

STANDARDS AND CRITERIA

FOR THE CARE OF PERSONS WITH
CONGENITAL BLEEDING DISORDERS
(REVISED APRIL 2002)



NATIONAL HEMOPHILIA FOUNDATION
for all bleeding and clotting disorders

MASAC Document #132

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CONGENITAL BLEEDING DISORDERS
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Revised and Approved by MASAC on March 24, 2002 and by the NHF Board of Directors on April 15, 2002.

In May 2000, the Chair of the Medical and Scientific Advisory Council of the National Hemophilia Foundation recommended regular revision of the *Standards and Criteria for the Care of Persons with Congenital Bleeding Disorders*. Last revised in 1994, this document outlines comprehensive standards of care and lists of services that should be provided by Hemophilia Treatment Centers (HTCs). The 1994 edition included an extensive discussion of the staff and services required for proper HIV care for men and women, hepatitis-related services, care for women with bleeding disorders, the need for culturally sensitive outreach programs, and access to clinical research protocols. Subsequent revisions have updated these recommendations in the context of new developments in product safety and efficacy, hepatitis and HIV therapy. This document will be revised periodically and may be viewed at <http://www.hemophilia.org/programs/masac/masac.htm>.

The National Hemophilia Foundation (NHF) consults qualified medical professionals before distributing any medical information. NHF does not, however, engage in the practice of medicine. NHF in no circumstance recommends particular treatments for specific individuals, and in all cases recommends that you consult your physician or local treatment center before pursuing any course of treatment.

THE COMPREHENSIVE HEMOPHILIA TREATMENT CENTER

The Hemophilia Treatment Center has been the model for the delivery of integrated, multi-disciplinary, comprehensive care. We believe its emphasis on early diagnosis and intervention to prevent disease complications provides an optimal health care delivery model for a complex chronic disease. This in turn reduces disability and should help the affected individuals realize their full potential. The network of HTC's has made a positive impact on the health and survival of people with bleeding disorders.

The following describes the composition, general and specific functions, availability and training of the Comprehensive Care Team. Depending on patient census and individual circumstances, varying functions and responsibilities may be combined so that each "member" does not necessarily represent a full time position.

The Treatment Center Team is divided into Core (described below) and Extended team members, (described in the Appendix). All Treatment Center Team members, both Core and Extended, must share a commitment to delivering high quality, patient oriented, culturally competent, comprehensive care of persons with congenital bleeding disorders. Professional practices should be consistent with guidelines, recommendations, and standards developed by the National Hemophilia Foundation (NHF) and its Medical and Scientific Advisory Council (MASAC).

THE CORE TEAM

A. General Requirements:

1. Functions.

- a). Each core team member should participate in each comprehensive clinic as a full member of the assessment team, including the post-clinic discussion, identifying and talking with the patient and family regarding their special needs and providing follow-up with patients as appropriate.
- b). Core team members should provide specialized assistance to persons and families with a congenital bleeding disorder for issues which arise during and between comprehensive clinic visits.
- c). Each core team member should function as an integral part of the team in confidential communication with other team members about patient care and program issues.

2. Availability.

- a). All core team members should have a defined percent of time clearly dedicated to the caring for persons with congenital bleeding disorders. The Medical Director,

Nurse Coordinator and Social Worker should, ideally, have the care of persons with congenital bleeding disorders as their primary clinical assignment.

- b). All core team members should have adequate time available for patient consultation during and outside comprehensive care clinic as needed.
- c). All core team members should regularly participate in team conferences, training, and program development.
- d). The Medical Director or a physician knowledgeable in the management of persons with congenital bleeding disorders must be available for consultation on a 24-hour basis.

3. Training.

- a). Within a year of being appointed to a Comprehensive Care Team, team members should demonstrate understanding of the treatment issues, both medical and psychological, facing the persons and families affected by congenital bleeding disorders. To accomplish these desired learning objectives, this professional training should be carried out: 1) on-the-job by other team members during regularly scheduled team meetings and clinic visits using a defined set of demonstrated skills and knowledge; 2) at professional meetings, and 3) through contact with other coagulation disorder professionals on a state, regional and national level. Documentation of training, skills achieved and demonstrated, evaluator, and recertification must be accomplished.
- b). Each core team member must make a commitment to maintain, update and document their knowledge and skills with respect to coagulation disorders and their complications, including HIV, hepatitis, etc. Administration must commit to providing opportunities for professional training and growth.

