\*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 Standard 1 of AmSECT’s Standards and Guidelines for Perfusion Practice.1 These Standards and Guidelines may also be superseded by the judgement of the healthcare professional taking into account the facts and circumstances of the individual case.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** | **Antiphospholipid Syndrome (APS)** | |
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| **PURPOSE:** | To provide a guideline and resource for the diagnosis andtreatment of antiphospholipid syndrome. | |
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| **TARGET POPULATION:** | Patients with Antiphospholipid Syndrome. | |
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| **DEFINITIONS:** | A phospholipid is a type of lipid molecule that is the main component of the cell membrane. Phospholipids are very prevalent, found in all cell membranes, including red blood cells and the endothelial vessel lining.  Antiphospholipid syndrome (APS) is an autoimmune disorder wherein the body produces antibodies that attack phospholipids.  APS occurs in 1-5% of the population, with half of these patients suffering also from other additional autoimmune disorders.  APS is a pro-inflammatory, prothrombotic state. A typical “APS cycle” starts with antibodies attacking phospholipids which damage cells and vessel linings which cause clot formation in response to injury. This in turn furthers the attack on those phospholipids and creates a massive clot formation and vessel occlusion.  Severity of the complication depends on location of the thrombus and degree of obstruction in the vessel or organ.  Symptoms/presentation: During an APS “cycle”   * Thrombocytopenia is common because the APS antibodies cause destruction to the platelets. In addition, the platelet count will be low due to the normal usage during the clotting process. * Patients often present with PE, DVT, Stroke, or MI. Valve vegetation is also common because stress/turbulent flow around valve leaflets cause trauma, exposing the phospholipids and activating clot aggregation.   APS Triggers of Thrombotic events:   * High cholesterol, immobility (bed rest), smoking, or surgery (CPB) * 60% of APS patients have post-op complications such as valve restenosis, early graft occlusion, CVA, PE, Limb ischemia.   Complications:   * Catastrophic APS occurs when repeated clotting events occur in a short time due to multiple insults and can cause damage to multiple organs. * Catastrophic APS has a 50% mortality rate and is more common on CPB cases where it was previously undiagnosed and no special measures were taken. * Most APS patients are undiagnosed, since diagnosis takes two positive samples at least 12 weeks apart, which isn’t feasible in urgent cases. * Unfortunately, with APS, coagulation tests are misleading. The antiphospholipid antibodies bind to the phospholipid sites leaving few binding sites for the protein in these coagulation tests. This creates a false negative, reading normal or hypo-coagulable instead of the true coagulopathy. * Tests that do work: PTT, HepXa, Heparin concentration dose response | |
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**POLICY:**

1. The typical APS patient is already in a pro-thrombotic, pro-inflammatory state and contact activation from the circuit will further stimulate the clotting cascade and create the potential for catastrophic APS.
2. The surgeon should determine the proposed protocol with the APS patient and caregiver team before the case.

**PERFUSION PUMP CONSIDERATIONS:**

**Modifications** that can be made to decrease the risk catastrophic APS clotting on cardiopulmonary bypass include:

1. Supra-therapeutic dose of heparin.
   1. With Target ACT >800 sec
   2. Frequent ACT checks, every 20min
   3. Draw Hep Xa levels once heparinized (depending on speed of lab and length of case, will be able to confirm heparin levels)
2. No antifibrinolytics (Amicar, TXA)
3. Only quarter reversal of Protamine, or less. More can always be given, too much may further create a hypercoagulable state.
4. Care should be taken with cell salvage unit and field items (basin) – clots will rapidly form
5. APS patients often have thrombocytopenia, but care should be taken if administering platelets after bypass to not trigger a massive thrombotic event.

Additional options available:

1. Utilizing ATIII replacement therapy to increase the effectiveness of heparin dose
2. Using Bivalirudin or a direct thrombin inhibitor
3. Heparin based tubing coating
4. Using HMS Hepcon machine for HDR and to monitor heparin concentration in addition to standalone ACTs.
   1. Manual Heparin titration using serial dilutions and ACT measurements has also been reported effective (4)

**PROCEDURE:**

The strategy for the perfusionist should be aimed at minimizing hypercoagulable state.

1. Maintain ACT >800 sec
2. Frequent ACT checks, every 20min
3. Do not give antifibrinolytics (Amicar, TXA)
4. Administer/recommend quarter dose reversal of Protamine, or less
5. Cell salvage – close attention and high drip rate of anticoagulation – will clot quickly
6. Blood product administration – proceed with caution, potential to trigger a massive thrombotic event.

**CLINICAL ASSESSMENT/SCREENING:**

1. Contraindications: None

# RELATED DOCUMENTS:

1. Angiomax/Bivalirudin Clinical Protocol

# REFERENCES:

1. Gomez, J.A., Cervera, R. Diagnosis and Classification of the Antiphospholipid Syndrome. Journal of Autoimmunity. 2013; 48-49:20-25.
2. Cho H, Jeon Y, Hong DM, Kim HJ, Min JJ. Anesthetic Management of Antiphospholipid Syndrome Patients who underwent Cardiac Surgery. Korean Journal of Anesthesiology. 2014; 66(2):164-168.
3. Wilson WA, Gharavi AE, Koike T, Lockshin MD, Branch DW, Oiette JC, et al. International consensus statement on preliminary classification criteria for definite antiphospholipid syndrome: report of an international workshop. Arthritis Rheum. 1999; 42:1309-11.
4. Seki T, Shingu Y, Sugiki H, Wakasa S, Katoh H, Ooka T et al. Anticoagulation Management during Cardiopulmonary Bypass in Patients with Antiphospholipid Syndrome. Journal of Artificial Organs. 2018; (1):1-4.

# 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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