European guidelines for the certification of Haemophilia Centres

07 June 2013

NOTICE

These Standards are designed to provide minimum guidelines for European Haemophilia Centres. These Standards are not intended to establish best practices or include all procedures and practices that Centres or individuals should implement if the standard of practice in the community or governmental laws or regulations establish additional requirements. Each Centre should analyze its practices and procedures to determine whether additional standards apply.

Contents

Introduction

This document has been produced by a group commissioned by the European Haemophilia Network (EUHANET) to set standards and criteria for the designation of Haemophilia Centres within Europe, in order to promote standardization of haemophilia and other inherited bleeding disorders care and then to ensure equity of treatment throughout our diverse continent. The document has been drawn up in consultation with both relevant professional organisations such as EAHAD (European Association for Haemophilia and Allied Disorders) as well as the European Haemophilia Consortium (EHC) and affiliated national patient bodies in Europe. Most of the funding for this project was provided by the European Commission via its Executive Agency for Health and Consumers (EAHC).

Background

Inherited bleeding disorders are rare but complex conditions to manage. The central philosophy of haemophilia care is that treatment should be delivered by a multidisciplinary team in a dedicated haemophilia centre. There are now data which indicate that outcome is improved when patients are managed in this way. Various health care professionals may need to be involved in the care of an individual patient, including not only a haematologist and nurse but also orthopaedic surgeons, physiotherapists, laboratory technicians, hepatologists and infectious disease specialists, dental surgeons, and psychologists. Advances in recent decades have led to great improvements in both longevity and quality of life of patients. At the same time, it is apparent that there are still great disparities in treatment between individual countries within Europe, in terms of the facilities available for treatment, the types of products used, as well as the amount of factor VIII and IX concentrate used.

Target patient group

This document refers to the management of patients with inherited bleeding disorders, covering haemophilia A and B (including female carriers), the rarer congenital deficiencies of other coagulation factors (such as fibrinogen and factors II, V, VII, X, XI and XIII), von Willebrand disease and inherited platelet defects.

The clinical phenotype of haemophilia is related to the baseline level of the relevant coagulation protein, which may be used to classify the severity. Patients with haemophilia may be classified as severe, moderate or mild. Severe haemophilia is defined by a baseline level of factor VIII (or IX) below 1 iu/dl (%). These patients typically experience repeated and spontaneous bleeding into joints such as the knees, elbows and ankles and require regular treatment. Subjects with a factor VIII (or IX) level between 1 and 5 iu/dl are defined as having moderately severe haemophilia and experience less frequent bleeding. Patients with levels above 5 iu/dl are classified as having mild haemophilia and usually experience bleeding only after injury (albeit minor) or trauma.

Methodology

A first draft of this document was prepared by the team members of the EUHANET Work package number 4 (WP4: Development of the standardisation criteria) based on a review and analysis of key reference documents including: three EUCERD recommendations on rare diseases[1-3], the European Principles of Haemophilia Care[4] launched at the European Parliament in January 2009, and information on the existing certification systems in use in European Union Member States.

The present document is the result of two consultation processes that widely involved European stakeholders. The first was launched in Prague during the EHC Conference, October 2012 and aimed to share principles and criteria as preliminary requirements to define the scope, approaches and rules for the production of a common set of EU standards on Haemophilia Centres for European countries. The main objective of the first consultation was to get feedback on the principles and criteria proposed by the WP4 Team for the production of Standards for Haemophilia Centres in EU MS. On the basis of the consensus reached and on the literature review, WP4 Team members started the production of Standards. With the agreement of the EUHANET Steering Committee, these standards were subject to a further round of consultation addressed to EU and non-EU MS stakeholders including clinicians, institutions and patients organizations. This second consultation process was launched at the 6th Annual Congress of the European Association for Haemophilia and Allied Disorders held in Warsaw, February 2013.

Delivery of haemophilia care

All patients with haemophilia and other related bleeding disorders should be registered and treated at designated European Haemophilia Centres. Patients should have access to a comprehensive care programme. This may be provided by a European Haemophilia Comprehensive Care Centre or by a European Haemophilia Treatment Centre which has established a formal relationship with one or more European Haemophilia Comprehensive Care Centres.

