

A case presentation – Haemoglobinopathies & patients who refuse blood products

Dr Katie Randall
Consultant Haematologist
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Patient SS

- 56 year old lady
- HbS/B-thalassaemia

HAEMOGLOBIN ELECTROPHORESIS			
HbA2		NA	%
HbF		26.9	%
Hb	*	76	g/L
Rbc	*	2.83	$\times 10^{12}/l$
MCV		85.2	fl
MCH	*	26.9	pg
MCHC		315	g/L
HPLC-Hb.Electrophoresis shows		HbS and HbF	
Haemoglobin S		66.1	%
Hb Electrophoresis			
SS or S beta thalassaemia with high HbF (known on hydroxycarbamide)			

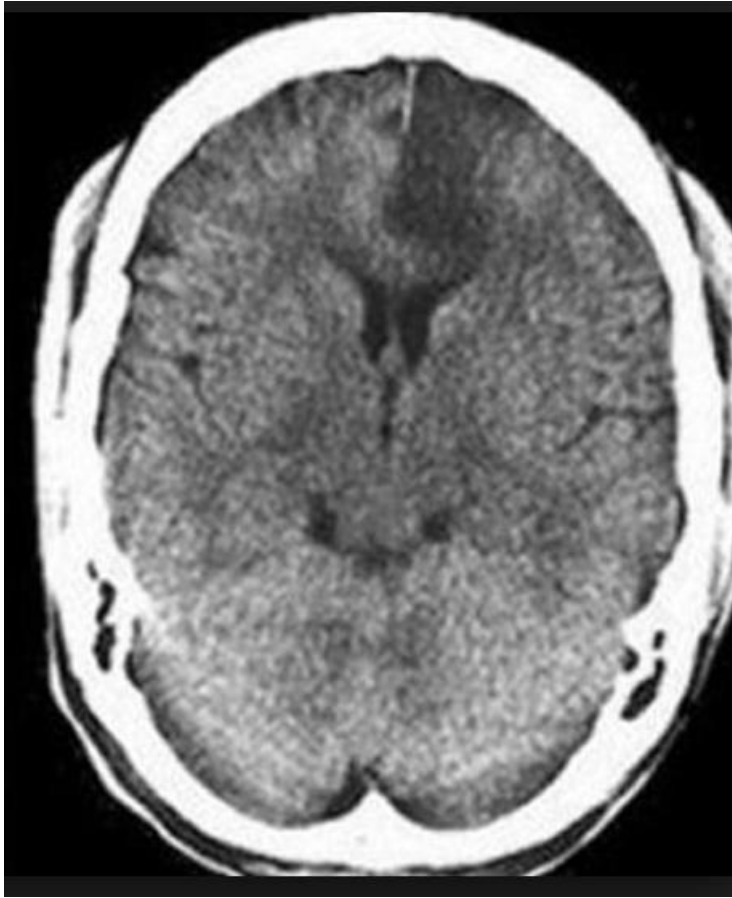
- Jehovahs witness

Patient SS

- Ulcerative colitis 1970s
– requiring surgery
- Pulmonary Embolisms
1998 & 2013 – on
rivaroxaban



