Guidelines for Risks and Prevention of Sudden Cardiac Death (JCS 2010)

– Digest Version –

JCS Joint Working Group

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Introduction to the Revised Guidelines

The best way to prevent sudden death is predicting the occurrence of sudden death and providing appropriate preventive measures. Since many cases of sudden death are arrhythmic death, the present guidelines describe disease conditions and typical clinical findings that may cause arrhythmic death and how to prevent sudden deaths in patients with such predicted findings. Since ventricular tachyarrhythmias (VTA) including ventricular fibrillation (VF) play an important role in the development of arrhythmic death, and the benefits of implantable cardioverter defibrillator (ICD) in preventing sudden death due to tachycardia have been demonstrated, the present guidelines mainly discuss the use of ICD in the treatment of potentially fatal tachycardia. Severe bradycardia and asystole, which also may cause sudden death, are described in the present guidelines as needed in relation to pathological conditions and diseases known to lead to bradyarrhythmia and asystole. The present guidelines is partly revised and reflect the newest findings to be included to the guidelines for the non-pharmacological and pharmacological treatment of arrhythmia of which the Japanese Circulation Society (JCS) are currently revising.

The present guidelines are written mainly for the use of cardiologists since pathological conditions and diseases that may cause arrhythmic death must be carefully assessed by expert cardiologists, and since ICD therapy, the most impor-

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There are laboratory examinations that are believed to be useful in identifying those patients at a high risk for sudden death, especially arrhythmic death. The majority of these examinations assess electrical activity of the heart and the activity of the autonomic nervous system. Clinical symptoms of cardiac dysfunction and heart failure are also important findings to identify patients at a high risk for sudden death. Although patients at a risk for sudden death can be identified (predicted) with these examinations, only a few of these examinations have been demonstrated to be useful in terms of primary prevention of sudden death in clinical research. The current usefulness of clinical findings and laboratory examinations in terms of predicting sudden death is listed by the class of recommendation, although there is some controversy as to the validity of methods used in the evaluation of predictive factors for arrhythmia and sudden death.

Classification of Recommendations

Class I: Conditions for which there is general agreement that a given clinical finding/laboratory examination is useful in predicting sudden death

Class IIa: Conditions for which there is some divergence of opinion, but weight of opinion is in favor of usefulness

Class IIb: Conditions for which there is some divergence of opinion, but weight of opinion is against usefulness

Class III: Conditions for which there is general agreement that the clinical finding/laboratory examination is not useful in predicting sudden death

Class III clinical findings/laboratory examinations are not described in this document as a rule.

1. Clinical Findings

Class I
- A decrease in ejection fraction ($\leq 30$ to $35\%$) in patients with ischemic and non-ischemic heart failure$^{1-3}$

2. Electrocardiogram

Class I
- Evaluation of ventricular arrhythmia with standard 12-lead electrocardiogram (ECG)$^7$

Class IIb
- Evaluation of patients with heart failure, cardiac hypertrophy (hypertrophic cardiomyopathy [HCM], hypertension, and aortic stenosis), and arrhythmogenic right ventricular cardiomyopathy (ARVC) based on QT dispersion (QTD)$^{4-6}$
- Evaluation of patients with syncope associated with long QT syndrome (LQTS) or HCM according to the magnitude of transmural dispersion of repolarization (TDR)$^7$

3. Heart Rate Variability

Class IIa
- Prediction of sudden death in patients after myocardial infarction (MI)$^9$

Class IIb
- Prediction of sudden death in patients with cardiomyopathy$^{9,10}$

4. Heart Rate Turbulence

Class IIb
- Prediction of sudden death in patients after MI and patients with heart failure$^{11}$

5. Baroreflex Sensitivity

Class I
- Prediction of sudden death in patients with cardiac dysfunction after MI$^{12}$

6. T-Wave Alternans

Class IIa
- Prediction of cardiac sudden death in patients after MI and in patients who have ischemic cardiomyopathy with cardiac dysfunction$^{13}$

Class IIb
- Prediction of cardiac sudden death in patients with (non-ischemic) dilated cardiomyopathy (DCM) or heart failure$^{14,15}$

7. Late Potential

Class IIb
- Prediction of sudden death in patients after MI$^{16}$

8. Cardiac Electrophysiological Study

Class I
- Patients after MI who have palpitation, near syncope or syncope suspected to be caused by VT
- Evaluation of patients following catheter ablation for the treatment of ventricular tachycardia (VT)
- Patients with syncope of unknown cause who have cardiac dysfunction, organic heart disease, a family history of sudden death and/or abnormal ECG findings

Class IIa
- Patients after MI who have nonsustained ventricular
tachycardia (NSVT) and a left ventricular ejection fraction (LVEF) of ≤40%.\textsuperscript{17}

- Patients with syncope that are suspected to be caused by bradycardia or tachyarrhythmia, but not confirmed with non-invasive examinations

\section*{9. Exercise Stress Test}

Class I
- Exercise induced VTA in patient suspected to have coronary disease
- Patients who have or are suspected to have exercise induced ventricular arrhythmia
- Abnormal blood pressure reactions in patients with HCM\textsuperscript{18}

Class IIa
- Evaluation of the efficacy of drug treatment or catheter ablation in patients with exercise induced ventricular arrhythmia

\section*{10. Genetic Tests}

Class IIa
- Cardiac ion channel gene mutations (LQTS mutations, especially LQT1, LQT2 and LQT3 mutations) and site of mutation\textsuperscript{19,20}

Class IIb
- Abnormalities of the genes of ryanodine receptor (RyR2) or calsequestrin (CASQ) in catecholamine-induced polymorphic ventricular tachycardia (CPVT)\textsuperscript{21}

\section*{II Prevention of Sudden Death}

This section lists major pathological conditions and diseases that are known to precede sudden death, and describes relationship between these conditions and arrhythmic death. This section also describes typical signs/symptoms and laboratory findings of the pathological conditions/diseases believed to cause sudden death as possible predictive factors and lists measures to prevent the patient at a high risk for sudden cardiac death as either primary or secondary prevention of arrhythmic death.

