Korea Hemophilia Foundation Registry Trends 1991-2014: Patient registry, demographics, health services utilization

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Background

With modern substitution treatments, the life expectancy and quality of life of individuals with hemophilia has markedly increased because of the improvements in the medical care, the use of clotting factor products , and effective management for contagious diseases. The Korea Hemophilia Foundation (KHF) has registered Korean hemophiliacs since 1991, and updated the cohort by establishing the network of medical information between affiliated clinics.

Methods

Experiences over the past 24 years are described regarding the KHF database from 1991 until 2014. Hemophilic arthropathy was defined as stage II or more by the Arnold-Hilgartner classification or as having deformity or limited motion of action in limbs joints. Hepatitis C was defined as HCV PCR positive with positive anti-HCV antibodies (Ab) and serum alanine aminotransferase (ALT) ≥ 50 IU/L. Hepatitis C carrier included hepatitis C, positive HCV PCR and/or positive anti-HCV Ab.

Results

From 1991 to 2014, the KHF registration patients grew about 200% (n=2,255). Registered hemophilia patients was approximately 1:20,000 males at 1995, which gradually increased over years, and up to 0.9:10,000 males at 2014, with 81% having factor VIII deficiency. Two third of registered patients had 'severe' disorder, 20% had moderate hemophilia and 11% was classified with mild deficiency. The registered patients with VWD grew by 107; they are now roughly one-twentieth of the population of hemophiliacs. The prevalence of hemophilic arthropathy decreased from 71% to 61% in patients with hemophilia A and from 63% to 40% in patients with hemophilia B. The prevalence of anti-HCV positivity has decreased from 49% to 30% in hemophilia A; from 48% to 19% in hemophilia B. In patients with severe hemophilia A, the overall prevalence of inhibitors was 17.2% in 2006, 10.6% in 2012, and 6.3% in 2014; in those with severe hemophilia B, the overall prevalence was 6.8%, 3.8%, and 3.1%, respectively.

Fig. 1. Status of registration in KHF by year 1991-2014

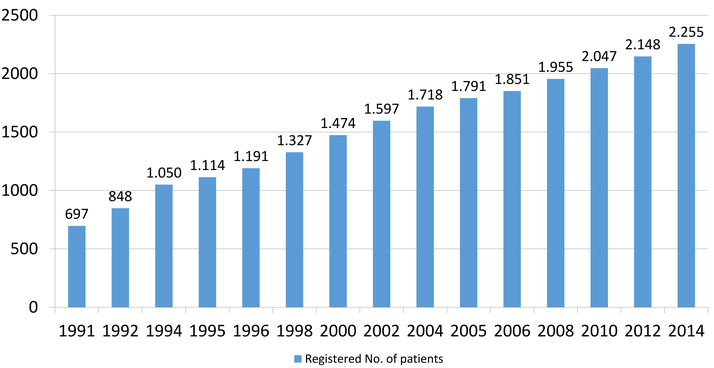

Table 1. Type of diseases in 2014 Korea Hemophilia Foundation (KHF) patients.

Diseases	Registered No. (%)	Male No.	Female No.
Hemophilia A	1,635 (72.5)	1,605	30
Hemophilia B	396 (17.6)	386	10
Von Willebrand disease (VWD)	107 (4.7)	65	42
Factor I deficiency	6 (0.3)	3	3
Factor V deficiency	6 (0.3)	2	4
Factor VII deficiency	36 (1.6)	19	17
Factor X deficiency	2 (0.1)	2	0
Factor XI deficiency	19 (0.8)	11	8
Factor XII deficiency	3 (0.1)	2	1
Factor XIII deficiency	5 (0.2)	1	4
Multiple factors deficiency	7 (0.3)	6	1
Acquired coagulation factor deficiency	8 (0.3)	5	3
Others	26 (1.2)	11	15
Unknown†		0	3
Total	2,255 (100)	2,114 (93.7)	141 (6.3)

Table 2. Age distribution of registered patients in KHF.

