



# World Bleeding Disorders Registry

## Hemophilia and VWD Data Set

February 2023

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## BASELINE DATA - VWD

*Unique form for VWD patients*

### Patient Consent

<b>Has the patient consented to participate in the WBDR?</b>	Yes
	No

### Demographics

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

## Diagnostic information

<b>Reason for diagnosis</b>	[dropdown menu]-one answer only Family History Symptoms of VWD (bleeding) Unknown
<b>VWD Type</b>	[dropdown menu] -one answer only -Type 1 VWD -Type 2 VWD -Type 3 VWD -Platelet-type VWD -Type Unknown  <i>If Type 1 VWD, please specify-one answer only</i> [dropdown menu] Type 1 (30-50 IU/dL + bleeding; or low VWF) Type 1<30 IU/dL Type 1 C Unclassified  <i>If Type 2 VWD, please specify-one answer only</i> 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

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

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	<i>If Other, please specify</i> [open text]
<b>BAT score</b>	[open field]

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### 2. FVIII coagulant activity assay

<b>FVIII coagulant activity assay</b>	[dropdown menu] -one answer only Done Note done, testing not available Not done, other reason Unknown
<b>Date of FVIII coagulant activity assay</b>	DD-MM-YYYY
<b>FVIII coagulant activity level (FVIII:C) (%)</b>	[open field] (integer, 0-100) <i>If level is unknown:</i> [checkbox] 'unknown'

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### 3. VWF antigen test (VWF:Ag)

<b>VWF antigen test (VWF:Ag)</b>	[dropdown menu]-one answer only Done Note done, testing not available Not done, other reason Unknown
<b>Date of VWF antigen test</b>	DD-MM-YYYY
<b>VWF antigen (VWF:Ag) level (%)</b>	[open field] (integer, 0-100) <i>If level is unknown:</i> [checkbox] 'unknown'

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### 4. VWF platelet-dependent activity

<b>VWF platelet-dependent activity</b>	[dropdown menu]-one answer only Done Note done, testing not available Not done, other reason Unknown
<b>VWF platelet-dependent activity test used</b>	[Dropdown menu] – one answer only VWF:RCo VWF:GPIbR VWF:GPIbM VWF:Ab, Unknown
<b>Date of VWF platelet-dependent activity test</b>	DD-MM-YYYY

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<b>VWF platelet-dependent activity level (%)</b>	[open field] (integer, 0-100) <i>If level is unknown:</i> [checkbox] 'unknown'
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### 5. VWF collagen binding (VWF:CB) activity

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<b>VWF collagen binding (VWF:CB) activity</b>	[dropdown menu]- one answer only Done Note done, testing not available Not done, other reason Unknown
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<b>Date of VWF collagen binding activity test</b>	DD-MM-YYYY
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<b>VWF collagen binding (VWF:CB) activity level (%)</b>	[open field] (integer, 0-100) <i>If level is unknown:</i> [checkbox] 'unknown'
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### 6. VWF multimer analysis

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<b>VWF multimer analysis</b>	[dropdown menu]-one answer only Done Note done, testing not available Not done, other reason Unknown
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<b>Date of multimer analysis</b>	DD-MM-YYYY
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<b>VWF multimer analysis result</b>	[dropdown menu] – one answer only -Normal -Loss of high molecular weight multimers (HMW) -Loss of high and intermediate molecular weight multimers -Absent -Other -Unknown <i>If other, please specify</i> [open text]
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### 7. Ristocetin-induced platelet aggregation (RIPA)

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<b>Ristocetin-induced platelet aggregation (RIPA)</b>	[dropdown menu] – one answer only Done Note done, testing not available Not done, other reason Unknown
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<b>Date of RIPA test</b>	DD-MM-YYYY
<b>Is there an enhanced response to low dose ristocetin?</b>	[dropdown menu]-one answer only -Yes -No -Unknown

### 8. VWF:FVIII B binding assay

<b>VWF:FVIII B binding assay</b>	[dropdown menu]-one answer only Done Note done, testing not available Not done, other reason Unknown
<b>Date of VWF:FVIII B binding assay</b>	DD-MM-YYYY
<b>VWF:FVIII B binding assay result</b>	[dropdown menu]-one answer only -Normal -Decreased binding -Unknown

## Clinical History

<b>Bleeding history</b>	[dropdown menu] Has had $\geq 1$ bleed in lifetime Has never had a bleed Unknown <i>If yes, please enter patient's age at first bleed (months)</i> [open text, integer] <i>If age at first bleed is unknown: [checkbox] 'unknown'</i>
<b>VWF Inhibitor History</b>	[dropdown menu] Inhibitor in the past Currently has an inhibitor Never had an inhibitor Unknown
<b>Treatment History</b>	[dropdown menu] -Has received treatment in the past -Never received treatment -Unknown  If answer is "has received treatment in the past" (optional field):