B. Core Team Members

- 1. Program Coordinator.** This person is primarily responsible for program development, program oversight, grants submission, program evaluation, and budget implementation. This person should be an MD, RN, MSW, or other qualified professional who coordinates communication inside and outside the team. The written job description should include a role as advocate for optimal patient care. Depending on patient load, these responsibilities may be combined with other core team member functions, but a coordinating role must be clearly defined.
- 2. The Hemophilia Nurse Coordinator.** A RN who has at least two years of hospital nursing experience should fill this position. Recent expertise in outpatient chronic care is preferable. The Hemophilia Nurse Coordinator should have training that includes: hemophilia physical assessment skills; infusion skills; patient/family education training;

and good verbal and written communication skills. This person's responsibilities should include nursing assessment of general health as well as bleeding-related problems; patient/family education; coordination and evaluation of home therapy programs; development and coordination of the patient plan of care; participation in quality assurance activities; facilitation of data collection and documentation; and participation in research studies. The Hemophilia Nurse Coordinator serves as a liaison between the Hemophilia Treatment Center and the community including primary care providers. This person also serves as a consultant to provide education to the lay population and professional groups seeking information about bleeding disorders.

3. **Medical Director.** This position should be filled by a physician with extensive training, knowledge and experience in the care of persons with bleeding disorders and treatment-related complications. This person will make medical decisions for the care of patients seen and participate as an active team member as noted above. A pediatrician, internist or other qualified physician may serve as the medical director. This person should seek consultation for problems or complications of hemophilia/bleeding disorders which fall outside of his/her area of expertise and assist in the coordination of other medical services on behalf of the patient.
4. **Physical Therapist.** A licensed physical therapist knowledgeable in the treatment of musculoskeletal conditions should be a member of the core team. The physical therapist will assess musculoskeletal and functional status, develop educational and exercise programs appropriate for home use, perform pre- and post-operative rehabilitation and conservative treatment protocols, and maximize independent functional status of all persons seen at the comprehensive clinic.
5. **Psychosocial Professional:** A licensed social worker should fill this position. The individual should have inpatient and outpatient hospital experience in the care of persons with bleeding disorders or other life threatening chronic illness and have experience on multidisciplinary medical teams. The social worker should be trained in crisis intervention and be knowledgeable about normal human development and institutional systems (e.g., social networks, schools, hospitals, community, etc.) impacting family functioning. This person will act as a primary resource to patients and their families, including sexual partners, and to the medical team. The social worker's patient responsibilities should include: psychosocial assessments, supportive counseling, health education, counseling resource referral, assistance with employment or insurance problems, loss and grief work, risk reduction, and safer sex education. As a primary resource to the medical team, the social worker should demonstrate candid and supportive communication, identify obstacles to optimal patient care (e.g., institutional rules, communication or personality styles, social circumstances, etc.), and facilitate effective problem solving and conflict resolution. The social worker should be a bridge to community-based organizations serving persons diagnosed with bleeding disorders, their families, and sexual partners.
6. **Case Manager:** This person helps identify appropriate providers and facilitates and coordinates services, while ensuring that available resources are being used in a timely and

cost-effective manner in order to obtain optimum value for both the client and the reimbursement source. Monitoring and evaluating options and services required to meet an individual's health needs as well as promoting quality cost-effective outcomes are also part of case management. Depending on patient load and complexity of required services, these responsibilities may be combined with those of other core team members, such as the nurse or psychosocial professional, but should be clearly delineated.

7. **Secretary:** This individual should provide adequate administrative and clerical support to the Core Team. Basic tasks would include typing correspondence, scheduling clinics, filing, grant or contract assistance, and telephone coverage. Additional tasks, pending adequate support, might include data entry, patient follow-up calls, clinic appointment scheduling, budget assistance, and IRB processing. Should funding permit and training allow, this person could play a key role in local database design and research protocol data access and entry.

COMPREHENSIVE CARE SERVICES PROVIDED BY THE CORE TEAM

I. DIAGNOSTICS AND THERAPEUTICS

A. Bleeding Disorder-related:

1. Comprehensive Evaluation: including but not limited to:

- a). **MEDICAL HISTORY, PHYSICAL EXAMINATION, AND THERAPEUTICS:** These should be age appropriate, culturally sensitive, and targeted towards the specific bleeding disorder and its complications. Core Team members should discuss the natural history of bleeding disorders and the risks, benefits and expected outcomes of various treatment regimens, including prophylaxis.
- b). **NURSING EVALUATION:** Core Team members should assess the patient's/family's understanding of the specific coagulation disorder, its management and potential complications as well as determine educational, support, and treatment follow-up needs. Consideration should be given to the patient's developmental stage, cultural and educational background, and individual patient and family characteristics that might impact on their understanding.
- c). **PHYSICAL THERAPY EVALUATION:** Comprehensive physical therapy evaluation will encompass range of motion, strength, orthopedic joint examination, pain assessment, sensation, posture, gait, functional status, and evaluation for assistive/adaptive devices. Growth and development will be assessed in an age-appropriate manner. Changes in status between comprehensive

visits will be highlighted. Appropriate treatment interventions will be developed and implemented based on evaluation findings. Imaging studies may be indicated to assess joint status.

