

Number of Patients with Coagulation Disorders in Japan: 2010 Annual Report from the Research Committee for the National Surveillance on Coagulation Disorders in Japan

Shinobu Tatsunami¹, Rie Kuwabara², Mieko Akita³, Junichi Mimaya⁴, Akira Ahirahata⁵, Masashi Taki⁶

¹Unit of Medical Statistics, St. Marianna University School of Medicine

²Institute of Radioisotope Research, St. Marianna University Graduate School of Medicine

³Department of Pediatrics, St. Marianna University School of Medicine

⁴Atami Public Health and Welfare Center

⁵Kitakyushu Yahata Higashi Hospital

⁶Department of Pediatrics, Yokohama City Seibu Hospital, St. Marianna University School of Medicine

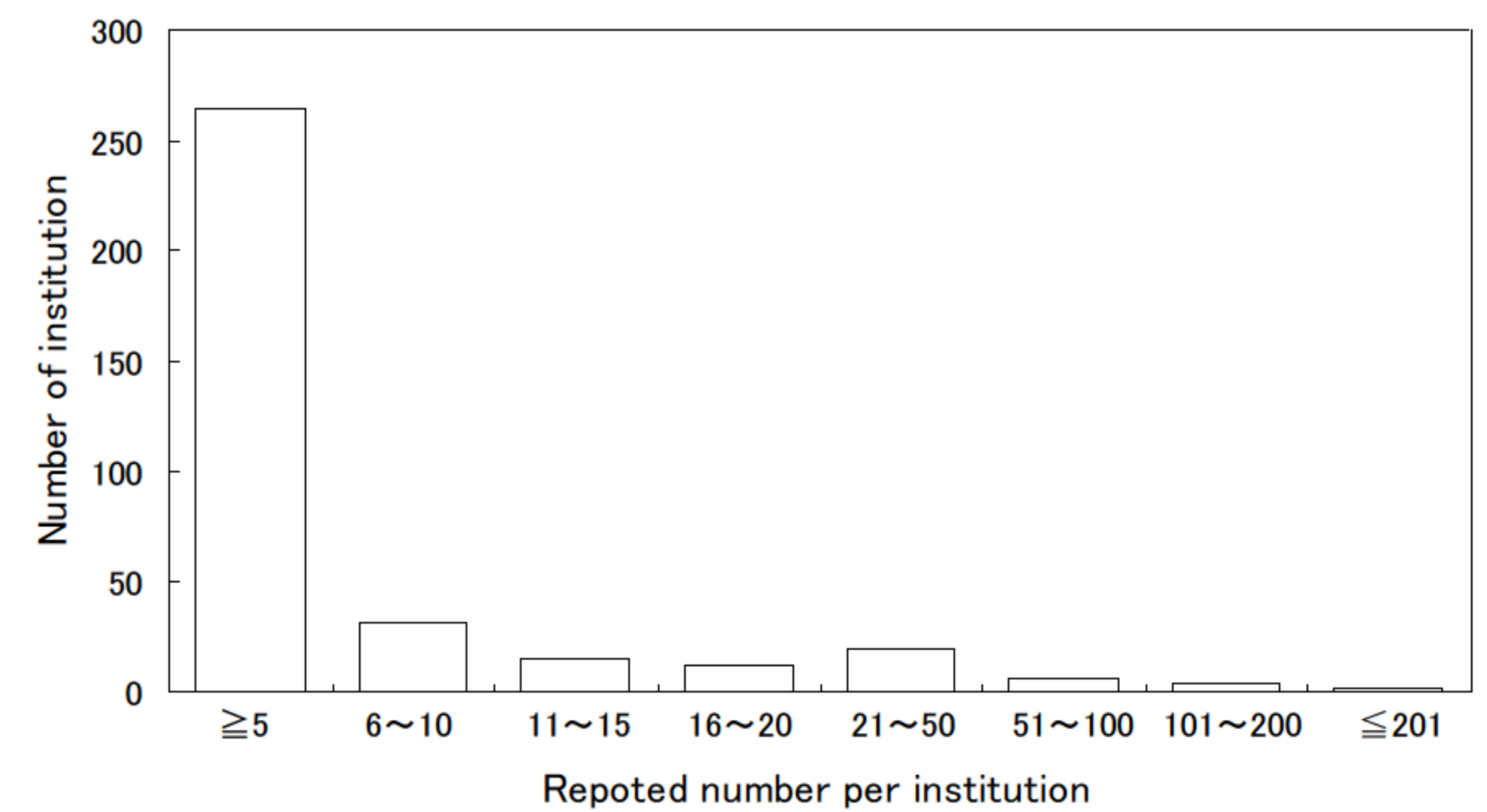
Objective: To report the collected number of Japanese patients with coagulation disorders as of May 31, 2010.

