

vember 1999 and November 2000. None of the patients with Shwachman-Diamond syndrome (SDS) had cytogenetic clones, and their bone marrow morphology did not show significant dysplastic features. Thus, p53 overexpression was the hallmark of the similarity between SDS and refractory anemia (RA). Dr Dror raises the interesting question regarding the relationship between this syndrome and RA. Although we agree with Dr Dror that SDS shares common features with RA, it may not be appropriate to classify all patients with this syndrome as having RA at the time of diagnosis. Other bone marrow failure syndromes, such as Fanconi anemia and Diamond-Blackfan anemia, share some features with RA as well.<sup>2,3</sup> We are concerned that labeling SDS as RA may prompt an aggressive mode of treatment that may not be supported by data other than these similarities. Moreover, there are some indications that myelodysplastic syndromes evolving from an inherited bone marrow disease may not have the same biological behavior as primary myelodysplastic syndromes in children.<sup>4</sup> Long-term prospective studies and the continued search for an underlying molecular defect for SDS should shed some light on this rare disease and its relationship to RA.

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*In Reply.*—We thank Dr Dror for his comments in support of our recent article.<sup>1</sup> Our patients were unselected, and included all of those whose samples were available between No-

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