- Estudio Electrofisiológico

# TERAPEUTICA

**“En prevención 1ria y 2ria de muerte súbita  
El CDI es la única medida terapéutica  
eficaz”**

**No hay estudios randomizados con CDI**

# M.SUBITA: PREVENCIÓN: CARDIODEFIBRILADOR

Ⓐ En Prevención Secundaria

Indicación clara

Clase I

M. Súbita Resucitado

TV espontánea

Ⓑ En Prevención Primaria

Basado en Juicio

Individual

Clase II b

\* Valorando Marcadores de Riesgo

\* Perfil clínico

Edad

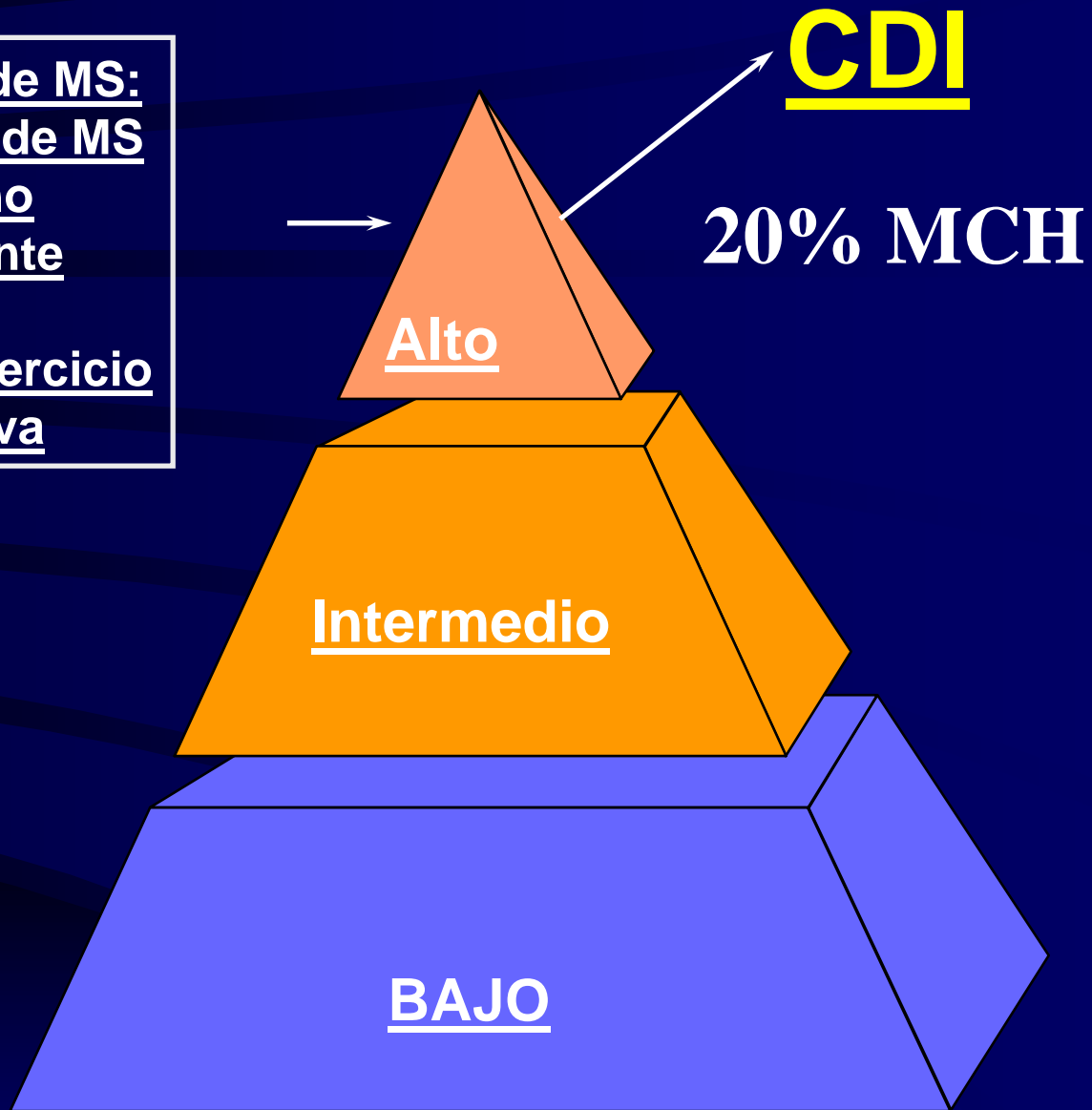
H. Familiar

Otros

\* Complicaciones del CDI

**Factores de Rx de MS:**  
Historia familiar de MS  
Genotipo maligno  
Síncope recurrente  
TVNS repetitiva  
Hipotensión c/ejercicio  
Hipertrofia masiva

