

# Transfusion Management of IgA deficiency

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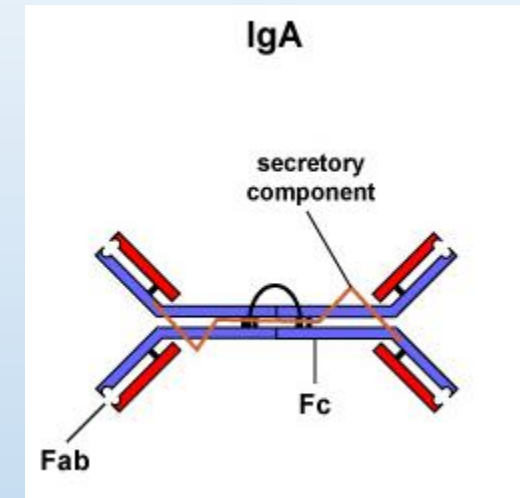
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# Our patient

- 63 year-old female with a duodenal stricture who was admitted with a bowel obstruction due to migrating duodenal stent
- Saturday afternoon, surgery intern calls the blood bank requesting 4 prbcs, 4 plasma and 4 platelets from an IgA deficient donor in preparation for surgery on Monday morning (blood type is AB positive)
- Patient reported to the surgeon a history of IgA deficiency
- Surgery is urgent and can only be delayed an extra day or two at most

# Selective IgA deficiency

- IgA deficiency is the most common immunodeficiency in Caucasians (1 in 500 to 700)
- Defined as IgA level  $< 7$  mg/dL in individuals older than 4 years of age
  - Only a minority of patients with selective IgA deficiency are severely deficient ( $< 0.05$  mg/dL)
  - Some patients with severe deficiency have a detectable anti-IgA



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# Selective IgA deficiency

- In severely IgA deficient patients, transfusion may result in an anaphylactic/anaphylactoid transfusion reaction
  - Anti-IgA may be naturally occurring without history of prior blood product exposure (although role of anti-IgA is unclear?)
  - Anaphylactic reactions can develop after a very small exposure to IgA, and may occur with the first transfusion
- Patients with less severe IgA deficiency (less than 7 mg/dL but greater than 0.05 mg/dL) may also have an increased risk for allergic transfusion reactions

# Transfusion options for IgA deficient patients

- Autologous products
- Washed or deglycerolized PRBCs or washed platelets
- Products collected from IgA deficient donors—through the American Rare Donor Program



# More history...

- The patient is quite knowledgeable about her medical history
  - She was diagnosed with IgA deficiency at the Mayo Clinic in the 1970s—they told her she can never have a regular blood transfusion or she will have a severe reaction
  - She has never had a blood transfusion in her life
  - She does not have any autologous products available
  - Several months ago she was supposed to have an elective cardiac surgery, but it was cancelled because they could not find appropriate blood products

# Labs

- Her hemoglobin was stable but low (around 8 g/dL)
- Her PT/INR were mildly elevated on admission (INR 1.5), but trending down
- Her platelet count was normal (around  $240 \times 10^9/L$ )
- Her primary care doctor ordered a celiac panel several years ago which reported an IgA level of less than 7 mg/dL (low sensitivity IgA test)
- We requested a high sensitivity IgA level and anti-IgA testing
- Contacted American Red Cross Rare Donor Program to determine options



<https://www.damas17podologia.es/>

**Table 1.** Guidelines for the distribution of IgA deficient plasma in emergency situations (needed within 4 days\*).†

	1	2	3	4	5	6	7	8	9	10	11
History of reaction	X	X	X	X	X	X	X	X			
No history of reaction ✓									X	X	X
No IgA level available	X										
Routine IgA tests—IgA deficient ✓		X	X	X	X				X	X	X
Sensitive IgA tests—no IgA			X	X	X					X	X
IgA present						X	X	X			
No anti-IgA performed ✓	X	X	X			X			X		
Anti-IgA present				X				X		X	
Anti-IgA not present					X		X				X
Decision‡	B,D	A,C	A,C	A	B	B,C	B	A	A,C	A	B

† Each of the columns represents an individual requesting scenario.

‡ Decision Key

- A. Release IgA deficient plasma
- B. Do not release IgA deficient plasma
- C. Ask for a pretransfusion sample for IgA and anti-IgA studies
- D. Ask to have a routine IgA level performed locally



# The plan

- Obtained 2 frozen rare donor units of IgA deficient group A plasma
- Planned for 4 washed PRBCs to be ready on morning of surgery
- Washed platelets were not an option, however we were optimistic that she would not need platelets given her normal platelet count
- Requested limited blood draws/use of pediatric phlebotomy tubes to limit hospital acquired anemia
- Based on lab results, patient was a candidate for IV iron and erythropoietin which were administered to treat her anemia

# The plan

- I communicated this plan to the surgeons, and they opted for a less invasive endoscopic procedure rather than an open procedure
- The main surgery was rescheduled for several weeks later after hemoglobin could be optimized
- No blood products were required for the endoscopic procedure

# While we were waiting for surgery...

- With IV iron and erythropoietin, hemoglobin improved to normal over several weeks!
- IgA levels returned as undetectable for all subclasses (high sensitivity testing)
- Anti-IgA testing could not be performed, however records were obtained from another hospital which had previously confirmed the presence of an anti-IgA

**Table 2.** Guidelines for the distribution of IgA deficient plasma in non-emergency situations (not needed within 4 days\*).†

	12	13	14	15	16	17	18	19	20	21	22
History of reaction	X	X	X	X	X	X	X	X			
No history of reaction									X	X	X
No IgA level available	X										
Routine IgA tests—IgA deficient		X	X	X	X				X	X	X
Sensitive IgA tests—no IgA			X	X	X					X	X
IgA present						X	X	X			
No anti-IgA performed	X	X	X			X			X		
Anti-IgA present				X				X		X	
Anti-IgA not present					X		X				X
Decision‡	B,C	B,C	B,C	A	B	B,C	B	A	B,C	A	B

\* The 4 day time frame is intended to allow for the time required for rapid shipment of serum samples and high sensitivity IgA and anti-IgA testing. Geographic and transportation circumstances may require more or less than 4 days for results.

† Each of the columns represents an individual requesting scenario.

‡ Decision Key

- A. Release IgA deficient plasma
- B. Do not release IgA deficient plasma
- C. Ask for a pretransfusion sample for IgA and anti-IgA studies
- D. Ask to have a routine IgA level performed locally

# Followup

- Because the hemoglobin improved with IV iron and erythropoietin, no transfusions were required perioperatively
- After the patient was discharged, we received a market withdrawal on one of the units of IgA deficient group A plasma; a subsequent donation had reactive screening tests for anti-HCV and Trypanosoma cruzi antibody; the unit was destroyed
- The other unit was returned to ARDP

# Summary

- When IgA deficiency is suspected, obtain a detailed patient history and carefully review lab results to determine if specialty products are required
- If the patient has a severe IgA deficiency, consider autologous products, washed products, or products from an IgA deficient donor
- Use blood management strategies to reduce the need for transfusion