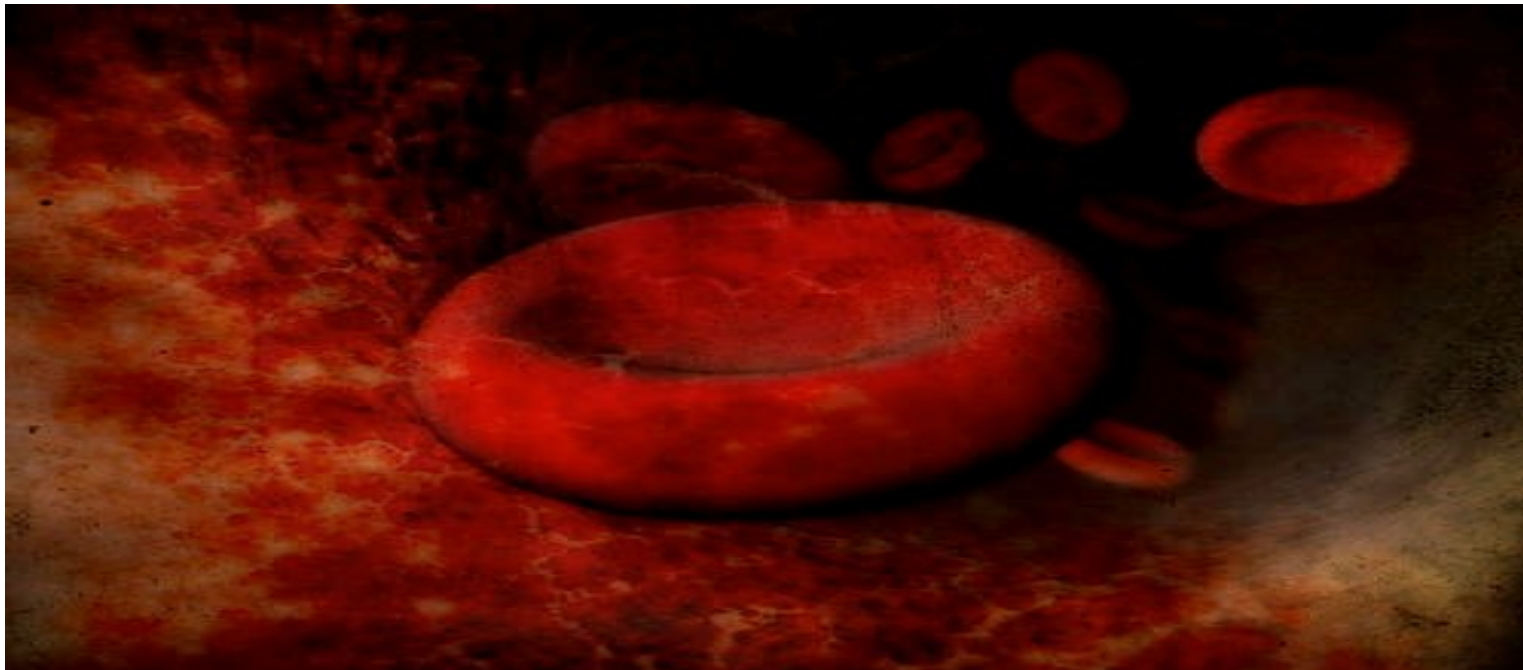


# The **BIG** Responder (revisited) Maureen McBrearty UCLH





**CAUTION**



**ON FIRST INVESTIGATION A DEGREE  
OF SPECULATION/ CONJECTURE  
APPLIED IN INTERPRETATION OF  
SOME FACTS & THEREFORE  
CONCLUSIONS REACHED(\*\*)**

# DISCLAIMER

Work in Progress!



Thus:

Theorem := I think I can prove it

Conjecture := Wishful thinking

Speculation := Nonsense

# It started with a phone call



- March 2010
- 32yr old female HbSS.
- Post Transfusion Jaundice Most Recent Tx. (1 month previously- Why no contact until now?)
- For Surgery in few months, may need pre-op exchange.
- For Molecular Genotyping at IBGRL

# UCLH Transfusion History-1



- **Group:** O ROr (cDe/cde) AST Negative
  - **Flag/Comment on LIS:** to receive O rr/ROr K- HbS- blood
  - **03/01/2007:** 2 units PRBC: - ?Ectopic pregnancy
- (**\*\*1** diagnosis confirmed? **\*\*2** Pro-active enquiry in old LIS & **\*\*3**? What Clinical Info on request form)

# UCLH Transfusion History-2



- **14/01/2008:** O Pos AST Negative
- **24/03/2008:** O Pos AST: +ve- Anti E- probably formed after 3/1/2007 – (Due to tx or ectopic??)
- **3/07/2008:** 2 units PRBC AST Neg
- **22/02/2010:** 2 units PRBC AST Neg , **POST TRANSFUSION JAUNDICE** (!?)