- d). **PSYCHOSOCIAL EVALUATION:** In addition to areas covered under #4 below, this should include basic assessment for adaptation to diagnosis (es).
- e). **GENETIC COUNSELING:** Core Team should obtain a detailed family history and record this in the patient's chart. Education and discussion about mode of inheritance, carrier testing, and reproductive decision-making options should be done in a confidential, sensitive, age appropriate and culturally competent manner. These activities may be delegated to a trained genetics counselor who functions as a Team member.
- f). **DENTAL EVALUATION:** The Core Team should provide basic education regarding oral hygiene and potential oral complications of bleeding disorders. Brochures about the importance of hygiene should be made available. The Social Worker should assist families or patients in identifying resources available for dental care. A close liaison with selected dentists, oral surgeons, and pedodontists is ideal. A List of Dental Resources should be made available to all families. This would include names, addresses, phone numbers, and referral information for dentists willing to care for patients with bleeding disorders. An annual dental exam by a dentist should be a part of Comprehensive care. The Team should coordinate with dental health professionals and patients who require invasive dental care. This will include discussion of factor replacement, pain control, universal precautions, and antifibrinolytics.
- g). **PERIODIC LABORATORY EVALUATIONS:** Periodic laboratory tests such as a complete blood count, inhibitor screen, and a basic chemistry panel including liver function tests should be obtained as indicated. Reliable and prompt testing by an affiliated, certified laboratory is mandatory. Basic tests such as a urinalysis and a skin test for tuberculosis should be obtained if not provided by the primary care physician.

Thought should be given to which services should be provided at each visit with respect to both provider recommendations and consumer requests.

- 2. **Treatment Products:** All products for treating congenital bleeding disorders per current MASAC guidelines should be readily accessible. These include latest generation plasma derived and recombinant factor VIII and IX concentrates, activated prothrombin complex concentrates, porcine factor VIII, recombinant factor VIIa, desmopressin acetate, and viral-attenuated concentrates containing von Willebrand factor. Contingency plans for dealing with potential product shortages should be created, consistent with national MASAC guidelines. Anti-fibrinolytic drugs and topical hemostatics should be available.

- 3. Home and Self-Infusion:** Education and training in home and self-infusion should be provided to all eligible patients and their families who prefer it. The eligibility requirements and expectations for home care as well as the efforts made to assist the family in fulfilling these should be documented in writing. Universal precautions should be part of all home self-infusion training and should be reviewed repeatedly. Family members who access central venous access devices must receive initial instruction in aseptic techniques, demonstrate mastery of these, and be periodically recertified in their use and care. (See MASAC guidelines at <http://www.hemophilia.org/programs/masac/masac.htm>.)
- 4. Psychosocial Services:** Consistent with the standards and criteria for care of persons with congenital bleeding disorders and their physical and/or psychological consequences, psychosocial services should be provided for all patients and their families. There should be developmentally appropriate services for male, female, pediatric, adolescent, young adult, adult, and elderly persons with bleeding disorders and complications thereof, including but not limited to HIV disease and hepatitis. At the initial interview and at subsequent visits, the following should be determined:

 - a). **MENTAL HEALTH:** Patients and their family or significant other should be assessed for difficulty with adjustment to or inability to accept or understand the patient's diagnosis. The risk of personal and family circumstances, e.g. suicide or homicide, domestic violence, child abuse, substance abuse should also be assessed. Referrals to specialized services should be made as appropriate.
 - b). **BASIC HUMAN NEEDS:** Adequacy of shelter, food, clothing, transportation, and phone service should be ascertained. With consent of the patient, referrals should be made to social service and community-based organizations as appropriate.
 - c). **FAMILY:** Names of family members should be recorded, along with the relationship to the person with the condition (the patient), and their availability to the patient. The records should identify who is the primary caregiver and who makes healthcare decisions when the patient is unable to, either by virtue of mental or physical incapacity or age (child).
 - d). **SOCIAL SUPPORT SYSTEM:** With the patient, sources of social support should be identified among the patient's family, neighborhood, school, other patients at the hemophilia treatment center, place of worship, place of employment, etc. Education and support on behalf of the patient and his/her family member(s) or significant other should be provided. With patient consent, appropriate community-based agency referrals should be made.
 - e). **INSURANCE/FINANCIAL:** Each patient's source of medical coverage and entitlement to benefit programs should be determined, and, with patient consent, referrals should be made as appropriate.

f). **LEGAL ISSUES:** Confidentiality consistent with standards of professional practice and with local, state and federal regulations should be maintained at all times. This includes assurance of informed consent and confidentiality in patient care and patient participation in medical or psychological research. Custody and guardianship issues for minors and/or patients with a court-appointed conservator should be confirmed. Education about and assistance of preparation of forms for "Durable Power of Attorney for Healthcare," living and other wills and "life plan" documents should be provided.

g). **EDUCATION:** Patients should be informed about the provider system, i.e. the roles of team members, access to hospital departments, the legislated rights of persons with special health conditions, etc. Patients should be educated about their rights and responsibilities. Their knowledge, attitudes and beliefs should be assessed and safer sex information provided within this framework.

h). **VOCATIONAL GUIDANCE:** The patient's vocational goals, attitudes towards work, career planning activities, educational history, etc. should be assessed. Functional limitations should be determined. Resources for career planning, vocational training or retraining should be identified and referrals made as appropriate.

