July 8, 2016

Ms. Carolyn W. Colvin
Acting Commissioner of Social Security
Social Security Administration
2100 M Street, MW
Washington, DC 20037

Dear Ms. Colvin,

The American Society of Hematology (ASH) appreciates the opportunity to submit comments to the Social Security Administration (SSA) in response to the Agency’s IdeaScale Campaign “Hematological Disorders.”

ASH represents more than 16,000 clinicians and scientists worldwide, who are committed to the study and treatment of blood and blood-related diseases. These diseases encompass non-malignant conditions, such as sickle cell disease (SCD), thalassemia, aplastic anemia, VTE, and hemophilia and malignant hematologic disorders, such as leukemia, lymphoma, and multiple myeloma. In addition, hematologists have been pioneers in the fields of stem cell biology, regenerative medicine, bone marrow transplantation, transfusion medicine, gene therapy, and the development of many drugs for the prevention and treatment of heart attacks and strokes. ASH membership is comprised of basic scientists, physician scientists, and physicians, who are working in diverse settings, including universities, hospitals, and private practices. ASH members also include hematologists who frequently render care to Social Security beneficiaries.

ASH’s comments below respond to the SSA’s questions regarding the SCD provisions in Chapter 7.00 Hematological Disorders - Adult.

Section 7.05 – ASH is concerned that this Section does not account for patients with SCD who manage their chronic and/or acute pain at home, in consultation with their physician. Pain is the most common clinical manifestation of SCD. Patients experience acute and chronic pain episodes that are superimposed on pre-existing chronic pain. The treatment of pain accounts for a large proportion of emergency department visits and hospitalizations of patients with SCD. However, patients frequently manage painful events at home, in consultation with their physician. Despite the fact that they do not physically go to a medical facility for care, the pain is still severe and requires pain medication, bed rest, fluids, etc.

ASH recommends that the SSA add a designation to Section 7.05 that broadens the criteria of an exacerbation of pain to include those episodes managed at home with oral opioids, but that are severe enough to significantly hinder activities such as education and employment. ASH would welcome the opportunity to work with the SSA as the Agency explores ways to expand this Section to assess how to most effectively account for SCD patients who manage their pain at home.
Section 7.05 – ASH is concerned that Section 7.05 does not consider chronic transfusion as "life-saving" for treating SCD. For patients with SCD, especially those who have had a stroke and are at significant risk for a reoccurrence, red blood cell transfusions are the standard of care for preventing future strokes in these patients. The patients who undergo regular blood transfusions for secondary stroke prophylaxis are typically a chronically ill group, who are likely to have other end stage organ diseases (e.g., pulmonary, cardiovascular, neurological or renal disease). Patients with SCD may also require regular blood transfusions for the prevention of frequent or severe episodes of vaso-occlusive pain, acute chest syndrome, or other end stage organ complications. This subset of patients with SCD who are being treated with chronic blood transfusions are probably the most severely affected group of SCD patients. ASH recommends that chronic blood transfusion therapy be added as an independent criterion for patients with SCD.

Response to Question Area 1 regarding new therapies to consider adding to the listings for hematological disorders – There are new curative approaches in the treatment of SCD, which include stem cell transplantation (SCT) from a haploidentical related donor and gene therapy. Gene therapy studies in patients with SCD are becoming more promising, and while currently not yet considered a standard of care, it would be helpful for the SSA to consider adding gene therapy to its current list of therapies that are covered under hematologic disorders. Currently, gene therapy includes myeloablative chemotherapy, which is similar to SCT. Since SCD patients, who receive SCT are considered to be disabled for at least 12 consecutive months from the date of transplantation, ASH recommends that the SSA change its list of therapies to include gene therapy with chemotherapy to be considered at a comparable level of disability, i.e., disabled at the same rate as SCT for at least one year.

Response to SSA’s Question Area 2: If a person has undergone chemotherapy-free SCT, should the person be considered disabled for one year? Or is there a different recovery period for someone who has undergone chemotherapy-free SCT and follow up treatment?

“Chemotherapy free” SCT includes the administration of radiation and immunosuppressive medication leading to the suppression of the bone marrow and of the immune system, analogous to the effects of other forms of SCT. While the complications are lower following chemotherapy free transplants, their application in sick adult SCD patients often results in a similar period of disability following a successful transplant. Patients should therefore be considered disabled for at least one year from the date of the SCT, similar to the period that is used for fully myeloblative transplants.

Sections 7.00 and 7.05 – ASH recommends that the SSA remove "comprehensive" when characterizing a sickle cell center from the following two sections. The majority of the facilities where adults with SCD receive care are not considered “comprehensive,” which means that they do not provide the spectrum of services often needed by SCD patients (i.e. social services, psychological support, education, and genetic counseling). The deletion of “comprehensive” would provide a better description of the type of facilities where adults with SCD receive care.

7.00 C. What are hemolytic anemias, and how do we evaluate them under 7.05?

2. The hospitalizations in 7.05B do not all have to be for the same complication of the hemolytic anemia. They may be for three different complications of the disorder. Examples of complications of hemolytic anemia that may result in hospitalization include osteomyelitis, painful (vaso-occlusive) crisis, pulmonary infections or infarctions, acute chest syndrome, pulmonary hypertension, chronic heart failure, gallbladder disease, hepatic (liver)
failure, renal (kidney) failure, nephrotic syndrome, aplastic crisis, and stroke. We will count the hours you receive emergency treatment in a comprehensive sickle cell disease center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

- **7.05 Hemolytic anemias, including sickle cell disease, thalassemia, and their variants (see 7.00C), with:**

  B. Complications of hemolytic anemia requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive sickle cell disease center immediately before the hospitalization (see 7.00C2).

- **Section 7.18** – SCD is a complex illness that affects multiple organs, and the numerous complications and manifestations of the disease must be considered when developing the criteria to determine disability. ASH recommends making the following additions to Section 7.18 to further account for these complications.

  **7.18 Repeated complications of hematological disorders (see 7.00G2) including those complications listed in 7.05, 7.08, and 7.10, but without the requisite findings for those listings, or other complications (for example, anemia, acute chest syndrome osteonecrosis, retinopathy, skin ulcers, stroke, splenic infarction, silent central nervous system infarction, cognitive or other mental limitation, or limitation of joint movement), resulting in significant, documented symptoms or signs (for example, pain, severe fatigue, malaise, fever, night sweats, headaches, joint or muscle swelling, or shortness of breath), and one of the following at the marked level (see 7.00G4)

Thank you for your consideration of ASH’s comments and recommendations. The Society looks forward to continuing to work with you as you finalize and implement the revisions for evaluating SCD. Please contact ASH Senior Manager of Government Relations and Public Health, Stephanie Kaplan (skaplan@hematology.org or 202-776-0544), if the Society can provide additional information or help.

Sincerely,

Charles S. Abrams, MD
President