European Haemophilia Centres

European Haemophilia Centres are of two types according to the services and facilities which they provide:

- European Haemophilia Treatment Centres (EHTCs) provide local routine care in conjunction with EHCCCs.
- European Haemophilia Comprehensive Care Centres (EHCCCs) provide the highest level of care and function as tertiary referral centres.

The functions and activities carried out by the two types of Centres and the agreements between them are described below.

European Haemophilia Treatment Centre (EHTC)

A EHTC should normally care for at least 10 people with severe haemophilia A or B or VWD type 3.

A European Haemophilia Treatment Centre (EHTC) carries out the following functions and activities:

- Provides care for patients, including diagnosis, treatment, follow-up and rehabilitation.
- Provides patients with safe and effective treatment products.
- Provides a 24 hour emergency treatment service.
- Provides basic diagnostic and monitoring laboratory support during normal working hours for the more common inherited bleeding disorders.
- Has access to multidisciplinary support, locally or in conjunction with EHCCC (physiotherapy and orthopaedics, surgery, dental care, hepatology, infectious diseases, obstetrics and gynaecology, paediatric facilities if children are treated, genetics, clinical psychology and social worker).
- Offers specific treatment for patients with inhibitors and immune tolerance in collaboration with a EHCCC.
- Provides advisory service, including genetic counselling, to patients and healthcare professionals.
- Promotes information and training programs on inherited bleeding disorders to patients and healthcare professionals.

European Haemophilia Comprehensive Care Centre (EHCCC)

A EHCCC should normally care for at least 40 people with severe haemophilia.

A European Comprehensive Care Haemophilia Centre (EHCCC) carries out the following additional functions and activities:

- Co-ordinates the delivery of haemophilia services - both in hospital and in the community including liaison with affiliated EHTCs.
- Provides a 24 hour advisory service for patients, families, hospital doctors, general practitioners and affiliated EHTCs health care professionals.
- Provides specialist care for patients with inhibitors, including surgery.
- Provides a diagnostic and reference laboratory service with a full repertoire of tests for the diagnosis and monitoring of inherited disorders of haemostasis.
- Provides a 24 hour laboratory service for clotting factor assays and inhibitors screens.
- Has access to orthopaedic and/or rheumatological service with provision of surgery.
- Has access to physiotherapy service.
- Has access to a specialised obstetric and gynaecological service for the management of haemophilia carriers and women with von Willebrand Disease and other hereditary bleeding disorders.
- Has access to paediatric facilities if children are treated.
- Has access to a genetic diagnosis service providing also carrier detection and antenatal diagnosis.
- Has access to dental service.
- Has access to hepatology and infectious diseases service for patients with HIV and/or viral hepatitis.
- Offers professional psychological support.
- Has access to social worker and welfare advice.
- Collates data (e.g. product usage, patient demographics).
- Participates in research, including clinical trials.

Standard requirements

1. General requirements

1.1 Facility

1.1.1 There must be dedicated disabled car parking spaces for patients/parents in the vicinity of the EHTC/EHCCC and appropriate disabled access throughout the haemophilia treatment area.

1.1.2 Within the EHTC/EHCCC, clinical treatment of patients takes place in dedicated clinical areas that must be comfortable, quiet and appropriately equipped. These areas must have facilities to allow confidential discussion between staff and patients.

1.1.3 Within the EHTC/EHCCC, there must be a secretariat where records of patients are kept available to the multidisciplinary team and emergency department.

1.2 General policy and objectives

1.2.1 The EHTC/EHCCC Director must formalise a work plan describing the mission, general objectives and policies of the Centre.

1.2.2 Consistent with general objectives and policies, and also in relation to critical issues resulting from systematic quality monitoring activities (see para. 1.9), the EHTC/EHCCC Director must develop specific quality improvement objectives to be assessed on a periodic basis.

1.3 Information about the Centre

1.3.1 The EHTC/EHCCC must draw up a document aimed at patients and which sets out at least the following:

- services offered;
- how to access the Centre;
- information about the centre staff and collaborating consultants and hospitals in the network.

1.4 Organization and staffing

The cornerstone of the treatment of haemophilia and other related bleeding disorders is comprehensive care delivered by a multi-disciplinary and specialised team.

1.4.1 The core team members of an EHTC/EHCCC consist of the following personnel:

- medical staff, who carry out routine and emergency treatment and follow-up clinical reviews;
- nursing staff, who co-ordinate much of the day to day treatment and supplies of coagulation factor concentrates;
- laboratory staff, who provide a diagnostic and factor replacement monitoring service.

