Scottish National Blood Transfusion Service
Policy Record

Ref: NATP CLIN 034 04
Cat: Clinical

Title: The transfusion management of patients found to be IgA deficient

Background:

This policy describes the management of transfusion support for patients with IgA-deficiency with or without preceding allergic transfusion reactions.

It does not provide advice on the general management of IgA-deficiency.

Key Changes from Previous Revision
Updated Contact details for NHSBT to request IgA-deficient products – now Barnsley instead of Sheffield as well as Filton. Reference to new version of policy: NATP Clin 047: The provision of Blood Components for Patients with a Special Transfusion Requirement. Update reference to NHSBT guidance document to new version IFN 486/1.6

Policy Agreement	CGSG: Not required as minor reference changes only.	BBWG: review requested at meeting on 15 FEB 2023– no policy updates required.
Supersedes Policy Ref:	NATP CLIN 034 03	
Date Of Implementation:	03 APR 2023	

Scottish National Blood Transfusion Service
Policy Record

Ref: NATP CLIN 034 04
Cat: Clinical

Management of transfusion support for patients with IgA-deficiency

Background

Primary selective Immunoglobulin A deficiency (IgAD) is defined as undetectable IgA levels (<0.07g/L) with normal levels of IgG and IgM. Primary IgAD is the most common immunodeficiency in humans, with an incidence of between 1 in 143 and 1 in 965. The majority of patients with IgAD are detected incidentally and are asymptomatic.

Patients with selective IgA deficiency can form antibodies if exposed to IgA which is present in blood components containing plasma (FFP, platelets, cryoprecipitate and red cells) as well as in intravenous immunoglobulin and anti-D Ig. Some patients develop antibodies without known exposure so the presence of anti-IgA antibodies is not thought to be predictive of anaphylactic transfusion tractions.

Transfusion reactions in IgA-deficient individuals: Reactions to IgA are typically allergic/anaphylactic in nature. The incidence of severe transfusion reactions in this context is extremely rare and appears to be confined to individuals with very low levels of IgA (4, and HT, verbal communication). The incidence of IgA anaphylactic transfusion reactions is estimated at 1:20000 – 1:47000 transfusions (5). Although some have argued against the evidence for the existence of this diagnostic entity. (3, 6)

Data from cumulative haemovigilance (SHOT Reports)
Only 1 case of IgAD in the context of an acute transfusion reaction (ATR) was reported to SHOT in the period of 2004-2009. In the 2010 SHOT report two cases of low IgA-levels were reported in the investigation of 62 ATRs: One with an allergic reaction and one with a febrile reaction (7). The SHOT reports for the period 2011-2019 included 14 cases of acute transfusion reactions where low anti-IgA levels were detected. Not all patients had anti-IgA antibodies detected and not all demonstrated the hyper-acute transfusion reaction typically associated with this diagnosis (7).

Patients with a past history of severe reactions are at greatest risk of further reactions.

Investigation of transfusion reactions in the context of suspected IgAD

If IgAD is suspected as a possible cause of an acute transfusion reaction, the IgA-level should be determined by an appropriate local hospital-laboratory. If IgA deficiency is confirmed, consideration should be made to testing for anti-IgA antibodies, though *the predictive value of the anti-IgA antibody level assessing the likelihood of reaction is poor. This test is not offered by SNBTS but can be performed by arrangement with NHSBT.*

Blood Components for IgA-deficient patients

Most IgA-deficient patients with or without anti-IgA-antibodies will not experience severe reactions with standard components.
Any requests for special components should always be discussed with SNBTS-Medical Staff. Specifically, information on the patient's transfusion history and general history of allergic reactions, reasons for measuring IgA-level, urgency of transfusion and likely future transfusion needs should be assessed.

General information on special components

All requests for special components such as plasma-reduced components or components from IgA-deficient donors must be discussed with and authorised by SNBTS-medical staff. Plasma-reduced components are recommended not only in many patients with known IgAD, but also for some other indications, esp. patients suffering with recurrent severe allergic/anaphylactic transfusion reactions due to other reasons (see SNBTS-policy NATP CLIN 047 02: Provision of Blood Components for Patients with a Special Transfusion Requirement. June 2020).

SNBTS: SNBTS can provide platelets in platelet additive solution (PAS). Production of this component takes 3-4 hours, and the component has a shelf-life of 24 hours. The protein content is usually less than 0.5 g per unit.

