

Brugada syndrome

PLACE 2022

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NO DISCLOSURES FOR THIS PRESENTATION

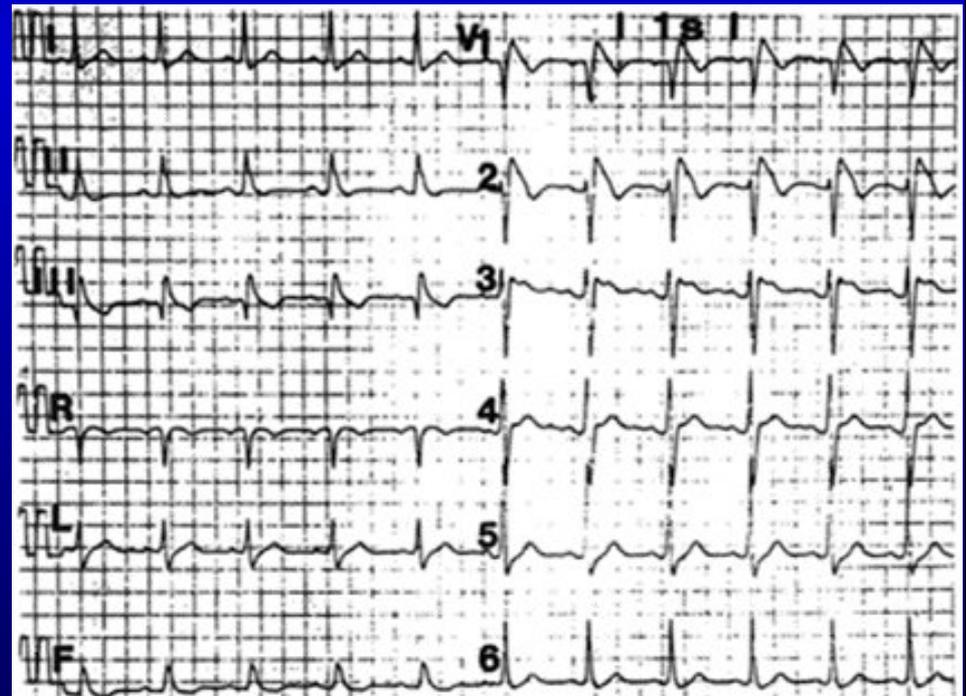
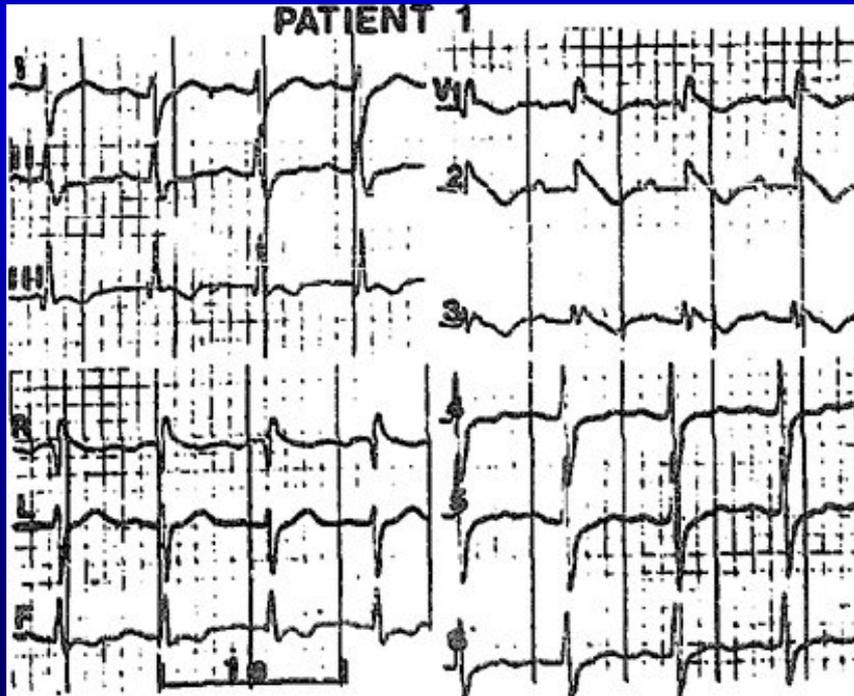
Right Bundle Branch Block, Persistent ST Segment Elevation and Sudden Cardiac Death: A Distinct Clinical and Electrocardiographic Syndrome

A Multicenter Report

PEDRO BRUGADA, MD, JOSEP BRUGADA, MD*†

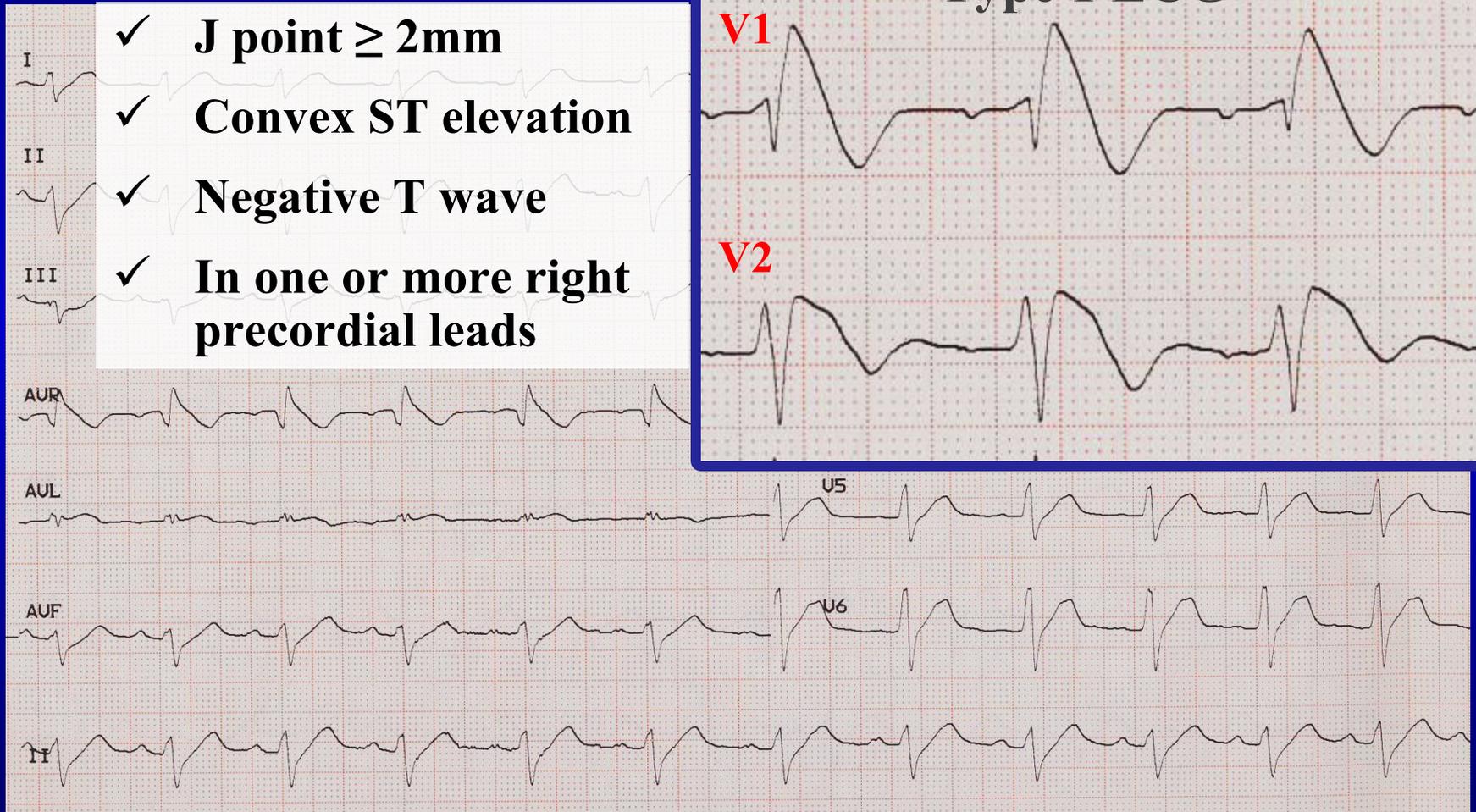
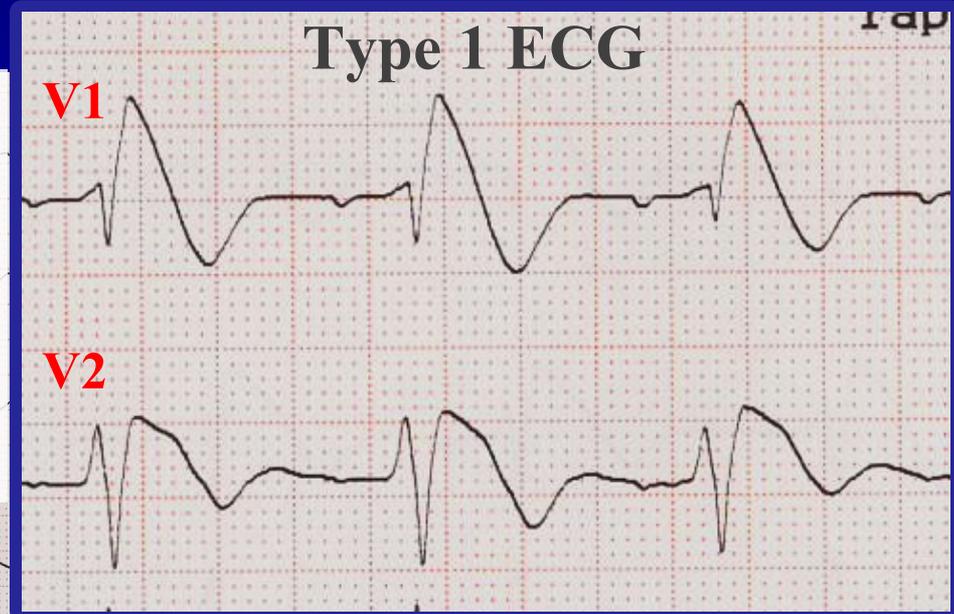
Aalst, Belgium and Barcelona, Spain