Not all members of the multidisciplinary team will be full-time employees of the EHTC/EHCCC itself: several will be members of other clinical departments and will collaborate with the Centre to provide a part-time clinical commitment.

In the case of an external laboratory, written agreements must be in place with the Centre (see para. 4).

1.4.2 There must be in place an organizational chart of key personnel and functions within the EHTC/EHCCC. The Director of the EHTC/EHCCC is responsible for assigning roles and responsibilities within the Centre.

1.4.3 There must be regular meetings between the multidisciplinary team members.

1.4.4 A data manager must be so designated by the EHTC/EHCCC Director.

1.5 Policies and procedures

1.5.1 The EHTC/EHCCC establishes and maintains policies and procedures addressing aspects of management and activities. These documents must include all elements required by these Standards and shall address at a minimum:

- organization of the Centre;
- patients' evaluation and treatment;
- personnel appraisal and continuing education;
- management and monitoring of facilities and equipment;
- supply and management of therapeutic products, laboratory reagents and medical devices;
- quality planning, evaluation and improvement;
- participation in clinical research.

1.5.2 The EHTC/EHCCC adopts a system for preparation, approval, implementation, review, revision and archival of all policies and procedures.

1.5.3 All policies and procedures are regularly reviewed in order to ensure the availability of appropriate and up-to-date references for personnel of the EHTC/EHCCC.

1.6 Record-keeping and data collection

Accurate recording of clinical information is essential for the effective delivery of haemophilia care. Data handling can be complex and EHTC/EHCCC should have in place the financial and human resources to facilitate the collection of information required.

1.6.1 The EHTC/EHCCC maintains a patient register with a clear indication of those patients who are on regular treatment and those who attend for regular review.

1.6.2 The EHTC/EHCCC ensures proper record keeping of all medical records related to patients.

In particular, the EHTC/EHCCC must prepare and update for each patient a file containing at least:

- general data of the patient;
- findings of each review;
- treatment plans;
- informed consents obtained from patient for their clinical details, if applicable;
- copies of correspondence with patient's general practitioner and, where appropriate, other specialists;
- any other relevant correspondence relating to the patient.

1.6.3 In the EHTC/EHCCC, all medical records must be kept in a confidential manner in accordance with applicable laws and regulations on data protection.

1.6.4 Emergency procedures must be in place in the EHTC/ECCC to ensure the proper performance of its activities in the event of medical data on electronic systems being temporarily unavailable.

1.6.5 Records related to quality management, training of personnel, facility and equipment maintenance or other general EHTC/EHCCC's issues must be retained in accordance with applicable laws and regulations, or defined policies and procedures.

1.6.6 The EHTC/EHCCC ensures the traceability of the personnel responsible for generating all critical records (e.g. medical records).

1.6.7 The EHTC/EHCCC identifies all the medical and management records to be maintained for established periods of time, also including them in specific lists, according to governmental or institutional policy, where applicable.

1.7 Personnel appraisal and continuing education

All staff within EHTC/EHCCC must have adequate knowledge and experience to adequately perform their assigned tasks and must comply with regulations regarding appraisal and continuing professional education which may be in place.

1.7.1 The EHTC/EHCCC identifies and formalises the skills and professional qualifications required for the personnel performing activities critical to quality of patient care and implements plans in order to guarantee their adequate training before they start working.

1.7.2 The EHTC/EHCCC Director must systematically identify the training needs for personnel operating within the Centre and plan training activities in order to guarantee that their skills are constantly updated and developed.

1.7.3 Within the EHTC/EHCCC, records must be retained relating to personnel training and competency.

1.7.4 The EHTC/EHCCC implements systems for the periodic assessment of the staff's skills in order to ensure that all members of staff can adequately perform the tasks assigned to them.

1.8 Supply and management of therapeutic products, reagents and medical devices

1.8.1 Procedures must be in place in the EHTC/EHCCC for the management of therapeutic products, laboratory reagents (if applicable), and medical devices in order to guarantee their correct selection, provision, inspection, storage under the appropriate environmental conditions and use.

1.8.2 The EHTC/EHCCC applies systems to prevent the use of expired medicinal products and, where applicable, laboratory reagents.

