Clinical Therapy in Patients with Aborted Sudden Cardiac Death due to Coronary Spasm

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Background: Coronary artery spasm may lead to the aborted sudden cardiac death (A-SCD). **Objectives:** We investigated the number of coronary vasodilators, including calcium channel blocker (CCB), implantable cardioverter-defibrillator (ICD) implantation and prognosis in patients with A-SCD due to coronary spasm in the real world. **Methods:** We recruited 98 patients (82 men, mean age of 59.6±13.3 years old) with A-SCD due to coronary spasm using a questionnaire. **Results:** Ventricular fibrillation (VF) as a cause of A-SCD was observed in 83 patients (84.7%), while pulseless electrical activity (PEA) was recognized in 12 patients (12.2%). ICD was implanted in 58 patients and appropriate ICD therapies were recognized in 8 patients (13.8%). There were no differences regarding medications between patients with and without ICD or between patients who survived and those who died. Mean follow-up duration was 27.1±19.9 months and three patients died. Mortality during the follow-up period was not different patients with ICD from those without ICD. The mean number of coronary vasodilators including CCB in patients with spasm provocation tests under medications were significantly higher than in those without. VF as an initial cause of A-SCD was recognized in all 3 patients who died, while PEA was a final cause of death in 2 of 3 patients. **Conclusions:** Clinical outcomes in patients with A-SCD due to coronary spasm is satisfactory under medical and mechanical therapy. Decision of requiring the ICD implantation in patients with A-SCD due to coronary spasm is a challenging clinical problem.

KEY WORDS: aborted sudden cardiac death, calcium channel blocker, coronary vasodilator, implantable cardioverter-defibrillator, spasm provocation test under the medication

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I. Introduction

Medical and mechanical therapy in patients with aborted sudden cardiac death (A-SCD) due to coronary spasm is controversial in the clinic^{1–4}. Medication, including calcium channel blocker (CCB) and other vasodilators, is the first line therapy, while an implantable cardioverter-defibrillator (ICD) is defined as class IIa when patients have resistant coronary spasm according to the 2018 Japanese Circulation Society (JCS)/Japanese Heart Rhythm Society guideline on non-pharmacotherapy of cardiac arrhythmias⁵. JCS guideline for the diagnosis and treatment of patients with vasospastic angina (VSA) mentioned that ICD implantation is one option for patients with A-SCD due to coronary spasm⁶. We previously reported that appropriate ICD therapies rescued 30 out of 33 patients with A-SCD due to coronary spasm, whereas 3 patients died: 2 due to PEA and 1 due to refractory coronary spasm⁷. ICD was effective in patients with A-SCD due to coronary spasm to suppress the next serious fatal events^{8,9}. In contrast, aggressive medications, including over two CCBs or more than 3 coronary vasodilators, were administered in just one-third of patients with A-SCD due to coronary spasm according to the previous report. ICD may not be always necessary for all patients with A-SCD due to coronary spasm. However, cardiologists have no answer for which A-SCD patients must receive ICD to suppress the next fatal events. Furthermore, cardiologists have no answer about the optimal medication to suppress the next fatal serious events.

In this article, we investigated the use of medications and ICD in patients with A-SCD due to coronary spasm. We also compared the prognosis and medication in patients with and without ICD. Furthermore, we investigated the usefulness of spasm provocation tests under the medications in A-SCD patients with coronary spasm.

II. Methods

We sent a questionnaire about patients with A-SCD due to coronary spasm at Mar 2019. This questionnaire was distributed among Japanese general institutions over 400–500 beds at random in 204 cardiology institutions including 101 hospitals in West Japan and 103 institutions in East Japan. The answer deadline for the questionnaire was Jun 2019. We investigated the number of SCDs due to coronary spasm, implantation of ICDs, and all spasm provocation tests, including the pharmacological spasm provocation tests under the medications, from Jan 2014 to Dec 2018 (Supplementary files 1–5).