1992



Brugada syndrome: type 1 pattern = diagnostic

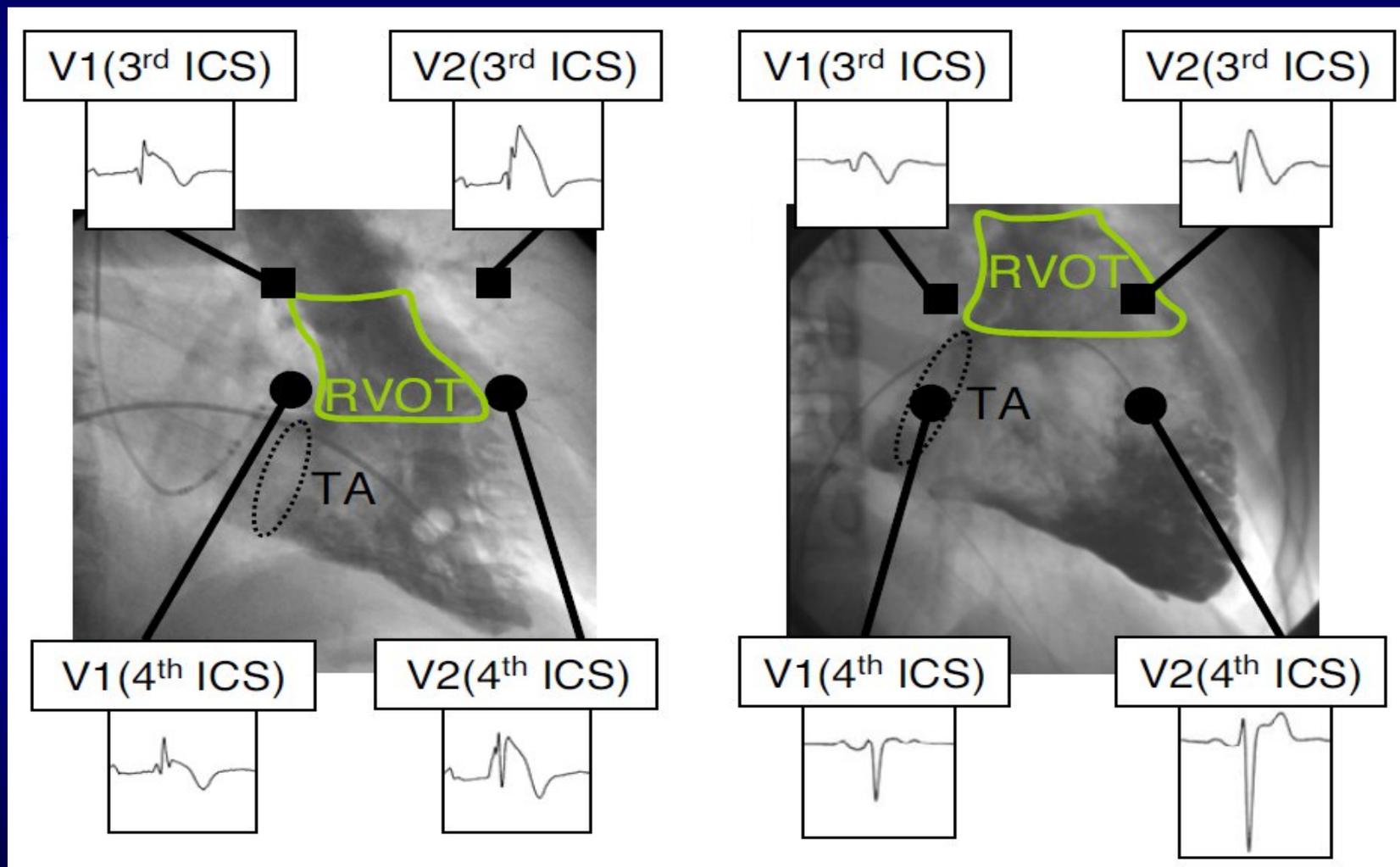
- ✓ J point $\geq 2\text{mm}$
- ✓ Convex ST elevation
- ✓ Negative T wave
- ✓ In one or more right precordial leads



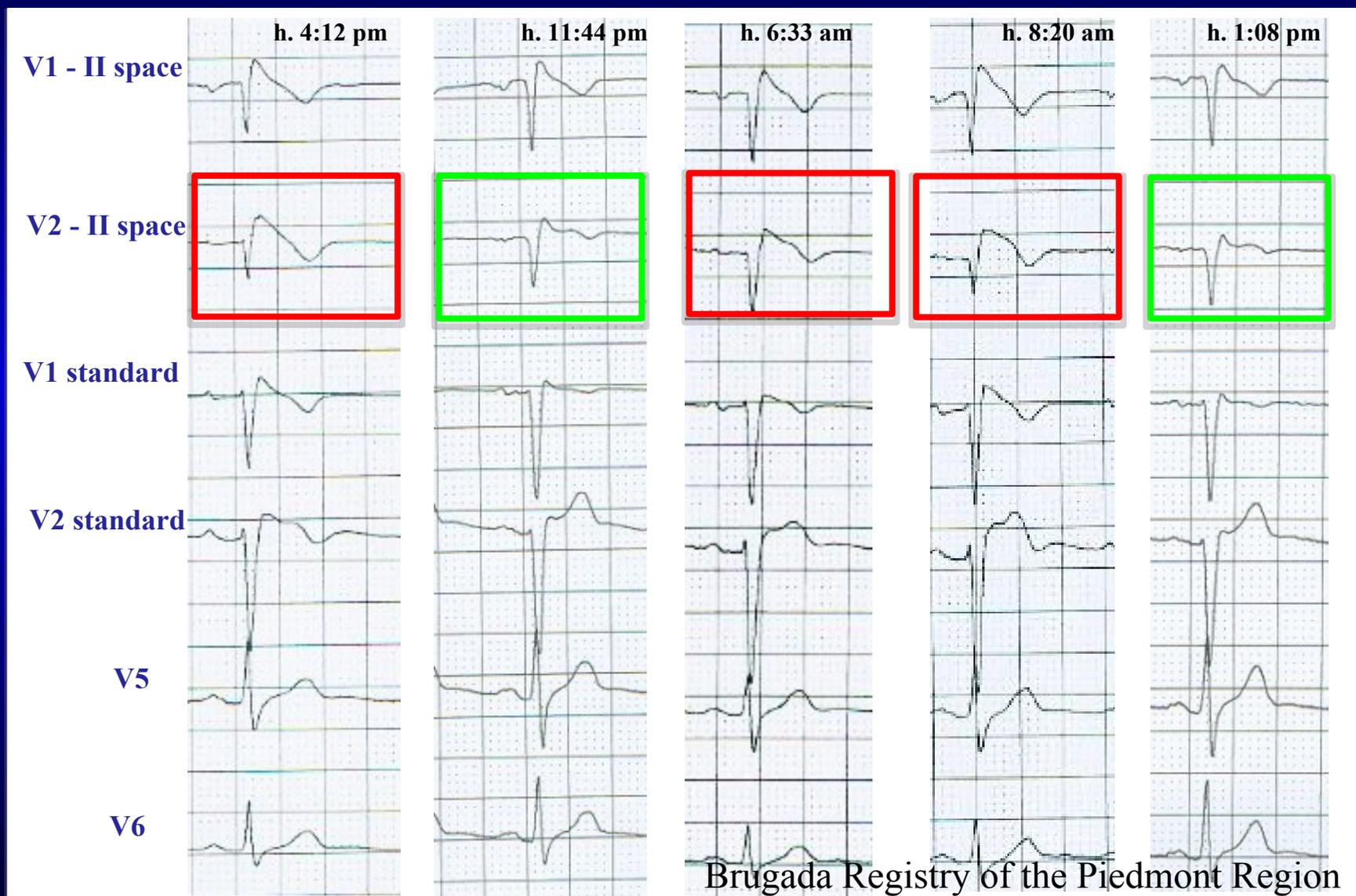
Brugada syndrome: suspicious patterns



Correlazione tra posizione del tratto di efflusso dal Vdx (RVOT) ed elettrodi esploranti



Intermittent spontaneous type 1 Brugada pattern at 12-lead 24-hour Holter monitoring

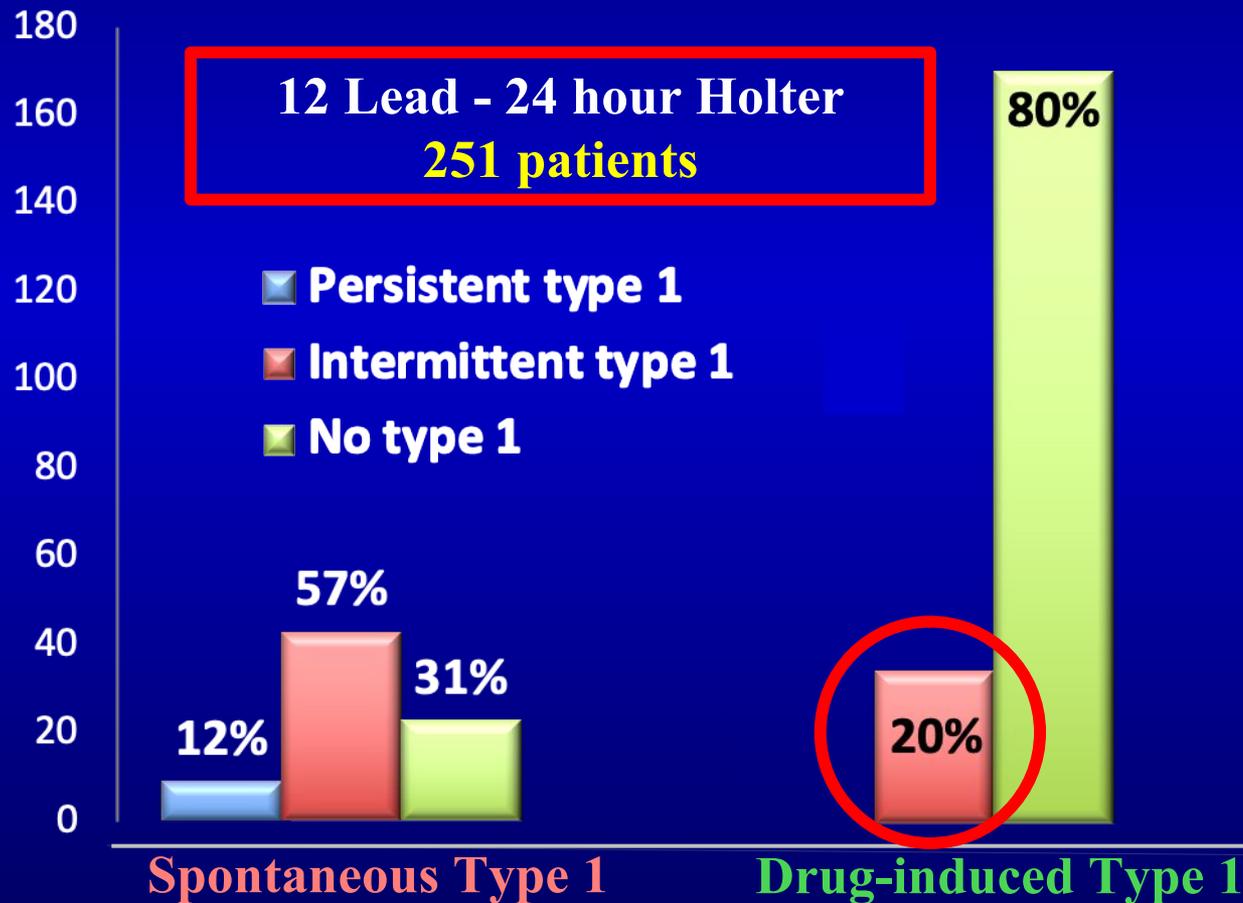


Prevalence of Type 1 Brugada Electrocardiographic Pattern Evaluated by Twelve-Lead Twenty-Four-Hour Holter Monitoring



Natascia Cerrato, MD^a, Carla Giustetto, MD^{a,*}, Elena Gribaudo, MD^a, Elena Richiardi, MD^b,
Lorella Barbonaglia, MD^c, Chiara Scrocco, MD^a, Domenica Zema, MD^a, and Fiorenzo Gaita, MD^a

