UDC
The Universal Data Collection Program
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Report on the Universal Data Collection Program

Includes data collected from May 1998 through August 2003

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The *Report on the Universal Data Collection Program* is accessible via internet at http://www.cdc.gov/ncbddd/hbd/surveillance.htm Confidential information, referrals, and educational material on hemophilia and other bleeding disorders are also available by calling the National Hemophilia Foundation's information line, HANDI, at 800-42-HANDI.

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Commentary

The two most common congenital bleeding disorders are von Willebrand disease (VWD) and hemophilia. VWD is caused by the defective synthesis or function of a protein, von Willebrand factor that is necessary for normal blood clotting. VWD occurs with equal frequency in men and women. Although the prevalence of this disease is not precisely known, it is estimated that between one and two percent of the population are affected. There are different types and severity of VWD. Symptoms include heavy or prolonged menstrual bleeding, easy bruising, frequent or prolonged nosebleeds, and prolonged bleeding following surgery, dental work, childbirth, or injury.

Hemophilia is caused by a defect in the gene located on the X chromosome that contains the genetic code for one of the clotting factor proteins necessary for normal blood clotting. A deficiency of factor VIII is referred to as hemophilia A or "classic" hemophilia. In contrast, a deficiency of factor IX characterizes hemophilia B, also known as Christmas disease. The defect usually occurs on one of the two female X chromosomes and results in a carrier state. When males have the defect on their only X chromosome, they have the disease. Thus, almost all of the approximately 17,000 people with hemophilia in the United States are male.

People with severe hemophilia can experience serious bleeding into tissues, muscles, joints, and internal organs, often without any obvious trauma. Repeated bleeding into joints without adequate treatment results in crippling chronic joint disease, one of the severe complications of bleeding disorders. In the mid-1970s, treatment for hemophilia was improved through the use of clotting factor concentrates, products made from the plasma of donated blood. However, because blood donations from thousands of donors are pooled together to make these products, many people with bleeding disorders were infected with hepatitis B and C viruses and with human immunodeficiency virus (HIV), the virus that causes AIDS, before the risk of disease transmission in blood products was recognized and prevention measures taken.

In 1975, Congress initiated federal funding to specialized hemophilia treatment centers (HTCs) to provide comprehensive care to people with bleeding disorders. Since 1986, the Centers for Disease Control and Prevention (CDC) has been involved with the hemophilia community through the HTC system, primarily through risk-reduction efforts aimed at preventing secondary infection of family members with HIV.

In 1991, CDC received a request from the National Hemophilia Foundation to expand their collaborative activities within the bleeding disorders community. Meetings with patients and hemophilia care providers were held during 1992 to determine the areas of highest priority. Based on recommendations from these constituents, a congressional mandate was issued to CDC, with the goal of reducing the human suffering and financial burden of bleeding disorders by focusing national emphasis on prevention and early intervention. The issues of greatest concern identified by the bleeding disorders community were: (1) the safety of the blood supply from infectious diseases and (2) the prevention of joint disease.

In response, CDC developed the Universal Data Collection Program (UDC). The purpose of UDC is two-fold: (1) to establish a sensitive blood safety monitoring system

among people with bleeding disorders and (2) to collect a uniform set of clinical outcomes information that can be used to monitor the occurrence of and potential risk factors for infectious diseases and joint complications.

People with bleeding disorders are enrolled in UDC by care providers in each of the nation's 140 federally funded HTCs. As part of the project, a uniform set of clinical data and plasma specimens is collected by HTC staff each year during each participant's annual comprehensive clinic visit. A portion of the plasma specimen is used to perform free screening tests for hepatitis A, B, and C viruses and for HIV. The remainder of the specimen is stored for use as needed in future blood safety investigations.

Enrollment in UDC began in May 1998. Information about eligibility requirements, enrollment procedures, and data collection can be found in the *Technical Notes* of this report. Participating HTCs are listed by region in the *Acknowledgements*. A regional map is included at the end of this report.

The purpose of this surveillance report is to disseminate the information being collected by UDC to public health workers, health educators and planners, other care providers, and patients in the bleeding disorders community. The report contains information about the demographic characteristics of the participants, their blood and factor product use, and the occurrence and treatment of joint and infectious diseases.

We hope that this information will prove useful to those involved in efforts to reduce or prevent the complications of these conditions.

The proper interpretation and appropriate use of surveillance data require an understanding of how the data are collected, reported, and analyzed. Therefore, readers of this report are encouraged to review the *Technical Notes*, beginning on page 18.

Highlights

This issue of the UDC Surveillance Report focuses on data collected from people who have been enrolled through August 2003.

Since May, 1998, 13,440 people with bleeding disorders have been enrolled. There have been 29,237 UDC visits. The overall national refusal rate is 7.9%.

The data presented in Figures 1 and 2 represents new enrollment in UDC. The decline of new enrollees who have either hemophilia or VWD represents the success of UDC in capturing more and more of the population over time.

Figures 3 and 4 show UDC visits over time and by region. UDC visits have increased over time and are expected to increase in 2003 over the previous year.

Figures 5 and 6 show the refusal rates in UDC from May 1998 through August 2003 by year and region. Refusal rates have generally declined over time, the exceptions being a rise in the refusal rate among people with hemophilia from 8.2% in 1998 to 10.5% in 1999 and also among people with VWD whose refusal rate rose from 9.6% in 1999 to 11.2% in 2000. At all other intervals for both bleeding disorders, refusal rates have declined over time. The overall refusal rate is 7.6% for people with hemophilia and 8.7% for people with VWD. Refusal rates through August 2003 in regions I through IX remained below 15%, with a low of 2.7% in Region VIII among people with hemophilia to a high of

14.9% among people with VWD in Region VII. In Region X refusal rates are unusually high due to low numbers of people enrolled in UDC.

Figure 7 shows the follow-up time for people enrolled in UDC. For the first time, more than 50% have follow-up time of over 1 year.

Figure 8 shows the number of people who have a first-time visit compared to those who have a follow-up visit for each year through August 2003. The number of people who have a first time visit have gone down as a proportion of total visits and those with follow-up visits have increased over time.

The distribution of demographic characteristics (Table 1) and sources of healthcare reimbursement (Table 2) remain consistent among people enrolled in UDC.

As depicted in Table 3, one quarter of people with hemophilia have mild disease, 22.7% have moderate disease, and 52.3% have severe disease. 70.9% of people with VWD are classified as having Type 1, 11.1% as having Type 2, 7.4% as having Type 3, and 10.7% are classified as having other or unknown type of VWD.

Table 4 shows the average number of bleeds by disease severity and prophylaxis use in people with hemophilia and VWD. For people with hemophilia, the reported bleeding frequency was similar for those who were using prophylaxis compared with those who received episodic care. On the other hand, people with severe disease on prophylaxis reported far fewer bleeds than those on episodic care. Among people with VWD, those with Type 3 VWD had the most bleeds. Bleeding was far more common in sites other than joints and much less common among people with hemophilia B.

In Table 5, 1200 (11.7%) people with hemophilia had an intravenous access device (IVAD) in the year previous to their most recent visit. Of these, 145 (12.1%) had an IVAD associated infection. 189 (29.2%) of the 652 people with hemophilia receiving any therapy for viral hepatitis had successful treatment of the disease.

Table 6 shows that the most common type of treatment used for all severity levels of hemophilia was episodic care. 11.2% of people with moderate disease used continuous prophylaxis and 8.2% of those with severe disease used intermittent prophylaxis.

Prevalence of inhibitors (Table 7) is highest among people with severe hemophilia A. Inhibitors are much less common among people with hemophilia B.

The proportion of overall factor product use (Table 8) has remained consistent throughout the surveillance period. The majority of people use recombinant products.

Figure 9 illustrates that people with hemophilia A who have mild disease have a lower prevalence of intra-cranial hemorrhage (ICH) over time than those with moderate or severe disease. With the exception of people with severe disease in 1998, the prevalence of ICH among people with all severity levels of disease and over all years was under 1% and relatively stable over the time period. In people with hemophilia B (Figure 10), there was more variability in the prevalence of ICH across severity levels and over time. This is probably a reflection of the smaller number of people with Hemophilia B enrolled in UDC.

In Table 9, nearly one-half of people with severe hemophilia report some restriction of activity. Data in Table 10 demonstrates that the average joint limitation value for people with Type 3 VWD (69.0) falls between the value for people with mild (55.6) and moderate (84.3) hemophilia.

Figures 11 and 12 show natural or acquired immunity to hepatitis A by age and region in people with hemophilia. Overall immunity rates are approximately 60%. Figure 13 shows that among people with VWD, the prevalence of natural or acquired immunity to hepatitis A averages around 40%, with people over the age of 60 having a higher prevalence of immunity.

Immunity to hepatitis B is illustrated in Figures 14, 15 and 16. As expected, immunity to this disease is higher (approximately 80% of people with hemophilia and 70% of people with VWD are immune) than to hepatitis A which is likely due to increased childhood vaccination.

Prevalence of hepatitis C infection for people with hemophilia and VWD is shown in Figure 17. Higher infection rates in adults reflect exposure to the disease prior to viral inactivation of factor products. Figure 18 shows that immunity to hepatitis A among people with hemophilia who are also infected with hepatitis C varies between 55-75% across regions.

Prevalence of HIV among people with hemophilia and VWD is shown in Figure 19. HIV infection is extremely low in all people with VWD; however, approximately one-third of people with hemophilia between the ages of 21 and 60 years are HIV-infected.

Tables 11 and 12 illustrate the number of people with hemophilia who use continuous prophylaxis by age group and severity. Continuous prophylaxis use drops off remarkably in people who are 21 or older. It is used most often by people with severe disease regardless of hemophilia type, less so by people with moderate disease, and rarely by people with mild disease. Among people with both moderate and severe disease, continuous prophylaxis use increases through the mid-teens and begins to decrease in the late teens, the decline in use occurring slightly earlier among people with severe type B hemophilia.

