Invited Review

Musculoskeletal Complaints: When Should We Consult a Pediatric Rheumatologist?

Author(s)	Hafize Emine Sönmez ¹ , Serife Gül Karadağ ² , Nuray Aktay Ayaz ³		
Affiliation(s)	¹ Kocaeli University Faculty of Medicine, Department of Pediatric Rheumatology, Kocaeli, Turkey ² Erzurum Regional Training and Research Hospital, Department of Pediatric Rheumatology, Erzurum, Turkey ³ Istanbul University, Faculty of Medicine, Department of Pediatric Rheumatology, Istanbul, Turkey		
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Abstract

This review aimed to summarize key points that might suggest rheumatologic diseases to physicians dealing with musculoskeletal (MSK) complaints. Evaluation of a child presenting with MSK findings requires a comprehensive, multidisciplinary, and systematic approach. In children with MSK complaints, detailed anamnesis, appropriate physical examination and joint examination, and the use of correct laboratory tests will be helpful for accurate diagnosis. The algorithm we have suggested for MSK complaints of children will be a guide for the physicians.

Keywords: Musculoskeletal complaints, joint pain, rheumatology, juvenile idiopathic arthritis

Introduction

Musculoskeletal (MSK) features account for 10%-20% of pediatric patients' complaints in primary care clinics.¹ It was shown that approximately 50% of the children and adolescents present with MSK complaints at least once during any period of their life.¹ The underlying causes are highly variable, ranging from benign muscular conditions to rheumatic diseases or malignancies. For instance, constitutional symptoms such as the presence of fever and fatigue, nature and location of pain, duration of morning stiffness, and presence of abnormal examination findings

may be helpful while finding out the underlying etiology. Some 'red flags' may guide the clinicians while making the differential diagnosis. A large number of patients with joint pain, fever of unknown origin, elevated acute phase reactants (APRs) and positive anti-nuclear antibody (ANA) are generally referred to a pediatric rheumatologist. However, underlying aetiologies are often found to be infectious, orthopedic, or traumatic conditions that can be recognized by a pediatrician.² Pediatric rheumatology has emerged as a subspecialty of pediatrics in the last 50 years



Correspondence: Hafize Emine Sönmez, Kocaeli University Faculty of Medicine, Department of Pediatric Rheumatology, Kocaeli, Turkey E-mail: eminesonmez@gmail.com



and then fellow education programs have been started all around the world. However, due to the insufficient number of specialists or centers in rural areas, patients still face problems when accessing appropriate care. Previous studies demonstrated that only half of the patients referred to rheumatology centers were finally diagnosed with rheumatic disease.³⁻⁵ Improvement

in the awareness of rheumatic diseases, as well as their differential diagnosis, will alleviate unnecessary consultations and lead to effective utilization of the capacity of pediatric rheumatologists. This review was organized to reveal key points that might suggest rheumatologic diseases to physicians dealing with musculoskeletal complaints.

History Taking

A proper anamnesis and a detailed physical examination contain many clues that will lead us to the

correct diagnosis. 'Listen to your patient; they are telling you the diagnosis' is a well-known and much-quoted aphorism. Nature, location, duration, and timing of pain, accompanying systemic and dermatologic features, exacerbating or relieving factors of complaints, family history of autoimmune or auto-inflammatory diseases usually concern important clues leading clinicians to make an accurate diagnosis while assessing children with MSK complaints. Since children are not just 'little adults', the assessment and management of children with MSK complaints require a special approach. It is difficult for younger children to define the location and character of pain, both because of the reflected pain and incomplete language development in early childhood. These points may cause family anxiety and may also make it complicated for the clinicians to make

a differential diagnosis. Segal et al.6 designed a decision support software to reduce diagnostic errors in pediatric rheumatology practice (www.simulconsult.com). Using such special software or following the clues for the diagnosis will greatly reduce unnecessary consultations (**Figure 1**). The differential diagnoses of MSK complaints are depicted in **Table 1**.

> With detailed anamnesis by taking into account the clues and with appropriate physical examination, almost all the patients can be diagnosed favourably. Some critical inquiries may help the clinicians in making an accurate diagnosis, one of the important points undoubtedly being the acute or chronic course of pain. New-onset pain is usually related to an acute disease such as septic arthritis, acute rheumatic fever (ARF), viral myositis, osteomyelitis, or trauma. The patient should be asked about

the recent history of infection for post-infection arthritis, trauma for orthopedic and mechanical conditions, or raw milk consumption for brucellosis.⁷⁻¹¹ In the presence of single joint involvement and fever, septic arthritis, osteomyelitis, soft tissue infections, and malignancies should be taken into account. In this case, the presence of fever, the history of the previous infection, and trauma should be questioned. While if a patient presents with single joint pain without fever, trauma, mechanical problems, toxic synovitis, and avascular necrosis may be the underlying etiology. Familial Mediterranean fever (FMF) should be considered in patients presenting with fever and recurrent non-erosive arthritis. The presence of migratory arthritis refers to ARF, patients with migratory arthritis should be questioned for a history of upper respiratory tract infection.7-11

