CORRESPONDENCE

Inherited Cardiac Arrhythmias: Diagnosis, Treatment, and Prevention

by Dr. med. Britt-Maria Beckmann, Dr. med. Dipl.-Biochem. Arne Pfeufer, Prof. Dr. med. Stefan Kääb in volume 37/2011

Exercise Tolerance in Genetic Cardiac Disorders

Cases of sudden cardiac death in sport in younger people are primarily caused by cardiac disorders with a genetic background, such as hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy (ARVC), familial dilated cardiomyopathy, and disorders of ion channels. Because of a lack of studies, reliable risk stratification is not possible. For this reason, any recommendations issued by specialist societies tend to be defensive. However, declaring participation in competitive sports as generally off limits, such as is recommended in the article by Beckmann et al (1), is not justified in terms of individual situations.

Athletes characterized as genotype-positive/phenotype-negative pose a particular challenge. US guidelines allow their participation in competitive sports (2), whereas European guidelines allow merely recreational sports (3). Since penetrance is variable, not every carrier of a pathogenic gene for example for hypertrophic cardiomyopathy develops pathological hypertrophy with an arrhythmogenic substrate. The argument in favor of competitive sports is supported by the fact that no data exist that would justify stopping athletes with a positive genotype, but a normal medical history and without any sign of a pathological clinical finding, from participating in competitive sports. Regular re-evaluation by a physician is however mandatory. The restrictive European recommendations are based on the hypothesis that competitive sports might trigger cellular mechanisms that induce a pathological phenotype (4). Ultimately, the controversial expert opinions show that especially in professional athletes, the decision whether the athlete is fit to compete must be made on an individual basis.

Differences between types of competitive sports should also be taken into consideration. Types of sports with low static and dynamic stress, such as golf or nine-pin bowling, can be pursued competitively even in the presence of arrhythmogenic genetic cardiac disorders. Again, the European guidelines are more restrictive and limit this to hypertrophic and dilated cardiomyopathy with a low risk profile (2, 3).

DOI: 10.3238/arztebl.2012.0236a

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Conflict of interest statement

The author declares that no conflict of interest exists.

In Reply:

In special circumstances, deviating from the recommendations with regard to competitive sports may be justified, especially for activities such as golf or bowling.

Ultimately, the decision lies with the treating specialist, who needs to consider all findings. Especially in asymptomatic relatives without any sign of subclinical disease (normal ECG and echocardiogram) with a positive genotype, a decision in favor of competitive sports may be justified (not in long QT syndrome [LQTS], catecholaminergic polymorphic ventricular tachycardia [CPVT], laminopathy [LMNA], and arrhythmogenic right ventricular cardiomyopathy [ARVC]).

In routine clinical practice, most patients are not professional athletes but children and adults who, at most, exercise to an average extent, and their quality of life is not restricted by not being able to pursue competitive sports. A relatively common exception to this rule, however, are adolescents, in whom the pertinent question concerns being allowed to play association soccer. Circumspect weighing-up of the risks and benefits, and, if required, psychological treatment, are of the utmost importance in this setting.

Since intense sports activities are a controllable, recognized risk factor, this should be communicated to persons affected by inherited cardiac arrhythmias (1).

The variable penetrance of the disorders described should not per se be used as an argument in favor of competitive sports. There are signs that in persons with the genotype who are primarily asymptomatic, competitive physical activities—especially those that entail high dynamic stress—may affect the course of their condition negatively. This is undisputed for most forms

of LQTS or CPVT, and there is evidence that this may exert a negative influence on the course of structural cardiac disorders, such as ARVC, or, in the case of cardiomyopathy due to mutations in the LMNA gene. Studies have shown that intense dynamic exercise, which was stopped an average of 15 years before the disorder manifested, constitutes a predictor for an unfavorable clinical course (2, 3).

DOI: 10.3238/arztebl.2012.0236b

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Conflict of interest statement

Professor Käb and Dr Beckmann have received third-party funding for research projects. ANR SCD Gene: 01 KU0907, M4 innovative therapeutic strategies for cardiac arrhythmias and drug safety: 01 EX 1021 E, NGFN Plus: 01 GS 0838.