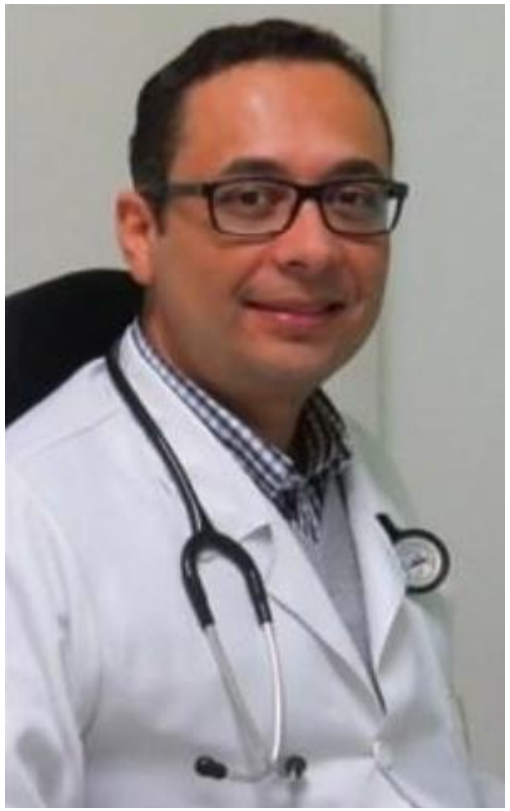
Case Report

Prezado Andrés, gostaria que submetesse esse surpreendente caso com padrão eletrocardiográfico incomum para o forum.

Dear Andrés, I would like you to submit this surprising case with an unusual electrocardiographic pattern to the forum.



Acácio F. Cardoso MD, PhD

Cardiology Service of the Nipo Brasileiro Hospital, São Paulo/SP, Brasil.

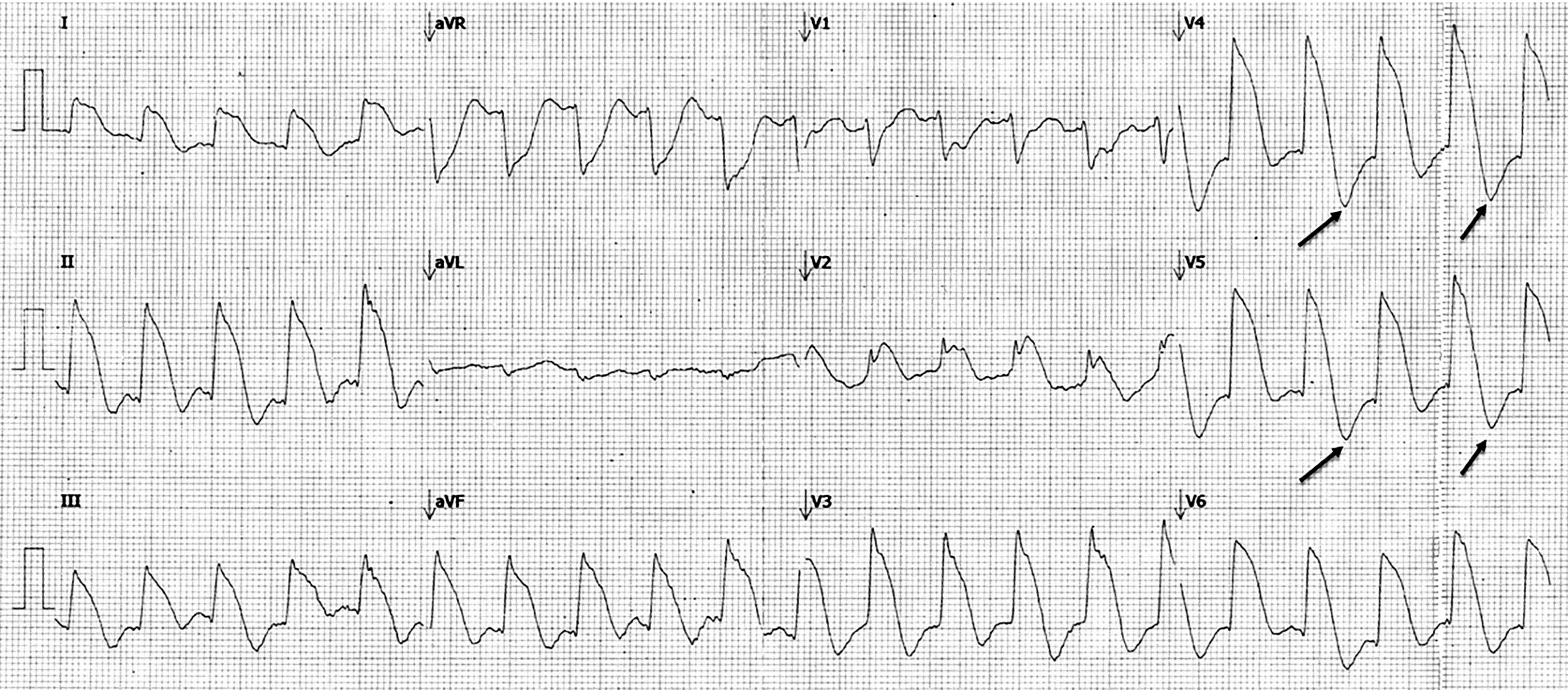
Case report

A 35 year-old-man, with a history of smoking and illicit drugs user (marijuana, cocaine, and inhalant solvents), under treatment with antipsychotic and antidepressant (haloperidol and escitalopram), after two days of many episodes of vomiting and diarrhea, he developed mental confusion and decreased level of consciousness and was admitted gasping in the emergency room. Shortly thereafter, he evolved with cardiorespiratory arrest. The patient was promptly attended and after six minutes of cardiac resuscitation maneuvers, adrenalin infusion, and two defibrillations the cardiac rhythm was recovered on the monitor. An ECG was performed and demonstrated a diffuse and atypical ST-segment elevation, and concomitant ST-segment depression in aVR and V1. Finally, a transient T-wave polarity is observed (ECG. 1). The initial laboratory tests showed a metabolic acidosis and a high level of serum lactate (lactate = 26mg/dl) associated with hypernatremia, hypokalemia, important leukocytosis, and a slight increase in troponin. Intubation and mechanical ventilation were required as well as fluid resuscitation, correction of metabolic acidosis, and antibiotics. One hour later the ECG was repeated and the ST-segment elevation was decreased by approximately fifty percent (ECG 2). No Q-wave was present at that moment and the coronariography was discharged. Six hours after the event the ECG alterations were completely normalized (ECG3). In the intensive care unit, the first echocardiogram showed an important left ventricular dysfunction (EF by Teichholz = 0.36) and a diffuse hypokinesis, which was totally recovered two days later (EF = 0.69). The coronary artery disease was ruled out by a coronary angiotomography. The patient progressed well and was discharged eight days later without any neurological sequelae.

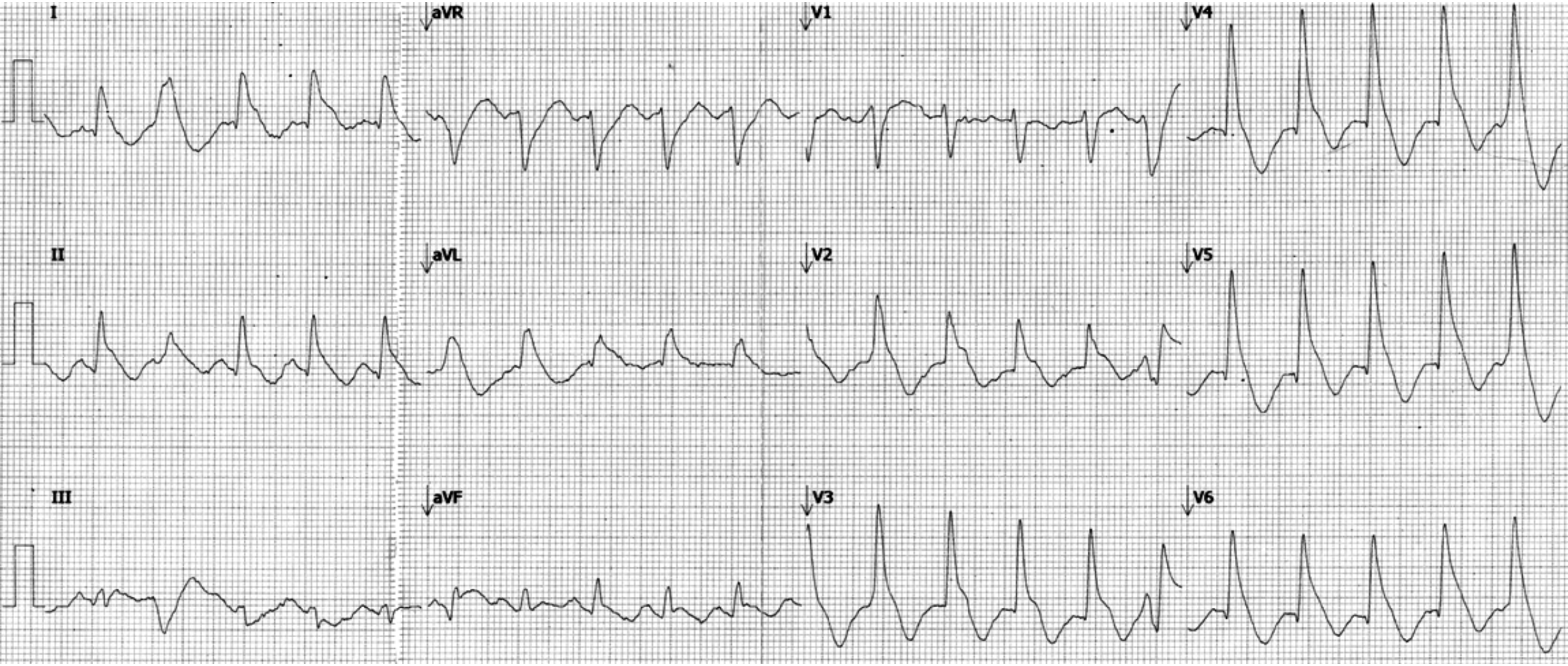
Spanish

Reporte de un caso

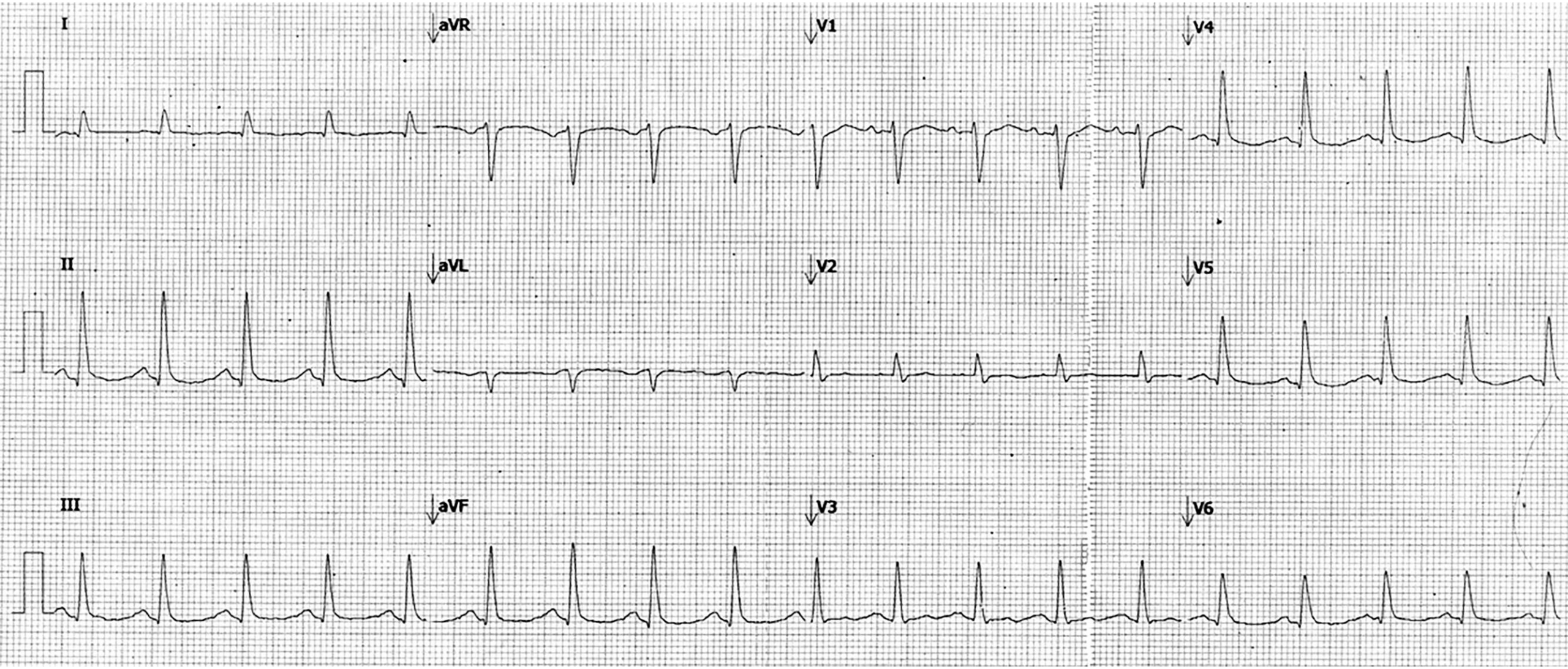
Un hombre de 35 años, con antecedentes de tabaquismo y consumo de drogas ilícitas (marihuana, cocaína y solventes inhalantes), en tratamiento con antipsicóticos y antidepresivos (haloperidol y escitalopram), después de dos días de numerosos episodios de vómitos y diarrea, desarrolló confusión mental y disminuyó el nivel de conciencia y fue admitido jadeando en la sala de emergencias. Poco después, evolucionó con paro cardiorrespiratorio. El paciente fue atendido de inmediato y después de seis minutos de maniobras de reanimación cardíaca, infusión de adrenalina y dos desfibrilaciones, el ritmo cardíaco se recuperó en el monitor. Se realizó un ECG y se demostró una elevación difusa y atípica del segmento ST y depresión concomitante del segmento ST en aVR y V1 (ECG. 1). Las pruebas de laboratorio iniciales mostraron una acidosis metabólica y un alto nivel de lactato sérico (lactato = 26 mg / dl) asociado con hipernatremia, hipocalemia, leucocitosis importante y un ligero aumento de troponina. Se requirió intubación y ventilación mecánica, así como reanimación con líquidos, corrección de acidosis metabólica y antibióticos. Una hora más tarde, se repitió el ECG y la elevación del segmento ST se redujo en aproximadamente un cincuenta por ciento (ECG 2). No hubo onda Q en ese momento y la coronariografía fue dada de alta. Seis horas después del evento, las alteraciones del ECG se normalizaron por completo (ECG 3). En la unidad de cuidados intensivos, el primer ecocardiograma mostró una importante disfunción ventricular izquierda (EF por Teichholz = 0,36) y una hipocinesia difusa, que se recuperó totalmente dos días después (EF = 0,69). La enfermedad de la arteria coronaria se descartó mediante una angiotomografía coronaria. El paciente progresó bien y fue dado de alta ocho días después sin ninguna secuela neurológica.



ECG1: First ECG recorded immediately after cardiorespiratory arrest recovery. What do the arrows mean?



ECG 2: ECG performed one hour later.



ECG 3: ECG performed six hours later.

Colleagues opinions

I think you well covered this topic in your last review of drug/ electrolyte abnormalities on the ECG. The wide bizarre QRS complex is likely SVT since it is so similar to later tracings and there is QRS alternans seen in precordial leads and lead 3. He was exposed to cocaine so it is possible that he had coronary spasm as well but think major issue is drugs with Na channel blockade. We are not told of K levels and this would be relevant. Eager to hear opinion of the group.

Melvin Scheinman MD

Brief trajectory: Dr. Melvin Scheinman, one of the pioneers of cardiac electrophysiology, was the first to perform catheter ablation on humans. His team was instrumental in developing radiofrequency energy applications for cardiac arrhythmias. Scheinman and his colleagues also developed techniques for modifying sinus node function in patients with inappropriate sinus tachycardia – a condition in which resting heart rate is abnormally high – and to cure patients with automatic junctional tachycardia, in which one area of the heart is leading to a too-fast beat.

Scheinman currently directs the cardiac genetic arrhythmia program, which is devoted to discovering new genes related to heart rhythm disorders.

Scheinman grew up in Brooklyn, New York, and earned an undergraduate degree at Johns Hopkins University, where he graduated first in his class. His medical education included Albert Einstein College of Medicine, residency training at the University of North Carolina at Chapel Hill and cardiology training at UCSF Medical Center.

Spanish

Querido Andrés: Creo que cubriste bien este tema en tu última revisión de las anormalidades de drogas / electrolitos en el ECG. Es probable que el complejo QRS ancho y extraño sea una taquicardia supraventricular, ya que el patrón es muy similar a los trazados posteriores

Se observan QRS alternantes en las derivaciones precordiales y en la derivación III. Por haber estado expuesto a la cocaína, es posible que también haya sufrido un espasmo coronario, no obstante, creo que la causa principal obedecería a bloqueo del canal de Na^+ por al efecto de drogas. No se nos informa de los niveles de K^+ y esto sería relevante.

Estoy con sobradas ganas de escuchar la opinión del grupo.

Melvin Scheinman MD

Portuguese

Caro Andrés, acho que você cobriu bem esse tópico em sua última revisão das anormalidades de drogas y eletrólitos no eletrocardiograma do ECG. É provável que o complexo QRS estranho e amplo corresponda a uma taquicardia supraventricular, uma vez que o padrão é muito semelhante aos traçados subsequentes.

QRSs alternantes se observam nas derivações precordiais e na derivação III. Por ter sido exposto à cocaína, também pode ter sofrido um espasmo coronariano que justificaria o padrão, no entanto, acho que a principal causa seria o bloqueio do canal de Na+ devido ao efeito das drogas. Nós não fuimos informados dos níveis de K+ o qual penso que seria relevante.

Estou ansioso para ouvir a opinião do grupo.

Portuguese

Resposta ao comentário do Professor Melvin

O nível de K^+ da entrada ao hospital era de 3,2mg/dl, o que nos parece pouco alterado para causar as alterações observadas no ECG.

Em relação ao espasmo coronário como causa, embora seja uma hipótese possível, acho que é improvável já que o paciente apresentou uma evolução de 48h precedendo o evento, isso implica a participação da doença de base (gastroenterite aguda) no evento final. É importante lembrar que a reversão das alterações de segmento ST- e onda T ocorreram após correção da volemia e acidose, o que diminuem a chance de espasmo coronariano como sendo responsável pela causa.

Vamos aguardar as outras opiniões do grupo.

Acácio Fernandes Cardoso MD PhD

English

Response to Professor Melvin's comment The K^+ level at admission to the hospital was 3.2 mg/dl, which seems little changed to cause the changes observed on the ECG. Regarding coronary spasm as a cause, although it is a possible hypothesis, I think it is unlikely since the patient had a 48-hour evolution preceding the event, this implies the participation of the underlying disease (acute gastroenteritis) in the final event. It is important to remember that the reversal of changes in the ST- and T-wave occurred after correction of blood volume and acidosis, which reduces the chance of coronary spasm as being responsible for the cause.

We will wait for the other opinions of the group.

Acácio Fernandes Cardoso MD PhD

Thank you,

Hola Potro

En el primer ECG presenta una TVMS

En el segundo un trastorno un específico de la conducción intraventricular. Con punto J elevado y QTc prolongado.

El tercero continúa con un punto J elevado y QT prolongado.

Como refirió el Dr Scheinman la cocaína produce bloqueo de los canales de Na^+ pudiendo provocar un patrón Brugada like. En este caso creo ha evidenciado una alteración de la repolarización maligna y QT prolongado.

No posee un ECG del paciente previo al alta?

Un abrazo y gracias por tan lindo caso.

Martín Ibarrola MD Buenos Aires Argentina

Hello Andres

In the first ECG he presents a sustained monomorphic ventricular tachycardia (SMVT)

In the second one , a nonspecific intraventricular conduction disturbance with J point and ST segment elevation and long QTc.

The third ECG continues with the same pattern .

As Dr Scheinman reported, cocaine blocks Na + channels and can cause a Bugada like pattern. In this case, I believe it has evidenced of an malignant repolarization and prolonged QT interval

Do you not have an ECG of the patient before discharge?

A hug and thanks for such a nice case.

Martín Ibarrola MD Buenos Aires Argentina

Caro Professor Riera: É uma honra opinar num caso que não conheço pertencente a um membro de minha equipe.
Trata-se de um caso muito interessante do Hospital Nipo-Brasileiro
Desde meu ponto de vista o paciente está em ritmo sinusal nos três traçados. O eixo elétrico do QRS em todos eles não se modificou.
Se observa distúrbio de condução intraventricular não específico com alargamento do QRS e relativo encurtamento do intervalo QT.
Como mais importante fator causante das modificações ocorridas na minha opinião é o elevadíssimo nível de lactato sanguíneo. Isto pode ocorrer na intoxicação por cocaína. Possivelmente a alteração metabólica causou hipercalemia intracelular e consequentemente parada cardíaca.
Obrigado pela Oportunidade
Jose Grindler Diretor do Serviço de Eletrocardiologia, do Hospital das Clínicas Faculdade de Medicina da Universidade de São Paulo FMUSP
jose.grindler@hc.fm.usp.br
English
Dear Professor Riera: It is an honor to express my opinion on a case I do not know that belongs to a member of my team.
This is a very interesting case from the Japanese-Brazilian Hospital
From my point of view, the patient is in sinus rhythm in the three traces. The QRS electrical axis in all of them has not changed.
A nonspecific intraventricular conduction disorder is observed with broad QRS interval and relative shortening of the QT interval.
As the most important factor causing the changes that occurred in my opinion is the extremely high level of blood lactate. This can occur in cocaine poisoning. Possibly the metabolic alteration caused intracellular hyperkalemia and consequently cardiac arrest.
Thanks for the opportunity
Jose Grindler Diretor do Serviço de Eletrocardiologia, do
Hospital das Clínicas Faculdade de Medicina da Universidade de São Paulo FMUSP
jose.grindler@hc.fm.usp.br

Estimado Andrés, pienso que por los antecedentes del pacientes (cocainómano) pudo tener un evento tromboembólico (con lisis espontánea) o espasmo coronario, y teniendo en cuenta el método del reloj, el eje del vector de lesión en plano frontal se dirige hacia abajo y a la izquierda, alrededor de + 60º y en plano horizontal hacia izquierda y atrás, por eso el vector de lesión se aleja de aVR y V1: probable arteria culpable la circunfleja. Sumado a las alteraciones hidroelectrolíticas, acidosis y maniobras de RCP. No se modifica la porción inicial del QRS, si el segmento ST (lesión mas onda "J" por la probable hipotermia), incluso recuerda un patrón de Aizawa (A**izawa, Y., Chinushi, M., Hasegawa, K., Naiki, N., Horie, M., Kaneko, Y., … Fukuda, K. (2013).** ***Electrical Storm in Idiopathic Ventricular Fibrillation Is Associated With Early Repolarization. Journal of the American College of Cardiology, 62(11), 1015–1019.*** **doi:10.1016/j.jacc.2013.05.030**), además de la prolongación del QTc producto de antipsicóticos y antidepresivos. Creo que tiene en todo momento ritmo sinusal 125 lpm y 107 lpm en 3º registro donde persiste el QTc prolongado, visible en V1 alrededor de 480 mseg. las fechas que tu señalas se deberían a la alternancia de onda T.

A la espera de las opiniones de los Maestros del Foro, me despido cordialmente

Juan Carlos Manzzardo MD Mendoza Argentina

Dear Andrés, I think that due to the history of the patient (cocaine addict) he could have had a thromboembolic event (with spontaneous lysis) or coronary spasm, and taking into account the clock method, the axis of the injury vector in the frontal plane is directed downwards and to the left, (around + 60º) and in a horizontal plane to the left and backward, therefore the injury vector moves away from aVR and V1: probable culprit left circumflex artery Added to the electrolytic disturbances, acidosis and CPR maneuvers.

The initial portion of the QRS complex is not modified, if the ST segment (lesion plus "J" wave due to probable hypothermia), even recalls an Aizawa pattern (**Aizawa, Y., Chinushi, M., Hasegawa, K., Naiki, N., Horie, M., Kaneko, Y.,… Fukuda, K. (2013). Electrical Storm in Idiopathic Ventricular Fibrillation Is Associated With Early Repolarization. Journal of the American College of Cardiology, 62 (11), 1015–1019 . Doi: 10.1016 / j.jacc.2013.05.030**), in addition to the prolongation of the QTc product of antipsychotics and antidepressants. I believe that he has sinus rhythm at all times 125 bpm and 107 bpm in the 3rd register where the prolonged QTc persists, visible in V1 around 480 msec. the dates you indicate are due to the alternance of the T wave characteristic of prolonged QT/QTc interval.

Waiting for the opinions of the Forum Masters, I cordially say goodbye

Juan Carlos Manzzardo MD

Hola. Tremenda secuencia de ECGs.!!!!

ECG 1. Complejo "QRS-ST-T triangular." El QRS es ancho por el supradesnivel del segmento ST. y no por QRS (RR 480 mseg). La parte inicial del QRS es rápida y luego se ensancha debido al ST y no al QRS, por lo que la deflexión previa al QRS podría ser una onda P y estaríamos en presencia de taquicardia sinusal?. Además tiene alternancia de la onda T. Este hayazgo es típico del síndrome del QT largo. El QRS-ST-T triangular se observa en oclusiones agudas arteriales coronarias (en este caso de tronco coronaria izquierda por lo difuso) y por espasmo coronario ya que no se demostró enfermedad obstructiva.

ECG 2. Es evolutivo del primero, con descenso del ST. y continua la alternancia de la onda T. RR 480 mseg. Igual FC al primero.

ECG 3. Repolarización prolongada que llega o sobrepasa la onda P, si es la P.

El consumo de drogas (cocaína) y fármacos antidepresivos-antipsicóticos asociado a alteraciones hidroelectrolíticas por vómito y diarrea debe ser tenido en cuenta. El laboratorio post paro cardíaco siempre dan alterados.

Sería interesante ver un ECG más bradicárdico para diferenciar mejor la repolarización ventricular.

Esperemos los comentarios de los expertos.

Saludos.

Oscar Pellizón.MD Universidad Nacional De Rosario. Santa Fe. Argentina
Cardiologo Universitario. Electrofisiologo. Doctor en Medicina

Hi. Tremendous sequence of ECGs. !!!!

ECG 1. Complex "QRS-ST-T triangular." The QRS is wide due to the ST segment elevation. and not by QRS (RR 480 msec). The initial part of the QRS is fast and then widens due to the ST and not the QRS, so the deflection prior to the QRS could be a P wave and we would be in the presence of sinus tachycardia. It also has alternation of the T wave. This finding is typical of long QT syndrome. Triangular QRS-ST-T is observed in acute coronary artery occlusions (in this case, the left main coronary artery because has a diffuse compromise) and coronary spasm since obstructive disease was not demonstrated.

ECG 2. It is evolutionary of the first, with ST segmente decrease. and the alternation of the T wave continues. RR 480 msec. Equal HR related to the first one.