<i>Table 1</i> The differential diagnoses of musculoskeletal complaints	
Traumatic Fractures	Neoplasia (Benign or malign)
Growing pains	Osteoid osteoma
Hypermobility-associated pain	Chondroblastoma
Diffuse idiopathic pain syndromes	Osteoblastoma
Juvenile fibromyalgia	Leukaemia
Complex regional pain syndromes	Lymphoma
Orthopaedic/mechanical	Neuroblastoma
Overuse injury	Ewing's sarcoma
Slipped upper femoral epiphysis	Osteosarcoma
Perthes' disease	Langerhans cell histiocytosis
Osgood Schlatter disease (tibial tuberosity),	Inflammatory
Scheuermann's disease	Transient synovitis of the hip
Leg length discrepancy	Juvenile idiopathic arthritis
Club foot	Reactive arthritis
Infectious	Acute rheumatic fever
Septic arthritis	Chronic recurrent multifocal osteomyelitis
Osteomyelitis	Systemic lupus erythematosus (SLE)
Discitis	Juvenile dermatomyositis
Lyme disease	Vasculitis
Brucella	Other
	Haemophilia
	Vitamin D deficiency/Rickets

Highlights

· Nature, location, duration, and

and

systemic

clues.

timing of pain, accompanying

features may concern important

· Inflammatory joint pain usually

Mechanical joint pain is more

likely to worsen with exercise.

long period of immobility.

occurs in the morning after a

dermatologic

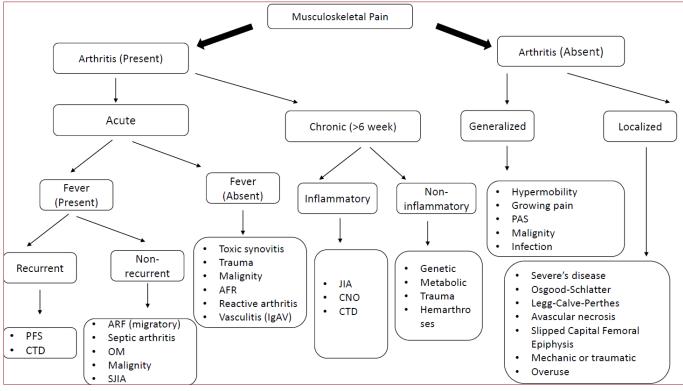


Figure 1. Algorithm for evaluation of children presenting with musculoskeletal complaints

CNO, chronic non-bacterial osteomyelitis; CTD, connective tissue disease; FMF, familial Mediterranean fever; IgAV, Immunoglobulin A vasculitis; JIA, juvenile idiopathic arthritis, PFS; Periodic fever syndrome, ARF; Acute rheumatic fever, OM; osteomyelitis, PAS; Pain Amplification Syndrome, SJIA; systemic juvenile idiopathic arthritis.

When the patients have chronic joint complaints, the character of the pain should be distinguished, that is whether it is of an inflammatory or mechanical type. Mechanical joint pain is more likely to worsen with exercise, while inflammatory joint pain usually occurs in the morning after a long period of immobility and improves throughout the daily exercise. Sports activities and lifestyles should be questioned for sports-related injuries (Table 2).7-11 Children with growing pains present with bilateral awakening leg pain, which is resolved through painkillers or massage. A physical examination of these children is normal and the pain is not accompanied by systemic symptoms such as fever, weight loss, and weakness. If clinicians follow the clues, they can easily distinguish the growing pain. For instance, growing pains are never present at the start of the day after waking and never restrict physical activities. Furthermore, children with growing pain do not limp and their growth development is normal.7-11

Table 2

Comparison of mechanical and inflammatory joint problems in children

	Mechanical joint problems	Inflammatory joint problems
Pain worsening with activity	+	-
Pain worsening with rest	-	+
Morning stiffness	-	+
Swelling	-/+	+
Loss of movement	_/+	+
Locking	+	-

Benign conditions present with pain resolving with rest and worsening with activity without obvious joint swelling, constitutional symptoms, and abnormal laboratory findings. Furthermore, extra-articular and systemic symptoms (fever, weight loss, fatigue) usually refer to systemic disease.^{7–12} Approximately 20% of children with leukemia present with MSK complaints at the diagnosis.^{13,14} Therefore, it is important to follow systemic symptoms such as fever, weight loss, and fatigue. Morning stiffness and unresponsive pain to painkillers often indicate serious conditions.

Juvenile idiopathic arthritis (JIA) is the most common reason for chronic arthritis worldwide. It is a heterogeneous group of disorders presenting with joint inflammation. The presence of morning stiffness for longer than 15 minutes was found to be a strong predictor factor to distinguish JIA among children with MSK complaints.¹⁵

Len et al.¹⁶ purposed a questionnaire for the early detection of convenient referral to pediatric rheumatology centers. This tool can guide clinicians for early referral of patients with chronic arthropathy to pediatric rheumatology centers. This tool consists of twelve questions and each question scores 1 point in the presence of positive response; if the final score is 5 points or above, the patient has to be referred to a rheumatologist. The affirmational tool questioned the following items:

- 1. Presence of swollen joints or articulations for the last 7 days.
- 2. Complaint of pain in the joints, muscles, or bones for the last 7 days without trauma.
- 3. Swollen joints lasting more than 30 days.
- 4. Complaint of pain in the joints lasting more than 30 days without trauma.
- 5. Presence of difficulties in closing the hands, folding the wrists, knees, or ankles.
- 6. Presence of limping in the last month.
- 7. Disabilities while playing or running.
- 8. Waking up with a complaint of pain in the joints.

