Transfusion Medicine Service support for Pregnant patients with Sickle Cell Anemia

A Review of Proactive Initiatives

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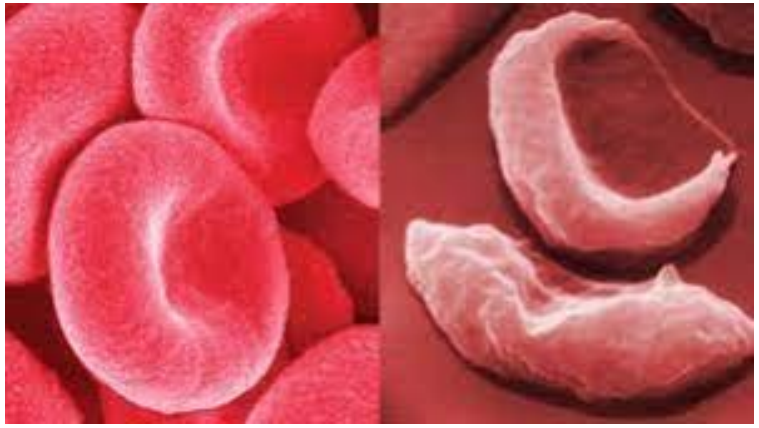


The Editors of Encyclopedia Britannica Sep 12, 2023https://www.britannica.com/science/sickle-cell-anemia#ref120899

Objective

To discuss proactive initiatives for pregnant sickle cell patients

- Patient history
- Routine specimen collection every 96 hrs
- RhD genotyping
- Full RBC genotype
- Transfusion support /preparedness

Notification

February 4th…

Diagnosis: Pregnant, Sickle cell disease

Patient History

"A full transfusion history should be taken from the patient at the booking appointment or first obstetric/hematology appointment and should include communication with the transfusion laboratory and national transfusion database to ensure there are no historical alloantibodies."

Oteng-Ntim E, Pavord S, Howard R, et al. 2021, 194:980-95.

Patient History

Direct questioning on manual and electronic forms has made it easier to draw out vital information:

“Previous transfusions?”

“Previous Rh immune globulin?”

“Patient has been transfused with red blood cells within the last three months?”

“Patient has been pregnant within the last three months?”

Reason for transfusion: Sickle cell disease

Historical record

Canadian Blood Services

Antibody identification/Phenotype/Genotype on record?

Anti-E

Phenotype on record (Rh, Kell, Duffy, Kidd)

