

# MATTERS ARISING

## Commented glossary for rheumatic spinal diseases

The glossary on rheumatic spinal diseases by a study group of the Committee of Pathology of EULAR<sup>1</sup> has certainly helped clarify many terms. This comment on spondylarthropathy is intended likewise to help with the terminology. The terms spondylarthritis and spondylarthropathy, as we understand it, are now both used to describe a partly heterogeneous group of diseases that have a number of features in common: familial aggregation, association with HLA B27 and probably other genetic factors, and several, partly overlapping, characteristic clinical symptoms.

The authors recommend use of the term 'spondylarthritis' originally proposed by Moll and Wright in 1974,<sup>2</sup> their main argument being that the term spondylarthritis (i) is original and historical, and (ii) emphasises the inflammation feature by including arthritis, while (iii) the term 'spondylarthropathy' can refer to any degenerative disease of the spine.

We believe that neither the term spondylarthritis nor spondylarthropathy can perfectly reflect the clinical and pathological background of this overlapping disease spectrum, and we prefer the term spondylarthropathy, for the following reasons:

- (1) The historical dimension and originality of the term spondylarthritis is unimpressive, as the authors have to correct the original definitions introduced by Wright *et al*<sup>3,4</sup> by excluding Whipple's disease and Behçet's disease.<sup>1</sup> We agree that these two diseases should be excluded from the spectrum because they lack HLA B27 association and have a distinctive clinical picture and pathogenesis.
- (2) In addition, the spectrum of clinical symptoms included in this conflation of spondylitis and peripheral arthritis, which had been listed by the 'Leeds group' in 1987,<sup>3</sup> has changed since the introduction of the criteria introduced by the European Spondylarthropathy Study Group (ESSG).<sup>4</sup> Features such as erythema nodosum and thrombophlebitis<sup>3</sup> are no longer considered essential to the spondylarthropathies.
- (3) These classification criteria have been developed and evaluated by leading European rheumatologists who agreed on the term spondylarthropathy.<sup>4</sup> Many other rheumatologists in Europe and the United States have approved these criteria, which have now gained wide international acceptance.<sup>5-8</sup>
- (4) An advantage of the term spondylarthropathy in clinical use is that it is applicable to a group of patients suffering spondylarthropathy that is now frequently reported as 'undifferentiated spondylarthropathy'.<sup>8,9</sup> When established criteria for more closely defined subcategories of spondylarthropathies such as ankylosing spondylitis are used,<sup>10</sup> these patients often received no proper diagnosis.
- (5) Arthritis need not be included in the general term, as not all patients with spondylarthropathy suffer arthritis (patients with inflammatory back pain, enthesopathy, uveitis). Of the 403 spondylarthropathy patients evaluated using the ESSG criteria,

only 35.3% had synovitis of the lower limbs, while 56.4% had enthesopathy (at any site). (6) Arthritis need not be included in the general term to exclude the so called degenerative diseases of the spine, as there has always been an argument as to whether these diseases should be primarily labelled as non-inflammatory, the problem is reflected in the differing terminology 'osteoarthritis' and 'osteoarthrosis'. From this point of view there is no clear advantage in using the term spondylarthritis.

(7) Use of the term spondylarthropathy for the spectrum of HLA B27 associated diseases discussed here excludes, by definition, degenerative diseases of the spine such as spondylarthrosis and spondylosis.<sup>4</sup> This is justified because spondylarthropathy has not been used for all arthropathies affecting the spine previously and there is no real need for a common term to describe these heterogeneous diseases.

(8) Other terms used to group rheumatological disease categories such as 'connective tissue diseases', which have been used for decades, are also far from being perfect.

In summary, no term is perfect, but agreement is needed. 'Spondylarthropathy' seems to us preferable because classification criteria using this term have been evaluated, the term is now frequently used, and it has a better chance of being accepted internationally.

