ATHN DATA DICTIONARY

VERSION 1.0 2/22/16

1 TABLE OF CONTENTS

2 ATHNDATASET PARTICIPANT

2.1 STATUS OF ATHNDATASET PARTICIPANTS

2.1.1 DEFINITION

ATHNdataset participants can have a status of active or inactive. Most of the analyses using the ATHNdataset will focus on active ATHNdataset participants who have authorized the sharing of their data for research through ATHN and who are receiving treatment from a hemophilia treatment center.

2.1.2 TYPE

Discrete variable.

2.1.3 VALUE

Active	25,262
Inactive	2,108

2.1.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

2.1.5 NOTE

All the following statistics include active ATHNdataset participants, herein referred to as "participant", by March 31, 2015 as the denominator unless specified otherwise.

2.1.6 USE

ATHNdataset, NHPCC (National Hemophilia Program Coordinating Center) Quality Metrics

2.2 HEMOPHILIA TREATMENT CENTER (HTC)

2.2.1 DEFINITION

HTCs are specialized health care centers that bring together a team of doctors, nurses, and other health professionals experienced in treating people with hemophilia, other bleeding disorders and clotting disorders.

2.2.2 TYPE

Discrete variable.

2.2.3 VALUE

Pick list (146 HTCs).

2.2.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing).

2.2.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF (My Life Our Future) Research Repository, HTDS.

2.3 REGION

2.3.1 DEFINITION

The regional designation corresponds to the names and geographic boundaries of regional networks as defined by the HRSA (Heath Resources and Services Administration) grant.

2.3.2 TYPE

Discrete variable.

2.3.3 VALUE

New England	3,299
Mid-Atlantic	1,852
Southeast	3,098
Great Lakes	8,519
Northern States	1,569
Great Plains	2,562
Mountain States	1,801
Western States	2,562

2.3.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

2.3.5 NOTE

New England Region includes Regions I and II. Mid-Atlantic Region is the same as Region III. Southeast Region include Region IV-N and IV-S. Great Lakes Region is the same as Region V-E. Northern States Region is the same as Region V-W. Great Plains Region includes Regions VI and VII. Mountain States Region includes Regions VIII and X. And Western States Region is the same as Region IX.

Region I includes states MA, CT, ME, VT, NH, and RI and Region II includes NY, NJ, and PR. Region III covers PA, DC, VA, DE, WV, and MD. Region IV-N includes NC, SC, KY, and TN and Region IV-S includes GA, FL, AL, and MS. Region V-E covers MI, OH, and IN. Region V-W includes WI, IL, MN, ND, and SD. Region VI covers TX, LA, AR, and OK and Region VII covers MO, IA, NE, and KS.

Region VIII includes CO, NM, AZ, UT, MT, and WY and Region X includes OR, WA, AK, and ID. Region IX covers CA, HI, GU, and NV.

2.3.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

3 DEMOGRAPHICS

3.1 DATE OF BIRTH

3.1.1 DEFINITION

The date on which a participant was born.

3.1.2 TYPE

Date variable.

3.1.3 VALUE

Date value (year, month, day).

3.1.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

3.1.5 NOTE

Date of birth is Personal Health Information (PHI) defined under HIPAA (Health Insurance Portability and Accountability of 1996). Date of birth is allowed to be included in a HIPAA compliant limited data set such as the ATHNdataset. Only year of birth, however, may be provided as part of a de-identified data set, except for dates indicative of ages over 89 years. Date of birth is most often used to derive time based variables (e.g., age at first bleed or age at first HTC visit). Full date of birth is released as a distinct variable only if required for an approved scientific purpose.

3.1.6 RELATED VARIABLE

Age, Year of Birth.

3.1.7 USE

ATHNdataset, NHPCC Quality Metrics, MLOF Research Repository.

3.2 YEAR OF BIRTH

3.2.1 DEFINITION

The year in which a participant was born.

3.2.2 TYPE

Discrete.

3.2.3 VALUE

Year integer (Y1918 – Y2015).

3.2.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

3.2.5 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

3.3 PLACE OF BIRTH

3.3.1 DEFINITION

The state or country where a participant was born.

3.3.2 TYPE

Discrete variable.

3.3.3 VALUE

Pick list (50 US states, and 204 foreign countries or territories).

3.3.4 COMPLETENESS

This variable is 95% complete (i.e., 5% missing value).

3.3.5 USE

ATHNdataset.

3.4 GENDER

3.4.1 DEFINITION

This data element reflects the gender of the participant.

3.4.2 TYPE

Discrete variable.

3.4.3 VALUE

Male	16,177
Female	9,080
Male to female	5
Female to male	0

3.4.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

3.4.5 NOTE

The gender of transgender patients can be recorded on the basis of their birth sex. In ATHNdataset, 16,182 (= 16,177 + 5) participants are considered as males and 9,080 are considered as females.

3.4.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

3.5 AGE

3.5.1 DEFINITION

This refers to the biological age of a participant. Ages based on events are included in sections of this document, such as age of diagnosis.

3.5.2 TYPE

Derived continuous variable.

3.5.3 VALUE

Mean	27 years
Median	20 years
Range	0-96 years

3.5.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

3.5.5 FORMULA

Age = (the given date – Date of Birth) / 365.25 if Date of Birth is given

OR

Age = (the given year – Year of Birth) if Year of Birth is only known.

3.5.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

3.5.7 RELATED VARIABLE

Date of Birth, Year of Birth, Age (Categorical)

3.6 AGE (CATEGORICAL)

3.6.1 DEFINITION

Several possible categories of Age measurement that appear often in the hemophilia literature.

3.6.2 TYPE

Derived discrete variable.

3.6.3 VALUE

0-2 years	766
3-12 years	6,077
13-18 years	4,907
19-29 years	5,013
30-49 years	4,434
50-74 years	3,641
75-89 years	402
90+ years	22

3.6.4 COMPLETENESS

This variable is 100% complete (i.e., 0% of missing value).

3.6.5 NOTE

0-2 years refer to $0 \leq Age < 3$, 3-12 years refer to $3 \leq Age < 13$, and etc. And 75+ years refer to $Age \geq 75$. Per HIPAA Privacy Rule, all ages over 89 years must be aggregated into a single category of age 90 or older.

3.6.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

3.6.7 RELATED VARIABLE

Age, Date of Birth, Year of Birth.

3.7 RACE

3.7.1 DEFINITION

This data element reflects the race of the participant. When possible, the participant was asked to identify the race or races he/she considers him/herself to be; otherwise, references in the medical record or observations by staff may have been used in the recording of this data element. Race categories are consistent with Federal Directive 15 as follows:

White	A person having origins in any of the original peoples of Europe, the Middle East, or North Africa.
Black or African American	A person having origins in any of the black racial groups of Africa.
Asian	A person having origins in any of the original peoples of the Far East, Southeast Asia, or the Indian subcontinent including, for example, Cambodia, China, India, Japan, Korea, Malaysia, Pakistan, the Philippine Islands, Thailand, and Vietnam.
Native Hawaiian or Other Pacific Islander	A person having origins in any of the original peoples of Hawaii, Guam, Samoa, or other Pacific Islands.
American Indian or Alaska Native	A person having origins in any of the original peoples of North and South America (including Central America), and who maintains tribal affiliation or community attachment.

3.7.2 TYPE

Discrete variable.

3.7.3 VALUE

White	20,832
Black or African American	2,811
Asian	669
Native Hawaiian or other Pacific Islander	113
American Indian or Alaskan Native	185
Mixed race*	191

3.7.4 COMPLETENESS

This variable is 98% complete (i.e., 2% missing value).

3.7.5 NOTE

Respondents have the option of reporting Unknown. Mixed race* is a derived value based upon the selection of more than one race.

3.7.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, HTDS.

3.8 ETHNICITY

3.8.1 DEFINITION

This data element reflects the ethnicity of the participant. When possible, the participant was asked to identify whether he/she considers him/herself to be Hispanic; otherwise, references in the medical record or observations by staff may have been used in the recording of this data element. Ethnicity categories are consistent with Federal Directive 15 as follows:

Hispanic, Latino/a, or Spanish origin	A person of Cuban, Mexican, Puerto Rican, South or Central American, or other Spanish culture or origin, regardless of race.
Not Hispanic, Latino/a, or Spanish origin	A person NOT of Cuban, Mexican, Puerto Rican, South or Central American, or other Spanish culture or origin.