The study protocol complied with the Declaration of Helsinki. Written informed consent was obtained before performing the pharmacological spasm provocation tests, and the protocol of this study was in agreement with the guidelines of the ethical

committee at our institution and institutional review boards of their institutions.

1. The definition of VSA and positive spasm and ischemic ECG changes

According to the JCS guidelines⁶, definite VSA was defined when myocardial ischemia or coronary spasm was obtained in examinations. Spontaneous coronary spasm or provoked positive spasm by pharmacological spasm provocation tests was defined as $\geq 90\%$ transient narrowing with ischemic findings. The degree of ST-segment depression was measured 80 msec after the J point. We considered a result to be positive when at least 1 of the following ischemic ECG changes was demonstrated during spontaneous spasm or pharmacological spasm provocation test: (1) ST-segment elevation of ≥ 0.1 mV in at least 2 contiguous leads; (2) ST-segment depression of 0.1 mV in at least two contiguous leads.

2. Statistical analysis

Data analysis was carried out with SPSS (version 22.0, IBM Japan, Ltd., Tokyo, Japan). All data were presented as the mean $\pm$ SD. Clinical characteristics were analyzed by the Fisher's exact test with correction or the Mann-Whitney U test. Univariate analyses were performed by Cox regression proportional hazards adjusted by age, gender, number of coronary vasodilators or calcium channel blockers, multivessel spasm, spontaneous spasm, ICD implantation and positive spasm provocation test under medications. Event-free survival curves from death were constructed using the Kaplan-Meier survival method. In all analyses, $p < 0.05$ was regarded as statistically significant.

III. Results

1. Collection rate

The percentage of reply for questionnaires was 16.7% (34/204). The response rate of questionnaire in East Japan was 13.6% (14/103), while the questionnaire collection rate in West Japan was 19.8% (20/101). Seven institutions (3.4%) answered that they could not obtain the questionnaire result due to busy daily works or strict ethics committees in their hospitals. The remaining 163 institutions (79.9%) had no answers.

2. Clinical characteristics in patients with A-SCD due to coronary spasm

We recruited 169 patients with A-SCD due to possible coronary spasm as shown in Fig. 1. However, a couple of institutions did not provide the precise data for 39 patients. Furthermore, 32 patients including 27 patients with suspected of VSA and 5 patients enrolled before Jan 2014 were excluded from our study subjects. Finally, as shown in Table 1, we recruited 98 patients

(mean age: 59.6 ± 13.3 years old, male: 83.7%) with A-SCD due to coronary spasm. Mean number of A-SCD per institution was 3.8 ± 3.8 patients. Organic stenosis was found in 7 patients (7.1%). Ventricular fibrillation (VF) was observed in 83 patients (84.7%), while pulseless electrical activity (PEA) was recognized in 12 patients (12.2%). One patient had unknown cause of A-SCD, while ventricular tachycardia was found in one patient. Complete atrioventricular block was recognized in one patient. Approximately three quarters patients (73 patients (74.5%)) had provoked spasm by pharmacological spasm provocation tests, while spontaneous coronary spasm was documented in 12 patients (12.2%) and 10 patients (10.2%) showed spontaneous ST elevation. A-SCD was documented in 3 (3.1%) definite VSA patients under medication. ICD was implanted in 58 patients (59.2%), including 5 patients complicated with Brugada syndrome and 2 patients with early repolarization syndrome. The mean follow-up period was 27.1 ± 19.9 months. During the follow-up periods, appropriate ICD therapies were recognized in 8 (13.8%) patients, including 5 VFs and 3 ventricular tachycardias.

Fig. 1 Flow chart of study subjects.