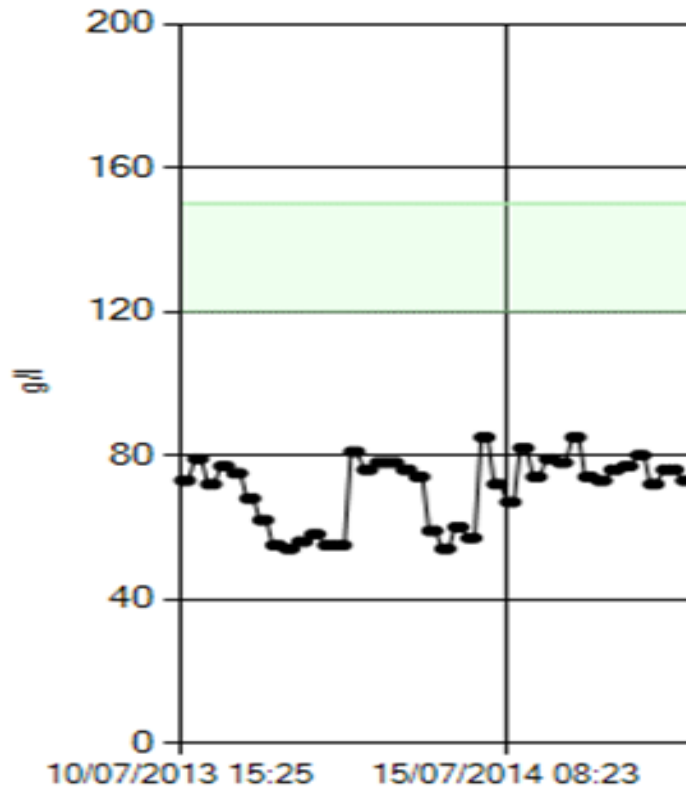
Patient SS



- 2007: Right frontal ischaemic stroke
- Declined blood products
- Started on hydroxycarbamide