Age (years)	Hemophilia A	Hemophilia B	VWD	Others	Total	Percent	
0~4	71	23	1	4	99	4.4%	0.00011
5~9	108	29	6	8	151	6.7%	
10~14	138	45	9	19	211	9.4%	
0~14					461	20.4%	
15~19	167	50	17	21	255	11.3%	0.00013
20~24	215	38	19	20	292	12.9%	
25~29	160	38	18	13	229	10.2%	
15~29					776	34.4%	
30~34	171	37	6	7	221	9.8%	
35~39	167	37	9	5	218	9.7%	
40~44	139	25	6	5	175	7.8%	
30~44					614	27.2%	
45~49	111	30	5	1	147	6.5%	
50~54	67	14	3	3	87	3.9%	
55~59	60	12	1	1	74	3.3%	
45~59					308	13.7%	
60~64	26	8	1	1	36	1.6%	
65~69	15	3	2	3	23	1.0%	
70~74	8	2	3	3	16	0.7%	
75~79	6	4	1	2	13	0.6%	
80~	6	1	0	1	8	0.4%	
≥60					96	4.3%	
Total	1,635	396	107	117	2,255	100%	

Table 3. Status of chronic hemophilic arthropathy in KHF hemophilia patients.

Hemophilia / Year	Severity: Severe	Moderate	Mild	Total
Hemophilia A				
1991-1999*				696/979 (71.1)
2006†	649/972 (66.8)	123/309 (39.8)	9/122 (7.4)	781/1,423 (54.9)
2010	761/1,015 (75.0)	156/324 (48.1)	25/162 (15.4)	942/1,501 (62.8)
2012	801/1,080 (74.2)	152/313 (48.6)	27/179 (15.1)	980/1579 (62.1)
2014	839/1,154 (70.6)	131/290 (45.2)	34/184 (18.5)	1,004/1,635 (61.4)
Hemophilia B				
1991-1999*				95/152 (62.5)
2006†	100/182 (54.9)	16/78 (20.5)	2/37 (5.4)	118/301 (39.2)
2010	132/176 (75.0)	19/46 (41.3)	27/46 (58.7)	178/268 (66.4)
2012	133/213 (62.4)	17/102 (16.7)	1/47 (2.1)	151/364 (41.4)
2014	137/224 (61.2)	17/123 (13.8)	3/46 (6.5)	157/396 (39.6)

Table 4. Status of hepatitis C in patients with hemophilia A

Diseases	Hepatitis C*	HCV PCR positive†	HCV Ab positive only‡	Overall (HCV positive)
Hemophilia A				
1991-1999 (n=1,137)§	128 (11.3)	-	430 (37.8)	558 (49.1)
2005 (n=1,381)§	175 (12.7)	122 (8.8)	257 (18.6)	554 (40.1)
2010 (n=1,473)§	40 (2.7)	124 (8.4)	345 (23.4)	509 (34.6)
2012 (n=1,518)§	37 (2.4)	104 (7.0)	360 (23.7)	501 (33.0)
2014 (n=1,635)§	35 (2.1)	84 (5.1)	482 (29.5)	601 (36.8)

Table 5. Status of hepatitis C in patients with hemophilia B

Diseases	Hepatitis C*	HCV PCR positive†	HCV Ab positive only‡	Overall (HCV positive)
Hemophilia B				
1991-1999 (n=192)§	26 (13.5)	-	66 (34.4)	92 (47.9)
2005 (n=287)§	30 (10.5)	20 (7.0)	48 (16.7)	98 (34.1)
2010 (n=328)§	6 (1.8)	16 (4.9)	65 (19.8)	87 (26.5)
2012 (n=344)§	4 (1.2)	14 (4.0)	64 (18.6)	82 (23.8)
2014 (n=396)§	3 (0.8)	11 (2.8)	76 (19.2)	90 (22.7)

Conclusions

Although the KHF patient base has grown much faster than the general Korea population, some patients have still been left out, especially those with VWD. Therefore, a more efficient and nationwide registry system should be fixed for the provision of comprehensive management for this disorder.