No genotype report

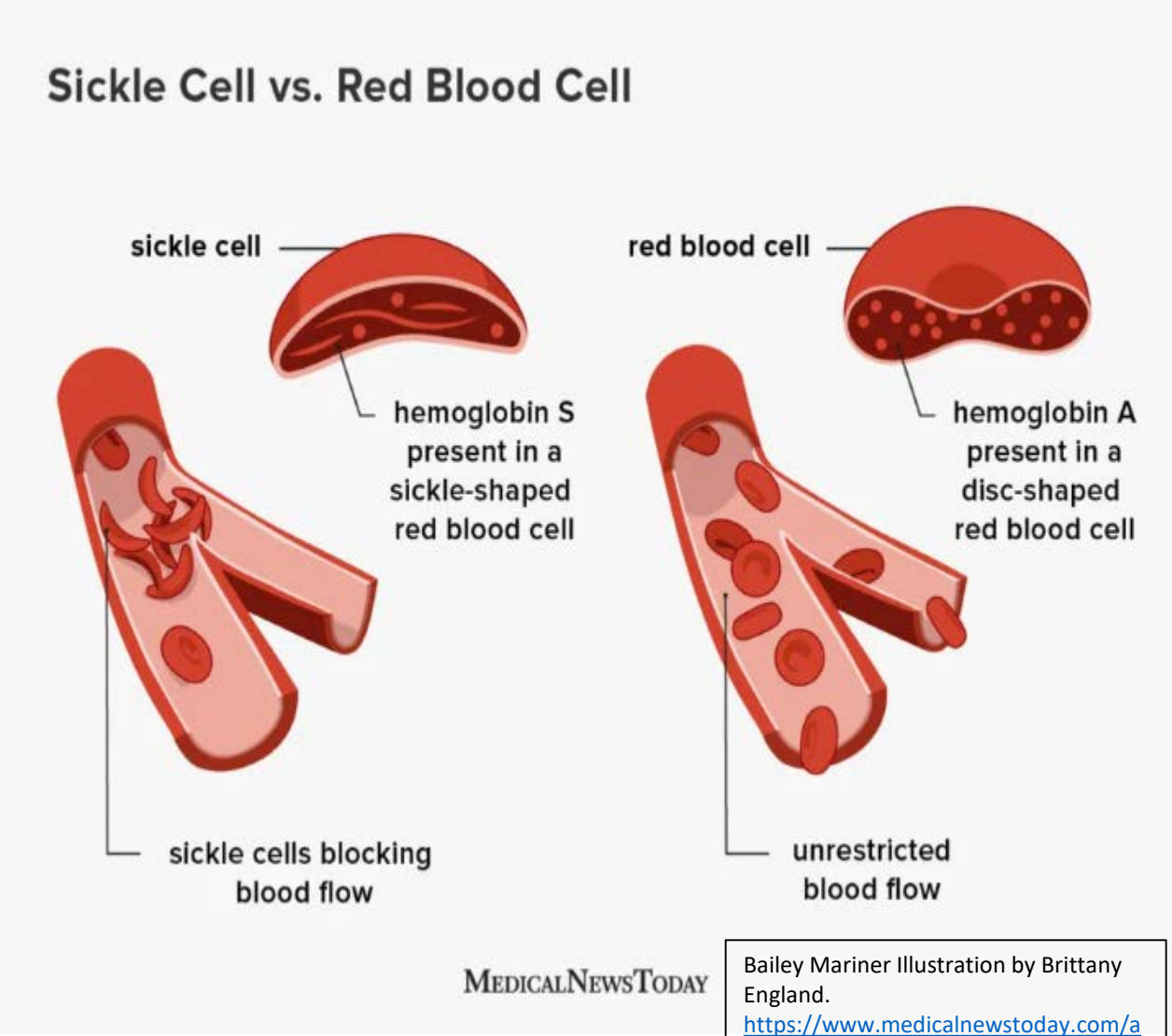
Sickle Cell Anemia and Pregnancy

In conditions of decreased oxygen availability, the hemoglobin S becomes rigid, rod-like, and fragile. The result is an abnormal hemoglobin which distorts the shape of red blood cells; the shape is described as a sickle, and is subject to increased hemolysis.

The abnormal shape of these red blood cells along with their increased adhesion results in physical blocking of small vasculature causing decreased oxygen delivery to vital organs.

The vasculature of the placenta may become obstructed resulting in adverse outcomes for mom and the fetus.

Shegekar T, Pajai S (June 30, 2023)
A Comprehensive Review of Pregnancy in Sickle Cell Disease.
Cureus 15(6): e41165.doi:10.7759/cureus.41165



Bailey Mariner Illustration by Brittany England.
https://www.medicalnewstoday.com/articles/sickle-cell-vs-normal-cell

Sickle Cell Anemia and Pregnancy

Terms used to describe the affects of pregnancy on patients with sickle cell disease are as follows:

Pregnancy

"…increased physical and psychological stress…"

"…dehydration…" [1]

"…worsening anemia…related [to] increased iron requirements and red cell turnover…which can precipitate a sickle crisis." [1]

"The physiological changes which take place in pregnancy…aggravate SCD leading to an increased risk of adverse effects for SCD mothers and neonates."

Delivery

"Birth, …is a profound physiological challenge…"

"the [increased] risk of dehydration, hypoxia, anemia, overexertion and significant metabolic derangement, which can all lead to a higher incidence of crisis."

Oteng-Ntim E, Pavord S, Howard R, et al.*Br J Haematol.* 2021, 194:980-95, Seaman CD, Yabes J, Li J, Moore CG, Ragni MV: *Thromb Res.* 2014, 134:1249-52.

