



Entrustable Professional Activities

EPA 2 for Pediatric Rheumatology

EPA 2: Manage Patients with Acute or Chronic Complex Multi-System Rheumatic Disease in an Ambulatory, Emergency, or Inpatient Setting

Supervision Scale for This EPA

1. Trusted to observe only
2. Trusted to execute with direct supervision and coaching
3. Trusted to execute with indirect supervision and discussion of information conveyed for most simple and some complex cases
4. Trusted to execute with indirect supervision but may require discussion of information conveyed for a few complex cases
5. Trusted to execute without supervision

Description of the Activity

Children with complex multi-system rheumatic disease constitute a challenging but critical activity for the rheumatologist. This core EPA for the rheumatologists requires the mastery of providing efficient and effective care to a large number of patients with suspected and confirmed autoimmune and auto-inflammatory disease. The focal components of this professional activity build upon those of managing both the clinic patient with more common rheumatic diseases as well as the more acutely ill hospitalized patient.

The specific functions which define this EPA include:

1. Developing knowledge of rheumatic autoimmune diseases, auto-inflammatory diseases, and associated musculoskeletal conditions specific to children, adolescents and young adults
2. Performing general and specialized history and physical examinations (e.g., rheumatologic review of systems and musculoskeletal examination) in order to care for patients with chronic complex multi-system rheumatic disease
3. Developing sound clinical reasoning that drives the creation of an appropriate differential diagnosis and evaluation
4. Applying guidelines, evidence-based literature, and/or consensus treatment plans to the care of patients
5. Developing and documenting a management plan that addresses the primary problem, all co-morbidities, as well as cost considerations
6. Coordinating care in different health care settings (e.g., ER, hospital) and with an interdisciplinary health care team
7. Placing the patient at the center of all management decisions
8. Demonstrating the ability to manage uncertainty (one's own as well as that of the patient and family)

Judicious Mapping to Domains of Competence

- Patient Care
- Medical Knowledge
- Practice-Based Learning and Improvement
- Interpersonal and Communication Skills



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- Professionalism
 Systems-Based Practice
 Personal and Professional Development

Competencies Within Each Domain Critical to Entrustment Decisions

| | |
|---------|---|
| PC 1: | Gathering information |
| PC 5: | Performing complete physical exams |
| PC 7: | Developing management plans |
| MK 1: | Demonstrating knowledge |
| PBLI 6: | Practicing EBM |
| ICS 1: | Communicating with patients/families |
| ICS 3: | Communicating with health professionals |
| SBP 2: | Coordinating care |
| PPD 8: | Dealing with uncertainty |

Context for the EPA

Rationale: Pediatric rheumatologists must be able to diagnose and manage patients with acute or chronic complex multi-system rheumatic disease in a variety of settings including outpatient clinic, the emergency room, and in the hospital. All of these settings will require coordination of care with a multidisciplinary health care team.

Scope of Practice: Care is provided in the ambulatory and inpatient setting. The pediatric rheumatologists also serve as a resource for telephone, electronic, and remote consultation, and care is occasionally supervised in-home or at remote sites. The patient population includes all patients with rheumatic, inflammatory, and autoimmune diseases, and associated musculoskeletal conditions followed regularly in the pediatric rheumatology clinic and patients evaluated as requested consultations. Patients will range in age from newborns to young adults. This document is intended to address the scope of knowledge and skills of the pediatric rheumatologist, with the understanding that the pediatric rheumatologist will often need to create and lead multidisciplinary teams and require evaluation and management input from other health care professionals, and participate in quality improvement.

