\*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 Standard 1 of AmSECT’s Standards and Guidelines for Perfusion Practice.1 These Standards and Guidelines may also be superseded by the judgement of the healthcare professional taking into account the facts and circumstances of the individual case.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** | **MALIGNANT HYPERTHERMIA** | |
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| **PURPOSE:** | To provide a guideline and resource for a patient with a previouslydiagnosed risk or sudden onset of Malignant Hyperthermia. | |
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| **TARGET POPULATION:** | Patients with a previously diagnosed risk or sudden onset of Malignant Hyperthermia. | |
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| **DEFINITIONS:** | Malignant Hyperthermia (MH) is a rare (1:100,000) disorder, found most prevalent in the younger, male population involving the regulation of calcium in skeletal muscle.  MH is a dominantly inherited disorder of skeletal muscle that upon exposure to potent volatile anesthetics (halothane, isoflurane, sevoflurane, desflurane, etc.) and succinylcholine; may lead to a serious life threatening reaction.  Ryanodine Receptor gene (RYR1) – the mutation of this gene is responsible for MH. This gene alteration causes an increased release of calcium from the sarcoplasmic reticulum of the skeletal muscle.  This increased calcium binding to the myofilament of the muscle causes irregular muscle contraction i.e.; rigidity. It’s this rigidity that leads to a hypermetabolic state. (Hyperthermia, tachycardia, tachypnea, hypercarbia, increased oxygen consumption, acidosis, hyperkalemia, muscle rigidity.)  Dantrolene binds to the mutated R1Y1 gene to reduce the activity in the sodium channels. Dantrolene (Dantrium) is available in 20 mg vials that require 60 cc of sterile water to reconstitute. The average adult may require 8-10 ampules.  Ryanodex is an FDA approved alternative to Dantrolene, available in 250 mg ampoules. These ampoules require only 5 cc of sterile water for reconstitution. | |
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**POLICY:**

1. All cardiac team members will be responsible for the early detection of the signs and symptoms of MH whenever possible.
2. Diagnosis of MH is often difficult pre-op and intraop because many of these signs and symptoms are masked by the abnormality of physiology that the basis of CPB offers.
3. The earliest clinical clue may be an increased end tidal CO2 level, despite an increased minute ventilation.
4. In addition, the team members shall be aware of the policies and procedures for the sudden onset of a MH diagnosis.

**PERFUSION PUMP CONSIDERATIONS:**

In order to prepare to adequately manage a MH patient, each patient should have the following in use and/or available urgently:

1. In line ABG monitoring
2. Ability to create hypothermia on CPB -heater/cooler available and ready for use
3. Patient temperature management (core/nasopharyngeal) until the patient leaves the operating room
4. Backup oxygenator, supplies for changeout
5. MH cart location- each institution should have a designated cart with all necessary supplies for an MH emergency. (KNOW the location of this cart in your institution)
6. Hemoconcentrator and supplies for possible active filtering of K+

**PROCEDURE:**

1. Diagnosis: Signs of hypermetabolism
   1. Tachycardia (high HR)
   2. Hypercarbia (high PCO2)
   3. Acidosis (low pH)
   4. Hyperthermia (high temperature)
   5. Decreased oxygen saturation (low PaO2 despite 100% FIO2)
   6. Unexplained increase in end-tidal CO2
   7. Unexplained significantly high lactate level
   8. Hyperkalemia (high K)
2. Other signs and symptoms
   1. Generalized muscle rigidity/hypercontractility
   2. Myoglobinuria - the presence of myoglobin (oxygen reserve in muscle cells)in the urine, usually dark red/brown urine and is associated with rhabdomyolysis or muscle destruction.
   3. Hyperkalemia
   4. Elevated CK-MB (Creatine Phosphokinase- MB = found in myocardium; therefore, used to indicate myocardial cell wall injury) normal values = 3-5% or 5-25 IU/L
3. Management
   1. Notification of diagnosis – MH cart urgently into OR
   2. Discontinue volatile agents (Isoflurane) and succinylcholine
      1. \*Anesthesia may switch to propofol and an activated charcoal filter.
   3. Call MHAUS hotline (1-800-644-9397)
   4. Hyperventilate
   5. Anesthesia to give Dantrolene 2.5 mg/kg into IV or CPB pump
      1. \*Anesthesia may continue post op drip 24 hours
      2. 1 mg/kg every 4-8 hours, for 24-48 hours
   6. Obtain ABG asap
   7. Actively cool patient – turn off blanket warmers
   8. If serum potassium is >5.9 mEq/L
      1. Calcium chloride IV 10 mg/kg
      2. Sodium Bicarbonate IV 1-2 mEq/kg
      3. 10 units Insulin and 50 ml 50% Dextrose IV
      4. Check hourly glucose levels
   9. Treat Arrhythmias- Anesthesia may use Amiodarone or Lidocaine
      1. Do not use Calcium Channel Blockers
   10. Once in unit, patient blood samples can be sent to the laboratory for additional screening/testing/diagnosis.
   11. ECMO is considered to be a viable option for safe, emergent recovery from MH.
       1. Once on ECMO, CRRT may be used for normalization of electrolytes

**CLINICAL ASSESSMENT/SCREENING**:

1. Contraindications: None
2. Laboratory diagnosis: MH is an in vitro (sample taken outside a living organism) contracture test or a DNA test. These lab samples are not run during an acute/emergency event, which stresses the necessity to be aware of the typical signs and symptoms, for early detection and treatment.

# RELATED DOCUMENTS:

1. n/a

# REFERENCES:

1. Butala, B., Busada, M., &amp; Cormican, D. (2018). Malignant Hyperthermia: Review of Diagnosis and Treatment during Cardiac Surgery with Cardiopulmonary Bypass. Journal of Cardiothoracic and Vascular Anesthesia, 32(6), 2771–2779. doi: 10.1053/j.jvca.2018.03.029
2. Phillips, T., Zimmermann, E., Dukatz, C., Tully, M., Aziken, N., Raman, J., … Sera, V. (2020). Use of Cardiopulmonary Bypass for Treatment of Malignant Hyperthermia. Journal of Cardiothoracic and Vascular Anesthesia, 34(3), 753–755. doi: 10.1053/j.jvca.2019.11.010

# 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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