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Task Force on Sudden Cardiac Death, European Society of Cardiology

Summary of Recommendations

S. G. Priori, E. Aliot, C. Blømstrom-Lundqvist, L. Bossaert, G. Breithardt, P. Brugada, J. A. Camm, R. Cappato, S. M. Cobbe, C. Di Mario, B. J. Maron, W. J. McKenna, A. K. Pedersen, U. Ravens, P. J. Schwartz, M. Trusz-Gluza, P. Vardas, H. J. J. Wellens and D. P. Zipes

The European Society of Cardiology has convened a Task Force on Sudden Cardiac Death in order to provide a comprehensive, educational document on this important topic. The main document has been published in the *European Heart Journal* in August 2001[1].

The Task Force has now summarized the most important clinical issues on sudden cardiac death and provided tables with recommendations for risk stratification and for prophylaxis of sudden cardiac death.

The present recommendations are specifically intended to encourage the development and revision of national guidelines on prevention of sudden cardiac death.

The common challenge for cardiologists, physicians of other medical specialties and health professionals throughout Europe is to realize the potential for sudden cardiac death prevention and to contribute to public health efforts to reduce its burden.

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Key Words: Cardiac arrest, sudden death, arrhythmias.

Definition

The term sudden cardiac death (SCD) has been used for several centuries and throughout this time different authors have debated how to define it most appropriately. SCD is defined as follows: 'Natural death due to cardiac causes, heralded by abrupt loss of consciousness within 1 h of the onset of acute symptoms; pre-existing heart disease may have been known to be present, but the time and mode of death are unexpected'[2]. A matter of debate has always been when an unexpected death should be called 'sudden' and 'how' the cardiac origin of the death should be ascertained. Several criteria have been proposed to link SCD to a specific 'mode' of death.

The clinical presentation of SCD is frequently used as a surrogate implying that a specific mechanism is involved. The more certain a specific mechanism is, the better preventive measures may be developed. Although it is true that in most cases of instantaneous death, such as after myocardial infarction, a tachyarrhythmia is the underlying cause, there are other mechanisms that may also lead to sudden death like aortic burst subarachnoidal aneurysm or rupture, cardiac rupture and tamponade, massive pulmonary embolism, and others. On the other hand, a death may still be arrhythmic in nature but may not occur suddenly, e.g. a patient who dies from the subsequent complications of an episode of sustained ventricular tachycardia after having been admitted into the hospital in haemodynamic collapse.

The key concepts that are central in the definition of sudden death are the non-traumatic nature of the event and the fact that sudden death should be unexpected and instantaneous. In order to limit sudden death to heart diseases, the word 'cardiac' has been added to forge the term 'SCD'. A further subclassification has been proposed to distinguish 'coronary' and 'non-coronary' SCD. The time frame used to describe the duration of the terminal event initially was 24 h but has subsequently been reduced to 1 h or even to an instantaneous event to render an arrhythmic mechanism more likely.

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Correspondence: Silvia G. Priori, MD, PhD, FESC, Chairman Task Force on Sudden Cardiac Death of the European Society of Cardiology, Fondazione Salvatore Maugeri, University of Pavia, Via Ferrata 8, 27100 Pavia, Italy. E-mail: spriori@fsm.it

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As a consequence, there has been a large inconsistency in the definitions used in the different clinical trials. The problems associated with defining the mode of death have been a matter of concern for many authors[3,4]. A very difficult issue is the classification of deaths that occur unwitnessed such as being found dead in bed. Most authors have erred in favour of classifying such events as SCDs, even though it is often impossible to define when the patient was last alive or for what duration he suffered any symptoms prior to death.

This document will propose recommendations for prevention of SCD that are based on results of trials and therefore will suffer from the unavoidable limitation of comparing studies that have used different definitions of sudden death. Furthermore, more recent trials have not analyzed the effect of devices and interventions on 'sudden cardiac death' but they have instead used 'arrhythmic death'. Conversely, not all sudden deaths are due to arrhythmias, specifically ventricular tachyarrhythmias.

In the analysis of trials, we have used whenever possible, the data specifically obtained in the subgroup having SCD as an end-point. When this was not available, data classified as arrhythmic death were used or when only cardiac mortality was available, it was assumed that a significant proportion of cardiac mortality was represented by arrhythmic death.

Recommendations are provided in the tables and are ranked as follows. Class I: conditions for which there is evidence that a given procedure is useful. Class II: there is conflicting evidence about the usefulness/efficacy of the procedure. Class IIa: weight of evidence in favour of efficacy. Class IIb: usefulness/efficacy less well established.