5. Services Specifically Related to Women:

a). **GYNECOLOGY & OBSTETRICS:** Women with a bleeding disorder and symptomatic carriers of a bleeding disorder should receive routine gynecologic/obstetric evaluation. Specific attention should be paid to complications of menarche, ovulation and pregnancy including estrogen therapy for menorrhagia, as well as issues relating to menopause and its complications. Core team providers should ensure communication with the obstetrician-gynecologist.

b). **SYMPTOMATIC CARRIERS OF HEMOPHILIA:** Appropriate replacement therapy or, as appropriate, treatment with DDAVP and antifibrinolytic therapy should be available to carriers with a bleeding tendency or a congenitally low factor level for surgical procedures, trauma, and hemorrhagic problems.

c). **GENETIC COUNSELING:** Women affected by a bleeding disorder and carriers or potential carriers of a bleeding disorder, as well as their partners, should receive genetic counseling about the inheritance of the disorder, options for carrier testing and prenatal diagnosis, and reproductive options. This should be done in a confidential, sensitive and culturally competent manner. The emotional impact of these issues should be recognized and appropriate psychosocial support given.

B. HIV-related Care:

- 1. HIV Antibody Testing:** HIV antibody testing should be provided for at-risk persons and their sexual partners, with pre- and post-test counseling by a trained HIV risk-

reduction specialist, ensuring confidentiality consistent with local legislation. Hepatitis and STD screening should also be provided to at-risk persons and their known sexual partners.

2. **Antiretroviral Therapy:** Antiviral therapy, prophylactic therapy, and up-to-date quality of care for HIV infection and AIDS case management including appropriate physical, laboratory, viral and immunologic monitoring should be provided. This may occur on site or by referral elsewhere depending on the patient's and provider's preferences, individual circumstances, and availability of services.
3. **Referral Network:** A referral network should be established for medical and psychosocial HIV-related problems, including but not limited to specialists in infectious disease, dermatology, psychology, psychiatry, pulmonology, gastroenterology, nutrition, neurology, dentistry and pain management who will be available to provide evaluations and recommendations and will develop a respect for and sensitivity to hemophilia and HIV-specific issues.
4. **Psychosocial Assessment:** Resources should be provided for: 1) psychosocial assessment and intervention, as well as stress reduction, for patients and their families regarding HIV infection; 2) stress reduction for center staff to enable them to provide optimal patient care; and 3) support for bereavement, death, and dying for patients, family, and staff.
5. **Women's Issues:**
 - a). **GYNECOLOGIC/OBSTETRIC:** Women affected by HIV infection should receive routine gynecologic and/or obstetric screening for and diagnosis and management of cervical dysplasia and cancer, pelvic inflammatory disease, candida vaginitis, and sexually-transmitted diseases. Pregnancy status should be considered prior to instituting any treatment.
 - b). **PREGNANCY ISSUES:** Women affected by HIV disease should receive support and counseling regarding HIV treatment options as well as reproductive and child bearing options in the setting of HIV infection. This will be done in a confidential, sensitive, supportive and culturally competent fashion.

C. Hepatitis-related Care:

1. **Annual Screening:** Periodic hepatitis screening including liver function testing and basic viral hepatitis serologies (hepatitis A, B, C) should be provided for patients with hemophilia and related bleeding disorders. In addition, all hepatitis B antigen positive individuals should be screened for delta hepatitis. Additional viral genotype and viral

load assays may be indicated. Patients should be counseled about additional risk factors for chronic liver disease such as the use of alcohol.

2. **Hepatitis Vaccines:** All patients and their contacts should be provided hepatitis A and B vaccination, if indicated, following current national recommendations and ACIP guidelines.
3. **Chronic Liver Disease Evaluation:** A basic diagnostic evaluation of chronic liver disease including viral serologies and additional commonly used laboratory tests for diagnosing other causes of chronic liver disease such as autoimmune hepatitis, metabolic and genetic disorders should be done. The patient may be referred to a major research or referral center for more expanded testing, including liver biopsy, if such testing is unavailable at the local Center.
4. **Chronic Liver Disease Treatment:** Appropriate treatment of chronic hepatitis, consistent with the most recent consensus guidelines, including consultation with, or referral to, specialists in liver disease should be available. For individuals with chronic liver disease due to HBV or HCV infection, liver biopsy may be appropriate and can be performed by a percutaneous or transjugular approach under appropriate factor coverage. Opportunity to participate in clinical research protocols for the therapy of chronic liver disease should be available.

