Community Blood Center

innovation • experience • expertise

Sickle Cell Disease and Delayed Hemolytic Transfusion Reactions

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Objectives

- Describe characteristics of sickle cell disease and delayed hemolytic transfusion reactions
- Describe the tests used to resolve the most complex serologic workups
- Apply recent research data to predict the risk of delayed hemolytic transfusion for transfusion of a sickle cell patient
- Select appropriate units for transfusion

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Sickle Cell Disease (SCD)

- Inherited group of red blood cell disorders
- Hemoglobin is abnormal
  - RBCs become hard and sticky
  - C-shaped
- Sickle cells die early
  - Shortage of RBCs
  - Stuck/clog small blood vessels

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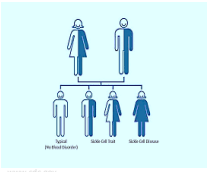
Inheritance



www.cdc.gov

HbSS
- 2 hemoglobin "S" genes
- More severe form

HbSC
- 1 hemoglobin "S" gene
- 1 hemoglobin "C" gene
- Milder form

HbAS
- 1 hemoglobin "S" gene
- 1 hemoglobin "A" gene
- Usually do not have any symptoms
- Can still pass to any children

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SCD Management and Treatment

- Treatment
  - Hydroxyurea + many new drugs are now available
  - Transfusion
- Transfusion
  - Decreases Hgb S levels
  - Reduces sickling
  - Prevents increase in blood viscosity
- Indication
  - Stroke prevention
    - Stroke is reduced to <10% if:
      - Hgb levels are 8-9 g/dL
      - Hgb S stays <30%

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Transfusion

pediatricsnationwide.org

- Chronic
  - Exchange transfusions every 4-6 weeks
  - Volume (# of units needed) determined by size of individual
- Occasional
  - Not monthly exchanges, only as needed
  - May just need 1 or 2 units transfused

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Risks

- Highest rates of red cell alloimmunization
  - Preferentially transfuse red cell antigen matched Rh and K- units
- Iron overload
  - Hepatic and cardiac dysfunction
- Delayed hemolytic transfusion reactions
  - May lead to hyperhemolysis

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Delayed Hemolytic Transfusion Reactions (DHTR)

- Induced by immunization against RBC antigens
- Favored by blood group polymorphism between donors and recipients
- Inflammation status may increase risk of alloimmunization

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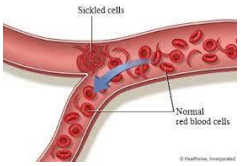
DHTR Presenting Symptoms

- DHTR in SCD patients is often hard to recognize/diagnose
  - Mimics vaso-occlusive crisis (VOC) symptoms
  - Alloantibodies may not be detectable yet
  - Additional transfusion may cause hyperhemolysis
    - Severe form of DHTR
    - Can lead to lethal multiple organ failure



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RESEARCH ARTICLE

WILEY AJH

Incidence and predictive score for delayed hemolytic transfusion reaction in adult patients with sickle cell disease

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Current Research Study

- Primary Aim
  - Single center observation study to determine the incidence of DHTR in the transfused sickle cell population
- Secondary Aim
  - Develop a score predicting DHTR for each transfusion episode

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Study Design

- Single-center observation study
- November 2011 – June 2014
- Adult SCD patients (>18)
- Pretransfusion analyses
  - ABO, Rh
  - Antibody screen / identification
  - Rh, Kell, Duffy, Kidd and MNS extended phenotype
- Each transfusion episode was recorded as an incidence
  - >8 days between transfusions
  - Led to larger transfusion episodes than patients enrolled

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Transfusion Protocols

Chronic (CTE) vs Occasional (OTE)	No history of antibodies	Known clinically significant antibodies	If patient typed C+
• Manual, automated, liquid, frozen, etc.	• Prophylactic Rh and K-matched RBCs	• Rh and K-matched and • Extended-matched RBCs (FY, JK, MNS)	• Genotyped for partial C • Given C-negative RBCs