3.8.2 TYPE

Discrete variable.

3.8.3 VALUE

Hispanic, Latino/a, or Spanish origin	3,098
Not Hispanic, Latino/a, or Spanish origin	21,906

3.8.4 COMPLETENESS

This variable is 99% complete (i.e., 1% missing value).

3.8.5 NOTE

Respondents have the option of reporting Unknown.

3.8.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, HTDS.

3.9 MARITAL STATUS

3.9.1 DEFINITION

This data element reflects the marital status of the participant.

3.9.2 TYPE

Discrete variable.

3.9.3 VALUE

Single	10,800
Married	3,365
Domestic Partnership	20
Divorced	332
Separated	45
Widowed	107

3.9.4 COMPLETENESS

This variable is 59% complete (i.e., 41% missing value).

3.9.5 NOTE

Respondents have the option of reporting Unknown.

3.9.6 USE

ATHNdataset.

3.10 1ST LANGUAGE

3.10.1 DEFINITION

This data element reflects the native language spoken by the participant.

3.10.2 TYPE

Discrete variable.

3.10.3 VALUE

Pick list (63 languages as well as Other Selection).

Five most common 1st languages below:

English	11,459
Spanish	751
Dutch	64
Arabic	29
German	14

3.10.4 COMPLETENESS

This variable is 50% complete (i.e., 50% missing value).

3.10.5 USE

ATHNdataset.

3.11 2ND LANGUAGE

3.11.1 DEFINITION

This data element reflects a language learned by the participant after his/her native language either as a resident of an area where the language is in general use or in an area where the language is not generally spoken.

3.11.2 TYPE

Discrete variable.

3.11.3 VALUE

Pick list (63 languages as well as Other selection).

Five most common 2nd languages:

English	559
Spanish	236
Vietnamese	9
Dutch	6
Cantonese	5

3.11.4 COMPLETENESS

This variable is 3% complete (i.e., 97% missing).

3.11.5 USE

ATHNdataset.

3.12 CURRENT STUDENT

3.12.1 DEFINITION

This data element reflects whether the participant is currently a student and includes the range of primary education to post college continuing education.

3.12.2 TYPE

Discrete variable.

3.12.3 VALUE

Yes	7,870
No	17,262

3.12.4 COMPLETENESS

This variable is 99% complete (i.e., 1% missing value).

3.12.5 USE

ATHNdataset.

3.13 EDUCATIONAL LEVEL

3.13.1 DEFINITION

This data element reflects the highest level of formal education completed by the participant. Categories are defined as follows:

None	No formal education, including children too young to attend a formal education program.
Pre-elementary	Pre-kindergarten or kindergarten or other formal education program prior to entering primary school (1st grade).
Primary/Secondary	Grades 1-12, or the equivalent from other countries and home-school programs. Enter the highest grade completed for grades 1-12.
GED	"General Educational Development" a test that may be taken by people who did not complete high school to demonstrate they have high school level skills; a passing score is equivalent to a high school diploma.
Technical school	Any post-high school or post-GED technical, vocational, or trade program.
2-year college degree	Completion of a 2-year program of study at a college or university culminating in one or more Associate degree/s (ex. AA, AS).
4-year college degree	Completion of a 4-year program of study at a college or university culminating in one or more Bachelor's degree/s (ex. BA, BS, AB).
Advanced degree	Completion of a program of study at a university beyond the Bachelor level culminating in one or more Masters (ex. MA, MS, MSW, MBA), Doctorate (ex. PhD, EdD, DrPH) or professional (MD, JD, DDS, DVM) degree/s.
Other	Any other level of formal education that does not fall into one of the other categories.

3.13.2 TYPE

Ordinal variable.

3.13.3 VALUE

None	125
Pre-elementary	1,504
Primary/Secondary	7,437
GED	73
Technical school	448
Some college	243
2-year college degree	1,134
4-year college degree	227
Advanced degree	457
Other	103

3.13.4 COMPLETENESS

This variable is 47% complete (i.e., 53% missing value).

3.13.5 NOTE

Respondents have the option of reporting Unknown.

3.13.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

3.14 EMPLOYMENT STATUS

3.14.1 DEFINITION

This data element reflects the participant's employment status. This does not include volunteer activities. Categories are defined as follows:

Employed full-time	Participant is working less than 35 hours per week.
Employed part-time	Participant is working less than 35 hours per week.
Employment status unknown	The patient's employment status is unknown.
Not Employed - Child	Participant is not working at all because they are a child.
Not Employed - Student	Participant is not working at all because they are a college student.
Not Employed - Homemaker	Participant is a homemaker and does not work outside of the home at all.
Not Employed – Able, but not currently working	Participant does not work and has no other listed reason for not working. This includes those able to work but who have been laid off or cannot find work.
Not Employed - Disabled	Participant does not work and is known to have qualified for disability income and is therefore prohibited from working by law.
Not Employed - Retired	Participant does not work at all and is of retirement age (usually >55).
Not Employed - Other	Participant does not work at all for a reason that is known but not listed in one of the other categories.
Not Employed – Unknown reason	Participant does not work but the reason is not known.

3.14.2 TYPE

Discrete variable.

3.14.3 VALUE

Employed full-time	3,774
Employed part-time	4,400
Not employed – child	1,300
Not employed – student	9,020
Not employed – homemaker	221
Not employed – able but not currently working	495
Not employed – disabled	883
Not employed – retired	546
Not employed – other	121
Not employed –unknown reason	43

3.14.4 COMPLETENESS

This variable is 69% complete (i.e., 31% missing value).

3.14.5 NOTE

Respondents have the option of reporting Employment Status Unknown.

3.14.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

4 DIAGNOSIS

4.1 DIAGNOSIS

4.1.1 DEFINITION

This data element reflects the participant's medical conditions identified by the HTC responsible for providing care.

4.1.2 TYPE

Discrete variable.

4.1.3 VALUE

Number of recorded primary diagnoses	24,831
Number of recorded secondary diagnoses	24,896

4.1.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

4.1.5 NOTE

The above tabulated values are calculated on the basis of the number of diagnoses as opposed to the number of participants. Participants may have more than one diagnosis. These tabulated values include primary and secondary diagnoses.

4.1.6 USE

ATHNdataset, Community Counts HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

4.1.7 RELATED VARIABLE

Primary Bleeding or Clotting Disorder.

4.2 DIAGNOSIS STATUS

4.2.1 DEFINITION

Diagnosis Status tells whether a specific condition is current or definitive.

4.2.2 TYPE

Discrete variable.

4.2.3 VALUE

Active	43,695
History of	831
Inactive	1,468
No history of	3,372
Presumed	9
Evaluation pending	36
Ruled out	128

4.2.4 COMPLETENESS

This variable is more than 99% complete (i.e., less than 1% missing value).

4.2.5 NOTE

The above tabulated values are calculated on the basis of the number of diagnoses as opposed to the number of participants. Respondents have the option of reporting Unknown.

4.2.6 USE

ATHNdataset, Community Counts HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

4.2.7 RELATED VARIABLE

Primary Bleeding or Clotting Disorder, Hepatitis C (HCV) Ever, HIV Ever, Inhibitor Ever, Diagnosis of Other Disease.

4.3 PRIMARY BLEEDING OR CLOTTING DISORDER

4.3.1 DEFINITION

This data element reflects the participant's main bleeding or clotting disorder diagnosis, as determined by the HTC.

4.3.2 TYPE

Derived discrete variable.

4.3.3 VALUE

Factor VIII deficiency	8,695
Factor IX deficiency	2,591
Von Willebrand disease	6,367
Rare disorders	1,174
Platelet disorders	1,679
Thrombophilia	1,392
Venous thrombosis	1,758
Other	1,355

4.3.4 COMPLETENESS

This variable is 98% complete (i.e., 2% missing value).

