Fontan VAD Management Protocol

**BACKGROUND**

In select cases of failing Fontan physiology, VAD has been demonstrated to be effective as a form of circulatory support. Physiologic optimization of VAD parameters in this unique population is likely to require individualization. There is limited published literature on VAD support of Fontan patients and we propose these recommendations based on collective clinical experience.

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**OBJECTIVES**

1. Optimize physiologic support in post-VAD Fontan patients to include minimization of central venous pressures (CVP) and maximization of effective (non-aortopulmonary collateral) cardiac output.
2. Better define circulatory physiology in post-VAD Fontan patients.

**PROTOCOL**

**PRE-OPERATIVE CONSIDERATIONS**

1. Indications.
   1. VAD support can be considered for Fontan patients with:
      1. Signs and symptoms of heart failure or other signs of Fontan failure not responsive to medical management, *and at least one of the following*
         1. Poor systemic ventricular systolic function
         2. Poor systemic ventricular diastolic function
         3. Atrioventricular valve regurgitation
   2. The role of VAD support in individuals with isolated PVR elevation is unclear
2. Pre-VAD Assessment
   1. Cath:
      1. Consider pre-VAD cardiac catheterization to assess pressures, PVR, Fontan obstruction and presence/severity of shunts (ie Aorto-pulmonary (AP) collaterals)
      2. In some cases, transcatheter closure of AP collaterals may be considered and any significant anatomic obstructions should be addressed, recognizing that gradients may be underestimated in the setting of low flow states and poor cardiac output
      3. Perform PVR reactivity testing if concern for PVR elevation, to help inform pulmonary vasodilatory use
   2. Imaging:
      1. Ventricular function and distal anatomy may be incompletely characterized by echocardiography
      2. Consider cardiac MRI to quantify systolic function, volumes, obstruction, flow differential, and collateral burden as well as anatomic data to inform device placement
      3. In patients who cannot have a CMR, ECG gated CT angiography provides anatomical and some functional data
      4. MRI or CT can be used for 3D modeling and virtual fit
3. Multi-organ system assessment:
   1. Liver disease is not a contraindication to VAD support but extent of liver disease should be thoroughly assessed, including cross-sectional imaging (CT or MRI), assessments for varices and porto-systemic shunts, and evaluation for HCC. If available, obtaining a baseline elastography (ultrasound or MRI) allows for serial evaluations post-VAD with potential prognostic implications
   2. Renal disease may be underestimated by creatinine alone, and other methods for evaluation of renal function are recommended (such as Cystatin C, timed urine collection, or nuclear GFR)
   3. Consider pre-VAD head imaging and detailed neurologic exam.
   4. In patients who are able, consider obtaining baseline functional assessment with cardiopulmonary exercise testing or 6-minute walk
   5. In patients who are able, consider conducting a frailty assessment either using Fried criteria or the Essential Frailty Toolset
   6. Consider obtaining a formal nutritional evaluation

