# INSIGHTS FROM A CASE

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ST6 Haematology
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# BACKGROUND

28 year old Afro-Carribean female

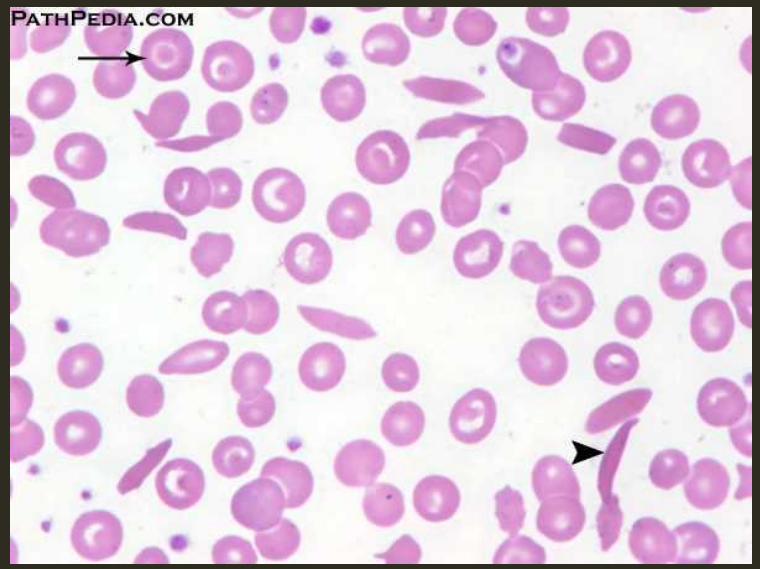
Hb SC disease

Came to UK aged 14 from Jamaica

Pains since the age of 8 years

Not diagnosed until age 18

Works as health care assistant



# ADMITTED AT 16:30

Presenting complaint:

- Severe pain in both knees not improving on co-codamol 30/500mg 2 tabs QDS and oramorph 10mg
- Systemically well with no respiratory complaints & no fevers
- Pain on and off for months since the birth of baby
  - 6 ½ months post partum
- Pain in knees, hips, ankles, shoulders
  - Several admissions to day unit for pain management
- Partial RCE 8 days ago – 3 units in ; 4 venesections

# CASE CONTINUED

## Past medical history

- Painful crisis 3-4 times per year
- Baseline Hb 90-110g/l
- Chronic splenomegaly
- Mild asthma
- Vit D deficiency
- 2 miscarriages – one age 18 and one ages 22 both at 12 weeks
- Emergency C-section on for failure to progress following planned induction of labour
  - Planned pregnancy
  - 38 weeks gestation
  - Estimated blood loss 500ml – not transfused

# TRANSFUSION HISTORY

Manual partial exchange done in pregnancy just prior to delivery

- 3 units given ; 4 venesections
- Poor venous access – lines required

None prior to that

A Rh D positive

- Extended red cell phenotype done

Antibody screen negative

# INITIAL ASSESSMENT

T 36.8

BP 140/80

HR 87

Sats 100% room air

RR 18

Examination unremarkable

No features of chest syndrome or infection

Hb 111
MCV 92.9
WCC 8.7
Plts 162
Neuts 5.6
Hb S 36.1 %  (50.3 %)

CRP 14  LDH 663
Na 138
K 4.3
Ur 4.1
Cr 65
Bili 34
ALP 79
ALT 49

# INITIAL MANAGEMENT

S/C morphine 5mg PRN

Regular paracetamol

Diclofenac with PPI cover

Thromboprophylaxis

IV fluids

Anti-emetics

Given pethidine by on-call SHO in evening

# DAY 2

Remains in pain – severe, worse on movement both knees, legs, feet

Remains systemically well

- Temp 37.2
- BP 145/90
- HR 78
- RR 14
- Sats 100% room air

Pain team contacted, IVI continued

- Switched to zomorph 30mg BD, oramorph 10-20mg PRN

# DAY 3

Seen 06:35am for chest pain and episode of haematuria

Appeared comfortable

- Temp 37.4
- BP 128/75
- HR 102
- RR 18
- Sats 92% room air

Plan: oxygen, ECG, monitor bloods mane, pain team r/v, monitor for further haematuria, given bisoprolol

