Adequate methods for measuring the range of movement at joints are essential for the definition of criteria used in the study of clinical problems associated with joint hypermobility. Scoring systems for hypermobility that survey a large number of joints in simple fashion are ideal for epidemiological studies in large populations. Latterly, investigators have devised sophisticated mechanical devices for the precise quantification of movement at a single joint. The greater precision afforded may be ideal for serial assessments in the same patient but this greater precision is of limited use in epidemiological work if the joint fails to mirror the status of laxity at other joints in the body. Moreover, a joint may display acquired hyperlaxity in compensation for a reduced range of movement at adjacent joints, for example in the vertebral column.

A recent trend has therefore been to return to scoring systems in which a reasonably large number of joints are assessed in simple fashion. Nevertheless, there still remains uncertainty about the value of new assessments proposed. The original scoring system, first devised by Carter and Wilkinson and modified by Beighton et al., even now is re-emerging as the simple method of first choice, particularly for the screening of large populations.

The definition of ‘generalised joint hypermobility’ still remains arbitrary, and rationally should reflect both the number of joints involved and the extent to which they move. Hypermobility may represent one extreme of a Gaussian distribution of joint laxity throughout the population. Scoring systems devised for measuring joint hypermobility have proved less satisfactory in the measurement of joint hypomobility. Attention has recently been directed at the factors that contribute to the range of joint movement, not only the shape of bony articulating surfaces, the inherited collagen structure and the tone and bulk of the restraining muscle, but also recently to their neurological control, particularly in respect of proprioception, which may be impaired. It is likely that future scoring systems will concentrate even more on aetiological aspects as we attempt to separate groups of patients who may be at particular risk of osteoarthritis.
2.1 Simple Scoring Systems for Hypermobility

The first scoring system was devised by Carter and Wilkinson\(^1\) in conjunction with their work on congenital dislocation of the hip. They defined generalised joint laxity as being present when three of the following tests were positive, provided both upper and lower limbs were involved:

1. Passive apposition of the thumb to the flexor aspect of the forearm
2. Passive hyperextension of the fingers so that they lie parallel with the extensor aspect of the forearm
3. Ability to hyperextend the elbow more than 10°
4. Ability to hyperextend the knee more than 10°
5. An excess range of passive dorsiflexion of the ankle and eversion of the foot

A more complex assessment was suggested by Kirk et al.,\(^3\) but in practice this proved to be too time-consuming for routine use. The system of Carter and Wilkinson\(^1\) was revised by Beighton and Horan\(^4\) for the measurement of joint laxity in persons with the Ehlers–Danlos syndrome (EDS). Passive dorsiflexion of the little finger beyond 90°, with the forearm flat on the table, was substituted for passive hyperextension of the fingers, as the latter test had proved too severe; the range of ankle movement was replaced by measurement of forward flexion of the trunk. Patients were given a score between 0 and 5.

Grahame and Jenkins\(^5\) modified this system to include passive dorsiflexion of the ankle beyond 15°. This was partly an adaptation to the particular subjects under study, half of whom are ballet dancers. Subsequently, Beighton et al.\(^2\) amended the 1969 system for use in an epidemiological survey of bone and joint disorders in an indigenous rural South African community. They employed the same tests, but gave one point for each side of the body for the paired tests. The range of scoring was thus between 0 and 9, with high scores denoting greater joint laxity. The manoeuvres used in this scoring system are listed below and depicted in Fig. 2.1:

1. Passive dorsiflexion of the little fingers beyond 90° (one point for each hand) – two points
2. Passive apposition of the thumbs to the flexor aspects of the forearm (one point for each thumb) – two points
3. Hyperextension of the elbows beyond 10° (one point for each elbow) – two points
4. Hyperextension of the knee beyond 10° (one point for each knee) – two points
5. Forward flexion of the trunk with knees fully extended so that the palms of the hands rest flat on the floor – one point

This method has found favour for the following reasons:

1. Scoring systems using hyperextension of the middle rather than the little finger exclude too many persons.
2. Scoring systems using ankle movements, although perhaps appropriate for dancers, are unlikely to show much variation between individuals in a normal population.
3. Scoring systems that include trunk and hip movement (composite joint movement) are more likely to reflect generalised articular laxity.
In a study on 502 normal adult indigenous South Africans (168 males; 334 females), 94% of the males and 80% of the females achieved scores of 0, 1 or 2. This range of movement might be regarded as normal for adults in this population. The majority of clinicians require a minimum score in adults of between 4/9 and 6/9 before accepting the diagnosis. Laxity decreases with age and a lower level may be more appropriate to an elderly population. At any age, females are more mobile than males. In both sexes the degree of joint laxity diminishes rapidly throughout childhood and continues to fall more slowly in adult life.

