A cross-sectional study of females with congenital bleeding disorders enrolled in the ATHNdataset

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Background

The number of women with bleeding disorders cared for at hemophilia treatment centers (HTCs) is increasing because of improved recognition of their distinct health-care needs.[1,2] However, there is still work to be done with regard to recognizing symptoms, ensuring accurate diagnoses and providing specific treatment.[1,3,4] An important step in improving care for women with bleeding disorders is identifying the cohort and evaluating who is currently being cared for at HTCs. The ATHNdataset is an important surveillance tool for persons with bleeding disorders cared for at HTCs. As of March 2016, over 29,000 male and female subjects are included in the dataset. In this study, we sought to report the demographic and clinical data of female participants in the ATHNdataset in order to better define the female population cared for at federally funded HTCs.

Study Design

The American Thrombosis and Hemostasis Network is a non-profit 501(c)3 organization founded in 2006 whose primary goal is to advance the care of individuals affected by bleeding and clotting disorders. The ATHNdataset is a secure HIPAA-compliant limited national health data set created to monitor trends and provide surveillance of those subjects cared for at participating HTCs. The ATHNdataset was queried to identify the following information for female participants with congenital bleeding disorders:

- primary bleeding disorder diagnosis
- demographic data
- bleeding event information
- treatment types
- viral infection
- mortality

Descriptive statistics were employed. Data presented is as of March 31, 2016. Deficiency was defined as <50% for factors II, V, VIII, VII, X, XI, XIII and <200 mg/dL for fibrinogen deficiency and <30% for either VWF:RCo or VWF:Ag for Von Willebrand disease (VWD) and 30–50% for low VWF.

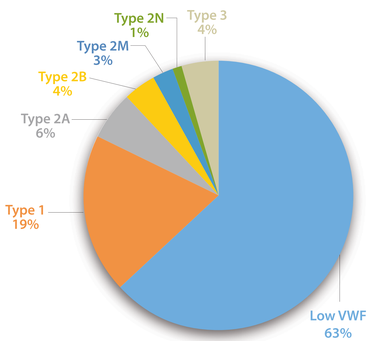
Results

	N (%)
Total Number of Patients Enrolled in ATHNdataset	29,005
Number of FEMALE Congenital Bleeding Disorder Patients in ATHNdataset	7265 (25.0)
Median Age of Cohort	21 y [IQR 15-37]
Race:	
White	6206 (85.4)
African American	598 (8.2)
Asian	153 (2.1)
Mixed Race	69 (0.9)
Native Hawaiian or Pacific Islander	26 (0.4)
American Indian or Alaska Native	60 (0.8)
Unknown	153 (2.1)
Ethnicity:	
Hispanic	878 (12.1)
Non Hispanic	6311 (86.9)
Unknown	76 (1.0)

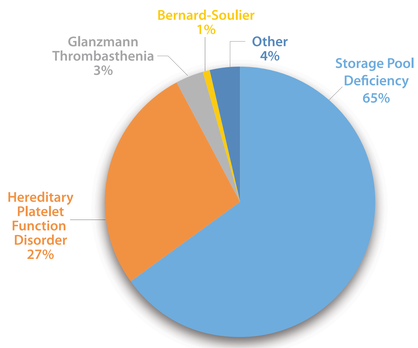
	N (%)
Primary Diagnoses for Cohort:	
VWD	4,620 (62)
Qualitative Platelet Disorders	1,295 (17.8)
Rare Bleeding Disorders	578 (8.0)
Hemophilia A	569 (7.8)
Hemophilia B	203 (2.8)

Rare bleeding disorders were defined as deficiency of factor II, V, combined V and VIII, VII, X, XI, XIII, or fibrinogen; Osler-Weber-Rendu syndrome.

VWD Subtypes

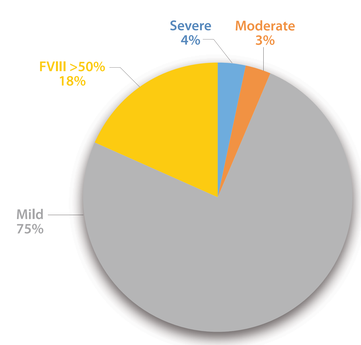


Platelet Disorders

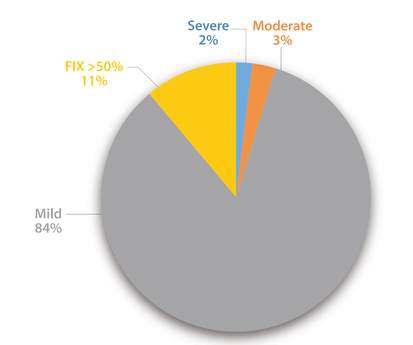


For subjects with a reported VWF activity, 23.3% meet NHLBI criteria for diagnosis of VWD. 76.7% would be classified as having low VWF.

Hemophilia A



Hemophilia B



Bleeding symptoms:

- 645 (8.9%) reported joint bleeding:
  rare disorders > VWD > hem A > hem B > qualitative platelet disorders
- 778 (10.7%) reported reproductive tract bleeding:
  Menstrual bleeding: 765
  Postpartum bleeding: 12
  Postmenopausal bleeding: 1

Medical treatment options utilized for reproductive tract bleeding:

- Desmopressin – 398 patients
- Aminocaproic Acid – 323 patients
- Tranexamic Acid – 216 patients
- Oral Contraceptive – 171 patients
- Plasma – 142 patients (all with VWD)
- IUD – 12 patients
- Depo Provera – 12 patients

Infectious complications:

- 10 women were reported to be infected with HIV (0.1% of cohort).
- 104 women were reported to be infected with HCV (1.4% of cohort).

Age at diagnosis:

- Median age at diagnosis was 13 years (std dev 15 yrs) among 5,438 diagnosis dates.
- Women with a qualitative platelet disorder or VWD were more likely to be diagnosed before 18 years than women with other disorders.

Conclusions

- The ATHNdataset provides a snapshot of females cared for at federally funded HTCs.
- VWD is the most common bleeding disorder in this cohort; however, the majority of patients with VWD do not meet NHLBI diagnostic criteria.
- Reproductive-type bleeding was reported in 10.7% of the cohort.
- Further work is needed to fully capture the impact of bleeding disorders and optimize diagnosis and treatment in women with bleeding disorders.

References

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