CONGENITAL HIP DISEASE:
OCCURRENCE AND ETIOLOGY

by

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In this paper I will summarize recent studies on the occurrence of congenital disease of the hip joint, one of the commonest skeletal disorders. I will also review current theories on the etiology of congenital dislocation of the hip (CDH), emphasizing those which discuss its relationship to persistent generalized joint laxity.

Congenital dislocation of the hip was described by Hippocrates and recognized by him as a congenital disorder, but the exact nature of the abnormality is still controversial. According to Rabin (1965), radiographs of newborns show too little ossification for accurate interpretation. In older children the defect appears as either frank dislocation or as a deformity of the acetabulum with underdevelopment of the femur but without complete dislocation. The latter condition is called hip dysplasia or subluxation. Both dislocation and dysplasia are considered to be manifestations of a single disease, but their relationship and the primacy of either condition is not yet clear. In various papers published before 1960, it was stated that hip dysplasia was the primary abnormality. According to this theory an increased degree of slope to the acetabular roof provides inadequate support for the femur. Dislocation results from the subsequent stress of motion and weight-bearing. In more recent studies, it has been proposed that the basic abnormality is hip dislocation which is present at birth. Many cases identified at birth become stable and normal within a few weeks, but a small percentage retain a
complete dislocation or show radiographic evidence of dysplasia. As Rabin notes (1965:P.2) under this concept "hip dysplasia, characterized by a shallow acetabulum, increased slope of the acetabular roof, and late development of the proximal femoral epiphysis, is secondary to and represents a recovery stage of hip dislocation". The recovery may proceed to normal or the patient may remain abnormal.

Hip abnormalities have been found in archaeological specimens from many parts of the world. Brothwell (1973:P.155) cites reports of congenital luxation (dislocation) of the hip in a Neolithic specimen from France, an ancient Peruvian, pre-Columbians in America, an early Iron Age skeleton in Greece, five specimens from one location in Nubia, and a Saxon skeleton from Guildown. Anderson (1965) describes a skeleton from Tehuacan which shows bilateral hip dislocation together with spina bifida and spondylolisthesis. Wells (1963) reports that osteoarthritis was the commonest disease identified in Anglo-Saxon and early Post-Saxon remains, but that there has been a disproportionately low occurrence of the disease in hip joints. One of the cases he examined exhibited CDH in the left hip with a flattening of the femoral head plus supero-posterior extension of the articular surface of the acetabulum.

The incidence of CDH varies in different populations according to several recent studies. Comparison of these studies is probably of limited value since the criteria for diagnosis were not standardized. Major sources of error include the age of the patient at examination, qualification of the examiner, criteria selected for diagnosis, quality and quantity
of birth records and neonatal or fetal death reports, and availability of follow-up data. The usual figures given for incidence of CDH are from Europe and North America. It is known to be rare among Negroes, Bantu, and Chinese, but has been reported as more common among Navajos, Lapps, and certain regions in Japan and northern Italy. Wells wrote in 1964 (P.40) "Congenital dislocation of the hip appears occasionally in cemeteries from most parts of the world, but it attains exceptional frequencies, up to 10%, in Apache, Salteaux, and some other Amerindian groups". In a 1950 report Corrigan and Segal found prevalence of CDH in a Manitoba Indian community of 60 per 1,000. These ranged in age from 2 to 74 years and the study did not include newborns, so that the neonatal incidence is unknown. In 1962, Barlow reported that one infant in sixty in Manchester, England, was born with instability in one or both hips. Of this number over 60% become normal within the first week of life, and 88% within the first two months. The remaining 12% represent true cases of CDH and will persist unless treated. The overall incidence in this population was 1.55 per 1,000. MacKenzie (1972) presented a table of comparisons of incidence of neonatal abnormal hips as published in recent literature:

<table>
<thead>
<tr>
<th>RESEARCHER/DATE</th>
<th>TOTAL EXAMINED</th>
<th>ABNORMAL HIPS</th>
<th>INCIDENCE PER 1,000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Palmen/1961</td>
<td>12,394</td>
<td>70</td>
<td>5.65</td>
</tr>
<tr>
<td>Rosen/1962</td>
<td>31,200</td>
<td>68</td>
<td>2.18</td>
</tr>
<tr>
<td>Barlow/1962</td>
<td>9,289</td>
<td>139</td>
<td>14.37</td>
</tr>
<tr>
<td>Stanislaviljevic/1964</td>
<td>6,000</td>
<td>29</td>
<td>4.84</td>
</tr>
<tr>
<td>Weissman/1966</td>
<td>16,841</td>
<td>45</td>
<td>2.68</td>
</tr>
<tr>
<td>Smaill/1968</td>
<td>6,000</td>
<td>24</td>
<td>4.0</td>
</tr>
<tr>
<td>MacKenzie/1970</td>
<td>76,675</td>
<td>1,671</td>
<td>21.80</td>
</tr>
</tbody>
</table>

