# National Haemophilia Database Dataset

## All Bleeding Disorders

Haemophilia Centre Name

Diagnosis

Title

Surname

Forename

Previous Surname

Previous Forename

Sex

NHS / CHI / HSC number

Date of Birth

Date of Death (if applicable)

Post-mortem performed (if applicable)

Postcode

Ethnicity

Weight (kg) / Date weighed

Bleeding disorder severity

Date first registered with NHD

Date first registered with Centre

Date first seen at the Centre

Normally Resident Overseas / Country

Born Overseas

Reason Seen

Clinical Bleeding History

Family History of the bleeding disorder

Genetic mutation / Date

Current Inhibitor / date detected

Previous inhibitor detected date

Family history of inhibitor

Current status (moved away / lost to follow-up etc.) / Date

General Practitioner Code

Home delivery / Home delivery provider

Surgery

Haemophilia Joint Health Score (HJHS) Score / Date

Treatment regimen (prophylaxis / on-demand / ITI) / start date

Usual prophylaxis dose / frequency

Bypass Prophylaxis / Date started

Patient has requested a copy of their NHD record

***Patient Factor Level(s)***

Factor type / level / date tested

## Additional data for Acquired Bleeding Disorders

Antibody Level

Date Antibodies Detected

Predisposing Condition

Previous Drugs

## Additional data for Von Willebrand Disease

Blood Group

Factor VIII:C result

VWF Activity result

VWF Antigen result

VWF Collagen Binding

FVIII Binding Activity

Platelet Count

VWD Type

Multimeric Analysis

Dates of blood tests

Family History of VWD

Ristocetin Induced Platelet Aggregation

## Quarterly Treatment Data

Every quarter Haemophilia Centres are required to submit information to the National Haemophilia Database about the treatment issued to patients, including products delivered to the patient's home. The following information is collected:

Name of product

Number of units issued

Patient's weight (kg)

Current inhibitor: Yes / No

Patient receives home delivery (Yes / No)

Home delivery provider

# Adverse Events Reports

**Allergic or Other Acute Event**

Event Date

Type of Event: Anaphylaxis / Rigors / Rash / Shortness of breath

Infusion information: Product / Batch number

Additional Blood Products? (Blood / FFP / Cryoprecipitate)

Time between dose and event

Lifetime exposure days

Has this happened previously - Yes / No

Outcome: Resolved / Alive with long-term disability / Death

Do you consider this relationship to the concentrate to be: Definite / Probable / Possible / Unlikely / Unrelated?

**Death Event**

Date of Death

Cause of death

Confirmed by Autopsy? - Yes / No

Inhibitor present at time of death - Yes / No

HIV \Positive - Yes / No

Hepatitis B Positive - Yes / No

Hepatitis C Positive - Yes / No

Evidence of cirrhosis (fibroscan > 13) or liver failure (hepatic decompensation)?

**Infection Event**

Event Date

Infection type: HIV / Hepatitis A, B and C / Parvovirus B19 / vCJD

Infusion information – Product / Batch number

Additional blood products - Yes / No

Last negative test date

First positive test date

Last exposure date

Do you consider this relationship to the concentrate to be: Definite / Probable / Possible / Unlikely / Unrelated?

**Inhibitor**

Reporting Consultant

Genotype

Relatives with inhibitors?

Relatives Details

Reason for test? Routine screen / Surgery / Poor response

Is this the first ever inhibitor? Yes / No

If no, has the inhibitor reappeared after Immune Tolerance Induction (ITI)? Yes / No

Previous inhibitor information (text box)

Date of last negative Inhibitor test *or* No previous inhibitor tests performed

***Inhibitor Potency (Bethesda Units)***

Anti-Human: 1st Positive Level / Date

2nd Positive Level / Date

Peak Inhibitor Titre / Date

Assay used

Laboratory normal range

Anti-Porcine: 1st Positive Level / Date

Peak Inhibitor Titre / Date

Any change in baseline FVIII/FIX? - Detail

Any change in bleeding pattern?