**SURGICAL CONSIDERATIONS**

Refer to separate *ACTION Harmonized Protocol on Patient and Device Selection*

**POST-OPERATIVE CONSIDERATIONS**

1. Post-op monitoring
   1. Lines: Optimizing blood flow through the Fontan circuit is critical, and requires in the first 3-5 days post-op:
      1. A reliable CVP catheter, *and either a* pulmonary arterial (PA) Swan-Ganz catheter, *or* an atrial line
      2. CVP line alone can be considered, especially if low concern for PVR issues.
   2. Monitor NIRS, UOP, and lactate closely in the first 24-48h post-op
   3. Trend mixed venous saturations
   4. If a CardioMems was previously implanted, it can be used to help guide postoperative management
2. Hemodynamic targets
   1. Cardiac index (CI): Recommend initial target of 3.5-4.5 L/min/m2 including both VAD and native output, to be titrated as needed to the filling pressures and hemodynamic requirements of the individual patient
      1. Patients’ native cardiac output will contribute a part of the total CI
      2. Higher CI (up to 6.5 L/min/m2) may be needed, especially in the presence of AP collaterals
      3. Target A-VO2 gradient <30%
   2. Blood pressure: May require higher target than other heart diseases post-VAD, as there may be detrimental physiologic changes which occur with excessive vasodilation. Depending on CVP (which typically is >10 mmHg with a Fontan circulation) doppler or mean arterial pressure targets of as high as 100-120 mmHg for continuous flow devices have been reported to be necessary to achieve adequate end-organ perfusion pressure (PP = MAP – CVP).
   3. CVP: Target CVP is based on a balance of decreasing systemic venous congestion while maintaining adequate VAD filling. Consider pulmonary vasodilators such as iNO in immediate post-operative period and sildenafil to help lower CVP.
3. Trouble-shooting: In cases of low cardiac output, consider the following:
   1. Inadequate preload

Causes:

* + 1. Volume status or bleeding
       1. Signs: Low CVP, low cardiac output, suction events, low flow alarm
       2. Treatment: Volume, hemostasis
    2. Elevated PVR
       1. Signs: Elevated CVP with low PCWP, hepatic congestion
       2. Treatment: consider pulmonary vasodilator therapy, fenestration creation, diuresis, optimize ventilation strategy
    3. Obstruction of pulmonary venous return
       1. Signs: Increased PCWP, increased pulmonary edema on CXR
          1. More frequently encountered with atrial cannulation/smaller patients
       2. Treatment: Surgical revision
  1. Tamponade

Causes:

* + 1. Pericardial effusion, tissue edema, oversized intracorporeal VAD
       1. Signs: Increased CVP, increased PCWP, decreased cardiac output
       2. Treatment: Volume resuscitation, opening chest. (Note: because TTE/TEE often inadequate for imaging, treatment of tamponade often requires proceeding with surgical intervention due to high index of suspicion without confirmatory imaging)
  1. Increased afterload

Causes:

* + 1. ↑SVR
       1. Signs: Decreased VAD flows, decreased power consumption, increased systemic blood pressure, increased pulsatility
       2. Treatment: Systemic vasodilator
    2. Thrombus: in either the inflow or outflow, obstructing flow into/out of the device
       1. Signs: uptrending power consumption and evidence of hemolysis
       2. Treatment: increase anticoagulation, thrombolytic therapy, or device change
  1. Ineffective Cardiac output

Causes:

* + 1. Excessive aortopulmonary collateral flow
       1. Signs: low Fick cardiac output or low mixed venous oxygen saturations (MVO2) with high VAD flows
       2. Treatment: cardiac catheterization for coiling, increase VAD speed
    2. Neo/Aortic Insufficiency
       1. Signs: low Fick cardiac output or low mixed venous oxygen saturations (MVO2) with high VAD flows
       2. TTE for assessment
       3. Treatment: Increase VAD flows typically will not overcome valve insufficiency; strongly consider surgical repair/replacement or catheter-based interventions (if thought amenable)
    3. Excessive Vasodilation
       1. Signs: end-organ hypoperfusion in the setting of elevated VAD-assessed cardiac output matched by Fick cardiac output. Consider milrinone accumulation (esp if impaired renal function), infection, vasoplegia
       2. Treatment: treat underlying etiology (ie, infection), vasopressin, methylene blue