4.3.5 NOTE

All the reported statistics above are associated with Diagnosis (see 4.1) and only limited to "active" or "history of" Diagnosis Status (see 4.2 above). The details of classifying each major bleeding or clotting disorder are provided in sections 4.4, 4.6, as well as sections 4.8-4.12. Here "Other" refers to the primary diagnosis, e.g., sickle cell disease, which is documented but cannot be grouped into any of seven distinctive types of bleeding or clotting disorder.

4.3.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

4.3.7 RELATED VARIABLE

Primary Diagnosis, Diagnosis Status, Factor VIII Deficiency, Factor IX Deficiency, Von Willebrand disease, Rare Disorders, Platelet Disorders, Thrombophilia, Venous Thrombosis.

4.4 FACTOR VIII DEFICIENCY

4.4.1 DEFINITION

Also called Hemophilia A, a genetic disorder caused by missing or defective factor VIII clotting protein.

4.4.2 TYPE

Discrete variable.

4.4.3 VALUE

Yes	8,695
No	16,561

4.4.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

4.4.5 NOTE

ATHNdataset applies SNOMED (http://www.ihtsdo.org/snomed-ct) for the purpose of recording diagnoses among participants. The number of factor VIII deficiency patients reported above corresponds to Primary Diagnosis = "Yes" as well as Diagnosis Status = "active" or "history of".

4.4.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

4.4.7 RELATED VARIABLE

Primary Bleeding or Clotting Disorder, Severity of Factor VIII Deficiency.

4.5 SEVERITY OF FACTOR VIII DEFICIENCY

4.5.1 DEFINITION

Severity is determined by the result of the baseline diagnostic test measurement of clotting factor activity. Individuals with < 1% factor VIII activity level are classified as having severe hemophilia A, those with 1-5% factor VIII activity level as having moderate hemophilia A, and those with > 5% but <50% of factor VIII activity level as having mild hemophilia A.

4.5.2 TYPE

Ordinal variable.

4.5.3 VALUE

Severe factor VIII deficiency	4,734
Moderate factor VIII deficiency	1,472
Mild factor VIII deficiency	2,349
Unknown	140

4.5.4 COMPLETENESS

This variable is 98% complete (i.e., 2% missing value).

4.5.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

4.5.6 RELATED VARIABLE

Factor VIII Deficiency, Factor VIII Baseline Diagnostic Test.

4.6 FACTOR IX DEFICIENCY

4.6.1 DEFINITION

Also called Hemophilia B or Christmas disease, a genetic disorder caused by missing or defective factor IX clotting protein.

4.6.2 TYPE

Discrete variable.

4.6.3 VALUE

Yes	2,591
No	22,665

4.6.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing value).

4.6.5 NOTE

ATHNdataset applies SNOMED (http://www.ihtsdo.org/snomed-ct) for the purpose of recording diagnoses among participants. The number of factor VIII deficiency patients reported above corresponds to Primary Diagnosis = "Yes" as well as Diagnosis Status = "active" or "history of".

4.6.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, MLOF Research Repository, HTDS.

4.6.7 RELATED VARIABLE

Primary Bleeding or Clotting Disorder, Severity of Factor IX Deficiency.

4.7 SEVERITY OF FACTOR IX DEFICIENCY

4.7.1 DEFINITION

Severity is determined by the result of the baseline diagnostic test measurement of clotting factor activity. Individuals with < 1% factor IX activity level are classified as having severe hemophilia B, those with 1-5% factor IX activity level as having moderate hemophilia B, and those with > 5% but <50% of factor IX activity level as having mild hemophilia B.

4.7.2 TYPE

Ordinal variable.

4.7.3 VALUE

Severe factor IX deficiency	829
Moderate factor IX deficiency	993
Mild factor IX deficiency	712
Unknown	57

4.7.4 COMPLETENESS

This variable is 98% complete (i.e., 2% missing value).

4.7.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, MLOF Research Repository, HTDS.

4.7.6 RELATED VARIABLE

Factor IX Deficiency, Factor IX Baseline Diagnostic Test.

4.8 VON WILLEBRAND DISEASE (VWD)

4.8.1 DEFINITION

Von Willebrand disease is a disorder caused by missing or defective von Willebrand factor, a clotting protein. This variable is a composite of VWD subtypes listed below.

4.8.2 TYPE

Discrete variable.

4.8.3 VALUE OF VWD SUBTYPE

VWD, type 1	5,099
VWD, type 1C	23
VWD, type 2	132
VWD, type 2A	307
VWD, type 2B	204
VWD, type 2M	133
VWD, type 2N	49
VWD, type 3	225
VWD, acquired	28
VWD, type unknown	167

4.8.4 COMPLETENESS

The subtype of VWD is 97% complete (i.e., 3% missing value).

4.8.5 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, Healthy People 2020, NHPCC Quality Metrics, HTDS.

4.8.6 RELATED VARIABLE

Primary Bleeding or Clotting Disorder.

4.9 RARE DISORDERS

4.9.1 DEFINITION

Rare bleeding disorders include deficiencies of coagulation factor I (fibrinogen), factor II (prothrombin), factor V, combined factor V and factor VIII, factor VII, factor X, factor XI, factor XII, factor XIII, congenital deficiency of vitamin K-dependent factors, hereditary connective tissue disorders like Ehlers-Danlos syndrome, PAI-1 (plasminogen activator inhibitor-1), as well as other inherited deficiency such as plasminogen deficiency, protein C deficiency, and protein S deficiency.

4.9.2 TYPE

Discrete variable.

4.9.3 VALUE OF RARE DISORDER SUBTYPE

Factor I deficiency	90
Factor II deficiency	10
Factor V deficiency	63
Factor V and VIII deficiency	7
Factor VII deficiency	368
Factor X deficiency	65
Factor XI deficiency	212
Factor XII deficiency	23
Factor XIII deficiency	66
Deficiency of vitamin K-dependent	
Ehlers-Danlos syndrome	33
PAI-1	51
Plasminogen deficiency	
Protein C deficiency	65
Protein S deficiency	86

4.9.4 COMPLETENESS

The subtype of Rare Disorders is 100% complete (i.e., 0% missing value).

4.9.5 NOTE

Cells with 5 or fewer cases are masked in above table as well as tables below where applicable.

4.9.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, HTDS.

4.9.7 RELATED VARIABLE

Primary Bleeding or Clotting Disorder.

4.10 PLATELET DISORDERS

4.10.1 DEFINITION

Platelet Disorders affect platelet function and impair their ability to start the process of blood clot formation.

4.10.2 TYPE

Discrete variable.

4.10.3 VALUE OF PLATELET DISORDER SUBTYPE

Bernard Soulier syndrome	11
Glanzmann thrombasthenia	66
Grey platelet syndrome	
Hermansky-Pudlak syndrome	13
Platelet function disorder,	464
Platelet function disorder,	7
Release defect	13
Storage pool deficiency	1,062
Platelet dense granule deficiency	
Platelet disorder, type unknown	41

4.10.4 COMPLETENESS

The subtype of Platelet Disorders is 98% complete (i.e., 2% missing value).

4.10.5 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry.

4.10.6 RELATED VARIABLE

Primary Bleeding or Clotting Disorder.

4.11 THROMBOPHILIA

4.11.1 DEFINITION

Thrombophilia is an abnormality of blood coagulation that increases the risk of blood clots in blood vessels.

4.11.2 TYPE

Discrete variable.

4.11.3 VALUE OF THROMBOPHILIAS SUBTYPE

Antiphospholipid antibodies	145
Antithrombin III deficiency	27
Factor V Leiden mutation	315
Hereditary elevated factor VIII	7
Homocysteine and MTHFR	63
Prothrombin G20210A mutation	73
Thrombophilia, hereditary	728
Thrombophilia, type unknown	34

4.11.4 COMPLETENESS

The subtype of Thrombophilia, is 98% complete (i.e., 2% missing value).

4.11.5 NOTE

The Thrombosis Committee has recommended that Thrombophilia is a genetic predisposition for a clinical thrombotic event and therefore discourage its use as a primary diagnosis. HTCs are being asked to report a Deep Vein Thrombosis (DVT) or Pulmonary Embolism (PE) as the Primary Diagnosis, where applicable. When the risk factor is reported, reporting of the more specific thrombophilia (i.e., rather than reporting thrombophilia, hereditary) is preferred.

