



AMERICAN SOCIETY OF HEMATOLOGY

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May 6, 2022

David Berglund, MD, MPH
Medical Officer / Classification and Public Health Data Standards
National Center for Health Statistics
ICD-10-CM Coordination and Maintenance Committee
3311 Toledo Road
Hyattsville, Maryland 20782

Dear Dr. Berglund,

I am writing on behalf of the American Society of Hematology (ASH) in support of the Shwachman-Diamond Syndrome Alliance's request for a new diagnosis code (ICD-10-CM) for Shwachman-Diamond Syndrome (SDS), a rare genetic multi-system disorder. Hematologists are often involved with SDS patients from the onset as many patients present with neutropenia and/or other cytopenias as an early manifestation of their disease. Since patients are at high risk of bone marrow failure/aplastic anemia, myelodysplastic syndrome (MDS), or acute myeloid leukemia (AML), longitudinal management of SDS patients by hematologists is critical to their care.

ASH represents more than 18,000 clinicians and scientists worldwide who are committed to the study and treatment of blood and blood-related diseases. These disorders encompass malignant hematologic disorders such as leukemia, lymphoma, and multiple myeloma, as well as non-malignant conditions such as sickle cell anemia, thalassemia, bone marrow failure, venous thromboembolism, and hemophilia. In addition, hematologists are pioneers in demonstrating the potential of treating various hematologic diseases and continue to be innovators in the field of stem cell biology, regenerative medicine, transfusion medicine, and gene therapy.

ASH agrees that a new ICD-10 code for SDS is needed. Currently, more than 30 ICD-10-CM codes are used to describe the symptoms and clinical manifestations associated with SDS. Some patients with SDS are likely coded as only having neutropenia (D70), for example, while others are likely coded under aplastic anemia, unspecified (D61.9) or pancytopenia (D61.81).

The creation of a new ICD-10-CM code to recognize SDS would greatly impact the SDS patient community as well as the global hematology clinical and research community. The current absence of an ICD-10-CM code for SDS is a substantial barrier in accurately and inclusively identifying SDS patients. This code is needed to facilitate more detailed knowledge related to natural history that can inform anticipatory guidance and the risks and benefits of therapy. Patients accurately coded with an SDS-specific ICD-10-CM code would more easily be flagged for further non-hematologic screenings (e.g., sub-clinical pancreatic screening) as well as enhanced myeloid malignancy screening. Patients with SDS have a high rate of MDS and AML but notably a much higher rate of mortality from MDS and AML than their non-SDS congenital neutropenia peers. Identification of SDS patients will more readily allow for both translational and bench research opportunities that are much needed to understand how best to treat patients with SDS. Additionally, an SDS-specific ICD-10-CM code would remove many insurance related barriers, such as access to granulocyte colony-stimulating factor (G-CSF), as well as help identify patients eligible for new clinical trials.

Thank you for your consideration. Please contact Suzanne Leous, Chief Policy Officer, at sleous@hematology.org with any questions.

Sincerely,

Jane N. Winter, MD
ASH President

2022

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