Finally, we agree that the German term Bechterew's disease, having once been popular for describing patients with very severe ankylosing spondylitis and a bad disease course, should be avoided, especially in early disease, because young patients should not be burdened with an unnecessarily pessimistic prognosis.

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- 1 Francois R J, Eulerink F, Bywaters E G L. Commented glossary for rheumatic spinal diseases, based on pathology. *Ann Rheum Dis* 1995; 54: 615-25.
- 2 Moll J M H, Haslock I, MacRae I, Wright V. Associations between ankylosing spondylitis, psoriatic arthritis, Reiter's disease, the intestinal arthropathies, and Behçet's syndrome. *Medicine* 1974; 53: 343-64.
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- 8 Braun J, Bollow M, Egges U, König H, Distler A, Sieper J. Use of dynamic magnetic resonance imaging with fast imaging in the detection of early and advanced sacroiliitis in spondylarthropathy patients. *Arthritis Rheum* 1994; 37: 1039-45.
- 9 Zeidler H, Mau W, Khan M A. Undifferentiated spondylarthropathies. *Rheum Dis Clin North Am* 1992; 18: 187-202.
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Any attempt to establish a glossary for rheumatological disease description is of interest, especially when conducted in a manner that transcends national borders.<sup>1</sup> Deciding to accept or reject a current usage, and suggesting new terms, such as the neologism 'spondylarthritis', represents an ambitious and stimulating approach. It did seem a little unusual not to find the osteoarthrosis-osteoarthritis terminology debate considered (although perhaps the authors feel it is resolved), while attempt was made to establish the category 'spinal rheumatoid arthritis.' If it is desired to establish such a term, it seems reasonable to characterise this term further, to ensure ability to distinguish rheumatoid arthritis and spondylarthropathy/spondylarthritis.

Spondylarthropathy/spondylarthritis is universally recognised on the basis of sacroiliac joint erosions and fusion, syndesmophytes, and zygapophyseal joint fusion,<sup>2-7</sup> findings which should allow at least a proportion of individuals with that category of arthritis to be readily distinguished from those with rheumatoid arthritis.<sup>2,3,5-11</sup> One obvious issue relates to the nature of axial disease.

In contrast with zygapophyseal joint fusion, valid identification of zygapophyseal joint erosions has been compromised by radiological artefacts.<sup>12</sup> The culprit proved to be the thin nature of zygapophyseal articular cortices. Thus loss of cortex as a result of eburnation from osteoarthritis was not radiologically distinguishable from that caused by erosion.<sup>12</sup> The fronts of resorption and remodelling that characterise the latter are below the resolution of clinical radiography equipment. This confusion led to the misconception that zygapophyseal joint erosion was occasionally found in rheumatoid arthritis. Validated analysis revealed that true erosions are specific for spondylarthropathy/spondylarthritis.<sup>12</sup>

Eric Bywaters' eloquent report and discussion of spinous process bursal involvement in rheumatoid arthritis<sup>13</sup> is quite different from the zygapophyseal phenomenon seen in spondylarthropathy/spondylarthritis. John Ball's article<sup>14</sup> (cited in the glossary<sup>1</sup>) commented on zygapophyseal joint fusion and erosion in rheumatoid arthritis. However, clinical (radiological) recognition of zygapophyseal joint erosion is fraught with artefact, precluding clinical reliability (as documented above)—and diagnosis must be questioned for those instances where fusion is reported.

The last issue pertains to diagnosis of rheumatoid arthritis and the lumper-splitter controversy.<sup>15</sup> The challenge relates to lack of axial joint involvement in 40-60% of the population with spondylarthropathy/spondylarthritis.<sup>2,3,5-7,16</sup> Many of this group are now distinguished from those with rheumatoid arthritis on the basis of normal periarticular bone density and presence of reactive new bone. It is unclear, however, if this perspective had developed by the 60s and 70s, when cited articles on rheumatoid spine disease<sup>1</sup> were published. Among any population with spondylarthropathy/spondylarthritis there are a few individuals with polyarticular disease. The presence of axial joint disease in at least some of those individuals facilitates diagnosis.<sup>17,18</sup>

If 'spinal rheumatoid arthritis' is to be considered a 'definition,' it seems premature to utilise the term until further characterisation ensures the ability to distinguish it from the changes of spondylarthropathy/

spondylarthritis. I look forward to the authors' continued effort.