1. Studies:
   1. Echocardiogram:
      1. Used to assess aortic valve opening, aortic and atrioventricular valve regurgitation, ventricular decompression, clots, and fenestration (if present) gradient
      2. Consider TTE in first 1-3 days post-op and weekly while on vasoactive or respiratory support
   2. Ramp Study (see attached worksheet): Using a ramp study to optimize VAD support can be considered. If performed, recommend using both hemodynamic (cath) and imaging (echo) assessments while VAD settings are titrated
      1. Indications for Ramp Study:
         1. Optimization recommended within 2 weeks post-op, 2-3 months postop or prior to discharge, and 6-12 months post-op
         2. Evidence of heart failure/elevated Fontan pressures, persistent symptoms, or any clinical deterioration
         3. Suspicion for device thrombus
         4. If Swan-Ganz catheter or atrial line in place, Ramp Study (with echo) recommended within 24 hrs post-op and with any change in clinical status
      2. Goals of Ramp Study:
         1. Decompression of ventricle and common atrium
         2. Minimize atrioventricular valve regurgitation (AVVR)
         3. No more than trivial aortic insufficiency
         4. Intermittent opening of the aortic valve
         5. Optimize Fontan pressures and PCWP
         6. Optimize AVO2 difference
      3. Safety:
         1. Ensure the patient is on therapeutic anticoagulation
         2. Ensure the ventricle and aortic root are free from thrombus
            1. Risk of thromboembolism with reduction in pump speed
         3. Allow ≥ 2 minute stabilization between speed changes
            1. When decreasing RPMs: monitor for increasing AVVR, increasing aortic valve opening (AoV), increases in Fontan pressures and PCWP, cyanosis (if fenestration). and any symptoms
            2. When increasing RPMs: monitor for impendence of flow into inflow cannula, changes in Fontan pressures, cyanosis (if fenestration), AoV not opening, increase in AI, suction events, and any symptoms
         4. Test endpoints: completion of test/desired outcome attained, suction event, hypotension, hypertension, increased cyanosis, symptoms
      4. Echocardiography during Ramp Study, suggested views (adapted from *ACTION Harmonized Protocol on Echocardiography for CF-VADs*, refer to this protocol for images):
         1. PLAX (2D, 3 beats): Ventricle internal diameter in diastole x3 beats
         2. PLAX of PSX (M-mode, 10 beats): Aortic valve opening (out of 10 beats)
         3. PLAX (Color, 3 beats): degree of AI
         4. PLAX or A4C (Color, 3 beats): degree of AVVR
         5. PSAX (2D, 3 beats): function
         6. A4C (2D, 3 beats): function
         7. PLAX, PSAX or A4C (Color, 3 beats): degree of AVVR
         8. A4C or PLAX (2D, Color, PW, CW): inflow cannula
         9. A4C or best view of fenestration if present (2D, PW): fenestration gradient
   3. Cardiac catheterizations:
      1. Recommended as part of Ramp Study as above (at 2 weeks post-op, 2-3 months postop or prior to discharge, and 6-12 months post-op).
      2. Consider assessing for and addressing AP collaterals, especially if elevated wedge/end-diastolic pressures with evidence of organ hypoperfusion and high VAD output due to AP collaterals based on above assessment.
      3. Consider placement of an implantable pulmonary arterial pressure monitoring device to guide diuretic and VAD management based on findings during in-house RAMP studies.

**PARA-CORPOREAL DEVICE CONSIDERATIONS**

The above guidelines generally refer to patients with intracorporeal CF devices, though many of the same principles apply to smaller Fontan patients with a para-corporeal device.

1. Fontan patients typically have high CI needs and larger Berlin pumps are required than in patients with biventricular physiology
2. Some centers report starting with a CentriMag (with Berlin cannulas) to determine cardiac output needs and then converting to a Berlin pump
3. Berlin Heart: Blood flow through the Fontan is continuous and if supported with a pulsatile pump, there is no flow into the pump during pump systole. Therefore, to ensure adequate unloading and to minimize atrial/pulmonary venous hypertension, consider targeting ~75% fill and shortening the percent of time in pump systole.

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***Disclaimer:*** *The ACTION network is focused on quality improvement efforts such as harmonizing best practice protocols, disseminating them among institutions, and helping centers to improve care practices at the local level. This protocol was developed as a consensus tool for pediatric VAD programs. The information in the protocols are based on center practices, individual opinions, experiences, and, where available, published literature. Centers may choose to adapt this protocol to include in their center-specific protocols with reference to ACTION with the understanding that these are meant as guidelines and not standard of care. (Revised: 06/09/2021)*

