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Melodysplastic syndrome in Korean children: Clinical findings and application of prognostic scoring systems

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Purpose: Myelodysplastic syndromes (MDS) in children are very rare and there is no widely accepted system for their diagnosis and classification. Thus, clinical characteristics, the most effective treatment and prognostic factors need to be further defined. This multicenter, retrospective study aimed to describe the characteristics and the disease courses of 75 MDS patients from 11 University Hospitals in Korea, seen between 1991 and 2001.

Methods: Kaplan-Meier probability of leukemic transformation and overall survival were plotted. And the usefulness of prognostic scoring systems, including French-American-British (FAB) classification, Bournemouth scoring system (BSS), and International Prognostic Scoring System (IPSS), and World Health Organization (WHO) in the prediction of transformation to acute myelogenous leukemia (AML) and overall survival was evaluated.

Results: The median age was 65 months (2-175 months) and the sex ratio was 2.6 : 1 (M : F). Fourteen patients (18.7%) were unable to be allocated into any subtype of FAB. The frequency of FAB subtypes in Korea was similar to that of Western countries except for higher proportion of refractory anemia (RA, 47.5%). Median survival was 54 months

with Kaplan-Meier 5-yr survival probability of 31.9% and 2-yr probability of transformation to AML was 23.7%. None of the FAB, BSS, IPSS, and WHO was capable of discriminating subgroup of patients for the prediction of survival. However, all of the FAB ($P = 0.004$), BSS ($P = 0.001$), IPSS ($P = 0.02$), and WHO ($P = 0.03$) were able to subdivide subgroups for the prediction of transformation to AML.

Conclusion: The characteristics of pediatric MDS in Korea were different from those of other countries, in light of the higher proportion of RA, the low percentage of inherited diseases, and the low percentage of cytogenetic abnormalities. However, the reasons of the differences were not clear. Moreover, none of the prognostic scoring systems, including IPSS, was reliably predictive of survival, reflecting differences from adult cases. With this multicenter study, we suggest the necessity of a prospective study for the classification and treatment. A newer, effective method should be developed for the prediction of disease progression and survival in pediatric MDS.

Key Words: Myelodysplastic syndrome, Children, Prognostic factors