Epidemiology

The single most important cause of death in the adult population of the industrialized world is SCD due to coronary disease. The first recorded rhythm in patients presenting with a sudden cardiovascular collapse is ventricular fibrillation (VF) in 75–80%, whereas bradyarrhythmias are thought to contribute to a minority of SCD. In about 5% to 10% of cases, SCD occurs in the absence of coronary artery disease or congestive heart failure.

Incidence rates of SCD ranging between 0·36 to 1·28 per 1000 inhabitants per year have been reported[5]. In these studies only witnessed victims seen or resuscitated by the emergency medical services are included; these data are therefore an underestimate of the incidence of SCD in the general population.

The incidence of SCD occurring out-of-hospital varies with age, gender and presence or absence of a history of cardiovascular disease. In males between 60 and 69 years of age and a prior history of heart disease, SCD rates as high as 8 per 1000 per year have been reported[6]. In Maastricht[7] a population-based study monitored all cases of out-of-hospital cardiac arrest occurring in victims between 20 and 75 years of age. An overall yearly incidence of SCD of 1 per 1000 was recorded. Overall, 21% of all deaths were sudden and unexpected in men and 14·5% in women. Eighty percent of out-of-hospital cases occurred at home and about 15% on the street or in a public place. Forty percent of SCDs were unwitnessed.

Myerburg and colleagues[8] reviewed the issue of the risk of SCD in population subgroups, and their contribution to the overall burden of SCD. Based on a figure of 300 000 SCDs/annum in the United States, the population incidence was just over 1/1000/year. Any intervention applied to the general population to reduce the risk of SCD would therefore be given to the 999/1000 individuals per annum who will not die suddenly in order to prevent the death of one individual. The cost- and risk-benefit ratios imply that only general lifestyle advice could be given on a population-wide basis. Of course, higher risk subgroups of the population can be identified. Asymptomatic individuals with multiple risk factors for coronary disease are at higher risk than the population at large, while individuals with manifest coronary artery disease are at still greater risk. As will be discussed below, subgroups of patients with coronary disease at still greater risk of SCD are identifiable on the basis of previous myocardial infarction, ischaemia, impaired left ventricular function and previous life-threatening ventricular arrhythmias. Identification and appropriate management of these patients is at the heart of modern cardiology, and is the subject of much of this review. However, subgroups with progressively greater annual risks of SCD comprise a progressively smaller proportion of the total numbers of SCDs in the population. The logical conclusion of these figures is that the greatest opportunity to reduce the population burden of SCD lies in the reduction in the prevalence of coronary artery disease in the population at large[9].

Most Western populations have a high prevalence of coronary atherosclerosis in middle-aged and elderly subjects. Since coronary artery disease is commonly asymptomatic or unrecognized, the general population will contain an unknown proportion of individuals with advanced coronary disease. Epidemiological studies have also reported a high prevalence of unrecognized myocardial infarction and left ventricular dysfunction in the community[10]. Individuals with unrecognized coronary artery disease will, by definition, not be amenable to the preventive measures available to those with manifest disease. However, they may be identified if coronary risk factor screening is undertaken either in a systematic or opportunistic fashion.

Risk factors for sudden cardiac death in the community

Population studies in many industrialized countries have demonstrated that the risk factors for SCD are predominantly the same as those for atherosclerotic

coronary disease, namely increasing age, male gender, family history of coronary artery disease, increased LDL cholesterol, hypertension, smoking and diabetes mellitus[11]. As a matter of fact SCD epidemiology is changing as CAD is more successfully managed with statins, aspirin and beta blockers. Some studies have attempted to identify risk factors which may specifically predict SCD as opposed to acute myocardial infarction or other manifestations of coronary disease in population subsets without recognized heart disease. Among the specific risk factors studied, increased heart rate[12,13] and heavy alcohol consumption have been reported in several studies.

Sudden cardiac death may occur as a consequence of an inherited genetic abnormality affecting key proteins of the heart. Diseases such as long QT syndrome, Brugada syndrome, hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, catecholaminergic polymorphic ventricular tachycardia or dilated cardiomyopathy are the best known examples of monogenic diseases predisposing to SCD. Evidence supporting the existence of a genetic 'susceptibility factor' predisposing to SCD has emerged from large-scale epidemiological studies that have demonstrated a familial association of SCD.

The practical implications of current knowledge about the genetic basis of SCD are to encourage the assessment of family history in survivors of SCD. In the presence of familial clustering of cardiac arrests or SCD, the presence of a monogenic disorder (Brugada syndrome, long QT syndrome, hypertrophic cardiomyopathy . . .) should be carefully evaluated, particularly if these events had occured at young age.