II. OUTREACH AND EDUCATION

A. Bleeding Disorder-Related Outreach and Education:

1. **Patient:** Culturally sensitive, educationally appropriate education about the coagulation disorder, its treatment and complications should be provided. A plan of care mutually acceptable to provider and consumer should be drawn up. The care plan should be routinely updated to reflect present and anticipated health care needs.
2. **Family Members:** Outreach should be done to family members including siblings and known sexual partners for diagnosis, treatment and counseling regarding congenital bleeding disorders and their complications.
3. **Culturally Sensitive Outreach:** Culturally sensitive outreach to diverse populations should be done by the members of the core team. Where feasible, an interpreter should be available at clinic and during hospitalizations to facilitate patient/provider communication. Proof of compliance with the Americans with Disabilities Act and Title VI of the Civil Rights Act must be shown. Literature usable by the major language groups in the service area should be available to patients and family members, including sexual partners.
4. **Community Education:** Education should be available for schools, health care professionals, employers, and other relevant agencies.

- 5. Home Infusion:** Home and self-infusion education and training should be available for all appropriate patients and families who request it regardless of background. Universal precautions should be specifically addressed as part of home infusion education and training. Training must emphasize demonstration of mastery of knowledge and technical skills required for safe and effective home treatment and self-infusion of coagulation factor concentrates.

B. HIV-related Outreach and Education:

- 1. Education on Transmission of STD Disease:** STD transmission education, including education about universal precautions, should be provided to all individuals at risk, their identified sexual partners and their families.
- 2. Safer Sex Education:** Safer sex education for at-risk persons, including adolescents, and their identified sexual partners should be provided as permitted by law and should be reviewed regularly. This includes risk behavior assessment and counseling regarding options, including abstinence and barrier protection, in a confidential, age appropriate, sensitive and culturally competent manner. Correct condom use should be demonstrated and information about access to condoms provided. If institutional policy prohibits direct demonstrations and distribution of safer sex products on site, an alternative method of providing this education should be provided.
- 3. HIV Knowledge, Attitudes, Beliefs and Behavior:** Assessment of HIV knowledge, attitudes, beliefs and behavior of patients, their known sexual partners, pregnant partners, and their families should be done by a trained professional in a confidential, sensitive and culturally competent manner.
- 4. Staff Education/Training:** A staff education and training plan related to HIV disease and services should be in place. Training should be consistent with requirements outlined in the Appendix.
- 5. Sexual Function Counseling:** Staff with expertise in sexual history-taking and counseling related to sexual function should be available to patients and their sexual partners.
- 6. Educational Materials:** Appropriate materials relating to sexually-transmitted diseases and HIV should be distributed to patients, partners, and families.
- 7. Women's Issues:** Psychosocial support and counseling should be offered to women affected by HIV disease and their known sexual partners regarding reproductive options and child-bearing in the setting of HIV infection.

C. Hepatitis-Related Outreach and Education:

1. **Hepatitis Counseling:** Counseling regarding hepatitis testing and modes of viral transmission should be provided to patients with hemophilia and related bleeding disorders, their family members and known sexual partners. This should include risks associated with currently available blood products and treatment, risks associated with alcohol consumption, and the risk of heterosexual and vertical transmission.
2. **Universal Precautions:** The importance of following universal precautions during home or self-infusion should be strongly emphasized and reviewed repeatedly. Achievement of desired learning outcomes should be documented.

D. Women's Issues in Outreach and Education:

1. **Bleeding Disorder Education:** Women affected by a bleeding disorder and carriers or potential carriers of a bleeding disorder should receive basic education about the bleeding disorder, options for treatment, and complications. The Center Team should play a leadership role in educating the lay public about bleeding disorders in women.
2. **Genetic Counseling and Reproductive Decision Making Options:** Women affected by a bleeding disorder and potential carriers should receive genetic counseling about the inheritance of the disorder, options for carrier testing and prenatal diagnosis, and reproductive options. This should be done in a confidential, age appropriate, sensitive and culturally competent manner. The emotional impact of these issues should be recognized and appropriate psychosocial support given.

III. COMMUNICATION

A. Bleeding Disorder-Related:

1. **Communication:** Providers should actively work to optimize communication between the team members and patients, consultants, primary care physicians, and other community based health care workers. Team members are expected to set aside clinic time to discuss findings, recommendations, and other relevant issues with patients and to give patients an opportunity to raise questions and concerns. Team members should encourage patients to communicate concerns, needs, or questions to the team both during and outside the routine clinic setting. Procedures and policies to assure above should be in place.
2. **Multidisciplinary Conference:** A multidisciplinary post-clinic conference including all of the core health care professionals attending the comprehensive clinic should take place to coordinate care. A written individualized treatment plan should document pertinent findings and recommendations. Copies of the treatment plan should be sent to the primary care physician, the patient, and to the local emergency room or urgent care center when applicable.

