

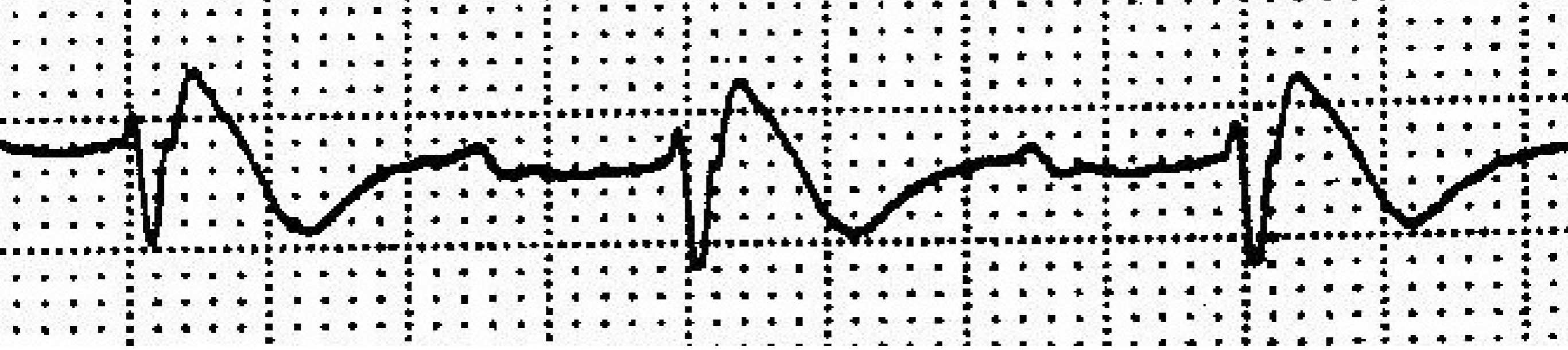
Voorlichtingsbijeenko  
mst  
25-05-2024

# Brugada syndroom



Erasmus MC  
Universitair Medisch Centrum Rotterdam





# Wat is Brugada syndroom



Amsterdam UMC  
Universitair Medische Centra



JACC Vol. 20, No. 6  
November 15, 1992:1391-6

1391

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**Right Bundle Branch Block, Persistent ST Segment Elevation  
and Sudden Cardiac Death: A Distinct Clinical and  
Electrocardiographic Syndrome**

**A Multicenter Report**

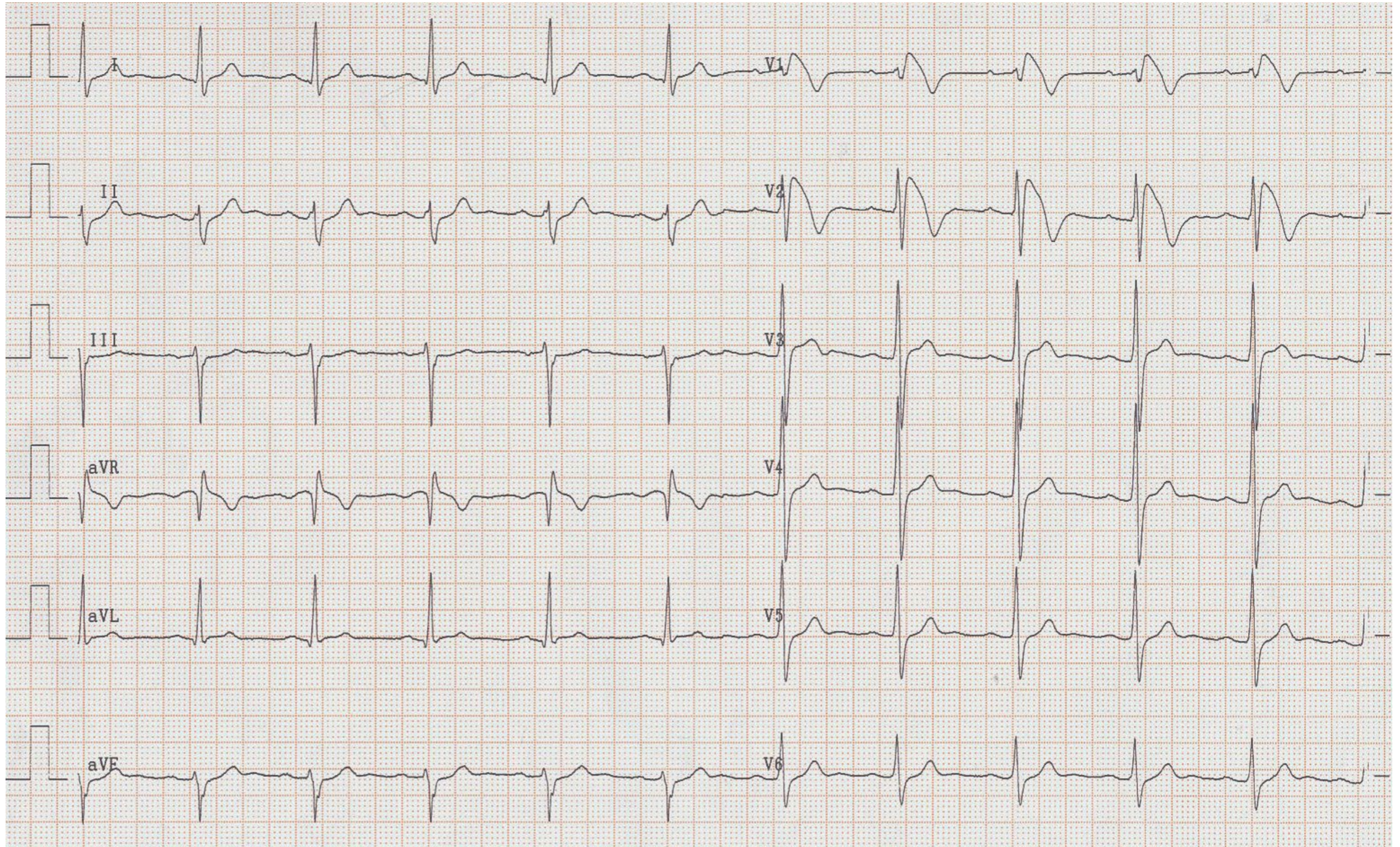
PEDRO BRUGADA, MD, JOSEP BRUGADA, MD\*†

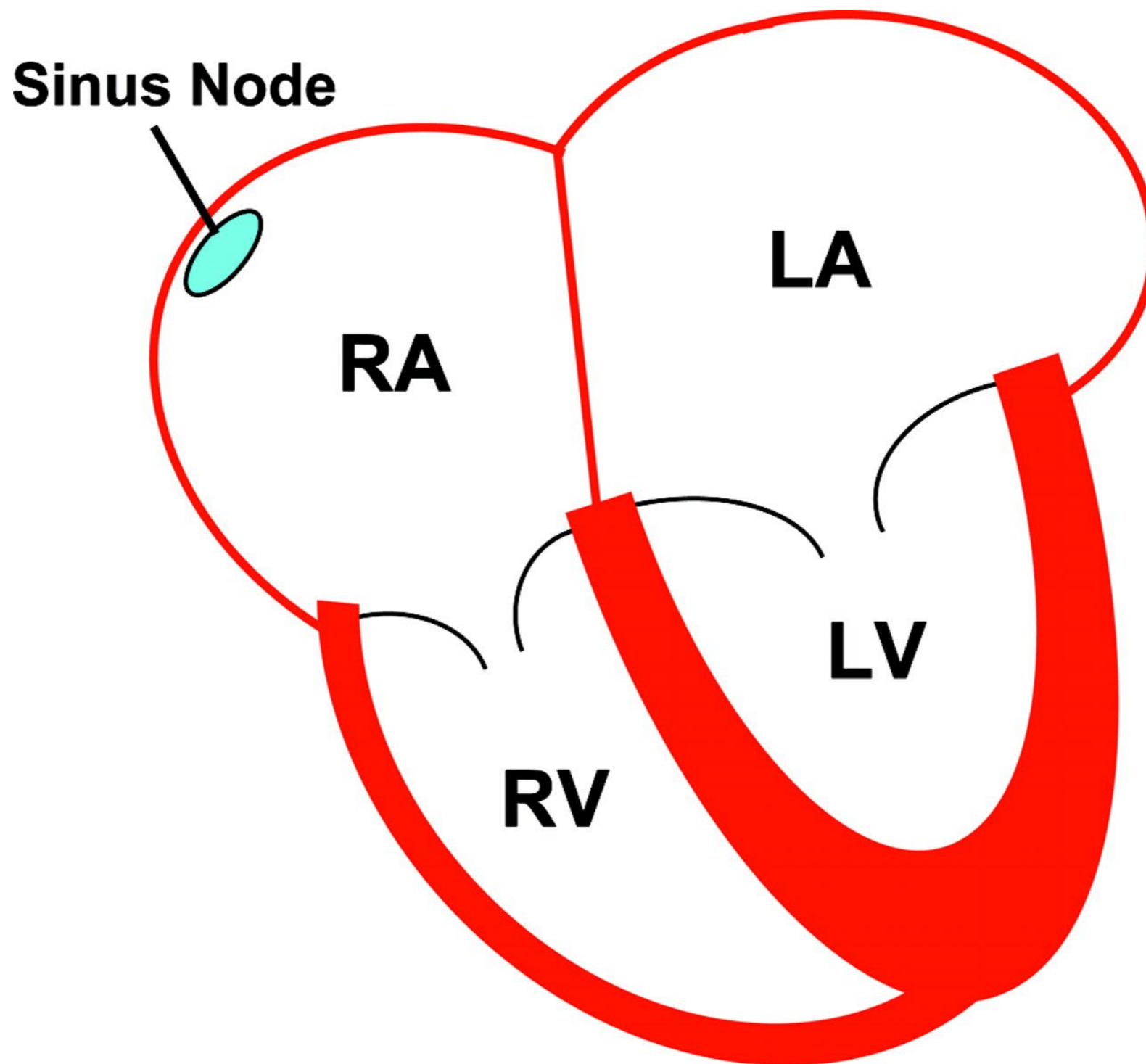
*Aalst, Belgium and Barcelona, Spain*

**Voor het eerst beschreven in 1992 door 2 broers**

**In dit stuk beschrijven ze 8 gereanimeerde patienten  
met een karakteristiek ECG**

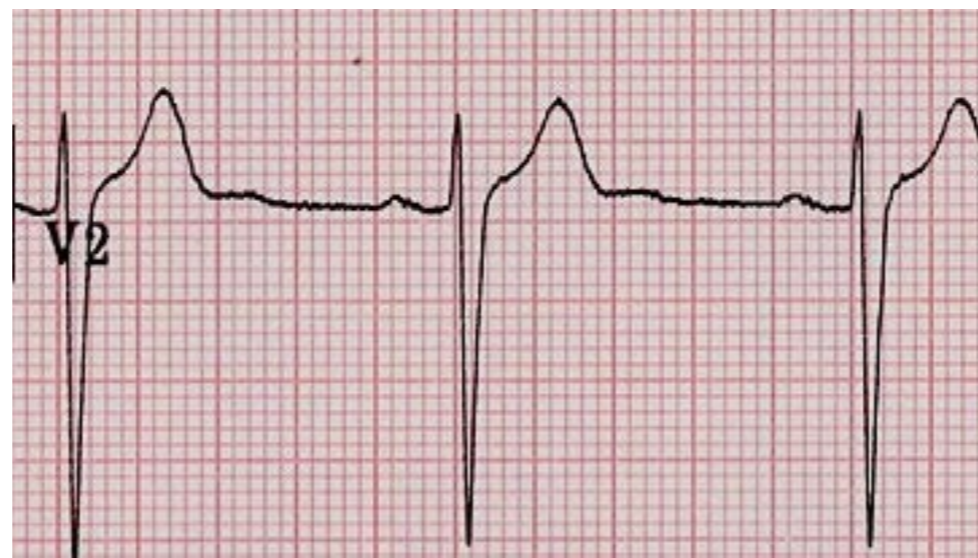
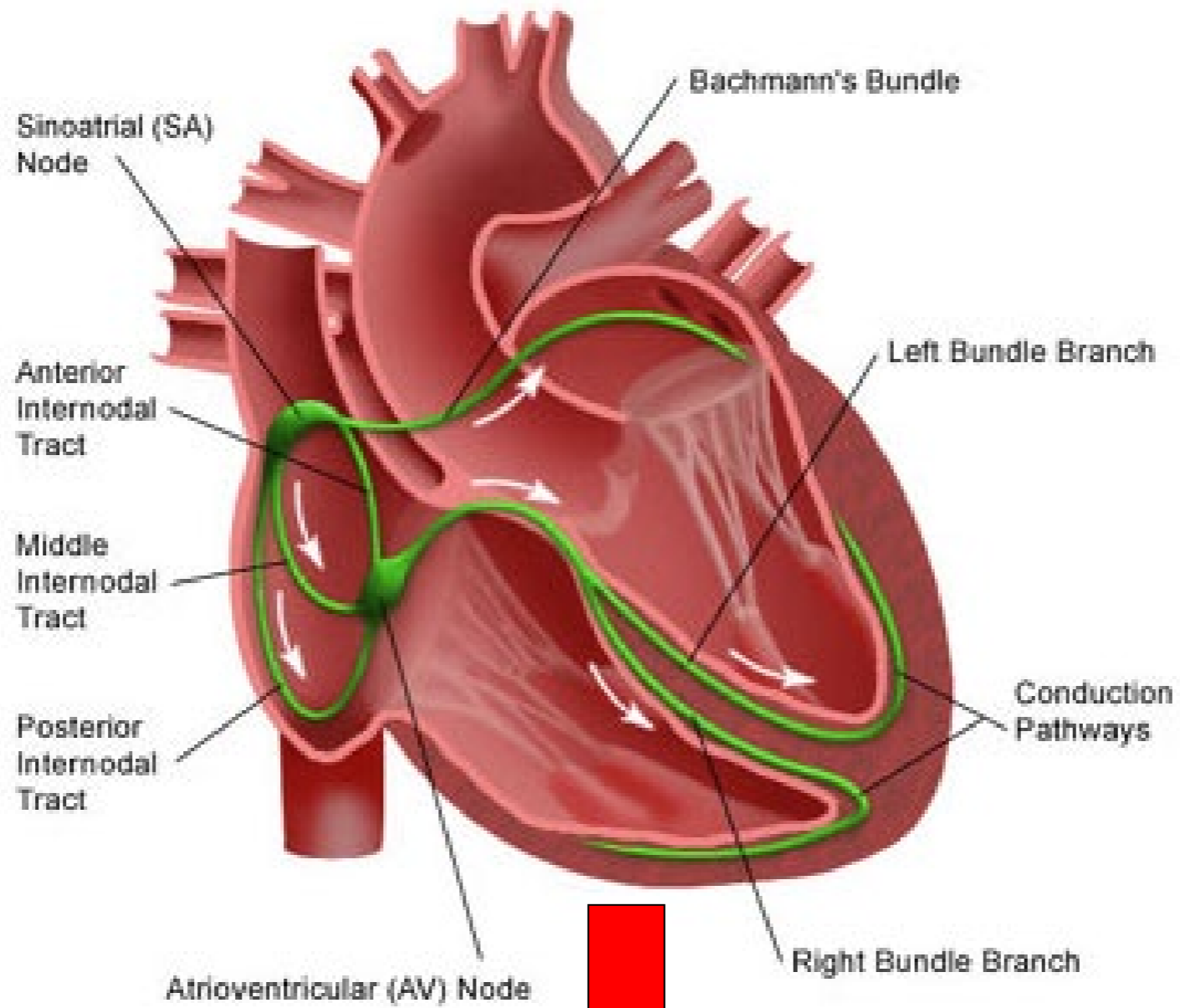
# Man 39 jaar (gereanimeerd)



