

during exercise stress testing or following catecholamine infusion.⁸ Recently, there have been reports of the use of diagnostic thresholds such as more than 10 premature ventricular contractions/minute,⁷ bigeminy,⁶ or couplets as the “minimum” ventricular arrhythmia, whose presence is indicative of a diagnosis of CPVT. In addition, although catecholamine infusion is not a totally reliable test, it continues to be used to enhance sensitivity in the diagnosis of this disease, especially when dealing with an index case. However, recent studies have reported the limited usefulness of catecholamine infusion because of its very low sensitivity (28%)^{7,9} and specificity⁹ for the diagnosis of CPVT. In one study, it was positive in 56 patients with a negative exercise stress test.⁷

The reality is that the available tests are insufficiently sensitive, and their negative predictive value is much lower than desired. However, the articles referred to in this letter reports results in terms of sensitivity (89%) and negative predictive value (93%) that do not agree with those reported to date, and convey the message that a negative exercise stress test rules out CPVT. These data probably require an in-depth study of a larger number of members of the family in question and, of course, cannot be extrapolated to other populations with other mutations in what, in our opinion, constitutes a selection bias.

In accordance with the recommendation of the scientific societies,⁵ a negative exercise stress test does not rule out CPVT. The disease can be confirmed by the presence of specific ventricular arrhythmias during exercise but, in the context of family screening, just 1 premature ventricular contraction is enough to render the results of the test abnormal, and probably justifies the introduction of preventive treatment with beta-blocker therapy. Moreover, the use of catecholamine infusion should be restricted to selected cases, and should not be included in the protocol to be applied on a general basis.

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Sensitivity and Negative Predictive Value of Treadmill Exercise Stress Testing for the Diagnosis of Catecholaminergic Polymorphic Ventricular Tachycardia. Response



Sensibilidad y valor predictivo negativo de la ergometría para el diagnóstico de la taquicardia ventricular polimórfica catecolaminérgica. Respuesta

To the Editor,

We wish to thank Ruiz Hernández and Wangüemert Pérez for their interest in our articles.^{1,2}

We used diagnostic criteria that were accepted at that time. With the recent consensus,³ doubts remain only about II:9, which requires the presence of *ventricular premature beats* or *bidirectional ventricular tachycardia* in relatives during the exercise stress test (EST).³ Other authors require *frequent isolated ventricular premature beats*.⁴ A family member, especially if older than 40 years, can have a few ventricular premature beats, despite having a negative genotype. Thus, the minimum number of ventricular premature beats is imprecise and two isolated premature beats appear to be insufficient to make a diagnosis of such importance for patients and their descendants.

The yield of EST varies (ranging from the 25% reported by Ruiz Hernández and Wangüemert Pérez to 100%),⁵ and perhaps depends on the point at which the mutation occurs. We had no intention of conveying the idea that a negative EST rules out the disease.^{1,2} A conclusive genetic study supports a positive EST and identifies carriers with a negative EST. In its absence, we perform Holter monitoring and an epinephrine test in relatives with a negative EST (not included in the consensus statement),³ and if they test negative, these individuals undergo follow-up with periodic EST. There are no studies on the value of epinephrine challenge in family members, but its usefulness has been documented in probands (which is included)³ and in families with cardiac arrest with preserved left ventricular ejection fraction⁶ and may justify its performance in the present context.

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