0	5%
1	7%
2	18%
3	35%



# CDI PREVENCIÓN 1ª

USA

1 FACTOR MAYOR  
(Salvo hipotensión)

B. Maron

EUROPA

2 FACTORES  
MAYORES

W. Mackena

**“Patients with hypertrophic  
cardiomyopathy treated with implantable  
defibrillator”**

**Aguinaga L, Anguera I, Mont Ll, Brugada  
J.**

**J Am Coll Cardiol April 1998, 31-5:20 C.**

-

# DROGAS

**“Ninguna droga es eficaz para disminuir la MS en la MCH”**

**Amiodarona para la FA**

# MH: ATLETAS

## RECOMENDACIONES

“Evidencia que evitar deporte competitivo disminuye el riesgo de muerte súbita”.

ACC/ESC Expert consensus document

# American College of Cardiology/European Society of Cardiology Clinical Expert Consensus Document on Hypertrophic Cardiomyopathy

A report of the American College of Cardiology Foundation Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines<sup>☆</sup>

Writing Committee Members, Barry J. Maron, (Co-Chair)<sup>a,b</sup>, William J. McKenna, (Co-Chair)<sup>a,b\*</sup>, Gordon K. Danielson<sup>a</sup>, Lukas J. Kappenberger<sup>a,b\*</sup>, Horst J. Kuhn<sup>b\*</sup>, Christine E. Seidman, Pravin M. Shah<sup>a</sup>, William H. Spencer III<sup>a</sup>, Paolo Spirito<sup>a,b\*</sup>, Folkert J. Ten Cate<sup>a,b\*</sup>, E. Douglas Wigle<sup>a</sup>

ACCF Task Force on Clinical Expert Consensus Documents Members, Robert A. Vogel, (Chair), Jonathan Abrams, Eric R. Bates, Bruce R. Brodie†, Peter G. Danias†, Gabriel Gregoratos, Mark A. Hlatky, Judith S. Hochman, Sanjiv Kaul, Robert C. Lichtenberg, Jonathan R. Lindner, Robert A. O'Rourke‡, Gerald M. Pohost, Richard S. Schofield, Cynthia M. Tracy†, William L. Winters Jr†

ESC Committee for Practice Guidelines Members\*, Werner W. Klein, (Chair), Silvia G. Priori, (Co-Chair), Angeles Alonso-Garcia, Carina Blomström-Lundqvist, Guy De Backer, Jaap Deckers, Markus Flather, Jaromir Hradec, Ali Oto, Alexander Parkhomenko, Sigmund Silber, Adam Torbicki

## Table of contents

Preamble .....	1966
Introduction .....	1966
Organization of committee and evidence review .....	1966
Purpose of this Expert Consensus Document .....	1966
General considerations and perspectives .....	1966
Nomenclature, definitions, and clinical diagnosis ..	1967
Obstruction to LV Outflow .....	1967
Genetics and molecular diagnosis .....	1968
General considerations for natural history, and clinical course .....	1970

## Symptoms and pharmacological management

strategies .....	1972
Beta-adrenergic blocking agents .....	1972
Verapamil .....	1973
Disopyramide .....	1973
Drugs in end-stage phase .....	1974
Asymptomatic patients .....	1974
Infective endocarditis prophylaxis .....	1974
Pregnancy .....	1974
Treatment options for drug-refractory patients .....	1974
Additional approaches to relieve outflow obstruction and symptoms .....	1976