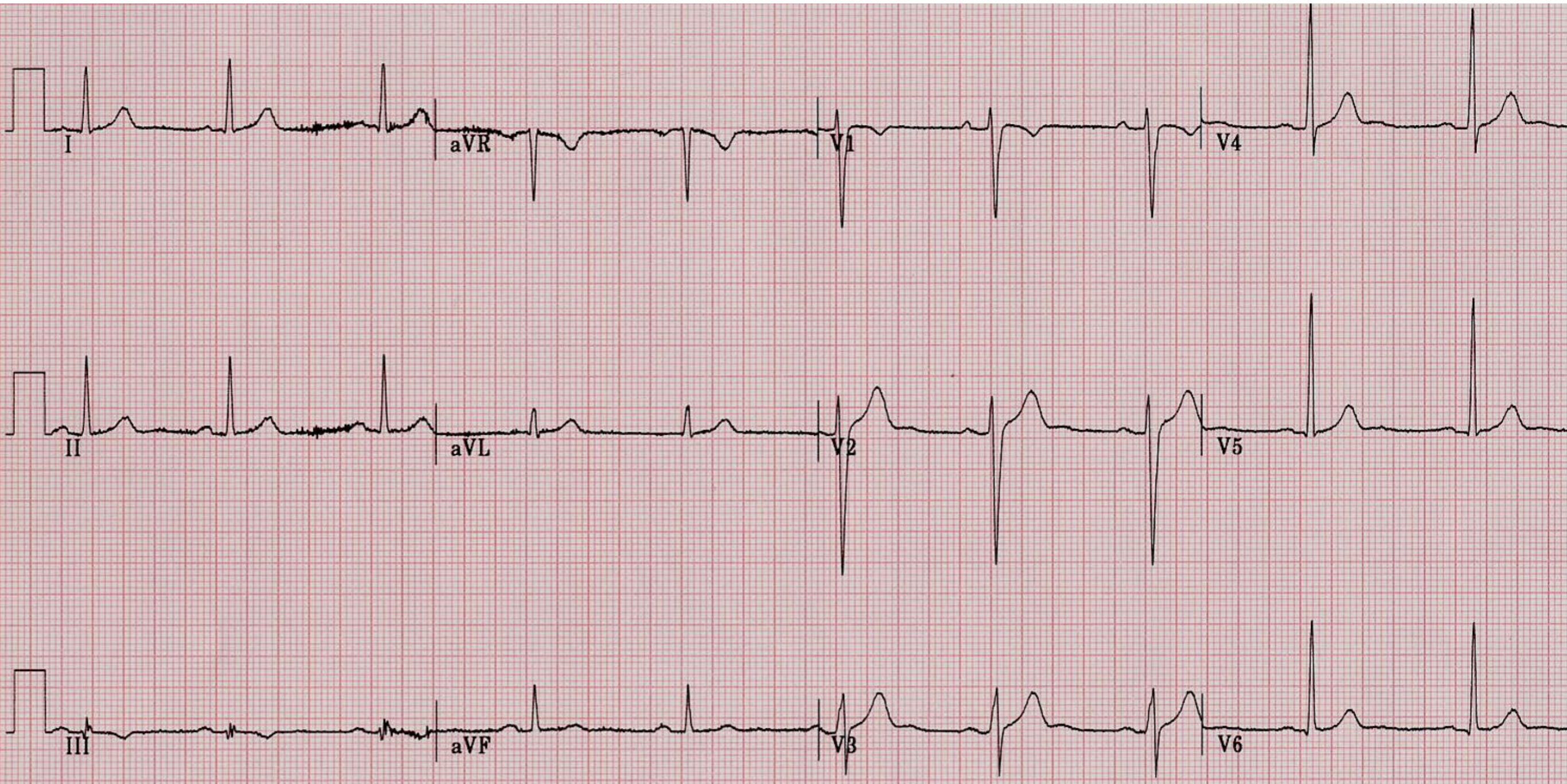


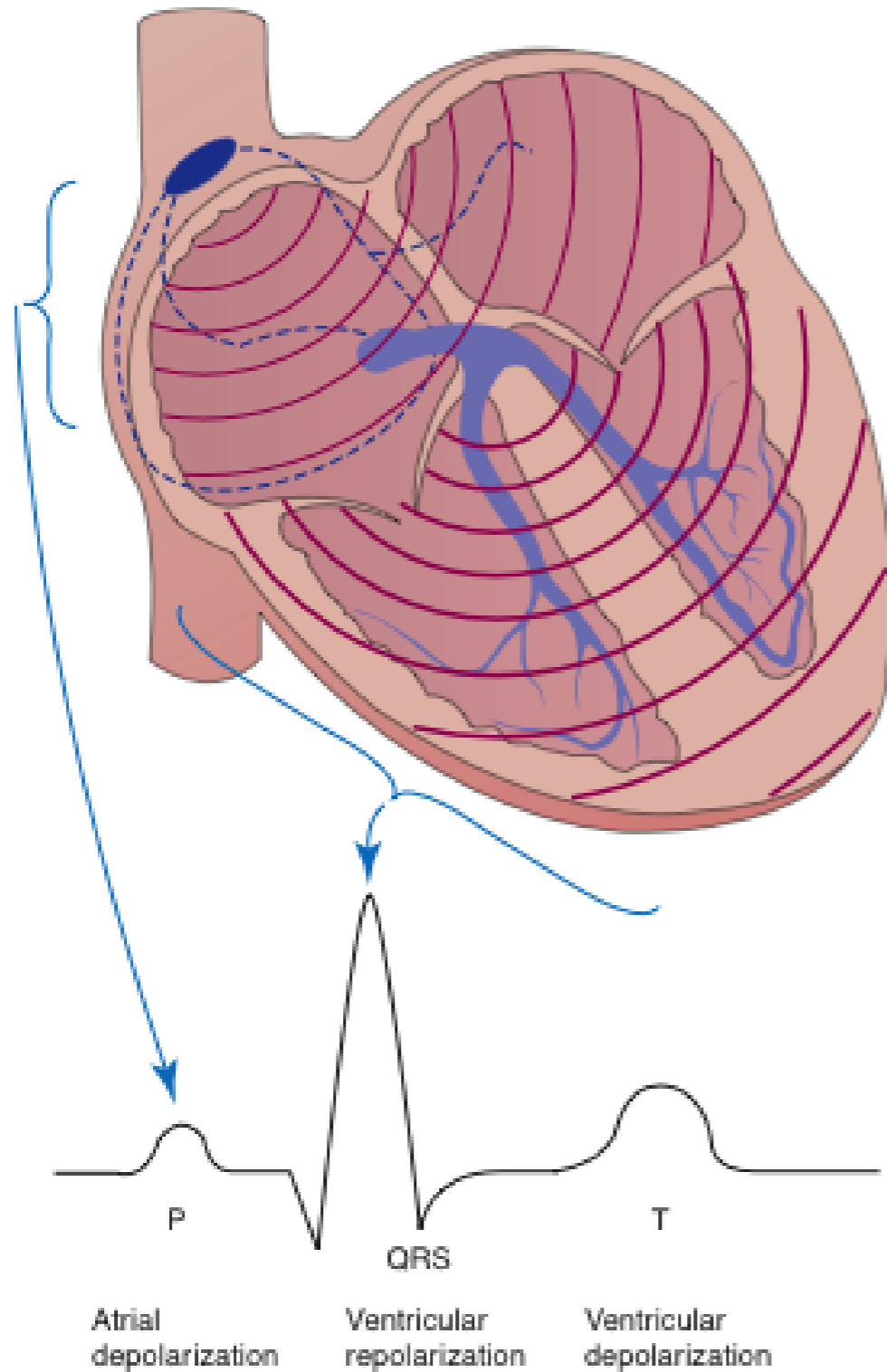
Cooper, J. M. *Circulation* 2005;112:e299-301e

