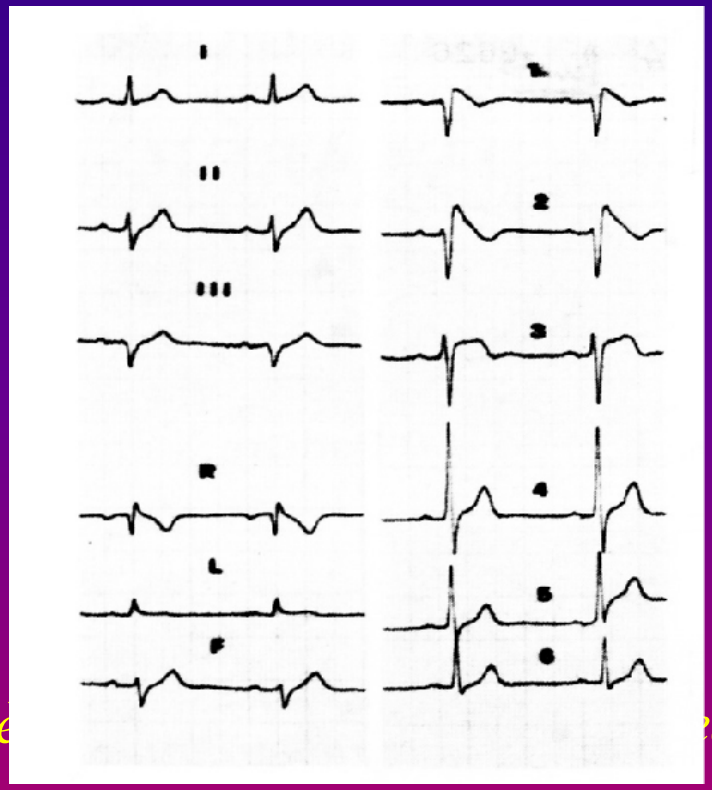
"Il pattern ECG di Nava-Martini-Brugada e la sindrome detta di Brugada".

Bortolo Martini,
Ospedale di Santorso (VI)

Roma 20-21 ottobre 2017

La sindrome della ripolarizzazione precoce nelle precordiali destre



- *Questa e … a Nava, 1988*

Gli studi su questa sindrome sono diventati una raccolta delle figurine e non una seria indagine scientifica che segua i dettami di Galileo. E' un album che più si arricchisce più la soluzione si allontana. Manca sempre l'ultima figurina, la più importante, ma esiste sul serio?

Ci si è dimenticati

- Che quasi tutte le scoperte sono state fatte analizzando accuratamente un singolo caso!
- Qualcuno si ricorda come è stato scoperto il diabete?

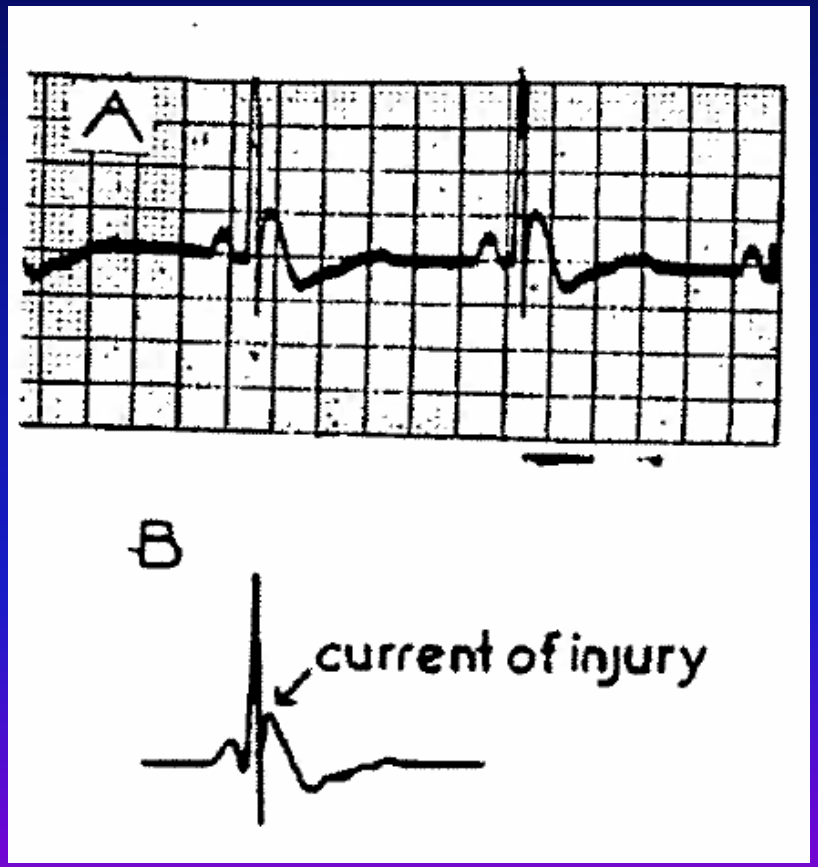
- *«Durante il periodo Rinascimentale*
- *il veneziano Vittorio Trincavella*
- *scopre il diabete assaggiando le urine»*

Experimental Hypothermia: Respiratory and Blood pH Changes in Relation to Cardiac Function

JOHN J. OSBORN[1]

From the Department of Pediatrics, New York University College of Medicine, New York City

1953



In 1953, Dr. John Osborn described the J wave as an "injury current" resulting in ventricular fibrillation during experimental hypothermia.

Osher's ECG

ELECTROCARDIOGRAPHIC PATTERN SIMULATING ACUTE MYOCARDIAL INJURY

By Harold L. Osher, M.D.
Portland, Maine

and

Louis Wolff, M.D.
Boston, Massachusetts

(From the Electrocardiographic Laboratory, Beth Israel Hospital, the Department of Medicine, Harvard Medical School, Boston, Massachusetts, and the Heart Disease Epidemiology Study, National Heart Institute, National Institutes of Health, Public Health Service, Department of Health, Education, and Welfare, Framingham, Massachusetts.)

Abnormal displacement of the S-T segment may be associated with a variety of clinical states. In myocardial injury the typical configuration consists of elevation and upward bowing of the S-T segment with symmetrical inversion of the T waves in leads facing the affected area. We have recently observed a similar electrocardiographic pattern in patients without clinical evidence of any acute myocardial process. The purpose of this paper is to point out the distinguishing characteristics of this pattern and to elucidate the mechanism of its production.

Our attention was first drawn to this pattern by the electrocardiograms reproduced in Figure 1. The patient, a 39-year-old white male, entered the Beth Israel Hospital for study on January 19, 1950, because of an episode, 2 weeks previously, of interscapular pain following exertion, and vague substernal discomfort 2 days prior to admission. Physical examination was entirely within normal limits with blood pressure of 120/80 and a normal sized heart with sounds of good quality and no murmurs. Chest roentgenogram and laboratory examination of urine and blood revealed no abnormalities. His hospital course was afebrile and symptom-free except for anxiety; there was no further chest pain and white blood cell counts and erythrocyte sedimentation rates remained normal. Initial electrocardiograms showed right bundle branch block (RBBB) with elevation of S-T segments and inversion of T waves in the right precordial leads, and were interpreted as consistent with anteroseptal injury. However, subsequent tracings showed persistence of the S-T-T pattern with only minor variations, rather than the serial changes typical of evolution of an acute process. The patient was discharged on February 6, 1950, and remained well for 8 months.

In November, 1950, during a period of emotional turmoil, he was again hospitalized because of mild substernal discomfort lasting one day. Clinical and laboratory examination failed to reveal any abnormality, and his hospital course was uneventful except for a few brief twinges of chest pain related to emotional upsets. The electrocardiograms were essentially unchanged and again failed to show progressive changes. The patient has remained well to date (April, 1953), and presents no objective evidence of heart disease although the unusual electrocardiographic pattern persists.

Figure 2 shows the electrocardiograms of a healthy 43-year-old white male (H.D.E.S. Case Number 4549)

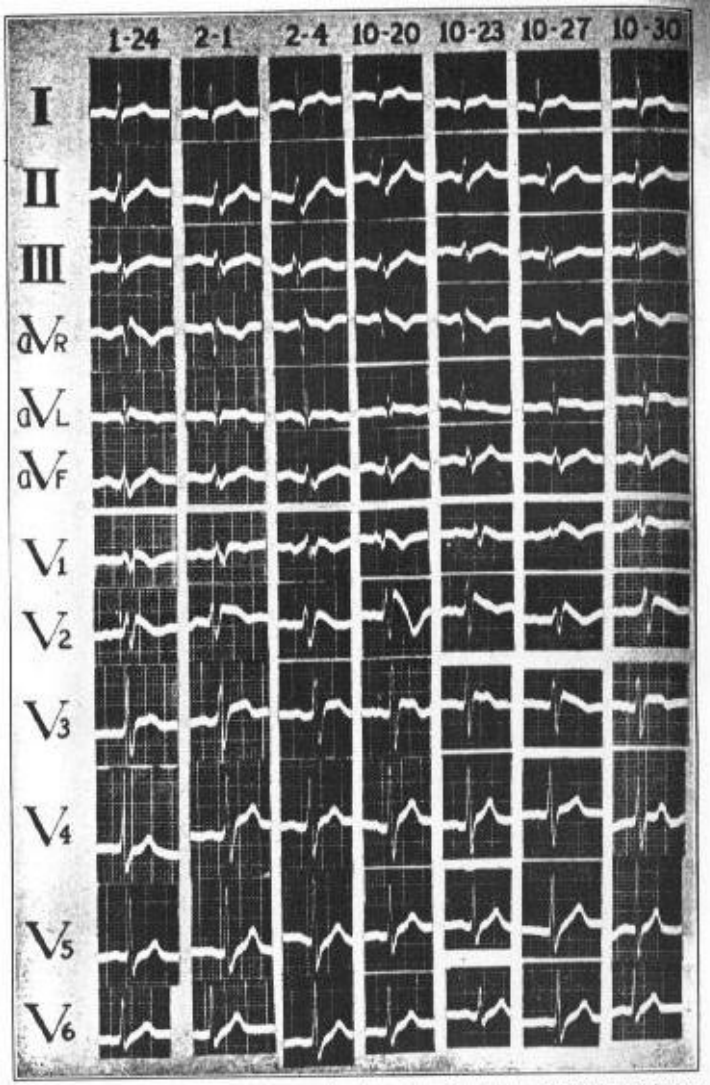


Fig. 1.—Electrocardiograms of a 39-year-old white male with mild atypical chest pain but no clinical evidence of heart disease. Note the wide QRS interval (0.12-0.13 sec.) with features of RBBB, and S-T segment elevation and T wave inversion in the right precordial leads, simulating the pattern of anteroseptal injury. The serial tracings show minor variations in the S-T-T configuration, but not the typical evolution seen with myocardial injury.

"questo è dovuto ad un ritardo di depolarizzazione focale di una porzione del ventricolo destro"

Osher HL, Wolff L. Electrocardiographic pattern simulating acute myocardial injury. Am J Med Sci 1953; 226: 541–5.

Harold osher

Louis Wolff, Sir John Parkinson and Paul Dudley

Pistas de una enfermedad orgánica subyacente en el síndrome de Brugada
Clues of an underlying organic substrate in the Brugada Syndrome

Bortolo Martini, Nicolò Martini, Manlio Marquez, Caterina Dorantes, Li Zhang, Guy Fontaine and Andrea Nava 2017

- On 20 October 1984 a 42 years old healthy and previous asymptomatic cook, quietly talking with the post officer outside his restaurant in Conegliano (the land of Prosecco) suddenly collapsed and a successful defibrillation was performed. The electrocardiogram (ECG) taken showed an ST elevation in the precordial leads (Figure 1) but a clever Cardiologist ruled out acute myocardial infarction. The patient is still alive and asymptomatic on beta blockers therapy.

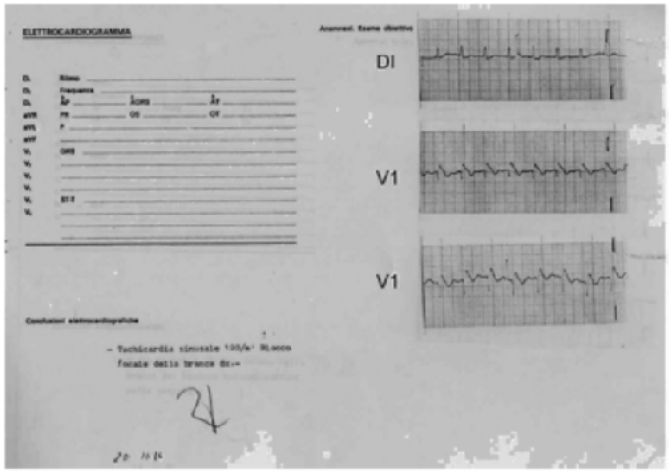
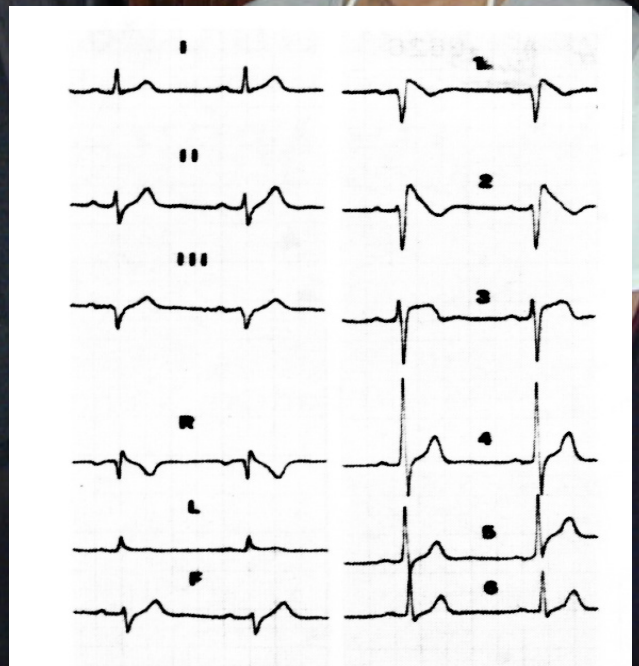


Figura 1 Derivaciones DI y V1 del primer paciente diagnosticado con el síndrome de muerte súbita, asociada a una morfología electrocardiográfica similar al bloqueo de rama derecha y elevación del segmento ST. Fue tomado poco después de un paro cardiaco reanimado mediante desfibrilación el 20 de noviembre de 1984. Es interesante que el cardiólogo lo interpretó como «taquicardia sinusal a 120 lpm y bloqueo focal de rama derecha».

Giuseppe Piccoli, Conegliano

Questo paziente, tuttora vivente ed asintomatico, ha avuto Un arresto cardiaco da fibrillazione ventricolare il 20 ottobre 1984, a 42 anni



LA REPOLARISATION PRECOCE DANS LES PRECORDIALES DROITES : TROUBLE DE LA CONDUCTION INTRAVENTRICULAIRE DROITE ?
CORRELATIONS DE L'ELECTROCARDIOGRAPHIE-VECTOCARDIO-GRAPHIEAVEC L'ELECTROPHYSIOLOGIE

A NAVA, B CANCIANI, M -L SCHIAVINATO, B MARTINI, G BUJA

Dep. de Cardio., Univ de Padoue (Italie)

Adresse : A Nava, Cattedra di Cardiologia, Policlinico università, via Giustiniani 2, 35100 Padova, Italia

MISES A JOUR CARDIOLOGIQUES-17° ANNEE 1988-XVII 5

p 157

1988-89!

Reprinted from AMERICAN HEART JOURNAL, St. Louis Vol. 118, No. 6, pp. 1203-1209, December, 1989, (Printed in the U.S.A.)

Ventricular fibrillation without apparent heart disease: Description of six cases

Since 1977, six patients (five males and one female), aged 14 to 35 years, resuscitated from ventricular fibrillation, were referred to our department for detailed evaluation, after exclusion of major cardiac pathologic conditions. Four patients had a family history of heart disease. Basic ECGs showed sinus rhythm in all of them. PR interval was prolonged in one. Two patients had complete and one had incomplete right bundle branch block. One patient had inverted t waves in V1-2 and late potentials. Three had an upslooping ST-T segment elevation in Vi.2. The cardio thoracic index was less than 0.5 in five and 0.50 in one. In one of the five patients studied, the clinical episode of ventricular fibrillation was reproduced by stimulation of the right ventricular outflow tract during electrophysiologic study. Results of cross-sectional echocardiography and angiography showed predominantly structural and wall motion abnormalities of the right ventricle in five patients and slight wall motion abnormalities of the left ventricle in two. Two patients also had mitral and tricuspid valve prolapse. Coronary arteries were normal in all five patients examined. Results of endomyocardial biopsy showed no abnormalities in one patient, fibrosis in two, and fibrolipomatosis in one. Two patients died during follow-up: autopsy was performed in one and results showed right ventricular cardiomyopathy. Thus in five of these selected patients with apparent idiopathic ventricular fibrillation, some abnormalities, predominantly of the right ventricle, were documented only after detailed investigation; however, clinical history and some nonspecific ECG abnormalities were factors in the diagnostic procedure. (AM HEART J 1989; 118:1203.)

Bortolo Martini, MD, Andrea Nava, MD, Gaetano Thiene, MD, Gian Franco Buja, MD, Bruno Canciani, MD, Roldano Scognamiglio, MD, Luciano Daliento, MD, and Sergio Dalla Volta, MD. *Padua, Italy*

Cosa è l'ECG di Nava?

PR>

HV allungato

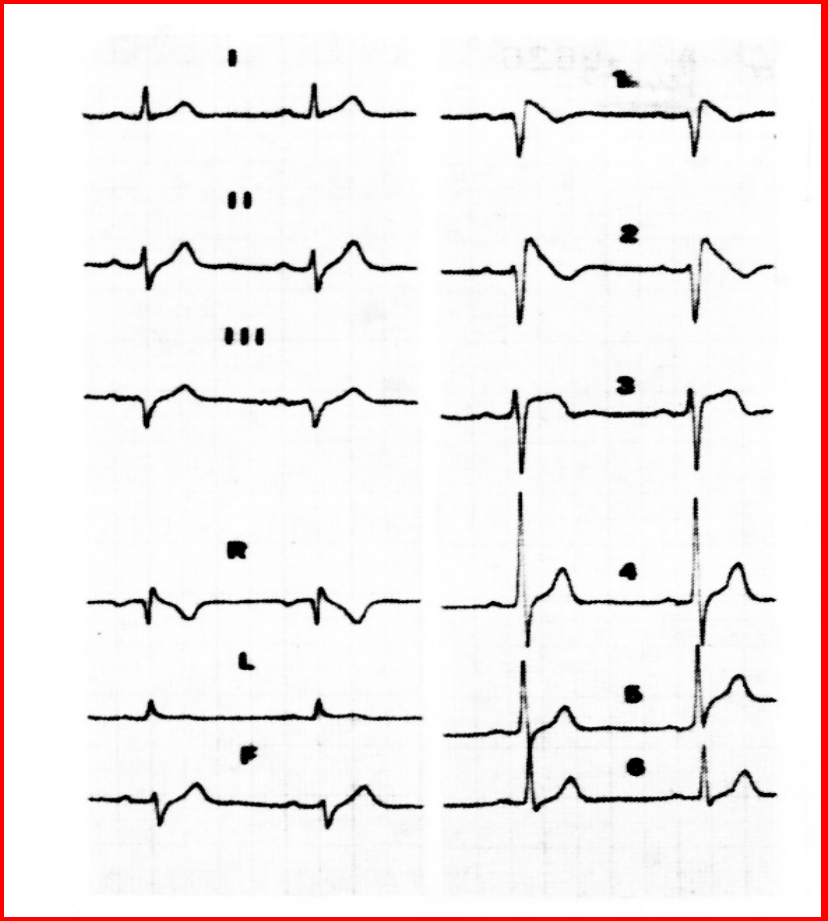
Deviazione assiale sn

T negative in V1-V2

Punto J arrotondato

ST a tenda in V1-V2

Assente o minima s in V6



LETTER TO THE EDITOR

To the Editor: A Long Lasting Electrocardiographic History

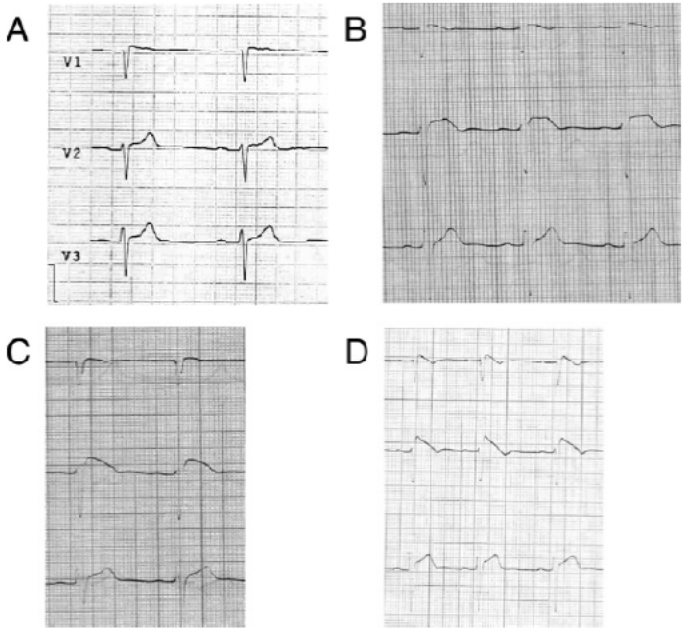


Figure 1 The ECG tracings in Figures 1 and 2 were not taken consecutively but were recorded in different years. **A:** Normal ECG. **B, C:** Dome-shaped ST-segment elevation. These traces were recorded when the patient was taking amiodarone. **D:** Coved ST-segment elevation.