Types and indications of preventive measures are listed by the class of recommendation.

\begin{table}
\centering
\begin{tabular}{|l|c|c|c|}
\hline
\textbf{Purpose of treatment/findings} & \textbf{Class I} & \textbf{Class IIa} & \textbf{Class IIb} \\
\hline
Cardiac arrest (VF) & ICD & Amiodarone* or sotalol* & \\
\hline
Patients with SVT who have syncope during tachycardia, or patients with LVEF <40% with a low blood pressure (<80 mmHg) & ICD & Amiodarone* or sotalol* & \\
\hline
Patients with hemodynamically stable SVT with underlying heart disease in whom drugs are not effective or contraindicated & ICD & Amiodarone* or sotalol* & \\
\hline
Patients with underlying heart disease and in whom SVT is no longer induced after catheter ablation & ICD & Amiodarone* or sotalol* & \\
\hline
Patients with SVT associated with underlying heart disease and a LVEF of ≥40% who are responding to drug treatment & ICD & Amiodarone* or sotalol* & ICD+effective drugs \\
\hline
\end{tabular}
\caption{Prevention of Sudden Death in Patients With Sustained Ventricular Tachycardia or Ventricular Fibrillation}
\end{table}

*Use to control VT/VF episodes in patients contraindicated for ICD therapy or patients already undergoing ICD therapy.

ICD, implantable cardioverter defibrillator; LVEF, left ventricular ejection fraction; SVT, sustained ventricular tachycardia; VF, ventricular fibrillation; VT, ventricular tachycardia.

Class III measures are not described in this guideline.

For example, in this guideline, ICD therapy and antiarrhythmic drugs are listed as Class I measures, but this does not mean that ICD therapy and antiarrhythmic drugs are similarly effective in the prevention of sudden death. This means that there is evidence or consensus that Class I measures decrease the incidence of arrhythmic death as compared with control groups. Limitation of exercise is a Class I measure to prevent exercise induced arrhythmia. Measures listed in the same class do not necessarily target the same goal of treatment. Physicians should be aware of this to select appropriate measures for individual patients.

\section*{1. Arrhythmias}

1. Cardiac Arrest (Resuscitated Cases)

Sustained ventricular tachycardia (SVT) and VF are the most frequent causes of cardiac arrest. Cardiac arrest frequently and the risk of sudden death is extremely high. ICD therapy is the most effective secondary preventive measure for sudden death.\textsuperscript{22-24}
### 2. Sustained Ventricular Tachycardia

In Japan, the underlying disease of SVT is old MI in about 30%, and non-ischemic heart disease in the majority of patients.²²,²³ SVT is treated mainly with procaineamide and lidocaine.²⁶,²⁷ Amiodarone and nifekalant are recommended for patients with refractory SVT.²⁷ Patients with a history of high-rate, hemodynamic deterioration of SVT require measures to prevent recurrence, and should be considered for the indication for ICD therapy first. Since patients with hemodynamically stable SVT associated with organic heart disease may often experience high-rate episodes or new onset of VT with different waveforms, their prognoses are not good and ICD therapy is thus recommended for them.²⁸ Patients following successful catheter ablation for the treatment of SVT are often indicated for ICD therapy since their long-term prognosis is unclear and recurrent SVT is occasionally observed (Table 1).

### 3. Premature Ventricular Contraction and Nonsustained Ventricular Tachycardia

When no heart diseases are present, the prognosis of individuals with PVC and NSVT is good, and these arrhythmias do not represent risk factors for sudden death. Whether primary prevention of sudden death is indicated for patients with PVC or NSVT depends on the type of underlying diseases, which are described in the following sections.

### 4. Bradycardia

Bradycardia accounts for about 10% of all deaths due to arrhythmia.²⁹,³⁰ Bradycardia gradually leading to asystole has been observed in patients with arrhythmical death.³⁰ The most common causes of bradycardia leading to arrhythmic death are sick sinus syndrome and atrioventricular block, which require treatment with a pacemaker.³¹–³³ The pacemaker treatment of these conditions is described in the Guidelines for Non-Pharmacological Therapy of Cardiac Arrhythmias reported by the JCS.²²

### 2. Cardiogenic Syncope (Table 2)

Cardiogenic syncope is defined as syncope other than those due to non-cardiac causes such as orthostatic syncope and syncopes due to vasovagal reflex and seizures.³⁴,³⁵ Cardiogenic syncope due to arrhythmia may be identified with the following conditions:

<table>
<thead>
<tr>
<th>Findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients in whom unstable SVT or VF is induced, and drug efficacy cannot be assessed</td>
<td>ICD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patients with underlying heart disease in whom hemodynamically stable SVT is induced and drug treatment and catheter ablation are ineffective</td>
<td></td>
<td></td>
<td>ICD</td>
</tr>
<tr>
<td>Patients with underlying heart disease and cardiac dysfunction in whom hemodynamically unstable SVT or VF is induced and drug efficacy has not been assessed</td>
<td></td>
<td></td>
<td>ICD</td>
</tr>
<tr>
<td>Patients with dilated or hypertrophic cardiomyopathy in whom hemodynamically unstable SVT or VF is not induced</td>
<td></td>
<td></td>
<td>ICD</td>
</tr>
<tr>
<td>Patients with a history of asystole associated with sick sinus syndrome or atrioventricular block</td>
<td>Pacemaker</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Note) Refer to the Guidelines for Non-Pharmacological Therapy of Cardiac Arrhythmias²² reported by the Japanese Circulation Society for the management of patients with bradycardia. The management of primary arrhythmia is described in a later section.