1.9 Quality planning, evaluation and improvement

The EHTC/EHCCC management is responsible for continuously monitoring the quality of the Centre's performance by establishing a system designed to identify the need for improvement and consequently implementing adequate organizational and technical changes.

1.9.1 The EHTC/EHCCC implements systems in order to guarantee a systematic monitoring of the quality of its performance for the identification of any critical situation that requires corrective actions, or for the identification of any unfavourable trends requiring preventive actions.

1.9.2 Procedures must be in place in the EHTC/EHCCC for detecting, evaluating, documenting and reporting any adverse events that occur in association with the administration of coagulation factor concentrates, e.g. allergic reactions and inhibitor development.

1.9.3 Procedures must be in place in the EHTC/EHCCC for managing complaints and for undertaking periodic surveys of patients' opinions about their care.

1.9.4 The EHTC/EHCCC conducts systematic clinical and quality audits to assess compliance with policies and procedures established within the Centre. The EHTC may organize audits in collaboration with the EHCCC.

1.9.5 Procedures must be in place in the EHTC/EHCCC for the identification and investigation of the cause of all critical issues and for the implementation of corrective or preventive actions.

1.9.6 The EHTC/EHCCC Director must conduct periodic analysis and global assessment of the EHTC/EHCCC's quality-related data, involving all the staff of the Centre in order to evaluate:

- adequacy of the EHTC/EHCCC's general policies and objectives (see para. 1.2);
- achievement of quality improvement objectives (see para. 1.2);
- outcome indicators (see para. 2.6);
- level of satisfaction of patients and their associations (see Standard 1.9.3);
- degree of application of policies and procedures within the Centre, resulting from clinical and quality audits (see Standard 1.9.4);
- adequacy of staff skills and need for training (see para. 1.7);
- adequacy of facilities and equipment (see para. 1.1).

1.9.7 Records of quality monitoring, evaluation and improvement must be maintained by the EHTC/EHCCC.

1.10 Participation in registries related to inherited and acquired bleeding disorders

Individual countries should be encouraged to create and update patient registries related to haemophilia and other related bleeding disorders.

Standard 1.10.2 applies only when a (regional/national) registry is established.

1.10.1 The EHTC/EHCCC maintains a database of patients under its care.

1.10.2 The EHTC/EHCCC participates in the regular collection and transmission to pertinent authorities of data obtained from registries of haemophilia and other related bleeding disorders. The data may be used to support epidemiological surveillance, pharmacovigilance, health planning and pharmacoeconomic evaluations. Data collection and transmission must comply with regulations concerning processing of sensitive data.

1.11 Participation in clinical research

1.11.1 The EHCCC participating in coagulation factor concentrate trials must record the products and number of units used in the trials.

1.11.2 The management of the records related to the trials must be maintained by the EHCCC in accordance with institutional policies and applicable laws and regulations.

2. Patient care

2.1 Awareness, information and education of patients and their families

Haemophilia and other related bleeding disorders have a significant impact on the patients and their family, leading not only to physical disability but also to problems with schooling, employment and relationships.

Patients and their carers should be encouraged to be active participants in, and assume appropriate responsibility for, their delivery of care. Effective haemophilia care can be optimised by the establishment of a close dialogue between the EHTC/EHCCC and patients.

2.1.1 The EHTC/EHCCC organizes in collaboration with other EHTCs/EHCCCs and patient associations events for the education and training of patients and their families, including home therapy/self-infusion.

2.1.2 Each patient must receive information about:

- the nature of the disease, treatment and possible complications;
- the EHTC/EHCCC and the multidisciplinary treatment team;
- contact details of EHTC/EHCCC;
- rights and obligations of the patient;
- local and national patient support organization.

The family of the patient may be involved.

2.2 Diagnosis of haemophilia and other related bleeding disorders and all forms of acquired haemophilia

2.2.1 The EHTC/EHCCC prescribes the necessary tests in cases of suspected bleeding disorders, according to national and, where relevant, international professional guidelines.

2.2.2 The diagnosis of a coagulation disorder must include disease type, severity, presence or absence of inhibitor and mode of inheritance.

2.2.3 The EHTC/EHCCC must issue a written report on diagnosis within one month from the initial review.

2.2.4 After diagnosis, the patient must be registered with the EHTC/EHCCC and, if applicable, with a regional/national registry.

