

**POST-OPERATIVE, POST-TRANSFUSION  
PAINFUL CRISIS IN A PATIENT WITH SICKLE CELL DISEASE**

Case study by Jim Perkins (©2009)

A 33 year old African-American man with sickle cell disease (SCD), phenotype SS, was referred to the blood bank for exchange transfusion prior to elective cholecystectomy. The patient had a 4 month history of crampy, right upper quadrant pain brought on by fatty meals. The pain radiated to the right shoulder and lasted for approximately 40 minutes. There was no associated nausea or vomiting. Ultrasound of the gallbladder confirmed a large stone. The patient had a history of multiple pain crises, the last of which occurred 4 months earlier. He had been transfused on several occasions, the last time 7 months earlier. Splenectomy was performed 7 years earlier, and he also had a history of a urinary tract infection. The remainder of the history was unremarkable.

Physical examination was unremarkable except for mild cardiomegaly and a liver edge 7cm below the costal margin.

Six days before surgery a partial exchange transfusion was performed in the outpatient clinic consisting of withdrawal of one unit of whole blood and transfusion of two units of RBCs. The blood group antibody detection test was negative. This brought his hemoglobin (hgb) to 11.2 from a baseline of 10.9 gm/dL. One day before surgery a second partial exchange transfusion of one unit of his whole blood for two units of RBCs packed cells was performed. This was repeated later that day. Again, no antibodies were detected in pretransfusion testing. On a blood specimen drawn the morning before surgery hgb electrophoresis revealed 59% hgb-A and 41% hgb- S, with a total hgb level of 12.7 and a hematocrit of 37.5.

Cholecystectomy was performed without apparent problem, yielding a pigmented gallstone. The estimated blood loss was 100mL.

On the night after surgery he spiked a temperature to 105°F, with a pulse rate of 100/min, respirations 18/min, and BP 130/70 mm Hg. He defervesced to 101°F with a cooling blanket one hour later. He was empirically started on penicillin and gentamycin every 4 hours. Blood cultures were obtained which ultimately proved negative. He continued to have intermittent fevers as high as 103°. He also began to complain diffuse pain. Macroscopic urinalysis (urine “dipstick” test) was 3+ for “blood” on the first and second post-operative days, and 1+ on the third day; microscopic urinalysis results are not available.

On the 4<sup>th</sup> post-operative day his hemoglobin level had fallen to 7.1, and a specimen was sent to the blood bank for pre-transfusion testing. The following serologic results were obtained:

**ABO and Rh Typing**

	Anti-			Test cells			Anti-		Interp
	A	B	A,B	A <sub>1</sub>	A <sub>2</sub>	B	D	D, cntl	
IS	0	0	0	1+	w+	1+	3+	0	O pos
RT, 30'				3+	1+	3+			

**Antibody detection test**

Cells	Saline			2 drops serum , LISS				Ficin-tx'd RBCs		
	IS	30' RT	4° C	IS	30' RT	AHG	CC	30' RT	AHG	CC
OI	0		0		w+	1+		1+	2+	
OII	0	0	0		1+	2+		2+	2+	
Auto	0	0	2+		0	w+		0	w+	
A <sub>1</sub>	1+	3+	3+							
A <sub>2</sub>	w+	1+	2+							
B	1+	3+	4+							
O Cord	0	0	0							