SCD in myocardial infarction and heart failure

A. Risk stratification

Both, non-invasive and invasive tests have been introduced to help stratify post-myocardial infarction (MI) patients according to their risk for SCD[14–24]. The decline in cardiac mortality in the thrombolytic era has enhanced a limitation inherent in risk stratification, namely the low positive predictive value of any test. This limitation is partly overcome when these tests are not used alone, although an inevitable decrease in sensitivity results. Despite use of a combination of different tests to improve their predictive value, the positive predictive accuracy rarely reaches more than 40% at reasonable levels of sensitivity. An additional limitation is represented by the fact that some of these variables are inter-related (e.g. different autonomic markers all exploring aspects of vagal control of sinus node function); thus, they compete with each other when placed in a multivariate or regression model.

There are variables whose specific value increases when moving from the general population after myocardial infarction to specific groups of patients. An example is represented by programmed electrical stimulation, which cannot be recommended for all post-MI patients but which acquires powerful prognostic value when used in patients with depressed LVEF and the presence of non-sustained ventricular tachycardia, particularly in patients with large infarcts.

Table 1 Risk stratification in post MI with or without HF

Class I	Class IIa	Class IIb
Demographic variables LVEF HRV or BRS LVV	PVCs VTns Resting heart rate	LP PES TWA HRT Patency of infarct related artery

MI = myocardial infarction; HF = heart failure; LVEF = left ventricular ejection fraction; HRV = heart rate variability; BRS = baroreflex sensitivity; LVV = left ventricular volume; PVCs = premature ventricular contractions; VTns = non-sustained ventricular tachycardia; HR = heart rate; LP = late potentials; PES = programmed electrical stimulation; TWA = T wave alternans; HRT = heart rate turbulence analysis.

The available data suggest that strong combinations result from the association of a marker of structural damage, such as depressed left ventricular ejection fraction, with markers of autonomic imbalance related to electrical instability, such as depressed heart rate variability or baroreflex sensitivity.

Clever and balanced use of risk stratification parameters will allow appropriate therapeutic strategies to be used successfully to reduce the incidence of SCD (Table 1).

B. Primary and secondary prevention

Due to the complex mechanisms leading to SCD, mainly due to ventricular tachyarrhythmias, a variety of therapeutic targets may be considered[25,26]. These may range from limitation of infarct size and prevention of a new ischaemic event (resulting from progression of coronary artery disease and plaque instability) to modulation of neuroendocrine activation, antiarrhythmic and antifibrillatory actions, all designed to prevent or terminate ventricular tachyarrhythmias.

The terms 'primary' and 'secondary' prophylaxis are used unconventionally in the context of ventricular arrhythmia. Therapy that is given in order to prevent a sustained ventricular arrhythmia in patients who have not yet suffered a life-threatening ventricular arrhythmia, but who are at high risk of such an arrhythmia, is usually described as 'primary' prophylaxis. Similar prophylactic therapy recommended for patients who have already suffered a cardiac arrest or syncopal/hypotensive ventricular tachycardia is known as 'secondary' prophylaxis.

It is important to point out that studies on the efficacy of drugs/interventions on specific 'modes' of death in

Table 2 Primary prevention in post MI with or without HF

	Class I	Class IIa	Class IIb
Post-MI	Beta-blockers ACE-inhibitors Lipid lowering drugs	PUFA Amiodarone	
MI+ LV dysfunction	Beta-blockers ACE-inhibitors Aldosterone receptor blockers	Amiodarone	
Haemodynamically tolerated VTs		Amiodarone Beta-blockers	ICD Ablation Surgery
EF ⩽40% + spont. VTns + VTs inducible at PES	ICD		

MI = myocardial infarction; HF = heart failure; PUFA = poly unsaturated fatty acids.

Table 3 Secondary prevention in post MI with or without HF

	Class I	Class IIa	Class IIb
VF	ICD		
Non-haemodynamically tolerated VTs	ICD	Amiodarone Beta-blockers	

MI = myocardial infarction; HF = heart failure; VTs = sustained ventricular tachycardia.

myocardial infarction and heart failure are dependent on the reliability and validity of the classification used. Accordingly, total mortality is probably the only reliable endpoint in MI and heart failure (HF) trials. As a consequence, treatment of patients should be aimed at reducing total mortality.

Prevention of SCD in patients with myocardial ischaemia and myocardial infarction with or without heart failure is based on the use of drugs without electrophysiological action such as beta-blockers, ACE-inhibitors, lipid-lowering agents, PUFA aldosterone receptor antagonists[31–37]. Among antiarrhythmic drugs, amiodarone may be indicated in post-MI patients and more specifically in patients with spontaneous, sustained, well tolerated ventricular tachycardia[38–43]. Based on results of clinical trials, the prophylactic use of the ICD is indicated in post-MI patients with an ejection fraction below 40% presenting spontaneous non-sustained ventricular tachycardia and inducible sustained ventricular tachycardial[19,44]. The ICD is also recommended in survivors of cardiac arrest for secondary prophylaxis of sudden cardiac death[39–41] (Tables 2 and 3).

