Table 2
Mobility according to ethnic origin: mean proportions with definite or marked hypermobility in each joint or group on the two sides in females aged 15–54 years

Joint	*Caucasian (n=81)*	*Negro (n=45)*
Elbows	18%	6%
PIPs II–IV	90%	74%
PIP V	46%	36%
DIPs II–V	79%	88%
IPs of thumb	52%	66%

only joints for which χ^2 indicated a significant difference were the second to fourth PIPs (e.g. $\chi^2=5{\cdot}56$; $0{\cdot}025>P>0{\cdot}01$ for third right PIP). Thus the clinical impression of laxity in Negroes appears to be incorrect.

This report began by drawing analogies with hypertension and hyperuricæmia. However, both these situations are relatively simple because at any one time only a single measurement is concerned. Hypermobility has been approached in a similar manner, considering the range of angular motion only in isolated joints. Thus far the analogies are very close. But any quantitative approach to disorders of joints is bedevilled by problems of clustering in space. The hypermobility concept relates not to individual joints in isolation, but to a composite of functional assessments of quite a number of joints. The first step towards a synthesis of the pattern one is trying to recognize may be accomplished by considering the interrelationship of angular motion in the PIPs and DIPs (Fig 2). At once a fascinating reciprocal pattern begins to emerge. In general, the PIP with the lowest frequency of hyperextension is associated with the DIP with the greatest frequency of hyperextension, and *vice versa*. However, this relationship is modified in detail both by the influence of the dominant limb, and by different performance in the various fingers, the middle finger usually being the most mobile. Theoretically it should be possible for us to take one further step, to relate the elbow to each and all of the 18 IP joints. The number of possible combinations in such an exercise is considerable, though, and our samples were not really large enough to take account of individual variations against such a background.

It only remains, therefore, to review briefly the implications of these findings when extended to the broader view of hypermobility. Take the five manœuvres by which Carter & Wilkinson (1964) defined hypermobility, requiring more than three of the manœuvres to be carried out. When they studied the entire diversity of human beings for these characteristics, those with what they categorized as persistent generalized joint laxity were in fact only the extreme of a distribution.

Does all this emphasis on the extreme of a distribution matter? I would submit that it does, because it alters one's appreciation of the entity one is considering and, more particularly, it has

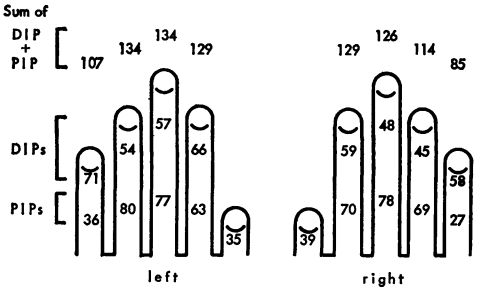

Fig 2 *Reciprocal mobility in proximal and distal interphalangeal joints. Proportions (%) with definite or marked hyperextensions (+ or + +). n=347 females*

important consequences on the manner in which the data can be analysed. Treating the condition as a graded rather than as a threshold attribute blunts the apparent precision of genetic analysis, and may also lead to very different conclusions (Wood 1971). Similarly, the testing for associations between hypermobility and other features is not so simple. Whether the extreme of a distribution can nevertheless constitute a significant cluster has not been established; more work is needed to resolve this fundamental conceptual problem.

Most of this report has related to angular rotation in a plane of normal motion. A different situation occurs with abnormal mobility such as lateral motion of the knee. This state of affairs could obviously be of great importance in contributing to damage in a weight-bearing joint, and perhaps this may be an all-or-none characteristic.

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Joint Hypermobility – Clinical Aspects

Hippocrates, in the fourth century BC, made the first known reference to hypermobility, when he described the Scythians as being 'so loose-limbed that they were unable to draw a bow-string or hurl a javelin'. It was only at the end of the 19th century, however, that hypermobility of joints was recognized as being of any clinical significance. I refer to the description by Tschernogobow in Moscow in 1892 of what we now refer to as the Ehlers-Danlos syndrome. He rightly attributed the association of hyperextensibility of

the skin and hypermobility and luxation of the joints to 'a fundamental and generalized inadequacy of connective tissue'. Four years later, Marfan (1896) described the syndrome which now bears his name.

