The following document was developed to provide a general understanding of the reasons for involving a pediatric rheumatologist in patient care and to identify circumstances when referral to a pediatric rheumatologist is appropriate. The ultimate objective in providing medical care for children with rheumatic diseases is to achieve the best possible health outcome in the most cost-effective setting.

Rheumatic diseases are an important cause of disability in childhood. Proper diagnosis and early aggressive intervention can minimize both short and long term morbidity of these conditions. Without proper therapy, acute rheumatic fever, systemic lupus erythematosus, dermatomyositis, progressive systemic sclerosis, and many forms of vasculitis can be fatal. Other conditions such as juvenile idiopathic arthritis and spondyloarthopathies which do not acutely threaten life, can be associated with lifetime disability. Rheumatic diseases in childhood differ from those in adults. There are important age related impacts of the diseases on the developing immune, neurologic and musculoskeletal systems. These chronic diseases have profound psychosocial effects on patients and their families.

The goals of treatment of childhood rheumatologic diseases are to control disease activity, preserve normal physical, social and emotional growth and development, minimize chronic disability and deformity, and achieve remission of disease. In pediatric rheumatic diseases, findings on physical examination often take precedence over laboratory findings in the establishment of a diagnosis and a treatment plan. Children and adolescents are often difficult to evaluate due to their development and behavioral stages; therefore the importance of a skilled examiner cannot be over emphasized.

Pediatric rheumatologists are physicians who specialize in providing comprehensive care to children with rheumatologic diseases and their families. They are pediatricians who have completed an additional 2-3 years of specialized training in pediatric rheumatology and are usually Board Certified in Pediatric Rheumatology. In some cases these physicians may have been trained initially as internists rather than pediatrics. Pediatric rheumatologists are specifically trained to be highly skilled in:

1. differential diagnosis in children and adolescents;
2. efficient use of diagnostic evaluations (including other consultative specialties) interventions in children and adolescents;
3. selecting the most appropriate therapy for children and adolescents with rheumatic diseases;
4. monitoring long term therapy for effectiveness and side effects unique to children and adolescents;
5. achieving favorable outcomes in terms of control of rheumatologic diseases and prevention of disability;
6. coordination of care for children and adolescents with multisystem diseases; and
7. caring for chronically ill children, adolescents and their families.

Most pediatric rheumatologists are located at university centers and work with a multidisciplinary team that includes one or more pediatric rheumatologist and other health care professionals who specialize in the treatment of rheumatologic diseases, such as registered nurses, physical therapists, occupational therapists and social workers.

A pediatric rheumatology center will also have available the services frequently needed by these patients such as nutrition, pediatric orthopedics, pediatric nephrology, pediatric ophthalmology, pediatric cardiology, child psychology/psychiatry, maxillo-facial surgery, pediatric dermatology, and physiatry.

The major strength of the multidisciplinary team is facilitating the achievement of the goals of treatment of childhood rheumatic diseases in the least costly setting. In those geographic areas of the country where visits to a pediatric rheumatologist or center can only be accomplished one to two times per year, a local adult rheumatologist may be part of this treatment team as well. Due to the limited availability of pediatric rheumatology services in many areas of the
country, adult rheumatologists who have training and experience in pediatric rheumatology should also be utilized as part of the multidisciplinary team to facilitate the achievement of the goals of treatment of childhood rheumatic diseases.

**Children and adolescents appropriate for referral to a pediatric rheumatologist:**

1. **Diagnostic Evaluation of Patients with Unclear Diagnoses**
   - Prolonged fever
   - Loss of function
     - inability to attend school
     - regression in physical skills
   - Normal laboratory findings but local or generalized pain and/or swelling
   - Abnormal laboratory findings but symptoms and/or examination do not fit clinical criteria for a specific rheumatic disease
   - Complaints not consistent with laboratory findings or physical examination
   - Unexplained physical findings such as rash, fever, arthritis, anemia, weakness, weight loss, fatigue or anorexia
   - Unexplained musculoskeletal pain
   - Undefined autoimmune disease

2. **Diagnostic evaluation and long-term management of:**
   - Juvenile idiopathic arthritis
   - Spondyloarthropathies
     - Ankylosing Spondylitis
     - Reactive arthritis
     - Arthritis associated with inflammatory bowel disease
   - Other arthritides
     - Lyme disease with arthritis
     - Post-infectious arthritis
   - Connective Tissue Diseases and related syndromes
     - Systemic lupus erythematosus
     - Juvenile Dermatomyositis or polymyositis
     - Mixed connective tissue disease
     - Scleroderma - systemic and localized
     - Sjögren syndrome
     - Anti-phospholipid antibody syndrome
   - Chronic vasculitides
     - Polyarteritis nodosa
     - Wegener granulomatosis
     - Behcet syndrome
     - Takayasu arteritis
     - Hypocomplementemic vasculitis
     - Hypersensitivity vasculitis
     - Cerebral vasculitis
     - Post-infectious vasculitis
   - Other Inflammatory Diseases
     - Autoinflammatory and Periodic fever syndromes
     - Chronic recurrent multifocal osteomyelitis
     - Relapsing polychondritis
     - Sarcoidosis
3. **Confirm diagnosis and help formulate and/or participate in a treatment plan for the following conditions:**
   - Acute rheumatic fever
   - Apophysitis
   - Cold
   - Complex autoimmune thrombocytopenia
   - Complex Regional Pain Syndrome
   - Erythromelalgia
   - Growing pains
   - Henoch-Schonlein Purpura
   - Hypermobility
   - Induced injury
   - Kawasaki disease
   - Osteoarthritis
   - Osteochondroses
   - Osteoporosis
   - Pain syndromes
   - Raynaud phenomenon
   - Secondary autoimmune hemolytic anemia
   - Serum sickness
   - Uveitis

4. **Diagnostic or treatment plan evaluation for autoimmune disorders associated with other primary diseases such as: immunodeficiency, neoplasm, infectious disease, endocrine disorders, genetic and metabolic diseases, post-transplantation, cystic fibrosis and arthritis associated with birth defects.**

5. **Provide second opinion or confirmatory evaluation when requested in certain cases where primary care physicians request expert opinion for families requiring subspecialty input to cope with disease process, accept treatment plan, allay anxiety and provide education.**

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