A-SCD: aborted sudden cardiac death, VSA: vasospastic angina, ICD: implantable cardioverter-defibrillator

Table 1 Patients' clinical characteristics of aborted sudden cardiac death due to coronary spasm

Number	98
Age (year)	59.6 ± 13.3
Male	82 (83.7%)
Organic stenosis (>75%)	7 (7.1%)
Follow-up duration (month)	27.1 ± 19.9
Cause of aborted SCD	
Ventricular fibrillation	83 (84.7%)
Ventricular tachycardia	1 (1.0%)
Pulseless electrical activity	12 (12.2%)
Complete atrioventricular block	1 (1.0%)
Unknown	1 (1.0%)
Diagnosis of spasm	
Provoked spasm by the pharmacological spasm provocation test	73 (74.5%)
Spontaneous spasm documented	12 (12.2%)
Spontaneous ST elevation documented	10 (10.2%)
VSA definite	3 (3.1%)
With ICD implantation	58 (59.2%)
Without ICD implantation	40 (40.8%)
With Brugada syndrome or early repolarization syndrome	7 (7.1%)
Appropriate ICD shocks	8 (13.8%)
Mean number of coronary vasodilators	2.2 ± 1.0
Mean number of calcium channel blockers	1.3 ± 0.5
Death	3 (3.0%)
With ICD implantation	2 (2.0%)
Without ICD implantation	1 (1.0%)

(SCD: sudden cardiac death, VSA: vasospastic angina, ICD: implantable cardioverter-defibrillator)

Table 2 Comparisons of clinical data between patients with and without ICD implantation

	With ICD implantation	Without ICD implantation	p value
Number	58	40	
Age (year)	57.5 ± 13.0	62.7 ± 13.3	0.0439
Male	50 (86.2%)	32 (80.0%)	0.4464
Organic stenosis (>75%)	5 (8.6%)	2 (5.0%)	0.4743
Follow-up duration (month)	29.3 ± 21.1	23.3 ± 17.4	0.1493
Cause of aborted SCD			
Ventricular fibrillation	53 (91.4%)	30 (75.0%)	0.0509
Ventricular tachycardia	1 (1.7%)	0	0.8609
Pulseless electrical activity	2 (3.4%)	10 (25.0%)	0.0019
Complete atrioventricular block	1 (1.7%)	0	0.8609
Unknown	1 (1.7%)	0	0.8609
Diagnosis of spasm			
Provoked spasm by the pharmacological spasm provocation test	44 (75.9%)	29 (72.5%)	0.7614
Spontaneous spasm documented	6 (10.3%)	6 (15.0%)	0.7402
Spontaneous ST elevation documented	7 (12.1%)	3 (7.5%)	0.6640
VSA definite	1 (1.7%)	2 (5.0%)	0.7591
With Brugada or early repolarization	7 (12.1%)	0	0.0561
Mean number of coronary vasodilators	2.1 ± 1.0	2.3 ± 1.0	0.4614
Mean number of calcium channel blockers	1.3 ± 0.6	1.3 ± 0.6	0.8898
Death	2 (3.4%)	1 (2.5%)	0.7591

(SCD: sudden cardiac death, VSA: vasospastic angina, ICD: implantable cardioverter-defibrillator)

The mean numbers of coronary vasodilators and CCBs at the baseline were 2.2 ± 1.0 and 1.3 ± 0.5 , respectively. During the follow-up periods, death was observed in two patients with ICD, while one patient without ICD died.

3. Comparisons between patients with and without ICD

As shown in Table 2, patients with ICD were younger than those without ICD. The frequency of VF in patients with ICD implantation was higher than in those without ICD but not significant, while the incidence of PEA in patients without ICD was significantly higher than in those with ICD (25.0% vs. 3.4%, $p=0.0019$). There were no differences regarding medications, including coronary vasodilators and CCBs, between the two groups. Mortality rate during the follow-up periods in patients with ICD was not different from that in those without ICD (3.4% vs. 2.5%, $p=0.7591$). Death-free survival in all patients with A-SCD who had ICD was not different from those without ICD, as shown in Fig. 2a. As shown in Fig. 2b, death-free survival in patients with A-SCD due to pure coronary spasm who had ICD implantation was similar to that in patients without ICD.

