

Medico legal Aspects of Sudden Cardiac Death

An Essay

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To
My parents, My wife and My daughter.

*without their patience,
understanding, support
and most of all love,
the completion of this work would not have been possible.*

Mokhtar Alkhrani

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ABSTRACT

Key Words: Sudden cardiac death, unexpected, medico legal aspects

Sudden cardiac death (SCD) is the most important challenge of modern cardiology. The importance of this problem is stressed by the fact that SCD from definition may occur suddenly in individuals that in spite of their heart disease are able to live a normal or near normal life. As in present times it can be prevented and the knowledge of pathological mechanisms as well as the correlation between SCD and associated diseases and risk factors is crucial.

Sudden cardiac death (SCD) is a major public problem accounting for approximately 20% of all mortality in the western world and up to 50% of overall cardiac mortality in developed countries.

The cause of SCD is different in different age groups. The risk factors are similar to those for coronary heart disease. In recent years, significant advances have been made in understanding the genetic basis of SCD.

The information necessary to establish a diagnosis of SCD is frequently lacking, because 80 percent of SCDs occur in the home environment, and up to 40 percent of sudden deaths are not witnessed.

In most cases of sudden death, cause and manner of death can be established, with many attributable to cardiac abnormalities evident at autopsy. A significant number of SCD, however, particularly in young people, remains unexplained following a comprehensive medico-legal investigation, including autopsy and laboratory tests.

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List of Abbreviations

ACC/AHA/HRS	The American College of Cardiology/American Heart Association/Heart Rhythm Society
AMVL	Anterior mitral valve leaflet
Ao	Aorta
ARVD/C	Arrhythmogenic Right Ventricular dysplasia Cardiomyopathy
BrS	Brugada syndrome
CACNA1	Calcium channel
CAD	Coronary artery disease
CASQ	Calsequestrin gene
CBN	Contraction band necrosis
CHD	Coronary heart disease
CPVT	Catecholaminergic polymorphic ventricular tachycardia
CT	Computed tomography
cTnT	Cardiac troponin T
DCM	Dilated cardiomyopathy
ECG	Electrocardiography
EF	Ejection fraction
EMD	Electromechanical dissociation
ESD	Exercise-related sudden death
FDCM	Familial DCM
FHC	Familial hypertrophic cardiomyopathy
GPD-L	Glycerol-3-phosphate dehydrogenase 1-like enzyme
HCM	Hypertrophic cardiomyopathy
HERG	H uman E ther-a-go-go R elated G ene
HF	Heart failure
HR	Heart rate
5-HT	5- hydroxytryptamine
ICD	Implantable cardioverter-defibrillator
IKI	Inward component of potassium delayed rectifier
IKr	Rapidly activating delayed rectifier
IKs	Slowly activating delayed rectifier
KCN	Cardiac potassium channel
KCJN2	Potassium voltage-gated channel gene, subfamily N2
KCNH2	Potassium voltage-gated channel gene, subfamily H2
KCNJ2	Potassium voltage-gated channel gene, subfamily J2
KCNQ1	Potassium voltage-gated channel gene, subfamily Q1
KvLQT1	Potassium voltage-gated channel
LA	Left atrium
LCA	Left coronary artery
LDL	Low density lipoprotein
LQTS	Long QT syndrome
LV	Left ventricular
LVD	Left ventricular dysfunction
LVEF	Left ventricular ejection fraction
LVFW	Left ventricular free wall
LVH	Left ventricular hypertrophy