An alternative scoring system was then developed. Based upon work by JP Contompasis, an American podiatrist, and described in detail by Poul and Fait, this scoring system is more complex than the modification by Beighton et al. of the Carter and Wilkinson scoring system. A multiple-point scoring system based on six manoeuvres, five of which replicate Beighton, its scores span from the normal to the hypermobile range with a maximum total of 72. Initial studies had suggested that it was highly correlated

**Fig. 2.1** Beighton et al. modification of the Carter and Wilkinson scoring system
with Beighton’s score ($r=0.92; p=0.0001$) in original work by the editors, and it had been claimed that it was particularly useful in the assessment of ligamentous laxity in children. The scoring system is described in detail elsewhere, but greater experience produced problems in measurement, particularly in the use of foot flexibility tests, the major feature on which it differed from the Beighton score. Since the Contompasis score takes significantly longer and, in spite of the theoretical greater sensitivity, conveys little more information, the score is now only occasionally used.

In a seminal paper Bulbena and colleagues compared Beighton’s modification with the original Carter and Wilkinson scoring system and the most popular scoring system used in France, to find the Beighton system as effective as any in measurement.

Recent studies have emphasised the difficulty in establishing joint hypermobility as a causative factor of symptoms in children whose joints in any case display an unusually large range of movement compared to adults. A further study on the high prevalence of joint laxity in West Africans has shown that joint hyperlaxity is substantially greater in a West African population than in almost any other population group in which it has been studied, yet is not associated with joint pain.

### 2.2 The Brighton Criteria for Hypermobility Syndrome

Although the measurement systems so far described suit the musculoskeletal system alone (and may be of particular value in measuring serial change), it became increasingly apparent that wherever abnormal collagen was ubiquitous throughout the body other organ systems would become involved. Moreover, certain individuals, particularly in different ethnic groups, would demonstrate striking hypermobility according to a scoring system but still remain asymptomatic.

It became clear that there was a need for a new scoring system that recognised all of these points. The Special Interest Group devoted to inheritable connective tissue disorders of the British Society for Rheumatology addressed this issue. As a result, criteria were proposed in Brighton in 1999, which were published the following year. These are shown in Table 2.1. Incorporating the Beighton score, still felt to be the best rapid assessment of musculoskeletal hypermobility, the presence of arthralgia for more than 3 months in four or more joints was allowed equal importance. A set of minor criteria was additionally proposed and, on the basis of pilot work, a number of major or minor criteria that needed to be fulfilled were decided.

The Brighton criteria have subsequently enjoyed extensive use. A study from Chile using the Brighton criteria suggested that true diagnosis in the majority of patients with joint hypermobility syndrome is often overlooked, a finding replicated in the UK. In the study from Chile it was noted that use of the Beighton criteria alone would have excluded 61% of patients who were identified by use of the Brighton criteria. It has been suggested that the criteria may yet benefit from further analysis and validation and even the ‘gold standards’ based on ‘a consensus of experts’ may be desirable, a point conceded by the original authors.

Nevertheless, there seems to be a consensus that the Brighton criteria represent a significant step forward in the quantification of hypermobility.
Table 2.1 The Brighton criteria for joint hypermobility syndrome

Major criteria
• A Beighton score of 4/9 or greater (either currently or historically)
• Arthralgia for longer than 3 months in 4 or more joints

Minor criteria
• A Beighton score of 1, 2 or 3/9 (0, 1, 2 or 3 if aged 50+)
• Arthralgia (>3 months) in one to three joints or back pain (>3 months), spondylolysis, spondyloysis/spondylolysthesia
• Dislocation/subluxation in more than one joint, or in one joint on more than one occasion
• Soft tissue rheumatism >3 lesions (e.g. epicondylitis, tenosynovitis, bursitis)
• Marfanoid habitus (tall, slim, span/height ratio >1.03, upper:lower segment ratio <0.89, arachnodactyly [positive Steinberg/wrist signs])
• Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring
• Eye signs: drooping eyelids or myopia or antimongoloid slant
• Varicose veins or hernia or uterine/rectal prolapse

The joint hypermobility syndrome is diagnosed in the presence of two major criteria, or one major and two minor criteria, or four minor criteria. Two minor criteria will suffice where there is an unequivocally affected first-degree relative.