Hypertrophic cardiomyopathy (HCM)

HCM is a relatively common cardiac disorder (adult prevalence about 1:500) in which sudden unexpected

Table 4 Hypertrophic cardiomyopathy

	Class I	Class IIa	Class IIb
Risk stratification	VTs VF	Fam Hist SCD Syncope Septal thickness >3cm VTns Hypotension at EST	High risk mutations
Primary prevention		ICD	Amiodarone
Secondary prevention	ICD		

VF = ventricular fibrillation; VTs = sustained ventricular tachycardia; Fam Hist SCD = familial history of sudden cardiac death; VTns = non-sustained ventricular tachycardia; Hypotension EST= hypotensive response on exercise stress test.

death is the more devastating component, occurring throughout life, but particularly in young, often asymptomatic patients[45]. A major focus is directed towards the identification of the small subset of HCM patients who are at high risk, so that therapeutic interventions to prevent SCD can be implemented[46,47]. The implant of an ICD for prevention of SCD is most strongly warranted for those patients with prior cardiac arrest (secondary prevention) and prophylactic use of the ICD is also supported in those individuals with two or more risk factors. Decisions regarding prophylactic treatment for primary prevention in HCM patients with a single risk factor may be individualized as the positive predictive accuracy for SCD is relatively low. Based on observational data, the prophylactic use of the ICD would appear at present to be the most appropriate treatment modality for the HCM patient judged to be at high risk[48], although amiodarone treatment may represent a pharmacological alternative to the ICD in some selected patients[49].

The evidence that lead to the proposed recommendations is mainly based on retrospective studies, small prospective studies and on the opinion of experts (Table 4).

Arrhythmogenic right ventricular cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is one of the major causes of SCD in the pre-coronary artery disease age group. Although predictive markers of SCD have not yet been defined in large prospective studies, SCD occurs more frequently in patients with extensive right ventricular changes and in those with left ventricular involvement[50]. Based on non-randomized studies, patients with sustained monomorphic ventricular tachycardia are thought to have a more favourable prognosis when treated medically. In patients with aborted SCD (secondary prevention), ventricular tachycardia unresponsive to antiarrhythmic drug therapy, and in high-risk patients with ventricular tachycardia, ICD therapy is considered appropriate[51]. The evidence that lead to the proposed recommendations is based on small studies or on the opinion of experts (Table 5).

Dilated cardiomyopathy

SCD due to malignant arrhythmias is the single most common cause of death in dilated cardiomyopathy (DCM). Few parameters have been identified as good predictors of SCD that can be reliably used for the risk stratification of DCM patients. Ejection fraction has been repeatedly identified as the most powerful predictor of outcome but its predictive accuracy has not been conclusively defined[52]. Occurrence of syncopal events is the other rather accurate indicator of risk of SCD[53].

Therapeutic strategies aimed at reduction of risk of SCD in patients with documented ventricular arrhythmias include ACE inhibitors, beta-blockers, amiodarone

Table 5 Arrhythmogenic right ventricular cardiomyopathy

	Class I	Class IIa	Class IIb
Risk stratification		VTs / VF RV dilatation RV dysfunction+ PES inducibility	Family History SCD LP+RV dysfunction VT PES Inducibilty
Primary prevention		ICD	Amtiarrhythmic drugs
Secondary prevention	ICD		

VTs = sustained ventricular tachycardia; VF = ventricular fibrillation; RV = right ventricular; LP = Late potentials; PES = programmed electrical stimulation; VT = ventricular tachycardia.

and the implantable cardioverter defibrillator[54]. Few studies have specifically investigated the role of non-antiarrhythmic drugs in DCM patients and it is commonly assumed (but not proven) that pharmacological treatment used in patients with progressive heart failure (with and without ischaemic substrate) is equally effective in patients with DCM. The use of the ICD for secondary prevention is considered appropriate and its prophylactic use in high risk patients for primary prevention of SCD is also recommended. The evidence that leads to the proposed recommendations is based on small studies or on the opinion of experts (Table 6).

Long QT syndrome

Long QT syndrome (LQTS) is associated with high risk of SCD. Risk stratification is mainly based on the history of syncopal events, TdP or cardiac arrest[55,56]. The duration of the corrected QT interval is a weaker predictor of major events. The clinical variants presenting association of the cardiac phenotype with syndactyly or with deafness (Jervell and Lange-Nielsen syndrome) have a more severe prognosis. Genetic defects on the cardiac sodium channel gene (*LQT3*) are also associated with higher risk of SCD[57].