# DAY 3

14:00-15:00

A bit drowsy, with pin-point pupils but appeared well

Given naloxone and zomorph reduced to 10mg BD

- Temp 37.1
- BP 128/70
- HR 138
- RR 18
- Sats 94% on 2L
- EWS 6

# DAY 3

18:53 reviewed re frank haematuria and EWS 11

- Temp 37.1
- BP 124/80
- HR 140
- RR 38
- Sats 100% on 4L

Extremely agitated

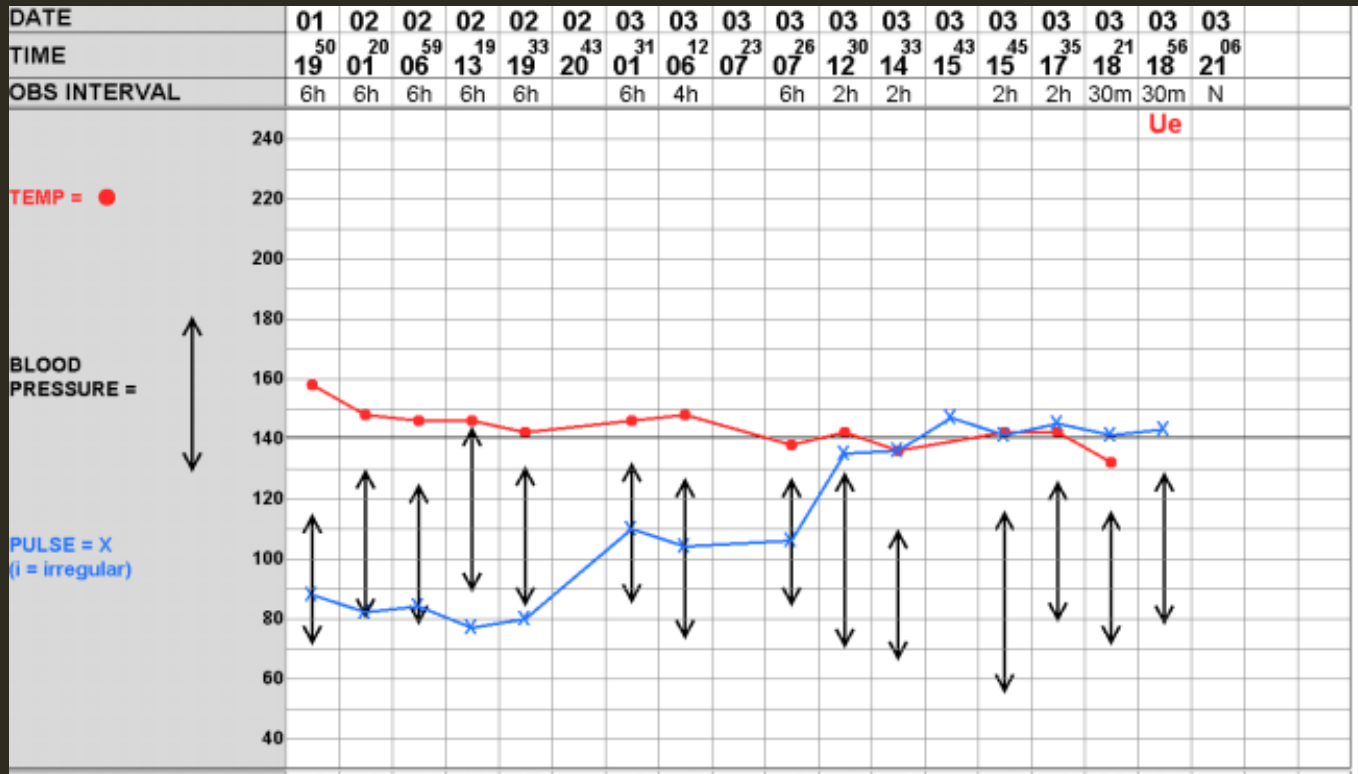
# DAYS 3 CONTINUED

Cardiac arrest call at 19:45

- Intubated promptly
- MHP activated
- 70 mins

Unfortunately unsuccessful

- Hb 24
- WCC 42.5
- Plts 90
- Na 145
- K 7.5
- Cr 194




| DATE | 01 | 02 | 02 | 02 | 02 | 02 | 03 | 03 | 03 | 03 | 03 | 03 | 03 | 03 | 03 | 03 | 03 |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| TIME | 19 50 | 01 20 | 06 59 | 13 19 | 19 33 | 20 43 | 01 31 | 06 12 | 07 23 | 07 26 | 12 30 | 14 33 | 15 43 | 15 45 | 17 35 | 18 21 | 18 56 | 21 06 |
| OBS INTERVAL | 6h | 6h | 6h | 6h | 6h | | 6h | 4h | | 6h | 2h | 2h | | 2h | 2h | 30m | 30m | N |
| Respiratory Rate | 18 | 16 | 16 | 14 | 17 | | 18 | 18 | | 18 | 18 | 18 | | 18 | 18 | 38 | 42 | |
| SaO2 (Oxygen Saturation) | 100 | 98 | 98 | 100 | 98 | | 96 | 92 | | 98 | 94 | 95 | | 94 | 94 | 100 | 99 | |
| FiO2 (Ltrs/Min or %) | Air | Air | Air | Air | Air | | Air | Air | | Air | 2.00L | Air | | 2.00L | 2.00L | 4.00L | 2.00L | |
| Pain Score (0=no pain-3=severe) | 1 | 0 | 0 | 0 | 2 | 1 | 0 | 2 | 0 | 0 | 1 | 1 | | 1 | 0 | 0 | 0 | |
| AVPU | A | A | A | A | A | | A | A | | A | A | A | | A | A | V | V | |
| Nausea / Vomiting | Y N | N N | N N | N N | N N | | N N | N N | | N N | N N | N N | | N N | N N | Y Y | N N | |
| Bowels opened (Bristol Stool Chart) | N | N | N | N | N | | N | N | | N | N | N | | N | N | N | N | |
| Urine (ml since last obs) | PU | PU | PU | PU | PU | | PU | PU | | PU | PU | NPU | | NPU | PU | 100 | NPU | |
| Weight (kg) | | | | | 69 | | | | | | | | | | | | | |
| VITALPAC EARLY WARNING SCORE (VIEWS) – Heart Rate | 0 | 0 | 0 | 0 | 0 | | 1 | 1 | | 1 | 3 | 3 | 3 | 3 | 3 | 3 | 3 | |
| Temperature | 0 | 0 | 0 | 0 | 0 | | 0 | 0 | | 0 | 0 | 0 | - | 0 | 0 | 0 | 0+ | |
| BP Systolic | 0 | 0 | 0 | 0 | 0 | | 0 | 0 | | 0 | 0 | 1 | - | 0 | 0 | 0 | 0 | |
| Respiratory Rate | 0 | 0 | 0 | 0 | 0 | | 0 | 0 | | 0 | 0 | 0 | - | 0 | 0 | 3 | 3 | |
| AVPU | 0 | 0 | 0 | 0 | 0 | | 0 | 0 | | 0 | 0 | 0 | - | 0 | 0 | 3 | 3 | |
