Calcium Pyrophosphate Deposition (CPPD) (formerly called Pseudogout)

Joint problems caused by crystals of a calcium salt called pyrophosphate may be one of the most misunderstood forms of arthritis. Joint problems seen with these crystals often are mistaken for gout and other conditions.

Proper diagnosis (detection) is important. Untreated calcium pyrophosphate deposition (CPPD) may lead to severe, painful attacks or chronic (long-term) pain and inflammation. Over time, joints may degenerate, or break down, resulting in chronic disability. Some treatment options for the arthritis pain do exist, but these do not treat the underlying crystal deposition. Some of the underlying causes are treatable and should be evaluated in people with CPPD (see causes below).

Fast facts
- The risk of CPPD greatly increases with age, but it can occur in young people, too.
- Proper diagnosis depends on detecting calcium pyrophosphate crystals in the fluid of an affected joint.
- Anti-inflammatory medications, which reduce pain and swelling, prevent or help relieve symptoms that you feel in your joints.

What is CPPD?
CPPD is a type of arthritis that, as the old name of pseudogout suggests, can cause symptoms similar to gout. Yet in CPPD, a different type of crystal deposit triggers the reaction.

CPPD can cause bouts of severe pain and swelling in one or more joints, which can limit activity for days or weeks. It also can cause a more lasting arthritis that mimics osteoarthritis or rheumatoid arthritis. The condition most often involves the knees, but can affect wrists, shoulders, ankles, elbows, hands or other joints.
What causes CPPD?
CPPD develops when calcium pyrophosphate crystals build up in a joint. Crystals deposit first in the cartilage (the tissue that cushions inside joints) and can damage the cartilage. The crystals also can cause inflammation that leads to joint pain, warmth and swelling.

In most cases, it is not clear why the crystals form, although crystal deposits clearly increase with age. Because the condition sometimes runs in families, genes likely play a role.

Other factors that can contribute to CPPD include:
- Excess iron storage (medical term: hemochromatosis)
- Low magnesium levels in the blood (hypomagnesemia)
- An overactive parathyroid gland (hyperparathyroidism)
- Some causes of excess calcium in the blood (hypercalcemia)
- A severely underactive thyroid (hypothyroidism)

Who gets CPPD?
The calcium pyrophosphate crystal deposits of CPPD affect about 3% of people in their 60s and as many as 50% of people in their 90s. The release of the calcium pyrophosphate crystals into the joint fluid can attract white blood cells, leading to a painful attack. Attacks of acute (short-lasting) arthritis can occur after injury to the joint or after joint surgery or other surgery. Such attacks also can happen without a clear reason.

Calcium pyrophosphate crystals often are found in the cartilage and even synovial fluids of older people who have no symptoms. Many people who have these crystal deposits will never have acute gout-like attacks or chronic arthritis. These crystals also are often present in people with osteoarthritis or coincidentally in other types of arthritis such as gout or joint infections.

How is CPPD diagnosed?
Diagnosis hinges on symptoms and medical test results. Your doctor may use a needle to take fluid (called synovial fluid) from a swollen or painful joint, to find out whether calcium pyrophosphate crystals are present. An X-ray of the joint may help detect whether calcium-containing deposits are present in the cartilage. Doctors call this X-ray appearance chondrocalcinosis, which is almost always due to CPPD.

Your doctor must rule out other potential causes of symptoms. These include gout, rheumatoid arthritis and joint infection. Your doctor also may do blood tests to check for some of the underlying causes listed above.

How is CPPD treated?
No treatment is available to dissolve the crystal deposits. For patients who have acute attacks, the doctor may prescribe nonsteroidal anti-inflammatory drugs, which are commonly called NSAIDs. NSAIDs like indomethacin (Indocin) and naproxen (Naprosyn) treat pain and swelling during severe attacks.
People with poor kidney function, who have a history of stomach ulcers and/or who are on blood thinners often cannot take NSAIDs. (See the fact sheet on NSAIDs.) For these patients, it may help to have your doctor drain the joint fluid and inject a corticosteroid into the affected joint. To try to prevent further attacks, low doses of colchicine (a medicine used more often for gout) or NSAIDs may prove effective.

Other medicines may help some patients during severe attacks of calcium pyrophosphate crystal arthritis or with the less common chronic inflammation that these crystals can cause. These drugs include hydroxychloroquine (Plaquenil, Quineprox), methotrexate (Rheumatrex, Trexall) or an “interleukin 1 beta antagonist” that can decrease inflammation, such as the biologic medicine anakinra (Kineret). Anakinra is approved by the government for treatment of rheumatoid arthritis.

Surgery to repair and replace damaged joints is an option in severe cases.

Prevention
Experts do not know how to prevent these crystals. If CPPD is due to some other medical problem, such as hemochromatosis or parathyroid problems, treatment of that condition may sometimes prevent CPPD from getting worse.

Points to remember
- CPPD may be hard to diagnose because the joint pain and other symptoms can mimic gout and other types of arthritis.
- Diagnosis is confirmed by using a microscope to see small calcium pyrophosphate crystals in joint fluid.
- Anti-inflammatory drugs can help lessen symptoms, but there is no way to get rid of the crystals.

The rheumatologist's role in the treatment of CPPD
Rheumatologists are actively researching the causes of CPPD to better prevent and treat this form of arthritis. Because people with CPPD tend to be older and more prone to side effects from anti-inflammatory medications, they may benefit from seeing a rheumatologist, who can offer expertise in using such drugs and other therapies.

Rheumatologists are experts at detecting CPPD and directing a team approach to diagnosis of underlying conditions, treatment, and efforts to help prevent possible joint damage these crystals may sometimes cause. This team approach is important, because the patient may need advice about surgery or may need more information and support from other health care providers. These include physical and occupational therapists and nurses.

To find a rheumatologist
For a list of rheumatologists in your area, click here.

Learn more about rheumatologists and rheumatology health professionals.
For more information
The American College of Rheumatology has compiled this list to give you a starting point for your own additional research. The ACR does not endorse or maintain these Web sites, and is not responsible for any information or claims provided on them. It is always best to talk with your rheumatologist for more information and before making any decisions about your care.

The Arthritis Foundation  
www.arthritis.org

MayoClinic.com Pseudogout Information  
www.mayoclinic.com/health/pseudogout/DS00717

National Institutes of Health MedlinePlus  

UpToDate Patient Information: Pseudogout  
http://www.uptodate.com/contents/pseudogout-beyond-the-basics?source=search_result&search=pseudogout&selectedTitle=1%7E3

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Written by H. Ralph Schumacher, MD, and reviewed by the American College of Rheumatology Communications and Marketing Committee.

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