ICD, implantable cardioverter defibrillator; SVT, sustained ventricular tachycardia; VF, ventricular fibrillation.

### 3. Heart Failure

The mortality of patients with heart failure is higher in patients with more severe disease by New York Heart Association (NYHA) classification.³⁶ In recent large-scale clinical studies of patients with heart failure, sudden cardiac death developed in 9 to 22% of the patients.³⁷–⁴¹ The percentage of sudden death cases among all-cause death cases is higher in patients with milder heart failure (NYHA Class I or II) than in severer patients (NYHA Class III or IV). The most likely causes of sudden cardiac death among patients with heart failure are SVT and VF.⁴²

About half of patients with out-of-hospital cardiac arrest (resuscitated) or patients with SVT (in Japan) are considered to have cardiac dysfunction.⁴³–⁴⁷ Patients with proven VF or SVT should undergo secondary preventive treatment mainly using an ICD.

In patients after MI, cardiac dysfunction is an independent predictive factor of poor prognosis. ICD therapy has been demonstrated to be useful as a primary preventive measure for sudden death in patients with cardiac dysfunction after MI.¹ ICD therapy has also been demonstrated to be useful in the primary prevention of sudden death in patients with DCM.²³ Although a number of studies have reported that amiodarone decreases the incidence of sudden death, some studies have denied such effect. β-blockers improve the prognosis and decrease the incidence of sudden death in patients with chronic heart failure.⁴⁵,⁴⁸ Angiotensin converting enzyme (ACE) in-
The one-year mortality of patients after MI was as high as 2%. Post-Myocardial Infarction with AMI. Elevation reported by the JCS describe how to treat VT/VF. Acute Coronary Syndrome without Persistent ST Segment elevation. Emergency medical service and treatment with an automated ECG monitoring and electrical defibrillation. Defibrillation by cardiogenic shock and pump failure.

The disease. Secondary VF may develop in association with patients with AMI, especially those in a very early phase of the condition leading to fatal arrhythmia. VF often develops in patients with heart failure. Catheter ablation may be effective in controlling monomorphic VT, when VT is no longer induced by programmed electrical stimulation. However, since many post-MI patients have polymorphic VT or cannot undergo electrophysiological mapping, patients often have to receive other treatment methods in addition to catheter ablation.

In Europe and the United States, supplementation of polyunsaturated fatty acids is recommended to prevent sudden death. Since the number of stenotic lesions, the results of reperfusion therapy during AMI, and the potency of the artery responsible for MI after treatment affect the incidence of arrhythmical accidents, it is important to improve myocardial ischemia including asymptomatic myocardial ischemia as much as possible.

3. Variant Angina

Patients with variant angina may experience angina attacks leading to fatal ventricular arrhythmia. Patients with variant angina often die from sudden death, which is believed to be associated with multi-vessel coronary spasm. The prognosis of patients with variant angina associated with VT is poor. Patients with a history of coronary spastic attacks should be treated with ICD therapy for secondary prevention.

### Table 3. Prevention of Sudden Death in Patients After Myocardial Infarction

<table>
<thead>
<tr>
<th>Clinical findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Secondary prevention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Patients with a history of SVT, VF or resuscitated cardiac arrest</td>
<td>ICD</td>
<td>Amiodarone, sotalol, catheter ablation</td>
<td></td>
</tr>
<tr>
<td>Primary prevention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Patients with syncope (+), NSVT (+) and LVEF &lt;40% in whom SVT or VF is induced and drug treatment is ineffective</td>
<td>ICD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Patients with syncope (-), NSVT (+) and LVEF &lt;35% in whom SVT or VF is induced and drug treatment is ineffective or drug efficacy has not been assessed</td>
<td>ICD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Patients with cardiac dysfunction (LVEF &lt;35%)</td>
<td>β-blockers, ACE inhibitors, anti-aldosterone drugs</td>
<td>ICD</td>
<td></td>
</tr>
</tbody>
</table>

Note: Drug treatment is generally believed to improve the prognosis, but no consensus has been achieved. ACE, angiotensin converting enzyme; ICD, implantable cardioverter defibrillator; LVEF, left ventricular ejection fraction; NSVT, nonsustained ventricular tachycardia; SVT, sustained ventricular tachycardia; VF, ventricular fibrillation.

### Table 4. Prevention of Sudden Death in Patients With Variant Angina

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary/secondary prevention</td>
<td>Calcium channel blockers</td>
<td>Nitrates, nicorandil</td>
<td>ICD</td>
</tr>
<tr>
<td>• Presence of SVT or VF during anginal attack</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
treated with calcium channel blockers to prevent recurrence of attacks. ICD therapy may be considered for patients who still have coronary spastic attacks after treatment with sufficient doses of calcium channel blockers and have severe ventricular arrhythmia, but it is unclear whether ICD therapy may improve the vital prognosis of these patients (Table 4).

### 5. Hypertrophic Cardiomyopathy

The annual mortality of patients with HCM has been reported as 1 to 2%. Although HCM does not necessarily represent a poor prognosis, more than half of death cases among patients with HCM are sudden death. HCM is an important cause of sudden death especially in young patients. Patients resuscitated from cardiac arrest and patients with SVT are at high risk for sudden death. HCM is an important cause of arrhythmia, but it is unclear whether ICD therapy may improve the prognosis of these patients (Table 4).

| Table 5. Prevention of Sudden Death in Patients With Hypertrophic Cardiomyopathy |
|------------------|------------------|------------------|
| Purpose of treatment/findings | Class I | Class IIa | Class IIb |
| **Secondary prevention** | | | |
| • Patients with cardiac arrest, SVT or VF | ICD | Amiodarone | |
| **Primary prevention** | | | |
| • Patients with more than one risk factor* | Limitation of exercise | ICD, amiodarone | |
| • Patients with one risk factor* | Limitation of exercise | Amiodarone | ICD |

*According to the DEFINITE study.