Washed Red cells (automated, closed system method) can usually be provided within 3 hours and have a shelf-life of 14 days. They will require crossmatching at the local hospital blood-bank.

Of note, transport times from JCC to local hospitals must be added to the times mentioned above for the production of plasma-reduced components, to give a practical supply time.

NHSBT: NHSBT keep a small stock of IgA-deficient red cells and FFP, and are able to contact a small number of IgA-deficient platelet and plasma donors. Any request to / contact with NHSBT must be mediated by SNBTS.
IgA-deficient cryoprecipitate is not available.

IgA deficient plasma is available from Barnsley Hospital Services (tel 0122 686 8061), or Filton Hospital Services: (tel 0117 912 (2)5724) Issue requires approval by an NHSBT consultant.

NB: For choice of immunoglobulin-products and other plasma derivatives it is advised to discuss patients with an immunologist and check information on IgA-content provided by the manufacturer.

Specific transfusion advice

Patients with a past history of severe transfusion reactions are at greatest risk of further reactions.

The consequences of a delay of transfusion incurred by seeking special components should always be balanced against the risk of a reaction. In an emergency, *standard* red cells in SAG-M and *standard* platelets should be used.

The clinical team looking after a patient with IgAD should confirm the diagnosis and discuss the indication for the transfusion and its urgency with their local Haematologist.
Patients with confirmed IgAD and previous transfusion reaction should be flagged up on the hospital laboratory computer system and notified to other blood banks in Scotland where systems are in place to do so.

Any requests for special components will then require a discussion between the clinical team, local Haematologist and SNBTS medical staff. SNBTS-medical staff must authorise any special components and point out practical issues such as their preparation time and shelf-life. SNBTS-medical staff must also inform relevant SNBTS-processing staff of the outcome of discussions.

Transfusions for IgA-deficient patients should be closely monitored in an area where severe allergic and anaphylactic reactions can be managed appropriately.
Depending on the circumstances, premedication with or early use of hydrocortisone and chlorpheniramine may be considered.

RBC TRANSFUSION

1. Transfuse with red cells from an IgA deficient donor if time permits.

2. If not, then transfuse with washed red cells.

3. In emergencies use standard SAGM RBC (11).

FFP/PLATELET TRANSFUSION

Where time permits use FFP / Platelets from IgA deficient donors or use washed platelets if in the emergency situation.

The management of **patients with IgAD but *no* history of acute transfusion reactions** is far less clear (9, 10). They rarely suffer serious transfusion reactions.

However, especially if they have suffered other allergic reactions or if they are likely to require repeated transfusions in the future, provision of plasma-reduced components should be considered as far as the urgency of the transfusion allows.

Decisions in these situations should be made on a case by case basis.

Future introduction of a highly sensitive assay for the identification of individuals with *severe* IgAD would help to focus support with special components to those patients at highest risk of transfusion reactions.

Literature

1. European Society for Immunodeficiencies: Definition of IgA deficiency, 2005.

2. Latiff AHA, Kerr MA. The clinical significance of immunoglobulin A deficiency. Ann Clin Biochem 2007; 44: 131-139

3. Lillic C, Sewell WAC. IgA-deficiency: what we should –or should not-be doing. J Clin Pathol 2001; 54: 337-338

4. Sandler SG. How I manage patients suspected of having had an IgA anaphylactic transfusion reaction. Transfusion 2006; 46: 10-13

5. Pineda AA, Taswell HF. Transfusion reactions associated with anti IgA-antibodies: Report of four cases and review of the literature. Transfusion 1975; 15: 10-15

6. Sandler S.G, Eder A.F., Goldman M., Winters J.L. The entity of immunoglobulin A-related anaphylactic transfusion reactions is not evidence based. Transfusion Vol. 55, January 2015 p.199- 205.

7. SHOT Annual Reports 2009 - 2019. Website: www.shotuk.org/shot-reports

8. Munks R, Booth JR, Sokal RJ. A Comprehensive IgA Service provided by a Blood Transfusion Centre. Immunohaematology 1998; 14: 155-160

9. NATP CLIN 047 05: Provision of Blood Components for Patients with a Special Transfusion Requirement. JAN 2022.

10. Tinegate H. Investigation and Clinical Management of suspected Reactions to immunoglobulin A (IgA). NHSBT Information 2019; INF 486/1.6

11. BSH guideline on the investigation and management of acute transfusion reactions, 2012. Website: www.bcshguidelines.com