

# **Pulmonic Ventricular Dysfunction Adult with Congenital Heart Disease**

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- I. Introduction
  - A. There are many potential causes of pulmonic ventricular dysfunction in the adult with congenital heart disease (CHD).
  - B. Ventricular dysfunction requiring intensive care management typically reflects an acute decompensation from the patient's baseline or a postoperative state.
  - C. Pulmonic ventricular dysfunction is more likely to be chronic in onset related to disease progression over decades or long term complications from previous reparative or palliative intervention.
  
- II. Lesions that may present with pulmonic ventricular failure
  - A. Right ventricular outflow tract obstruction
    - 1. Tetralogy of Fallot
    - 2. Pulmonary atresia/intact ventricular septum with sinusoids
    - 3. Pulmonary atresia/ventricular septal defect (VSD)
    - 4. Supravalvar pulmonic stenosis
    - 5. Right ventricle (RV) to pulmonary artery (PA) conduit stenosis
  - B. Pulmonary regurgitation
    - 1. Tetralogy of Fallot s/p repair with transannular patch
    - 2. Failure of pulmonary valve regurgitation (PVR)/RV-PA conduit with homograft
  - C. Right ventricular volume/pressure overload
    - 1. Ventricular septal defect (VSD)
    - 2. Atrial septal defect (ASD)
    - 3. Patent ductus arteriosus (PDA)
    - 4. Atrioventricular septal defect (AVSD)
  - D. Pulmonary artery hypertension
    - 1. Eisenmenger's
    - 2. Pulmonary vascular obstructive disease (PVOD)
    - 3. Idiopathic pulmonary hypertension (IPHTN)
    - 4. Thromboembolic event
    - 5. Mitral stenosis/cor triatiatum
    - 6. Severe pulmonary vein obstruction
  - E. Coronary artery injury/occlusion
    - 1. Anomalous right coronary artery in tetralogy of Fallot
    - 2. Coronary artery sinusoids in pulmonary atresia/intact ventricular septum (PA/IVS)
    - 3. Ischemic heart disease as comorbidity
  - F. Trauma

1. RV trauma/contusion
  2. Adult respiratory distress syndrome (ARDS)
  - G. Tricuspid regurgitation
    1. Ebstein's anomaly
  - H. Diastolic dysfunction
    1. Prolonged volume overload from shunt
    2. Hypertrophied RV from longstanding right ventricular outflow tract obstruction (RVOTO)
    3. Myopathy
- III. Critical Thinking
- A. Assessment related to underlying congenital heart lesion
    1. Repaired or palliated defect
      - a. Residual defect at time of repair
      - b. Complication at time of repair
      - c. Volume/pressure overload from unrepaired shunt
      - d. Pressure overload from unrelieved stenosis
      - e. Degree of muscles resection needed for repair
      - f. Need for ventriculotomy for repair
  - B. Long term complications of congenital heart lesion
    1. Progression of any disease state
      - a. Conduit stenosis/regurgitation
      - b. Progressive PR/RV dilation
      - c. PVOD
      - d. Occlusion/stenosis of graft/PA's/stents
      - e. Baffle leak
    2. Thromboembolic event
    3. Arrhythmia
      - a. Atrial flutter/fib
      - b. Ventricular tachycardia/fibrillation
      - c. Sick sinus syndrome
      - d. Heart block
    4. Infectious complication
      - a. Endocarditis
    5. Co-morbidities
      - a. Diabetes
      - b. Coronary artery disease
      - c. Syndromes
        - i. Trisomy 21
        - ii. DiGeorge
        - iii. CHARGE
        - iv. VACTERL
        - v. Turner's
      - d. Pulmonary disease
        - i. Bipap
        - ii. Home oxygen therapy
        - iii. Inhalation treatment

- e. Smoking history
    - f. Reactive airway disease
    - g. Obesity
    - h. Substance abuse
    - i. Connective tissue disorders
    - j. Metabolic diseases
    - k. Neuromuscular disorders
  - C. Mechanical Causes
    - 1. Pneumothorax
    - 2. Hemothorax
    - 3. Effusion
    - 4. Atelectasis
    - 5. Pneumonia
- IV. Clinical Assessment
  - A. General Appearance
    - 1. Early recognition with ongoing and continual reassessment for response to therapy of subtle changes key
    - 2. Color
    - 3. Perfusion
    - 4. Glasgow Coma Scale/ Level of Consciousness
      - a. Pain and sedation scales
    - 5. Jugular venous distension (JVD)
    - 6. Peripheral edema
  - B. Vital signs
    - 1. Heart rate/rhythm
      - a. Extra heart sounds
      - b. Murmur
    - 2. Blood pressure in relation to pulmonary artery pressure if line present
    - 3. Pulse oximetry
    - 4. Respiratory rate
      - a. Work of breathing
      - b. Bilateral breath sounds
  - C. Filling pressures
    - 1. Pulmonary artery (PA) and relationship to arterial pressure
    - 2. Left atrial (LA) and transpulmonary gradient (RA-CVP minus LA)
    - 3. Central venous pressure (CVP) for RV filling and diastolic compliance in absence of tricuspid regurgitation
    - 4. Temperature
    - 5. Urine output
  - D. End organ assessment
    - 1. Abdomen
      - a. Bowel
      - b. Organomegaly
    - 2. Neurologic
    - 3. Renal
- V. Diagnostic Evaluation