Table 13 shows the prevalence of overweight and obese people enrolled in UDC. People with hemophilia and VWD aged 13 to 19 years are almost twice as likely to be overweight as children of the same age group in the U.S. population. Obesity, which could only be compared for those 20 or older, is at a similar rate as in the U.S. population for people with hemophilia but appears higher for people with VWD than the general population.

Figure 1. New enrollment in UDC, May 1998 –August 2003

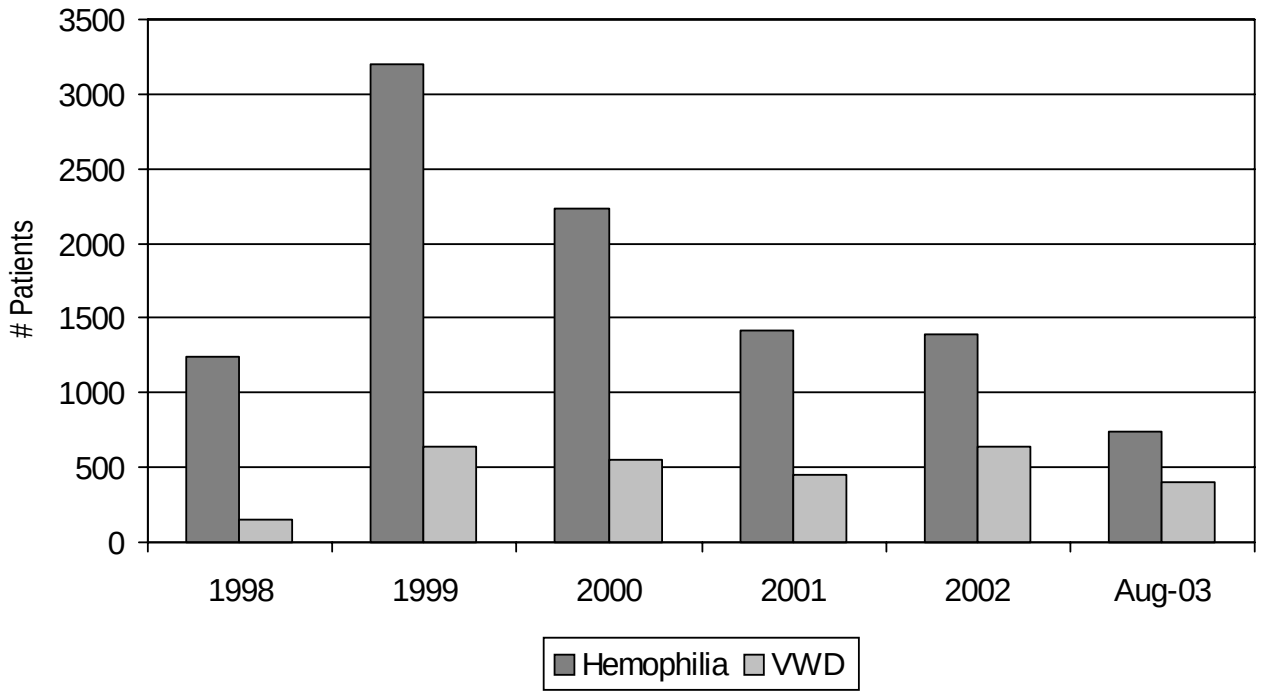
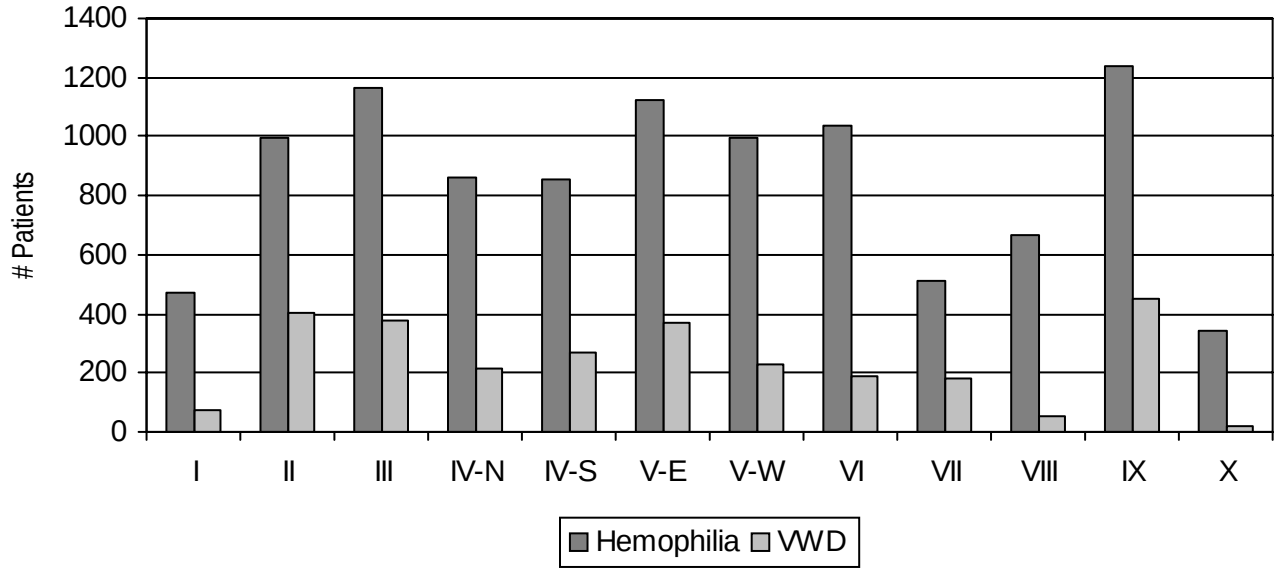


Figure 2. Total patients enrolled in UDC by region* through August 2003



*See map (page33) for regional designations.

Figure 3. UDC visits by year, May 1998-August 2003

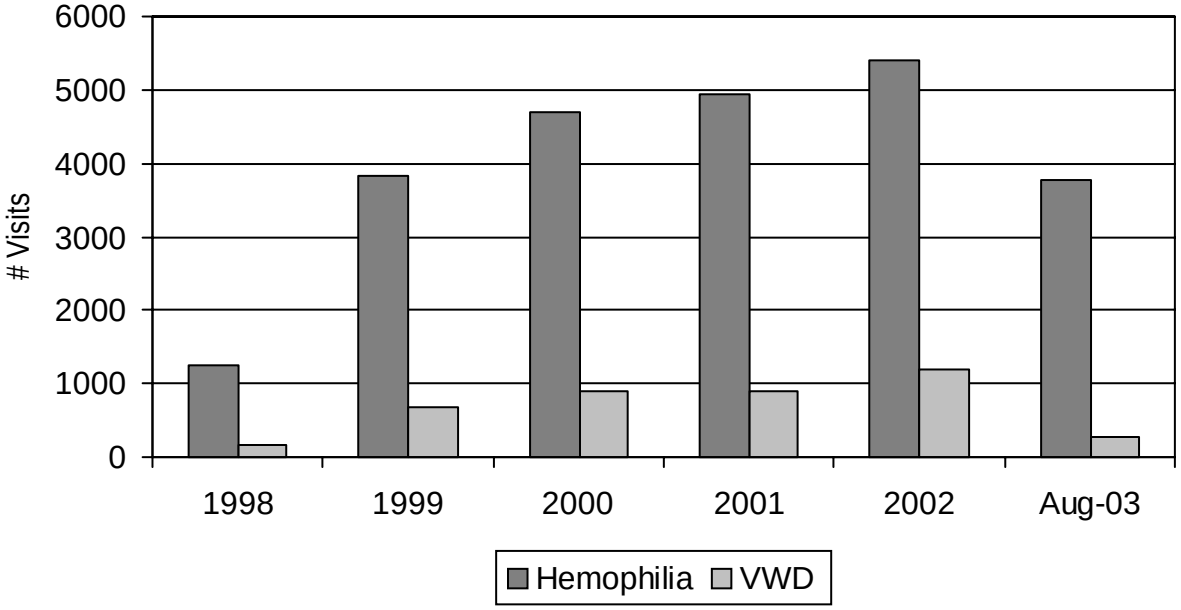
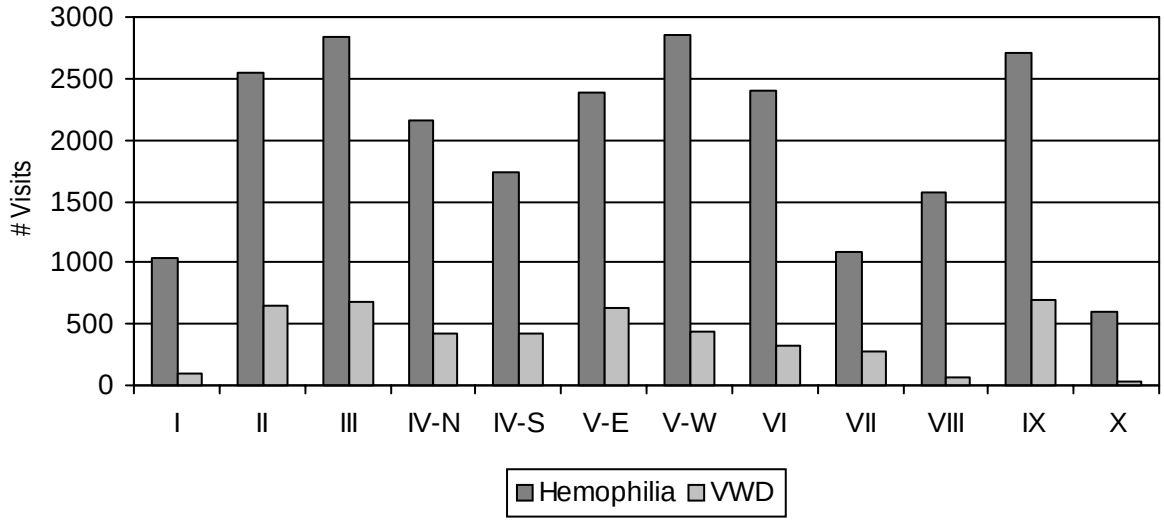


Figure 4. Total UDC visits by region* through August 2003



*See map (page 33) for regional designations.