4.11.6 USE

ATHNdataset, HTDS

4.11.7 RELATED VARIABLE

Primary Bleeding or Clotting Disorder.

4.12 VENOUS THROMBOSIS

4.12.1 DEFINITION

Venous thrombosis is a blood clot that forms within the vein.

4.12.2 TYPE

Discrete variable.

4.12.3 VALUE

Abdominal vein (mesenteric, portal, or renal) thrombosis	67
Cerebral venous sinus thrombosis	22
Deep vein thrombosis	604
Deep vein thrombosis of lower	511
Deep vein thrombosis of upper	124
Pulmonary embolism	415
Pulmonary vein thrombosis	
Splenic vein thrombosis	
Superior vena cava syndrome	
Retinal vein thrombosis	
Subclavian vein thrombosis	
Superior sagittal sinus thrombosis	
Venous thrombosis, type unknown	3

4.12.4 COMPLETENESS

The subtype of Venous Thrombosis is almost 100% complete (i.e., less than 1% missing value).

4.12.5 NOTE

Centers are encouraged to report Deep Vein Thrombosis with more specificity. For example: The Community Counts project has adopted the AHRQ (Agency for Healthcare Research and Quality) categorization based on site of DVT (abdominal, upper extremity, lower extremity).

4.12.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry.

4.12.7 RELATED VARIABLE

Primary Bleeding or Clotting Disorder.

4.13 AGE AT BLEEDING/CLOTTING DISORDER DIAGNOSIS

4.13.1 DEFINITION

This data element reflects the age at which the participant was diagnosed.

4.13.2 TYPE

Derived continuous variable.

4.13.3 VALUE OF MEAN DIAGNOSIS AGE

Factor VIII deficiency	7 years
Factor IX deficiency	9 years
Von Willebrand disease	13 years
Rare disorders	15 years
Platelet disorders	13 years
Thrombophilia	38 years
Venous thrombosis	32 years

4.13.4 COMPLETENESS

This variable is 77% complete for Primary Diagnosis of Factor VIII Deficiency, 78% complete for Factor IX Deficiency, 75% complete for Von Willebrand Disease, 76% complete for Rare Disorders, 88% complete for Platelet Disorders, 80% complete for Thrombophilia, and 80% complete for Venous Thrombosis.

4.13.5 FORMULA

Age of Bleeding Disorder Diagnosis = Year of such a diagnosis – Year of Birth.

4.13.6 USE

ATHNdataset.

4.14 HEPATITIS C (HCV) EVER

4.14.1 DEFINITION

Hepatitis C is a pathogenic RNA virus that preferentially infects liver hepatocytes causing liver fibrosis and hepatocellular carcinoma. HCV consists of 7 genotypes (Type 1 is most prevalent in the U.S. ~ 70%). It is most commonly reported as a co-morbidity to one of the primary bleeding or clotting disorders resulting from contaminated plasma derived factor replacement products in the 1980s.

4.14.2 TYPE

Discrete variable.

4.14.3 VALUE OF HCV DIAGNOSIS STATUS

HCV, active	2,104
HCV, history of	80
HCV, inactive	213
HCV, no history of	380
HCV, ruled out	
HCV, status unknown	78
No HCV diagnosis	22,404

4.14.4 COMPLETENESS

This variable is 99% complete (i.e., 1% missing value).

4.14.5 NOTE

HCV Ever includes any cases with a diagnosis of HCV with a Diagnosis Status = "active" or "history of".

4.14.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, HTDS.

4.14.7 RELATED VARIABLE

HCV Antibody Test, HCV RNA Test.

4.15 HIV EVER

4.15.1 DEFINITION

HIV stands for human immunodeficiency virus which can lead to acquired immunodeficiency syndrome, or AIDS. It is most commonly reported as a co-morbidity to one of the primary bleeding or clotting disorders resulting from contaminated plasma derived factor replacement products in the 1980s.

4.15.2 TYPE

Discrete variable.

4.15.3 VALUE OF HIV DIAGNOSIS STATUS

HIV, active	695
HIV, history of	
HIV, no history of	439
HIV, ruled out	
HIV, status unknown	90
No HIV diagnosis	24,033

4.15.4 COMPLETENESS

This variable is 99% complete (i.e., 1% missing value).

4.15.5 NOTE

HIV Ever includes any cases with a diagnosis of HIV with a Diagnosis Status = "active" or "history of".

4.15.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, HTDS.

4.15.7 RELATED VARIABLE

HIV Antibody Test, HIV Antigen Test, CD4 Cell Test, HIV Viral Load Test.

4.16 INHIBITOR EVER

4.16.1 DEFINITION

Inhibitors develop when the body's immune system stops accepting the factor (factor VIII for hemophilia A and factor IX for hemophilia B) as a normal part of blood. Approximately 15-20% of people with hemophilia will develop an antibody, called an inhibitor, to the product used to treat or prevent bleeding episodes. Inhibitors are more likely to occur in the first 50 exposure days to treatment.

4.16.2 TYPE

Discrete variable.

4.16.3 VALUE OF INHIBITOR DIAGNOSIS STATUS

Inhibitor, active	672
Inhibitor, history of	273
Inhibitor, inactive	260
Inhibitor, no history of	2,400
Inhibitor, ruled out	11
Inhibitor, evaluation pending	6
Inhibitor, presumed	
Inhibitor, status unknown	12
No inhibitor diagnosis	21,627

4.16.4 COMPLETENESS

This variable is almost 100% complete (i.e., less than 1% missing value).

4.16.5 NOTE

Inhibitor Ever includes any cases with a diagnosis of inhibitor with a Diagnosis Status = "active" or "history of" or "inactive'.

4.16.6 USE

ATHNdataset, Community Counts HTC: Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, HTDS.

4.16.7 RELATED VARIABLE

Inhibitor Titers.

4.17 AGE AT HCV, HIV, OR INHIBITOR DIAGNOSIS

4.17.1 DEFINITION

It is the biological age of acquiring hepatitis C virus, HIV or inhibitor.

4.17.2 TYPE

Derived continuous variable.

4.17.3 VALUE OF MEAN DIAGNOSIS AGE

HCV	30 years
HIV	25 years
Inhibitor	10 years

4.17.4 COMPLETENESS

This variable is 57% complete for HCV Ever patients, 54% complete for HIV Ever patients, and 65% complete for Inhibitor Ever patients.

4.17.5 FORMULA

Age of HCV, HIV, or Inhibitor Diagnosis = Year of such a diagnosis – Year of Birth.

4.17.6 USE

ATHNdataset.

4.18 DIAGNOSIS OF OTHER DISEASE

4.18.1 DEFINITION

This could be any secondary condition other than primary bleeding or clotting disorder, HIV, HCV, or inhibitor.

4.18.2 TYPE

Discrete variable.

4.18.3 VALUE

Let's take the example of finding diabetes patients in ATHNdataset. The related SNOMED descriptions are listed as follows:

Diabetes mellitus, active	149
Diabetes mellitus, history of	
Diabetes mellitus type 1, active	24
Diabetes mellitus type 1, inactive	
Diabetes mellitus type 2, active	52

4.18.4 COMPLETENESS

We are unable to evaluate completeness of other diagnoses because they are recorded on an as needed basis.

4.18.5 USE

ATHNdataset.

5 LABORATORY TEST

5.1 FACTOR VIII BASELINE DIAGNOSTIC TEST

5.1.1 DEFINITION

This blood test measures the amount of factor VIII in the blood in order to establish the diagnosis of hemophilia A. The test is performed prior to treatment with factor replacement products. The test result is used to determine if the person has hemophilia A and the severity of the disorder. Normal level of factor VIII plasma is 50-200%. Less than 1% of normal level of factor VIII is considered as having severe disorder, 1-5% is considered as having moderate disorder, and >5% - <50% as having mild disorder.

5.1.2 TYPE

Continuous variable.