Curricular Components That Support the Functions of the EPA

1. Developing knowledge of rheumatic autoimmune diseases, auto-inflammatory diseases, and associated musculoskeletal conditions specific to children, adolescents, and young adults

Such diseases include but are not limited to:

- Juvenile Idiopathic Arthritis (all subtypes)
- Inflammatory Eye Disease
- Idiopathic Inflammatory Myositis
- Systemic Vasculitis (Henoch-Scholein Purpura, Anti-neutrophil cytoplasmic antibody [ANCA] associated vasculitis, Polyarteritis nodosa, Kawasaki disease, Takayasu arteritis, Behçet disease, central nervous system vasculitis)



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- Systemic Lupus Erythematosus and Related Disorders
 - Scleroderma, local and systemic and Related Disorders
 - Autoinflammatory diseases
 - Primary immunodeficiencies and other disorders associated with inflammatory and autoimmune manifestations
 - Arthritis related to infection and associated conditions
 - Noninflammatory musculoskeletal conditions
2. Performing general and specialized history and physical examinations (e.g., rheumatologic review of systems and musculoskeletal examination) in order to care for patients with chronic complex multi-system rheumatic disease
- Obtains a thorough clinical history, relevant review of systems, and assess functional status of children, adolescents, and young adults with rheumatic and musculoskeletal disease symptoms
 - Assimilates, organizes, and succinctly summarizes all pertinent previously obtained medical information from the Emergency Department, Primary Care Physician, outside hospital, and/or other subspecialists
 - Performs and interprets a complete general physical examination
 - Performs and accurately interprets the examination of all joints, periarticular structures, peripheral nerves, and muscles
 - Identifies extra-articular findings associated with rheumatic and autoimmune diseases, and inflammatory and associated musculoskeletal conditions
 - Identifies ocular manifestations and complications of rheumatologic and musculoskeletal disease in children, adolescents, and young adults
 - Frequently reassesses to assure correct diagnosis and/or response to therapy
3. Developing sound clinical reasoning that drives the creation of an appropriate differential diagnosis and evaluation
- Develops a differential diagnosis in patients presenting with signs and symptoms of potential rheumatologic diseases and outlines further testing necessary to establish the correct diagnosis
 - Explains the indications for and costs of laboratory tests, imaging studies, and procedures to establish a diagnosis of rheumatic and autoimmune diseases, and inflammatory and associated musculoskeletal conditions
 - Accurately interprets the results of laboratory tests, imaging studies, pathology, and other studies
 - Reassesses to assure appropriate diagnosis
4. Applying guidelines, evidence-based literature, and/or consensus treatment plans to the care of patients
- Develops a clinical question using the PICO format
 - Searches the literature for evidence focusing on the highest grade evidence available
 - Interprets the evidence in light of its grade
 - Applies the evidence to the care of the patient given the particular context for that patient
 - Adheres to evidence-based practice and/or commonly accepted guidelines for comorbidity screening and toxicity monitoring



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5. Developing and documenting a management plan that addresses the primary problem, all comorbidities, as well as cost considerations
 - Understands the rationale for, risks, benefits, and costs of different therapies used in the management of these diseases
 - Develops an appropriate treatment plan for the care of a patient with a rheumatologic problem integrating medications (oral, injectable, infused), other therapies when appropriate (counseling, physical therapy referral, occupational therapy referral), and when necessary other consultation.
 - Demonstrates the ability to explain the rationale and the risks/benefits for the treatment plan and engage patients and parents in shared decision making
 - Knows the clinical pharmacology of each medication, understands the dosing, pharmacokinetics, metabolism, mechanisms of action, side effects, drug interactions, compliance issues, costs, and use in patients of child-bearing potential
 - Explains the indications for and performs arthrocentesis
 - Recognizes and manages ocular manifestations and complications of rheumatologic and musculoskeletal diseases in collaboration with ophthalmologists
 - Assesses and manage pain
 - Identifies physical impairment, relates the impairment to the observed functional deficits, and prescribes appropriate physical therapy and occupational therapy to achieve goals to improve the defined impairment
 - Identifies and addresses barriers to adherence to treatment plan
 - Reassesses patients over time, recognizing treatment related adverse events, and alters the treatment plan accordingly
 - Determines the need for medication dose escalation, reduction, or withdrawal, or additional medication initiation
 - Identifies when referral to other specialists is indicated, for example:
 - Nephrologist for lupus nephritis or vasculitis with glomerulonephritis
 - Ophthalmologist for juvenile idiopathic arthritis or others at risk for ocular inflammation
 - Dermatologist for cutaneous manifestations of lupus, dermatomyositis, scleroderma, or vasculitis
 - Hematologist for blood dyscrasias
 - Cardiologist for myocarditis and/or pericarditis
 - Collaborate and communicate effectively with other subspecialists and providers to co-manage patients receiving immunomodulatory therapy
6. Coordinating care in different health care settings (e.g., ER, hospital) and with an interdisciplinary health care team
 - Managing the acutely ill hospitalized patient with either a rheumatic disease or high suspicion for rheumatic disease.
 - Initiates immunosuppressive therapy in the patient without a defined rheumatic disease if other diagnostic possibilities have been ruled out or when such therapy can reasonably be expected to not cause deterioration in patient clinical status
 - Co-manages with a general medical team if indication for hospitalization is not due to patient's primary rheumatic disease
 - Co-manages with critical care team in patients who require such care



