Haemoglobin disorder

Contents

This page was last modified on 7 November 2014, at 23:37.

Condition

Inherited haemoglobin disorders are endemic through Africa, South-East Asia and the Caribbean and the Mediterranean, where the high allelic frequency offers some protection against infectious disease, particularly malaria. They are conventionally divided into disorders of haemoglobin structure (e.g. sickle cell) and haemoglobin production (e.g. the thalassaemias).

By their nature, inherited haemoglobin disorders will always be acquired the recipient of a haematopoietic stem cell transplant, assuming engraftment occurs. However, many carrier states and compound heterozygotes have a clinical silent disease which may be well-tolerated by the recipient. In addition, for ethnic minority recipients who may have an extremely limited choice of donors, the risk of an acquired haemoglobinopathy may be outweighed by the chance of cure.

See here for

Individual at Risk

Recipient

Guidance for CORD BLOOD DONATION

Must not donate if: mother or father homozygous or heterozygous for inherited haemoglobin disorders and infant affected.

Discretionary: If the cord blood or infant/child is tested for the condition and the infant is shown to be unaffected or heterozygous (trait), accept and inform the transplant centre.

Guidance at RECRUITMENT

QUALIFIED, see below

Acceptability at CT / Work-Up

QUALIFIED, see below

Guidance

Each donor with a haemoglobinopathy should be assessed by a physician on a case-by-case basis. In general:

Unacceptable

Thalassaemia major or intermedia

Sickle cell disease (HbSS, HbSC, HbSBthal, HbSD)

High affinity haemoglobin

Other clinically significant structural or functional haemoglobinopathies.

Acceptable if haemoglobin is within normal sex-adjusted range, at the discretion of the transplant centre

Alpha or beta thalassaemia trait

Sickle cell trait

HbC, HbDPunjab/Oman, HbE traits

Other asymptomatic traits or compound heterozygotic haemoglobinopathies e.g. HbC/a-thal trait.

Sickle cell trait

There is no evidence of clinically significant sickling during mobilized PBSC collection in those with sickle cell trait. However, clumping of sickle cells has been observed during post-collection processing. In these circumstances, it is advisable that donors with sickle cell trait should donate by BM.

References

Kang EM, Areman EM, David-Ocampo V, Fitzhugh C, Link ME, Read EJ et al. Mobilization, collection, and processing of peripheral blood stem cells in individuals with sickle cell trait. Blood 2002; 99(3): 850-5.

Castro O, Hardy KP, Winter WP, Hornblower M, Meryman HT. Freeze preservation of sickle erythrocytes. Am J Hematol 1981; 10(3): 297-304.

Meryman HT, Hornblower M. Freezing and deglycerolizing sickle-trait red blood cells. Transfusion 1976; 16(6): 627-32.

Adler BK, Salzman DE, Carabasi MH, Vaughan WP, Reddy VV, Prchal JT. Fatal sickle cell crisis after granulocyte colony-stimulating factor administration. Blood 2001;97(10):3313-4.