Pediatric Rheumatology for the Primary Care Clinicians-Recognizing Patterns of Disease

Charles H. Spencer, MD,[a] and Anjali Patwardhan, MD[b]

This review presents a diagnostic approach to musculoskeletal and rheumatic diseases in children for primary care clinicians. The focus is on juvenile idiopathic arthritis (JIA) as the major arthritis disease in children. It is necessary to know the personalities of these JIA categories. It is also crucial to be able to recognize the common infectious, orthopedic and mechanical, malignant, genetic, other rheumatic diseases, and other miscellaneous syndromes that can mimic JIA. To do so requires recognition of clinical patterns using a thorough musculoskeletal and rheumatic history and repeated complete physical exams with emphasis on the musculoskeletal exam. It also requires targeted and limited laboratory testing with careful follow-up over time.

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Background

Musculoskeletal and rheumatic diseases in children are common and often debilitating.[1–3] It is estimated that 1–4/1000 children worldwide have a juvenile arthritis disease.[1] A recent report estimated that 294,000 children in the US have juvenile arthritis diseases or 1/250 children, more than many other better-known chronic diseases in childhood.[2] The incidence of musculoskeletal illnesses also may be increasing.[3–5] Thus there is some evidence that these musculoskeletal illnesses do need more attention.

As the World Health Organization noted in 2003, musculoskeletal and rheumatic diseases in adults and children do not receive much attention because they are infrequently fatal but remain the number one cause of suffering and morbidity in the world.[6] Specifically, juvenile idiopathic arthritis has significant morbidity and can be painful, disable a child, limit quality of life, add to the financial stress of families, and have a substantial societal cost as well, reflecting a major personal and societal "burden."[6–12]

Despite these reports and accumulating evidence of the impact of musculoskeletal and rheumatic disease, medical education in musculoskeletal and rheumatic diseases remains inadequate in the United States, United Kingdom, and probably in other countries as well.[13–16] It appears that a minority of medical students and residents graduate competent in musculoskeletal medicine in the United States. This deficiency is especially striking in the average clinician's inability to perform a competent musculoskeletal physical exam in children and adults.[17–22]

In addition, there is ample evidence that documentation of a musculoskeletal exam for pediatric inpatients is infrequent.[23–26] A study in the UK in 2002 noted that 4% of 257 pediatric admissions had a documented musculoskeletal exam compared to >90% cardiac and pulmonary exam.[27] As musculoskeletal education has been deficient in the US and the UK and probably elsewhere in the world, this deficit appears to result in inappropriate musculoskeletal referrals crowding already busy rheumatology, orthopedic, and sports medicine physician practices as well as delayed and diminished quality of medical care for children and adults who actually do have significant musculoskeletal diseases.[28–32] While critically important, improving musculoskeletal and rheumatic education, particularly in the neglected area of pediatric rheumatology and musculoskeletal medicine, will require a multifaceted approach beyond the scope of this paper.

The goal of this article is to provide a basic educational primer for diagnosis of musculoskeletal and rheumatic diseases in children. Easier diagnosis may lead to early treatment and possibly better disease outcomes. This primer may prove useful for primary

From the [a]Nationwide Children's Hospital, Ohio State University, Columbus, OH; and [b]School of Medicine, University of Missouri, Columbia, MO.

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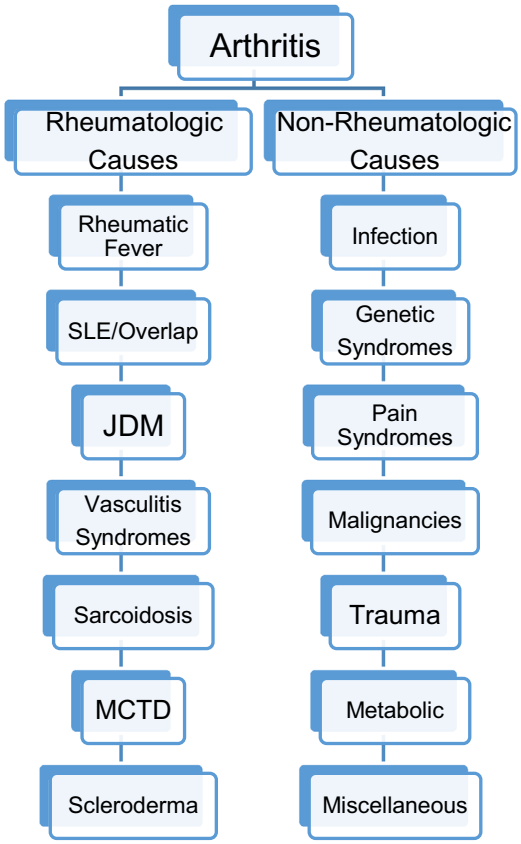