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- Determines when patient can be safely discharged to home based on clinical status and patient/family ability to manage disease and treatment at home
 - Managing patients with acute or chronic complex rheumatic disease in the emergency setting.
 - Assesses disease activity status through targeted history of present illness, physical exam, and laboratory and radiologic testing
 - Recognizes urgent/life threatening rheumatologic conditions such as:
 - Macrophage activation syndrome
 - Catastrophic antiphospholipid antibody syndrome (APA)
 - Pulmonary embolus, pulmonary hemorrhage, or myocardial Infarction in systemic lupus erythematosus
 - Pericarditis associated with a rheumatic condition
 - Cerebrovascular accident complicating a rheumatic condition
 - Determines whether input from other subspecialists is required
 - Distinguishes disease flare from adverse treatment effect
 - Determines need for medication dose escalation, reduction, or withdrawal, or additional medication initiation
 - Assesses whether patient's current clinical status would improve with inpatient admission
 - Arranges post-visit follow up as needed
7. Placing the patient at the center of all management decisions
- Explains the management to patients and families
 - Invites questions from family
 - Includes family in shared decision-making
 - Communicates effectively with other members of the multidisciplinary team, consultants, and other providers
8. Demonstrating the ability to manage uncertainty (one's own as well as that of the patient and family)
- Consults with other experts in the field
 - Develops consensus with colleagues
 - Discusses inherent uncertainty in diagnosis with patient and referring physician
 - Frequently reassesses history, physical exam, and laboratory data

Problems **generally** within the scope of pediatric rheumatology practice (based on prevalence and potential morbidity) where the role of the rheumatologist is to recognize, evaluate, and treat or determine that the patient needs referral to a different subspecialist for more specialized treatment.

- Juvenile Idiopathic Arthritis (all subtypes)
- Inflammatory Eye Disease
- Idiopathic Inflammatory Myositis



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- Systemic Vasculitis (Henoch-Scholein Purpura, Anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis, Polyarteritis nodosa, Kawasaki disease, Takayasu arteritis, Behçet disease, central nervous system vasculitis)
- Systemic Lupus Erythematosus and related disorders
- Scleroderma, local and systemic and related disorders
- Autoinflammatory diseases
- Primary immunodeficiencies and other disorders associated with inflammatory and autoimmune manifestations
- Arthritis related to infection and associated conditions.
- Noninflammatory musculoskeletal conditions

Problems that **generally** require further consultation where the role of the subspecialist is to recognize, provide preliminary evaluation, and refer/co-manage.

- Nephrologist for lupus nephritis or vasculitis with glomerulonephritis
- Ophthalmologist for juvenile idiopathic arthritis or others at risk for ocular inflammation
- Dermatologist for cutaneous manifestations of lupus, dermatomyositis, scleroderma, or vasculitis.
- Hematologist for blood dyscrasias
- Cardiologist for myocarditis and/or pericarditis

(Collaboration and communication with other subspecialists and providers to co-manage patients receiving immunomodulatory therapy is critical.)

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Curricular Components Authors

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