These surveys were all northern European and all were done after
1960 in response to the work of von Rosen (1962) and Barlow (1962) who demonstrated the possibility of diagnosis of CDH at birth.

Diagnosis at birth is based on the following criteria:

1. Ortolani's test - with hips and knees flexed the hips are slowly abducted. In dislocation the femoral head will slip back into the acetabulum with a visible and palpable "click".

2. Asymmetry of the hips and gluteal folds.

3. Restricted abduction with abduction contracture on opposite side.

4. Tendency to dislocation with manipulation.

In his study of a Navajo population of 2,300 at Many Farms, Arizona, Rabin (1965) divided the target population into two groups: adults aged 30 to 50 years and children aged one to six years. These groups were examined and radiographed for evidence of hip dysplasia and dislocation. The dysplasia rate was 0.7% in children and 2.6% in adults. Rabin's study offered an unusual opportunity to observe the natural history of untreated CDH as no treatment had been available for the adults and treatment was rejected by Navajo parents for their children. Observation of children through the time of the study showed that three-fourths of the cases diagnosed as dysplastic showed marked, spontaneous improvement to normal or near normal. In the adult population aged 30 to 50 there was no significant increase in the occurrence of osteoarthritis except for a few cases of dislocation or severe dysplasia. The Navajos did not consider the disease particularly incapacitating or worth treating unless there was associated pain. They sometimes recognized the disease as congenital and they explained these cases in
various ways: the pregnant woman who had been butchering had cut through the animal's joints, or the husband had twisted a horse's legs while branding or castrating it. The characteristic limp was often assumed to be caused by an injury in which cases, the parents would sometimes resort to a Navajo curing ceremony. The offer of surgery for CDH was generally refused. In general, the Navajo did not accept the idea that CDH is a disease since an affected individual could still function freely in the traditional society.

Other groups of Navajos were among the Indians examined by radiologists at the Phoenix Indian Medical Center. Goldman (1972) published a report on the evaluation of pelvic x-rays taken for various reasons between 1955-1970. Both untreated and unrecognized CDH was found among all age groups, but complaints related to the disorder were rare.

The high prevalence of untreated CDH in Corrigan and Segal's 1950 study of Indians at Island Lake, Manitoba, also presented an opportunity to observe the course of the untreated disease. The Island Lake population in 1949 totaled 1,253 and was composed of three distinct totems and many different families. Diagnosis on the basis of history, physical examination, and radiography disclosed 44 living cases of CDH. The characteristic history was of a lurching or waddling gait dating from the first attempt to walk. The physical examination showed obliquity of the hips, asymmetry of the inguinal, labio-femoral, and gluteal folds, shortening of the affected leg from one-quarter to two and one-half inches in unilateral cases, displacement of the greater trochanter of the femur in relation to the anterior superior spine of the ilium and the ischial tuberosity, and in bilateral
cases extreme lumbo-sacral lordosis. In all cases there was a limitation in the range of abduction of the affected hip and a positive Trendelenburg sign (the inability to maintain the pelvis in a level position when weight is borne on the affected hip). The positive Trendelenburg sign is apparently the consequence of the disorientation of the levering action of the gluteus medius. Radiography of the pelvis with the subject standing showed hip dislocation in 43 of the 44 cases. The femoral head or its epiphysis was displaced above the level of the acetabulum. In young children the epiphysis was located external to a line perpendicular from the outer edge of the acetabular roof and above a line through the upper limit of the ischial bones. In this group the proportion of unilateral to bilateral dislocations was 6:5 and the proportion of females to males affected was 6.5:1. Genealogical studies were made to determine the relationships of affected individuals. A genealogical chart was prepared to show the examined propositi and all relatives living or deceased who were suspected of having the disease because of an anomalous gait. A total of 94 known or suspected cases appear on the chart and all are connected by blood relationship. Corrigan and Segal concluded that there was strong presumptive evidence that the primary etiology is genetic.