ECG 3. Prolonged repolarization that reaches or exceeds the P wave, if it is P.

The consumption of cocaine with antidepressant-antipsychotic drugs associated with electrolyte disturbances due to vomiting and diarrhea must be taken into account. The laboratory post cardiac arrest always give altered.

It would be interesting to see a more bradycardial ECG to better differentiate ventricular repolarization.

Let's wait for the experts' comments.

Regards.

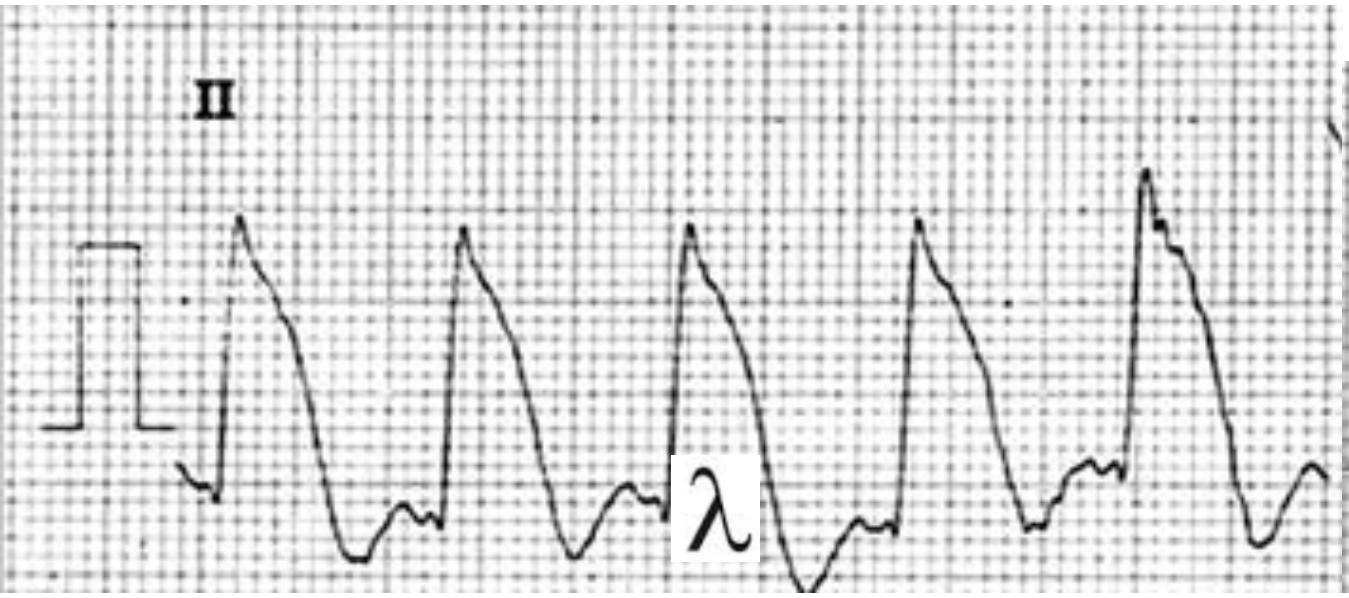
Oscar Pellizón MD PhD

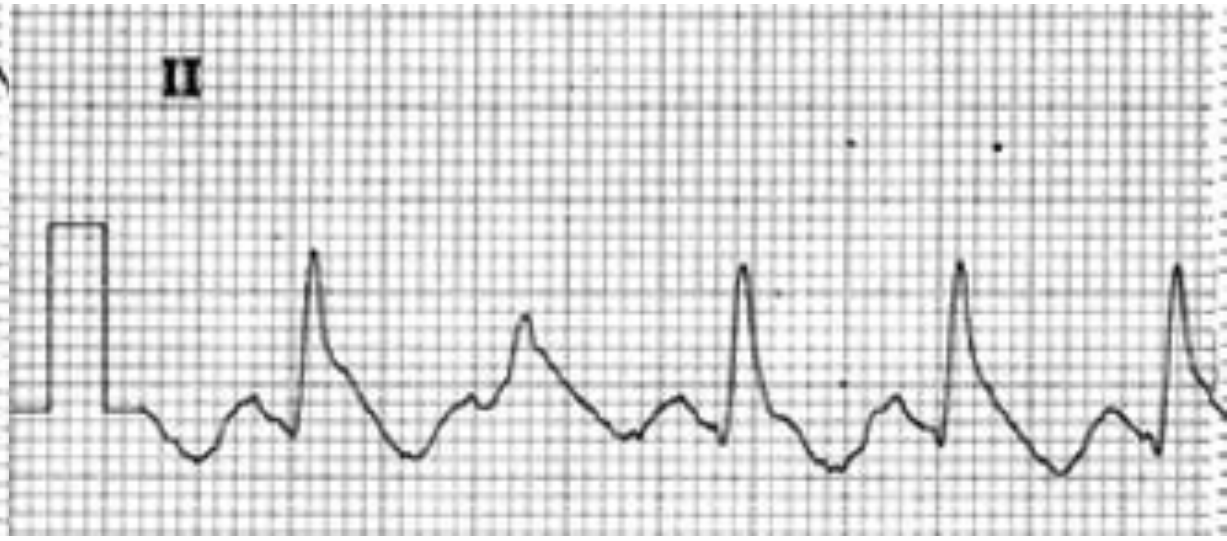
Final comments

by Andrés Ricardo Pérez-Riera MD, PhD& Acácio F. Cardoso MD, PhD

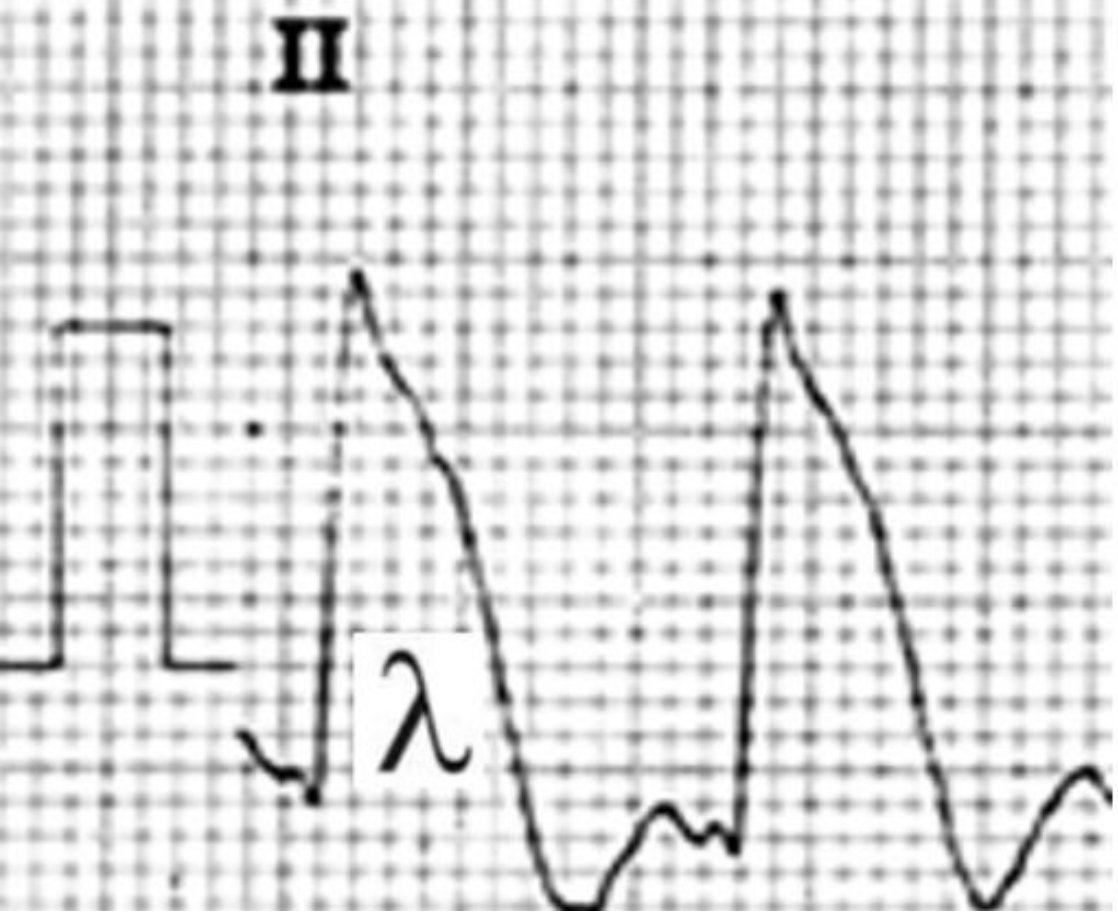
Broad ST-T Abnormalities on ECG

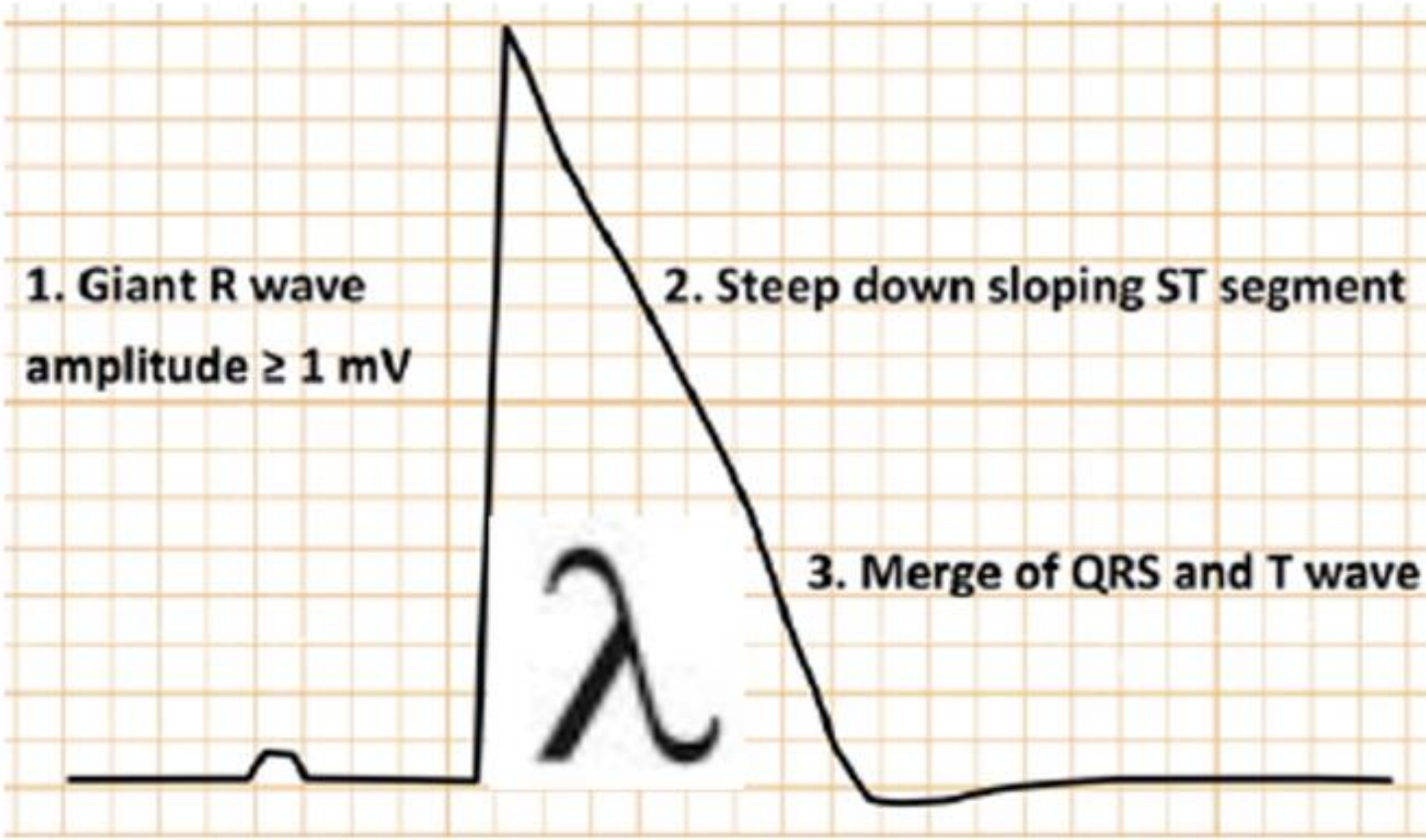
1. **Lambda wave or End-QRS slurring**
2. **End-QRS notching**
3. **Tombstone pattern**
4. **Shark Fin pattern. Giant R wave or Triangular QRS-ST-T wave**
5. **Transiet ST-segment elevation immediately after direct-current (DC)**
6. **Coronary vasoespasm (Prinzmetal Angina)**
7. **Brugada syndrome, Brugada phenocopies, and acquired Brugada syndrome(drugs)**
8. **Spiket Helmet Signal**
9. **Immediately after Direct-Current (DC) countershock**
10. **Pericardites, bundle brunch block, LVH, etc)**





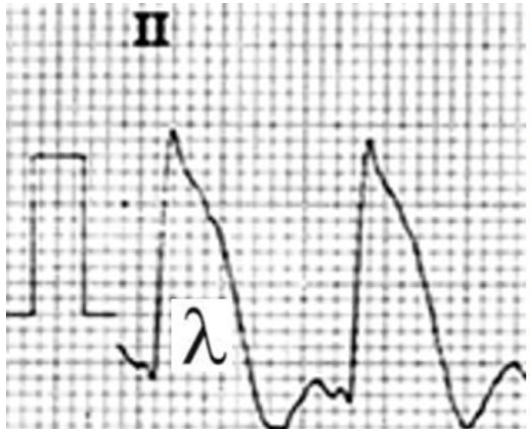
One of the ST segment abnormalities is a lambda-like ST-elevation pattern, which was first described by our group sixteen years ago **(Riera AR, Ferreira C, Schapachnik E, et al. Brugada syndrome with atypical ECG: downsloping ST-segment elevation in inferior leads. J Electrocardiol 2004;37:101–4.)** and was further characterized by Gussak et al. **(Gussak I, Bjerregaard P, Kostis J. Electrocardiographic "lambda" wave and primary idiopathic cardiac asystole: a new clinical syndrome? J Electrocardiol 2004;37:105–7.)**. On ECG the QRS complex, with its both slurry up-sloping and steep down-sloping limbs, resembles the Greek letter lambda**. (Guangqiang Wang, MS,a,∗ Na Zhao, MS,b Chuanhuan Zhang, MS,a Shu Zhong, BS,a and Xuexun Li, Msa. Lambda-like ST-segment elevation in acute myocardial infarction triggered by coronary spasm may be a new risk predictor for lethal ventricular arrhythmia Medicine (Baltimore). 2018 Dec; 97(49): e13561. Published online 2018 Dec 10. doi: 10.1097/MD.0000000000013561)**. Lambda wave probably shares sometimes the same electrophysiological mechanism than "spike helmet" sign (SHs) eventually.



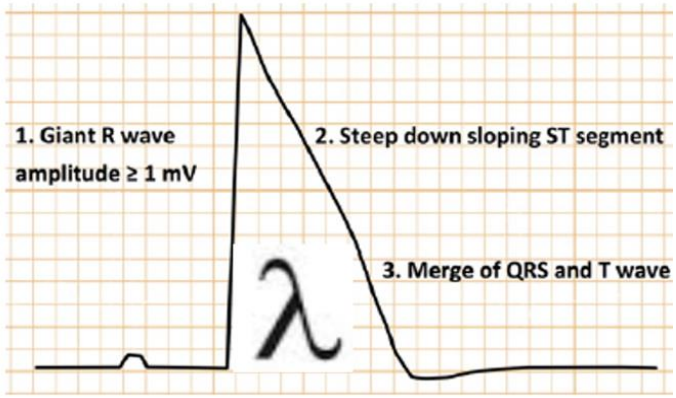


Lambda wave of the present case and Schematic representation of the Shark Fin pattern or triangular QRS-ST-T

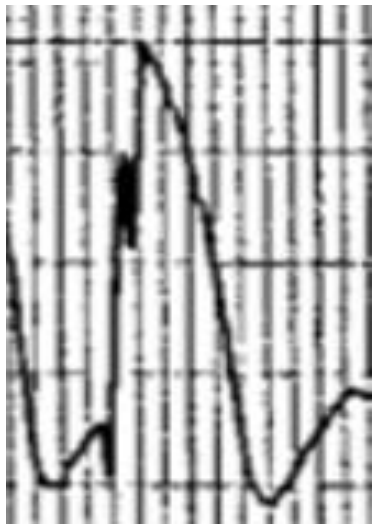
Lambda wave = Extreme Early repolarization pattern variant End-QRS slurring = Shark Fin pattern = Triangular QRS-ST-T = Type 1 Brugada pattern or coved type= Transient ST-segment elevation immediately after direct-current (DC) = Spiket Helmet Signal= Tombstone pattern = Coronary vasospasm Prizmetal



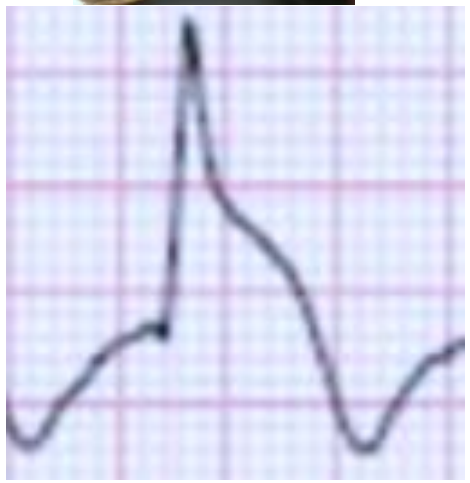
The present case



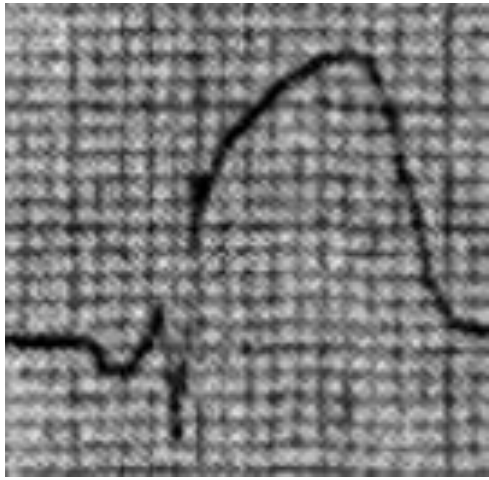
Shark Fin pattern or triangular QRS-ST-T



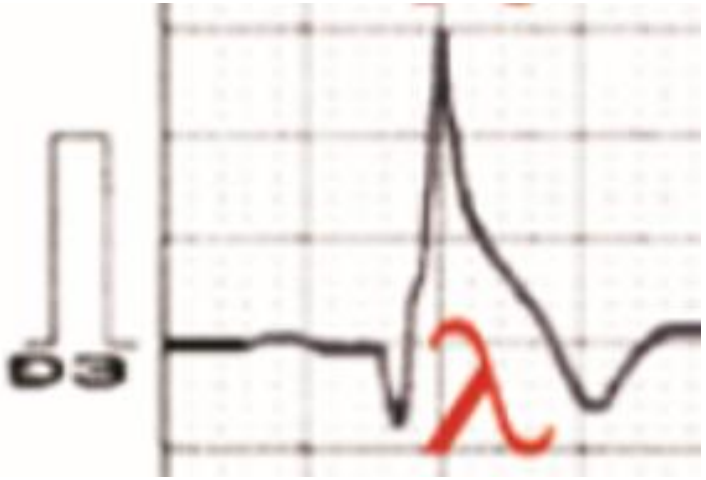
Immediately after Direct-Current (DC) countershock



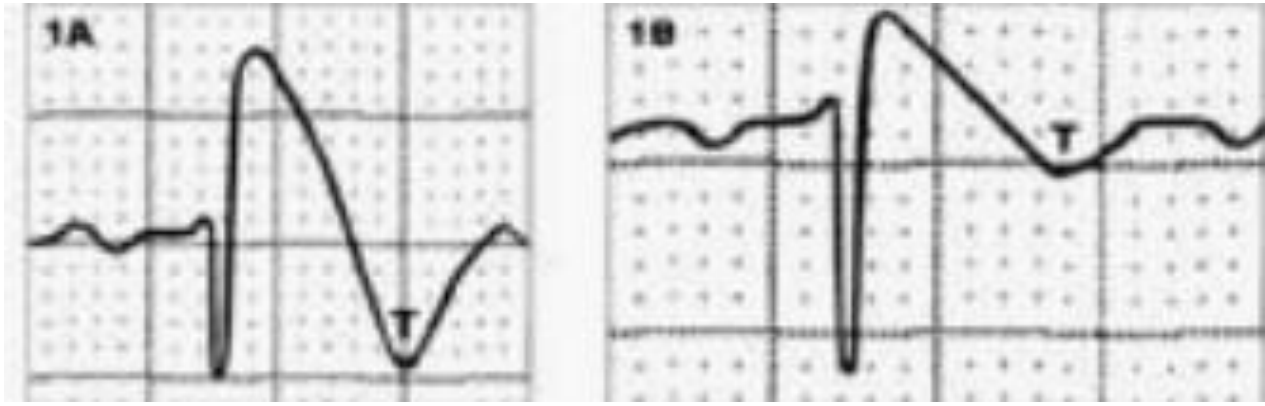
Spiket Helmet Signal



Tombstone pattern

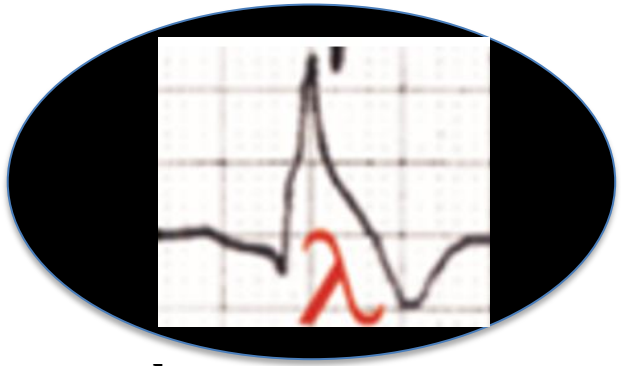


Lambda wave or End-QRS slurring

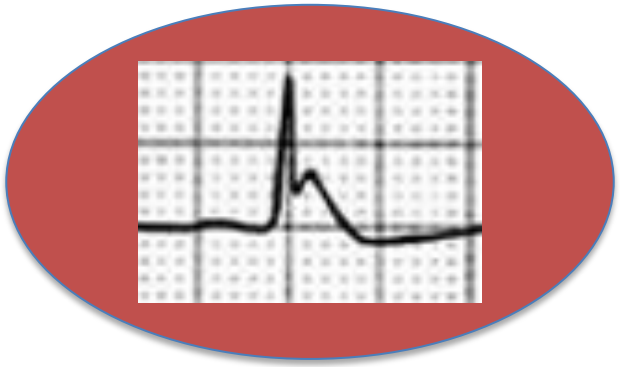


Type 1 Brugada pattern, coved type or Brugada sign

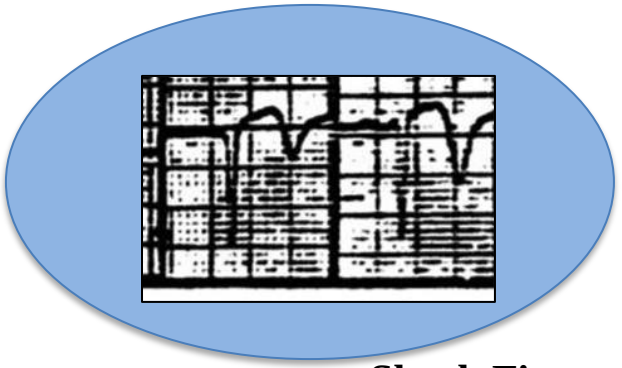
Lambda wave or End-QRS slurring,



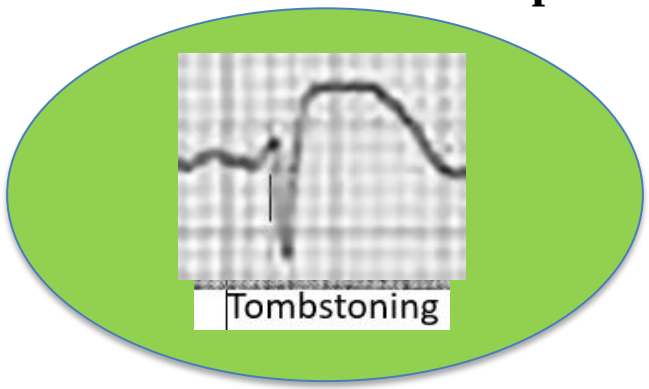
End-QRS notching



Prinzmetal Angina

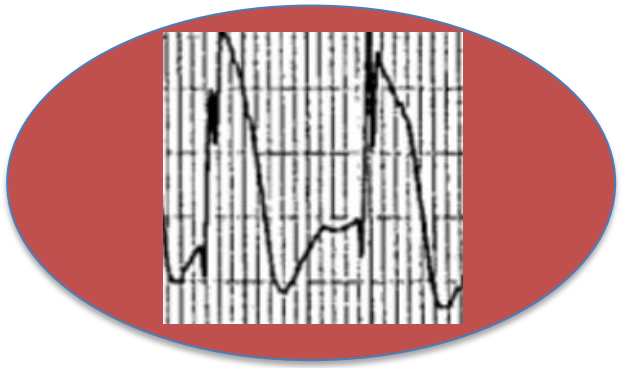


Tombstone or tombstone pattern

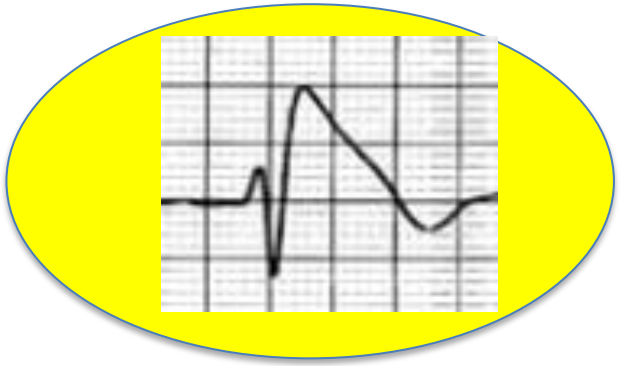


"Shark Fin", Lambda wave or End-QRS slurring, End QRS notching, spontaneous type 1 Brugada pattern, The “Spiked Helmet” Sign, Tombstone or tombstone pattern, Transient ST-Segment Elevation Immediately after Direct-Current (DC) and Prinzmetal angina are deadly ominous ECG Signs with similar changes in ST-segment.