LVNCCM	Left ventricular noncompaction cardiomyopathy
LVOT	Left ventricular outflow tract
MA	Methamphetamine
MDMA	Methylenedioxymethamphetamine
MI	Myocardial infarction
MMWR	Morbidity & Mortality Weekly Report
MRI	Magnetic resonance imaging
MSCT	Multislice computed tomography
MVP	Mitral valve prolapse
MYBP	Myosin binding protein
MYH	Myosin heavy chain
MYL	Myosin light chain
NBT	Nitroblue tetrazolium
N-Terminal	Amino terminal pro-brain natriuretic peptide
BNP	
PHSCD	Potentially hereditary sudden cardiac death
PMNs	Polymorphonuclear leukocytes
PVCs	Premature ventricular complexes
QTc	QT interval, corrected for heart rate
RCA	Right coronary artery
RCM	Restrictive cardiomyopathy
RCPATH	Royal College of Pathologists
RV	Right ventricle
RyR	Cardiac ryanodine receptor
SADS	Sudden Adult Death Syndrome
SCA	Sudden cardiac arrest
SCD	Sudden cardiac death
SCN	Cardiac sodium channel
SCN5A	Cardiac sodium channel gene, subfamily 5A
SIDS	Sudden infant death syndrome
SOFT	Society of Forensic Toxicologists
SQTS	Short QT syndrome
SUD	Sudden unexplained death
TdP	Torsades de pointes
TNNI	Cardiac troponin I
TNNT	Cardiac troponin T
TTC	Triphenyltetrazolium chloride
VF	Ventricular fibrillation
VS	Ventricular septum
VT	Ventricular tachycardia
VTC	Ventricular tachycardia
WHO	World Health Organization
WPW	Wolf-Parkinson-White

Introduction:

A major component of forensic pathology is the ability to recognize and interpret injury and determine its role in causing death, yet the majority of cases investigated by medical examiners or coroners are sudden deaths caused by natural disease. In some instances, there will be a well-documented history of potentially lethal natural disease, allowing for a straightforward cause of death determination. Unfortunately, the majority of cases present without ante-mortem documentation of significant pathology (**Dowling, 2005**).

Sudden death is defined as a witnessed or unwitnessed natural death occurring unexpectedly within 24 hours of a previously normal state of health. In sudden death, the immediate cause is almost always to be found in the cardiovascular system, even though topographically the lesion is not in the heart or great vessels (**Saukko & Knight, 2004**).

Sudden cardiac death (SCD) describes the unexpected natural death from a cardiac cause within a short time period from the onset of symptoms in a person without any prior condition that would appear fatal. The definition of SCD includes the time interval from onset of the symptoms leading to collapse and then to death, the unexpected nature of the event, and the specific cause of death. More recent definitions have focused on time intervals of 1 hour or less, which normally identify SCD populations having a 90 percent or more proportion of arrhythmic death. Because 80 percent of SCDs occur in the home environment, and up to 40 percent of sudden deaths are not witnessed, the information necessary to establish a diagnosis of SCD is frequently lacking (**Reynold et al., 2007**).

Sudden cardiac death (SCD) is a major public problem accounting for approximately 20% of all mortality in the western world. In the USA alone, SCD afflicts 450,000 people each year **(Ruey et al., 2008)**.

The World Health Organization reported an annual incidence of SCD of 1.9 per 1000 persons in men and 0.6 per 1000 persons in women **(Reynold et al., 2007)**.

SCD rates in developing countries are considerably lower, paralleling the rates of ischemic heart disease which is the most common substrate for SCD; In Egypt, for example, the mortality rate for ischemic heart disease alone is 21% **(WHO, 2002)**.

Egypt's Mortality statistics of cardiac arrest extrapolated in 2004 by WHO revealed that the total number of SCDs were 21,892 cases **(WHO, 2004)**.

It is very likely that a death that is delayed by hours will not be referred to the medico legal authority, as the diagnosis may well have made, and a death certificate can be completed by the attending doctors **(Richard, 2003)**.

However, several surveys in various countries have shown that where a physician offers a cause of death without the benefit of autopsy findings, the error rate is of the order of 25-50 percent, even in deaths in hospital **(Bowker et al., 2003)**.

In case where a clinical doctor can not so certify, the death is usually reported for medico legal investigation. In many countries such notifications form by far the largest proportion of medico legal autopsies reaching to 80% of coroner's autopsies, the reminder being suicides, accidents & homicides **(Gallagher, 2007)**.

After a properly conducted autopsy, a small proportion of cases will not reveal a cause of death. This is probably of the order of 2–5%. However, before the death is recorded as unascertained, it is important that appropriate ancillary investigations have been conducted. However, even with a full and thorough investigation, some natural disease processes such as cardiac conduction abnormalities will result in a negative autopsy. If investigated properly, a cause of death may still be identified (**Milroy, 2007**).

Aim of the Work:

This work aims at throwing light on disease processes & spectrum of causes of sudden or unexpected cardiac death, focusing specifically on its autopsy diagnosis as commonly encountered by forensic pathologists especially in negative autopsies.