# Haem/Chem Results



	21/2/2010 (pre Tx)	24/2/2010 (post Tx)	24/3/2010
Hb (g/dl)	6.6	9.2	8.4
Retics ( $\times 10^9/l$ )	N/A	482	385
Bilirubin ( $\mu\text{mol/L}$ )	24	31	40
Potassium ( $\text{mmol/L}$ )	3.7	3.6	3.8
LDH ( $\text{iu/L}$ )	750	818	n/a

# NHSBT Investigation Results



- (a) Anti E detected by IAT**
- (b) Anti Cw detected by IAT**
- (c) Anti M by detected IAT**
- (d) Anti Jkb detected by IAT**



# Another Phone Call



- “ These don’t look too bad, we would be able to get blood with no real problem”
- “Well.....why do you want to transfuse”
- Red Cell MDT discussion...

# Patient History

32 yr old female HbSS, frequent crises, although not required much transfusion

Weight 125.4kg

Height 1.67m therefore ideal weight= 65kg

BMI= 45

Wants Bariatric Surgery.

First seen in bariatric clinic: **10/12/2008**.

Depression in past, refused treatment.

Seen in Haematology Clinic **11/12/2008**, no documentation or discussion on subject

# Patient History

**17/6/2009:** Psychological Assessment Not yet suitable for surgery.

**7/7/2009:** Admitted PV Bleeding.

**20/7/2009:** Referred to urologist for recurrent UTIs – still on going.

**7/10/2009:** Bariatric MDT – proceed with surgery, “will need Haematology Review Prior to surgery in view of sickle cell disease” (nss)

# Patient History

## **5/02/2010-Time of Original Presentation:**

- Numerous Ongoing Bariatric Clinic Consultations.
- One admission for painful Crisis.
- Several letters from Haematology Consultants.
- Plan if Surgery proceeds..

# Advice From NHSBT

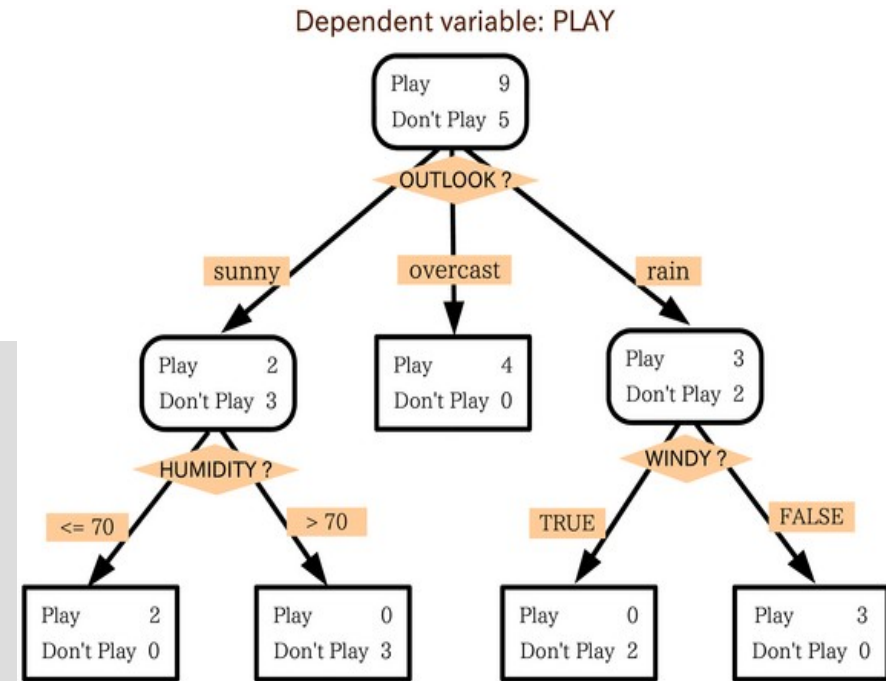


- Select E-, Cw-, M-, K-, Jkb- and S- (HbS -)
- 4 Weeks notice requested.
- Samples for genotyping.
- “Although the patient seems to be a high responder, it is difficult to predict whether she may form other additional antibodies”

# THE DECISION

Bariatric MDT:  
Medical Pathway if  
Possible –

“if she forms more antibodies then it may jeopardise her future ability to be transfused”



# Revisited-

- **03/01/2007** Ectopic Pregnancy  
Confirmed **BUT** 1 unit transfused =  
E+
- **14/01/2008:** O Pos AST Negative
- **24/03/2008:** O Pos AST: +ve- Anti E

# Revisited

- **3/07/2008:** O Pos AST Neg:
- 2 units PRBC both units rr, *M+*, 1 *Jkb+*
- **22/02/2010:** O Pos AST Neg:
- 2 units PRBC both units rr, 1 *M+* , 1 *Jkb+*.
- ***Post Transfusion Jaundice***
- **Not transfused since Feb 2010**



# Revisited

- **Phenotyping finally completed:**
- M- N+ S- s+ P1+ Lua- Lub+ K- k+ Kpa- Kpb+ Lea- Leb+ Fya- Fyb- Jka+ Jkb- Cw- Js(b+)
- Information on patient status/diagnosis July 2008
- **Not transfused since Feb 2010**

Thank You