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- 1 François R J, Eulerink F, Bywaters E G. Commented glossary for rheumatic spinal diseases, based on pathology. *Ann Rheum Dis* 1995; 54: 615–25.
- 2 Rothschild B, Martin L, eds. *Paleopathology: disease in the fossil record*. London: CRC Press, 1993.
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- 14 Ball J. Enthesopathy of rheumatoid and ankylosing spondylitis. *Ann Rheum Dis* 1971; 30: 213–23.
- 15 Rothschild B M. The challenge of diagnosing arthritis from skeletal remains. *Clin Exp Rheumatol* 1995. In press.
- 16 Rothschild B, Woods R. Spondyloarthropathy. Erosive arthritis in representative defleshed bones. *Am J Phys Anthropol* 1991; 85: 125–34.
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Reading the article by François, Eulerink and Bywaters<sup>1</sup> reminded us of a Latin sentence we learned as students: *Tres faciunt collegium sed non universitatem litterarum*—in this context: at least three students have to attend to make it a lecture, but three professors, teachers or experts, etc cannot expect their opinion to be shared all over the world. Thus the commented glossary by François and colleagues is not a consensus article, but merely a recommendation of experts asking for discussion.

We would like to make our contribution.

Basically there are three pathological bony spurs to be found on the spine:<sup>2</sup> the spondylophyte in degenerative disc disease, the syndesmophyte in ankylosing spondylitis, and the parasyndesmophyte in psoriatic spondylitis and in spinal involvement of Reiter's syndrome.

The authors mentioned the parasyndesmophyte; however, there is missing any reference to the two diseases mentioned above of which parasyndesmophytes are indicative. They do mention paravertebral ossification, but no mention is made that this is an inaccurate synonym of parasyndesmophyte. Paravertebral ossifications *sensu lato* are also seen with fibrodysplasia ossificans progressiva and with tetra- and paraplegia.

François and colleagues prefer the acronym SAPHO syndrome. In 1987, Chamot *et al*<sup>3</sup> described 'Le syndrome acné pustulose hyperostose ostéite' and used the acronym SAPHO—that is, this acronym reflects the first letters of this rather long denomination of this syndrome. In principle, an acronym is used as a secondary term and artificial word, but this seems to be not the case with SAPHO. In 1988, Benhamou *et al* (including Chamot)<sup>4</sup> defined the acronym SAPHO in a different way, to adopt it for English literature: synovitis-acne-pustulose hyperostosis-osteomyelitis syndrome. This may speak in favour of the authors' inventive abilities, but from a scientific point of view the conversion remains questionable and seems confusing.

Use of the well substantiated and established denomination 'acquired hyperostosis syndrome',<sup>5,6</sup> instead of SAPHO syndrome, pustulotic arthro-osteitis or more than 40 other synonyms<sup>7</sup> of the same syndrome, is rejected by François and colleagues, on the grounds that 'this term might be confused with diffuse idiopathic hyperostosis'; this sentence requires the addition of the phrase 'if used by amateurs' to complete it.

