“You’re stuck with me, baby”…

Hereditary Persistence of Fetal Hemoglobin

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Patient History

- 29 y/o Caucasian female.
- G2P1 approximately 8 months pregnant.
- presents to hospital with complete placenta previa and actively bleeding.
- No history of blood transfusions.
- Blood type O Rh negative.
- First pregnancy: C section delivery of O Rh Pos baby girl. No complications.
- Patient received 1 prenatal and 1 antenatal RHIG with first pregnancy.

Laboratory Results

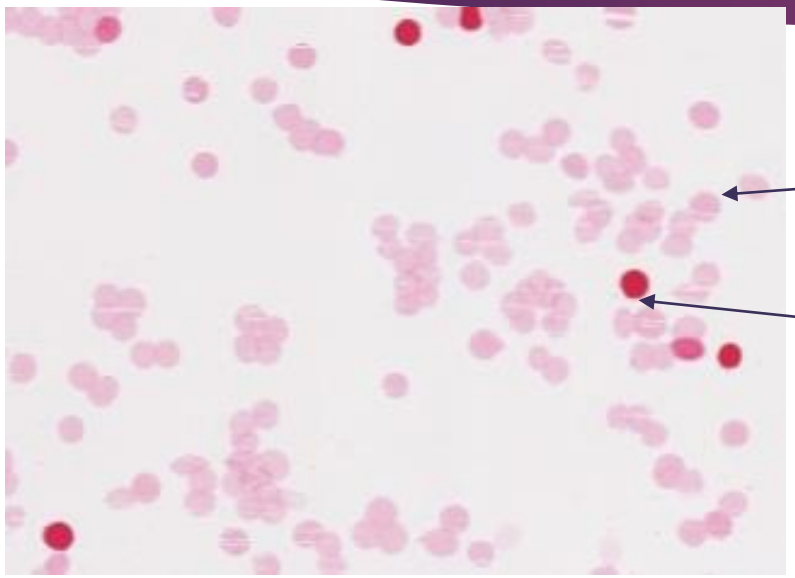
- **ABORh : O Rh negative**
- **Antibody Screen: Negative**
- **CBC: nothing remarkable**
- **Kleihauer Betke: Positive**

Procedure	Result	C	Units
Instr WBC	11.7		x10'3/microL
WBC	11.7 H		x10'3/microL
RBC	3.64 L		x10'6/microL
HGB	12.1		g/dL
HCT	33 L		%
MCV	92		fL
MCH	33		pg
MCHC	36		g/dL
RDW	11.8		%
RDW SD	39.8		fL
Platelet	283		x10'3/microL
MPV	9.2		fL
Neutro Auto	70		%
Lymph Auto	22		%
Mono Auto	6		%
Eos Auto	1		%
Basophil Auto	0		%
Imm Gran Auto	1 H		%
Neutro Absolute	8.1 H		x10'3/microL
Lymph Absolute	2.6		x10'3/microL
Mono Absolute	0.7 H		x10'3/microL
Eos Absolute	0.1		x10'3/microL
Basophil Absolute	0.0		x10'3/microL
Imm Gran Absolute	0.1 H		x10'3/microL

Kleihauer Betke Principle

- A quantitative test performed using mother's blood to detect a fetal maternal hemorrhage.
- Adult hemoglobin (HbA)dissolves out of the cells making them appear as ghost cells or "negative"
- Because it is acid resistant, Fetal hemoglobin (HbF) retains the Erythrosin stain making them a dark reddish pink or "positive".