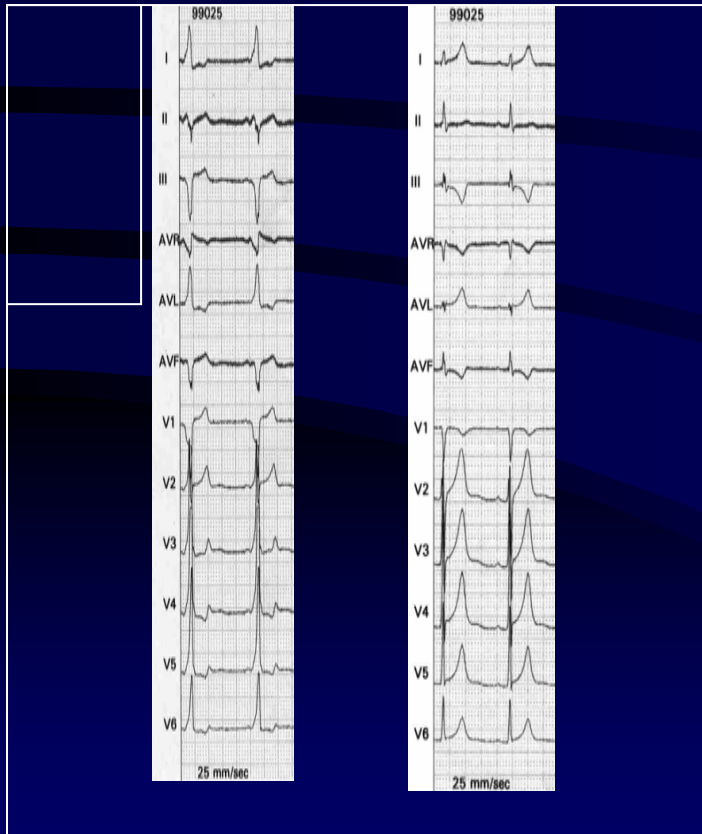
# MUERTE SUBITA

## CASO CLINICO



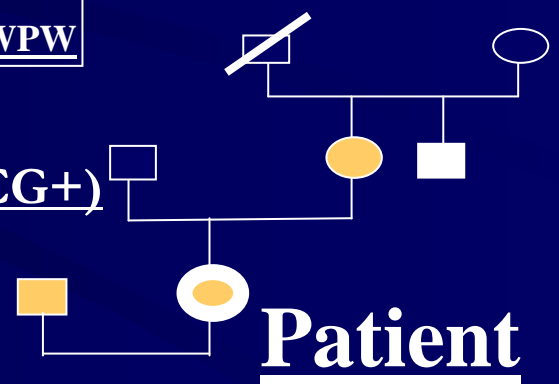
# “ CLINICAL GENOTYPE-PHENOTYPE IN A PATIENT WITH FAMILIAL WPW ”