	<input type="checkbox"/> – multiple answers possible -Factor/Cryoprecipitate/Plasma -Desmopressin -Hormonal treatment -Anti-fibrinolytic -Unknown
<b>Prophylaxis History</b>	-Has received prophylaxis in the past -Never received prophylaxis -Unknown  <i>If has received prophylaxis in the past:</i> Please enter patient's age at start of first prophylaxis (months) <input type="text"/> [open text, integer] <i>If age at start of first prophylaxis is unknown:</i> <input type="checkbox"/> 'unknown'

## Genetic testing

<b>Date of genetic testing</b>	DD-MM-YYYY
<b>DNA sequencing technique</b>	<input type="text"/> [dropdown menu] Sanger sequencing of a specific domain NGS analysis MLPA analysis Whole genome sequencing Other Unknown
<b>If Other, Please specify DNA sequencing technique</b>	<input type="text"/> [open field]
<b>DNA Variant type</b>	<input type="text"/> [dropdown menu]  Inversion Large structural variant ( $\geq 50$ bp) Nonsense Frameshift Small insertion or deletion (indel) (<50 bp) Splice Missense Synonymous Promoter UTR Other Unknown
<b>If Other Please specify DNA variant type</b>	<input type="text"/> [open field]

## Blood Group

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**Blood group**

A  
B  
O  
AB  
Unknown

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**Rh Factor**

Positive  
Negative  
Unknown

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# BASELINE DATA – HEMOPHILIA

*Unique for Hemophilia Patients*

## Patient Consent

<b>Has the patient consented to participate in the WBDR?</b>	Yes
	No

## Demographics

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

## Diagnostic information

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

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

## Clinical history

<b>Bleeding history</b>	<p>[dropdown menu]</p> <p>Has had <math>\geq 1</math> bleed in lifetime</p> <p>Has never had a bleed</p> <p>Unknown</p> <p><i>If yes, please enter patient's age at first bleed (months)</i></p> <p>[open text, integer]</p> <p><i>If age at first bleed is unknown: [checkbox] 'unknown'</i></p>
<b>Joint bleeding history</b>	<p>[dropdown menu]</p> <p>Has had <math>\geq 1</math> joint bleed in lifetime</p> <p>Has never had a joint bleed</p> <p>Unknown</p> <p><i>If yes, please enter patient's age at first joint bleed (months)</i></p> <p>[open text, integer]</p> <p><i>If age at first joint bleed is unknown: [checkbox] 'unknown'</i></p>
<b>Treatment History</b>	<p>[dropdown menu]</p> <p>Has received treatment in the past</p> <p>Never received treatment</p> <p>Unknown</p>
<b>Prophylaxis History</b>	<p>Has received prophylaxis in the past</p> <p>Never received prophylaxis</p> <p>Unknown</p> <p><i>If has received prophylaxis in the past:</i></p> <p>Please enter patient's age at start of first prophylaxis (months)</p> <p>[open text, integer]</p> <p><i>If age at start of first prophylaxis is unknown: [checkbox] 'unknown'</i></p>
<b>Inhibitor History</b>	<p>[dropdown menu]</p> <p>Inhibitor in the past</p> <p>Currently has an inhibitor</p> <p>Never had an inhibitor</p> <p>Unknown</p>
<b>Has any family member ever had an inhibitor?</b>	

## Genetic testing

<b>Date of genetic testing</b>	DD-MM-YYYY
<b>DNA variant</b>	[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
<b>DNA Variant type</b>	[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

## Blood Group

<b>Blood group</b>	A B O AB Unknown
<b>Rh Factor</b>	Positive Negative Unknown

# BASELINE VISIT & FOLLOW-UP VISIT

*For both VWD & Hemophilia patients*

## Bleeding events assessment

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

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

## Target joints

<b>Target joints</b>	Yes No Unknown
<i>If yes, indicate the number of target joints</i>	[open field]

## Inhibitor assessment

<b>Inhibitor assessment in the past 6 (BV) months or since last visit (FUV)</b>	[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
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## Admission to hospital

<b>Admission to hospital in the past 6 months (BV) or since last visit (FUV)</b>	Yes No Unknown
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## Treatment

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

### Adverse event

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

### Pregnancy

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

<b>Start date</b>	DD-MM-YYYY
<b>Number of days hospitalized</b>	[open field]
<b>Reason for hospitalization</b>	<ol style="list-style-type: none"> <li>1. Not related to bleeding disorder</li> <li>2. Related to bleeding disorder</li> </ol> <p><i>If related to bleeding disorder, multiple choice (checkbox):</i></p> <ul style="list-style-type: none"> <li>Intracranial hemorrhage</li> <li>Intra-abdominal bleeding</li> <li>Gastrointestinal bleeding</li> <li>Neck hematomas</li> <li>Psoas muscle bleeding</li> <li>Other muscle bleeding</li> <li>Joint Bleeding</li> <li>Soft tissue bleeding</li> </ul>