(1) Recurrent syncopal attacks (especially syncopes occurring in children during exercise)\(^{51,63,64}\)

(2) A first-degree family history of sudden death or multiple family history of sudden death\(^{61,64,65}\)

(3) Complication with NSVT\(^{66-68}\)

(4) Significant left ventricular wall thickening (maximum thickness ≥30 mm)\(^{69}\)

(5) A decrease or insufficient increase (≤20 mmHg) in systolic blood pressure during exercise stress test\(^{18,59,70}\)

The indication for primary prevention of sudden death should be determined according to the presence of above risk factors (Table 5). In the Guidelines for Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy reported by the JCS,\(^{71}\) primary prevention is indicated according to whether VT/VF is induced during EPS.

| Table 6. Prevention of Sudden Death in Patients With Dilated Cardiomyopathy |
|------------------|------------------|------------------|
| Purpose of treatment/findings | Class I | Class IIa | Class IIb |
| **Secondary prevention** | | | |
| • Patients with SVT or VF | ICD | Aldosterone antagonists | Amiodarone |
| • Patients with syncope (+), LVEF ≤40% in whom VT/VF is induced, and drug treatment is ineffective | ICD, aldosterone antagonists | Amiodarone |
| • Patients with syncope (-), LVEF ≤40% in whom VT/VF is induced, and drug treatment is ineffective | ICD, amiodarone |
| • Patients with syncope (+), LVEF ≤40% in whom VT/VF is induced, and drug efficacy has not been assessed | ICD, amiodarone |
| • Patients with LVEF ≤36% and NSVT or frequent PVC (≥10 times/hr)* | ICD, amiodarone |
| • Patients with LVEF ≤30% and NSVT | Amiodarone |

*According to the DEFINITE study.

ACE, angiotensin converting enzyme; DEFINITE, DEFibrillators In Non-Ischemic Cardiomyopathy Treatment Evaluation; ICD, implantable cardioverter defibrillator; NSVT, nonsustained ventricular tachycardia; SVT, sustained ventricular tachycardia; PVC, premature ventricular contraction; VT, ventricular tachycardia; VF, ventricular fibrillation.

### 6. Other Heart Diseases Associated With Cardiac Hypertrophy

Left ventricular hypertrophy is observed in patients with hypertension, those with aortic stenosis and those with athlete’s heart. The incidences of cardiovascular accidents such as sudden death, arrhythmia, heart failure, MI and stroke are higher in individuals with left ventricular hypertrophy than those without it.\(^ {72-74}\) It is unclear that left ventricular hypertrophy per se increases the risk for sudden death or not, while the incidence of sudden death is considered to be high among patients with left ventricular hypertrophy complicated with arrhythmia,\(^ {75-78}\) coronary artery disease,\(^ {79,80}\) or heart failure.\(^ {81,82}\)

Secondary prevention of sudden death with ICD therapy is indicated for patients with left ventricular hypertrophy associated with SVT or VF, but the results of primary prevention have not been reported.
7. Dilated Cardiomyopathy

Major causes of death of patients with DCM are heart failure (about 50%) and sudden death (30 to 40%). Many cases of sudden death are caused by VT or VF, but severe bradycardia may lead to sudden death. Patients with a history of SVT or VF are indicated for secondary prevention using an ICD. Some patients with monomorphic SVT may be successfully treated with catheter ablation.

A multivariate analysis of the prognosis of patients with DCM has revealed that a history of SVT or VF and LVEF are risk factors for sudden death in this patient population. The incidence of arrhythmic accidents is high among patients with NSVT and a LVEF of <30%. In the analysis of DCM patients implanted with an ICD, the incidence of arrhythmia is high among those with low LVEF (<30%). It has been demonstrated that ICD therapy is effective as primary prevention of sudden death in patients with a LVEF of ≤36% associated with either NSVT or frequent PVC.

All-cause mortality and incidence of sudden death are high among patients with DCM complicated with left bundle branch block. Currently, the types and indications of primary prevention of sudden death among patients with DCM are determined according to the symptoms, cardiac function, presence/absence of SVT and VF induced during EPS, and so on (Table 6).

8. Arrhythmogenic Right Ventricular Cardiomyopathy

ARVC is a disease with unknown cause that is characterized with fatty infiltration in the right ventricle (often extending to the left ventricle) and VT originated from the right ventricle. Patients with ARVC start to show right heart failure in their 40 to 50s. In Europe and the United States, ARVC should be suspected in cases of sudden death or cardiac arrest in patients under 35 years old. In Japan, ARVC is the underlying disease of SVT in about 10% of patients.

The incidence of arrhythmic accidents is high among patients with extensive abnormal right ventricular wall motion, those with VT induced during EPS, and those affecting the right and left ventricles. Monomorphic VT is commonly induced and catheter ablation is often successful, but long-term outcome of catheter ablation remains unknown. ICD therapy is the first-choice secondary preventive method. During a 3-year follow-up of patients undergoing ICD therapy, ICD was activated in about half of the patients. In the primary prevention, the indication for ICD therapy should be determined according to the presence/absence of SVT induced during EPS with lesions of larger size and a family history of sudden death (Table 7).

