**Departmental Clinical** 

**Protocol Manual**

**Clinical Protocol CP.XX.xxx**

\*The intent of this product is to be a resource; not a replacement for institutional protocols. Standard 1 of AmSECT’s Standards and Guidelines for Perfusion Practice.1 These Standards and Guidelines may also be superseded by the judgement of the healthcare professional taking into account the facts and circumstances of the individual case.

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| **SUBJECT/TITLE:** | **SICKLE CELL DISEASE** |
| **PURPOSE:** | To provide a guideline and resource for a patient with a previously diagnosed risk of Sickle Cell Disease. Cardiopulmonary bypass in the presence of abnormal sickle cell disease/hemoglobin S (Hgb-S) causes the cells to move slowly, clump together, eventually breaking down resulting in anemia and blood clots. Regardless of the disease state, homozygous or heterozygous, the surgeon should work together with the cardiac surgical team to ensure a safe and effective procedure is provided to all patients.  |
| **TARGET****POPULATION:** | Patients diagnosed with Sickle Cell Disease.  |
| **DEFINITIONS:** | Sickle cell disease is a hereditary, autosomal recessive hemoglobinopathy resulting from a mutant form of a β-globin gene. Normally, hemoglobin A (the most common form of hemoglobin in adults) consists of 2α and 2β chains. Hemoglobinopathy is a genetic defect of the hemoglobin molecule. Hb S is insoluble, is not malleable and forms crystals when exposed to low oxygen tension. Deoxygenated Hb S will aggregate to form long polymers that can occlude microcirculation. This patient population may have a vaso-occlusive history, ulcers, pulmonary embolisms, or strokes. Hb S shifts the oxygen- dissociation curve to the right. In patients with sickle cell disease, the mutant form of the β-globin produces hemoglobin S (Hb S). * Patients who are heterozygous for hemoglobin S possess the sickle cell trait (HbAS). (25-45% of Hb is HbS)
* Patients who are homozygous for hemoglobin S have sickle cell disease (HbSS).

There are two other hemoglobinopathies involving hemoglobin S: * HbSC
* HbS/β-thalassemia.

Clinical expression of the sickle cell disease is variable and even carriers of the trait (Hb AS) can produce manifestations as severe as patients who are homozygous for the disease (HbSS). Therefore it is important to recognize variables that play a role in sickling while on cardiopulmonary bypass. Sickling can be caused by various factors: * Hypoxemia
* Acidosis
* Hypothermia
* Infection
* Stagnation
* Hemolysis
* Presence of high percentage of HbS erythrocytes
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**POLICY:**

1. Patients with the sickle cell trait may be placed on CPB safely if oxygen saturation is kept at an increased level, acidosis is prevented, and hypothermia is limited.

2. Sickling occurs at an arterial hemoglobin saturation below 85%. Therefore, it may be necessary to remove some of the blood before bypass and replace it with adequate blood components.

3. Delivery of cold blood cardioplegia could result in capillary sludging, inadequate distribution of cardioplegia, poor myocardial protection and depressed myocardium function.

**PERFUSION PUMP CONSIDERATIONS:**

**Modifications** that can be made to decrease the risk of a sickling crisis on cardiopulmonary bypass may include:

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| **Hypoxemia** | Maintain adequate hemoglobin saturation * High PaO2
	+ (>85%) for homozygous
	+ (>40%) heterozygous.
* High SvO2(>75%)

Continuous arterial and venous blood gas monitoring is recommended |
| **Acidosis** | Maintain pH within normal parameters 7.35-7.45 while on CPB |
| **Hypothermia**  | Maintain normothermia  |
| **Infection** | Use of sterile technique  |
| **Stagnation** | Maintain high blood flows at a CI of 2.4 to prevent capillary sludging * Hemodilution may also reduce sludging
* Use of vasodilators, if needed, to maintain high flows

Use of crystalloid cardioplegia (Plegisol, Custodiol or del Nido) may help decrease the risk of stagnation within the coronaries Anesthetic assistance to maintain MAP >60mmHg * Minimal use of vasoconstrictors if possible
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| **Hemolysis** | Use of a centrifugal pump may decrease damage to RBCs Cell Saver blood is commonly discarded  |
| **Presence of High % of HbS erythrocytes**  | Exchange transfusions for patients that are HbSS to a target HbS <30% may reduce risk of sickling |

**PROCEDURE:**

The strategy for the perfusionist should be aimed at preventing the sickling crisis and its damage:

1. Maintain an adequate hemoglobin saturation

a. >85% for homozygous disease (high Pa02)

b. >40% for heterozygous trait (high SVO2)

2. Maintain pH 7.35 – 7.45

3. Maintain normothermia

4. Maintain high flows at least 2.4 Cardiac Index

5. Deliver crystalloid cardioplegia (Plegisol, Custodial) \*see example below 6. Maintain a MAP >60 mmHg

7. Centrifugal pump

8. Cell Saver – do not return cell saver blood

9. Possible exchange transfusion \*see example below

**Exchange Transfusion:**

1. Patients with sickle cell disease experience sickling at saturation drops below 85%, it therefore may be necessary to remove some of the blood before bypass and replace it with adequate blood components. An exchange transfusion may be deemed necessary if the patient has sickle cell disease with a fractional HbS >30%.