2.2.5 Each patient must be issued with a medical emergency card, which contains at least basic information about his/her diagnosis as well as contact details of the EHTC/EHCCC.

2.3 Therapy of haemophilia and other related bleeding disorders and all forms of acquired haemophilia

2.3.1 Treatment programme

2.3.1.1 A tailored treatment programme must be drawn up by the EHTC/EHCCC for each patient, detailing the therapeutic product, dosage and regimen, on the basis of patient's individual response and bleeding episodes. Patients' views must be taken into consideration.

2.3.1.2 All treatments offered by the EHTC/EHCCC must be in line with national and, where relevant, international professional guidelines. Patients with haemophilia must be treated with coagulation factor concentrates or desmopressin (DDAVP) and not cryoprecipitate or plasma.

2.3.1.3 The treatment programme and clinical records must comply with all legal requirements which may apply to provision of consent.

2.3.1.4 The care of children with haemophilia and other inherited bleeding disorders is complex and should only be carried out in the EHTC/EHCCC by clinical staff specifically trained in the care of children with inherited bleeding disorders.

2.3.1.5 Transfer from paediatric to adult care is a particularly sensitive time for the teenager with a hereditary bleeding disorder, particularly if the adult Centre is at a different hospital. The EHTC/EHCCC must follow a protocol for transition from paediatric to adult care.

2.3.2 Prophylaxis

2.3.2.1 Prophylactic treatment should be available in the EHTC/EHCCC to patients with severe haemophilia as it has been shown to prevent chronic joint disease onset and progression. Bleeding episodes should be monitored and documented in order to define a tailored treatment programme.

2.3.3 Home treatment plan

Wherever appropriate, the care of patients with haemophilia and other inherited bleeding disorders should be delivered in the home setting which will minimise hospital attendance and absence from school and work, enabling them to live as normal a life as possible.

2.3.3.1 The EHTC/EHCCC draws up and periodically updates a home treatment plan for each patient based upon the patient's individual clinical needs.

2.3.3.2 Patient and his family must be instructed as to the importance of recording all bleeding and treatment episodes. EHTC/EHCCC staff must assess the theoretical knowledge and practical competence of the patient before embarking upon home treatment and subsequently at regular intervals as part of the follow-up process (see para. 2.4).

2.3.3.3 The EHTC/EHCCC provides patients with written instructions and/or instruments for recording data on the infusion of therapeutic products at home or in the ambulatory care setting.

2.3.3.4 The EHTC/EHCCC must have a system in place for the monitoring of factor usage by patients on home treatment.

2.3.4 Treatment of acute bleeds and prevention

2.3.4.1 The EHTC/EHCCC must have 24 hour access to an adequate stock of supplies of all treatment products tailored to the type and number of treated patients in order to ensure continuity of care as well as the appropriate and timely treatment of haemorrhagic episodes.

2.3.5 Emergencies, treatment outside normal working hours

2.3.5.1 The EHTC provides 24-h medical cover by formalized arrangements with other departments and/or designated EHCCC.

2.3.5.2 The EHCCC provides 24-h expert haemophilia medical cover.

2.3.5.3 Protocols covering emergencies and arrangements for patients presenting outside normal working hours must be in place in the EHTC/EHCCC, aimed at both patients and medical staff.

2.3.5.4 Patients must be informed by the EHTC/EHCCC of whom they should contact in the event of an emergency or in case treatment is needed outside normal working hours.

2.3.6 Elective surgery

2.3.6.1 Elective major surgery in non-inhibitor patients and elective surgery in patients with inhibitors must only be carried out in EHTCs/EHCCCs with experience of such cases.

2.3.7 Treatment of patients with inhibitors, including immune tolerance

2.3.7.1 All patients who develop inhibitors must be reviewed in a EHCCC and the level of antibody titre determined.

2.3.7.2 Notifications of new cases must be reported by the EHTC/EHCCC to the regional/national registry, if applicable, and other pharmacovigilance programmes (such as EUHASS).

2.3.7.3 An individualised treatment programme for patients with a high inhibitor titre must be drawn up by a EHCCC. The extent to which these patients are treated at a local EHTC must be determined on an individual basis through discussion between the two Centres.

2.3.7.4 In the individualised treatment programme for patients with a high inhibitor titre, immune tolerance induction therapy should be available as soon as possible after inhibitor development. The option of long term prophylaxis with bypassing agents should be considered in patients who fail to respond to immune tolerance.

