Reply to the Editor-Carnitine and Ventricular Repolarization: A Complex Link?

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Dr Acampa discussed the link between QT interval abnormalities, ventricular arrhythmias, the autonomic nervous system (ANS), and metabolic disorders associated with the low level of carnitine. First, in Rett syndrome the level of carnitine is low but not null as in primary carnitine deficiency (PCD). Rett syndrome (RS) is a severe neurodevelopmental disorder associated with a high frequency of seizure (68%) in contrast to PCD. It is known that the frequency of sudden death is high in the epileptic population.[1] Second, Rasmussen et al[2] and Guideri et al[3] did not report abnormalities of the QT interval in PCD and RS. De Biase[4] and Schimmenti[5] noted one patient with prolonged QT interval and ventricular arrhythmias, but a genetic mutation in the main genes involved in long QT syndrome was not ruled out. Third, short QT syndrome published by Brugada is rare and probably underestimated. Recently, several abnormalities of the T wave were reported in PCD.[6] The effect of the ANS on ventricular repolarization and ventricular arrhythmias is well known. All conditions associated with ANS dysfunction favor cardiac arrhythmias. However, in the Guideri study[3] the low frequency/high frequency ratio was unchanged after carnitine supplementation, demonstrating a limited effect of carnitine supplementation on heart rate variability. Finally, the electrophysiological effects of acyl-L-carnitine and L-carnitine are different[7]: in vitro carnitine has no effect in contrast to acyl-L-carnitine, which increases the potassium current I_{Kr} that plausibly decreases the QT interval. These effects should be studied in the intact heart.

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