- A. Noninvasive testing
    - 6. Physical examination
    - 7. Chest x-ray
    - 8. EKG/pacemaker interrogation
    - 9. Stress test
    - 10. Echocardiogram
    - 11. Pulmonary function tests
    - 12. ETCO<sub>2</sub> and SVO<sub>2</sub>
    - 13. Pulse oximetry
  - B. Laboratory testing
    - 1. Electrolytes
    - 2. Hepatic function
    - 3. Kidney function
    - 4. Coagulation studies
    - 5. BNP
    - 6. CBC/platelets
    - 7. Serum lactate
  - C. Imaging
    - 1. Cardiac catheterization
    - 2. CT
    - 3. MRI/MRA
    - 4. Lung-perfusion scan
- VI. Treatment
- A. Residual lesion
    - 1. Surgical intervention for progression of valvar regurgitation or stenosis
    - 2. Surgical intervention for residual shunt or baffle leak
    - 3. Surgical intervention for prosthetic valve or conduit stenosis
    - 4. Supportive care
      - a. Fever
      - b. Pain/sedation
  - B. Arrhythmia
    - 1. Antiarrhythmic medications
    - 2. Atrial pacing and atrial antitachycardia pacemaker
    - 3. ICD
    - 4. Biventricular pacing
    - 5. Cardioversion
    - 6. Ablation
    - 7. Electrolyte replacement
  - C. Preload
    - 1. Diuretics
    - 2. Volume replacement
      - a. Choice of volume based on baseline HCT and paO<sub>2</sub>
      - b. Avoid underfilling of hypertrophied ventricle or single ventricle physiology
  - D. Pulmonary antihypertensives (afterload reducers)
    - 1. Medications

- a. Bosentan
      - b. Remodulin
      - c. Prostacyclins
      - d. Tadalafil
      - e. Sildenafil
      - f. Cialis
      - g. Nitric oxide
      - h. Flolan
    - 2. Alveolar hypoxemia
      - a. Oxygen
      - b. Hematocrit
  - E. Ventricular contractility
    - 1. Medications
    - 2. Reverse acidosis
    - 3. Assist device (AD)
      - c. RVAD/LVAD
      - d. Extracorporeal membrane oxygenation (ECMO)
  - F. Transplant
    - 1. Heart
    - 2. Lung
    - 3. Heart and lung
    - 4. Special considerations
      - b. Surgical reconstruction preformed and issues with transplant surgery
      - c. Antibody formation from multiple transfusions earlier in life
      - d. Collateral formation from long term cyanosis, repeated operations or chest tube insertions
- VII. Associated Complications
- A. Anticoagulation
    - 1. Thromboembolic event
    - 2. Bleeding risk/hemorrhage
  - B. Neurologic
    - 1. Stroke
    - 2. Seizures
  - C. Renal
    - 1. Electrolyte abnormalities
  - D. Nutrition
    - 1. Increased demand, decreased intake
  - E. Endothelial dysfunction
- VIII. Special considerations
- A. Single ventricle physiology
    - 1. Fontan
      - a. 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).*

- b. 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).*
2. Systemic right ventricle
  - a. Anatomic variations of LV versus RV
  - b. Anatomic variations of aortic valve (AV)
- B. Cyanotic heart disease
- 1. Palliated congenital heart disease unable to move to repair
    - a. Patient's with aorto-pulmonary shunts
    - b. Patient's with bidirectional Glenn
  - 2. AV malformations
  - 3. Effects of hypoxemia
    - a. Polycythemia
    - b. Platelet dysfunction
    - c. Elevated PVR
    - d. Brain abscess
    - e. Stroke
    - f. Thromboembolic phenomena

**References:**

Gatzoulis MA et al. (2003) *Diagnosis and Management of Adult Congenital Heart Disease*. Churchill Livingstone.

Furhman BP, Zimmerman JJ: *Pediatric Critical Care*, ed, 3, St. Louis, 2006, Mosby.

Mavroudis C and Backer CL: *Pediatric Cardiac Surgery*, ed, 3, St. Louis, 2003, Mosby.

Wheeler DS, editor: *Pediatric Critical Care medicine: Basic Science and Clinical Evidence*, 2008, Springer-Verlag.

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