Brugada ECG with very tall R waves Porminent anterior forces – 2012

Prof. Dr. Hanno Tan

Dear Andres,

How are you? I really enjoyed your paper on VCG in Brugada syndrome. Great work, congratulations once again!

Arthur and I would like to ask your opinion about a patient who is now admitted to our department.
It's a 17 year old Brazilian boy (,, normal build) who was born in the vicinity of Sao Paolo and adopted by Dutch parents. He moved to the Netherlands at the age of 2. He suffered out-of-hospital cardiac arrest from VF while at rest (on vacation with his parents). There were no apparent causes for VF. His history is unremarkable and he uses no drugs. His family history is unknown. Our workup has included the following:

1) 12-lead ECG: conduction slowing and very tall R waves in V1-V2 (attached)
2) ajmaline testing: type 1 Brugada ECG and further very prominent conduction slowing at 30 mg ajmaline, at which point the test was stopped. We also measured VCG during ajmaline testing.
3) signal averaged ECG: late potentials present
4) cardiac MR: normal

The ajmaline test clearly is positive, evoking a clear Brugada-type respons. While this strongly suggests Brugada syndrome (along with his conduction slowing and the absence of structural abnormalities that can be detected by cardiac MR), his very tall R waves in V1-V2 are unlike any other Brugada syndrome patient known to us. We also consider the possibility of a structural disease that cannot be detected with the methods that we employed, and that is associated with a positive ajmaline test. One could think of Chagas disease or ARVC. We would be quite curious about your ideas on this. Have you ever seen a similar case? Would VCG analysis help (if so, would you need the original digital VCG files?)?

Best

Hanno Tan

Estimados colegas este es un desafío que nos envía el Professor Hanno Tan un destacado pesquisador de la escuela de Amsterdam cuyo jefe es el Profesor Arhtur Wilde. Hanno ha escrito importantes manuscritos principalmente en referencia a las mutaciones del gen SCN5A.
Uno de ellos de gran impacto en la prestigiosa revista Nature.
Cuando vi el trazado confieso que me quedé perplejo y feliz porque a mi criterio junta en un solo paciente lo que vengo estudiando hace más de 1 década: el síndrome de Brugada y el bloqueo de la fibras medias del ramo izquierdo o “Left Septal Fascicular Block”
El trazado pertenece a un joven de 17 anos nacido en Brasil próximo de Sao Paulo y adoptado por una pareja de Holandeses. El vive en Holanda desde su tierna edad (2

años) Cuando gozaba de vacaciones con sus padres adoptivos sufrió una súbita e inesperada parada cardiaca durante el descanso. Su historia nada nos dice en especial no es drogadicto, pero su historia familiar es desconocida.
El ECG de 12 derivaciones muestra alteración dromótropa y muy altas ondas R en V1-V2
El test provocativo con ajmalina originó un patrón tipo 1 y una severa alteración dromótropa. También realizou VCG durante la ajamlina.

El equipo considero la posibilidad de cardiopatia estructural como ARVD?C o Chagas disease

Andres R. Pérez Riera.