Pulmonic Ventricular Dysfunction
Adult with Congenital Heart Disease

Louise Callow, RN, MSN, CPNP;
University of Michigan, CS Mott Children's Hospital

Introduction

- There are many potential causes of pulmonic ventricular dysfunction in the adult with congenital heart disease (CHD).
- Ventricular dysfunction requiring intensive care management typically reflects an acute decompensation from the patient's baseline or a postoperative state.
- Pulmonic ventricular dysfunction is more likely to be chronic in onset related to disease progression over decades or long term complications from previous corrective or palliative intervention.

Lesions that may present with pulmonic ventricular failure

- Right ventricular outflow tract obstruction
    - Tetralogy of Fallot
    - Pulmonary atresia/intact ventricular septum with sinusoids and coronary dependent circulation
    - Pulmonary atresia/ventricular septal defect (VSD) with or without multiple aortopulmonary collaterals (MAPPCA's)
    - Supravalvar pulmonary stenosis
    - Right ventricle (RV) to pulmonary artery (PA) conduit stenosis
    - Valvar pulmonary stenosis
    - Infundibular stenosis, double chamber right ventricle
- Pulmonary regurgitation
    - Tetralogy of Fallot s/p repair with transannular patch
    - Failure of RV-PA conduit with homograft
    - S/P balloon or surgical relief of PV stenosis with resultant PVR
    - Tetralogy of Fallot with absent pulmonary valve
    - Endocarditis of previously competent valve
- Right ventricular volume/pressure overload
    - Ventricular septal defect (VSD)
    - Atrial septal defect (ASD)
    - Patent ductus arteriosus (PDA)
    - Atrioventricular septal defect (AVSD)
    - Moderate to severe pulmonary regurgitation
    - Large aortopulmonary shunt such as Pott's or Waterston anastomosis
- Pulmonary artery hypertension (See Adult Guidelines on Pulmonary Hypertension and Eisenmenger Syndrome)
    - Eisenmenger Syndrome
    - Pulmonary vascular obstructive disease (PVOD)
    - Idiopathic pulmonary hypertension (IPHTN)
    - Thromboembolic event

    - Mitral stenosis/cortriatiatum
    - Severe pulmonary vein obstruction
- Coronary artery injury/occlusion
    - Anomalous right coronary artery in tetralogy of Fallot
    - Coronary artery sinusoids in pulmonary atresia/intact ventricular septum (PA/IVS)
    - Ischemic heart disease as comorbidity
    - Anomalous origin of the left coronary artery
    - Stenosis of coronary artery/coronary ostia s/p arterial switch operation
- Trauma
    - RV trauma/contusion
    - Adult respiratory distress syndrome (ARDS)
- Tricuspid regurgitation
    - Ebstein's anomaly (See Defect Guideline on Ebstein's Anomaly)
    - Injury to the tricuspid valve during surgical repair of VSD, AVSD
    - Endocarditis
- Diastolic dysfunction
    - Prolonged volume overload from aortopulmonary shunt or pulmonary regurgitation
    - Hypertrophied RV from longstanding right ventricular outflow tract obstruction (RVOTO)
    - Myopathy

Critical Thinking

- Assessment related to underlying congenital heart lesion
    - Repaired or palliated defect
        - Residual defect at time of repair
        - Complication at time of repair
        - Volume/pressure overload from unrepaired shunt
        - Pressure overload from unrelieved stenosis
        - Degree of muscle resection needed for repair
        - Need for ventriculotomy for repair
- Long term complications of congenital heart lesion
    - Progression of any disease state
        - Conduit stenosis/regurgitation
        - Progressive PR/RV dilation
        - PVOD
        - Occlusion/stenosis of graft/PA's/stents
        - Baffle leak
        - Progression tricuspid regurgitation with repair AVSD
    - Thromboembolic event
        - Clotting disorder
        - Septic emboli
    - Arrhythmia (See Adult Guidelines for Arrhythmias in ACHD)
        - Atrial flutter/fib
        - Ventricular tachycardia/fibrillation

        - Sick sinus syndrome
        - Heart block
    - Infectious complication
        - Endocarditis
        - Myocarditis
    - Co-morbidities
        - Diabetes
        - Coronary artery disease
        - Syndromes
            - Trisomy 21
            - DiGeorge
            - CHARGE
            - VACTERL
            - Turner's
            - Noonan's
            - William's
        - Pulmonary disease
            - BiPAP
            - Home oxygen therapy
            - Inhalation treatment
            - Smoking history/COPD
            - Reactive airway disease
            - Obesity
            - Substance abuse
            - Connective tissue disorders
            - Metabolic diseases
            - Neuromuscular disorders
            - Cystic fibrosis
    - Mechanical Causes
        - Pneumothorax
        - Hemothorax
        - Effusion
        - Atelectasis
        - Pneumonia
        - Side effect medications such as amiodarone

Clinical Assessment

- Early recognition with ongoing and continual reassessment for response to therapy key to good outcome
- General Appearance
    - Color
    - Perfusion
    - Glasgow Coma Scale/ Level of Consciousness
        - Pain and sedation scales
    - Jugular venous distension (JVD)