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Diagnosis of DHTR

- Detection (24 hours to 25 days post transfusion)
  - Clinical declarative of patient/clinician-observed criteria
    - VOC
    - Dark urine
    - Onset or worsening of anemia symptoms
- Confirmation
  - Significant decrease in HbA (>50%)
  - Total Hb levels (>30%)
  - No immunohematology results factored in
    - Detectable antibodies not always found in DHTR cases

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DHTR Incidence Results

- During the 30 months
  - 311 patients
  - 694 transfusions
    - 360 OTE (221 patients)
    - 334 CTE (118 patients)
- 15 DHTR reported
  - Incidence was:
    - 4.2% over 3 years
    - 6.8% per patient
  - OTE associated with a higher risk of DHTR

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DHTR Transfusion Characteristics

- 26 cases
  - 15 cases from single-center study
  - 11 cases referred during the same time period
    - Also due to OTE transfusions
- Patient Data
  - Age: 19-63 years (mean age of 33.5 ± 9 years)
  - 66% women
  - 80.2% originated from Sub-Saharan Africa
  - Transfusions triggers
    - 45.5% acute complications
    - 31.5% pregnancy

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DHTR Outcomes

- 3 patients died (11.5%)
- 8 developed classically significant antibodies
- 9 developed nonclassically significant antibodies
- 9 no new antibodies

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DHTR vs No DHTR Transfusions

		345 OTE- No DHTR	26 OTE - DHTR
Number of previous transfusions		0-546	0-71
Known immunizations (antibodies)	No history	66%	27%
	Nonsignificant or Rh/K	25%	35%
	Significant	10%	38%
History of DHTR		12 (3.5%)	8 (30%)
RBCs transfused	Liquid	333 (97%)	18 (69%)
	Frozen	3 (1%)	6 (23%)

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What Is the Difference?

Chronic Transfusions

- Not associated with DHTR
- Benefit from more extensive matching protocols
- Pretransfusion immunization status
  - Higher numbers of immunized patients than occasional group
  - More likely to form nonsignificant antibodies and/or Rh/Kell
    - 3% significant

Occasional Transfusions

- Associated with higher risk of DHTR
- May not get antigen matching protocol
  - Limited time
  - History unknown
- Pretransfusion immunization
  - More likely to be nonimmunized
  - More likely to form significant antibodies
    - FY, JK, MNS (11.6%)

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Predictive Risk Groups

Classification

- Low Risk
  - Score <8
- Intermediate Risk
  - Score 8-14
- High Risk
  - Score >14

Application to CTE Transfusions

- 334 CTEs
- Low risk
  - 95.5%
- Intermediate
  - 4.5%
- High
  - None

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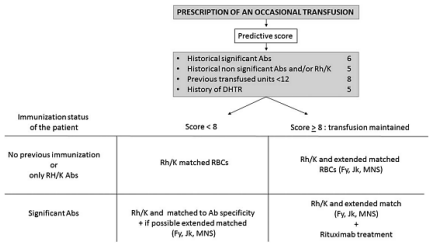
Transfusion Strategy



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Using the Transfusion Predictive Score

- Applied for all occasional transfusion episodes
- First transfusion episode in patients enrolled in a chronic transfusion protocol
- Low risk (<8) = safely transfuse
- Intermediate and high risk = evaluate closely
  - If score >14; consider rituximab to prevent new immunization and may decrease DHTR risk

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Concerns

- Strategy had a very good negative predictive value (NPV)
  - Patient with a score under 8 have low risk of DHTR
- 4 of 26 DHTR cases would have been missed if strategy applied
  - No history of DHTR
  - >12 units of previous RBCs
  - 1 patient had no history of immunization
  - 1 of the patients considered low risk actually died

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DHTR Risk?

