ECG Contest

Response to ECG, December 2019

Respuesta al ECG de diciembre de 2019

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This patient has a marked QT prolongation (≈ 520 ms) with T-wave macroalternans, and he presented with a cardiorespiratory arrest from which he receoved,[1] and so answer 1 is incorrect. This is an autosomal-recessive condition with a poor prognosis, which is more frequent when consanguinity is present, and which characteristically includes congenital deafness[2] (answer 2, correct). Given the family history of sudden cardiac death in early childhood, and the electrocardiographic abnormalities observed, diagnosis of obstructive hypertrophic cardiomyopathy is unlikely (answer 4, incorrect). Timothy syndrome, although a congenital long QT syndrome, is an autosomal-dominant form that is associated with other disorders, such as mental retardation, autism, dysmorphy, calcium metabolism disorders, and immunodeficiency, and so answer 3 is also incorrect.

REFERENCES

1. Goldenberg I, Moss AJ, Peterson DR, et al. Risk factors for aborted cardiac arrest and sudden cardiac death in children with the congenital long-QT syndrome. *Circulation.* 2008;117:2184–2191.
2. Giudicessi JR, Ackerman MJ. Prevalence and potential genetic determinants of sensorineural deafness in KCNQ1 homozygosity and compound heterozygosity. *Circ Cardiovasc Genet.* 2013;6:193–200.

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