World Bleeding Disorders Registry

Hemophilia and VWD Data Set

V 1.4 - April 2025

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BASELINE DATA - VWD

Unique form for VWD patients

Patient Consent

Has the patient consented to participate in the WBDR?	Yes No

Demographics

Date of registration in WBDR	*Automatically filled*
Date of birth	DD-MM-YYYY
Sex	Male Female Other Unknown
Age at diagnosis in years	[integer]
Country of residence	List of countries
Bleeding disorder	*Automatically filled* Hemophilia VWD Hemophilia & VWD
Clinic	Automatically filled
Date of first visit to HTC	DD-MM-YYYY

Diagnostic information

Reason for diagnosis	[dropdown menu] Family History Symptoms of VWD (bleeding) Unknown
VWD Type	[dropdown menu] -Type 1 VWD -Type 2 VWD -Type 3 VWD -Platelet-type VWD -Type Unknown *If Type 1 VWD, please specify* [dropdown menu] Type 1 (30-50 IU/dL + bleeding; or low VWF) Type 1<30 IU/dL Type 1 C Unclassified *If Type 2 VWD, please specify* Type 2A Type 2B Type 2M Type 2N Unclassified

Diagnostic tests

1. Bleeding Assessment Tool (BAT) Score pre-diagnosis or at the time of diagnosis

Bleeding Assessment Tool Score – pre-diagnosis or at the time of diagnosis	[dropdown menu] Done Not done Unknown
Date of BAT score	DD-MM-YYYY
BAT used	[Dropdown menu] ISTH MCMDM-1VWD Condensed MCMDM-1VWD Self-BAT Other Unknown

	If Other, please specify [open text]
BAT score	[open field]

2. FVIII coagulant activity assay

Date of FVIII coagulant activity assay	DD-MM-YYYY
FVIII coagulant activity level (FVIII:C) (%)	[open field] (integer, 0-100) *If level is unknown:* [checkbox] 'unknown'

3. VWF antigen test (VWF:Ag)

Date of VWF antigen test	DD-MM-YYYY
VWF antigen (VWF:Ag) level (%)	[open field] (integer, 0-100) *If level is unknown:* [checkbox] 'unknown'

4. VWF platelet-dependent activity

VWF platelet-dependent activity test used	[Dropdown menu] VWF:RCo VWF:GPIbR VWF:GPIbM VWF:Ab, Unknown
Date of VWF platelet-dependent activity test	DD-MM-YYYY
VWF platelet-dependent activity level (%)	[open field] (integer, 0-100) *If level is unknown:* [checkbox] 'unknown'

5. VWF collagen binding (VWF:CB) activity

Date of VWF collagen binding activity test	DD-MM-YYYY
VWF collagen binding (VWF:CB) activity level (%)	[open field] (integer, 0-100) *If level is unknown:* [checkbox] 'unknown'

6. VWF multimer analysis

Date of multimer analysis	DD-MM-YYYY
VWF multimer analysis result	[dropdown menu] -Normal -Loss of high molecular weight multimers (HMW) -Loss of high and intermediate molecular weight multimers -Absent -Other -Unknown *If other, please specify* [open text]

7. Ristocetin-induced platelet aggregation (RIPA)

Date of RIPA test	DD-MM-YYYY
Is there an enhanced response to low dose ristocetin?	[dropdown menu] -Yes -No -Unknown

8. VWF:FVIIIB binding assay

Date of VWF:FVIIIB binding assay	DD-MM-YYYY
VWF:FVIIIB binding assay result	[dropdown menu] -Normal -Decreased binding -Unknown

Clinical History

Bleeding history	[dropdown menu] Has had ≥1 bleed in lifetime Has never had a bleed Unknown *If yes*, please enter patient's age at first bleed (months) [open text, integer] *If age at first bleed is unknown:* [checkbox] 'unknown'

