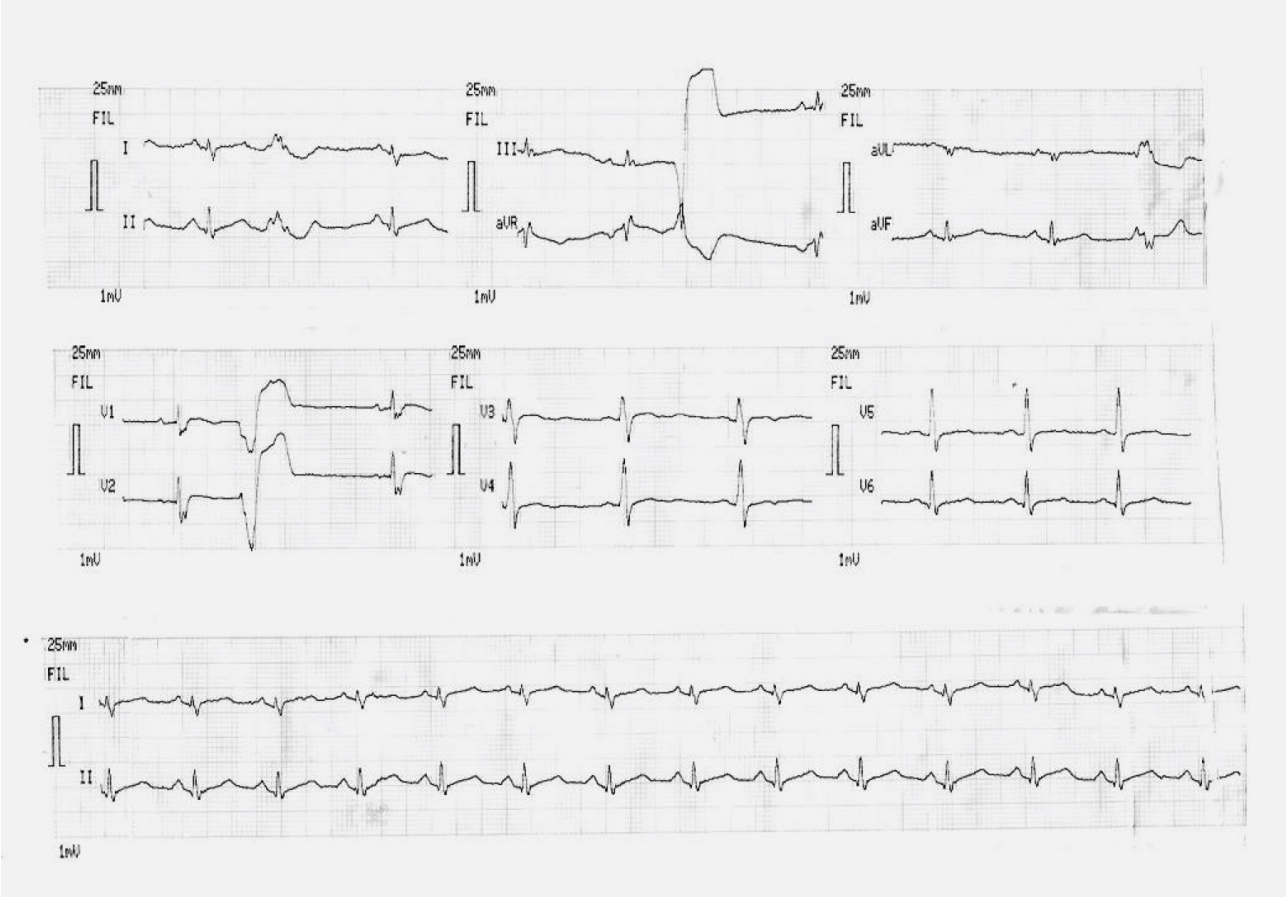
# Joven de 17 años asintomático con ECG muy patológico – 2016

Dra. Paola González

Hola a todos. Les presento el caso de un chico de 17 años que consulta por un apto físico para realizar tenis profesional. Asintomático y sin antecedentes familiares. Presenta el siguiente ECG.

