Patterns of Incidence Rates of Myelodysplastic Syndromes in SEER 21 Regions: 2001 – 2016

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Myelodysplastic syndrome (MDS) is a poorly understood malignancy consisting of a diverse group of bone marrow disorders. The disorder is defined by how it impacts stem cells in the blood. The stem cells will either fail to mature, leading to accumulation of immature blood cells (blasts) or become defective (dysplastic cells), leading to formation of faulty blood cells with a shortened life span. Prior to 2000, MDS was considered preleukemic conditions because a high proportion of patients with MDS eventually progress to acute myeloid leukemia (AML). In 2001, after it was reclassified as a neoplasm, MDS became a reportable malignancy in the Surveillance, Epidemiology, and End Results (SEER) Program. Since then, all participating registries have been tracking MDS occurrence in their catchment areas. Estimates of disease incidence, derived from such surveillance data, are important in understanding the burden of disease, whereas changes in patterns of incidence over time can provide valuable insight on effects of early screening as well as changes in disease diagnosis and coding guidelines. In the case of MDS, while annual incidence rates have been reported, changes in rates over time have not been analyzed. Since it first became a reportable malignancy, several new diagnostic tools have become available, which have improved our understanding of this disease, as reflected by multiple changes in the WHO classification guidelines since 2001. This analysis was performed to assess trends in overall incidence rates for MDS from 2001 to 2016 and assess any difference that may exist across sex, age groups, race/ethnicity, and histologic type.

Incidence data from SEER 21 regions for 2001-2016 period were used to estimate incidence rates using SEER*Stat, version 8.3.6. MDS cases diagnosed during 2001-2016 with the following ICD-O-3 histology codes were included: 9980, 9982-9986, 9989, 9991-9992. All incidence rates were age-adjusted using the 2000 US standard population. Rates were estimated for the total population as well as by histology, sex, age, and race/ethnicity, applying coding changes implemented in 2010.

A total of 86,146 MDS cases were diagnosed during the study period, with an age-adjusted average incidence rate of 4.7 cases per 100,000 population. The majority (~61%) were classified under MDS NOS (ICD-O-3: 9989). Annual rates steadily increased from 3.7/100,000 in 2001 to 5.6/100,000 in

2010, then slowly declined to 3.8/100,000, making a slightly wide inverted v-shaped pattern. This pattern was observed for both sexes, all racial/ethnic groups, and among the 65 and over age group. When the rates were assessed separately by histology, this pattern was observed for MDS NOS (9989), but not for other MDS subtypes.

The patterns of incidence rates observed during the study period are dominated by the incidence rates for MDS NOS, a provisional diagnosis given before all diagnostic information becomes available to indicate a specific diagnosis. The decline in rates since 2010, observed mainly for the MDS NOS subtype, is most likely a reflection of the following types of diagnostic and coding changes, most of which occurred in 2010 to incorporate WHO's 2008 guidelines: 1) definitive diagnostic methods were applied to identify specific subtypes and/or to rule out MDS, 2) therapy-related MDS (9987) is no longer captured under MDS, and 3) diagnosis criteria for AML were expanded to include cases with 20%-30% bone marrow blasts, which means that most cases previously defined as refractory anemia with excess blast in transformation (9984) are now classified as AML. Further analysis is warranted to conclusively determine all factors leading to the changes observed.