

Introduction to Rheumatology and Calcium Pyrophosphate Deposition Disease (CPPD, Pseudogout)

[See online here](#)

In rheumatology, we talk mainly about disorders of the musculoskeletal system, connective tissue diseases, inflammatory disorders, and bone disease. Musculoskeletal pain is a common feature in most rheumatological disorders and might be caused by arthritis or soft tissue inflammation. In this article, you will get an introduction to rheumatology and pseudogout and gain all the essential information for your next medical exam.



An Introduction to Rheumatology

Rheumatology is the branch of medicine associated with evaluation and treatment of people who have disorders of muscles and joints including autoimmune diseases. Most rheumatological disorders have local effects on the involved joints in addition to systemic features. Therefore, when assessing a patient with a rheumatological disorder, it is important to **focus on the local structures involved in the disease** in addition to following a systematic approach to exploring any associated systemic features.

Pain is a common symptom of rheumatological disorders. Pain might be caused by either nerve damage or tissue damage. Pain due to nerve damage is known as [neuropathic pain](#)

and is not covered in this article. **Nociceptive pain is the term experts use** to refer to pain related to tissue damage. Inflammation and infection are two common causes of tissue damage and both can present with pain.

In large joints, there are many structures that can become inflamed and cause musculoskeletal pain. In a patient with the rheumatological disorder, the pain might be caused by bursitis, enthesitis, tendinitis, tenosynovitis, synovitis, ligamentous injury, capsulitis, or capsule injury.

The definitions of these terms are given below:

Caused by	Definition
Bursitis	Inflammation of the bursa
Enthesitis	Inflammation of the point of insertion of a tendon onto the bone
Tendinitis	Inflammation of the tendon of a muscle
Tenosynovitis	Inflammation of the tendon of a muscle and its synovial sheath
Synovitis	Inflammation of the synovial lining of a joint
Capsulitis	Inflammation of the capsule of a joint

Clinical Evaluation in Rheumatology

The clinical history and physical examination are the most important tools available to the doctor to form a diagnosis of a rheumatological disease and management of the patient (maybe long-term management). The most important symptoms of rheumatological diseases include pain, swelling of a joint, redness and warmth of the involved structure, stiffness of a joint, joint deformity, and systemic symptoms. The systemic symptoms of the rheumatological disease include fever and fatigue.

The physical examination in rheumatology is the same as that of orthopedics and can be summarized in three words: look, feel, and move.

You should look for the following abnormalities/changes in your physical examination:

- Bony deformities and deformities in the alignment of long bones
- Color and appearance of the skin over the affected joints indicating inflammation
- Wasting of muscles
- Soft tissue abnormalities including joint swelling and subcutaneous nodules

You should feel the following abnormalities in your physical examination:

- The temperature of the skin, looking for warmth
- Swelling due to inflammation of a joint or any of the soft-tissue structures around the joint
- Tenderness

Finally, you should move the joints to see if one can elicit or relieve the patient's pain. The range of motion of a joint can be tested with an active and passive movement that can help the physician to assess the disability and deformity caused by the illness.

Common Rheumatological Disorders

In this article, we will focus on crystal deposition arthritis: gout and pseudogout. It is, however, quite important to mention the different types of rheumatological disorders you might encounter in your practice before we discuss crystal deposition arthritis.

Arthritis is the inflammation of joints. The following rheumatological disorders present with arthritis in most patients – Osteoarthritis which might be primary or secondary to trauma that can lead to wear and tear of the joints involved. Crystal arthritis which includes gout and pseudogout. Other rheumatological disorders are Rheumatoid arthritis and Spondyloarthropathies, which include ankylosing spondylitis, psoriatic arthritis, reactive arthritis, and inflammatory bowel disease associated with arthritis. The diseases that are associated with the diseased immune system also come under rheumatological disorders such as rheumatoid arthritis, systemic lupus erythematosus.

Connective tissue diseases are also considered as rheumatological disorders. They can also present with arthritis:

- [Systemic lupus erythematosus](#)
- Systemic sclerosis
- Poly-dermatomyositis
- Mixed connective tissue disease

Vasculitis is a group of inflammatory conditions that are characterized by the inflammation of small, middle, and/or large blood vessels. They include:

- Giant cell arteritis; also known as temporal arteritis
- Takayasu
- Polyarteritis nodose
- Wegener's vasculitis

Gout

Gout is characterized by the **deposition of monosodium urate crystals in the bone, joints, and soft tissue**. Patients might develop severe bone and joint destruction. Gout is considered a disorder of metabolism permitting uric acid or urate to accumulate in tissues and blood. There are usually elevated serum levels of urate. This happens due to either too much intake of uric acids and the inability of the kidney to excrete them completely. When tissues are fully saturated, the urate salts precipitate, creating crystal-like structures. These crystals deposit in the skin and other tissues of joints termed as tophi. These crystals are less soluble under acidic conditions and low temperatures, as evidently observed in cool, peripheral joints, for example, the metatarsophalangeal joint of the big toe.