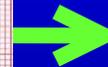
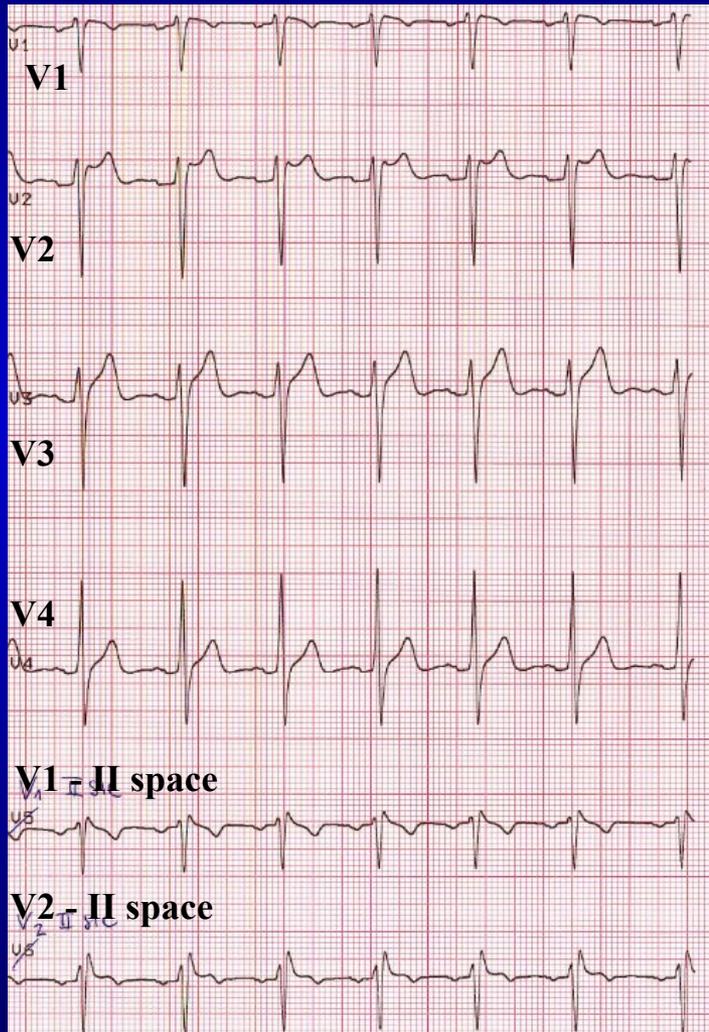
Am J Cardiol 2015; 115:



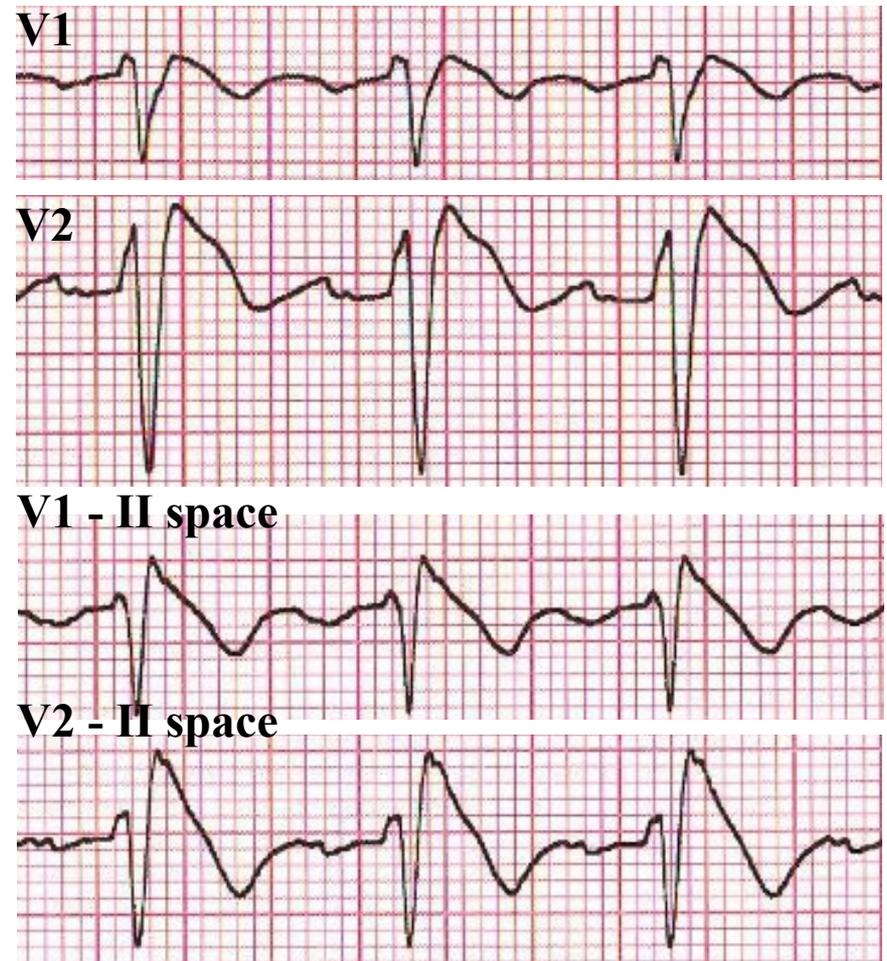
Drug challenge with Na⁺ channel blockers

(ajmaline 1 mg/kg IV over 5 -10 min o flecainide 2 mg/kg IV over 10 min)

Basal ECG



Ajmaline infusion (1mg/kg in 5 min)



**Which and How
to treat
Brugada patients**

Risk factors for arrhythmic events

Most patients with arrhythmic events are between 20 and 65 years old

	Probst et al. (1029 pts)	Sieira et al. (343 pts)
Aborted sudden death	HR= 12.4, p<0.001	HR= 10.9, p<0.01
History of <u>syncope</u>	HR = 3.4, p=0.002	HR = 3.7, p<0.01
<u>Spontaneous type 1 ECG</u>	HR = 1.8, p=0.04	HR = 2.7, p<0.01
EP-Study	HR= 1.9, p=0.05	HR = 4.7, p<0.01
Male gender	NS	HR = 2.7, p=0.02
SCN5A mutations	NS	-

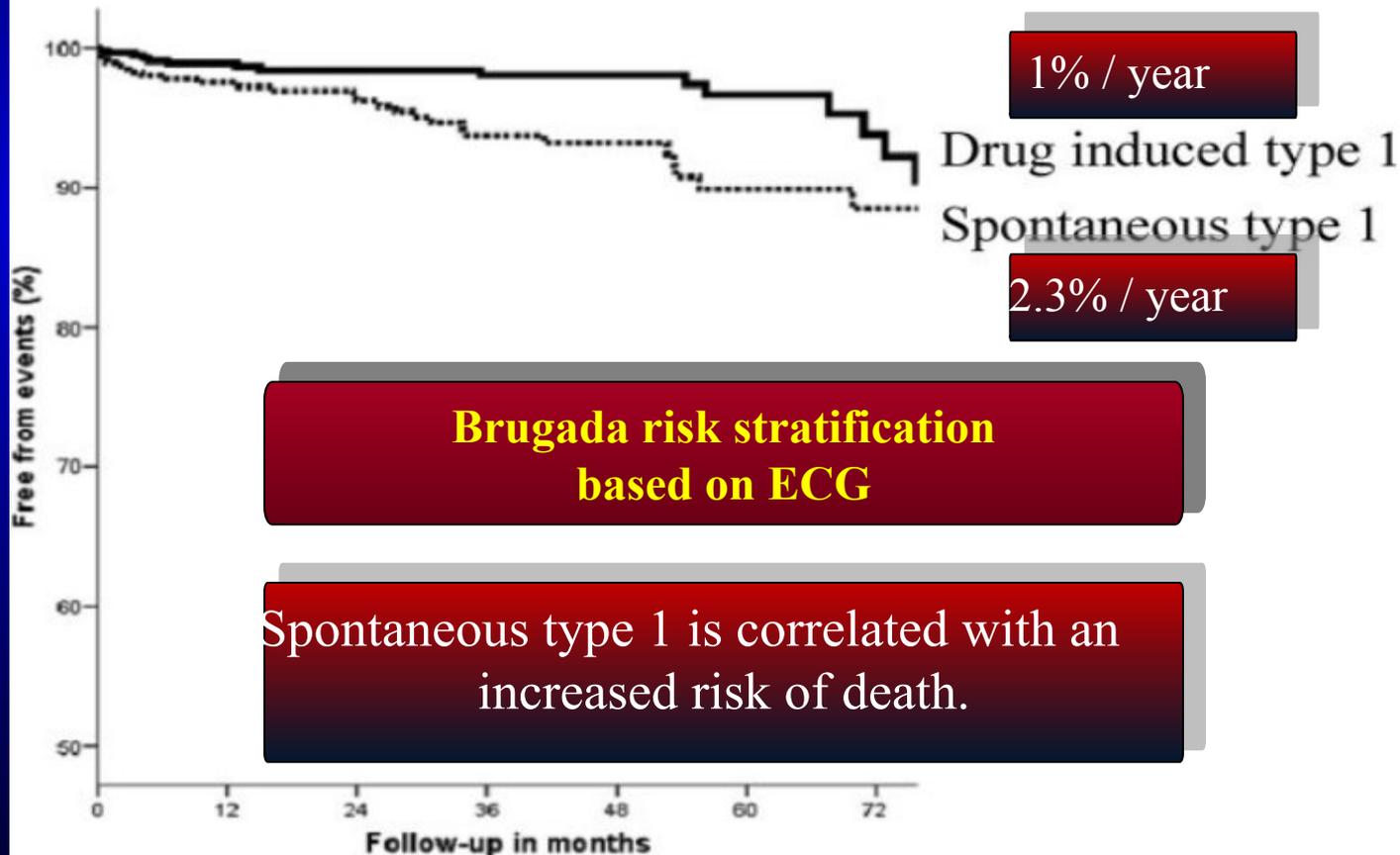
aSD, syncope and spontaneous type 1 ECG pattern are known as significant risk factors