# Electrical System of the Heart



# Normaal hartfilmpje (ECG)





**Sinusknop**

**boezem**

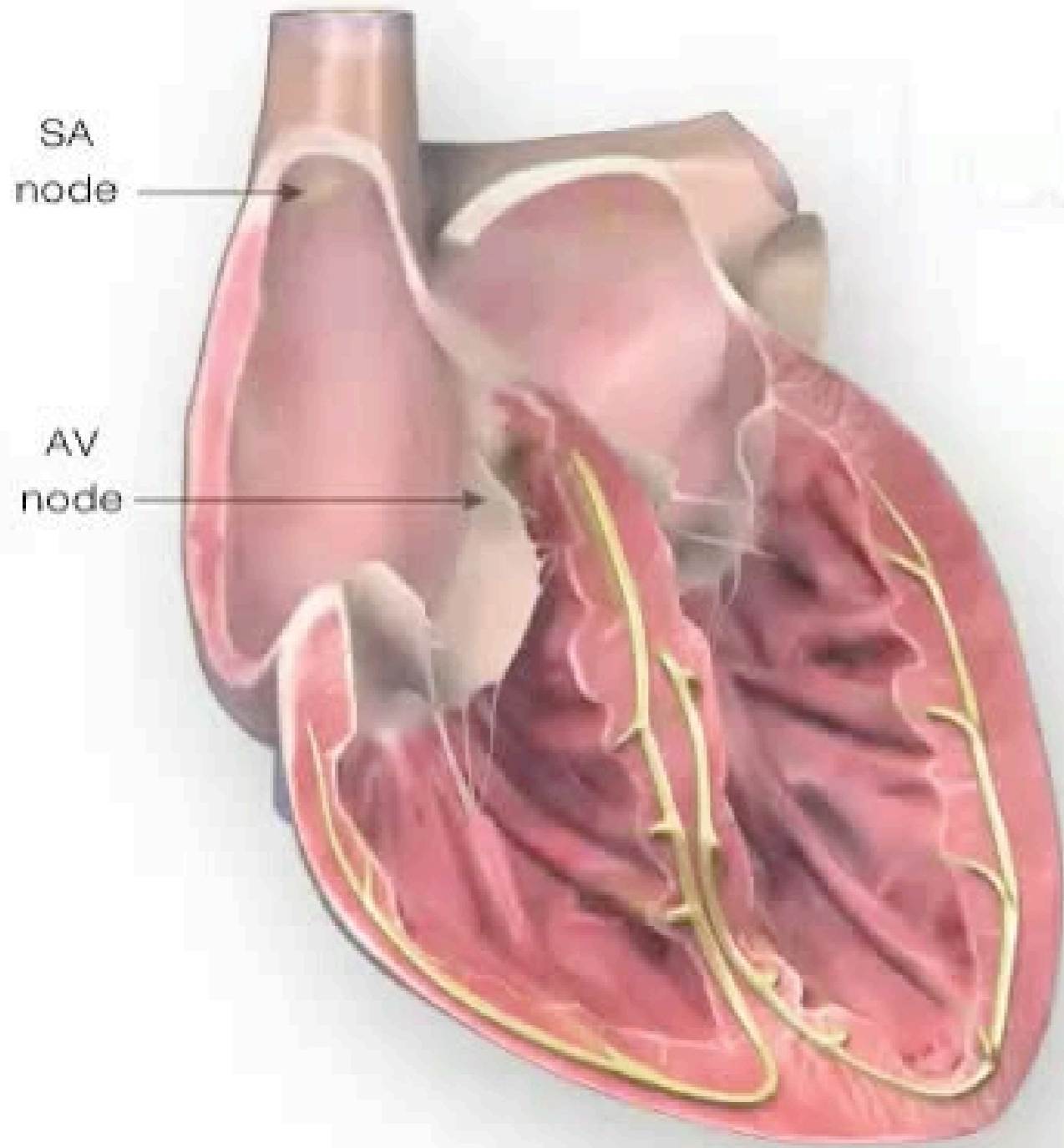
**AV knoop**

**Geleidingsyteem**

**hartkamers (L + R)**

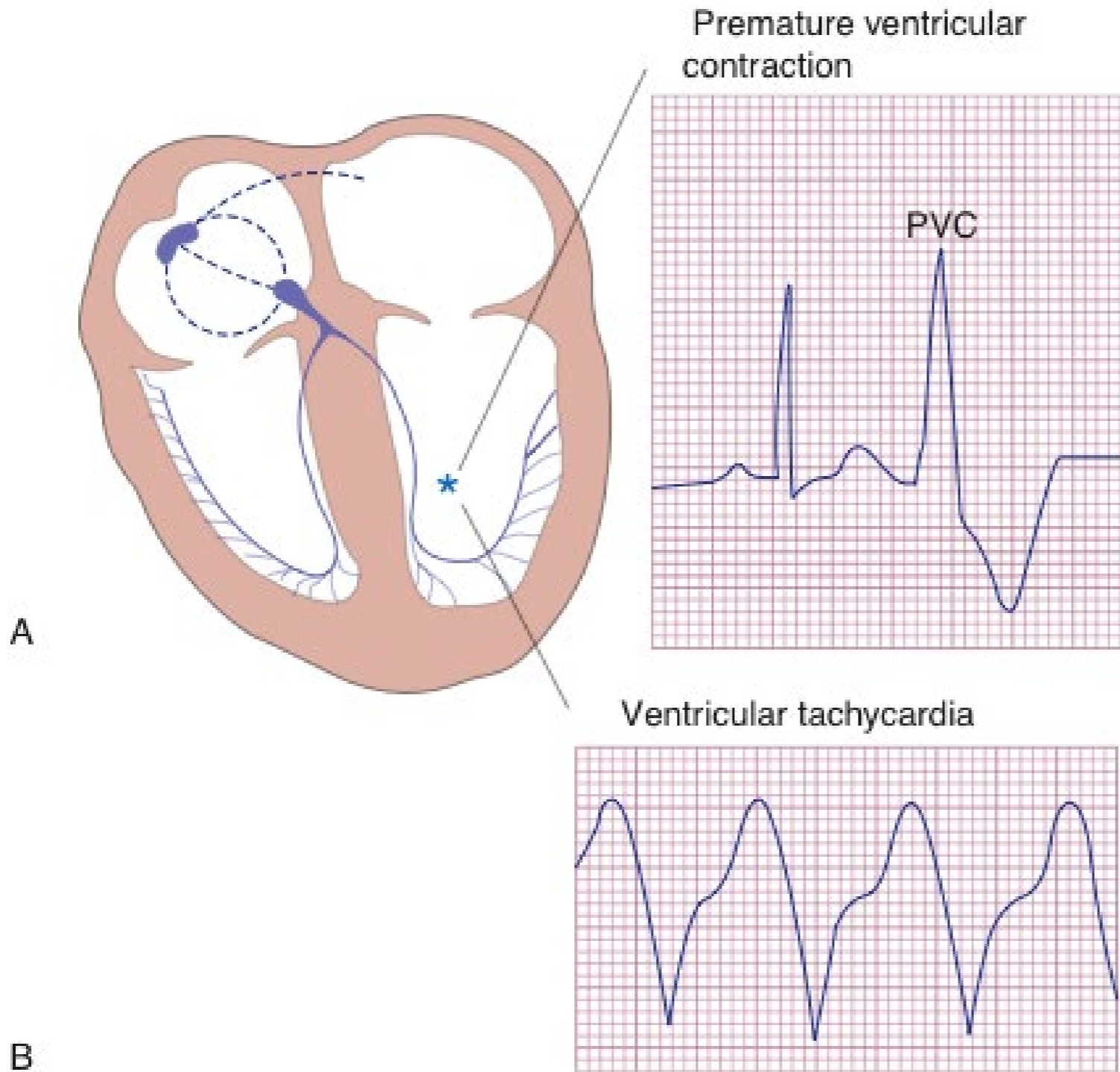


# ***De elektrische impuls en het ECG***



**SA node: Sinus Knoop**

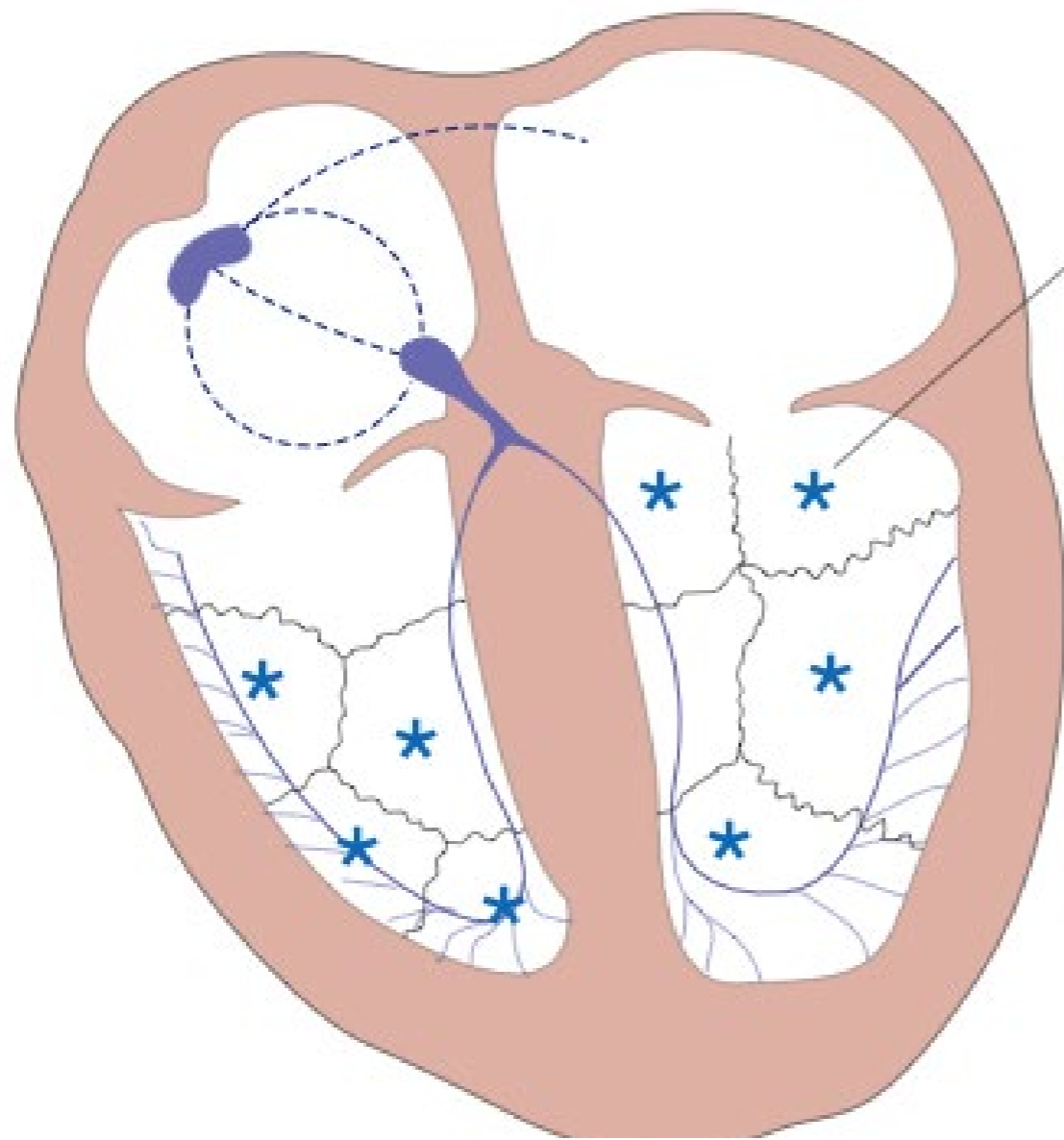
**AV node: Atrio-Ventricular knoop**



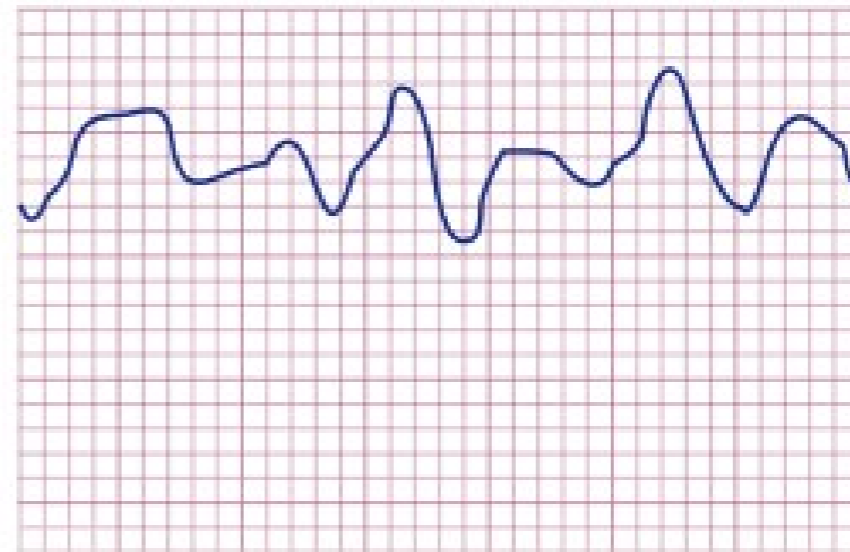
# Ritmestoornis uit de Ventrikel

Man 68 jaar, hartkloppingen, 4 jaar geleden hartinfarct



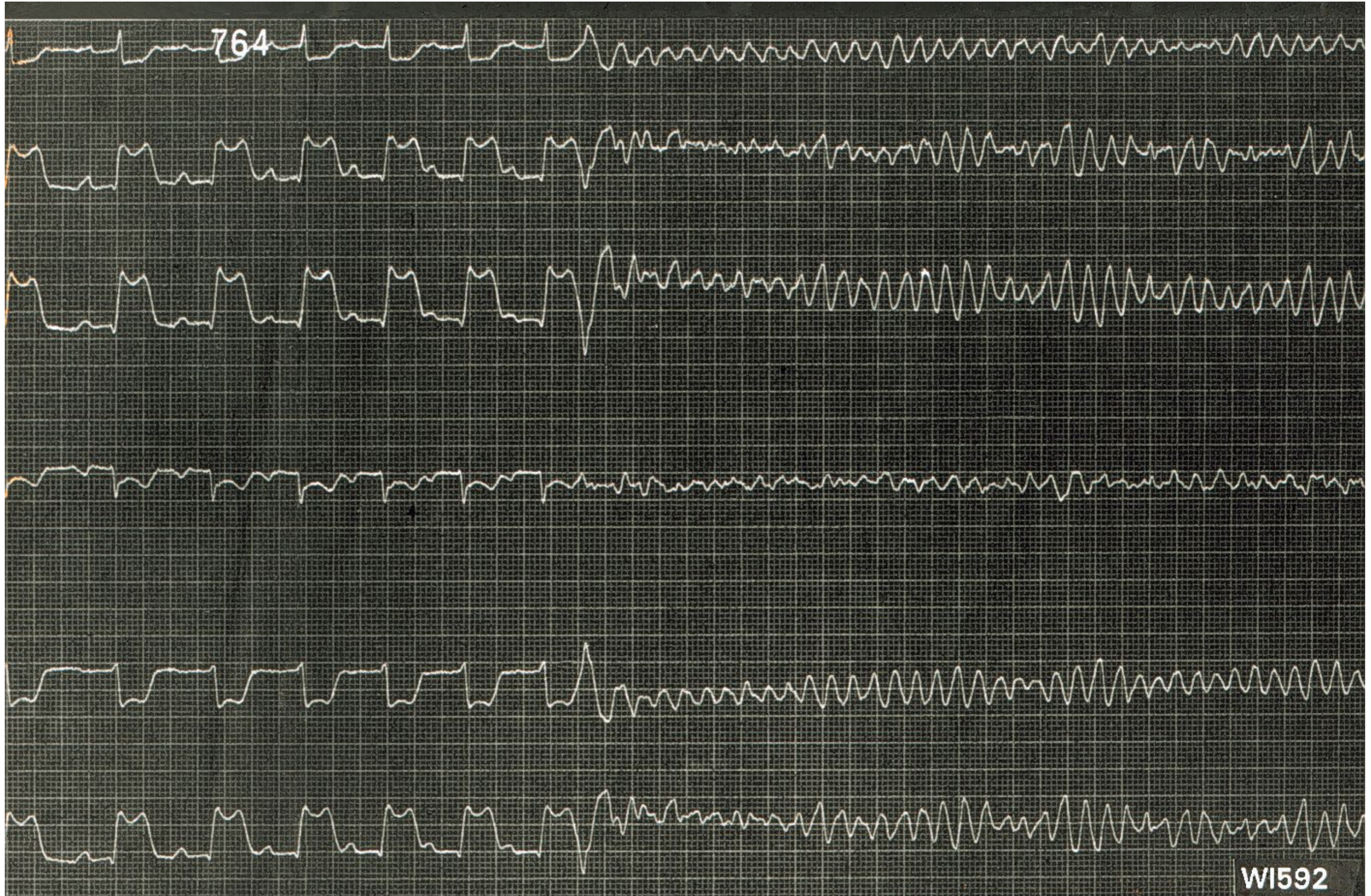


Ventricular fibrillation



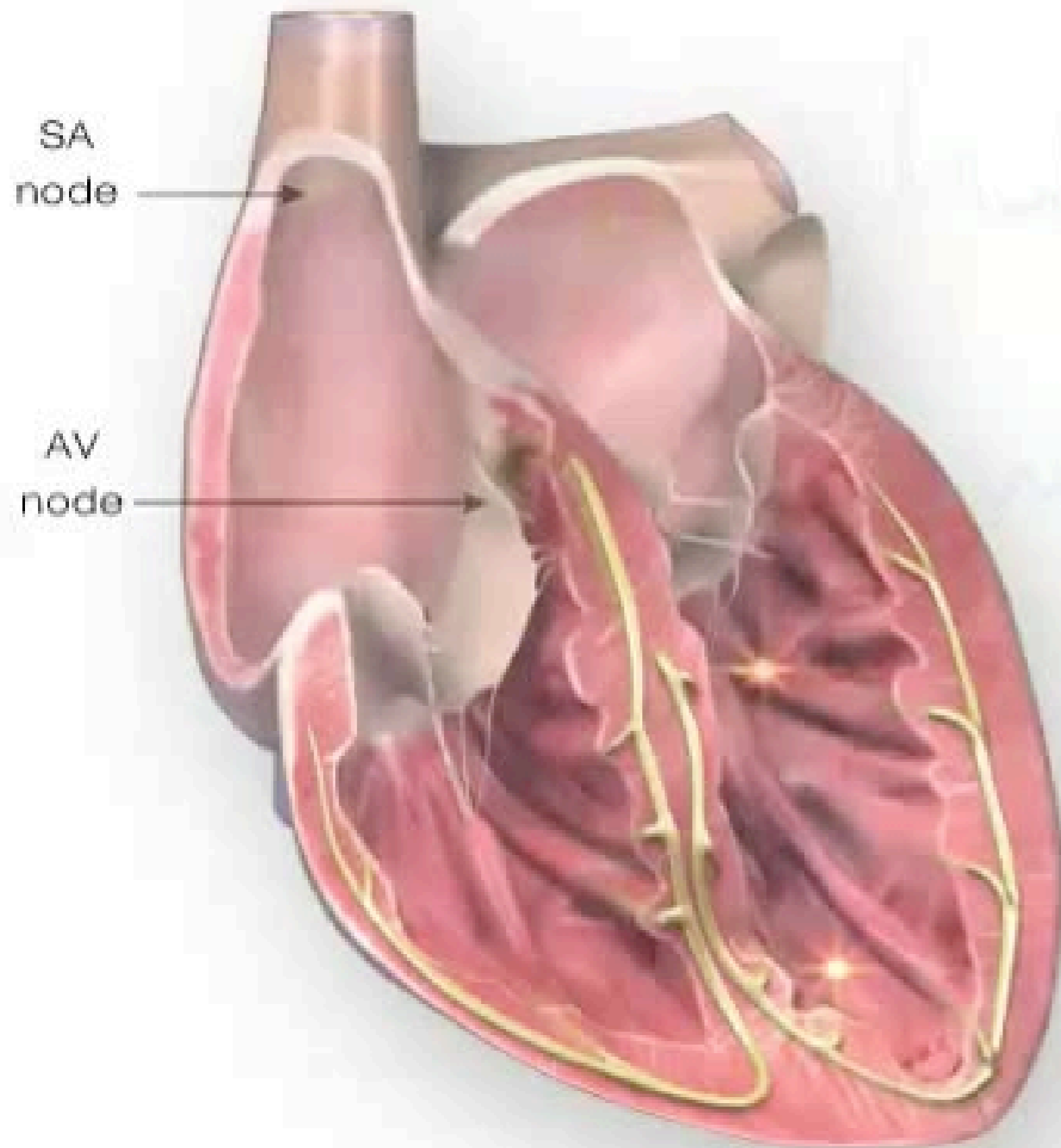
# Ritmestoornis uit de Ventrikel

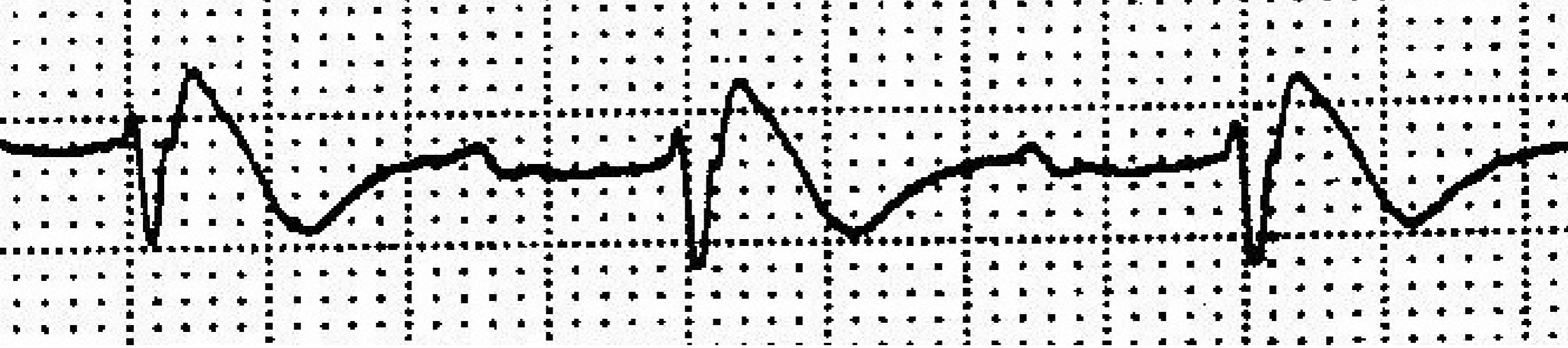
# ventrikel fibrilleren



# *Ventrikel fibrilleren*

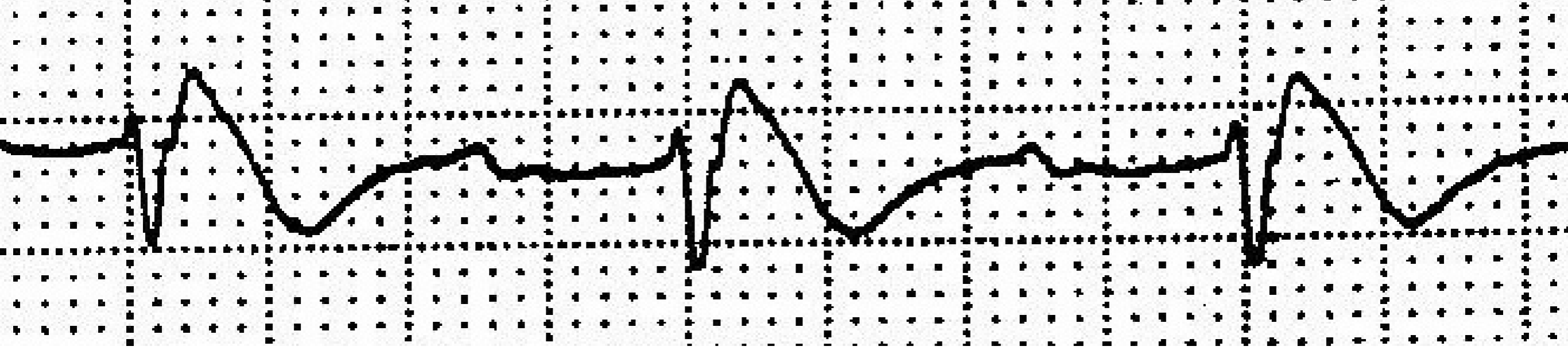
*- De meest voorkomende dodelijke ritmestr.-*