5.1.3 VALUE

< 1% of normal factor VIII level	4,734
≥ 1% and ≤ 5% of normal factor VIII level	1,472
> 5% and < 50% of normal factor VIII level	2,349

5.1.4 COMPLETENESS

98% of hemophilia A patients have records of factor VIII baseline diagnostic test (i.e., 2% missing value).

5.1.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics, MLOF Research Repository, HTDS.

5.1.6 RELATED VARIABLE

Severity of Factor VIII Deficiency.

5.2 FACTOR IX BASELINE DIAGNOSTIC TEST

5.2.1 DEFINITION

This blood test measures the amount of factor IX in the blood in order to establish the diagnosis of hemophilia B. The test is performed prior to treatment with factor replacement products. The test result is used to determine if the person has hemophilia B and the severity of the disorder. Normal level of factor IX plasma is 50-200%. Less than 1% of normal level of factor IX is considered as having severe disorder, 1-5% is considered as having moderate disorder, and >5% - <50% as having mild disorder.

5.2.2 TYPE

Continuous variable.

5.2.3 VALUE

< 1% of normal factor IX level	829
≥ 1% and ≤ 5% of normal factor IX	993
> 5% and < 50% of normal factor IX	712

5.2.4 COMPLETENESS

98% of hemophilia B patients have records of factor IX baseline diagnostic test (i.e., 2% missing value).

5.2.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, MLOF Research Repository, HTDS.

5.2.6 RELATED VARIABLE

Severity of Factor IX Deficiency.

5.3 HCV ANTIBODY TEST

5.3.1 DEFINITION

When a person is infected with HCV, the immune system produces antibodies against the virus. Antibodies to HCV can be detected in the blood, usually within two or three months after the virus enters the body. If a person is positive for HCV antibodies, he or she has been exposed to the virus in the past. In the case of the signal-to-cut-off (s/co) ratio presented below, once the signal reaches above 3.8, it was found that there was a 95% chance that the test provided a true positive HCV antibody result.

5.3.2 TYPE

Continuous variable.

5.3.3 VALUE OF SIGNAL-TO-CUT-OFF-RATIO

> 3.8	664
≥ 1 and ≤ 3.8	197
> 0 and < 1	756
= 0	17,053

5.3.4 COMPLETENESS

HCV antibody test was reported for 50% of subjects with hepatitis C.

5.3.5 USE

ATHNdataset.

5.3.6 RELATED VARIABLE

HCV Ever.

5.4 HCV RNA TEST

5.4.1 DEFINITION

HCV contains RNA, which is the genetic material that helps it replicate, i.e., make more copies of itself. A health care provider can order an HCV RNA test to figure out a person's HCV viral load (the amount of HCV currently present in a measurement of blood). Various methods are used to detect HCV RNA, including TMA (transcription-mediated amplification), PCR (polymerase chain reaction), and bDNA (branched DNA). Viral load testing using PCR or TMA are more sensitive than bDNA testing.

5.4.2 TYPE

Counting variable.

5.4.3 VALUE OF VIRAL LOAD

> 800,000 IU/L	553
≥ 615 and ≤ 800,000 IU/L	691
≥ 0 and < 615 IU/L	7,228

5.4.4 COMPLETENESS

HCV RNA test was reported for 30% of subjects with hepatitis C.

5.4.5 NOTE

A viral load of less than 615 IU/L (international units per liter) means there is no detectable hepatitis C virus or it's too low to detect. Additionally, a viral load of more than 800,000 IU/L is high and less than 800,000 IU/L is low. During treatment, a falling viral load is an indication that treatment is succeeding.

5.4.6 USE

ATHNdataset, NHPCC Quality Metrics.

5.4.7 RELATED VARIABLE

HCV Ever.

5.5 HIV ANTIBODY TEST

5.5.1 DEFINITION

When person is infected with HIV, their body responds by producing special proteins to fight the infection called antibodies. It generally takes 2 to 8 weeks for your body to produce antibodies, but in some cases it can take up to six months. A HIV Antibody Test looks for these antibodies in blood, saliva or urine. If antibodies to HIV are detected, it means a person has been infected with HIV.

5.5.2 TYPE

Continuous variable.

5.5.3 VALUE OF SIGNAL-CUT-OFF-RATIO

> 1	67
≥ 0.9 and ≤ 1	13
≥ 0.5 and < 0.9	85

5.5.4 COMPLETENESS

This test was reported for 37% of subjects with HIV.

5.5.5 NOTE

Signal-cut-off-ratios vary from instrument to instrument and also from experiment to experiment. S/co ratios less than 0.9 would be HIV negative. Values between 0.9 and 1.0 would be borderline. And values above 1.0 would be preliminarily positive. These preliminarily positive tests require a confirmatory test before someone is told they are HIV positive.

5.5.6 USE

ATHNdataset.

5.5.7 RELATED VARIABLE

HIV Ever.

5.6 HIV ANTIGEN TEST

5.6.1 DEFINITION

This test can detect HIV up to 20 days earlier than HIV antibody tests. It checks for HIV antigen, a part of the virus that shows up 2-4 weeks after infection. A positive result for the antigen allows treatment to begin earlier and the patient to avoid infecting others. These are blood tests only.

5.6.2 TYPE

Discrete variable.

5.6.3 VALUE

Positive	0
Negative	990

5.6.4 COMPLETENESS

This test was reported for 3% of subjects with HIV.

5.6.5 NOTE

A negative result might mean that a person is not infected with HIV or that the level of antigen is below the detectable limits of the test. If someone suspects that s/he has been exposed to HIV or if s/he is at an increased risk, repeat testing with a combination of HIV antigen/antibody tests is recommended.

5.6.6 USE

ATHNdataset.

5.6.7 RELATED VARIABLE

HIV Ever.

5.7 CD4 CELL TEST

5.7.1 DEFINITION

A CD4 cell is a type of lymphocyte. Lymphocytes are a type of white blood cell. About 15 to 40 percent of your white blood cells are lymphocytes. And they are some of the most important cells in your immune system protecting you from viral infections. Since HIV targets T-helper immune cells, in order to monitor the health of the patient's immune system, a doctor will check CD4 count, the number of CD4 cells in a sample of blood.

5.7.2 TYPE

Counting variable.

5.7.3 VALUE

> 500 cells/mm^3	1,248
≥ 200 and ≤ 500 cells/mm^3	1,253
< 200 cells/mm^3	2,513

5.7.4 COMPLETENESS

CD4 test was reported for 40% of subjects with HIV.

5.7.5 NOTE

A normal CD4 count is more than 500 cells per cubic millimeter (mm^3). The doctor will probably start treatment by the time a CD4 count is under 500 cells/mm^3. If a subject's CD4 count drops to below 200/mm^3, she or he is said to have full-blown AIDS. CD4 counts rise when treatment of HIV infection is effective.

5.7.6 USE

ATHNdataset.

5.7.7 RELATED VARIABLE

HIV Ever.

5.8 HIV VIRAL LOAD TEST

5.8.1 DEFINITION

The HIV viral load test is used primarily to monitor HIV infection over time. It is a quantitative measurement of HIV nucleic acid (RNA) that reports how many copies of the virus are present in the blood. Evidence shows that keeping the viral load levels as low as possible for as long as possible decreases the complications of HIV disease, slows the progression from HIV infection to AIDS, and prolongs life.

5.8.2 TYPE

Counting variable.

5.8.3 VALUE

> 5,000 copies/mL	157
≥ 500 and ≤ 5000 cells/mm^3	123
≥ 40 and < 500 copies/mL	781
< 40 copies/mL	686

5.8.4 COMPLETENESS

CD4 test was reported for 40% of subjects with HIV.

5.8.5 USE

ATHNdataset.

5.8.6 RELATED VARIABLE

HIV Ever.

5.9 HIGH/LOW INHIBITOR TITER

5.9.1 DEFINITION

The presence of an inhibitor is usually confirmed using a specific blood test called the Bethesda inhibitor assay. The assay is now often performed using the Nijmegen modification of the original method to improve test accuracy. The amount of antibody can be measured using this test, and is reported as a number of Bethesda units (BU), or a Bethesda titer.