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Other bleeding  
 Thromboembolic event  
 Acute heavy menstrual bleed  
 Hemorrhagic ovarian cyst  
 Primary post-partum hemorrhage  
 Secondary post-partum hemorrhage  
 Epistaxis  
 Surgery  
 Other

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If Other or Surgery, please specify [open text]

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### Treatment module

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

	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
<b>End date</b>	DD-MM-YYYY
<b>Reason for stopping</b>	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
<i>If other</i> Please specify adverse event	[open field]
Interventions:	[open field]

### Pregnancy module

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

### Height & Weight

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

## Comorbidities

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

<b>If Cancer, please specify</b>	Lung Breast Colorectum Prostate Stomach Liver Other
<b>If Cancer = Other Please specify</b>	[open field]

### Employment, education, marital status

<b>Employment status</b>	[dropdown menu] <ul style="list-style-type: none"> <li>• Employed full-time</li> <li>• Employed part-time (due to Bleeding Disorder)</li> <li>• Employed part-time</li> <li>• Not employed (due to Bleeding Disorder)</li> <li>• Not employed</li> <li>• Long term sick leave (due to Bleeding Disorder)</li> <li>• Long term sick leave</li> <li>• Student</li> <li>• Retired (due to Bleeding Disorder)</li> <li>• Retired</li> <li>• Other</li> </ul>
<b>How many days during the past 6 months was the patient not able to perform daily activities (e.g., school, work, other)</b>	[open field]
<b>Marital status</b>	<ul style="list-style-type: none"> <li>• Married/Living together</li> <li>• Separated/Divorced</li> <li>• Widow</li> <li>• Single</li> <li>• Prefer not to answer</li> </ul>

# 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

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

<b>Strength</b>	<p>0: Holds test position against gravity with maximum resistance (gr.5)</p> <p>1: Holds test position against gravity with moderate resistance (but breaks with maximal resistance) (g.4)</p> <p>2: Holds test position with minimal resistance (gr 3+) or holds test position against gravity (gr.3)</p> <p>3: Able to partially complete ROM against gravity (gr.3/2+) or able to move through ROM gravity eliminated (gr 2-)</p> <p>4: Trace (gr.1) or no muscle contraction (gr.0)</p> <p>Non-evaluable</p>
<b>Comments</b>	[open field]
<b>Global gait</b>	
<b>Global gait (walking, stairs, running, hopping on 1 leg)</b>	<p>0: All skills are within normal limits</p> <p>1: One skill is not within normal limits</p> <p>2: Two skills are not within normal limits</p> <p>3: Three skills are not within normal limits</p> <p>4: No skills are within normal limits</p> <p>Non-evaluable</p>

## Joint Disease

<b>Date</b>	DD-MM-YYYY
<b>How often since the last annual visit has the patient used a cane, crutches or walker for ambulation or mobility?</b>	Never Intermittently Always
<b>How often since the last annual visit has the patient used a wheelchair for mobility?</b>	Never Intermittently Always
<b>How many days since the last annual visit has the patient missed work or school?</b>	[open field]
<b>Has the patient experienced a joint infection since the last annual visit?</b>	Yes No If Yes, specify joint [open field]
<b>How far can the patient walk unaided?</b>	Less than 100 metres; 100-500 metres; 500 metres - 1Km; >1 Km
<b>Check the statement which best describes the patient's current overall activity level</b>	<ul style="list-style-type: none"> <li>• Unrestricted school/work and recreational activities</li> <li>• Full school/work with limited recreational activity levels due to pain, loss of motion or weakness</li> <li>• Limited school/work and recreational activity levels due to pain, loss of motion or weakness</li> <li>• Limited school/work and recreational activity levels, and self-care activity levels due to pain, loss of motion or weakness</li> <li>• Requires assistance from another person for school/work, self-care, and unable to participate in recreation due to pain, loss of motion or weakness</li> </ul>
<b>Comments</b>	[open field]



## Range of motion (ROM)

Date	DD-MM-YYYY
ROM measured	Yes
	No
	<i>If no, please specify:</i>
	Acute bleed Post-op restrictions Other medical reason
Record ROM Endpoint	Joint & Normal Range
	Flexion 135° Extension 0° Hyperextension Pronation Supination 80°
Left, right	
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*