## Brugada syndrome:

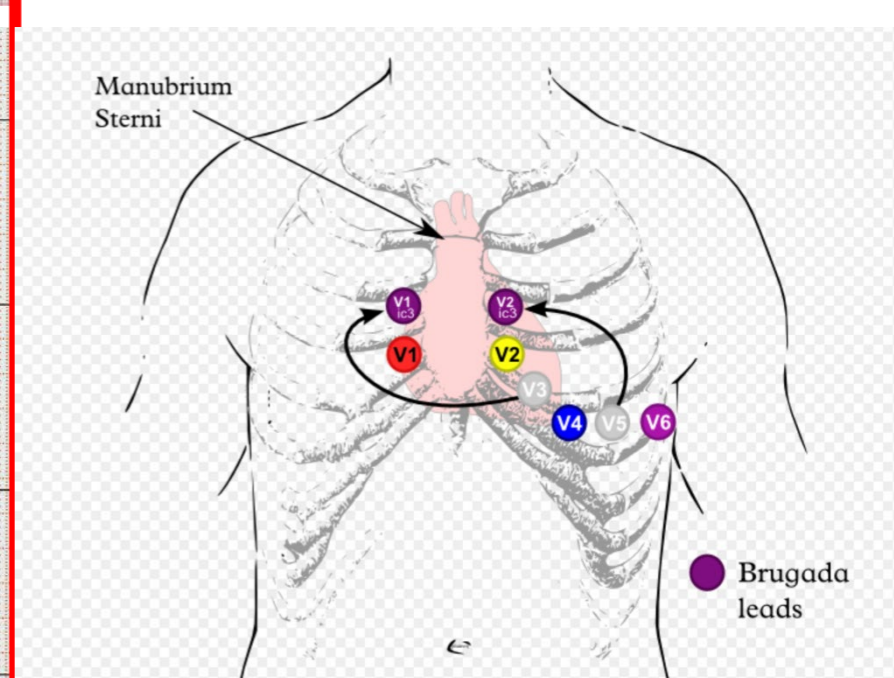
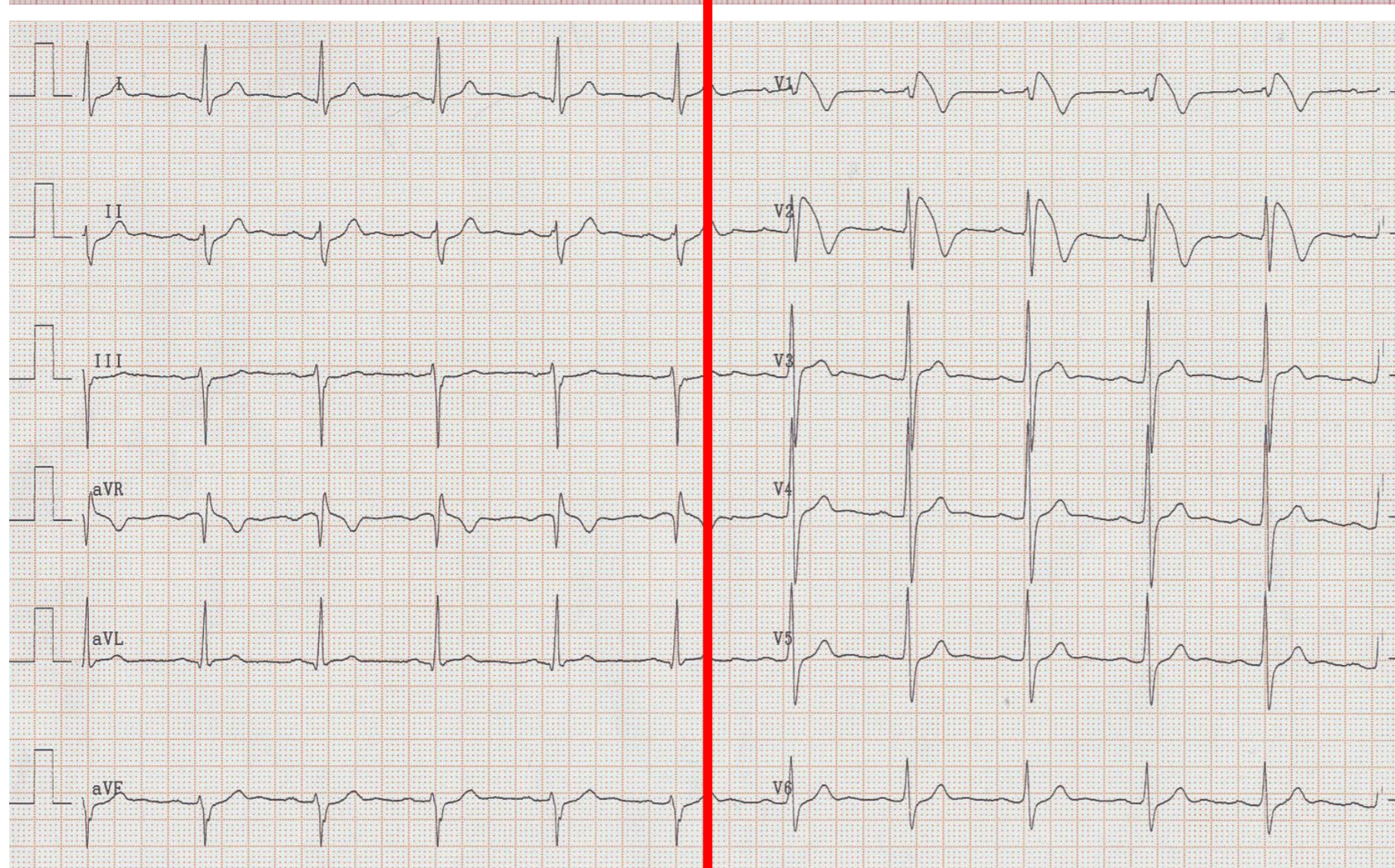
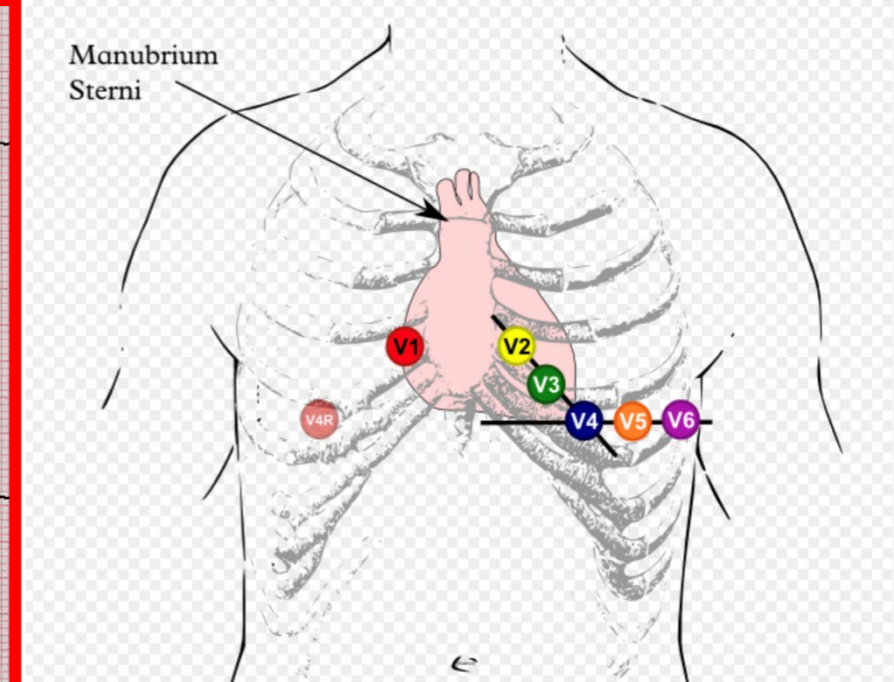
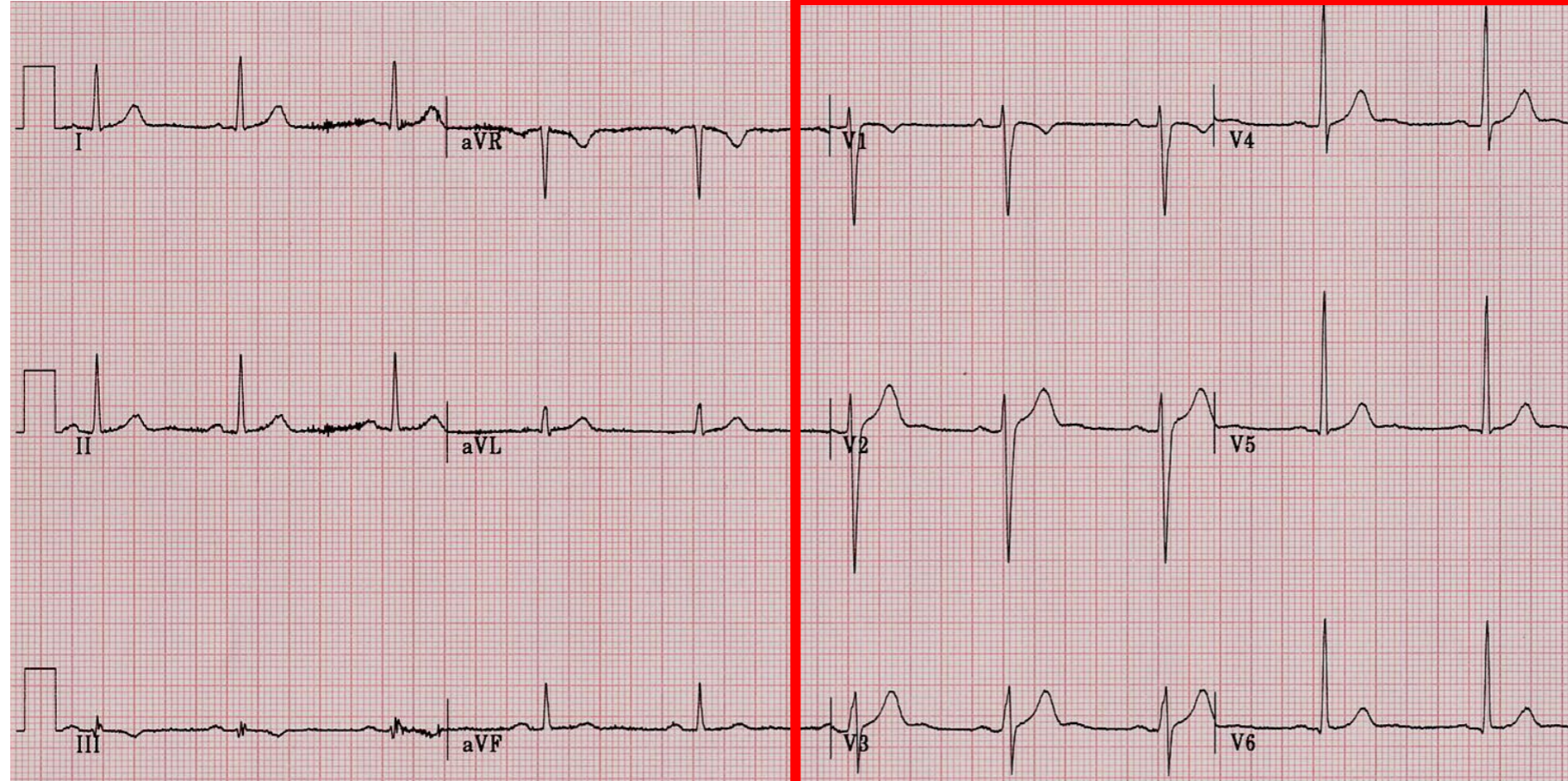
- ♥ heeft een kleine kans op kamerfibrilleren
- ♥ die niet goed te voorspellen is
- ♥ pompfunctie is en blijft eigenlijk goed



# Brugada syndrome

**Hoe stellen we de diagnose?**





# Special Report

## Brugada Syndrome Report of the Second Consensus Conference

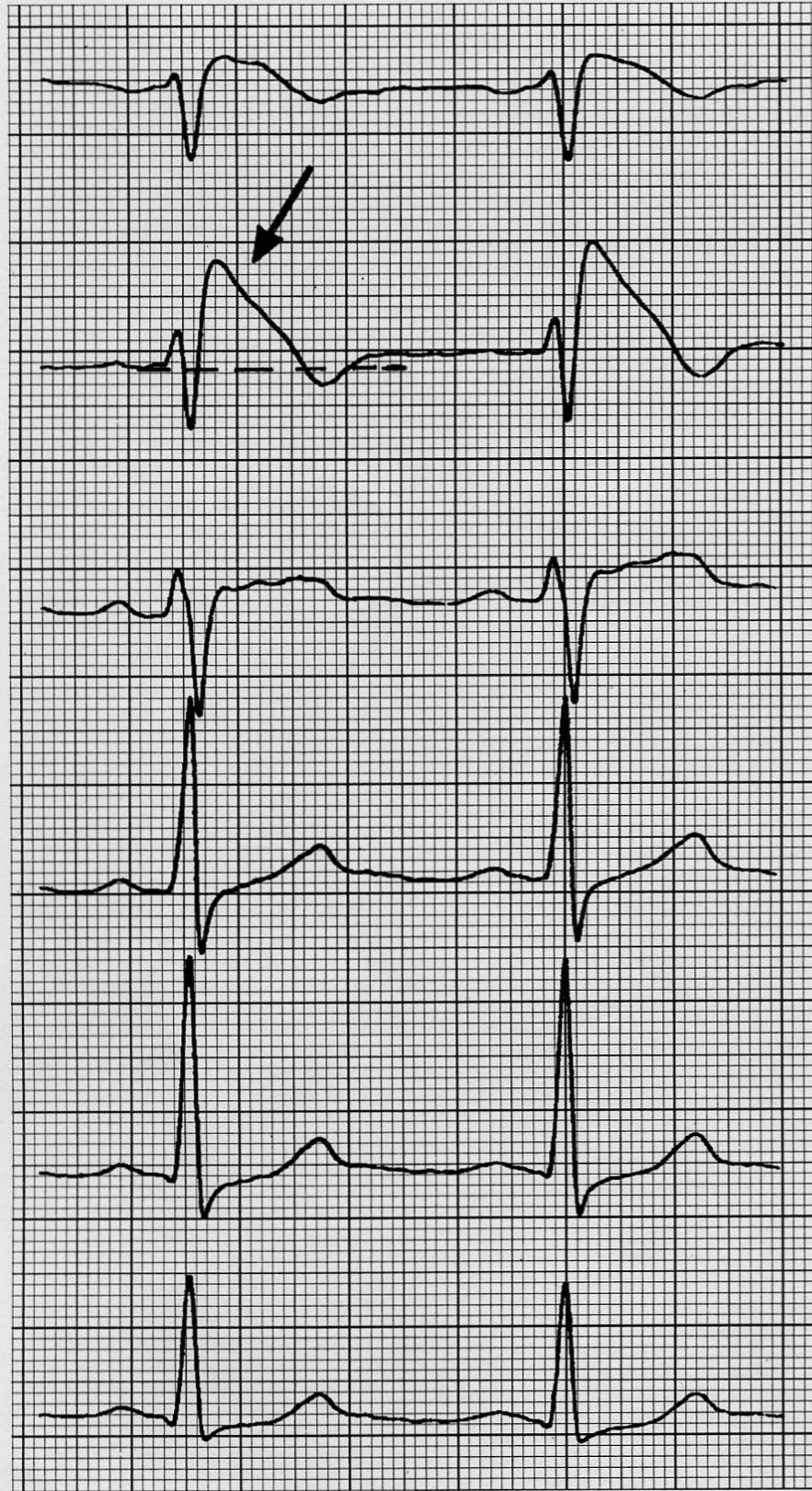
*Endorsed by the Heart Rhythm Society and the European Heart  
Rhythm Association*

Charles Antzelevitch, PhD; Pedro Brugada, MD, PhD; Martin Borggrefe, MD, PhD;  
Josep Brugada, MD; Ramon Brugada, MD; Domenico Corrado, MD, PhD; Ihor Gussak, MD, PhD;  
Herve LeMarec, MD; Koonlawee Nademanee, MD; Andres Ricardo Perez Riera, MD;  
Wataru Shimizu, MD, PhD; Eric Schulze-Bahr, MD; Hanno Tan, MD, PhD; Arthur Wilde, MD, PhD