Figure 5. Refusal rates in UDC by year, May 1998 – August 2003

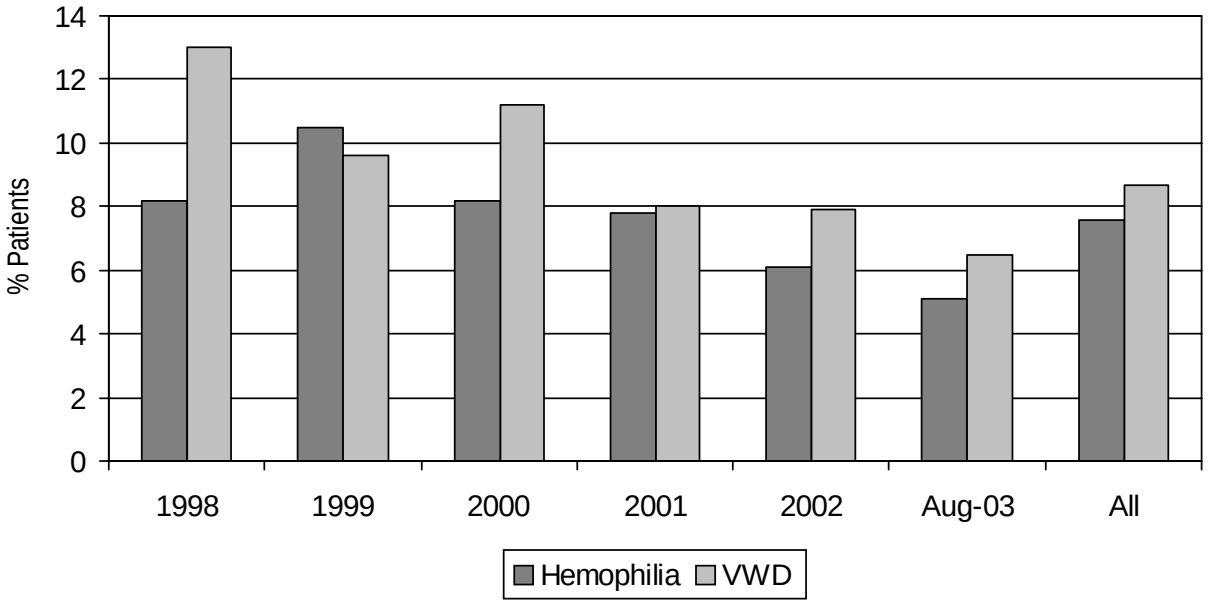
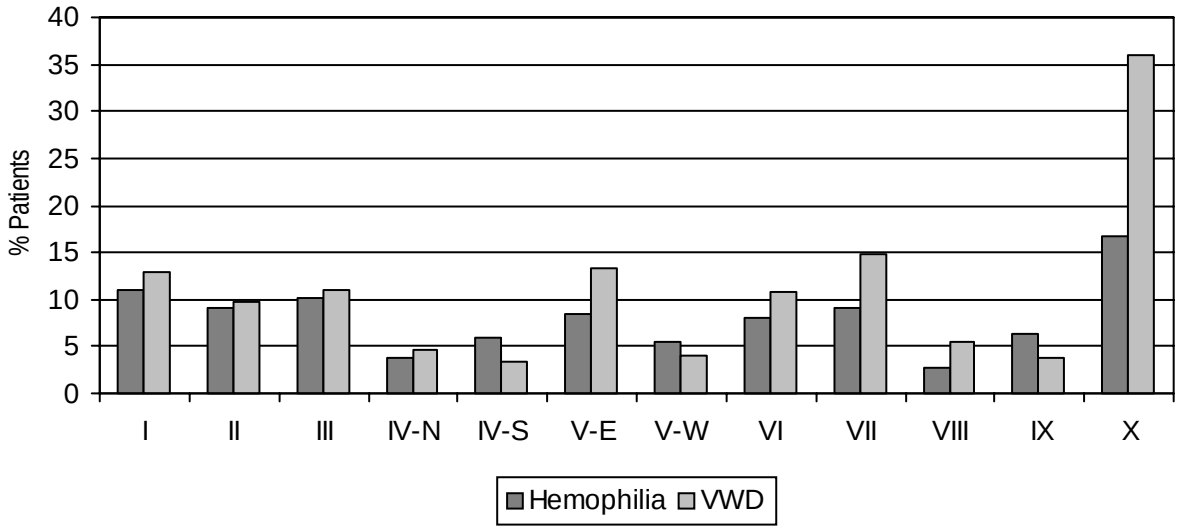


Figure 6. Refusal rates in UDC by region*, May 1998 – August 2003



*See map (page33) for regional designations.

Figure 7. Number of years of follow-up for people enrolled in UDC

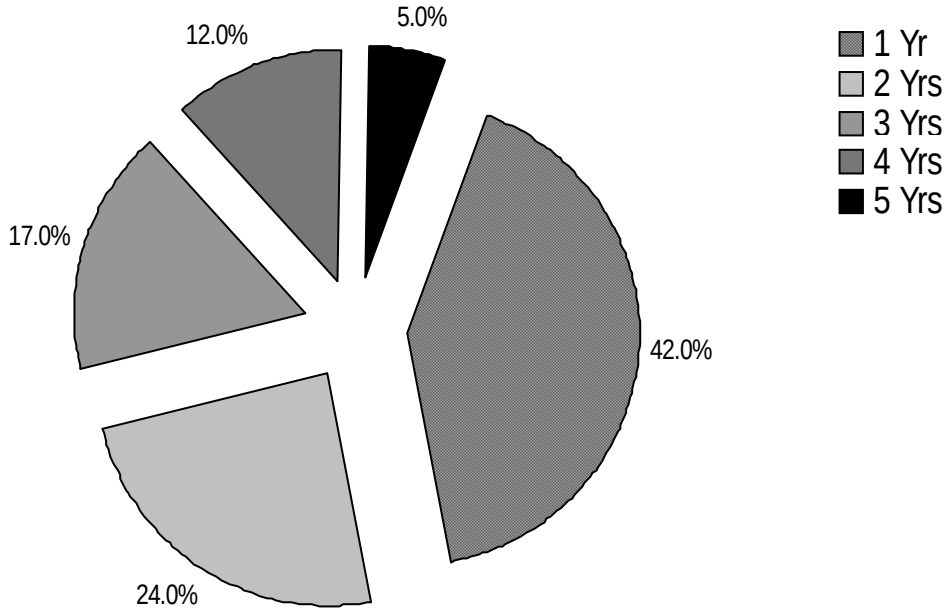


Figure 8. Visits by UDC participants through August 2003

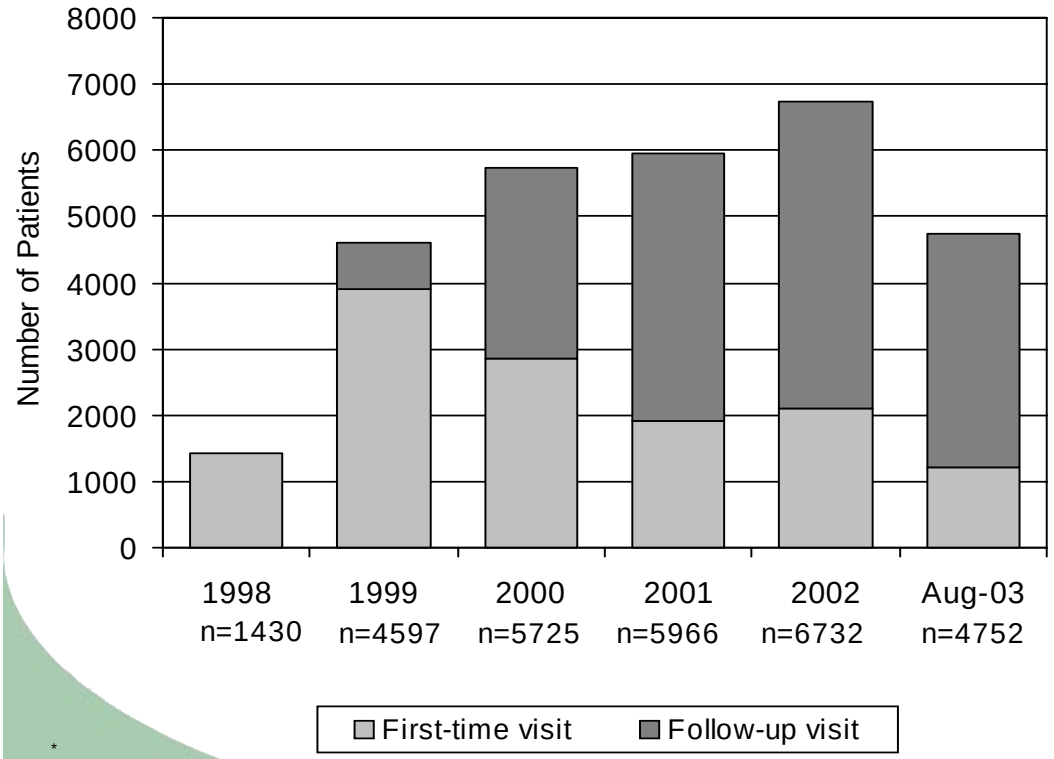


Table 1. Demographic characteristics of people* enrolled in UDC

	Hemophilia A (n =8087)		Hemophilia B (n=2183)		VWD (n = 2441)	
Characteristic	**Number**	**Percent**	**Number**	**Percent**	**Number**	**Percent**
Age Group (yrs)						
2-10	1763	21.8	447	20.5	538	18.9
11-20	2576	31.9	629	28.8	1113	39.1
21-40	2269	28.1	585	26.8	608	21.4
41-60	1200	14.8	407	18.6	436	15.3
>60	279	3.5	115	5.3	149	5.2
Race/Ethnicity						
White	5541	68.5	1632	74.8	2104	74.0
African American	1014	12.5	242	11.1	167	5.9
Hispanic	1053	13.0	209	9.6	355	12.4
Asian/Pacific Islander	220	2.7	33	1.5	77	2.7
Native American	59	0.7	17	0.8	19	0.7
Other	200	2.5	50	2.3	122	4.3
Sex						
Male	7915	97.9	2106	96.5	1185	41.7
Female	172	2.1	77	3.5	1659	58.3

*Sixty-five people were reported to have both hemophilia and VWD (these people are included in analyses as hemophilia patients only and not VWD patients). A total of 326 people had a bleeding disorder other than hemophilia or VWD.

