Case Study
An Unusual Case of Anti-JK3 Alloantibody and Implications for Pregnancy Management

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Patient History

• 32 year old Caucasian woman from Mennonite community
• Recently moved to Columbia, MO from PA
• Presented to OB/GYN in Oct 2015 at approximately 21 weeks gestation
• 9 prior pregnancies with 4 1\textsuperscript{st} trimester spontaneous abortions
• Known anti-Jk3, and anti-E from outside hospital record
Jk (Kidd) Antigen

- Part of urea transporter on RBCs
  - Human urea transporter 11 (HUT11)
- Transports urea across cell membranes in the hypertonic renal medulla
  - Prevents cells from shrinking and swelling
## Prevalence

<table>
<thead>
<tr>
<th>Phenotype</th>
<th>Whites</th>
<th>Blacks</th>
<th>Asians</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jk(a+b-)</td>
<td>26</td>
<td>52</td>
<td>23</td>
</tr>
<tr>
<td>Jk(a+b+)</td>
<td>50</td>
<td>40</td>
<td>50</td>
</tr>
<tr>
<td>Jk(a-b+)</td>
<td>24</td>
<td>8</td>
<td>27</td>
</tr>
<tr>
<td>Jk(a-b-)</td>
<td>&gt;0.1</td>
<td>&gt;0.1</td>
<td>&gt;0.1</td>
</tr>
</tbody>
</table>
• Jk(a-b-) or null phenotype is rare
  – Discovered in 1959: antibody against both Jka and Jkb in a female patient
    • Called antibody anti-Jk3
  – Increased prevalence in Polynesians
    • 0.9% overall, 1.4% in Niueans
  – Increased prevalence in Finns
    • More rare than in Polynesians
Where is Polynesia?
Where is Niue?
Anti-Kidd Antibodies

- Found in antibody mixtures
- Mostly IgG
- About half are able to fix complement → extravascular/intravascular hemolysis
- Transient and exhibit dosage
- Common cause of delayed HTRs
  - Likely due to transient nature on detection
- Rarely cause acute HTRs
Anti-Kidd and HDFN

- Rarely cause severe HDFN
- Anti-Jka first described in 1951; detection of antibodies against unknown RBC antigen in Mrs. Kidd during pregnancy
  - Led to fatal HDFN
- As of 2012 there were only 13 reported cases of Anti-Jk3 related HDFN
  - Most were mild, rarely fatal
  - Not previously described in a Mennonite community
Back to the Patient

• Original panel: plasma reacted with all test cells – negative auto-control
  – Kidd antigens are essentially ubiquitous

• Sample sent to ARC reference lab for antibody identification
  – Serum initially reacted with all test cells
  • No reaction with Jk3 negative cells
Red Cell Phenotypes

• Mother
  - Type: O +
  - C +
  - Fya +
  - K1 +
  - c (little) –
  - E –
  - Jka –
  - Jkb –

• Father
  - Type: unknown
  - C –
  - Fya +
  - K1 –
  - c (little) +
  - E +
  - Jka +
  - Jkb +
Implications

• 100% chance of having a Kidd antigen
• At least a 50% chance of having E
  – HDFN may occur from anti-E alone, but usually mild
• RBCs needed for mother, baby once born, and possibly before delivery for intrauterine transfusion (IUT)
  – In a similar case in 2014 there were only 2 Jk3 negative frozen units available
Autologous Units

• The patient had 3 autologous frozen units at a private institution in PA.

• Couldn’t ship frozen to us due to their use of a closed system (APC 215).

• Good news: expiration date extended to 14 days from 24 hrs after thaw in closed system.

• Bad news: 2-3 days for thawed units to arrive at our blood bank.
Request at ARC

- Request placed with ARC for O positive Jka/Jkb/E/c (little) negative units
- For possible IUTs we preferred O neg units
  - Very difficult to find O neg/c (little) neg units
  - We settled for units that were O positive
Frozen Units

• 1 frozen unit was found in Hawaii
  – Personal escort was denied and the unit was flown to our ARC facility in St. Louis (2 hour drive away)
  – Expiration in 24 hours after thaw begins and several hours to get to us
Directed Donation by Family

- Tested siblings were all blood type A
- One sister was pregnant and not tested
- Father type A
- Mother type O
- None went in for antigen testing
ARC located one appropriate donor in CA willing to donate a fresh unit
  - This donor actually works for the ARC
• We planned to collect unit in January for February due date
Timing

<table>
<thead>
<tr>
<th>Unit Location</th>
<th>Status</th>
<th>Time To Arrive</th>
<th>Shelf Life Post Thaw</th>
</tr>
</thead>
<tbody>
<tr>
<td>3 autologous units in PA</td>
<td>Frozen</td>
<td>2-3 days</td>
<td>14 days</td>
</tr>
<tr>
<td>1 unit in St. Louis</td>
<td>Frozen</td>
<td>7 hours</td>
<td>24 hours</td>
</tr>
<tr>
<td>1 possible unit in CA</td>
<td>Fresh</td>
<td>1-2 days</td>
<td>42 days</td>
</tr>
</tbody>
</table>

- Fresh units (≤ 7 days old) preferred for fetal/neonate transfusions
  - When to collect?
Complications

• Normally for IUT we wash units to increase HCT to 80%
  – We would not do this because then shelf life drops to 24 hours
• Irradiation
  – Required for units going to baby, but not mom
  – Shortens shelf life to 28 days
  – ARC irradiates units for us or emergency protocol in radiology
    • We chose to use our protocol when needed
More Problems

• In December, fetal ultrasounds became concerning for fetal anemia

• 12/18/15 fetal US PSV: 54.3 cm/s, 1.24 MoM
• 12/23/15 fetal US PSV: 65 cm/s, 1.43 MoM
• 12/30/15 fetal US PSV: 78.4 cm/s, 1.65 MoM

• On 12/23 we requested the fresh unit be collected to have on site for an emergency/possible urgent intrauterine transfusion
Birth

- OB/GYN elected for C-Section on 12/31 at 34 weeks due to more evidence of fetal anemia
- By then, we had 1 fresh unit on site
- Physician was uncomfortable with only 1 unit
  - We requested 1 autologous unit from PA 12/30
  - Arrived in 1 day, just before procedure
- Patient elected to have a tubal ligation
Outcome

• Procedure performed without transfusion
• Live male infant (2450 g, APGAR 6 at 1 min, 8 at 5 min) born with resuscitation performed by pediatrics
• Day of life 2, started on 2 banks of phototherapy (total bilirubin 7.9 mg/dL)
  – Discontinued 1 bank on DOL 5
  – Stopped on DOL 6 (total bilirubin 6.1)
• Autologous unit discarded
• Fresh unit released into general inventory
  – Fresh unit had passed the cutoff for freezing
Baby Screening

• Blood type: O positive
• DAT positive for IgG
• Pan-reactive eluate
• Cord blood cells type as E antigen positive
  – Presence/contribution to hemolytic anemia of anti-E undetermined due to pan-reactivity
Summary

• Very few reported cases of anti-Jk3 in pregnancy
• Most have been mild, requiring phototherapy
• This is another case supporting this trend
• Many facets of care to consider
  – Communication is key
  – With clinicians, patients, blood bank personnel, blood collection agency, etc.
Thank You!
References