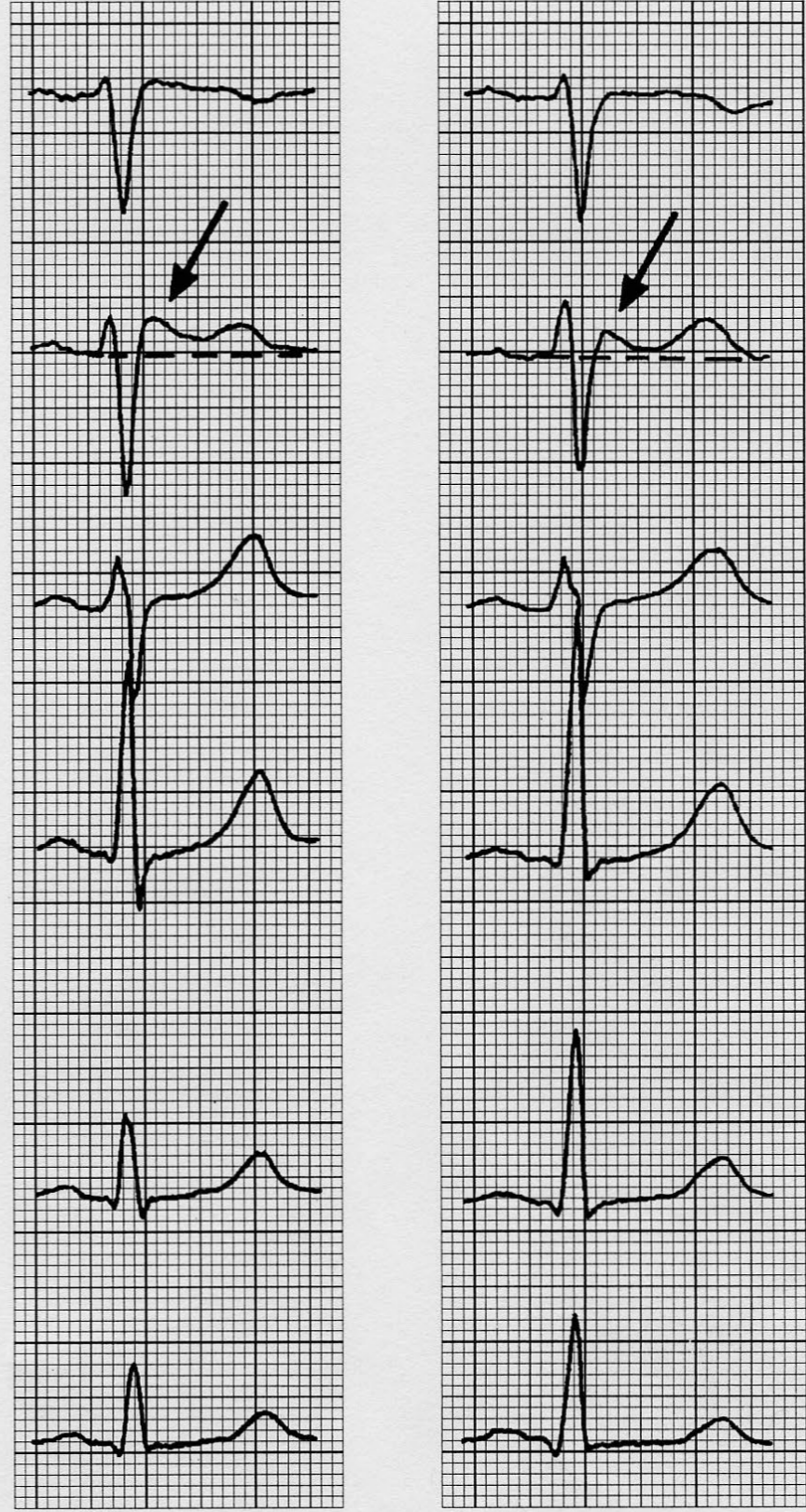
**Heart Rhythm 2005;2:429-440**



# Type 1



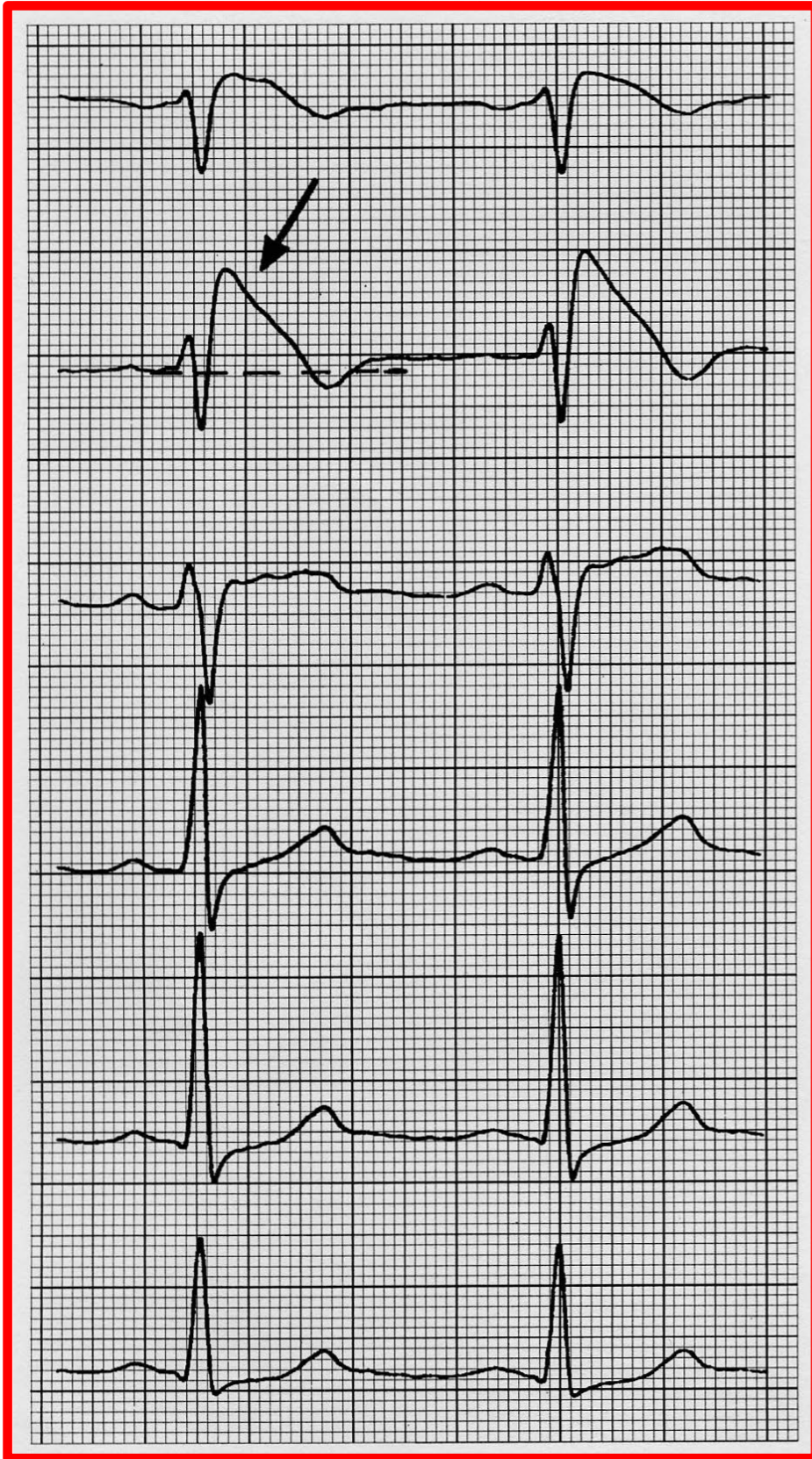
# Type 2 & 3



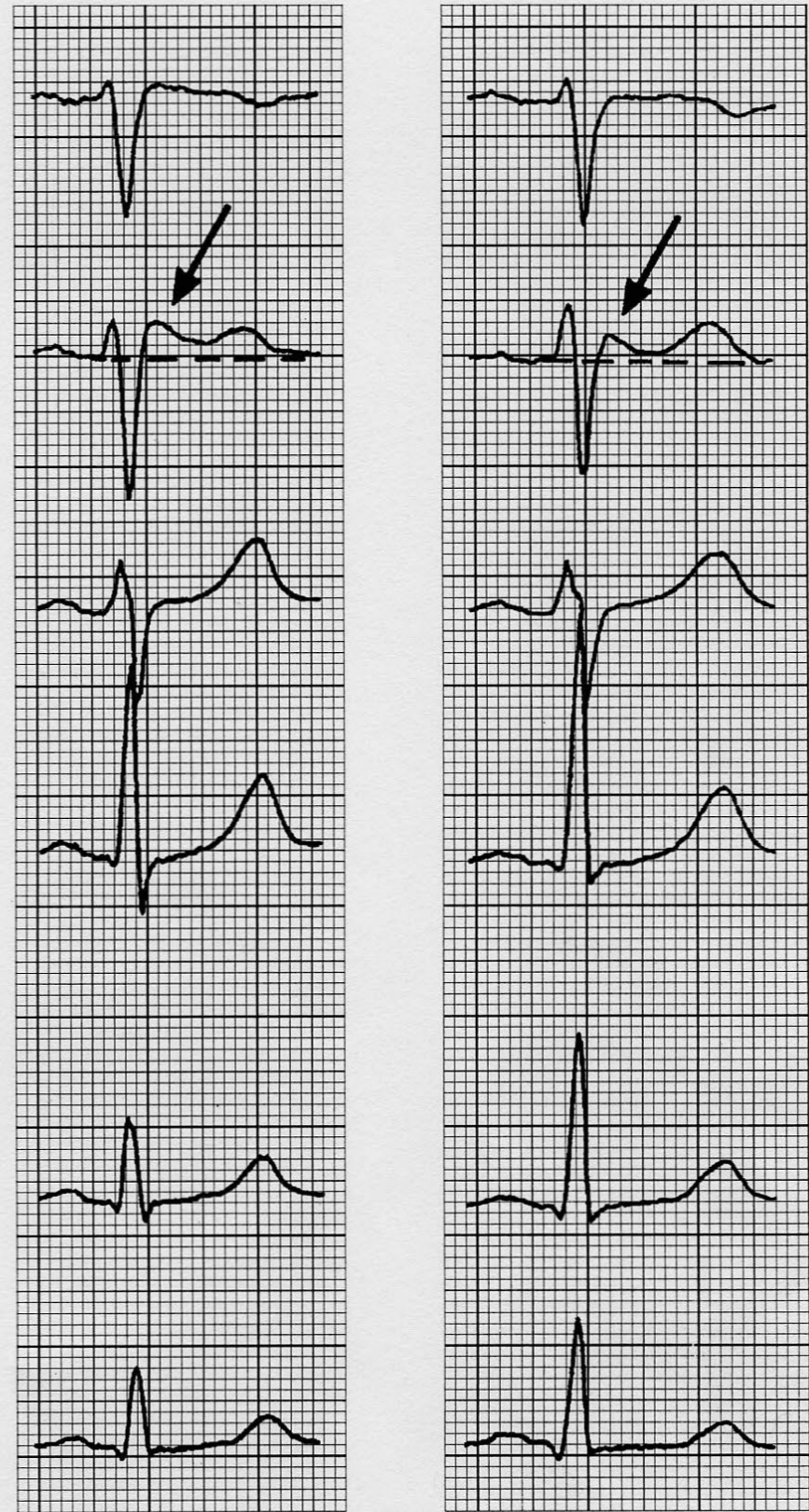
1 mV

500ms

# Type 1

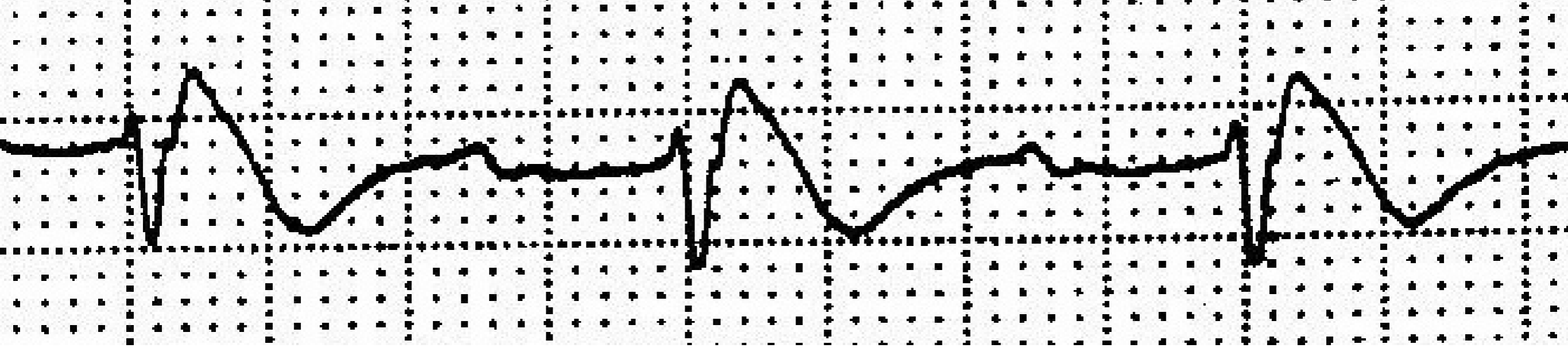


# Type 2 & 3



1 mV

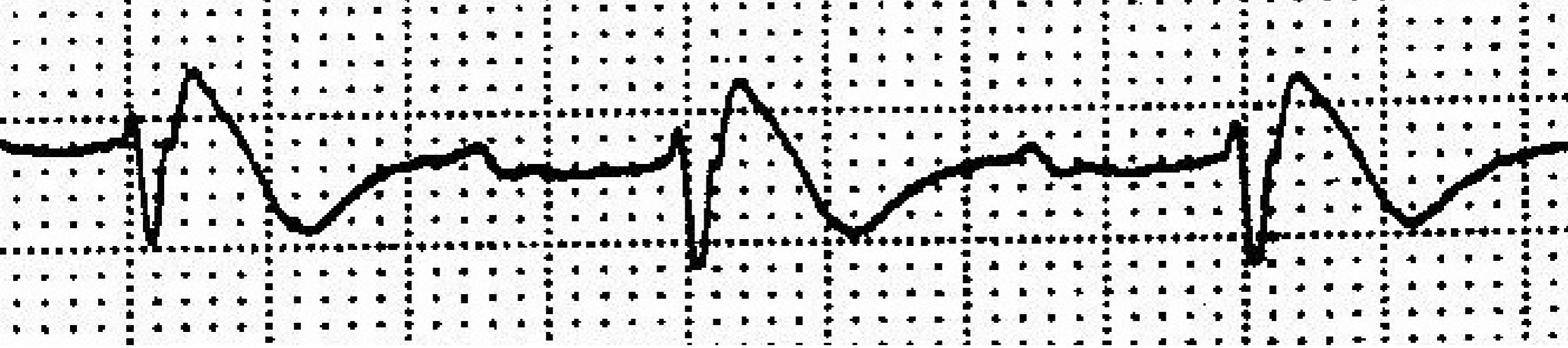
500ms



## De diagnose Brugada syndrome gesteld:

2005

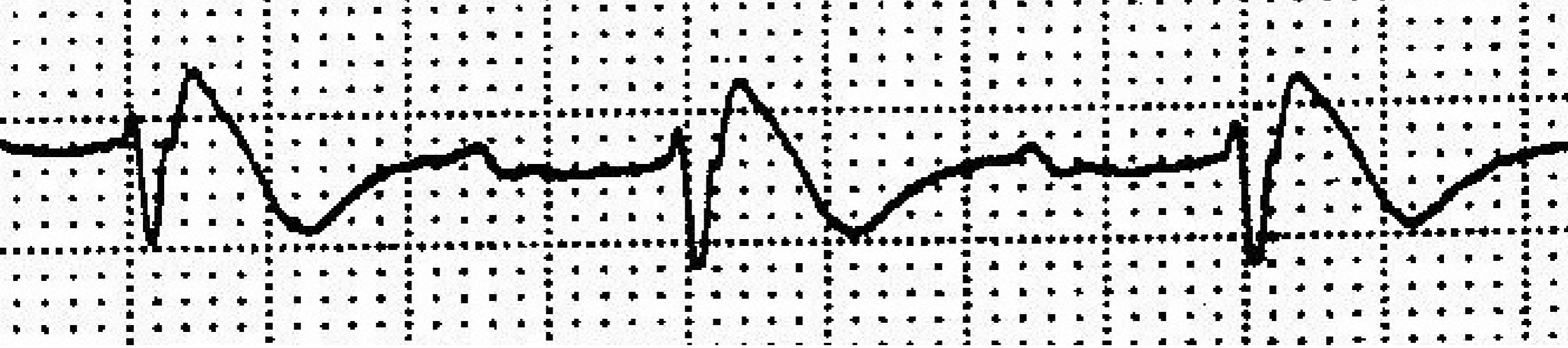
- ♥ Type 1 ECG ( $\pm$  medicatie) **en**
- ♥ gedocumenteerde ritmestoornissen
- ♥ Familie verhaal van jong plotseling overlijden
- ♥ Type 1 ECG in familieleden
- ♥ wegraking(en)
- ♥ ritmestoornissen opgewekt tijdens onderzoek



# De diagnose Brugada syndrome gesteld:

2013

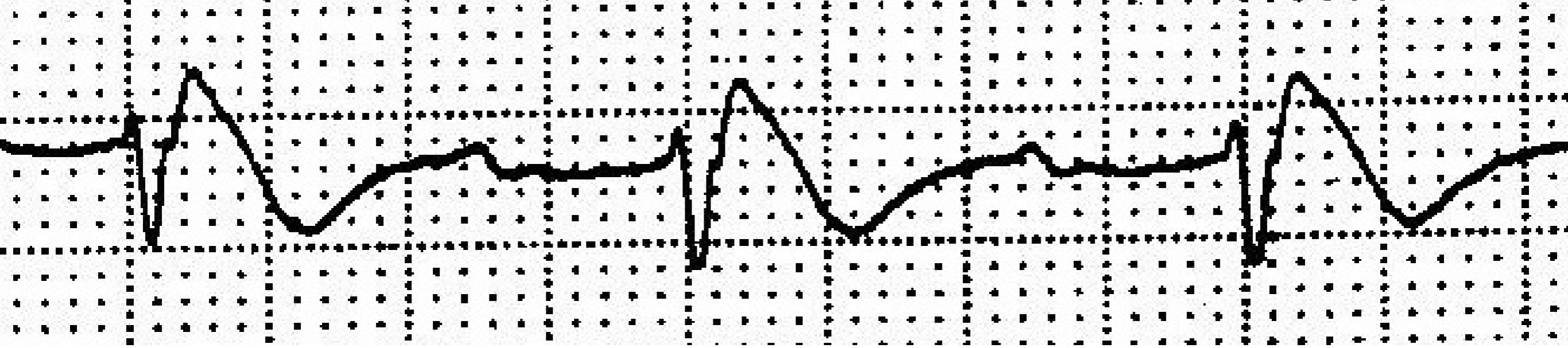
- ♥ Type 1 ECG ( $\pm$  medicatie)
- ♥ Zonder extra dingen



# De diagnose Brugada syndrome gesteld:

2016

♥ Type 1 ECG (zonder medicatie)

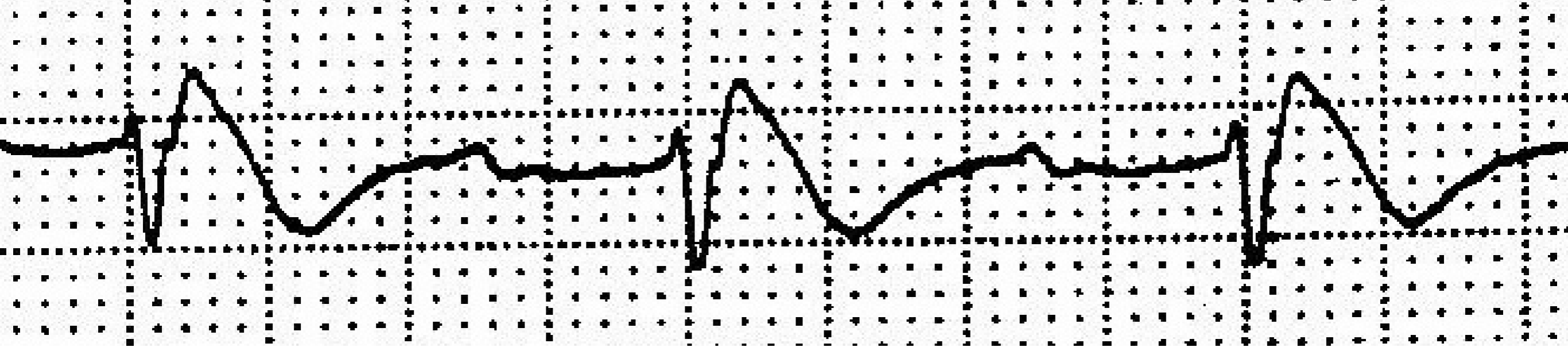


# De diagnose Brugada syndrome gesteld:

2016

- ♥ Type 1 ECG (door medicatie) **en**
- ♥ gedocumenteerde ritmestoornissen
- ♥ Familie verhaal van jong plotseling overlijden
- ♥ Type 1 ECG in familieleden
- ♥ wegraking(en)
- ♥ ritmestoornissen opgewekt tijdens onderzoek



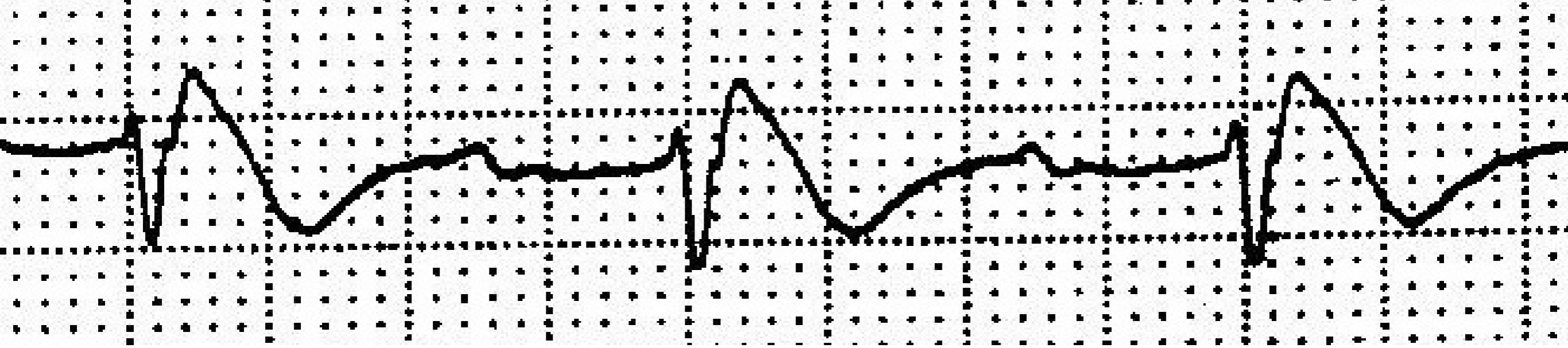


Waarom een verschil tussen Type 1 ECG  $\pm$  medicatie?

2016

Omdat gebleken is dat:

- ♥ 3-4% van alle mensen positief reageert
- ♥ bij bepaalde ziektebeelden % nog veel hoger is



Waarom een verschil tussen Type 1 ECG ± medicatie?

