Editorials

Time to take hypermobility seriously (in adults and children)

A third of a century has elapsed since the hypermobility syndrome (HMS) appeared on the rheumatological horizon [1]. From the outset it was perceived more as a curiosity than as an entity that could have significant (let alone serious) import. In both children and adults it aroused more wonder at the seemingly bizarre contortionist manoeuvres patients could perform (to the misplaced amusement of medical students and their teachers) than a quest for appreciating and alleviating the adverse aspects of the condition. There is now abundant evidence from papers published in peer-reviewed journals in many countries to demonstrate the serious impact that hypermobility can have on peoples’ lives [2]. There is no longer (if there ever was) any justification for regarding hypermobility as merely a circus act [3].

The original description of Kirk et al. [1] defined the HMS as the occurrence of ‘musculoskeletal symptoms in the presence of generalised joint laxity in otherwise normal subjects’. It was diagnosis by exclusion, the phrase ‘otherwise normal’ implying the absence of other identifiable rheumatic disease. Yet much has been learnt since to change our concept of the condition. It has been established from a number of studies [4, 5], first that hypermobility is more often pauciarticular than polyarticular and, secondly that it does not have to be generalized to cause symptoms [6]. Even a single hypermobile joint may suffer any or all of the consequences of laxity, including a tendency to dislocate, develop traumatic synovitis or premature osteoarthritis, or it may just hurt for no visibly obvious reason [2]. Thirdly, the HMS, as seen from the clinical perspective, far from occurring in ‘otherwise normal subjects’, has all the hallmarks of a forme fruste of a heritable disorder of connective tissue (HDCT). That is to say, it is imbued with features that overlap with its more serious cousins—the Marfan and Ehlers–Danlos (EDS) syndromes and osteogenesis imperfecta—though in general its features are milder and lesser in degree [7]. As life expectancy is not reduced, the use of the term ‘benign joint hypermobility syndrome’ (BJHS) has largely replaced the earlier HMS [8]. The British Society for Rheumatology (BSR) Special Interest Group on the HDCTs has recently published a revised set of validated diagnostic criteria for the BJHS [9], which many authorities now believe to be identical with the hypermobility type of the EDS (formerly EDS type III [10]). The BJHS represents a complex mix of acute, recurrent or recalcitrant, widespread soft tissue lesions of traumatic origin, recurrent joint subluxations and/or dislocations, often commencing in childhood or adolescence and continuing into adult life. This painful existence may be compounded by as yet undetermined neurophysiological influences, possibly including nociceptive enhancement. Clues to this possibility are provided by the discovery of two seemingly unrelated, though tantalizing, observations, namely that patients with BJHS/EDS have diminished joint proprioceptive acuity [11] and are less responsive than normal to the local anaesthetic effects of lignocaine [12]. Failure to receive effective intervention not infrequently results in a complicating chronic pain syndrome [13], fibromyalgia [14] and depression [15]. Later in life, premature osteoarthritis may add to the burden of morbidity [2].

In this issue of *Rheumatology* we publish three papers concerned with hypermobility. A paper by Grahame and Bird, is a survey of perceptions about the HMS carried out among UK-based consultant rheumatologist members of the BSR early in 1999 [16]. The questionnaire probed deeply and widely for details of estimated clinic prevalence, criteria for diagnosis, treatments used and their efficacies, attitudes to the syndrome, and its impact on the lives of affected individuals and on the community as a whole. At 76%, the response rate was unusually high, so that the findings may be taken as truly representing the opinion of the BSR consultant membership as a whole. The findings are disturbing on several counts. First, they reveal a distinct polarization of perceptions of the prevalence, significance and impact of the syndrome. Secondly, there appears to be a total lack of consensus on what criteria to use in diagnosis. Thirdly (and perhaps most worrying of all), by their answers the respondents have demonstrated a distinct lack of familiarity with the recently published literature, in particular on the questions of the impact on patients’ lives in terms of the effects of chronic pain and the psychosocial sequelae [15, 17, 18].

The second Editorial in this issue is by S. Gurley-Green, entitled ‘Living with the hypermobility syndrome’, which was presented in 1999 at the XIVth EULAR Congress in Glasgow [19]. The author is a former Chairperson of the Hypermobility Syndrome Association, a patient self-group. In it she describes as dispassionately as possible, and on the basis of her own experiences and those of her members, what it is like to have the hypermobility syndrome—the everyday pains, the disruptions to family and professional life, the frustrations and the hopelessness borne by many sufferers. Not least is the barrier of disinterest and lack of knowledge confronting patients in their dealings with
their medical carers. Let us hope that the readers of this journal will heed the paper’s messages.

The third Editorial in this issue of the journal is a comprehensive review of the BJHS in children and adolescents by Murray and Woo [20]. The importance of hypermobility is gaining increasing recognition among paediatricians and paediatric rheumatologists, and there is increasing evidence that, for many adults with the BJHS, symptoms commence in their early years [15]. There can be few rheumatic diseases that scale the age spectrum in quite such a dramatic way. Taken together, these three papers provide an instructive and salutary read.

What is the way forward? If doctors would only take the trouble to look for joint hypermobility and other stigmata of the BJHS in the course of their routine examination of the locomotor system (as has been suggested in a major international rheumatology textbook published recently [21]), the condition would certainly be diagnosed more frequently. But, alas, recognition alone is not enough: it is, in fact, only a start. The BJHS is not an easy condition to treat. Physiotherapy forms the mainstay of treatment but has to be tailored to the needs of intrinsically vulnerable tissues, otherwise it may aggravate rather than relieve. Analgesics and non-steroidal anti-inflammatory drugs are generally ineffective, and surgery can be counter-productive unless the surgeon is aware of the underlying condition and the need to take additional precautions in handling and suturing tissues. The respondents to the questionnaire [16] acknowledge that conventional therapeutic approaches are often disappointing. Innovative approaches are clearly required. There have been some encouraging recent developments [22]. Stabilizing lax joints by appropriate exercises has been shown to increase stability, reduce pain and diminish hypermobility [23]. Improving proprioceptive acuity has been shown to be more effective in the treatment of anterior cruciate ligament deficiency than conventional quadriceps exercises [24]. In the presence of a chronic pain syndrome, a comprehensive pain management programme (incorporating cognitive behavioural therapy techniques) has effected improvements in quality of life, walking speed, pain intensity, distress, depression severity and confidence [25]. This is a fertile area for research in rheumatology. For a more detailed account of an evidence-based approach to the therapy of the BJHS, readers are referred to a recently published review [22].

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References
