<table>
<thead>
<tr>
<th>1. <strong>EPA Title</strong></th>
<th>Diagnosis and management of patients with congenital or acquired cardiac problems</th>
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<td>2. <strong>Description of the Activity</strong></td>
<td>Pediatric cardiologists, completing training, are expected to be able to diagnose and manage a broad range of congenital and acquired cardiac problems. These may include but are not limited to cyanotic congenital heart disease (CHD) in the newborn, left to right shunt lesions; such as, atrial septal defect (ASD), ventricular septal defect (VSD), patent ductus arteriosus (PDA), outflow obstruction lesions (e.g. aortic stenosis, pulmonary stenosis, coarctation), cardiomyopathies, Kawasaki Disease, dyslipidemia and cardiac manifestation of genetic syndromes (e.g. Down, Marfan, Turner, Noonan, Williams, DiGeorge).</td>
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The specific functions which define this EPA include:

- Knowing and understanding the natural and unnatural history, as well as unique cardiovascular anatomy and physiology seen in CHD
- Obtaining a complete history and physical examination
- Obtaining a thorough family history with a focus on genetic abnormalities associated with an important risk of heart disease in childhood
- Developing a prioritized differential diagnosis
- Knowing the risks and benefits of non-invasive and invasive evaluation of infants, children and young adults with congenital or acquired cardiovascular disease and applying these when ordering diagnostic testing in a cost effective manner
- Developing a management plan that incorporates medical therapy, interventional catheter procedures, and surgical intervention as well as addressing the psychosocial aspects of acute chronic disease.
- Communicating with and counseling the patient/family regarding immediate, mid- and long-term management
- Evaluating and managing acquired cardiac conditions found in pediatric patients
- Identifying and applying key evidence-based guidelines
- Recognizing cardiac conditions and indications that might require additional sub-specialty expertise
3. Judicious mapping to domains of competence

- Patient Care
- Medical Knowledge
- Practice-based Learning and Improvement
- Interpersonal & Communication Skills
- Professionalism
- Systems-based Practice
- Personal and Professional Development

4. Competencies within each domain critical to entrustment decisions

- PC 1: Gathering information
- PC 5: Performing complete physical examination
- PC 6: Using optimal clinical judgment
- PC 9: Counseling patients and families
- MK 1: Demonstrating knowledge
- ICS 1: Communicating with patients/families
- ICS 4: Working as a member of a health care team

5. Curricular components that support the functions of the EPA (knowledge, skills and attitudes needed to execute this EPA safely):

Rationale: The fundamental goal of clinical pediatric cardiology training is to acquire the diagnostic skills necessary to provide optimal inpatient, outpatient and consultative care to the fetus, infant, child and young adult with congenital or acquired cardiovascular disease.

Scope of practice: Diagnosis and management of congenital and acquired pediatric cardiovascular disease can occur in a variety of clinical settings, including the outpatient clinic, the inpatient wards, the newborn nursery, and the neonatal, pediatric, and cardiovascular intensive care units. The fundamental skills of history taking and physical examination form the basis for correctly initiating diagnostic assessment and determining management options appropriate for the individual patient. Knowledge of cardiac anatomy and physiology, the natural history of untreated and treated congenital and acquired cardiovascular disease, the risks and benefits of standard diagnostic tests, as well as current evidence-based guidelines for management of congenital or acquired cardiovascular disease, all establish the foundation for optimal patient care.

The curricular components listed in this document adhere to current guidelines, such as those listed in the reference section below. Furthermore, there is the expectation of continued self-directed learning toward ongoing advances in diagnosis and treatment.

Setting: Diagnosis and management in the following settings: inpatient, outpatient, consultation, routine and acute/emergent or intensive care environment.

Patient population: fetus, infant, child, adolescent, and adult

Curricular Components that support the functions of the EPA:

Knowing and understanding the natural and unnatural history as well as the unique cardiovascular anatomy and physiology seen in CHD

• Demonstrates knowledge of the pathophysiology and natural history of left to right shunt lesions, cyanotic CHD, valve abnormalities and other congenital systemic and arterial vascular abnormalities.

• Demonstrates knowledge of diagnosis, care and management of critical CHD

• Knows the different congenital coronary artery abnormalities, including pathophysiology and modes of presentation of congenital coronary artery abnormalities.

Obtaining a complete history and physical examination

• Demonstrates skills for obtaining a complete, age-specific history.

• Knows normal age-related vital sign values, including pulse, respirations, expected values for four-extremity blood pressures and pre- and post-ductal oxygen saturations.

• Performs a successful and complete cardiac physical evaluation, including auscultation in infants, toddlers, children and young adults.
  • Accurately describes the location and intensity of cardiac sounds, clicks, snaps, murmurs, rubs and gallops.
  • Differentiates stridor, wheeze, rales and rhonchi on auscultation of lungs.