2.3.8 Treatment of patients with chronic viral infections

2.3.8.1 In the EHTC/EHCCC, patients with chronic hepatitis and/or HIV infection must be reviewed at least once a year by a specialist physician and be offered appropriate therapy in accordance with accepted guidelines.

2.3.9 Treatment of patients with acquired haemophilia and acquired vWD

Acquired haemophilia and acquired von Willebrand's disease are rare conditions but often associated with serious haemorrhagic manifestations and other underlying disorders. Management of these patients is thus particularly challenging.

2.3.9.1 An individualised treatment programme for patients with these rare acquired disorders must be drawn up by a EHCCC in accordance with accepted guidelines. The extent to which these patients are treated at a local EHTC must be determined on an individual basis through discussion between the two Centres.

2.4 Periodic clinical and multi-disciplinary review

All registered patients must be offered a regular clinical review. The EHTC/EHCCC must have a system in place to organize these clinics.

2.4.1 EHTCs and EHCCCs that enter into shared patient care arrangements must ensure that all registered patients with disorders classified as severe have a multidisciplinary review performed at least once a year.

2.4.2 Patients with frequent bleeding episodes, or complications such as inhibitors, arthropathy or chronic viral infections, as well as young children, may require more frequent review in the EHTC/EHCCC, in accordance with accepted guidelines.

2.4.3 Patients with moderately severe or mild haemophilia must be invited by the EHTC/EHCCC for review at least once every two years.

2.4.4 The EHTC/EHCCC must have a protocol in place for multidisciplinary evaluation during each review designed to monitor complications (e.g. inhibitor, arthropathy, chronic liver disease, HIV infection and any comorbidity related to ageing such as cardiovascular diseases and tumours).

2.4.5 A letter must be drafted after each follow-up which includes the following information:

- current clinical problems;
- treatment regimen, highlighting any changes since last review;
- results of relevant laboratory and other tests such as imaging;
- date of next review.

This letter must be sent to the patient's general practitioner and other doctors involved in the management of the patient. Copies must also be filed in the patient's clinical records and sent to the patient.

2.4.6 The EHTC/EHCCC must keep records of non-attendance.

2.5 Genetic services

Identification of the underlying genetic abnormality causing haemophilia in a family facilitates the identification of carriers in the wider family. This information can also be used to provide antenatal diagnosis in pregnant carriers (although there are both legal and cultural restrictions which may apply in some countries). Carriers of haemophilia may themselves have low levels of factor VIII (or IX) and thus may have clinical problems similar to mild haemophilia.

2.5.1 The mutation (or other underlying genetic abnormality) within a family affected by haemophilia must be identified by the EHTC/EHCCC.

2.5.2 The EHTC/EHCCC must have a formal relationship with a genetic laboratory so that all patients and families have access to genetic testing.

2.5.3 It is recognised that genetic testing requires sophisticated technology which is not universally available throughout countries within the territory of the European Union. The EHTC/EHCCC which cannot provide basic genetic services locally must establish collaborative links with other Centres (which may be located in other countries) to ensure the availability of these important tests to their own patients and their families.

2.5.4 Families and individuals must have access to genetic counselling. Written informed consent needs to be documented by the EHTC/EHCCC before genetic tests are performed. This must include specific consent for storage of samples and to share relevant results with other family members.

2.5.5 In the EHTC/EHCCC, each family must have a genetic file separate from the main notes and each individual must have a section of this file that can be kept confidential.

2.5.6 In the EHTC/EHCCC, potential female carriers of haemophilia must be offered genetic counselling and testing to confirm their status when they are old enough to understand the issues involved and give informed consent.

2.5.7 The level of factor VIII (or IX) must be measured by the EHTC/EHCCC in all known or potential carriers of haemophilia before surgery or other invasive procedures.

2.5.8 Pregnancy in known or potential carriers of haemophilia must be supervised in the EHTC/EHCCC that has specific expertise in the area. At all stages of pregnancy there must be close collaboration between the obstetric and haemophilia staff. A documented care plan for the delivery and aftercare of any infant at risk of having a bleeding disorder must be established. Appropriate haemostatic agents for both mother and infant must be available for immediate use if necessary. In the case of a male baby, cord blood must be tested as soon as possible after birth.