Table 2. Sources* of health care reimbursement listed by people enrolled in UDC

	Hemophilia (n = 10270)		VWD (n = 2844)	
Reinbursement source	**Number**	**% of Total**	**Number**	**% of Total**
Commercial Insurance	1853	18.0	613	21.6
Commercial Insurance HMO	1949	19.0	675	23.7
Commercial Insurance PPO	2124	20.7	632	22.2
Medicare	861	8.4	149	5.2
Medicare HMO	82	0.8	27	1.0
Medicaid	2145	20.9	364	12.8
Medicaid HMO	583	5.7	208	7.3
CHAMPUS	74	0.7	35	1.2
State high risk plan	290	2.8	41	1.4
Other	1359	13.2	392	13.8
Uninsured	394	3.8	86	3.0

*Some people may have listed more than one source of reimbursement.
HMO = Health maintenance organization; PPO = Preferred provider organization

Table 3. Disease severity of people enrolled in UDC

	Number	Percent
Hemophilia	**10270**	
Mild	2567	25.0
Moderate	2332	22.7
Severe	5371	52.3
VWD	**2844**	
Type 1	2015	70.9
Type 2	315	11.1
Type 3	211	7.4
Other/Unknown	303	10.7

Table 4. Bleeding episodes* among people enrolled in UDC by prophylaxis use and disease severity

No Prophylaxis

	Hemophilia			VWD		
	Mild n = 2553	Moderate n = 2074	Severe n = 3677	Type 1 n = 2003	Type 2 n = 312	Type 3 n = 205
Bleeding site						
Joint *	0.6 (± 2.6)	3.2 (± 7.9)	8.6 (± 12.4)	0.1 (± 1.1)	0.2 (± 1.5)	1.7 (±5.0)
Muscle*	0.2 (± 0.9)	0.9 (± 2.3)	2.0 (± 4.8)	0.1 (± 1.1)	0.1 (± 0.9)	0.4 (±1.3)
Other*	0.8 (± 4.9)	1.4 (± 6.5)	1.7(± 6.3)	3.4 (± 10.1)	3.6 (± 15.3)	5.9 (±14.0)
All sites						
Mean (±SD)	1.7 (±5.8)	5.5 (±11.0)	12.4 (±15.7)	3.7 (±10.2)	3.9 (±15.3)	8.0 (±15.5)
Median	0	2	8	0	1	2

Prophylaxis Used

	Hemophilia	
	Moderate n =258	Severe n = 1694
Bleeding Site		
Joint *	3.3 (± 7.0)	2.9 (± 7.4)
Muscle*	0.9 (± 2.0)	0.8 (± 2.0)
Other*	1.1 (± 2.2)	1.2 (± 4.8)
All sites		
Mean (±SD)	5.3 (8.6)	4.9 (9.7)
Median	3	2

*Values are mean (±SD) number of bleeding episodes experienced during the 6-month period preceding the UDC visit

Table 5. Liver disease and intravenous access device infections among people enrolled in UDC

	Hemophilia (n = 10270)		VWD (n = 2844)	
	Number	% of Total	Number	% of Total
Risk Factors for liver disease				
Past/present hepatitis B virus infection	609	5.9	22	0.8
Past/present hepatitis C virus infection	1401	13.6	72	2.5
History of alcohol abuse	409	4.0	12	0.4
Other	151	1.5	22	0.8
None	8429	82.1	2744	96.5
Signs or symptoms of liver disease (During the last year)				
Jaundice	70	0.7	2	0.1
Ascites	65	0.6	6	0.2
Varices	54	0.5	5	0.2
Other	88	0.9	9	0.3
None	10065	98.0	2827	99.4
Laboratory markers of liver disease				
Chronically elevated ALT/AST levels	1484	14.5	64	2.3
Elevated prothrombin time in the last year	187	1.8	66	2.3
Therapy for chronic viral hepatitis				
Any therapy	652	6.4	34	1.2
Successful therapy	189	29.2*	10	30.3*
Intravenous access device (IVAD)				
Used an IVAD in the last year	1200	11.7	56	2.0
IIVAD infection in the last year	145	12.1**	1	1.8**

*Percent of people who received any therapy for chronic viral hepatitis.
**Percent of people who used an IVAD in the last year.

Table 6. Treatment type for people with hemophilia enrolled in UDC

	Mild		Moderate		Severe	
Treatment	Number	Percent	Number	Percent	Number	Percent
Episodic care	2530	98.5	1982	85.0	3163	58.9
Intermittent Prophylaxis	22	0.9	86	3.7	441	8.2
Continuous Prophylaxis*	14	0.5	257	11.0	1686	31.4
TOTAL	**2567**		**2332**		**5371**	

*Prophylaxis is considered continuous when administered on a regular basis and is expected to continue indefinitely.

Table 7. Prevalence of current inhibitors by titer* among people with hemophilia enrolled in UDC

	Hemophilia A			Hemophlia B		
Severity	Number	Low titer	High titer	Number	Low titer	High Titer
Mild	2000	17 (0.9%)	5 (0.3%)	567	0 --	0 --
Moderate	1572	49 (3.1%)	18 (1.1%)	760	1 (0.1%)	3 (0.4%)
Severe	4515	255 (5.7%)	241 (5.3%)	856	17 (2.0%)	24 (2.8%)

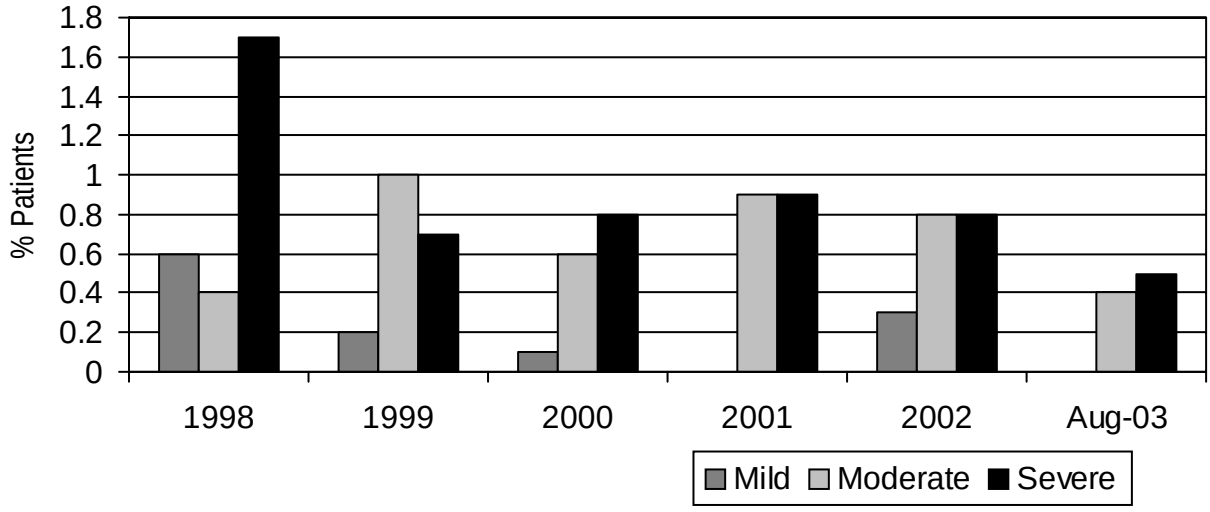
Inhibitor titer is determined by the highest reported inhibitor titer for any visit.
Low titer is defined as an inhibitor level of 0.5 – 5 Bethesda units (BU).
High titer is defined as an inhibitor level of >5 BU.

Table 8. Blood and factor products used* by people enrolled in UDC

	Hemophilia A n = 8087		Hemophilia B n = 2183		VWD n = 2844	
Treatment product	Number	Percent	Number	Percent	Number	Percent
Recombinant factor	5035	62.3	1229	56.3	13	0.5
Monclonal factor VIII	1441	17.8	4	0.2	4	0.1
Other human factor VIII	174	2.2	3	0.1	545	19.2
Porcine factor VIII	10	0.1	0	--	0	--
Purified factor IX	6	0.1	454	20.8	0	--
Prothrombin complex	28	0.4	30	1.4	0	--
Activated prothrombin complex	283	3.5	21	1.0	0	--
Cryoprecipitate of FFP	49	0.6	7	0.3	31	1.1
Desompressin	650	8.0	9	0.4	1181	41.5
None used	956	11.8	491	22.9	1098	38.6

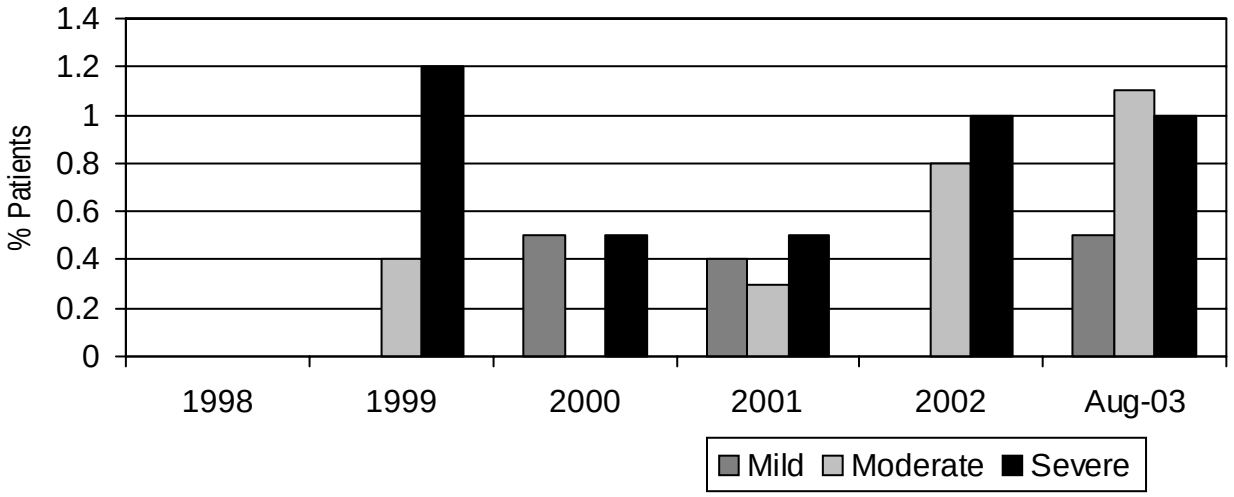
*Any use of the product(s) during the 12-month period preceding UDC enrollment.
NOTE: Individuals may have used more than one type of treatment product.