March 19th (…over 1 month later)

Patient first registered as an outpatient

TMS receives their first specimen:

	Anti-A	Anti-B	Anti-D	D ctrl		A1 cell	B cell	ABO/Rh interp
Patient	4+	0	2+	0		0	4+	A

ABO/Rh : A ??? (anti-D : 2+)

Antibody screen: Negative

RhD genotyping

Rh D genotyping is used to classify a patient's weak D phenotype as that of an individual who can either form anti-D or not

Patients with weak D type 1, 2, or 3 cannot form anti-D and are registered as Rh D positive

Patients with Weak D types other than 1, 2, and 3 are registered as Rh D negative

RhD genotyping

Canadian Blood Services, Transfusion Medicine Report dated 2023-04-03:

Date Collected: 2023-03-19

Test Performed	Results:	Comment
RHD Genotyping	Weak D Type 1 (RHD*01W.1)	

Remarks:
RHD genotyping results indicate that this patient should be treated as an an RhD positive individual. Testing was performed using Immucor's RHD Molecular BeadChip Test which is an in vitro diagnostic test licensed by Health Canada.

RhD genotyping

In a proactive effort to differentiate underlying weak D phenotypes as either anti-D forming or not, Mount Sinai hospital submits sample for Rh D genotyping from all untested pregnant women in the sickle cell population regardless of their anti-D reaction strength.

RBC genotyping

Full RBC genotyping can provide additional insight into the donor RBC phenotype requirement of the patient:

(1) Antigen negative for high incidence antigens

(2) RhCE altered alleles

Dinardo, C., Kelly, S., Dezan, M., *et al.:Transfusion*. 2019 October ; 59(10): 3228–3235.

Sunday, March 28th (…2 weeks later)

Patient arrives in ER…

(1) TMS receives 2nd specimen… results consistent with historical record

(2) Upon review of the electronic order comments, the following details are revealed:

Transfused Red Cells in Last 3 Months : Yes

(3) A request for patient history is submitted to the hospital where the patient was last transfused. The patient history report received revealed…

L**ast transfused 2 units on March 22.**

Antenatal and Postnatal care

Patients with sickle cell disease often receive red blood cell transfusions

High risk of developing multiple alloantibodies

High risk of transfusion reaction

Transfusion considerations

Sickle Cell Disease Hemoglobin Values

Lab Test	Healthy adult	Normal SCD patient	Case study
Hemoglobin	120 – 150 g/L	60 – 110 g/L	75 g/L

Patient discharged March 29th

SCD.(2020, October 30).*Blood Tests for Sickle Cell Disease.* https://sickle-cell.com/blood-test (accessed September 4,2023)

June 25th (… 2 months later)

Blood product request for exchange transfusion

9 units

Diagnosis: Acute Chest Syndrome

https://www.inovablood.org/get-involved/host-a-blood-drive/steps-to-success/

RBC Transfusion Requirement for Patients with Sickle Cell Anemia

Without an antibody

Rh and Kell antigen matched units

Immediate spin and IAT/gel crossmatch compatible

With an antibody

Rh, Kell, Kidd, Duffy, MNS antigen matched units

Immediate spin and IAT/gel crossmatch compatible

Adult Exchange Transfusion

Unit centrifuged to separate red cells from supernatant

Most of the supernatant is removed

Final product: Hct = 0.85 L/L or 85% +/- 5

Deliver the least amount of blood with the highest oxygen carrying capacity in the shortest amount of time

Replace sickle cells with normal cells to improve blood circulation and therefore oxygen delivery

August 15th (…2 months later)

Delivery

Hgb = 69 g/L

Blood product request for 1 PRBC unit

Patient delivered baby boy

No transfusion required

https://unsplash.com/images/people/baby

August 16th

Hgb = 50 g/L (>24 hrs post delivery)

Received 1 units of PRBCs post delivery

In Summary…

Pregnant women with sickle cell disease are classified as high risk

Transfusion medicine service assists in best outcomes as it relates to…

Acquiring Patient history

Maintaining indate specimen

Providing safest blood (patient genotyping, providing antigen matched, crossmatch compatible PRBCs)

Thank You !