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The Patient

- **Patient Information**:
  - 41-year-old
  - African American
  - Female
- **Diagnosis:**
  - Anemia, Sickle Cell Anemia
- **History:**
  - Anti-C, anti-E, anti-K, Anti-S, anti-Js^{a}, anti-Jk^{b}
  - Cold autoantibody
  - Warm autoantibody
  - HLA antibody
- **Hospital Reports:**
  - Positive DAT
  - Antibody Screen: 1-3+ reactivity in all cells
- **Transfusion:**
  - Want 3 units to transfuse

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ABO/Rh						
ABO Group					Rh Type	
	Anti-A	Anti-B	A_1 Cells	B Cells	Anti-D	Control
IS	0	4+	4+	0	3+	0

- IRL types patient as B Rh positive.

Direct Antiglobulin Test			
Poly	IgG	C'	Saline
(+)	(+)	(+)	(0)

- DAT is weakly positive.

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Report

- History:
  - Warm autoantibody
  - Cold autoantibody
  - HLA antibody
  - Anti-C, anti-E, anti-K, anti-Jk^{b}, anti-S, anti-Js^{a}
- Eluate
  - No alloantibodies detected
- Plasma
  - No new alloantibodies detected

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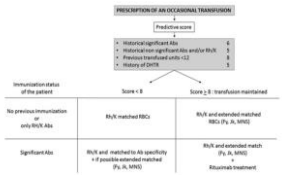
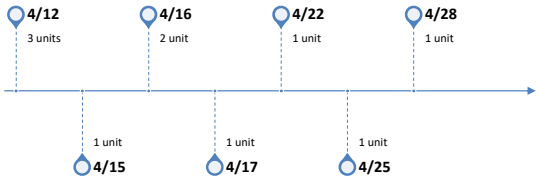
Transfusion Predictive Score – Should they transfuse?



- History of nonsignificant antibodies and/or Rh/K= 5
- Historical significant Abs = 6
- Total Score = 12

- Intermediate Risk

Transfusions



Antibody Identification – Part 2

- **Patient Information**:
  - 41-year-old
  - African American
  - Female
- **Diagnosis:**
  - Sickle cell crisis
- **History:**
  - Anti-C, anti-E, anti-K, Anti-S, anti-Js^a, anti-Jk^b
  - Cold autoantibody
  - Warm autoantibody
  - HLA antibody
- **Hospital Reports:**
  - Positive DAT
  - Antibody Screen: 2+ reactivity in all cells
- **Transfusion:**
  - Want 1 unit to transfuse

ABO/Rh						
ABO Group					Rh Type	
	Anti-A	Anti-B	A_1 Cells	B Cells	Anti-D	Control
IS	0	4+mf	4+	0	3+mf	0

- IRL types patient as B Rh positive.

mf = mixed field due to transfusion of Group O, Rh pos and Rh neg RBCs.

Direct Antiglobulin Test			
Poly	IgG	C'	Saline
1+	1+	(+)	(0)

- DAT is weakly positive.

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Antibody Panel

	Rh					Kell			Duffy		Kidd		Lewis		MNS					Plasma Results	
	D	C	E	c	e	K	k	Js^a	Fy^a	Fy^b	Jk^a	Jk^b	Le^a	Le^b	M	N	S	s		5' RT	PEG IAT
1	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	+	0	+		0	(0)
2	0	0	0	+	+	0	+	0	+	0	+	0	0	+	+	+	0	+		0	1+
3	0	0	0	+	+	0	+	0	0	+	+	0	0	+	0	+	0	+		0	1+
4	+	0	0	+	+	0	+	0	0	0	+	0	+	0	+	0	0	0		0	(0)
5	0	0	0	+	+	0	+	0	+	0	+	0	+	0	0	+	0	+		0	1+
6	0	0	0	+	+	0	+	0	0	+	+	0	+	0	+	+	0	+		0	1+
7	0	0	0	+	+	0	+	0	0	+	+	0	0	+	+	0	0	+		0	1+
8	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	0	0	+		0	(0)
AC																				0	1+