Figure 9. Prevalence of intra-cranial hemorrhage* in people with hemophilia A by severity, May 1998- August 2003



*Occurrence since last annual visit

Figure 10. Prevalence of intra-cranial hemorrhage* in people with hemophilia B by severity, May 1998- August 2003



*Occurrence since last annual visit

Table 9. Joint complications among people enrolled in UDC

	Hemophilia			VWD		
	Mild n (%)	Moderate n (%)	Severe n (%)	Type 1 n (%)	Type 2 n (%)	Type 3 n (%)
Target joint *	151 (5.9)	523 (22.4)	1953 (36.4)	29 (1.4)	6 (1.9)	38 (18.0)
Invasive procedure	79 (3.1)	78 (3.3)	498 (9.3)	27 (1.3)	2 (0.6)	15 (7.1)
Joint infection	15 (0.6)	7 (0.3)	69 (1.3)	17(0.8)	2 (0.6)	2 (0.6)
Used cane	315 (12.3)	508 (21.7)	1614 (30.1)	107 (5.1)	16 (5.1)	37 (17.5)
Used wheelchair	55 (2.1)	131 (5.6)	534 (9.9)	30 (1.5)	9 (2.9)	12 (5.7)
Any activity restriction	406 (15.8)	652 (28.0)	2199 (41.0)	161 (8.0)	31 (9.8)	57 (27.0)

*Please see Technical Notes (page 27) for the definition of a target joint

Table 10. Joint limitations among people enrolled in UDC

	Hemophilia			VWD		
	Mild	Moderate	Severe	Type 1	Type 2	Type 3
Number of patients	2347	2113	4647	1866	291	187
Mean indicator* value	55.6	84.3	148.8	19.3	26.2	69.0
Standard deviation	106.9	152.2	212.1	77.3	77.8	110.0

*Indicator is the total number of degrees of range of motion less than normal for five joints. The joint motions measured and normal values used (in parentheses) are: hip extension (30); hip flexion (120); knee flexion (135); knee extension (0); shoulder flexion (180); elbow flexion (150); elbow extension (0); elbow pronation and supination (80); ankle dorsiflexion (20); ankle plantar flexion (50). Any hyperextension of the knee or elbow is included in the calculation. In UDC, limitations in knee and elbow extension are recorded as negative numbers. Patients with missing measures for any of the joints are excluded from the analyses. As an example, patients with mild hemophilia have on average 55.6 degrees less than normal range of motion across ten joints. Because the sum of all of the normal measures is 1,690 degrees, this represents an overall 3.3% loss in range of motion.

Figure 11. Prevalence of natural or acquired immunity to hepatitis A virus among people with hemophilia enrolled in UDC

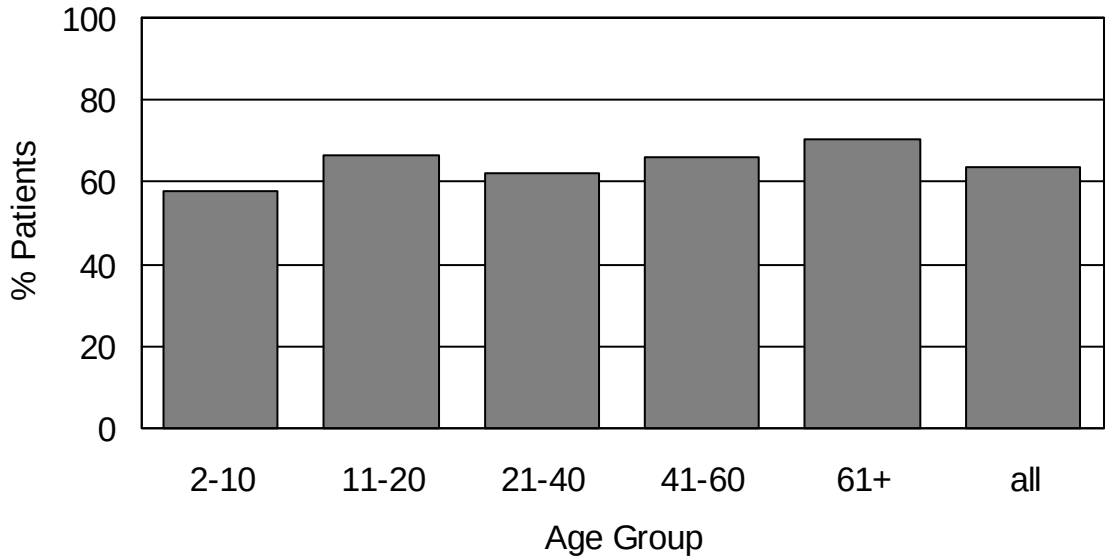


Figure 12. Regional distribution of natural or acquired immunity to hepatitis A virus among people with hemophilia enrolled in UDC

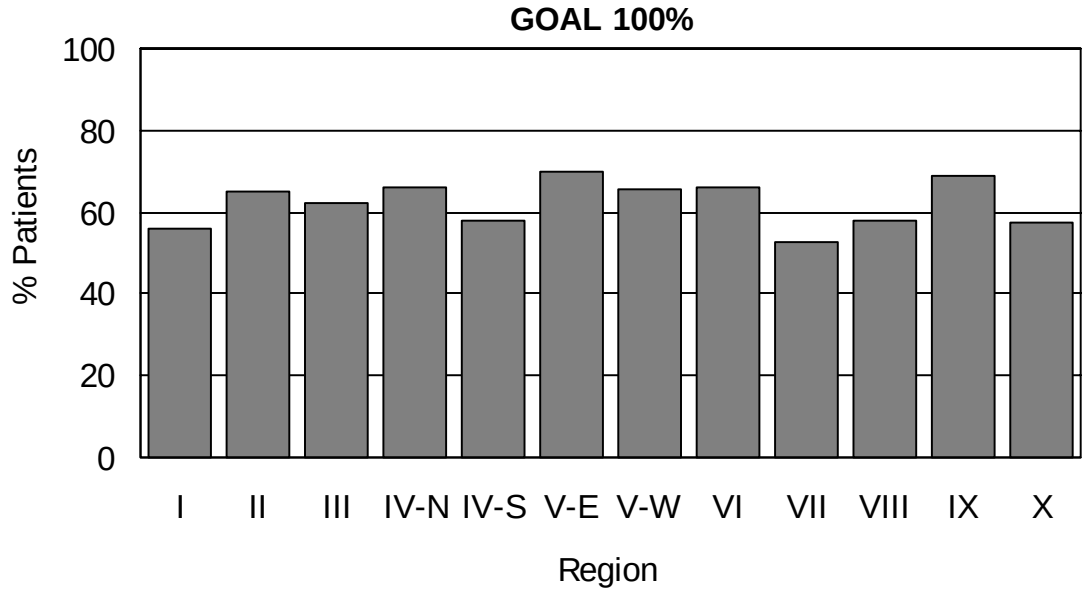


Figure 13. Prevalence of natural or acquired immunity to hepatitis A virus among people with VWD enrolled in UDC

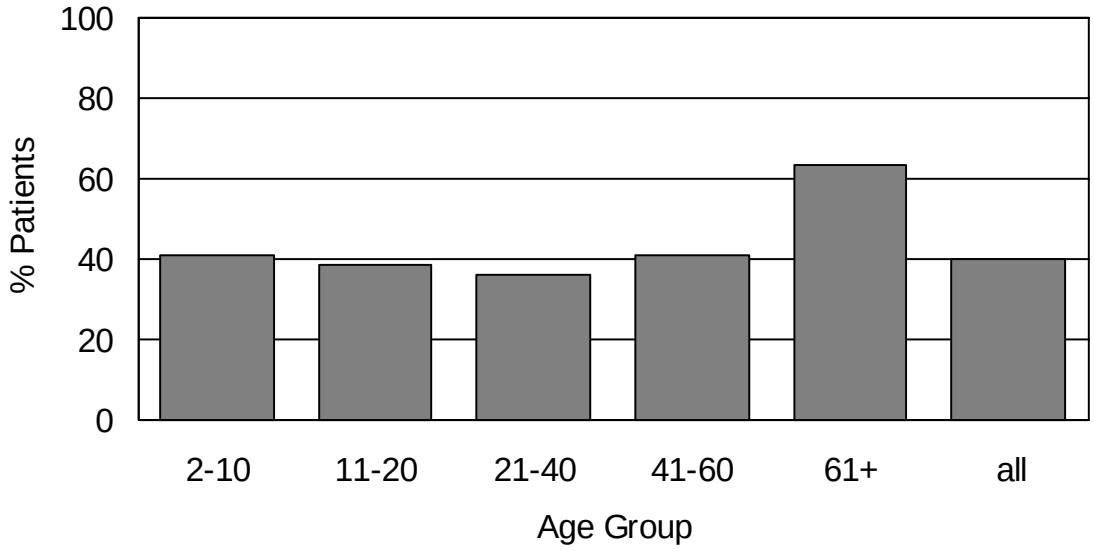


Figure 14. Prevalence of natural or acquired immunity to hepatitis B virus among people with hemophilia enrolled in UDC