Life-style adjustment is very important in prevention of sudden cardiac death in all categories of patients with LQTS (symptomatic, asymptomatic, and silent carriers of the genetic defect). Life style measures include avoidance of strenuous physical exercise (including competitive sports), avoidance of QT-prolonging agents should also be enforced in all patients[56]. Primary prevention of sudden cardiac death is mainly based on treatment with beta-blockers[58]; the implantable cardioverter defibrillator is recommended in secondary prevention (cardiac arrest survivors) and in patients experiencing cardiac events on full dose beta-blocker therapy.

No randomized trial is available. However, large prospective registries with very long follow-up are available and have provided the basis of most of the recommended strategies for risk stratification and management (Table 7).

Brugada syndrome

The diagnosis of Brugada syndrome (BS) is established in the presence of spontaneous or induced ST segment elevation in leads V_1–V_3 with/without right bundle branch block. Risk stratification is still ill defined and the role of programmed electrical stimulation to identify high-risk patients is debated[59,60]. Cardiac arrest occurs mainly in males in the third–fourth decade of life: up to 80% of victims of cardiac arrest had experienced a syncopal event. It is therefore considered appropriate to include among high-risk patients those with a history of syncope. In survivors of cardiac arrest the implantation of an ICD is recommended, the prophylactic use of the ICD in high risk patients is warranted but this approach

Table 6 Dilated cardiomyopathy

	Class I	Class IIa	Class IIb
Risk stratification	VTs VF	Syncope	↓ EF VTns
Primary prevention	ACE-inhibitors Beta-blockers	ICD Aldosterone receptor blockers	Amiodarone
Secondary prevention	ICD ACE-inhibitors Beta-blockers	Aldosterone receptor blockers	Amiodarone

VF = ventricular fibrillation; VTs = sustained ventricular tachycardia; EF = ejection fraction; VTns = non-sustained ventricular tachycardia.

Table 7 Long QT syndrome

	Class I	Class IIa	Class IIb
Risk stratification	TdP / VF / CA Syncope JLN LQT3	QTc > 600ms CE in infants Post-partum Synd. +AV block TWA Female Gender	Fam Hist SCD ↑ QT dispersion
Primary prevention	Avoid QT prolonging drugs Avoid sport(*) Beta-blockers(*)		LCSD Pacemaker
Secondary prevention	ICD + beta-blockers + Avoid QT prolonging drugs + Avoid sport		

(*) IIa in pt. without syncope or silent gene carriers; TdP = torsades de pointes; VF = ventricular fibrillation; CA = cardiac arrest; JLN = Jervell and Lange Nielsen; CE = cardiac event; TWA = macroscopic T wave alternans; Fam Hist SCD = familial history of sudden cardiac death; Synd = syndactyly; LCSD = left cardiac sympathetic denervation.

is limited by the lack of reliable indicators at risk. Given the limited number of studies on this disease, the evidence used to provide recommendations derives from small multi-centre non-randomized studies with short follow-up and is therefore largely based on the opinion of experts (Table 8).

Table 8 Brugada syndrome

	Class I	Class IIa	Class IIb
Risk stratification	VF - VT	Syncope Family history of SCD	VTs - VF inducibility
Primary prevention	ICD in pt. with syncope/ VT		ICD in asymptomatic pt. inducible by PES
Secondary prevention	ICD		

VF = ventricular fibrillation; VT = ventricular tachycardia; VTs = sustained ventricular tachycardia; PES = programmed electrical stimulation.

Catecholaminergic polymorphic ventricular tachycardia

The natural history of catecholaminergic polymorphic ventricular tachycardia (CPVT) is still poorly defined because large studies are not available. The disease is associated with a high risk of SCD at young age but risk stratification parameters are missing[61]. Inducibility at PES is not considered as an accurate predictor of outcome. History of syncope, previous occurrence of cardiac arrest, rapid and sustained runs of ventricular tachycardia on Holter recording or during exercise stress test are regarded as predictors of risk of major arrhythmic events. Treatment is based on beta-blockers even if recurrence of ventricular arrhythmias has been reported; the implantable defibrillator is indicated in secondary prevention of cardiac arrest while its value in primary prevention is unknown. Since no large prospective studies are available, the recommendations presented are based on the opinion of experts (Table 9).

Aortic stenosis

Among all patients dying of aortic stenosis (AS), death is sudden in about 20%. In the absence of cardiac symptoms, survival is excellent without valve replacement. The prognostic value of different haemodynamic and electrophysiological testing is limited. This information comes only from small observational studies[62,63]. Asymptomatic patients with haemodynamically severe AS should be followed up frequently and carefully and surgical therapy should be undertaken as soon as the patient develops symptoms. In patients presenting with sustained ventricular tachyarrhythmias implantation of ICD should be considered[62]. Recommendations are based on small studies and on opinion of experts (Table 10).