4. Comparisons of clinical data between patients who survived or who died

Death was observed in 3 patients, while the 95 patients remained alive during the follow-up periods. Patients who died were older than those who survived (68.7 ± 6.8 vs. 59.3 ± 13.4 year old, $p=0.0276$). VF as a cause of A-SCD in patients who survived was not different from that in those who died (84.2% vs. 100%, $p=0.9807$). Furthermore, there were no differences regarding medications, including mean number of coronary vasodilators and CCBs, between the two groups.

5. Comparisons of clinical data between patients with and without Brugada and early repolarization syndrome

There is no difference except mean number of CCB between the two groups. Mean number of CCB in patients with Brugada and early repolarization syndrome was markedly lower than those in patients without it (0.9 ± 0.4 vs. 1.3 ± 0.5 , $p=0.0136$).

6. Comparison of clinical data between patients with and without appropriate ICD therapies

Appropriate ICD therapies were observed in 8 patients. There were no differences between the two groups. No death was observed in patients with appropriate ICD therapies, whereas two

Fig. 2 Kaplan-Meier survival curve in patients with A-SCD due to coronary spasm.

a: with Brugada syndrome and early repolarization syndrome.

b: without Brugada syndrome and early repolarization syndrome.

Table 3 Spasm provocation data in patients with aborted sudden cardiac death due to coronary spasm

	ACH	ER	RCA positive	LCX positive	LAD positive	LMT positive	1 vessel spasm	2 vessel spasm	3 vessel spasm	Multiple spasm
With ICD implantation (n=44)	39	7	23 (52.3%)	15 (34.1%)	33 (75.0%)	3 (6.8%)	17 (38.6%)	19 (43.2%)	8 (18.2%)	27 (61.4%)
Without ICD implantation (n=29)	19	8	20 (69.0%)	9 (31.0%)	16 (55.2%)	0 (0%)	15 (51.7%)	10 (34.5%)	4 (13.8%)	14 (48.3%)
P value			0.1560	0.7856	0.0776	0.4045	0.2701	0.4573	0.6205	0.2701
TOTAL (n=73)	58	15	43 (58.9%)	24 (32.9%)	49 (67.1%)	3 (4.1%)	32 (43.8%)	29 (39.7%)	12 (16.4%)	41 (56.2%)

(ACH: acetylcholine, ER: ergonovine, RCA: right coronary artery, LCX: left circumflex artery, LAD: left anterior descending artery, ICD: implantable cardioverter-defibrillator)

patients without appropriate ICD therapies died due to PEA.

7. Spasm provocation data

During the 5 years, pharmacological spasm provocation tests were performed in 5,027 examinations in 34 institutions. Table 3 shows the spasm provocation results in patients with A-SCD due to coronary spasm. There was no difference regarding the incidence of provoked positive spasm between patients with and without ICD. Left main trunk spasm was observed in 3 patients with ICD implantation, while patients without ICD implantations had no left main trunk spasm.

8. Clinical findings in patients who had spasm provocation tests under medications

As shown in Table 4, spasm provocation tests under the medications were performed in 18 (18.4%) out of 98 patients. Provoked positive spasm recognized before medications were attenuated or disappeared in 7 patients after the tests under

medications. However, provoked spasm was not attenuated in 11 patients irrespective of performing the spasm provocation tests under medications. The mean number of coronary vasodilator in patients with spasm provocation tests under medications was significantly higher than in those without (3.0 ± 1.1 vs. 2.0 ± 0.9 , $p=0.0036$). The mean number of CCBs in patients with spasm provocation tests under medication was also remarkably higher than in those without it (1.8 ± 0.5 vs. 1.2 ± 0.5 , $p=0.0010$). During the follow-up periods, there were no death in all 18 patients with spasm provocation tests under medications, while appropriate ICD therapies were observed in 3 out of 8 patients with ICD. Two ventricular tachycardias and one ventricular fibrillation were recovered after the appropriate ICD therapies.