Long-Term Prognosis of Patients Diagnosed With Brugada Syndrome

Results From the FINGER Brugada Syndrome Registry

1029 pts
mean f-up 3 years

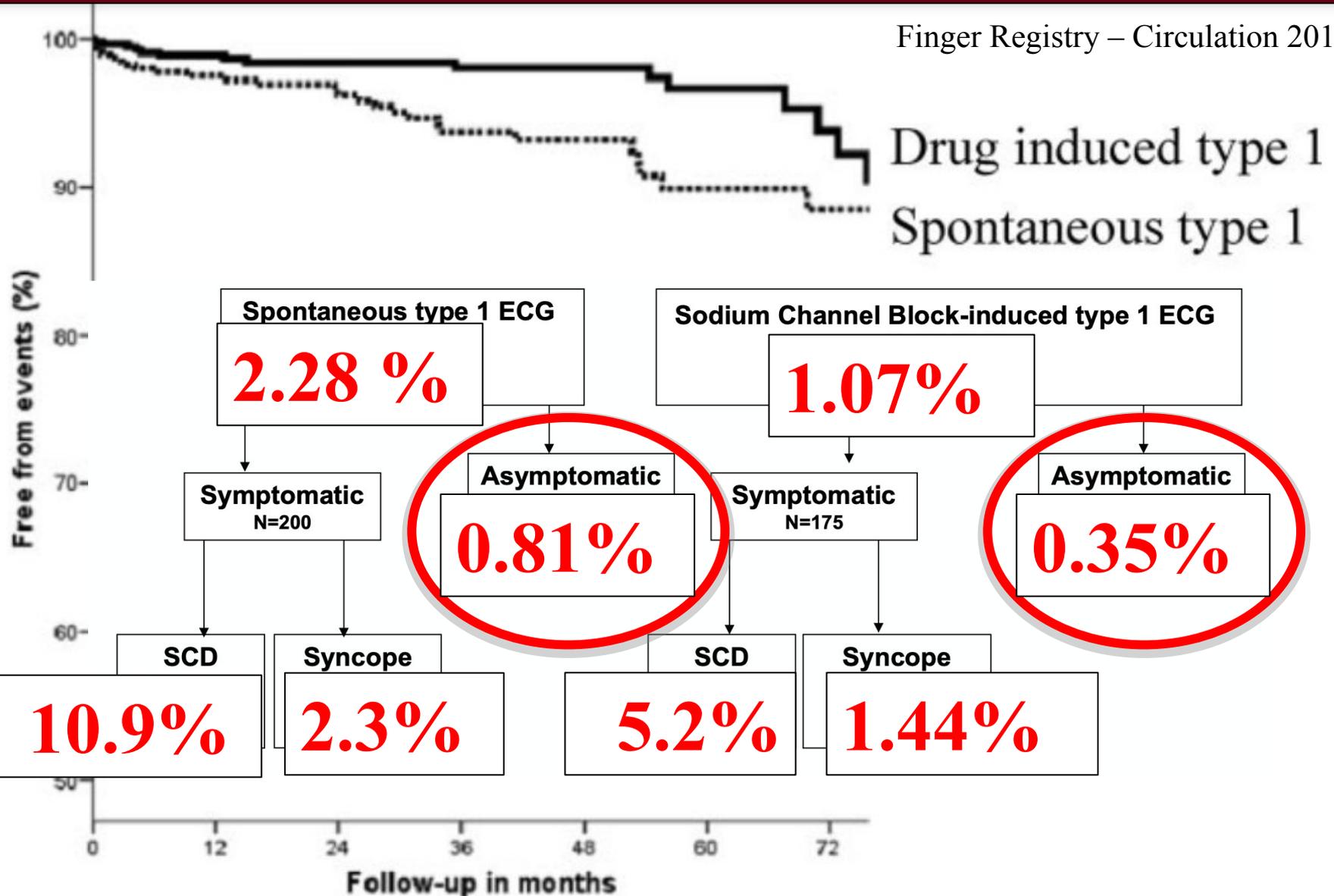
V. Probst, MD, PhD, C. Veltmann, MD, L. Eckardt, MD, P.G. Meregalli, MD, F. Gaita, MD, H.L. Tan, MD, PhD, D. Babuty, MD, PhD, F. Sacher, MD, C. Giustetto, MD, E. Schulze-Bahr, MD, PhD, M. Borggrefe, MD, PhD, M. Haissaguerre, MD, P. Mabo, MD, PhD, H. Le Marec, MD, PhD, C. Wolpert, MD, PhD, and A.A.M. Wilde, MD, PhD *Circulation* 2010



Brugada risk stratification

ECG & SYMPTOMS BASED

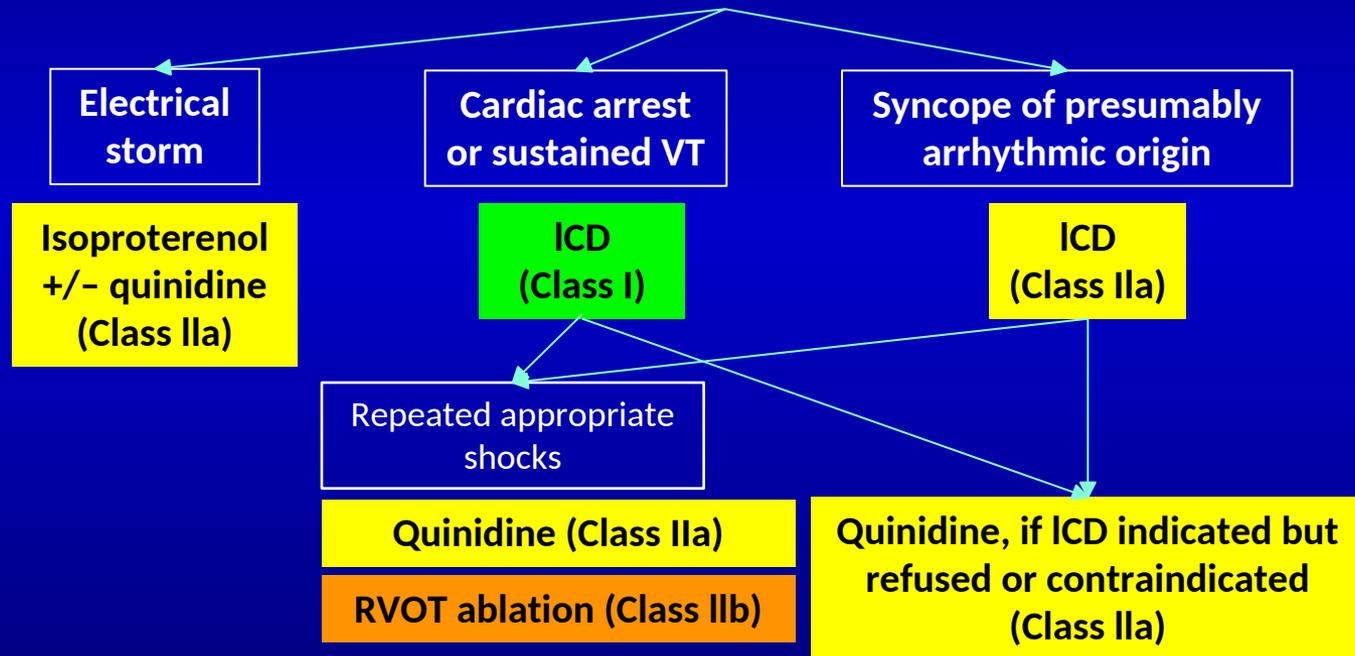
Finger Registry – Circulation 2010



Management of Brugada patients

- Avoid drugs that may induce or aggravate ST segment elevation (www.Brugadadrugs.org)
- Avoid cocaine and excessive alcohol intake
- Immediately treat fever with antipyretic drugs. (Class I)

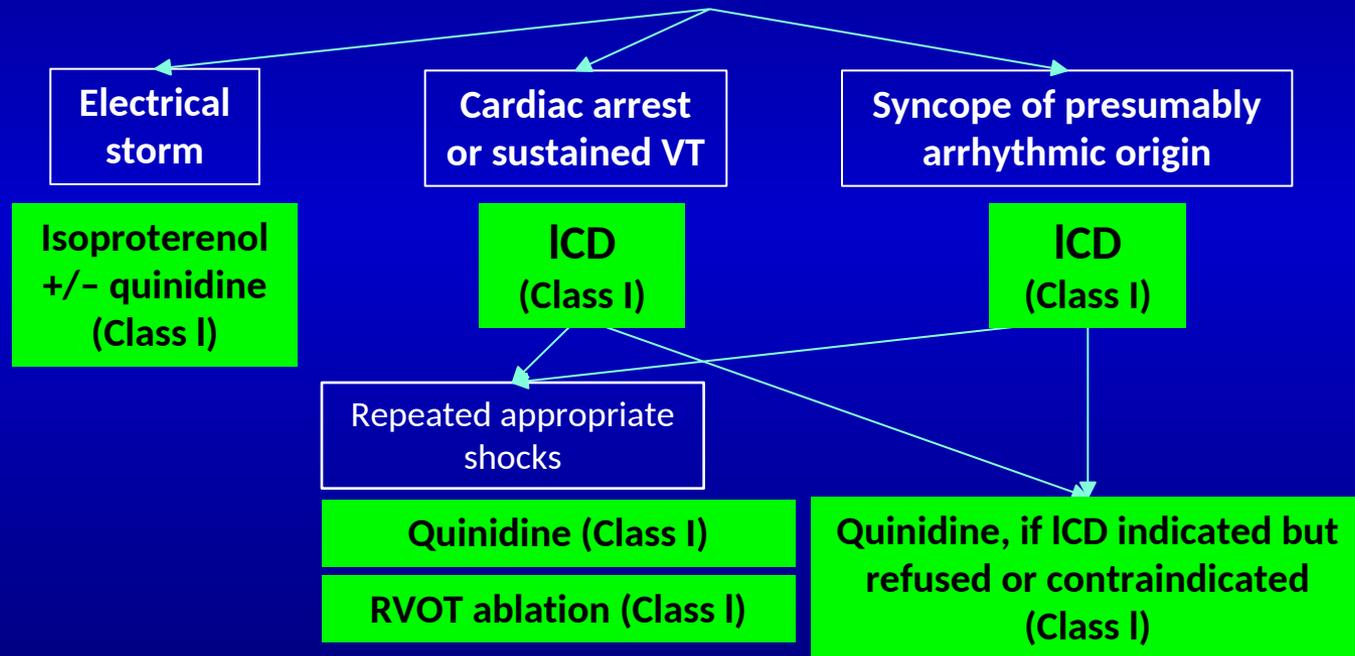
Symptomatic



Management of Brugada patients

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Symptomatic



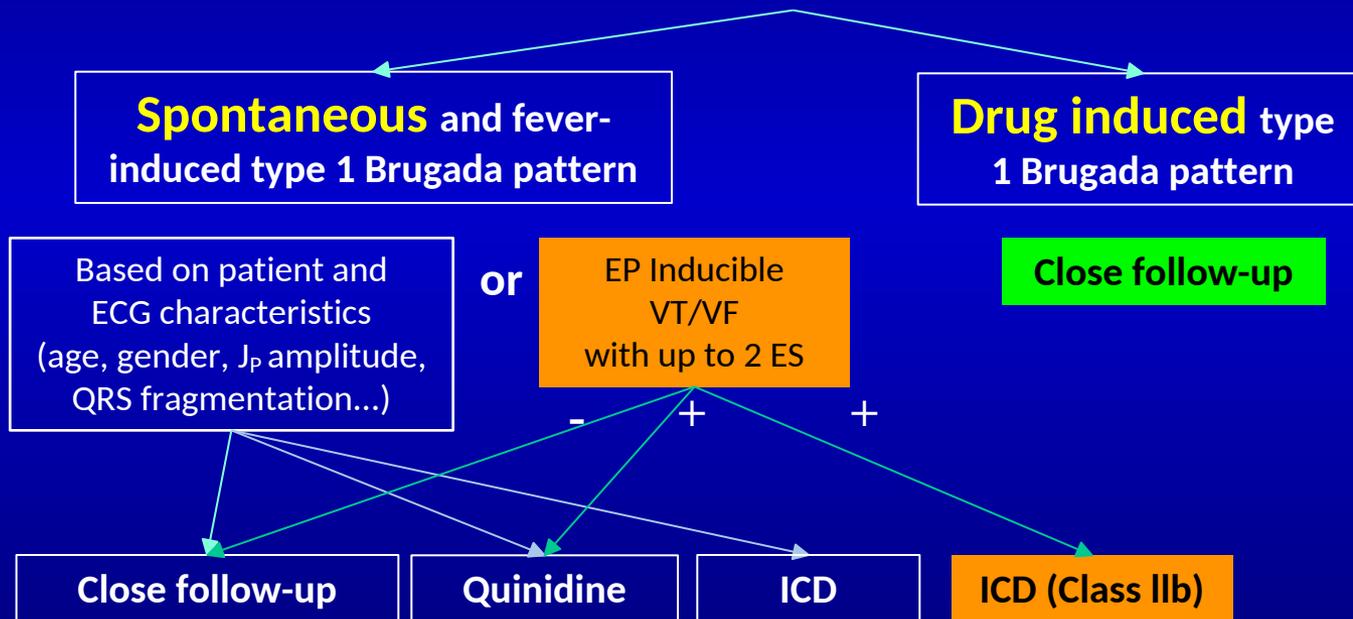
Antzelevitch et al. Europace 2017 Expert Consens Conference;19:665-69

