Hernia in Congenital Dislocation of the Hip
Enquiry was also made regarding hernia both in the index patients and in their families. The figure for inguinal hernia in males to the age of 15 years is usually quoted as about 9 per 1,000 or nearly 1%. In the congenital dislocation of the hip survey the figure for males was about 7% and it must be remembered that many children were not yet 15 years of age. It was also interesting to note that inguinal herniæ occurred in about 5% of their fathers and brothers.

Thus, one etiological factor in congenital dislocation of the hip is likely to be familial hypermobility of joints and this is probably a dominant trait. It is particularly a feature of neonatal dislocation but is also present in many cases of dislocation presenting at a later age.

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Is Hypermobility a Discrete Entity?
Hypermobility of joints is characteristic of the Ehlers-Danlos syndrome, and is the hallmark of the hypermobility syndrome described by Kirk *et al.* (1967). The implication of these statements is that hypermobility is a discrete entity. In other words, that people either exhibit hypermobility of joints or they do not. The other side of this coin is that limitation of motion is also a discrete phenomenon. I wish to question the validity of these concepts.

Conceptual appreciation in medicine undergoes a sequential development as far as biological characteristics are concerned. Cochrane has represented this process graphically (Wood 1971, Fig 1). When a characteristic is first linked with a disease, it is often concluded that the diseased and non-diseased states are qualitatively different from each other with regard to the characteristic. The everyday necessity of making a binary decision, treatment required or not required, predisposes to such a simplistic view.

With the passage of time the stark dichotomy gets eroded by gradual appreciation that there are distributions of the characteristic, both in the diseased and in the nondiseased. Finally, it is conceded that the characteristic is distributed continuously, so great is the overlap between the two states. However, there persists a reluctance to discard the notion that underlying the skewed distribution are the two discriminated occurrences.

This whole pattern of development reflects stages in the progressive acquisition of knowledge, as experience increases. To bring this pattern to life you have only to remember the celebrated controversy over the nature of hypertension, with which the names of Platt (1959) and Pickering (1963) are associated – is hypertension a discrete entity or only the extreme of a distribution? A similar conflict arose about the significance of hyperuricæmia (Neel 1968). The epidemiologist, by examining a representative sample, is protected from the intermediate biases.

My interest in the distribution of mobility in a joint was sparked off by observations on the Ehlers-Danlos syndrome. Two population samples of females in Buffalo, USA, were examined with my colleagues Floyd Green and David Sackett (Green *et al.* 1965), and the data in this report are derived from this work. However, I have comparable data from a British sample of both sexes in the Rhondda Fach, and in general the findings were similar in this group.

We restricted our attention to joints that move in only one plane, and the bulk of our data relates to the elbow and the interphalangeal joints of the upper limb. We made preliminary studies with a goniometer, but our alignment of the arms of this instrument in relation to the axis of the limb showed unacceptable variability. Inter-observer variation was much less with judgments made by standardized procedures and recorded on a seven-point ordinal scale (Fig 1).

The elbow provides a good example (Table 1). Although the neutral position was the limit of passive extension in more than half the individuals, the pattern of this distribution is nevertheless within the family described as normal or Gaussian. Two important conclusions stem from these observations. First, the mobility of a joint is a continuously distributed variable. In other words, neither hypermobility nor limitation of motion are discrete phenomena. Furthermore, in

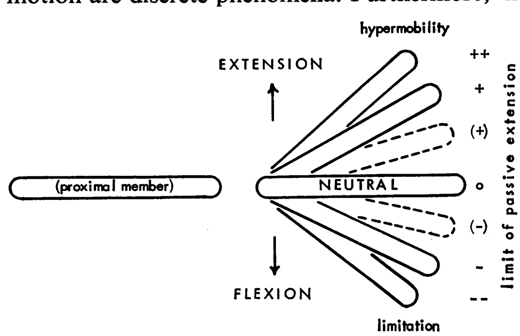

Fig 1 *Mobility of a joint which moves in only one plane, recorded on a seven-point ordinal scale. The neutral position was categorized as zero and departures from this as doubtful (symbol in parentheses), definite (single symbol) and marked (double symbol), hypermobility being indicated by plus and limitation by minus*

Table 1

Angular motion of elbow, proximal interphalangeal joints (PIPs) and distal interphalangeal joints (DIPs) in 347 females

	Limit of passive extension (percentage of observations ●)						
Joint	++	+	(+)	0	(−)	−	− −
Right elbow	0	12	14	55	10	8	0·3
Left elbow	0	16	17	55	7	5	0·3
PIPs II–IV ■	6	67	9	16	1	1	0·1
PIP V ■	1	31	18	40	5	4	1
Third left PIP (age-specific):							
15–24 years	14	77	0	9	0	0	0
25–34 years	9	68	17	6	0	0	0
35–44 years	5	71	6	11	7	0	0
45–54 years	4	64	9	16	4	3	0
55–64 years	4	62	11	13	3	7	0
65 and over	0	49	14	26	4	7	0
DIPs II–V ■	6	51	14	28	1	0·2	0
IPs of thumb ■	5	32	14	49	0	0	0

● Percentages rounded to whole numbers, unless <0·5
■ Mean of observations in each joint or group on the two sides

the elbow at least, not much more than half the individuals conform to the so-called norm, with extension as far as the neutral position.