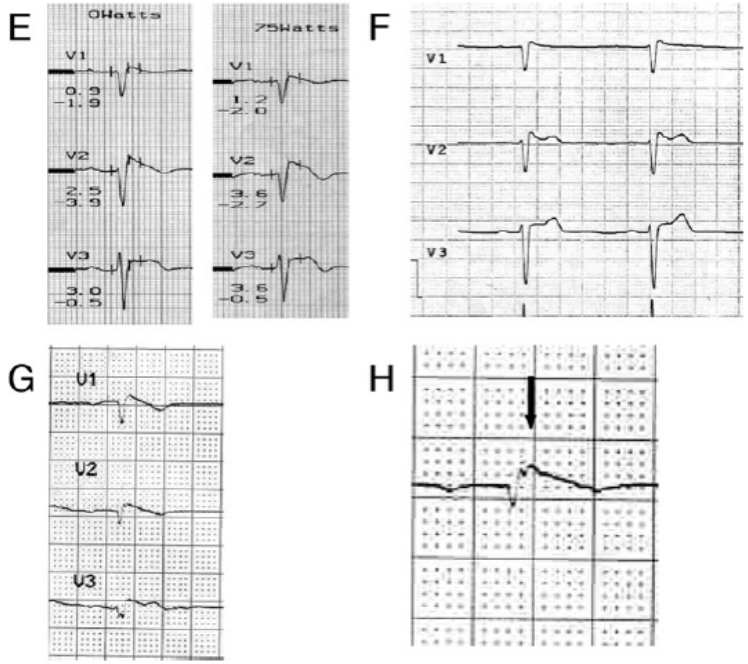
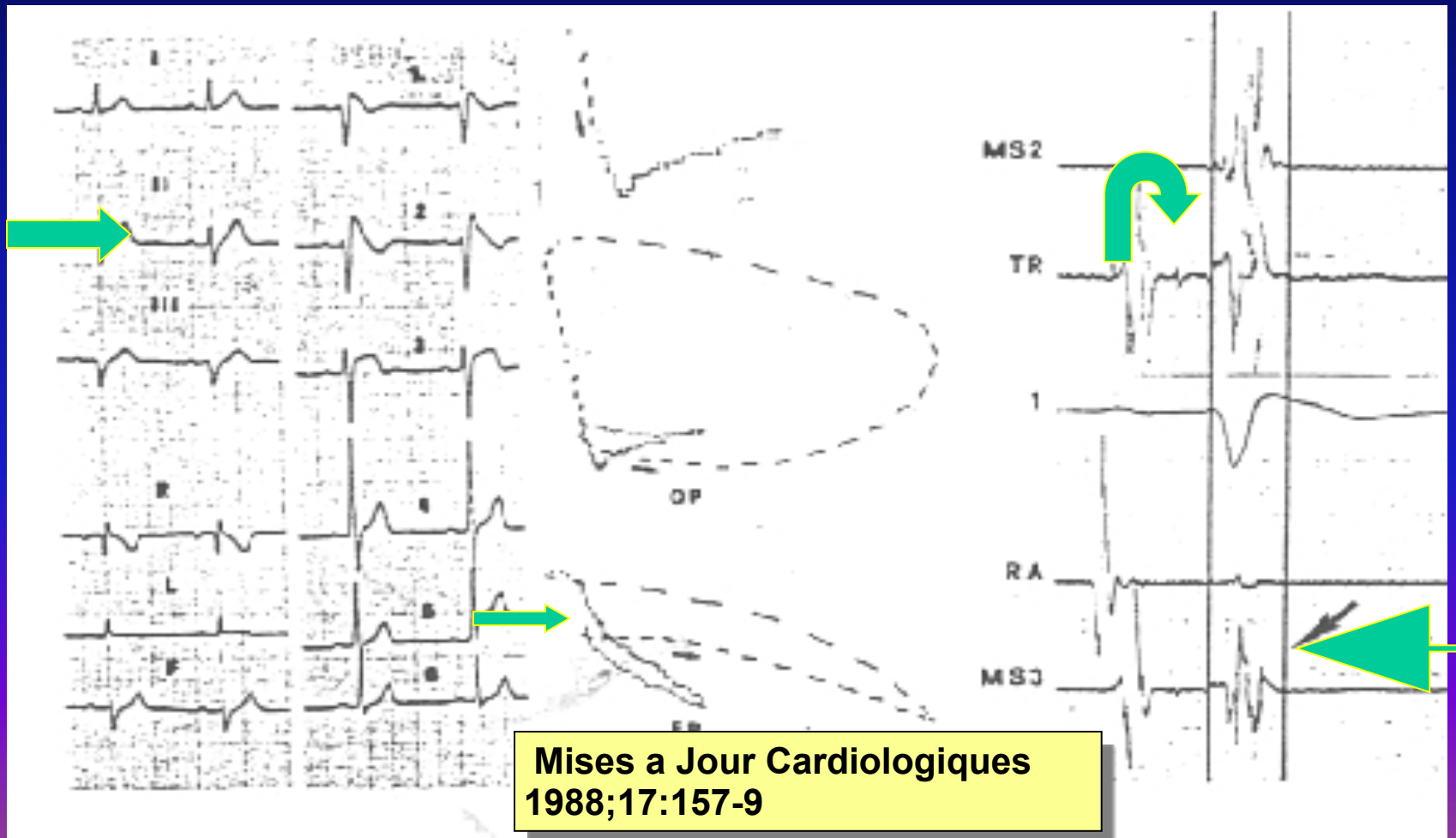


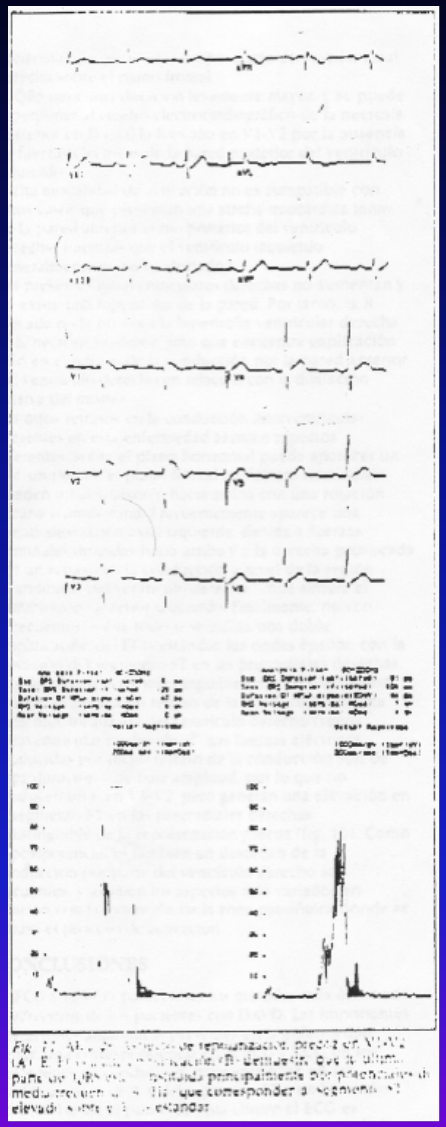
Figure 2 **E:** Coved ST-segment elevation accentuated during stress test. **F:** Saddle-back ST-segment elevation. **G:** Epsilon wave in V2, which is best seen in the larger magnification at **H** (arrow).

Martini B. Heart & Rhythm 2010

ECG, VCG, Endocardial recordings



Mises a Jour Cardiologiques
1988;17:157-9



Presence of late potentials
Confirms ECG, VCG,
Endocavitary ECG suspicion
of
Depolarization Abnormality

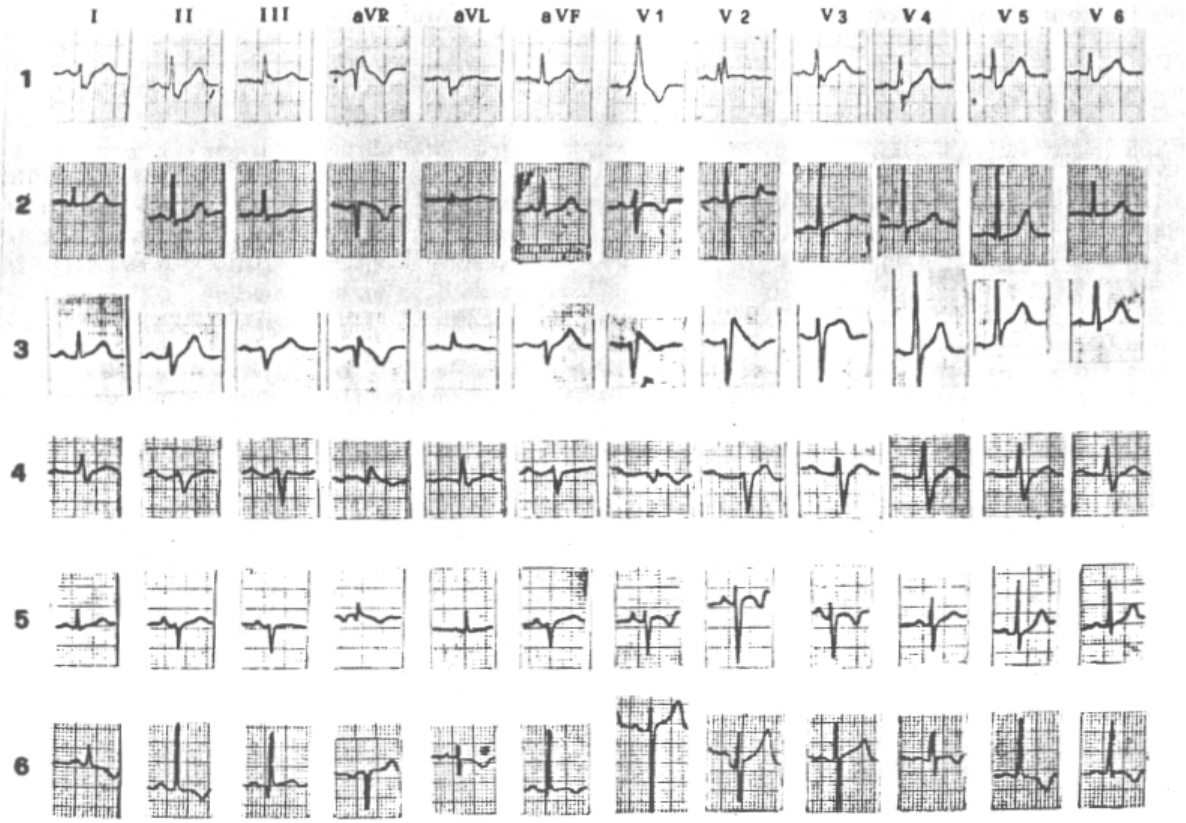
Nava 1992
Arch Cardiol Mex

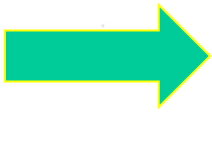
Ventricular fibrillation without apparent heart disease: Description of six cases

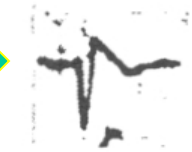
Bortolo Martini, MD, Andrea Nava, MD, Gaetano Thiene, MD, Gian Franco Buja, MD, Bruno Canciani, MD, Roldano Scognamiglio, MD, Luciano Daliento, MD, and Sergio Dalla Volta, MD. *Padua, Italy*

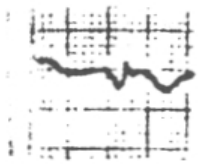
In this article <u>all</u> the ECG pattern of the syndrome were described

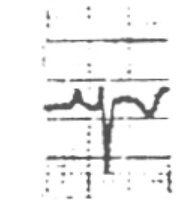
December 1989
American Heart Journal











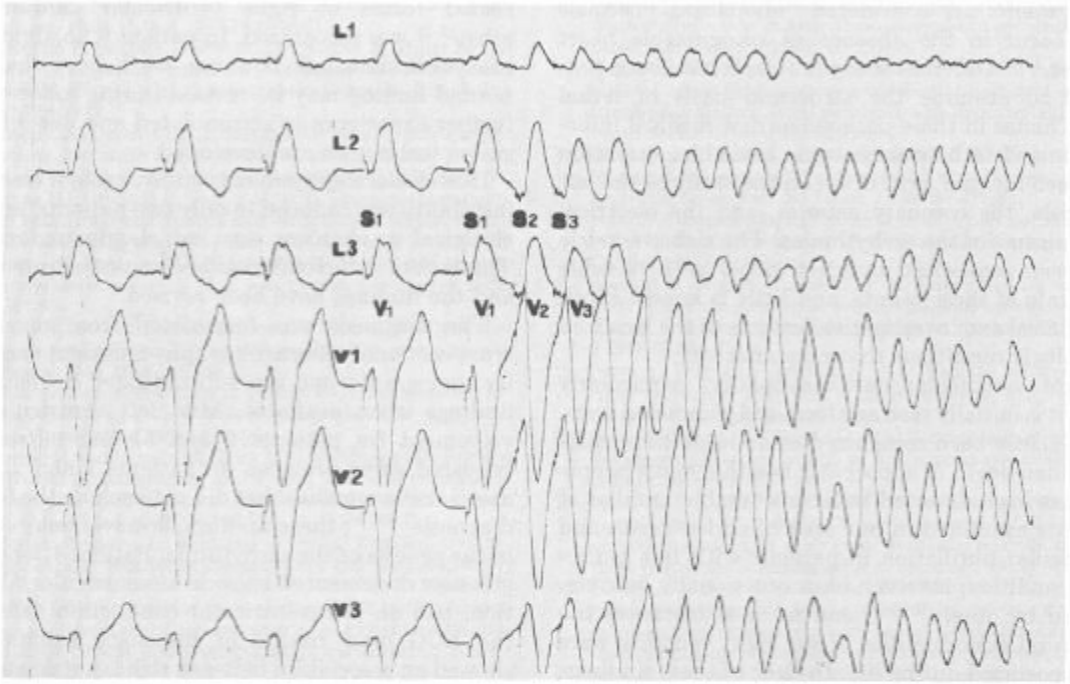


Fig. 2. Electrophysiologic study in patient 1: ventricular tachycardia was induced during pacing at level of pulmonary infundibulum, which degenerated in ventricular fibrillation.

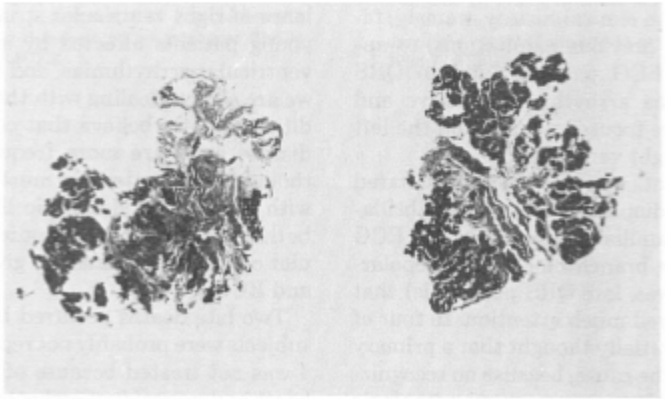


Fig. 3. Endomyocardial biopsy from patient 1: moderate fibrosis is visible in two fragments. (Azan Mallory stain; original magnification ×10.)

Martini 1989
AHJ

Familial Cardiomyopathy Underlies Syndrome of RBBB, ST Segment Elevation and Sudden Death

Corrado D, Nava A, Buja G, Martini B, Thiene G.

JACC 1996;27:443-8

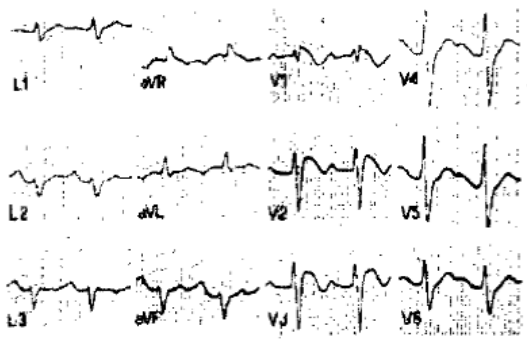


Figure 2. The 12-lead basal electrocardiogram of the proband, recorded nearly 2 years after the episode of aborted sudden death.

