PSEUDO HYPOTHERMAL J WAVE ("OSBORN WAVE") IN SYMPTOMATIC PROFESSIONAL ADOLESCENT ATHLETE

Dr. Andrés R. Pérez Riera

Male, 17-year-old teenager, of Asian ethnicity ("sansei", grandchild of native Japanese). Professional baseball player for 8 months now. He practices regularly and daily the mentioned physical activity since his 10 years of age.

Main complaint: the brother says (they sleep in the same room) that approximately 5 h before, during night rest, at 3 AM, he uttered an intense guttural noise followed by sudden episode compatible with tonic-clonic seizures accompanied by anal and urinary sphincter relaxation.

Risk factors: he never smoked, he denies alcohol consumption or use of illegal drugs. He denies having diabetes, hypertension, dysthyroidism, and dyslipidemias.
Recent normal full "check up" demanded by the club.
He denies having cerebral dysrrhythmia, seizures. He denies allergic diathesis.

Family history: a cousin on his father side suffered sudden death due to unknown causes when he was 25 years old during night sleep.
He has a younger 12-year-old brother, using antiepileptic drugs since he was small, due to presenting tonic-clonic seizure events, followed up by a neurologist and using phenobarbital regularly. Even using antiepileptic drugs regularly he had "two seizure episodes".

Physical examination: good general state, pink cheeks, eupneic, no fever (axillary temperature 36.5°C). height, 175m, 62 kg, normal BMI.
Normal location and characteristics of ictus, rhythmical and normal heart sounds, without murmurs, and significant regular bradycardia of 40 bpm. BP 100/60. Normal jugular and abdominal pressure without visceromegaly.
Palpable peripheral pulse without edema or cyanosis.

ECG below
2-D transthoracic echo with color Doppler: normal

Request ambulatory Holter monitoring, Cardiac Magnetic Resonance image or magnetic resonance imaging of the heart with gadolinium infusion, signal-averaged ECG (SA-ECG) or high-resolution electrocardiography and VCG
I'm waiting for your valuable opinions.

Andrés Ricardo Pérez-Riera

PORTUGUESE LANGUAGE

PSEUDO ONDA J HIPOTÉRMICA ("ONDA DE OSBORN") EM ATLETA ADOLESCENTE PROFISSIONAL

Adolescente, 17 anos, masculino, da etnia asiática ("sansei" neto de Japoneses nativos).
Esportista Professional de baseball há 8 meses. Pratica regularmente y diariamente a mencionada atividade física desde os 10 anos de idade
Queixa principal: Refere o irmão (dorme no mesmo quarto), que aproximadamente 5h, atrás durante o repouso noturno sendo as 3 horas da madrugada preferiu um intenso ruído gutural seguido de súbito episódio compatível com convulsões tónico-clónicas acompanhadas de relaxamento esfincteriano fecal e urinário.
Fatores de risco: nunca fumou, nega consumo de álcool ou drogas ilícitas. Nega diabetes, hipertensão arterial, distiroidismo e dislipidemias.
Recente "check up" completo exigido pelo clube normal.
Nega disritmia cerebral, convulsões. Nega diáteses alérgicas
História familiar: um primo da linha paterna teve morte súbita de causa desconhecida aos 25 anos durante o sono noturno.
Um irmão menor de 12 anos em uso de anticomiciais desde pequeno por apresentar eventos convulsivos tónico-clónicos em acompanhamento com neurologista e em uso regular de fenobarbital. Mesmo em uso regular do anticomicial teve "dois episódios" convulsivos mesmo estando em uso regular do medicamento.

Exame físico: Bom estado geral, corado, eupnéico, acianótico, afebril, (temperatura axilar 36,5º) Altura 175, 62 kg, IMC normal.
Ictus de localização e características normais, bulhas rítmicas e normo-fonéticas, sem sopros e bradicardia regular significativa de 40bpm. PA 100/60. Jugulares e abdome normal sem visceromegalias
Pulsos periféricos palpáveis sem edemas nem cianose.

Ecocardiograma transtorácico bidimensional com Doppler colorido: Normal.
Solicitamos VCG, Ressonância nuclear magnética do coração com infusão de gadolíneo, ECG-AR,
Estou esperando vossas valiosas opiniões.
Andrés Ricardo Pérez-Riera.