Secondly, there was 30% more hyperextension in the left elbow than in the right, and about 40% less limitation to compensate for this. These observations held true whether one considered only definite changes, or if one included the doubtful limits as well. Moreover, this was not a function of the biases of a particular observer, because 88% of the individuals in these samples were examined independently by a second observer, and the same broad pattern emerged. As only 7% of the sample admitted to being sinistrals, and a further 4% to being ambidextrous or uncertain which was their dominant hand, one cannot escape the conclusion that the range of motion in the elbow tends to be greater in the non-dominant limb.

The distribution of angular motion in the proximal interphalangeal (PIP) joints differed from that seen in the elbow (Table 1). In most of the PIP joints 66–75% of the individuals showed hyperextension, and in about 6% of them this was marked in extent. The fifth PIP showed a marked contrast; only one-third of the individuals exhibited hypermobility. One is aware of the relative frequency of congenital flexion deformities of the fifth PIP, but it is interesting to note that this occurs in a joint that is generally less mobile than its fellows. As was observed in the elbow, the PIP joints of the third, fourth and fifth digits on the left showed hyperextension more frequently, and limitation less frequently, than did those on the right. However, the reverse was true of the index finger, where mobility appeared to be greater on the dominant side.

In contrast to the elbow, where age appeared to have relatively little effect on the range of motion, mobility in all the PIPs showed an age gradient (Table 1). The proportion that showed hyperextension diminished with increasing age, and *pari passu* the proportions mobile only to the neutral position and with limitation both increased as age increased. In fact limitation of the PIPs was observed only in those over 35 years old.

The distribution of angular motion in the distal interphalangeal (DIP) joints resembled that in the PIPs (Table 1), although hyperextension was not quite so frequent. In consequence the neutral position was the limit of motion in almost one-third, whereas this occurred in only one-sixth of the second to fourth PIPs. Restriction of movement was less frequent in the DIPs. In a class on its own was the interphalangeal (IP) joint of the thumb. The marked bimodality of the distribution is very evident (Table 1). Most striking, however, was the lack of even a suspicion of limitation in the thumb; the proportion reaching the neutral position was almost 50%.

The DIPs on the left side showed greater mobility than did those on the right, although the difference was least marked in the index finger. In the thumb the position was reversed, with the IP joint on the right exhibiting hyperextension slightly more frequently. Thus the dominant thumb and index fingers are the notable exceptions to the general pattern of greater mobility on the non-dominant side. All these joints, though, showed the same inverse changes with age that were noted in the PIPs, hyperextension occurring less frequently with increasing age.

Two important local variations were studied. Heberden's nodes are an obvious factor, but the other, variations in the axis of the distal phalanx in a lateral plane, is perhaps less well-recognized – how the distal phalanges of the ulnar digits often tend to deviate radially, and those of the radial digits to deviate towards the ulnar side. However, both these characteristics occurred more frequently than did limitation of the DIPs, even of questionable degree. Thus their relationship to angular motion is not clear-cut, apart from noting that both appear to be associated with lesser ranges of movement.

More relevant to the hypermobility concept is the question of race. The second sample examined included 81 Caucasians and 45 Negroes. Although the Ehlers-Danlos syndrome does not appear to have been described in the Negro, McKusick (1960) refers to the clinical impression that Negroes are more loose-jointed than whites. In the DIPs and the interphalangeal joints of the thumb hyperextension was more frequent in the Negro, but in the PIPs and the elbows hyperextension was less frequent, and limitation of movement more common, than was observed in the Caucasians (Table 2). No formal hypotheses concerning the effects of ethnicity were formulated in advance of this work, so that conventional tests of significance are scarcely appropriate. Furthermore, the samples studied were not as large as one might wish for. It is interesting, though, that the

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

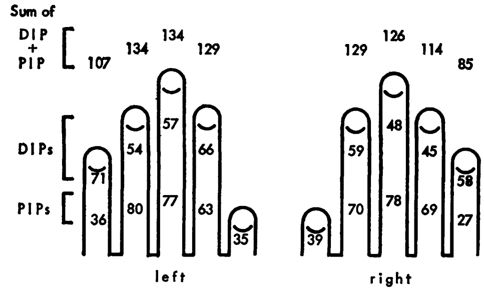

Fig 2 *Reciprocal mobility in proximal and distal interphalangeal joints. Proportions (%) with definite or marked hyperextensions (+ or + +). n=347 females*

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