NAC ENDORSEMENT: ICTMG GUIDELINES ON RBC SPECIFICATIONS FOR PATIENTS WITH HEMOGLOBINOPATHIES

ENDORSEMENT SUBCOMMITTEE

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Publication Date:	November 17, 2020
Date of Last Revision:	February 26, 2024

Cite As:

Shih A, Fesser J, Musuka C, Webert K. NAC Endorsement: ICTMG Guidelines on RBC Specifications for Patients with Hemoglobinopathies [Internet]. Ottawa: National Advisory Committee on Blood and Blood Products; 2020 November 17 [updated 2024 02 26; cited YYYY MM DD]. Available from: https://nacblood.ca/en/endorsements

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Sickle cell disease and beta-thalassemia are inherited red blood cell (RBC) disorders, where patients often require both acute and chronic RBC transfusion therapy, to treat and prevent life-threatening complications. However, alloimmunization and associated hemolytic transfusion reactions are well-documented challenges in this multiply transfused patient population. Prophylactically choosing matched RBCs for transfusion prevents alloimmunization and fresher units have been suggested to provide benefit, but there is no international consensus given the conflicting balance of incremental patient benefit and RBC availability. Therefore, the International Collaboration for Transfusion Medicine Guidelines (ICTMG) set to provide guidance through a comprehensive literature review by a multidisciplinary international team of experts.

ICTMG requested that the NAC review the guidelines for endorsement, where four transfusion medicine experts from the NAC independently utilized the AGREEII (Appraisal of Guidelines for Research and Evaluation) tool to assess the methodological rigour and quality in which the guideline was developed. The guideline achieved scores deemed acceptable by NAC membership, which was at least 50% in all Domains and a minimum score of 70% in the Domain of Rigour of Development (Domain 3). This review occurred before the formation of the Endorsement Subcommittee, though the standard used in this review is in line with the process detailed in the *NAC Guideline Endorsement Framework*.

The NAC endorsed the ICTMG guidelines on RBC specifications for patients with hemoglobinopathies in 2020 with no updates to the guidelines as of writing. Although many subspecialty hemoglobinopathy clinics in Canada already practice to this standard, the information provided in this guideline will be notably useful in disseminating best practice to centres that do not regularly care for these patients.

The British Society of Haematology (BSH) Guidelines Committee published a position paper in 2020 on this guideline (originally published in 2018), where the authors agreed with the recommendations except that patients with hemoglobinopathies who are alloimmunized should be routinely provided extended phenotype-matched RBCs (CcEe-, K-, Fy^a-, Fy^b-, Jk^a-, Jk^b-, and Ss-matched), if feasible. The BSH Committee notes that routinely requesting blood to be matched beyond Rh and K antigens as well as the antigens to which they have developed an antibody would place considerable demands on Blood Services to support these patients. The NAC Endorsement Subcommittee agrees that extended phenotype-matching is not considered feasible if either delays/reductions in transfusion or reducing supply for difficult to match patients occur, though extended-phenotype matched units should be provided where possible. Consultation with Canadian Blood Services is needed if not feasible in the Canadian setting (except for Québec).

In addition, the ICTMG guideline does not mention recommendations regarding age of blood or HbS-negative units specifically.

- Based on the points below, the NAC Endorsement Subcommittee suggests age restrictions are not routinely required and that extended-phenotype matching requirements should supersede requirements regarding the age of blood.
    - The BSH position paper suggests that its prior recommendations and other UK guidance should stand in regards to age of blood. The BSH guideline published in 2017 recommends “blood provided for SCD patients should be HbS negative and, where possible, should be <10 days old for simple transfusion and <7 days old for exchange transfusion but older blood may be given if the presence of red cell antibodies makes the provision of blood difficult”. The United Kingdom Thalassaemia Society recommendations, previously published in 2016 and updated in 2023, now suggests “Over the recent decades, improvements have been made in the way blood is processed, stored and issued...Age restrictions have therefore been removed for blood required for patients with haemoglobinopathies, including patients with thalassaemia in the UK.”
- The prevalence of HbS is low in Canada, where extended-phenotype matching requirements should supercede requirements regarding HbS-negative units, notably at sites that do not offer red cell exchange.

Guideline Citation:

Compernolle V, Chou ST, Tanael S, et al. Red blood cell specifications for patients with hemoglobinopathies: a systematic review and guideline. Transfusion. 2018 June: 58(6)1555-66. Available from: https://doi.org/10.1111/trf.14611

Other Relevant Citations:

Trompeter S, Massey E, Robinson S. Position paper on International Collaboration for Transfusion Medicine (ICTM) Guideline ‘Red blood cell specifications for patients with hemoglobinopathies: a systematic review and guideline’. British Journal of Haematology. May 2020; 189(3):424-427. Available from: https://doi.org/10.1111/bjh.16405

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Aneke J, Barth D, Ward R, Pendergrast J, Kuo K, Cserti-Gazdewich C. The rationale for abandoning sickle trait screening of red blood cell units for patients with sickle cell disease. Transfusion Medicine. 2019 December; 29(6):466-467. Available from: https://doi.org/10.1111/tme.12603

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ICTMG Webpage Link:

https://www.ictmg.org/hemoglobinopathies-1