9. Clinical and angiographic findings in patients who died

As shown in Table 5, 3 patients died within 2 years after A-SCD. Three coronary vasodilators were administered in one patient, while one case had two CCBs. Although VF was recog-

Table 4 Clinical findings and spasm provocation tests without medication and under medication in 18 patients with aborted sudden cardiac death due to coronary spasm

No	Age	Sex	Cause	ICD	Spontaneous spasm or ST change	Spasm provocation test without medication	Spasm provocation test under medications	Number of coronary vasodilators	Number of CCB
1	66	M	VF	no		ER(R: 20/L: 32) #2(d) #13(d)	ER(R: 20/L: 32) no spasm	2	2
2	49	F	CAVb	yes	ST ele in INF	Not implemented	ACh(L: 20/50/100/200) #7(f) #11(d)	3	2
3	52	M	VF	yes		ACh(R: 20/L: 20) #4(d) #5(f) #7(f)	ACh after ER(R: 80/L: 200) #7(f)	3	2
4	68	M	VF	no	ST ele in ANT	ACh(R: 50/L: 20) #3(t) #6(t) #11(d)	ACh after ER(R: 80/L: 200) #3(f)	3	2
5	73	M	VF	no	Negative T in ANT	ACh(R: 25/L: 25) #2(t) #7(d) #11(d)	ACh(R: 25/L: 25/50/100) #2(f) #8(f) #13(d)	3	2
6	69	M	VF	yes		ACh(R: 20/L20/50/100) #2(t) #7(subt) #13(d)	ACh(R: 20/L: 20/50/100) #2(t) #7(f) #13(d)	2	1
7	64	M	VF	yes		ACh(R: 20/50/L: 20/50) #1/4(d) #8/9/11/12(d)	ACh(R: 20/50/L:20/50) #1/4(d) #8/9/11/12 (d)	4	2
8	42	M	VF	yes		ER(R: 20/40/L: 40/80) #2/3(d) #6(d)	ER(R: 10/20/30/L: 10/20) #7(d)	4	2
9	82	F	VF	yes		ER(R: 20/50/L: 20/50/100) #7(f)	ER(R: 20/30/L: 30/30) no spasm	5	3
10	45	M	VF	no		ACh(R: 20/50/L: 20/50/100) #4(d) #7(d)	ACh(R: 20/50/L: 20/50/100) #4(d) #7(d)	1	1
11	38	F	VF	yes		ACh(R: 20/50/L: 20/50/100) #4(d) #7(d)	ACh(R: 20/50/L: 20/50/100) #4(d) #7(d)	1	1
12	25	M	VF	yes		ACh(R: 20/50/L: 20/50/100) #4AV(d) #8(d)	ACh(R: 20/50/L: 20/50/100) #4AV(d) #8(d)	2	1
13	69	F	VF	no		ACh(R: 20/50) #2(subt) #4PD(subt)	ACh(R: 20/50) #2(subt) #4(subt)	2	1
14	69	F	VF	no		R(20/50/100) #4PD(subt)	R(20/50/100) #4PD(subt)	4	2
15	64	M	VF	no		ACh(R: 20/50/100/ L: 20/50/100) #1(t)	ACh(R: 20/50/100/ L: 20/50/100) #1(t)	4	2
16	64	M	VF	no	#1(subt) spasm	Not implemented	ACh(R: 20/50/L: 20/50/100) no spasm	3	2
17	67	M	VF	no	#1/6/11(f) spasm	Not implemented	ACh(L: 20/50/100) #13(d)	4	2
18	57	M	VF	no	VSA definite	Not implemented	ACh(L: 20/50) #7(d) #14(d)	4	2
Mean	59			(8)				3.0 ± 1.1	1.8 ± 0.5

(ICD: implantable cardioverter-defibrillator, CCB: calcium channel blocker, M: male, F: female, VF: ventricular fibrillation, CAVb: complete atrio-ventricular block, INF: inferior, ANT: anterior, VSA: vasospastic angina, (f): focal spasm (subt): subtotal, (d): diffuse spasm, (t): total, ACh: acetylcholine, ER: ergonovine, R: right coronary artery, L: left coronary artery)

nized in all 3 patients as an initial diagnosis, no final cause of VF was found. PEA was observed in 2 patients as a final cause of death.

