Goal 1: The Distinction of Immune Thrombocytopenic Purpura (ITP) from other Forms of Thrombocytopenia

This includes consideration of features of thrombocytopenia that would suggest “so-called” nonimmune thrombocytopenia. The first part of the first aim is to distinguish systemic illnesses that might include thrombocytopenia.

The second part of this first aim is to recognize features leading to the diagnosis of nonimmune thrombocytopenia.

Goal 2: To Learn Management of Nonimmune Thrombocytopenia (NIT)

In general, for management of NIT the options are limited to DDAVP, Amicar, platelet transfusion, and more recently, recombinant VIIa. The keys are recognizing which patients are sufficiently affected not to use DDAVP and when the risk of life-or-organ-threatening bleeding is high enough to justify platelet transfusion. Patients with antiplatelet antibodies or those with serious but not life-threatening bleeding may try FVIIa; insufficient data is available to diagnose who will respond and who may develop thrombosis with it. The biggest issue is usually to decide on whether any treatment is required. Finally, location-specific treatment may be used, such as Amicar for dental work or nosebleeds, birth control pills or Provera or estrogen for vaginal bleeding, or vaporizers, packing and, if necessary, cautery for epistaxis.

Goal 3: To Identify When Appropriate to Treat ITP Patients, as well as Treatment Options

This is a considerably debated area and therefore, there is no clear right answer in most cases. For this reason, recognizing the appropriate choice of treatment is important. This includes being able to understand the rate of platelet increase associated with the different therapies, as well as the other advantages and disadvantages of each therapy with regard to duration of effect and to toxicities. This is a complex ever-changing field with many alternatives because no one alternative is a clearly superior alternative. The fellow needs to learn about all of the potential choices and what the pros and cons of each one are.

The fellow learns these skills primarily on the outpatient rotation at NewYork-Presbyterian Hospital-Weill Medical College of Cornell University. The fellow will see a number of patients on the ITP Service during the rotation, including patients who have nonimmune thrombocytopenia. By seeing a number of these patients, presenting their diagnosis, treatment and care, and discussing their disease progression, the fellow will learn from experience to handle the three objectives described above. The third one is the most complicated in certain ways, and the fellow benefits from the number of complex cases sent to the ITP Service for second opinions.

Comprehensive Hemophilia Service

Hemophilia Training

The goals for this experience are expertise in:

the recognition, diagnosis, and management of hemophilia and von Willebrand disease, and rare congenital bleeding disorders;
management of dental procedures/surgery in the above;
management of joint disease in the above;
diagnosis and treatment of inhibitors in hemophilia;
gene therapy options for hemophilia and other disorders;
the diagnosis and management of bleeding disorders in women;
the psychosocial aspects of care for the chronically ill patient;
and familiarity with legislative/advocacy issues with regard to hemophilia.

The goals will be achieved by an intensive clinical experience in hemophilic comprehensive and acute care.

The Regional Comprehensive Hemophilia Diagnostic and Treatment Center currently cares for patients with hemophilia and other coagulopathies.

The clinical program is currently expanding its patient care services. An Isotopic Synovectomy Program has been developed in collaboration with Pediatric Rheumatology and Orthopedics at the Hospital for Special Surgery. It is the first of its kind in the New York State area, and it is anticipated that it will become a regional referral center for outpatient radionuclide synovectomies for hemophilia and rheumatology patients.

In addition, program expansion to include NewYork Hospital-based pediatric dentistry is underway. Although this program currently cares for all NYPH-WMC's bleeding disorder patients, the goal of the current project is to have dental residents and fellows rotate through the hemophilia clinic to further their education on the implications of bleeding diatheses on dental care.

Finally, the clinical program has added physiatry to its consultative services.