2. In patients with SCD, the fractional concentration of HbS can be as high as 70-98%. In patients with SCT, the fractional concentration of HbS is generally less than 50%. 3. If an exchange transfusion is necessary, you will need:

a. PRBC’s

b. FFP

c. 25% albumin

d. Plasmalyte-A

Volume can be removed from the patient via a Y connector to a separate reservoir off of the venous line while transfusing the blood prime at the beginning of CPB.

Careful calculation of the predicted blood volume of the patient will assist in knowing how much volume should be removed from the patient in order to reach an estimated HbS <30%.

**For Total body washout:**

1. Connect a Y connector in the venous pump line that is connected to a cell salvaging device. The platelets and the plasma can then be separated from the faulty red blood cells for reinfusion

2. Packed RBCs, FFP, 25% Albumin and Plasmalyte A will be used to prime the heart lung machine.

3. The patient must be infused with volume via the arterial cannula to replace the native blood that is removed.

4. On bypass, the oxygen saturation is kept high, acidosis is prevented and normothermia is maintained.

**THALASSEMIA:**

Thalassemia is a defect in hemoglobin that causes early red blood cell destruction and anemia, leading to a decreased oxygen delivery. This is a hereditary disorder of Mediterranean African and Asian ancestry. Hemolysis can occur when patients receive quinidine or sulfonamides.

1. Autologous blood should be salvaged prior to surgery and transfused as needed. 2. The surgeon will ensure the perfusionist takes the steps necessary to ensure adequate oxygenation.

a. Increase FIO2 as needed

b. Increase blood flow

c. Ensure the hematocrit remains adequate by hemoconcentrating and/or giving donor blood.

**CLINICAL ASSESSMENT/SCREENING:**

A. Contraindications: None

**RELATED DOCUMENTS:**

A. Cardioplegia Solution Delivery System Set-Up and Administration

**REFERENCES:**

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2. Daaboul, D., Yuki, K., Wesley, M. and DiNardo, J. (2011). Anesthetic and Cardiopulmonary Bypass Considerations for Cardiac Surgery in Unique Pediatric Patient Populations: Sickle Cell Disease and Cold Agglutinin Disease. World Journal for Pediatric and Congenital Heart Surgery, 2(3), pp.364-370.

3. Hoffbrand, A. and Moss, P. (2011). Essential Haematology: 6 th Edition. West Sussex: Wiley-Blackwell; pp.99-104.

4. Maddali, M., Rajakumar, M., Fahr, J., Albahrani, M. and Amna, M. (2006). Cardiopulmonary Bypass Without Preoperative Exchange Transfusion in Sicklers. Asian Cardiovascular &amp; Thoracic Annals, 14(1), pp.51-56.

5. Staikou, C., Stavroulakis, E. and Karmaniolou, I. (2014). A narrative review of peri-operative management of patients with thalassaemia. Anaesthesia, 69(5), pp.494-510.

6. Yousafzai, S., Ugurlucan, M., Al Radhwan, O., Al Otaibi, A. and Canver, C. (2009). Open Heart Surgery in Patients With Sickle Cell Hemoglobinopathy. Circulation, 121(1), pp.14-19.

7. Mullins F, Ott S, Mister N, Travis J. Sickle Cell Hemoglobin C Disease Patient Undergoing Coronary Artery Bypass Grafting with Complete Exchange Blood Transfusion during Cardiopulmonary Bypass. J Extra Corpor Technol.

2018;50(2):117–119.

**DISCLAIMER:**

In emergency situations, immediate life support measures of whatever appropriate nature come first with attention turning to measures described in this protocol/process as soon as possible and practical.

This is a minimal protocol/process that may be exceeded at any time based on the judgment of the involved patient care personnel.

This protocol/process encourages high quality patient care but observing it cannot guarantee any specific patient outcome.

This protocol/process is subject to revision from time to time, as warranted by the evolution of technology and practice.

Review period: Review as changes occur or per institutional protocol.

Original hard copies and computer copies of this protocol are stored under the supervision of the Chief Perfusionist, Department of Cardiovascular Perfusion.

Documents relating to patient care standards are developed according to the accepted hospital standards.

**APPROVED BY:** *(signature of CMO and CNE only required)*

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| **Source:**  | *(originating department/committee)*  |
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