Joint hypermobility syndrome is excluded by the presence of Marfan or Ehlers–Danlos syndromes (other than the EDS hypermobility type (formerly EDS III) as defined by the Ghent (1996) and the Villefranche (1998) criteria respectively). Criteria Major 1 and Minor 1 are mutually exclusive as are Major 2 and Minor 2.

Recently, Hakim and colleagues have also devised and validated a simple five-point questionnaire that can be used in general practice to alert suspicion to the presence of hypermobility.

2.3 General Principles of More Precise Measurement at Selected Joints

A complete evaluation of a new technique for the precise measurement of movement a single joint requires the following:

1. Statements on the inter- and intra-observer error of the method
2. Consideration of the influence of age on the range of observed movement
3. A study of sex-determined differences
4. Indication of whether specialist groups have been included in the survey population (for instance, physiotherapists are often used for such studies but are likely to form a highly selected group by virtue of their training)
5. Consideration of the influence of the dominant side

A hinge goniometer provides the simplest method for measuring the range of movement at a hinged joint. There are difficulties in positioning such an instrument accurately and a spirit-level device is often more appropriate. The Loebl hydrogoniometer was the first such devised described. The MIE clinical goniometer (Fig. 2.2) is an example of a similar device that is currently manufactured. Providing the patient is correctly positioned, the instrument can be used to record the arc of movement at any joint.
Fig. 2.2  (a) A clinical goniometer capable of measuring arcs of movement in any direction (manufactured by MIE Medical Research Ltd, 6 Wortley Moor Road, Leeds LS12 4JF, UK).  
(b) The goniometer in use
Recent modifications include instruments such as the Myrin goniometer, which resembles an aircraft gyrocompass; however, this instrument is expensive and lacks the sensitivity and precision of the simpler device.

When surface goniometry is correlated with movement measured radiologically, goniometry frequently proves to be inadequate. The skin, fat and soft tissues may distend and cause markers on the skin to move less or more than the underlying bones. Correlation coefficients between angular bony movement at the joint determined radiologically and movement of the overlying skin are rarely provided.

A comprehensive account of techniques for measuring joint movement throughout the body is described in a booklet published by the American Academy of Orthopaedic Surgeons. Diagrams of suitable methods for using goniometers to determine the arcs of movement at all joints in the body are given, together with ‘normal’ values, but the coefficients of variation for these measurements, both between serial assessments and in the same observer and between different observers, are not provided. A volume of Clinics in Rheumatic Disease, edited by V. Wright, devotes one chapter to the measurement of movement at each major joint in the body. Available methods are compared and the most suitable selected. This is used to define the normal range of movement at each joint in males and females, usually in 10-year cohorts. Estimations of inter-observer and intra-observer variation are provided.

Some additional devices have been championed for more sophisticated measurement of the range of movement. For the shoulder, an electromagnetic movement sensor has been devised and validated. At the hip, a plurimeter has been devised and validated, providing a relatively inexpensive measure for the range of movement at this joint and one that might be of particular use in primary care.

Regular training undoubtedly affects the range of movement, due either to alteration in muscle control or to stretching of the joint capsule. Atha and Wheatley showed the effect of training to be a source of greater variation in passive goniometry at larger joints; investigators therefore need to specify whether an individual is warmed up or participating in a physical training programme designed to increase the range of movement. Such changes have been further quantified in studies on athletes, which drew attention to the way in which the range of joint movement could be altered by ‘warm up’ and this varied according to the experience and skill of the athlete. Programmes were then introduced to stabilise unstable joints by the use of regular exercises. These were also shown to be effective.