Pediatric Coagulation/Thrombophilia Training

The goals of this experience are development of expertise in the etiology, diagnosis, and treatment of the following areas:

congenital/acquired bleeding disorders other than hemophilia;
congenital/acquired thrombophilic disorders;
acquired bleeding/thrombotic disorders in the pediatric cancer/SCT patient;
thrombotic disorders in the adult population (arterial and venous). The goals will be accomplished as follows:

Outpatient Clinic Experience

Pediatric patients with possible bleeding disorders will be screened through a weekly coagulation/thrombophilia clinic. This clinic evaluates all physician referrals and patient self-referrals from both within and outside the extensive Weill-Cornell Hospital network system. A major initiative of NYPH has been the incorporation of pediatric thrombophilia care into the Hemophilia Diagnostic and Treatment Center mission. The active anticoagulation service follows pediatric (and some adult) patients on Coumadin/Low Molecular Weight Heparin long-term therapy. Teaching, home/clinic lab monitoring, and supportive care for these patients are coordinated by Hemophilia Diagnostic and Treatment Center nurses. The Pediatric Hematology/Oncology Service handles approximately 300 consults per year (25 to 30 each month) in a.) pediatric inpatients; b.) ER patients; c.) oncology and SCT patients at NYPH and MSKCC. The majority (80 percent) of these consults are for disorders in hemostasis and thrombosis.

Pediatric Ward/ER/MSKCC Consult Service

Clinical Coagulation Lab Experience

The goals for this rotation are the following:

- expertise in the procedure/performance of all coagulation assays;
- thorough understanding of the technical issues affecting the performance of each assay;
- expertise in laboratory QA/QC issues;
- interpretation of coagulation results within the context of the patient's clinical history/exam.

The Clinical Coagulation Laboratory at NYPH performs the complete range of coagulation assays using state-of-the-art robotics. Molecular diagnostics capacity is currently being developed.

Thalassemia Syndromes/Hemoglobinopathies

Director: Sujit Sheth, MD

The fellowship training program in the thalassemia syndromes is incorporated into two months of outpatient rotation and two months of inpatient rotation, which take place during the first year of clinical training. During these rotations, the goals and objectives of the fellows' training include expertise in the following specific topic areas:

- Understanding of diagnostic issues relating to the thalassemia syndromes:
  - differentiation of the alpha, beta, and other forms of thalassemia;
  - interpretation of hemoglobin electrophoresis patterns and red blood cell size in thalassemia as they relate to the newborn, the older child, and to the adult;
  - genetic counseling for patients with thalassemia syndromes and prenatal diagnosis;
  - diagnosis of hypersplenism;
  - interpretation of the peripheral blood smear in thalassemia syndromes work-ups;
  - diagnosis and evaluation of iron metabolism, hemochromatosis, and iron homeostasis;
  - evaluation of cardiovascular function in thalassemia;
  - evaluation of pulmonary function in thalassemia;
  - evaluation of growth and development in thalassemia;
  - evaluation of osteoporosis in thalassemia;
  - evaluation of hepatitis in thalassemia;
  - radiographic findings in thalassemia;
  - indications for a chronic transfusion program;
  - identification of risk factors and complications of transfusion therapy;
  - evaluation of potential iron chelating agents;
  - evaluation of desferrioxamine iron chelation safety and efficacy.
- Understanding of basic issues relating to the pathophysiology of thalassemia disorders:
  - hemoglobin switching in human development;
  - biochemical properties of abnormal hemoglobins and developmental patterns in thalassemia;
  - pathophysiology of chronic anemia in the thalassemia syndromes;
  - pathologic basis of hypersplenism;
  - pathologic basis of hemochromatosis;
  - iron and immune function;
  - pathologic basis of osteoporosis.
- Understanding of current guidelines for treatment of the thalassemia syndromes:

transfusion therapy;
iron chelation therapy;
bone marrow transplantation;
cord blood stem cell collection;
prenatal diagnosis;
treatment of febrile episodes;
judicious use of antibiotics;
vitamin supplementation: ascorbic acid, folate, and vitamin E;
pneumococcal vaccines;
indications for splenectomy and cholecystectomy;
chronic end organ failure.

Inpatient Rotation (NYPH)

During the inpatient rotations, the fellows take active care of patients with acute complications of thalassemia disorders, as well as those patients admitted to the Clinical Research Center on protocol studies. The inpatient team consists of one attending physician, three to four rotating residents, and the fellow. During these rotations, the fellows learn the basic principles of diagnosis and treatment of various complications of thalassemia.

The fellows' involvement with each patient includes:

a daily review of problems and assessment of the severity of the thalassemia complications;

a daily physical examination with particular attention to evaluation of signs and symptoms of infection, exacerbated anemia, or complications of iron overload;

review of medications and their potential untoward effects;

review of laboratory results;

development of a daily treatment plan;

review of pertinent literature; and

interaction with pediatric and adult consultative services.