2016

En dus

- ♥ is de test **niet betrouwbaar genoeg** om aan te tonen dat iemand Brugada syndroom heeft
- ♥ de test is **wel betrouwbaar** om het onwaarschijnlijk te maken

# Shanghai 2016

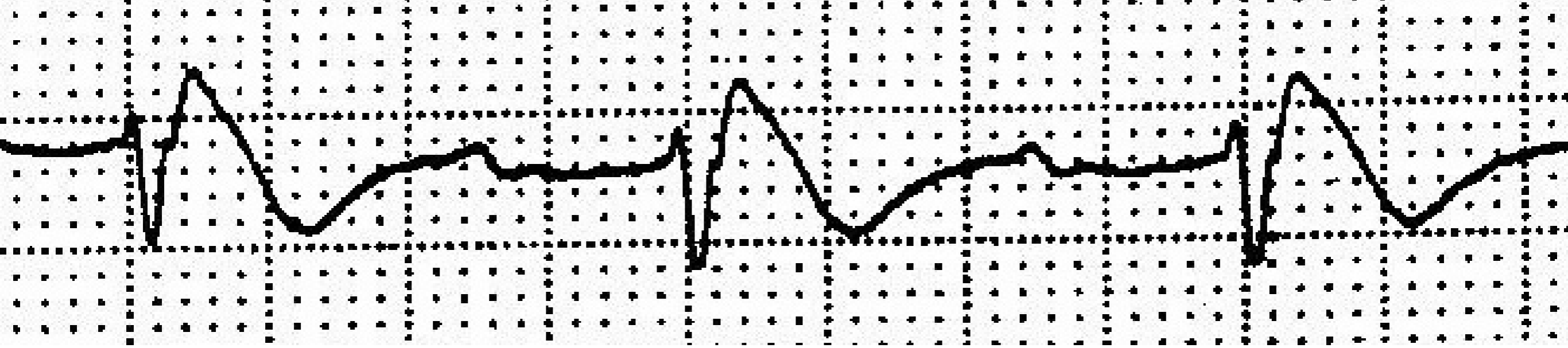
## J-Wave syndromes expert consensus conference report: Emerging concepts and gaps in knowledge

Charles Antzelevitch, PhD, FHRS,<sup>\*</sup> Gan-Xin Yan, MD, PhD,<sup>†</sup> Michael J. Ackerman, MD, PhD,<sup>‡</sup>  
Martin Borggrefe, MD,<sup>§</sup> Domenico Corrado, MD, PhD,<sup>¶</sup> Jihong Guo, MD,<sup>#</sup> Ihor Gussak, MD, PhD,<sup>\*\*</sup>  
Can Hasdemir, MD,<sup>††</sup> Minoru Horie, MD,<sup>‡‡</sup> Heikki Huikuri, MD,<sup>§§</sup> Changsheng Ma, MD,<sup>¶¶</sup>  
Hiroshi Morita, MD, PhD,<sup>##</sup> Gi-Byoung Nam, MD, PhD,<sup>\*\*\*</sup> Frederic Sacher, MD, PhD,<sup>†††</sup>  
Wataru Shimizu, MD, PhD,<sup>‡‡‡</sup> Sami Viskin, MD,<sup>§§§</sup> Arthur A.M. Wilde, MD, PhD, FHRS<sup>¶¶¶</sup>

**Heart Rhythm 2016, Europace 2017, J of Arrhythmia 2016**

**Shanghai criteria**





Waarom een verschil tussen Type 1 ECG ± medicatie?

2016

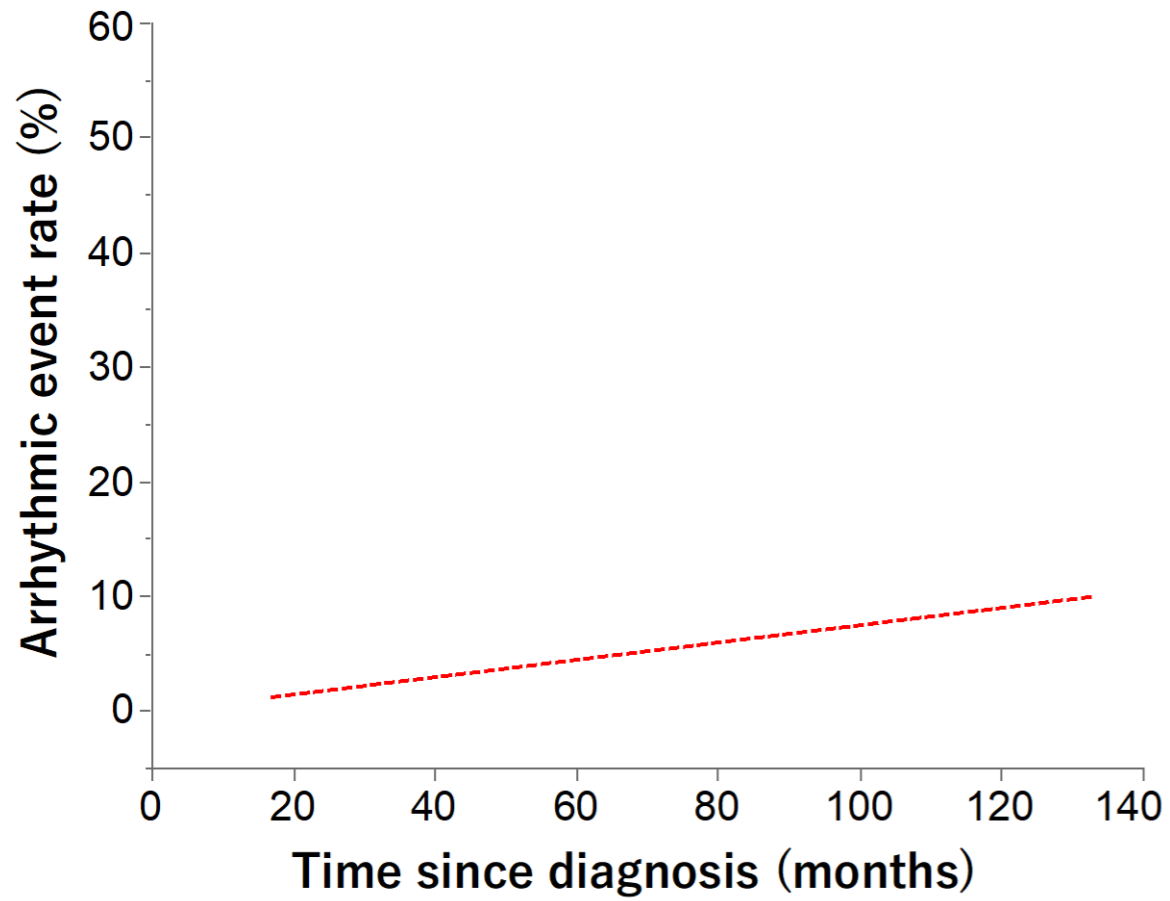
Omdat gebleken is dat:

- ♥ 3-4% van de mensen positief reageert
- ♥ bij bepaalde ziektebeelden % nog veel hoger is
- ♥ risico op ritmestoornissen wezenlijk anders

# Asymptomatic patients

- Spontaneous type 1
- Drug-induced type 1

A



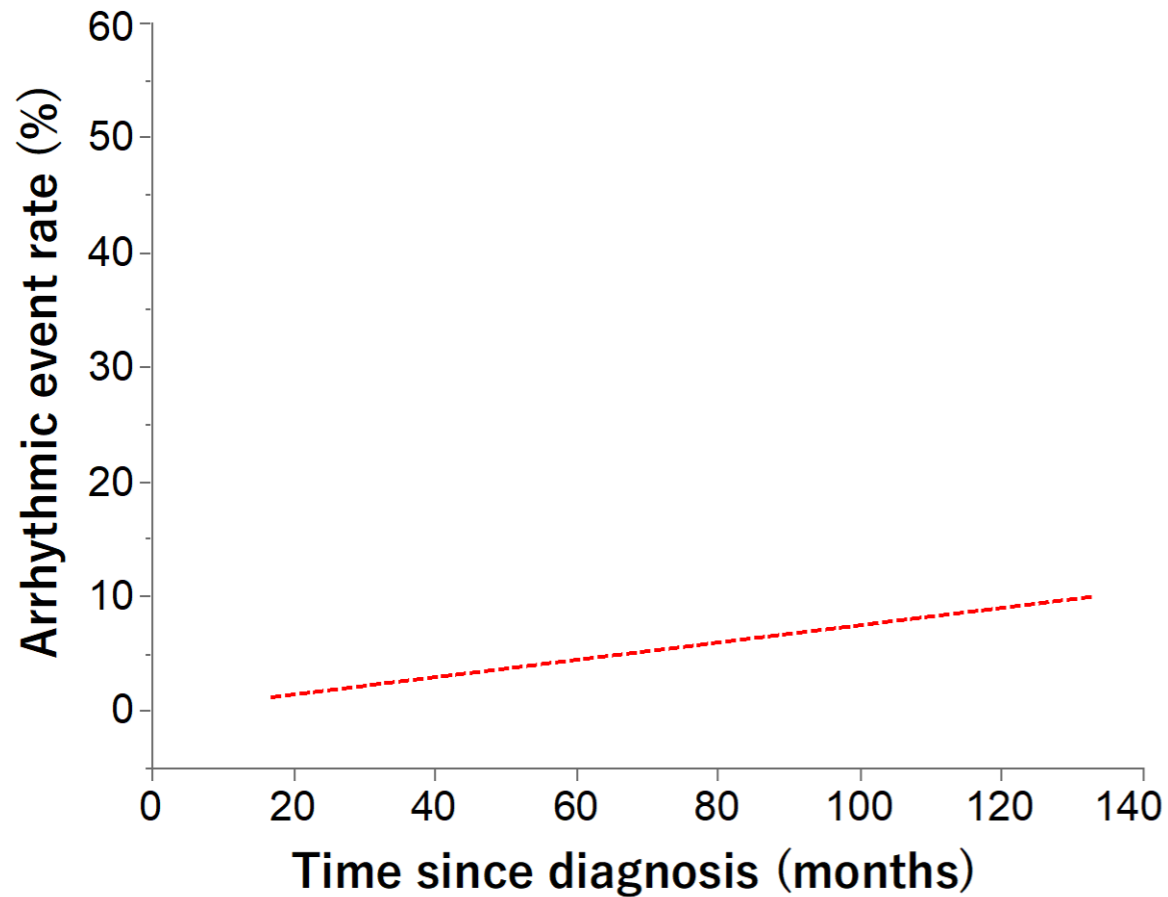
**0.29% vs 0.88% / year**

# Asymptomatic patients

# Symptomatic patients

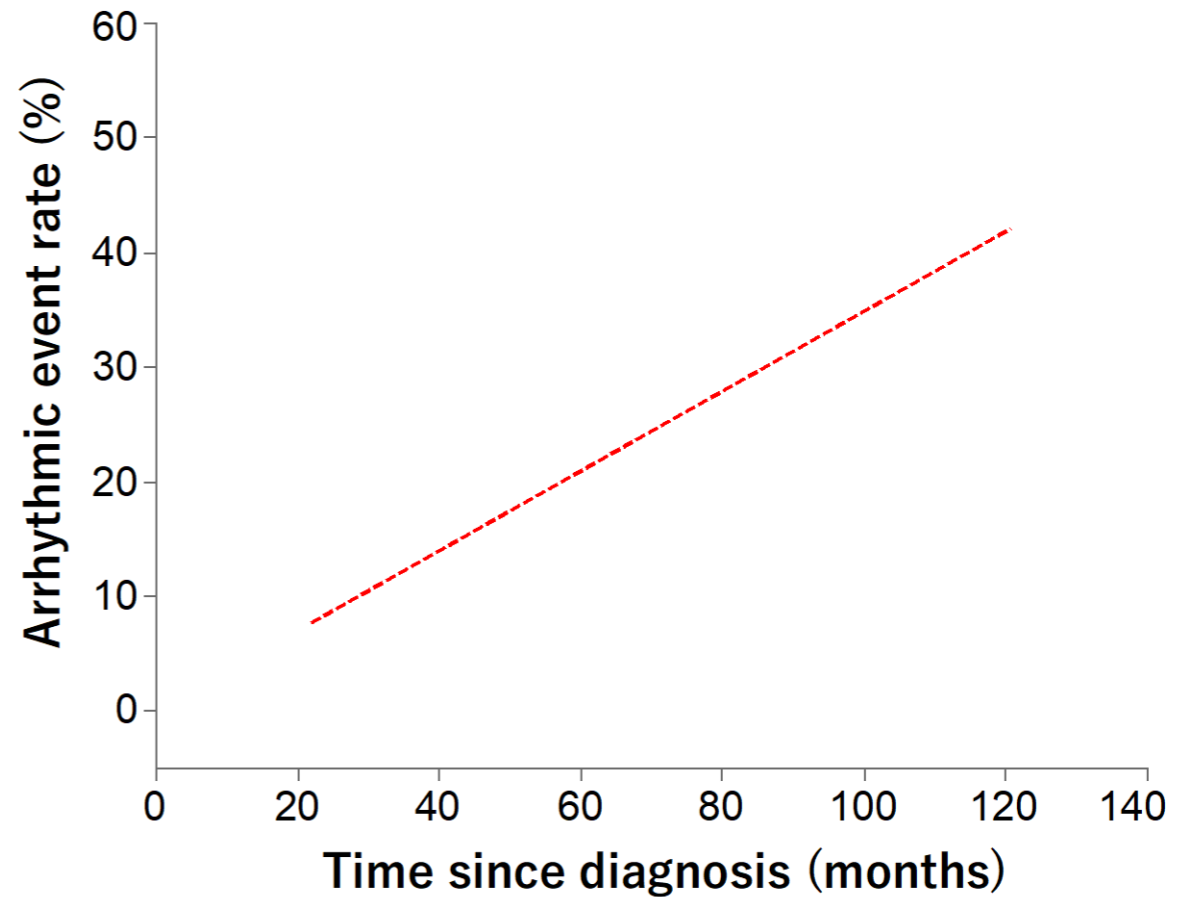
Spontaneous type 1  
Drug-induced type 1

### A



**0.29% vs 0.88% / year**

### B



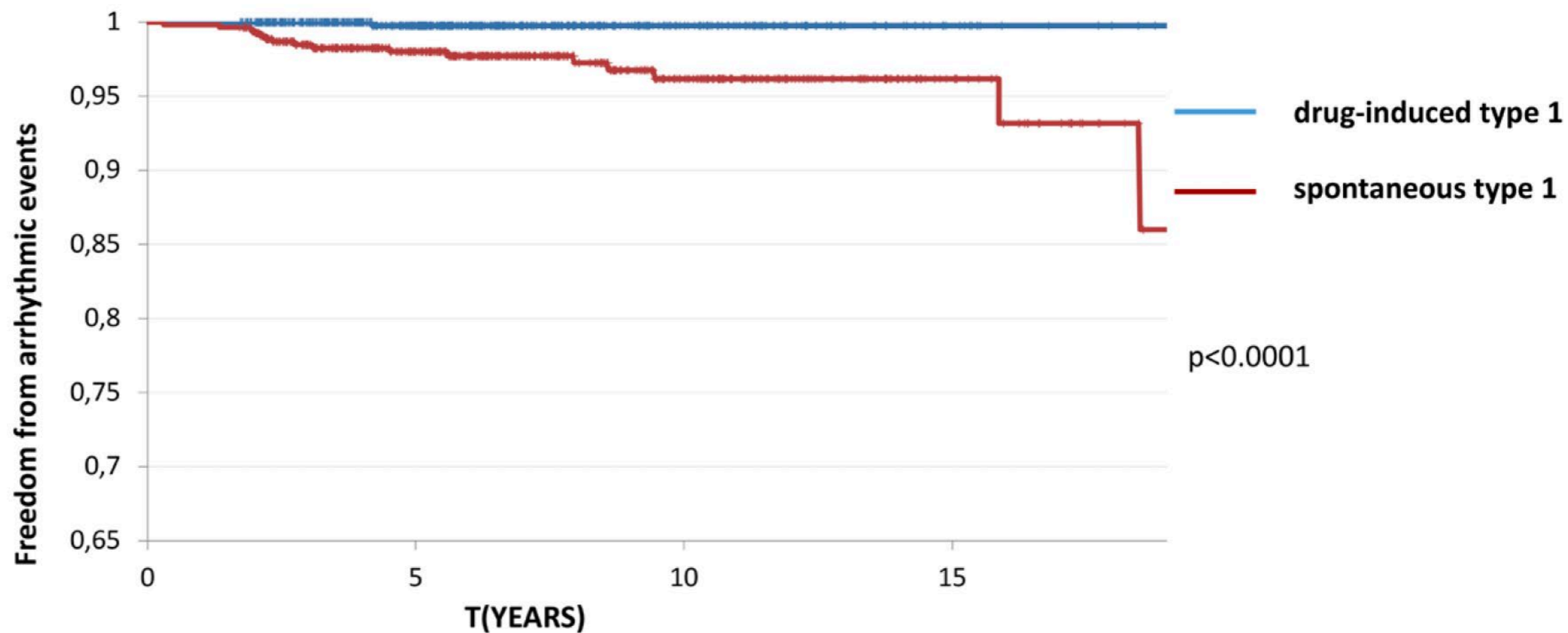
**1.34% vs 4.08% / year**

## ORIGINAL RESEARCH ARTICLE

## Asymptomatic Patients With Brugada ECG Pattern: Long-Term Prognosis From a Large Prospective Study

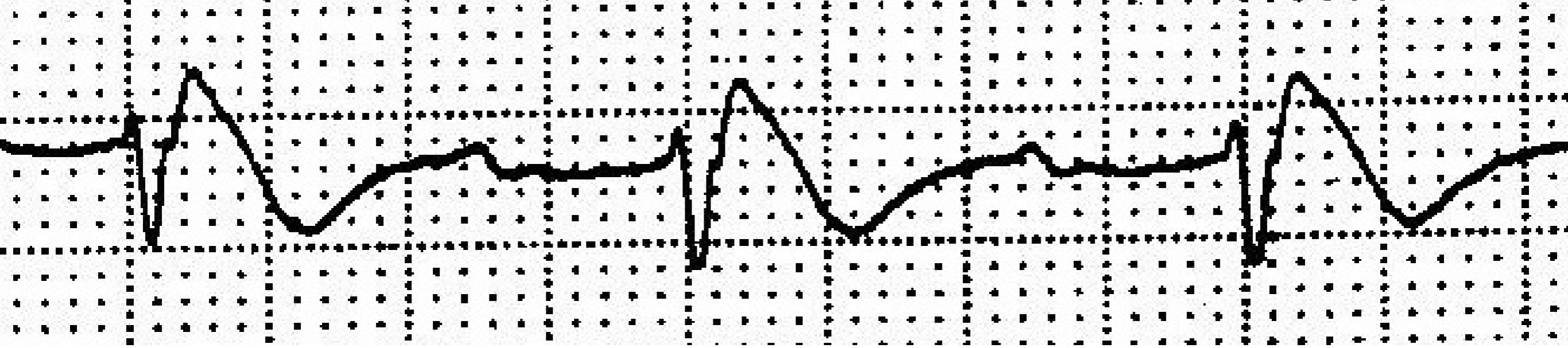
Fiorenzo Gaita<sup>1</sup>, MD\*; Natascia Cerrato<sup>1</sup>, MD\*; Carla Giustetto<sup>1</sup>, MD\*; Annamaria Martino, MD, PhD; Laura Bergamasco<sup>1</sup>, PhD; Michele Millesimo<sup>1</sup>, MD; Lorella Barbonaglia, MD; Paula Carvalho, MD; Domenico Caponi, MD; Andrea Saglietto<sup>1</sup>, MD; Giacomo Bonacchi<sup>1</sup>, MD; Francesca Bianchi, MD; Elisa Silvetti<sup>1</sup>, MD; Cinzia Crescenzi<sup>1</sup>, MD; Stefano Canestrelli, MD; Melissa De Maio, MD; Gaetano Maria De Ferrari, MD; Giuseppe Musumeci<sup>1</sup>, MD; Francesco Rametta, MD; Marco Scaglione, MD; Leonardo Calò<sup>1</sup>, MD