3. **Liaison with Lay Community:** There should be a lay hemophilia advisory group which includes representation of the demographic profile of the patient population. A procedure for seeking regular input from this advisory group at least semiannually should be established and fostered. Records should be maintained documenting input received from the advisory group, as well as the response to these recommendations. A close working relationship with the local NHF chapter or hemophilia organization should be established and maintained.

B. HIV-Related Communication:

1. **Interdisciplinary Communication:** The health care professionals primarily responsible for providing health care for the patient are responsible for insuring that regular communication regarding diagnosis, therapy and prognosis occurs between themselves, subspecialists, the patient, and his/her family. All referrals and their outcome should be documented.
2. **Community Network:** A community network should be in place to link community-based organizations with the center to maximize resources and minimize duplication of services.

- C. Hepatitis-Related Communication:** The health care professionals primarily responsible for the patient's care should establish a close working relationship with gastroenterologists and nutritionists with hepatitis expertise. The particular significance of hepatitis-related coagulation defects in patients with a congenital coagulation disorder should be addressed. The risks and benefits of diagnostic procedures, especially when invasive such as liver biopsy, and therapy, both accepted and experimental, should be clearly explained to the patient.

- D. Women's Issues:** Because women with coagulation disorders usually represent a minority of the patients cared for at a comprehensive center, and because coagulation disorders in women may also affect reproductive and family-caring aspects of their life, staff members should address these issues with sensitivity. Regular communication regarding the medical and psychosocial aspects of diagnosis, treatment, reproductive status, genetic testing, and HIV-related problems should be available to every female patient. These discussions should be confidential, supportive, age-appropriate, and culturally sensitive. Referrals and their outcome should be documented and coordinated by center staff.

IV. FACILITY CONSIDERATIONS

- A. Space:** A designated Center office should be available for comprehensive team staff where records, educational materials, and supplies can be kept. Private space should be available for consultation. A meeting room should be available for Team meetings.
- B. Integrated Medical Records:** Patient records containing all relevant data from comprehensive clinic visits, consultations, laboratory studies and radiologic examinations

should be available on site. Shadow charts or files must be secured and kept confidential. Computer security must be maintained. The organization of the medical records and electronic registries or databases should allow for the protection of confidential information following current institutional, state and federal regulations.

- C. Protocols:** Protocols for management of specific bleeding episodes, teaching of home therapy, assurance of universal precautions and safe needle disposal in the home, etc. should be readily available and centrally located.
- D. Laboratory:** There should be an affiliated, certified, reliable coagulation laboratory capable of performing all coagulation assays. The laboratory should be able to provide assays on an emergency basis when needed for patient care. The laboratory should have the facility to freeze plasma at -80°C to prevent degradation of VIII:C and properly pack and ship samples to be sent to referral labs. Referral laboratories must properly process samples received and properly thaw samples rapidly at 37°C .

V. COMPREHENSIVE CARE REFERRAL SERVICES

Patients needing sophisticated services that require very specific expertise beyond that available at a Comprehensive Care Center may be referred to larger, more specialized Referral Centers. It is not expected that any one Referral Center will have expertise in all areas. On the contrary, it is anticipated that each Referral Center will concentrate on providing one or two such services and that Centers throughout the country will establish close linkages with each other to maximize resources and prevent duplication of efforts. Referral patterns should be clearly documented for all services. Referrals may be made at the request of the consumer. Examples of some sophisticated services that may be offered include:

A. Bleeding disorder-related:

- 1. Surgery:** Surgeons able to perform high-risk surgery on individuals with bleeding disorders and sensitive to the special care requirements they may present.
- 2. Orthopedics:** Availability of specialized orthopedic techniques such as arthroscopic synovectomy in children or radionucleotide synoviorthesis.
- 3. Physical therapy/Rehabilitation:** Expertise in special rehabilitative programs and devices.
- 4. Genetics:** On-site DNA based carrier testing or prenatal diagnosis.
- 5. Psychology/Psychiatry:** Availability of a psychologist and psychiatrist for ongoing, more intensive counseling, neuropsychological testing, biofeedback training, or other specialized services.

6. **Laboratory:** Availability of specialized coagulation tests such as human and porcine inhibitor titers, vWF multimeric analysis, factor VIII or IX gene mutational analyses, and chromogenic factor assays.
7. **Research:**
 - a) Basic and clinical research in the area of bleeding disorders, including experimental protocols of new therapeutic agents and gene therapy when available.
 - b) Psychosocial research examining the psychosocial impact of bleeding disorders on individuals and their families.
 - c) When possible, other facilities treating patients with congenital bleeding disorders should be invited to join in collaborative research so that trials may be available to a larger number of affected individuals.
 - d) There should be documented outreach to other Centers to make research results as broadly available as possible.

B. HIV-related:

1. **Immunology Program:** A strong program in immunology with a focus on AIDS research and treatment.
2. **Clinical Trials:** Designation as an AIDS Clinical Trial Unit or subunit, or participation in other competitively funded clinical, therapeutic research. There should be documented outreach to other facilities treating individuals with congenital bleeding disorders enabling them to register patients in clinical trials.
3. **Psychosocial Research:** Research examining the psychosocial impact of HIV/AIDS on individuals and/or families.