9. Other Myocardial Disorders

SVT and VF may develop as a complication of cardiac sarcoidosis, muscular dystrophy, chronic lung disease, progressive systemic sclerosis or diabetes mellitus, and so on. It is difficult to predict occurrence of sudden death, SVT or VF in these patients. When the presence of these conditions is confirmed or strongly suspected, the use of an ICD should be considered.

10. Brugada Syndrome

Brugada syndrome is a disorder characterized by right bundle branch block pattern with ST segment elevation in V1 to V3 leads on ECG, and may cause sudden death mainly due to VF at night. Brugada syndrome is considered to be consistent with sudden unexpected nocturnal death syndrome in Southeast Asia and “Pokkuri disease” in Japan. History of VF or syncope represents a significant risk factor for sudden death in patient with Brugada syndrome, and ICD therapy is indicated for such patients. It has been reported that quinidine is effective in preventing arrhythmic attacks and activations of the ICD in patients with Brugada syndrome. The need of primary prevention including ICD therapy is usually determined in each institution on the basis of ECG findings and presence/absence of VF induced during EPS, and...
Table 9. Prevention of Sudden Death in Patients With Congenital Long QT Syndrome

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Secondary prevention • Patients with VF or cardiac arrest</td>
<td>ICD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary prevention • Patients meeting ≥2 of the 3 criteria of (1) a history of TdP or syncope,</td>
<td>ICD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(2) a family history of sudden death, (3) not responding to β-blockers</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Patients responding to β-blockers with a history of TdP or syncope or</td>
<td>β-blockers</td>
<td></td>
<td></td>
</tr>
<tr>
<td>a family history of sudden death</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ICD, implantable cardioverter defibrillator; TdP, torsades de pointes; VF, ventricular fibrillation.

Table 10. Indications of Drug Treatment in Patients With Congenital Long QT Syndrome

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Patients with a history of syncope (especially those with LQT1 or LQT2 mutation)</td>
<td>β-blockers</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Asymptomatic patients with QT prolongation, congenital deafness, being</td>
<td>β-blockers</td>
<td></td>
<td></td>
</tr>
<tr>
<td>neonates/infants, having a family history of sudden death of siblings, and</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>anxiety for sudden death or strong desire for treatment by patients or their family</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Asymptomatic patients without congenital deafness, or a family history of</td>
<td>Mexiletine</td>
<td>β-blockers</td>
<td></td>
</tr>
<tr>
<td>sudden death of siblings, and who either have LQT3 mutation with a history of syncope or</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>have LQTS not responding to monotherapy with β-blockers</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Note) Exercise and drugs prolonging QT interval (antiarrhythmic drugs, tricyclic antidepressants and antihistamines) are contraindicated in all patients.

Table 11. Prevention of Sudden Death in Patients With WPW Syndrome

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Secondary prevention • Patients with VF, cardiac arrest or syncope</td>
<td>Catheter ablation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary prevention • Patients with a R-R interval during atrial fibrillation of ≤250msec</td>
<td>Catheter ablation</td>
<td>Amiodarone, Class Ia and Ic drugs</td>
<td></td>
</tr>
<tr>
<td>• Patients with a refractory period of antegrade conduction over the accessory pathway of ≤270msec, patients with multiple accessory pathways, patients with a family history of sudden death or athletes</td>
<td>Catheter ablation</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

VF, ventricular fibrillation; WPW, Wolff-Parkinson-White.

11. Congenital Long QT Syndrome

The congenital LQTS is a group of genetic arrhythmic disorders that cause syncope and sudden death due to ventricular arrhythmia characterized by QT prolongation and torsades de pointes (TdP), and include Romano-Ward syndrome and Jervell and Lange-Nielsen syndrome. Since cardiac arrest is the first manifestation of the congenital LQTS in 10% of patients, it is quite important to predict and prevent cardiac arrest. To date, 12 different genetic mutations (referred to as LQT1 through LQT12) have been reported to be related to congenital LQTS. The prevalence is highest for LQT1, which is followed by LQT2 and LQT3. Patients with LQT1, LQT2 and LQT3 account for most of the patients with congenital LQTS.

In male patients, arrhythmic accidents often occur before adolescence, and the incidence of arrhythmic accidents is higher in male than female patients. During adolescence and thereafter, the incidence of cardiac accidents is higher in female than male patients. Cardiac accidents during childhood are more common in boys than girls with LQT1 mutation, but there are no sex differences among patients with LQT2 and LQT3 mutations. Since cardiac event (eg, syncope, cardiac arrest or sudden death) by the age of 40 years was 63%, 46% and 18% in patients with LQT1 (n=112), LQT2 (n=72) and LQT3 (n=62) mutations, respectively, and the mortality was 4% in patients with LQT1 and LQT2 mutations and as high as 20% in patients with LQT3 mutation. Congenital LQTS patients with a history of cardiac arrest are indicated for ICD therapy combined with β-blockers and limitation of exercise (Tables 9,10).

The risk of occurrence of TdP is high among patients with LQT2 mutation and a QTc of >500 msec and male patients with LQT3 mutation. Patients with recurrent syncope despite treatment with β-blockers, and patients with a family history of sudden death are at a high risk for cardiac events. TdP often develops during exercise, and especially swimming, among patients with LQT1 mutation; in association with mental stress, sudden auditory stimuli or immediately after child-
birth among those with LQT2 mutation; and during sleep among those with LQT3 mutation. In patients with LQT2 mutation, the risk of occurrence of TdP is high among patients with mutations in the pore region of the human-ether-related gene (HERG gene). Primary prevention consists of limitation of exercise and treatment with β-blockers. Mexitelina is effective for patients with LQT3 mutation, and pacing is used for patients with bradycardia.