Methods: We utilized the database of the Research Committee for the National Surveillance on Coagulation Disorders in Japan, which is the only national registry of Japanese hemophiliacs.

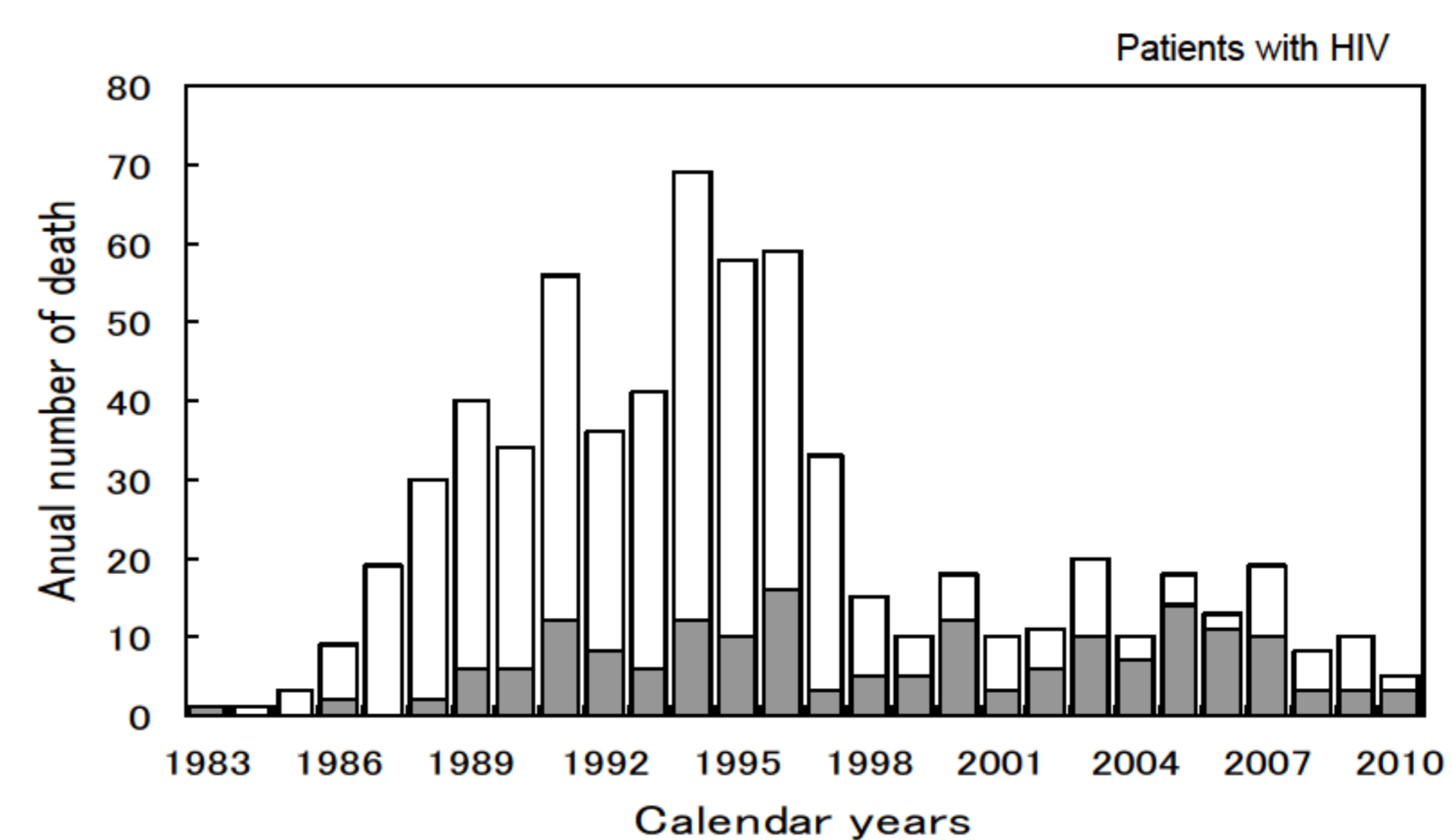
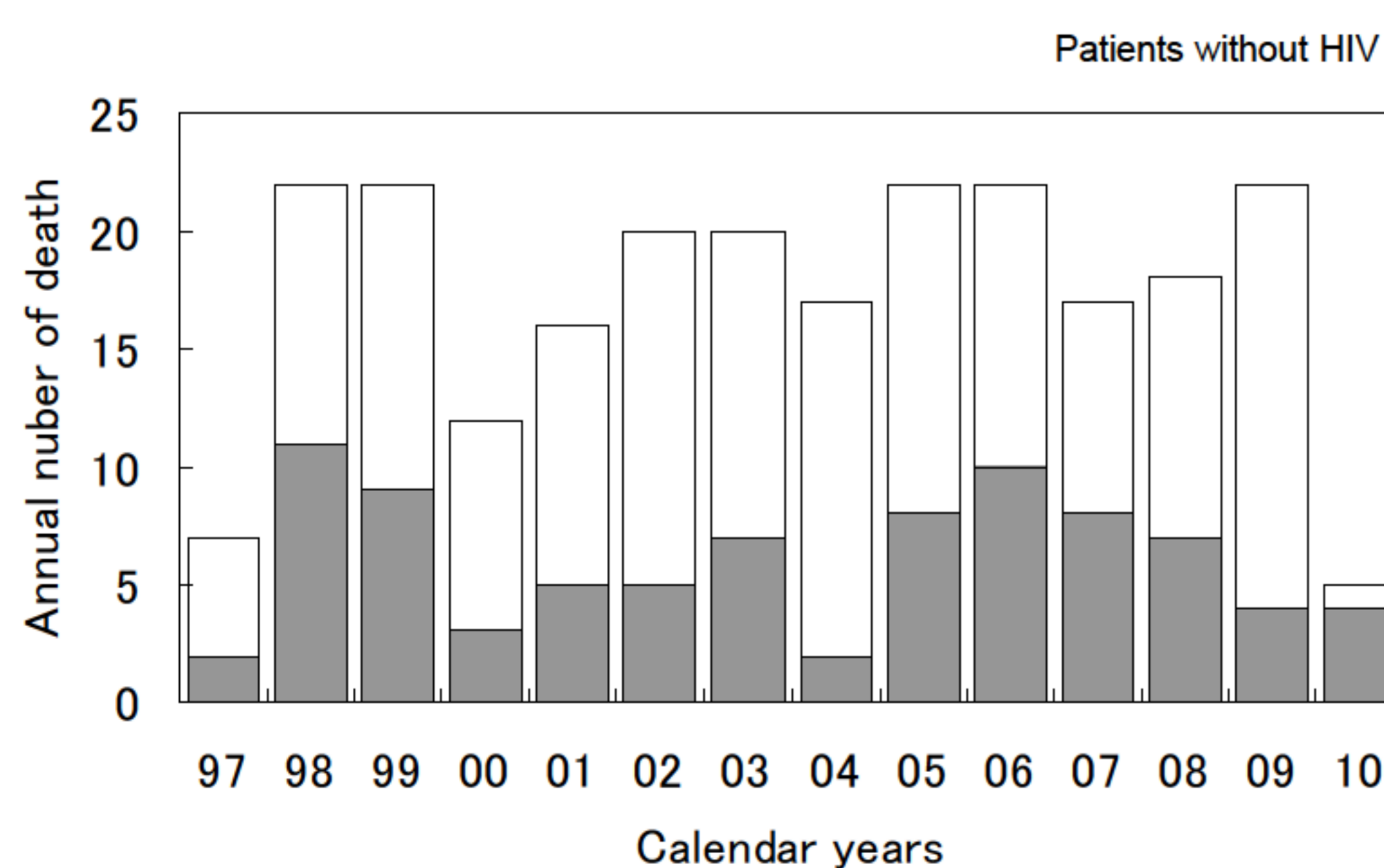