Luis Aguinaga. Heart Rhythm Society 2006



FAMILIAL WPW

- Affected (ECG+)
- No affected
- ◻ Dead



Patient

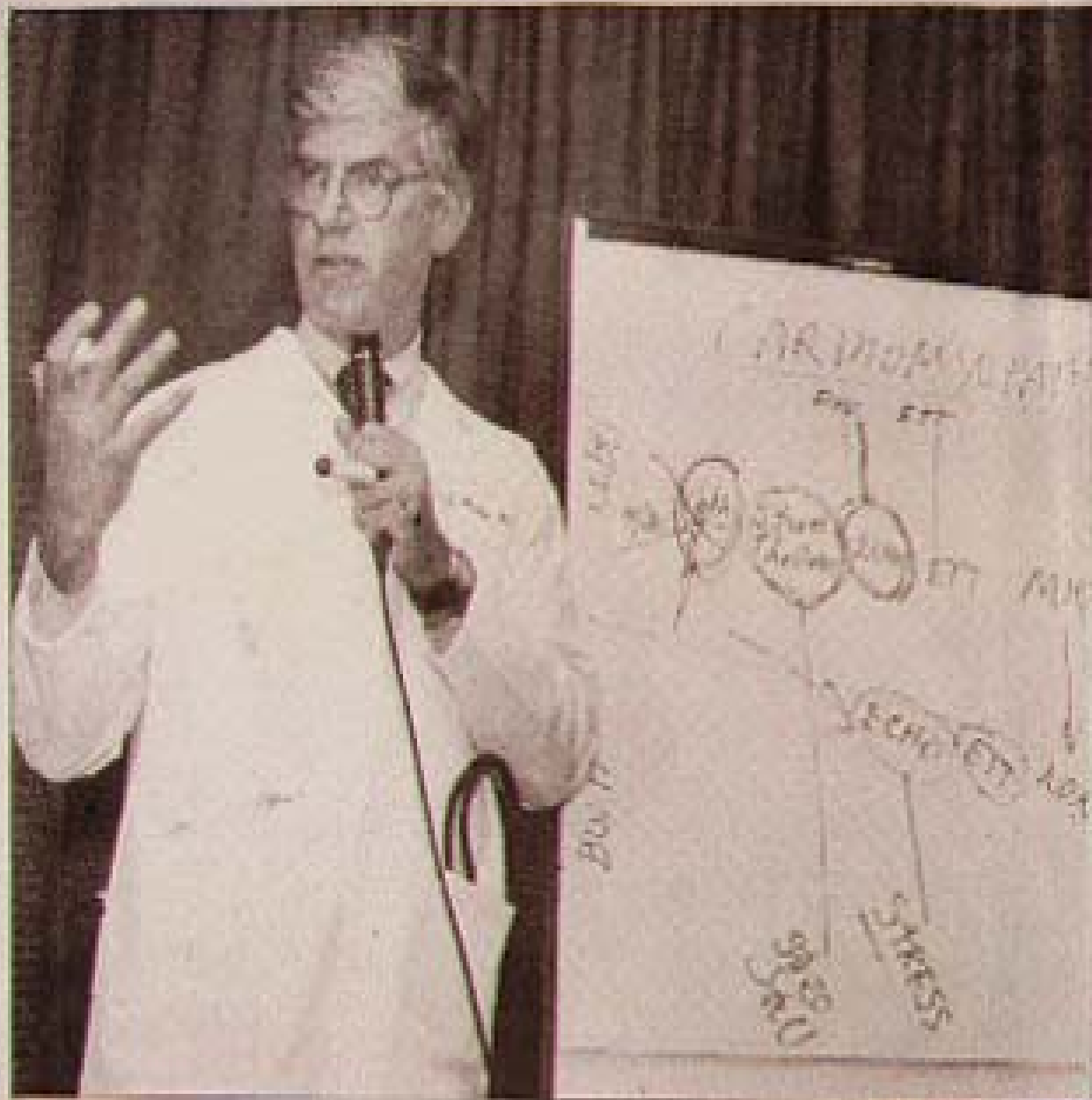
ECG BASAL    +    -    +

# Reggie Lewis

- **Lipotimias**.....4 meses
- **Síncope**.....playoff...29/4/93
- Hospital Boston
  - Hipertrofia septo-apical
  - **TVNS monitor.**
  - Indican EEF.....CDI....Fin de competencia
  - “dream team” de médicos

# Reggie Lewis

- Otra opinión
- Tilt-test positivo
- Retorna al deporte competitivo



Dr. Gilbert H. Mudge explains his diagnosis at a May 10 press conference, contradicting the "Drum Team" of cardiologists that concluded Reggie Lewis had cardiomyopathy.

¡¡Muere el 27/7/93

jugando al basketball....!!

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**REGGIE LEWIS**  
**MS-1993**

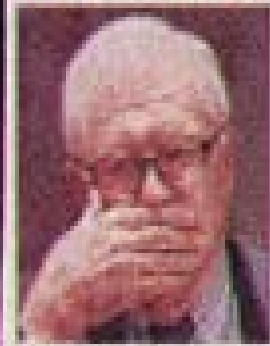


134 pages  
30 cents

# The Boston Globe

THURSDAY, APRIL 8, 2004

## CARDIOLOGIST DOUBTED MUDGE DIAGNOSIS



Dr. Eugene Braunwald (above), former boss of Dr. Gilbert H. Mudge Jr., testified yesterday that he had misgivings about Mudge's diagnosis of Caffery over Reggie Lewis and worried that, if Mudge were wrong, Lewis could die. Braunwald, a reluctant witness, appeared during the trial of the magnate and attorney from Lewis's death in 1981. Mudge and Lewis's widow, Debra Harris-Lewis, are at right. 32



WILLIAM H. WOOD



*“El futuro no es una página en blanco sino una  
fe de erratas”*

M. Benedetti

FIN

“Comprender es la suprema perfección y la verdadera sabiduría”

Heráclito, siglo VI a.de C