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INTRODUCTION

The **SAPHO syndrome** (synovitis, acne, pustulosis, hyperostosis, and osteitis) is a rare disease (<1/10,000) with skin and musculoskeletal manifestations. The typical dermatological manifestations (DM) are palmoplantar pustulosis (PPP), vulgar psoriasis (VP) and severe acne (SA).

Musculoskeletal manifestations (ME) are diverse, including the anterior chest wall (ACW), spine, temporomandibular joint, and peripheral joints. It is considered as part of spondyloarthropathies (SpA), however, no significant relationship with HLA-B27 has been found.

For treatment, non-steroidal anti-inflammatory drugs (NSAIDs), disease-modifying drugs (DMARDs) and bisphosphonates (BPP) have been traditionally used. More recently, new therapies such as biological therapy (BT) and other molecules are being used more frequently.

OBJETIVES: To describe the clinical and diagnostic characteristics of patients with SAPHO syndrome evaluated in Rheumatology services in Spain.

METHODS

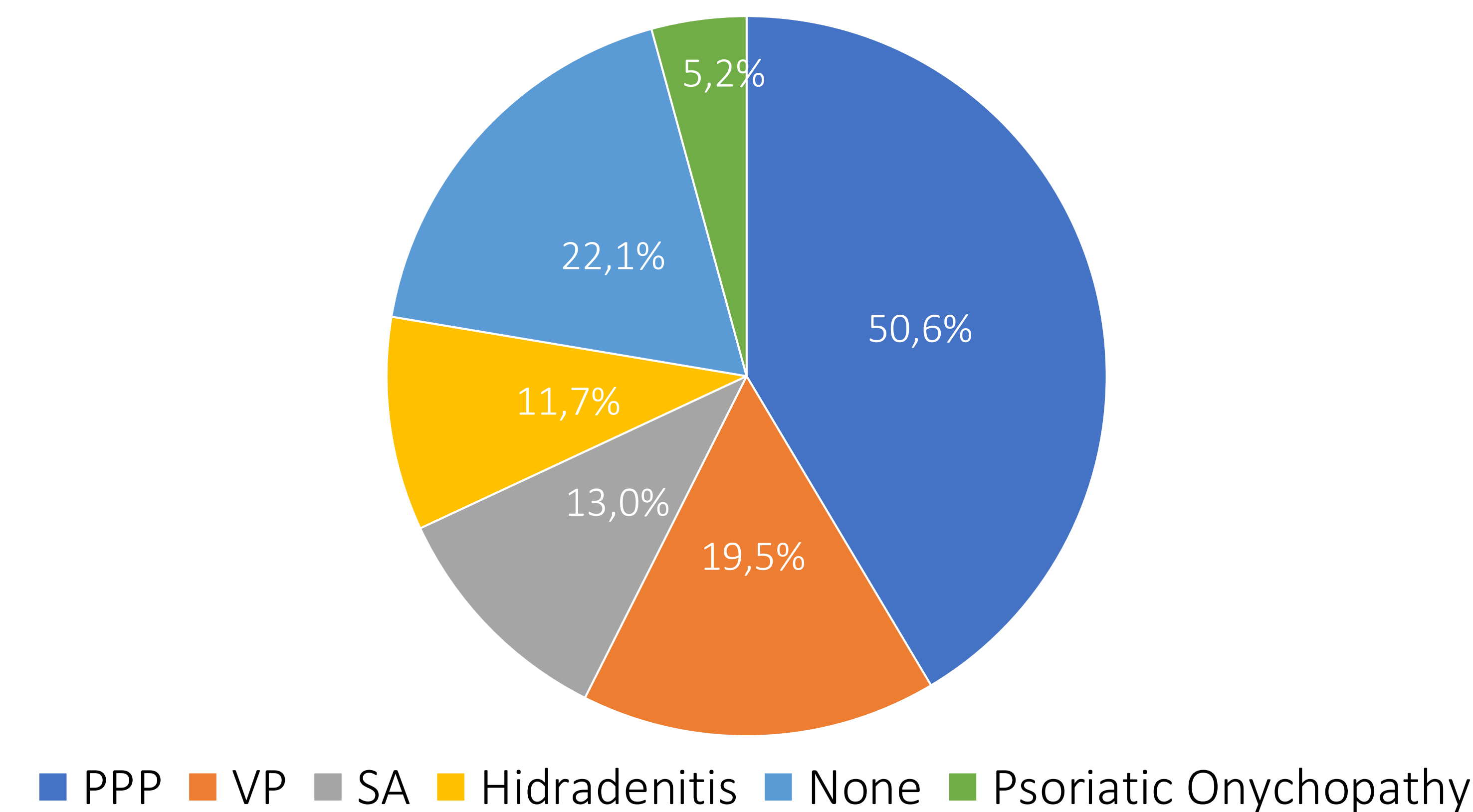
Descriptive, cross-sectional, retrospective, multicenter study, carried out in Rheumatology services of several hospitals in Spain.

RESULTS

77 patients (75% women) with a mean age of 50.27 years (± 15.46) were included. 94.8% of the patients were Caucasian, 2.6% Latin American and 2.6% Arab.

The mean age of diagnosis was 43 years (± 15.22). The most frequent initial diagnosis was SAPHO in 59.7%, followed by Psoriatic Arthritis 16.9%, SpA 7.8%, Undifferentiated Arthritis 6.5%, Chronic Recurrent Multifocal Osteomyelitis 5.2%.