VWF Inhibitor History	[dropdown menu] Inhibitor in the past Currently has an inhibitor Never had an inhibitor Unknown
Treatment History	[dropdown menu] -Has received treatment in the past -Never received treatment -Unknown If answer is "has received treatment in the past" (optional field): [checkbox] – multiple answers possible -Factor/Cryoprecipitate/Plasma -Desmopressin -Hormonal treatment -Anti-fibrinolytic -Unknown
Prophylaxis History	-Has received prophylaxis in the past -Never received prophylaxis -Unknown *If has received prophylaxis in the past:* Please enter patient's age at start of first prophylaxis (months) [open text, integer] *If age* at start of first prophylaxis *is unknown:* [checkbox] 'unknown'

Genetic testing

Date of genetic testing	DD-MM-YYYY
DNA sequencing technique	[dropdown menu] Sanger sequencing of a specific domain NGS analysis MLPA analysis Whole genome sequencing Unknown Other
If Other, Please specify DNA sequencing technique	[open field]
DNA Variant type	[dropdown menu] Inversion Large structural variant (≥ 50 bp)

	Nonsense Frameshift Small insertion or deletion (indel) (<50 bp) Splice Missense Synonymous Promoter UTR Other Unknown
If Other **Please specify DNA variant type**	[open field]

Blood Group

Blood group	A B O AB Unknown
Rh Factor	Positive Negative Unknown

BASELINE DATA – HEMOPHILIA

Unique for Hemophilia Patients

Patient Consent

Has the patient consented to participate in the WBDR?	Yes No

Demographics

Date of registration in WBDR	*Automatically filled*
Date of birth	DD-MM-YYYY
Sex	Male Female Other Unknown
Country of residence	List of countries
Bleeding disorder	*Automatically filled* Hemophilia VWD Hemophilia & VWD
Clinic	*Automatically filled*
Date of first visit to HTC	DD-MM-YYYY

Diagnostic information

Reason for diagnosis	[dropdown menu] Family History Symptoms of VWD (bleeding) Unknown
Date of Diagnosis	DD-MM-YYYY
Hemophilia Type	A B Unknown
Hemophilia Severity	Mild (>5%) Moderate (1-5%) Severe (<1%) Unknown
Factor level test	Done Not done No capacity to measure at HTC Unknown

Factor VIII or IV level (%)	[open field]
Date of test	DD-MM-YYYY

Clinical history

Bleeding history	[dropdown menu] Has had ≥1 bleed in lifetime Has never had a bleed Unknown *If yes*, please enter patient's age at first bleed (months) [open text, integer] *If age at first bleed is unknown:* [checkbox] 'unknown'
Joint bleeding history	[dropdown menu] Has had ≥1 joint bleed in lifetime Has never had a joint bleed Unknown *If yes*, please enter patient's age at first joint bleed (months) [open text, integer] *If age at first joint bleed is unknown:* [checkbox] 'unknown'
Treatment History	[dropdown menu] Has received treatment in the past Never received treatment Unknown
Prophylaxis History	Has received prophylaxis in the past Never received prophylaxis Unknown *If has received prophylaxis in the past:* Please enter patient's age at start of first prophylaxis (months) [open text, integer] *If age* at start of first prophylaxis *is unknown:* [checkbox] 'unknown'
Inhibitor History	[dropdown menu] Inhibitor in the past Currently has an inhibitor Never had an inhibitor Unknown
Has any family member ever had an inhibitor?	Yes No

	Unknown

Genetic testing

Date of genetic testing	DD-MM-YYYY
DNA variant	[dropdown menu] Intron 22 inversion Intron 1 inversion Not Done Unknown Other If Other, please specify DNA variant
If Other **Please specify DNA variant**	[open field]
DNA Variant type	[dropdown menu] Inversion Large structural variant (≥ 50 bp) Nonsense Frameshift Small insertion or deletion (indel) (<50 bp) Splice Missense Synonymous Promoter UTR Other Unknown If Other, please specify DNA variant type
If Other **Please specify DNA variant type**	[open field]