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Acid Eluate

	Rh					Kell			Duffy		Kidd		Lewis		MNS				Eluate
	D	C	E	c	e	K	k	Js^a	Fy^a	Fy^b	Jk^a	Jk^b	Le^a	Le^b	M	N	S	s	PEG IAT
1	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	+	0	+	(0)
2	0	0	0	+	+	0	+	0	+	0	+	0	0	+	+	+	0	+	2+
3	0	0	0	+	+	0	+	0	0	+	+	0	0	+	0	+	0	+	2+
4	+	0	0	+	+	0	+	0	0	0	+	0	+	0	+	0	0	0	(0)
5	0	0	0	+	+	0	+	0	+	0	+	0	+	0	0	+	0	+	2+
6	0	0	0	+	+	0	+	0	0	+	+	0	+	0	+	+	0	+	2+
7	0	0	0	+	+	0	+	0	0	+	+	0	0	+	+	0	0	+	2+
8	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	0	0	+	(0)

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What Next?

- Warm Autoantibody
- Cold Autoantibody
- High Incidence Antibody
- Multiple Antibodies

Blood Group	Antigen	Results	Comments
Rh	c	+	
	C	0	
	e	+	
	E	0	
	V	+	
	VS	+	
Kell	K	0	
	k	+	
	Kpa	0	
	Kpb	+	
	Jsa	0	
	Jsb	+	
Duffy	Fya	0	
	Fyb	W*	Not at risk for anti-Fy^a
Kidd	Jka	+	
	Jkb	0	
MNS	M	+	
	N	0	
	S	0	
	s	+	
	U	+	
Lutheran	Lua	0	
	Lub	+	
Diego	Dia	0	
	Dib	+	
Colton	Coa	+	
	Cob	0	
Dombrock	Doa	0	
	Dob	+	
	Hy	+	
	Joa	+	
Landsteiner-Wiener	LWa	+	
	LWb	0	
Scianna	Sc1	+	
	Sc2	0	

- **Human Erythrocyte Antigen (HEA) Phenotype by DNA Analysis Report**
  - Sample contains GATA mutation resulting in loss of Fy^b expression on RBCs
    - Individuals not expected to make anti-Fy^b
  - Patient is positive for common high incidence antigens
    - Examples: anti-Js^b and anti-U

Acid Eluate

	Rh					Kell			Duffy		Kidd		Lewis		MNS				Eluate
	D	C	E	c	e	K	k	Js^a	Fy^a	Fy^b	Jk^a	Jk^b	Le^a	Le^b	M	N	S	s	PEG IAT
1	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	+	0	+	(0)[4]
2	0	0	0	+	+	0	+	0	+	0	+	0	0	+	+	+	0	+	2+
3	0	0	0	+	+	0	+	0	0	+	+	0	0	+	0	+	0	+	2+
4	+	0	0	+	+	0	+	0	0	0	+	0	+	0	+	0	0	0	(0)[4]
5	0	0	0	+	+	0	+	0	+	0	+	0	+	0	0	+	0	+	2+
6	0	0	0	+	+	0	+	0	0	+	+	0	+	0	+	+	0	+	2+
7	0	0	0	+	+	0	+	0	0	+	+	0	0	+	+	0	0	+	2+
8	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	0	0	+	(0)[4]