¿Qué les parece?

Paola González



# OPINIONES DE COLEGAS

Ese ECG es muy anormal y altamente sugestivo de ARVC. (Cardiomiopatía del VD). Debe estudiarse.

Sergio Pinski

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Hello, ARVC must be excluded – epsilon wave, slow upstroke in S wave in V1-V2? (Right axis deviation also, extrasystole from right ventricle?).

Regards

Kjell Nikus

Tampere, Finland

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Ecg muy patológico para un joven de 17 años. En principio que deje de jugar hasta tener estudios diagnósticos.

Saludos.

Oscar Pellizzón

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Hola a todos ¿algún diagnóstico diferencial de ARVD?

Un saludo

Martín Ibarrola

Dear Paola I agree all colleagues that it is necessary others studies: High ressolution ECG, ECO, CMRI + ECG + genetic screening in proband and family members.

The baseline ECG shows:

I) Premature ventricular contractions (PVCs) with two morphologies:

A) Left bundle branch block-like pattern with positive R wave in leads: III and aVF what suggested PV Carising from RVOT

B) In II we observe a PVC with negative QRS. Consequently, the PVCs are polymorphic.

II) Parietal block on right precordial leads: a delayed S wave upstroke ≥ 55 msec in leads V1 – V3

III) Clear fragmented QRS (fQRS) evident in aVL, V1, V2 and aVL (multipes leads).

Comments: In ARVD/C fragmented QRS (fQRS) has a high diagnostic value similar to epsilon potentials by a highly amplified and modified recording techniques, such as right precordial leads ECG (R-ECG) and Fontaine leads (F-ECG) (**Peters 2008**).

fQRS refers to the 'slurs or notches' appeared on the R or S wave or if the total QRS complex had ≥ 4 spikes. fQRS can be registered as a normal variant mainly in seniors endurance athlete heart if it appeared randomly in just a few leads. fQRS presenting in multiple leads is more likely pathologic. The underlying cause is the regional delay in propagation of ventricular depolarization (**Monta 2008**). fQRS is highly prevalent in ARVC/D patients when applied to amplified and modified ECG recording techniques, including the use of the Fontaine Leads System (**Peters 2008: Hurst 1998**).

In real world practice, nevertheless, most ECGs available from ARVC/D patients and family members were obtained by using only the standard ECG recording technique. fQRS is easily recognizable from standard ECGs (S-ECG) and they are much more common in ARVC/D patient when compared with control subjects. Among them a notch before the end of R or S wave is characteristic, seen in 51% of ARVC/D vs 26% in controls. In ARVC/D, fQRS is often seen in multiple leads (**Zhang 2014**). Such changes, however, are common in control subjects as well. In the latter, the QRS complex is wider (**Dechering2013**).

fQRS complex, with various morphology, has been described as a diagnostic criterion of ARVC/D. Since fQRS is also prevalent in other types of cardiomyopathies (both ischemic and non-ischemic) (**Das 206; 2010**). fQRS is induced by radiotherapy in patients with breast cancer (**Adar 2015**), and in normal subjects, its use in ARVC/D diagnosis is limited.

Andrés R. Pérez Riera

Hola a todos.
Les comento que le hice una ergometría insuficiente detenida por agotamiento muscular. Fca máxima 174, ITT 22620, carga 900 kgm. Asintomático con EV polimórficas aisladas en el esfuerzo y en reposo de dos focos con imagen de BRI y BRD.
Ecocardiograma: Rao 33 AI 34 Sp 10 PP 9 DDVI 56 DDVD 37 con FSVI y FSVD conservada.

Le solicité un Holter que mostró un FC media de 127, mínima de 47 y máxima de 127. EV 4068 con 148 duplas, 2 tripletas, 2 bigeminias, 55 trigeminias. Las EV son de 2 focos de VD y VI. El paciente hizo su vida diaria sin jugar al tenis y completamente asintomático.

Paola González

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Hello Andrés:

Can make an analysis of diferential diagnosis of ARVD?