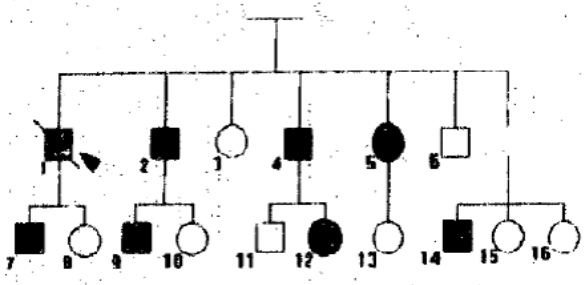
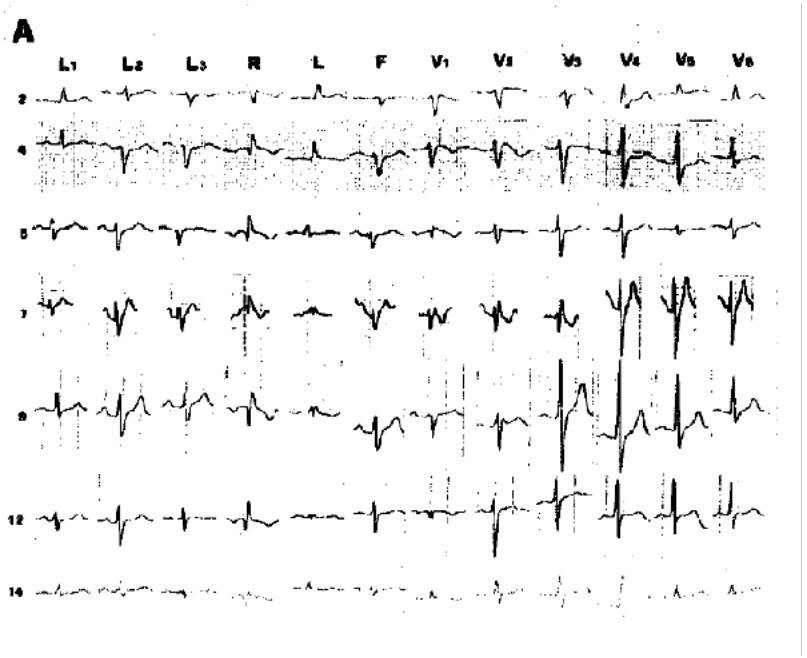
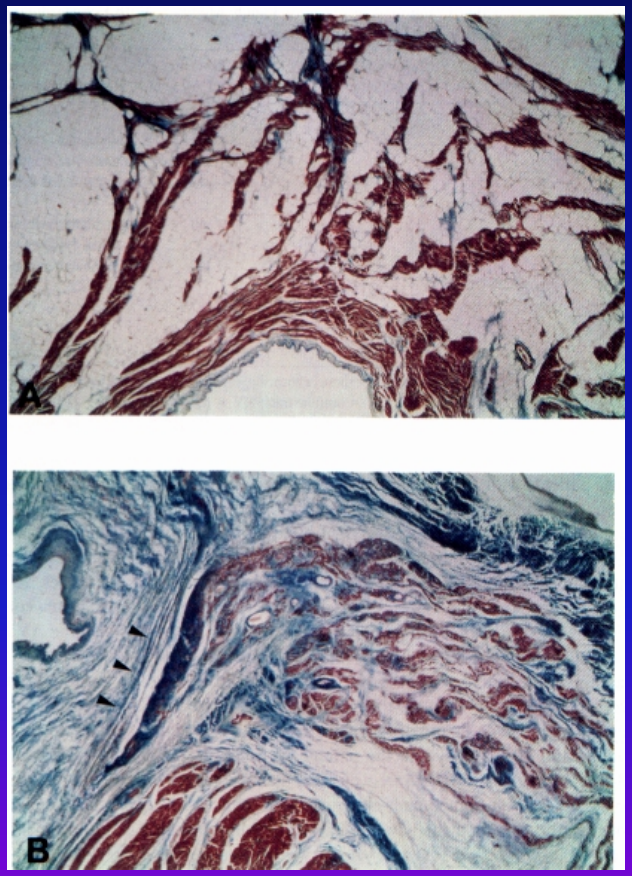


Figure 1. Family pedigree. **Numbers** indicate cases. **Circles** represent women and **squares** represent men. **Arrowhead** indicates the proband. **Diagonal bars** indicate deceased family members. Affected members are represented by **solid circles** and **squares**. Nonaffected and noninvestigated members are represented by **open** and **gray symbols**, respectively.

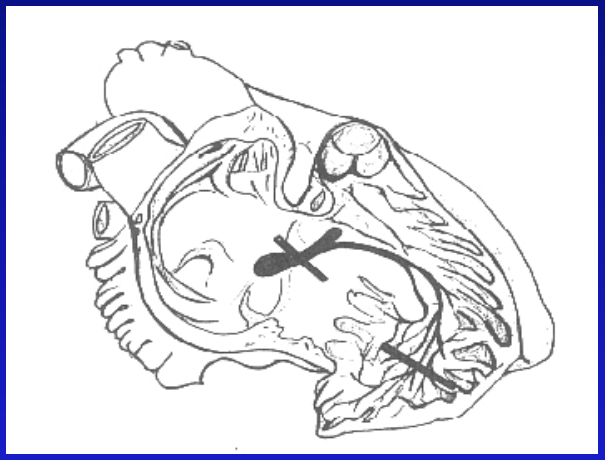


Syndrome of right bundle branch block, ST Segment elevation and sudden death: evidence of an organic substrate

Bortolo Martini, Domenico Corrado, Andrea Nava and Gaetano Thiene. 1997

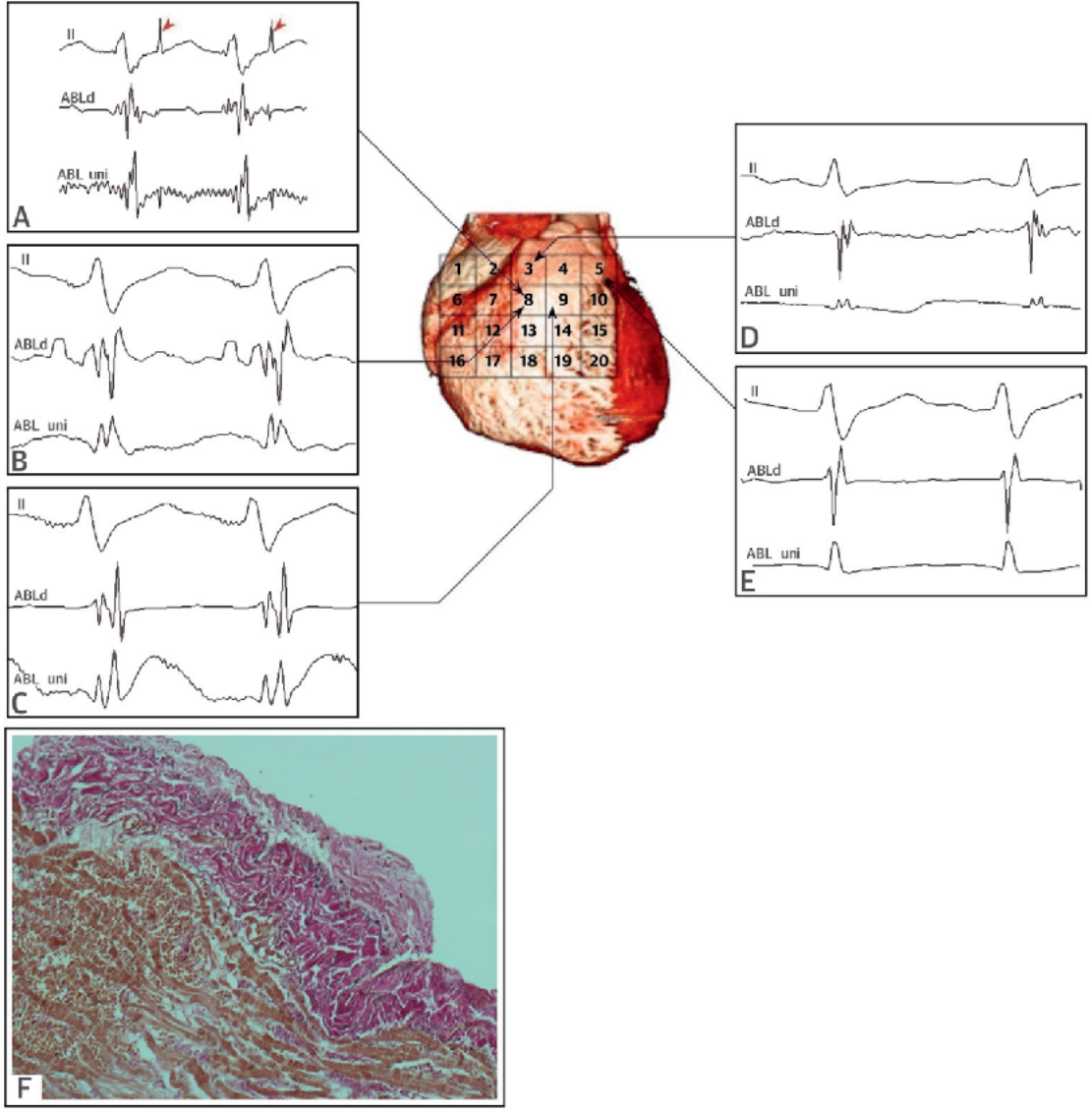


- **A: atrophy, fibrosis, adiposis of the RVFW**



- **B: severe fibrosis of the bifurcating His bundle with sclerotic interruption of right bundle branch**

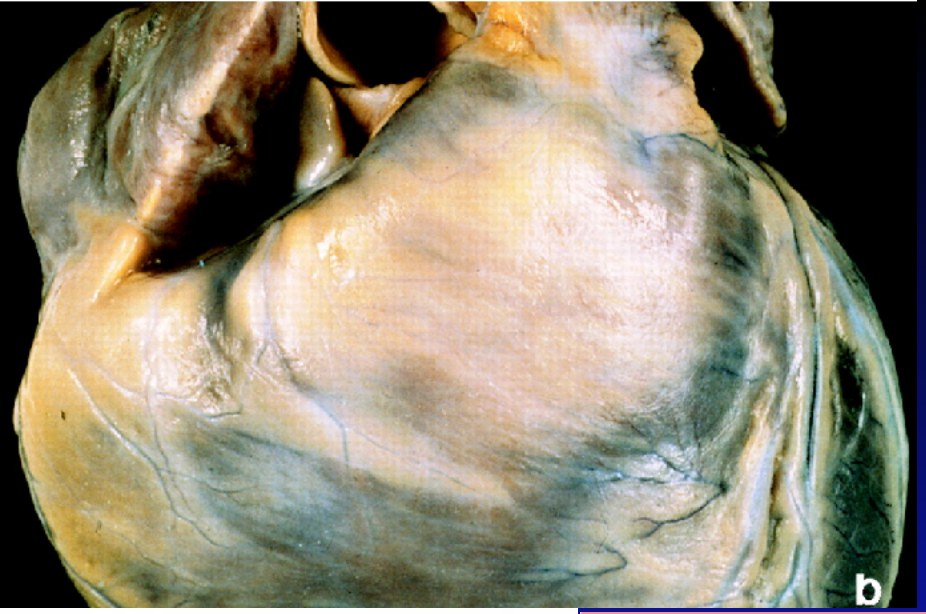
Fibrosis, Connexin-43, and Conduction Abnormalities in the Brugada Syndrome.
Nademanee- Wilde Jacc 2015



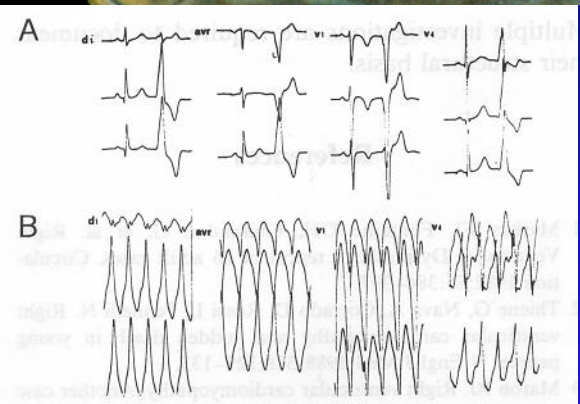
Computed tomography scan of the heart (center) of in vivo BrS patient V2 showing an anatomical grid over the anterior RVOT. ECG lead II and a distal bipolar (0.4 mV/cm voltage scale at 30- to 300-Hz filter settings) and unipolar (5 mV/cm voltage scale at 0.05- to 300-Hz filter settings) electrogram at labeled sites are given in surrounding panels, with pacing stimuli indicated by red arrowheads. Abnormal fractionated electrograms are on the (A to C) left and normal electrograms on the (D to E) right. (F) Epicardial biopsy and histology (PSR) at the site of the abnormal electrogram shows epicardial fibrosis with focal finger-like projections of collagen into myocardium. ABL d ¼ distal bipolar ablation catheter electrogram; ABL uni ¼ unipolar ablation catheter electrogram; BrS ¼ Brugada syndrome; RVOT ¼ right ventricular outflow tract; other abbreviations as in Figure 1.

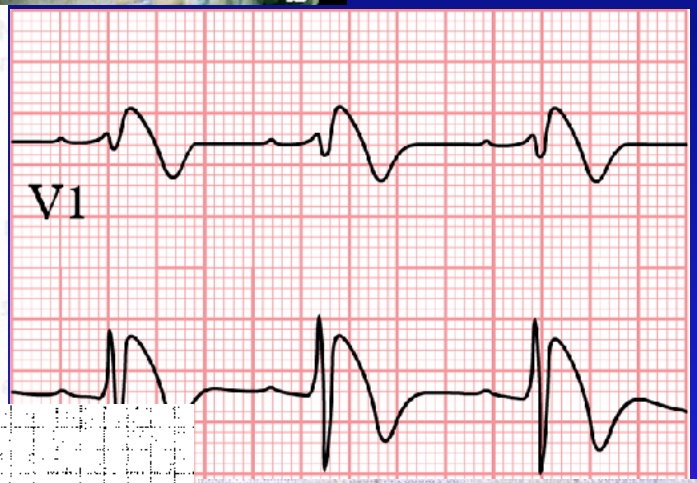
Perchè

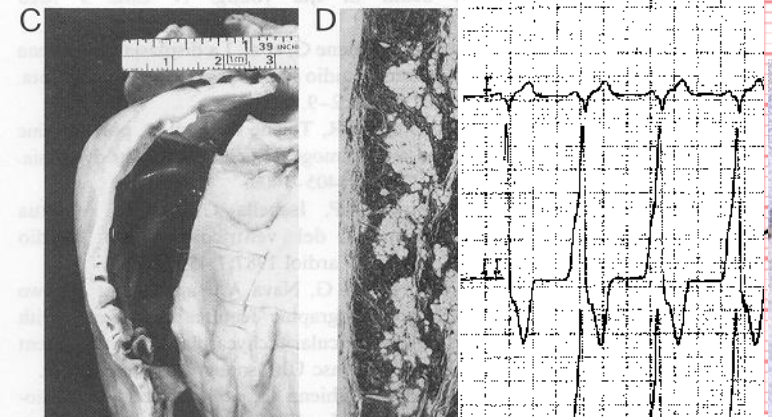
- **Il tratto del flusso del ventricolo destro è la madre di tutte le aritmie idiopatiche?**



La madre delle aritmie idiopatich







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

Objectives. The objectives of this study were to present data on eight patients with recurrent episodes of aborted sudden death unexplainable by currently known diseases whose common clinical and electrocardiographic (ECG) features define them as having a distinct syndrome different from idiopathic ventricular fibrillation.

Background. Among patients with ventricular arrhythmias who have no structural heart disease, several subgroups have been defined. The present patients constitute an additional subgroup with these findings.

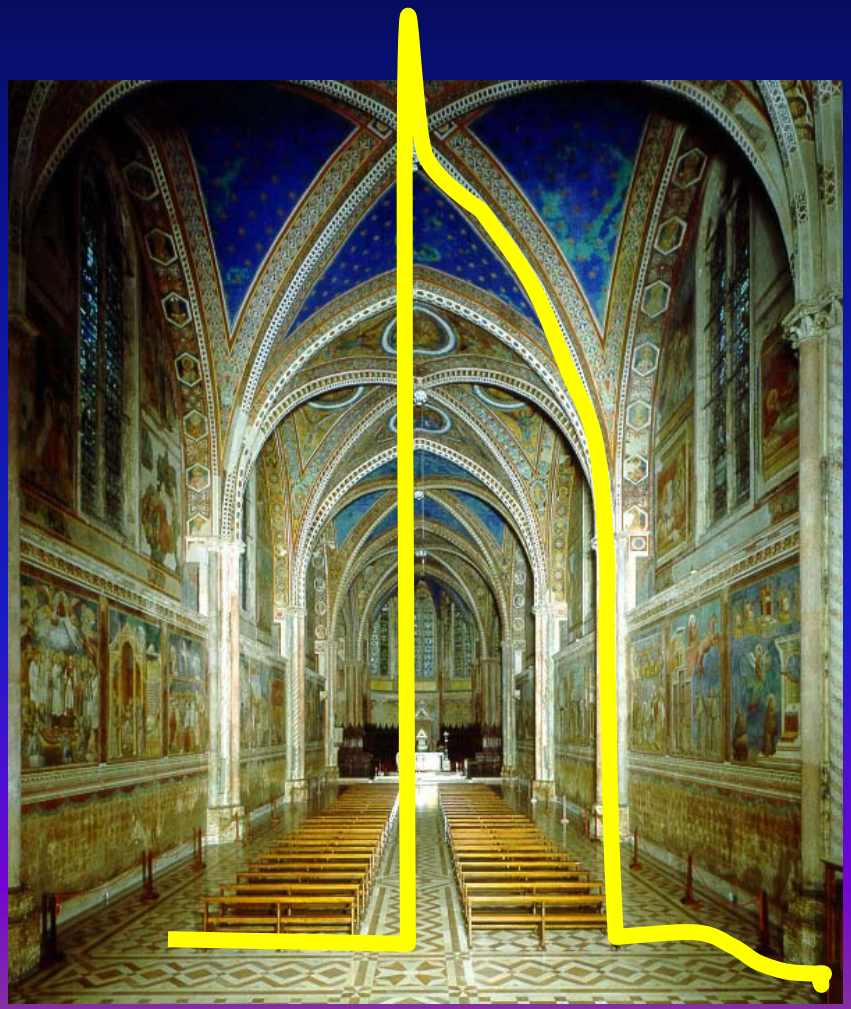
Methods. The study group consisted of eight patients, six male and two female, with recurrent episodes of aborted sudden death. Clinical and laboratory data and results of electrocardiography, electrophysiology, echocardiography, angiography, histologic study and exercise testing were available in most cases.

Results. The ECG during sinus rhythm showed right bundle branch block, normal QT interval and persistent ST segment elevation in precordial leads V_1 to V_2–V_3 not explainable by electrolyte disturbances, ischemia or structural heart disease. No histologic abnormalities were found in the four patients in whom ventricular biopsies were performed. The arrhythmia leading to (aborted) sudden death was a rapid polymorphic ventricular tachycardia initiating after a short coupled ventricular extrasystole. A similar arrhythmia was initiated by two to three ventricular extrastimuli in four of the seven patients studied by programmed electrical stimulation. Four patients had a prolonged HV interval during sinus rhythm. One patient receiving amiodarone died suddenly during implantation of a demand ventricular pacemaker. The arrhythmia of two patients was controlled with a beta-adrenergic blocking agent. Four patients received an implantable defibrillator that was subsequently used by one of them, and all four are alive. The remaining patient received a demand ventricular pacemaker and his arrhythmia is controlled with amiodarone and diphenylhydantoin.

Conclusions. Common clinical and ECG features define a distinct syndrome in this group of patients. Its causes remain unknown.

(J Am Coll Cardiol 1992;20:1391–6)

La sacra sindrome «dei Brugada»?



Stephen M. Stigler is Professor of Statistics at the University of Chicago.

Stigler's law

- no scientific discovery is named after its original discoverer.

THE CLEVELAND CLINIC FOUNDATION

LERNER RESEARCH INSTITUTE

April 30, 2000

Bortolo Martini, M.D.

Department of Cardiology
Ospedale Civile di Thiene
Thiene
Italy

Dear Dr. Martini:

I read a commentary article from you in *Circulation*. This is how I know your address. I am the senior and corresponding author on the *Nature* paper (Genetic Basis and Molecular Mechanism for idiopathic ventricular fibrillation) (please see the enclosed reprint).

As you know, many people now call idiopathic ventricular fibrillation with right bundle branch block and ST-segment elevation as Brugada syndrome. In as I understand, you were the first to describe this syndrome. The disease should be at least Martini-Brugada syndrome or just Martini syndrome. But, sometimes politics does penetrates basic science.

I have moved my laboratory from Baylor to the Cleveland Clinic Foundation. My wife, Qiuyun Chen, Ph.D., who was the first author on the Nature paper, has also moved to Cleveland. My laboratory is continuing to look for new genes and mutations for IVF with STE. Dr. Glenn Kirsch, who is the co-first author on the Nature paper is also in Cleveland, and we are continuing our long-time collaboration. So, we have a very strong team at Cleveland for genetics of cardiovascular disease. By the way, the Cleveland Clinic Foundation has been ranked FIRST for Cardiology five years in a row in the USA.