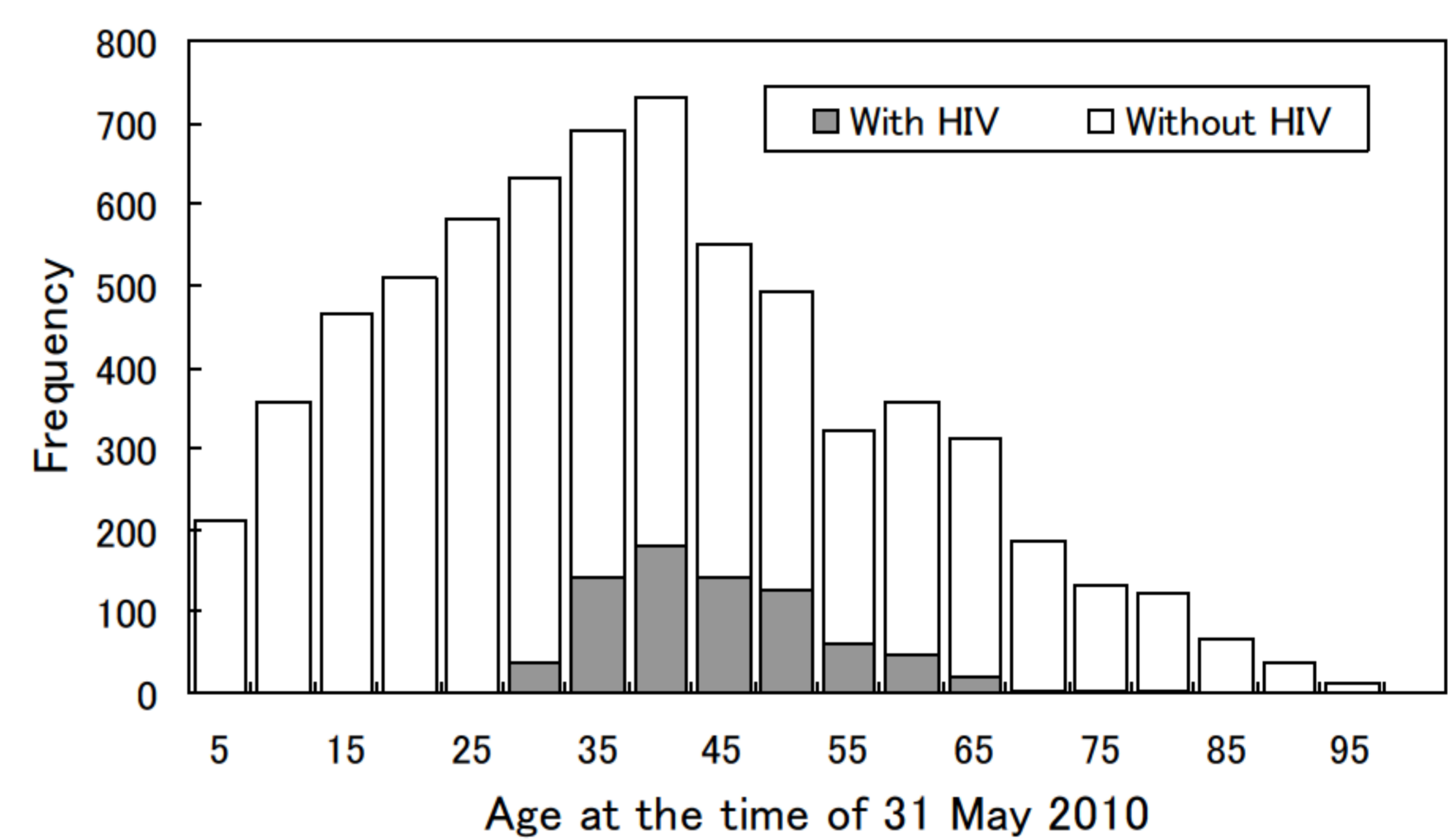
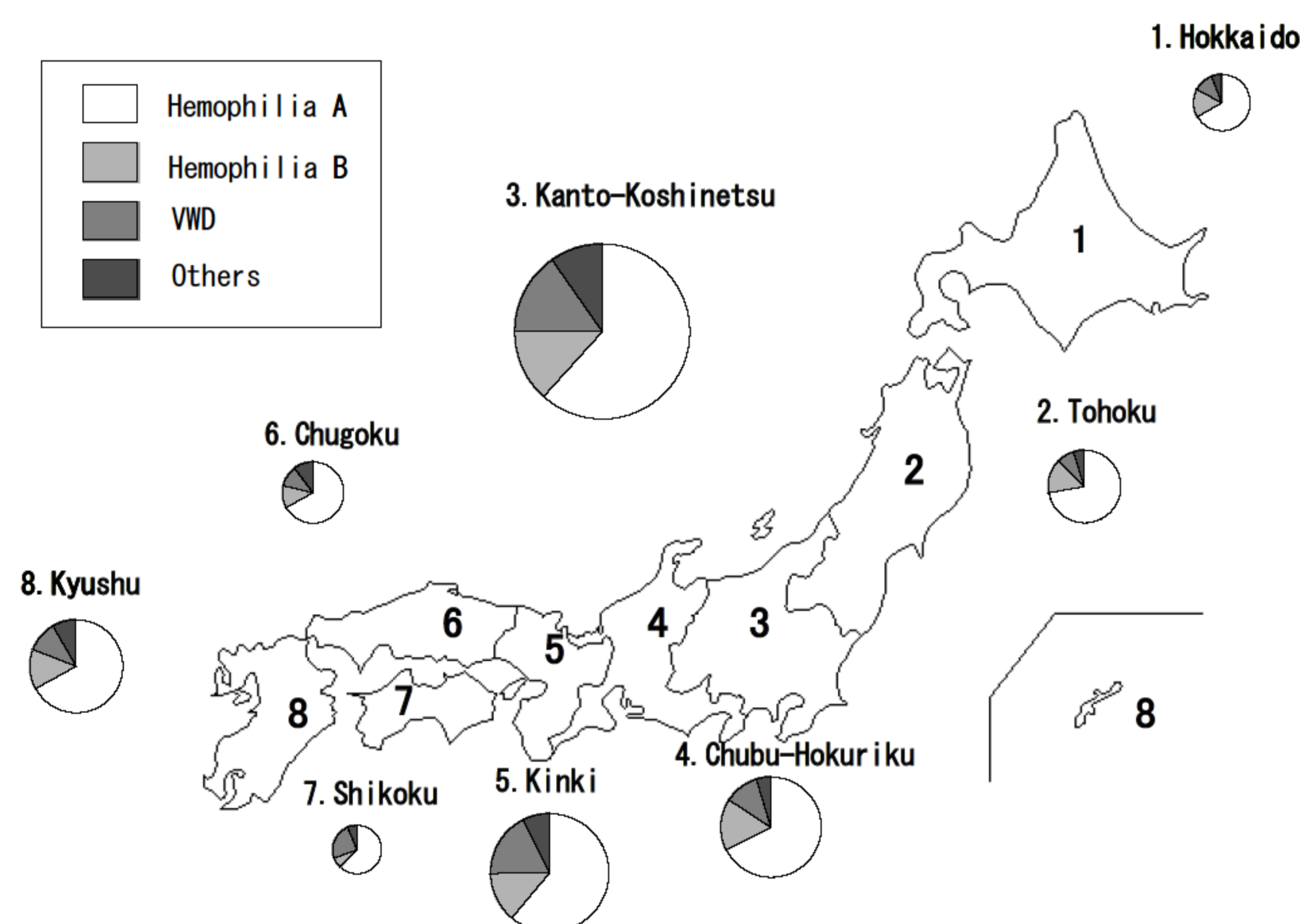
Table 1. Number of patients with coagulation disorders in Japan