FIG 1. Differential diagnosis of the pediatric rheumatologic arthritis universe.

care and emergency physicians. The reader is referred to a musculoskeletal screening exam for children (pGALS) of Foster et al.[33] and her website (http://www.pGALS.org).

As noted in medical education research, recognition of clinical patterns is crucial to correct diagnosis.[34,35] This article will demonstrate how the pediatric rheumatologic universe is organized (Fig. 1), and how to recognize the distinctive patterns and personalities of each of the rheumatic and non-rheumatic problems that must be considered in the differential diagnosis.

Clinical Patterns for Diagnosis

JIA—The Center of the Universe

Juvenile idiopathic arthritis is the center of the universe of a pediatric rheumatologist (Fig. 1). The most common diagnosis of the children seen regularly in our clinics is JIA.[36,37] It is useful to understand all problems that can cause a child to have arthritis or a painful extremity in terms of how they differ from JIA.

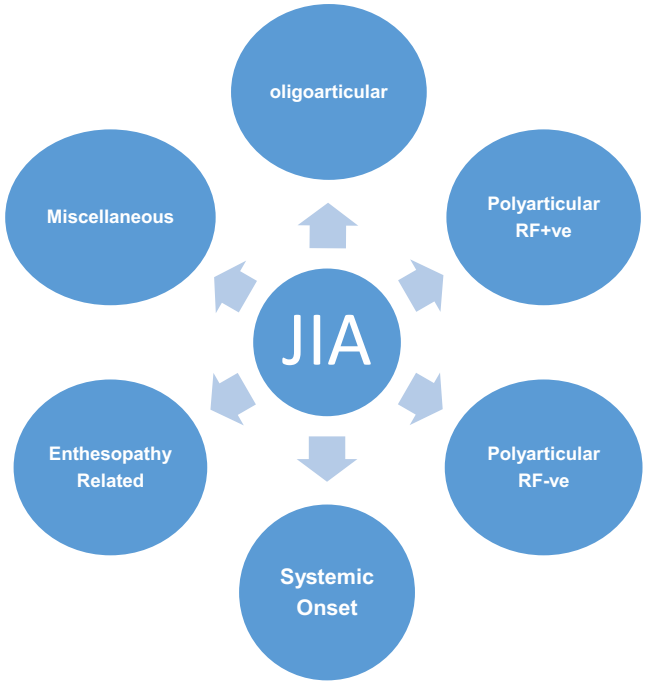

FIG 2. Classification of juvenile idiopathic arthritis.

JIA is chronic synovitis of childhood and includes the traditional juvenile rheumatoid arthritis subtypes[38] as well as other synovitis diseases. This term JIA has supplanted JRA as the appropriate nomenclature for the chronic arthritis diseases in childhood. A child only has JIA when he/she meets the criteria for JIA and has no other rheumatic or non-rheumatic illness (Fig. 2). The types of JIA include systemic onset, rheumatoid factor-positive polyarticular, rheumatoid factor-negative polyarticular, oligoarticular, extended oligoarticular, psoriatic, enthesopathy with arthritis, and a miscellaneous other category.[39–42]

The diagnostic criteria for the classic JRA subtypes of JIA (systemic, polyarticular-5 joints or more, oligoarticular-4 joints or less) include that the child must have had day-in, day-out arthritis for a minimum of 6 weeks. The arthritis is not a transient, episodic arthritis but a persistent swelling, limitation, or tenderness/pain in a joint each day. The arthritis is documented best by the presence of joint swelling. Limitation of joint movement is suggestive as well, but must be supported by the presence of swelling or tenderness/pain. Joint pain or arthralgia is not sufficient for JIA diagnosis as there are many causes of joint and extremity pain. The 6 week requirement reflects the reality that children may have brief, transient synovitis episodes (traumatic, viral, and post-viral) that