For low titer inhibitors, the amount in the blood is less than 5 BU. The number and therefore strength of the inhibitor is low. People with low titer inhibitors can sometimes continue to use factor VIII or factor IX products to treat bleeds; they just need a lot more of it. Low titer inhibitors can sometimes resolve on their own.

For high titer inhibitors, the amount of inhibitors found in the blood is at least 5 BU. The number and therefore strength of the inhibitor is high. People with a high titer inhibitor get no benefit from factor VIII or factor IX, no matter how much they infuse.

5.9.2 TYPE

Discrete variable.

5.9.3 VALUE

Hemophilia A, high titer	1,072
Hemophilia A, low titer	4,158
Hemophilia B, high titer	29
Hemophilia B, low titer	708

5.9.4 COMPLETENESS

1,533 factor VIII deficiency subjects as well as 683 factor IX deficiency subjects have inhibitor titer records.

5.9.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

5.9.6 RELATED VARIABLE

Inhibitor Ever.

6 VISIT

6.1 VISIT TYPE

6.1.1 DEFINITION

This data element captures the nature of clinic visits as well as utilization of telephone and written communications between HTCs and participants. The total number of visit counts by 3/31/2015 was 442,581.

6.1.2 TYPE

Discrete variable.

6.1.3 VALUE

Comprehensive care visit	125,227
Consultation	9,408
Diagnostic evaluation	185
Emergency room	10,411
Hospitalization (inpatient)	10,886
Hospitalization (outpatient)	1,473
Office visit	103,547
Non-office visit	7,903
Study visit	3,875
Program/Event	915
Telephone contact	153,095
Written communication	15,656

6.1.4 COMPLETENESS

100% of visit records contain visit type information (0% missing).

6.1.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

6.2 VISIT AGE

6.2.1 DEFINITION

This refers to the biological age of participants in relation to clinic visits.

6.2.2 TYPE

Derived continuous variable.

6.2.3 VALUE

Mean	20 years
Median	14 years
Interquartile range	6-28 years

6.2.4 COMPLETENESS

This variable is 100% complete (0% missing).

6.2.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

6.2.6 FORMULA

Visit Age = Year of Visit – Year of Birth.

6.3 HEIGHT AT COMPREHENSIVE CARE VISIT (CCV)

6.3.1 DEFINITION

This data element reflects the mean, median and interquartile range of participant height values documented during a comprehensive care visit.

6.3.2 TYPE

Continuous variable.

6.3.3 VALUE

Mean	1.5 m
Median	1.7 m
Interquartile range	1.4-1.8 m

6.3.4 COMPLETENESS

This variable is 93% complete (7% missing).

6.3.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

6.3.6 RELATED VARIABLE

BMI at CCV.

6.4 WEIGHT AT CCV

6.4.1 DEFINITION

This data element reflects the mean, median and interquartile range of participant weight values documented during a comprehensive care visit.

6.4.2 TYPE

Continuous variable.

6.4.3 VALUE

Mean	58 kg
Median	60 kg
Interquartile range	29-81 kg

6.4.4 COMPLETENESS

This variable is 94% complete (6% missing).

6.4.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

6.4.6 RELATED VARIABLE

BMI at CCV.

6.5 BODY MASS INDEX (BMI) AT CCV

6.5.1 DEFINITION

This data element reflects the mean, median and interquartile range of participant body mass index measurements documented during a comprehensive care visit. The standard weight status categories for adults are underweight (<18.5), normal weight (≥18.5 to <25), overweight (≥25 to <30), obese (≥30).

6.5.2 TYPE

Derived Continuous variable.

6.5.3 VALUE

Mean	23
Median	22
Interquartile range	17-27

6.5.4 COMPLETENESS

This variable is 87% complete (13% missing).

6.5.5 FORMULA

BMI = Weight (kg) / (Height (m) × Height (m))

6.5.6 NOTE

BMI is interpreted differently for children and teens, even though it is calculated using the same formula as adult BMI. For people whose age below 18, their BMI classification is based upon percentile information among their peers sharing similar age and gender information.

6.5.7 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

6.5.8 RELATED VARIABLE

Height at CCV, Weight at CCV.

6.6 PULSE RATE AT CCV

6.6.1 DEFINITION

This data element reflects the mean, median and interquartile range of participant pulse rate (heart beats per minute) values documented during a comprehensive care visit.

6.6.2 TYPE

Continuous variable.

6.6.3 VALUE

Mean	86 beats/min
Median	81 beats/min
Interquartile range	71-94 beats/min

6.6.4 COMPLETENESS

This variable is 56% complete (44% missing).

6.6.5 USE

ATHNdataset.

6.7 BLOOD PRESSURE AT CCV

6.7.1 DEFINITION

This data element reflects the mean, median and interquartile range of participant blood pressure (BP) values documented during a comprehensive care visit. BP is comprised of systolic and diastolic values.

6.7.2 TYPE

Continuous variable.

6.7.3 VALUE

Mean systolic/diastolic	119/69 mmHg
Median systolic/diastolic	117/68 mmHg
Interquartile range systolic/diastolic	106/60-129/77 mmHg

6.7.4 COMPLETENESS

This variable is 59% complete (41% missing).

6.7.5 USE

ATHNdataset.

6.8 BODY TEMPERATURE AT CCV

6.8.1 DEFINITION

This data element reflects the mean, median and interquartile range of participant body temperature values documented during a comprehensive care visit. Body temperature measures body's ability to generate and get rid of heat.

6.8.2 TYPE

Continuous variable.

6.8.3 VALUE

Mean	97.8°F
Median	97.9°F
Interquartile range	97.3-98.4°F

6.8.4 COMPLETENESS

This variable is 49% complete (51% missing).

6.8.5 USE

ATHNdataset.

6.9 RESPIRATORY RATE AT CCV

6.9.1 DEFINITION

This data element reflects the mean, median and interquartile range of participant respiration rate values documented during a comprehensive care visit. The respiration rate is the number of breaths a person takes per minute. The rate is usually measured when a person is at rest and simply involves counting the number of breaths for one minute.

6.9.2 TYPE

Continuous variable.

6.9.3 VALUE

Mean	20 breaths/min
Median	20 breaths/min
Interquartile range	16-20 breaths/min

6.9.4 COMPLETENESS

This variable is 41% complete (59% missing).

6.9.5 USE

ATHNdataset.

7 BLEED

7.1 AGE AT BLEEDING

7.1.1 DEFINITION

This data element reflects the exact age when bleeding occurs. The total number of bleed events recorded on 3/31/2015 ATHNdataset was 204,188.

7.1.2 TYPE

Derived continuous variable.

7.1.3 VALUE

Mean	24 years
Median	20 years
Interquartile range	10-35 years

7.1.4 COMPLETENESS

This variable is 100% complete (0% missing).

7.1.5 FORMULA

Age at Bleeding = Year of Bleeding – Year of Birth.

7.1.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

7.2 BLEED LOCATION

7.2.1 DEFINITION

This data element reflects categories of anatomical locations in which bleeding occurred.

7.2.2 TYPE

Discrete variable.

7.2.3 VALUE

Joint	91,854
Muscle	25,402
Oral/nasal	22,810
Soft tissue	14,175
Head	2,563
Genitourinary	2,019
Spine	59
Others	14,928

7.2.4 COMPLETENESS

This variable is 85% complete (15% missing).

7.2.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

7.2.6 RELATED VARIABLE

Joint Bleeding.

7.3 JOINT BLEEDING

7.3.1 DEFINITION

This data element reflects the joint anatomical locations in which bleeding occurred. Joint bleeds are bleedings that occur in the space where two bones meet. They happen most often in the ankles, elbows, and knees.

7.3.2 TYPE

Discrete variable.

7.3.3 VALUE

Ankle	29,885
Elbow	24,985
Knee	19,487
Hip	6,901
Shoulder	5,713
Wrist	2,625
Finger	813
Toe	640
Other joints	235

7.3.4 COMPLETENESS

This variable is 99% complete (1% missing).

7.3.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, Healthy People 2020, NHPCC Quality Metric.

7.3.6 RELATED VARIABLE

Bleed Location.