Paola performed another studies at the moment.

Best Regards

Martín Ibarrola

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I agree with the opinion of Dr Nikus. It would be interesting to perform the derivations of Fontaine.

Regards

Raimundo Barbosa Barros

i do not like this ECG at all..or let us say differently.. I love this ECG but the patient having this ECG is in trouble..nothing looks good and normal there..not the P wave not the QRS not the uggly VPC.. My feeling is that this young patient has a cardiomyopathy that could be an ARVD....This is my only diagnosis at the present time...The diagnosis should be made by simple echo that should be very anormal... Of course I maybe wrong… Personnally I have not seen such ECG at this age..

Professor Bernard Belhansen

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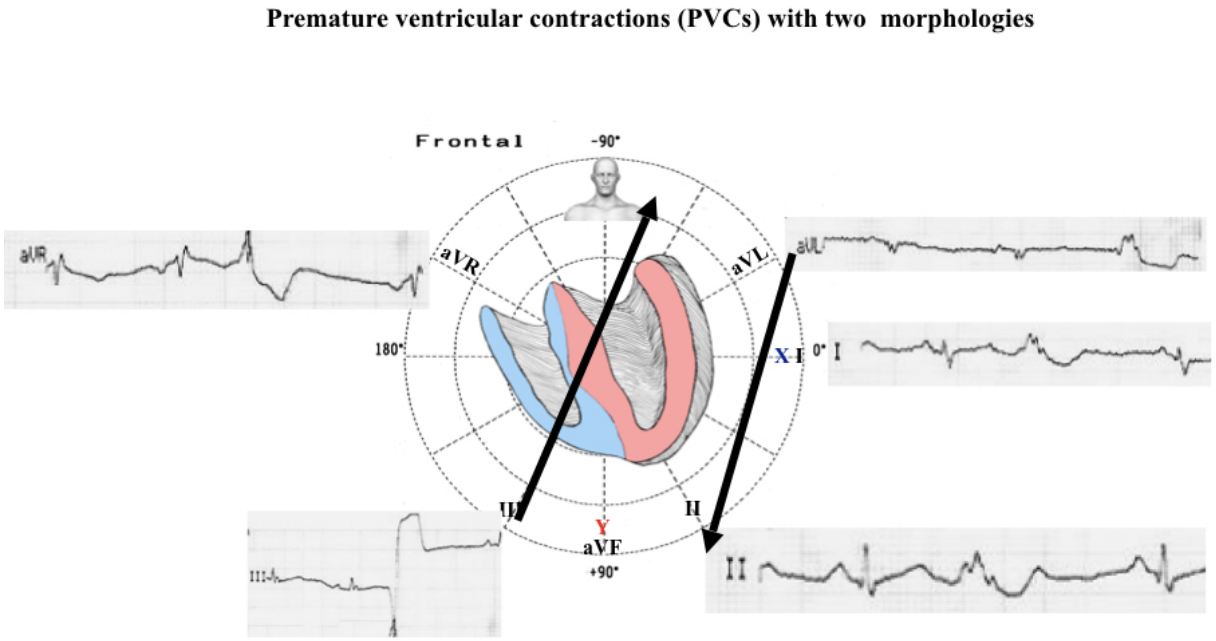
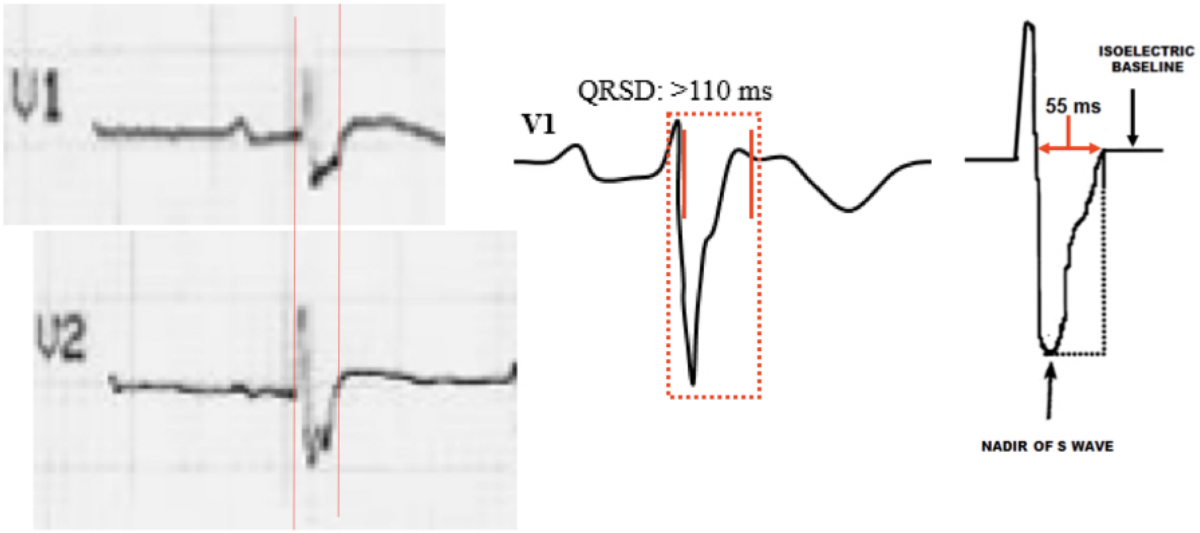
Prezada Paola estos son otras opiniones Y mi lúdica explanation

