

The Patient with Sickle Hemoglobin

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Sickle cell trait exists in approximately 10% of the negro population and is generally considered a benign condition. A recent report by Jones, et al.¹ again points out that sickle hemoglobin, even in combination with normal adult hemoglobin imposes an added threat to its carrier. (Figure 1) That general and thoracic surgery can be performed in these patients with a relatively low mortality and morbidity has been reported.^{2,3}

Open-heart surgery poses special problems in acid-base balance, oxygenation, perfusion and temperature control that are not common to other types of surgery. Since 1965, 22 patients with sickle hemoglobin have undergone open-heart surgery by the University of Tennessee, Cardiovascular Surgery Department. Two of the more recent patients will be presented in detail to illustrate the problems, and in some cases, how to avoid them.

Case Reports

Case 1—W. C., an 18 year old female was seen in November, 1971 with a history of recurrent episodes of heart failure which would respond to bed rest, diuretics, and digitalis. She had been known to have a harsh systolic murmur at the apex since late childhood. At cardiac catheterization, she was shown to have severe mitral valve insufficiency and valve replacement was recommended. While awaiting surgery, she had a "seizure" characterized by "drawing" of the left side of the face. Cultures of her blood revealed a micro-aerophilic streptococcus and she was treated for 6 weeks with intravenous antibiotics.

Mitral valve replacement with a Starr-Edwards prosthesis was performed on March 2, 1972. Hemoglobin electrophoresis showed that she had A-S hemoglobin; thus, preparations were made to obtain samples for determination of the number of sickle cells and blood gases during the perfusion. She tolerated the surgery well and was discharged on the 13th post-operative day. Discharge medications included Coumadin, digitalis, and an antibiotic for long-term rheumatic fever prophylaxis.

Case 2—L.M.K., a 42 year old female was seen in June, 1970 with progressive dyspnea, pedal edema and fatigue. Her history included rheumatic fever at age 8 years, and a heart murmur of many years duration. She had had cardiac catheterization in 1963 which showed severe aortic valve insufficiency and mild aortic stenosis.

SICKLE CELL ANEMIA



SICKLE CELL TRAIT



Figure 1: Drawing showing degree of sickling in vitro of S-S and A-S red blood cells. (courtesy of Dr. L. W. Diggs)

A repeat catheterization in October, 1968 demonstrated worsening of the condition. Left ventricular end diastolic pressure was 32mmHg. At the time she was seen in 1970, it was felt that aortic valve replacement was necessary in view of her worsening clinical condition. She was known to have sickle cell trait (A-S Hemoglobin); thus, preparations were made to monitor all parameters during and immediately after surgery.

The aortic valve was replaced with a Starr-Edwards aortic valve prosthesis on July 29, 1972, using mild hypothermia (30°C) and without coronary perfusion. Defibrillation was accomplished with some difficulty, but with the establishment of normal sinus rhythm, adequate cardiac output was achieved. A satisfactory course was maintained for 8 hours, at which time, she removed her endotracheal tube. Mild hypoventilation occurred with resultant hypoxia and acidosis. At 22 hours following surgery, she experienced a cardiac arrest. Adequate cardiac output could not be maintained, and she died 33 hours following surgery.

Discussion

Both patients were given very little pre-operative sedation. Adequate hydration and oxygenation were maintained throughout the operative period. The Bentley Solid Bubble Oxygenator and Sarns Roller Pump were used for by-pass. Prime solution consisted of 1000cc D5W, 1000cc L.R., and 1000cc of fresh heparinized whole blood. Cannulation was through the ascending aorta and flow rates were approximately 60cc/Kg of body weight. (See page 28)

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Open-Heart Surgery In The Patient With Sickle Hemoglobin



Sodium bicarbonate was added to the oxygenator as needed to adjust pH. Arterial blood samples are routinely obtained for sickling determinations and blood gas studies. A modification of the Sherman⁴ procedure as described by Barreras and Diggs⁵ was used for these determinations. Irreversibility of the sickled cell has been described by many authors.^{6, 7}

Harris and co-workers⁸ reported three cases in which congenital heart lesions occurred in patients with S-S hemoglobin. Open-heart repair was successfully accomplished in two of their cases. Dilution of the S-hemoglobin⁹ and adequate hydration¹⁰ were utilized in both of their cases. Strict attention was paid to the blood gases in each of our patients and blood was obtained to determine the amount of sickling with each gas determination.

Chart 1 shows the correlation of the pH and pO₂ with the percentage of sickling in case 1. A smooth course is evidenced by the normal pH and pO₂ during the operative and post-operative period. Note that an attempt was made to keep her slightly alkalotic.⁵

Chart 2 shows the overall course of case 2. All the samples outlined in this chart were arterial. As she started to get into difficulty 20 hours following surgery, frequent arterial blood gas determinations were done, and the percent of sickling was determined in both arterial and venous blood.

Chart 3 shows the findings surrounding the cardiac arrest. It is quite evident from the chart that acidosis plays a major role in the developing of sickling. It is also evident that patients with only the sickle cell trait can have massive amounts of sickling under the right conditions.

Time on by-pass was 53 minutes for case 1 and 115 minutes for case 2. Post-operative determinations of free plasma hemoglobin were 33.6 and 42mgm% respectively. This denotes very little hemolysis under the conditions of the surgery.

Summary

Two cases with A-S hemoglobin undergoing open-heart surgery are presented. General problems and techniques of perfusion and care are discussed. In one case, the severity of sickling under proper conditions is pointed out, even in the sickle cell trait patient. It is not a condition to be taken lightly.

CHART 1
Correlation of Blood Gases and Sickling in Case 1

TIME	pH	pO ₂	PERCENT SICKLING		QUESTION-ABLE
			Irreversible	Reversible	
Before By-Pass	7.415	219	0	0	0
On By-Pass	7.443	215	0	0.6%	0.8%
After By-Pass	7.371	140	0	0.2%	1.2%
6 Hours Post-Op	7.500	140	0.2%	1.0%	0.2%
12 Hours Post-Op	7.535	104	0	0	0
24 Hours Post-Op	7.450	108	0	0.4%	0.4%

CHART 2:
Correlation of Blood Gases and Sickling in Case 2

TIME	pH	pO ₂	PERCENT SICKLING
Before By-Pass	7.30	114	0%
On By-Pass	7.44	418	0%
6 Hours Post-Op	7.46	114	0%
12 Hours Post-Op	7.39	69	1-2%
22 Hours Post-Op	7.15	74	28%

CHART 3:
Case 2, Arterial Blood Gases Correlated with Both
Arterial and Venous Sickling
Before, During, and After Cardiac Arrest.

HOURS POST-OP	pH	pO ₂	VENOUS SICKLING	ARTERIAL SICKLING
20	7.39	72	35%	22%
22	7.15	74	37%	28%
22.5	7.56	67	12.5%	8%
23	7.38	418	3%	<1%
24	7.40	219	<1%	<1%
25	7.34	77	6.5%	<1%

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