7.4 ANNUAL BLEED RATE (ABR)

7.4.1 DEFINITION

ABR is the number of bleeds a person experiences over the course of one year.

7.4.2 TYPE

Derived counting variable.

7.4.3 VALUE

ABR = 1	9,672
ABR = 2	4,452
ABR = 3	2,825
ABR = 4	1,989
ABR = 5	1,302
ABR = 6	1,419
ABR > 6	7,830

7.4.4 COMPLETENESS

This variable may encounter the problem of under-reporting.

7.4.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

7.5 ANNUAL JOINT BLEED RATE (AJBR)

7.5.1 DEFINITION

AJBR refers to the number of joint bleeds a person has in one year.

7.5.2 TYPE

Derived counting variable.

7.5.3 VALUE

AJBR = 1	5,550
AJBR = 2	2,542
AJBR = 3	1,462
AJBR = 4	1,029
AJBR = 5	630
AJBR = 6	682
AJBR > 6	3,588

7.5.4 COMPLETENESS

This variable may encounter the problem of under-reporting.

7.5.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

8 MEDICATION / BLOOD PRODUCT

8.1 MEDICATION NAME

8.1.1 DEFINITION

The medication name can be generic name or brand name of the drug listed on the prescription. The former refers to the chemical makeup of the drug whereas the latter the name of a drug under which is sold or advertised.

8.1.2 TYPE

Discrete variable.

8.1.3 VALUE

17,764 ATHNdataset participants	A total of 113,938 prescriptions
	2,580 unique medications names

8.1.4 COMPLETENESS

70% of active participants have at least 1 prescription record..

8.1.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

8.1.6 RELATED VARIABLE

Factor Replacement Therapy, Bypassing Agent, Immune Tolerance Induction (ITI).

8.2 MEDICATION TREATMENT TYPE

8.2.1 DEFINITION

The following medication treatment types are available as part of the medication record in the ATHNdataset: 1) Prophylaxis-Continuous; 2) Prophylaxis-Continuous with bypassing agents plus ITI; 3) Prophylaxis-Menstrual bleeding; 4) Prophylaxis-Event-based, short time, or intermittent; 5) Episodic (On-demand); 6) Immune tolerance induction; 7) Other - Non-hemostasis / thrombosis medication; 8) Unknown.

Listed below are definitions for each treatment type.

Prophylaxis - Continuous	Currently use any treatment product on a regular basis to prevent any bleeds or to maintain tolerance to factor or both (even if adherence to the regimen isn't perfect), and is expected to do so indefinitely.
Prophylaxis - Continuous with bypassing agents plus ITI (Immune Tolerance Induction)	Currently uses factor VIII or factor IX concentrates on a regular basis for the primary purpose of eradicating an inhibitor AND uses bypassing agents on a regular basis to prevent any bleeds (even if adherence to the regimen isn't perfect), and is expected to do so indefinitely.
Prophylaxis - Menstrual bleeding	Currently uses treatment product regularly in anticipation of and to prevent excessive menstrual bleeding.
Prophylaxis - Event-based, short time, or intermittent	To prevent anticipated bleeds associated with an event (such as a dental procedure or surgery) or an activity (such as a sports event) on an intermittent basis, repeatedly over a short period of time or on a single occasion; OR on a regular basis to prevent any bleeds, and is expected to do so for an extended period of time (weeks to months) but not indefinitely (such as a sports season). This may be as follow-up to a bleed where the treater determines that a short course of prophylaxis is indicated.
Episodic	Currently uses treatment product ONLY for treatment of bleeds.

Immune tolerance Induction (ITI)	Currently uses factor VIII or factor IX concentrates on a regular basis for the primary purpose of eradicating an inhibitor (even if adherence to the regimen isn't perfect) and does not also use bypassing agents for continuous prophylaxis.
Other - Non-hemostasis / thrombosis medication	This can be used to describe the treatment type of non-bleeding/clotting drugs.
Unknown	Medication treatment type is not specified.

8.2.2 TYPE

Discrete variable.

8.2.3 COMPLETENESS

This variable is 98% complete (i.e., 2% missing).

8.2.4 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

8.2.5 RELATED VARIABLE

Factor VIII Deficiency Continuous Prophylaxis, Factor IX Deficiency Continuous Prophylaxis.

8.3 FACTOR REPLACEMENT THERAPY

8.3.1 DEFINITION

The basic treatment to stop or prevent bleeding in people with hemophilia A and B is factor replacement therapy. This is the infusion (injection into the bloodstream) of factor VIII and IX concentrates to prevent or control bleeding. These concentrates come from two sources: 1) human plasma (a component of blood); 2) a genetically engineered cell line made by DNA technology, called recombinant. Long acting products are a special group of recombinant products.

Brand names of factor VIII and IX products approved for use in the U.S. are listed in the following table:

Brand names of recombinant factor VIII concentrates
Advate, Adynovate, Eloctate, Helixate FS, Kogenate FS, NovoEight, Recombinate, ReFacto AF, Xyntha.
Brand names of plasma derived factor VIII concentrates
Alphanate, Humate-P, Koate DVI, Hemophil, Monoclate-P, Monarc-M.
Brand names of recombinant factor IX concentrates
Alprolix, BeneFix, IXinity, Rixubis.
Brand names of plasma derived factor IX concentrates
AlphaNine SD, Bebulin VH, MonoNine, Profilnine SD, Proplex T.

8.3.2 TYPE

Discrete variable.

8.3.3 VALUE

Prescriptions of recombinant factor VIII concentrates	39,536
Prescriptions of human plasma derived factor VIII concentrates	7,239
Prescriptions of recombinant factor IX concentrates	10,641

Prescriptions of human plasma derived factor IX concentrates	2,299

8.3.4 NOTE

All the counts in above table are irrespective of treatment type. Also note, once the number of long-acting products includes more than one product, ATHN will consider separating into a separate group. Treatment type qualifies the reason that the prescription was written.

8.3.5 COMPLETENESS

52% of the prescriptions are for factor VIII and factor IX replacement products.

8.3.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

8.3.7 RELATED VARIABLE

Medication Name, Factor Concentrate dosage, Factor Replacement Frequency.

8.4 BYPASSING AGENT

8.4.1 DEFINITION

If someone has hemophilia and high titer levels of inhibitors, they are often treated with bypassing agents. Instead of replacing missing factor, bypassing agents go around (or bypass) the need for factor VIII or factor IX, since response to factor is blocked by the body's immune system. This helps the body form a clot and stop bleeding.

The following table lists all the recombinant bypassing products currently available to treat hemophilia with inhibitors:

Bypassing Agents
Autoplex T, Feiba, NovoSeven RT.

8.4.2 TYPE

Counting variable.

8.4.3 VALUE

Prescriptions of recombinant bypassing agent	4,096

8.4.4 COMPLETENESS

4% of the prescriptions are for recombinant bypassing products.

8.4.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

8.4.6 RELATED VARIABLE

Medication Name.

8.5 IMMUNE TOLERANCE INDUCTION (ITI)

8.5.1 DEFINITION

With ITI therapy, factor concentrate is given regularly over a period of time until the body is trained to recognize the treatment product without reacting to it. When immune tolerance induction is successful, the inhibitors disappear and the patient's response to factor concentrates returns to normal.

8.5.2 TYPE

Discrete variable.

8.5.3 VALUE

Prescriptions of immune tolerance induction	2,566

8.5.4 COMPLETENESS

2% of the prescriptions are for immune tolerance induction.

8.5.5 NOTE

A prescription is flagged as immune tolerance induction if Medication Treatment Type is equal to "Immune Tolerance Induction".

8.5.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

8.5.7 RELATED VARIABLE

Medication Name.

8.6 DDAVP

8.6.1 DEFINITION

DDAVP (desmopressin acetate) is an antidiuretic hormone that helps stop bleeding. In patients with mild hemophilia, it can be used for joint and muscle bleeds, for bleeding in the mucous membranes of the nose and mouth, and before and after surgery. It comes in an injectable form and a nasal spray (Stimate).

8.6.2 TYPE

Counting variable.

8.6.3 VALUE

Prescriptions of DDAVP	6,962

8.6.4 COMPLETENESS

6% of the prescriptions are for DDAVP.