Treatment before inhibitor development: None / Routine on demand / Routine prophylaxis / Intensive

Additional Blood Products? (Blood / FFP / Cryoprecipitate)

***Estimate the following:***

Time interval between first replacement and inhibitor:

Total lifetime exposure days to any FVIII/IX concentrate

*Factor Replacement details*: (approx. dates and amounts of blood products issued over the previous two years): Date From - To / Product name / Number of units

Comment (include batch information where available)

***Concomitant Events***

Date From - To

Comment

## Intracranial Haemorrhage (ICH) (Paediatric)

Event Date

Platelet function abnormality (if applicable):

Platelet count (if low):

Family history of inherited bleeding disorder (known before diagnosis): Yes / No / Unknown

Family history of ICH: Yes / No / Unknown

### *Perinatal history (mandatory for those with ICH < 1yr of age):*

Gestation at delivery (weeks)

Mode of delivery: Vaginal - spontaneous / Forceps / Ventouse/ Lower segment Caesarean section – Elective / Lower segment Caesarean section – Emergency

Mobility at time of event: Pre-mobile / Early mobile (crawling, cruising) / Mobile

### *Details if ICH Event:*

Site of ICH: Subdural / Intracerebral / Subarachnoid

Previous history of ICH: Yes / No

Inhibitor present at the time of ICH: Yes / No

Preceding trauma: Yes / No, if yes: Minor head bump / Major trauma

On prophylaxis prior to ICH: Yes / No, if yes: Prophylaxis with FVIII/FIX / Prophylaxis with FEIBA/rFVIIa

### *Management:*

Surgical intervention: Yes / No, if yes: Craniotomy & Evacuation / VP Shunt or drain / Both of the above / Unknown

Duration of intensive factor replacement therapy (days)

Post Intensive Phase Treatment: ITI / On Demand / Prophylaxis

### *Short term outcome:*

Status: Alive/Deceased

Neurological sequelae: Yes / No, if yes: Gross motor weakness / Seizures / Cognitive problems / Other – comment

Inhibitor development after ICH: Yes / No

Useful Additional information

**Intracranial Haemorrhage (Adults)**

Event Date

Platelet function abnormality (if applicable):

Platelet count (if low):

Family history of inherited bleeding disorder: Yes / No / Unknown

Family history of ICH: Yes / No / Unknown

***Details if ICH Event:***

Site of ICH: Subdural / Intracerebral / Subarachnoid

Previous history of ICH: Yes / No

Inhibitor present at the time of ICH: Yes / No

Preceding trauma: Yes / No, if yes: Minor head bump / Major trauma

On prophylaxis prior to ICH: Yes / No, if yes: Prophylaxis with FVIII/FIX / Prophylaxis with FEIBA/rFVIIa

Was a vascular abnormality found? Yes / No, if yes: Aneurysm / Arterio-venous malformation

Other Risk Factors: Hypertension / Smoking / Diabetes / Amyloid / None

***Management:***

Surgical intervention: Yes / No, if yes: Craniotomy & Evacuation / VP Shunt or drain / Both of the above / Unknown

Duration of intensive factor replacement therapy (days)

Post Intensive Phase Treatment: ITI / On Demand / Prophylaxis

***Short term outcome:***

Status: Alive/Deceased

Neurological sequelae: Yes / No, if yes: Gross motor weakness / Seizures / Cognitive problems / Other – comment

Inhibitor development after ICH: Yes / No

Useful Additional information

**Malignancy Event**

Event Date

Malignancy diagnosis

Did the patient ever undergo radioactive synovectomy? – Yes / No / Don't know

In the last 10 years did the patient receive: Plasma derived concentrate or FFP or Cryoprecipitate / Recombinant concentrate / Both of the above / None of the above

Is the patient HIV Positive: Yes / No

Hepatitis B Positive: Yes / No

Hepatitis C Positive: Yes / No

**Neurological Event**

Event Date

Event type: Motor Neurone Disease (Amyotrophic Lateral Sclerosis) / Sporadic CJD / Variant CJD / Other Neurological Event

Patient received concentrate / Emicizumab / DDAVP / FFP / Platelets in previous 3 months?

If yes, is the patient on prophylaxis treatment?