C. Hepatitis-related:

1. **Hepatologist:** A gastroenterologist with special expertise in liver disease, including the performance of liver biopsies for individuals with bleeding disorders.
2. **Research:** Methodologically sound research into the prevalence, natural history and prevention of blood transfusion-related hepatitis as well as protocols for treatment of chronic liver disease.
3. **Psychosocial Research:** Research to examine the impact of hepatitis and its treatment on patients with bleeding disorders.

D. Referral Services for Women:

1. **Genetics:** On-site carrier testing and prenatal diagnosis, including appropriate pre- and post-test counseling.
2. **HIV-related:** Obstetrical, perinatal and pediatric health care providers experienced in managing HIV-infected women during pregnancy, labor and delivery, as well as her child in the immediate postpartum period.
3. **Psychosocial:** Family-oriented medical and psychosocial services, including case management, for HIV-infected women and their children.
4. **Obstetrical and Gynecological:** Wherever possible, routine gynecologic/obstetrical evaluation should be available at the comprehensive visit. If not, then both basic gynecologic/obstetrical evaluation and specialist consultation should be available for the complications of menarche, ovulation, pregnancy and delivery, and menopause.
5. **Research:** Research protocols may be available for diagnosis/treatment of women with congenital bleeding disorders, HIV infection, pregnancy, menstrual disorders and other reproductive issues.

APPENDIX A: EXTENDED TEAM MEMBER REQUIREMENTS

A. General: All extended team members should meet the following guidelines:

1. Functions:

- a). Each extended team member should participate whenever possible in comprehensive clinic as a full member of the assessment team including the post-clinic discussion, identifying needs related to their specialty with patient and family and providing follow-up with patients for identified problems. If the team member is unable to attend clinic, their services can be provided outside the clinic, but written documentation should be included in the comprehensive clinic written summary.
- b). Extended team members should be available to provide specialized assistance to persons and families with a congenital bleeding disorder for issues which arise between comprehensive clinic visits.
- c). Extended team members should be readily available to core team members on a consultation basis. There should be regular, written communication between core and extended team members about both patient care and program issues.

2. Availability: All extended team members should have time available to the core team for consultation and to patients for referrals.

3. Training: Within one year of becoming part of the hemophilia treatment center team, each extended team member should demonstrate understanding of the medical and psychosocial issues of the families affected by congenital bleeding disorders. Core team members should carry out this professional training on the job, and, if possible, professional training opportunities outside the center should be made available to the extended team members.

B. Bleeding Disorder-Related Extended Team Members:

1. **Coagulation Laboratory Director:** This individual should assist the Core Team in the diagnosis of patients with complex bleeding disorders. The Lab Director must ensure that timely, reliable, and accurate factor assays are available for diagnosis and management of patients.
2. **Pharmacist:** If the HTC provides on-site factor or ancillary supplies, a responsible pharmacist must ensure that an adequate supply of both plasma-derived and recombinant products is available for both inpatient and outpatient dispensing. Should the on-site or contracted pharmacy provide home factor delivery, the pharmacist should also ensure timely and dependable delivery of preferred products and ancillaries. Should product shortages occur or be anticipated, the pharmacist and Team will work together to ensure that all patients have factor to treat bleeding episodes.

3. **Dentist:** A dentist who has training, knowledge, and experience in the care of persons with bleeding disorders and treatment-related issues should fill this position. He/she should be part of the comprehensive clinic visit. If a dentist is not physically available at the time of the visit, the dental evaluation can be done at another time, but written documentation should be included in the comprehensive clinic visit written record and the patient's care plan.
4. **Genetics Counselor:** A person trained in genetic counseling, testing and prenatal diagnosis should fill this position. Laboratory tests may be sent to an outside facility.
5. **Orthopedist:** An orthopedic surgeon skilled in emergency and reconstructive procedures should be available to the core team for referral and consultation. The orthopedist should have or be willing to obtain training, knowledge and experience in the care and management of persons with bleeding disorders. The orthopedist will work with the patient and treatment center team in pre- and post-operative planning and goal setting. Surgical procedures will be carried out according to accepted standards of orthopedic and hematologic guidelines.
6. **Obstetrician/Gynecologist:** An obstetrician/gynecologist with expertise in the diagnosis and management of bleeding disorders and complications of menarche, ovulation and pregnancy, including hormone therapy for menorrhagia, should fill this position.
7. **Other:** Health care providers representing other sub-specialties should be available by local consultation or outside referral as deemed necessary by the provider or requested by the consumer.

