NATIONAL HAEMOPHILIA DATABASE – DATA SET

The National Haemophilia Database is required to collect the following Data by the DoH. In the past, they have also required us to collect data on HIV but not Hepatitis C, apart from where a case of Hepatitis C has occurred. In the event of this, this will be reported using an Adverse Events form. We have recently been asked to collect detailed data on potential vCJD exposure including batch numbers and volumes used. This data collection is ongoing and since entries will have to include all registered patients, even those never treated with blood products as well as those affected, the whole process involves more than 20,000 patients. Data entry into the database has subsequently commenced and is still in progress.

NB: It is important to note that, with a very few exceptions, we have never held central data on factor VIII/IX product batch numbers used by patients. Neither have we ever held data on Hepatitis C status, apart from where a case of hepatitis C is known historically. Since these two items are the commonest focus of interest from patients requesting extracts from the database, it is important to emphasise this limitation in the historical dataset.

Name

Patient Unique ID	Used to link patient records with other tables
Name	
Soundex	

General patient information

FIELD NAME	**EXPLANATION**
Patient Unique ID	Used to link patient records with other tables
Sex	Male or Female
Basic factor level	Factor level for the diagnosed deficiency
DOB	
Severity	Severity classification of disorder
Year First Registered	Year first registered with the NHD
Inhibitor detected	Yes / No
Year Inhibitor First Detected	Year of first inhibitor detection
Inhibitor form received and date	
HB s AG Carrier	
Resident Overseas	Yes / No
Date of Death	
Cause of Death	
History of Jaundice/Hepatitis	Yes / No
Post-Mortem	Yes / No
NHS number	
Old NHS	
No Oxford Diagnosis	No longer used
Oxford Reg No	No longer used
Home Post Code	
GP code	
Date of Death - Not known at Centre	Used if information is obtained from source other than a Haemophilia Centre
Cause of Death - Not known at Centre	Used if information is obtained from source other than a Haemophilia Centre

Quarterly Treatment Record

FIELD NAME	EXPLANATION
Patient Unique ID	Used to link patient records with other tables
Treatment Year and quarter	
Centre	Centre from which treatment was issued
Regular Home Treatment	Yes / No
Regular Prophylaxis	Yes / No
No Inhibitor This Year	Yes / No
Inhibitor Present This Year	Yes / No
No Dose This Year	Yes / No
Visitor	Yes / No
Other Treatment Used	Yes/ No If Yes, please specify
Material used	
rFVIIa dose given	Used dose in µg/kg: ≤90, 90-180, 180-270, ≥270
Number of doses Time period between doses	
Material units used	
Surgery in treatment quarter	Yes / No
Home Delivery	Yes / No
Home Delivery Provider	
Weight (kg)	
Ethnicity	
Inhibitor Family History	
Malignancy Diagnosed	
Auto Immune Diagnosed	
Interferon Received	

Record of centres at which a patient is registered (patients can be registered at more than one Haemophilia Centre)

FIELD NAME	EXPLANATION
Patient Unique ID	Used to link patient records with other tables
Centre	Name of Haemophilia Centre
Gene Mutation Known	Yes / No
Gene Mutation Date	Date mutation detected
No Longer Seen	Yes / No – used when patients transfer
No Longer Seen - Date	Date
Date Registered	Date registered at individual Haemophilia Centres
Date first seen	Date first seen at individual Haemophilia Centres

Adverse Events

Adverse events are now collected monthly, although historically they were collected on a quarterly basis. The following events are collected with the date of event and details of the event (free text).

- HIV Transmission
- Non-A, non-B or Hepatitis C Transmission
- Hepatitis B Transmission
- New Inhibitor
- Thrombotic Arterial/venous Event/DIC: Date/Location/diagnosis/Symptoms/Outcome For thromboembolic events: Completion of targeted NN follow-up request forms
- Transfusion Reaction
- Other Event- specified (Other adverse drug reaction related to the use of rFVIIa)

Inhibitor Events, however, require a form to be completed which contains the following information:

Centre
Email
NHS No
NHD No
Surname
Forename
Date of Birth
Hospital No
Reporting Consultant
Severity
Genotype
Relatives with inhibitors
First inhibitor detection
Reason for test
Date of first detection of Anti-Human Inhibitor
Inhibitor potency (BU)
Anti-Human at first detection
Date of detection of maximum Anti-Human Inhibition
Inhibitor potency (BU)
Anti-Human at maximum
Date of first detection of Anti-Porcine Inhibitor
Inhibitor potency (BU)
Anti-Porcine at first detection
Date of detection of maximum Anti-Porcine Inhibitor
Inhibitor potency (BU)
Anti-Porcine at maximum
Any Change in Baseline
Any Change in bleeding pattern?
Details

Treatment before inhibitor development
Treatment interval between first replacement and inhibitor
Treatment days before inhibitor development

Factor Treatment History Records
From Date
To Date
Product
Units
Comment

Concomitant Events
From Date
To Date
Product

This form is currently being reviewed by the Inhibitor Working party to assess the way in which Inhibitor Surveillance is undertaken through the NHD. They suggest a revision of the current data set that is collected. This is due to regulatory agencies in Europe and USA requesting that national haemophilia databases survey for inhibitor risk specifically in relation to changes of factor VIII concentrate in PTPs with severe haemophilia A. In addition to this, they request standardisation of data collection between countries so that aggregate data from national databases can be pooled.

The current value of the data collected by the NHD is limited because equivalent data are not collected on patients who do not develop inhibitors. It is therefore not possible to establish whether any potential risk factors for inhibitor development are present in the UK cohort. In the proposed revised inhibitor surveillance all PTPs with severe haemophilia A will be followed for possible risk factors for inhibitors for a defined period of time or until inhibitor development. This will establish the rate of inhibitor formation in patients who have changed concentrate compared to those that do not change. Analysis will be done with respect to potential risk factors for inhibitor development

There will be two new Inhibitor Surveillance forms. Currently there is a Neonatal Management form used to collect data on patients aged <3 years at registration. These patients will be tracked for 50 exposure days & information will be requested on a 3 monthly basis until the 50 exposure days have been completed. This form contains the following information:

General Information

NHD no
Ethnicity
Factor Levels
Type
Levels
Units
Family History of inherited bleeding disorders
Screened at Birth: Yes/No
If no, age at diagnosis: Months

Pregnancy and Delivery

Born at obstetric unit affiliated to Haemophilia centre: Yes/No
Gestation at delivery: Weeks
Mode of delivery
Duration of labour (if known): Hours

Pre-Natal History

Cranial USS performed?: Yes/No
If yes, what age performed?: Months
Indication: Routine
Bleeding (definite or suspected)
If bleeding, provide details:
USS Result: Normal/Abnormal If abnormal, provide details:
Vitamin K given?: Yes/No
Route: IM/IV/PO
Breast Fed?: Yes/No
Prophylaxis with factor concentrate following delivery?: Yes/No
Indication: Preterm/Routine/Trauma
Comments:
Bleeding during 1st month of life?: Yes/No
Severity: Minor (treatment required)/Moderate-Severe (treatment)

All patients with severe haemophilia over the age of 10 who have up to 50 exposures will have the following data requested on a 3 monthly basis:

Date

Period start: dd/mm/yyyy
Period end: dd/mm/yyyy
Has the patient received any vaccinations in the last period? Yes/No
If yes which ones?

Bleeds

Total number of bleeds in the period for the following: Haemarthrosis Other soft tissue Mucosal / minor bleeds Intracranial Other life-threatening bleed (specify)

Exposure days

Please ONLY fill in which exposure numbers 1-50 have been given since last form

Indication key: **M**inor bleed eg mucosal, **L**ife threatening bleed including intracranial, **S**urgery/Procedure, **P**rophylaxis, **C**ontinuous **I**nfusion Exposure Day No: Date: Factor name: Dose IU: Recent weight (Kg): Indication (see key):

If surgery was it by C/I: Yes/No
Was extravasation possible? Yes/No
Intercurrent Infection: Yes/No

Proposed quarterly returns data collection extension

Date of change of concentrate
Surgical procedures requiring inpatient stay for >2 days
Type of operation
Total FVIII usage in first 2 weeks,
Use of continuous infusion (yes/no)
Known autoimmune disease
Interferon use (yes/no)

The following fields are not related to individual patient records

Quarterly Returns - Material used per diagnosis

Centre
Year
Diagnosis
Date Form Completed
Material
Units