Curricular Components That Support the Functions of EPA 4: Diagnose, Initially Manage, and Refer Children with Advanced or End Stage Heart Failure and/or Pulmonary Hypertension to Experts for Medical Therapy, ECMO, Ventricular Assist Devices, and/or Cardiac Transplantation

1. Performing a comprehensive evaluation for the etiologies of heart failure and pulmonary hypertension, including clinical and laboratory assessment and noninvasive, invasive, and genetic testing
   - Performs the initial evaluation and management of the child with PH in the outpatient ambulatory setting
   - Performs the initial evaluation and stabilization of the hemodynamically compromised patient with PH
   - Knows the indications, risks, and benefits of medications used for the treatment of PH
   - Demonstrates knowledge of the indications and appropriate timing of referral to a dedicated specialist in pediatric or adult PH for advanced care and the evaluation and treatment of the ambulatory child with AHF
   - Provides the initial evaluation and stabilization of the hemodynamically compromised patient with heart failure
   - Administers medications approved or commonly used for the treatment of heart failure
   - Refers to dedicated subspecialist for consideration of advanced management options including mechanical circulatory support and transplantation evaluation
   - Co-manages cardiology care with a specialized center of a recipient who has undergone transplantation
   - Demonstrates knowledge of the patterns of heritable cardiomyopathies and genetic tests relevant to cardiomyopathies

2. Demonstrating expertise in the interpretation of hemodynamic testing and the implications for therapeutic intervention
   - Pulmonary Hypertension
     - Discriminates between normal and abnormal pulmonary vascular physiology, including the “neonatal transition” in pulmonary vascular resistance
     - Applies knowledge of the distinction between pulmonary hypertension and elevated pulmonary vascular resistance (i.e., hypertensive pulmonary vascular disease) in the care of patients
     - Demonstrates knowledge of pulmonary vascular pathophysiology, including the physiological and clinical meaning of “reactivity” to vasodilators
     - Demonstrates knowledge of the indications/contraindications for repair of congenital cardiac lesions in the presence of pulmonary vascular disease
     - Performs key components of an initial evaluation of PH, utilizing appropriate noninvasive and invasive imaging to discriminate and evaluate multiple etiologies, including: persistent pulmonary
hypertension of the newborn, congenital heart disease (including Eisenmenger physiology), PH associated with left heart disease, PH associated with chronic lung disease, and idiopathic PAH

- Demonstrates knowledge about the indications, risks, benefits, and outcomes of lung or heart lung transplantation in children with PH
- Demonstrates the skills to plan an appropriate referral for consultation with a dedicated specialist in pediatric or adult PH
- Demonstrates the skills to counsel families regarding the acute and chronic care of the child with pulmonary hypertension

- Heart Failure
  - Demonstrates knowledge of and recognizes the etiology, signs, and symptoms of heart failure in children
  - Evaluates diagnosis utilizing appropriate noninvasive and invasive imaging and genetic, metabolic, and electrophysiological testing and provides initial treatment for pediatric patients with heart failure of various etiologies
  - Demonstrates knowledge and use of the indications, mechanism of actions, risks, and benefits associated with the FDA-approved medications for use in adults with heart failure and medications commonly used in pediatric patients with heart failure
  - Plans appropriate consultation with a dedicated specialist in pediatric advanced heart failure therapies

3. Initiating target treatment for heart failure and for pulmonary hypertension
   - Demonstrates knowledge of the Consensus Guidelines for the Treatment of Heart Failure (American Heart Association [AHA] and International Society of Heart and Lung Transplantation [ISHLT])
   - Demonstrates knowledge about the incidence of the common complications associated with immunosuppressive medications used in patients following heart transplantation
   - Demonstrates knowledge about the use of FDA-approved medications in adults with PAH and medications commonly used in pediatric patients with PAH, including understanding of important drug interactions and adverse effects of medications

4. Knowing when to refer these patients with disease refractory to medical therapy to heart failure or pulmonary hypertension experts for advanced therapies for disease refractory to medical therapy, including ECMO, assist devices, and heart and/or lung transplantation
   - Demonstrates knowledge of indications, risks, and benefits of advanced heart failure therapies
   - Knows limitations and searches the literature for the highest grade of evidence available
   - Applies the evidence to and knows when to refer children with advanced or end stage heart failure and/or pulmonary hypertension to experts for medical therapy, ECMO, ventricular assist device, and/or cardiac
transplantation

5. Counseling patient and families regarding prognosis and treatment options
   • Establishes the level of understanding of the patient and family about the disease process
   • Engages the patient, family, and team members in shared decision making
   • Engages with the patient/family in deciding on the right time to refer to a subspecialist for a second opinion and further counseling

6. Coordinating multidisciplinary care with other subspecialties such as neonatal or critical care, cardiothoracic surgery, genetics, respiratory care, nursing, social work, and child life
   • Communicates with colleagues in multiple disciplines to coordinate care
   • Works in interprofessional teams to improve patient care and quality of life
   • Provides access to resources or other team members who can access resources for the patient/family

7. Participating in the cardiac care of a patient before, during, and after transplantation
   • Co-manages the patient along with the consultant
   • Opens a dialogue with the consultant that allows for a bidirectional exchange of information after each patient encounter

8. Managing medications and social challenges of medication compliance in outpatient follow-up of patients
   • Discusses medication check list with patients
   • Assesses medication noncompliance (social challenge)
   • Gauges health literacy of patients/families and counsels accordingly to help them take control over aspects of their disease such as medication adherence
   • Engages members of the interdisciplinary team to help with medication adherence

Problems that generally require consultation where the role of the general cardiologist is to recognize, provide preliminary evaluation, and refer. This list depends greatly on context in which one practices. Those generalists practicing in areas where access to subspecialists is difficult will likely provide more of the care and may do so with telephone advice from a trusted colleague.

   • Children with Inotropic dependency (HF)
   • Children with heart failure who require mechanical ventilation (HF)
   • Children with inotropic dependency +/- organ dysfunction (renal, liver) who might require mechanical support (HF)
   • Patients requiring more than one inotrope (HF)
• Newborns and infants with diaphragmatic hernia (PHN)
• Newborns and infants with persistent pulmonary hypertension (PHN)
• Children with Eisenmengers (PHN)
• Infants and children who may need pulmonary hypertension medications (pulmonary vasodilator medication, not including nitric oxide) (PHN)

Curricular Components Authors

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