Shark Fin



Transient ST-Segment Elevation Immediately after Direct-Current (DC)



Type 1 Brugada pattern V1/V2

The “Spiked Helmet”

Lambda wave (“shark fin”) pattern in patients with apical ballooning syndrome and unstable hemodynamics. characteristic minimal notch separating QRS complex from ST and the ST-elevation amplitude exceeding R-wave amplitude (**Madias, J. E. (2018). “Spiked Helmet” electrocardiogram sign in a patient with takotsubo syndrome: Similarities with a previously described marker. American Journal of Emergency Medicine, 36, 1696 10.1016/j.ajem.2018**.01.025). “Spiked Helmet” ECG sign in a patient with apical ballooning syndrome : Similarities with a previously described marker by Samadov et al (**Samadov, F. , Gasimov, E. , Aliyev, F. , & Isayev, E. (2018). The, “Spiked Helmet” sign - A potential relationship to Takotsubo cardiomyopathy. American Journal of Emergency Medicine, 36, 345.e5–345.e7. 10.1016/j.ajem.2017.11.041**). However, we can hypothesize substantial differences between Lambda wave in apical ballooning syndrome with both SHs sign and Lambda wave initially described by **Kukla (Kukla, P. , Jastrzebski, M. , Sacha, J. , & Bryniarski, L. (2008). Lambda-like ST segment elevation in acute myocardial infarction - a new risk marker for ventricular fibrillation? Three case reports. Kardiologia Polska, 66, 873–877. discussi[on 877-8**.) and Riera (**Riera, A. R. , Ferreira, C. , Schapachnik, E. , Sanches, P. C. , & Moffa, P. J. (2004). Brugada syndrome with atypical ECG: Downsloping ST-segment elevation in inferior leads. Journal of Electrocardiology, 37, 101–104. 10.1016/j.jelectrocard.2004.01**.002 . From a clinical perspective, despite the general poor prognosis characterizing subjects with Lambda wave ECG, Lambda wave aspect was originally associated with malignant arrhythmias; in Tarantino study, severe ST-elevation was, instead, predominantly associated with hemodynamic instability. Additionally, ST-elevation in their population occurred in elderly women with apical ballooning syndrome, whereas the Lambda wave and “Spiked Helmet” ECG sign apparently regarded mainly younger males (**Kukla et al., 2008; Riera et al., 2004; Samadov et al., 2018**), in different clinical settings. Furthermore, if compared with “classic” lambda wave by Riera et al. and Kukla et al. ECGs found in Tarantino cohort are characterized by a less evident notch between QRS and ST, and ST-elevation amplitude often exceeding R-wave amplitude, just resembling more a “shark fin” than either Lambda wave or “Spiked Helmet” ECG sign (Figure slide 22).

Tarantino "shark fin" sign, instead, in some aspects rather reminds "tombstone" ST-elevation, an ECG pattern associated with proximal stenosis of the left anterior descending coronary artery and large anterior ischemia (**Sinha, M. K. , Dasgupta, D. , & Lyons, J. P. (2004). "Tombstone" ST segment elevation of acute myocardial infarction. Postgraduate Medical Journal, 80, 276 10.1136/pgmj.2003.010751**). However, "shark fin" sign might hypothetically feature high-risk hemodynamics in apical ballooning syndrome patients with transient severe LV dysfunction transient LV dysfunction rather than represent just myocardial ischemia real myocardial ischemia. Therefore, ST-elevation exceeding R wave amplitude - which is probably initially attenuated by apical ballooning syndrome itself (**Madias, J. E. (2014). Transient attenuation of the amplitude of the QRS complexes in the diagnosis of Takotsubo syndrome. European Heart Journal Acute Cardiovascular Care, 3, 28–36. 10.1177/2048872613504311**), and ST-elevation amplitude exceeding R-wave amplitude, probably transiently attenuated by apical ballooning syndrome , could be useful in distinguishing two different ECG patterns. Furthermore, tall J-waves are known to possibly anticipate ST-elevation in apical ballooning syndrome (**Santoro, F. , Ieva, R. , Ferraretti, A. , De Gennaro, L. , Michele, L. , Di Biase, M. , & Brunetti, N. D. (2013). Diffuse ST-elevation following J-wave presentation as an uncommon electrocardiogram pattern of Tako-Tsubo cardiomyopathy. Heart and Lung, 42, 375–378. 10.1016/j.hrtlng.2013.05.002**). It might be therefore of interest to investigate possible correlations between gender, age, and specific ECG morphologies which stem from different pathophysiological pathways, all falling into the wider spectrum of Nonacute Coronary Syndrome related ST-elevation patterns (NASTEP), but with different etiologies and different electrophysiology.

Eighty percent of Lambda wave patients suffered from cognitive decline, therefore it was nearly impossible to objectively and reliably record the real incidence of chest pain. The principal symptom in two patients was chest/epigastric discomfort, which was considered as an angina

equivalent. Because of unstable hemodynamics, nitroglycerin was not tested. However, intravenous diuretics administered to treat pulmonary congestion could explain the fluctuant pattern of ST-elevation, probably through the same mechanism of nitroglycerin that is preload variations.

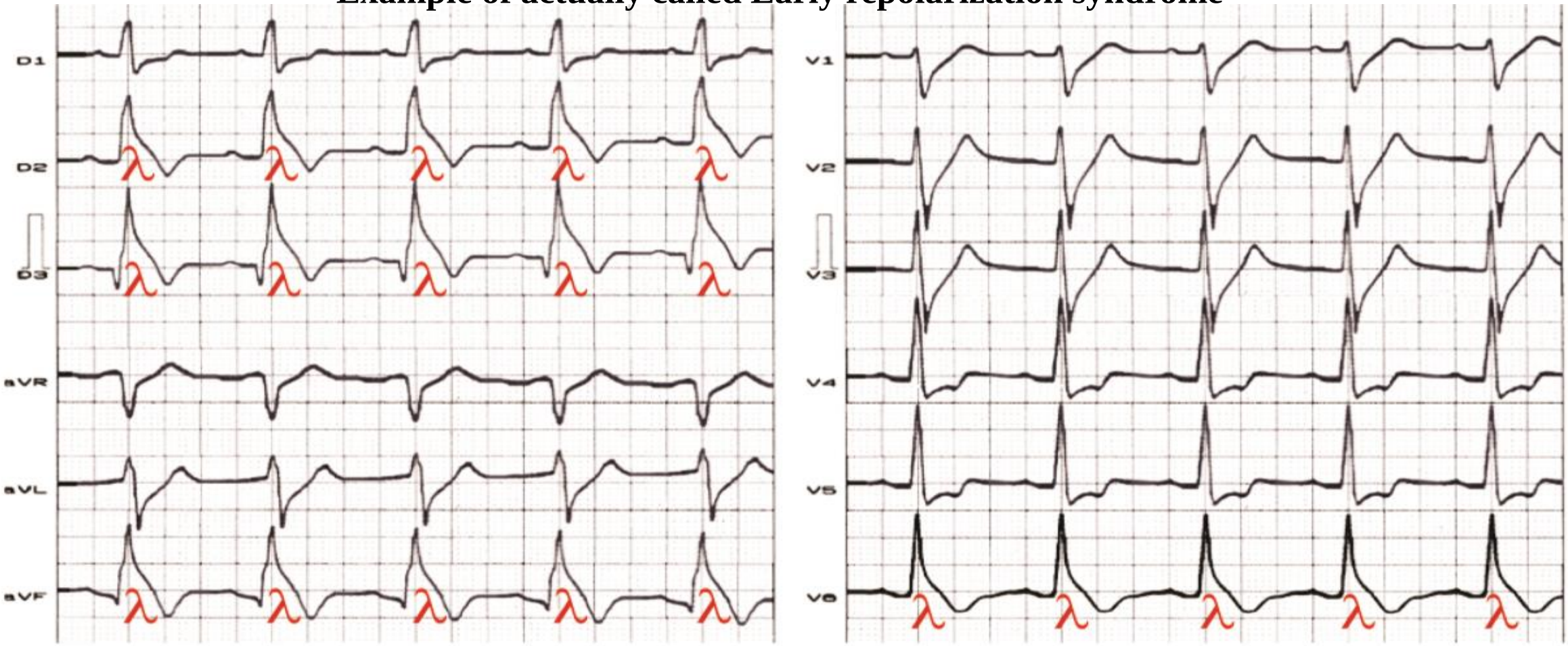
Serial transthoracic echocardiograms were obtained at the presentation and every other day in each patient as per protocol. The Lambda wave group presented only typical apical ballooning syndrome with basal hyperkinesia. A noninvasive method to assess sympathetic status would be helpful to better monitor in-hospital stay and prevent complications. Future studies can be jointly carried out in this direction.

Unfortunately, pre- apical ballooning syndrome ECGs can be compared in only two patients: one already reported in Figure 4b (T**Tarantino, N. , Santoro, F. , Guastafierro, F. , Di Martino, L. F. M. , Scarcia, M. , Ieva, R. , … Brunetti, N. D. (2018). "Lambda-wave" ST elevation is associated with severe prognosis in stress (takotsubo) cardiomyopathy. Annals of Noninvasive Electrocardiology, July, e12581 10.1111/anec.12581**), and a transient apical ballooning syndrome -related reduction of QRS amplitude was notable. Moreover, two patients out of five deceased during in-hospital stay, therefore no ECG could be obtained after discharge. The remainders were followed up via telephone follow-up.

War helmets, Greek Letters Lamda and shark fins: the conundrum of nonacute coronary syndrome related ST-elevation patterns (NASTEP) still remains largely unresolved.

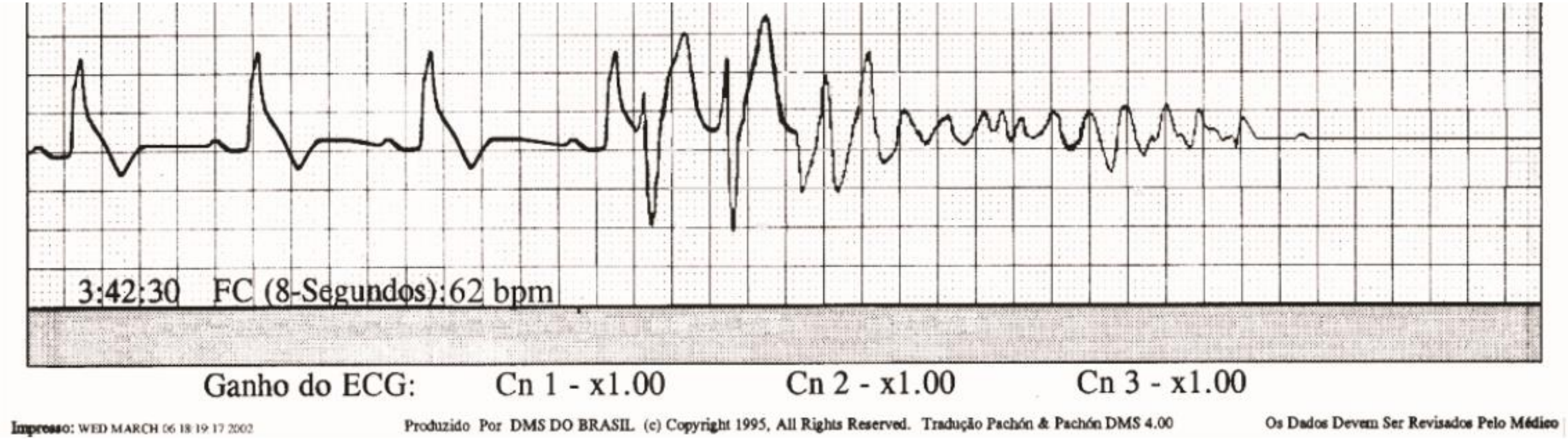
The spiked-helmet sign (SHS) was first described by Littmann at al in 2011 as a pseudo and transitory ST-segment elevation related to a severe non-cardiac illness clinical conditions, without cardiac biomarkers elevation and a high mortality course (**Littmann L, Monroe MH. The "spiked helmet" sign: a new eletrocardiographic marker of critical illness and high risk of death. Mayo Clin Proc 2011;86(12):1245-6**.). In this series, 6 of 8 patients died after the ECG index within 1 to 10 days. Initially described as ST-segment elevation restricted to inferior leads, new patterns have been reported with the involvement of multiple electrocardiographic leads (**Darek C, Hoshiar A, Adrian B. An ominous ECG signal in critical care. Circulation 2020;141:2106-2109**.). The morphology shown on the ECG resembles Pickelhaube, a spiked helmet worn in the 19th and 20th centuries by Prussian and Germany military. As other conditions related to ST-segment elevation.

Example of actually called Early repolarization syndrome



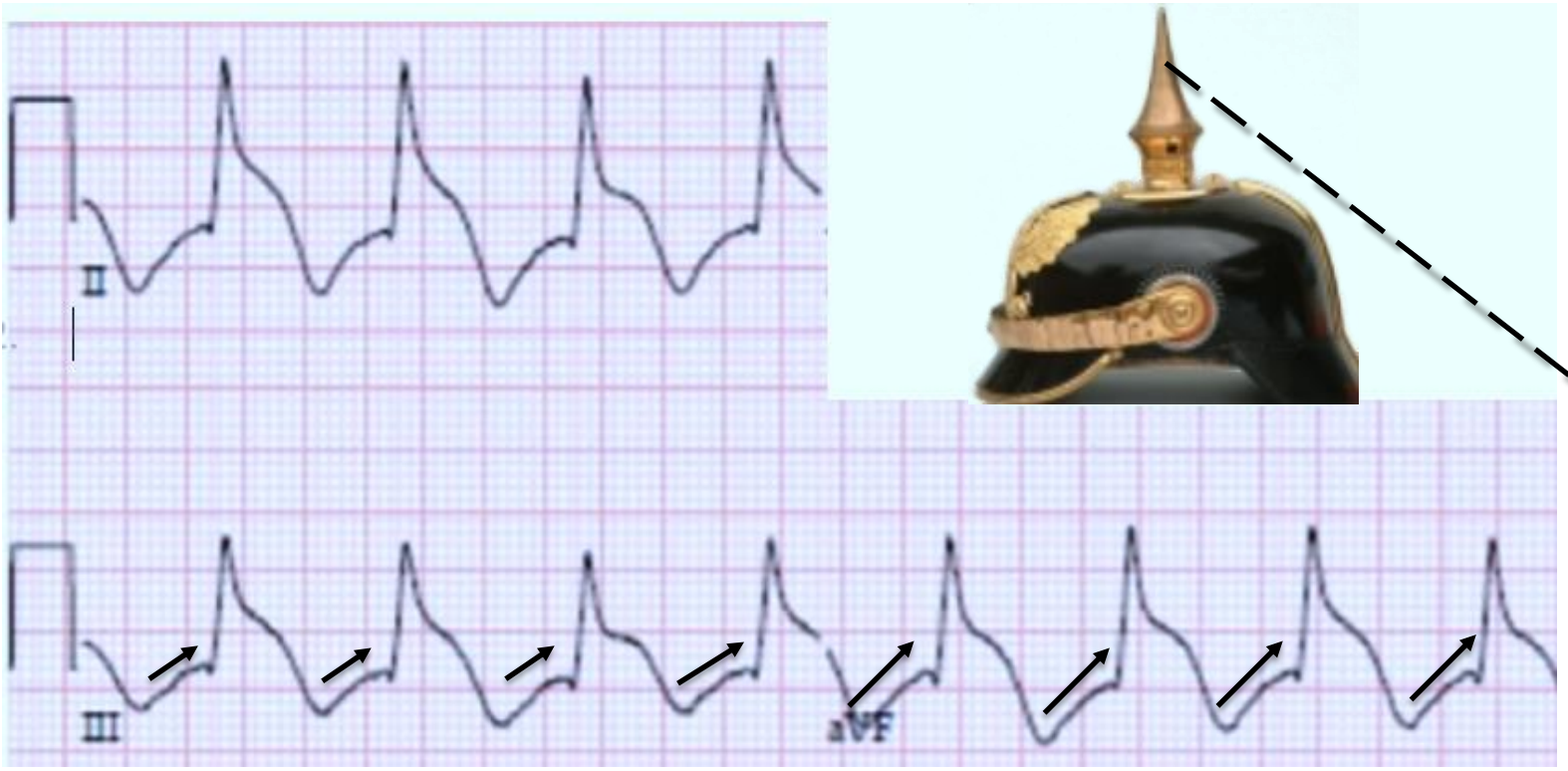
The ECG shows persistent ST segment elevation in the inferior and apical lateral leads, associated to concomitant reciprocal or mirror image in the anterior wall that was not modified with the use of sublingual nitrate in absence of hypothermia, electrolyte imbalance or ischemia. This Lambda wave correspond to J wave and/or QRS slurring The "J wave" (also referred to as "the Osborn wave," "the J deflection," or "the camel's hump") is a distinctive deflection occurring at the QRS-ST junction. In 1953, Dr. John Osborn described the "J wave" as an "injury current" resulting in VF during experimental hypothermia.

Riera AR, Ferreira C, Schapachnik E, et al. Brugada syndrome with atypical ECG: downsloping ST-segment elevation in inferior leads. J Electrocardiol. 2004 Apr; 37: 101-104.

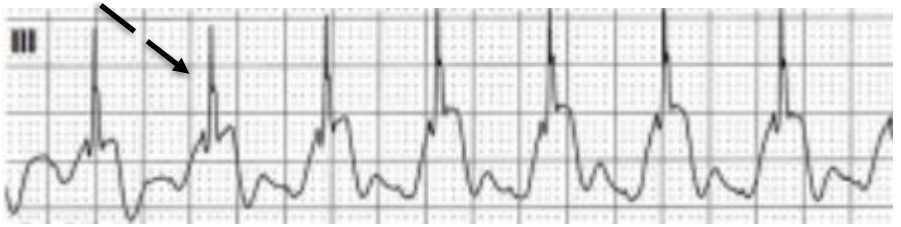


Holter monitoring recorded the final event, manifest by PVT episode with initial short-coupling ventricular premature contractions (R on T) that ended quickly in VF and asystole. Pattern 1C of repolarization has been observed in acute myocardial infarction by Kukla et al (**Piotr Kukla 1, Marek Jastrzebski, Jerzy Sacha, Leszek Bryniarski. Lambda-like ST segment elevation in acute myocardial infarction - a new risk marker for ventricular fibrillation? Three case reportsKardiol Pol 2008 Aug;66(8):873-7; discussion 877-8.**). These authors raised the hypothesis that the "Lambda-like ST" could be a new marker of risk of acute infarction with ST segment elevation.

The “Spiked Helmet” Sign (SHS): Clinical scenario critical noncardiac illness.; Lead affected: multiple, main inferior lead II, III and aVF.; mortality rate: high in-hospital death. 75% to 95%.; **Laslo Littmann, MD, PhD and Michael H. Monroe, MDThe “Spiked Helmet” Sign: A New Electrocardiographic Marker of Critical Illness and High Risk of DeathMayo Clin Proc. 2011 Dec; 86(12): 1245–1246. doi: 10.4065/mcp.2011.0647** ECG Characteristic: unique pattern of apparent STEMI whose presence was found to be associated with critical illness and very high risk.



This characteristic ECG pattern was named for its resemblance to the Prussian military helmet, the Pickelhaube (**Derek Crinion, Hoshiar Abdollah, Adrian Baranchuk. An Ominous ECG Sign in Critical Care. Circulation. 2020;141:210621092020 https://doi.org/10.1161/CIRCULATIONAHA.120.047427**



The elevation of the isoelectric line precedes the QRS, followed by a sharp R wave and then convex to the top ST-segment elevation followed by negative T wave. Exclusively registered in the inferior leads SHS mimics STSEAMI infarction. However, the upward shift preceding the QRS that can appear to align with the ST-segment elevation is not consistent with acute coronary syndrome (**Laslo Littmann, MD, PhD and Michael H. Monroe, MD. The “Spiked Helmet” Sign: A New Electrocardiographic Marker of Critical Illness and High Risk of DeathMayo Clin Proc. 2011 Dec; 86(12): 1245–1246. doi: 10.4065/mcp.2011.0647).**

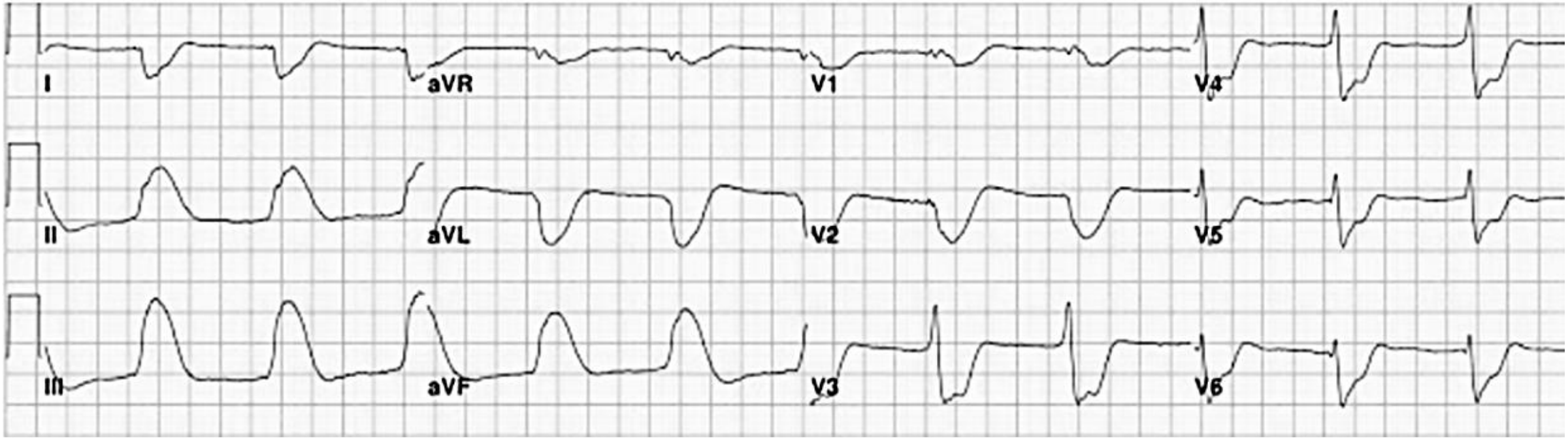
SHS can mimic an acute myocardial infarction. The absence of Q-wave formation and T-wave inversion after the ST-segment regression are elements that can help to distinguish these two clinical situations.

The main features on ECG are shown as an elevation to the isoelectric line that precedes the QRS, followed by a sharp R wave and then a convex ST-segment elevation (**Darek C, Hoshiar A, Adrian B. An ominous ECG signal in critical care. Circulation 2020;141:2106-2109**.).. The pathophysiologic mechanisms related to these ECG patterns are not totally clarified. The previous giant T-U waves that advance over the next QRS and or a prolongation of the repolarization that are superimposed to a faster heart rate, are attributed to some authors(**Laundon RK, Littmann L. Spiked helmet pattern ST elevation in subarachnoid hemorrhage. J Eletrocardiol 2019;52:96-8.**). The initial cases were identified in abdominal and thoracic pathologies and associated with muscle artifacts and an acute increased pressure in these cavities. Subsequently, other cases involving acute brain bleeding, severe metabolic disorders, and sepses states, pointed to participation in the critical illness with intense adrenergic discharge as a final pathway for this presentation. It can be reinforced by other hyperadrenergic states related to SHS as observed after the stellated ganglionar ablation (**Aliyev F, Abdulkerimov V, Gul EE, Samedov F, Isayev E, Ferecov E. Spiked helmet sign after percutaneous left stellate ganglion ablation in a patient with long QT syndrome. J Eletrocardiol 2017;50(6):944-6**.) and taktsubo cardiomyopathy[5].(**Samadov F, Gasimov E, Aliyev F, Isayev E. Yhe "spiked helmet" sign – a potencial relationship to Takotsubo cardiomyopathy. Am J Emerg Med 2018;36(2) [345 e345-345 e347**)

The macro T-wave alternans is a rare manifestation on ECG and reflects important dispersion of repolarization that precedes TdP and ventricular fibrillation. (**Sanjiv MN, T-wave alternans and the susceptibility to ventricular arrhytmias. JACC 2006;17:269-81**.) Commonly, this pattern is seen in a high risk congenital and acquired long QT syndromes and announces the Torsades Pointes.

beginning. In our case, the macro T-wave alternans was observed on ECG after an aborted ventricular fibrillation event (non documented). This presentation demonstrates that SHS works as a potential marker of ventricular repolarization prolongation associated with a fast heart rate in critical illness. These findings are according to the previous report by A. Simon et al that demonstrates a SHS case related to Taktsubo Cardiomiophaty, manifested through QT prolongation, T-wave alternans, and self-limited Torsade de Pointes (**Sanjiv MN, T-wave alternans and the susceptibility to ventricular arrhytmias. JACC 2006;17:269-81**.) Together, these cases alert us to the high risk of malignant ventricular arrhythmias in this scenario and collaborate to understand the repolarization abnormalities related to possible sudden death mechanisms in these acute and severe diseases.

Case report: A 75-year-old man collapses to the ground in cardiac arrest while shopping with his wife. Medically trained bystanders happen to witness the event and begin CPR right away. Paramedics rush to the scene and find the man to be in VF. He is intubated and shocked 3 times prior to arrival in the ED. He comes in with CPR in progress via LUCAS device and is now in slow PEA. An intra-arrest arterial line is placed. After 3 more rounds of chest compressions there is a sudden spike in ETCO2 and the A-line shows a BP of 70/40 mmHg. Bedside Echo reveals what appears to be stunned myocardium with low LVEF. on. Vasopressor infusions are started.

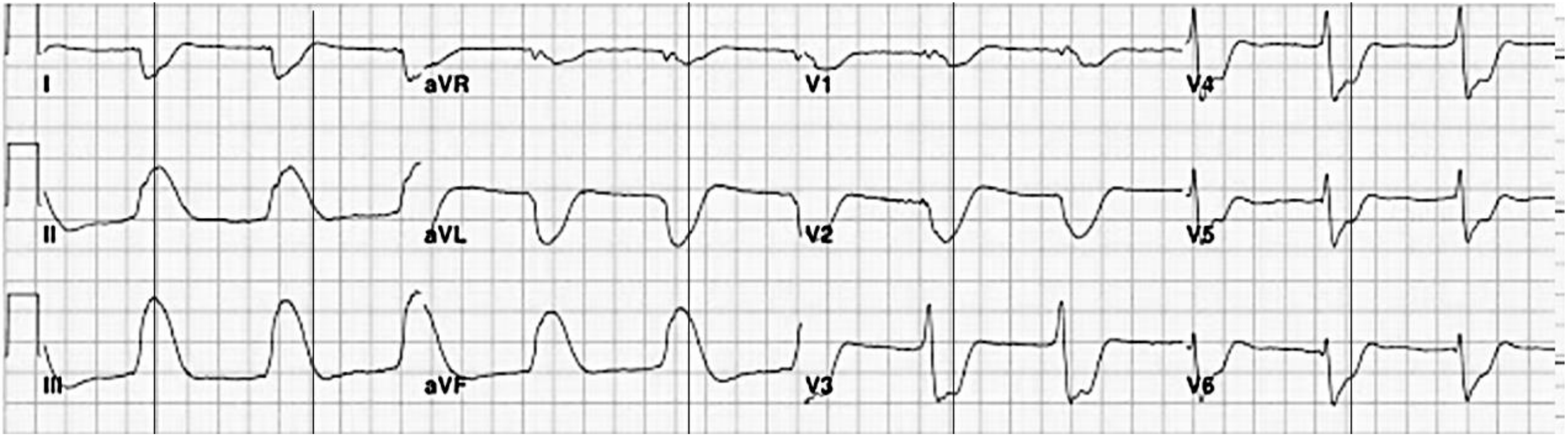


Broad regular junctional rhythm with very broad complexes (HR65 bpm) This wide one should think of toxicologic or metabolic causes of arrest. In particular severe hyperkalemia. But take a close look at the unique morphology of these particular complexes: notice how they look remarkably like Shark Fins. Another observation is bifasicualr block (RBBB+LAFB) While lacking amplitude, the QRS complex in lead V1, appears to be a triphasic, and associated with wide terminal S waves in all lateral leads. One could debate whether terminology is best served by calling this IVCD vs RBBB — but given the rS pattern in lead I (*with steep decline and predominant negativity from the S wave in this lead*) + predominant R waves in each of the inferior leads — I submit that the most logical explanation is *combined* RBBB/LPHB. They are not QRS complexes but rather a combination of QRS and T-wave. What they represent is massive ST-deviation! This is a junctional rhythm with massive ST-Elevation in leads the inferior leads , with concomitant massive reciprocal ST-depression in lateral leads I & aVLamd V4 to V6. There is also ST-Depression in the precordial leads maximal in V2-V4 consistent with latero basal involvement. What you are looking at is a Massive Inferolaterobasal STEMI!