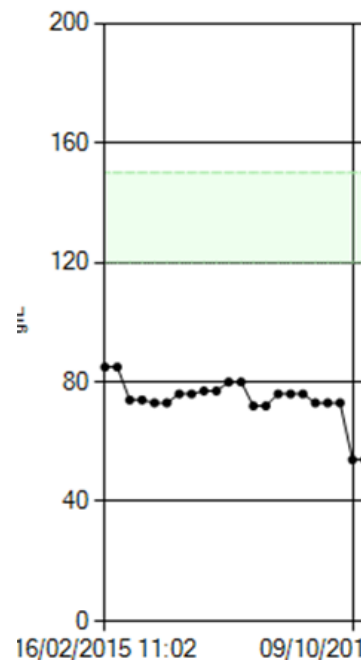
Patient SS

- Hb typically 70-80g/l



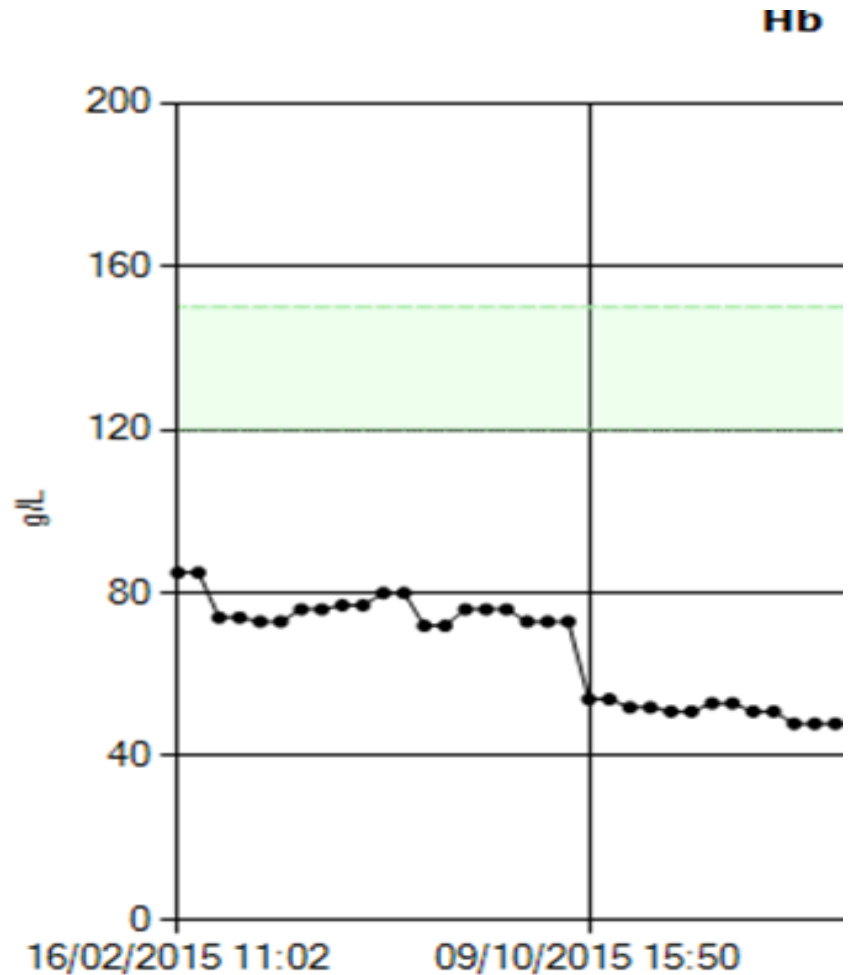
Patient SS

- Number of hospital admissions throughout 2015 with sickle painful crises
- Hb largely stable



Patient SS – Oct 2015

- Admitted with sickle painful crisis
- Typical presentation – pain affecting chest & arms
- Tired++, jaundiced
- Hb 43g/l



Patient SS – Oct 2015

- Hydroxycarbamide dose increased
- Started erythropoietin
 - darbepoietin 150mcg once weekly
- Seen by sickle team @ UHCW



