ARRHYTHMOCGENIC SUDDEN DEATH IN THE MODERN SETTING SIDS. AN OVERVIEW

Authors: Rossi L, MD, Matturri L, MD, PhD, Ottaviani G, MD - Institute of Pathology, University of Milan, Italy

Besides the quoted pathogenetic backgrounds of cardiac SIDS, and even overcoming the statistical exaggerations of Schwartz’ group who arbitrarily put forward the LQTS (1), the present authors stand convinced that the presence of accessory AV pathways can play an important role, in an unpredicted but significant number of cardio-arrhythmogenic SIDS. These authors themselves studied an uncommonly numerous case group of SIDS who exhibited (and likely succumbed to) as a unique histopathological evidence, accessory AV communications, however ‘silent’ on the clinical plane, since they lacked ECG-confirmed tachy-arrhythmias (2). To favour their own conviction, the present uphelders of this thesis do point out the anatomoclinical knowledge that most of the high-risk tachy-arrhythmias in early infancy show a typical ECG of a junctional reentry, together with the inherent implications of a potential or actual degeneration into ventricular fibrillation. The fact that the lethal event, or its threat, would not be recorded electrically, depends on the very same circumsidential ‘limitations’ emphasised by Schwartz and co-workers among the over 34,000 EKGs of the never-ascertained suspicion of an LQTS pathogenesis; indeed, this was only documented by objective proof 2 years after the publication of the original article. In this article no case (i.e. less than 1,34,000) ever presented with confirmed risk neither of arrhythmic death nor of related dysfunctions. To confirm the missing reliability of Schwartz’s thesis, even after the sporadic and numerically limited set of inherent controls by genetic investigations (3); yet, accurate as they were, such controls did not take into consideration the newer data concerning the genetic background of high-risk dysrhythmias, both sudden and lethal, as causes of SIDS beyond cases of cardiac failure and/or hypertrophy. Such gene, namely the K+CHIPz (or Kv channel-interacting protein or Ito) provides the Ca-independent regulation of K+onic channels during early repolarization, thereby playing a key role in regular myocardial electrical activation. For this reason such a genetic mechanism configurates a plausible alternative to the exclusivist Schwartz’ hypothesis focused on the equation SIDS/LQTS alone. Recent studies have also borne out further close clinico-patho-genetic analogies with arrhythmogenic lethal late repolarization, attended by fetal developmental impairments of the conduction system, often resulting into accessory AV pathways, whose high frequency in SIDS has been documented in the present authors’ cases and is entirely consistent with junctional tachycardia (2). It is important to underline, in this connection, that Hai-Chien Kuo and al.(4) emphasize the possible role of the conduction system among the ontogenic substrates of the Ito’s abnormalities, such a genetic clinico-pathological suggestion is further substantiated by the recent work of one of the present authors (5) on the life-threatening potential of WPW syndrome, whose commonplace accessory AV pathway substrate was proven to occasionally rely on genetic familiarity (6). All in all, under the light of these late findings, which alligne among the developmental backgrounds of an arrhythmogenic SIDS the postnatal persistence of AV junctional accessory pathways, one must better re-consider, critically, the overemphasised and overintrinsic LQTS/SIDS equation of the Schwartz’ school. Indeed, the basic arguments of this, rely on data that are being overcome, at the probable unawaresness of the upholders themselves.

REFERENCES