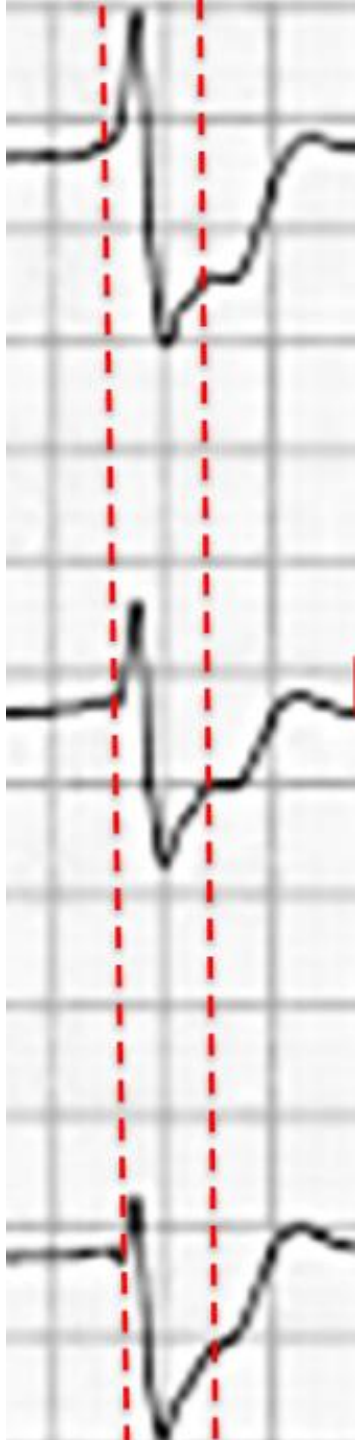
The key to seeing and understanding how these complexes represent profound ST-deviation lies in delineating the end of the QRS complex. The problem is that with this unique morphology, the QRS complex and T-wave merge together as a result of *extreme* STdeviation, and the two become indistinguishable. But remember this: If you can find the J point (end of the QRS and beginning of repolarization in one lead, you can find the end of the QRS in any lead. Look in all 12 leads and find one which clearly shows the end of the QRS. Here lead V5 happens to show the beginning and end of the QRS very nicely. Now all you have to do is simply draw a line straight up from this point (J-point) in V5, and you can find the same point in any lead.

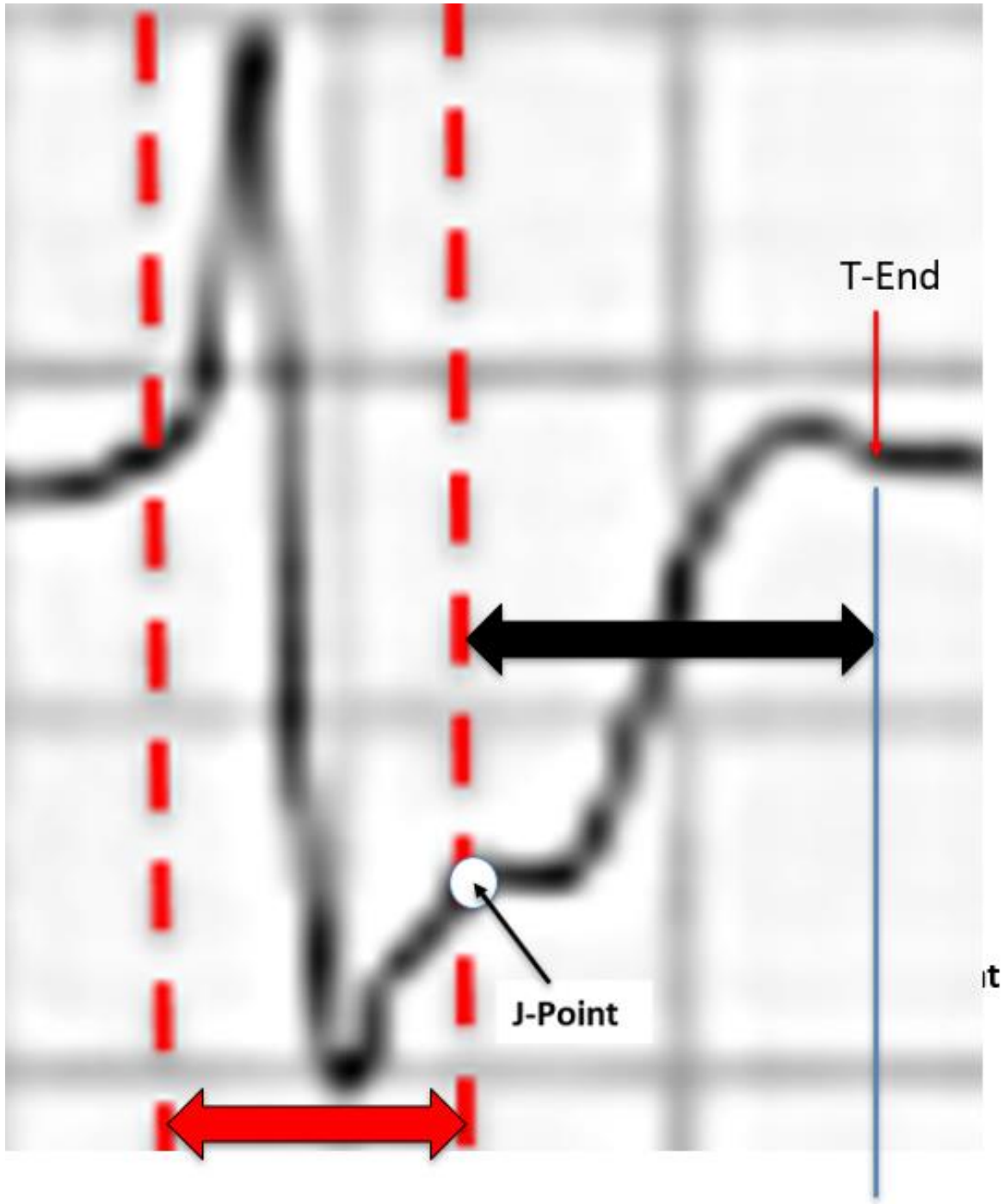
Shark fins pattern

Synonymous: Giant R wave or Triangular QRS-ST-T wave



The *profound* ST-seviations suddenly become glaringly obvious and you can now easily appreciate the classic pattern of Massive Infero-lateral STEMI!



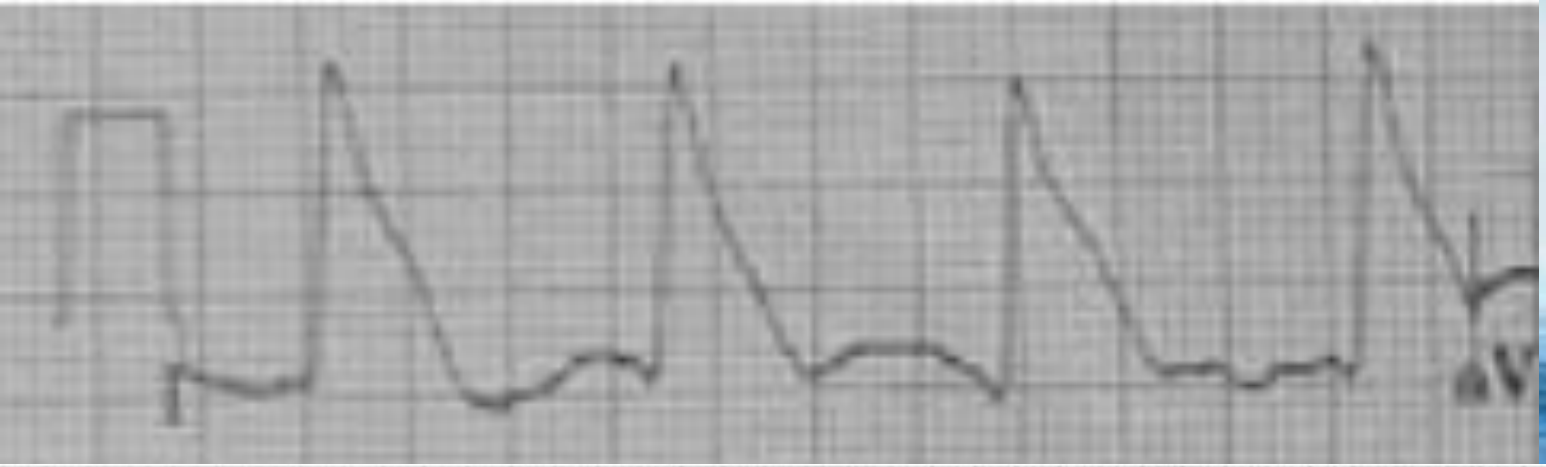


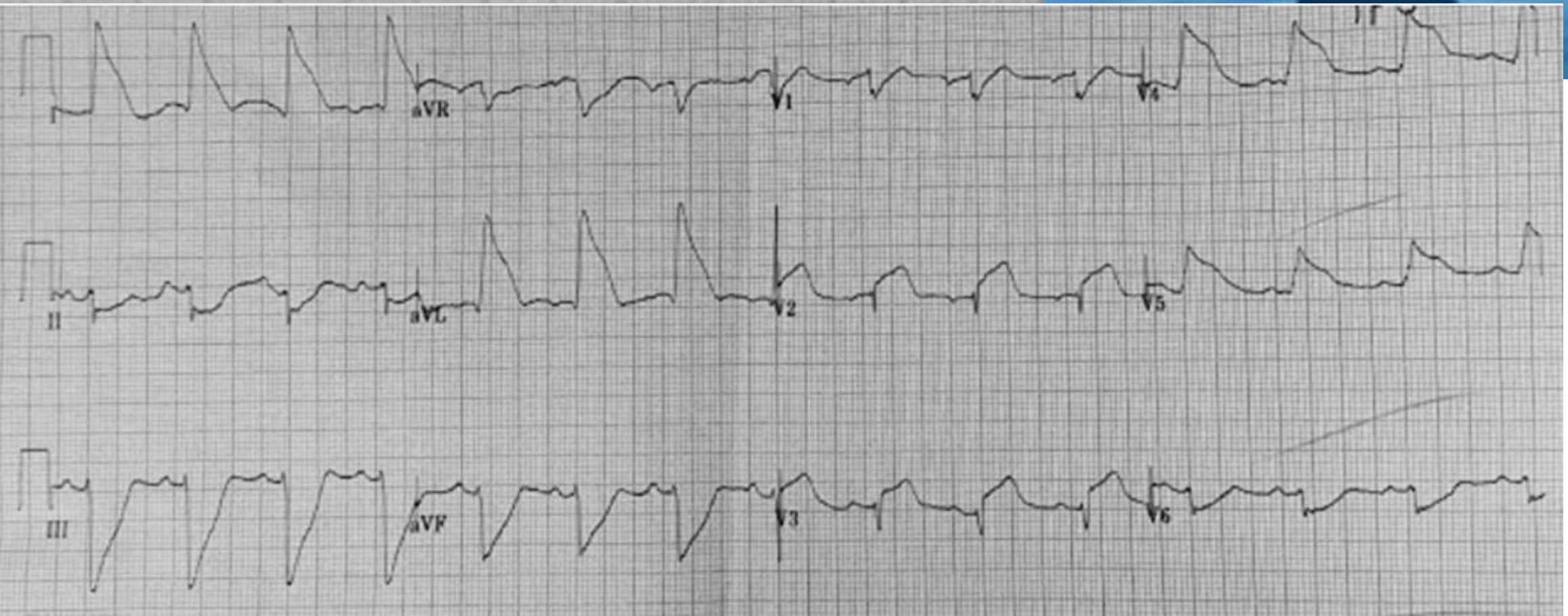
Depolarization (QRS)

Repolarization (ST-T)

The patient was given Aspirin and loaded with Ticagrelor via OG tube and the Cath Lab was immediately activated. Unfortunately, this was met with significant resistance. The Cardiology team was not familiar with this ECG phenomenon and there was an ongoing concern for hyperkalemia. Point-of-care laboratory testing was performed and revealed a normal K^+. Despite the normal lab result there was persistent concern for hyperkalemia. (It is worth noting that it is not uncommon for labs to show falsely elevated K^+. due to specimen hemolysis, but a falsely normal K^+. is exceedingly rare!) With hyperkalemia an ongoing focus, there was continued delay in catheterization. The patient was given multiple doses of calcium, insulin, glucose, and multiple ampules of sodium bicarbonate without any response or improvement of shock. He subsequently became bradycardic and a decision was made to pursue transvenous pacemaker insertion. In the process, the patient arrested once again and required an additional round of CPR to regain a perfusing circulation. Given persistent shock the decision was eventually made to proceed with coronary angiography, which revealed a 100% thrombotic RCA occlusion. During attempts to open and stent this culprit lesion the patient arrested yet again. Unfortunately this time he was unable to be resuscitated. The literature on this distinct ECG phenomenon is scant. Therefore, its incidence is unknown. Presumably many cases go unrecognized and are mistaken for conduction abnormalities, metabolic derangements, or toxicologic insult. From the cases that have been described, Shark Fin appears to be an ominous sign with a strikingly poor prognosis. A term that has been used in the literature to describe Shark Fin morphology is "Giant R-waves". This designation is suboptimal for a few reasons. Firstly, Shark Fin morphology represents extreme ST-*Deviation* which encapsulates both ST-Elevation as well as ST-Depression. ECG territories with Shark Fin reciprocal depression will not have R-waves, but rather S-waves. More importantly, the term "Giant R-wave" is problematic because it has also been used in the literature to refer to R-waves that are only mildly prominent and come nothing close in size or morphology to the ECG phenomenon described here.

"Shark Fin“, "Giant R-wave“ "Triangular QRS-ST-T waveform"

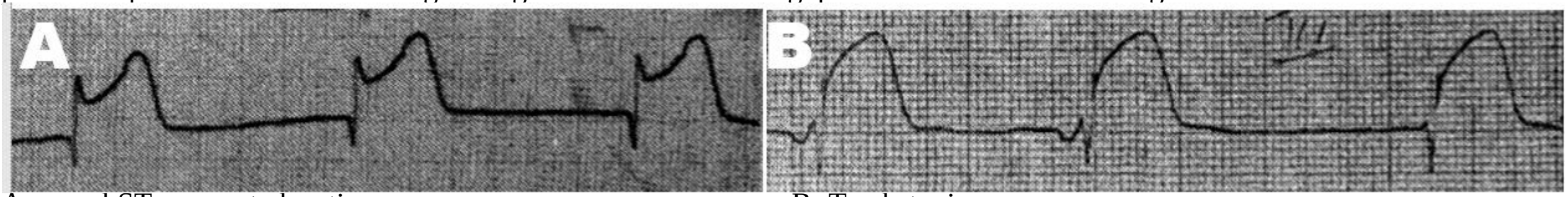




Drs Birnbaum and Schakowsky in JAMA editorial(**Birnbaum Y, Sclarovsky S. "Tombstoning" of ST segment in acute myocardial infarction. Lancet 1993; 342: 1494.**) wrote related to SiR- Wimalaratna manuscript (**H S Wimalaratna. "Tombstoning" of ST segment in acute myocardial infarction. Lancet. 1993 Aug 21;342(8869):496. DOI: 10.1016/0140-6736(93)91622-s**) witch describes the prognostic importance of a special ECG pattern of myocardial ischemia, which he termed "tombstoning". He states that the prognostic value of the various shapes of ST segment deviation during acute MI has not been assessed. Birnbaum and Schakowsky previously have reported the prognostic importance of the initial ECG pattern in patients with first acute MI involving the anterior wall (**Birnbaum Y, Sclarovsky S, Blum A, Mager A, Gabbay U. Prognostic significance of the initial electrocardiographic pattern in patients with a first acute anterior wall myocardial infarction. Chest 1993; 103: 1681-87. 2**). They studied 147 consecutive patients admitted in the early ECG stages of first anterior wall MI were assigned to one of three groups according to the pattern of the admission ECG: **Group A,** tall symmetrical abnormal T waves in the involved leads without ST elevation or major changes in the terminal portion of the QRS complex; **Group B,** abnormal T waves and ST elevation (> 0-1 1 mV) in two or more adjacent leads without major changes in the terminal portion of the QRS; and **Group C,** abnormal T waves and ST elevation with distortion of the terminal portion of the QRS complex (emergence of the J-point at a level above the lower half of the R-wave, or disappearance of S-wave in leads **Birnbaum Y, Sclarovsky S, Ben-Ami R, et al. Polymorphous ventricular tachycardia early after acute myocardial infarction. Am J Cardiol 1993; 71: 745-49.)(Sclarovsky S, Mager A, Kusniec J, et al. Electrocardiographic classification of acute myocardial infarction. Israel J Med Sci 1990; 26: 525-31.)**

Wimalaratna wrote that the ST SE is one of the earliest indicators of AMI, although it may occur in other causes of myocardial ischaemia.1 Various shapes of the ST segment in myocardial ischamia have been described, but the prognostic value in acute infarction has not been assessed. The term "tombstoning" is used by experienced junior doctors to describe a certain shape of the ST segment in the ECG of patients with AMI and

is noted on admission in some cases. This provocative term is often used to communicate to colleagues a grave prognosis of the patient in question. The tombstoning ST segment has some striking particular characteristics. Figure below



A= usual ST segment elevation, B=Tombstoning

The ST segment is convex upwards and has a fast rise-time, and these changes are seen in all the leads that have ST SE . The peak of the convex ST segment is often higher than the preceding R wave, which is of short duration (often less than 40ms) and small in amplitude. The ST segment merges with the ascending limb of the following T wave and therefore the T wave cannot be identified separately. Inversion of the T wave is not noted in tombstoning tracings. To evaluate the significance of this observation, SiR-Wimalaratna examined ECGs of 100 consecutive patients admitted with a history of AMI to intensive care in a district general hospital. The diagnosis was confirmed by rising title of cardiac enzymes (aspartate aminotransferase) over 3 consecutive days. 37 subjects were excluded because they had normal enzyme concentrations. The remaining 63 were divided into two groups according to the shape of the ST segment: tombstoning (n = 6) and usual ST changes (n = 57). Prognosis was assessed according to the number of complications that occurred during the first 7 days of hospital stay.

Complications monitored were: hypotension on admission (systolic blood pressure < 90 mm Hg), significant fall in blood pressure

(> 30% of admission pressure), ventricular or supraventricular tachyarrhythmias, PVCs of > 15 per min, complete heart block or new development of bundle branch block, cardiogenic shock as indicated by need to administer dopamine, hospital stay longer than 7 days because of complications, and death during first 7 days. All 6 patients with tombstoning ECGs had three or more complications. 4 died within 7 days. Only 16 (28%) patients with normal ST changes had two or more complications, and 2 of these patients died. Those who had complications but no tombstoning showed a varying degree of ST SE. 9 patients with no significant ST segment change had no complications. The difference between the two groups, as assessed by Fisher's exact test, was significant (p=0001). This preliminary study supports the notion that tombstoning may be a sign of bad prognosis in patients with AMI. Awareness of this variation may help save lives by prompt action. It could be argued that tombstoning is merely the presentation of a "hyper-acute state" or early change of the STSE after MI. However, the patients with these specific ST changes at admission have a poor prognosis. The mechanism of this particular ST change is difficult to explain, but it is likely to represent extensive and rapid myocardial damage after the ischemic episode. Although there was a statistically significant difference between the groups, numbers were small and not well balanced.

Bahattin et al studied 106 patients with tombstoning ECG patterns, concluding that infarction size is larger; left ventricular ejection fraction and preinfact angina are lower, and in-hospital complications are higher.(**Bahattin Balci 1, Osman Yesildag Correlation between clinical findings and the "tombstoning" electrocardiographic pattern in patients with anterior wall acute myocardial infarction. Am J Cardiol. 2003 Dec 1;92(11):1316-8. doi: 10.1016/j.amjcard.2003.08.014**.). Tombstoning ECG patterns (TOMB-ST) was observed in 25% of patients with STEMI and was associated with an increased mortality rate, higher incidence of HF, VF, decreased LVEF, increased mortality with independent of the total amplitude of ST(**Piotr Kukla 1, Dariusz Dudek, Kazimierz Szczuka. "Tombstoning" of ST segment in acute myocardial infarction -- effect on clinical courseKardiol Pol. 2006 Mar;64(3):275-80; discussion 281**.)

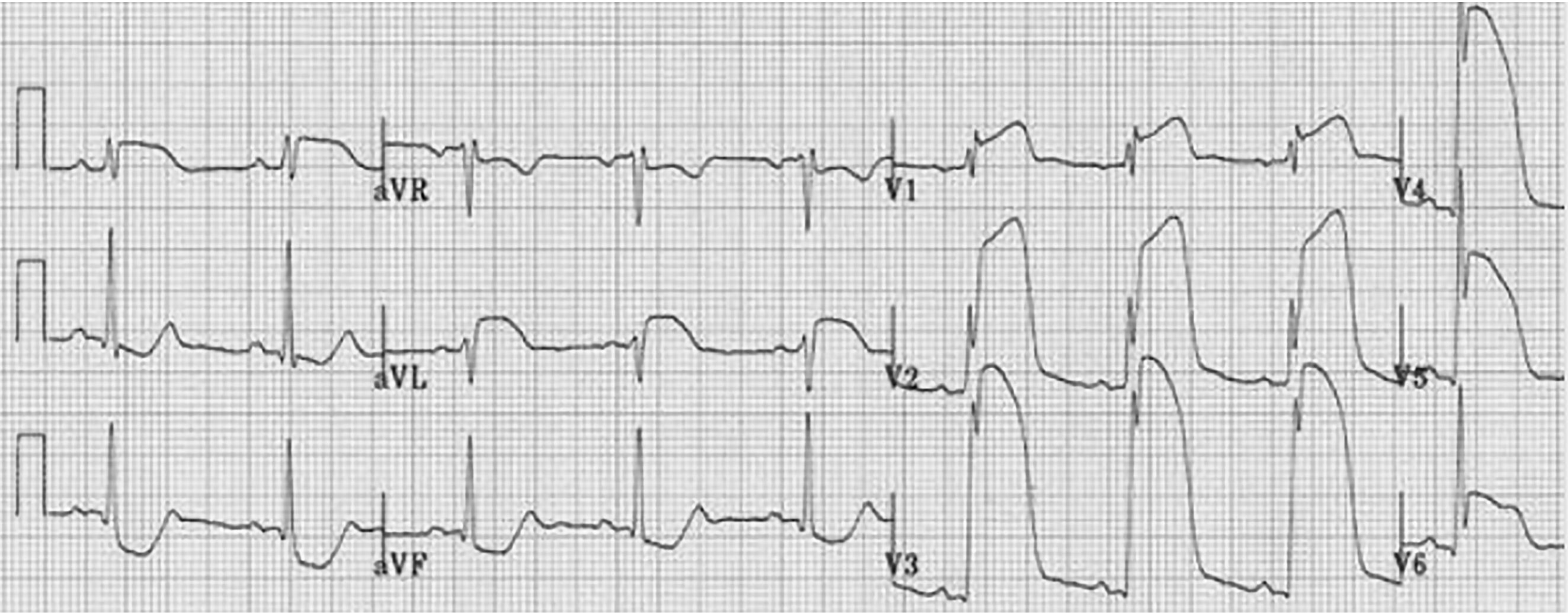
Tombstone criteria

The R wave is absent or its duration is <40ms with minimal amplitude;

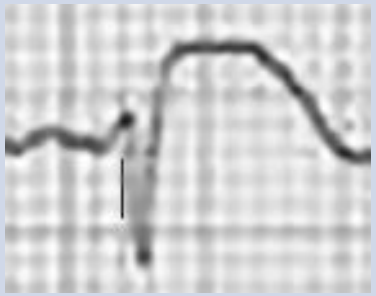
The ST segment is convex upward and merges with the descending limb of the R wave or the ascending limb of the QS wave;

The peak of the convex ST segment is higher than whatever remains of the R wave; and

The convex ST segment merges with the ascending limb of the T wave.

