

The Relevance of Scianna in Science Transfusion

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Edição 15/2021
27 Maio a 3 Junho

Introduction

- The Scianna blood group was discovered in 1962
- It consists of seven antigens recognized by The International Society of Blood Transfusion:
 - Sc1 and Sc3 are high frequency
 - Sc2 and Sc4 (Rd) are low frequency.
 - Three other antigens are the Sc5 (STAR), Sc6 (SCER) and Sc7 (SCAN).
- They are expressed by the erythroblast membrane-associated glycoprotein (ERMAP), which is encoded on chromosome 1p34.2.

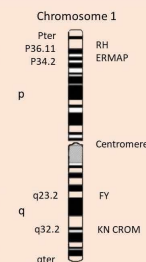


Fig. 1: Genomic Location of the ERMAP Gene in Chromosome 1

ERMAP

- The ERMAP protein is expressed in the RBC and its function is not certain even though it is known that it might have a function in the erythropoiesis.
- ERMAP Is a Member of the Butyrophilin-like Family of the Immunoglobulin Superfamily**

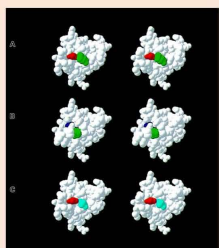


Fig. 2: Stereo View of a 3D Model of the ERMAP Immunoglobulin Domain

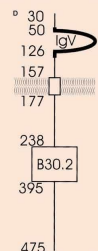


Fig. 3: Schematic Representation of the Protein Topology

- (A) In the wild-type ERMAP expressing Sc1 but lacking Rd antigens, there is a Gly (red) at position 57 and a Pro (green) at position 60;
- (B) The Sc2 antigen is caused by an Arg (blue) at position 57;
- (C) The Rd antigen by an Ala (light blue) at position 60;
- (D) The topology of the ERMAP protein.

- Sc3** • Nucleotide variation is located at position 103
- Sc5** • Nucleotide variation is located at position 47
- Sc6** • Nucleotide variation is located at position 81
- Sc7** • Nucleotide variation is located at position 35

Antigens

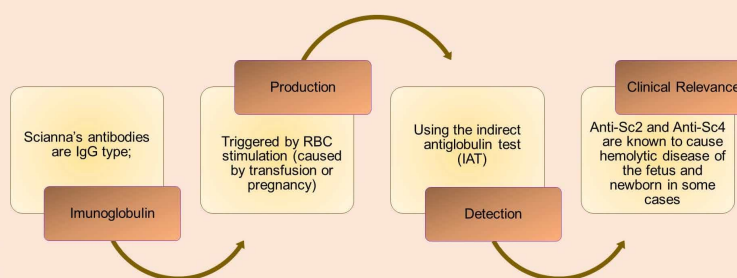
- In most of the cases, Sc antigens are resistant to cin plus papain, trypsin, α-chymotrypsin and sialidase.
- The Scianna blood group has various possible phenotypes through the combination of its antigens. Their prevalence in two different populations is described on Table 1.

Phenotype	Caucasians	Blacks
Sc: 1, -2	99%	100%
Sc: 1, 2	1%	0%
Sc: -1, 2	Very Rare	0%
Sc: 1, -2, 4	Very Rare	Very Rare
Sc: 1, 2, 4	Very Rare	0%
Sc: 1, -2, -5	Very Rare	0%
Sc: -1, -2, -3 (Null)	Very Rare	0%

Table 1: Percentage prevalence of Scianna Phenotypes

Antibodies

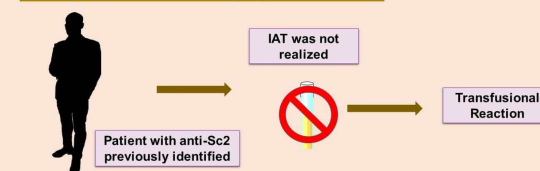
- The main obstacle to detect Sc antibodies is the lack of serologic and RBCs reagents.
- The most logical approach would be to use recombinant Sc protein during pretransfusion testing to detect antibodies and establish a threshold titer.



Clinical Relevance

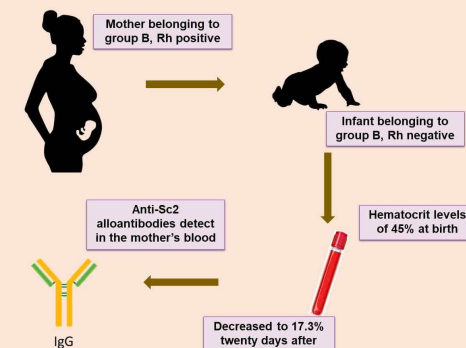
Antibodies to the Scianna blood group are not known to cause any severe hemolytic reaction, although in some rare cases it has been reported their clinical relevance:

• Hemolytic transfusion reaction (HTR) due to anti-Sc2:



- The patient's symptoms included fever, rigors, nausea, and abdominal pain.
- The patient's plasma showed a phagocytic index greater than 50% in a monocyte monolayer assay, that correlated very well with clinical relevance.

• Hemolytic disease of the newborn related to anti-Sc2:24



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