Andrés R. Pérez Riera

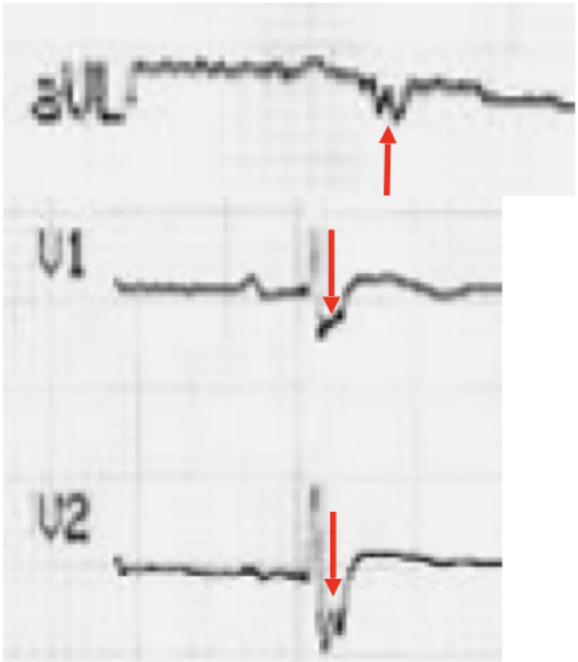


**Parietal block on right precordial leads: a delayed S wave upstroke ≥ 55 msec in leads V1 – V3**




**QRSD of $V_1$+$V_2$+$V_3$ /$V_4$,$V_5$, $V_6$ or ≥ 1.2 In ARVC this parameter is positive in 65% of cases. QRS prolongation located in right precordial leads**

**Fragmented QRS(fQRS) in aVL, V1, V2.**



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This is an ARVD period!

The epsilon waves is shown in leads III and aVF. The ST-segment of lead II is not clean---too blurry to tell there.

Yes ARVD battery test is recommended.

Here I'm inviting two biggest ARVD GURUs, hoping to hear their comments.

Thanks much for asking,

Li Zhang

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Hola a todos los que opinaron: yo ya conocía el caso y mi diagnostico fue el de una ARVD.

La Resonancia no la confirma, más una infiltración miocárdica por un proceso miocárdico, en este caso una supuesta perimiocarditis no suele dar trastornos segmentarios sino de

distribución homogénea por afectación de todo el miocardio, por eso es que no me convence una miocarditis viral.

La discordancia del ECO (operador dependiente), y desconociendo en que centro se realizó la RNM, con el electro característico de ARVD. Es que motivo la presentación.