10. Univariate Cox hazard regression analyses for death

No determinant factors for death-free survival was found.

IV. Discussion

In this article, we showed the satisfactory clinical outcomes in patients with A-SCD due to coronary spasm under medical and mechanical therapies. However, appropriate ICD therapy was recognized in just 8 (13.8%) patients, including 5 VFs and 3 ventricular tachycardias, whereas the remaining 86.2% of patients had no appropriate ICD therapy. There were no differences regarding the number of coronary vasodilators or CCBs between patients with and without ICD or between patients who survived and those who died. The mean number of coronary vasodilators or CCBs in patients with spasm provocation tests under medica-

tions was significantly higher than the number without tests. No deaths during the follow-up period were recognized in patients with spasm provocation tests under medications. Death due to PEA was observed in 2 patients with ICD implantation, although VF was the initial cause of A-SCD in all 3 died patients. Cardiologists should reconsider the medications administered in patients with A-SCD due to coronary spasm. Furthermore, cardiologists should select patients with A-SCD who require ICD to suppress the next fatal events due to pure coronary spasm.

1. Coronary vasodilators and CCBs

According to previous data in patients with A-SCD due to coronary spasm, the mean coronary vasodilator numbers were 2.6 ± 0.9 (Ahn et al)¹⁰, 1.5 ± 0.7 (Manero et al)¹¹, or 2.0 ± 0.8 (Sueda et al)⁷. In this study, the mean number of coronary vasodilators was 2.2 ± 1.0 , and the mean number of CCBs was 1.3 ± 0.5 . The mean number of coronary vasodilators in this study was similar to that in VSA patients without A-SCD in the study by

Table 5 Clinical and angiographic findings in patients with death who survived from aborted sudden cardiac death once time

No	Age	Sex	Initial diagnosis	Organic stenosis	ICD	Spasm provocation test or spontaneous spasm/ST elevation	Duration (month)	Number of coronary vasodilators	Number of CCB	Cause of death
1	74	M	VF	no	yes	Spontaneous Cx spasm	11	2	2	PEA
2	61	M	VF	no	yes	ACh(R: 50/L: 100) #3(d) #6(d)	15	1	1	PEA
3	71	M	VF	no	no	ACh(L: 50) #8/13(t) #1-3 spontaneous spasm	11	3	1	Unknown
Total	68			0	2		12.3 ± 2.3	2.0 ± 1.0	1.3 ± 0.6	

(ICD: implantable cardioverter-defibrillator, CCB: calcium channel blocker, M: male, F: female, VF: ventricular fibrillation, PEA: pulseless electrical activity, Cx: circumflex artery, ACh: acetylcholine, R: right coronary artery, L: left coronary artery, (d): diffuse spasm, (f): focal spasm, (t): total spasm, VSA: vasospastic angina)

Ahn et al (2.3±0.8)¹⁰. Variant angina is one category of higher VSA disease activity. Compared with non-variant angina, cardiologists may administer abundant coronary vasodilators to suppress the angina attacks in patients with active variant angina. Variant angina is along the same lines as coronary spasm. However, A-SCD due to coronary spasm may be another special category of VSA. These patients may have a higher risk type. Cardiologists should reconsider their thought about medications in patients with A-SCD due to coronary spasm.

2. ICDs

Although ICDs were implanted in more than half of patients with A-SCD due to coronary spasm in this study, appropriate ICD therapies were found in just 8 patents (13.8%). This number was lower than that reported by Sueda et al but not significant (24.1% (33/137) vs 13.8% (8/58), $p=0.1068$)⁷. In this article, we could not find any difference about medications between patients with and without ICDs. ICD implantation is not a final therapy for patients with A-SCD due to coronary spasm. Cardiologists do not currently have a precise selection tool for ICD implantation. Decision of requiring ICD implantation in patients with A-SCD due to coronary spasm is an unsolved clinical problem.

3. Clinical outcomes compared with other diseases except for congenital monogenetic cardiac disease after ICD implantation as a secondary prevention

According to previous reports, 23%-34.4% of patients without congenital monogenetic cardiac disease died in the secondary prevention after ICD implantation, and appropriate ICD therapy was recognized in 37%-39.2% of these patients during median follow-up period of approximately 5 years^{12, 13}. In our study, mortality was 3.1% during median follow-up of 27.1 months in patients with A-SCD due to coronary spasm, whereas death rate was 3.4% during median follow-up of 29.3 months in patients with A-SCD due to coronary spasm after the implantation of ICD. The frequency of appropriate ICD therapy in this study was remarkably lower than previous reports. Furthermore, the clinical

outcomes in patients with A-SCD due to coronary spasm were satisfactory under optimal medical and mechanical treatment.

4. Spasm provocation tests under medications

We already reported spasm provocation tests under medications in patients with refractory VSA on emergency admission and with A-SCD due to coronary spasm¹⁴⁻¹⁸. In this study, spasm provocation tests under medications were observed in 18 patients. All 18 patients were alive and had significantly large number of coronary vasodilators and CCBs. Provoked spasm attenuated or disappeared in 7 (38.9%) patients, while the remaining 11 patients (61.1%) had no attenuation of provoked spasm irrespective of spasm provocation tests under medications. In the real world, cardiologists have no method or strategy to define the real coronary spasticity or effectiveness of coronary vasodilators to suppress serious catastrophic coronary spasm. In this study, we could not prove the direct efficacy of spasm provocation tests under medications when we decided to implant an ICD in patient with A-SCD due to coronary spasm.

5. Study limitations

There are several limitations in this study. The first limitation is that sample size was small, and the response rate of questionnaire was 16.7% of all 204 institutions. In 2004 and 2005, we sent the questionnaire concerning the number of cases of coronary arteriography, percutaneous coronary intervention, invasive/non-invasive spasm provocation tests, and the clinical status of coronary spasm to the members of the JCS in 120 cardiology hospitals in the Chugoku/Shikoku area and in 1,177 cardiology hospitals in all of Japan¹⁹⁻²¹. The surveys were returned from 45 hospital (37.5%) and 208 hospitals (17.7%), respectively. Compared with the previous questionnaires, the collection rate in this article was not different from all Japan questionnaires. The second limitation is missing precise characteristics about coronary risk factors and serum data in this study subjects with A-SCD due to coronary artery spasm because of questionnaire flaw. The

third limitation is also missing data about the value of left ventricular ejection fraction, side effect of ICD implantation including infection and inappropriate ICD therapies because of questionnaire flaw. Further study concerning medications and the necessity for an ICD will be necessary for characterizing A-SCD due to coronary spasm in the future.

V. Conclusions

Clinical outcomes in patients with A-SCD due to coronary spasm is satisfactory under medical and mechanical therapy. The incidence of death in patients with ICD was not different from those without it. Appropriate ICD therapies were observed in just 13.8% of patients, whereas the remaining 86.2% of patients had no mechanical actuations. Medications were reinforced and no death was observed in patients with spasm provocation tests under medications. Decision of requiring the ICD implantation in patients with A-SCD due to coronary spasm is an unsettled clinical problem.

Appendix

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Conflicts of interest

The authors declare that they have no conflict of interest.

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