### 12. Wolff-Parkinson-White Syndrome

It has been reported that symptomatic Wolff-Parkinson-White (WPW) syndrome is observed in 1 to 2/1,000 individuals, the incidence of sudden death is 0.02 to 0.15%/year, and the incidence of VF is about 3 to 4 fold of the incidence of sudden death. Even in patients with asymptomatic WPW syndrome, VF may develop in rare cases during the first episode of atrial fibrillation. WPW syndrome may usually be completely treated with catheter ablation and the risk of sudden death will disappear after treatment (Table 11).

The incidence of VF tends to be higher in young male patients. The risk of VF is high among patients with a history of atrial fibrillation or reciprocating tachycardia. It has been reported that about half of patients experience VF during the first episode of atrial fibrillation. It is believed that VF develops in 20 to 40% of patients with multiple Kent bundles. The presence of Ebstein’s anomaly is considered a risk factor for sudden death in patients with WPW syndrome.

On the other hand, the risk of sudden death is considered low in patients with intermittent WPW syndrome and patients in whom delta waves disappear after intravenous administration of ajmaline or procainamide. Patients at a high risk of transition from atrial fibrillation to VF may be identified on the basis of the refractory period of antegrade conduction over the accessory pathway or the shortest R-R interval during atrial fibrillation induced during EPS.

### 13. Catecholamine-Induced Polymorphic Ventricular Tachycardia

The first episodes of CPVT often manifest during childhood after infancy as exercise induced polymorphic VT or VF. CPVT is not associated with organic heart diseases, and there is no sex difference in incidence. In patients with CPVT, exercise and intravenous isoproterenol induce a gradual increase in PVC, which further lead to polymorphic or bidirectional VT, very fast polymorphic VT (350 to 400/min), and finally to VF. Recently, genetic mutations of RyR2 and CASQ2 genes have been found in patients with CPVT.

Secondary prevention is indicated for patients in whom VF or polymorphic VT was observed. The use of an ICD as primary prevention is indicated for patients with a family history of sudden death or a history of syncope. Pharmacological preventive treatment mainly consists of β-blockers. When treatment with β-blockers is insufficient, calcium channel blockers such as verapamil may be effective in some cases. In addition to these drugs, strict limitation of exercise is important. Male patients with RyR2 mutation and patients with CASQ2 gene mutation need early implantation of an ICD (Table 12).

### 14. Other Arrhythmias

#### 1. Non-Brugada Type Idiopathic Ventricular Fibrillation

Some cases of idiopathic VF do not represent ECG findings characteristic to Brugada syndrome. It is important to differentiate non-Brugada type idiopathic VF from Brugada syndrome of which characteristic ECG findings may vary and be normalized over time. Cases of polymorphic VT initiated in the Purkinje’s fiber have been recently reported, and are successfully treated with catheter ablation.

#### 2. Polymorphic Ventricular Tachycardia Triggered by Short-Coupled Ventricular Premature Contraction

This condition develops as very short-coupled PVC (<250 msec in many cases), which leads polymorphic VT to VF.

#### 3. Polymorphic Ventricular Tachycardia Resulting From Ventricular Parasystole

There are cases of polymorphic VT originating from a ventricular parasystolic rhythm. If it is transient, polymorphic VT terminates without causing further changes, while if it persists, it may lead to VF.

---

*Table 12. Prevention of Sudden Death in Patients With Catecholamine-Induced Polymorphic Ventricular Tachycardia*

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Secondary prevention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Patients with VF or cardiac arrest</td>
<td>ICD</td>
<td>+ β-blockers</td>
<td>+ Flecainide</td>
</tr>
<tr>
<td></td>
<td></td>
<td>+ Calcium channel blockers</td>
<td></td>
</tr>
<tr>
<td>Primary prevention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Syncope</td>
<td>ICD, left stellactomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Male patients with RyR2 mutation or patients with CASQ2 mutation</td>
<td>β-blockers</td>
<td>+ Calcium channel blockers</td>
<td>+ Flecainide</td>
</tr>
<tr>
<td>• Children with a family history of sudden death, NSVT or syncope</td>
<td>β-blockers</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

(Note) Exercise should be limited in all patients.

CASQ2, calsequestrin 2 gene; ICD, implantable cardioverter defibrillator; NSVT, nonsustained ventricular tachycardia; RyR2, ryanodine receptor; VF, ventricular fibrillation.
4. Ventricular Tachycardia/Fibrillation Associated With Short QT Intervals

A few cases of VT/VF associated with subnormal short QT intervals on ECG have been reported.\textsuperscript{146} The use of an ICD is indicated for secondary prevention of sudden death in these patients, and catheter ablation may cure this condition in some cases. It is difficult to predict high risk patients indicated for primary prevention.

15. Valvular Heart Disease

1. Aortic Stenosis

Aortic stenosis is the most common valvular heart disease causing sudden death. It has been reported that sudden death occurs in 15 to 20% of adult patients with aortic stenosis (mean age: 60 years) and 44 of 70 patients died from sudden death during survey.\textsuperscript{147,148} The most common causes of sudden death in patients with aortic stenosis are considered VF and SVT. Patients with SVT and VF (after cardiopulmonary resuscitation) should undergo secondary prevention mainly with ICD therapy.

Although aortic pressure gradient is an effective measure of severity of valvular stenosis, it is not useful in predicting sudden death. Arrhythmia findings of Holter ECG correlate with the interventricular septal wall thickness, left ventricular mass, and the decrement of LVEF.\textsuperscript{149–152} Patients with a QTD of $\geq 70$ msec are at high risks of developing syncope and cardiac arrest.\textsuperscript{5,153}

Patients with hemodynamically significant valvular stenosis are indicated for surgery (Table 13).

