Northern Ireland Blood Transfusion Service

POLICY DOCUMENT

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Document Title: NORTHERN IRELAND BLOOD TRANSFUSION SERVICE POLICY FOR HAEMOGLOBINOPATHY TESTING OF BLOOD PRODUCTS AND OTHER NON-MANDATORY TESTING

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Document Authorisation

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This policy has been screened for equality implications as required by Section 75 and Schedule 9 of the Northern Ireland Act 1998.

CROSS REFERENCES

This Policy refers to the following documents:

Doc Type	Doc. No.	Title
APPENDIX	1	Review of HbS testing of Red Cell Products at NIBTS

Key Change From Previous Revision:
Equality screening statement updated.
Appendix updated for 2021 data.

1 STATEMENT

- This policy outlines NIBTS' approach to Haemoglobinopathy testing of blood donations within Northern Ireland. A review of the population demographics of Northern Ireland and the rationale for policy decision making is included as an appendix.

- NIBTS has previously not screened donors for HbS based on risk assessment carried out in 2012 which indicated screening was not required due to the demographics of the population within NI. However, these demographics have changed significantly with those residents from ethnic groups doubling between 2011 and 2021(The Northern Ireland Census 2021 Report: Main statistics for Northern Ireland Statistical bulletin Ethnic group- 22 September 2022) and the data from the antenatal haemoglobinopathy screening programme undertaken in 2022 in the BHSCT indicated that the prevalence of positive sickle cell or thalassaemia carriers was 2.7% amongst total pregnancies.

2 OVERVIEW

- Sickle Cell Disease is an inherited condition characterised by the presence of Haemoglobin S. HbS exists in a homozygous state (S/S) known as a sickle cell anemia or in a heterozygous state as sickle cell trait. Homozygous individuals commonly exhibit symptoms of severe haemolytic anemia or vascular occlusion. Heterozygous individuals are usually asymptomatic carriers.

- HbS and other haemoglobinopathies are particularly common, though not exclusively, in people with an African or Caribbean family background. These mutations can also be found in the Indian /Saudi Arabian subcontinent and other areas such as Southern Europe. These are serious and lifelong health conditions, although treatment can help manage many of their symptoms.

- Current blood transfusion guidelines recommend the use of HbS negative blood components for intrauterine transfusion, neonatal exchange transfusion and for the transfusion of children and adults with haemoglobinopathies. In fact, JPAC Guidelines state each Blood Establishment should have a protocol that "Ensures the use of donations which are HbS screen negative for the manufacture of whole blood and red cell components for intrauterine transfusion, neonatal exchange transfusion and for the transfusion of children and adults with haemoglobinopathy. Also, clinical practice guidelines of the British Society of Haematology states that all components for infants (up to 1 year) should be HbS negative.

- The data from the Northern Ireland Census 2021 Report (Main statistics for Northern Ireland Statistical bulletin Ethnic group- 22 September 2022) clearly reflects the changing diversity in the population in Northern Ireland, at least in the Belfast area, and maybe in the rest of the region, where this will impact on NIBTS, not only in term of increasing blood demand, as the transfusion requirement for these patients is higher, but also for screening blood donors and components issued to them or those at higher risks.

3 RESPONSIBILITY

3.1 Medical Director.

4 POLICY

4.1 GENERAL POLICY

- A review of the population demographics for Northern Ireland has been undertaken and is included as an appendix to this policy.

- Taking into consideration the ethnic mix of the donor population in Northern Ireland and the requirements of the Red Book, BSQR and the Donor Selection Guidelines , it is recommended that NIBTS test for HbS for all components issued as recommended by JPAC "Ensures the use of donations which are HbS screen negative for the manufacture of whole blood and red cell components for intrauterine transfusion, neonatal exchange transfusion and for the transfusion of children and adults with haemoglobinopathy".

4.2 REVIEW OF POLICY

- This policy should be reviewed in the light of any significant update of available information on the population demographics of Northern Ireland, such as the population census.

4.3 REFERENCES

- The attached appendix paper ("Review of HbS testing of Red Cell Products at NIBTS") provides further detail on the decision-making process involved in this policy and provides a full reference list of documents accessed.

5 EQUALITY SCREENING AND ACCESSIBILITY

This policy has been drawn up and reviewed in light of the statutory obligations contained within Section 75 of the Northern Ireland Act (1998). In line with this statutory duty of equality this policy has been screened against particular criteria. If at any stage of the life of the policy there are any issues within the policy which are perceived by any party as creating adverse impacts on any of the groups under Section 75 that party should bring these to the attention of the Head of HR & Corporate Services.