## Asymptomatic Brugada patients: spontaneous vs induced type 1 ECG



Number at risk

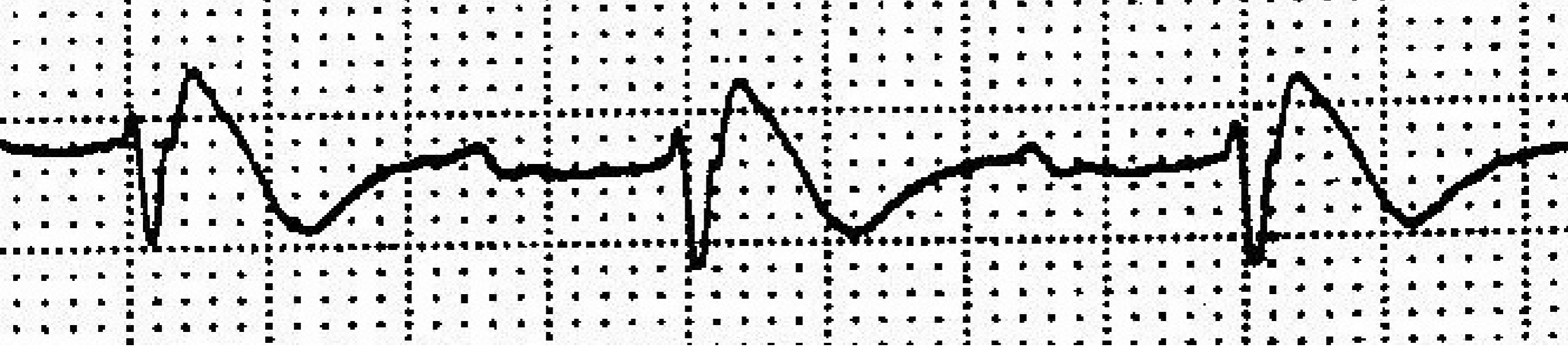
<b>induced type 1</b>	610	350	86	17
<b>spont. type 1</b>	539	379	211	43



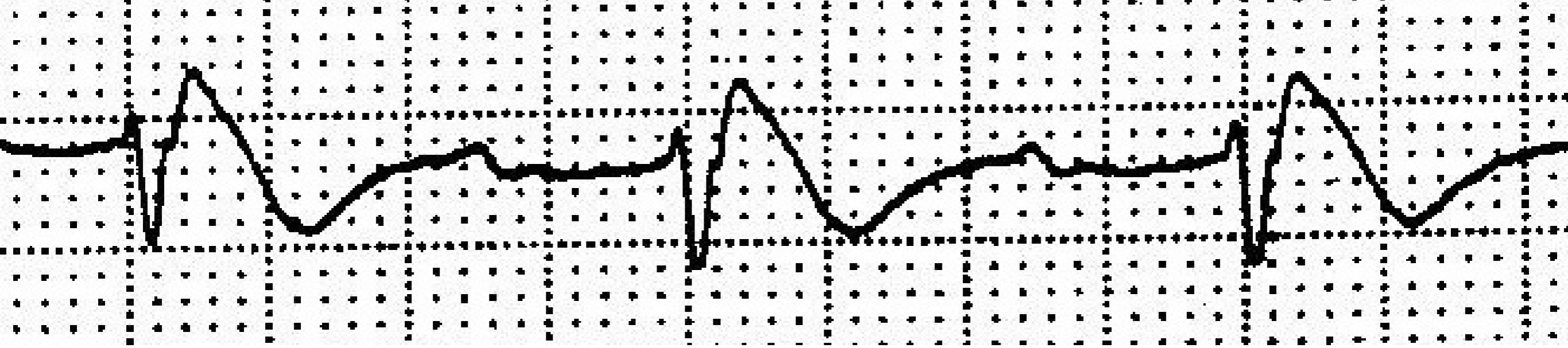
De Shanghai criteria betekenen dus dat de diagnose Brugada syndroom nu nog alleen gesteld wordt op basis van een spontaan type 1 ECG

Of als alleen de medicatie test positief in aanwezigheid van extra criteria





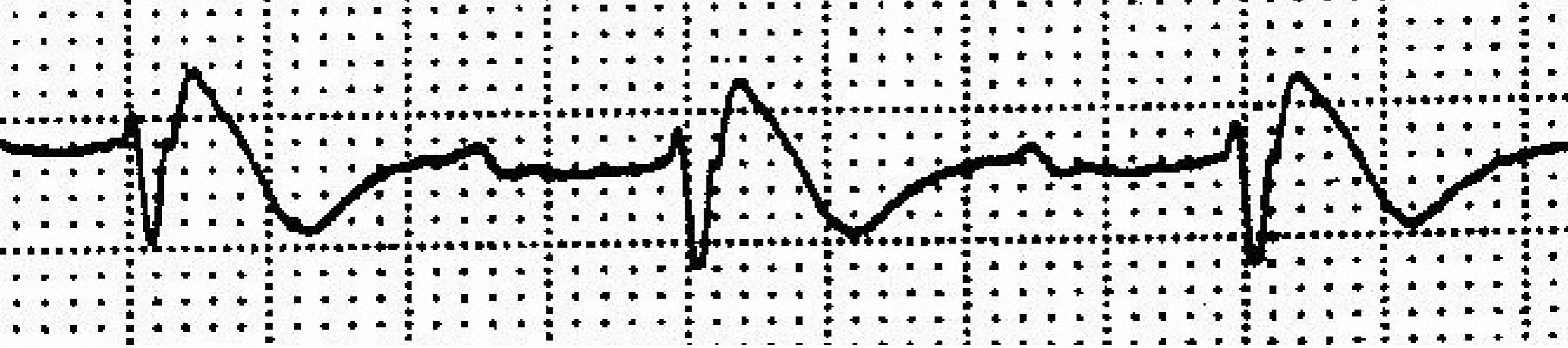
Al met al kan dit dus betekenen dat als bij U in het verleden de diagnose Brugada syndroom is gesteld op basis van alleen een positieve medicatie test daar nu anders over gedacht wordt.



Al met al kan dit dus betekenen dat als bij U in het verleden de diagnose Brugada syndroom is gesteld op basis van **alleen** een positieve medicatie test daar nu anders over gedacht wordt.

Dit is niet zo als:

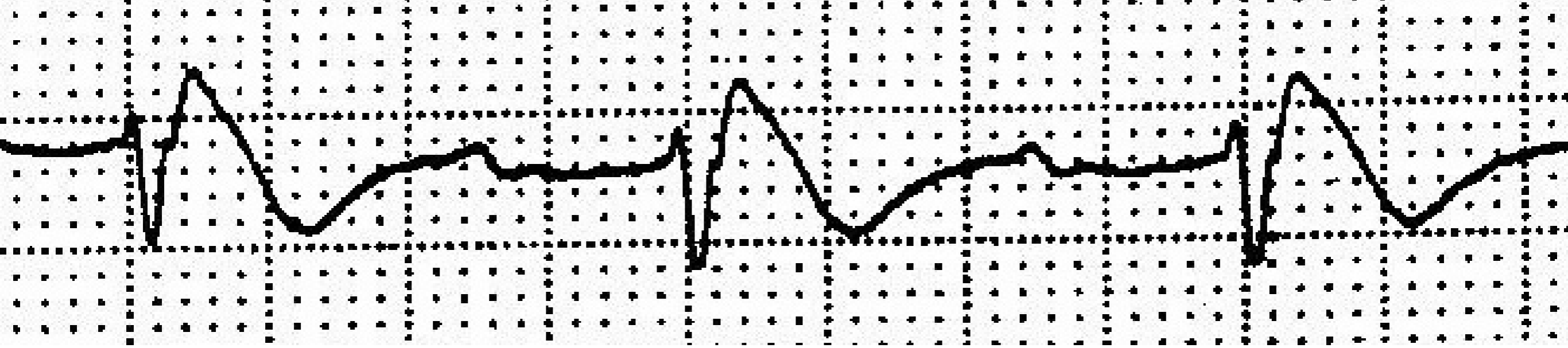
- ♥ als er ook sprake is van ernstige ritmestoornis.
- ♥ Familieleden met zeker BrS



Voor de behandeling maakt het echt uit:

- ♥ of het ECG spontaan aanwezig is
- ♥ of door medicatie opgewekt is

Actieve behandeling nodig (ICD, medicatie)?



Voor de te nemen voorzorgen maakt het niet echt uit:

- ♥ of het ECG spontaan aanwezig is
- ♥ of door medicatie opgewekt is

Bij koorts ECG en bepaalde medicatie vermijden

# Waarom is nog meer belangrijk een juiste diagnose te stellen

*Effect op iemand (recente Deense studie):*

Bij mensen, zonder depressie/angst stoornis, die de diagnose Brugada synd. kregen

- ♥ Ontwikkelde 16% een depressie/angst stoornis
- ♥ nog meer bij BrS patiënten met klachten

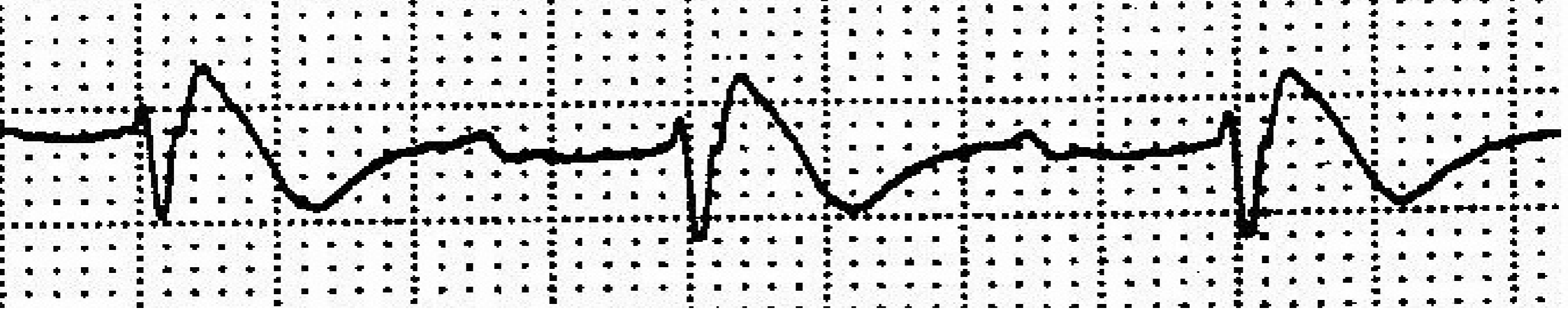
# Waarom is nog meer belangrijk een juiste diagnose te stellen

*Effect op iemand (recente Belgische studie):*

Bij mensen, zonder depressie/angst stoornis, die door medicatie test de diagnose BrS kregen

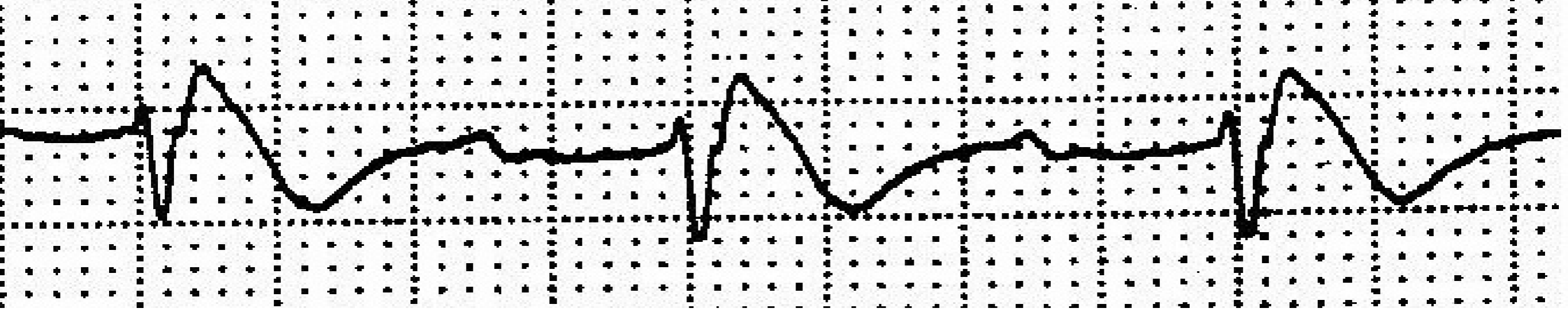
- ♥ toename angst/stress nivo's
- ♥ 49% tot een nivo waarbij verder onderzoek zou moeten
- ♥ 28% tot een nivo waarbij grote kans op echt probleem\*

\*: bijna 18% in de Belgische populatie



## Conclusies:

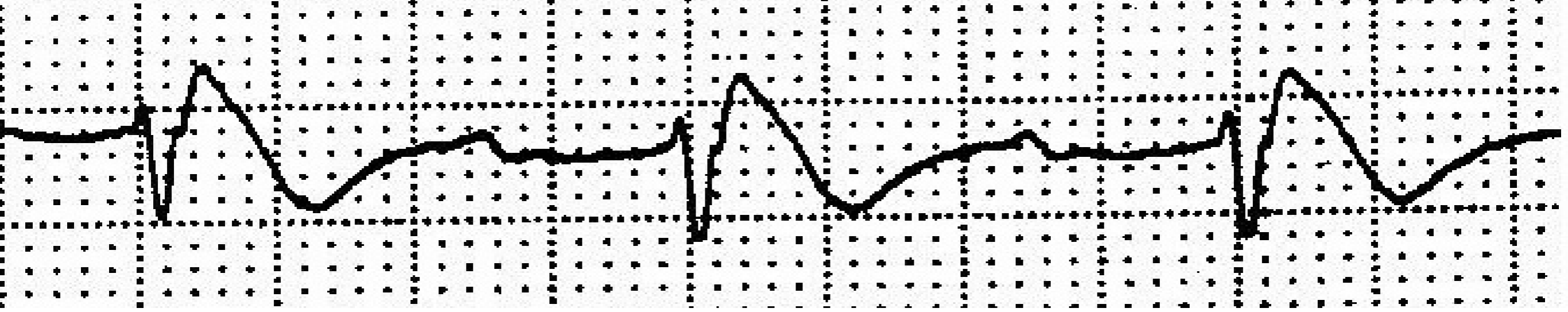
- ♥ Brugada syndroom is een ECG diagnose
- ♥ en behoeft een spontaan type 1 ECG
- ♥ preciese definitie is veranderd in de jaren



## Conclusies:

- ♥ Brugada syndroom is een ECG diagnose
- ♥ en behoeft een spontaan type 1 ECG
- ♥ Medicatie-geïnduceerd ECG  $\neq$  Brugada syndr.
- ♥ Medicatie-geïnd. ECG heeft echt laag risico
- ♥ en behoeft geen genetisch testen





## **Tenslotte uw aandacht voor 2 onderzoeken:**

- ♥ Biobank cardiogenetica (formulieren buiten)
- ♥ Vragenlijst voor pten met diagnose na test (vragenlijst in envelop)

- **Vragenlijst voor het berekenen van Shanghai score.**
- **Om meer duidelijkheid te krijgen over de diagnose Brugada syndroom bij patiënten die deze diagnose in het verleden hebben gekregen na een ajmaline provocatietest.**
- **Vragenlijst zit in de envelop.**
- **Wij vragen U deze in te vullen en in te leveren bij de tafel na de klapdeuren.**

**Naam:**  
**Geboortedatum:**

**Contactgegevens**  
-Email:  
-Telefoonnummer:

**Medische voorgeschiedenis:**

1. Bent u wel eens flauwgevallen sinds bij u de diagnose Brugada syndroom is gesteld?  
 Ja  
 Nee  
 Zo ja, was dit te herleiden naar een uitlokkende factor (zoals bloed zien, lang staan, warmte, emotie/stress)?  
|
2. Zijn er bij u gevaarlijke hartritmestoornissen gevonden sinds de diagnose Brugada syndroom is gesteld?  
 Ja  
 Nee  
 Zo ja, weet u welke soort hartritmestoornis?
3. Heeft u sinds de diagnose Brugada syndroom een pacemaker of ICD gekregen?  
 Ja, Pacemaker  
 Ja, ICD  
 Nee  
 In geval van ICD, heeft u een ICD-shock gehad?