The microscopic examination of the synovial fluid reveals monosodium urate crystals. Polarized light microscopy shows extracellular birefringent needle-shaped urate crystals. The destruction of the bone might present with a large tophus.

Management of Gout

Gout is managed through the following three stages:

- Administration of prophylaxis for the prevention of acute flares.
- Early treatment of acute gout attack

- Reducing the excessive accumulation of urate to prevent gouty arthritis and tissue deposition of urate crystals.

Treatment of an Acute Gout Attack

The treatment of an acute gout attack includes **nonsteroidal anti-inflammatory drugs, oral prednisone, or colchicine**. The nonsteroidal anti-inflammatory drugs are considered as first-line therapy for an acute gout attack. They should be started within 24 hours of the onset of symptoms for optimum efficacy.

Oral prednisone might be needed in complicated cases that are not responsive to nonsteroidal anti-inflammatory drugs alone. NSAIDs are not recommended for the patients having a history of renal or hepatic disease, bleeding problems or ulcers in the stomach and cardiovascular disease. Colchicine is quite effective in managing the acute attacks of gout, especially if started within the first 24 hours of symptoms' onset.

Prophylactic Treatment of Gout

The long-term management of gout focuses on **reducing serum uric acid levels**. This is achieved with allopurinol or any of the newer uric acid reducing agents:

- Febuxostat
- Probenecid
- Lesinurad
- Pegloticase

Complications of gout

These complications include

- Nervous system and spinal cord impingement.
- Fractures in gout affected joints.
- chronic degenerative arthritis
- Increased susceptibility to primary and secondary infections
- Urate and uric acid nephropathy
- Development of renal stones

Pseudogout: Calcium pyrophosphate dihydrate crystal deposition disease

Calcium pyrophosphate dihydrate crystal deposition disease is the third common inflammatory disease of the joints. The Calcium pyrophosphate dihydrate (CPP) crystals that produce pseudo gout contains a combination of inorganic pyrophosphate and calcium. The inorganic pyrophosphate is produced in large part by ectonucleotide phosphodiesterase pyrophosphatase (ENPP1), a catalytic enzyme found in chondrocytes of cartilage, and the pyrophosphate is transferred by the membrane transporter.

In pseudogout, there is calcium pyrophosphate dihydrate crystals deposition in the periarticular tissues of joints and soft tissue. It can induce remarkable damage to the affected joints. The symptoms of pseudogout are a pain, swelling, and heat in the affected joint. In contrast to gout, **laboratory investigations in pseudogout are usually negative**.

An x-ray of the affected joint reveals the calcification of the cartilage and soft tissue of the inflamed area. The examination of the synovial fluid shows the characterized crystals. The deposition of the crystals might involve the bursa, the hyaline cartilage, fibrocartilage, capsule, synovium or tendon of the affected joint.

Conditions associated with crystal deposition pose an increased risk of osteoarthritis. Pseudogout is more common in the elderly and its symptoms resemble symptoms of osteoarthritis or rheumatoid arthritis that are long-lasting. The synovial fluid contains the characteristic crystals and is usually blood-stained. Soft-tissue calcifications can be readily seen on plain radiography of the affected joint. The joints that are most commonly affected by pseudogout are shoulders, elbows, wrists, hands, ankles, and others.

Treatment of Pseudogout

The main focus of the treatment is a cessation of the inflammation. The treatment of pseudogout is usually more aggressive than that of gout and usually includes **combination therapy**. Nonsteroidal anti-inflammatory drugs are usually combined with a corticosteroid for optimum efficacy. If there is significant joint effusion, joint aspiration might be needed. Cortisone injection can help in reducing inflammation.

Joint aspiration might lead to the inoculation of the synovial fluid with bacteria and infection. However, it is still recommended to perform a joint aspiration for the evaluation of the synovial fluid to reliably exclude septic arthritis, a rare but serious condition.

Colchicine is given in small daily doses with optimal hydration to ensure long-term prevention of recurrent pseudogout.

References

Arthritis Foundation:

<https://www.arthritis.org/about-arthritis/types/calcium-pyrophosphate-deposition-disease-cppd/>

Arthritis Foundation:

<https://www.arthritis.org/about-arthritis/types/gout/treatments/types.php>

University of Pretoria, Department of Library Services, An Introduction to Rheumatology

Legal Note: Unless otherwise stated, all rights reserved by Lecturio GmbH. For further legal regulations see our [legal information page](#).