**Direct antiglobulin test**

IS	AHG		
	Poly	>IgG	>C'
	w+, mf	w+, mf	0

**Serum panel**

Lot #1018	Rh	MNSs							P	Lewis		Lutheran		Kell				Duffy		Kidd		Xg	LISS					FICIN						
Cell	Rh	D	C	E	c	e	V	M	N	S	s	P1	Le <sup>a</sup>	Le <sup>b</sup>	Lu <sup>a</sup>	Lu <sup>b</sup>	K	k	Kp <sup>a</sup>	Js <sup>a</sup>	Fy <sup>a</sup>	Fy <sup>b</sup>	Jk <sup>a</sup>	Jk <sup>b</sup>	Xg <sup>a</sup>	Cell	IS	RT	37°	AHG	CC	37	AHG	CC
1	r'r	0	+	0	+	+	0	+	+	+	0	+	0	+	0	+	0	+	0	0	+	+	0	+	+	1	0	0	0	vw+		2+	vw+	
2	R1R1w	+	+	0	0	+	0	0	+	0	+	+	0	+	0	+	0	+	0	0	+	+	0	+	0	2	0	0	w+	1+		2+	w+	
3	R1R1	+	+	0	0	+	0	+	+	+	0	+	0	+	0	+	0	+	0	0	0	+	+	0	0	3	0	0	0	vw+		1+	vw+	
4	R2R2	+	0	+	+	0	0	+	+	+	+	+	+	0	0	+	0	+	0	0	+	+	0	+	+	4	0	0	1+	2+		2+	3+	
5	r''r	0	0	+	+	+	0	0	+	0	+	w	0	+	0	+	+	+	0	0	+	+	+	0	+	5	0	0	1+	1+		2+	2+	
6	rr	0	0	0	+	+	+	+	+	+	+	+	+	0	0	+	0	+	0	0	0	0	+	+	+	6	0	0	0	0	2+	0	0	2+
7	rr	0	0	0	+	+	0	+	0	0	+	+	+	0	0	+	+	+	0	0	0	+	0	+	+	7	0	0	0	0	2+	0	0	2+
8	rr	0	0	0	+	+	0	0	+	0	+	+	+	0	0	+	0	+	0	+	0	+	+	0	+	8	0	0	0	0	2+	0	0	2+
9	rr	0	0	0	+	+	0	+	0	+	+	0	0	0	0	+	0	+	0	0	+	0	+	+	+	9	0	0	0	0	2+	0	0	2+
10	Ro	+	0	0	+	+	0	+	+	0	+	+	0	+	0	+	0	+	0	0	+	0	+	+	+	10	0	0	0	0	2+	0	0	2+
11	Cord	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	11	0	0	0	w+		0	vw+	
AC																										AC	0	0	0	vw+		0	vw+	



**DISCUSSION QUESTIONS:**

1. What antibody(ies) is(are) present in the patient's post-transfusion serum? What is present in the eluate? Is any further serologic workup needed to prove your answer?
2. What immunohematologic problem is occurring based on this sequence of events and the patient's laboratory data? What additional laboratory tests would help make this diagnosis?

**ADDITIONAL TESTING:**

Stored tubing segments from the 6 previously transfused units of RBCs were retrieved and tested for the C and E antigens with the following results:

<b>RBCs</b>	<b>Day transfused</b>	<b>Anti-C</b>	<b>Anti-E</b>
<b>Unit #1</b>	<b>-6</b>	<b>0</b>	<b>3+</b>
<b>Unit #2</b>	<b>-6</b>	<b>3+</b>	<b>3+</b>
<b>Unit #3</b>	<b>-1</b>	<b>0</b>	<b>3+</b>
<b>Unit #4</b>	<b>-1</b>	<b>3+</b>	<b>0</b>
<b>Unit #5</b>	<b>-1</b>	<b>3+</b>	<b>3+</b>
<b>Unit #6</b>	<b>-1</b>	<b>3+</b>	<b>0</b>
<b>R1R2 pos control</b>		<b>4+</b>	<b>4+</b>
<b>rr neg control</b>		<b>0</b>	<b>0</b>

The antibody detection test was repeated on the patient specimens drawn before the exchange transfusions. Both were negative. The final diagnosis was of a delayed hemolytic transfusion reaction due to anti-C and anti-E.

**DISCUSSION QUESTIONS REGARDING THE TRANSFUSION MANAGEMENT OF SICKLE CELL DISEASE:**

3. Is there any connection between this patient's reaction to transfusion and his painful sickle cell crisis? Do you think that this exchange transfusion was indicated? Assuming that the transfusion was appropriate, how might this transfusion reaction have been avoided?