No presento síntomas de pericarditis, no presentaba frote según refieris Paola, no cuenta con serologias virales y tampoco con test genético.

¿Qué otros signos de infección viral encontraste en los análisis, como el recuento de glóbulos blancos y la formula? ¿ESD y valor de PCR?

Además de troponina y BNP?

Me gustaría conocer además de lo que expresé ¿qué opinan los expertos?

Un saludo

Martín Ibarrola

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El Dx diferencial incluye sarcoidosis. Un PET con FDG es útil para ver áreas de inflamación. ARVC no está descartada. El estudio genético todavía está indicado.

El realce tardío ¿es transmural, epicárdico, o medio miocárdico?
Gran caso. La enseñanza es que él ECG era muy anormal.

Sergio Pinski

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Este es un DD aveces dificil Esto puede ayudar

**Cardiac Sarcoidosis versus Arrythtmogenic Right Ventricular Dysplasia/Cardiomyopathy**

Andrés Ricardo Pérez-Riera MDPhD.

Patients with cardiac sarcoidosis may present with clinical and morphological features similar to ARVC/D or cardiomyopathy (Ott 2003). Sarcoidosis is an inflammatory granulomatosis entity of unknows cause, characterized by multisystemic involvement. Practically no organ is immune to sarcoidosis; most commonly, in up to 90% of patients, it affects the lungs. (Hoitsma 2004). The most commonly involved organ in sarcoid related death has been reported to be the lung in western countries, while it was the heart in the Japanese autopsy series. (Iwai 1994).

The diagnosis of myocardial sarcoidosis is difficult and frustrating. Its clinical manifestations depend on the location and extentof granulomatous inflammation, and the symptoms and signs rangeamong benign arrhythmias, heart block, intractable CHF, intense chest pain, to fatal VF. (Sharma 2003).

The ECG finding may be normal or may reflect every degree ofblock of the atrioventricular junction and bundle of His and every type of arrhythmia along with nonspecific ST-T-wavechanges (Flemming, 1994).

Cardiac sarcoidosis should be considered in all young patients with unexplained conduction disorders, (Kollermann 2001) CHF or in cases of SCD (Lip 1996).

In extensive forms are frequently pseudo myocardial infarction patterns with pathological Q waves on ECG. (Shindo 1998).

MRI abnormalities, consisting of cardiac signalintensity and thickness, with the following threepatterns:

1) Nodular;

2) Focal increase in signal on gadolinium diethylenetriaminepentaacetic acid-enhanced, T1-weighted images;

3) Focal increased signal on T2-weighted images without gadolinium uptake.

The improvement or stabilityof the MRI findings is correlated with clinical features.

With corticosterotherapy, the MRI images improved either partially or completely, whereas.

The cardiac MRI may find its usefulness as a guide toobtaining EMB specimens and to monitoringthe response of the disease to treatment.

The study is small and lacks a correlation of myocardial histology with MRI features. However, the study clearly calls for a large multicenter trial.

The most significant drawback of MRI is that the patient with a pacemaker and/or automatic ICD will not beable to take advantage of it. In such patients, $^{201}$Tl scanning remains the test for assessing myocardial damage.

Cardiac PET using (18) F-FDG under fasting conditions (fasting (18) F-FDG PET) is a promising technique for identification of cardiac sarcoidosis and assessment of disease activity. The methodology can detect the early stage of cardiac sarcoidosis, in which fewer perfusion abnormalities and high inflammatory activity are noted, before advanced myocardial impairment. The sensitivity of fasting (18) F-FDG PET in detecting cardiac sarcoidosis was 100%, significantly higher than that of (99m)Tc-MIBI SPECT (63.6%) or (67)Ga scintigraphy (36.3%). The accuracy of fasting (18) F-FDG PET was significantly higher than (67) Ga scintigraphy. (Okumura 2004.).

An EMB is preferable, but the procedure has sensitivity aslow as 20% (Uemura, 1999). Others author referred sensitivity approximately of 50% thus, the search for a safe, reliable, and easily available diagnostic test for cardiac sarcoidosis continues. The

pathological feature is the presence of noncaseating granulomas that eventually form fibrotic scars.

