Case Study: When Hemoglobinopathies Collide

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Objectives

- Discuss Jsb antigen/antibody
- Discuss challenges associated with antibodies to high incidence antigens
- Discuss Hemoglobin C trait and disease

Admission

- 21 y/o male with Sickle Cell Anemia (Hgb SS) presents to KU ER c/o chest and hip pain.
- Patient is admitted--suspected Acute Chest Syndrome.
- Admitting hemoglobin value was 10.9 g/dL. No blood needed…**YET…**
- History check: "*Sickle Patient*," only has one antibody. No big deal?

Uh oh…

- …the antibody is Js^b

Js^B Antigen frequency

Phenotype	White	Black
Js(a-b+)	100%	80%
Js(a+b+)	Rare	19%
Js(a+b-)	0%	1%

Antibodies to Js^B

- Kell system
- Clinically significant
- Known to cause severe HDFN
- Also may lead to acute or delayed HTR

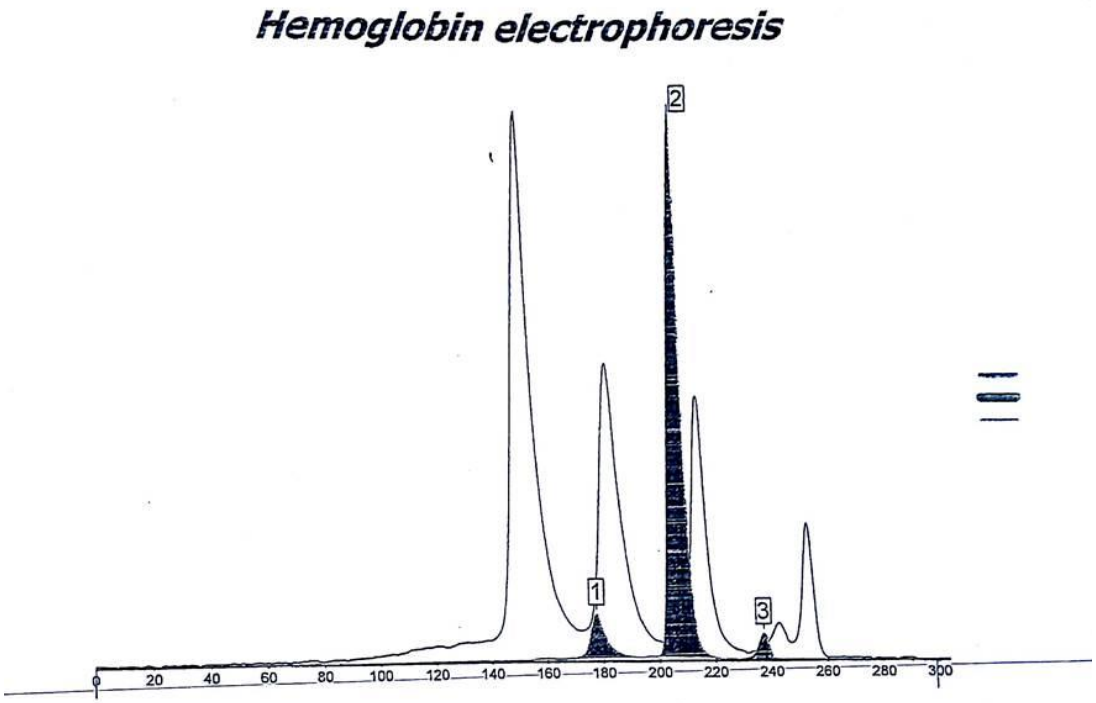
Patient Needs Blood…

	Admission	3 days post admit	*Reference Ranges*
Hemoglobin	10.9	6.8	*13.5-16.5 g/dL*
Hematocrit	31.0	19.1	*40-50%*
Total Bilirubin	2.8	12.7	*0.3-1.2 mg/dL*

Preparing for RBC Exchange

- RBC exchange ordered
- KU policy: In addition to matching for antibodies and providing Sickledex negative units, match for C, c, E, e, K, Fya in sickle cell disease population.
- Try to fully match this patient- don't want more antibodies
- Need units of E, K, Jsb, Fya, Fyb, Jkb negative, Sickledex negative pRBCs…STAT.