2.4 Back and Spinal Mobility

The spine is a complex set of joints. Restrictions of movement at one site, either inherited or acquired by disease, may result in compensatory hyperlaxity at adjacent vertebrae leaving the overall range of movement, as measured by surface techniques, unaltered. Troup et al. used photography to study movement of the lumbar spine and hips in a sagittal plane, and a full review appears elsewhere. One-dimensional
measures involve skin distraction techniques such as Schober’s method, as modified by Macrae and Wright. Plumbline techniques have also been described and lumbar sagittal mobility may be measured by flexicurves. The latter method has an intra-observer and inter-observer variation of 9% and 15%, respectively. A hydrogoniometer is probably the most satisfactory instrument, though more complex spondylometers are available. Three-dimensional techniques include stereoradiography, vector stereography and three-dimensional optical systems. All have been reviewed recently in comprehensive fashion and reference values for normal regional lumbar sagittal mobility have been published.

It is of interest that, although most studies at peripheral joints have clearly shown that hypermobility is more prevalent in females than in males, this does not appear to be so for the lumbar spine. Thus, Loebl and Troup have both shown that spinal movement is approximately equal in both sexes, while a seminal paper by Macrae and Wright showed the male spinal mobility to be greater than that in females. The reason for this is not clear.

In a study correlating low back symptoms with lumbar sagittal mobility (Burton and Group, unpublished results), flexicurves were used in 958 individuals aged from 10 to 84 years. Both hypermobility and hypomobility of the lumbar spine were identified as risk factors for low back trouble, though, as ascertained by questionnaire, current sufferers were more likely to be relatively hypomobile.

The Polhemus Navigation Sciences 3Space Isotrak system has been used to measure the range of movement in the lumbar spine. This proved valuable in detecting minor changes in spinal movement throughout the 24-h period that were attributed to the natural circadian variation. Although expensive, sophisticated, and only available in small numbers of centres, this system may provide insight into diurnal variation of symptoms arising from the spine that are a feature of subjects with hyperlaxity of the spine and also intrude on clinical practice.

2.5 Rotation in the Limbs

Haskard and Silman have devised fixed-torque screwdrivers that measure forearm and lower limb rotation in epidemiological studies. Inter-observer variation has been validated and is low. One such device measures forearm rotation and another leg rotation. Fairbank et al. devised a goniometric assessment involving six joints. Special jigs were constructed for the measurement of hip rotation and tibial rotation.

2.6 Movement at the Metacarpophalangeal Joint

The metacarpophalangeal (MCP) joint is easily accessible and also forms a component part of conventional scoring systems. Harris and Joseph developed a radiological technique for measuring the range of extension at the MCP joint and
Loebl described a mechanism for abducting the fingers to investigate movement at the MCP joints. Grahame and Jenkins described a simple spring device that applied a predetermined force (2 lb (0.91 kg)) to the fifth MCP joint. Applied to the relaxed patient, this force mimicked the passive range of movement measured in the clinical scoring system. It had good reproducibility but only quantified movement to the nearest 30°.

The Leeds finger hyperextensometer (Fig. 2.3) can be used for either hand. It allows greater precision in quantification of the range of movement and has good inter-observer and intra-observer reliability. The hyperextensometer applies a torque of 2.6 kg cm\(^{-1}\). The device can be used in epidemiological surveys as it is portable, light and inexpensively constructed. It is of particular value in serial assessments of joint laxity in the same patient and has been used to provide the first demonstration of enhanced peripheral joint laxity prior to parturition in pregnant females. An increased level of serum relaxin has been noted in pregnant women who have pelvic ligament pain. Since relaxin levels are known to be high at the end of pregnancy when peripheral joint laxity was demonstrated, it remains a possibility that this hormone may be directly related to the development of rheumatological symptoms arising from hyperlax ligaments. A finger arthrograph quantifies the resistance encountered when the index finger is moved in sinusoidal fashion at a constant speed through a pre-selected angle of displacement and is of value in measuring stiffness.

Most recently, an electronic gravity goniometer has been developed for determining the passive range of movement of the four MCP joints by the use of preset fixed torques. This may represent an improvement on the hyperextensometer. The arthrograph has also been revisited and a microprocessor-controlled arthrograph devised. In addition to the greater accuracy provided, a novel feature is the movement of the MCP joint in a lateral rather than a flexion/extension plane.