The table below shows the principal differences between the two entities:

| | Cardiac Sarcoidosis | ARVC/D |
|---|---|---|
| Family history: | Absent. | Present in 30% to 50% of cases. When the disease is identified genetic screening should be performed among patient's family members. |
| Gender (M/F): | 1 to 1. | 2.9 to 1 |
| Age at presentation: | Young or middle-aged adults. | Adolescents and young adults, perhaps<br>There are rare references in childhood |
| Multisystemic involvement: | Yes. | No. |
| Chest pain: | Intense chest pain is refered. | No. |
| Clinical myocardial restrictives features: | Posible. | No. |
| Mitral regurgitation: | Is common. | Only in late estage with involvement of LV. |
| Pseudo myocardial infarction patterns on ECG: | Fequent in extensive forms. | No. |
| Chest roentgenogram: | Bilateral hiliar linphoadenopathy. | Eventually RV cardiomegaly. |
| Lungs affectation: | In up to 90% of | No. |

| | | |
|---|---|---|
| | patients.<br>Cor pulmonale is frequent. | |
| Pathological features: | Noncaseating granulomas that eventually form fibrotic scars. | Typical fibro-fatty replacement of the RV myocardium on dysplasia triangle. |
| More comum cardiac sites involved: | LV free wall and interventricular septum. | RVOT, RVIT, and apex of RV. |
| Pericardial effusion: | Are not uncommon. | Absent. |
| Improved MRIimages with corticosteroids: | Yes. | No. |
| Therapy with corticosteroids, hydroxychloroquine, methrotexate or cyclophosphamide: | Sometime are indicated.<br>(Mitchell 1997). Immunosuppressive and anticytokine treatments can be effective in severe systemic sarcoidosis and should be considered in sight-threatening disease. | No. |

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Hola a todos.

Desde ya muchas gracias por todos los comentarios y muchas gracias Andrés por la maravillosa explicación del diagnostico diferencial de sarcoidosis y displasia.

De los datos que me consultaron, previo a la resonancia le pedi un análisis de sangre que informó 7000 GB con formula conservada, hormonas tiroideas normales y Chagas negativo. Todo el resto normal.

En la resonancia el realce tardio fue en subepicardio e intramiocardio de los segmentos afectados.

Cuando lo vea le voy a solicitar serologia viral y PCR y ESD.

¿Tendría que pedirle un PET para descartar sarcoidosis?

Paola González.

El diagnóstico de sarcoidosis es importante porque el tratamiento con corticoesteroides puede frenar la progression y la evolución a la fibrosis.

El PET muestra inflamación, inflitración cellular; no necesariamente diferencia entre sarcoidosis y otra causa de myocarditis.

Puede ser útil una tomografía de tórax. La mayoría de los pacientes con sarcoidosis cardíaca tienen compromiso pulmonar o de ganglios mediastinales.

Este es un caso para considerar la biopsia endomiocárdica. Tratatando de llevar el biotomo a las áreas que se ven comprometidas en el septo en la MRI. La biopsia tiene muy alta especifidad pero baja sensibilidad. Debe ser interpretada por un expert, en un centro que hagan transplantes cardíacos.

Se puede aumentar la sensibilidad de la biopsia haciendo al mismo tiempo un mapeo de voltaje del VD y llevando el biotomo a zonas de bajo voltaje y fraccionamiento de las señales endocárdicas. Es engorroso pero vale la pena.

Consideraría una interconsulta con un expert, por ejemplo, en Bs As, el Dr. Sergio Perrone.