Blood Group

Blood group	A B O AB Unknown
Rh Factor	Positive Negative Unknown

BASELINE VISIT & FOLLOW-UP VISIT

For both VWD & Hemophilia patients

Bleeding events assessment

Baseline Visit (BV): Total number of bleeds experienced in the past 6 months **Follow-up Visit (FUV): Total number of bleeds experienced since the last clinic visit**	[open field]
BV: Location of bleeds experienced in the past 6 months **FUV: Location of bleeds experienced since the last clinic visit**	[checkbox] – multiple answers possible Joints Muscles CNS Heavy menstrual bleeding Gastrointestinal bleeding Neck/Throat Other
If Neck/throat **Indicate total number of neck/throat bleeds**	[open field]
If neck/throat **Of these neck/throat bleeds, how many were traumatic?**	[open field]
If heavy menstrual bleeding, **Indicate number of heavy menstrual bleeds**	[open field]
If gastrointestinal bleeding, **Indicate number of gastrointestinal bleeds**	[open field]
If gastrointestinal bleeding, **Of these gastrointestinal bleeds, how many were traumatic?**	[open field]
If joints **Indicate number of joint bleeds**	[open field]
If joints **Of these joint bleeds, how many were traumatic?**	[open field]
If muscles **Indicate number of muscle bleeds**	[open field]

If muscles **Of these muscle bleeds, how many were traumatic?**	[open field]
If CNS **Indicate total number of CNS bleeds**	[open field]
If CNS **Of these CNS bleeds, how many were traumatic?**	[open field]
If Other **Indicate number of bleeds for Other**	[open field]
If Other, **Of these other bleeds, how many were traumatic?**	[open field]

Target joints

Target joints	Yes No Unknown
***If yes*, indicate the number of target joints**	[open field]

Inhibitor assessment

Inhibitor assessment in the past 6 (BV) months or since last visit (FUV)	[dropdown menu] -Yes, inhibitor testing done -Suspected based on clinical response, testing not available -Suspected based on clinical response, testing not done -Not suspected, inhibitor testing not done -Unknown

Admission to hospital

Admission to hospital in the past 6 months (BV) or since last visit (FUV)	Yes No Unknown

Treatment

BV: Treatment received in the past 6 months	Yes No Unknown
FUV: any new treatment started, or change in existing treatment, since last clinic visit	No access to treatment products at the time of bleed(s)

Adverse event

Adverse events in the past 6 months (BV) or since last visit (FUV)	Yes No Unknown

Pregnancy

BV: In the past, has the patient given birth, had a pregnancy termination or miscarriage? **FUV: Since last visit, has the patient given birth, had a pregnancy termination or miscarriage?**	[dropdown menu] Yes No Unknown

Inhibitor Assessment module

Date of test	DD-MM-YYYY
Inhibitor test result	Positive Negative
Titre (BU/ml)	[open field]
Type of test	Bethesda Nijmegen-Bethesda Mixing study (aPTT study) Other Unknown

Hospital Admission module

Start date	DD-MM-YYYY
Number of days hospitalized	[open field]
Reason for hospitalization	1. Not related to bleeding disorder 2. Related to bleeding disorder *If related to bleeding disorder, multiple choice (checkbox):* • Intracranial hemorrhage • Intra-abdominal bleeding • Gastrointestinal bleeding • Neck hematomas • Psoas muscle bleeding • Other muscle bleeding • Joint Bleeding • Soft tissue bleeding

	• Other bleeding • Thromboembolic event • Acute heavy menstrual bleed • Hemorrhagic ovarian cyst • Primary post-partum hemorrhage • Secondary post-partum hemorrhage • Epistaxis • Surgery • Other If Other or Surgery, please specify [open text]

Treatment module

Start date	DD-MM-YYYY
Indication	Prophylaxis On demand ITI Surgery/procedure Trauma with no known bleed Selective Bleed Prevention (i.e. before activity) Other Unknown
Drug	[dropdown menu] *List of drugs*
Strength	[open field]
Unit	BU/ml mg IU VWF:RCo units/kg Other
Number of intakes	[open field]
Time units	Daily Every other day Every third day Every fourth day Once per weekTwice per week Three times per week Loading dose, once per week, for Emicizumab