**2.7 Joint Proprioception**

With increasing realisation that this is relevant to hypermobility, efforts have been directed to its accurate quantification. Ferrell and colleagues in Glasgow have designed a sophisticated rig to quantify proprioception at the knee joint and have shown in a sophisticated series of studies that this is impaired in hypermobility syndrome, that enhanced proprioception ameliorates symptoms and that musculoskeletal reflex function is also altered in hypermobility. The rig used, however, is not portable. Parallel work has shown that proprioception is impaired both in inflammatory and degenerative arthritis and intriguingly, improved after joint replacement, probably because of surgical tightening of the capsule.

A portable proprioceptometer has been devised for use in the hand and is currently being used in studies of hypermobile individuals, as well as on musicians and typists. Proprioception has also recently been demonstrated to be abnormal in hypermobile children.
Fig. 2.3  A finger hyperextensometer for the quantification of joint laxity. The finger of the subject is hyperextended at the metacarpophalangeal joint by the application of a pre-set fixed torque. The resultant angle of the hyperextension is read off on the dial.
2.8 Correlations Between Scoring Systems Used in Assessing Joint Laxity

A comparison has been made between the Carter and Wilkinson\(^1\) scoring system, as modified by Beighton et al.,\(^2\) the Leeds finger hyperextensometer and a ‘global index’ constructed by using goniometry to assess the range of movement at almost all the joints in the body. This comparison follows the guidelines suggested by the American Academy of Orthopaedic Surgeons\(^22\) and sums the measured arcs of movement.\(^55\) Individuals were selected from different sporting groups thought to reflect more generalised hyperlaxity than that seen in the normal population. Beighton et al.’s modification of the Carter and Wilkinson system correlated well with the global index, endorsing the value of a simple scoring system that could be applied to large populations.\(^56\) The hyperextensometer appeared to convey more applied information in an accurate fashion, emphasising that the range of movement at a single joint does not necessarily correlate with overall joint laxity.

Silman et al.\(^57\) have confirmed the Gaussian distribution in joint mobility that can be measured with fixed-torque measuring devices. Subsequently, a family study showed that, although the fixed-torque devices reliably reflected anticipated epidemiological findings in Asian families, the index finger hyperextensometer produced different results. They concluded that both genetic and constitutional factors affect mobility independently at certain sites.\(^58\) Fairbank et al.,\(^41\) using goniometry at 6 different joints in a group of 446 normal adolescents, concluded that there was a weak but significant correlation between the ranges of movement at each of the different joints measured, except for elbow hyperextension.

2.9 Variation of Joint Laxity Within Populations

A major development in the epidemiology of hypermobility has been the demonstration that the range of movement at a given joint is observed as a Gaussian distribution throughout the population.\(^59\) It is no longer acceptable to consider hypermobility as an ‘all or nothing’ phenomenon and it becomes logical to define hypermobile individuals as those who comprise a certain extreme proportion of the normal population. The cut-off point for hypermobility remains arbitrary, but it is our impression that the majority of musculoskeletal complaints attributable to hypermobility occur in the most supple 5\% or 10\% of the population.

The range of normal joint movements decreases rapidly throughout childhood and more slowly in adulthood. This observation has been confirmed in children in Edinburgh,\(^60\) in a South African population\(^2\) and in London children.\(^61\) Joint laxity continues to diminish throughout adult life.\(^3\) The joints of females were found by several authors to be more lax than those of age-matched males,\(^2,42,60\) though this finding has been disputed by Silverman et al.\(^61\) and is not always seen in the spine, as previously described. Laxity may be localised to a small number of joints or a
single joint. The concept of pauci-articular hypermobility has been reviewed in detail by Larsson et al.\textsuperscript{62}

Although few comparative studies have been carried out, there is a strong clinical impression of a racial variation in joint mobility. For instance, Indians show more hyperextension of the thumb than Africans, who in turn have greater hyperextension than Europeans.\textsuperscript{42} A similar result has been obtained by comparing the finger joints of different ethnic groups in Southern Africa.\textsuperscript{63} The question of inter-ethnic variation could be resolved by large-scale comparative studies employing the techniques discussed in this chapter.