2. Mitral Valve Prolapse

Mitral valve prolapse had attracted interest as a cause of sudden death since it was the single and only abnormal finding on autopsy in patients died from sudden death of unknown cause.\textsuperscript{154,155} However, it was then found that sudden death is extremely rare among patients with mitral valve prolapse without mitral regurgitation.\textsuperscript{156,157} The incidence of sudden death in patients with mitral regurgitation (1.8%/year) is estimated 50 to 100-fold higher than those without mitral regurgitation. The risk for sudden death among patients with mitral valve prolapsed cannot be predicted with the presence/absence of arrhythmia.\textsuperscript{158} Also, QTD and the presence/absence of ventricular arrhythmia induced during EPS are not considered helpful in predicting sudden death. Although the presence/absence of mitral regurgitation significantly affects the incidence of sudden death, the significance of echocardiography in risk assessment has not been investigated in detail. Patients with SVT or VF are indicated for secondary prevention using an ICD (Table 14).

3. Patients After Prosthetic Valve Replacement

It has been reported that the incidence of sudden death during the late phase after valve replacement with the St. Jude Medical prosthesis ranged 0.5 to 2.4%,\textsuperscript{159,160} while the incidence of sudden death after replacement with bioprosthetic valves was as low as 0.2 to 1%.\textsuperscript{161} It is believed that sudden death in patients using mechanical valves is often related to heart failure, MI or fatal arrhythmia.\textsuperscript{162} Secondary prevention is essential for patients with VT/VF, but prediction and primary prevention of sudden death among patients after valve replacement have not been established yet.

### Table 13. Prevention of Sudden Death in Patients With Aortic Stenosis

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIA</th>
<th>Class IIB</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Secondary prevention</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• SVT or VF</td>
<td>ICD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Surgical repair of stenotic valves</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Primary prevention</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Patients with ventricular arrhythmia in whom SVT or VF is induced</td>
<td>Surgical repair of stenotic valves</td>
<td>Amiodarone</td>
<td></td>
</tr>
<tr>
<td>• Patients with severe valvular stenosis</td>
<td>Surgical repair of stenotic valves</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ICD, implantable cardioverter defibrillator; SVT, sustained ventricular tachycardia; VF, ventricular fibrillation.

### Table 14. Prevention of Sudden Death in Patients With Mitral Valve Prolapse

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIA</th>
<th>Class IIB</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Secondary prevention</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• SVT, VF or cardiac arrest</td>
<td>ICD</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ICD, implantable cardioverter defibrillator; SVT, sustained ventricular tachycardia; VF, ventricular fibrillation.

III. Prevention of Sudden Death in Children

The most common causes of sudden cardiac death in children in Japan are cardiomyopathy, congenital heart diseases and arrhythmias.\textsuperscript{162,163} HCM is the most common cardiomyopathy leading to sudden death in children, and arrhythmias leading to sudden death include VF, complete atrioventricular block, LQTS and WPW syndrome. Major pathological conditions and diseases are described in the following subsections.

1. Sudden Infant Death Syndrome

Several factors and causes have been pointed out for sudden death in infants. Although there is an opinion that sudden death with known causes should not be included in sudden infant death syndrome (SIDS), this section includes sudden death with known causes to ensure appropriate management.
1. **Long QT Syndrome**  
In an investigation of a relationship between SIDS during neonatal period and the QT prolongation, infants with a QTc $\geq 440$ msec were at a high risk of sudden death (Table 15).^{164}

2. **SCN5A Gene Mutation**  
SCN5A gene mutation was reported in about 2% of infants died from SIDS.^{165-167} Since it is difficult to predict sudden death, patients with SCN5A gene mutation are indicated for secondary prevention mainly using an ICD. However, it is difficult to implant an ICD to children with small body size.

3. **Impulse Conduction Disorders**  
Fasciculoventricular tracts were significantly frequently observed during autopsy of infants died from SIDS.^{168,169} It is difficult to predict such findings. Children with a history of cardiac arrest due to atrioventricular block are indicated for pacemaker. Primary prevention is recommended for children with familial impulse conduction disorders (Table 16).

4. **Abnormal Fatty Acid Metabolism**  
It has been reported that disorders of mitochondrial fatty acid $\beta$-oxidation cause accumulation of long-chain acylcarnitine, which may lead to VT, atrial tachycardia, sinus node dysfunction, atrioventricular block, left bundle branch block and other types of arrhythmia responsible for SIDS.^{170} Appropriate dietary therapy and drug treatment are recommended for children with abnormal fatty acid metabolism (Table 17).

### Table 15. Prevention of Sudden Infant Death Syndrome in Infants With Long QT Syndrome

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Secondary prevention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Infants resuscitated from SIDS with a QTc of $\geq 440$ msec</td>
<td>ICD</td>
<td>$\beta$-blockers</td>
<td></td>
</tr>
<tr>
<td>Primary prevention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Infants whose elder siblings died from SIDS with a QTc of $\geq 440$ msec</td>
<td>ICD</td>
<td>$\beta$-blockers</td>
<td></td>
</tr>
<tr>
<td>• Infants without a family history of SIDS with a QTc of $\geq 440$ msec</td>
<td>$\beta$-blockers</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ICD, implantable cardioverter defibrillator; SIDS, sudden infant death syndrome.