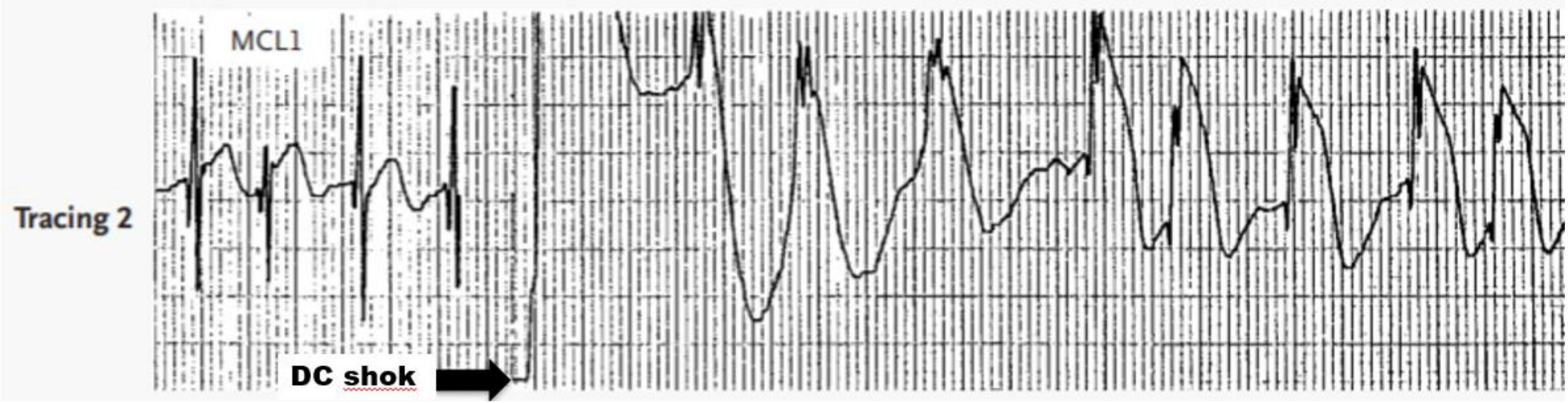


ECG changes in the differentiation of ischemic and non-ischemic ST elevation(Pericarditis, takotsubo cardiomyopathy and ER) in patients with acute chest pain and at least 0.1 mV or 1mm ST elevation.(1)

	Ischemic ST elevation	Non-ischemic ST elevation
Reciprocal ST depression Reciprocal effect or mirror image	Was associated with an ischemic diagnosis	ST depression in aVR and chest-lead PR depression were associated with a non-ischemic diagnosis.
Chest-lead PR depression	Uncommon in STEMI (12%) Only possible if atrial infarction associated	38%
ST-segment convex upward 	Present in 22% of STEMI cases	It is observed only in 9% of cases
QRS terminal distorsion	40%	7%
ST depression in lead II (≥0.025 mV)	Present in 40% of patients with STEMI	Absent
ST depression in lead I (≥0.025 mV)	Present in 83% of patients with STEMI	Absent

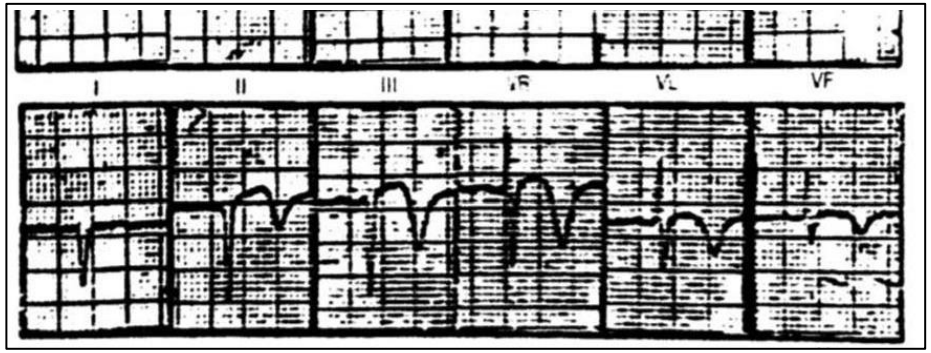
1. **Thomas Lindow, Olle Pahlm, Ardavan Khoshnood, Ingvar Nyman, Daniel Manna, Henrik Engblom, Annmarie Touborg Lassen, Ulf Ekelund. Electrocardiographic changes in the differentiation of ischemic and non-ischemic ST elevation. cand Cardiovasc J. 2020 Apr;54(2):100-107. doi: 10.1080/14017431.2019.1705383.**

Electrical cardioversion is one of the causes of ST-segment elevation. The prevalence varies between 15.4% and 48% and are more often after induced by monophasic DC shocks and in patients with lower left ventricular systolic function. The recording leads of ST changes are in accordance with an anterolateral location near the desfibrillation patches. Rumeau et al (Int J Cardiol 2011;148:341–6.) found that ST changes were only observed in precordial leads except V6, being more prominent in V1 in 5%, V2 in 70%, V4 in 15%, V5 in 5%, and V4R in 5%, without significant difference between ST depression or elevation. Although several theories have been proposed for postshock ST deviation, the exact mechanism has not been clearly established. The most convincing theory is the electroporation. This phenomenon would provoke a transient sarcolemmal microlesions, which short circuit the menbrane causing immediate despolarization and significant ionic exchange across the cell menbrane. Finally, the ST segment elevation post-DC cardioversion is transient short-lived phenomenon, being maximal just after the DC shock with mean duration of 60 seconds, and Usually resolving 5 minuts postshock. Ref.: P Shan et al., American Journal Of Emergency Medicine, 2014.



Tracing 2: Patient with Transient ST-Segment Elevation Immediately after Direct-Current (DC) countershock to the precordium.

Prinzmetal Angina



In 1950, Prinzmetal et al described a new syndrome of precordial pain of ischemic origin that usually occurs at rest without an evident classical trigger as exercise, stress, etc. The pain episodes often occur in consecutive or nearly consecutive days, mainly at night or in the early hours of the morning. Ventricular arrhythmias often occur at the maximum ST-elevation, Ventricular fibrillation and sudden death rarely occurs. The hypothesis to explain this syndrome was that transient coronary Vasoconstriction induces sudden, but transient and usually total oclusion of one or rarely more than one coronary artery. The pain can controlled with nitroglycerin and very rarely evolves to acute myocardial infarction. The coronary arteries usually appear normal or show insignificant obstruction. The ischemic episodes typically last for a short period of minutes and can be triggered by tobbaco, cocaine, hyperventilation, among others.

Prinzmetal Angina

Changes of Repolarization: ST-segment and T Wave Changes

1. Increased height of the R wave, witch appears in all cases.
2. Coincident S wave diminution or disappearance in all cases.
3. Up-sloping TQ segment in two third of cases.
4. Alternans of the elevated ST-segment and negative T wave deepness in 20% of cases (Fig).

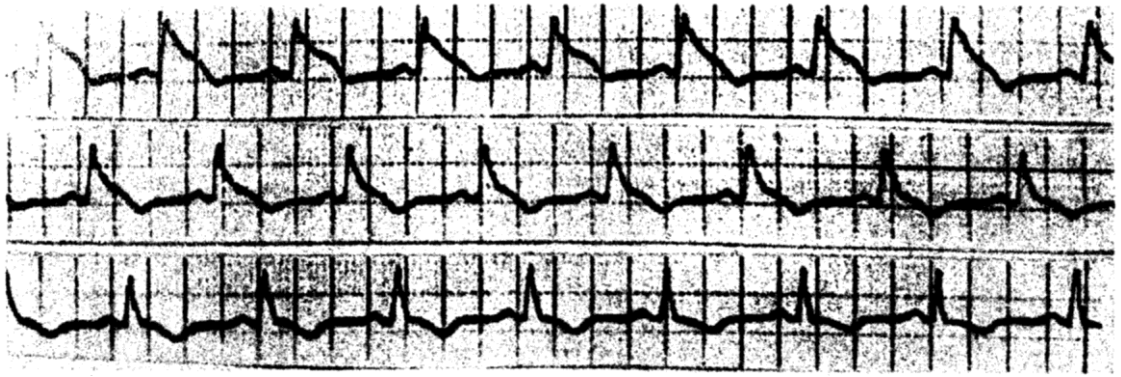


Fig.: Prinzmetal angina on Holter monitoring. See the T wave inversion at the end of the episode. Lamda wave

Ref.: Bayes de Luna, at al. ANE. September 2014.

An early repolarization (ER) pattern was previously considered a benign ECG pattern, until it was shown to be associated with idiopathic VF in three separate case-control studies in 2008 (**Haïssaguerre M, Derval N, Sacher F, et al.: Sudden cardiac arrest associated with early 281 repolarization. N Engl J Med 2008; 358:2016–2023. 282; Nam G-B, Kim Y-H, Antzelevitch C: Augmentation of J waves and electrical storms in patients 283 with early repolarization. N Engl J Med 2008; 358:2078–2079. 284 3; Rosso R, Kogan E, Belhassen B, et al.: J-point elevation in survivors of primary ventricular 285 fibrillation and matched control subjects: incidence and clinical significance. J Am Coll 286 Cardiol 2008; 52:1231–1238**.). Subsequently, researchers found that ER was also associated with all-cause mortality, cardiac death, and SCD in the general population (**Tikkanen JT, Anttonen O, Junttila MJ, et al.: Long-term outcome associated with early 288 repolarization on electrocardiography. N Engl J Med 2009; 361:2529–2537. 289; Rollin A, Maury P, Bongard V, et al.: Prevalence, prognosis, and identification of the malignant 290 form of early repolarization pattern in a population-based study. Am J Cardiol 2012; 291 110:1302–1308. 292 6; Sinner MF, Reinhard W, Müller M, et al.: Association of early repolarization pattern on ECG 293 with risk of cardiac and all-cause mortality: a population-based prospective cohort study 294 (MONICA/KORA**). **PLoS Med 2010; 7:e1000314; Cheng Y-J, Lin X-X, Ji C-C, et 295 al.: Role of Early Repolarization Pattern in Increasing Risk of Death. J Am Heart Assoc 2016; 296 5:e003375. 297 8; Haruta D, Matsuo K, Tsuneto A, et al.: Incidence and prognostic value of early repolarization 298 pattern in the 12-lead electrocardiogram. Circulation 2011; 123:2931–2937**). However, some studies found no link between ER and adverse events (**Cheng Y-J, Lin X-X, Ji C-C, et 295 al.: Role of Early Repolarization Pattern in Increasing Risk of Death. J Am Heart Assoc 2016; 296 5:e003375**). Consequently, researchers attempted to distinguish benign ER patterns from patterns that associate with more unfavorable prognoses (**Olson KA, Viera AJ, Soliman EZ, Crow RS, Rosamond WD: Long-term prognosis associated with J-point elevation in a large middle-aged biracial cohort: the ARIC study. Eur Heart J 301 2011; 32:3098–3106; Rollin A, Maury P, Bongard V, et al.: Prevalence, prognosis, and**

identification of the malignant form of early repolarization pattern in a population-based study. Am J Cardiol 2012; 291 110:1302–1308; Roten L, Derval N, Maury P, et al.: Benign vs malignant inferolateral early repolarization: Focus on the T wave. Heart Rhythm 2016; 13:894–902). Furthermore, other studies examined whether the prognosis associated with ER varies across different patient subgroups (**Sinner MF, Reinhard W, Müller M, et al.: Association of early repolarization pattern on ECG with risk of cardiac and all-cause mortality: a population-based prospective cohort study (MONICA/KORA). PLoS Med 2010; 7:e1000314; Olson KA, Viera AJ, Soliman EZ, Crow RS, Rosamond WD: Long-term prognosis associated with J-point elevation in a large middle-aged biracial cohort: the ARIC study. Eur Heart J 301 2011; 32:3098–3106; Adler A, Rosso R, Viskin D, Halkin A, Viskin S: What do we know about the "malignant form" of early repolarization? J Am Coll Cardiol 2013; 62:863–868**). ER was associated with cardiac mortality, particularly among younger middle-aged subjects, whereas in studies among older subjects ER was not associated with an excess risk (**Sinner MF, Reinhard W, Müller M, et al.: Association of early repolarization pattern on ECG with risk of cardiac and all-cause mortality: a population-based prospective cohort study (MONICA/KORA). PLoS Med 2010; 7:e1000314. 7; Hisamatsu T, Ohkubo T, Miura K, et al.: Association between J-point elevation and death from 309 coronary artery disease--15-year follow up of the NIPPON DATA90. Circ J 2013; 77:1260– 310 1266**). In young adult populations, ER is a prevalent finding and considered a benign phenomenon (**Ilkhanoff L, Soliman EZ, Prineas RJ, et al.: Clinical characteristics and outcomes associated with the natural history of early repolarization in a young, biracial cohort followed to middle age: the Coronary Artery Risk Development in Young Adults (CARDIA) study. Circ 314 Arrhythm Electrophysiol 2014; 7:392–399. 315; Sager SJ, Hoosien M, Junttila MJ, et al.: Comparison of inferolateral early repolarization and its electrocardiographic phenotypes in pre- and postadolescent populations. Am J Cardiol 2013; 317 112:444–448. 318; Lanza GA, Mollo R, Cosenza A, et al.: Prevalence and clinical correlates of early repolarization and J wave in a large cohort of subjects without overt heart disease. J Electrocardiol 2012; 320**

45:404–410). Whether age affects the risk of SCD associated with ER in adult subjects remains unclear.

Arttu Holkeri et al present in 2019 their investigation of the association between ER and SCD, cardiac mortality, and all-cause mortality in a Finnish general population cohort conducted in 1978–1980. and examine whether this association differs between subjects younger than 50 years old and those ≥50 years. Furthermore, the authors assess whether sex impacted the risk associated with ER in these age groups.(**Holkeri, A., Eranti, A., Haukilahti, M. A. E., Kerola, T., Kentta¨, T. V., Tikkanen, J. T., … Huikuri, H. V. (2019). *Impact of age and sex on the long-term prognosis associated with early repolarization in the general population. Heart Rhythm.* doi:10.1016/j.hrthm.2019.10.026**) These authors concluded that among adults aged 30–50 years, ER associates with SCD. In particular, women under 50 years old with ER exhibited a higher risk of SCD, while ER was not associated with SCD among men.

Definition of early repolarization pattern following 2015 modified consensus paper

(Macfarlane PW, Antzelevitch C, Haissaguerre M, et al.: The Early Repolarization Pattern: A 324 Consensus Paper. J Am Coll Cardiol 2015; 66:470–477.) (Tikkanen JT, Junttila MJ, Anttonen O, et al.: Early repolarization: electrocardiographic phenotypes associated with favorable long-term outcome. Circulation 2011; 123:2666–2673) (Tikkanen JT, Anttonen O, Junttila MJ, et al.: Long-term outcome associated with early 288 repolarization on electrocardiography. N Engl J Med 2009; 361:2529–2537.)

An ER pattern si defined as an end QRS notch or slur on the downward slope of the prominent R-wave at the J-point, with an amplitude of ≥0.1 mV or 1mm measured with respect to the true baseline determined as the TP segment of the precedent beat and QRS duration <120ms.

Figure next slide

A subject's ECG is considered positive for ER if an ER pattern is present in either ≥2 of the inferior (II, III, or aVF) or ≥2 of the lateral (I, aVL, V4, V5, or V6) leads. An ER amplitude was classified as ≥0.1 mV, but 110 ms, a pacemaker 108 rhythm, or rare ECG findings not representing the general population.

In summary ER pattern can be diagnosed in the presence of J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG.

Definition of Early Repolarization Syndrome

ER syndrome is diagnosed in the presence of J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG **in a patient resuscitated from otherwise unexplained VF/polymorphic VT**. ER syndrome can be diagnosed in an SCD victim with a negative autopsy and medical chart review with a previous ECG demonstrating J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG . The magnitude of the J-point elevation may have prognostic significance. Either slurred or notched J-point elevation ≥0.2 mV is relatively rare in the general population but appears to be associated with an increased risk (**Tikkanen JT, Anttonen O, Junttila MJ, et al. Long-term outcome associated with early repolarization on electrocardiography. N Engl J Med 2009;361: 2529–2537**). Furthermore, J-point elevation in idiopathic VF patients is of greater amplitude and ECG lead distribution compared to those with an established cause of cardiac arrest (**Derval N, Simpson CS, Birnie DH, et al. Prevalence and characteristics of early repolarization in the CASPER registry: cardiac arrest survivors with preserved ejection fraction registry. J Am Coll Cardiol 2011;58:722–728**). The available data also suggest that transient changes in the presence and amplitude of J-point elevation portends a higher risk for VF (**Haissaguerre M, Derval N, Sacher F, et al. Sudden cardiac arrest associated with early repolarization. N Engl J Med 2008;358:2016–2023**). A horizontal or descending ST segment following J point elevation is associated with a worse outcome in the general population.(**Tikkanen JT, Junttila MJ, Anttonen O, et al. Early repolarization: electrocardiographic phenotypes associated with favorable long-term outcome. Circulation 2011;123:2666–2673**)

This observation has been very helpful in distinguishing idiopathic VF patients from matched controls and is a key aid in clinical decision making (**Rosso R, Glikson E, Belhassen B, et al. Distinguishing "benign" from "malignant early repolarization": the value of the ST-segment morphology. Heart Rhythm 2012;9:225–229**).

According to the consensus document (**Macfarlane PW, Antzelevitch C, Haissaguerre M. The early repolarization pattern: a consensus paper. J Am Coll Cardiol. 2015;66:470–477**), it is generally recommended that ST-segment elevation should be defined as early repolarization only in the presence of end-QRS notch or slur on the downslope of a prominent R-wave, and QRS duration < 120 ms. If end-QRS notching or slurring is absent, it is recommended that this finding should not be described as early repolarization. In this sense, J point elevation without end-QRS notching or slurring should be defined as non specific ST-segment elevation.[3] end-QRS notching or slurring in the terminal QRS complexes are essential for identifying early repolarization. In conclusion, early repolarization pattern has an arrhythmogenic potential that may cause ventricular fibrillation. However, it should be clearly defined based on the consensus document to avoid misdiagnosis. In the absence of end-QRS notching or slurring, J point elevation should not be defined as early repolarization. Different interpretations of "J-point elevation" were considered and classified following definitions, the amplitude of the peak of an end QRS notch is denoted **pkQRSn** and the amplitude at the **• Type 1 pkQRSn ≥0.1 mV and STj ≥0.1 mV and ST-segment upward sloping.**

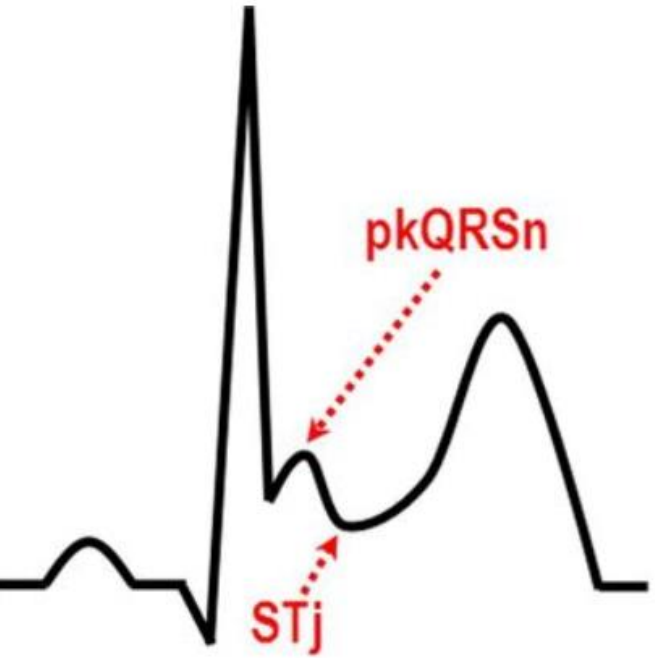
• Type 2 pkQRSn ≥0.1 mV and STj <0.1 mV.

• Type 3 onQRSs ≥0.1 mV and STj ≥0.1 mV and ST-segment upward sloping.

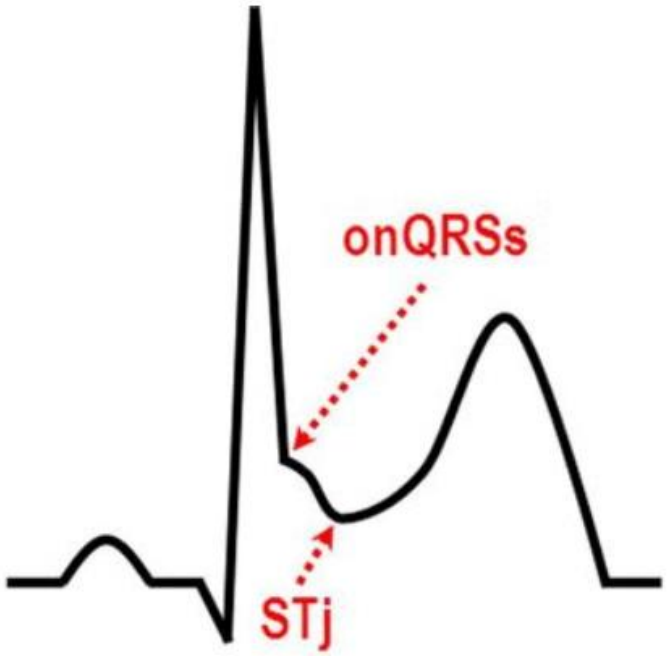
• Type 4 onQRSs ≥0.1 mV and STj <0.1 mV.

• Type 5 No QRS notching or slurring and STj ≥0.1 mV and ST-segment: Currently this pattern.

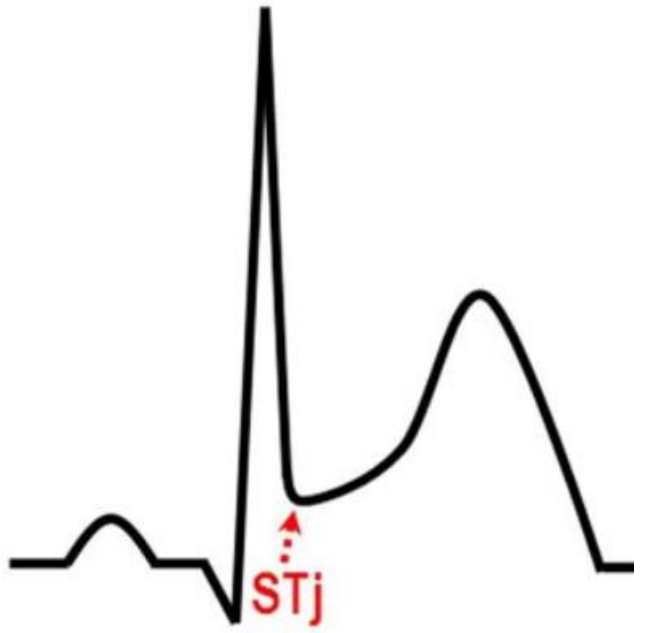
ER Notched **ER**



Types 1 & 2



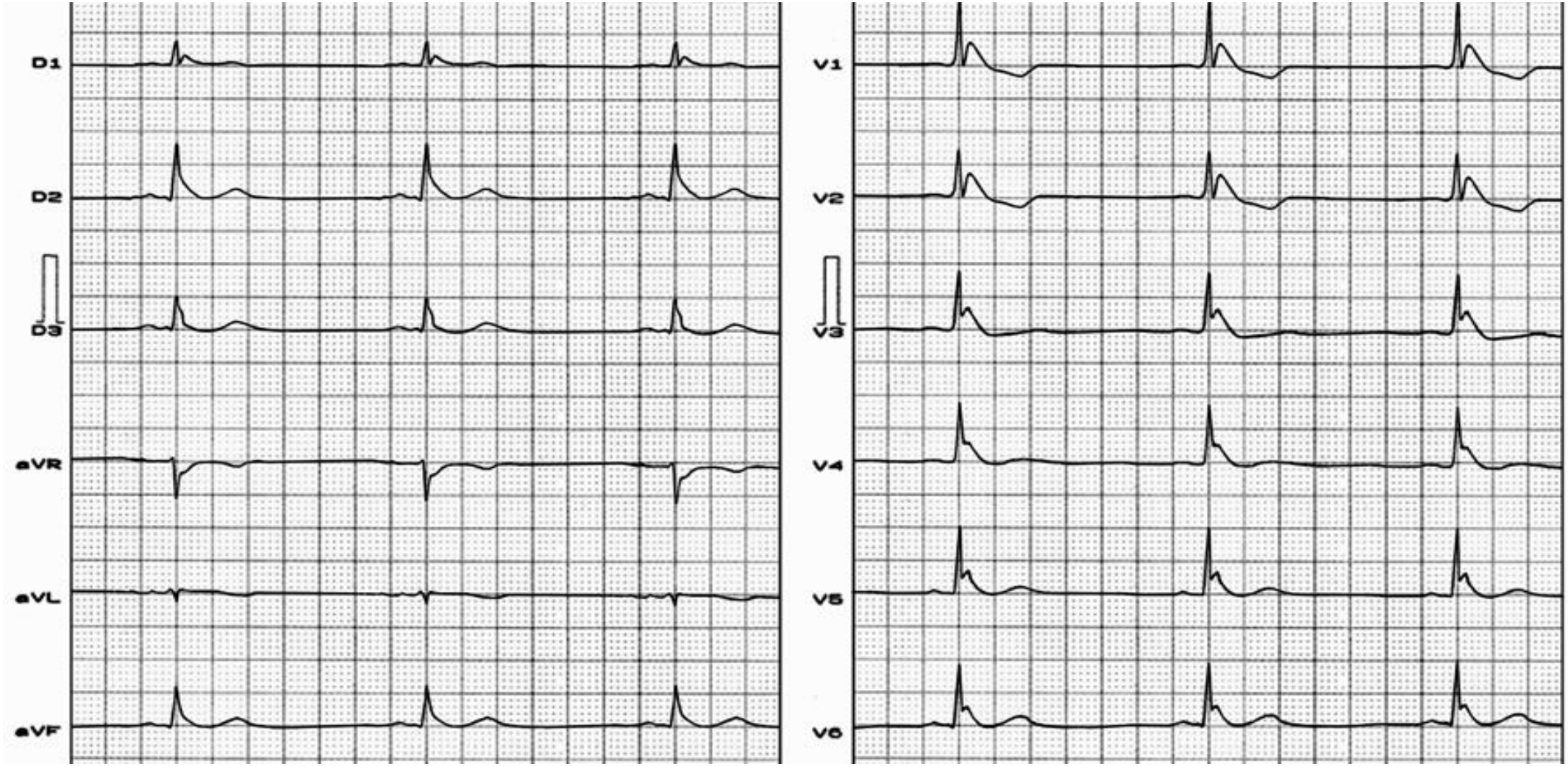
Types 3 & 4



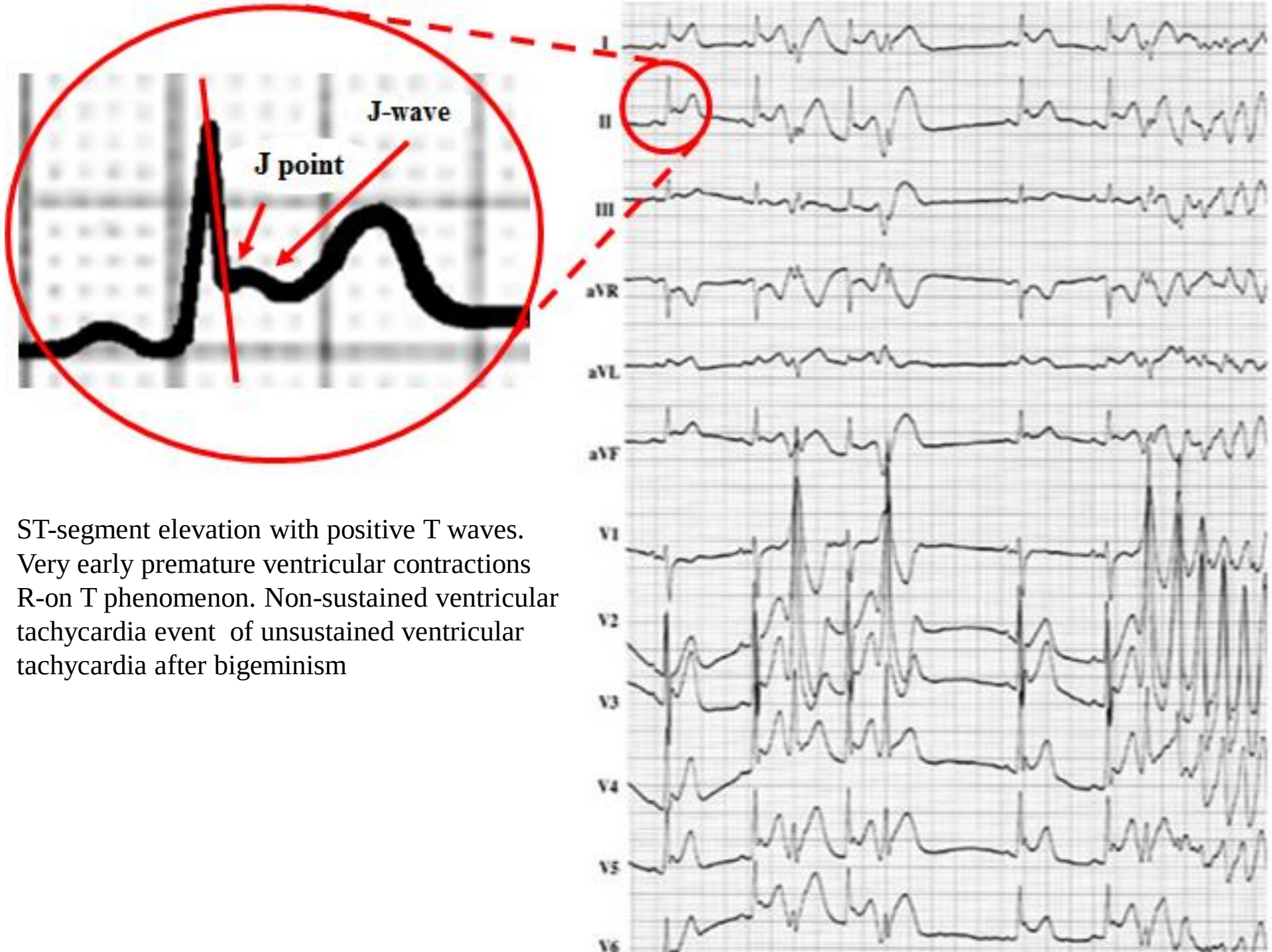
Type 5

onQRSs; Onset QRSs

Malignant Early Repolarization Pathological J or malignant waves of idiopathic ventricular fibrillation associated with early repolarization pattern (*ERP): the "Haïssaguerre pattern"*