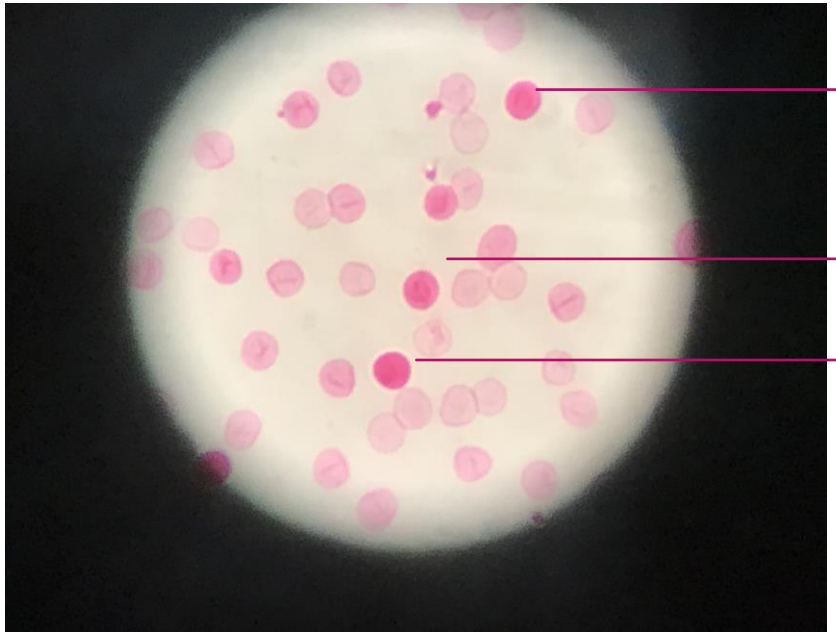
Interpretation of Kleihauer Results



Indications For Kleihauer Testing

- To quantitate Rhig necessary to protect a Rh negative mother against Rh positive immunization from fetal maternal bleed.
- Used as a follow up for positive qualitative fetal screening “rosette” test.
- For mother’s who’s babies are weak D positive (DU). “rosette” tests may not detect the weakly reacting Rh positive cells if they are present.
- To determine if there is a presence of fetal maternal hemorrhage from trauma in an Rh negative OR positive mother.

Back to our mom's first Kleihauer Betke result….



Doing the math…. # fetal cells/total cells counted

15/1018= 1.4% fetal cells

1.4 x50 *= 70 mL fetal bleed

* 5000mL is mother's arbitrarily assigned blood volume

How much rhig ?

3 vials administered...

Mom's second admission 1 month later…

Due to bleeding, mom returns to hospital.

Ultra sound is performed.

Per notes: placenta previa is still noted and baby appears to be progressing.

Kleihauer Betke is ordered.

Result is positive. 95mL fetal bleed is detected.

How much rhig?

4 vials Administered..

3 weeks later

Delivery day!

Mom arrives for scheduled c section. Type and crossmatch for 2 units of RBC's and a baby O neg CPDA irradiated RBC is on hold as well.

A healthy O Rh positive baby girl is born! (with a 3+ DAT using gel method).

Per policy: a screen for fetal maternal bleed is performed on all Rh negative mother's who give birth to a Rh positive baby

A qualitative "rosette" test was performed and found to be negative.

Where's The Fetal Cells?

The detective work….

Although a "rosette" test does not detect fetal cells, it *is* designed to detect Rh positive red blood cells.

***If* the baby had a bleed *and* is Rh positive, we should have a positive screening test.**

Therefore….

The 2 previous positive Kleihaur Betke results do *not* correlate with the negative screening test..

One more Kleihaur Betke (not because they are fun)

We performed one more Kleihaur Betke test:

- Using the post partum sample collected for the "rosette" test.
- Multiple blood bankers viewed the slide and all agreed very positive!
- We also reviewed the previous 2 Kleihaur Betke stained slides to be sure we did not have a technical error..

ALL 3 Kleihaur Betke stains correlated: positive fetal cells

Time to send out…

We are suspecting there really ***isn't*** a fetal bleed at all… Is mom making her own fetal hemoglobin?

Per Sure-Tech manufacture's insert: *"Hemoglobin F may occur in relatively high concentrations in adult patients with certain inherited disorders of erythropoiesis such as thalassemia major and sickle cell anemia. In adults with hereditary persistence of fetal hemoglobin, the concentration of HbF is approximately 26% of total red blood cells in adults possessing these and other hemoglobin abnormalities."*

Flow cytometry testing is performed. The result is…..

28.12% of cells appear to be adult cells that express HbF.

*If clinically indicated, hemoglobin electrophoresis is recommended to detect hereditary persistence of fetal hemoglobin.

Confirmation.

Mom and baby were discharged.

7 months post partum, We asked the patient to return for Hemoglobin electrophoresis for our research purposes..

Results of the Hemoglobin electrophoresis
Hemoglobin F (Fetal Hb) 2.7%

Thank you to our friends at ARUP laboratories who supported this presentation.

ARUP LABORATORIES

Hereditary Persistence of Fetal Hemoglobin

- HPFH is a benign, rare, congenital condition affecting the synthesis of fetal hemoglobin.
- Fetal hemoglobin (HbF) is the primary hemoglobin formed in the fetus comprising of 95% of total hemoglobin until about 36 weeks gestation.
- At birth, a baby has approximately 50% HbF. Hemoglobin chains convert to adult hemoglobin HbA during the first year of the infants life.
- Individuals with HPFH, have a mutant gene that inhibits synthesis of hemoglobin A and A2 allowing for the continued production of HbF.

Sources

- Fetal Hemoglobin [package insert]. St Louis, MO; Sure-Tech Diagnostic Associates INC; 2016.
- Conley, C. L., Weatherall, D. J., Richardson, S. N., Shepard, M. L., & Charache, S. (1963). Hereditary Persistence of Fetal Hemoglobin: A Study of 79 Affected Persons in 15 Negro Families in Baltimore. Blood, 21(3), 261-281. Accessed April 02, 2018.