Wynne-Davies (1970a) conducted a survey of 589 children examined at orthopedic clinics in Edinburgh and Glasgow or collected from a large random sample of hospital records with the purpose of comparing the factors affecting neonatal and late-diagnosis CDH. As noted before the incidence of neonatal CDH appears to fluctuate more widely than some more easily recognized
deformities. Wynne-Davies gives figures from recent surveys in England and Scotland ranging from 3.9 per 1,000 to 15 per 1,000 live births (in the latter study, Barlow/1962, half of the hips were classed as unstable, not dislocated -- see chart, P. 3). Surveys for CDH before neonatal diagnosis became common indicated an incidence in the British Isles of about 1 per 1,000 and Wynne-Davies accepts this figure as reasonable for late-diagnosis CDH. All of the identified cases in her study were analyzed for environmental and genetic factors previously proposed as affecting the incidence of CDH.

Environmental factors reported to have significance in the incidence of CDH include paternal age, birth order, pregnancy history, season of birth, and social class. In Wynne-Davies' study no significant difference could be attributed to paternal or maternal age, but the frequency in first-born children was highly significant for both neonatal and late-diagnosis groups. An increased number of breech presentations or versions was reported in this and earlier studies, but breech presentation is also more common in first-borns. CDH births were more common in the winter months in both neonatal and late-diagnosis cases. In social classes 1 and 2 (i.e., professional and managerial), there was a significant increase in the neonatal CDH group, even though it is likely that in recent years a larger proportion of lower classes are seen regularly at clinics.

Wynne-Davies discusses three aspects of the heredity of these cases: family incidence, inheritance of acetabular dysplasia, and inheritance of joint laxity. The proportion of affected first degree relatives was significantly higher than
expected. The figures for affected parents are lower than for sibs and children, partly due to the lack of neonatal diagnosis in the parents' generation and partly because it is probable that fewer CDH patients than normal individuals marry. There were too few twins in this survey to draw any conclusions, but larger studies in the past reflected a concordance of 35% in monozygotic twins compared with a 3% in dizygotic twins. MacKenzie (1972) surveyed the literature on twin studies and reported a rate of concordance in monozygotic twins of 42.7% and in dizygotic twins of 2.8%. The pattern of inheritance in first degree relatives could be dominance of reduced penetrance or polygenic. More affected second degree relatives were reported on the maternal side than on the paternal, but Wynn-Davies suggests that this probably reflects a greater knowledge on the part of the interviewed mother of events in the preceding generation of her own family. There was a rapid drop in incidence among third degree relatives which would indicate polygenic rather than dominant inheritance.

Wynne-Davies believes that the configuration of the acetabulum is inherited as a multiple gene system. The measurements of the shallowness of the acetabulum in the children in this survey were not found to be meaningful because radiographic assessment in the absence of complete ossification was unsatisfactory. However, the parents of 162 of the index patients were radiographed and compared with a control series on the assumption that if CDH has a genetic component some deviation from normal should also appear in the parents. It was found
that the apparently normal parents of the late-diagnosis CDH patients had a significantly shallower acetabulum than the control group. Figures for parents of the neonatal group showed the same trend but not at a statistically significant level.

In this survey lax joints were demonstrable not only in the index patients but in parents, sibs, and children in numbers significantly higher than in a control survey. This apparent disorder of the connective tissue occurred in a high proportion of both sexes and all generations, suggesting a possible dominant inheritance. Wynne-Davies considers it probable that two independent gene systems are operable in CDH. One is a polygenic system affecting development of the acetabulum which has in the past contributed to a high proportion of late-diagnosis CDH, and the other concerns the laxity of the hip joint capsule (as well as other joint capsules) which influences the great majority of cases of neonatal CDH and is probably of dominant inheritance. Any group of neonatal CDH patients will contain cases with both etiologies with a preponderance of perhaps 80% of the joint laxity type, and any group of late-diagnosis CDH patients will also contain both etiologies but with a preponderance of the primary acetabular type. Since these traits are common in the general population, some patients will have both defects, and in the few families in which it could be shown that both defects were present, there appeared to be an increased risk of CDH. Tentative calculations for the heritability of each defect were made on the basis of incidence in first degree relatives. For the acetabular dysplasia type, assuming a population incidence of 1 per 1,000 and
noting the proportion of affected relatives to be 3.6%, the heritability was found to be nearly 80%. For the joint laxity type the population incidence was assumed to be 4 per 1,000 and the proportion of affected sibs was 13.4%, giving a heritability in excess of 100%.