Antibody Panel

	Rh					Kell			Duffy		Kidd		Lewis		MNS				Plasma Results	
	D	C	E	c	e	K	k	Js^a	Fy^a	Fy^b	Jk^a	Jk^b	Le^a	Le^b	M	N	S	s	5' RT	PEG IAT
1	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	+	0	+	0	(0)[4]
2	0	0	0	+	+	0	+	0	+	0	+	0	0	+	+	+	0	+	0	1+
3	0	0	0	+	+	0	+	0	0	+	+	0	0	+	0	+	0	+	0	1+
4	+	0	0	+	+	0	+	0	0	0	+	0	+	0	+	0	0	0	0	(0)[4]
5	0	0	0	+	+	0	+	0	+	0	+	0	+	0	0	+	0	+	0	1+
6	0	0	0	+	+	0	+	0	0	+	+	0	+	0	+	+	0	+	0	1+
7	0	0	0	+	+	0	+	0	0	+	+	0	0	+	+	0	0	+	0	1+
8	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	0	0	+	0	(0)[4]
AC																			0	1+

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What Do We Know

- Have a genotype already
  - List of potential antibodies the patient can form
- Patient already has an anti-C, anti-E, anti-K, anti-Jk^b, anti-S, anti-Js^a
- Antigens to consider:
  - Fy^a, Do^a
  - Fy^b (?)

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Selected Cell Panel

	Rh					Kell			Duffy		Kidd		Lewis		MNS				Dombrock		Plasma Results		Eluate Results
	D	C	E	c	e	K	k	Js^a	Fy^a	Fy^b	Jk^a	Jk^b	Le^a	Le^b	M	N	S	s	Do^a	Do^b	PEG IAT		PEG IAT
1	+	0	0	+	+	0	+	0	0	0	+	0	0	0	0	+	0	+	0	+	(0)[4]		(0)[4]
2	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	+	0	+	+	0	(0)[4]		(0)[4]
3	+	0	0	+	+	0	+	0	+	0	+	0	+	0	+	+	0	+	+	+	1+		1+
4	+	0	0	+	+	0	+	0	0	0	+	0	0	+	0	+	0	+	0	+	(0)[4]		(0)[4]
5	0	0	0	+	+	0	+	0	0	+	+	0	0	+	0	+	0	+	+	0	(0)[4]		(0)[4]

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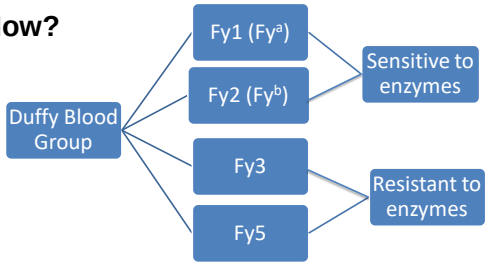
What Do We Know

- Eluate
  - Reactivity with:
    - Fy(a+b-) and Fy(a-b+) cells
- Plasma
  - Reactivity with:
    - Fy(a+b-) and Fy(a-b+) cells

Next Steps..

- Plasma Testing
  - Determine what antibody in the Duffy blood group system is causing reactivity

How?



Enzyme Treatment

- Ficin treated RBCs tested with patient's plasma

	Rh					Kell			Duffy		Kidd		Lewis		MNS				Plasma		
	D	C	E	c	e	K	k	Js^a	Fy^a	Fy^b	Jk^a	Jk^b	Le^a	Le^b	M	N	S	s	PEG IAT	Ficin 30'37C	Ficin IAT
1	+	0	0	+	+	0	+	0	0	0	+	0	0	+	+	+	0	+	(0)$^{\surd}$	1+	(0)$^{\surd}$
2	0	0	0	+	+	0	+	0	+	0	+	0	0	+	+	+	0	+	1+	1+	1+
3	0	0	0	+	+	0	+	0	0	+	+	0	0	+	0	+	0	+	1+	1+	1+
4	+	0	0	+	+	0	+	0	0	0	+	0	+	0	+	0	0	0	(0)$^{\surd}$	1+	(0)$^{\surd}$
5	0	0	0	+	+	0	+	0	+	0	+	0	+	0	0	+	0	+	1+	1+	1+
6	0	0	0	+	+	0	+	0	0	+	+	0	+	0	+	+	0	+	1+	1+	1+
7	0	0	0	+	+	0	+	0	0	+	+	0	0	+	+	0	0	+	1+	1+	1+