	Maintenance dose, once per week, for Emicizumab Maintenance dose, every 2 weeks, for Emicizumab Maintenance dose, every 4 weeks, for Emicizumab Once every 2 months Other
End date	DD-MM-YYYY
Reason for stopping	Treatment failure (not incl. inhibitors) Adverse event (not incl. inhibitors) Inhibitors Switch treatment Change of dose/regime On demand treatment complete Other ITI: partial success ITI: success ITI: other Loading dose, Emicizumab

Adverse Events module

Date of onset	DD-MM-YYYY
Date of resolution	DD-MM-YYYY
Events	Infection Allergic reaction including anaphylaxis Thromboembolic event Thrombotic microangiopathy Lack of efficacy Injection site reaction Other
If other Please specify adverse event	[open field]
Interventions:	[open field]

Pregnancy module

Date of delivery or miscarriage/termination	DD-MM-YYYY
Bleeding during pregnancy	[dropdown menu] Yes No Unknown
Pregnancy outcome	[dropdown menu] – single answer Live birth Miscarriage/termination
***If live birth*, Type of delivery**	C-section Vaginal
***If live birth,* Primary postpartum hemorrhage (first 24 hours)**	[dropdown menu] Yes No Unknown
***If live birth,* Secondary postpartum hemorrhage (after 24 hours)**	[dropdown menu] Yes No Unknown
***If miscarriage/termination*, Excessive bleeding in case of a miscarriage or termination**	[dropdown menu] Yes No Unknown

Height & Weight

Height (cm)	[open field]
Weight (kg)	[open field]

Comorbidities

HIV status	[dropdown menu] Positive Negative Unknown Testing not done
If Positive **Date of HIV-positive diagnosis**	DD-MM-YYYY
If Positive **Is HIV positive status related to bleeding disorders treatment?**	Yes No Unknown
HCV status	[dropdown menu] No infection Active infection (HCV PCR+) Infection resolved with treatment Infection resolved spontaneously Testing not done Unknown
If Active infection (HCV PCR+) **Date of diagnosis**	DD-MM-YYYY
If Infection resolved with treatment or Infection resolved spontaneously **Date infection was resolved**	DD-MM-YYYY
Has the patient been diagnosed with any other disease?	None Respiratory Disease Cardiovascular Disease Hypertension Kidney disease Cancer Diabetes Osteoarthritis Osteoporosis Rheumatoid arthritis Obesity Anxiety Depression Chronic liver disease Dementia Immunosuppression Endometriosis Other
***If Other,* please specify**	[open field]

***If Cancer*, please specify**	Lung Breast Colorectum Prostate Stomach Liver Other
If Cancer = Other **Please specify**	[open field]

Employment, education, marital status

Employment status	[dropdown menu] • Employed full-time • Employed part-time (due to Bleeding Disorder) • Employed part-time • Not employed (due to Bleeding Disorder) • Not employed • Long term sick leave (due to Bleeding Disorder) • Long term sick leave • Student • Retired (due to Bleeding Disorder) • Retired • Other
How many days during the past 6 months was the patient not able to perform daily activities (e.g., school, work, other), since the last clinic visit?	[open field]
Marital status	• Married/Living together • Separated/Divorced • Widow • Single • Prefer not to answer

Functional Scales

For both VWD & Hemophilia patients

Hemophilia Joint Health Score (HJHS)

Data in the table below are collected for all 6 joints: Elbow, ankle, and Knee.

Joint assessment

Date	DD-MM-YYYY
Time	HH:MM
Name of physiotherapist	[open field]
Swelling	0: none 1: Mild 2: Moderate 3: Severe Non-evaluable
Swelling duration	0: no swelling or <6 months 1: ≥ 6 months Non-evaluable
Muscle Atrophy	0: none 1: Mild 2: Moderate 3: Severe Non-evaluable
Crepitus on motion	0: none 1: Mild 2: Moderate 3: Severe Non-evaluable
Flexion loss	0: <5° 1: 5°-10° 2: 11°-20° 3:>20° Non-evaluable
Extension loss	0: <5° 1: 5°-10° 2: 11°-20° 3:>20° Non-evaluable
Joint pain	0: No pain through active range of motion 1: No pain through active range; only pain on gentle overpressure or palpation 2: Pain through active range Non-evaluable

