### Curricular Components for Pulmonary EPA

<table>
<thead>
<tr>
<th>1. EPA Title</th>
<th>Care of children with chronic respiratory disease</th>
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<tbody>
<tr>
<td>2. Description of the activity</td>
<td>Children with chronic respiratory illness requiring long-term care pose a challenging but critical activity for the pediatric pulmonologist. Scope of practice is the management of the respiratory system as a component of multi-system disease in children with diseases that include asthma, chronic lung disease of infancy, cystic fibrosis, lower respiratory tract infections, sleep disorders, chronic ventilatory assistance, aspiration syndromes, congenital anomalies of the respiratory system, and other diseases such as pulmonary hypertension, interstitial lung disease, and hemosiderosis. The specific functions which define this EPA include:</td>
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<td>• Identifying and applying evidence based guidelines to diagnose and manage children with chronic respiratory disease</td>
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<td>• Managing home respiratory equipment and effectively translating its use to more acute settings as needed</td>
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<td>• Assessing co-morbid conditions which may be a consequence of the disease process, the medications used to treat the disease, and the emotional, behavioral and mental health problems that often accompany chronic disease</td>
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| 3. Judicious mapping to domains of competence | _X_ Patient Care  
_X_ Medical Knowledge  
___ Practice-Based Learning and Improvement  
_X_ Interpersonal & Communication Skills  
___ Professionalism  
_X_ Systems-Based Practice  
_X_ Personal & Professional Development |
| 4. Competencies within each domain critical to | PC 6: Using optimal clinical judgment  
PC 7: Developing management plans  
PC 8: Performing procedures |

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

Rationale: Pediatric pulmonologists must be able to manage children with complex chronic respiratory illness that requires long-term care.

Scope of Practice: Care for patients with chronic respiratory disease may begin with newborns, including those with cystic fibrosis identified by newborn screen, premature infants who develop severe bronchopulmonary dysplasia, and children born with congenital anomalies of the respiratory system. Other chronic respiratory diseases in children that can present from early childhood through adolescence include chronic aspiration, severe asthma, recurrent infections, sleep disorders, and those that require chronic ventilatory assistance. Care for infants and children with rare diseases, including pulmonary hypertension, interstitial lung disease, and hemosiderosis, may pose particular challenges. This document is intended to address the scope of knowledge and skills of a pediatric pulmonologist who manages children with chronic respiratory disease.

Curricular components that support the function of the EPA:

Developing a long-term management plan with the patient/family that incorporates input of the many professionals within the health care team (e.g., nurses, nutritionists, physical therapists, speech therapists, social workers, and case managers)
- Reviews problems within the scope of the pediatric pulmonologists for chronic care management.
- Identifies and engages with members of the multi-disciplinary team.
- Recognizes ambiguities that exist in the long-term management plan as clinical conditions change and the goals of the family evolve.

Identifying and applying evidence based guidelines to manage children with chronic respiratory disease
- Develops a clinical question.
- Searches the literature for evidence-based guidelines.
- Interprets the evidence in light of grade and relative importance.
- Applies the evidence to care of a particular patient.

Managing home respiratory equipment and effectively translating its use to more acute settings as needed
- Knows the variety of respiratory equipment available for home use.
- Applies the use of specific equipment to different disease states.
- Educates the patient/family about the need for and use of the equipment.
- Arranges home health visits to ensure effective use of the equipment.

Assessing co-morbid conditions which may be a consequence of the disease process, the medications used to treat the disease, and/or the emotional, behavioral and mental health problems that often accompany chronic disease

- Interviews the patient and family using a biopsychosocial framework to understand the full impact of the chronic illness beyond the physical signs and symptoms.
- Lists disease processes as well as psychosocial, behavioral or mental health problems that require multi-disciplinary input and other subspecialty input.
- Manages the complex medication regimen, titrating medications for the individual patient to maximize disease treatment, and identify and minimize possible medication side effects that impact chronic care management.

Utilizing necessary procedures, such as pulmonary function testing and bronchoscopy, for evaluation of children with complex respiratory disease

- Interprets pulmonary function testing in long-term management.
- Performs bronchoscopy to guide long-term management.
- Identifies necessary procedures to be performed by other subspecialists.

Advocating for quality care, which includes involvement of palliative care providers and transition to adult providers

- Involves intra and interdisciplinary care teams to maximize the care of the whole child in the context of the family circumstances.
- Enlists palliative care, when appropriate.
- Initiates and facilities transition to adult providers when appropriate, preparing the patient and family for the transition and ensuring their readiness.

Problems generally within the scope of the pulmonologist’s practice (based on prevalence and potential morbidity) where the role of the pulmonologist is to recognize, evaluate and treat including:

- Severe asthma
- Severe bronchopulmonary dysplasia
- Cystic fibrosis
- Pediatric interstitial lung disease
- Lung transplant recipients
- Patients requiring chronic ventilatory support
- Primary Ciliary Dyskinesia

Problems that generally require consultation where the role of the pulmonologist is to recognize, provide preliminary evaluation and refer/co-manage. This list depends greatly on the context in which one practices. Those pulmonologists 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 subspecialist as needed.
- Chronic aspiration
- Congenital anomalies of the airways
- Immunodeficiency that results in chronic respiratory infections
- Neuromuscular disease, including spinal muscular atrophy and muscular dystrophy
- Rheumatologic lung disease
- Pulmonary complications of cancer and hematopoetic cell transplant
- Pulmonary hypertension
- Sleep disorders