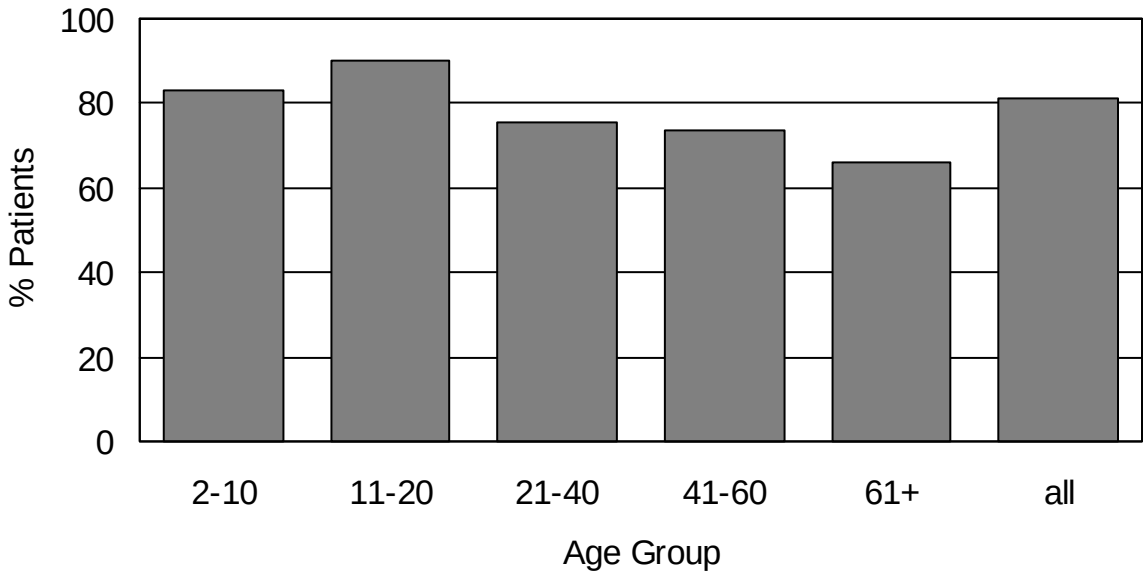


Figure 15. Regional distribution of natural or acquired immunity to hepatitis B virus among people with hemophilia enrolled in UDC

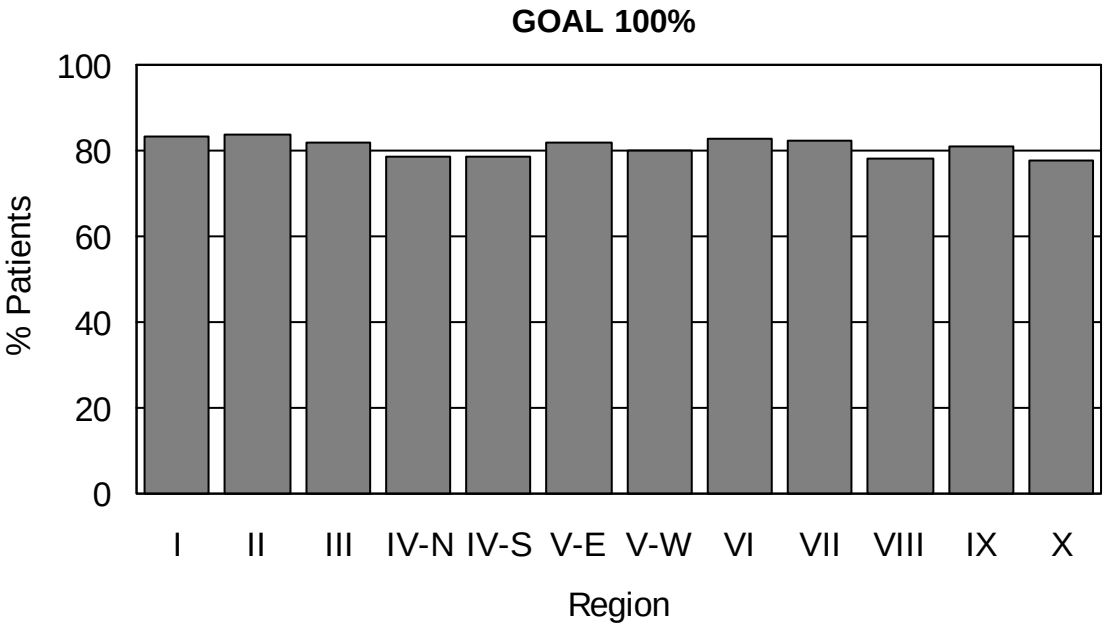


Figure 16. Prevalence of natural or acquired immunity to hepatitis B virus among people with VWD enrolled in UDC.

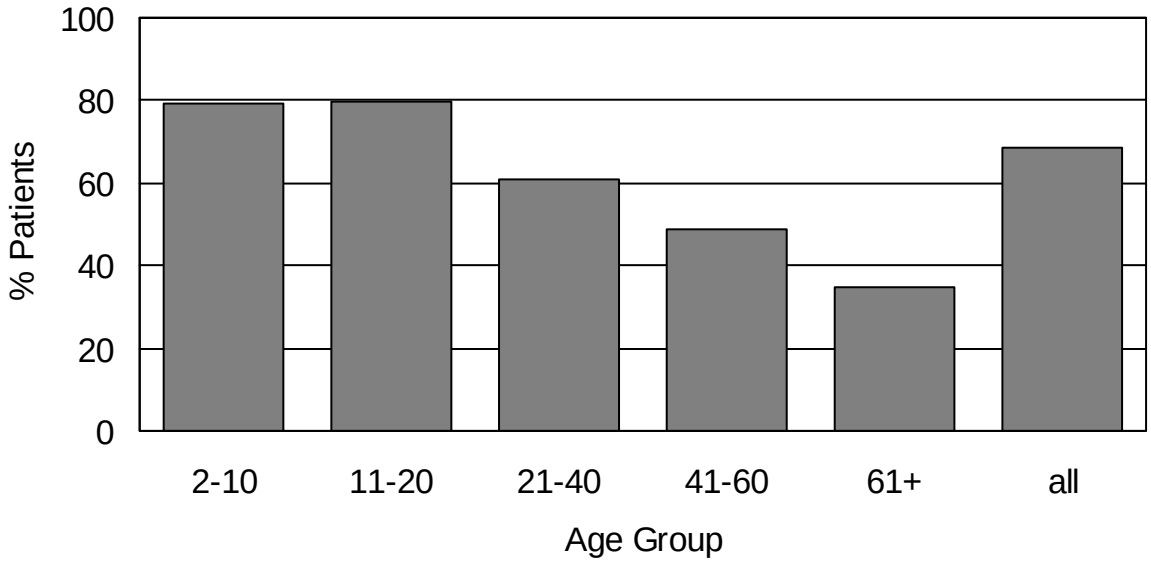


Figure 17. Prevalence of hepatitis C virus infection among people with bleeding disorders enrolled in UDC

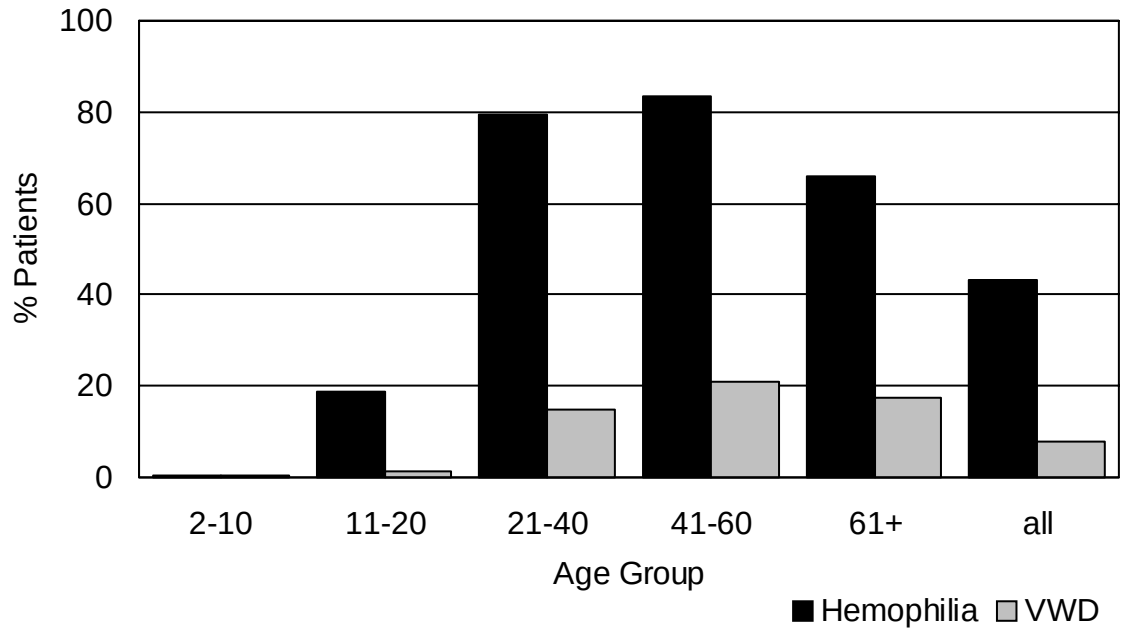


Figure 18. Prevalence of natural or acquired immunity to hepatitis A among people with hemophilia and infected with hepatitis C enrolled in UDC