Time between last dose and neurological event (minutes / hours / days)

Infusion information – Product name / Batch number

Additional Blood Products? (Blood / FFP / Cryoprecipitate)

Comment

**Poor Efficacy Event**

Event Date

Event information

**Thrombotic Event**

Event Date

Event type: Angina / Deep vein Thrombosis / Myocardial Infarction / Pulmonary Embolism / Thrombotic Microangiopathy / Thrombotic Stroke / Transient Ischemic Attack

Patient received concentrate / Emicizumab / DDAVP / FFP / Platelets in previous 3 months?

If yes, is the patient on prophylaxis treatment?

Time between last dose and neurological event (minutes / hours / days)

Infusion information – Product name / Batch number

Additional Blood Products? (Blood / FFP / Cryoprecipitate)

Was thrombosis associated with a central venous catheter? - Yes / No

If yes, choose: Broviac Line, Hickman Line, Jugular/subclavian central line, Portacath, other

Did the patient have surgery in the last 3 months? - Yes / No

If yes, state surgery type

If yes, was prophylaxis given?

***Risk Factors***: Thrombophilia / Pregnancy / Oral contraceptive pill / Hormone replacement therapy / Diabetes / Smoking – (current, former, never), Hyperlipidaemia, BMI > 30

Any personal past history of MI or Stroke or DVT or PE / Any first-degree relatives with MI or Stroke / Any first-degree relatives with DVT or PE / HIV positive / On HAART / Hypertension / Atrial Fibrillation

**Other Event not specified above**

Event Date

Details of the event

# Immune Tolerance Induction (ITI) Survey

Only collected in minimally-treated (less than 150 exposure days) patients with Haemophilia A who develop an inhibitor

**Baseline inhibitor data**

Date of diagnosis of inhibitor

Inhibitor titre at diagnosis

Exposure days at inhibitor detection

Product at inhibitor development

Family history of inhibitor

**After one month of inhibitor detection**

ITI commenced?

Date commenced

If no, reason for delay

Product for ITI

Regimen at start of ITI: 50/kg, every other day / 100/kg, Daily / 200/kg, Daily / Weaning FVIII dosing / Other

Inhibitor titre at start

Historical peak titre

Weight kg / Date weighed

**Follow-up questions, requested 3, 6, 9 & 12 monthly from ITI start date**

Is there a sustained downward trend in inhibitor titre? Yes / No

Latest Weight (kg) / Date weighed

Current Bethesda Units (BU) / Date of current BU

Peak BU after starting ITI (Only required in 1st 3-month follow-up)

Date of peak BU after starting ITI (Only required in 1st 3-month follow-up)

Current Product

Current Regimen: 50/kg, every other day / 100/kg, Daily / 200/kg, Daily / Weaning FVIII dosing / Other

Adjuvant therapy used: Yes / No

If adjuvant used: Rituximab / Other (please give details)

Portacath or equivalent inserted: Yes / No

Portacath required changing: Yes / No

Portacath related bacteraemia: Yes / No

Outcome: Ongoing ITI with First Line / Ongoing ITI with Second Line / Partial Remission / Total Remission / ITI Failure

**Outcome - Upon total remission or ITI failure**

Negative Bethesda: Yes / No

Date of 1st Neg Bethesda

Date of 2nd Neg Bethesda

If no negative Bethesda, titre at end of 1st ITI

Tolerance achieved: measurable trough on ≤50iu/kg alt daily or half-life >7hrs: Yes / No

Date tolerance achieved

Partial tolerance achieved: Neg Bethesda but >50iu/kg alt daily required: Yes / No

Date of failure

Date of relapse if applicable

Bypassing agent use during ITI?

## Acquired Haemophilia A Survey

**Baseline Acquired Haemophilia A Data**

Factor VIII:C at diagnosis

Bethesda Unit (BU) at diagnosis

Anti-porcine FVIII BU (if known)

Presenting to another hospital and asking your advice: Yes / No

Transferred to your hospital: Yes / No

Comorbidity at presentation: Hypertension requiring medication / Diabetes / IHD / Pregnancy / post-partum / Autoimmune condition / Active malignancy / Malignancy thought to be in remission / None / Other (please give details)

Previously resident in: Nursing home / Sheltered Accommodation / Own home with care package / Independent living in own home / Other (please give details)

Red Cell transfusion requirement upon diagnosis

Bleeding Pattern (choose all which apply): Muscle bleed / GI bleed / Intracranial Bleed / Haemarthosis / Pulmonary / Nasopharyngeal / Subcutaneous / Urogenital / Iatrogenic / Other (give details)

Bypassing agent required at presentation: Yes / No

First line immunosuppression choice (multiple choice): Steroid / Cyclophosphamide / MMF / Rituximab / Other (please give details)

**Follow-up questions, requested 3, 6, 9 & 12 monthly**

Alive: Yes / No

If no,

Cause of death (if known) / Date of death (if known)

Cause of death related to immunosuppression?