Al-Khatib et al. [2017 AHA/ACC/HRS Guidelines on VA/SCD \(Circulation\)](#)

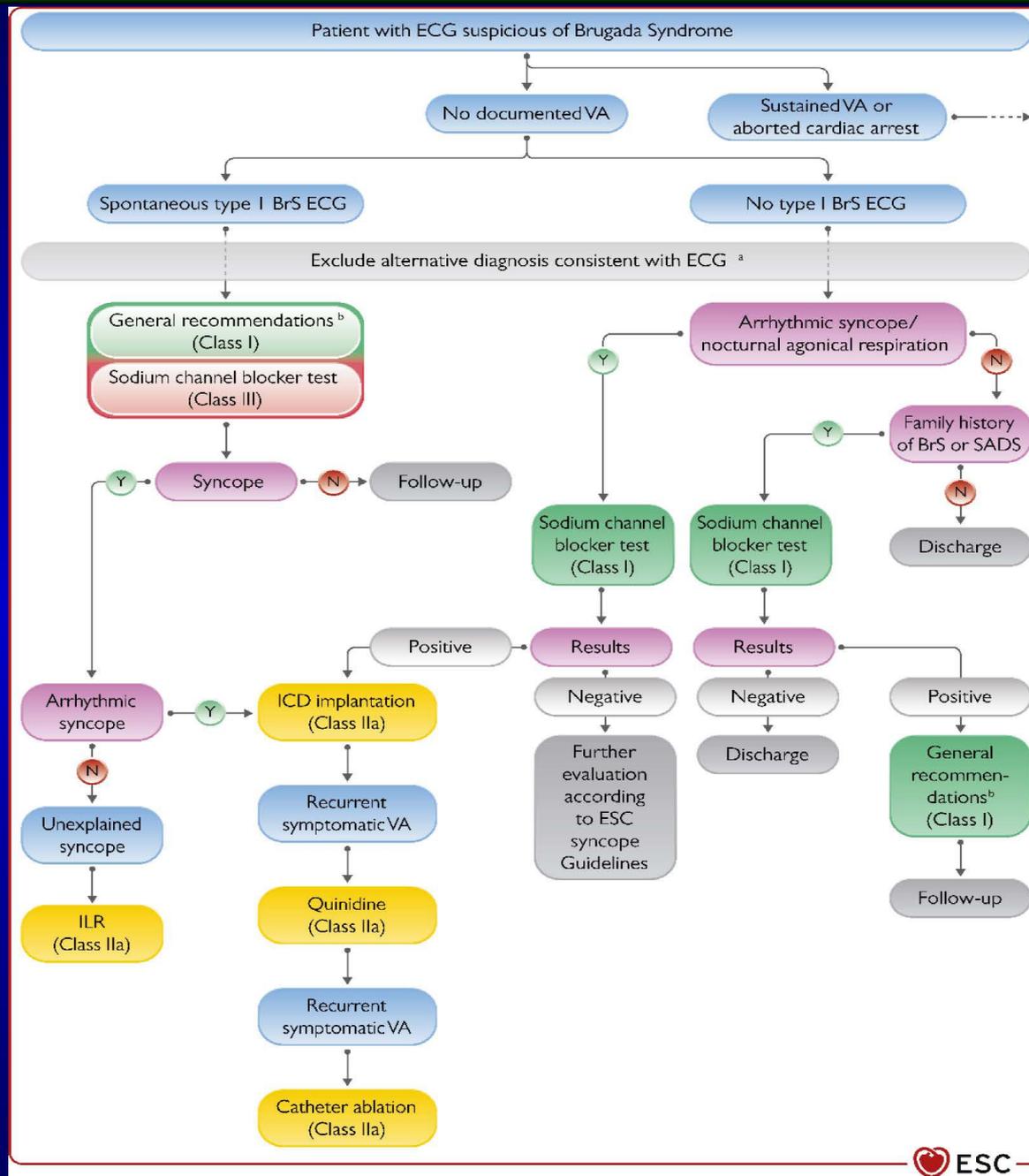
Management of Brugada patients

- Avoid drugs that may induce or aggravate ST segment elevation
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Asymptomatic



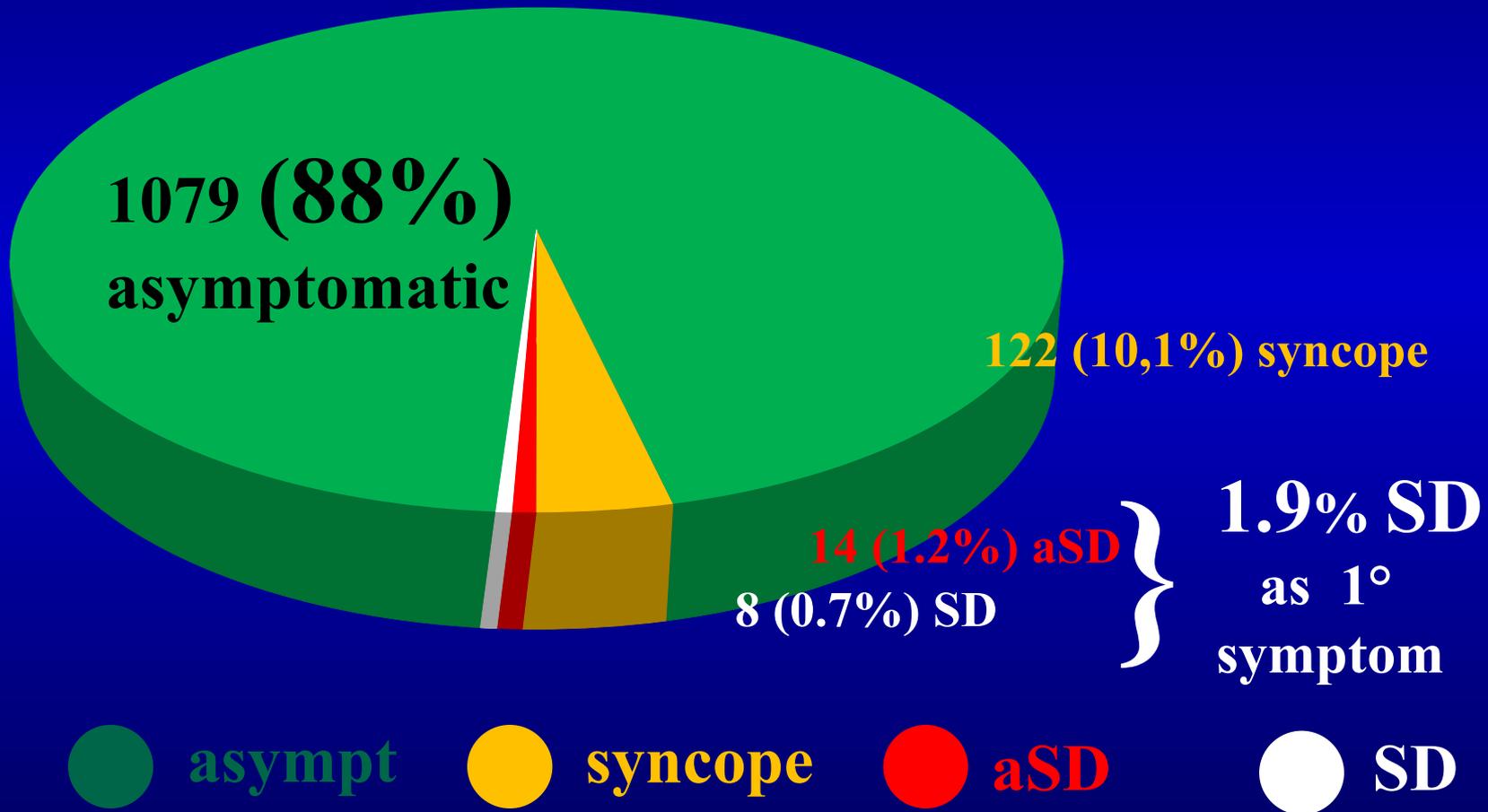
Clinical guidelines lack specific recommendations for asymptomatic pts



Brugada Piedmont Registry results at 2022

Total **1223** pts

enrolled from 2001 to 2018 and followed until 2022



How we treated ALL the patients with Brugada ECG pattern

Avoid drugs that may increase the ST segment elevation www.brugadadrugs.org

Drug-Induced Brugada-Like ECG Patterns	Psychotropic drugs
Antiarrhythmic drugs	Tricyclic antidepressants ²¹⁸
Na ⁺ channel blockers	Amitriptyline, ^{217,218} Nortriptyline, ¹⁵¹ Desipramine, ¹⁴⁹ Clomipramine ¹⁵⁰
Class IC drugs (Flecainide, ^{12,15,142,207,208} Pilsicainide, ^{148,209} Propafenone ²¹⁰)	Tetracyclic antidepressants
Class IA drugs (Ajmaline, ^{12,211} Procainamide, ^{12,13} Disopyramide, ^{4,13} Cibenzoline ^{212,213})	Maprotiline ²¹⁷
Ca ²⁺ channel blockers	Phenothiazine
Verapamil	Perphenazine, ²¹⁷ Cyamemazine.
β -Blockers	Selective serotonin reuptake inhibitors
Propranolol intoxication ²¹⁴	Fluoxetine ²¹⁸
Antianginal drugs	Lithium ¹⁵⁷
Ca ²⁺ channel blockers	Other drugs
Nifedipine, diltiazem	Histaminic H1 receptor antagonists
Nitrate	Dimenhydrinate ¹⁵²
Isosorbide dinitrate, nitroglycerine ²¹⁵	Diphenhydramine ²¹⁹
K ⁺ channel openers	Cocaine intoxication ^{153,220}
Nicorandil	Alcohol intoxication

Modified from Antzelevitch et al.⁵⁶ and Shimizu²²¹ with permission.