Strength	0: Holds test position against gravity with maximum resistance (gr.5) 1: Holds test position against gravity with moderate resistance (but breaks with maximal resistance) (g.4) 2: Holds test position with minimal resistance (gr 3+) or holds test position against gravity (gr.3) 3: Able to partially complete ROM against gravity (gr.3/2+) or able to move through ROM gravity eliminated (gr 2-) 4: Trace (gr.1) or no muscle contraction (gr.0) Non-evaluable
Comments	[open field]

Global gait

Global gait (walking, stairs, running, hopping on 1 leg)	0: All skills are within normal limits 1: One skill is not within normal limits 2: Two skills are not within normal limits 3: Three skills are not within normal limits 4: No skills are within normal limits Non-evaluable

Joint Disease

Date	DD-MM-YYYY
How often since the last annual visit has the patient used a cane, crutches or walker for ambulation or mobility?	Never Intermittently Always
How often since the last annual visit has the patient used a wheelchair for mobility?	Never Intermittently Always
How many days since the last annual visit has the patient missed work or school?	[open field]
Has the patient experienced a joint infection since the last annual visit?	Yes No If Yes, specify joint [open field]
How far can the patient walk unaided?	Less than 100 metres; 100-500 metres; 500 metres - 1Km; >1 Km
Check the statement which best describes the patient's current overall activity level	• Unrestricted school/work and recreational activities • Full school/work with limited recreational activity levels due to pain, loss of motion or weakness • Limited school/work and recreational activity levels due to pain, loss of motion or weakness • Limited school/work and recreational activity levels, and self-care activity levels due to pain, loss of motion or weakness • Requires assistance from another person for school/work, self-care, and unable to participate in recreation due to pain, loss of motion or weakness
Comments	[open field]

Range of motion (ROM)

Date	DD-MM-YYYY
ROM measured	Yes No *If no, please specify*: Acute bleed Post-op restrictions Other medical reason
Record ROM Endpoint **Left, right**	Joint & Normal Range Flexion 135° Extension 0° Hyperextension Pronation Supination 80°
Left ROM value	[open field]
Right ROM value	[open field]
Orthopedic brace	Yes No If yes, please specify for Left, right.
Ancillary therapy **Left knee, right knee**	Orthodesis Joint replacement Orthoscopic synovectomy Open synovectomy Radioisotopic synovectomy Other invasive procedure
Target joint	Yes No If yes, please specify Left, Right, left & right

WFH Score (Gilbert) – Appendix I

Functional Independence Score in Hemophilia (FISH) – Appendix II

COVID-19

For both VWD & Hemophilia patients

Was this patient tested for COVID-19?	Yes No Not tested but presumed to have been infected based on symptoms
Did the patient have Antibody test?	Positive Negative Not done
Was the patient admitted to hospital due to COVID-19?	Yes No
Was the patient ventilated?	Yes No
Patient status	Alive Deceased
Were there any bleeding complications related to COVID-19?	Yes No
Was the patient on any of the following treatments at time of diagnosis/symptoms?	Hemlibra (Emicizumab) Fitusiran Marstacimab Conzisumab Gene Therapy Other
Hemophilia treatment during infection	Unchanged Initiation of prophylaxis Intensified prophylaxis No access to treatment products due to COVID19 Other

Mortality

For both VWD & Hemophilia patients

Date of death	DD-MM-YYYY
Primary cause of death	Intracranial hemorrhage Bleeding (excluding intracranial) Thromboembolic event Pregnancy-related event HIV Liver disease Cancer Infection including pneumonia Cardiac COVID-19 Unknown Other
***If other,* please specify**	[open field]
Source of information	Death certificate Physician Family member Other

Appendix I – WFH Score (Gilbert)

THE CLASSIFICATION RECOMMENDED BY THE ORTHOPEDIC ADVISORY COMMITTEE OF THE WORLD FEDERATION OF HEMOPHILIA

The clinical evaluation includes patient data, joint evaluation and physical evaluation as detailed below:

I. Patient Data

1. Age
2. Factor deficiency (VIII, IX, etc.)
3. Factor level
4. Inhibitor (Yes or No)
5. Mode of treatment

O = No, or minimal transfusion therapy
E = Episodic transfusion for most of all bleeding episodes M = Maintenance or prophylactic therapy
(H) = Added after E or M indicates that the patient is on a homeself-transfusion program

Example: 16: VIII: <1:NO:E(H)

A 16-year-old patient, factor VIII deficient, with a level of less than 1%. He does not have an inhibitor and treats at home on an episodic basis.