In a separate paper based on the same survey Wynne-Davies (197 Ob) asked the following questions:

1) What is the role of generalized joint laxity in CDH?

2) What is the role of delayed ossification of the femoral head and is it primary or secondary?

3) What is the place of primary acetabular dysplasia in CDG?

Summarizing the results from the parallel report (197 Oa), she found a higher proportion of children with CDH were lax-jointed than in the control group, and a higher proportion of neonates with CDH and their first-degree relatives had joint laxity than did the late-diagnosis group. A comparison of the ages at appearance of the ossification center of the femoral head in a control survey and in neonatal CDH patients indicated no significant delay in affected hips. Since this paper (197 Ob) is more concerned with the pathology of CDH Wynne-Davies discusses acetabular dysplasia in detail. The degree of acetabular dysplasia is determined through radiographic measurement of the CE angle, defined as follows (P. 705):

"The head of the femur is a sphere and thus its centre can be found. This point is joined on the left and right sides, a perpendicular erected and a second line drawn from the center to the edge of the acetabulum. The angle between
is the CE angle and is a measure of the coverage of the femoral head and of the depth of the acetabulum."

(Figure 1) An attempt was made to measure the "normal" hips of unilateral CDH patients, but satisfactory measurement of incompletely ossified acetabula was impossible. Instead, the parents of the index patients were radiographed and compared with a control group of radiographs, as described in the previously discussed paper. Histograms of the raw data show a slight but insignificant shift in distribution to a shallower acetabulum for the parents of index patients. It is known that the CE angle increases gradually throughout adult life, so, in order to eliminate the age effect a mean angle was calculated for each age in the control group and degrees of deviation from the mean were determined. In this way a significant shift to a shallower acetabulum was shown in the parents of the late-diagnosis CDH patients, along with a smaller shift for parents of the neonatal group, not at a significant level.

In discussing the significance of the findings of acetabular dysplasia, Wynne-Davies suggests two possibilities: 1) Acetabular configuration is an inherited character (probably multiple
gene) which allows easy dislocation and discourages maintenance of reduction. 2) The CDH parent, though apparently normal, was lax-jointed in infancy with dislocation or subluxation producing a secondary shallow acetabulum. There is no direct evidence to determine which factor is significant. However, the groups of CDH parents with and without acetabular dysplasia were compared as to several clinical factors and Wynne-Davies states that the dysplastic group has a more "genetic" look (1900b, P. 714). In addition it was determined that as the degree of dysplasia becomes more severe the proportion of relatives with dislocation increases, a result to be expected in a disorder of multiple gene inheritance.

A more recent paper by Wynne-Davies (1972) summarizes several reports of familial concentrations of CDH and the possible role of familial joint laxity and primary acetabular dysplasia as genetic factors. Environmental factors are still considered important in the development of disease in susceptible persons. There have been several studies investigating the relationship to breech births or late versions, as well as to birth order, it being presumed that a first-born child is more constricted in a primaparous uterus and that breech position, especially with knees extended, is conducive to dislocation of the hip. All these surveys have shown an increased incidence associated with breech presentation and primaparity. Hummer and MacEwen (1972) report an increased incidence of torticollis and CDH in the same patient significantly higher than expected. In a group of 70 children, aged 3 weeks to 1 year, with torticollis, there was an incidence of 5% CDH plus an additional 15% with congenital subluxation, whereas the expected co-occurrence is calculated
to be 0.006%. The report suggested a parallel etiology, uter-
ine compression, might account for the unexpected conjunction
of these defects.

According to Wynne-Davies (1972) several centers have re-
ported a significant seasonal increase in the number of CDH
children born during the winter months. In some cases this
increase may have been related to the fact that infants are
more tightly wrapped during the winter months, a situation
which forces the hips into extension and tends to maintain
any early dislocation which would have reduced itself if the
child were able to kick and maintain a greater degree of flexion.
This factor may account for some of the increased frequency among
races which swaddle their newborn such as the Lapps and the
Navajo. However, there is also an increased incidence in winter
months apparent in newborns who have not been swaddled. The
wrapping or swaddling theory cannot account for these.