Table 1 lists the group of hereditary diseases with which hypermobility is now known to be associated. Many of these are familiar conditions, and I will comment briefly only on the less familiar ones.

Achard's syndrome (arachnodactyly with mandibulofacial dysostosis) was distinguished from Marfan's syndrome by Parish (1960) at a meeting of this Section. The marfanoid hypermobility syndrome (Walker *et al.* 1969) is a combination of the skeletal features of Marfan's syndrome with gross hypermobility of joints and marked hyperextensibility of the skin. The other features of Marfan's syndrome, such as involvement of the aorta or dislocation of the lens, are lacking, as are the tendency to skin-splitting and the molluscoid pseudotumours seen in the Ehlers-Danlos syndrome.

Homocystinuria (Schimke *et al.* 1965) is an inborn error of metabolism caused by a deficiency of the enzyme cystothionine synthetase, and is characterized by ectopia lentis, thrombosis in medium-sized blood vessels, osteoporosis and, frequently, mental retardation. Hyperlysinæmia (Ghadimi *et al.* 1965) combines joint hypermobility with gross physical and mental retardation.

The term 'hypermobility syndrome' was coined by Kirk *et al.* in 1967 to describe generalized joint laxity causing symptoms in an otherwise normal subject. The first report indicating a familial tendency in this condition was that of Finkelstein (1916) who called the condition 'joint hypotonia'. Where it occurred in infants he distinguished it from amyotonia congenita, with which such cases had probably previously been confused. Key, in St Louis in 1927, confirmed the familial tendency; and referring to the same condition, Hass & Hass, in 1958, used the term 'arthrochalasis multiplex congenita'.

Table 1
Etiology of joint hypermobility

(1) Marfan's syndrome	(5) Marfanoid hypermobility syndrome
(2) Achard syndrome (Parish 1960)	(6) Homocystinuria
(3) Osteogenesis imperfecta	(7) Hyperlysinæmia
(4) Ehlers-Danlos syndrome	(8) The hypermobility syndrome

Table 2
Criteria for hypermobility (Carter & Wilkinson 1964)

(1) Passive apposition of thumb to flexor aspect of forearm
(2) Passive hyperextension of fingers so that they lie parallel to extensor aspect of the forearm
(3) Ability to hyperextend the elbow beyond 10 degrees
(4) Ability to hyperextend the knee beyond 10 degrees
(5) An excessive range of passive dorsiflexion of the ankle and eversion of the foot

So far I have considered only hereditary hypermobility. It has been suggested both by Callegarini (1957) and Levine (1958) that hypermobility is more frequent in patients with rheumatic fever; Kirk *et al.* (1967) also found this to be the case. More recently, Bluestone and his co-workers (1971) at the Hammersmith Hospital have observed in 42 acromegalic patients an increased overall spine and hip mobility compared with controls.

The question arises, in any discussion of this nature, of what constitutes the normal range of joint movement. Clearly there is a wide physiological variation. In children, for instance, there is a far greater range of spinal and peripheral joint movement than in adults, though genu recurvatum and hyperextensibility of the elbow is outside the norm even in children.

There is considerable confusion about racial variations. Dr Wood (p 690) found no difference between subjects of Caucasian and Negro origin in Buffalo, and the incidence of congenital genu recurvatum in various groups (Wong 1966, Charif & Reichelderfer 1965) indicates no statistically significant differences. On the other hand Schweitzer (1970) reported differences in the extensibility of the metacarpophalangeal joints of the fingers and the interphalangeal joint of the thumb between some but not all of the five ethnic groups tested in South Africa. Those of Indian extraction showed greatest extensibility.

Harris & Joseph (1949) also found significantly greater hyperextensibility of the interphalangeal joint of the thumb in people of African and Indian origin than in Europeans. Gross hyperextensibility of this joint was more common in Indians than in the other two groups. No such differences were seen in the first metacarpophalangeal joint. Interestingly enough the same workers noted significant differences between men and women in the same ethnic group.

The major problem in investigating this subject is that of establishing suitable criteria by which to judge the existence and degree of hypermobility. One method, suggested by Carter & Wilkinson (1964), scores the ability of the individual to perform certain movements (Table 2). Using their criteria, they found that 7% of 300 children, aged 6 to 11, showed a score of 4 or more.