II. ***Joint Evaluation*** **(of the nonbleeding joint)**

1. Pain	0-3
2. Bleeding	0-3
3. Physical examination	0-12
4. Radiologic evaluation	0-13

If the limb described requires an aid to ambulation, the following letters should be added at the end of the evaluation:

B = Brace or orthosis
C = Cane
CR = Crutches
WC = Wheelchair

PAIN

0: No pain
No functional deficit
No analgesic use (except with acute hemarthrosis)

1: Mild pain
Does not interfere with occupation nor with activities of daily living (ADL)
May require occasional non-narcotic analgesic

2: Moderate pain
Partial or occasional interference with occupation or ADL Use of non-narcotic medications
May require occasional narcotics

3: Severe pain
Interferes with occupation or ADL
Requires frequent use of non-narcotic and narcotic medications

BLEEDING

This is measured by the number of minor and major hemarthroses *per year*.
0 = None
1 = No major, 1-3 minor
2 = 1-2 major or 4-6 minor
3 = 3 or more major or 7 or more minor

Guidelines

Minor	*Major*
Mild pain	Pain
Minimal swelling	Effusion
Minimal restrictions of motion	Limitation of motion
Resolves within 24hrs of treatment	Failure to respond within 24hrs

PHYSICAL EXAMINATION

This is based on an additive score of 0-12 with 0 being a normal join and 12 being most affected. An (S) is added after the number if a chronic synovitis is clinically diagnosed

Swelling	0 or 2+(S)
Muscle atrophy	0-1
Axial deformity	0-2
Crepitus on motion	0-1
Range of motion	0-2
Flexion contracture	0 or 2
Instability	0-2

Guidelines

Swelling:

0 = None
2 = Present
(S) = Added after score if chronic synovitis is present

Muscle atrophy:

0 = None or minimal (<1cm)
1 = Present

Axial deformity (measured only at knee or ankle):

Knee

0 = Normal = 0-7° valgus
1 = 8-15° valgus or 0-5° varus
2 = >15° valgus or >5° varus

Ankle

0 = No deformity
1 = Up to 10° valgus or up to 5° varus
2 = >10° valgus or >5° varus

Crepitus on motion:

0 = None
1 = Present

Range of motion:

0 = Loss of 10% of total full range of motion (FROM)
1 = Loss of 10-33 1/3% of total FROM
2 = Loss of >33 1/3% of total FROM

Flexion contracture:

Measured only at hip, knee, or ankle
0 = <15° FFC (fixed flexion contracture)
2 = 15° or greater FFC at hip or knee or equines at ankle

Instability:

0 = None
1 = Noted on examination but neither interferes with function nor requires bracing
2 = Instability that creates a functional deficit or requires bracing

Appendix II – Functional Independence Score in Hemophilia (FISH)

FUNCTIONAL INDEPENDENCE SCORE IN HEMOPHILIA (FISH)

Performance based instrument

Patient Name:	**Patient Code:**
	Today (dd/mm/yyyy):____/__ /____.
A.Self Care	
1. Eating and grooming	◯1 ◯2 ◯3 ◯4
2. Bathing	◯1 ◯2 ◯3 ◯4
3. Dressing	◯1 ◯2 ◯3 ◯4
B. Transfers	
4. Chair	◯1 ◯2 ◯3 ◯4
5. Squatting	◯1 ◯2 ◯3 ◯4
C. Locomotion	
6. Walking	◯1 ◯2 ◯3 ◯4
7. Stairs (12 - 14 steps)	◯1 ◯2 ◯3 ◯4
8. Running	◯1 ◯2 ◯3 ◯4
Total Score	

Scores range from 1 - 4 depending on the degree of independence (see scoring key)

Comments