I would like to invite you, Dr. Gaetano Thiene, and your other colleagues to join our team on this project. If you have families and patients with idiopathic VT or VF, please send blood samples to us for genetic research. Needless to say, I will include you and your colleagues as co-authors on publications from this collaboration.

If you have any questions, please feel free to contact me.

Yours sincerely,

Qing Wang, Ph.D.
Assistant Staff of Molecular Genetics, Molecular Cardiology, and Cardiology

1-216-445-0570 (Office)
1-216-444-2682 (fax)
wangq2@ccf.org (e-mail)

9500 Euclid Avenue, Cleveland, Ohio 44195

Sfortunatamente
La politica ha invaso
La Medicina

Scoperte Italiane e nomi Stranieri in Aritmologia

Malattia	Scopritore	Nome corrente
Sincope da BAVC	Mercuriale, Morgagni	Stokes-Adam
Fascio Accessorio	Paladino	Kent bundle
BAV di grado 2	Luciani	Wenckebach Mobitz 2
LQTS	Romano	Ward-Romano
RBBB+ST+ Morte Improvvisa	Nava-Martini-Thiene	Brugada

CARDIOVASCULAR CENTER AALST

Prof. Dr. Pedro Brugada

Aalst, March 18, 1999

Dear Doctor Martini,

Thank you very much for your recent letter and for sending me a reprint of your article which was published in the Am Heart J 1989;1118:1203-1209

In our publication from 1992, we did not refer to your report because we were not aware of it. Lateron, when we became aware of your publication we did not include it in our group because you state very clearly that your patients had right ventricular dysplasia.

With best personal regards,

Prof. Dr. Pedro Brugada

«nel 1992 non ci eravamo
Accorti della vostra scoperta
Del 1989»

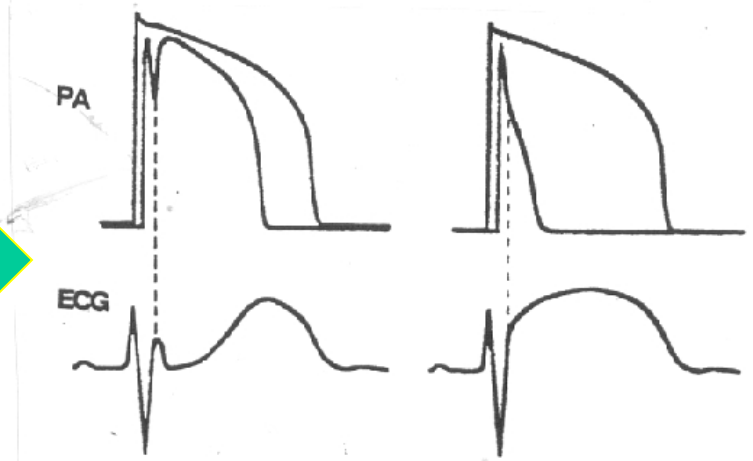
Published Functional Theories

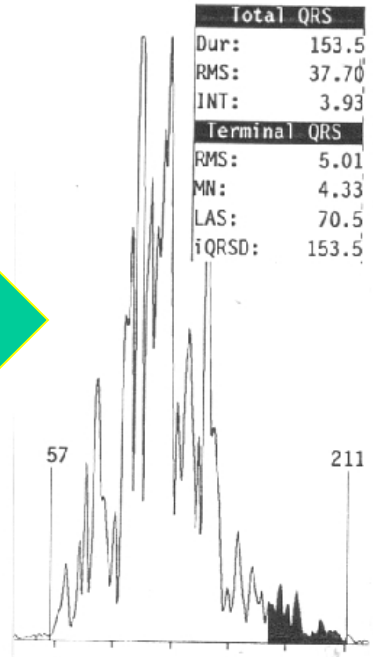
- 1992: “prolonged HV suggest His-Purkinje disease”. “Marked dispersion of refractory periods or extreme anisotropic conduction
- 1994: disorder related to “M cells”
- 1996: IT0 channels involvement
- 1998 mutations of SCN5A genes inducing eterogenicity in epicardial and endocardial AP in 50% of pts with the ECG
- ***The available data suggest that the B.S is a familial primary electrical disease caused by a defect in an ion channel gene, resulting in premature repolarization of some right ventricular epicardial sites.***
- 2002 “the morphological abnormalityes are secondary to electrical conduction defect and abnormal repolarization”
- 2002 “loss of the action potential dome,because it creates a hibernation-like state, may over long periods of time lead to mild morphological changes, which include lipid accumulation and fibrosis

What is the pathopysiology of these ECGs?? Two theories

? ? ?

functional repolarization abnormality





organic depolarization abnormality

Repolarization or Depolarization abnormality?

- «However we hope that Dr Martini no longer ignore the existence of a functional disorder underlying the syndrome of right bundle branch block, right precordial ST-segment elevation, and sudden death.
- Arthur Wilde, Circulation 1999

- Tukkie R, Sogaard P, Vleugels J, De Groot I, Wilde AM, Tan H.:
- Delay in Right Ventricular Activation Contributes to Brugada Syndrome. Circulation 2004;109:1272-1277.)

JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY
© 2016 BY THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION
PUBLISHED BY ELSEVIER

VOL. 67, NO. 1, 2016
ISSN 0735-1097/$36.00
http://dx.doi.org/10.1016/j.jacc.2015.10.020

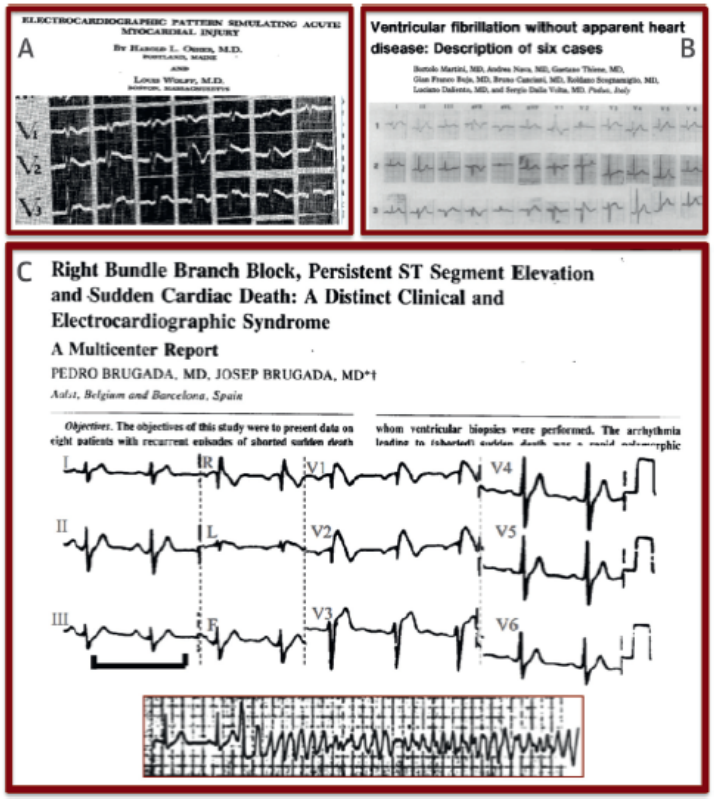
REVIEW TOPIC OF THE WEEK

A Tale of 2 Diseases

The History of Long-QT Syndrome and Brugada Syndrome

Ofer Havakuk, MD, Sami Viskin, MD

Initial Reports of Brugada Syndrome



(A) First presentation of electrocardiograms nowadays defined as type I Brugada syndrome in 3 ostensibly healthy men with a pattern "resembling myocardial infarction," followed for years by Osher and published in 1953 (49). **(B)** Presentation of 6 patients with idiopathic ventricular fibrillation, including 1 with "type-I Brugada-like pattern," by Martini et al. (50). **(C)** Description of 8 patients with "right bundle branch block, persistent ST-segment elevation and sudden death" by Pedro and Josep Brugada in 1992 (48), an entity soon to become recognized as Brugada syndrome.

La sindrome «di Brugada»
Sta crollando la «cupola»?

- **Altri aspetti ECG**

Dinamic ST behaviour

30-50% of pts with ths syndrome. How many healthy subject with the ecg???

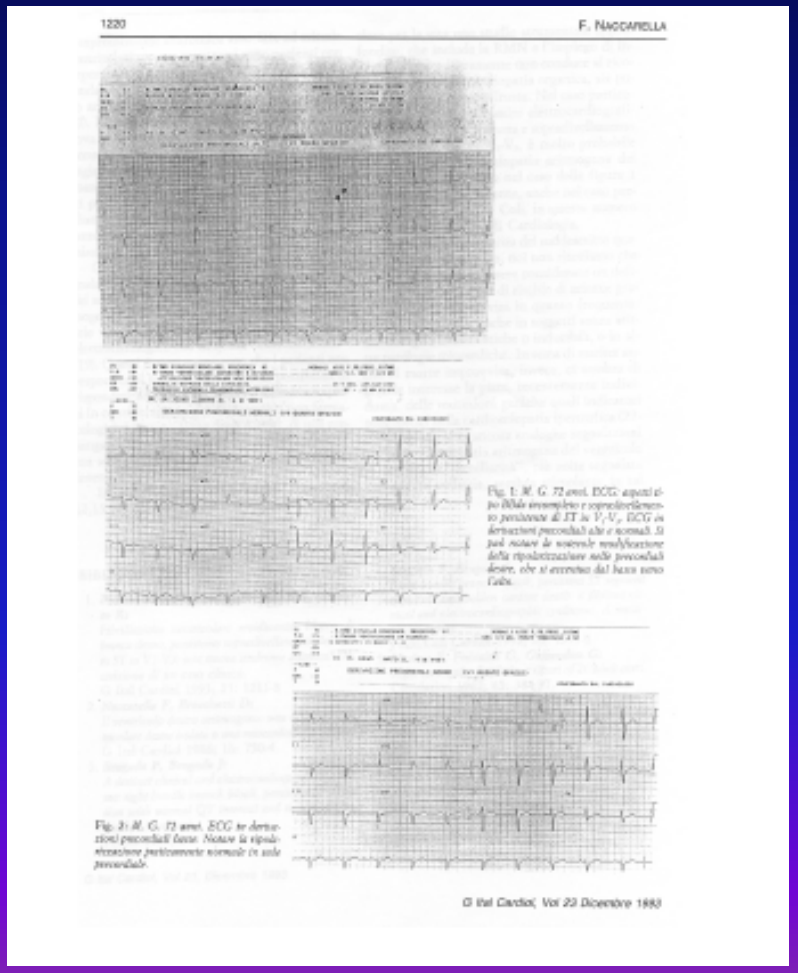
- **GII SPAZI INTERCOSTALI**

COMMENTO EDITORIALE - EDITORIAL COMMENT

G Ital Cardiol 1993; 23: 1219-1222

ARITMIE VENTRICOLARI MALIGNE IN PAZIENTI CON BLOCCO DI BRANCA DESTRA E PERSISTENTE SOPRASLIVELLAMENTO DI ST IN V_1-V_3: PROBABILE CARDIOMIOPATIA ARITMOGENA DEL VENTRICOLO DESTRO

FRANCO NACCARELLA



Prima dimostrazione che
Il tipico ecg può essere
Registrato nelle precordiali
Alte.

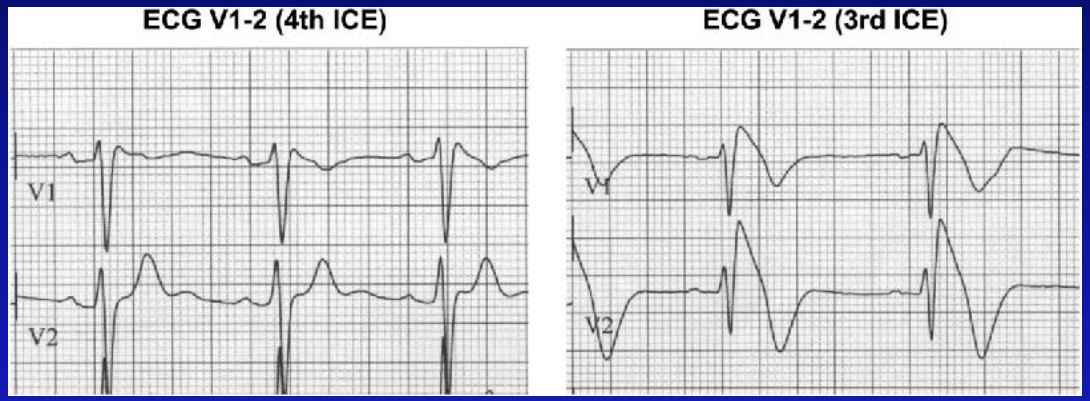
Brugada Syndrome 2012

Paola Berne, MD; Josep Brugada, MD, PhD

Figure 3.

(Left) Basal ECG shows a suggestive but not diagnostic ECG pattern. When V1 and V2 are placed in the 3rd intercostal

space (ICE; Right), a type 1 BS pattern (diagnostic) is observed.

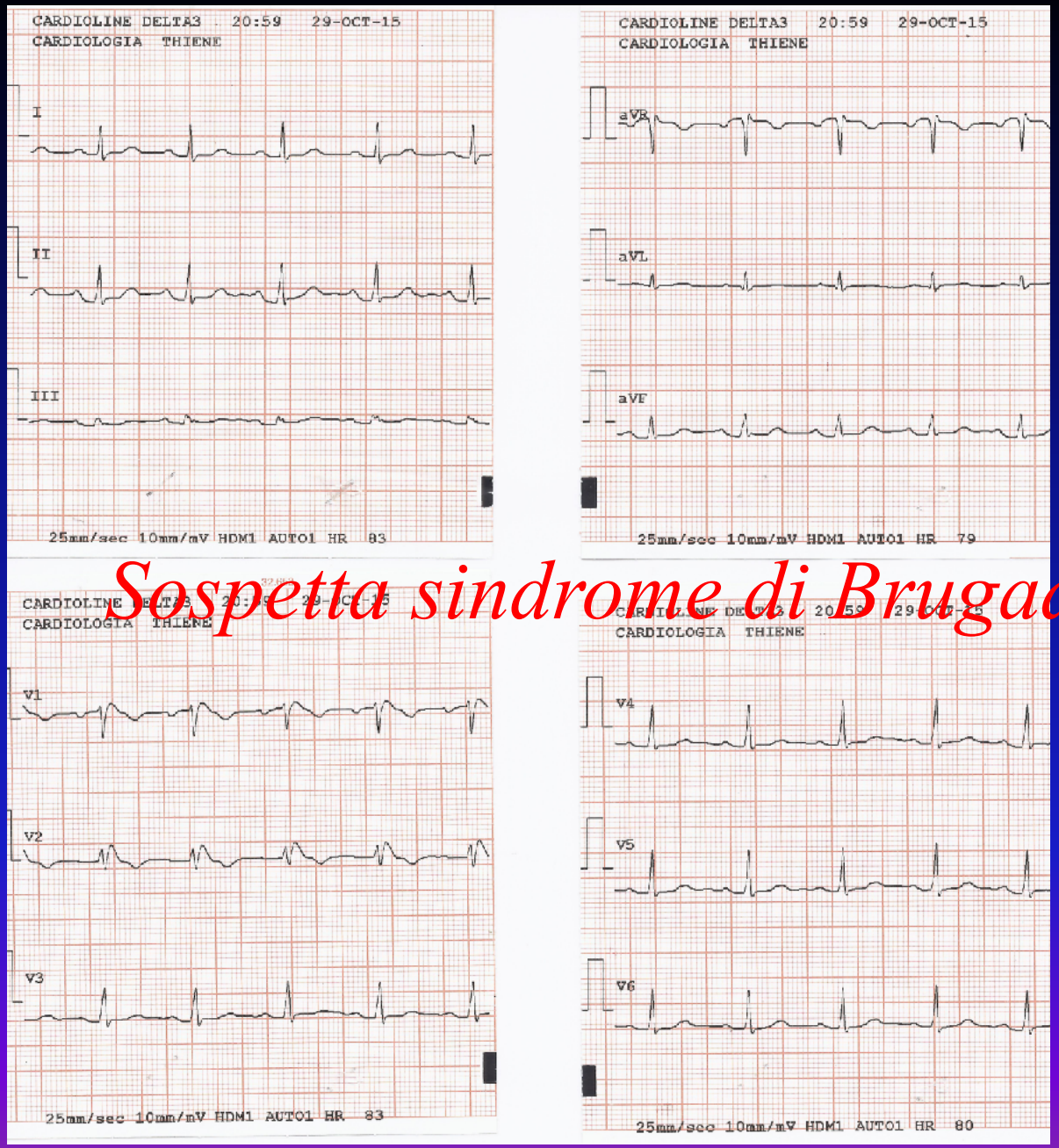


Clinical presentation and outcome of Brugada syndrome diagnosed with the new 2013 criteria (Curcio, Priori JCE 2016)

- Take home message!
- **Su 300 (loro) soggetti con minime anomalie di base 4 hanno tipo 1 nelle precodiali alte spontaneamente e 60 dopo ajmalina!!**
- **CONCLUSION: This study demonstrates that the use of new diagnostic criteria for BrS allows increasing the diagnostic yield by 20% and that the arrhythmic risk is low when BrS can be established only in High-ICS.**

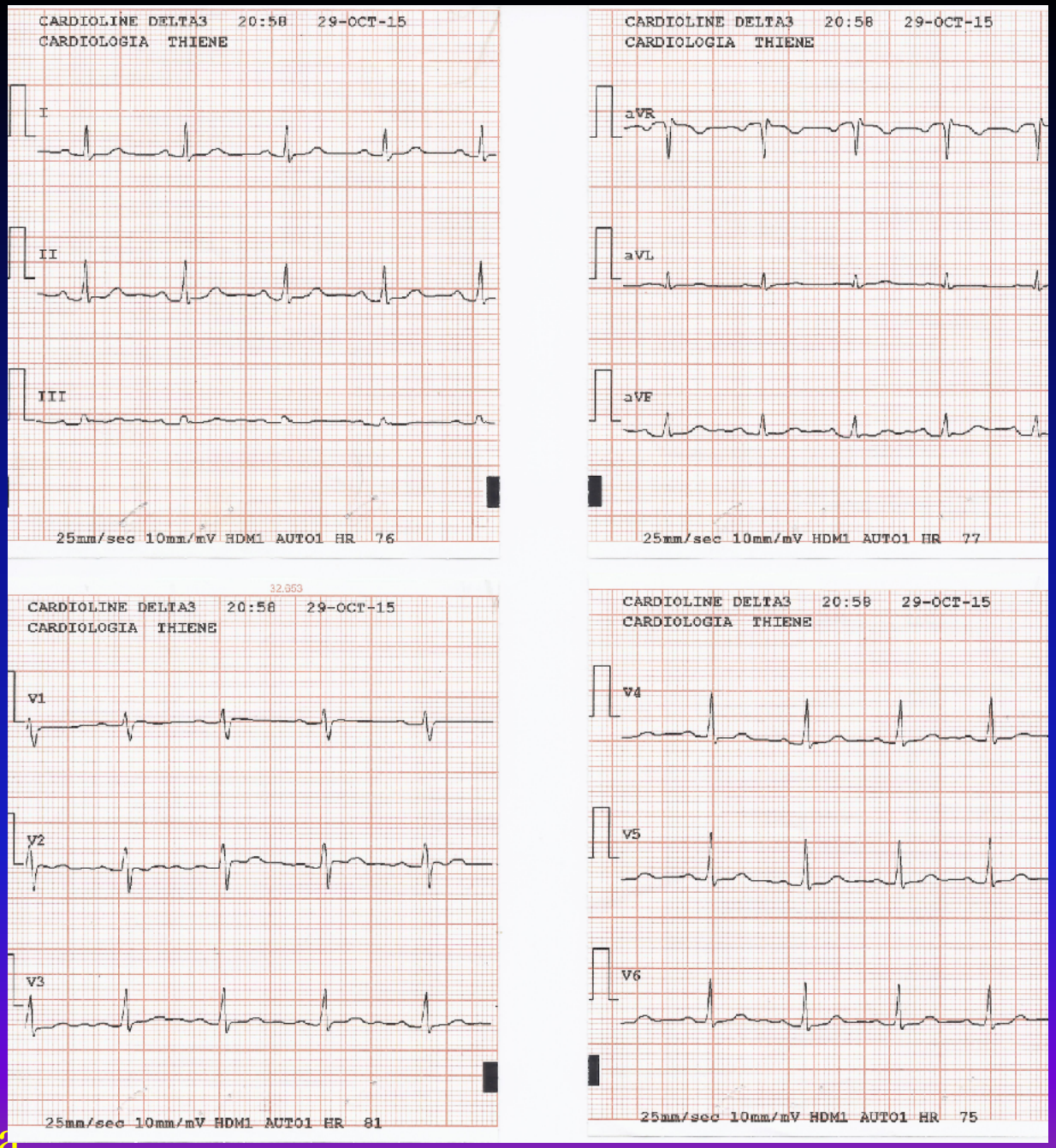
Gasiero D. 29 anni, femmina

- Asintomatica. Reperto occasionale.
- Nessuna familiarità
- Holter negativo
- Eco normale
- Diagnosi: «sindrome di Brugada»!!
- Consiglio terapeutico: lista di farmaci da evitare in particolare con febbre