Mitral valve prolapse

MVP is usually benign, its link with SCD has been suggested but never conclusively demonstrated[64]. Accordingly, no data are available to define prophylactic interventions that may reduce the risk of SCD. No single finding is a consistent predictor of cardiac arrest. Most cases of SCD seem to involve patients with previous cardiac arrest or syncope, a family history of SCD at a young age, and mitral valve redundancy. Other clinical, echocardiographic and electrocardiographic markers, including electrophysiological study, do not appear to be valuable in determining a high-risk subgroup[65]. In survivors of cardiac arrest use of an ICD should be considered. These conclusions are based on data from small observational studies and the consensus of experts (Table 11).

Anomalous origin of coronary arteries

SCD occurs most commonly in individuals with anomalous origin of the left main coronary artery from the right or non-coronary sinus of Valsalva. Therefore, special care should be taken to evaluate young patients

Table 9 Catecholaminergic polymorphic ventricular tachycardia

	Class I	Class IIa	Class IIb
Risk stratification	VF	Fam Hist SCD VTns / syncope in paediatric age	Syncope
Primary prevention		Beta-blockers	ICD
Secondary prevention	ICD + beta-blockers	Beta-blockers	

VF = ventricular fibrillation; Fam Hist SCD = familial history of sudden cardiac death; VTns = non-sustained ventricular tachycardia.

Table 10 Aortic stenosis

	Class I	Class IIa	Class IIb
Risk stratification	Syncope Angina	VA and PES inducibility ↓ Exercise tolerance LV dysfunction	Severity of stenosis
Primary prevention	Surgery	Amiodarone	
Secondary prevention	ICD		

VA = ventricular arrhythmias; PES = programmed electrical stimulation; LV = left vertricle.

with chest pain resembling angina. Surgical intervention appears to be the most appropriate treatment modality in patients who are at high risk for SCD[66,67]. Data were derived from a limited number of small observational studies and consensus of experts (Table 12).

Myocardial bridging

Long-term prognosis of isolated myocardial bridges appears to be excellent but in some cases they may cause ventricular tachyarrhythmias and SCD[68]. In

Table 11 Mitral valve prolapse

	Class I	Class IIa	Class IIb
Risk stratification	VTs VF	Fam Hist SCD Redundant / Myxomatous valve leaflets	Long QT Frequent / complex PVCs PES Inducibility MV regurgitation LP
Primary prevention			
Secondary prevention	ICD		

VTs = sustained ventricular tachycardia; VF = ventricular fibrillation; Fam Hist SCD = familial history of sudden cardiac death; PVCs = premature ventricular complexes; PES = programmed electrical stimulation; LP = Late potentials.

Table 12 Anomalous origin of coronary artery

	Class I	Class IIa	Class IIb
Risk stratification	VF	Young pt. with: Angina Postive exercise stress test	
Primary prevention	Surgery		
Secondary prevention	Surgery		

VF = ventricular fibrillation; Positive exercise stress test = ischaemic ST segment on Exercise Stress Test.

symptomatic patients, coronary angiography, Doppler flow analysis and intravascular ultrasound are used to characterize myocardial bridging. Medical treatment with beta-blockers, surgery, angioplasty or stenting may be the therapeutic alternatives.

This information is based on a limited number of small observational studies and a consensus opinion

Table 13 Myocardial bridging

	Class I	Class IIa	Class IIb
Risk stratification	VF Symptomatic VT	Myocardial ischaemia	
Primary prevention	Surgery in ischaemic patients	Beta-blockers	
Secondary prevention	Surgery in ischaemic patients		

VF = ventricular fibrillation; Symptomatic VT = symptomatic ventricular tachycardia.

of experts was the primary source of recommendation[69] (Table 13).

Wolff–Parkinson–White syndrome

In patients with Wolff–Parkinson–White (WPW) syndrome natural history studies have reported SCD rate of 0·15%/year which comes from atrial fibrillation with a rapid ventricular response which degenerates into ventricular fibrillation. SCD survivors tend to be symptomatic, have short (<250 ms) RR intervals during atrial fibrillation and multiple or postero-septally located accessory pathways. An electrophysiological study with induction of atrial fibrillation and determination of RR intervals between pre-exicted QRS complexes has a high sensitivity but limited specificity and positive predictive value[70]. These data are derived from well-designed analyses of non-randomized studies. The non-invasive tests (intermittent pre-excitation, loss of pre-excitation during exercise or with medication by antiarrhythmic agents) are not very helpful in risk stratification. This information is based on relatively small observational studies. Catheter ablation is recommended in patients at risk of SCD, especially those who were resuscitated from ventricular fibrillation or had clinical atrial fibrillation with rapid ventricular responses[70]. Indications for procedure therapy are based on expert consensus and clinical experience (Table 14).