The Northern Ireland Blood Transfusion Service is committed to the promotion of equality of opportunity for staff, donors and service users. We strive to ensure that everyone is treated fairly and that their rights are respected at all times. We believe that it is important that our policy is understood by all those whose literacy is limited, those who do not speak English as a first language or those who face communication barriers because of a disability. On request it may be possible to make this policy available in alternative formats such as large print, Braille, disk, audio file, audio cassette, Easy Read or in minority languages to meet the needs of those not fluent in English.

6 TRAINING REQUIREMENTS

- NIBTS Medical Consultants and Specialty Doctor should have read and understood this policy.

APPENDIX 1

Review of HbS testing of Red Cell products at NIBTS

- Sickle cell disease is a chronic haemolytic anaemia caused by a point mutation in the β-globin gene which causes insolubility of HbS in its deoxygenated state. These insoluble chains crystallise in the red cells causing sickling and vascular occlusion.
- There are several types of sickle cell disease, depending on which types of haemoglobin mutations have been inherited. The most common are: Sickle Cell Anaemia (SS), Sickle-Haemoglobin C Disease (SC disease), Sickle β^+ thalassaemia and Sickle β^0 thalassaemia.
- Sickle Cell trait (HbAS) is an inherited condition in which both haemoglobin A and S are produced in the red blood cells, always more A than S. Sickle cell trait is not sickle cell disease; individuals with sickle cell trait are generally healthy and may not even be aware of their 'carrier' status.
- The HbS mutation originated in at least 4 places in Africa and in the Indian/Saudi Arabian subcontinent. It exists in all countries of Africa and in areas where Africans have migrated. It is most common in West and Central Africa where as many as 25% may have sickle cell trait (AS) and 1-2% of all babies are born with a form of the disease. Sickle cell disease is also present in Portuguese, Spaniards, French Corsicans, Sardinians, Sicilians, mainland Italians, Greeks, Turks and Cypriots. The HbS gene also appears in most of the Near and Middle East countries including Lebanon, Israel, Saudi Arabia, Kuwait and Yemen, and as such, sickle cell disease constitutes an international health problem.

Donor Selection Guidelines:

- WB&C-DSG Edition 203, release 66(October 2022) specifies *Must not donate* for sickle cell or thalassaemia disease. There is a discretionary accept for donors with haemoglobin traits (i.e. heterozygous for condition) with the note that they must not donate as a component donor (risk of sickling in the apheresis equipment) and they are not suitable for blood for intrauterine or neonatal use. There is also a much increased 'filter fail' rate with donations from sickle trait (HbAS) donors.

Risks associated with transfusing HbS blood:

The risks are two-fold:

1. Transfusion of HbS blood to neonates, particularly as a large volume /exchange transfusion.
2. Transfusion of HbS trait blood to a child or adult with Sickle cell disease.

Estimates of sickle cell (HbSS) patients living in N. Ireland:

- A recent update identified 34 patients with sickle cell disease currently living in Northern Ireland, of which, at least one is a child requiring regular blood exchange. Data taken from the most recent Census (2021) has shown that 3.4% of the population, or 65,600 people, belonged to minority ethnic groups. This is around double the 2011 figure (1.8% – 32,400 people) and four times the 2001 figure (0.8% – 14,300 people). With this increase in minority ethnic groups, will come an increase in potential donors and also patients requiring treatment/management due their anaemias.

- In addition, the antenatal haemoglobinopathy screening programme undertaken in the BHSCT detected 120 positive sickle cell or thalassaemia carriers amongst 568 women screened out of 4372 pregnancies. This clearly reflects the changing diversity in the population in Northern Ireland, at least in the Belfast area, and maybe in the rest of the region. This data makes 2.7% of the total pregnancies are carriers of haemoglobinopathy, suggesting that Belfast is becoming a high prevalence area. This will impact on NIBTS, not only in term of increasing blood demand, as the transfusion requirement for these patients can be extremely high, but also for screening blood donors and components issued to them or those at higher risk.

Risk of transfusing blood from a donor not identified as Sickle trait (HbAS):

- These indicate that there is now potentially an increasing number of patients who require HbS negative blood and are not receiving the optimal standard of care, as an increasing number of donors may now be HbS positive and this can lead to potential patient harm.

Recommendations:

On review of the best available data at present, it is recommended that NIBTS tests for HbS for the components as per JPAC recommendations.

References:

Northern Ireland Census 2021*, accessed at* https://www.nisra.gov.uk/