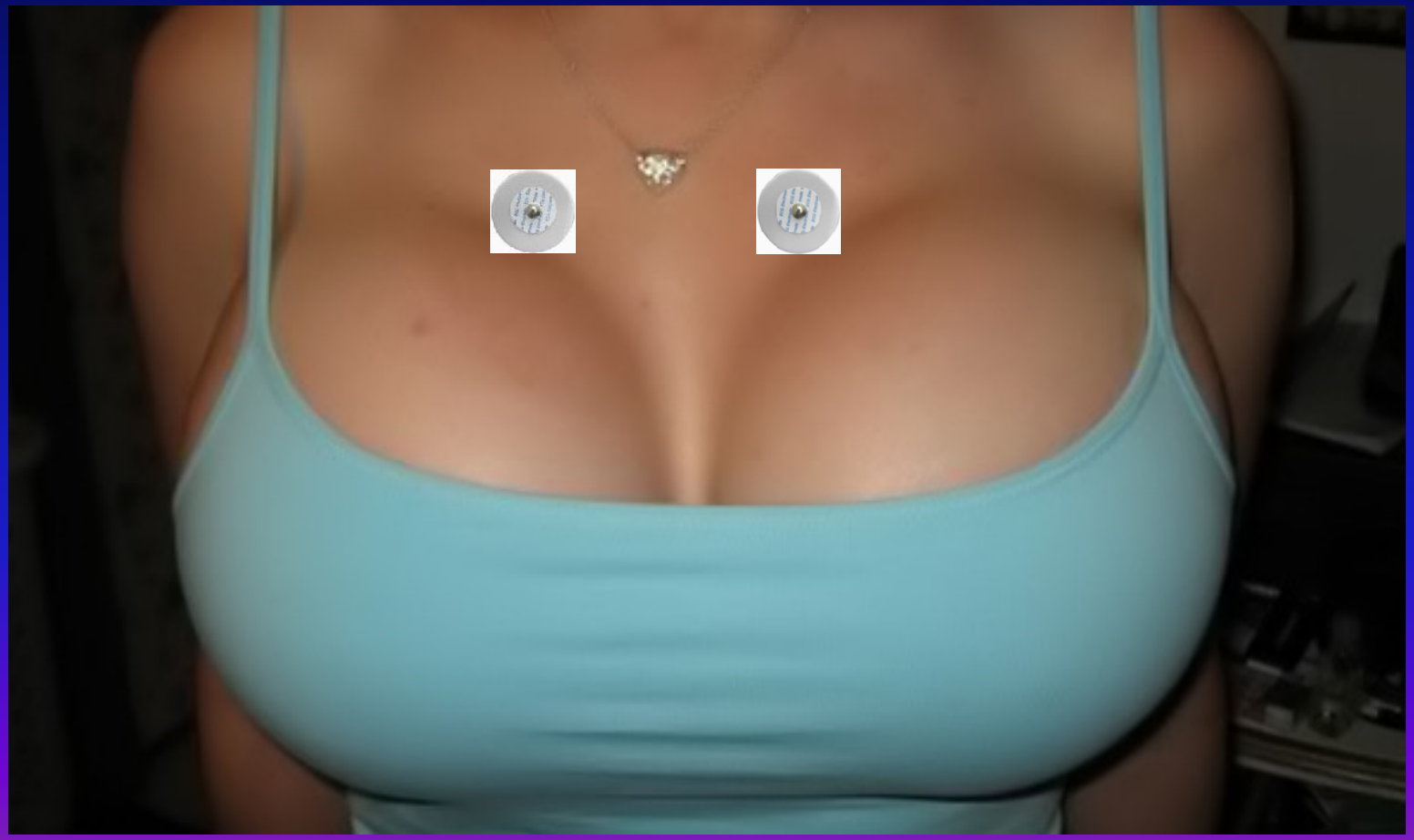
Sospetta sindrome di Brugada!

Gasiero
D. ♀
29 anni



Gasiero D
29 yof
Nuova visita

Causa della sindrome dopo 2 mesi di panico: elettrodi pudicamente messi in secondo spazio!



Drug challenge!

- **AJMALINE**

1219 Ajmaline unmasks apparent right bundle branch block and ST segment elevation in V1–V3 in patients with 'idiopathic ventricular fibrillation'

J. Brugada, P. Brugada [1], R. Brugada [2]. *Arrhythmia Unit, Hospital Clinic, University of Barcelona, Spain;* [1] *Cardiovascular Center, OLV Hospital, Aalst, Belgium;* [2] *Baylor College of Medicine, Houston, TX, USA*

It has been described that an electrocardiographic (ECG) pattern of apparent Right Bundle Branch Block (RBBB) and ST segment elevation in leads V1 to V3 is associated with sudden cardiac death (SD) in patients (pts) without demonstrable structural heart disease. It has also been described that these ECG abnormalities can transiently normalize during follow-up. To study whether these ECG abnormalities can be unmasked by pharmacological interventions, we studied the effects of a single dose of i.v. ajmaline in the ECG of pts with the described syndrome and transient normalization of the ECG and we compared it with the effects on pts previously classified as "idiopathic ventricular fibrillation" and with normals.

Methods: A single i.v. dose of 1 mgr/kg of ajmaline was administered during 5 min to: Group A) 5 pts with the RBBB + ST elevation + SD during transient normalization of the ECG; Group B) 7 pts previously classified as "idiopathic ventricular fibrillation", and; Group C) 10 control pts with no history of syncope or SD and no structural heart disease.

Results: Ajmaline administration reproduced the ECG abnormalities previously observed but now transiently normalized in all pts in group A. Ajmaline administration produced a similar abnormal ECG pattern in 4 out of the 7 pts in group B with "idiopathic ventricular fibrillation". In none of the 10 control pts in group C this pattern was observed after ajmaline administration.

Conclusions: Ajmaline administration is a simple tool to unmask the RBBB + ST segment elevation in pts previously diagnosed as idiopathic ventricular fibrillation. Pending confirmation in a larger study group, these data suggest that some of the pts previously diagnosed as "idiopathic ventricular fibrillation" might suffer the RBBB + ST segment elevation + SD syndrome.

1996, PACE, Eur H J

Martini 2000

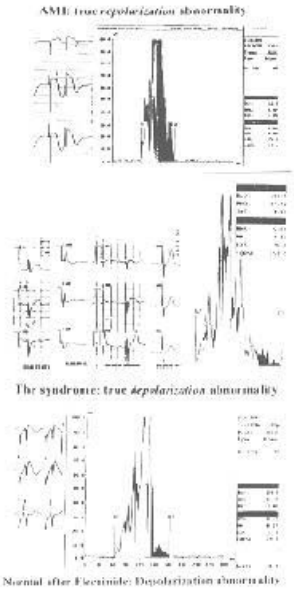


Figure 4: *The same ECG patttern : j wave and coved ST, in three different pts. In the upper trace, a pt with acute myocardial infarction do not show late potentials, as the st elevation is a true repolarization abnormality. In the middle trace a pt with the syndrome shows important late potentials, definitely consistent with depolarization abnormality, and conduction disturbance. The same in a pt. with the st elevation induced by flecainide, in the lower trace.*

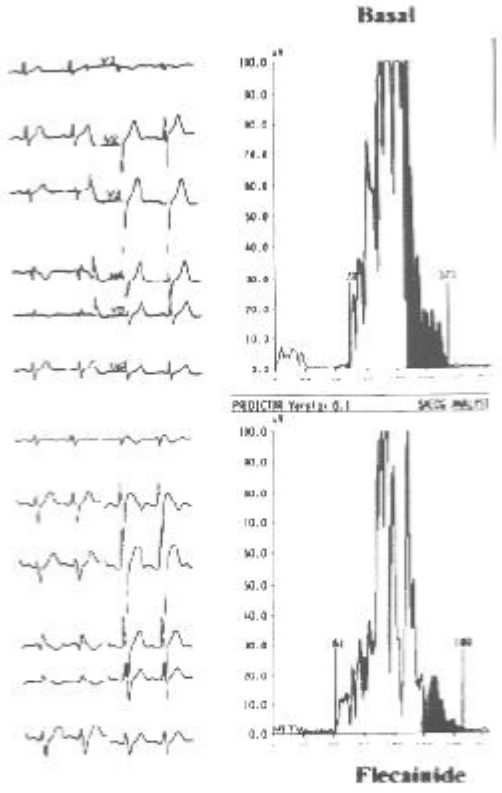


Figure 5: *Normal subject belonging to a family of sudden death and autoptical demonstration of right ventricular cardiomyopathy. Flecainide induces in this pt the typical ECG pattern, which is definitely related to abnormal late potentials, clearly consistent with depolarization abnormality, and conduction disturbance.*

Brugada Syndrome

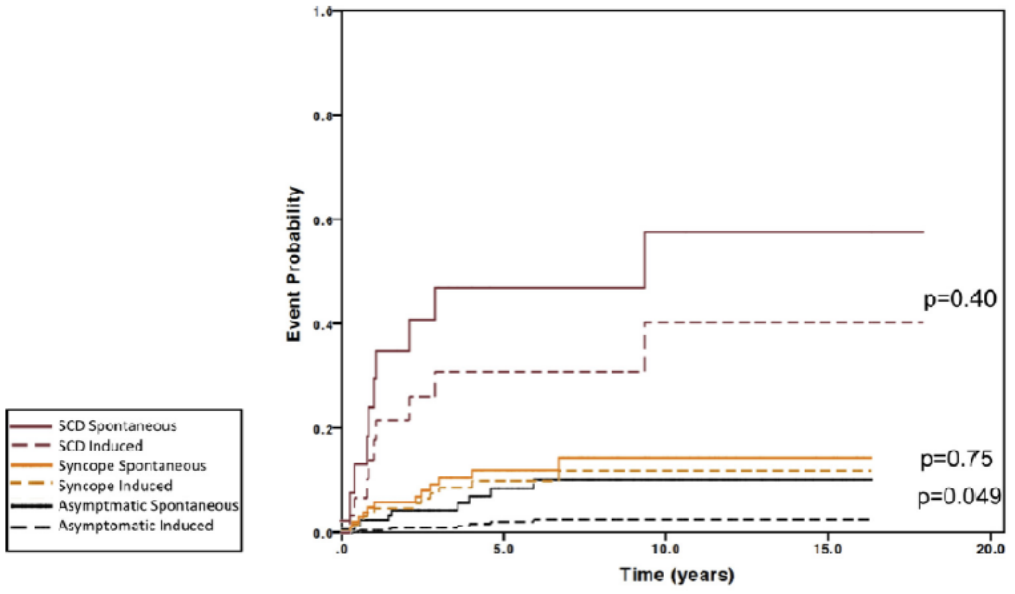
CHARLES ANTZELEVITCH, PH.D.

PACE 2006

- La specificità dei farmaci 1c nell'identificare la sindrome è incerta. La conversione indotta da farmaci da tipo 3 a tipo 1 è inconclusiva per la diagnosi di sindrome

Long-term prognosis of drug-induced Brugada syndrome

Juan Sieira 2017



Asymptomatic	Spontaneous	44	28	13	3
	Drug induced	244	109	51	4
Syncope	Spontaneous	26	18	9	4
	Drug induced	86	32	12	2
SCD	Spontaneous	8	2	1	1
	Drug induced	13	2	2	1

Figure 2 Risk of events by electrocardiographic pattern and symptoms at presentation. SCD = sudden cardiac death.

Test all'ajmalina?

- Un'altra bufala?

- **Alla spasmodica ricerca del «tipico» ECG.**
- **L'ultima figurina!!!**

Table 1 ST segment abnormalities in leads V_1–V_3

	Type 1	Type 2	Type 3
J-wave amplitude	≥2 mm	≥2 mm	≥2 mm
T-wave	Negative	Positive or biphasic	Positive
ST-T configuration	Coved type	Saddle back	Saddle back
ST segment (terminal portion)	Gradually descending	Elevated ≥1 mm	Elevated <1 mm

1 mm = 0·1 mV, the terminal portion of the ST-segment refers to the latter half of the ST-segment.

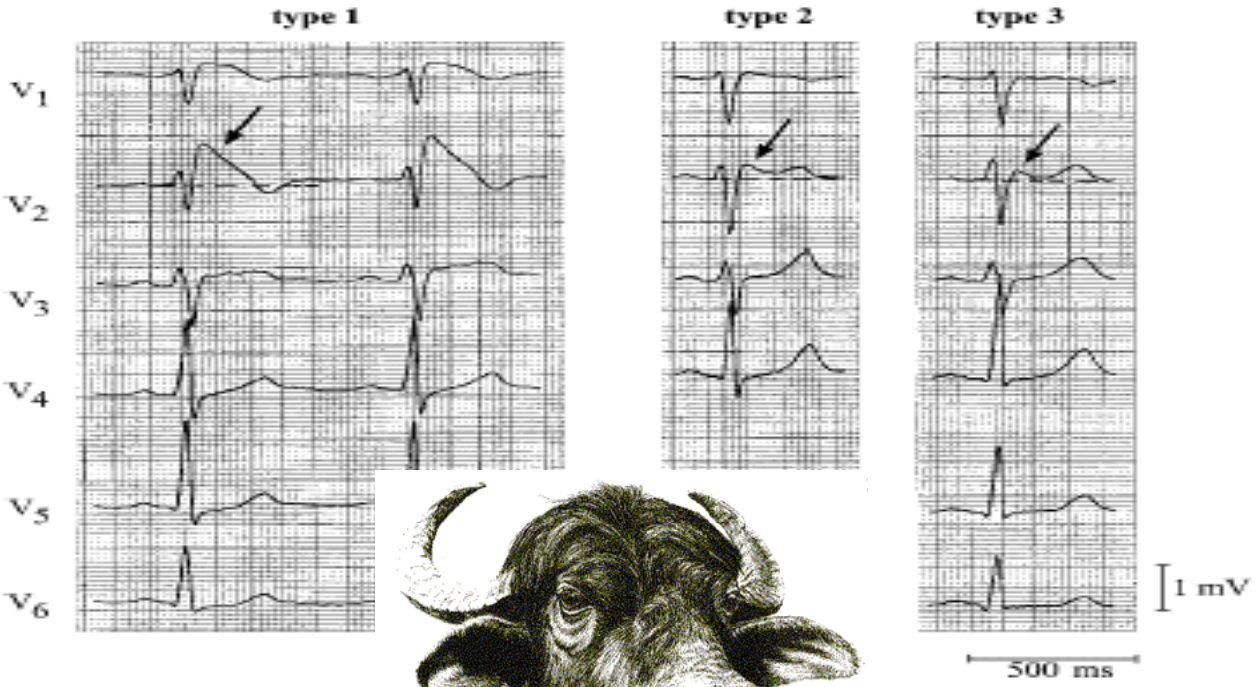


Figure 1 ***Precordial leads of a resuscitated patient with Brugada syndrome.*** **Note the dynamic ECG changes in the course of a couple of days. All three patterns are shown. Arrows denote the J-wave. The left panel shows a clear type 1 ECG. Between 7 February 1999 and 13 February 1999 types 2 and 3 appear.**

Consensus Report

Proposed Diagnostic Criteria for the Brugada Syndrome

Wilde et al Circulation 2002

This was not a classification but a case report!!

Pedro Brugada, MD, PhD, FESC; Ramon Brugada, MD; Josep Brugada, MD, PhD
Circulation 2005

- Gli studi più recenti hanno evidenziato che solo l'ECG tipo 1 è diagnostico per la sindrome.
- Sfortunatamente (SIC!!!!), molte serie includono individui con un ECG tipo 2 e 3 che probabilmente non hanno la sindrome.