• Recognizes abnormalities of cardiac performance by palpation of precordium, suprasternal notch, abdomen and peripheral pulses.

• Recognizes and differentiates infants, children and young adults with primary cardiac disease from those with cardiac disease as part of a complex systemic disease process.

• Recognizes signs of respiratory distress and signs of impending respiratory failure

• Recognizes physical signs of low cardiac output and signs of impending cardiac arrest
  • Differentiates central from peripheral cyanosis and cardiac from non-cardiac causes of cyanosis.
  • Differentiates acute from chronic hypoxemia.

Obtaining a thorough family history with a focus on genetic abnormalities associated with an important risk of heart disease in childhood

• Obtains a thorough family history focusing on congenital and acquired heart disease.

• Recognizes common genetic syndromes, and their associated cardiac abnormalities.

• Recognizes important non-cardiac abnormalities associated with certain genetic abnormalities or syndromes.

• Demonstrates knowledge of the various methods of genetic testing that are available, along with appropriate and cost-effective use of this testing where applicable.

Developing a prioritized, differential diagnosis

• Knows the signs/symptoms of CHD and acquired heart disease across the spectrum of ages from neonate to young adult.

• Synthesizes clinical findings into an appropriate, prioritized differential diagnosis.

Knowing the risks and benefits of non-invasive and invasive evaluation of infants, children and young adults with congenital or acquired cardiovascular disease, and applying these when ordering diagnostic testing in a cost effective manner

• Prioritizes and orders diagnostic testing to optimize knowledge acquisition, minimize patient risk and discomfort, and be cost-effective.

• Interprets an electrocardiogram (EKG) for acute and/or chronic changes of rate, rhythm, P or QRS axis, ventricular hypertrophy, atrial enlargement, PR interval, QRS duration, corrected QT intervals, ST and T wave changes.

• Knows the indications for obtaining, and has the skills to interpret, chest X-rays based upon the differential diagnosis.
• Knows the indications for, and has the skills to perform and interpret, transthoracic echocardiograms based upon the differential diagnosis.
• Knows the indications and risks, and has the skills to interpret, hemodynamic and angiographic data obtained from diagnostic and interventional cardiac catheterization.
• Knows the indications and risks and has the skills to interpret advanced imaging such as CT angiograms and cardiac magnetic resonance imaging (MRI).

Developing a management plan that incorporates medical therapy, interventional catheter procedures and surgical intervention as well as addressing the psychosocial aspects of acute and chronic disease.

• Demonstrates the skills to develop appropriate initial management plans based upon physiology, including recognizing ductal-dependent lesions and utilizing PGE1 appropriately.
• Knows and understands the medical and surgical interventions for the various types of acyanotic and cyanotic congenital heart disease, including the timing of the interventions, and expected/unexpected complications.
• Knows and understands the medical and surgical interventions for the various types of acquired heart disease, including the type and timing of interventions, and expected/unexpected complications.
• Coordinating both acute and chronic care with interprofessional management teams, to meet the medical, social, developmental, behavioral, mental health, educational and financial needs of the patient and family.

Communicating with and counseling the patient/family regarding immediate, mid- and long-term management

• Communicates the clinical findings, diagnosis and management plan to the patient/family, using understandable terms and concepts, and avoiding the use of excessive jargon.
• Explains and interprets the test results for the patient/family in a timely manner.

Evaluating and managing acquired cardiac conditions found in pediatric patients

• Evaluates and manages basic arrhythmias (tachy/bradyarrhythmias) based on the type of arrhythmia, mechanisms of action, and available therapies.
• Evaluates and manages cardiac masses, tumors, pericardial disease.
• Evaluates chest pain (cardiac and noncardiac) in children and adolescents and manages chest pain of cardiac origin.
• Evaluates and manages syncope.
• Evaluates for cardiac etiology and effects of systemic hypertension
• Evaluates and manages hypercholesterolemia and other dyslipidemias.
• Evaluates and manages Kawasaki Disease.
• Evaluates and manages rheumatic heart disease and its associated cardiac manifestations/complications.
• Evaluates and manages infective endocarditis.

Identifying and applying key evidence-based guidelines

• Conducts a search to find and adhere to current clinical practice guidelines, if available, for management of the various types of congenital and acquired heart disease.
• Explains the various grades of evidence, searches the literature for evidence focusing on the highest-grade evidence available.
• Finds and assesses medical literature for optimal management strategies in the absence of evidence and guidelines.

**Recognizing cardiac conditions and indications that might require additional sub-subspecialty expertise**

• Recognizes and refers/co-manages patients with cardiomyopathy and end stage heart failure.
• Recognizes and refers/co-manages primary pulmonary hypertension.
• Recognizes and refers/co-manages patients with complex arrhythmias
• Refers as needed for pacemaker assessment and management.
• Recognizes limitations and demonstrates help seeking behavior by referring patients to invasive or non-invasive imaging experts as needed.

Reference: