

Original Article

Relationship between the Cumulative Incidence of Kawasaki Disease and the Prevalence of Electrocardiographic Abnormalities in Birth-Year Cohorts

Kunio Kawai^{1,2}, Mayumi Yashiro², Yosikazu Nakamura², and Hiroshi Yanagawa²

¹Kouno Clinic, Fukui, Japan

²Department of Public Health, Jichi Medical University, Shimotsuke, Tochigi, Japan

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ABSTRACT

Background: Kawasaki disease (KD) causes systemic vasculitis and coronary aneurysms. It frequently results in electrocardiographic (ECG) abnormalities of short duration. Cardiac sequelae persist beyond the acute stage in a few patients. There are many areas to be investigated concerning the effects on the vascular system of patients suffering from KD and its sequelae.

Methods: The cumulative incidences of KD and its cardiac sequelae were calculated in birth-year cohorts, using data obtained from KD nationwide surveys. The results were compared with the prevalence of ECG abnormalities detected in cardiac examinations conducted at primary and secondary schools for each birth-year cohort. This comparison allowed observation of relationships in these trends for each birth-year cohort.

Results: The cumulative incidence of late-stage cardiac sequelae gradually declined. However, there were increases in the cumulative incidence of ECG abnormalities and in the cumulative incidences of KD and acute-stage cardiac disorders related to KD.

Conclusions: The results suggest that even among children without late cardiac sequelae, KD can have a persistent effect on the cardiovascular system. It thus appears necessary to extend clinical observation of children with a history of KD, even if they developed only acute-stage cardiac lesions.

Key words: Kawasaki disease; mucocutaneous lymph node syndrome; cumulative incidence; birth-year cohort; prevalence of electrocardiographic abnormalities

INTRODUCTION

Since the first report on Kawasaki disease (KD), in 1967, there have been 19 nationwide surveys conducted in Japan from 1970 through 2006.¹ These biennial retrospective incidence surveys investigated patients with KD who visited target hospitals for the first time. The medical facilities that were requested to participate in the survey were hospitals with pediatric departments and 100 or more beds, or those specializing in pediatrics but with fewer than 100 beds.

The 19th nationwide survey of Kawasaki disease investigated the period from January 2005 through December 2006. Of the 2183 hospitals that were requested to participate, 1543 reported 20475 patients with KD during this time. The percentage of KD patients recorded in these surveys was reported to exceed 80% of all KD patients.²

From the early days of these surveys, vasculitis-related cardiac sequelae, such as coronary aneurysms, were noted,

and electrocardiographic (ECG) abnormalities attracted attention. It was found that 72% of KD patients developed ECG abnormalities, although some only temporarily, according to Asai et al.³ Although ECG abnormalities are not investigated in the nationwide surveys of KD, the surveys do request a list of patients who received a diagnosis of KD, together with their cardiac sequelae—such as coronary aneurysm, giant coronary aneurysm, coronary dilatation, coronary stenosis, valvular dysfunction, and myocardial infarctions.

In these nationwide surveys, the annual number of KD patients per 100 000 children aged 0 to 4 years is reported. These data are useful for pediatricians who wish to observe the annual epidemiologic trends in KD.^{4,5} However, it is important that general physicians who perform cardiac examinations in primary and secondary schools know the numbers of students and children who suffer from cardiac sequelae due to KD within a single school grade. For this

Address for correspondence: Kunio Kawai, Kouno Clinic, 7-31-1 Kaburaki, Minamiechizen-chou, Fukui 915-1113, Japan (e-mail: k-m-kawai@sun.vipa.ne.jp).
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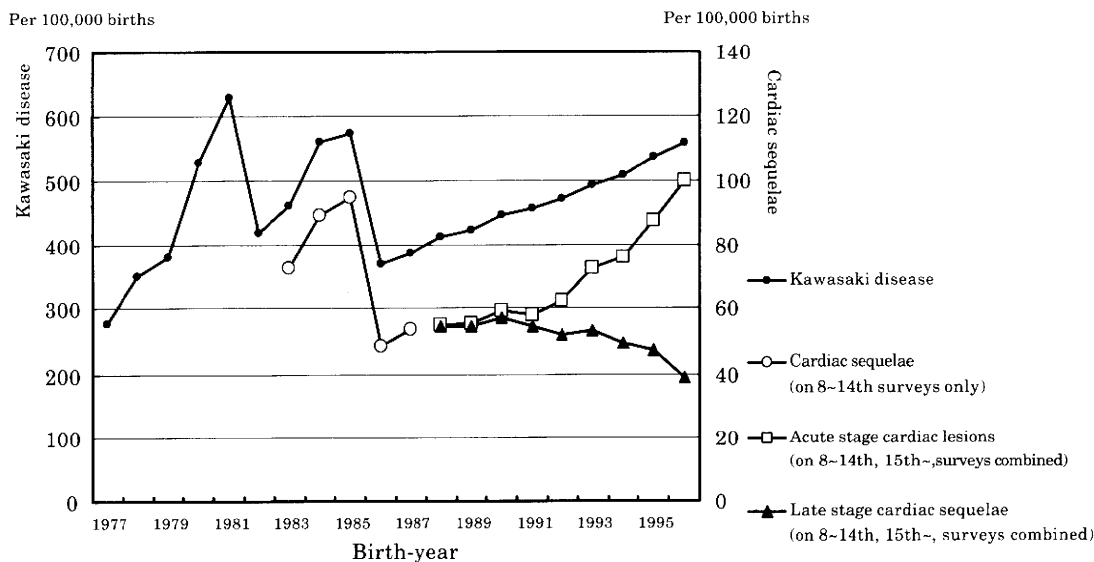


Figure 1. Cumulative incidences of Kawasaki disease and cardiac sequelae at age 10 years, by patient birth year. The cumulative incidences of acute cardiac lesions and late-stage cardiac sequelae were calculated by combining data from the 8th to 14th nationwide surveys with those from the 15th and later nationwide surveys.

purpose, reports on annual incidence are not informative enough to identify cardiac sequelae several years after onset. Previous epidemiologic studies indicated that the age-specific incidence of KD peaks at approximately age 1 year. Thus, several years will elapse before the child reaches school age. Cumulative data on the incidence of KD are necessary for general physicians. For internists, it is more important to know the trend in KD incidence for the patient's age-specific cohort.

We propose a new approach. Using data collected in the nationwide surveys on KD, KD patients were grouped by year of birth, ie, birth-year cohort. The cumulative incidences of KD, acute-stage cardiac lesions, and cardiac sequelae were calculated. Studies have reported the cumulative incidence of KD for each birth-year cohort⁶ and the cumulative incidences of patients with acute-stage cardiac lesions and cardiac sequelae due to KD in each birth-year cohort.⁷

Using these data, we examined the relationship between the cumulative incidence of KD in each birth-year cohort and the prevalence of abnormalities found during school cardiac examinations among these cohorts.

METHODS

The subjects of the current study were selected from those reported to the 8th to 19th nationwide surveys of KD, which investigated the period from January 1983 through December 2006. They were 153 718 patients who presented for KD treatment for the first time; thus, patients suffering from recurrences were excluded. Those with no record of cardiac complications were assumed to be free of such sequelae.

Starting with the 8th nationwide survey (July 1982 through December 1984), a new question related to cardiac sequelae

was added to the survey. Beginning with the 15th nationwide survey (January 1997 through December 1998), the survey sheet was designed in such a manner that sequelae could be recorded on 1 of 2 forms: one for cardiac abnormalities that developed during the early stage (within 1 month) of the disease, referred to as acute-stage cardiac lesions, and the other for detectable persistent cardiac abnormalities (after the first month), referred to as late-stage cardiac sequelae.

In our study of the cumulative incidence of KD in birth-year cohorts, patients with KD were classified by their birth year, and the cumulative number of patients aged 0 to 9 years for the previous 10 years was expressed as the number per 100 000 individuals born in the respective year. The cumulative number of 0-year patients is the number of those who had KD during the period from January through December of that birth year; therefore, the cumulative number of patients for 10 years represents the number of those with KD from January of their birth year through December of the year when the patients were age 9 years. Similarly, the cumulative incidence of those with cardiac sequelae from KD in each birth-year cohort was expressed as the 10-year cumulative incidence of those having either acute- or late-stage lesions due to KD between the ages of 0 and 9 years. Thus, those identified as having cardiac sequelae in one of the items included on the 8th through 14th surveys (time of observation, January 1983 through December 1996) were classified as having cardiac lesions in both the acute and late stages, as shown in Figure 1.

Cardiac sequelae were covered by one item on the questionnaire included in the 8th through 14th nationwide surveys. The subject of this item could be regarded as late-stage lesions, which are investigated in an item added to the questionnaire for the 15th survey. However, after the change

of questionnaire, the reported number of patients with late-stage lesions due to KD suddenly dropped in the 15th survey, as compared with the number of the patients with such lesions reported in the 8th through 14th surveys. This indicates that the numbers for patients with cardiac sequelae in the 8th through 14th surveys included those with the acute-stage lesions. Therefore, in the 8th through 14th surveys, the number of patients with "cardiac sequelae" was greater than the number of those with late-stage lesions and lower than the number of patients with acute-stage lesions. It is necessary to accumulate data for several years to calculate the cumulative incidence of KD.

Those listed with "cardiac sequelae" in one of the items included in the 8th through 14th surveys were counted as having both acute- and late-stage cardiac lesions, and in the cohorts from 1988 to 1996, provisional cumulative incidence rates of acute-stage cardiac lesions and late-stage cardiac sequelae of KD in the birth-year cohort were calculated.

These data on KD were compared with those obtained from the annual Report of the Statistical Survey on School Health published by the Ministry of Education, Culture, Sports, Science,⁸ which categorizes its results by student birth year. In the Statistical Survey on School Health, there are data on the prevalence of cardiac diseases and cardiac anomalies, hereafter referred to as the prevalence of cardiac diseases. The prevalence of ECG abnormalities is also reported. The prevalence of cardiac diseases and the frequency of ECG abnormalities revealed by school cardiac examinations within a single school grade can be characterized as the prevalence of abnormalities among a birth-year cohort of pupils. These comparative studies were conducted to identify changes in birth-year cohorts by observing the cumulative incidence of KD and the prevalence of cardiac diseases and ECG abnormalities in related examinations.

The prevalence of cardiac disease and cardiac anomalies was surveyed for each school year. Starting in 1995, ECG examinations were given to children aged 6, 12, and 15 years, which corresponds to entry into elementary, junior high school, and senior high school, respectively. The proportions of those who had cardiac diseases for each school year and the proportions of ECG abnormalities found at ages 6 and 12 were converted to percentages or number per 100 000 for each birth-year cohort. Secular trends were analyzed by comparing them against those for the cumulative incidence of KD for each birth-year cohort, which were obtained from the nationwide surveys.

Next, we focused on ECG examinations given at age 6, and the graphs of the secular trend in the prevalence of cardiac diseases and ECG abnormalities observed in cardiac examinations at schools were compared against the cumulative incidence of acute-stage cardiac lesions and cardiac sequelae due to KD, obtained from the nationwide surveys, in which the incidences were listed for each birth-year cohort. The cumulative number of patients with cardiac

lesions at age 6 years was calculated for each patient birth-year cohort by totaling the number of those suffering from cardiac sequelae due to KD between January of their birth year and December of the year when they became 5 years of age.

RESULTS

Figure 1 shows graphs of the cumulative incidence of KD per 100 000 births and the cumulative incidences of cardiac lesions and late-stage cardiac sequelae at age 10 years for each birth-year cohort, according to the nationwide surveys. The secular trend for the cumulative incidence of cardiac sequelae for birth-year cohorts before 1990 parallels that for the cumulative incidence of KD. After 1991, there were rises in both the cumulative incidences of acute-stage cardiac lesions and KD; however, the cumulative incidence of late-stage cardiac sequelae gradually declined after 1991.

Figure 2 shows the percentages of cardiac diseases and ECG abnormalities detected at school health examinations at age 6, 12, and 15 years, adjusted for birth-year cohort. The prevalence of ECG abnormalities was higher at ages 12 and 15 than at 6, while prevalences at ages 12 and 15 were generally similar. When prevalence was compared by birth year, a general increase was noted. Linear regression analysis revealed that the frequency of ECG abnormalities, adjusted for birth-year cohort, increased significantly ($P < 0.05$). In contrast, the prevalence of cardiac diseases showed few age-related differences at ages 6, 12, and 15 years. Sequential comparisons by birth year also revealed a generally stable pattern, with deviations limited to 0.5% to 1%. There was no statistically significant trend in the prevalence of cardiac diseases adjusted for birth-year cohort.

Figure 3 shows the secular trend in the prevalence of ECG abnormalities and cardiac diseases detected at school health examinations in children aged 6 and 12 years compared with the cumulative incidence of KD obtained from the nationwide surveys. Examined sequentially by birth year, the graph for the prevalence of ECG abnormalities gradual increased in a manner similar to that for the cumulative incidence of KD. At age 12, the change faithfully reflected the sudden dramatic increase in the number of KD patients born in 1985. In addition, among the cohorts from the years 1986 to 1995, the slope of the curve for the prevalence of ECG abnormalities is similar to that for the cumulative incidence of KD. However, after 1995, there was a sharp increase in the cumulative incidence of KD at age 6, whereas the prevalence of ECG abnormalities only slightly increased. The change in the prevalence of cardiac diseases did not parallel the trend in the cumulative incidence of KD.

The graphs for the cumulative incidences of acute-stage cardiac lesions and late-stage cardiac sequelae due to KD for the birth-year cohort at age 6 (from nationwide surveys) were compared with those for the prevalence of ECG abnormalities

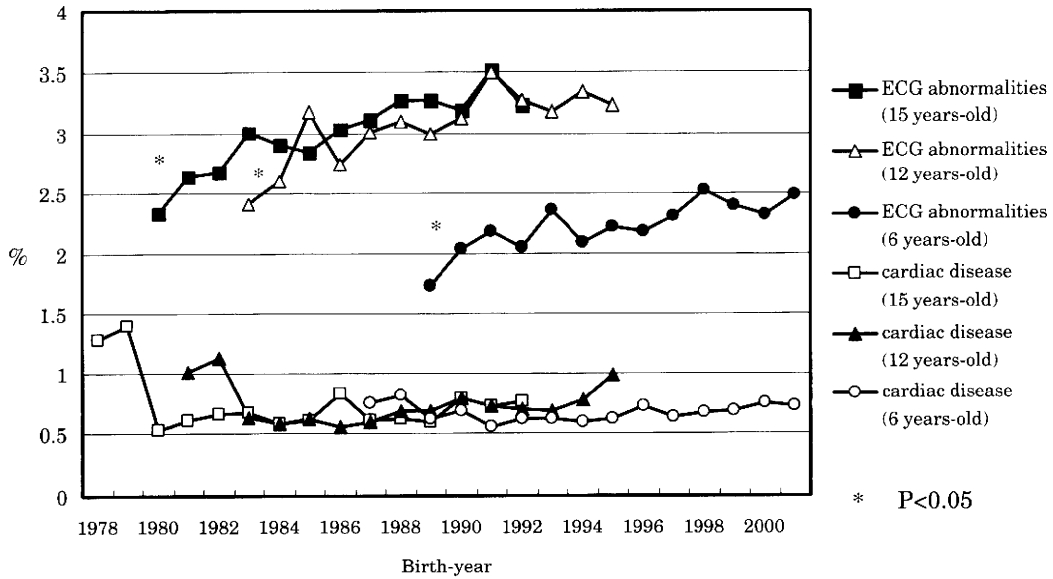


Figure 2. Percentages of cardiac disease abnormalities on electrocardiography (ECG), by birth-year cohort, among students aged 6, 12, and 15 years (data obtained from the Statistical Survey on School Health). Linear regression analysis was conducted to investigate the statistical significance of the observed trends.

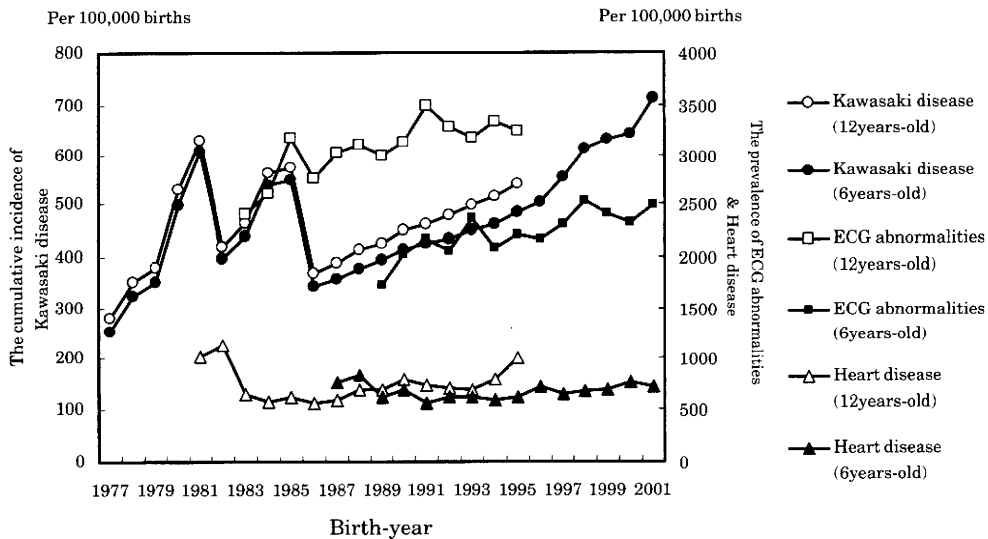


Figure 3. Cumulative incidence of Kawasaki disease and the prevalences of cardiac disease and abnormalities on electrocardiography (ECG), by birth-year cohort, among students aged 6 and 12 years.

and heart diseases detected at school health examinations (Figure 4). Since 1997, the changes in the cumulative incidence of acute-stage cardiac lesions were accurately reflected in the occurrence of ECG abnormalities.

The graph for the development of cardiac diseases was not similar to those for the cumulative incidence of acute-stage cardiac lesions or those for cardiac sequelae.

DISCUSSION

The data on the prevalence of ECG abnormalities and cardiac diseases in the current study were gathered from the annual

Report of the Statistical Survey on School Health (1993–2007).⁶ The report is prepared by extracting samples, rather than by studying the entire student population. In the latest report, the 2007 Statistical Survey on School Health, the data on students were extracted from 22.4% of Japanese students aged 5 to 17 years. In the process of sample extraction, schools are first stratified by prefecture or other legislative district, type of school, and number of students. Within each group thus stratified, simple randomization was used to determine the schools at which a survey was to be conducted. The rates of occurrence of cardiac diseases and anomalies and ECG abnormalities are reported annually. It can

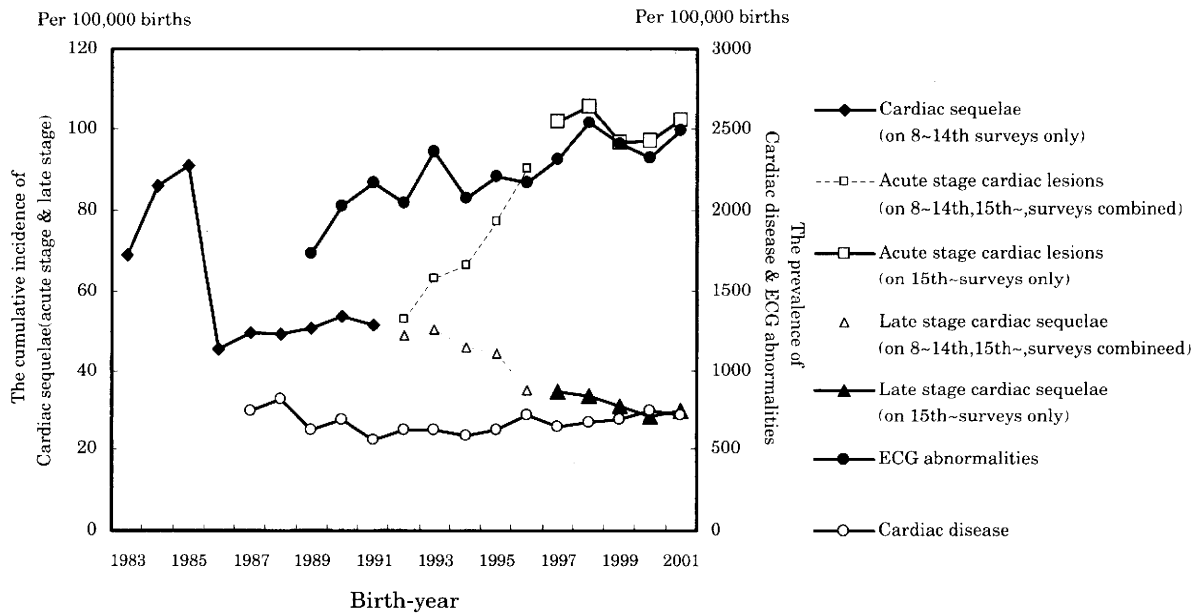


Figure 4. Cumulative incidence of cardiac sequelae of Kawasaki disease and the prevalences of cardiac disease and abnormalities on electrocardiography (ECG), by birth-year cohort, among students aged 6 years. The cumulative incidences of acute cardiac lesions and late-stage cardiac sequelae were calculated by combining data from the 8th to 14th nationwide surveys with those from the 15th and later nationwide surveys.

be assumed that these rates generally agree with overall incidences among Japanese students. If one posits that the students in a single grade level constitute a population group all born within a single year, it is plausible that the occurrence of a cardiac abnormality among the first-grade students, ie, children aged 6 years, may be replaced by the occurrence of a cardiac abnormality developing 6 years later in the cohort of individuals who were born 6 years earlier. The data on the occurrence of diseases thus obtained by adjusting for each birth-year cohort were studied by comparing the cumulative incidence of KD and related cardiac sequelae, both for acute- and late-stage lesions, of a birth-year cohort that had been reported earlier.

The cardiac examination at elementary schools is carried out from April through June for first-graders, when they are 6 years of age. Therefore, the cumulative incidence was calculated by using data until December of the year when the patients were 5 years of age.

Secular trends in the prevalence of cardiac diseases for birth-year cohorts detected at school health examinations show a relative lack of change at ages 6, 12, and 15 years and trivial increases among cohorts. This feature of the prevalence of cardiac diseases appears to differ from that of the cumulative incidence of KD and its acute-stage cardiac lesions and cardiac sequelae in data obtained from the nationwide surveys. However, the graphs for the prevalence of ECG abnormalities in birth-year cohorts that were found in school cardiac examinations show changes similar to those for the cumulative incidence of KD. According to the annual incidence reports of nationwide surveys, there were 3

epidemics of KD, in 1979, 1982, and 1986. Moreover, we previously reported that rises in the cumulative incidence of KD for each birth-year cohort were noted among the birth-year cohort 1 year before the years of these 3 outbreaks. If one examines the data for age 12 years, the prevalence of ECG abnormalities in each birth-year cohort appears to increase, which corresponds to the rising trend for the third outbreak in the 1985 cohort. For the data for the 1986 to 1995 cohort, like the graph for the cumulative incidence of KD, the prevalence of ECG abnormalities at ages 6 and 12 years in the birth-year cohorts has been increasing. In the graphs for the prevalence of ECG abnormalities at age 6 and the cumulative incidence of KD after the 1996 cohort, the patterns are different. However, they both showed a tendency to increase.

In comparison with the graph for the cumulative incidence of acute cardiac lesions at age 6 (according to data from the nationwide survey), the prevalence of ECG abnormalities found during school cardiac examinations exhibited changes similar to those for cohorts dated 1997 or later. This similarity is noteworthy. As stated before, for those cohorts for 1996 or earlier, the method for calculating the cumulative morbidity of acute cardiac lesions was modified because of a change in the questions posed in the nationwide survey. KD patients who would have been classified with cardiac sequelae in the previous survey were now classified as having acute cardiac lesions. Therefore, the numbers for the cumulative incidence of acute-stage lesions before the 1996 cohort should be regarded as provisional. After 1997, however, the figures for the cumulative incidence included only those patients with

acute-stage cardiac lesions due to KD, based on results after the 15th survey. Therefore, we may safely assume that patterns are similar for the prevalence of ECG abnormalities detected at school health examinations and the cumulative incidence of acute cardiac lesions obtained from nationwide surveys.

The cumulative incidence of late-stage cardiac sequelae has continued to decline, which differs from the secular trends of the 2 abovementioned graphs.

The current study was limited to a comparison of the cumulative incidence of KD among birth-year cohorts obtained from nationwide surveys and the prevalence of ECG abnormalities calculated from the results of the Statistical Survey on School Health. This study is not analytic; rather, it uses a descriptive approach in which statistical tests are not used in comparing the cumulative incidence of KD with the prevalence of ECG abnormalities. In addition, the nationwide surveys of KD were based on the general population, not selected samples.

We have come to realize that a case-control study between a group of KD patients and a normal group is needed. Hirata et al investigated students who had a cardiac examination during their first year of junior high school, at age 12 years, in Tochigi Prefecture.⁹ In their comparison of children with a history of KD and normal individuals, the prevalence of ECG abnormalities was significantly higher in the former. They also noted that 57% of ECG abnormalities in the former were due to an incomplete right bundle-branch block and right axial deviation. The prevalence of ECG abnormalities was significantly higher in the KD patients, a finding which parallels the data presented in this study. Naturally, there are many causes for ECG abnormalities: they are not limited to KD, but may also be caused by ischemic disease or myocarditis. However, because the sample size in nationwide surveys of KD is large, it is likely that the similarity in the secular trend of the 2 graphs indicates that there is some relationship between them.

Asai et al³ investigated the clinical course of KD and found that ECG abnormalities occur at a high rate among patients with KD. In the nationwide surveys, those patients with acute-stage cardiac lesions or cardiac sequelae are equivalent to those who developed coronary artery aneurysm or giant coronary aneurysm, coronary dilatation, coronary stenosis, valvular dysfunction, or myocardial infarction, all of which were due to KD. Therefore, there were more patients with a temporary ECG abnormality than there were KD patients with acute-stage cardiac lesions in those surveys. For example, in the 2001 cohort at age 6 years the cumulative incidence of KD was 713 per 100 000 births and that of acute-stage cardiac lesions was 102.2. The prevalence of ECG abnormalities was 2490 per 100 000 births. The prevalence of congenital heart disease in Japan was reported to be 0.8% to 1.0% per live birth.^{10,11} The cumulative incidence of KD is up to one half that of ECG abnormalities acquired after birth. Thus,

particular attention should be paid to KD patients with temporary ECG abnormalities.

Before embarking on a case-control study, it would be useful to utilize the national survey to ascertain if there is a correlation between the cumulative incidence of KD-related pathologies in each birth-year cohort and the morbidities from other diseases stratified by age.

The first report on KD patients was published approximately 48 years ago. Thus, a number of KD patients have reached adulthood. They consult with internists rather than pediatricians. Therefore, important statistical information on KD should also be provided to general practitioners.

In the current study, fluctuations in the graph for the prevalence of ECG abnormalities at age 6 closely resembled changes in the cumulative morbidity of acute-stage cardiac lesions after 1997. Moreover, the secular trend seen in the graph for the prevalence of ECG abnormalities at age 12 was consistent with the trend for KD. This suggests that KD has a sustained impact on the cardiovascular system, not only in those few KD patients with cardiac sequelae but also in those without evident cardiac dysfunction.

Long-term careful follow-up of KD patients is necessary, not only in those with cardiac sequelae, but also in those who developed only acute-stage cardiac lesions and no cardiac sequelae.

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第20回川崎病全国調査成績

川崎病全国調査担当グループ（特定非営利活動法人日本川崎病研究センター）

やしろまゆみ なかむらよしかず うえはらりてい やながわ ひろし
屋代真弓 中村好一 上原里程 柳川 洋

はじめに

1970年以来、2年に1回の間隔で19回にわたって、川崎病全国調査が行われてきた^{1)~33)}。今回、2007～2008年の2年間の患者を対象に実施した第20回川崎病全国調査の成績がまとまった。

2年間の調査成績より、報告患者数、初診年月分布、性・年齢分布、地域分布、診断、家族歴、再発例、死亡例、急性期の心障害例、後遺症例、初診時病日、 γ -グロブリン治療開始時病日、容疑例主要症状の数、 γ -グロブリン治療、追加治療（ γ -グロブリン投与、ステロイド投与、インフリキシマブ投与、免疫抑制薬投与）、心臓以外の合併症などの疫学像ならびに臨床像を明らかにしたので、これまでに得られた36年間の調査成績と比較しながら、その概要を報告する。

方法

第20回川崎病全国調査は、2007年1月1日～2008年12月31日の2年間に小児科を標榜する100床以上の病院、および小児科のみを標榜する100床未満の専門病院を受診した川崎病初診患者を対象に、郵送（一部、インターネットサーベイランス参加の施設にはメールでも依頼）により実施した。

施設の選定は、前回使用した医療機関のリストに、その後、現在までの変更を更新したものをを用いた。対象候補施設数は2,150カ所であっ

た。

本調査は、自治医科大学疫学研究倫理審査委員会の承認を得て実施した（2008年11月11日、疫08-39）。

調査結果

1. 回収率

依頼状、調査票などを送付した2,150施設のうち、廃院などの連絡があった48施設を除く2,102施設を調査対象とした。回答は1,540施設から得られ、回収率は73.3%であった。そのうち、ホームページより調査票をダウンロードしてExcelファイルで回答した施設が95カ所、インターネットサーベイランス参加施設で登録済みの還元ファイルを使用して回答した施設が50カ所であった。なお、サーベイランス参加施設でも調査票郵送により協力したところもある。

回答があった1,540施設のうち、患者報告があった施設は972施設（回収施設の63.1%）であった。回収率は、都道府県によって52.0～87.5%の開きがみられた。

2. 年次推移

今回の調査で報告された2年間の患者数は、2007年11,581人、2008年11,756人の合せて23,337人であり、前回の調査と合せると4年連続で1万人以上の患者が報告された。

性別患者数は男13,523人、女9,814人で、2年間平均の罹患率は0～4歳人口10万対216.9（男245.4、女187.0）であった。

患者数の性比（男/女）は1.38、罹患率の性比

表1 性別患者数，罹患率，死亡数，致命率の推移

年次	患者数			0～4歳人口10万対年間罹患率*			死亡数 (致命率%)
	計	男	女	計	男	女	
～1964	88	58	30	1.1	1.4	0.8	—
1965	61	33	28	0.8	0.8	0.7	—
1966	79	49	30	1.0	1.2	0.8	—
1967	101	60	41	1.2	1.4	1.0	2 (1.98)
1968	310	177	133	3.7	4.1	3.2	6 (1.94)
1969	461	281	180	5.3	6.3	4.3	9 (1.95)
1970	887	527	360	10.1	11.8	8.4	10 (1.13)
1971	804	480	324	8.7	10.1	7.1	10 (1.24)
1972	1,135	658	477	12.0	13.5	10.4	16 (1.41)
1973	1,524	928	596	15.6	18.4	12.5	34 (2.23)
1974	1,963	1,157	806	19.7	22.6	16.7	20 (1.02)
1975	2,216	1,332	884	22.3	26.1	18.3	16 (0.72)
1976	2,337	1,406	931	23.9	28.0	19.6	15 (0.64)
1977	2,798	1,706	1,092	29.3	34.8	23.5	17 (0.61)
1978	3,459	2,064	1,395	37.7	43.7	31.2	14 (0.40)
1979	6,867	3,987	2,880	78.0	88.1	67.3	34 (0.50)
1980	3,932	2,317	1,615	46.5	53.4	39.2	8 (0.20)
1981	6,383	3,677	2,706	78.3	87.9	68.2	16 (0.25)
1982	15,519	8,762	6,757	196.1	215.8	175.4	46 (0.30)
1983	5,961	3,441	2,520	77.3	86.9	67.1	15 (0.25)
1984	6,514	3,790	2,724	86.0	97.5	73.9	17 (0.26)
1985	7,611	4,430	3,181	102.1	116.4	87.1	10 (0.13)
1986	12,847	7,250	5,597	176.8	194.7	157.9	18 (0.14)
1987	5,256	3,066	2,190	73.8	84.0	63.1	9 (0.17)
1988	5,217	3,056	2,161	75.3	86.0	64.1	4 (0.08)
1989	5,591	3,251	2,340	83.6	94.7	71.9	8 (0.14)
1990	5,706	3,268	2,438	88.1	98.4	77.3	12 (0.21)
1991	5,677	3,354	2,323	90.1	103.8	75.7	7 (0.12)
1992	5,544	3,250	2,294	89.9	102.8	76.4	2 (0.04)
1993	5,389	3,155	2,234	89.1	101.6	75.9	11 (0.20)
1994	6,069	3,574	2,495	101.1	115.9	85.4	2 (0.03)
1995	6,107	3,548	2,559	102.6	116.4	88.2	6 (0.09)
1996	6,424	3,691	2,733	108.4	121.6	94.6	4 (0.06)
1997	6,373	3,690	2,683	108.0	122.0	93.2	9 (0.14)
1998	6,593	3,799	2,794	111.5	125.3	96.9	2 (0.03)
1999	7,047	4,102	2,945	119.6	135.8	102.6	3 (0.04)
2000	8,267	4,758	3,509	141.1	158.5	122.8	5 (0.06)
2001	8,113	4,588	3,525	138.8	153.2	123.7	0 (—)
2002	8,839	5,156	3,683	151.9	172.8	130.0	2 (0.02)
2003	9,146	5,281	3,865	159.2	179.2	138.2	4 (0.04)
2004	9,992	5,778	4,214	175.9	198.3	152.4	4 (0.04)
2005	10,041	5,868	4,173	181.0	206.5	154.2	1 (0.01)
2006	10,434	6,024	4,410	191.4	215.8	165.9	1 (0.01)
2007	11,581	6,684	4,897	215.3	242.6	186.6	2 (0.02)
2008	11,756	6,839	4,917	218.6	248.2	187.4	4 (0.03)
計	249,019	144,350	104,669	—	—	—	435 (0.17)

*：罹患率の計算には人口動態統計の分母に用いる日本人口（5年ごとの国勢調査人口および各年次の推計人口で、人口動態統計に掲載されているもの。ただし、2008年は2007年の推計人口）を用いた。前回調査の2006年は2005年の人口を用いたので今回2006年の人口で修正した

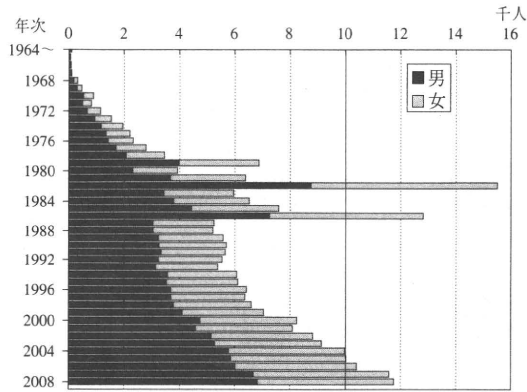


図1 年次別, 性別患者数

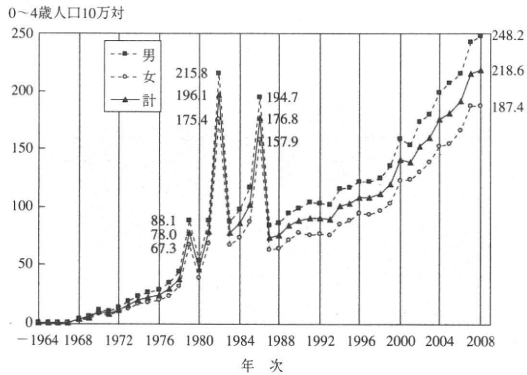


図2 年次別, 性別罹患率

は1.31で男のほうが多かった。過去19回に報告された患者を含めると、2008年12月末までの患者数は合計249,019人（男144,350人、女104,669人）になった。

川崎病患者数の年次推移は、表1、図1に示すとおりである。1979年、1982年、1986年の3回にわたる全国規模の流行がみられた。その後、年次とともに増加傾向が続き、2000年の患者数は第1回目の流行年（1979年）の1.2倍となり、2005年には10,000人を超え、さらに2007年には11,000人を超え、近年になって急勾配で増加している。

罹患率の年次推移をみると、表1、図2に示すように、さらに近年の増加傾向が目立ち、0～4歳人口10万対罹患率は2007年は215.3、2008年は218.6であり、両年ともに史上最高値となっ

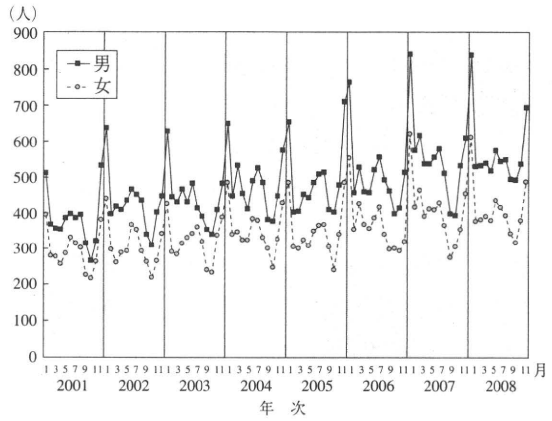


図3 年次別, 月別, 性別患者数

た。

3. 月別推移

最近8年間の月別、性別患者数を図3に示す。患者数は各年とも同じような季節変動を示し、すべての月で男が多くなっている。また、10月は少なく、1月は多かった。とくに、2006年からの3年間の1月は以前に比べて極端に多くなっていた。また、夏にも若干の患者数の増加が観察された。

4. 性・年齢分布

患者数の性・年齢別分布をみると、3歳未満の者の割合は全体の65.9%（男68.7%、女64.1%）であった（表2）。2007～2008年平均の性・年齢別罹患率では、男女とも月齢9～11カ月にピーク（人口10万対男449.4、女305.5）をもつ一峰性の山（女は月齢12～14カ月にやや減少）がみられた。前回実施した2005～2006年の成績では月齢6～8カ月にピークがみられ、高齢へのわずかなずれがみられた。罹患率の性比は、月齢12～14カ月の者でもっとも大きく1.56であり、月齢0～2カ月の者でもっとも小さく1.02であった（図4）。

5. 地域分布

2年間の患者住所都道府県別報告数をもっとも多かったのは東京で2,358人、次いで神奈川県1,921人、愛知1,706人、大阪1,620人の順であっ

た。2007年、2008年の年次別都道府県別罹患率を0～4歳人口10万対の割合で計算した（都道府県別罹患率の分母は2005年国勢調査人口を使用し、全国の罹患率は各年次の推定人口を用いて計算した。ただし2008年は前年の人口を使用）。両年とも罹患率が高いところは、栃木、東京、神奈川などであった。2007年の罹患率が高いところは、和歌山、徳島、群馬などである。2008年の罹患率が高いところは、福井、奈良、熊本などであり、局地的に患者数の増加があったと考えられる。2年間とも低いところは岩手、沖縄、長崎などであった（表3）。

2005～2008年の各年について、都道府県別罹

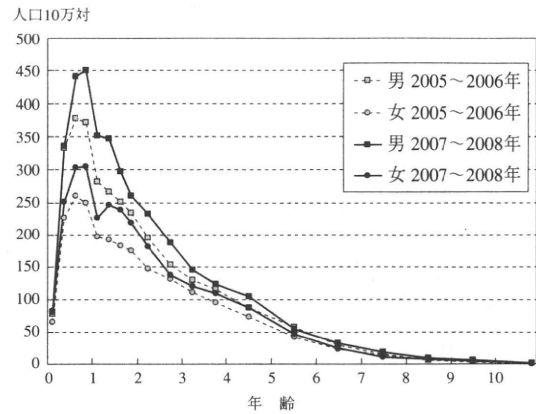


図4 年次別、性別、年齢別罹患率 (2005～2006年平均、2007～2008年平均)

表2 年齢別、年次別、性別患者数および罹患率（人口10万対）

年齢	患者数											
	総数				2007年				2008年			
	総数	罹患率*1	男	女	総数	罹患率*1	男	女	総数	罹患率*1	男	女
総数*2	23,337	216.9	13,523	9,814	11,581	215.3	6,684	4,897	11,756	218.6	6,839	4,917
0～2カ月	439	81.1	223	216	221	81.7	112	109	218	80.6	111	107
3～5カ月	1,594	294.6	934	660	772	285.4	447	325	822	303.9	487	335
6～8カ月	2,023	373.9	1,222	801	983	363.4	603	380	1,040	384.5	619	421
9～11カ月	2,052	379.3	1,247	805	1,024	378.6	615	409	1,028	380.0	632	396
12～14カ月	1,550	290.8	963	587	757	284.1	463	294	793	297.6	500	293
15～17カ月	1,588	297.9	949	639	748	280.7	455	293	840	315.2	494	346
18～20カ月	1,430	268.3	812	618	727	272.8	412	315	703	263.8	400	303
21～23カ月	1,277	239.6	713	564	646	242.4	360	286	631	236.8	353	278
2歳～	2,153	206.8	1,231	922	1,094	210.2	620	474	1,059	203.5	611	448
2歳6カ月～	1,689	162.2	991	698	861	165.4	509	352	828	159.1	482	346
3歳～	1,440	133.0	806	634	729	134.6	412	317	711	131.3	394	317
3歳6カ月～	1,257	116.1	684	573	656	121.1	355	301	601	111.0	329	272
4歳～	2,135	96.3	1,185	950	1,037	93.6	569	468	1,098	99.1	616	482
5歳～	1,186	50.9	659	527	558	47.9	298	260	628	53.9	361	267
6歳～	681	29.2	389	292	330	28.3	187	143	351	30.1	202	149
7歳～	358	15.4	231	127	182	15.6	122	60	176	15.1	109	67
8歳～	190	8.2	106	84	103	8.8	52	51	87	7.5	54	33
9歳～	118	5.1	69	49	62	5.3	33	29	56	4.8	36	20
10歳以上	177	1.5	109	68	91	1.5	60	31	86	1.4	49	37

*1：罹患率の計算には2007年人口動態統計の分母に用いる日本人人口を用いた

*2：総数の罹患率の計算には0～4歳日本人人口を用いた

表3 患者住所都道府県別, 年次別, 性別患者数および罹患率 (0~4歳人口10万対)

	2005年				2006年				2007年				2008年			
	患者数			罹患率*	患者数			罹患率*	患者数			罹患率*	患者数			罹患率*
	総数	男	女		総数	男	女		総数	男	女		総数	男	女	
全国*2	10,041	5,868	4,173	181.0	10,434	6,024	4,410	191.4	11,581	6,684	4,897	215.3	11,756	6,839	4,917	218.6
1:北海道	392	230	162	177.4	385	239	146	174.2	488	285	203	220.8	461	257	204	208.6
2:青森	112	69	43	193.1	89	53	36	153.4	92	42	50	158.6	99	62	37	170.7
3:岩手	43	27	16	75.4	47	26	21	82.5	47	26	21	82.5	74	48	26	129.8
4:宮城	89	56	33	87.3	132	86	46	129.4	201	119	82	197.1	201	115	86	197.1
5:秋田	58	30	28	141.5	55	27	28	134.1	90	49	41	219.5	70	42	28	170.7
6:山形	110	70	40	215.7	91	56	35	178.4	117	56	61	229.4	118	74	44	231.4
7:福島	148	81	67	159.1	187	109	78	201.1	214	113	101	230.1	229	115	114	246.2
8:茨城	225	133	92	170.5	240	127	113	181.8	217	124	93	164.4	226	127	99	171.2
9:栃木	177	106	71	194.5	200	133	67	219.8	226	145	81	248.4	229	151	78	251.6
10:群馬	167	100	67	181.5	180	112	68	195.7	237	134	103	257.6	214	125	89	232.6
11:埼玉	505	253	252	160.3	464	267	197	147.3	531	318	213	168.6	532	309	223	168.9
12:千葉	597	369	228	225.3	622	360	262	234.7	613	357	256	231.3	628	338	290	237.0
13:東京	956	549	407	200.4	983	533	450	206.1	1,184	686	498	248.2	1,174	702	472	246.1
14:神奈川	883	520	363	224.1	908	531	377	230.5	949	549	400	240.9	972	568	404	246.7
15:新潟	150	100	50	151.5	148	80	68	149.5	194	116	78	196.0	178	106	72	179.8
16:富山	47	33	14	97.9	52	30	22	108.3	59	34	25	122.9	75	44	31	156.3
17:石川	147	90	57	277.4	134	83	51	252.8	112	69	43	211.3	100	51	49	188.7
18:福井	84	47	37	227.0	96	61	35	259.5	76	47	29	205.4	104	57	47	281.1
19:山梨	44	28	16	115.8	67	37	30	176.3	54	32	22	142.1	48	30	18	126.3
20:長野	189	111	78	189.0	208	118	90	208.0	225	127	98	225.0	244	134	110	244.0
21:岐阜	150	87	63	156.3	146	88	58	152.1	197	105	92	205.2	183	99	84	190.6
22:静岡	272	165	107	157.2	353	172	181	204.0	341	204	137	197.1	325	188	137	187.9
23:愛知	639	356	283	180.5	723	432	291	204.2	878	493	385	248.0	828	484	344	233.9
24:三重	152	90	62	181.0	142	79	63	169.0	201	105	96	239.3	168	106	62	200.0
25:滋賀	145	75	70	210.1	114	65	49	165.2	97	55	42	140.6	95	59	36	137.7
26:京都	227	129	98	202.7	213	118	95	190.2	254	159	95	226.8	222	128	94	198.2
27:大阪	786	460	326	199.0	779	454	325	197.2	796	457	339	201.5	824	479	345	208.6
28:兵庫	536	312	224	211.9	525	297	228	207.5	545	318	227	215.4	603	368	235	238.3
29:奈良	108	61	47	180.0	106	64	42	176.7	131	74	57	218.3	161	83	78	268.3
30:和歌山	111	71	40	264.3	78	48	30	185.7	118	68	50	281.0	93	55	38	221.4
31:鳥取	27	14	13	103.8	44	27	17	169.2	47	30	17	180.8	39	22	17	150.0
32:島根	41	24	17	132.3	35	21	14	112.9	55	31	24	177.4	55	33	22	177.4
33:岡山	124	73	51	140.9	156	85	71	177.3	126	79	47	143.2	134	78	56	152.3
34:広島	237	147	90	183.7	250	141	109	193.8	264	151	113	204.7	304	176	128	235.7
35:山口	98	63	35	160.7	96	54	42	157.4	99	57	42	162.3	127	68	59	208.2
36:徳島	44	25	19	137.5	58	42	16	181.3	86	47	39	268.8	71	40	31	221.9
37:香川	77	48	29	171.1	68	42	26	151.1	74	48	26	164.4	107	71	36	237.8
38:愛媛	118	70	48	193.4	136	67	69	223.0	147	82	65	241.0	129	75	54	211.5
39:高知	47	24	23	151.6	62	38	24	200.0	51	28	23	164.5	68	33	35	219.4
40:福岡	328	187	141	147.1	366	221	145	164.1	477	275	202	213.9	472	277	195	211.7
41:佐賀	40	27	13	100.0	48	28	20	120.0	65	40	25	162.5	70	47	23	175.0
42:長崎	76	44	32	134.4	104	65	39	162.5	88	56	32	137.5	83	47	36	129.7
43:熊本	150	92	58	185.2	148	88	60	182.7	177	100	77	218.5	211	131	80	260.5
44:大分	124	71	53	243.1	137	78	59	268.6	97	58	39	190.2	108	62	46	211.8
45:宮崎	50	32	18	98.0	52	29	23	102.0	45	24	21	88.2	73	50	23	143.1
46:鹿児島	114	62	52	148.1	101	57	44	131.2	110	65	45	142.9	140	78	62	181.8
47:沖縄	95	57	38	115.9	104	55	49	126.8	87	46	41	106.1	86	46	40	104.9
48:国外	2	0	2	-	2	1	1	-	2	1	1	-	1	1	0	-

*1: 都道府県別罹患率は2005年国勢調査人口を用いて計算した

*2: 全国の罹患率は各年次の推計人口を用いて計算した (ただし2008年は2007年の推計人口を使用)

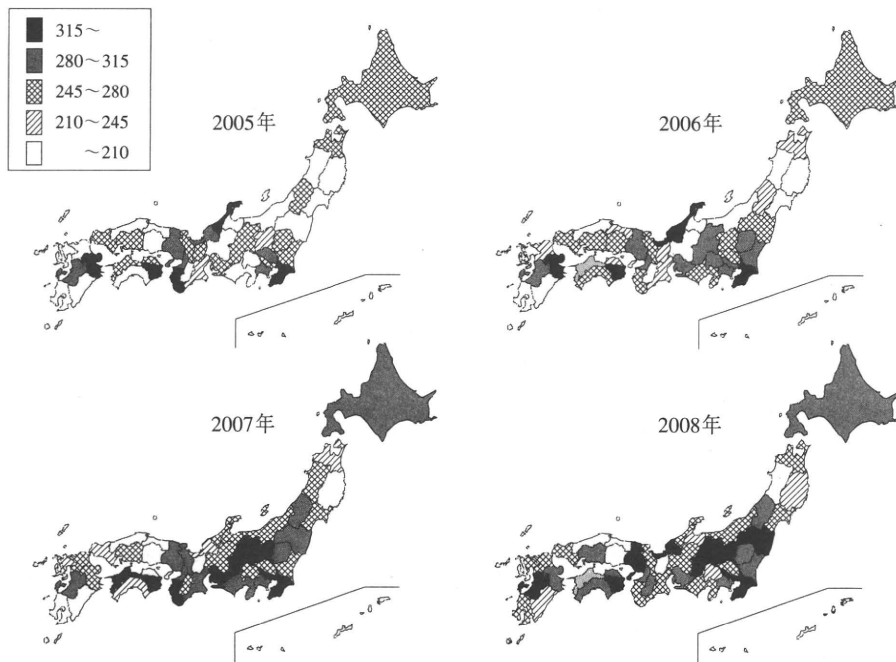


図5 年次別、都道府県別罹患率

罹患率の地図を作成した(図5)。都道府県によって回収率が異なるので、未回収施設も同じ患者数があると仮定して回収率を100%に補正し、0～4歳人口10万対罹患率の地域差を示した。2005年に罹患率の高い地域は、和歌山、大分、石川と関東、近畿、九州の一部の県であったが、2006年にはそれらの周辺地域に拡がり、関東甲信越では罹患率の上昇が目立った。2007年には、関東甲信越のみでなく近畿、四国でも罹患率の上昇がみられ、東北、北海道でも高い罹患率が観察されるようになった。この傾向は2008年にはより顕著になり、さらに中国、四国、九州南部に高率地域が拡がった。

6. 診断

診断基準への一致度をみると、定型例79.8% (男79.8%, 女79.8%), 不定型例2.8% (男3.1%, 女2.4%), 容疑例17.4% (男17.1%, 女17.8%)であった。前回より定型例、不定型例が減少し、容疑例が増加した。年齢別にみると、2歳未満の若年齢では容疑例の割合が比較的高く、また

5歳以上の年長児でもその割合が高かった(表4)。

なお、定型例(調査票では「确实A」とした)は「川崎病診断の手引き 改訂版」(2002年2月に診断の手引きが改訂され、第17回川崎病全国調査から改訂5版を使用)に示された六つの主要症状のうち五つ以上の症状を伴う者、不定型例(「确实B」)は四つの症状しか認められなくても、経過中に断層心エコー法もしくは心血管造影法で冠動脈瘤(いわゆる拡大を含む)が確認され、他の疾患が除外された者をいう。また、容疑例(「容疑」)は上記のいずれにも合致しないが、主治医が川崎病の疑いありと診断して全国調査に報告した者をいう。

今回の調査では、容疑例について主要症状の数を調査項目に加えたが、その結果、主要症状は四つがもっとも多く65.4%、次いで三つ26.1%、二つ5.9%、一つ0.8%、不明1.8%であった。性別にみても同様の割合であった。年齢別には、10歳以上を除き高年齢になるほど三つ以上の症

表4 年齢別，性別診断の確実度

		総数 (%)	定型例 (%)	不定型例 (%)	容疑例 (%)
総数		23,337 (100)	18,620 (79.8)	648 (2.8)	4,069 (17.4)
性別	男	13,523 (100)	10,791 (79.8)	413 (3.1)	2,319 (17.1)
	女	9,814 (100)	7,829 (79.8)	235 (2.4)	1,750 (17.8)
年齢別	0～5カ月	2,033 (100)	1,402 (69.0)	80 (3.9)	551 (27.1)
	6～11カ月	4,075 (100)	3,009 (73.8)	110 (2.7)	956 (23.5)
	1歳～	5,845 (100)	4,641 (79.4)	167 (2.9)	1,037 (17.7)
	2歳～	3,842 (100)	3,272 (85.2)	89 (2.3)	481 (12.5)
	3歳～	2,697 (100)	2,317 (85.9)	51 (1.9)	329 (12.2)
	4歳～	2,135 (100)	1,814 (85.0)	54 (2.5)	267 (12.5)
	5歳～	1,186 (100)	988 (83.3)	35 (3.0)	163 (13.7)
	6歳～	681 (100)	549 (80.6)	26 (3.8)	106 (15.6)
	7歳～	358 (100)	285 (79.6)	9 (2.5)	64 (17.9)
	8歳～	190 (100)	139 (73.2)	9 (4.7)	42 (22.1)
	9歳～	118 (100)	93 (78.8)	4 (3.4)	21 (17.8)
	10歳以上	177 (100)	111 (62.7)	14 (7.9)	52 (29.4)

表5 性別，年齢別容疑例の主要症状の数

		患者数 (容疑例) (%)	主要症状の数 (%)				
			1個	2個	3個	4個	不明
総数		4,069 (100)	32 (0.8)	239 (5.9)	1,063 (26.1)	2,661 (65.4)	74 (1.8)
性別	男	2,319 (100)	22 (0.9)	134 (5.8)	622 (26.8)	1,499 (64.6)	42 (1.8)
	女	1,750 (100)	10 (0.6)	105 (6.0)	441 (25.2)	1,162 (66.4)	32 (1.8)
年齢別	0～5カ月	551 (100)	13 (2.4)	49 (8.9)	150 (27.2)	331 (60.1)	8 (1.5)
	6～11カ月	956 (100)	5 (0.5)	71 (7.4)	286 (29.9)	583 (61.0)	11 (1.2)
	1歳～	1,037 (100)	9 (0.9)	60 (5.8)	245 (23.6)	700 (67.5)	23 (2.2)
	2歳～	481 (100)	1 (0.2)	20 (4.2)	118 (24.5)	328 (68.2)	14 (2.9)
	3歳～	329 (100)	1 (0.3)	14 (4.3)	87 (26.4)	223 (67.8)	4 (1.2)
	4歳～	267 (100)	0 (—)	13 (4.9)	73 (27.3)	174 (65.2)	7 (2.6)
	5歳～	163 (100)	1 (0.6)	5 (3.1)	39 (23.9)	116 (71.2)	2 (1.2)
	6歳～	106 (100)	0 (—)	1 (0.9)	27 (25.5)	77 (72.6)	1 (0.9)
	7歳～	64 (100)	0 (—)	3 (4.7)	8 (12.5)	52 (81.3)	1 (1.6)
	8歳～	42 (100)	0 (—)	2 (4.8)	9 (21.4)	30 (71.4)	1 (2.4)
	9歳～	21 (100)	0 (—)	0 (—)	6 (28.6)	15 (71.4)	0 (—)
	10歳以上	52 (100)	2 (3.8)	1 (1.9)	15 (28.8)	32 (61.5)	2 (3.8)

表6 性別、初診時年齢別、診断別死亡例の割合

総数		総数	死亡例	(%)
		23,337	6	0.03
性別	男	13,523	5	0.04
	女	9,814	1	0.01
年齢別 初診時	0～5カ月	2,033	4	0.20
	6～11カ月	4,075	1	0.02
	1歳	5,845	1	0.02
診断別	定型例	18,620	5	0.03
	不定型例	648	0	—
	容疑例	4,069	1	0.02

状をもつ患者の割合が高かった(表5)。

7. 家族歴

同胞例ありの割合は報告患者中1.4%(男1.4%, 女1.5%)であった。

両親のいずれかに川崎病の既往歴ありは165人(男103人, 女62人)報告され, 報告患者中0.7%(男0.8%, 女0.6%)であった。既往歴を有する両親の内訳は父76人, 母85人, 不明4人であった。これらは前回報告[102人(父45人, 母55人)]よりも多かった。

8. 再発例

再発例の割合は, 報告患者中3.5%(男3.6%, 女3.4%)であった。

性・年齢別にみると, 男女とも5歳までは年齢とともに再発患者の割合が増加していた。

9. 死亡例

死亡例は2年間で6人(男5人, 女1人)報告され, 致命率は0.03%であった。死亡例の初診時年齢は4人が6カ月未満の若年児で, そのうち3人が発病後1カ月以内の急性期の死亡であった。診断別には5人が定型例であった(表6)。

10. 心障害例

心障害については, 発病後1カ月以内に出現した「急性期」と1カ月以降も残存する「後遺症」に分けて調査を実施した。

報告患者中, 急性期の割合は11.0%(男

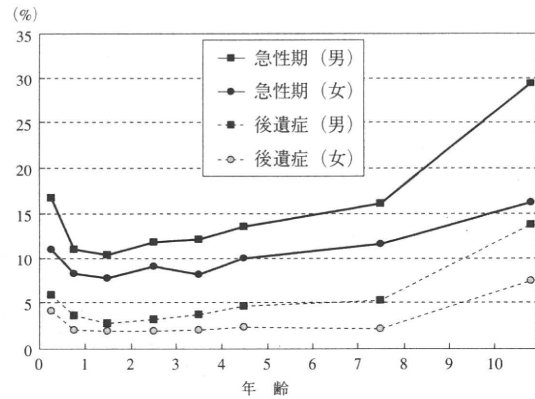


図6 性別、年齢別心障害の出現率

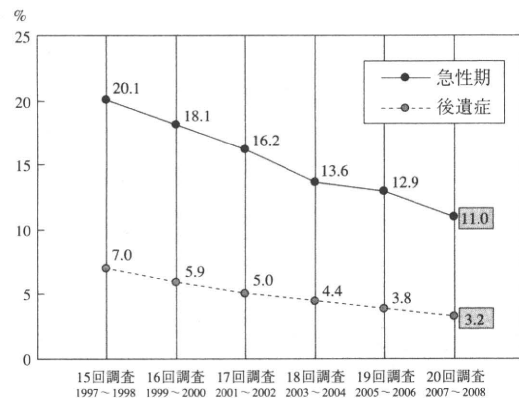


図7 心障害の出現割合の年次推移

12.5%, 女9.0%), 後遺症の割合は3.2%(男3.9%, 女2.3%)であり, 急性期の割合は前回よりも減少した。今回も後遺症は急性期に比べて男女とも1/3以下に減少していた。両者とも男が高率を示し, 性・年齢別にみると男女とも若年児と高年児が高く, 急性期, 後遺症ともに1歳に窪みをもつゆるやかなU字型のカーブを示していた(図6)。どちらの割合も, 第15回調査(1997～1998年)から第20回調査(2007～2008年)の間に半分ちかくに減少していた(図7)。

報告患者に占める急性期の心障害種類別割合は, 冠動脈の拡大8.54%, 弁膜病変1.64%, 瘤1.21%, 巨大瘤0.25%, 狭窄0.03%, 心筋梗塞0.01%であった。前回と比べて, 狭窄以外はいずれも減少した。性別にみると, 弁膜病変以外

表7 種類別, 性別, 年齢別心障害の出現率

		総数 (%)	巨大瘤 (%)	瘤 (%)	拡大 (%)	狭窄 (%)	心筋梗塞 (%)	弁膜病変 (%)	
急性期	総数	23,337 (100)	58 (0.25)	282 (1.21)	1,992 (8.54)	8 (0.03)	3 (0.01)	383 (1.64)	
	性別	男	13,523 (100)	44 (0.33)	203 (1.50)	1,331 (9.84)	5 (0.04)	3 (0.02)	212 (1.57)
		女	9,814 (100)	14 (0.14)	79 (0.80)	661 (6.74)	3 (0.03)	0 (-)	171 (1.74)
	年齢別	2歳未満	11,953 (100)	24 (0.20)	147 (1.23)	969 (8.11)	2 (0.02)	3 (0.03)	164 (1.37)
		2歳以上	11,384 (100)	34 (0.30)	135 (1.19)	1,023 (8.99)	6 (0.05)	0 (-)	219 (1.92)
後遺症	総数	23,337 (100)	59 (0.25)	188 (0.81)	435 (1.86)	5 (0.02)	2 (0.01)	114 (0.49)	
	性別	男	13,523 (100)	48 (0.35)	138 (1.02)	307 (2.27)	4 (0.03)	2 (0.01)	68 (0.50)
		女	9,854 (100)	11 (0.11)	50 (0.51)	128 (1.30)	1 (0.01)	0 (-)	46 (0.47)
	年齢別	2歳未満	11,953 (100)	23 (0.19)	91 (0.76)	233 (1.95)	1 (0.01)	1 (0.01)	47 (0.39)
		2歳以上	11,384 (100)	36 (0.32)	97 (0.85)	202 (1.77)	4 (0.04)	1 (0.01)	67 (0.59)

はすべて男で高かった。出現率を2歳未満と2歳以上の2区分に分けてみると、巨大瘤、冠動脈の拡大、狭窄、弁膜病変の出現率は2歳以上で高率にみられた。

報告患者に占める後遺症の種類別割合は、冠動脈の拡大1.86%、瘤0.81%、弁膜病変0.49%、巨大瘤0.25%、狭窄0.02%、心筋梗塞0.01%であった。性別にみると、すべて男で高かった。とくに巨大瘤は、男が女の3.2倍の出現率であった。出現率を2歳未満と2歳以上の2区分に分けてみると、巨大瘤、瘤、狭窄、弁膜病変の出現率は2歳以上で高率にみられた。とくに巨大瘤は、2歳以上が2歳未満の1.7倍の出現率であった。

心障害の種類別の観察では、瘤、冠動脈の拡大、狭窄、弁膜病変は、急性期に比べて後遺症では減少していたが、巨大瘤では減少がみられなかった(表7)。

11. 初診時病日および γ -グロブリン治療開始時病日

患者の初診日は第4病日をもっとも多く24.6%であり、第4病日までに受診した者は67.0%であった。2歳未満と2歳以上の2区分に分けてみると、第4病日までに受診した者は2歳未満では71.4%を占めていたが、2歳以上では62.4%であり、2歳以上の年長児の受診が遅れる傾向がみ

られた。

γ -グロブリンの投与開始日は第5病日をもっとも多く37.5%であった。年齢別にみると、2歳未満に早く投与を開始する傾向がみられ、第5病日までに投与を開始された者の割合は2歳未満では69.3%、2歳以上では59.6%であった(表8)。

12. γ -グロブリン治療

γ -グロブリン治療を受けた者は87.0%(男87.4%、女86.6%)であった。

γ -グロブリン1日あたりの投与量は、1,900~2,099 mg/kgの者がもっとも多く77.0%、次いで900~1,099 mg/kgが19.7%であった。投与期間は1日をもっとも多く87.5%、次いで2日11.6%であった。 γ -グロブリンの1日投与量と使用日数から計算した使用総量は、1,900~2,099 mg/kgがもっとも多く88.5%、次いで900~1,099 mg/kgが8.5%、1,700~1,899 mg/kg、2,100 mg/kg+がともに1.1%であった。前回に比べてさらに2,000 mg/kg×1日の短期間大量投与が増加し、治療を受けた者の9割近くを占めていた(表9, 図8)。

13. 追加治療

追加治療(γ -グロブリン投与)の割合は、初回 γ -グロブリン使用例のうち16.5%であった。診断別では定型例が多く、男女別では男で多

かった。2歳未満と2歳以上の2区分に分けてみると、2歳以上が多かった。

追加治療（ステロイド投与）の割合は全体の5.0%であった。診断別では不定型例がやや多く、性別では男が多かった。2歳未満と2歳以上の2区分に分けてみると、2歳以上が多かった。

追加治療（インフリキシマブ投与）の割合は全体の0.4%であった。診断別では不定型例がやや多く、性別では男が多かった。2歳未満と2歳以上の2区分に分けてみると、2歳以上が多かった。

追加治療（免疫抑制薬投与）の割合は全体の

0.2%であった。診断別では定型例が多く、性別では男が多かった。2歳未満と2歳以上の2区分に分けてみると、2歳未満が多かった（表10）。

14. 心臓以外の合併症

今回の調査ではじめて心臓以外の合併症を調べた結果、肝障害（入院時のASTとALTがともに50 IU/L以上）27.4%，胆嚢腫大1.6%，関節痛または関節炎1.1%，無菌性髄膜炎0.55%，麻痺性イレウス0.45%，播種性血管内凝固（DIC）0.081%，顔面神経麻痺0.0043%であった。

診断別では、関節痛または関節炎、無菌性髄

表8 年齢別、初診時およびγ-グロブリン投与開始時病日の分布

		総数 (%)	2歳未満 (%)	2歳以上 (%)
*1 初診時	総数	23,332 (100)	11,949 (100)	11,383 (100)
	第1病日	1,151 (4.9)	763 (6.4)	388 (3.4)
	第2病日	3,478 (14.9)	2,094 (17.5)	1,384 (12.2)
	第3病日	5,261 (22.5)	2,770 (23.2)	2,491 (21.9)
	第4病日	5,741 (24.6)	2,899 (24.3)	2,842 (25.0)
	第5病日	4,057 (17.4)	1,920 (16.1)	2,137 (18.8)
	第6病日	1,999 (8.6)	861 (7.2)	1,138 (10.0)
	第7病日	819 (3.5)	320 (2.7)	499 (4.4)
	第8病日	373 (1.6)	140 (1.2)	233 (2.0)
	第9病日	184 (0.8)	56 (0.5)	128 (1.1)
	第10病日以上	269 (1.2)	126 (1.1)	143 (1.3)
*2 γ-グロブリン投与時	総数	20,290 (100)	10,238 (100)	10,052 (100)
	第1病日	12 (0.1)	7 (0.1)	5 (0.0)
	第2病日	188 (0.9)	130 (1.3)	58 (0.6)
	第3病日	1,128 (5.6)	738 (7.2)	390 (3.9)
	第4病日	4,150 (20.5)	2,383 (23.3)	1,767 (17.6)
	第5病日	7,608 (37.5)	3,836 (37.5)	3,772 (37.5)
	第6病日	3,991 (19.7)	1,807 (17.6)	2,184 (21.7)
	第7病日	1,848 (9.1)	757 (7.4)	1,091 (10.9)
	第8病日	735 (3.6)	291 (2.8)	444 (4.4)
	第9病日	304 (1.5)	121 (1.2)	183 (1.8)
	第10病日以上	326 (1.6)	168 (1.6)	158 (1.6)

*1：初診時病日不明5人を除く23,332人を集計した

*2：γ-グロブリン使用例20,313人のうち1日投与量、投与日数、投与開始時病日不明23人を除く20,290人を集計した

膜炎、麻痺性イレウス、DICは不定型例でやや多く、肝障害、胆嚢腫大は定型例が多かった。

性別では関節痛または関節炎、無菌性髄膜炎、肝障害は女で多く、胆嚢腫大、麻痺性イレウスは男が多かった。

年齢を2歳未満と2歳以上の2区分にして観察すると、関節痛または関節炎、胆嚢腫大、麻痺性イレウス、DICは2歳以上で多く、無菌性髄膜炎、肝障害は2歳未満が多かった。

顔面神経麻痺は1例のみ（6歳の男の定型例）にみられた（表11）。

要約

①2年間の報告患者数は23,337人〔2007年11,581人（男6,684人，女4,897人），2008年11,756人（男6,839人，女4,917人），性比1.38〕

で、2008年12月末までの患者数は合計249,019人（男144,350人，女104,669人）となり、2000年以降、患者数は急上昇を続けている。

②0～4歳人口10万対罹患率は、2007年215.3（男242.6，女186.6），2008年218.6（男248.2，

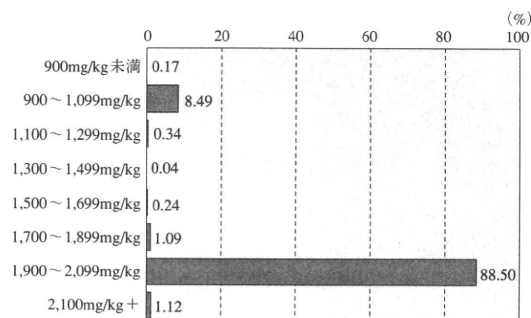


図8 γ-グロブリン使用総量（初回）の分布
γ-グロブリン使用例20,313人のうち1日投与量、投与人数不明9人を除く20,304人を集計

表9 γ-グロブリン1日投与量 (mg/kg) 別、投与日数の分布

	総数および%		1日	2日	3日	4日	5日	
	総数	%						
総数*	20,304 (100)	100	17,765 (87.5)	2,346 (11.6)	41 (0.2)	21 (0.1)	131 (0.6)	
～299 mg/kg	17	0.08	3	3	1	1	9	
300～499 mg/kg	143	0.70	3	8	13	9	110	
500～699 mg/kg	27	0.13	4	3	6	9	5	
700～899 mg/kg	30	0.15	12	11	3	0	4	
900～1,099 mg/kg	4,006	19.73	1,712	2,272	17	2	3	
1,100～1,299 mg/kg	71	0.35	58	12	1	0	0	
1,300～1,499 mg/kg	7	0.03	6	1	0	0	0	
1,500～1,699 mg/kg	34	0.17	33	1	0	0	0	
1,700～1,899 mg/kg	199	0.98	199	0	0	0	0	
1,900～2,099 mg/kg	15,642	77.04	15,607	35	0	0	0	
2,100 mg/kg+	128	0.63	128	0	0	0	0	
再掲	200 mg/kg	11	0.05	0	2	1	0	8
	400 mg/kg	130	0.64	2	8	12	9	99
	1,000 mg/kg	3,872	19.07	1,636	2,215	16	2	3
	2,000 mg/kg	15,209	74.91	15,175	34	0	0	0
	その他	1,082	5.33	952	87	12	10	21

*：γ-グロブリン使用例20,313人のうち1日投与量、投与日数不明9人を除く20,304人を集計した
()内は横向き%を示す