Sinus node and atrio-ventricular conduction disturbances

SCD may be attributable to bradyarrhythmic mechanisms in as many as 15–20% of cases. Importantly, significant numbers of bradyarrhythmic patients with impaired LV function suffer SCD due to the development of ventricular tachyarrhythmias[71].

Intraventricular conduction disturbances have been associated with bradyarrhythmic deaths but when the conduction defect is caused by irreversible structural abnormalities, SCD may be due to ventricular tachyarrhythmias. Intraventricular conduction disturbances have been associated with bradyarrhythmic deaths, while SCD could also be caused by ventricular tachyarrhythmias in those patients with conduction defects[72]. Cardiac pacing undoubtedly improves the symptoms of bradyarrhythmic patients and may limit mortality[73,74] (Table 15).

Athletic heart

Sudden and unexpected death in young trained athletes is predominantly due to underlying and usually unsuspected congenital cardiovascular disease. The most important of these appear to be hypertrophic cardiomyopathy, anomalous origin of coronary artery and arrhythmogenic right ventricular cardiomyopathy. Screening strategies for asymptomatic normal populations of trained atheletes can detect certain abnormalities, but the power of identification is enhanced considerably by the incorporation of non-invasive testing (i.e. 12-lead ECG or echocardiography)[75,76]. Removal of atheletes with cardiovascular disease from competition and de-training may decrease risk. Consensus panel guidelines and criteria governing such clinical decision-making are available. Due to the nature of the subject, much of the assembled data and conclusions have necessarily been based on uncontrolled, retrospective and inferential observations.

Table 14 Wolff–Parkinson–White

	Class I	Class IIa	Class IIb
Risk stratification		< 250ms AF CL < 270ms ant. RP of AP Multiple APs	Loss of pre-excitation with ajmaline
Primary prevention	Ablation in AF + fast conduction through the AP	Ablation in asympt. pt.w. - fam hist of SCD - athletes	Amiodarone 1A, 1C, AA drugs
Secondary prevention	Ablation		

AF CL = cycle length of atrial fibrillation; ant. RP = anterograde refractory period; AP = accessory pathway; SCD = sudden cardiac death; AA = antiarrhythmic.

Table 15 Conduction system abnormalities

	Class I	Class IIa	Class IIb
Acquired AV block		III° AVB II° AVB type II Syncope Coexistant HD/HF	
Congenital III° AV block	Syncope Long QT interval Congenital HD		
Chronic bi-fascicular or tri-fascicular block	Coexistent HD/HF	Syncope HV ⩾100ms or inf.H block PES inducibility	

AVB = atrio-ventricular block; HD = heart disease; HF = heart failure; Inf. H = infra-Hisian; PES = programmed electrical stimulation.

Drug-induced torsades de pointes

The steps to be recommended for increasing the awareness of pro-arrhythmic risks associated with established and new drugs include[77]:

Detailed list of all drugs associated with QT-prolongation;
For new drugs, data on block of K^+ channels (HERG, etc.) are mandatory;

Avoidance of co-administration of drugs prolonging the QT-interval;
Avoidance of drugs that interfere with metabolism and excretion;
Avoidance of drugs that produce TdP-promoting conditions (hypokalaemia, bradycardia).

The absolute incidence of cardiotoxicity of any drug must be judged in relation to the severity of the treated disease: a high risk may be perfectly acceptable when treating a life threatening condition whereas even a very low incidence reported for non-sedating antihistamines is not acceptable as these drugs are widely prescribed for minor complaints.

Out-of-hospital resuscitation

Survival after cardiac arrest (CA) varies from less than 5% to 60% according to the characteristics of the cardiac arrest event (e.g. cardiac aetiology or not, witnessed or not, VF or not). The results of cardiopulmonary resuscitation (CPR) are influenced not only by the resuscitation efforts but also by the conditions before initiation of CPR. Outcome from cardiac arrest is a complex interplay of so-called 'fate factors' (e.g. age, underlying disease) and 'programme factors' (e.g. time interval to basic life support and to defibrillation). It is now generally accepted that the time to electrical defibrillation is the single most important determinant of survival after cardiac arrest.

In areas where early defibrillation by ambulance personnel is implemented, more patients are found in VF at the time of the intervention resulting in a higher hospital discharge rate of 25–28%[78].