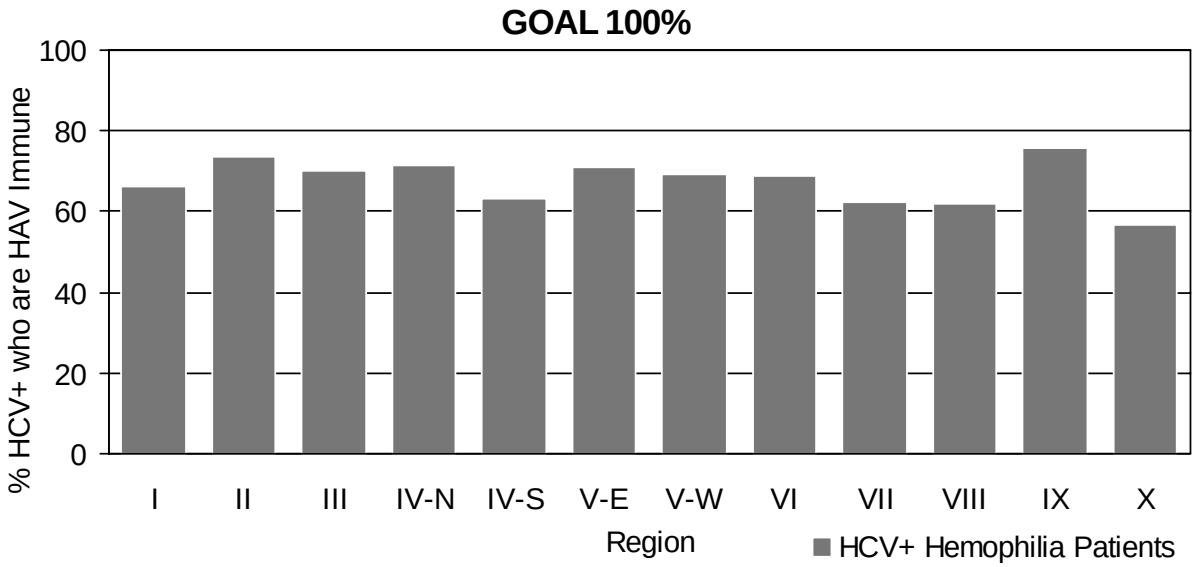


Figure 19. Prevalence of human immunodeficiency virus infection among people with bleeding disorders enrolled in UDC

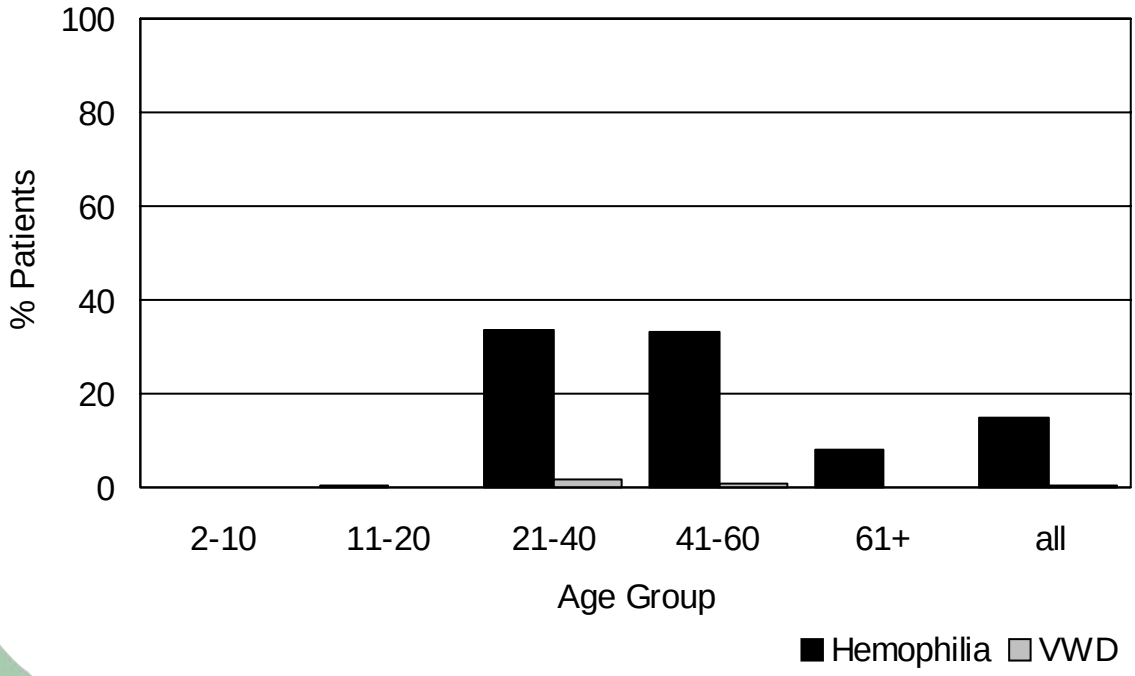


Table 11. Hemophilia A: Percent of patients on continuous prophylaxis

	Level of Severity					
	Mild		Moderate		Severe	
Age Group (years)	Total	N (%)	Total	N (%)	Total	N (%)
2-5	82	1 (1.2)	97	10 (10.3)	371	174 (46.9)
6-10	271	3 (1.1)	266	56 (21.1)	676	391 (57.8)
11-15	304	3 (1.0)	277	71 (25.6)	794	443 (55.8)
16-20	289	2 (0.7)	252	45 (17.9)	660	254 (38.5)
21+	1054	1 (0.1)	680	32 (4.7)	2014	222 (11.0)

Table 12. Hemophilia B: Percent of patients on continuous prophylaxis

	Level of Severity					
	Mild		Moderate		Severe	
Age Group (years)	Total	N (%)	Total	N (%)	Total	N (%)
2-5	23	0 (0.0)	52	3 (5.8)	65	26 (40.0)
6-10	69	1 (1.5)	110	10 (9.1)	128	59 (46.1)
11-15	82	1 (1.2)	127	18 (14.2)	123	54 (43.9)
16-20	75	0 (0.0)	103	5 (4.9)	119	31 (26.1)
21+	318	2 (0.6)	368	8 (2.2)	421	40 (9.5)

Table 13. Prevalence of overweight and obese persons in UDC and the US population

Overweight

Age Group	Hemophilia	VWD	US Population*
2-12	15.5%	17.0%	13.7%
13-19	18.2%	17.2%	11.5%
20+	32.4%	29.7%	32.6%

Obese

Age Group	Hemophilia	VWD	US Population*
20+	22.4%	31.9%	22.3%

*Based on data from the third National Health and Nutrition Examination Survery

Technical Notes

Eligibility Requirements

To participate in UDC, patients must receive care in a federally funded HTC and meet at least one of the following criteria: (1) be 2 years of age or older with a bleeding disorder due to congenital deficiency or acquired inhibitors in which any of the coagulation proteins is missing, reduced, or defective and has a functional level of less than 50 %; or (2) be 2 years of age or older with a diagnosis by a physician of von Willebrand disease. Individuals specifically excluded from participation in UDC include persons with any of the following: (1) an exclusive diagnosis of a platelet disorder, (2) thrombophilia, or (3) coagulation protein deficiencies due to liver failure.

Data Collection

UDC data are collected during a participant's "annual visit", which ideally should occur once each calendar year (January—December), with the interval between visits as close as possible to 12 months. Data are collected according to guidelines and definitions detailed in surveillance manuals provided to HTC staff by CDC. Informed consent for participation is obtained each year. Demographic information and reasons for refusal are obtained using a patient refusal form for all eligible people who decline to participate. To protect patient confidentiality, all data sent to CDC do not contain personal identifying information, but rather use a unique 12-digit code that is generated by a computer software program supplied to HTCs by CDC.

Eligible participants are registered into UDC through a registration form completed by HTC staff; information collected on this form includes patient demographic, diagnostic, and historical information. Month and year of birth are used to calculate age on the last day of the current year. Information on race and ethnicity is obtained from clinic records and might be based either on self-report or on observations made by care providers. During the annual visit, clinical information is recorded on a standardized data collection form (annual visit form). In addition to information about education, employment status, and health insurance, data are also collected about the type of treatment (episodic vs. prophylactic), presence and treatment of inhibitors, the number of bleeding episodes experienced (based on infusion logs or patient recall), the type and brand name of all factor concentrates or other treatment products used, and whether or not clotting factor is infused at home.

Information regarding infectious diseases is also collected, including risk factors and clinical signs, symptoms, and laboratory markers of liver disease. Data are also recorded about any therapy for chronic hepatitis; the status of vaccination for hepatitis A and B viruses; and, among patients with an intravenous access device, the occurrence of a device-associated infection. People $\geq$16 years of age who are HIV-infected are asked several questions concerning risk-reduction activities, including partner testing and condom use.

Data are also collected on joint disease, including the use of walking aids, the occurrence of joint infections, and measures of impact of joint disease on daily activities. During the visit, range of motion measurements on five joints (hip, knee, shoulder, elbow, and ankle) are taken by a physical therapist or other trained health care

provider according to detailed guidelines provided in a reference manual supplied by CDC. All health care providers performing these measurements are trained and certified by regional physical therapists who have themselves received centralized training. In addition, information about whether a particular joint is a "target joint" or whether the participant has required the use of an orthopedic appliance or has undergone an invasive orthopedic procedure is collected. In UDC, a target joint is defined as a joint in which recurrent bleeding has occurred on four or more occasions during the previous 6 months.

All data collection forms are sent overnight to CDC where they are then key entered into a computer database using double-entry software to minimize data entry errors. Data are then screened for omissions, inconsistencies, and unusual values that possibly represent abstraction or data-entry errors. Error reports are generated and faxed to the HTC, where a designated UDC contact uses available information to resolve discrepancies and complete missing data items.

Laboratory Testing

During the annual visit, a blood specimen is obtained from each participant in UDC. Each specimen is processed by HTC personnel according to guidelines provided by CDC that are designed to minimize the effects of storage and shipment on subsequent analyses. Samples are shipped overnight to the CDC Serum Bank where they are aliquoted and stored. A portion of each specimen is sent to the Eugene B. Casey Hepatitis Laboratory at Baylor College of Medicine in Houston, Texas. A second portion is sent to the HIV testing laboratory at CDC. The remainder of the specimen is stored in the CDC Serum Bank for future blood safety investigations, as needed.

Testing for hepatitis A, B, and C viruses follows algorithms designed to determine with the highest probability the patient's status with regard to exposure to or infection with these viruses. Information provided by HTC staff on a Laboratory Form, including the results of previous local testing and vaccination history, is used by personnel at the testing laboratory to provide a detailed interpretation of the test results.

