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Prognostication in Myelodysplastic Syndromes: Beyond the International Prognostic Scoring System (IPSS)

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To the Editor:

We read with interest the comprehensive review "Myelodysplastic Syndromes: Therapy and Outlook" by Lyons published in ***The American Journal of Medicine*** in July 2012.[1] We agree with Lyons that the International Prognostic Scoring System (IPSS) is a valuable prognostic tool in myelodysplastic syndromes. Nonetheless, there is increasing evidence that a significant group of patients with IPSS low-risk myelodysplastic syndromes has a more aggressive disease with shorter survival than predicted. Identification of such patients would allow for increased surveillance and possibly impact their disease through earlier intervention with disease-modifying therapies.

The IPSS was originally derived from a cohort of untreated patients at time of diagnosis, and it did not account for multilineage dysplasia, transfusion-dependence, or severity of cytopenias.[2] To overcome these limitations, the World Health Organization Prognostic Scoring System incorporated transfusion-dependence and multilineage dysplasia in a flexible scoring instrument to be used at different time points and further refine prognosis in myelodysplastic syndromes.[3] The MD Anderson group developed a new prognostic model (MDAS) by adding age, performance status, and degree of thrombocytopenia.[4] The model was externally validated where 25% of IPSS low-risk myelodysplastic syndromes patients were up-staged by the MDAS.[5] Four subgroups among the IPSS low-risk myelodysplastic syndromes patients were identified with significantly different median overall survival (OS) of 93, 53, 31, and 18 months.[5] A specific low-risk MDAS model also was proposed.[6] Patients were stratified into 3 categories using the sum of points generated from cytogenetics, hemoglobin level, platelet count, bone marrow blast percentage, and age. Bejar et al[7] validated the low-risk MDAS model in an independent cohort of patients, and separated these patients into 3 risk categories with significantly different OS.

Recognizing these shortcomings, a revised IPSS (IPSS-R) was recently proposed.[8] The IPSS-R used 5 rather than 3 cytogenetic prognostic subgroups, split the low bone marrow blast percentage, and used the depth of cytopenias to generate 5 prognostic categories. While this system still needs further confirmation, it has become clear that efforts to better refine prognosis beyond the IPSS are critically important to help clinical decision-making. It also is expected that additional molecular, epigenetic, and immunologic determinants will

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contribute to improving our prognostic tools and, more importantly, allow tailoring therapy accordingly.

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