Colleagues opinions

Dear Andrés

The ECG that you are showing to us is clearly "pathological." We could say that is normal. Represents an early repolarization syndrome, and strikingly this pattern occurs in almost all leads. It could also be confused with a Brugada, but it is not typical type 1 Brugada ECG pattern.

The presence of changes in the QRS-ST in the inferior and lateral can be found in up to 10% of the general population and is very common in athletes. These alterations until recently were considered benign. But studies of "healthy" patients who presented with primary VF cardiac arrest suggested an association between ST elevation and the slurring of the QRS in the inferior and lateral and the risk of VF. These changes suggested that athletes were of imbalance between the sympathetic and the parasympathetic. Although today we know from experimental studies that have alterations in the Ito current.

This patient is symptomatic, and there are familiar with sudden death and a brother with similar symptoms.

All proposed studies, I think that will give normal, although is necessary to rule out structural heart disease.

If this were my patient I wound conduct an isoproterenol infusion to observe what happens with the repolarization.

Still we do not have much knowledge about this "new entity".

In principle, I would suspend competitive sport.

The question I ask myself is: a new episode symptomatic cardiac arrest might be?. Therefore, the implantation of an ICD therapy would not be appropriate until we know more about this condition?
Perhaps Dr. Antzelevich enlightens us on this issue. In addition to the many colleagues in this prestigious forum with expertise in these patients.

Embrace and waiting for opinions.

Oscar A. Pellizzon Argentina MD.

The ECG shows classic changes of early repolarization in almost all leads. The J wave is quite pronounced and the ST segment is flat to depressed which according to both

Finnish "prospective" study and that of Viskin portends a poorer prognoses. He should have a 24 hour Holter recording to see whether there are high density PVCs wich trigger episodes of Polmorphic VT. The symptoms are quite worrisome especially as they occurred during sleep. Similarly the family history is also quite worrisome. Think he should have genetic testing as several genes (ie genes encoding for Ikatp as well calium channel proteins) have been associated with J wave syndrome. He needs a defibrillator.

Melvim Sheiman

Dear Andres and Colleagues

This indeed appears to be a classical case of Type 3 Early Repolarization Syndrome (ERS). Based on presenting symptoms, the patient is at risk for SCD. In addition to consideration of an ICD, quinidine therapy would be advisable. If quinidine is not well tolerated, cilostazol might be a reasonable alternative. It would be of interest to gauge the response to exercise, which would be expected to normalize the ECG and the response to vagal maneuvers, which might be expected to accentuate the ERS phenotype and provoke polymorphic VT. Most importantly, genetic screening should be performed to determine the underlying genetic basis. We would be pleased to provide this service at no cost. Instructions for collection and delivery of blood together with informed consent forms can be downloaded from

http://www.mmrl.edu/genetic-screening/

Best regards

Charlie Antzelevitch

This is not ER (early repolarization). This patient has what we call MER "monstrous early repolarization," and he almost died a few nights ago because the nocturnal seizures that he had were very likely due to ventricular fibrillation that miraculously terminated spontaneously (as often happens in idiopathic VF).
The differential diagnosis is nocturnal epilepsy. In this regard, a very important aspect of the clinical history is missing: If after the seizures he regained consciousness rather quickly and became alert within minutes (in other words, if within minutes he was capable of responding to question and understood where he was) then this cannot be a grand-mal seizure and self-terminating ventricular fibrillation can be diagnosed with confidence. The reverse however, is not true: although a prolonged state of sleepiness and confusion is regarded as characteristic of the postictal period after epilepsy, malignant ventricular arrhythmias may be long enough to cause transient anoxic brain damage. With this electrocardiogram, this young boy should be observed in intensive care until definitive therapy is started.

Quinidine is the drug of choice. I would do an electrophysiologic study which will most likely demonstrate inducible VF. I would then start quinidine at the highest tolerated dose and repeat the EPS after a few days. If he is non-inducible with a very aggressive protocol of extrastimulation, then he has the option of quinidine therapy without ICD implantation.
ICD without quinidine is not a good option because these patients tend to have VF-storms that are often fatal even with an implanted ICD.

Sami Viskin (Tel Aviv).