Cardiac arrest usually happens at home (about 2/3), in male patients aged >50 years of age (about 3/4) and during daytime (about 3/4 between 8–18 h). In most reports on out-of-hospital cardiac arrest presenting with VF, cardiac arrest has been witnessed in 2/3 in cases. People are more likely to survive out-of-hospital cardiac arrest when activation of the Emergency Medical Service (EMS)-system, basic cardiopulmonary resuscitation (CPR), defibrillation and advanced care occur as rapidly as possible. The concept of 'the chain of survival'[79] describes the interventions that are needed for optimal survival.

- The first link in the chain of survival, 'early access' is essential to bring trained people and appropriate equipment, i.e. the defibrillator, quickly to the patient. This includes recognition of the collapse, decision to call, calling and dispatch, and can be strengthened by public education and availability of an efficient emergency communication system.
- The importance of the second link, 'early CPR' has been shown abundantly. Bystander CPR is able to maintain the heart some 10–12 min longer in VF. Basic CPR is efficient to sustain life until early arrival of trained and equipped people, and is therefore a bridge to first defibrillation.
- The most crucial link is 'early defibrillation'. Initially, out-of-hospital defibrillation was only performed by medical and paramedical personnel, but recently the automated external defibrillator (AED) has allowed the reliable use by the first-line trained ambulance personnel and laymen. First tier ambulances arrive many vital minutes before arrival of the second tier. Primary rescue teams, such as police, security personnel and fire fighters are present at the scene several minutes before the first tier ambulance of the EMS-system. In remote areas (airplanes, cruise ships, trains) members of the crew are the only ones who can administer a defibrillatory shock within seconds or minutes. To shorten the time to defibrillation, rescuers in the community other than physicians or paramedics should have access to defibrillation.
- Early defibrillation is of high value as long as the other links of the 'chain of survival' do not fail. In systems, where access time is excessively long, only disappointment can be expected.
- The fourth link 'early advanced life support' implies early intervention of a well-trained and well-equipped team, working with specially equipped ambulances or rapid intervention vehicles.

Defibrillation of the heart is the only effective treatment of VF and pulseless VT. The time between the onset of VF and the first defibrillating shock is the most important variable of the efficacy of this treatment. The objective of the management of out-of-hospital cardiac arrest is to provide electrical defibrillation of the heart as soon as possible after collapse.

The introduction of the automated external defibrillator (AED) has allowed less trained emergency medical technicians to deliver electric shocks in cases of out-of-hospital VF or VT, often many minutes before the arrival of the medical intervention team[80,81]. This strategy is also known as 'first responder defibrillation' (Table 16).

Conclusion

Although SCD remains a serious public health hazard, major developments in risk stratification and therapy have now made it possible to identify many of those at risk and to provide effective prophylactic treatment. However, the implementation of novel and effective risk stratification and of therapies known to reduce the risk of SCD has been slow and inconsistent. The SCD Task Force has attempted to draw together in one document the substantial evidence-base both for risk stratification and for prophylactic treatment against SCD. The widespread introduction of these recommendations into clinical practice should reduce, but it will not eliminate SCD.

It is recognized that most of the success in defining risk and proving therapy has so far been achieved in patient groups with considerable pre-existing cardiac disease. Much more work is needed and expected in larger populations with less or no apparent heart

Table 16 Automatic external defribillators

	Class I	Class IIa	Class IIb
Out-of-hospital defibrillation	Shock delivery within 5 min of EMS call receipt	Use by health-care providers with a duty to perform CPR	Use of AED in children >8y or >25kg
In-hospital defibrillation	Shock delivery within 3 min of collapse	Availability of equipment and trained responders throughout hospital	
Public access defibrillation		Use by security personnel (police, airline, firefighters, ...)	Use by family members of high risk individuals

disease. Effective identification and treatment of these subjects will then lead to a very substantial reduction in SCD in the general population. Epidemiological and clinical investigations in this arena are already underway and will provide much further information on which to base comprehensive strategies for the elimination of SCD.

The most effective long-term treatment that is currently available for SCD is the implantable cardioverter defibrillator. This therapy is generally more effective than drug-based treatments but has not been uniformly adopted, probably because of differing medical priorities in communities that have limited resources. This document emphasizes the outstanding success of ICD therapy and provides cogent information and argument that supports investment in this treatment. It is recognized that ICD therapy cannot be proved against every other treatment in every condition. Obviously some sensible extrapolation is justifiable.

The Task Force expects further development in the therapy for the prevention and emergent treatment of SCD. Improvements in automatic external defibrillators, implantable cardioverter defibrillators and 'anti-arrhythmic' drugs will certainly lead to even more effective treatment of those at risk of SCD. In due course, it will clearly be necessary to reconvene the Task Force on SCD to reconsider the inevitably more comprehensive evidence base that will accumulate in the next few years.

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