- 9. Presence of difficulties in daily activity due to pain in the joints.
- 10. Presence of deformity in any joint.
- 11. Presence of fever for more than 30 days without any apparent cause.
- 12. Presence of rash followed by swelling or pain in the joints.

Physical Examination

Paediatric Gait, Arms, Legs and Spine (pGALS) has been introduced as a simple and quick screening tool for the evaluation of the MSK system in children.¹⁷ This tool is recommended as the initial assessment while evaluating a child with MSK complaints. The accuracy of this tool in Turkish children was tested and its Turkish version was shown as a valid, acceptable, and practical screening test.¹⁸ pGALS is based on methodology as a "copy me" approach. The clinicians show the movements and wait for the patient to imitate them, thus allowing a rapid assessment of the entire MSK system. In the presence of an abnormal finding in pGALS, patients should be examined in more detail.¹⁹

A comprehensive physical examination should be performed to exclude non-rheumatic conditions. Skin evaluation may provide some clues for accurate diagnosis. For instance, the presence of erythema marginatum or subcutaneous nodules with migratory polyarthritis in a child with a history of throat infection is typical for the diagnosis of ARF. The presence of palpable purpura, especially on the lower extremities, in a patient with arthritis and subcutaneous edema will direct the clinician to the diagnosis of immunoglobulin A (Ig A) vasculitis only by inspection. Furthermore, severe pain in a single joint, redness and increased heat on the affected joint should suggest septic arthritis to physicians. Physical examination including palpation, evaluation of active and passive movements of the joints, and muscle strength examination may reveal the source of pain as either being of joint or muscle origin. Malignancies should be considered in children with insidious onset of symptoms such as weight loss, weakness, fever, widespread extremity pain awakening from sleep at night, and abnormal physical findings such as ecchymosis and petechiae, hepatosplenomegaly, and lymphadenopathy.7-12

Although connective tissue diseases are rare in children, children with systemic lupus erythematosus (SLE), juvenile dermatomyositis (JDM), juvenile scleroderma, or mixed connective tissue disease may present with arthritis. In patients, especially adolescent girls, presenting with skin findings such as malar rash and photosensitivity concomitant to arthritis and/or myositis the diagnosis of SLE should be kept in mind. In addition, JDM should be considered in children with symmetric proximal muscle weakness and skin findings such as heliotropic rash and Gottron's papules. Therefore, detailed systemic examination, both skins assessmentand MSK examination has great importance in patients with musculoskeletal complaints.

Children with hypermobility usually suffer from chronic MSK pain. The Beighton score is a popular screening technique for hypermobility. It is a nine-point scale

including 5 manoeuvres as follows: 1) Passive dorsiflexion and hyperextension of the fifth metacarpophalangeal joint beyond 90°, 2) Passive apposition of the thumb to the flexor aspect of the forearm, 3) Passive hyperextension of the elbow beyond 10°, 4) Passive hyperextension of the knee beyond 10°, 5) Palms of the hands resting flat on the floor.7–12 A positive Beighton score for children is at least 6 out of the 9 points.

By considering the age factor, a detailed joint examination guides the presence of tendinitis, apophysitis, or arthritis. For instance, the calcaneal growth plate is stressed by the Achilles tendon (Sever's disease) in younger children and older children due to the growth of the immature skeleton apophysitis located to the tibial tubercle or the inferior patellar pool (Osgood-Schlatter disease) ensues.²⁰

Laboratory Evaluation

A normal erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels in a child with chronic joint pain may help to exclude the diagnosis of infections and malignancies. A blood smear should be performed to exclude hematologic malignancies in patients with MSK complaints and systemic features. In the presence of elevated lactate dehydrogenase (LDH) levels, malignancy should be ruled out. Autoantibodies such as ANA and rheumatoid factor (RF) may be positive in healthy children.²⁰ The positivity rate of ANA and RF in healthy Turkish children was reported to be 4% and 3%, respectively.²¹ For this reason, testing ANA or RF is not recommended as a screening test to rule out rheumatologic disease in the primary care setting.^{10–12} Furthermore, the antistreptolysin O (ASO) titer should be used to confirm the diagnosis of ARF but it should be kept in mind that ASO titer peaks between three to eight weeks after a streptococcal infection and may remain high for several months.²² Consequently, the presence of ANA and RF antibodies or elevated ASO does not directly refer to rheumatological diseases and their usage as a screening test causes unnecessary costs and concerns, as well.

Conclusion

In children with MSK complaints, detailed anamnesis, appropriate physical examination and joint examination and the use of correct laboratory tests will provide early access to accurate diagnosis and also reduce the burden of pediatric rheumatology clinics.

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