
Cardiopulmonary Bypass on Patients with Sickle Cell Hemoglobin

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Abstract

The purpose of this paper is to present briefly the background and physiology of sickle cell states, with special emphasis on sickle cell trait patients who require cardiopulmonary bypass surgery and the importance of the perfusionist's role in managing these patients during bypass.

Introduction

Sickle cell disease is a genetic disorder characterized by an abnormal hemoglobin molecule which can distort erythrocytes into sickle cells.¹ While some reserve the term, "sickle cell disease," for the homozygous state (SS) only, others use it in a broader or generic sense to include all conditions in which hemoglobin S is present. By the latter definition, the sickle cell disease states would include not only sickle cell anemia (SS), but also the usually benign "carrier" state (AS), doubly het-

erozygous hemoglobinopathies (as SC, SD), and S-Thalassemia.

There are two basic types of disease processes which result in the faulty manufacture of a hemoglobin polypeptide chain. In one (abnormal hemoglobins), the polypeptide chain produced is abnormal and in the other (thalassemias), the polypeptide chain is normal but produced in insufficient quantity.²

The disease is thought to have arisen in Arabia in Neolithic times and from there to have spread eastward and westward. Today it is found in parts of India, East and West Africa, the West Indies, and among the American Negroes.³ Worldwide distribution is such that sickle cell disease occurs in areas where malaria had at one time been quite prevalent. Sickle cell provided resistance, though not immunity, to malaria and was beneficial to those who carried the gene.⁴ In the United States, some 50,000 to 100,000 Negroes have the disease and more than 2,000,000 are carriers. This is equivalent to 8-11% of the Negro population.¹

The sickle cell disease process affects every organ in the body and has major symptoms of anemia, joint disease, leg ulcers, and periods of severe pain. These painful episodes, or crises, last anywhere from hours to days, occur at unpredictable intervals, and represent one of the most devastating aspects of the disease.¹ Under normal conditions, the sickle cell trait produces

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few symptoms but is a potential threat to the successful management of patients undergoing procedures such as cardiopulmonary bypass which expose them to certain environments which magnify this problem.³

Factors which Precipitate Sickling_____

The ease with which sickling occurs depends upon several factors. Hypoxemia induces sickling of erythrocytes, which increases the viscosity of the blood. Sickling is also enhanced by acidosis and hypothermia. With the increase in viscosity of the whole blood and the formation of sickle cell thrombi, a vicious cycle is started in which the factors of stasis, hypotension, lowered pH, and continuing oxygen uptake combine to increase the number of sickled cells and prolong the stasis.⁵

Consequences of Intravascular Sickling_____

Intravascular sickling produces vaso-occlusion and infarction. Small blood vessels supplying various organs and tissues can be partially or completely blocked by sickled erythrocytes. Such occlusion of the circulation may result in intravascular thrombi, hemolysis, and dysfunction, as well as permanent tissue or organ damage. Anemia results mainly from the shortened survival time of the red blood cell from trauma within the circulation and hemolysis.¹

During cardiopulmonary bypass surgery, inadequate oxygenation of arterial blood, hypoperfusion of part or parts of the body, or acidosis, even for short periods of time, can trigger the vicious cycle and result in interference with tissue perfusion which might be fatal. In addition, severe hemolysis may result from subjecting the already fragile sickle cell to the trauma of extra-corporeal circulation.⁶

Methods of Preventing Sickling on Bypass_____

The above-mentioned complications can be minimized on bypass by careful attention to perfusion parameters and slightly revising technique. Partial exchange transfusion prior to bypass reduces the number of circulating cells containing sickled hemoglobins (S). This can be achieved by removing blood and replacing it with normal donor blood, or other priming fluid from the oxygenator prior to starting extra-corporeal perfusion. This exchange transfusion should be carried out slowly to prevent a sudden drop in the circulating blood

volume and cardiac output. The blood can be diverted to a cell saver to be separated, as suggested by Black and Dearing.⁷ Since the plasma of sickle cell patients is normal, it can be spun down and retransfused to preserve the patient's own platelets and other clotting factors. This transfusion not only augments oxygen-carrying capacity, but reduces the number of circulating sickle cells. Normal transfused cells do not participate in the sickling phenomenon.⁵

Normal arterial blood pO₂ should be maintained with efficient and abundant tissue perfusion. One way to determine adequate perfusion is by monitoring venous pO₂. It is important to maintain oxygen saturation above 97% because it has been shown² that cells show sickling shape changes with oxygen saturation of hemoglobin in the ranges of 30–90%.

Acidosis should be prevented or immediately corrected. Even maintaining a slightly alkalotic state might be preferred as a prophylactic measure to prevent sickling of the red cells.

Hypothermia should be minimized, as it presents one mechanism for the development of capillary stasis and increased blood viscosity. It is recommended⁸ that sickle cell patients should not be cooled any lower than 30–32°C. Cooling should be performed evenly and slowly to prevent vasoconstriction and hypoperfusion.

A carefully selected prime mixture should include some blood to mix with the patient's blood and hemodilute to lower the blood viscosity. The study by Valitis (cited in Howells et al., 1972²) showed that the oxygen dissociation curve of the cells which have been stored in an acid-citrate-dextrose medium at 4°C is shifted to the left. Therefore, blood should be as fresh as possible. Transfusion of blood stored for seven days or more results in a similar change in the oxygen dissociation curve of the recipient's blood. As a result, less oxygen can be released from the blood to the tissue.

Adequate arterial pressure should be maintained. Arterial hypoperfusion predisposes to sickling and should be avoided. The recommended mean arterial pressure is 65–80 mmHg. Maintaining an adequate level of anesthesia will prevent the patient from awakening and shivering, causing constriction and extremely high pressure.

Abundant and repeated perfusion of the coronary circulation with appropriate cardioplegic solution should be provided. This prevents stasis of blood in the myocardium. Oxygen tension can drop to a very low

level when the aorta is occluded, assuming special importance in those patients whose red cells contain hemoglobin S and who are, therefore, susceptible to sickling during anoxia. This could clog the myocardial circulation, increase stasis, and produce myocardial infarction with grave complications.

Conclusion

The presence of sickle cell disease or sickle cell trait with all of its risks of sickling, thrombosis, hemolysis, and infarction does not contraindicate putting a patient on extra-corporeal bypass. Knowledge of this disease, together with proper care and judgment, can yield good results.

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