Research

Screening of long Q-T syndrome in patients with congenital sensorineural hearing loss (Jervell and Lange Neilesen syndrome):prevention of fatal events

abstract

The idiopathic long Q-T syndrome is an infrequently occurring disorder in which affected individuals have an unusual electrocardiographic repolarization abnormality presenting as syncope or loss of consciousness related to ventricular tachycardia or fibrillation. Congential long Q-T prolongation can be associated with congenital deafness in an autosomal recessive manner (Jervell and Lange-Nielsen syndrome). The purpose of this stuff was to screen this electrocardiographic abnormality in deaf-mute school children in our population, which has not been yet performed. Of 1190 patients with hearing loss, 779 had congenital sensorineural deafness (CSD), aged 13±3.8 yrars (4-24), 63% female and 37% male. The family history of deafness was as follows: Cardiac axis deviation was found in 56(7%) patients. Electrical conduction abnormalities were found in 12(15%) patients, Wolff-Parkinson-White syndrome, sinus bradycardia, and sinus aryhmia were found in 2(0.25%), 4(0.5%), and 3(0.38%) patients, respectively. The Q-T interval, and Q-Tc duration were 312.6±28.9ms(200-500ms. median 320ms), and 383.6±29.3ms (232-527ms, median 413ms), respectively. Long Q-T syndrome was found in 4(0.5%) patients (3F,and 1M). Two of these 4 patients had total deafness and 2 had profound hearing loss. None of the patients with mild deafness had Q-T prolongation. Only one of these patients were symptomatic, and had been treated as a case of epilepsy for several years.This data supports the presence of long Q-T syndrome in patients with sensorineural hearing loss in our population, so routine electrocardiographic screening of anyone with congenital deafness is warranted to prevent subsequent associated cardiac arrythmias and sudden cardiac death.

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