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- 1 François R J, Eulerink F, Bywaters E G L. Commented glossary for rheumatic spinal diseases, based on pathology. *Ann Rheum Dis* 1995; 54: 615–25.
- 2 Dihlmann W. *Radiologic atlas of rheumatic diseases*. New York: Thieme Inc, 1986; 176, 228, 256.
- 3 Chamot A M, Benhamou C L, Kahn M F, Beranek L, Kaplan G, Prost A. Le syndrome acné pustulose hyperostose ostéite (SAPHO). Résultats d'une enquête nationale. 85 observations. *Rev Rhum* 1987; 54: 187–96.
- 4 Benhamou C L, Chamot A M, Kahn M F. Synovitis-acne-pustulosis hyperostosis-osteomyelitis syndrome (Sapho). A new syndrome among the spondyloarthropathies? *Clin Exp Rheumatol* 1988; 6: 109–12.
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- 6 Dihlmann W, Schnabel A, Gross W L. The acquired hyperostosis syndrome: a little known skeletal disorder with distinctive radiological and clinical features. *Clin Invest* 1993; 72: 4–11.
- 7 Kahn M F, Kahn M A. The SAPHO syndrome. *Baillière's Clin Rheumatol* 1994; 8: 333–62.

**AUTHORS' REPLY:** We have been very interested by the comments made by several readers on the paper 'Commented glossary for rheumatic spinal diseases, based on pathology'.

We agree with Drs Braun and Sieper that neither spondylarthritis nor spondyloarthropathy are really adequate terms for the group of

diseases to be covered, especially if not all patients exhibit spinal disease or peripheral arthritis. 'B27-related-diseases' might be better, but is not perfect either, because B27 has only a statistical link to the disease and is not present in every patient.

Our working group did not support 'spondyloarthropathy' because its etymological meaning is 'any vertebral or spinal disease'. We are aware that people use it in a more restricted sense, but why call a hospital a 'building' instead of a hospital?

It is the responsibility of people who propose new terms to elaborate them according to existing rules<sup>1</sup> and to avoid using any term for any disease without taking account of the significance of the word proposed.

We do not deny the value of the criteria elaborated by the European spondyloarthropathy study group (ESSG); we just regret that the ESSG adhered to a term that we consider inappropriate.

Taking account of the weight of common practice—our fifth methodological rule—we proposed a term that would not be too different from spondyloarthropathy. We recommended two alterations.

The first is the inclusion of a connective –o–, which is already very much used in the USA.<sup>2–4</sup> In spondyloarthropathy, the connective –o– before the vowel indicates the association between spinal and another arthritis. Incidentally, the ESSG criteria, but not always the term spondyloarthropathy, gained international recognition: Khan, quoted by Braun and Sieper, uses spondyloarthropathy.

The second alteration consists in replacing arthropathy by arthritis, which is more accurate. We all agree that degenerative conditions may exhibit some low grade inflammation and that chronic inflammatory diseases are exposed to secondary osteoarthritis and to mechanical factors, but surely nowadays nobody believes it useful to go back to Beneke<sup>5</sup> who called degenerative spinal conditions 'spondylitis', and to Marie and Léry<sup>6</sup> who considered ankylosing spondylitis (AS) to be a form of 'spondylitis'?

To Dr Rothschild, we want to emphasise that our neologism is not 'spondylarthritis', which means spinal arthritis, but 'spondyloarthritits'.

Dr Rothschild casts doubt on the existence of rheumatoid lesions in the spine. His observations are based on defleshed bones. Study of fresh cadavers seems much more pertinent to the description of evolving and early changes.

Histopathologists have actually observed synovitis, pannus, and rheumatoid granulomata in the discal<sup>7–11</sup> and zygapophysal<sup>10</sup> joints of rheumatoid arthritis patients. Their observations, however, were mainly anecdotal. To our knowledge, a true systematic and comparative study of the spinal changes associated with AS and rheumatoid arthritis (RA) is still lacking. The fact that a minority of RA patients develop zygapophysal fusion is insufficient to change their diagnosis to one of spondyloarthritits; there are indeed several ways to reach ankylosis. RA and AS spinal changes are not characterised by a single pathognomonic lesion, but by a typical constellation of partly shared findings.

RA spinal changes also differ from degenerative changes. Eulerink *et al* have published a macroscopic study of 44 rheumatoid cervical spines compared with 44 control cervical spines matched for age and gender.<sup>12</sup> They did report significant