

Validated classification criteria for calcium pyrophosphate deposition (CPPD) disease

February 1 2024

Phase 1: Item generation

- Scoping literature review
- Patient co-investigators & Combined Expert Committee (CEC) suggestions

Phase 2: Item reduction &

- · CEC meeting to discuss "difficult" items
- Advisory Group discussions to reduce items & refine definitions
- Item rating exercise by individual CEC members

reduction & refinement

Phase 3: Item weighting &

threshold

score

- Derivation cohort recruitment & adjudication
- Patient profile ranking exercise
- Associations between potential items and CPPD disease
- · Draft classification criteria framework
- Item weights via multi-criterion decision analysis exercise
- Threshold score determination

Phase 4: Validation

- Validation cohort recruitment & adjudication
- · Sensitivity and specificity calculated

ACR/EULAR CPPD Classification Criteria

Overview of the ACR/EULAR classification criteria for CPPD disease across the 4 Phases. Credit: *Annals of the Rheumatic Diseases* (2023). DOI: 10.1136/ard-2023-224575



Calcium pyrophosphate deposition (CPPD) disease is a common, symptomatic arthritis, but until now there have been there are no validated classification criteria. This has hampered research in this common disease that becomes more frequent with increasing age.

To address this, EULAR—The European Alliance of Associations for Rheumatology—has worked with the American College of Rheumatology (ACR) to develop the first-ever classification criteria, now <u>published</u> in the *Annals of the Rheumatic Diseases*.

People with CPPD disease have diverse presentations, and understanding of this common disease has lagged other types of arthritis—partly because there have been no validated classification criteria. These types of criteria are important to help recruit patients into <u>clinical trials</u>, compare findings between studies, and to perform consistent research.

Additionally, diagnosis of CPPD disease has been difficult because it has historically relied on finding evidence of visible calcification on X-ray, but modern advanced imaging techniques are more sensitive.

Together, these two gaps have left CPPD without a practical gold standard in the clinic. Addressing this could help to develop better clinical understanding and options and move the disease forward for patients.

To develop validated classification criteria, EULAR and the ACR set up an international collaborative working group. The goal was to develop a framework that would help investigators to reliably identify people with CPPD disease to include in research studies.

To begin, a list of possible criteria items was developed based on the



available literature and expert input. This was then discussed among a group of experts, and the list condensed and refined. The key criteria that were sufficient to classify someone with arthritis as CPPD disease were presence of calcium pyrophosphate crystals in <u>synovial fluid</u> or the presence of crowned dens syndrome (a painful arthritis affecting the neck).

In the absence of calcium pyrophosphate crystals or crowned dens syndrome, a score based on the presence of clinical and imaging findings can also distinguish CPPD disease. The EULAR and ACR working group developed a classification tool that uses the presence of clinical features, <u>metabolic disorders</u>, and laboratory and imaging investigations.

Each item has a weighted number of points, and people with an overall score greater than 56 on a 0–100 point scale are classified as having CPPD. When tested in 251 people with CPPD, and 162 with another similar disease, these new criteria had a sensitivity of 99.2% and specificity of 92.5% in the validation cohort.

These new classification criteria for CPPD disease represent the first validated criteria set for the condition, with robustly validated performance characteristics. EULAR hopes that future studies of CPPD will use these as inclusion criteria for participant screening and enrollment.

More information: Abhishek Abhishek et al, The 2023 ACR/EULAR classification criteria for calcium pyrophosphate deposition disease, *Annals of the Rheumatic Diseases* (2023). DOI: 10.1136/ard-2023-224575

Provided by EULAR



Citation: Validated classification criteria for calcium pyrophosphate deposition (CPPD) disease (2024, February 1) retrieved 27 April 2024 from https://medicalxpress.com/news/2024-02-validated-classification-criteria-calcium-pyrophosphate.html

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