**Toestemming en contact:**

4. Wanneer bent u voor het laatst onder controle geweest bij een cardioloog?
5. In welk centrum was dit?
6. Mogen wij de laatste brief opvragen bij dit ziekenhuis?  
 Ja  
 Nee

**Handtekening**

**Datum**

.....

.....

**Opmerkingen/Extra informatie: (voeg hier eventuele opmerkingen of extra informatie toe)**



**Thank you**





**Table 2** Proposed Shanghai Score System for diagnosis of Brugada syndrome

	Points
I. ECG (12-Lead/Ambulatory)	
A. Spontaneous type 1 Brugada ECG pattern at nominal or high leads	3.5
B. Fever-induced type 1 Brugada ECG pattern at nominal or high leads	3
C. Type 2 or 3 Brugada ECG pattern that converts with provocative drug challenge	2
<i>*Only award points once for highest score within this category. One item from this category must apply.</i>	
II. Clinical History*	
A. Unexplained cardiac arrest or documented VF/polymorphic VT	3
B. Nocturnal agonal respirations	2
C. Suspected arrhythmic syncope	2
D. Syncope of unclear mechanism/unclear etiology	1
E. Atrial flutter/fibrillation in patients <30 years without alternative etiology	0.5
<i>*Only award points once for highest score within this category.</i>	
III. Family History	
A. First- or second-degree relative with definite BrS	2
B. Suspicious SCD (fever, nocturnal, Brugada aggravating drugs) in a first- or second-degree relative	1
C. Unexplained SCD <45 years in first- or second-degree relative with negative autopsy	0.5
<i>*Only award points once for highest score within this category.</i>	
IV. Genetic Test Result	
A. Probable pathogenic mutation in BrS susceptibility gene	0.5
<b>Score (requires at least 1 ECG finding)</b>	
≥3.5 points: Probable/definite BrS	
2–3 points: Possible BrS	
<2 points: Nondiagnostic	

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**3.5 pts**



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A. First- or second-degree relative with definite BrS	2
B. Suspicious SCD (fever, nocturnal, Brugada aggravating drugs) in a first- or second-degree relative	1
C. Unexplained SCD <45 years in first- or second-degree relative with negative autopsy	0.5
<i>*Only award points once for highest score within this category.</i>	
IV. Genetic Test Result	
A. Probable pathogenic mutation in BrS susceptibility gene	0.5
<b>Score (requires at least 1 ECG finding)</b>	
≥3.5 points: Probable/definite BrS	
2–3 points: Possible BrS	
<2 points: Nondiagnostic	



**2 pts**

**Table 2** Proposed Shanghai Score System for diagnosis of Brugada syndrome

	Points
I. ECG (12-Lead/Ambulatory)	
A. Spontaneous type 1 Brugada ECG pattern at nominal or high leads	3.5
B. Fever-induced type 1 Brugada ECG pattern at nominal or high leads	3
C. Type 2 or 3 Brugada ECG pattern that converts with provocative drug challenge	2
<i>*Only award points once for highest score within this category. One item from this category must apply.</i>	
II. Clinical History*	
A. Unexplained cardiac arrest or documented VF/polymorphic VT	3
B. Nocturnal agonal respirations	2
C. Suspected arrhythmic syncope	2
D. Syncope of unclear mechanism/unclear etiology	1
E. Atrial flutter/fibrillation in patients <30 years without alternative etiology	0.5
<i>*Only award points once for highest score within this category.</i>	
III. Family History	
A. First- or second-degree relative with definite BrS	2
B. Suspicious SCD (fever, nocturnal, Brugada aggravating drugs) in a first- or second-degree relative	1
C. Unexplained SCD <45 years in first- or second-degree relative with negative autopsy	0.5
<i>*Only award points once for highest score within this category.</i>	
IV. Genetic Test Result	
A. Probable pathogenic mutation in BrS susceptibility gene	0.5
<b>Score (requires at least 1 ECG finding)</b>	
≥3.5 points: Probable/definite BrS	
2–3 points: Possible BrS	
<2 points: Nondiagnostic	



**5 pts**

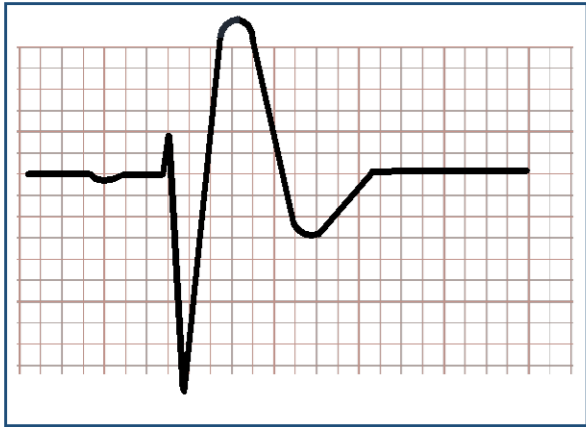
# Use, misuse, and pitfalls of the drug challenge test in the diagnosis of the Brugada syndrome

**Arthur A.M. Wilde** <sup>1,2\*</sup>, **Ahmad S. Amin**<sup>1,2</sup>, **Hiroshi Morita** <sup>3,4</sup>,  
**and Rafik Tadros** <sup>5</sup>

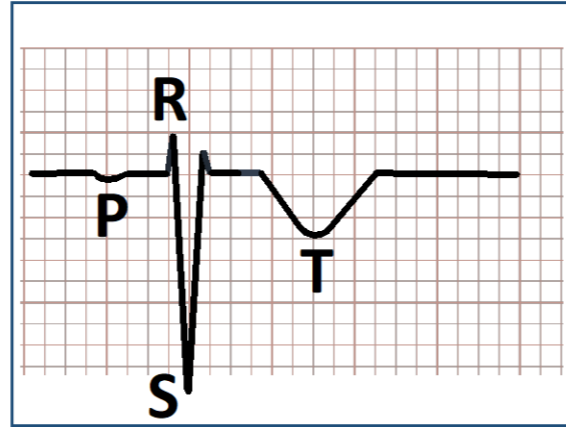
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**Spontaneous**



**Lead V<sub>1</sub> baseline**



**Brugada syndrome**



**All-comers:  
2,3 - 2,9%  
per year**

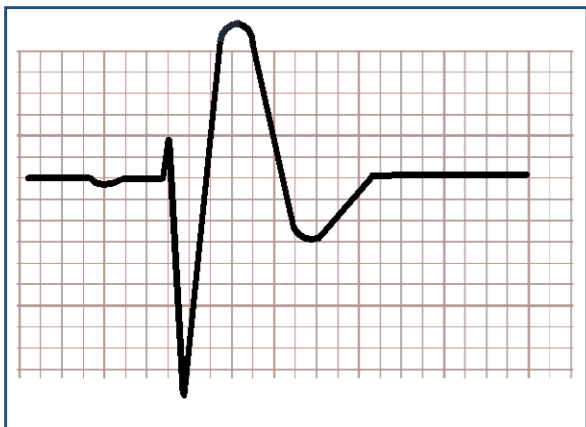
**Asymptomatic:  
0,5 - 1,2%  
per year**

**ECG**

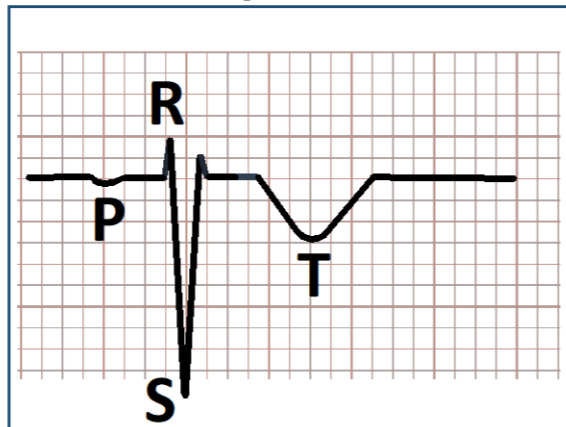
**CONDITION**

**RISK OF LETHAL  
ARRHYTHMIC EVENTS**

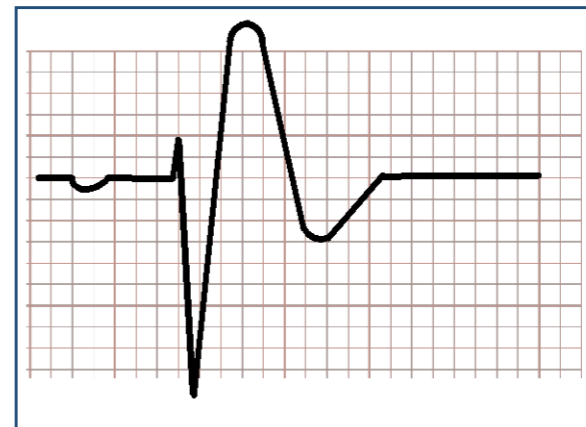
## Spontaneous



## Lead V<sub>1</sub> baseline



## SCBT



ECG

Brugada syndrome

Reduced conduction reserve in RVOT  
*(heterogeneous)*

CONDITION

All-comers:  
2,3 - 2,9%  
per year

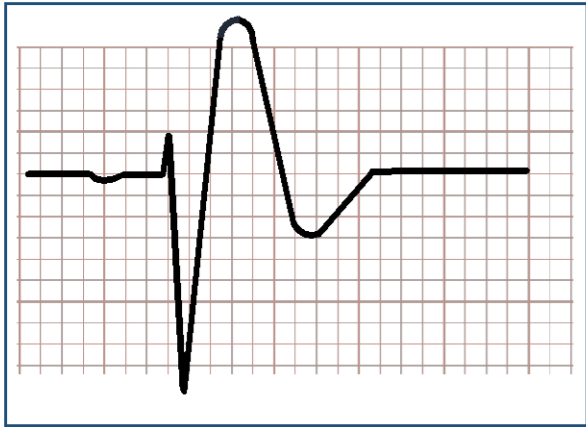
All-comers:  
1,07 - 1,22%  
per year

Asymptomatic:  
0,5 - 1,2%  
per year

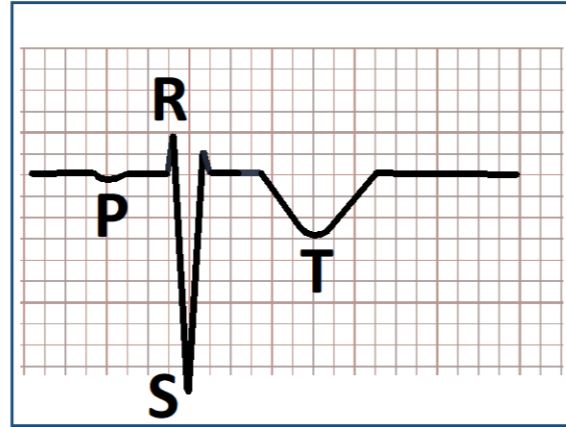
Asymptomatic:  
0 - 0,4%  
per year

RISK OF LETHAL  
ARRHYTHMIC EVENTS

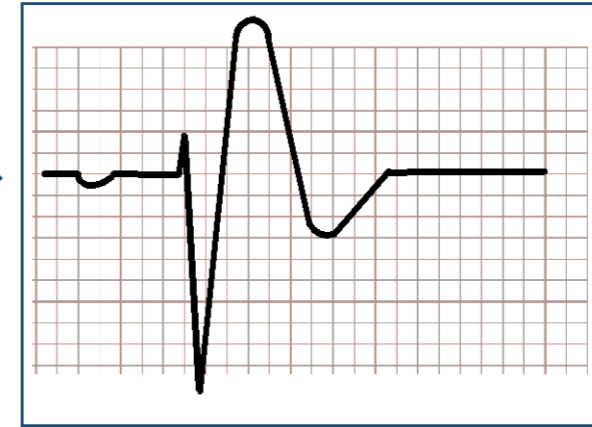
## Spontaneous



## Lead V<sub>1</sub> baseline



## SCBT



ECG

**Brugada syndrome**

**% with Brugada syndrome\* depends on indication for SCBT\*\***

**Reduced conduction reserve in RVOT**  
*(heterogeneous)*

**CONDITION**

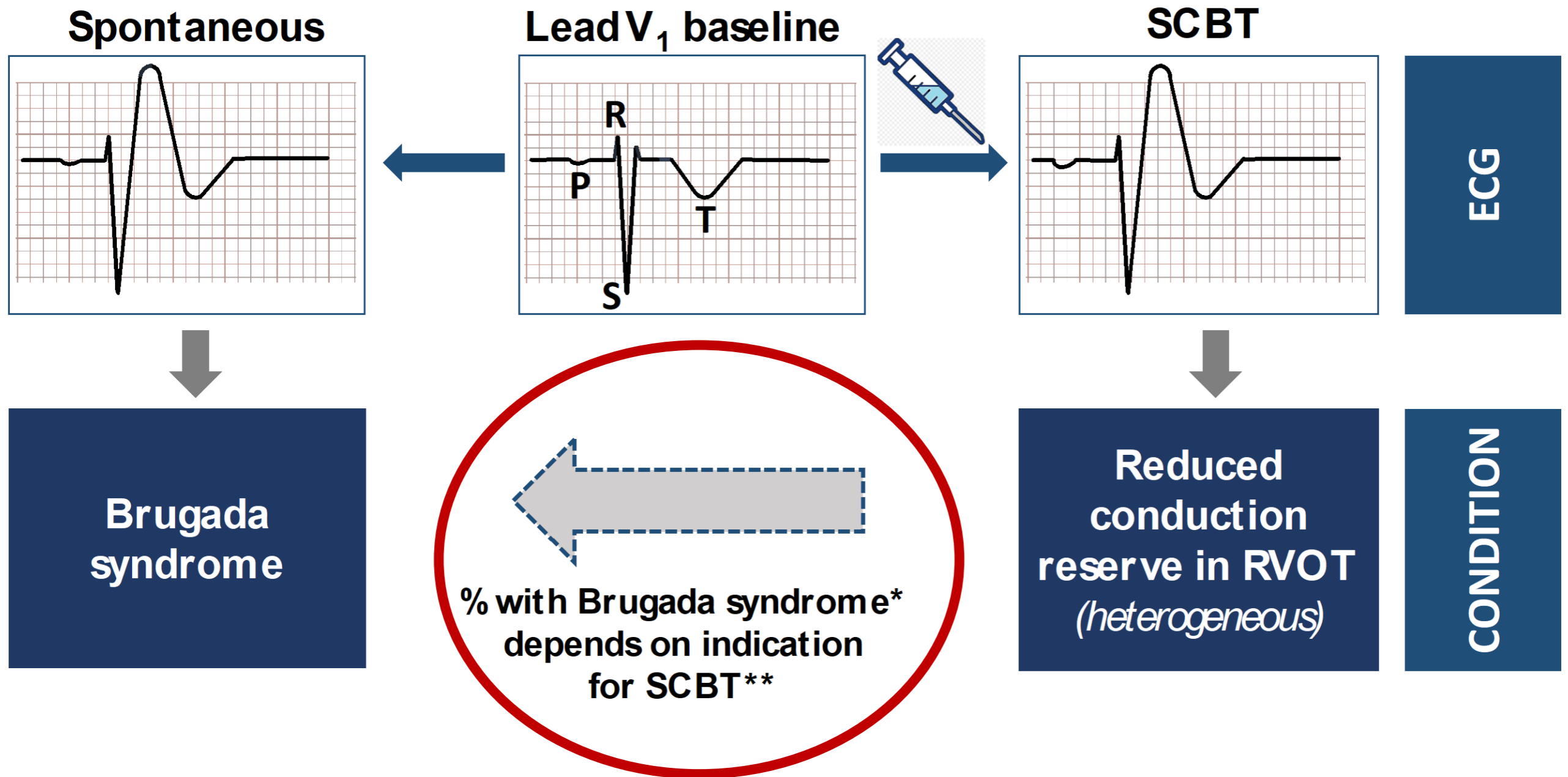
**All-comers:  
2,3 - 2,9%  
per year**

**All-comers:  
1,07 - 1,22%  
per year**

**Asymptomatic:  
0,5 - 1,2%  
per year**

**Asymptomatic:  
0 - 0,4%  
per year**

**RISK OF LETHAL  
ARRHYTHMIC EVENTS**



## Indication for SCBT, % with BrS:

- ♥ Fam member BrS pt:  $\pm \geq 95\%^*$
- ♥ random pt:  $< 0.2\%?$  (type 1&2)\*\*

\*Population tested by Pappone?, \*\*: overall, large geographic variation