Promptly treat **fever**

How we treated Brugada patients – before 2013

1. aSD

ICD

if still arrhythmic events: add HQ

2. unexplained SYNCOPE

EPS +

ICD

EPS -

Follow-up/loop recorder +/- HQ

3. ASYMPTOMATIC/
neurally mediated syncope
spontaneous or induced type 1

EPS +

HQ

EPS +

ICD

EPS -

F-up

EPS -

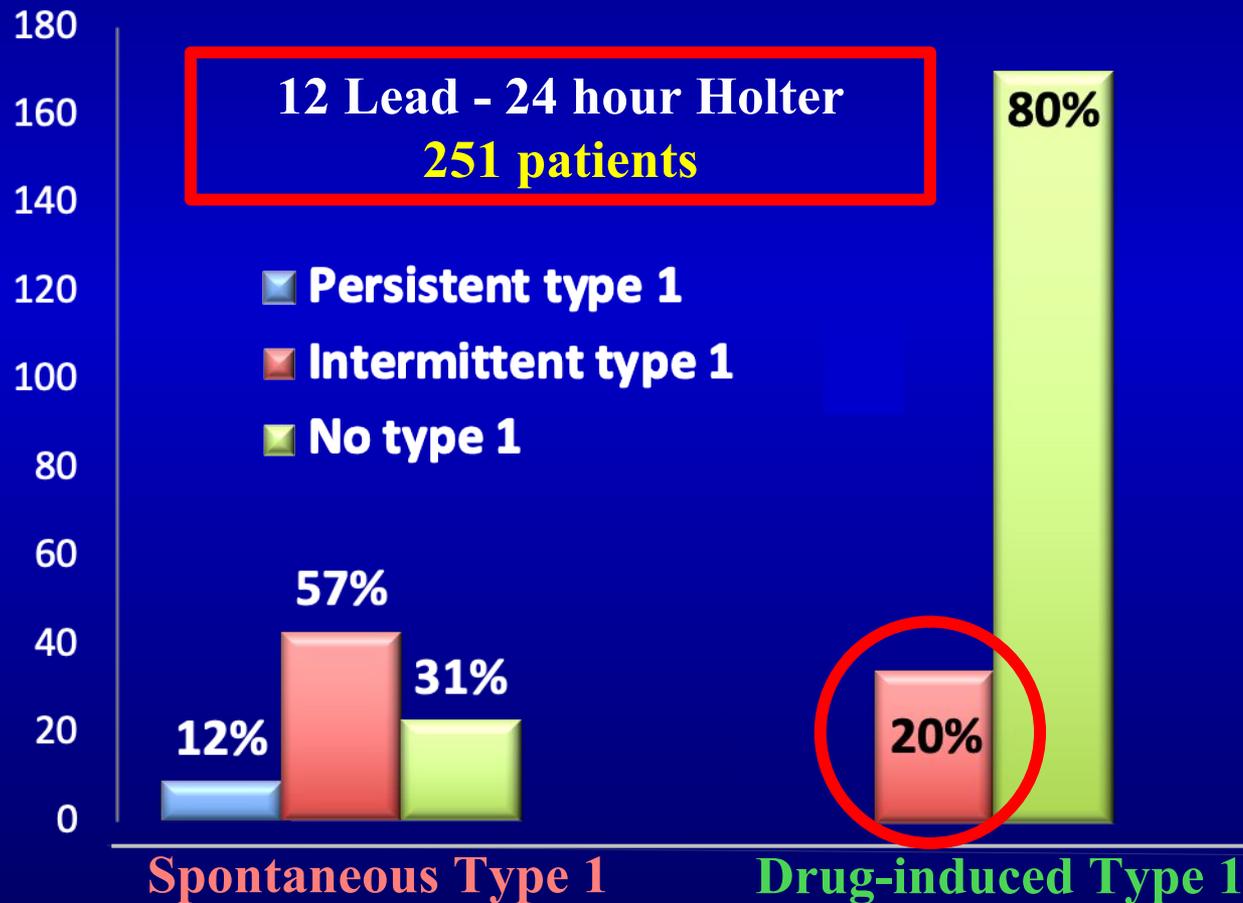
F-up

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Am J Cardiol 2015; 115:



ICD-related complications

Implantable cardioverter-defibrillator in Brugada syndrome: Long-term follow-up

Ibrahim El-Battrawy^{1,2} | Gretje Roterberg¹ | Volker Liebe¹ | Uzair Ansari¹ |
Siegfried Lang^{1,2} | Xiaobo Zhou^{1,2} | Martin Borggrefe^{1,2} | Ibrahim Akin^{1,2}

747 patients from 11 research papers diagnosed with BrS and treated with **transvenous ICD** implantation 2007 and 2018

- ✓ inappropriate shocks 18%
- ✓ lead failure 5.4%
- ✓ infection 4%
- ✓ psychiatric problems 1.5%

From 2013 **S-ICD** is a reliable alternative to transvenous ICD for both secondary and primary prevention

Epicardial RVOT ablation

Prevention of Ventricular Fibrillation Episodes in Brugada Syndrome by Catheter Ablation Over the Anterior Right Ventricular Outflow Tract Epicardium

Koonlawee Nademane, MD; Gumpanart Veerakul, MD; Pakorn Chandanamatta, MD; Lertlak Chaothawee, MD; Aekarach Ariyachaipanich, MD; Kriengkrai Jirasirojanakorn, MD; Khanchit Likittanasombat, MD; Kiertijai Bhuripanyo, MD; Tachapong Ngarmukos, MD

Circulation 2011, 123: 1270-79

Brugada Syndrome Phenotype Elimination by Epicardial Substrate Ablation

Josep Brugada, MD*; Carlo Pappone, MD, PhD*; Antonio Berruezo, MD, PhD; Gabriele Vicedomini, MD; Francesco Manguso, MD, PhD; Giuseppe Ciconte, MD; Luigi Giannelli, MD; Vincenzo Santinelli, MD

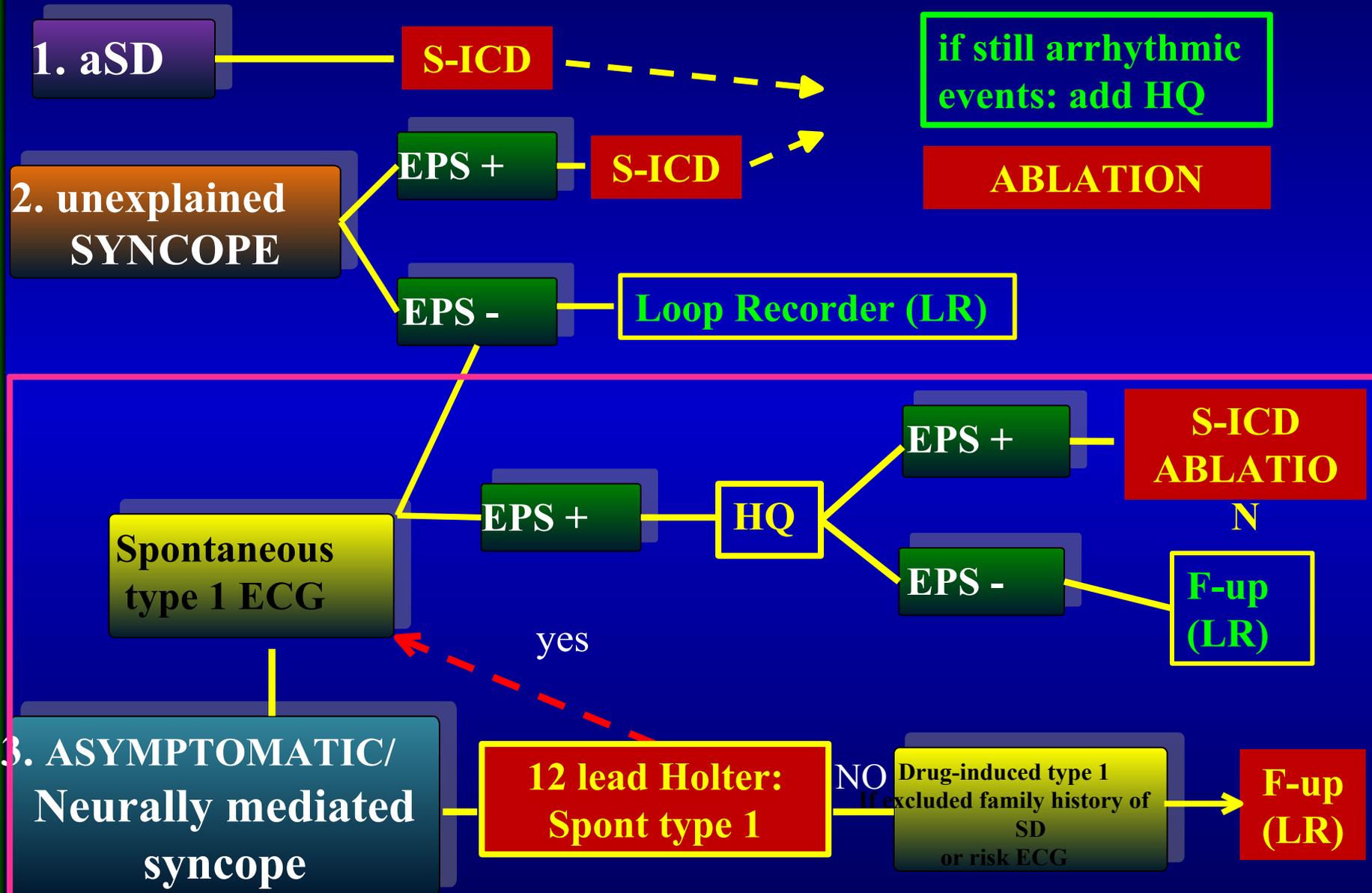
Circ Arrhythm El 2015

Electrical Substrate Elimination in 135 Consecutive Patients With Brugada Syndrome

Carlo Pappone, MD, PhD*; Josep Brugada, MD, PhD*; Gabriele Vicedomini, MD; Giuseppe Ciconte, MD; Francesco Manguso, MD, PhD; Massimo Saviano, MD; Raffaele Vitale, MD; Amarild Cuko, MD; Luigi Giannelli, MD; Zarko Calovic, MD; Manuel Conti, MD; Paolo Pozzi, Eng; Andrea Natalizia, PhD, Eng; Simonetta Crisà, Eng; Valeria Borrelli, PhD; Ramon Brugada, MD, PhD; Georgia Sarquella-Brugada, MD, PhD; Marco Guazzi, MD; Alessandro Frigiola, MD; Lorenzo Menicanti, MD; Vincenzo Santinelli, MD