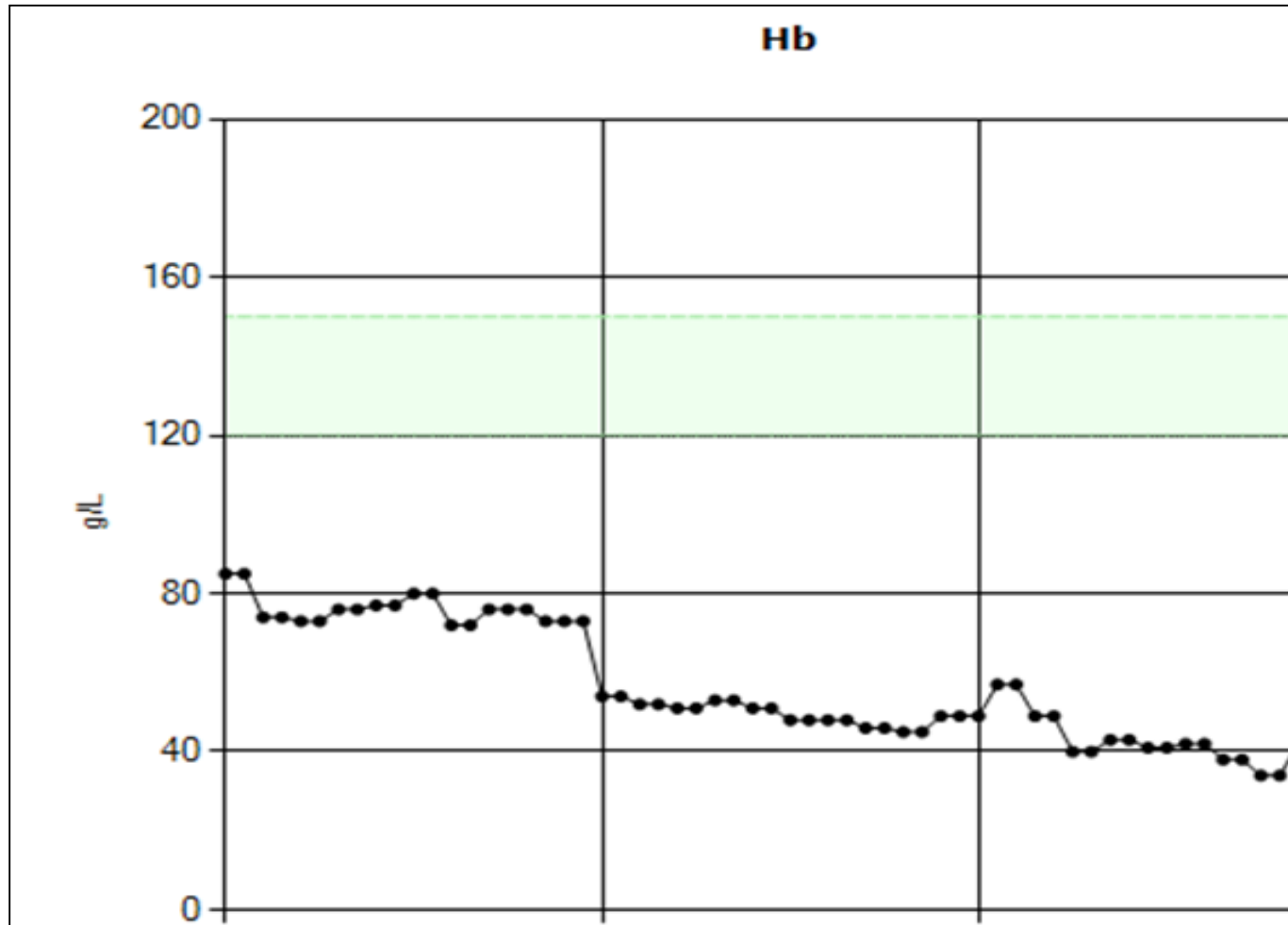
Patient SS – early Dec 2015

- Admitted acutely unwell
- SOB, chest pain
- Uncertain compliance with medication
- Hb 40g/l, platelets low
- Very weak & tired
- Discussed again re blood products including meeting with JW elders
 - Patient had a clear & valid advanced directive

Patient SS – Dec 2015

- Increasingly unwell over next few days
- Confined to bed
- Observations unstable
- Concerns over cardiac reserve
- Reviewed by intensive care team
- Lengthy discussions re blood products, resuscitation

Hb nadir – 32g/l



What did we do?

- Stopped hydroxycarbamide:
 - ? Contributing to low Hb
- Bed rest
- Minimised blood letting:
 - Paediatric bottles
 - Limited frequency of testing
- Ferritin 89 → Iron replacement
- B12 (already on folic acid)
- Increased darbepoetin
- Followed 'patients who refuse blood products' policy