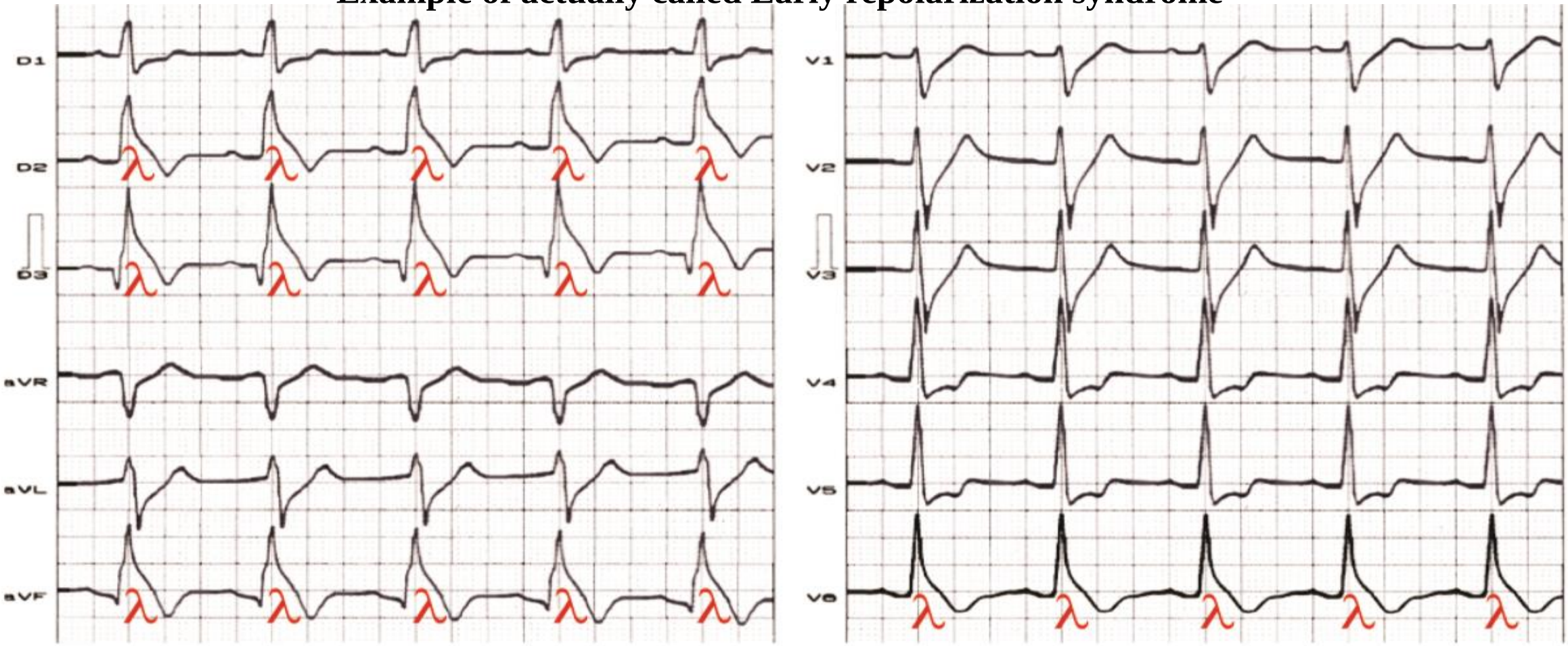


Subtype 3 shows an ER pattern registered globally in the inferior, lateral and right precordial leads. This variant is associated with the highest level of risk for the development of VF storms (**Nam 2008**). In subtype 3, the Brugada waves may be seen together with giant J waves in other ECG leads. Although the Brugada waves are not called ER, their underlying mechanism is identical to that of the ER patterns.



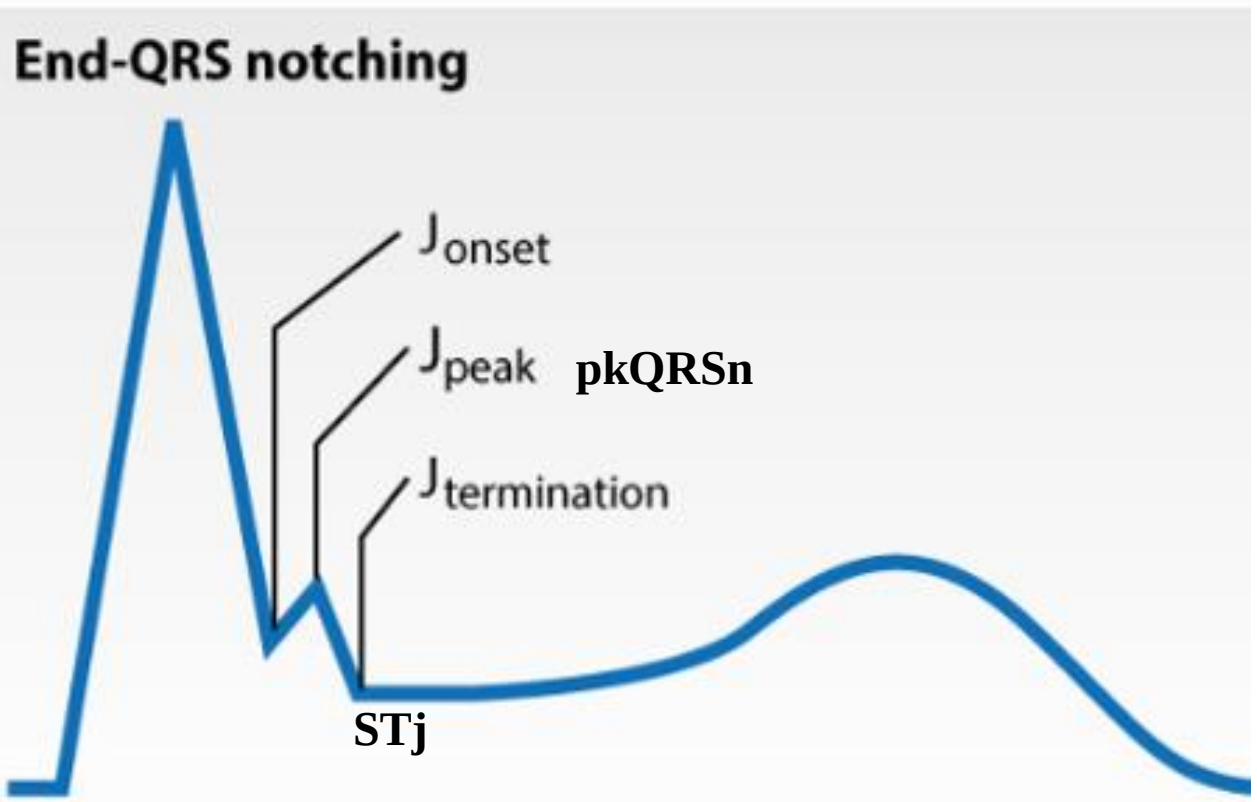
ST-segment elevation with positive T waves. Very early premature ventricular contractions R-on T phenomenon. Non-sustained ventricular tachycardia event of unsustained ventricular tachycardia after bigeminism

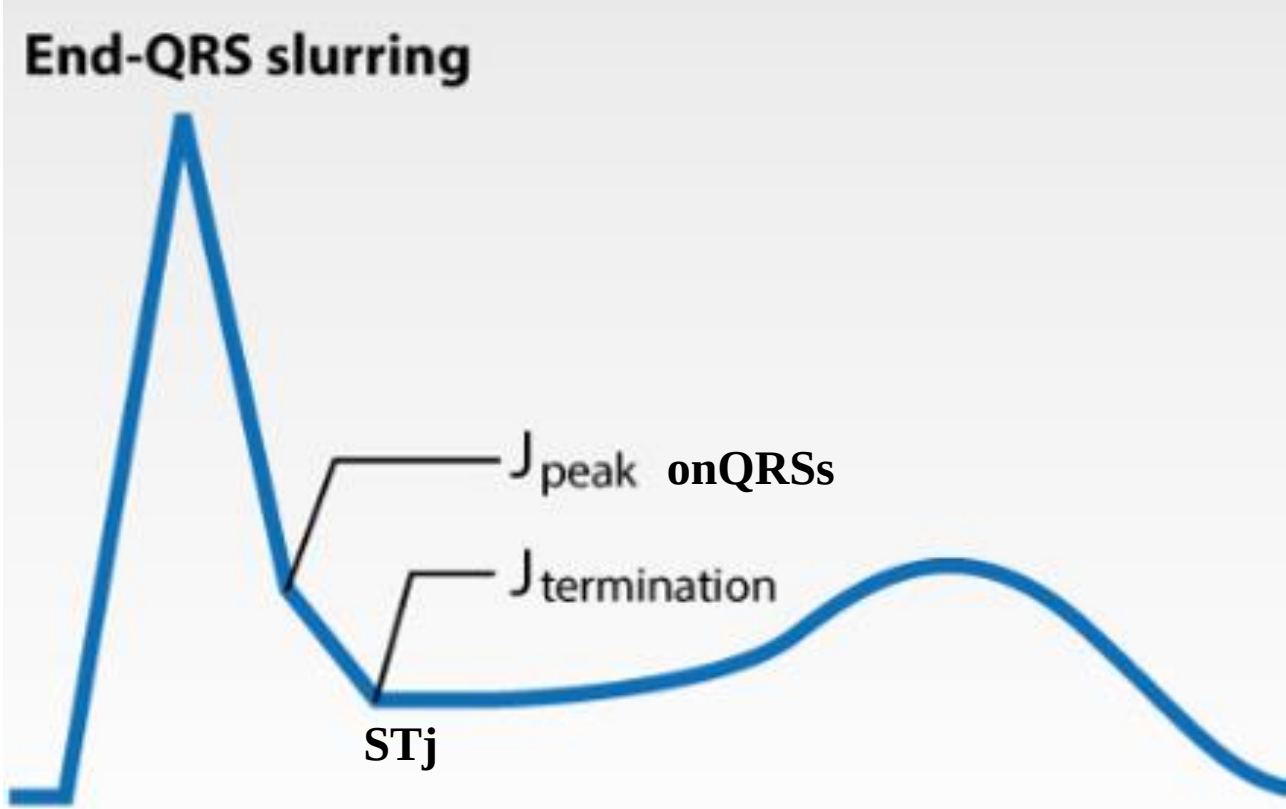
Example of actually called Early repolarization syndrome

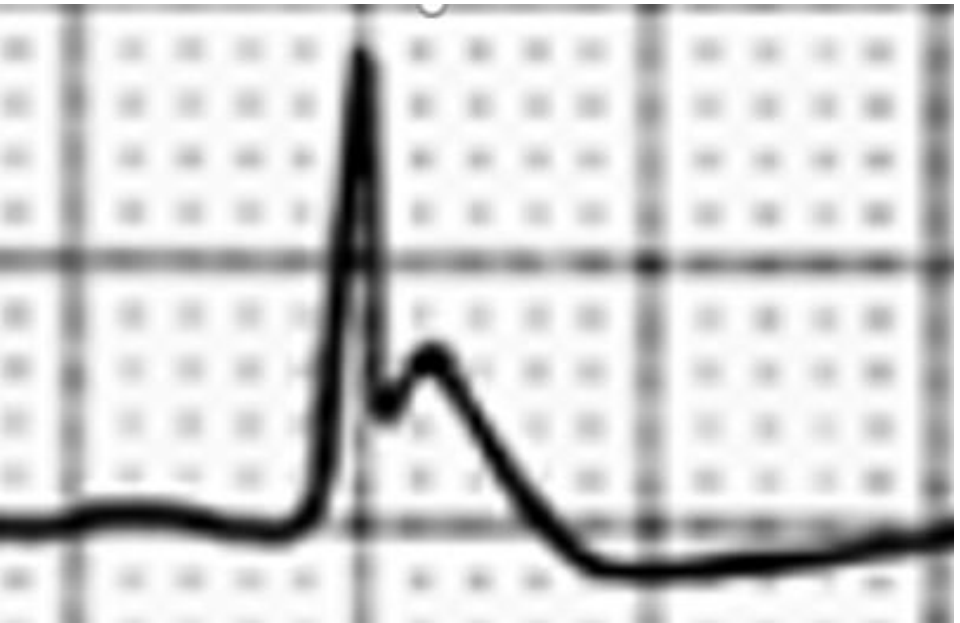


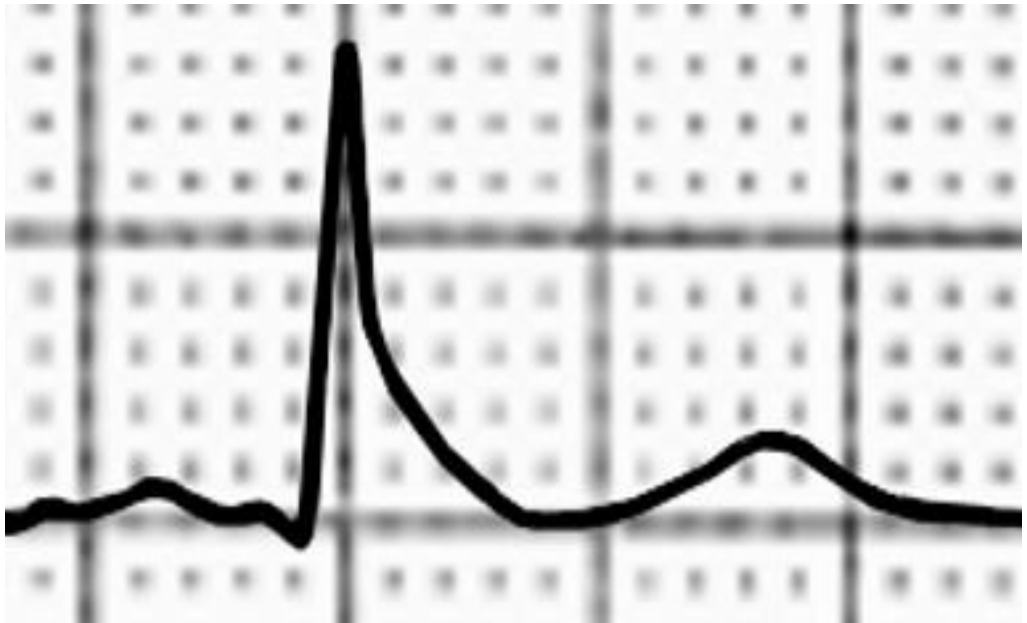
The ECG shows persistent ST segment elevation in the inferior and apical lateral leads, associated to concomitant reciprocal or mirror image in the anterior wall that was not modified with the use of sublingual nitrate in absence of hypothermia, electrolyte imbalance or ischemia. This Lambda wave correspond to J wave and/or QRS slurring The "J wave" (also referred to as "the Osborn wave," "the J deflection," or "the camel's hump") is a distinctive deflection occurring at the QRS-ST junction. In 1953, Dr. John Osborn described the "J wave" as an "injury current" resulting in VF during experimental hypothermia.

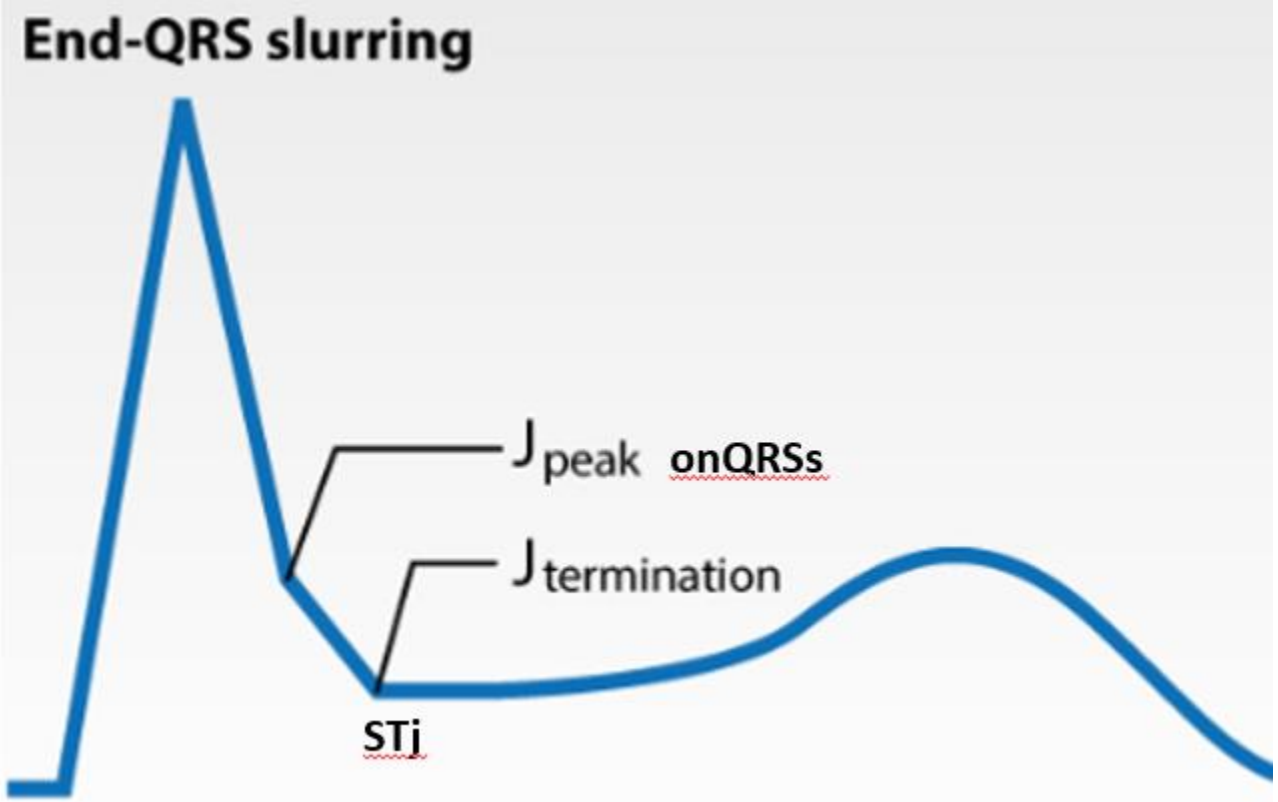
Riera AR, Ferreira C, Schapachnik E, et al. Brugada syndrome with atypical ECG: downsloping ST-segment elevation in inferior leads. J Electrocardiol. 2004 Apr; 37: 101-104.

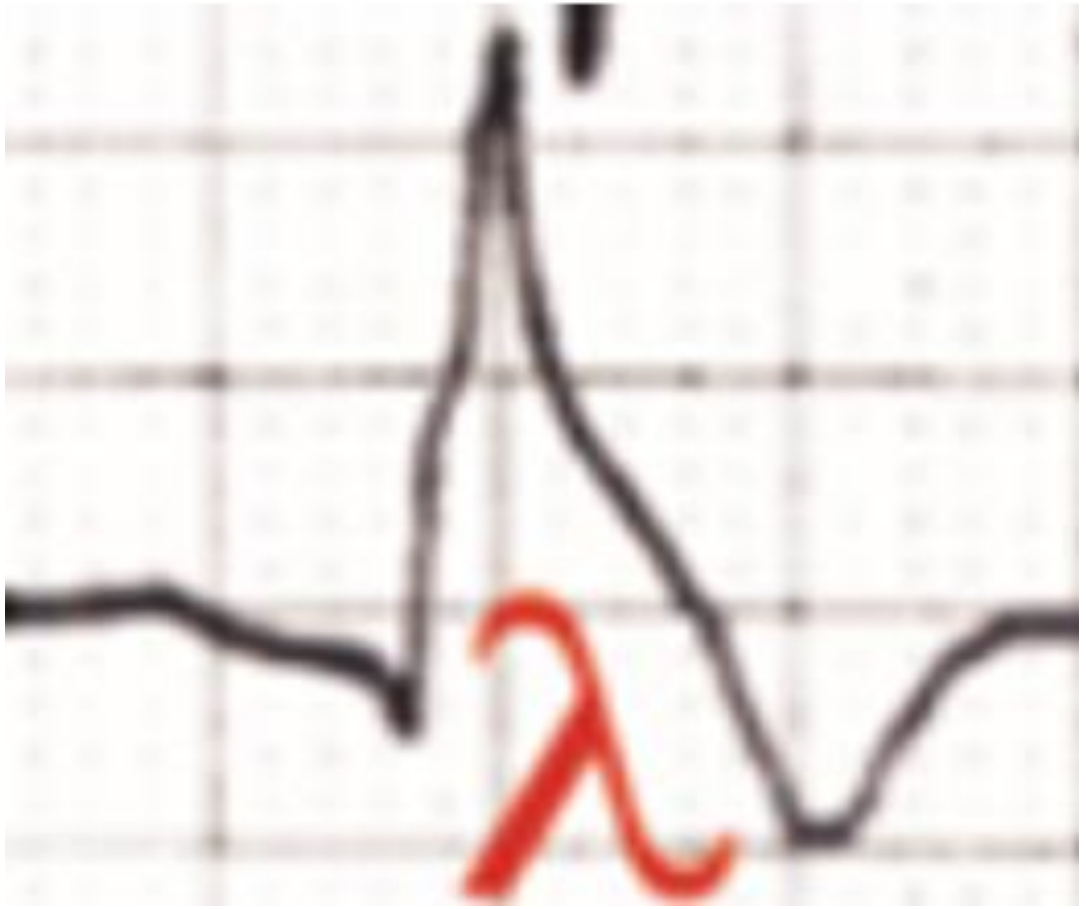

End-QRS notching
J onset
J peak
pkQRSn
J termination
STj


End-QRS slurring
J peak
onQRSs
J termination
STj









Currently we consider that End-QRS slurring is the same as Lambda wave

Inferolateral J-Wave Syndromes

Early repolarization pattern indicates a distinct ECG phenotype affecting the junction (J-wave) between the QRS complex and the ST segment in inferolateral leads. It was initially described as a benign ECG finding or in association with hypothermia. (**Tomaszewski W. Changements electrocardiographiques observes chez un homme mort de froid. Arch Mal Coeur 1938; 31:525–8.**) Subsequently, many conditions producing this phenotype have been described such as hypercalcemia, acute ischemia, brain injury, and others. (**Gussak I, Antzelevitch C. Early repolarization syndrome:clinical characteristics and possible cellularand ionic mechanisms. J Electrocardiol 2000;33:299–309.**) The link with an increased risk of arrhythmic death was demonstrated in sporadic cases and in case-control studies of unexplained SCD then finally in association with various types of structural heart disease. (**Otto CM, Tauxe RV, Cobb LA, et al. Ventricular fibrillation causes sudden death in southeast Asian immigrants.Ann Intern Med 1984;101:45–7.**) (**Aizawa Y, Tamura M, Chinushi M, et al. Idiopathic ventricular fibrillation and bradycardia-dependent intraventricular block. Am Heart J 1993; 126:1473–4.**) (**Haissaguerre M, Derval N, Sacher F, et al. Sudden cardiac arrest associated with early repolarization. N Engl J Med 2008; 358:2016–23**.) (**Tikkanen JT, Anttonen O, Junttila MJ, et al. Long term outcome associated with early repolarization on electrocardiography. N Engl J Med 2009;361: 2529–37**.) (**Rosso R, Kogan E, Belhassen B, et al. J-point elevation in survivors of primary ventricular fibrillation and matched control subjects: incidence and clinical significance. J Am Coll Cardiol 2008; 52:1231–8.**) (**Antzelevitch C, Yan GX. J-wave syndromes: Brugada and early repolarization syndromes. Heart Rhythm 2015; 12:1852–66.**) (**Macfarlane PW, Antzelevitch C, Haissaguerre M, et al. The early repolarization pattern: a consensus paper. J Am Coll Cardiol 2015;66:470–7.**) The most recent mapping data in humans, provide evidence for heterogeneity of substrates, which underlie inferolateral J-wave syndromes. For a comprehensive review of risk stratification and clinical management of J-wave syndromes the readers are guided the following manuscripts:

1. **Viskin S, Rosso R, Halkin A. Making sense of early repolarization. Heart Rhythm 2012; 9:566–9.**
2. **Obeyesekere N, Krahn AD. Early repolarization – what should the clinician do? Arrhythm Electrophysiol Rev 2015;4:96–9.35.**
3. **Nademanee K, Haissaguerre M, Hocini M, et al. Ventricular Fibrillation substrates and Electrophysiological Abnormalities In Early Repolarization Syndrome: A Tale Of Two Phenotypes. HRS San Francisco. Heart Rhythm Scientific Sessions S-PO01-131. 2019.).**

Diagnosis of early repolarization and inferolateral J-wave syndromes

Expert consensus recommendations (**Viskin S, Rosso R, Halkin A. Making sense of early repolarization. Heart Rhythm 2012;9:566–9.**) defined the syndrome (the term indicating symptomatic patients) as follows:

(1) the presence of J-point elevation ≥1 mm in greater than or equal to 2 contiguous inferior and/or lateral leads of a standard 12-lead ECG in a patient resuscitated from otherwise unexplained VF; an SCD victim with a negative autopsy and medical chart review with a previous ECG demonstrating J-point elevation ≥1 mm in greater than or equal to 2 contiguous inferior and/or lateral leads of a standard 12-lead ECG.

(2) The ECG pattern (asymptomatic subjects) is defined by a distinct J-wave or J-point elevation greater than or equal to 1 mm that is a notch or a slur, with or without ST-segment elevation.

This definition may be sometimes ambiguous owing to the small amplitude and spontaneous changes of the J-wave. Strong inspiration or Valsalva maneuvers can amplify J-wave possibly by changing the heart shape and position relative to the chest. The dynamic changes in J-waves during cycle-length prolongation, either unchanged pattern or amplification, are an important sign to evaluate, as it relates to the individual substrate underlying the J-wave.