Each item is then reviewed with the attending physician on a daily basis.

A manual of guidelines for diagnosis and treatment of thalassemia is included in the annual fellowship manual.

Outpatient Rotation (NYPH)

Fellows routinely attend the Outpatient Thalassemia Clinic, which is held three days each week, as well as on Tuesday evenings and alternate Thursday evenings.

In clinic fellows have the opportunity to evaluate new patients with thalassemia, as well as other non-thalassemia hemoglobinopathies requiring transfusion therapies. All new patients are seen by the fellows for diagnostic evaluation. Previously diagnosed patients are seen for quarterly comprehensive medical and psychosocial visits on Thursdays.

The fellows write the transfusion and medication orders for each clinic and are responsible for coordinating and evaluating all the new thalassemia patients. In addition, fellows are responsible for coordinating quarterly visits for returning patients in the Thursday AM and alternate Thursday PM clinics. The fellows work along with the psychologist, social worker, genetic counselor, and nurse practitioner as necessary. Fellows review patient's treatment plans with the attending physician. Patient evaluations include history, physical examination, review of medications, interpretation of laboratory results, writing prescriptions, and determining return visits. Upon review with the attending physician, treatment plans are developed and modified as necessary.

Sickle Cell Service

The fellowship training program in sickling disorders is incorporated into two months of outpatient rotation and two months of inpatient rotation, which take place during the first year of training. During these rotations, the goals and objectives of the fellows' training include mastery of the following specific topics:

Understanding of diagnostic issues relating to the hemoglobinopathies:

- interpretation of hemoglobin electrophoresis in the newborn;
- interpretation of hemoglobin electrophoresis in the older child and adult;
- genetic counseling for patients with sickling disorders or sickle cell trait;
- diagnosis of functional asplenia;
- differentiation of infection and infarction;
- diagnosis of gallbladder disease;
- diagnosis of osteomyelitis;
- evaluation of cardiovascular function in sickle cell disease;
- evaluation of pulmonary function;
- identification of risk factors for cerebrovascular accidents;
- interpretation of radiographic findings in sickle cell disease;
- interpretation of the peripheral blood smear in sickling syndromes.

Understanding of basic issues relating to the pathophysiology of sickling disorders:

- hemoglobin switching in human development;
- biochemical properties of abnormal hemoglobin;
- pathologic basis of functional asplenia;
- pathophysiology of chronic anemia in the sickling disorders;
- pathologic basis of acute splenic sequestration crisis;
- pathologic basis of the hyperhemolytic episode;
- pathologic basis of transient bone marrow aplasia;
- pathophysiology and manifestations of the acute vasoocclusive episode;
- theories of the pathologic basis of stroke in sickle cell disease.

Understanding of current guidelines for treatment of sickle cell disease:

- judicious use of antibiotics;
- folate supplementation;
- pneumococcal vaccines;
- hydration;
- pain management;
- treatment of febrile episodes;
- transfusion therapy;
- indications for oxygen supplementation;
- gallbladder disease;
- preparation for surgery;
- environmental issues;
- chronic end organ failure.

Inpatient Rotation (NYPH)

During the inpatient rotations, the fellows take active care of patients with sickling disorders. The inpatient team consists of one attending physician,

three to four rotating residents, and the fellow. During these rotations, the fellows learn the basic principles of diagnosis and treatment of various complications of sickle cell disease. Their involvement with each patient includes:

- daily review of problems and assessment of their severity;
- daily physical examination with particular attention to evaluation of signs and symptoms of infection, exacerbated anemia, or of a vasoocclusive crisis;
- review of medications and their potential untoward effects;
- review of laboratory results;
- development of a daily treatment plan;
- review of pertinent literature.

Each item is then reviewed with the attending physician on a daily basis.

A manual of guidelines for diagnosis and treatment of sickling disorders is included in the annual fellowship manual.

Outpatient Rotation (NYPH)

Fellows routinely attend Sickle Cell Clinic, which is held twice monthly. In clinic they have the opportunity to evaluate new patients with sickling disorders, as well as other non-sickling hemoglobinopathies. Previously diagnosed patients are also seen for follow-up and evaluated by fellows. Evaluation includes history, physical examination, review of medications, and interpretation of laboratory results. Upon review with the attending physician, treatment plans are developed and modified as necessary.