8.6.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

8.6.6 RELATED VARIABLE

Medication Name.

8.7 FACTOR CONCENTRATE DOSAGE

8.7.1 DEFINITION

The dosage of factor VIII or factor IX product is determined based on weight, type of disorder, severity, desired factor level, and the type of bleeding episode the person is experiencing. A hematologist will calculate dosage needed for major, minor and prophylactic doses. The dosage will be indicated in units of factor. In addition, dosage may be adjusted based on how the person's body uses the factor product.

8.7.2 TYPE

Derived continuous variable.

8.7.3 VALUE

Mean	1,767 units
Median	1,250 units
Interquartile range	60-2,600 units

8.7.4 COMPLETENESS

79% of the prescriptions associated with factor VIII and factor IX replacements include dosing units (i.e., 21% missing).

8.7.5 USE

ATHNdataset.

8.7.6 RELATED VARIABLE

Factor Replacement Therapy.

8.8 FACTOR REPLACEMENT FREQUENCY

8.8.1 DEFINITION

This data element reflects the prescribed interval at which medications, including factor VIII or factor IX concentrates, are to be used.

8.8.2 TYPE

Ordinal variable.

8.8.3 VALUE

5 most commonly observed frequency patterns:

3 times per week	10,274
2 times per week	6,134
Every other day	5,601
Daily	3,553
Weekly	2,219

8.8.4 COMPLETENESS

49% of the prescriptions associated with factor VIII and factor IX replacements include dosing frequency information (i.e., 51% missing).

8.8.5 USE

ATHNdataset.

8.8.6 RELATED VARIABLE

Factor Replacement Therapy.

8.9 HEMOPHILIA A PATIENTS CONTINUOUS PROPHYLAXIS

8.9.1 DEFINITION

Prophylaxis is the regular infusion of clotting factor concentrates in order to prevent bleeding. Prevention of bleeding is now the goal of treatment, allowing people with hemophilia to remain active and participate more fully in daily life. The Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation (NHF) recommends prophylaxis as an optimal therapy for children with severe hemophilia A. See definitions of continuous prophylaxis in Section 8.2.

8.9.2 TYPE

Discrete variable.

8.9.3 VALUE

Yes	3,479
No	5,159
Unknown	57

8.9.4 COMPLETENESS

This variable is 99% complete (i.e., 1% missing).

8.9.5 NOTE

A factor VIII deficient participant is included as being on prophylaxis if the Medication Treatment Type is either "Prophylaxis-Continuous" or "Prophylaxis- Continuous with bypassing agents plus ITI" or "Prophylaxis-Menstrual bleeding" AND the prescription ending date is not given.

8.9.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

8.9.7 RELATED VARIABLE

Medication Treatment Type, Hemophilia A Continuous Prophylaxis Products.

8.10 HEMOPHILIA A PATIENTS CONTINUOUS PROPHYLAXIS PRODUCTS

8.10.1 DEFINITION

For hemophilia A patients, prophylaxis products include recombinant factor VIII concentrates, human plasma derived factor VIII concentrates, bypassing agents, or DDAVP.

8.10.2 TYPE

Discrete variable.

8.10.3 VALUE

Recombinant prescriptions	4,950
Plasma derived prescriptions	338
Bypassing agent prescriptions	141
DDVP prescriptions	11

8.10.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing).

8.10.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry, NHPCC Quality Metrics.

8.10.6 RELATED VARIABLE

Hemophilia A Patients Continuous Prophylaxis.

8.11 HEMOPHILIA B PATIENTS CONTINUOUS PROPHYLAXIS

8.11.1 DEFINITION

Prophylaxis is the regular infusion of clotting factor concentrates in order to prevent bleeding. Prevention of bleeding is now the goal of treatment, allowing people with hemophilia to remain active and participate more fully in daily life. The Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation (NHF) recommends prophylaxis as an optimal therapy for children with severe hemophilia b. See definitions of continuous prophylaxis in Section 8.2.

8.11.2 TYPE

Discrete variable.

8.11.3 VALUE

Yes	612
No	1,953
Unknown	26

8.11.4 COMPLETENESS

This variable is 99% complete (i.e., 1% missing).

8.11.5 NOTE

A factor IX deficient participant is included as being on prophylaxis if the Medication Treatment Type is either "Prophylaxis-Continuous" or "Prophylaxis- Continuous with bypassing agents plus ITI" or "Prophylaxis-Menstrual bleeding" AND the prescription ending date is not given.

8.11.6 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

8.11.7 RELATED VARIABLE

Medication Treatment Type, Hemophilia B Continuous Prophylaxis Products.

8.12 HEMOPHILIA B CONTINUOUS PROPHYLAXIS PRODUCTS

8.12.1 DEFINITION

For hemophilia B patients, prophylaxis products include recombinant factor IX concentrates, human plasma derived factor IX concentrates, bypassing agents, or DDAVP.

8.12.2 TYPE

Discrete variable.

8.12.3 VALUE

Recombinant prescriptions	686
Plasma derived prescriptions	117
Bypassing agent prescriptions	17

8.12.4 COMPLETENESS

This variable is 100% complete (i.e., 0% missing).

8.12.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

8.12.6 RELATED VARIABLE

Medication Treatment Type, Hemophilia B Patients Continuous Prophylaxis.

9 PROCEDURE/SURGERY

9.1 JOINT REPLACEMENT

9.1.1 DEFINITION

Joint replacement procedures are designed to replace damaged cartilage and any associated loss of bone structure.

9.1.2 TYPE

Counting variable.

9.1.3 VALUE

Number of joint replacements	522

9.1.4 COMPLETENESS

1.3% of active participants have at least 1 joint replacement procedure.

9.1.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

9.2 RADIONUCLIDE SYNOVECTOMY

9.2.1 DEFINITION

Radionuclide synovectomy is a method of treatment for several acute and chronic inflammatory joint disorders. A small amount of a beta-emitting radionuclide is injected into the affected joint delivering a radiation dose to the synovia.

9.2.2 TYPE

Counting variable.

9.2.3 VALUE

Number of radionuclide synovectomies	392

9.2.4 COMPLETENESS

0.8% of active ATHNdataset participants have at least 1 radionuclide synovectomy surgery.

9.2.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

9.3 OPEN SYNOVECTOMY

9.3.1 DEFINITION

Synovectomy is an operation performed to remove partial or all the synovial membrane of a joint. A Synovectomy may be performed as an open surgical procedure or with the aid of arthroscopy (see Section 9.4).

9.3.2 TYPE

Counting variable.

9.3.3 VALUE

Number of open synovectomies	194

9.3.4 COMPLETENESS

0.6% of participants have at least 1 open synovectomy surgery.

9.3.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

9.3.6 RELATED VARIABLE

Arthroscopic Synovectomy.

9.4 ARTHROSCOPIC SYNOVECTOMY

9.4.1 DEFINITION

This is a minimally invasive surgical procedure on a joint in which an examination and treatment of damage is performed using an arthroscope, an endoscope that is inserted into the joint through a small incision.

9.4.2 TYPE

Counting variable.

9.4.3 VALUE

Number of arthroscopic synovectomies	510

9.4.4 COMPLETENESS

1.4% of participants have at least 1 open synovectomy surgery.

9.4.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

9.4.6 RELATED VARIABLE

Open Synovectomy.

9.5 ARTHRODESIS

9.5.1 DEFINITION

Arthrodesis is the artificial induction of joint ossification between two bones by surgery.

9.5.2 TYPE

Counting variable.

9.5.3 VALUE

Number of arthrodesis	147

9.5.4 COMPLETENESS

0.4% of participants have at least 1 arthrodesis surgery.

9.5.5 USE

ATHNdataset, Community Counts: HTC Population Profile, Mortality Report, Registry.

9.6 HYSTERECTOMY

9.6.1 DEFINITION

A hysterectomy is a procedure to remove a woman's uterus.

9.6.2 TYPE

Counting variable.

9.6.3 VALUE

Number of hysterectomies	150

9.6.4 COMPLETENESS

1.6% of female participants have had a hysterectomy.

9.6.5 USE

ATHNdataset.