A study on joint mobility among university students in Iraq has shown a relatively high prevalence of individuals scoring 4/9 on Beighton et al.’s\textsuperscript{2} modification of the Carter and Wilkinson\textsuperscript{1} scale, the right (usually dominant) side being significantly less mobile than the left side, whatever the hypermobility score.\textsuperscript{64} Comparable data from an age-matched group of English university students has shown a lower prevalence of hypermobility using the same scoring system.\textsuperscript{65} When English Caucasian subjects were compared with Asian Indians and a group of patients suffering from a variety of inherited disorders, including Ehlers–Danlos Syndrome (EDS) and osteogenesis imperfecta, Asian Indians were significantly more mobile than English Caucasians. Males and females with EDS were hypermobile, but only females with osteogenesis imperfecta (and female relatives of those with severe or lethal osteogenesis imperfecta) showed excess joint laxity.\textsuperscript{66}

Studies with the hyperextensometer in Europe have defined normal curves for laxity at the MCP joint in relation to age and sex and then correlated hyperlaxity with various orthopaedic diseases.\textsuperscript{67} The frequency of occurrence of generalised ligamentous laxity has been defined in a Czechoslovakian population (Poul J and Fait M 1989, personal communication). In 890 healthy children, the Contompasis criteria\textsuperscript{6} defined the variability of generalised ligamentous laxity in relation to age and sex. It was found that pathology was most likely to develop in subjects who exhibited two standard deviations from the mean. This study failed to show a discrete clinical abnormality of connective tissue – it was felt that rheumatic or orthopaedic symptoms could occur in any individuals, providing their overall hyperlaxity exceeded a certain degree. This favours mechanical rather than biochemical aetiology for symptoms arising from joint hypermobility. A study from Yugoslavia (I. Jajic 1988, personal communication), in which 632 schoolchildren were surveyed, confirmed the greater prevalence of joint hypermobility, as measured by the Beighton et al.\textsuperscript{2} scoring system, in schoolgirls compared to age-matched schoolboys.

\subsection*{2.10 Clinical Applications of Scoring Systems}

Both the Beighton\textsuperscript{2} and Contompasis scoring systems have been used to quantify laxity in a study of 58 consecutive patients presenting to a rheumatology clinic with putative benign joint hypermobility syndrome (BJHS). There appeared to be no great prevalence of cardiac, bone, skin or eye abnormalities in this group, helping to
differentiate it from more serious hereditary disorders of connective tissue. An epidemiological study in rheumatology clinics has evaluated 130 consecutive new patients with joint hypermobility. Musculoskeletal problems were the main reason for referral, and there was a statistically significant association between diffuse joint hypermobility and osteoarthritis, supporting the hypothesis that joint hypermobility predisposes to musculoskeletal disorders, particularly osteoarthritis.

More contentious is whether joint hypermobility is associated with fibromyalgia or even a cause of that condition. Criteria for fibromyalgia have recently been proposed by the American College of Rheumatology. This has allowed the correlation of scoring systems for mobility with diagnostic criteria for fibromyalgia in schoolchildren, which showed an apparent strong association between the two conditions, certainly in schoolchildren. In a group of adults in Oman, there was less correlation between widespread musculoskeletal symptoms in any age group and joint mobility scores though specific diagnostic criteria for fibromyalgia were not sought. Other studies have suggested there may be an association between hypermobility and fibromyalgic type symptoms in adults, and if further studies confirm this, exercise programmes to reduce hyperextension of joint capsules and other soft tissues may become a recognised part of the treatment of fibromyalgia.

The new international Nosology of heritable disorders of connective tissue from Beighton and colleagues has replaced the earlier classification, first proposed in 1986, though not published until later. This has defined the benign joint hypermobility syndrome as an entity quite discreet from rarer and more serious inherited abnormalities of connective tissue such as the EDS, some variants of which cause much greater involvement of body structures other than joints, particularly the blood vessels.

### 2.11 Joint Hypolaxity

Restricted movement of the joint has been recognised clinically in association with certain diseases, particularly diabetes mellitus. Reduced movement has been reported in the hand, shoulder, wrist, elbow and ankle. The scoring systems currently used for hyperlaxity have evolved specifically for this need and prove not to be particularly suited to the detection and measurement of joint hypolaxity.

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2012, XVI, 204 p., Hardcover
ISBN: 978-1-84882-084-5