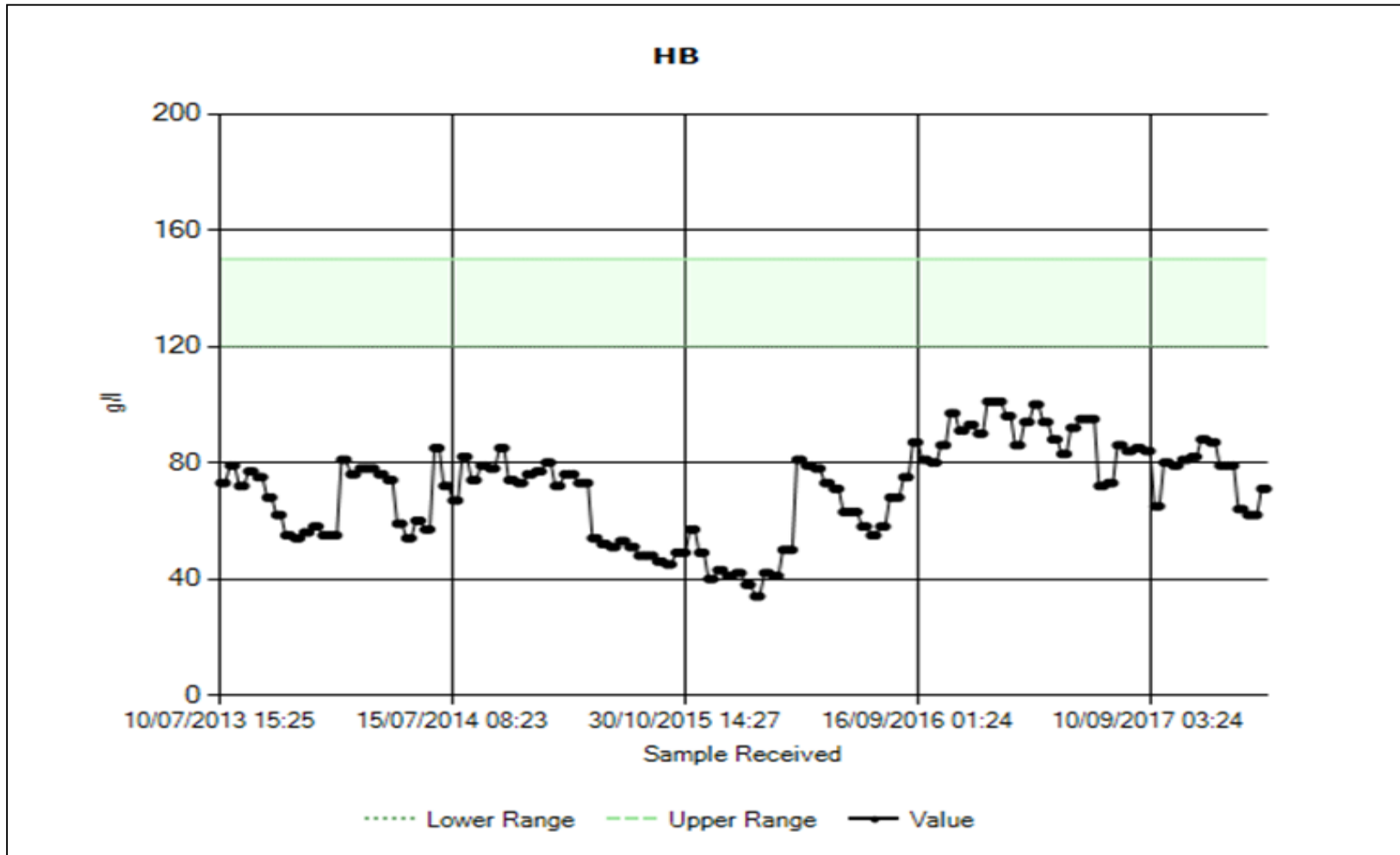
Patient SS

Date	Haemoglobin
3/12/15	40
6/12/15	38
10/12/15	32
14/12/15	41
20/12/15	50
10/01/16	68
22/01/16	81

Patient SS - progress

- Back on hydroxycarbamide (lower dose)
- Continues on erythropoietin
- Hb largely maintained
 - Issues with compliance

Patient SS - progress



Jehovah's witnesses

- Levitus 17:10

“As for any man...who eats any sort of blood...I shall indeed cut him off from among his people”

- Believed to be an act of blood eating that excludes the offender from eternal life in heaven

Jehovah's witnesses

- 7.5million active members worldwide:
 - Estimated 130,000 in the UK
- Individuals can vary in their choice of products would/wouldn't accept

NOT ACCEPTABLE (‘PRIMARY COMPONENTS OF BLOOD’)	IS ACCEPTABLE	MAY OR MAY NOT BE ACCEPTABLE (‘MATTERS OF CONSCIENCE’)
Whole blood (an autologous predonation)	Crystalloids	Albumin
Red cells	Synthetic colloids, e.g. dextran	Immunoglobulin
Platelets	Hydroxyethyl starch (Hexastarch)	Vaccines
White cells	Gelatines (Haemacel, Gelofusine)	Coagulation factors (non- recombinant)
Plasma (FFP)	Recombinant products e.g. G-CSF, † Epo, Coagulation Factors	Haemodilution
		Intraoperative cell salvage
		Organ transplantation

- Close liaison with patient and documentation around decisions
- Utilise JW Hospital liaison teams
- Employ principles of patient blood management

“Many of the techniques developed for use in Jehovah’s Witness patients will become standard practice in years to come in an effort to conserve blood stocks and reduce the need for transfusion”.

- Royal College of Anaesthetist’s Continuing Medical Education Journal
- Management of Anaesthesia for Jehovah’s Witnesses 2nd Edition, Concluding remarks, 2005

Thank you....any questions?