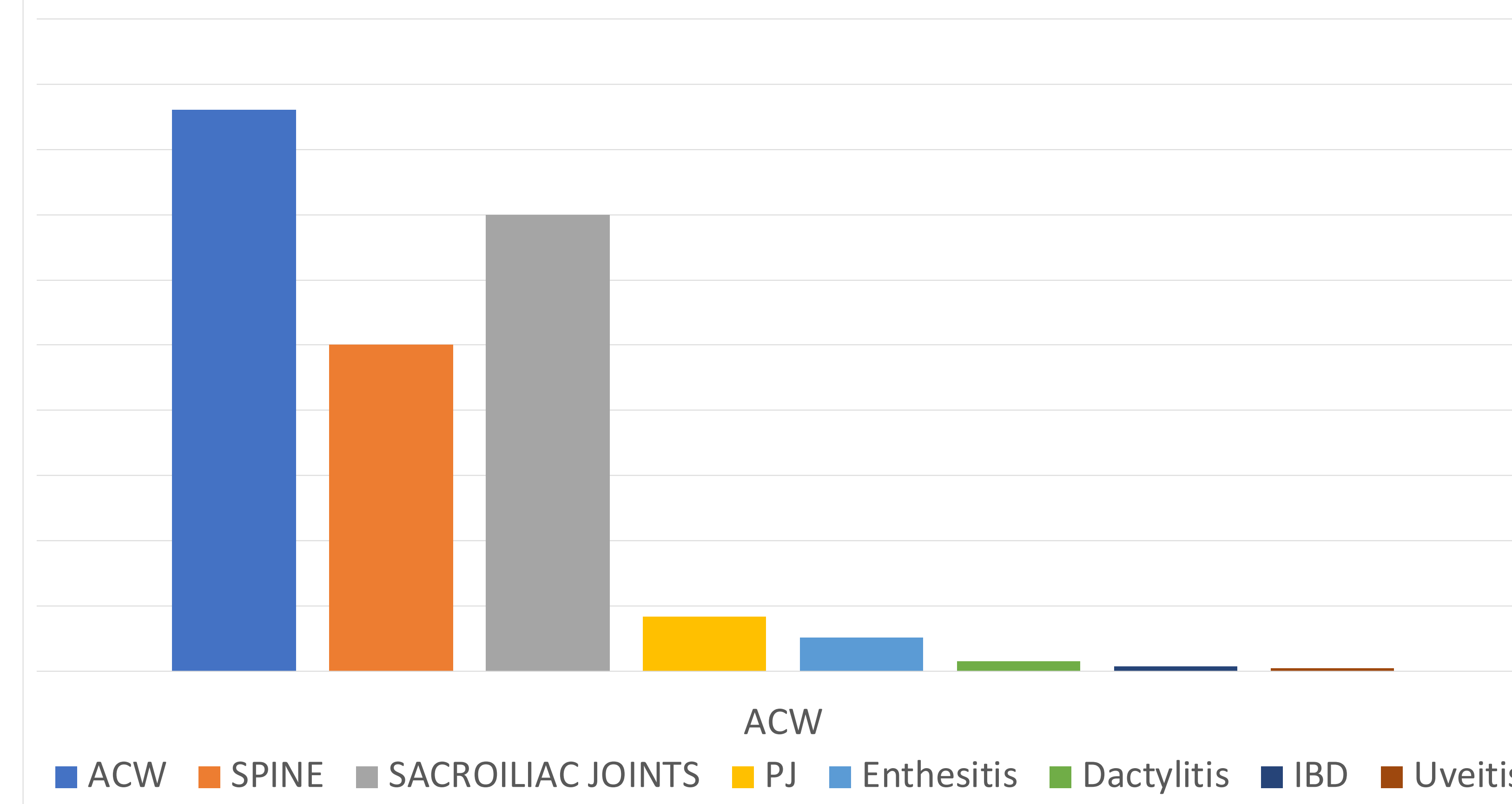
Figure 1. Dermatological manifestations (%)



The age of onset of skin symptoms was 38.22 years (± 12.95) and MSK was 38.64 years (± 13.17).

The most frequent location of MSK compromise was the ACW with 76.6%, followed by the spine in 36.4%, sacroiliac joints 26 and PJ in 42.9%, with the oligoarticular form being the most frequent in the latter. Among other manifestations related to SpA, 26% presented enthesitis, 7.8% dactylitis and rarely IBD (3.9%) along with uveitis (2.6%).

Figure 2. Musculoskeletal manifestations (%)



Analytically, 12% were HLA B27+ and only 4% were HLA cw6+. Acute phase reactants were slightly elevated, RCP 10.54 (± 16.76), ESR 23.58 mm (± 27.84).

Diagnostic imaging tests for diagnosis were performed in almost all patients, the most frequent being XR in 84.4% followed by MR 71.4%, scintigraphy 67.5%, CT 57.1% and finally US in 39%.

The culture of lesions in our cohort was only performed in 13 patients and the bone biopsy in 7.

The most frequent course of the disease was the relapsing remitting (45.5%) followed by the chronic form (33.8%) and only 16.9% presented a single outbreak of the disease.

18.2 of the patients required hospitalization due to disease activity.

CONCLUSION

- The mean age at presentation of DM and MSK symptoms was similar.
- The most common DM was PPP, although less frequently than that described in other series.
- The most frequent MSK involvement was ACW. PJ involvement was frequent compared to other series.
- There are currently no series that report uveitis as an associated manifestation of SAPHO.
- More than 40% of the patients in our cohort required biological therapy synonymous with the severity of this disease in some cases

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