Case Studies

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Case 2
Case 2 Background

- Young female patient with HbSC
- 36/40 gestation
- Presented with fever, hypoxia, chest and bone pain
- Hb 9.5g/dL
- Previously transfused 4 months earlier
Is this most likely to be a sickle cell crisis? - Q

1. Yes

2. No

3. Not sure
Is this most likely to be a sickle cell crisis? - A

1. Yes
   - Green Bar

2. No
   - Red Bar

3. Not sure
   - Blue Bar
Treatment

- Diagnosis: sickle cell crisis
- Symptoms worsened and patient required emergency C/Section
- 2 unit transfusion post delivery
  - E-, K-
  - O D positive, R_{1}r (DCe/cde)
- Discharged Hb 10.1 g/dL
10 days later…

**Pre-delivery**
- Fever, hypoxia, pain (chest; bone)
- Negative antibody screen
- Hb 9.5g/dL
- Previously transfused 4 months earlier
- Hb 10.1g/dL on discharge

**10 days post delivery**
- Fever, hypoxia, pain (chest; bone)
- Negative antibody screen
- Hb 6.7g/dL
- Previously transfused 10 days earlier
Is this most likely to be a sickle cell crisis? - A

1. Yes

2. No

3. Not sure
Is this most likely to be a sickle cell crisis? - A

1. Yes  23%
2. No  40%
3. Not sure  37%
Exchange transfusion

Diagnosis:
sickle cell crisis

- Given 6 unit exchange transfusion, E-, K-
  - Hb ↑ from 6.7 to 9.4 g/dL
  - HbS level ↓ from 48% to 21%

24 hours post exchange

- Continuing fever, pain, dysnoea
- Hb ↓ 7.1g/dL
- Spherocytes noted on blood film
Is this most likely to be a sickle cell crisis? - Q

1. Yes

2. No

3. Not sure
Is this most likely to be a sickle cell crisis? - A

1. Yes
2. No
3. Not sure
Finally…

- Diagnosis: Sickle cell crisis
- Following day Hb ↓ 5.2g/dL

- Clinical review 7 days later

? Transfusion Reaction
Transfusion reaction investigation results

Pre-exchange sample
- Antibody screen confirmed negative
- DAT positive
  - IgG + C3
- Anti-Jk<sup>a</sup> detected in an eluate

Post-exchange sample
- Antibody screen confirmed negative
- DAT positive
  - IgG + C3
- Anti-Jk<sup>a</sup> detected in an eluate

2/2 post delivery units were Jk(a+)
4/6 units for exchange were Jk(a+)
What was the diagnosis post-exchange - Q

1. Continuing sickle cell crisis?

2. Delayed HTR?

3. Acute HTR?

4. Something else
What was the diagnosis post-exchange - A

1. Continuing sickle cell crisis?  
   -12%

2. Delayed HTR?  
   -51%

3. Acute HTR?  
   -32%

4. Something else  
   -5%
SHOT report and outcome

- Reported to SHOT as acute HTR
- Probably delayed HTR on readmission
- Probably also acute HTR post exchange
- Probably also sickle cell crisis
Lessons

If a patient with sickle cell disease presents with symptoms of sickle cell crisis but a disproportionate fall in Hb 3 -14 days post transfusion consider a diagnosis of DHTR

Include a DAT and eluate as part of the investigation