In Beighton & Horan's improved criteria (1969) the second and last tests have been changed: passive dorsiflexion of the little finger to 90 degrees, and forward flexion of the trunk so that the hands rest flat on the floor, replace passive dorsiflexion of the fingers and wrist and excessive dorsiflexion of the ankle respectively.

The Consequences of Hypermobility

(1) *Trauma:* Recently Nicholas (1970) has studied 139 members of the New York Jets football team

and found that the 39 loose-jointed members had eight times the incidence of ruptured knee ligaments requiring surgery, when compared with the remaining 100 players who were tight-jointed.

(2) *Recurrent dislocations:* As long ago as 1882, Macleod, in the *Glasgow Medical Journal*, recorded the case of Charles Warren, a professional contortionist whose joints were so lax that he could dislocate and reduce most of his joints at will. His father, and two of his own children, possessed the same facility. Joint dislocation is a frequent occurrence in Marfan's syndrome, and occurred in 26 of Beighton's 100 cases of Ehlers-Danlos syndrome (Beighton & Horan 1969). Carter & Sweetnam (1958) noted an association between recurrent dislocation of the patella and generalized joint laxity. Subsequently Carter & Wilkinson (1964) showed a striking association between generalized hypermobility and the occurrence of congenital dislocation of the hip.

(3) *Effusions:* Sutro (1947), a medical officer in the American forces, recorded 5 cases of recurrent effusions (3 in the knee, 2 in the ankle) occurring without obvious trauma, in healthy soldiers who showed generalized joint hypermobility. Persistent effusions occurred in 20% of Beighton's Ehlers-Danlos patients.

We have recently studied one hypermobile patient. The joint aspirate was a clear, viscous fluid, with a low cell count, suggesting a low-grade synovitis. She showed a hitherto unrecorded complication of this syndrome, namely a large Baker's cyst.

(4) *Premature osteo-arthrosis:* There have been several reports suggesting an association between hypermobility and premature osteo-arthrosis. We have recently seen two patients with polyarticular osteo-arthrosis and generalized hypermobility. To prove that this is an etiological relationship would require a long-term prospective study, which is no mean undertaking.

So far I have dealt with the adverse effects of hypermobility. What are its possible benefits? To what extent does hypermobility improve physical performance? In the field of sport, it seems no coincidence that as many as 28% of the New York Jets footballers were hypermobile. Nicolo Paganini, the famous violinist, undoubtedly owed part of his outstanding technical ability to the fact that his joints were hypermobile. In a physiological notice to the Academy of Paris in 1831, Dr Bennati wrote of Paganini: 'The hand is no larger than normal, but because of the flexibility of all its joints, its reach could be doubled'. Despite his slender appearance, the cast of his right hand shows that he did not have Marfan's syndrome.

Ballet dancers show striking mobility of the hips and spine. Is this solely the result of training, or does an inherent, generalized hypermobility confer a positive advantage which results in individuals showing this trait being selected for ballet training? In an attempt to resolve this question, assisted by Miss Jean Jenkins, I recently studied joint mobility in 53 senior students from the Royal Ballet School in London. The results were compared with those obtained by examining an identical number of similarly-aged student nurses at Guy's Hospital. This study will be reported in detail elsewhere (Grahame & Jenkins 1971); it can only be summarized here.

Six observations were made on each subject, according to Beighton's five criteria (Beighton & Horan 1969), plus Carter's criterion 5 (Carter & Wilkinson 1964). The results showed a significantly higher incidence of hypermobility in the dancers in respect of all the joints measured, with the exception of dorsiflexion of the little finger. The differences were most marked in the lower limbs. No one would dispute that in the course of their training ballet dancers strive to enhance the range of movement of their spines and lower limbs, and this could of course explain the differences seen in these movements. I am told on good authority, however, that hyperextension of the elbows and knees, and apposition of the thumb to the forearm, are not movements that ballet dancers perform. This suggests that a more generalized inherent joint hypermobility exists, though why the metacarpal phalangeal joints should not take part in this process is difficult to explain, and is being considered further.

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