    - Peripheral edema
- Vital signs
    - Heart rate/rhythm
        - Extra heart sounds
        - Murmur, especially new murmur
    - Blood pressure in relation to pulmonary artery pressure
    - Pulse oximetry, end tidal CO2
    - Respiratory rate
        - Work of breathing
        - Bilateral breath sounds
        - Position of comfort
    - Filling pressures
        - Pulmonary artery (PA) and relationship to arterial pressure
        - Left atrial (LA) and transpulmonary gradient (RA-CVP minus LA)
        - Central venous pressure (CVP) for RV filling and diastolic compliance in absence of tricuspid regurgitation
        - Temperature
        - Urine output
    - End organ assessment
        - Abdomen
            - Bowel
            - Organomegaly
        - Neurologic
        - Renal
        - Exercise tolerance

Diagnostic Evaluation

- Noninvasive testing
    - Physical examination
    - Chest x-ray
    - EKG/pacemaker/ICD interrogation
    - Exercise stress test
    - Echocardiogram
    - Pulmonary function tests
    - ETCO2
    - Pulse oximetry
- Laboratory testing
    - Serum electrolytes
    - Hepatic function
    - Kidney function
    - Coagulation studies
    - BNP/troponin
    - CBC/platelets
    - Serum lactateSVO2
- Imaging
    - Cardiac catheterization
    - CT/CT angiogram

- MRI/MRA
- Lung-perfusion scan

Treatment

- Residual lesion
  - Surgical intervention for progression of valvar regurgitation or stenosis
  - Surgical intervention for residual shunt or baffle leak
  - Surgical intervention for prosthetic valve or conduit stenosis
  - Surgical intervention for complication of previous surgery i.e. coronary stenosis with arterial switch operation
  - Supportive care
    - Fever
    - Pain/sedation
- Arrhythmia (See both Adult and Peds/Neo Arrhythmia Guidelines)
  - Antiarrhythmic medications
  - Atrial pacing and atrial antitachycardia pacemaker
  - Pacemaker implantation
  - ICD
  - Biventricular pacing
  - Cardioversion
  - Radio frequency/cryoablation surgical or interventional cardiology
  - Electrolyte replacement
- Preload
  - Diuretics
  - Volume replacement
    - Choice of volume based on baseline HCT, PaO2, SVO2
    - Avoid underfilling of hypertrophied ventricle or single ventricle physiology
    - Avoidance of positive pressure ventilation if possible
- Pulmonary antihypertensives (afterload reducers)
  - Endothelin receptor antagonist
    - Bosentan
    - Ambrisentan
  - Phosphodiesterase 5 inhibitors
    - Sildenafil
    - Tadalafil
  - Prostacylins
    - Treprostinil (Remodulin)
    - Epoprostanil (Flolan)
  - Nitric oxide
  - Alveolar hypoxemia
    - Oxygen
    - Hematocrit
    - Maintain functional residual capacity
- Ventricular contractility
  - Medications

        - Pulmonary vasodilators
        - Vasoactive agents
    - Reverse acidosis
    - Assure electrolytes in normal range and replace if not
    - Assist device (AD)
        - RVAD/LVAD
        - Extracorporeal membrane oxygenation (ECMO) (See Peds/Neo VA ECMO Guidelines)
- Transplant
    - Heart
    - Lung
    - Heart and lung
    - Special considerations
        - Surgical reconstruction previously performed and issues with reconstructive transplant surgery
        - Antibody formation from multiple transfusions earlier in life
        - Collateral formation from long term cyanosis, repeated operations or chest tube insertions

Associated Complications

- Anticoagulation
    - Thromboembolic event
    - Bleeding risk/hemorrhage
- Neurologic
    - Stroke
    - Seizures
- Renal
    - Electrolyte abnormalities
- Nutrition
    - Increased demand, decreased intake
- Endothelial dysfunction

Special considerations

- Single ventricle physiology
    - Fontan
        - Protein losing enteropathy (PLE*)*
          A complication following the Fontan procedure where protein is lost in the gut, manifesting as ascites, peripheral edema, pleural and pericardial effusions. It is exacerbated by high systemic venous pressures. (Gatzoulis et al., 2003).
        - Plastic bronchitis
          A rare, serious complication; noninflammatory, mucinous casts formed in the trachea and bronchi that can produce airway obstruction and asphyxia; may be related to unfavorable post-Fontan hemodynamics with increased CVP and a fragile lymphatic system. (Gatzoulis et al., 2003).

    - Systemic right ventricle
        - Anatomic variations of LV versus RV
        - Anatomic variations of pulmonary and tricuspid valve (PV and TV)
- Cyanotic heart disease (See Adult Guidelines on Eisenmenger Syndrome)
    - Palliated congenital heart disease unable to move to repair
        - Patient's with aorto-pulmonary shunts
        - Patient's with bidirectional Glenn or hemiFontan
        - Palliative Mustard/Senning
    - AV malformations
        - Pulmonary
    - Effects of hypoxemia
        - Polycythemia
        - Platelet dysfunction
        - Elevated PVR
        - Brain abscess
        - Stroke
        - Thromboembolic phenomena

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Reviewed/Updated
8/2015
L. Callow