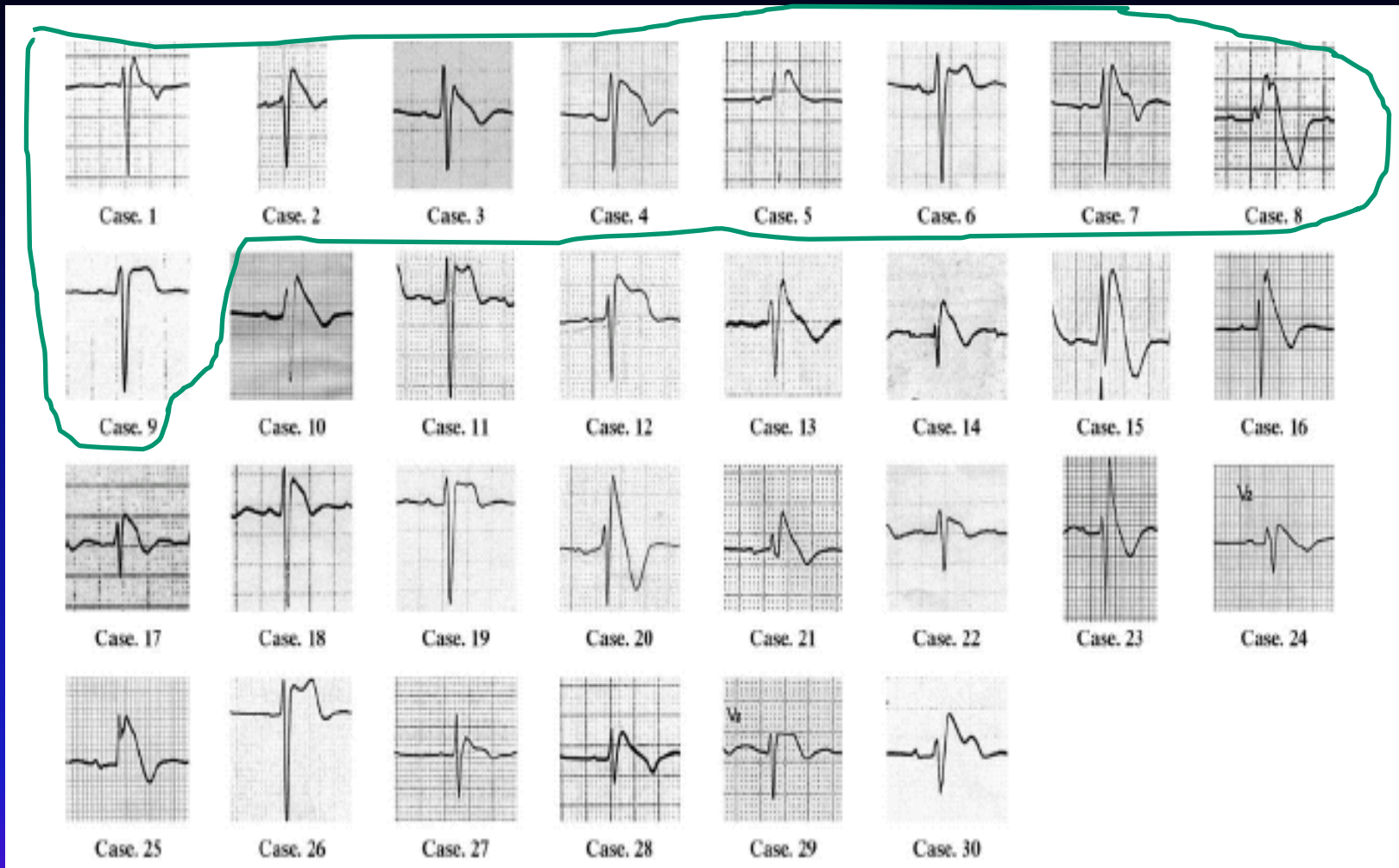
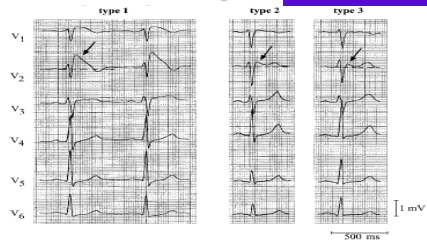
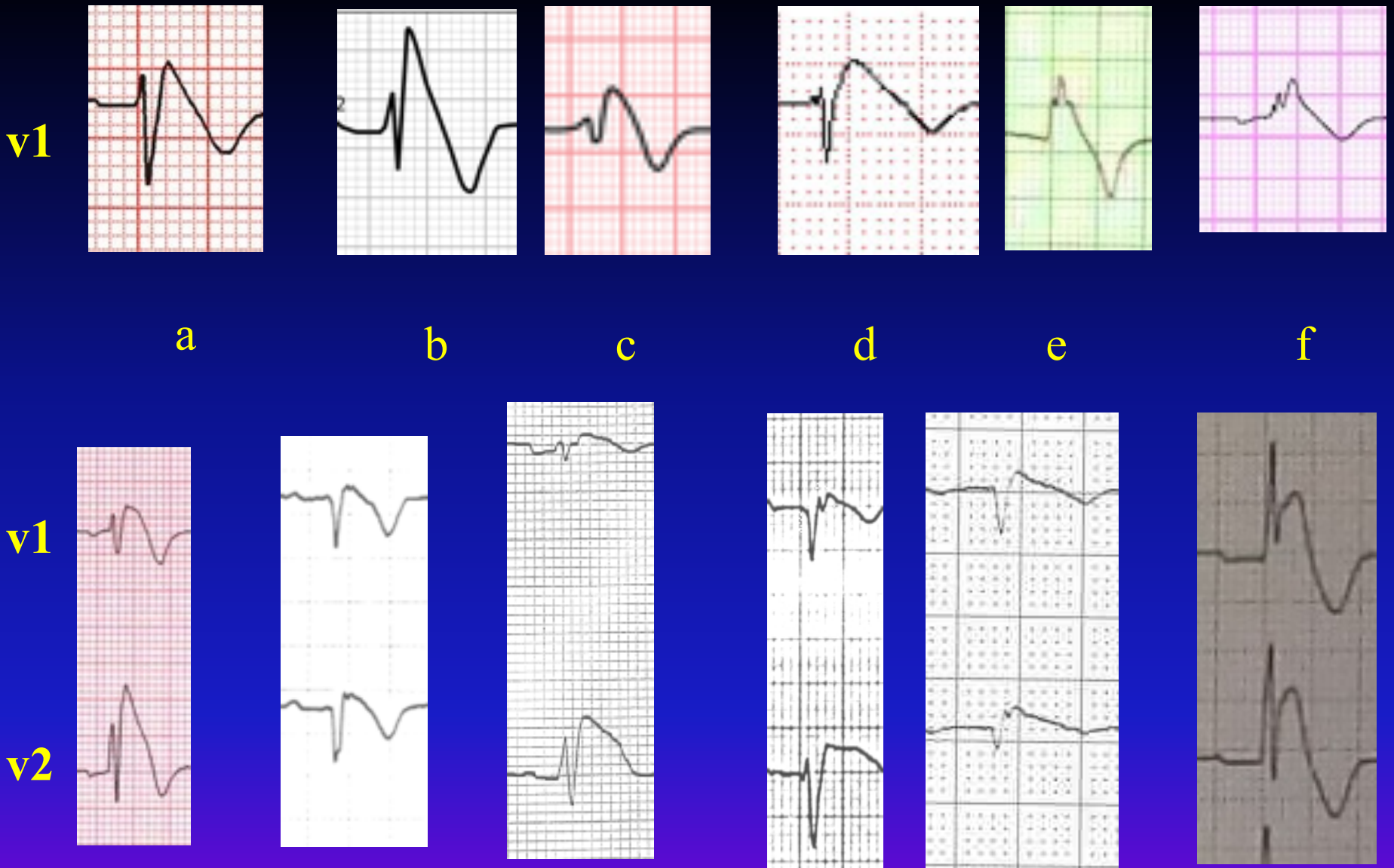


Fig 1. Lead V_2 ECGs in patients with the characteristic Brugada syndrome ECG at the time of presentation. Twenty-four patients had the 'coved type' ST-segment elevation and 6 patients had 'saddle-back type' ST-seg

Itoh Jap Circ J 2001

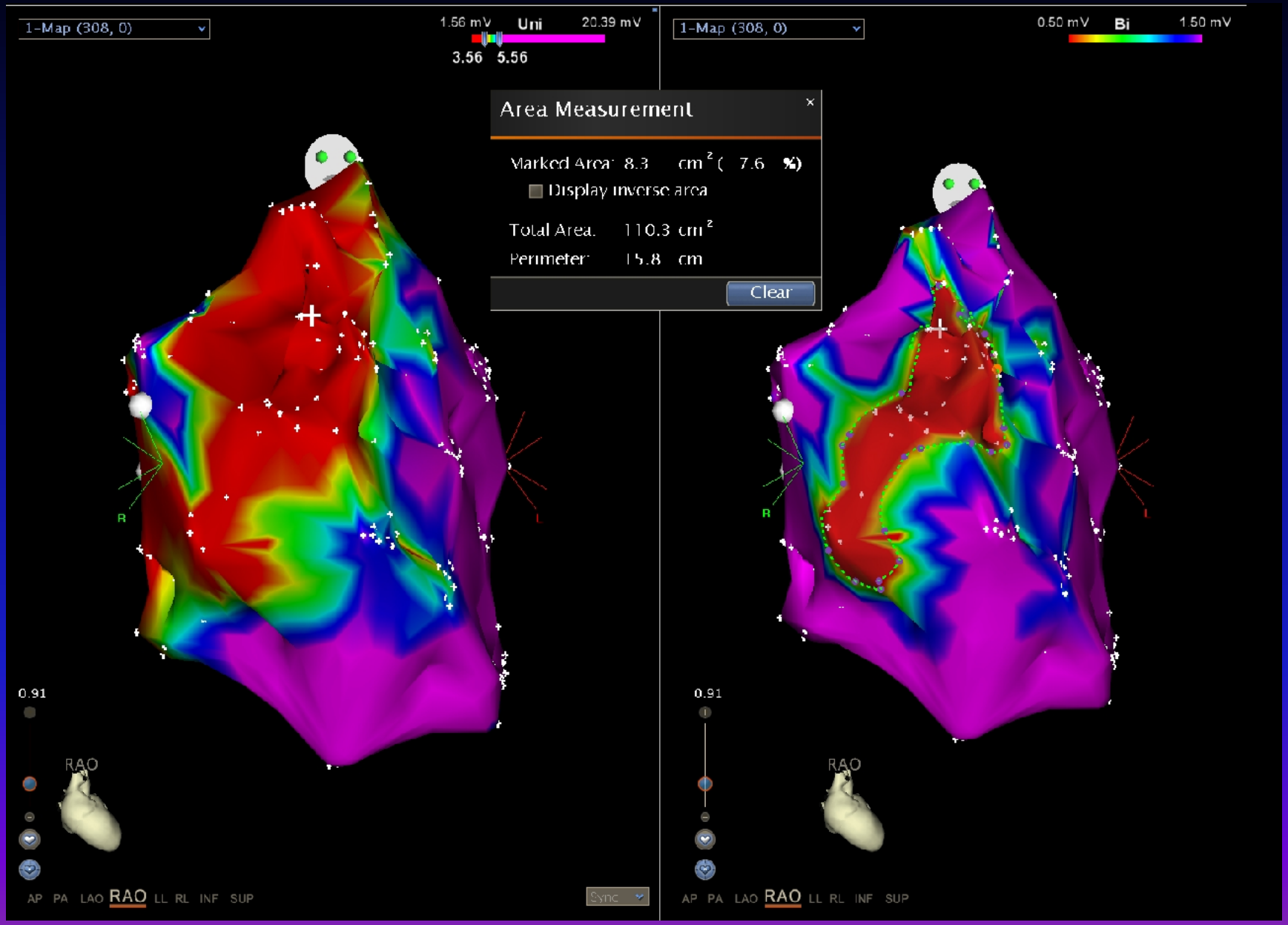
True syndrome evidentiated





Martini 2013

Delise 2007: The first report of an E.A.M.



Differential Diagnosis of rSr' Pattern in Leads V1-V2. Comprehensive Review and Proposed Algorithm

Adrian Baranchuk 2015

- Uno dei maggiori dilemmi è la diagnosi differenziale di una morfologia tipo rSr' in V1-V2.

Differential Diagnosis of rSr' Pattern in Leads V1-V2. Comprehensive Review and Proposed Algorithm

Adrian Baranchuk 2015

- Data from the Copenhagen City Heart Study showed a prevalence of 4.7% in men and 2.3% in women without apparent cardiovascular disease. This "normal" rSr' pattern is more frequent in younger subjects, with male preponderance and by definition is not a precursor of complete (proximal) RBBB. The r' is of fast ascent/descent inscription and the mechanism is a peripheral conduction delay with late activation of the pulmonary conus and posterobasal portion of the left ventricle.

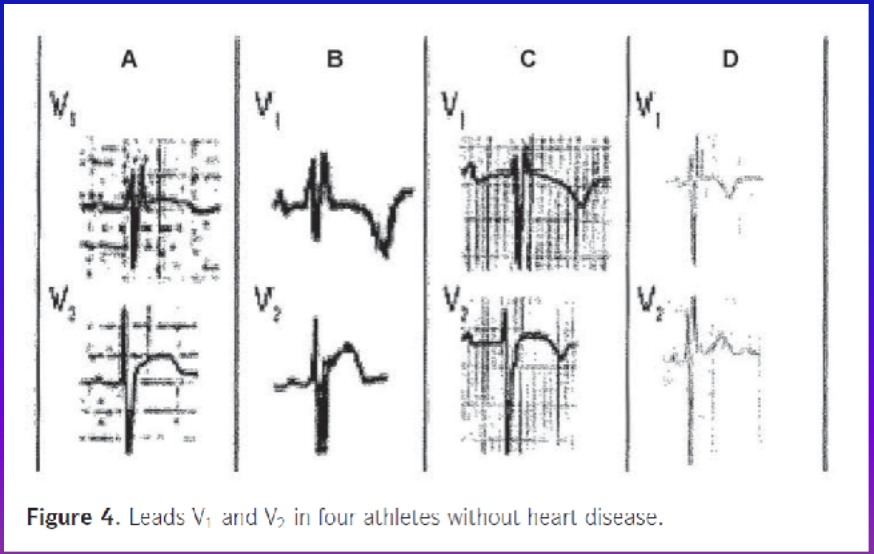


Figure 4. Leads V_1 and V_2 in four athletes without heart disease.

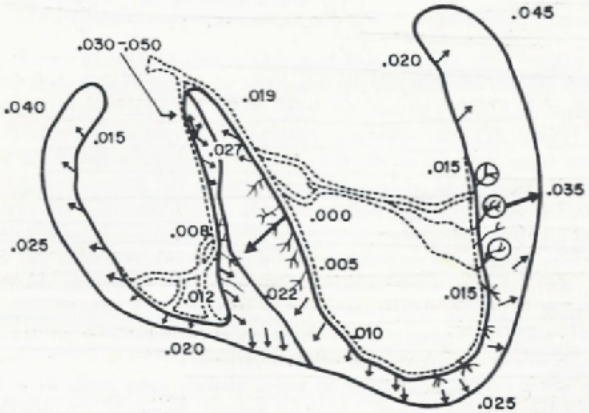


Fig. 20.

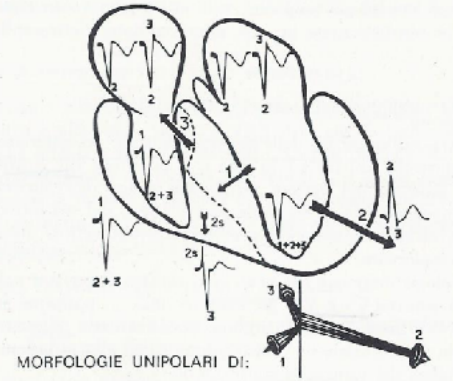


Fig. 21.

Il vettore basale o vettore 3 della attivazione ventricolare normale è diretto verso destra, in alto e dorsalmente (figura 21). Esso fugge dall'epicardio ventricolare sinistro producendo un potenziale negativo (onda s). La grandezza del vettore 3 è all'incirca eguale, o lievemente maggiore, a quella del vettore settale.

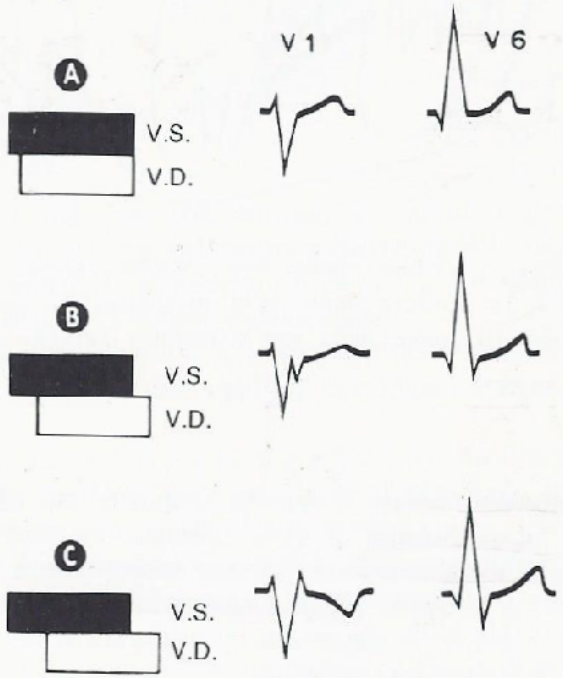
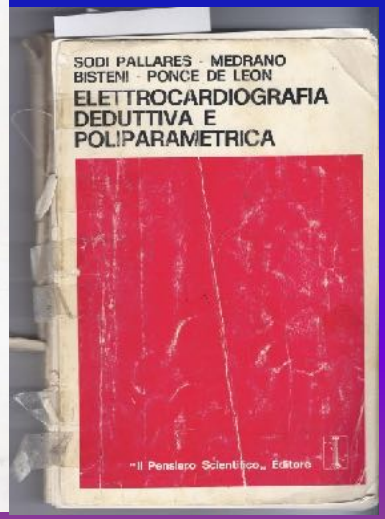
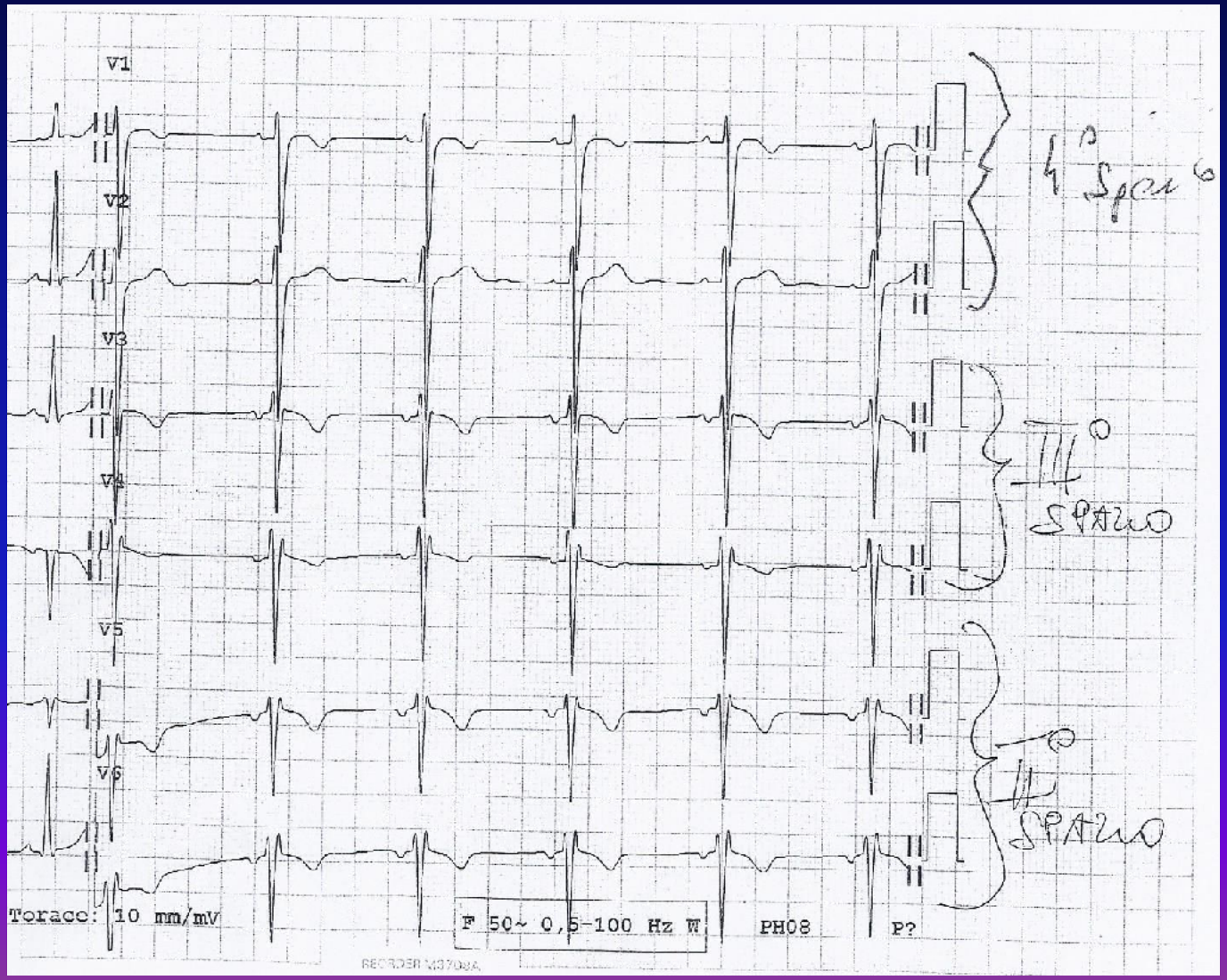


Fig. 55.



