Challenges in the Management of a Patient with anti-JMH

Kylie Rushford
Monash Health
Monash Medical Centre
Case Presentation – Mr JP

- Mr JP
- Date of Birth 25-Nov-1930
- Presented to ENT with hoarse voice
- Left vocal cord paralysis
- CT scan of head, neck and chest
- Identified a large aneurysm arising from the aortic arch
Mr JP – Vascular Team

- CT angiogram shows a large 6.7 cm saccular aortic arch pseudoaneurysm arising from the aortic arch
- Quite close to the supra-aortic trunk
- Requires an angiogram to plan surgery, and then a graft
Pre Operative Bloods

- Seen in pre anaesthetic clinic
  - FBE
  - Coagulation profile
  - UEC
  - LFT
  - Group and Hold
Group and Hold

• O Rh(D) Negative

• AutoVue Screen (0 - 4 scoring system)
  • SC I  2
  • SC II  3
  • SC III 2
## Panel Results

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*Monash Health*
## Panel Results - CSL Panel A

### Panel A

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Different Techniques

• Tube screen using anti-IgG, no LISS
  • SC I 0.5 / 0.5 / 2
  • SC II 0.5 / 0.5 / 2
  • SC III 0.5 / 0.5 / 2

• DAT BioVue Tube
  • Polyspecific 0.5 1
  • IgG 0.5 1
  • C3b,C3d 0 0
## Panel Results - CSL Panel C

### Panel C

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## More Cells - Phenocell C

### Phenocell C 0.8% Antigen Composition Sheet

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**Batch No:** 9653 064

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Monash Health
Extra Testing

- DiaCidel (acid-glycine) eluate non-reactive

- 3 x Autoabsorption (RAMPEG), 3% Panel B

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- Transfusion Record (August 1997)
  - O Rh(D) Negative
  - No antibodies detected
### Phenotyping

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More Testing

- Papain IAT screen
  - SC I 0
  - SC II 0
  - SC III 0
Communication with Vascular Team

- No blood available
- Procedure cancelled

- Sample sent to the Reference Red Cell Serology Laboratory at the Blood Service
First Blood Service Report

- Anti-JMH detected by PEG-IAT and BioRad LISS/Coombs cards
- Titre of 16
- Does not react by enzyme-IAT

- Some reactions at saline 22°C and with enzyme
- ? Suggestive of a cold reactive antibody
- Anti-JMH does not cause haemolytic transfusion reactions
First Blood Service Report

- Group O Rh(D) Negative cde (rr)
- K-k+, Kp(a-b+), Fy(a-b+), Jk(a+b+), M+N+, S+s+, Lu(a-b+), Co(b-), JMH-
Second Blood Service Report

• Anti-JMH detected by PEG-IAT and BioRad LISS/Coombs cards

• No other antibodies present

• Give O Rh(D) Negative cde/cde K Neg IAT crossmatch compatible or “least incompatible”. Consider giving Fy(a) Neg blood.
Blood Group Systems

- **Glycophorins, Kell**
  - A to D
  - Lutheran, LW, Knops, Indian, Vel

- **RhD, RhCcEe, RhAG, Kidd, Diego, Colton, Gill, Kx, RAPH, Junior, Lan**

- **Duffy**

- **GPI anchored**
  - Type 5

- **Polytopic (multi-pass)**
  - Type 3
Semaphorin 7A

Neural and immune functions

### JMH System

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Anti-JMH

- Anti-JMH is a rare antibody.
- JMH Negative phenotype is usually acquired rather than inherited.
- No nucleotide changes in SEMA7A gene in the acquired type.
- Sema7A deficiency on red cells due to a post-transcriptional mechanism

Will the Patient Haemolyse?

- Numerous cases where patients with anti-JMH have been transfused with JMH+ blood with no adverse events
- One case of an acute intravascular haemolytic transfusion reaction caused by anti-JMH
- Select “least incompatible” red cells

Laboratory Plan 1

• Remove anti-JMH by phenotype matched RAMPEG absorption to ensure that there are no underlying allo-antibodies

• Cross match O Rh(D) Negative rr K Neg Fy(a) Neg units with the adsorbed plasma

• Plasma adsorbed using R_1R_1, R_2R_2 and rr cells - no change in the reactivity of the anti-JMH.
Laboratory Plan 2

• Treat panel cells and donor cells with
  • Papain
    • Destroys Fy\textsuperscript{a}, Fy\textsuperscript{b}, M, N, S, s, Xg\textsuperscript{a}, Ch, Rg, Ina, Inb, En\textsuperscript{a}, Pr, and JMH
  • DTT
    • Destroys Kell system, Knops, Dombrock, Lutheran, Cartwright, LW\textsuperscript{a} and JMH
Patient Blood Management

Optimise Red Cell Mass

- Pre op
  - Detect/treat anaemia
  - Treat sub optimal iron stores
  - Treat other haematinic deficiencies
  - Cease medications

- Intra op
  - Time surgery with optimisation of Hb

- Post Op
  - Manage anaemia
  - Manage medications
Patient Blood Management

Minimise blood loss and bleeding

• Pre op
  • Identify/treat bleeding risk
  • Minimise phlebotomy
• Intra op
  • Meticulous haemostasis/surgical technique
  • Cell salvage techniques
  • Avoid coagulopathy
  • Patient positioning/warming
  • Pharmacological agents
• Post Op
  • Manage post op bleeding
  • Keep patient warm
Patient Blood Management

Harness and Optimise Physiological Reserve of Anaemia

- **Pre op**
  - Estimate tolerance of blood loss
  - Patient’s bleeding history
- **Intra op**
  - Optimise ventilation and oxygenation
- **Post Op**
  - Maximise oxygen delivery
  - Treat infections promptly
  - Restrictive transfusion strategies
Patient Blood Management Plan

- Document baseline haemolytic parameters
- Monitor parameters if the patient is transfused
- Notify Haematology Registrar if the patient is transfused.
- Anti-JMH still present
- Papain and DTT testing showed no other underlying alloantibodies
- Crossmatched units available
- All ready to go!!
What Happened

• Surgery cancelled
• Re scheduled to some time later

• Surgery cancelled
• Lost the computer link between Monash Health and Dandenong Hospital
• Re scheduled
What Happened - Third Time Lucky

- Endoluminal thoracic aneurysm graft successfully performed
- Post op patient admitted to ICU for monitoring
- Haemodynamically stable
- Hb went from 136 to 109 g/L post op
- Back to 120 g/L by discharge (12 days post op)
Acknowledgments

- Michael Parker
- Sarah Hotchin
- Jennifer Condon
- Norah Lee
- Erica Wood
Any Questions?