High-density mapping of the J-wave

In contrast to experimental wedge models focusing on repolarization abnormalities, (**Antzelevitch C, Yan GX. J-wave syndromes: Brugada and early repolarization syndromes. Heart Rhythm 2015; 12: 1852–66.**) recent high-density electrogram mapping in humans both in vivo and ex vivo experimental conditions provide evidence that a spectrum of heterogeneous substrates, related either to delayed depolarization or to early repolarization abnormalities, underlie inferolateral J-wave syndromes.

Clinical mapping data have been collected from patients with inferolateral J-wave syndrome in 3 centers. (**Haissaguerre M, Nademanee W, Hocini M, et al. Depolarizationversus repolarization abnormality underlyinginferolateral J wave syndromes–new conceptsin sudden cardiac death with apparently normal hearts. Heart Rhythm 2018;16(5):781–90.**). These patients had no apparent SHD on MRI, and most were referred for VF recurring despite antiarrhythmic drugs including quinidine. A J-wave syndrome combined with a BrS was observed in a subset, either spontaneously or with sodium channel blocker. High density endocardial and epicardial (2000–6000 recorded points) electrograms were performed during sinus rhythm in bipolar and unipolar mode.

A specific attention was paid to the electrograms coincident with the timing of J-wave. The electrograms occurring within the J-wave were considered as belonging to depolarization if they were in temporal continuity of the surrounding depolarization field and sharp. They were considered as indicating ventricular repolarization if there was no continuity with surrounding depolarization electrograms and a slow pattern (hump) in unipolar mode. (**Boineau J. The early repolarization variant-an electrocardiographic enigma with both QRS and J-STT abnomalies. J Electrocardiol 2007;40:3.e1-10.**) (**Coronel R, Casini S, Koopmann TT, et al. Right ventricular fibrosis and conduction delay in a patient with clinical signs of Brugada syndrome: a combined electrophysiological, genetic, histopathologic, and computational study. Circulation 2005;112: 2769–77**.)

J-wave due to delayed depolarization abnormality

In patients with an isolated inferolateral J-wave syndrome, the J-wave was the expression of an area of delayed depolarization in the inferior part of ventricles in 15% to 30% of cases (depending on the inclusion center), whereas an early repolarization was the cause of J-wave in the others (70%–85%). The true prevalence of depolarization versus repolarization abnormality varied between the inclusion centers and was also biased as the patients could be referred after quinidine failure, which may have selected more patients with delayed depolarization. Delayed-depolarization J-waves were recorded at the sites of terminal activation in the RV or LV. Most of the abnormal electrograms are found in epicardium, whereas 3 patients had abnormal electrograms recorded endocardially and epicardially. The terminology of early repolarization here is erroneous and "inferolateral J-wave" is more appropriate. An example of a late depolarization J-wave is shown in Fig.in TWO next slide. In contrast, in the patients with combined J-wave and BrS, J-wave was consistently caused by a delayed depolarization. The abnormal electrograms were recorded in inferior RV and similar to those in the RVOT, suggesting structural alteration from the same pathogenesis. The absence of ST elevation in inferior leads, in contrast to V1-V2 leads, may be due to specific properties of the RVOT tissue, more prone to develop marked repolarization changes secondary to altered depolarization. **(Martini M, Nava A, Thiene G, et al. Ventricular fibrillation without apparent structural heart disease. Am Heart J 1989; 118:1203–9.) (Hoogendijk MG, Potse M, Linnenbank AC, et al. Mechanism of right precordial ST-segment elevation in structural heart disease: excitation failure by current-to-load mismatch. Heart Rhythm 2010;7: 238–48.) (Benoist D, Charron S, Dubes VN, et al. Arrhythmogenic Molecular Substrate In The Healthy Right Ventricular Outflow Tract Heart Rhythm Scientific Sessions C-PO01-04. 2017.)**

(Bernus O, Walton RD, Hof T, et al. A Wide Spectrum Of Substrates Underlie J-wave Syndromes In Humans. HRS Chicago. Heart Rhythm Scientific SessionsS-MP13-05. 2019.) Ajmaline testing was performed in all patients of the cohort and resulted in J-wave amplification or ST elevation in the inferior leads in only 5 patients. All 5 patients had a delayed depolarization, whereas no patient with early repolarization had J/ST wave amplification on Ajmaline. The results mentioned earlier are in keeping with the high prevalence of J-waves described in various cardiomyopathies including noncompaction or arrhythmogenic right ventricular cardiomyopathies. In this context, it is likely that most of the J-waves express delayed depolarization. The ectopy morphology is essential to document on the 12-lead ECG as it guides mapping techniques by allowing focus on the area of interest. When originating from the left Purkinje fibers, the PVCs are narrow (<120 ms) with a right bundle right bundle-branch block morphology. They demonstrate right or left axis deviation when originating from the anterior or the posterior Purkinje fibers, respectively. Morphology changes are frequently observed in the left Purkinje PVCs, indicating different exit sites or origins and thus a need for larger ablation. When originating from the right Purkinje arborization, the PVCs have a left bundle-branch block morphology and left axis. (**Wilde AAM, Garan H, Boyden PA. Role of the Purkinje system in heritable arrhythmias. Heart Rhythm 2019; 16:1121–6.**) They display a rapid initial deflection but have a wider QRS duration than left Purkinje PVCs, as the right Purkinje has a more limited spatial arborization. More discrete PVC morphology changes are observed, as compared with left Purkinje PVC.

In sinus rhythm, distal Purkinje potentials precede the QRS complex by less than or equal to 15 ms. Longer intervals indicate a fascicular origin (with the risk of creating significant QRS changes if targeted for ablation). Special care should be taken during catheter manipulation to avoid inadvertent bumping of the right bundle, which will conceal Purkinje potentials within the local electrograms, the left-bundle-branch being much less vulnerable. When they are rare, PVCs can be inducible by creating postpacing (atrial or ventricular) pauses and/or drug infusion such as isoproterenol (1–2 mcg/kg/min) or class I drugs.

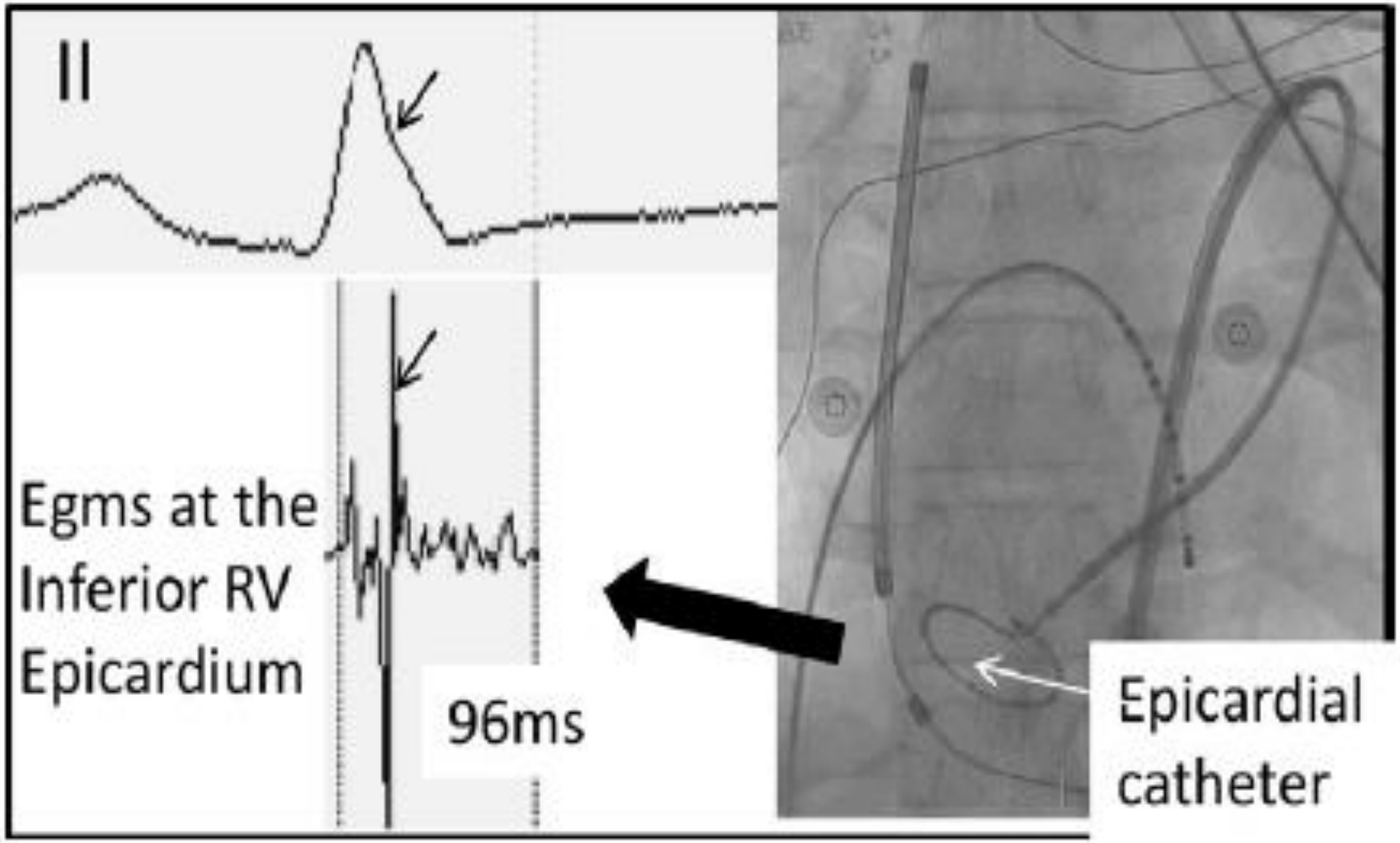


Fig. . A case of inferolateral J-wave syndrome due to abnormal depolarization. Epicardial depolarization in the inferobasal right ventricle occurs at the J-wave onset (small arrow) and is prolonged by low-voltage fragmented electrograms coincident with the J-wave body. Fluoroscopic anterior view shows the recording of circular 20-pole catheter in the epicardial inferobasal right ventricle and an endocardial decapolar catheter along the septum. ER, early repolarization.

Repetitive reentry in the Purkinje network

Reentry in the proximal Purkinje system has been described in the form of monomorphic ventricular tachycardia, either as bundle branch reentry or as left fascicular reentry. Polymorphic VT has also been reported, with all QRS complexes being associated with Purkinje activity (see Fig. 1). The authors have observed 5 patients, survivors of IVF, who had neither short-coupling Purkinje ectopy nor microstructural myocardial alteration. A distinct electrophysiologic response was however observed during programmed stimulation at the distal left fascicular system. No VF or repetitive activity was induced by stimulation from the right ventricle (in 4 of 5 patients), while pacing (S2-S3 extrastimuli) from the left posterior fascicle, and could consistently induce repetitive polymorphic VT within the distal Purkinje system. The polymorphic QRS complexes were associated with distal Purkinje potentials on a beat-to-beat basis, whereas the proximal Purkinje fascicle and bundle branch potentials were slower or absent (excluding a bundle branch reentry). The variations in ventricular cycles were preceded by a similar change in Purkinje cycles; and arrhythmia termination was preceded by the disappearance of Purkinje activity (Fig. 2). Two of these cases have been published recently (**Haissaguerre M, Vigmond E, Stuyvers B, et al. Ventricular arrhythmias and the His-Purkinje system. Nat Rev Cardiol 2016;13:155–66.**). These observations suggest that pacing performed near the distal left posterior Purkinje captures and invades retrogradely the Purkinje network. Although triggered activity cannot be ruled out, the repetitive responses are likely reentries induced in peripheral Purkinje system, with a gradual shift in trajectory and ventricular exit as demonstrated by computer modeling studies. These responses have been considered as clinically abnormal by the authors, as they are uncommon in their experience and have not been reported in the unique study evaluating left ventricular pacing in control patients. (**Haissaguerre M, Vigmond E, Stuyvers B, et al. Ventricular arrhythmias and the His-Purkinje system. Nat Rev Cardiol 2016;13:155–66.**) Further investigations are needed to confirm their pathologic significance and whether this electrophysiologic protocol may offer methods to reveal Purkinje abnormalities that can underlie VF susceptibility.

MICROSTRUCTURAL MYOCARDIAL SUBSTRATE IN IDIOPATHIC VENTRICULAR FIBRILLATION

In high-resolution experimental setups, VF requires the continuous formation of reentry for its maintenance, of which a critical determinant is the presence of normal (fiber arrangement) or abnormal (fibrosis) structural heterogeneities. In cardiomyopathic posttransplant human hearts, reentry has been shown to self-perpetuate formant cycles, as it can anchor to a myocardial scar. (**Gray RG, Pertsov AM, Jalife J. Spatial and temporal organization during cardiac fibrillation. Nature 1998; 392:75–8.**) (**Hsia H, Callans DJ, Marchlinski FE. Characterization of endocardial electrophysiological substrate in patients with nonischemic cardiomyopathy and monomorphic ventricular tachycardia. Circulation 2003; 108:704–10.**) The authors have used multielectrode body surface recordings (ECGi) in clinics to identify the reentries during ongoing VF. Because patients with IVF are free of SHD, VF reentries were expected to be distributed homogeneously across both ventricles. However, a clustering of reentries was observed, and mapping of these regions in sinus rhythm revealed, in some of them, abnormal electrogram characteristics, indicating the presence of “microstructural” cardiomyopathic alteration.

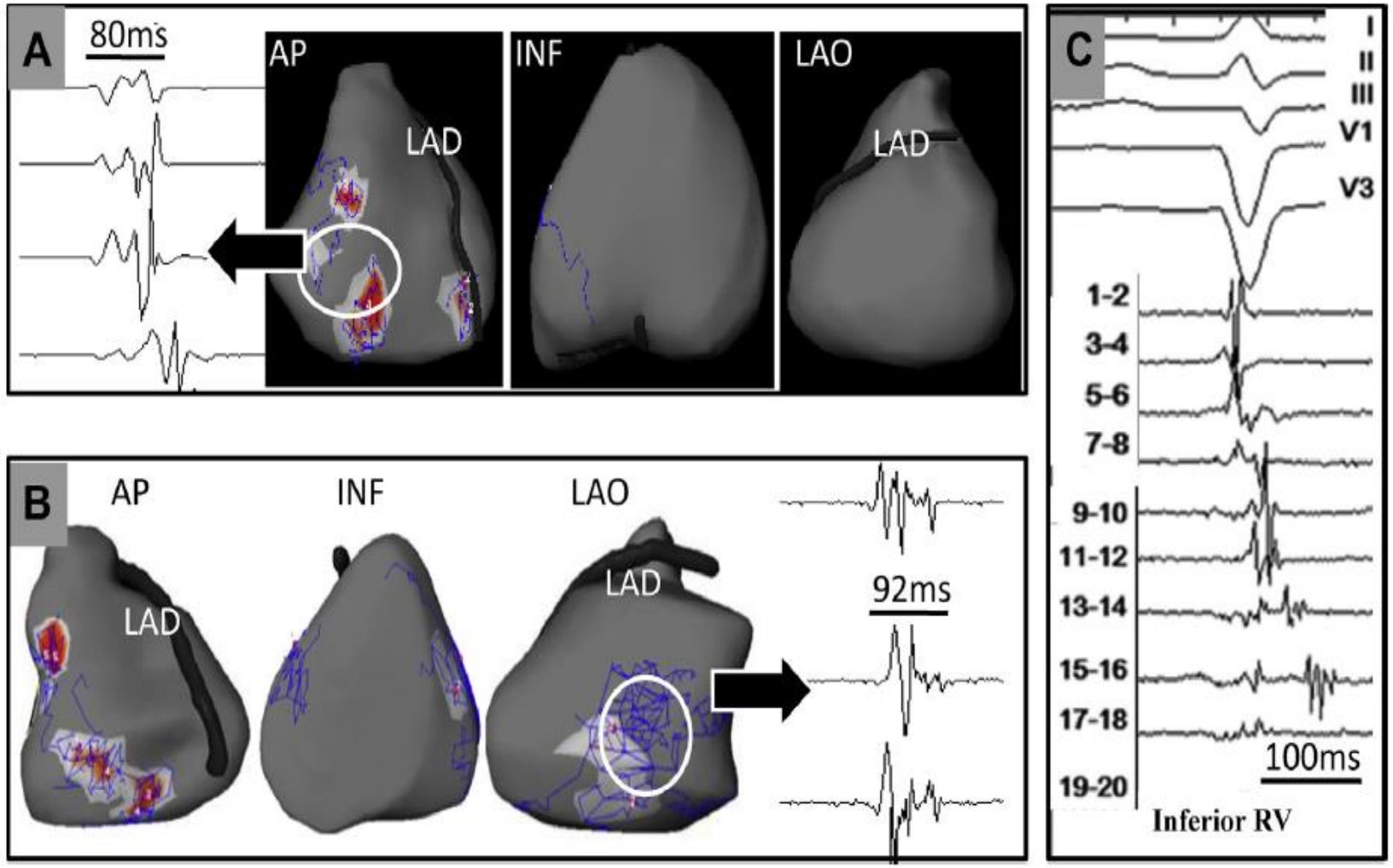
Invasive Electrogram Mapping in Sinus Rhythm

Text from Professor Michel Haïssaguerre

High-density recordings of endocardial and epicardial electrograms were performed using multispline catheter (Lasso or PentaRay catheter with 2 mm interelectrode distances) in the epicardium and PentaRay or decapolar catheters in the right and left endocardium. A transseptal or retroaortic approach was performed to access the endocardial left ventricle and a subxyphosternal approach to access into the pericardial space. The objective was to analyze electrograms present at the main driver areas (with the maximal number of rotations) compared with nondriver regions. Electroanatomical mapping was performed using magnetic geolocalization (CARTO system, Biosense Webster, CA); in a prior study, the authors mapped a mean of 590 _ 403 (endocardial right ventricle), 547 _ 292 (endocardial left ventricle), and 2081 _ 1278 (epicardium) sites per patient. Electrogram criteria were identical to those defining fibrotic and cardiomyopathic tissue during mapping of ischemic or dilated cardiomyopathies. Areas of low-amplitude electrograms were delineated in bipolar (less than 1 mV) and unipolar modes (less than 8.3 and 5.5 mV in LV and RV respectively) on 3-dimensional ventricular reconstruction (CARTO system, BiosenseWebster, CA). Because low-amplitude electrograms can be due to normal fat tissue on the epicardium, epicardial electrograms were only considered abnormal if they harbored fragmented signals with a duration > 70 ms with more than 3 components or split/late potentials. (**Soejima K, Stevenson WG, Sapp JL, et al. Endocardial and epicardial radiofrequency ablation of ventricular tachycardia associated with dilated cardiomyopathy: the importance of low-voltage scars. J AmColl Cardiol 2004; 43:1834–41.**) (**Cano O, Hutchinson M, Lin D. Electroanatomic substrate and ablation outcome for suspected epicardial left ventricular tachycardia in nonischemic cardiomyopathy. J Am Coll Cardiol 2009; 54:799–808**.) (**Janse MJ, Kléber AG. Electrophysiological changes and ventricular arrhythmias in the early phase of regional myocardial ischemia. Circ Res 1981;49: 1069–81**.) Greater mapping density was acquired in cases of abnormality to delineate the abnormal surface area.

Results

In the authors' current experience, myocardial areas manifesting low-amplitude and prolonged fractionated signals were found in 32 of 48 (67%) patients. The abnormal tissue was clustered in 1 or 2 areas, whereas 4 patients had scattered abnormal areas. In a previous study the abnormal surface area covered a mean of 13_5 cm^2, ranging from 6 to 22, representing 3.9±1.7% of the total ventricular surface. This finding is in keeping with prior experimental studies, which showed that even small ventricular lesions in the range of 4 cm^2 are sufficient to promote VF. (**Kubota I, Lux RL, Burgess MJ, et al. Activation sequence at the onset of arrhythmias induced by localized myocardial warming and programmed premature stimulation in dogs. J Electrocardiol 1988; 21:345–54.**) (**Tomaszewski W. Changements electrocardiographiques observes chez un homme mort de froid. Arch Mal Coeur 1938; 31:525–8.**) The RV was the structure preferentially harboring the abnormal area, whereas the LV or the septum was less frequently affected (Fig. 3 in next slide).



Spatial location of abnormal electrogram areas in 3 patients with IVF. Two patients had spontaneous (A) or induced (B) sustained VF, with VF drivers mapped using body surface mapping. The trajectories of unstable and stable reentries are shown by the blue curves and red areas, respectively in 3 views, whereby the anterior view shows the right ventricle and the left anterior oblique view shows the left ventricle. The abnormal signals recorded in sinus rhythm during epicardial mapping are present within the white dotted contours, which collocated with a dominant area of VF reentries, in the epicardial right (B) or left (C) ventricle. All other ventricular regions show narrow signals indicating healthy underlying tissue. Case C had no VF inducible by electrical stimulation. An area of prolonged and late electrograms was detected in the epicardial inferior right ventricle. AP, anteroposterior; LAD, left anterior descending; LAO, left anterior oblique; RV, right ventricle.

J-wave due to early repolarization

In this subset of patients there was no depolarization electrogram coincident with J-wave, but low-frequency signals (hump) were present particularly in unipolar recordings.

Electrocardiographically, **the increase of J-waves during cycle-length prolongation was likely the most specific sign associated with early repolarization syndrome**

figure

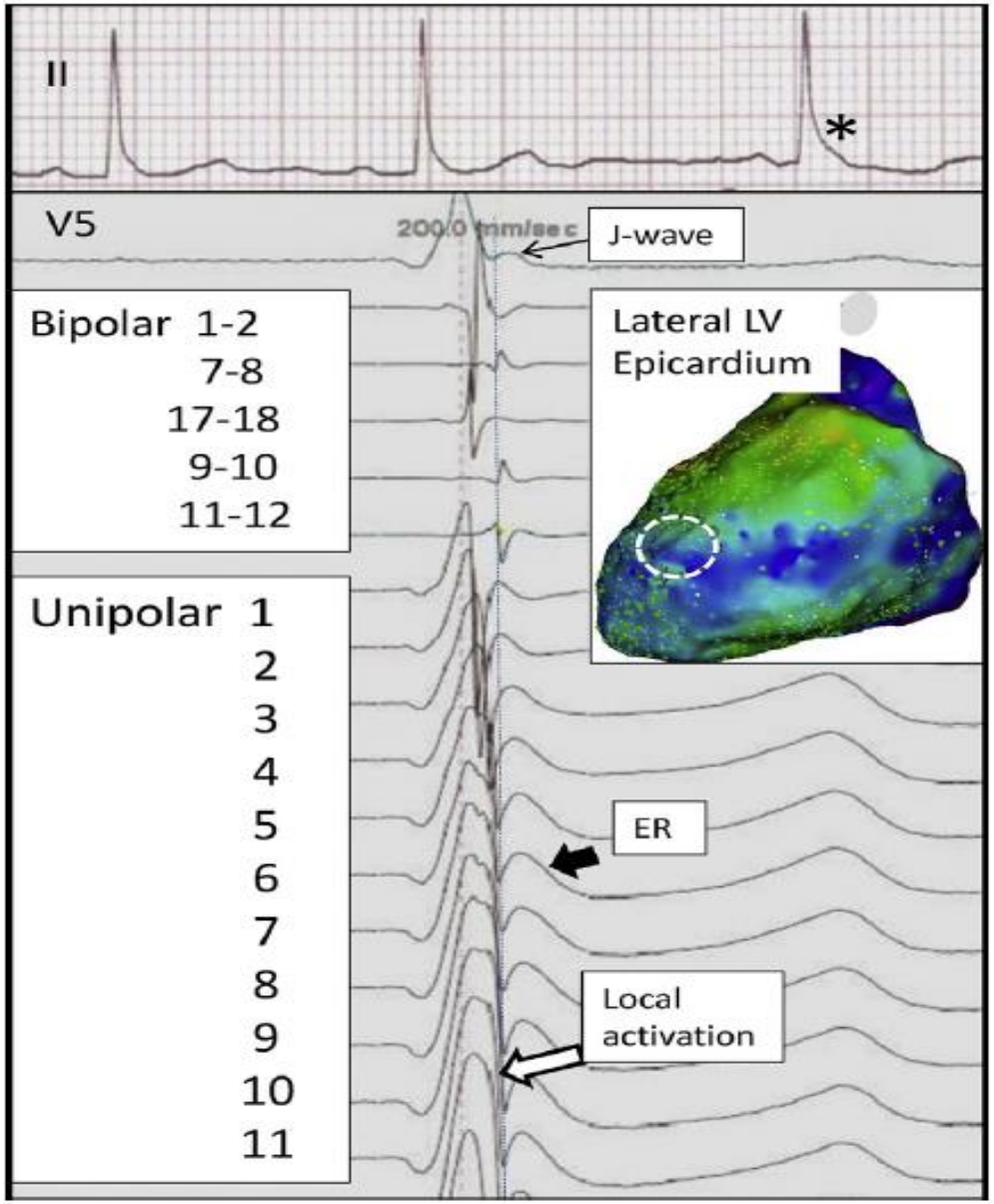


Fig. . A case of inferolateral J-wave syndrome due to ERS. The ECG shows J-wave amplification occurring in the third beat (asterisk) following a longer cycle length. Electrograms are recorded during epicardial mapping within the white dotted contour. Bipolar and unipolar recordings show the sharp local depolarization coincident with the very onset of J-wave (vertical line). The electrocardiographic J-wave is coincident with low-frequency ("ER") signals in unipolar recordings 1 to 11, which cover the entire half-inferior surface of left ventricle in this patient.

The early repolarization potentials were dominantly located epicardially in the inferior septum and adjacent left ventricle including the regions of papillary muscles. The J-wave mechanism is likely here a voltage gradient across the ventricular wall during early phase of repolarization, as shown by Antzelevitch and Yan. (**Antzelevitch C, Yan GX. J-wave syndromes: Brugada and early repolarization syndromes. Heart Rhythm 2015; 12:1852–66**.) However, the current clinical mapping techniques do not allow to measure repolarization parameters precisely and to demonstrate a gradient of repolarization or a phase 2 reentry. In patients with spontaneous VF initiation, the authors found an important triggering role of the Purkinje system further confirmed by favorable outcome after ablation. However, current techniques do not allow to measure repolarization parameters precisely and to demonstrate a gradient of repolarization or a phase 2 reentry. In patients with spontaneous VF initiation, the authors found an important triggering role of the Purkinje system further confirmed by favorable outcome after ablation (Fig. 6).