| SATS | 0 | 0 | 0 | 0 | 0 | | 0 | 2 | | 0 | 1 | 1 | - | 1 | 1 | 0 | 0 | |
| O2 Therapy | 0 | 0 | 0 | 0 | 0 | | 0 | 0 | | 0 | 2 | 0 | - | 2 | 2 | 2 | 2 | |
| TOTAL EWS | 0 | 0 | 0 | 0 | 0 | | 1 | 3 | | 1 | 6 | 5 | - | 6 | 6 | 11 | 11+ | |

*Height*
**1m 54cm**
**02-Nov-2016**
**19:33**

*Weight*
**69kg**
**02-Nov-2016**
**19:33**

*Target weight*
**not set**

*BMI*
**29.1**

## KEY

### GENERAL
**OW** = Off ward

### UNMEASURABLE READINGS
**Uc** = Unmeasurable/ Patient condition
**Ue** = Unmeasurable/ Equip unavailable
**U** = Unmeasurable/ Other reason
**R** = Unmeasurable/ Patient refused

### AVPU
**A** = Alert
**V** = Voice
**P** = Pain
**U** = Unresponsive
**C** = Confusion

### GCS
**Best Eye open**

# DIFFERENTIAL DIAGNOSIS

Acute haemolysis
- Delayed haemolytic transfusion reaction / hyperhaemolysis

Acute sequestration
- Splenic / hepatic / pulmonary (chest syndrome)

Aplastic crisis

Acute blood loss

Sepsis

# POST-MORTEM

Cause of death

- Intravascular haemolysis
- Sickle cell anaemia

Haemoglobin staining of major arteries

Report of haematuria

# HYPERHAEMOLYSIS

A type of delayed haemolytic transfusion reaction

- Rare outside of that context

Typically seen 4-20 days post red cell transfusion

Destruction of transfused red cells and patients own red cells

SCD, Thalassaemia, myelofibrosis, MDS

# HYPERHAEMOLYSIS SYNDROME

## Clinical features

Pain that mimics VO crisis

Fever

(Jaundice)

Haemoglobinuria

## Laboratory features

Hb < pre-transfusion

Reticulocytopenia

↑LDH, Bil

↑Ferritin

DAT +/-

New alloantibody +/-

# LEARNING POINTS

Recognition of acutely unwell patient

- Not always so obvious when young and fit – compensate well

Delay in doing repeat bloods – not done on day 2 or 3 until she arrested (difficult veins)

Haematuria was not reported to seniors

- Educate patients on recognition & reporting of change in urine colour

Pain was very difficult to control

# LEARNING POINTS

## Transfusing patients with Sickle cell disease

- Only when absolutely necessary
  - Risk of alloimmunisation
  - Risk of rare but potentially fatal hyperhaemolysis
  - Hyper-viscosity

# ACKNOWLEDGEMENTS

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# REFERENCES

Win, Nay. Hyperhaemolysis syndrome in Sickle Cell disease. Expert Rev. Hematol. 2(2), 111-115 (2009)

# THANK YOU