	Hemophilia A	Hemophilia B	VWD	Others	Total
Patients living without HIV	3804	777	937	499	6017
(Male)	3778	765	430	261	5234
(Female)	26	12	507	238	783
Patients living with HIV	590	175	7	3	775
(Male)	590	175	2	0	767
(Female)	0	0	5	3	8
Total	4394	952	944	502	6792
(Male)	4368	940	432	261	6001
(Female)	26	12	512	241	791

(Dated on 31 May 2010)



Results: The numbers of patients as of 31 May 2010 were as follows: hemophilia A, 4394 patients (male 4368, female 26); hemophilia B, 952 (male 940, female 12); von Willebrand's disease (VWD), 944 (male 432, female 512); and hemophilia-related disorders, 502 (male 261, female 241). The prevalence of hemophilia (hemophilia A and hemophilia B) was 8.7 male hemophiliacs per 100,000 Japanese males. Regarding the percentages with HIV or HCV infection in patients with coagulation disorders, 10.2 percent of them had been infected with both HIV and HCV, 0.2 percent HIV only, and 30.5 percent HCV only. The number of newly registered patients in the 2010 surveillance was 177. Their age distribution was as follows: age ≤5 years, 65; 6-10 years, 18; 11-15 years, 12; 16-20 years, 4; and 21 years ≤ age, 78. The annual numbers of deaths in the interval from 1998 to 2010 varied between 12 and 22 in HIV-negative patients, and between 8 and 20 in HIV-positive patients. The proportion of deaths with a report of critical liver disease among 402 accumulated deaths was 29% and 55% in HIV-negative or -positive patients, respectively ($p < 0.001$). Among the deaths with critical liver disease, infection with HCV had been identified in 88.2 percent of patients.



Conclusions: There were 177 newly reported patients in 2010. However, they included 116 patients older than 5 years old. Therefore, continuing surveillance is still inevitably necessary to accurately comprehend the prevalence of coagulation disorders in Japan. Death with critical liver disease triggered by HCV has been the most important problem among the present Japanese patients with coagulation disorders.