Cause of death related to bleeding?

Last known FVIII:C level / Date of assay

Last known BU level / Date recorded

If died or Unknown status - no further data required. If alive, continue below

Current FVIII:C / date recorded

Most recent anti human BU / date recorded

Most recent anti porcine BU (if done) / date recorded

In complete remission (FVIII:C normal, Bethesda neg, off immunosuppression): Yes / No

Continued immune treatment: Yes / No

If no, date of cessation:

Current immune therapy: Steroid / Cyclophosphamide / MMF / Rituximab / Other (please give details)

Current dosing:

New bleeding episodes requiring haemostatic intervention since presentation: Yes / No

If yes, how many bleeding episodes?

New treatment related toxicity: Yes / No

If yes, what type: Infection / Glucose intolerance / Mental health (incl. depression, psychosis) / Other (please give details)

If discharged, where to: To long term nursing home / Short term rehab facility / Sheltered accommodation / Own home with care package / Independent living in own home / Other (please give details)

Required readmission? Yes / No

Estimated total time as hospital inpatient since diagnosis: None / Up to a week / 1 week - 1 month / 1 - 2 months / >2 months

Total bypassing agent use

# Hepatitis C Dataset

Received factor concentrates before 1988 (Yes / No)

Received blood components before 1992 (Yes / No)

HCV antibody (Pos. / Neg.)

HCV PCR (Pos. / Neg.)

Cirrhosis (Yes / No)

Liver Failure or progressed chronic liver disease (Yes / No)

Hepatocellular carcinoma (Yes / No)

On-going screening for hepatocellular carcinoma (Yes / No)

Liver Transplant (Yes / No)

# Haemtrack Dataset

***Patient Information***

Forename *(Mandatory)*

Surname *(Mandatory)*

Date of birth *(Mandatory)*

Email address *(Mandatory)*

Phone number(s) *(Optional)*

Address *(Optional)*

Preferred treatment product

Registered Haemophilia Centre *(Mandatory)*

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***Clinical Information*** *(optional – can be entered by the Haemophilia Centre Staff*

Diagnosis

Severity

Factor Level

Treatment Regimen (Prophylaxis / Immune Tolerance / On Demand)

Patient's home treatments managed by centre? (Yes / No)

Weight (kg) / Date weighed

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***Patient User Account Information***

Username

Password *(encrypted)*

Memorable question *(encrypted)*

The database records the time and date of when all data are submitted

***Treatment Information***

The device used to enter the treatment (App, Web, Entered by Centre staff on behalf of patient from paper records) is recorded.

Treatment Date & Time

Product

Infusion Information – Batch number / number of vials / vial size / units

Treatment Reason: New Bleed / Follow-Up Bleed / Prescribed Treatment / Routine Prophylaxis / Surgery / Prior to or After an Activity or Event / Physiotherapy / Immune Tolerance / Other / Inpatient

***Additional information for New Bleed Treatments***

Time between bleed & treatment (hours)

Cause of bleed (Spontaneous / Surgery or Dental / Trauma or Activity)

Severity of bleed (Not stated / Minor (not so bad) / Major (very bad) / Life or Limb Threatening)

Did this bleed interfere with a planned activity?

Site of bleed (Joint / Mouth / Muscle or Soft Tissue / Other)

Location of bleed (e.g. Rt Shin / Lt Calf / Lt Shoulder / Lt Cheek / Blood in vomit / Nose bleed)

Pain Scale (graded from 1 – 6 or 'not stated')

Images can be uploaded securely *(optional)*

***Clinical Study Name***

Only if consented to a registered clinical study