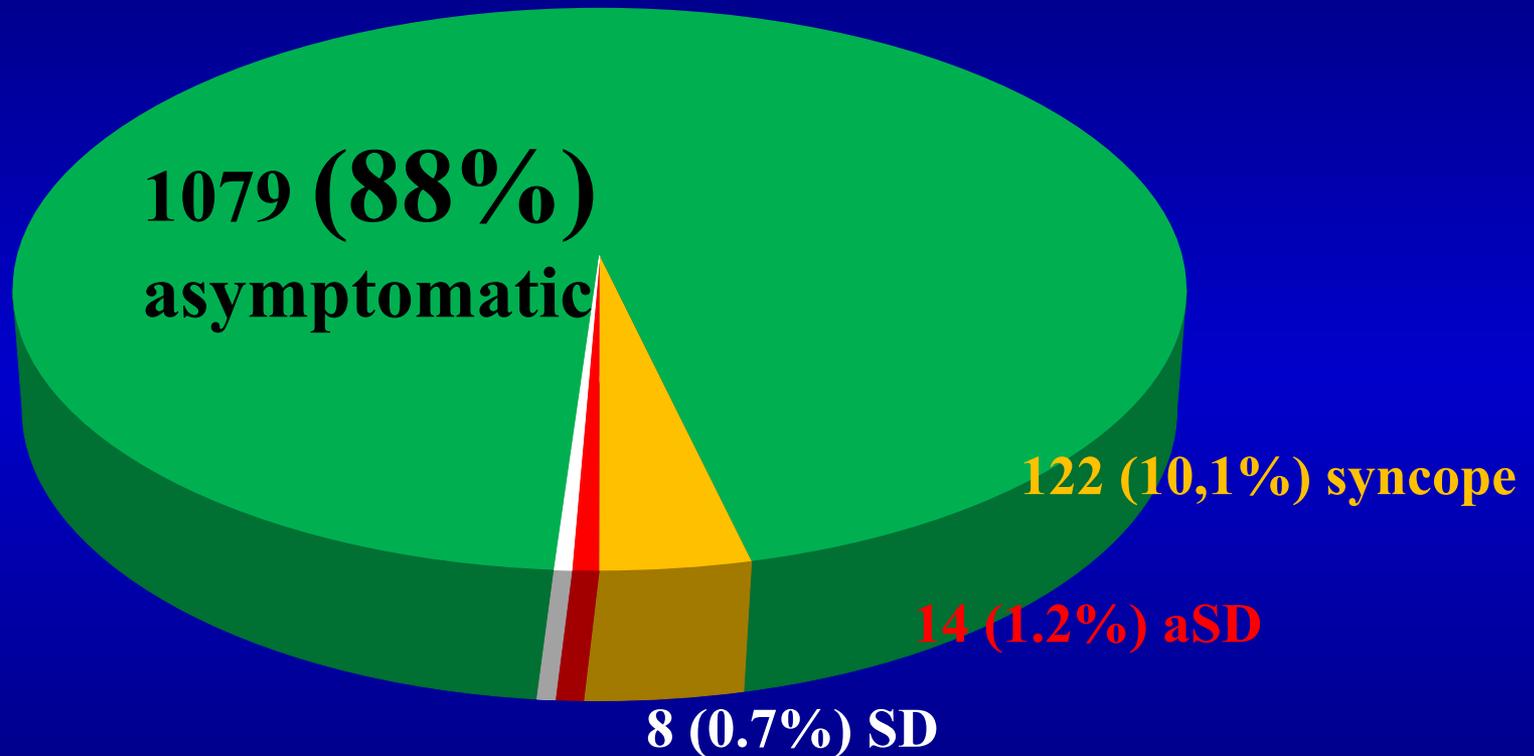
Circ Arrhythm El 2017

How we treated Brugada patients after 2013



Brugada Piedmont Registry results at 2022

Total **1223** pts (-8 SD) **1215**
enrolled from 2001 to 2018 and followed until 2022



● **asympt**

● **syncope**

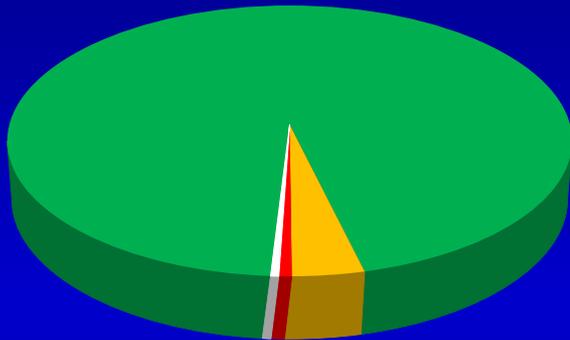
● **aSD**

● **SD**

Brugada Piedmont Registry



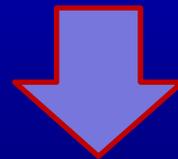
pts with history of **aborted SD**



14 (1.1%)