rSr’ in upper precordial leads



Incomplete rbbb

Φυσιολογικό ΗΚΓ Αθλητή **Σ. Brugada**

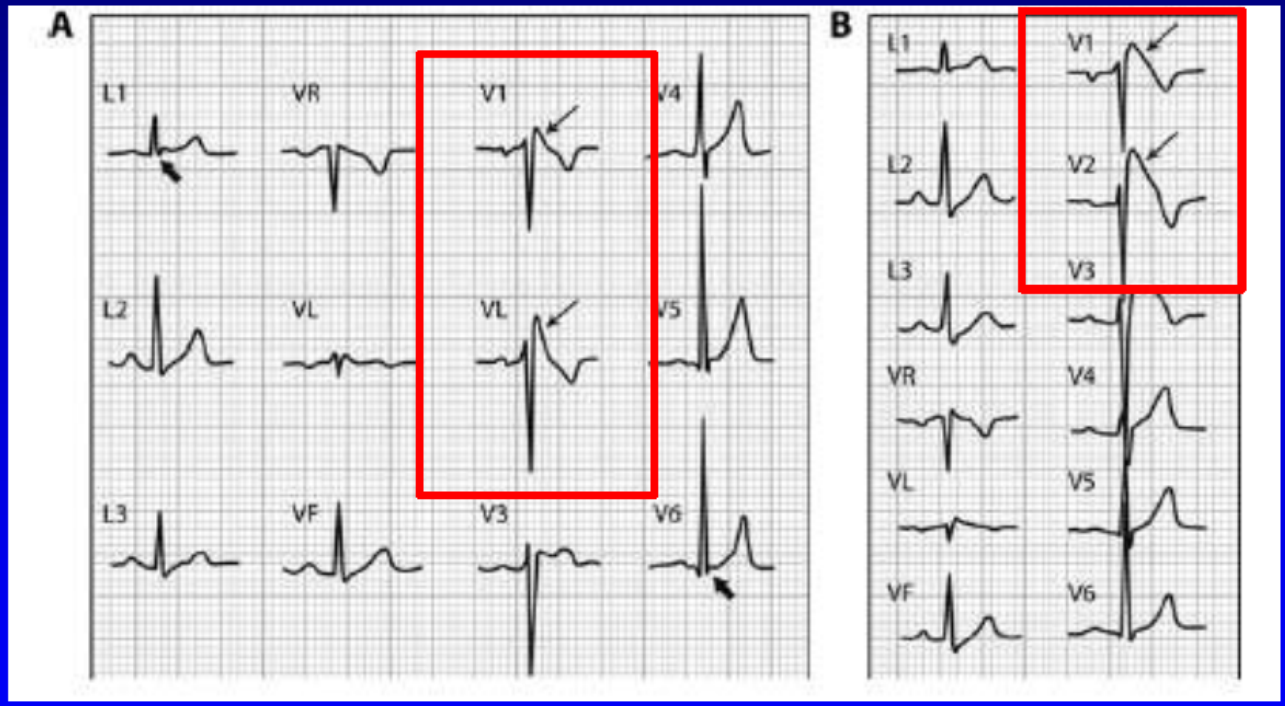
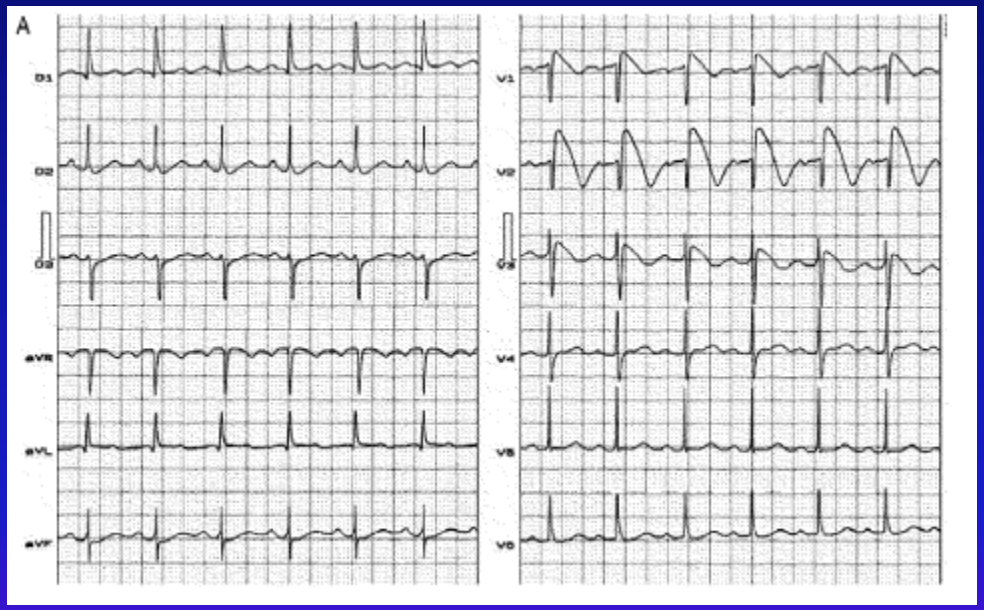
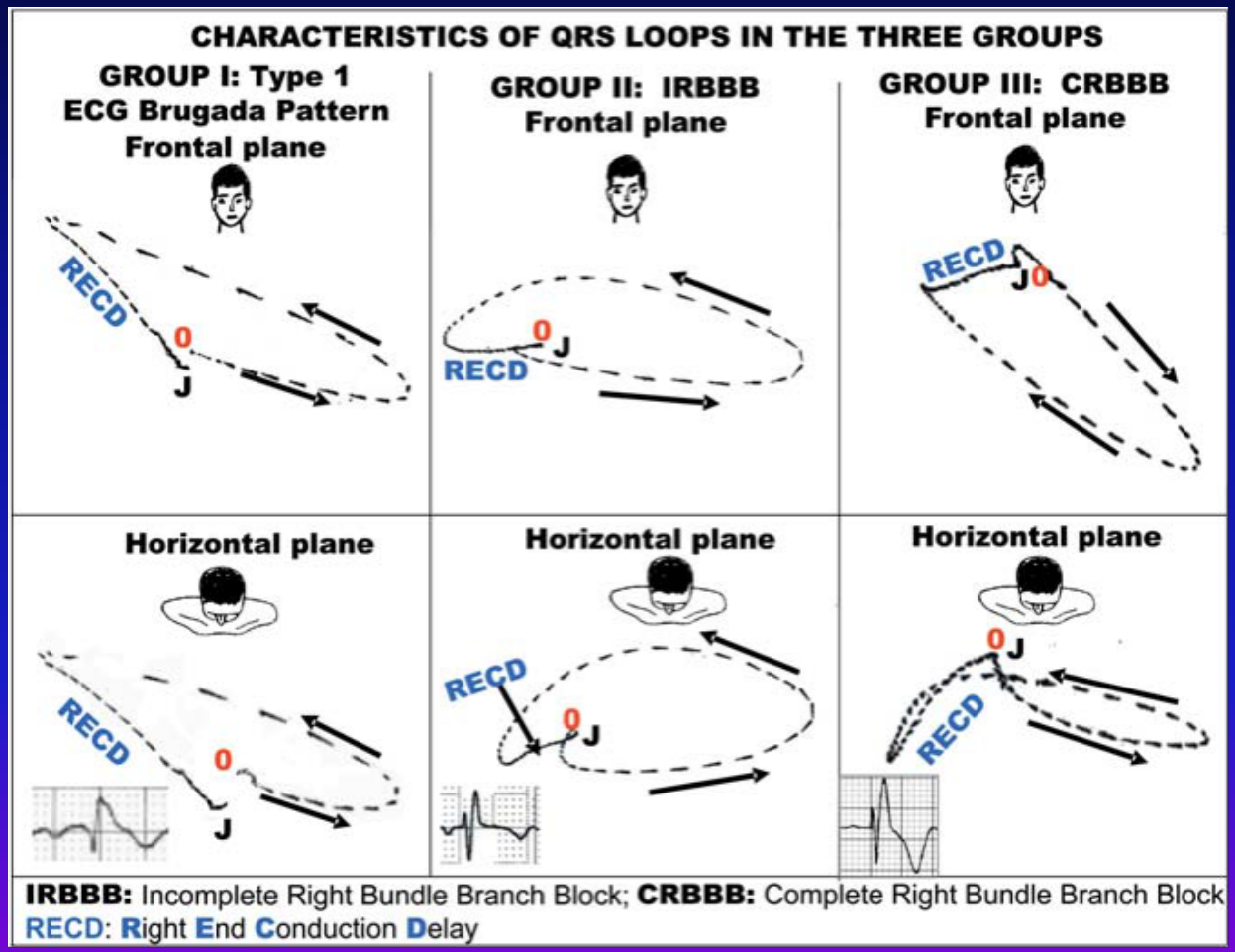


Figure 1 (A) Electrocardiogram of a patient with Brugada syndrome depicting Brugada type 1 electrocardiogram pattern. Note the absence of concomitant broad S wave in the left leads (I, aVL, V_5, and V_6) making the diagnosis of associated right bundle branch block highly unlikely.



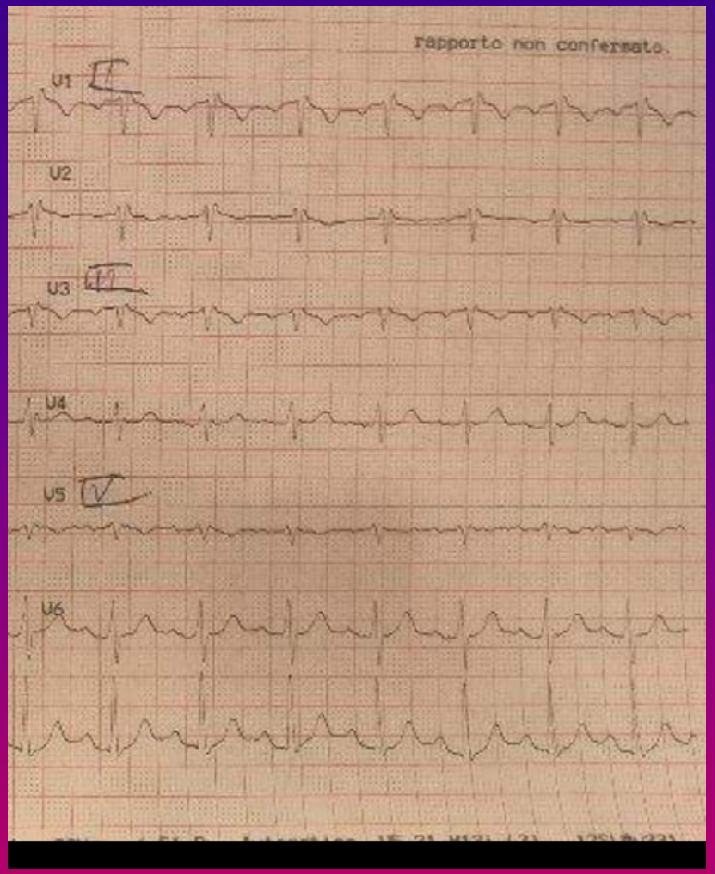
No s wave in V6

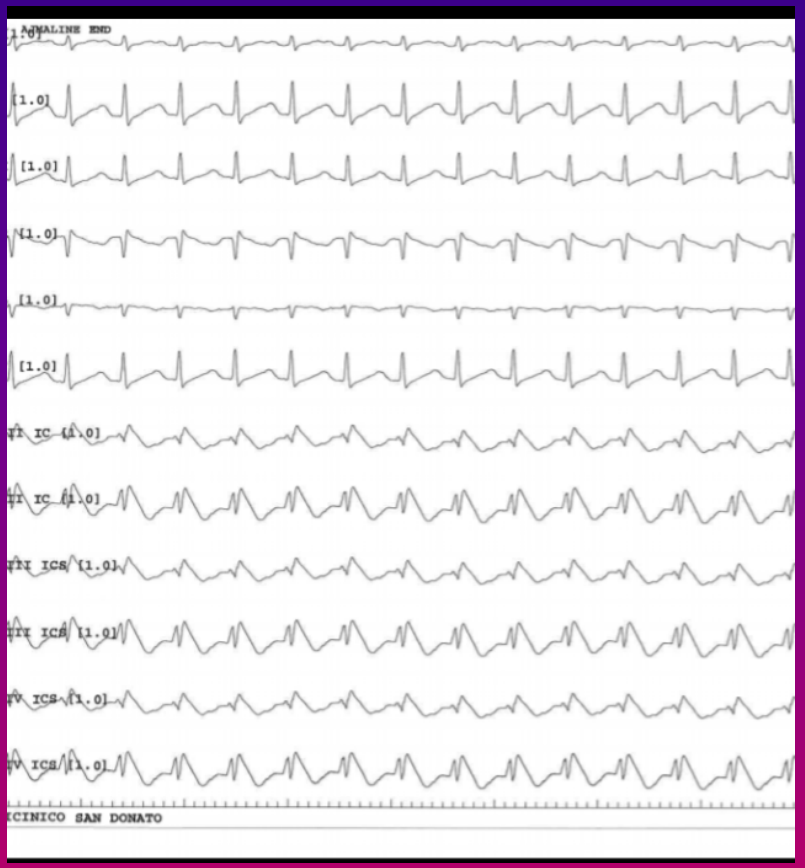
Figura 2 Ansa del QRS in tre gruppi. Il Gruppo BrS; ritardo terminale destro situate nel quadrante superiore del piano frontale e nel quadrante destro posterior del piano orizzontale. I gruppi IRBB e CRBB: ritardo di conduzione destro situato nel quadrante inferiore del piano frontale e nel quadrante anteriore del piano orizzontale.



Easy diagnosis ???

vorrei raccontare di un episodio : una giovane paziente viene da me in studio eseguo un ecg e trovo un semplice bbdx incompleto ,lei non contenta va dal mitico professor XX che le diagnostica una sindrome di brugada di tipo 2, esegue test farmacologici sconvolgendo gli standard della medicina in pratica utilizza solo l ajmalina ,vcon un dosaggio di ajmalina maggiore 1,5 mg per kg in soli 4 minuti ...e per concludere esegue il test tramite un catetere femorale posizionando il catetere infusore di farmaco appena al di sotto della vena cava inferiore. La paziente successivamente è stata seguita da noi con un test alla flecainide eseguito secondo il protocollo ed è risultata negativa ma oramai lei è andata in depressione ..

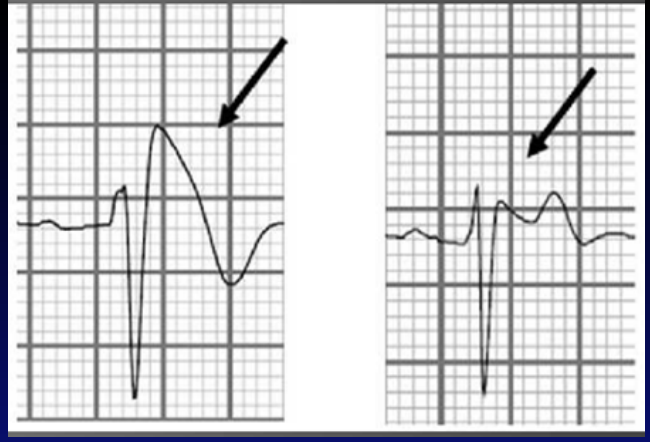




Brugada syndrome is not an ECG

- Bortolo Martini, Heart & Rhythm 2017

Serious data

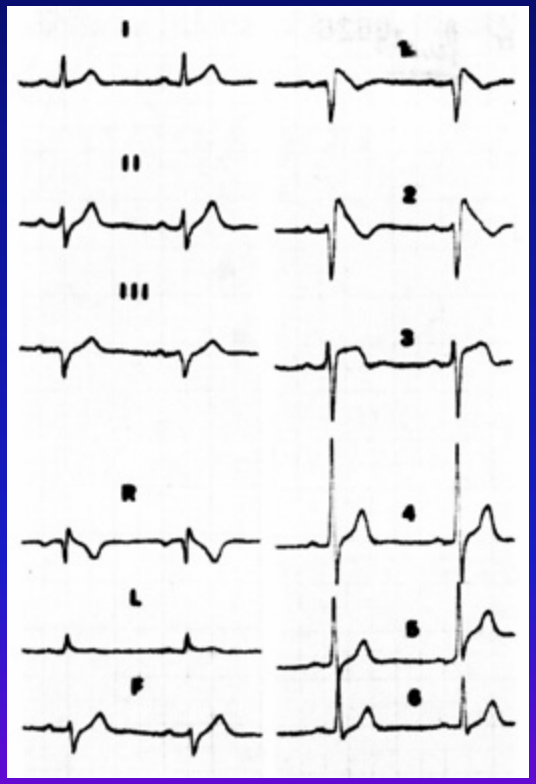


- The prevalence in seriuous studies is:

- 0.05% to 0.2% for type 1 BrS and
- 1.0% to 6.0% for non–type 1 BrS

- N.B: in Italy 30.000-120.000 type 1
- And 600.000- 3.600.000 type 2

Cosa è la sindrome della «ripolarizzzione precoce nelle precordiali destre»?



Questo è un ECG non una sindrome !!!!!!!!!!!!!!!!!!

Questo è un evento clinico e non un ECG !!!!!!! (circa 150 casi)

2015 ESC Guidelines for the management of patients with ventricular arrhythmias d the prevention of sudden cardiac death The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC) 2015

- Authors/Task Force Members: Silvia G. Priori*(Chairperson) (Italy) Carina Blomstro¨ m-Lundqvist*(Co-chairperson) (Sweden) Andrea Mazzanti† (Italy), Nico Bloma (The Netherlands), Martin Borggrefe (Germany), John Camm (UK), Perry Mark Elliott (UK), Donna Fitzsimons (UK), Robert Hatala (Slovakia), Gerhard Hindricks (Germany), Paulus Kirchhof (UK/Germany), Keld Kjeldsen (Denmark), Karl-Heinz Kuck (Germany), Antonio Hernandez-Madrid (Spain), Nikolaos Nikolaou (Greece ne M. Norekva° l (Norway), Christian Spaulding (France), and Dirk J. Van Veldhuisen (The Netherlands)

Recommendations	Class[a]	Level[b]	Ref.[c]
Brugada syndrome is diagnosed in patients with ST-segment elevation with type 1 morphology ≥2 mm in one or more leads among the right precordial leads V1 and/or V2 positioned in the second, third, or fourth intercostal space, occurring either spontaneously or after provocative drug test with intravenous administration of sodium channel blockers (such as ajmaline, flecainide, procainamide or pilsicainide).	I	C	This panel of experts

Syndrome=ECG!!

Authors «triage» according to their exerience!!

MalaCardiologia: interesse o ignoranza?

- Ischemia

- Sindrome del QT lungo

- Sindrome di Brugada

World Journal of Cardiology

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World J Cardiol 2017 September 26; 9(9): 737-741

DOI: 10.4330/wjc.v9.i9.737

ISSN 1949-8462 (online)

MINIREVIEWS

Brugada type 1 electrocardiogram: Should we treat the electrocardiogram or the patient?