C. HIV-related Extended Team Members:

1. **HIV/Infectious Diseases Expert:** This position should be filled by an internist, pediatrician, or infectious disease specialist with specific interest, expertise, and experience in the care and management of individuals with HIV infection. This individual should be knowledgeable about the diagnosis, prevention and treatment of infectious complications of HIV disease. This should include skill in administration of antiviral, anti-infective, and prophylactic agents, monitoring for adverse drug effects, and willingness to provide experimental antiviral or immunomodulating agents as appropriate or available, including through experimental protocols. This person should have compassion and sensitivity to the medical and psychological needs of HIV-infected individuals and have a willingness to work in collaboration with the primary hemophilia treatment center physician to assure that adequate hemostasis is maintained throughout prescribed treatments or procedures and to carefully assess potential hepatotoxicity of treatments.

2. **Obstetrician/Gynecologist:** A physician with expertise in the management of women with HIV disease should fill this position. This will usually be an obstetrician-gynecologist but could be a family medicine practitioner or internist depending on specific circumstances. This individual should be willing to discuss reproductive options and child-bearing in the setting of HIV infection in a confidential, sensitive, culturally competent manner.
3. **Dentist:** An individual with a special interest in the prevention, diagnosis and treatment of oral manifestations of HIV disease, as well as familiarity with the management of individuals with bleeding disorders, should fill this position.
4. **Nutritionist:** A registered dietician with training, knowledge and experience in the nutritional needs of persons with HIV infection and bleeding disorders should fill this position. If possible, this person should have experience in direct patient care of HIV-infected individuals.

D. Hepatitis-related Extended Team Members:

1. **Hepatologist:** This position should be filled by a gastroenterologist with specific expertise in hepatitis. This person should be skilled in performing transjugular or percutaneous liver biopsies or refer to a colleague who is skilled and experienced in these procedures.
2. **Nutritionist:** This position should be filled by a registered dietician with training, knowledge and experience in the nutritional needs of persons with hepatitis and bleeding disorders.

APPENDIX B: CARE FOR PERSONS WITH THROMBOPHILIA

- A. **General.** Thromboembolic events (TE) comprise a major cause of morbidity and mortality. Accurate and prompt diagnosis are keys to successful therapy and prevention of complications of venous or arterial thromboembolism. Recent discoveries of common inherited mutations that predispose to such events, e.g. factor V Leiden, have placed the consulting hematologist in new roles. Among hematologists consulted to care for patients with TE events, those who regularly participate in the care of patients with bleeding disorders are uniquely skilled, knowledgeable and experienced in blood coagulation problems and clinical practice. Skills and knowledge of preventive medicine, carrier detection, genetic and prenatal counseling, patient education, blood product use, complications of therapy, and therapy of bleeding that are used in the care of individuals with bleeding disorders are easily applied to the care of people with thrombophilia. Thus, these guidelines are intended for expansion of services by HTC teams to individuals with inherited thrombophilic disorders.
- B. **Core Team.** The Core Team shall consist of a hematologist, pediatric hematologist, nurse coordinator, psychosocial professional, and coagulation laboratory director. Addition of a genetics counselor and physical therapist would be ideal.
- C. **Support.** Medically accepted support activities include readily available, on-site diagnostic imaging studies for thromboembolism including, but not limited to, Doppler ultrasound, CT, MRT, MRA, venography, arteriography, and ventilation-perfusion scanning.
- D. **Laboratory requirements.** An affiliated, reliable, certified coagulation laboratory should provide basic tests of the thrombophilic state, including but not limited to the following:
1. Protein C activity
 2. Protein S activity (free)
 3. Antithrombin III activity
 4. APC resistance
 5. Lupus anticoagulant (two different methods)
 6. Anticardiolipin antibodies, anti-phospholipid functional assay
 7. Fibrinogen (Clauss)
 8. Thrombin and reptilase times (dysfibrinogenemias)
 9. FDP or D-dimer
 10. Assay for heparin-dependent antiplatelet antibodies

A chemistry laboratory should provide homocysteine levels and lipid profiles (HDL, triglycerides, cholesterol, etc.)

A molecular genetics laboratory should provide studies for factor V Leiden (R506Q), prothrombin 20210A, and MTHFR.

Additional studies performed at the affiliated lab or by referral labs might include:

1. Protein C antigen
2. Protein S antigen
3. Antithrombin III antigen

4. Plasminogen assay
5. Tissue plasminogen activator (activity and antigen)
6. PAI-1 (activity and antigen)
7. Markers of thrombin generation, e.g. FP-A and B, prothrombin F1.2
8. Markers of platelet activation, e.g. beta-thromboglobulin, PF-4
9. Fibrinogen antigen assay

Future studies that might become more clinically relevant may include soluble tissue factor, soluble thrombomodulin, thrombin activable fibrinolysis inhibitor (TAFI), tissue factor pathway inhibitor (TFPI), protein Z, and platelet function analyzers.

E. Education. The Core team should design and implement an educational program for patients, family, and other health care providers about the unique aspects of congenital thrombophilic disorders, including natural history, signs and symptoms, diagnosis, therapy, genetic counseling, and preventive aspects.