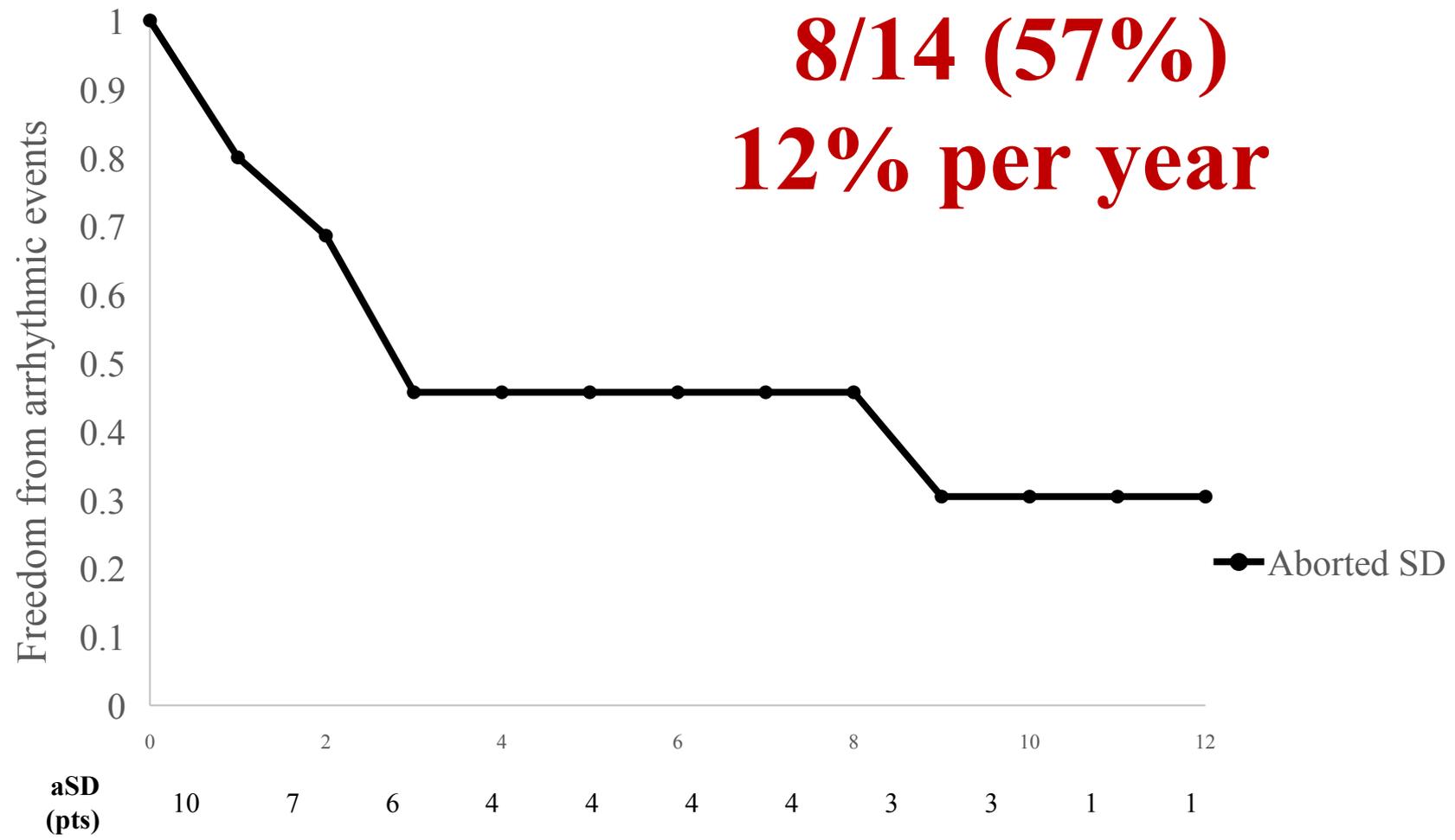
13 ICD

1 child € HQ and loop recorder

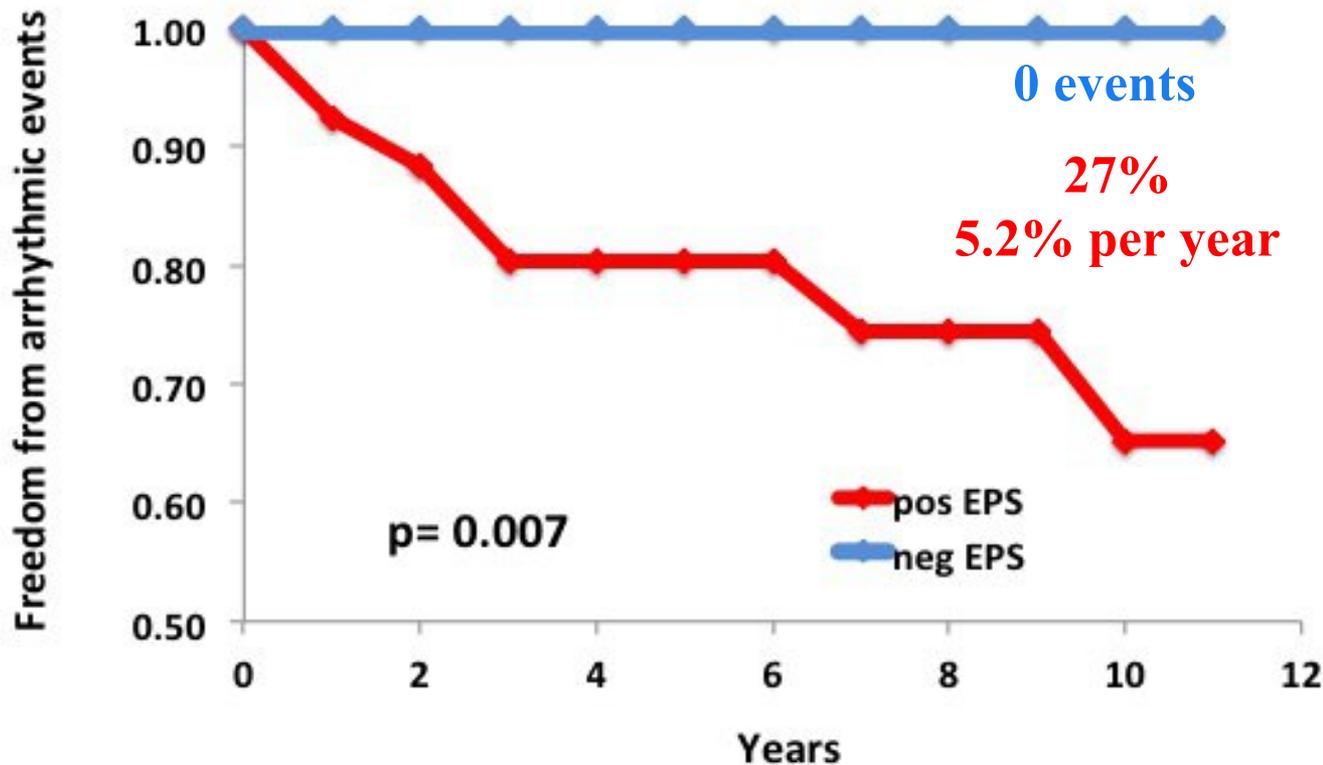


NO DEATHS

Arrhythmic events in pts with aSD



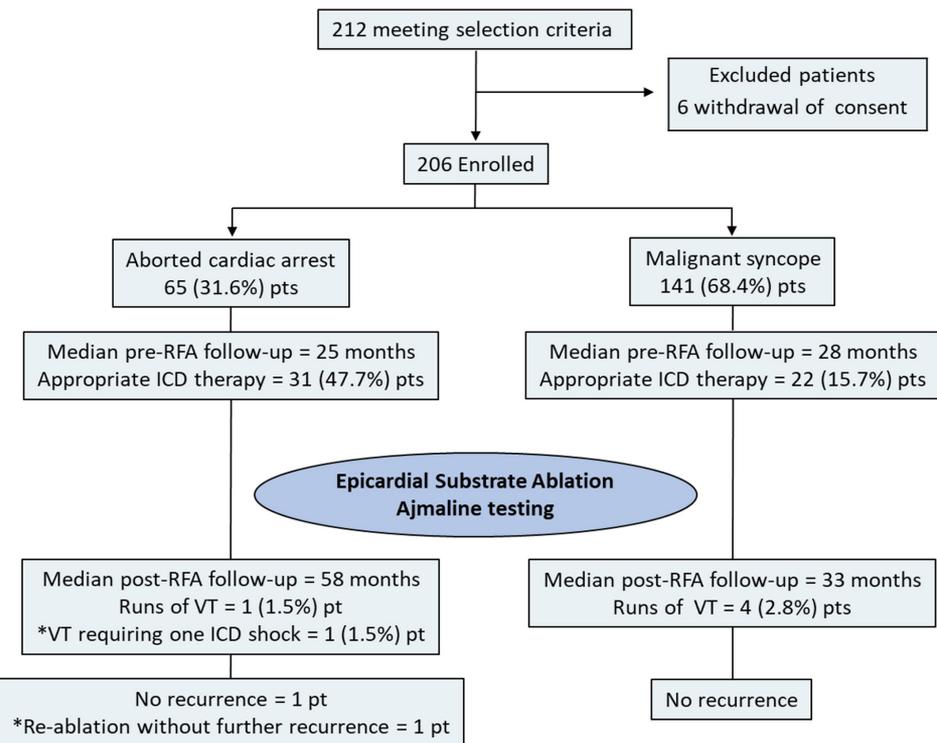
Role of **EPS** in pts with UNEXPLAINED SYNCOPES



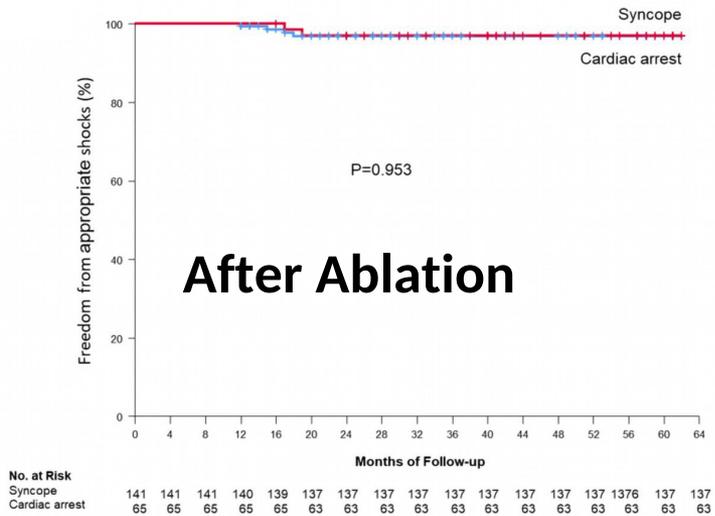
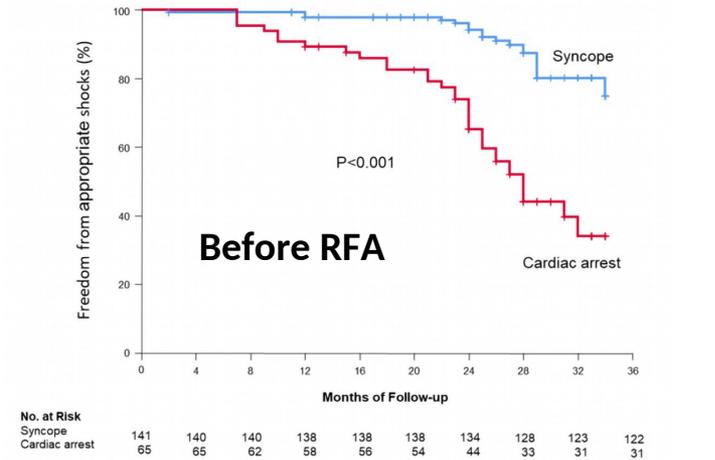
**In 2022
one SD**

pos EPS	26	23	22	20	17	15	14	13	11	8	7
neg EPS	31	28	22	17	14	14	10	8	5	3	2

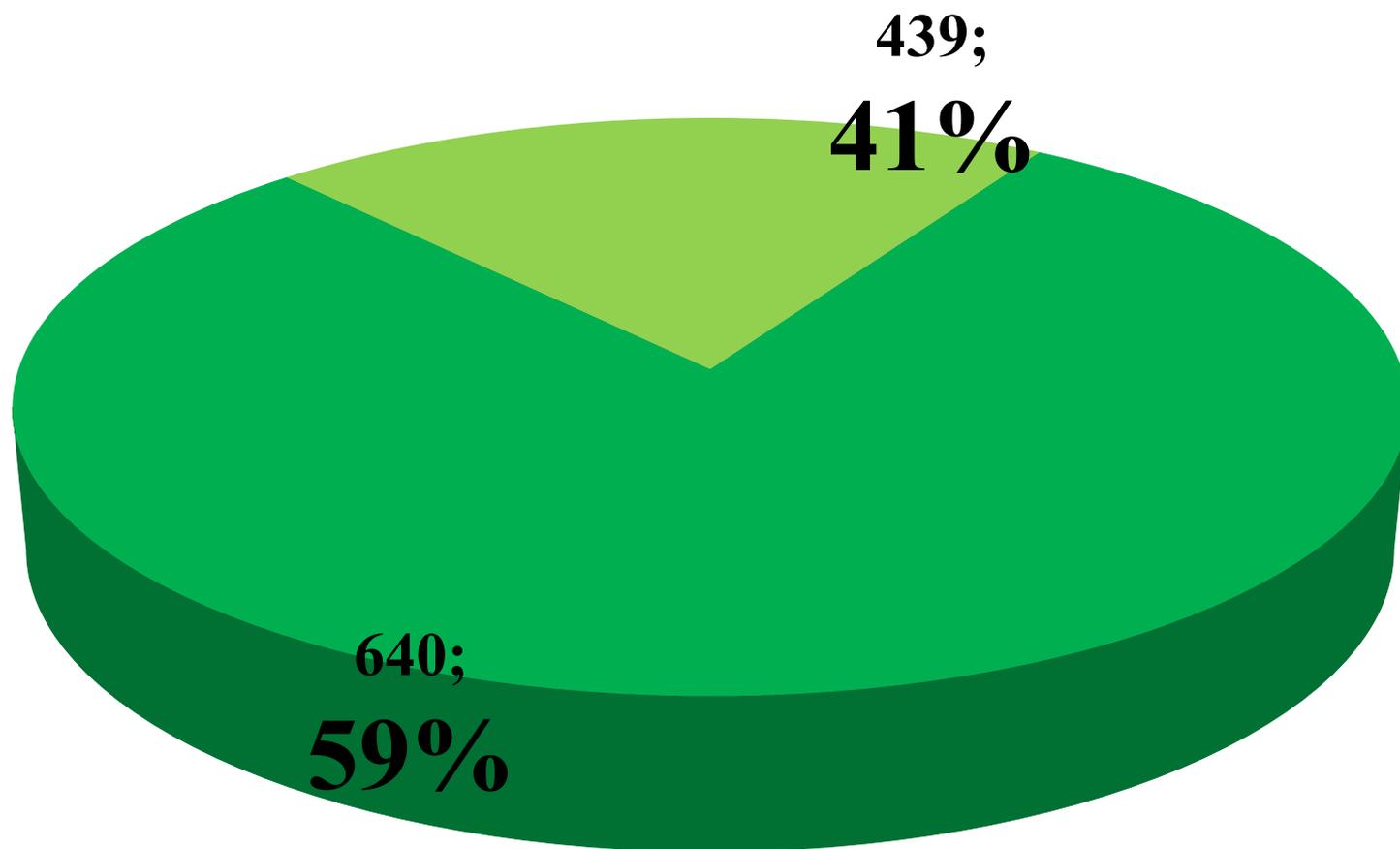
Catheter Ablation in 212 Symptomatic BrS



under review. Courtesy of Prof. C. Pappone



1079 Asymptomatic Brugada patients



■ spont type 1 ■ induced type 1

Asymptomatic pts with SPONTANEOUS type 1

439 pts

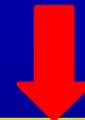
NO EPS
131 (30%)

ICD
2
1.5%

HQ
12
9.3%

Ablation
0

NO tp
117
89%



4 (3,5%)
2SD+ 2aSD

Asymptomatic pts with SPONTANEOUS type 1

439 pts
(41%)

EPS: yes
308 (70%)

EPS +
99 (32%)

ICD
54 (55%)

HQ
47+5 (53%)
Stop in 14 (29%)

Ablation
12 (12%)

NO tp
4 (4%)



5/58 (8,6%)
shocks

NO DEATHS

Asymptomatic pts with SPONTANEOUS type 1

439 pts
(41%)

EPS: yes
308 (70%)

EPS -
209 (68%)

ICD
13
6%

HQ
5
2%

Ablation
1

NO tp
190
92%

3/190
(1,5% SD)

Asymptomatic pts with INDUCED type 1

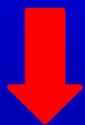
640 pts

Eventi 2 (0.3%)

NO EPS
488 (78%)

EPS +
22 (18%)

EPS -
112 (82%)

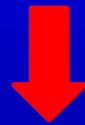


1 syncope



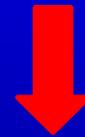
0.2%

1 SD



0.4%

1 SHOCK



**NO
DEATH**

35 events out of 1215 (2,9)
Sudden Death 9 (0.7%)



14 aSD
(13 ICD)
8/14 events
(57%)
NO SD

123 syncope
(54 ICD)
13/123 events
(11%)
1 SD

1079 (91 ICD)
asymptomatic
14/1079 events
(1.3%)
6 scocks
8 SD (2 aSD)

We have properly stratified and treated the symptomatic patients, but we still have to improve risk stratification and treatment of the asymptomatic patients

Conclusions

Clinics, ECG markers and EPS help to better identify higher risk patients but are not enough to identify all pts at risk of SD

Ablation, considering its good results and the low rate of complications, should be considered increasingly in symptomatic patients with Brugada syndrome.

Asymptomatic patients with Type I Brugada, with current therapy , still present a low (3,5 %) but not negligible risk of SD.

E.P.stratification reduces the risk of death from 3.5% to 1.3%