Editorial

The Need to Take a Fresh Look at Criteria for Hypermobility

Joint hypermobility is a phenotypic feature shared by most (if not all) heritable disorders of connective tissue (HDCT), and there are abundant reasons for requiring reliable and accurate criteria for their precise diagnosis. First and foremost it is imperative to recognize diseases with potentially life-threatening consequences, such as the Marfan syndrome and vascular-type Ehlers-Danlos syndrome (formerly known as EDS type IV), if lives (in particular, young lives) are not to be needlessly lost. Few would argue with this assertion. However, at the more "benign" end of the hypermobility spectrum confusion arises in clinical assessment: there is still a widely held view among rheumatologists that hypermobility, as identified using the Beighton 9-point or a similar scale[1], predominantly represents the upper end of a spectrum of normal range joint motion. The weight of evidence accumulated over the last 25 years, however, supports the notion that, at least in the clinical setting, hypermobility is the outward manifestation of an underlying (albeit mild) systemic HDCT, indistinguishable from, if not identical to, the hypermobility type EDS (formerly EDS type III)[2]. Moreover, there is accumulated evidence of connective tissue laxity identified in various sites throughout the body, including the skin, eye, skeleton, heart, and — more familiar to the rheumatologist — the locomotor apparatus[3].

The original description of the syndrome[4] comprised 2 elements, hypermobility and associated symptoms. Forty years on, the scope of the definition has broadened: the 1998 Brighton Criteria (published in 2000 in *The Journal*[5]) is the successor instrument for classification and diagnosis of what has now become the (benign) joint hypermobility syndrome (BJHS); the criteria include the systemic phenotypic features (Table 1), while retaining the 2 elements of the original definition (hypermobility plus symptoms), as well as the Beighton score, albeit in a more flexible format.

Yet publication of the Brighton Criteria has not led to the anticipated and hoped for greater acknowledgment, understanding, or recognition of JHS among fellow rheumatologists[6]. Paradoxically, however, several investigators have usefully applied the Brighton Criteria to define cohorts of JHS patients for selection into studies, and the results have considerably expanded our fund of knowledge of the wide-ranging ramifications of JHS beyond the confines of the musculoskeletal system or the boundaries of rheumatology. Important newly opened areas of neurophysiology include joint proprioception impairment[7,8], lack of efficacy of local anesthetics[9], and autonomic dysfunction[10]. In addition, advances now extend to the physical therapy of JHS[11] and aspects of performing arts medicine[12].

One unexpected consequence of the application of the Brighton Criteria to research has been the finding of unexpectedly high prevalences of JHS among unselected routine

Table 1. 1998 Brighton classification/diagnostic criteria for the benign joint hypermobility syndrome (BJHS)[5]. BJHS is diagnosed in the presence of 2 major criteria, or one major and 2 minor criteria, or 4 minor criteria. Two minor criteria suffice where there is an unequivocally affected first-degree relative. BJHS is excluded by presence of Marfan or Ehlers-Danlos syndrome (other than the EDS hypermobility type, formerly EDS III) as defined by the Ghent 1996[18] and the Villefranche 1998[19] criteria, respectively. Criteria Major 1 and Minor 1 are mutually exclusive, as are Major 2 and Minor 2.

MAJOR CRITERIA
1. Beighton score ≥ 4/9 (currently or historically)
2. Arthralgia for longer than 3 months in ≥ 4 joints

MINOR CRITERIA
1. Beighton score = 1, 2, or 3/9 (0, 1, 2, 3 if age > 50 yrs)
2. Arthralgia (≥ 3 months) in 1 to 3 joints, or back pain (≥ 3 mo), spondylosis, spondylolysis/spondylolisthesis
3. Dislocation/subluxation in more than one joint, or in one joint on more than one occasion
4. Soft tissue rheumatism, ≥ 3 lesions (e.g., epicondylitis, tenosynovitis, bursitis)
5. Marfanoid habitus (tall, slim, span/height ratio > 1.03, upper:lower segment ratio < 0.89, arachnodactily (positive Steinberg/wrist signs)
6. Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring
7. Eye signs: drooping eyelids or myopia or antimongoloid slant
8. Varicose veins or hernia or uterine/rectal prolapse

See Diagnostic criteria for general joint hypermobility, *page 798*, and Epidemiology of general joint hypermobility, *page 804*

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rheumatology outpatient referrals both in Chile[13] and in the UK[14]. In the UK, rheumatologists' estimates of the number of cases seen annually[15] strongly suggest that the true diagnosis in a majority of patients with JHS is overlooked (up to 95%). If these data are confirmed, the specialty of rheumatology worldwide will justly hang its corporate head in shame.

It is timely that in 2 articles in this issue of *The Journal* Remvig, Jensen, and Ward, in a critical but balanced and painstaking fashion, address diagnostic criteria for hypermobility and BJHS. Their first article[16] describes a computer-driven literature search pertaining to ranges of normal joint motion, localized (or pauciarticular) and generalized hypermobility, and the BJHS. They draw attention to the methodological shortcomings and lack of statistical sophistication apparent in many published reports, in particular the range of cutoff points for the Beighton score used by different authors and a lack of reproducibility and validating studies. Their proposed solution is to develop a set of "gold standards" based on "a consensus of experts."

The second article concentrates on existing criteria for BJHS[17], examines the Brighton Criteria in depth, and concludes that a further revision is needed "to gain better data but also greater international acceptance." Their goals also include "a better definition of normal joint ROM among population-based cohorts sorted according to age, gender, and race; establish cutoff levels that accurately portray group differences, and to implement longitudinal and cross-sectional cohort-based diagnostic and treatment studies." These are worthy goals, ones that few would argue with. The authors generously conclude that "in the meantime the existence of BJHS can be accepted using the present criteria."

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