Pre Exchange Hemoglobin Electrophoresis

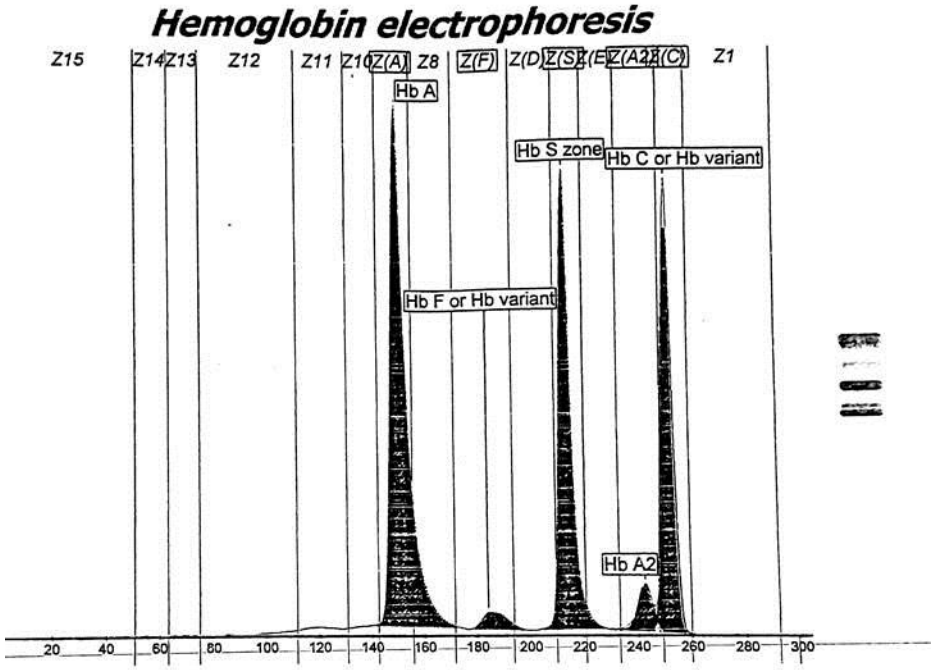


Fractions	%
1	8.4
2	88.3
3	3.3

RBC Exchange

- 11 units ordered (high number due to deglycing)
- All 11 units were found frozen in KC!
  - One broke while thawing
- Exchange goes *SMOOTHLY*

Post Exchange Hemoglobin Electrophoresis



Fractions	%
Hb A	43.8
Hb F or Hb variant	2.2
Hb S zone	28.9
Hb A2	3.2
Hb C or Hb variant	21.9

Wait…What Happened?

- First thoughts: Clerical error? Specimen mix up?
  - Specimen verified-correct specimen, no clerical errors
- Second thoughts: Exchange related?
  - Let's call up our blood supplier

Post Exchange Followup

- Call the blood supplier!
- All units are from ONE donor.
- Donor is most likely an asymptomatic carrier- still allowed to donate!

Hemoglobin C

- 1 in 40 African Americans carry the gene for Hgb C trait
- Hemoglobin C trait:
  - Generally asymptomatic
  - No treatment needed
- Hemoglobin C disease:
  - Mild, hemolytic anemia
  - Occasional mild jaundice, joint pain, splenomegaly, gallstones
- Hemoglobin SC:
  - Not a concern for this patient
  - Hbg C does not deform as deform the red cell as severely as Hgb S
  - Fewer vaso-occlusive crises
  - Some complications more severe: retinopathy, osteonecrosis, priapism

Patient Follow-up

- Patient discharged a few days later
- Follow-up visits at outpatient clinic are unremarkable
- Seems to be back at his baseline

References

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