### Table 16. Prevention of Sudden Infant Death Syndrome in Infants With Impulse Conduction Disorders

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Secondary prevention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Resuscitated infants</td>
<td>Pacemaker</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary prevention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Infants with familial impulse conduction disorders</td>
<td>Pacemaker</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Table 17. Prevention of Sudden Infant Death Syndrome in Infants With Abnormal Fatty Acid Metabolism

<table>
<thead>
<tr>
<th>Purpose of treatment/findings</th>
<th>Class I</th>
<th>Class IIa</th>
<th>Class IIb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary prevention</td>
<td>Common preventive methods:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Carnitine transporter deficiency</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• CPT-II deficiency</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• CA translocase deficiency</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• LCAD deficiency</td>
<td>+L-carnitine</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• VLCAD deficiency</td>
<td>+L-carnitine</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• LCHAD/trifunctional protein deficiency</td>
<td>+L-carnitine</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

CA translocase, carnitine-acylcarnitine translocase; CPT-II, carnitine palmitoyltransferase type II; LCAD, long-chain acyl-coenzyme A dehydrogenase; LCHAD, long-chain 3-hydroxyacyl-CoA dehydrogenase; VLCAD, very-long-chain acyl-CoA dehydrogenase.

1. WPW Syndrome  
Although there are electrophysiological findings suggesting a high risk of sudden death (VF), it has been pointed out sudden death in children cannot be predictable with EPS.^{119,123,171} Curative treatment using catheter ablation is indicated for patients with WPW syndrome to prevent sudden death (see Table 11).

2. Ventricular Tachycardia and Catecholamine-Induced Polymorphic Ventricular Tachycardia  
Sudden death due to VT is observed even in children under 5 years.^{172} ICD therapy is indicated for children with VT/CPVT, but implantation is not feasible due to small body size and lead troubles associated with bodily movement and development. It has been reported that catheter ablation is effective. The treatment are similar to those recommended for adult patients.^{124,132,139} See the section of CPVT in adults.
3. Long QT Syndrome

LQTS is one cause of sudden cardiac death due to arrhythmia in children. ECG check-ups in schools are common in Japan, and the prevalence of asymptomatic QT prolongation is estimated as 1 in 1,200 children. Symptoms are believed to develop in 1 in 10 children with asymptomatic LQTS. Onset of LQTS during neonatal period and infancy is prevalent among patients with LQT2 or LQT3 mutations, and mexiletine has been reported to be effective in the treatment of LQTS. Poor compliance with drug treatment is a risk factor of arrhythmic accidents.

Patients who still have arrhythmias during treatment with antiarrhythmic drugs such as β-blockers and mexiletine, a pacemaker or an ICD is implanted. There are no reports of large-scale studies in children with LQTS.

Treatment strategies for LQTS are determined according to the results of genetic tests, family histories of sudden death and syncope and ECG findings (eg, exercise stress ECG and Holter ECG). Since it is well known that children with LQT1 mutation often experience drowning or near-drowning accidents during swimming, swimming is commonly prohibited or limited in these children. It is important to provide appropriate guidance on exercise (Table 18). See the section of SIDS for the relationship between LQTS and SIDS.

The Research Committee on Management of Children with LQTS of Japanese Society of Pediatric Cardiology and Cardiac Surgery is conducting a prospective study of children with asymptomatic LQTS, and is expected to propose guidelines for primary prevention for such children.

3. Commotio Cordis

Commotio cordis is a noticeable condition defined as sudden death triggered by relatively mild mechanical impact to the chest observed among athletes. The cause of death is...
believed to be arrhythmias triggered by blunt trauma to the chest wall. Commotio cordis often develops in players of baseball, soft ball and ice hockey, but has also been reported during football, soccer, rugby, lacrosse, boxing and karate as well as when someone deliver a knee kick to the chest or hit the chest with a hand or fist.184,185 Prompt cardiopulmonary resuscitation should be initiated. Chest protectors are used to prevent commotion cordis, but the benefit is limited since it has been reported that 28% of individuals died from commotio cordis wore chest protectors.185 Further safety measures such as using softer balls should be considered (Table 19).

### 4. Congenital Heart Diseases

Sudden death after surgery for congenital heart diseases has been observed.186,187 The incidence of sudden death after surgery is relatively high in patients with tetralogy of Fallot and patients with complete transposition of great arteries.187–189 Secondary prevention is indicated for patients who experienced syncope, symptomatic VT or resuscitated from near-miss sudden death. Since criteria for ICD therapy in children with congenital heart diseases are not available, guidelines for adult patients should be referred.22

#### 5. Pediatric Hypertrophic Cardiomyopathy

HCM is the most important cause of sudden death in children.91,190–194 Children with HCM with a history of resuscitated from near-miss sudden death, syncope or symptomatic VT or a first-degree family history of sudden death are indicated for ICD therapy according to the criteria in each institution or ACC/AHA/NASPE recommendations.117

It is unclear that whether septal myectomy, pacemaker implantation and percutaneous transluminal septal myocardial ablation (PTSMA) used for adult patients with HCM are effective secondary preventive methods for children or not.190,195 Drug treatment is recommended as both primary and secondary prevention. β-blockers, calcium channel blockers, disopyramide (or cibenzoline) are used (Table 20).71,196 It has been reported that highdose β-blocker treatment (5 to 23 mg/kg/day) is highly effective,160 but some studies have denied such effects.71

Pediatric HCM is characterized by the higher prevalence of heart failure and poorer prognosis among children in whom HCM developed during infancy than those with later onset.196,197 Physicians should refer to the School Activity Management Table published by the Japanese Society of School Health for level of exercise limitation and lifestyle guidance.71
6. Kawasaki Disease

There are no sufficient data about arrhythmic deaths among patients in the acute phase of Kawasaki disease. The Japanese Society of Kawasaki Disease, the JCS and the American Academy of Pediatrics among other societies have published detailed guidelines for the management of patients with Kawasaki disease. Patient should be assessed for severity of illness and appropriate treatment strategies according to these guidelines (Tables 21,22).

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