Testing for HIV follows algorithms designed to determine patient status with regard to infection with HIV-1 and HIV-2. The results of all laboratory testing are reported to the HTC using the CDC unique code that can be matched to the patient only by HTC staff.

Mortality Reporting

Deaths occurring among all HTC patients (regardless of whether they have been enrolled in UDC) are reported to CDC using a mortality form. Data collected include age at death, sex, race or ethnicity, type and severity of disease, and whether or not blood products had been used during the year prior to death. Additionally, information about the death, including the date, cause (primary and contributing), and whether or not an autopsy was performed, is also collected.

Tabulation and Presentation of Data

Data in this report are provisional. The data represent the most current data available from an on-going surveillance project. Future reports will include expanded data tables to cover subsequent surveillance periods and will provide the results of more detailed analyses of available data and findings from special studies.

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New Haven, CT
Maine Medical Center
Scarborough, ME
Dartmouth-Hitchcock Hemophilia Center
Lebanon, NH
Rhode Island Hospital
Providence, RI
Vermont Regional Hemophilia Center
Burlington, VT
UCONN Hemophilia Treatment Center
Farmington, CT
Boston Hemophilia Center- Children's Hospital
Boston, MA
Boston Hemophilia Center- Brigham and Women's
Boston, MA

Region II
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Weill Medical College of Cornell University
New York, NY
Puerto Rico Hemophilia Treatment Center
San Juan, PR
UMDNJ-Robert Wood Johnson University Hospital
New Brunswick, NJ
St. Michael's Medical Center
Newark, NJ
Mary M. Gooley Hemophilia Center, Inc.
Rochester, NY
SUNY Upstate Medical University-Adult Program
Syracuse, NY
SUNY Upstate Medical University-Pediatric Program
Syracuse, NY
Hemophilia Center of Western New York – Adult
Buffalo, NY
The Regional Comprehensive Hemophilia & von Willebrand Treatment Center
Albany, NY
UHSH Blood Disorder Center
Johnson City, NY
Long Island Jewish Medical Center
New Hyde Park, NY
Mount Sinai School of Medicine
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Hemophilia Center of Western New York – Pediatric
Buffalo, NY
Newark Beth Israel Medical Center
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Georgetown University Hospital
Washington, DC
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University of Virginia Hospital
Charlottesville, VA
Virginia Commonwealth University
Richmond, VA
Children's Hospital of the King's Daughters
Norfolk, VA
Cardeza Foundation Hemophilia Center
Philadelphia, PA
Christiana Care Health Services
Newark, DE
Hemophilia Center of Central Pennsylvania
Hershey, PA
Lehigh Valley Hospital
Allentown, PA
Hemophilia Center of Western Pennsylvania
Pittsburgh, PA
West Virginia University Medical Center
Morgantown, WV

Charleston Area Medical Center
Charleston, WV
Johns Hopkins University Medical Center
Baltimore, MD
Children's Hospital of Philadelphia Specialty Center
Voorhees, NJ
Penn Comprehensive Hemophilia Program
Philadelphia, PA
University of Virginia Hospital Adult Hemophilia Program
Charlottesville, VA

Region IV-N
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Winston-Salem, NC
Norton Kosair Children's Medical Center
Louisville, KY
Brown Cancer Center
Louisville, KY
University of Kentucky Hemophilia Treatment Center
Lexington, KY
East Carolina University
Greenville, NC
Children's Hospital of Palmetto
Richland Memorial
Columbia, SC
University of Tennessee - Memphis
Memphis, TN
East Tennessee Comprehensive Hemophilia Center
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Vanderbilt University Medical Center
Nashville, TN
St. Jude Research Hospital
Memphis, TN
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Miami Comprehensive Hemophilia Center - Pediatrics
Miami, FL
University of Florida
Gainesville, FL
Children's Healthcare of Atlanta at Scottish Rite
Atlanta, GA
Medical College of Georgia - Adult
Augusta, GA
University of Mississippi Medical Center
Jackson, MS
University of Alabama Birmingham
Medical Center
Birmingham, AL
Miami Comprehensive Hemophilia Center - Adult
Miami, FL
Children's Rehabilitation Services
Mobile, AL
Children's Rehabilitation Services
Birmingham, AL
Emory University Hemophilia Program Office
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Munson Medical Center
Traverse City, MI
West Michigan Pediatric at Bronson
Kalamazoo, MI
Hemophilia Clinic of West Michigan Cancer Center
Kalamazoo, MI
Eastern Michigan Hemophilia Treatment Center
Flint, MI

DeVos Children's Hospital
Grand Rapids, MI
Ohio State University Medical Center
Columbus, OH
Cincinnati Children's Hospital Medical Center
Cincinnati, OH
University of Cincinnati Medical Center
Cincinnati, OH
Forum Health, Youngstown Hemophilia Center
Youngstown, OH
Columbus Children's Hospital
Columbus, OH
Northwest Ohio Hemophilia Treatment Center
Toledo, OH
Dayton Children's Medical Center
Dayton, OH
Indiana Hemophilia and Thrombosis Center
Indianapolis, IN
Michigan State University Center for Bleeding Disorders & Clotting Disorders
East Lansing, MI
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Cleveland, OH
Children's Hospital Medical Center of Akron
Akron, OH
DMC Karmanos Cancer Institute
Detroit, MI
University of Michigan Hemophilia and Coagulation Disorders
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Children's Memorial Hospital
Chicago, IL
Comprehensive Bleeding Disorders Center
Peoria, IL
Fairview University Hemophilia and Thrombosis Center
Minneapolis, MN
Mayo Comprehensive Hemophilia Center
Rochester, MN
MeritCare Hospital DBA Roger Maris Cancer Center
Fargo, ND
Hemophilia Outreach Centre
Green Bay, WI
Gundersen Clinic
LaCrosse, WI
American Red Cross-Badger Center for Bleeding Disorders
Madison, WI
Sacred Heart Hospital
Eau Claire, WI
Rush Children's Hospital
Chicago, IL
Michael Reese Hospital - Adult
Chicago, IL
South Dakota Children's Specialty Clinics
Sioux Falls, SD
Comprehensive Center for Bleeding Disorders
Milwaukee, WI
Cook County Children's Hospital
Chicago, IL

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Arkansas Center for Bleeding Disorders
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Oklahoma City, OK
Fort Worth Comprehensive Hemophilia Center
Ft. Worth, TX
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Kansas City, MO
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Hemophilia Treatment Center
Columbia, MO
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Palo Alto, CA
Alta Bates Medical Center
Berkeley, CA
Hemophilia and Thrombosis Center of Hawaii
Honolulu, HI
University of California at Davis
Sacramento, CA
University of California, San Francisco
San Francisco, CA
Orthopedic Hospital of Los Angeles
Los Angeles, CA
Children's Hospital, San Diego
San Diego, CA
Children's Hospital of Orange County
Orange, CA
Children's Hospital Oakland
Oakland, CA
City of Hope National Medical Center
Duarte, CA
Hemophilia and Thrombosis Center of Nevada
Las Vegas, Nevada
Guam Comprehensive Hemophilia Care Program
Hagatna, GU
Children's Hospital of Central California
Madera, CA
Hemophilia Treatment Center of Las Vegas
Las Vegas, NV

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Oregon Hemophilia Treatment Center
Portland, OR
Alaska Hemophilia Program
Anchorage, AK
Idaho Regional Hemophilia Center
Boise, ID

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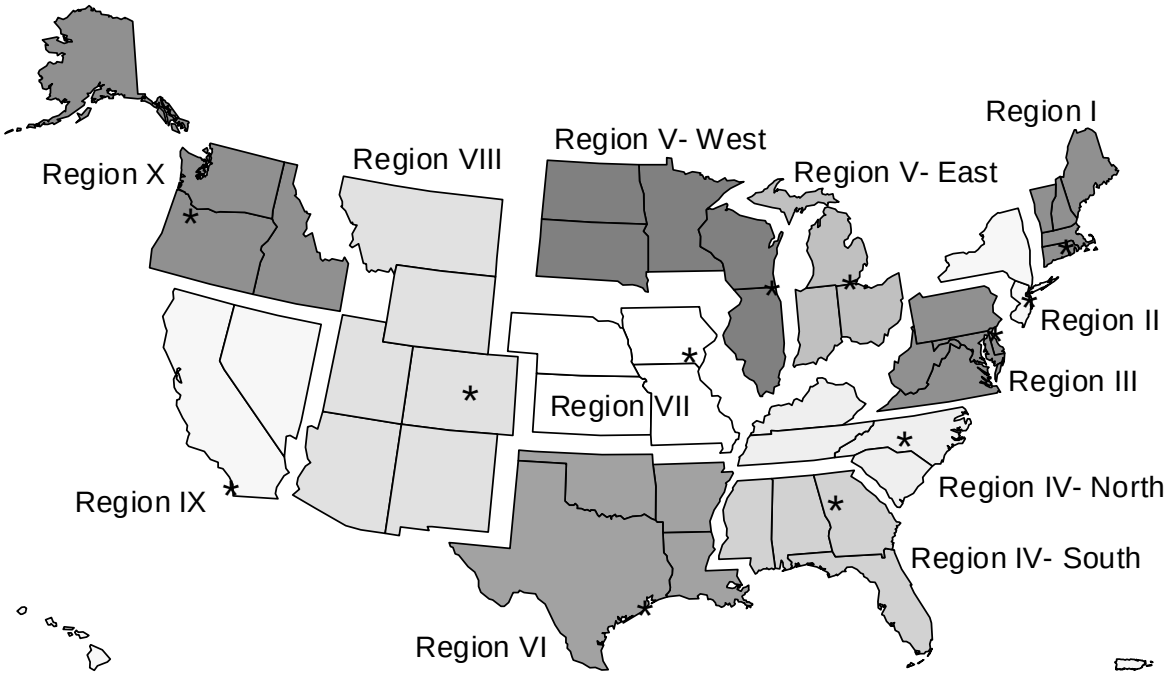
Hemophilia Treatment Center Regions



*Location of regional core center.

Department of Health and Human Services
Centers for Disease Control and Prevention