\*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 Standard 1 of AmSECT’s Standards and Guidelines for Perfusion Practice.1 These Standards and Guidelines may also be superseded by the judgement of the healthcare professional taking into account the facts and circumstances of the individual case.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** | **COLD AGGLUTININS** | |
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| **PURPOSE:** | To provide a guideline and resource on the diagnosis andtreatment of patients with cold agglutinins disease. | |
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| **TARGET POPULATION:** | Patients with cold agglutinins disease. | |
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| **DEFINITIONS:** | Cold agglutinins is an autoimmune disease characterized by the reaction of serum antibodies with antigens on the surface of red blood cells at cold temperatures.  Pathophysiology of the disease is temperature dependent but can occur at any temperature below 37 degrees Celsius.  Agglutination is dependent on two factors:  ● the concentration of the specific antibody  ● the temperature at which agglutination occurs  Once the blood temperature goes below the thermal amplitude, it causes activation of the autoantibodies. Once the antibodies are activated, they bind to red blood cells initiating the complement pathway of the coagulation cascade.  Agglutination and hemolysis can have catastrophic consequences on and off CPB including myocardial infarction, renal failure, hemolytic anemia, and thrombosis. | |
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**POLICY:**

1. Cold agglutinins is a very rare, yet very serious patient condition in which the perfusionist must make modifications to standard CPB practices in order to improve patient care.
2. Whether a diagnosis of cold agglutinins is made prior to surgery or the presence of agglutinins is observed unexpectedly during the case, the perfusionist should communicate his/her observations to the heart team and the surgeon will determine the plan of action.

**PROCEDURE:**

If the presence of cold agglutinins is determined preoperatively, then consider the following:

1. Careful intraoperative temperature monitoring to include core (bladder or rectal), nasopharyngeal, skin
   1. During CPB: arterial and venous blood, cardioplegia, myocardium.
2. Maintenance of patient temperature above activation thermal threshold
   1. Warm operating room
   2. Warm all fluids and anesthetic gases.
   3. Normothermic or mild hypothermia during CPB
3. Myocardial Protection Techniques:
   1. Cold crystalloid solution (see below)
   2. V-Fib with continuous perfusion
4. Warm Continuous Cardioplegia
5. In extreme cases:
   1. Preoperative plasma exchange (plasmapheresis)
   2. Total body washout and/or exchange transfusion

**Crystalloid Cardioplegia Delivery**: utilize a setup that allows switching between blood and crystalloid solution and hot/cold delivery options.

1. Prime cardioplegia circuit with crystalloid according to your Policy and Procedure
2. Isolate cardioplegia circuit to deliver only crystalloid.
3. Prime and de-bubble cardioplegia delivery system and bridge.
4. Administer 250-300 ml WARM high potassium CRYSTALLOID CARDIOPLEGIA to flush blood out of the coronaries.
5. Administer COLD high potassium CRYSTALLOID cardioplegia.
6. The total dosage of cardioplegia and use of topical ice/slush should be determined by surgeon protocol.
7. If bi-caval cannulation is utilized, the right atrium can be opened and to suction the cold high potassium crystalloid cardioplegia to the waste suction off the surgical field.
8. Administer subsequent doses of low potassium cold crystalloid cardioplegia according to surgeon protocol.
9. Prior to removal of the cross clamp, administer a dose of WARM CRYSTALLOID CARDIOPLEGIA, followed by a dose of WARM BLOOD ONLY CARDIOPLEGIA.
   1. Dosage according to surgeon protocol.

If the presence of cold agglutinins is *determined intraoperatively*, then;

1. Raise core temperature to normothermia.
2. Utilize warm retrograde cardioplegia to washout the myocardium.

**CLINICAL ASSESSMENT/SCREENING:**

1. Contraindications: None

# RELATED DOCUMENTS:

1. n/a

# REFERENCES:

1. Atkinson VP, Soeding P, Horne G, Tatoulis J. Cold Agglutinins in Cardiac Surgery: Management of Myocardial Protection and Cardiopulmonary Bypass. Ann Thorac Surg 2008;85:310–1.
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. Kypson AP, Warner JJ, Telen MJ, Milano CA. Paroxysmal cold hemoglobinuria and cardiopulmonary bypass. Ann Thorac Surg 2003;75:579-81.
4. Panos A, Murith N, Myers PO, Kalangos A. Aortic arch repair and cold-reactive agglutinins: what to do? Ann Thorac Surg. 2007;84(:1403-4.
5. Hoffman JW Jr, Gilbert TB, Hyder M. Cold agglutinins complicating repair of aortic dissection using cardiopulmonary bypass and hypothermic circulatory arrest: case report and review. Perfusion. 2002;17:391-4.

# 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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