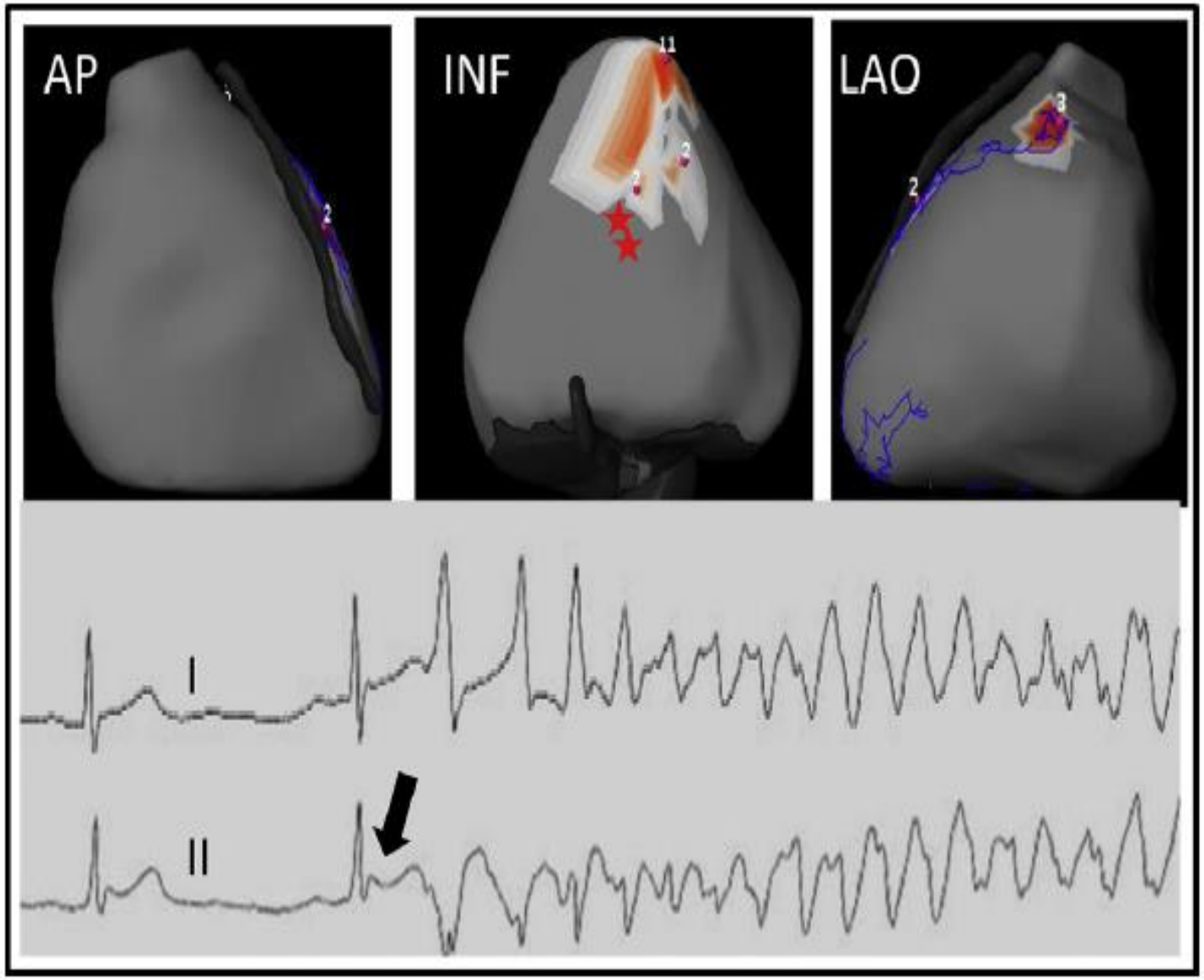


Fig. Location of VF reentries in a 14-year-old patient with early repolarization (arrow). Noninvasive mapping is performed during the initial 2 seconds following spontaneous initiation of VF (ECG shown). The initial 2 beats seem as focal breakthroughs (2 red stars), whereas the subsequent 15 beats seem as reentries (in red) clustered in the inferior septal area (inferior view). The latter (epicardial) location is compatible with an exit from the (endocardial) left posterior Purkinje fascicle. INF,inferior.

J-wave in human optical mapping

In human hearts with electrocardiographic J-wave patterns obtained through an organ donor program at the University Hospital. Epi- and endocardial optical mapping and microelectrode recordings were performed on either the left or the right ventricle. In one subject, the J-wave phenotype was due to delayed conduction in the LV basal region with 80% activation time prolongation compared n order to elucidate the substrates underlying J-wave syndrome, the authors' team had the unique opportunity to investigate with control (P<.05) and was exacerbated with increasing pacing frequency. For the other 2 subjects (who were siblings), J-waves increased during bradycardia and were associated with a heterogeneous increase of the action potential notch (phase 1 repolarization) and delayed action potential plateau on the right and/or left ventricular endocardium, but not epicardium, with no associated conduction abnormalities. These unique human heart data demonstrate that a variety of individual conduction and repolarization substrates, not limited to the epicardium, can underlie J-wave syndromes in humans. (**Haissaguerre M, Vigmond E, Stuyvers B, et al. Ventriculararrhythmias and the His-Purkinje system. Nat Rev Cardiol 2016;13:155–66**.)

SUMMARY

Arrhythmogenic diseases leading to SCD in apparently normal hearts may be underlined by fundamentally different substrates despite similar ECG phenotypes (such as J-wave) or the absence of phenotype (IVF). An important category is emerging in which the primary substrates maintaining VF are localized depolarization abnormalities due to **microstructural myocardial alterations**. Their pathogenesis is potentially of multiple causes, altering myocardial cells or their connections or leading to intervening fibrotic, inflammatory, or fatty tissue. A common feature of these substrates is that they constitute a target for efficient catheter ablation. In a second category of SCD with structurally normal hearts, the primary VF substrates are repolarization abnormalities (long/short or early repolarization) for which drugs are the most appropriate therapy.

In a third category the arrhythmia, mechanism seems dominated by "focal" activities from Purkinje or myocardial origin, such as

CPVT or a subset of IVF. This simplified classification (**Table. below**) does not exclude overlapping mechanisms. It puts together arrhythmogenic diseases, which are clinically distinct but share similar modes of pathogenesis and therapy, and potentially common genetic variants.

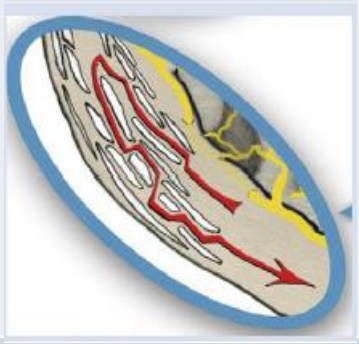
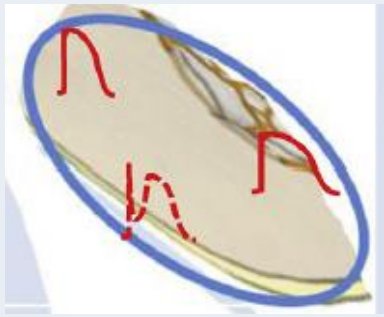
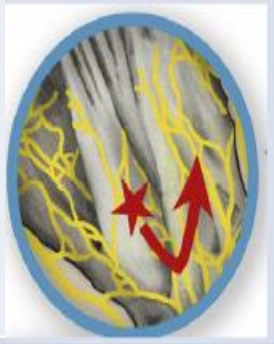
	Conduction Abnormality	Repolarization Abnormality	Excitation Abnormality
Primary VF substrate	Structural Abnormality, Heterogeneity of depolarization	Electrical Abnormality, Heterogeneity of repolarization	Rapid or multifocal ectopic activity
			
Diagnostic	Localized prolonged Despolarization		Consistent ectopy at arrythmia initiation and no myocardial abnormality
Type	1. Brugada Syndrome 2. Inferolateral J-wave 3. IFV with localized structural abnormality.	I. Long QT II. Short QT III. Early Repolarization	• IVF from Purkinje or myocardial foci • CPVT • Accidental commotio cordis, electrocution, drugs.
Therapy	Ablation	Drugs	Drugs or Ablation

Table - The spectrum of arrhythmogenic diseases leading to SCD in apparently normal hearts, in 3 categories, based on the primary pathogenesis.

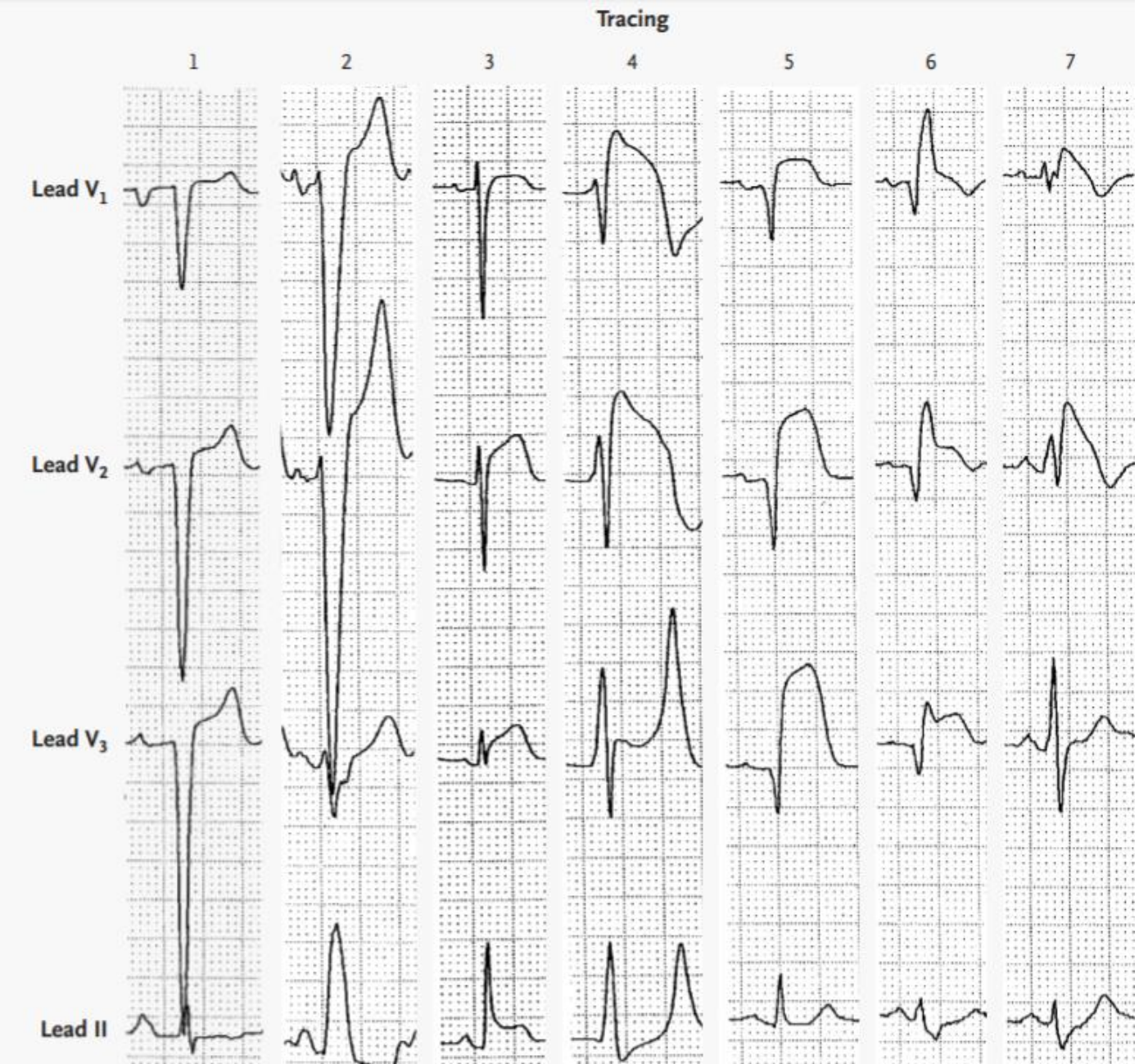
J wave syndromes

	Inherited				Acquired	
Characteristics	**ERS type 1**	**ERS type 2**	**ERS type 3**	**BrS**	**Ischemia mediated VT/VF**	**Hypothermia mediated VT/VF**
Average age of first event	35 years				30-40 years	
Anatomic location	Anterolateral left ventricle	Inferior left ventricle	Left and right ventricles	RVOT	Left and right ventricles	Left and right ventricles
Leads displaying J point/J wave	I, V4-V6	II, III, aVF	Global	V1-V3	Any of 12 leads	Any of 12 leads
Response of J wave/ST elevation to Bradycardia or pause	↑	↑	↑	↑	NA	NA
Response of J wave/ST elevation to Na-channel blockers	↓→	↓→	↓→	↑	NA	NA
Male predominance	75%			80%		
VT/VF	Rare. Common in healthy athletes	Yes	Yes, electrical storm	Yes	Yes	Yes
Sex dominance	Male	Male	Male	Male	Male	Male
Response to quinidine					Limited data	
J wave/ST elevation	↓	↓	↓	↓		

	Inherited				Acquired	
Characteristics	**ERS type 1**	**ERS type 2**	**ERS type 3**	**BrS**	**Ischemia mediated VT/VF**	**Hypothermia mediated VT/VF**
VT/VF	↓	↓	↓	↓		↓
Response to isoproterenol			Limited data		NA	NA
J wave/ST elevation	↓	↓		↓		
VT/VF	↓	↓		↓		
Gene mutations	CACNA1C, CACNB2B	KCNJ8, CACNA1,CACNB2B KCNJ8, CACNA1, CACNB2B	CACNA1C	SCN5A, CACNA1C, CACNB2B, GPD1L, SCN1B, KCNE3, SCN3B, KCNJ8, CACNA2D1, KCND3, MOG1, ABCC9, HCN4, KCNH2, KCNE5	SCN5A	Not available.

ERS: Early repolarization syndrome; BrS: Brugada syndrome; VT: Ventricular tachycardia; VF: Ventricular fibrillation; NA: Not available

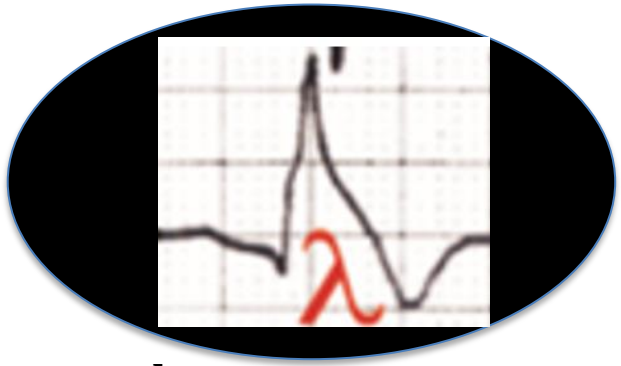
Miscellany



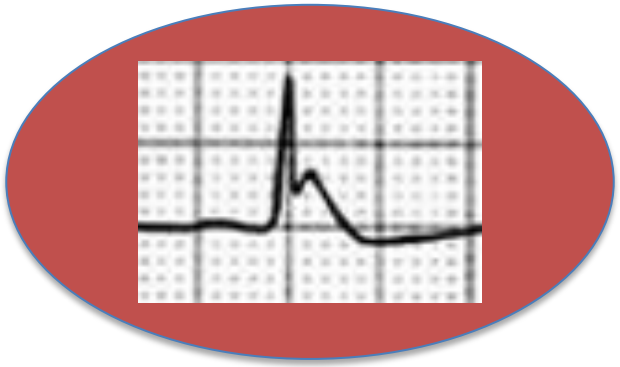
Electrocardiograms showing ST-segment elevation in various conditions.

1. **Tracing 1** is from a patient with LVH
2. **Tracing 2** is from a patient with LBBB
3. **Tracing 3,** from a patient with acute pericarditis, is the only tracing with ST-segment elevation in both precordial leads and lead II and PR-segment depression.
4. **Tracing 4** shows a pseudo infarction pattern in a patient with hyperkalemia. The T wave in V3 is tall, narrow, pointed, and tented. “Tour Eiffel T-wave”
5. **Tracing 5** is from a patient with acute anteroseptal infarction.
6. **Tracing 6,** from a patient with acute anteroseptal infarction and right bundle-branch block, include the remaining R' wave and the distinct transition between the downstroke of R' and the beginning of the ST segment. T
7. **Tracing 7**, from a patient with the Brugada syndrome, shows Type 1 pattern(coved): and J point and ST-segment elevation ≥2mm limited to V1 and V2. The ST segment begins from the top of the R' and is downsloping folowed by negative T wave.

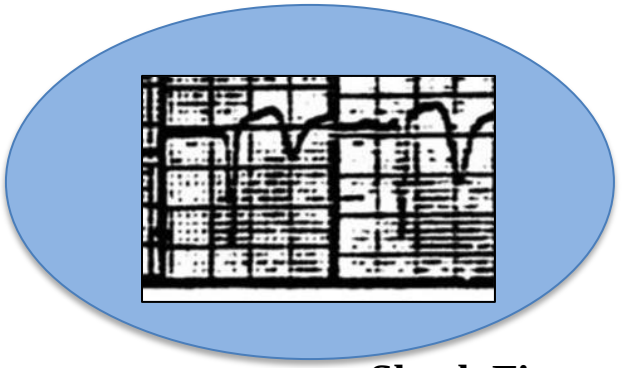
Lambda wave or End-QRS slurring,



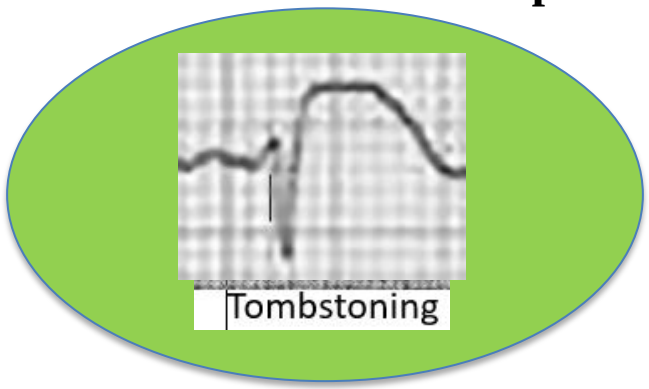
End-QRS notching



Prinzmetal Angina

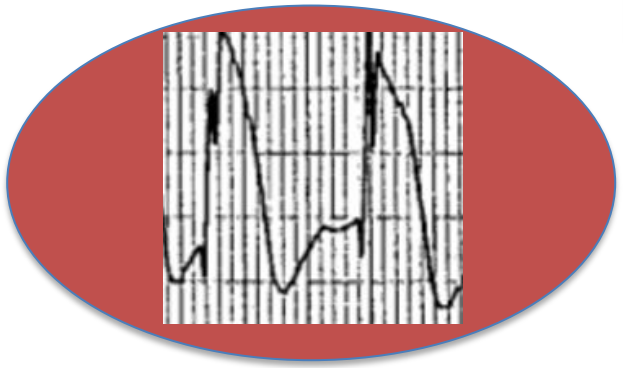


Tombstone or tombstone pattern

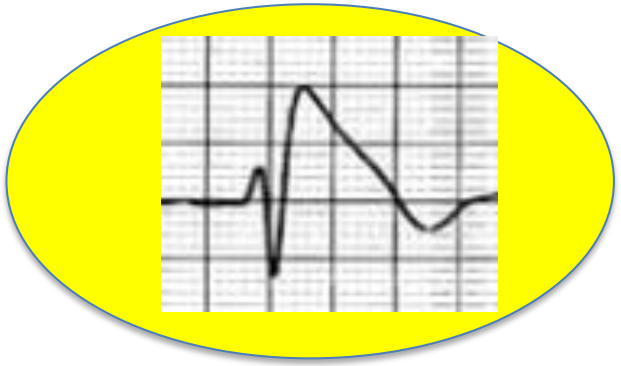


"Shark Fin", Lambda wave or End-QRS slurring, End QRS notching, spontaneous type 1 Brugada pattern, The “Spiked Helmet” Sign, Tombstone or tombstone pattern, Transient ST-Segment Elevation Immediately after Direct-Current (DC) and Prinzmetal angina are deadly ominous ECG Signs with similar changes in ST-segment.

Shark Fin

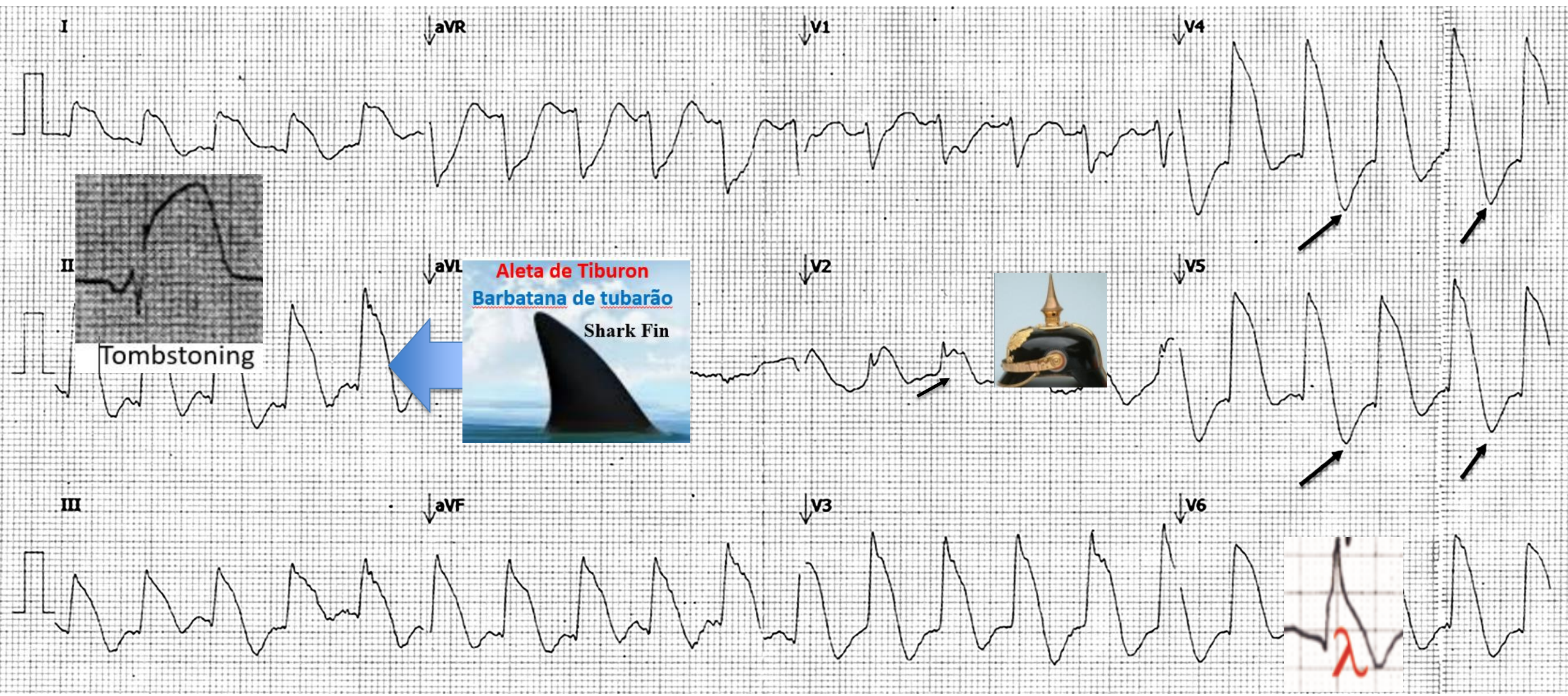


Transient ST-Segment Elevation Immediately after Direct-Current (DC)



Type 1 Brugada pattern V1/V2

The “Spiked Helmet”



Our case ECG1: First ECG recorded immediately after cardiorespiratory arrest recovery. What do the arrows mean? They show macrocopic T-wave alternans (TWA)

Final Conclusion

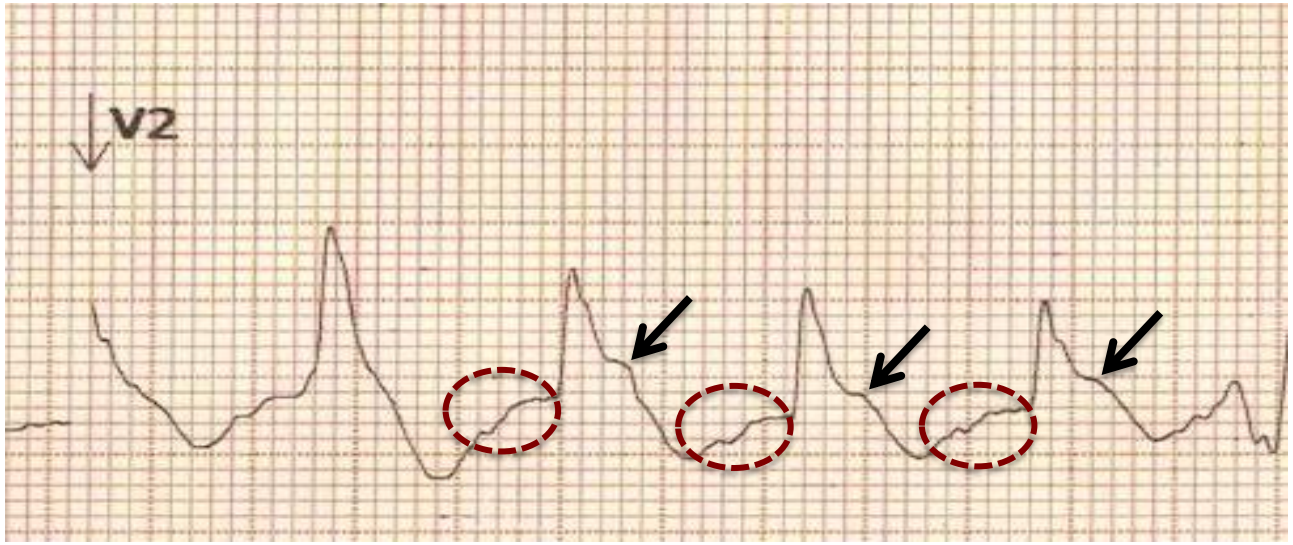


Fig. : The main ECG findings on SHS. The previous upward isoelectric line (red circles)is followed to convex ST elevation (arrows).

Final Considerations

- I believe that Prinzmetal angina caused by cocaine poisoning should enter the main differential diagnosis. However, I would like to point out that the patient reported that he had been abstinence from cocaine for 2 months, the event was not preceded by angina pain and the ECG changes were diffuse (although it does not exclude, it makes the possibility less likely, since it would require a vasospasm of multiple arteries). Another interesting fact is that the event lasted 6 hours and there was no increase in myocardial necrosis biomarkers. These 2 data also speak against Prinzmetal´s angina, since episodes of vasospasm usually last only minutes and for such a prolonged ischemia event, a significant increase in ckmb and troponin was expected, which did not occur in this case.
- Regarding the effects of cocaine, it is known that this drug can affect all ion channels of action potential. An important sodium channels block is observed in patients exposed to high doses of cocaine. However, in our case the phase I of the action potential was not affected, since the upward phase of the QRS is maintained. This data testifies against the acute effects of cocaine on sodium channels. Another relevant fact is that the patient is tachycardic. In cocaine intoxication, blockage in the sodium and potassium currents leads to bradycardia and QT prolongation and this would be one the reasons for an arrhythmic event.

Spiked Helmet: A sign or a Syndrome?

In my humble opinion, the so-called Spiked Helmet signal should be considered a syndrome as it includes ECG changes associated with severe clinical events in patients with hyperadrenergic status and significant metabolic disorders. Another issue is that ECG changes depend on the stage of manifestation of the syndrome, on our case the spiked was almost amputated by the extreme presentation. Finally, considering all aspects discussed, the most likely hypothesis for the case would be a **Spiked Helmet Sign/Syndrome.**

Thank you very much for your contribution to this case.

My sincere thanks to Prof. Dr. Pérez-Riera for his invaluable collaboration!

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This is my granddaughter Luzia. Is there anyone more beautiful than her? Andrés a happy grandfather