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

# Mortality

*For both VWD & Hemophilia patients*

<b>Date of death</b>	DD-MM-YYYY
<b>Primary cause of death</b>	Intracranial hemorrhage Bleeding (excluding intracranial) Thromboembolic event Pregnancy-related event HIV Liver disease Cancer Infection including pneumonia Cardiac COVID-19 Unknown Other
<b><i>If other, please specify</i></b>	[open field]
<b>Source of information</b>	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<br>No functional deficit<br>No analgesic use (except with acute hemarthrosis)  |
| 1: | Mild pain<br>Does not interfere with occupation nor with activities of daily living (ADL)<br>May require occasional non-narcotic analgesic     |
| 2: | Moderate pain<br>Partial or occasional interference with occupation or ADL Use of non-narcotic medications<br>May require occasional narcotics |
| 3: | Severe pain<br>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*

Mild pain

Minimal swelling

Minimal restrictions of motion

Resolves within 24hrs of treatment

#### *Major*

Pain

Effusion

Limitation of motion

Failure to respond within 24hrs

## PHYSICAL EXAMINATION

This is based on an additive score of 0-12 with 0 being a normal joint 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

<b>Patient Name:</b>	<b>Patient Code:</b>
	Today (dd/mm/yyyy): ___ / ___ / ____.
<b>A. Self Care</b>	
<b>1. Eating and grooming</b>	<input type="radio"/> 1 <input type="radio"/> 2 <input type="radio"/> 3 <input type="radio"/> 4
<b>2. Bathing</b>	<input type="radio"/> 1 <input type="radio"/> 2 <input type="radio"/> 3 <input type="radio"/> 4
<b>3. Dressing</b>	<input type="radio"/> 1 <input type="radio"/> 2 <input type="radio"/> 3 <input type="radio"/> 4
<b>B. Transfers</b>	
<b>4. Chair</b>	<input type="radio"/> 1 <input type="radio"/> 2 <input type="radio"/> 3 <input type="radio"/> 4
<b>5. Squatting</b>	<input type="radio"/> 1 <input type="radio"/> 2 <input type="radio"/> 3 <input type="radio"/> 4
<b>C. Locomotion</b>	
<b>6. Walking</b>	<input type="radio"/> 1 <input type="radio"/> 2 <input type="radio"/> 3 <input type="radio"/> 4
<b>7. Stairs (12 - 14 steps)</b>	<input type="radio"/> 1 <input type="radio"/> 2 <input type="radio"/> 3 <input type="radio"/> 4
<b>8. Running</b>	<input type="radio"/> 1 <input type="radio"/> 2 <input type="radio"/> 3 <input type="radio"/> 4
<b>Total Score</b>	

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

**Comments**

## Appendix III – Quality of life EQ-5D-5L

*For both VWD & Hemophilia patients*



**Health Questionnaire**

**English version for the USA**



Under each heading, please check the ONE box that best describes your health TODAY.

**MOBILITY**

- I have no problems walking
- I have slight problems walking
- I have moderate problems walking
- I have severe problems walking
- I am unable to walk

**SELF-CARE**

- I have no problems washing or dressing myself
- I have slight problems washing or dressing myself
- I have moderate problems washing or dressing myself
- I have severe problems washing or dressing myself
- I am unable to wash or dress myself

**USUAL ACTIVITIES** (e.g. work, study, housework, family or leisure activities)

- I have no problems doing my usual activities
- I have slight problems doing my usual activities
- I have moderate problems doing my usual activities
- I have severe problems doing my usual activities
- I am unable to do my usual activities

**PAIN / DISCOMFORT**

- I have no pain or discomfort
- I have slight pain or discomfort
- I have moderate pain or discomfort
- I have severe pain or discomfort
- I have extreme pain or discomfort

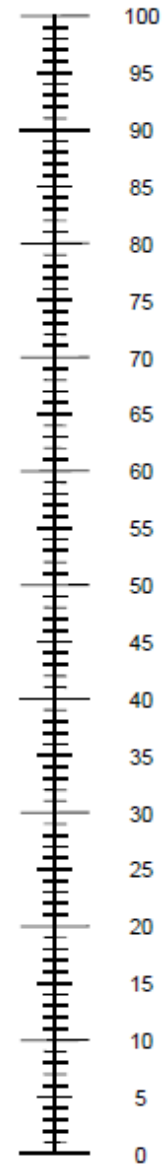
**ANXIETY / DEPRESSION**

- I am not anxious or depressed
- I am slightly anxious or depressed
- I am moderately anxious or depressed
- I am severely anxious or depressed
- I am extremely anxious or depressed

- We would like to know how good or bad your health is TODAY.
- This scale is numbered from 0 to 100.
- 100 means the best health you can imagine.  
0 means the worst health you can imagine.
- Mark an X on the scale to indicate how your health is TODAY.
- Now, please write the number you marked on the scale in the box below.

YOUR HEALTH TODAY =

The best health  
you can imagine



The worst health  
you can imagine