Cordialmente

Sergio Pinski

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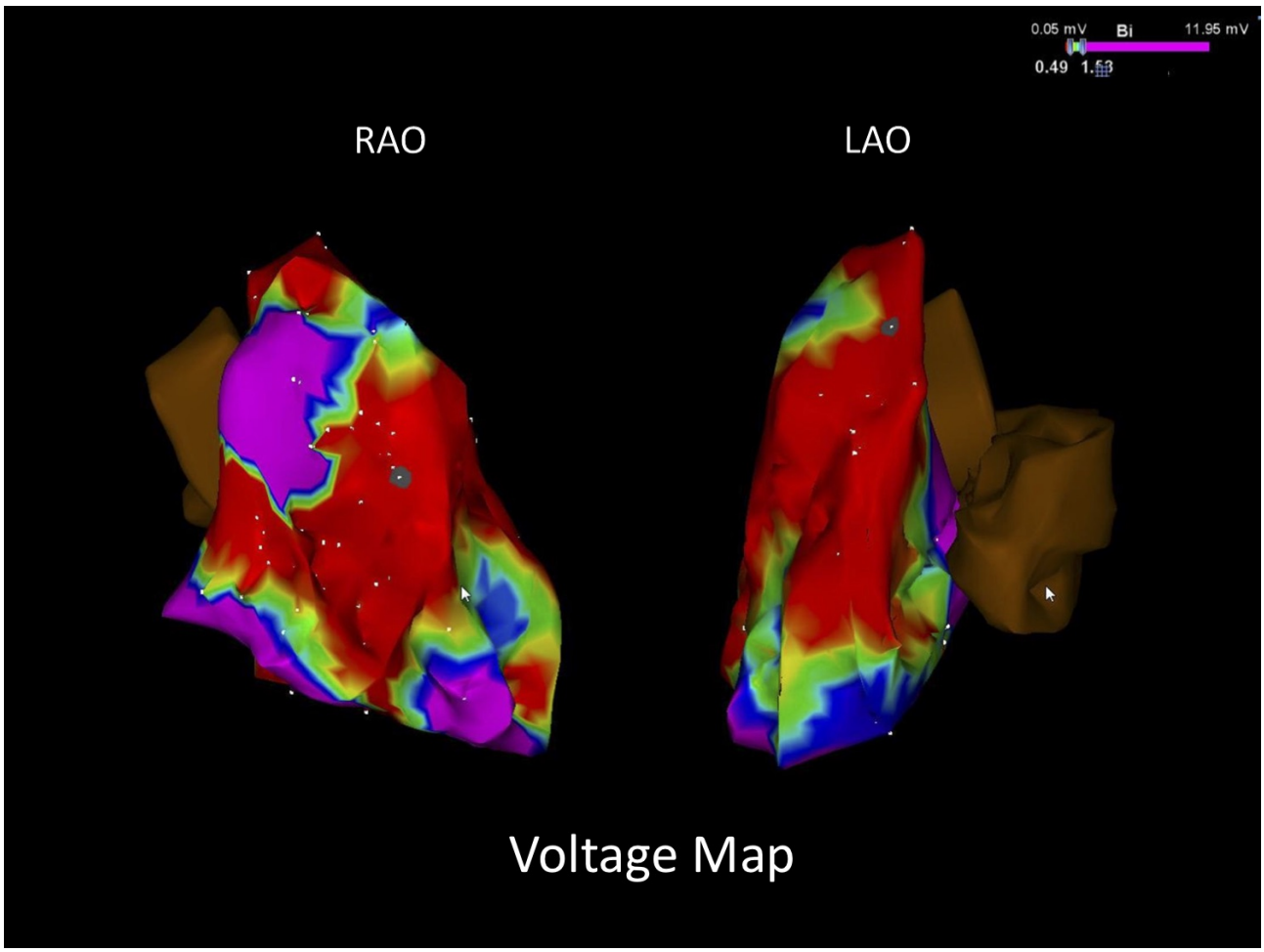
Este es uno de mis pacientes con Sarcoidosis que inicialmete parecía displasia arritmogenica del VD.

El mapeo del ventrículo derecho muestra en rojo las zonas de bajo voltaje que representan fibrosis y perdida de miocitos cardiacos.

Mejoro significamente luego del tratamiento con corticoesterioides y ablación de TV.

Saludos,

Mario González


0.05 mV
Bi
11.95 mV
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RAO
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Voltage Map