Pietro Delise, Giuseppe Allocca, Nadir Sitta

Europace (2017) 0, 1–9
doi:10.1093/europace/eux226

CLINICAL RESEARCH

Clinical outcome of patients with the Brugada type 1 electrocardiogram without prophylactic implantable cardioverter defibrillator in primary prevention: a cumulative analysis of seven large prospective studies

Pietro Delise[1-3]*, Vincent Probst[2], Giuseppe Allocca[3], Nadir Sitta[3], Luigi Sciarra[4], Josep Brugada[5], Shiro Kamakura[6], Masahiko Takagi[7], Carla Giustetto[8], and Leonardo Calo[4]

1) Brugada type 1 electrocardiogram: Should we treat the electrocardiogram or the patient?
2) Clinical outcome of patients with the Brugada type 1 electrocardiogram without prophylactic implantable cardioverter defibrillator in primary prevention: a cumulative analysis of seven large prospective studies

P. Delise 2017

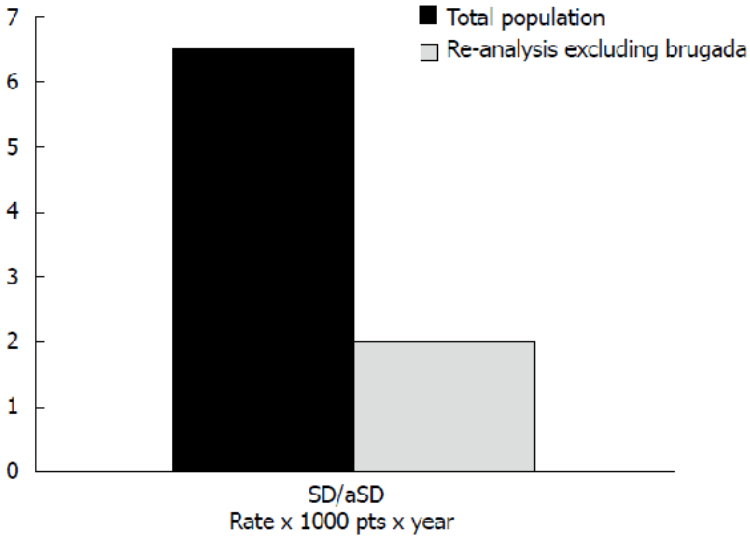
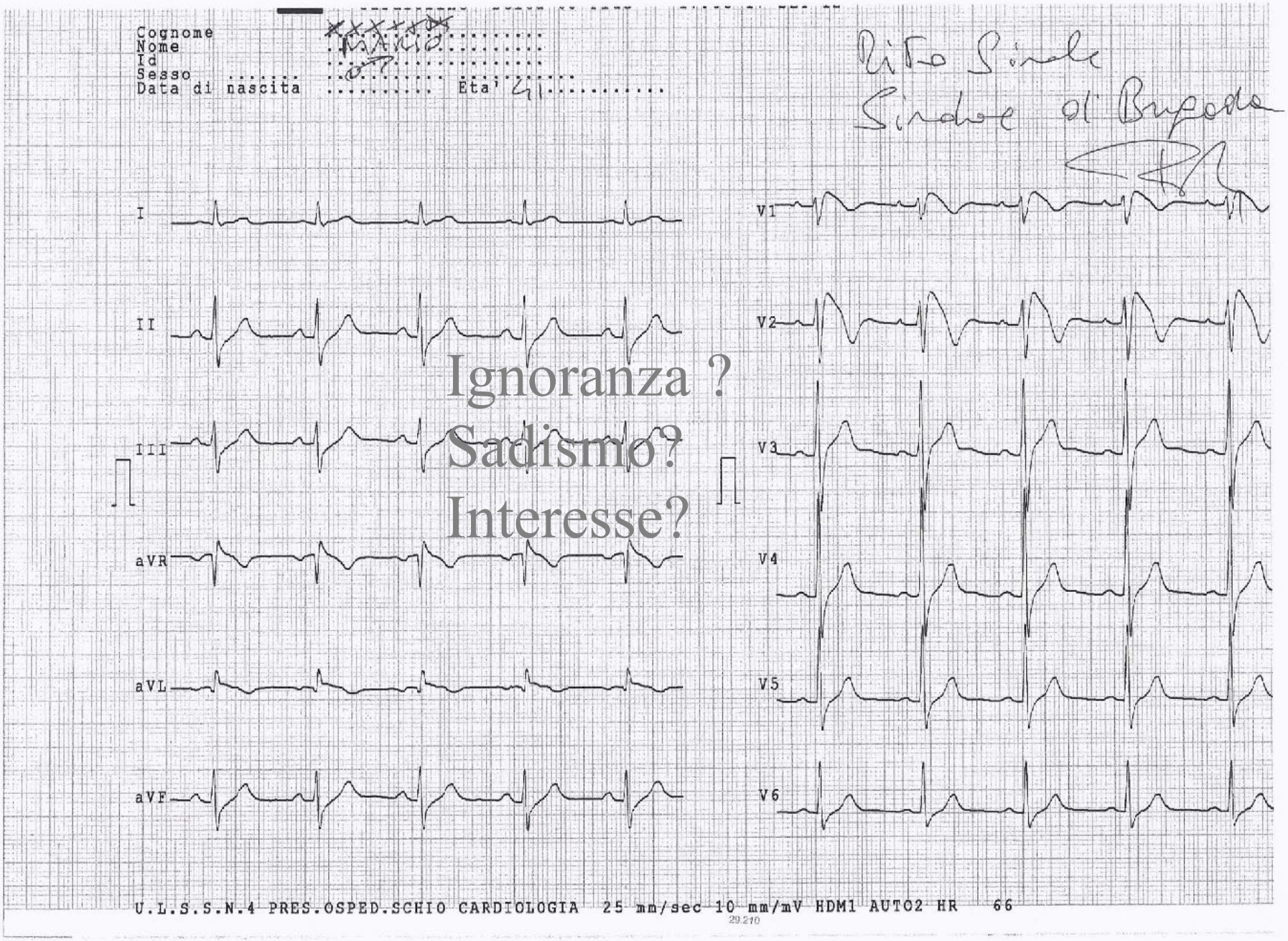
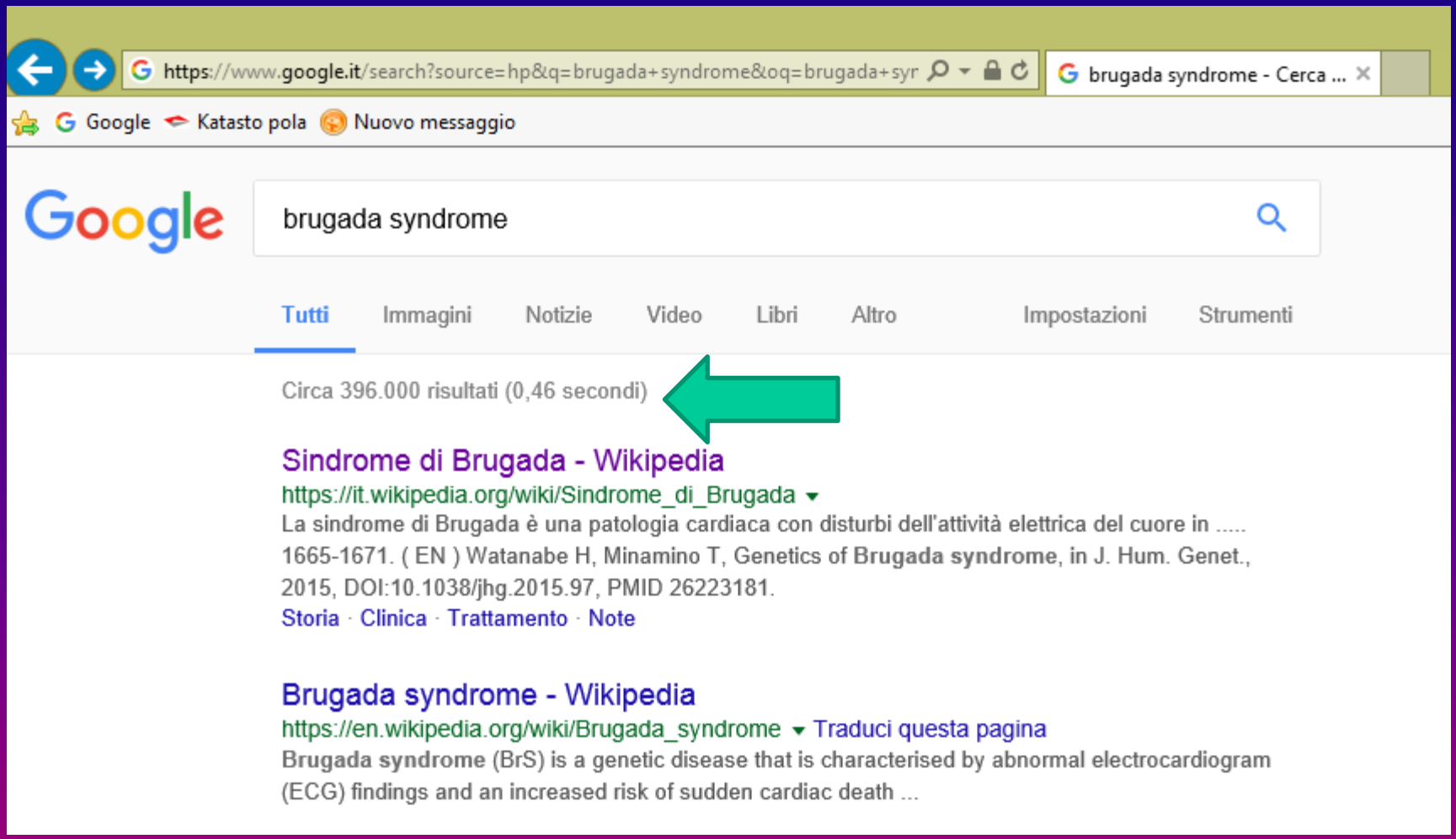


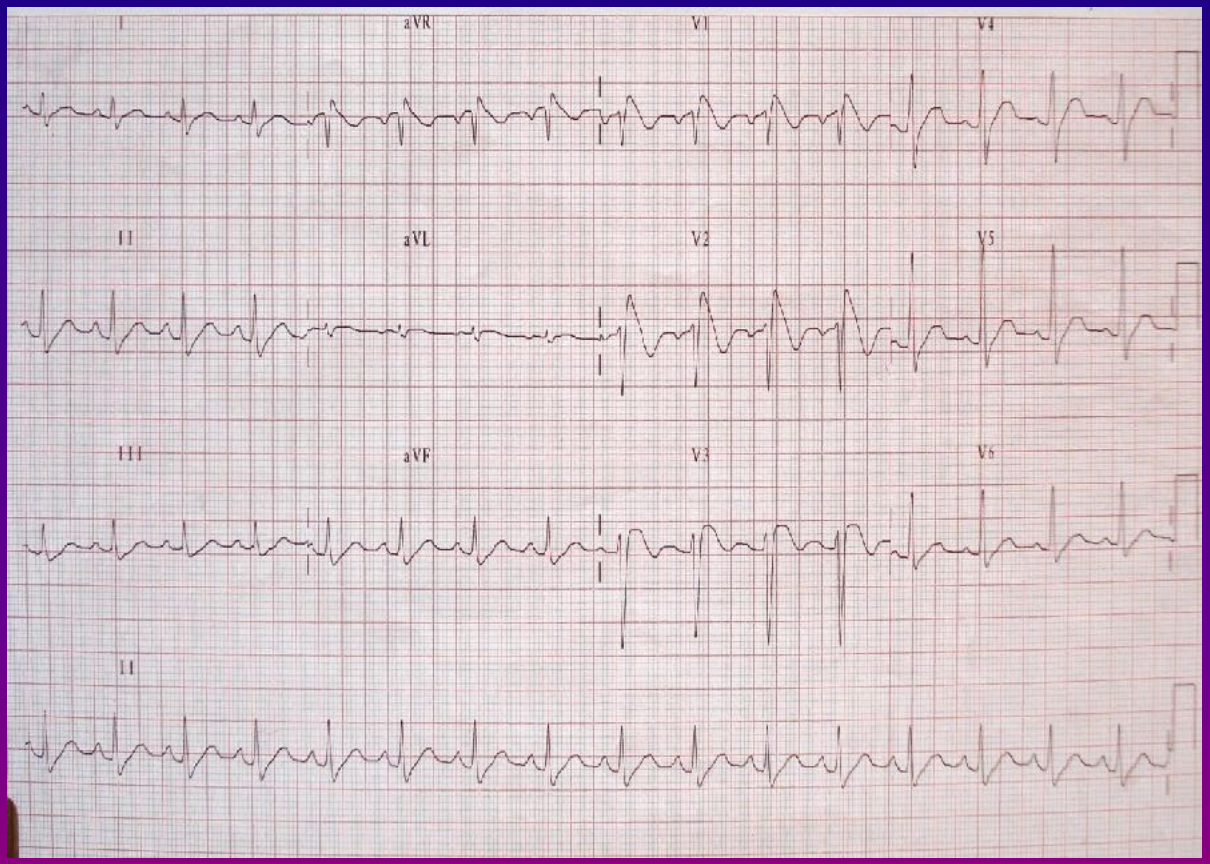
Figure 1 Incidence of sudden cardiac death/aborted sudden cardiac death × 1000 patientsxyear in subjects with type 1 Brugada type 1 electrocardiogram pattern without implantable cardioverter defibrillator. Cumultive analysis of 1366 patients including and excluding the paper of Brugada *et al*[7] from Delise *et al*[24] modified.

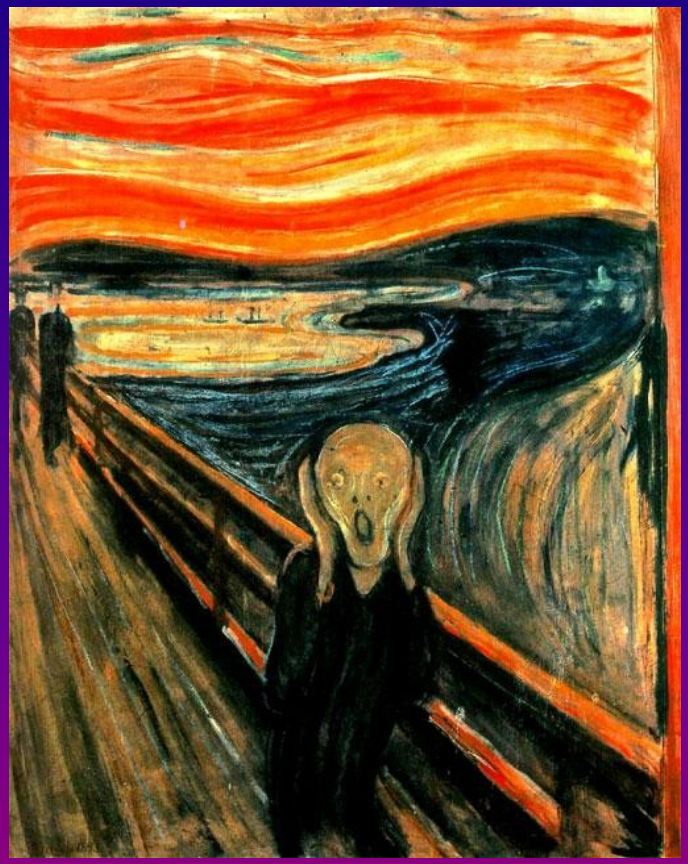
On excluding the Brugada series, the annual incidence of SD/aSD in the remaining 1198 patients fell to 0.22% in the total population and to 0.38 and 0.06% in spontaneous and drug-induced Br type 1, respectively

- Il mondo reale
- o «surreale»

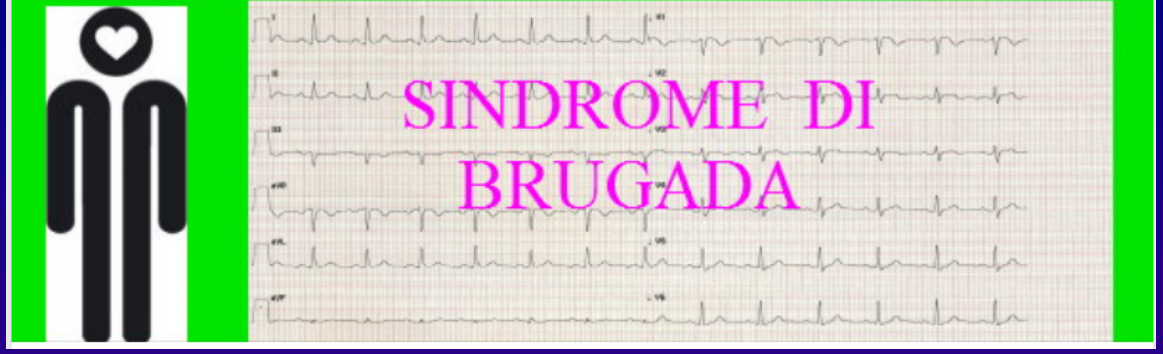

Cognome
Nome
Id
Sesso
Data di nascita
Età
Rito Sindr
Sindrome di Brugada
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
Ignoranza ?
Sadismo?
Interesse?
U.L.S.S.N.4 PRES.OSPED.SCHIO CARDIOLOGIA 25 mm/sec 10 mm/mV HDM1 AUTO2 HR 66
29.210


https://www.google.it/search?source=hp&q=brugada+syndrome&oq=brugada+syr
brugada syndrome - Cerca ...
Google
Katasto pola
Nuovo messaggio
Google
brugada syndrome
Tutti
Immagini
Notizie
Video
Libri
Altro
Impostazioni
Strumenti
Circa 396.000 risultati (0,46 secondi)
Sindrome di Brugada - Wikipedia
https://it.wikipedia.org/wiki/Sindrome_di_Brugada
La sindrome di Brugada è una patologia cardiaca con disturbi dell'attività elettrica del cuore in .....
1665-1671. (EN) Watanabe H, Minamino T, Genetics of Brugada syndrome, in J. Hum. Genet.,
2015, DOI:10.1038/jhg.2015.97, PMID 26223181.
Storia · Clinica · Trattamento · Note
Brugada syndrome - Wikipedia
https://en.wikipedia.org/wiki/Brugada_syndrome
Traduci questa pagina
Brugada syndrome (BrS) is a genetic disease that is characterised by abnormal electrocardiogram
(ECG) findings and an increased risk of sudden cardiac death ...





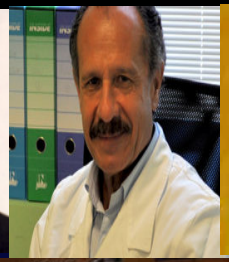
The horror channel!!

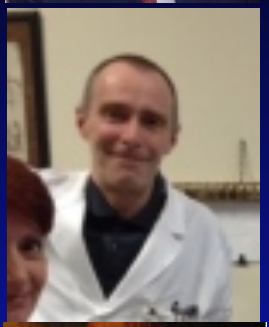


- Ciao a tutti e grazie x avermi accettato. Ecco la mia storia: nel 2011 faccio l ecc nessun sintomo, solo controllo, e il mio cardiologo scrive " non tipico x fenomeno di Brugada", cerco su Internet e scopro il terrore, voglio un altro consulto e mi reco da un altro medico che mi smonta holter, ecocardio, elettrocardiogramma, prova di sforzo... e mi rassicura dicendomi che devo stare tranquillo e che è una specifica del mio elettrocardiogramma, tranquillizzato vivo serenamente, mai singoli, mai aritmia. A dicembre del 2016 quindi 5 anni dopo, vado consigliato da amici, da un Aritmologo, che mi stronca..... mi dice patten Brugada tipo 2, e mi suggerisce il test alla flecanide per capire se realmente affetto da sindrome e se passa al tipo 1. A gennaio faccio il test positivo gia con mezza dose. Diagnosi: soggetto asintopatico, a basso rischio. Gelo, paura, ansia, pensi a tutto, ma soprattutto ai figli e alla famiglia. Su internet la confusione aumenta, Chi parla di cure e chi nega, unica soluzione un ICD, sto impazzendo.... AIUTO....

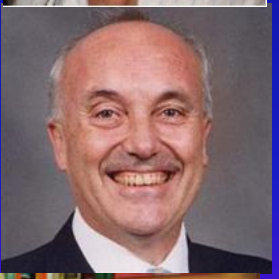











La sindrome della **«Ripolarizzazione precoce nelle Precordiali destre»**

Date a Cesare quel che è di Cesare!