2.6 Outcome indicators

2.6.1 All Centres delivering haemophilia care must collect information concerning the outcome of treatment. In order for this to be possible, patients (or parents in the case of children) must agree to provide the raw data requested.

2.6.2 The EHTC/EHCCC must agree the precise parameters to be collected with the appropriate regional and/or national authorities.

It is recommended that as a minimum the following be recorded:

- Units of coagulation factor concentrate used by each patient per year.
- Number of new bleeding episodes (including breakthrough bleeds in the case of patients on prophylaxis).
- Adverse events possibly related to treatment (inhibitors, viral infections, poor efficacy of treatment, etc.).
- Mortality and causes of death.

Other parameters which may be collected include:

- Joint scores.
- Quality of life measurement.
- Number of days missed from school or work due to bleeds.
- Trough levels in patients on prophylaxis.

3. Advisory service

3.1 The EHCCC provides a continuous emergency medical advisory service.

3.2 The EHTC/EHCCC provides an advisory service to patients and their families, as well as other professionals and caregivers who treat the patients during normal working hours.

4. Network of clinical and specialised services in conjunction with the haemophilia team

4.1 The EHTC must establish a formal relationship with one or more EHCCCs. Many smaller EHTCs play a critical role in providing effective emergency care at a local level for patients with haemophilia and other related bleeding disorders. However patients may need to attend the EHCCC for more comprehensive elements of care (e.g. elective surgery in patients with inhibitors). The level of collaboration will depend upon the degree of expertise available in the EHTC.

4.2 The EHTC/EHCCC must guarantee between them an integrated approach to patient multidisciplinary comprehensive care. The support available must include at least the following specialities:

- Physiotherapy and orthopaedics (patients with severe haemophilia must be reviewed by a physiotherapist and an orthopaedic surgeon periodically depending on individual clinical circumstances).
- Surgery.
- Dental care.
- Paediatrics (children must be followed up by health care professionals specifically trained in the care of children).
- Hepatology, Infectious diseases (patients exposed to HIV and chronic liver disease must be followed up by appropriate specialists).
- Obstetrics and Gynaecology (known or potential carriers of haemophilia who are pregnant must be supervised in Centres that have specific expertise in the area).
- Genetics (all people with haemophilia and related bleeding disorders must have access to specialised genetic services for inheritance counselling and mutational analysis to enable confirmation of diagnosis, determination of carrier status and antenatal fetal testing).
- Psychosocial support, particularly regarding provision of social welfare, occupational therapy and counselling services (patients with haemophilia and related bleeding disorders and their family members often have complex psychological issues).

If the above-mentioned specialist services are not provided by the EHTC, arrangements for their provision must be put in place by the EHCCC.

4.3 The relationship between the EHTC/EHCCC and the structures that provide these specialist services must be regulated by specific formal agreements. Protocols must define how to access services and name the physicians involved in providing continuity of care. There must also be diagnostic and therapeutic protocols, protocols on the use of haemostatic agents and protocols to define the processes of exchange of medical information and data collection.

4.4 The EHTC/EHCCC has access to a laboratory, which may be either internal or external to the Centre, which provides at least the following coagulation tests (see table below which also stipulates the TAT* allowed):

Tests	EHCCC		EHTC	
		24 hr service		24 hr service
PT, APTT, Thrombin time and mixing studies	YES TAT: within 3 hours	YES	YES TAT: within 3 hours	-
Factor VIII and IX assays	YES TAT: within 6 hours	YES	YES	-
Inhibitor screen	YES TAT: within 12 hours	YES	YES	-
Fibrinogen, VWF activity and factor II, V, VII, X, XI assays	YES TAT: within 12 hours	YES	YES	-
Platelet aggregation	YES	-	-	-
VWF antigen and multimers	YES	-	-	-

* TAT (turnaround time: completion time from sample collection to result reporting)

The Turnaround Time for laboratory tests carried out must be agreed in writing between the clinical and laboratory services and be subject to monitoring.

The laboratories that perform the above-mentioned tests must participate in an accredited external quality assurance scheme in haemostasis.

4.5 The EHTC/EHCCC, in collaboration with other Centres and/or patient associations, organizes periodic training events and updates for associated services in order to optimize diagnostic and therapeutic approaches (e.g. specialist services, emergency and other hospital departments, family physicians and paediatricians, pharmacy).

References

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