platelet recovery and number of required plasma exchanges.

Conflict-of-interest disclosure: The authors declare no competing financial interests.

REFERENCES

Transplantation

Comment on Hassan et al, page 3615

HCT survival in ADA-SCID: what’s the buzz?

Sanny K. Chan and William T. Shearer  Baylor College of Medicine; Texas Children’s Hospital

In this issue of Blood, Hassan et al have turned the spotlight on hematopoietic stem cell transplantation (HCT) of adenosine deaminase (ADA)–deficient severe combined immunodeficiency (SCID). They opened up the curtain of beliefs on this therapy that enables facts to be separated from fiction.

Hassan et al show what common beliefs concerning stem cell immunoreconstitution of ADA-SCID are true or not. To be helpful to the expectations of parents of such children, they also raise the critical issue of quality of life and educational and employment accomplishments as important goals not being sufficiently assessed. The results of this study are powerful and persuasive, as it is the only definitive analytical study of a large number of ADA-SCID children given HCT. ADA enzyme replacement and ADA gene therapy are also considered alternate therapies. The authors point out that the ADA deficiency affects many cells and the remedy for the immune cell lineages may not apply to other cell types, such as the central nervous system and respiratory system.

One hundred six ADA-SCID patients were entered into this multicenter analysis of the outcomes of their HCT therapy. The most prominent risk factor for a good outcome is the lack of perfect HLA matching as noted by an earlier publication. In the present study, the only definitive analysis of a large number of ADA-SCID children given HCT, ADA enzyme replacement and ADA gene therapy are also considered alternate therapies. The authors point out that the ADA deficiency affects many cells and the remedy for the immune cell lineages may not apply to other cell types, such as the central nervous system and respiratory system.
responses, suggesting T:B-cell cooperation even with HLA disparity.

The need for such definitive studies as the Hassan et al article grows more intense with the advent of universal screening for all forms of SCID.\textsuperscript{9,10} Seven states and 1 territory have adopted the T-cell receptor excision circle (TREC) test of newborns by DNA analysis of dried blood spots on a Guthrie card. Early results indicate a higher incidence of SCID (1 in 40 000-60 000 vs previous estimates of 1 in 100 000 live births). With more SCID infants coming to diagnosis, including ADA-SCID infants, the need for analyses such as the Hassan et al study becomes urgent.

We thank Hassan et al for their monumental effort in trying to sort out the successes and failures of HCT for ADA-SCID, and performing statistical analysis of their results to better guide physicians and patients in making informed choices of therapy for their special children.

Conflict-of-interest disclosure: The authors declare no competing financial interests.

REFERENCES

HCT survival in ADA-SCID: what's the buzz?

Sanny K. Chan and William T. Shearer