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- Anti-Fy3
  - **Negative** with cord cells
  - Positive with Rh_{null} cells
- Anti-Fy5
  - Positive with cord cells
  - **Negative** with Rh_{null} cells

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Rh Null and Cord Cells

		Rh					Kell		Duffy		Kidd		Lewis		MNS				Plasma	Eluate
		D	C	E	c	e	K	k	Fy^a	Fy^b	Jk^a	Jk^b	Le^a	Le^b	M	N	S	s	PEG IAT	PEG IAT
1	Cord Cells	0	0	0	+	+	0		+	+	+	0					0	+	(0)$^{\surd}$	(0)$^{\surd}$
2	Cord Cells	+	0	0	+	+	0		0	+	+	0					0	+	(0)$^{\surd}$	(0)$^{\surd}$
3	Rh null	0	0	0	0	0	0		0	+	+	0	+	0	+	0	0	+	1+	1+
4	Rh null	0	0	0	0	0	0	+	+	+	+	0	0	0	+	+	0	+	1+	1+

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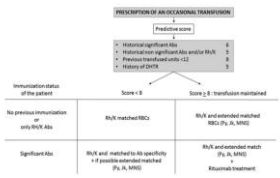
Are We Done?

Report

- History:
  - Warm autoantibody
  - Cold autoantibody
  - HLA antibody
  - Anti-C, anti-E, anti-K, anti-Jk^{b}, anti-S, anti-Js^{a}
- Eluate and Plasma
  - Probable anti-Fy3
- Results provide serologic evidence of a delayed transfusion reaction due to anti-Fy3

Transfusion Predictive Score – Take 2



- History of nonsignificant antibodies and/or Rh/K= 5
- Significant antibodies = 6
- History of DHTR = 5
- Total Score = 16

- **High Risk**

Transfusion

- Recommend:
  - E-negative, C-negative, K-negative, Js(a-), Fy(a-b-), Jk(b-), S-negative blood negative with the patient's plasma.
- Transfused:
  - 1 E-,C-,K-,Js(a-),Fy(a-b-),Jk(b-),S- units were transfused several days after workup
    - Prevalence:
      - In Caucasian population: 0.15%
      - In African American population: 10.9%

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Case Follow Up

- Antibody Identification performed again
  - 10 days later (May):
    - Diagnosis: Sickle cell crisis with other complications
    - Current Hgb 4.3 g/dL
    - DAT Negative
    - No new alloantibodies
    - Hospital called and wanted to discuss possibility of an exchange transfusion
    - No transfusions as patient was potentially entering hyperhemolysis
  - 33 days later (June):
    - Diagnosis: sickle cell crisis
    - DAT Negative
    - Cold autoantibody demonstrating again
    - No new alloantibodies
    - 1 E-C-K-Js(a-)Fy(a-b-)Jk(b-)S- unit transfused

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Hyperhemolysis

- Severe form of delayed hemolytic transfusion reaction
- Clinicians should have a high awareness for suspicion of hyperhemolysis in sickle cell patients
- Critical as transfusions in hyperhemolytic episode can accelerate hemolysis and cause life-threatening anemia
- Recommendations:
  - Stop transfusions, if possible
  - IVIG and steroids

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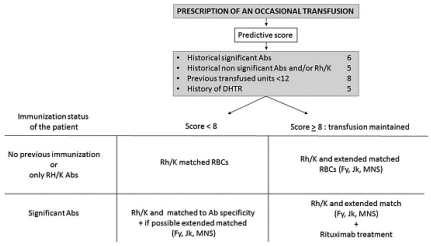
Transfusion Strategy



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Objectives

- Describe characteristics of sickle cell disease and delayed hemolytic transfusion reactions
- Describe the tests used to resolve the most complex serologic workups
- Apply recent research data to predict the risk of delayed hemolytic transfusion for transfusion of a sickle cell patient
- Select appropriate units for transfusion

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