A study done in Israel in 1962-67 (Chen, 1970), reported
on the incidence of CDH in newborns. The rate was found to be
significantly higher among infants born during the cold months.
However, comparison of weather data in this study with that in
England during similar studies showed that the temperature range
of the "cold" season in Israel is comparable to the "warm" season
in England, ruling out a suggestion that temperature variation
is somehow a factor. Chen and associates formulated a hypothesis
concerning the gestational age of susceptibility (the 4th or 5th
month) to an unknown seasonal factor.

It has been proposed in several papers that the effect of
maternal hormone secretion on the fetus may be a factor affecting
joint laxity and consequently the predisposition to dislocation. Woolf (1968) cites a study of urinary excretion of hormones in 26 normal and 11 CDH infants which showed an increased level of estrone and estradiol in affected infants. The relationship between hormone levels and joint laxity is described by Woolf as follows: "A developing fetus receives estrone and progesterone across the placenta. The fetal adrenal glands also produce progesterone, and the fetal ovaries produce estrone. These hormones produce minor degrees of joint laxity by their direct action on fetal ligaments. These hormones also stimulate the fetal uterus to produce relaxin*, which further accounts for the greater degree of joint laxity in females. An enzyme system present in the normal fetal liver converts estrone and progesterone to inactive products which enter the maternal circulation and are excreted. It is proposed that a failure of this enzyme system results in fetal hormonal imbalance, especially in the female. The consequence is generalized ligamentous laxity, which may initiate the chain of events leading to symptomatic hip disease" (P.435-6).

McKenzie (1972) reports that similar studies have failed to confirm this hypothesis. Wilkinson (1963) performed a series of experiments on young female rabbits attempting to reproduce human intro-uterine factors: joint laxity by injecting estrone and progesterone; and breech malposition by splinting of the knee in extension, since extension of the knees is thought to be the most harmful factor in the breech position. He was able to demonstrate laxity in the hip capsule, development of a limbus (a fold in the

*This statement is probably inaccurate. See Williams, 1974, P.383.
capsule between the femoral head and acetabulum which often presents reduction), and posterior dislocation of the hip joint.

To clarify some of the controversial aspects of the pathogenesis of CDH, McKibbin (1970) performed a detailed anatomical study of an infant with bilateral CDH who died a few hours after birth. Particular attention was given to the role of femoral anteversion (anterior rotation during fetal development) and the orientation of the acetabulum. McKibbin discusses the theory presented by Le Damany in 1908 that "when the leg is in the anatomical position the stability of the hip is influenced by the relationship to the sagittal plane of both the femoral neck and the acetabulum, so that if the former faces significantly forwards (anteversion) and the acetabulum is also inclined to the front a stable articulation may be impossible" (McKibbin, P.148). Measurements of femoral neck orientation have since then received frequent attention with ingenious techniques devised to measure the living subject. However, several studies of acetabular orientation have produced a diverse range of normal values. McKibbin suggests that lack of standard orientation of the pelvis in measurement has contributed a possible source of difference, for some were measured with the brim of the pelvis horizontal instead of in the anatomical position. "Since the acetabulum not only faces forward but downwards, it is obvious that alterations in the relationship of the plane of the pelvic brim to the horizontal will also alter the sagittal orientation of the obliquely placed acetabulum" (P.149). Standardization of measurement with the pelvis in normal anatomical position is de-
sirable, since it is in that position that dislocation is of concern. Normal anatomical position places the top of the pubic symphysis in the same vertical plane as the anterior superior spine of the ilium, and is conveniently reproduced in a dissected specimen by laying the pelvis prone so that these landmarks are simultaneously in contact with a level table top. The angle of the acetabulum to the sagittal plane is then read from an adjustable protractor held in the vertical plane against the greatest diameter of the acetabulum parallel to a line drawn between the two anterior iliac spines.

McKibbin presents the findings from dissection of this specimen of CDH in the context of the theories of etiology found in the literature. The concept of a primary acetabular dysplasia is one of the oldest proposed causes, but he found no evidence of dysplasia in this dissection and concludes that such dysplasia is secondary to dislocation. Another group of theories relates to Le Damany's concept of incompatible anteversion of the femoral neck and the acetabulum during early fetal development. These theories propose that failures in reciprocal alignment of the femoral neck and the acetabulum during early growth of the fetus may predispose the hip to dislocation. McKibbin says that these theories all imply that actual dislocation occurs after birth when the hip is first extended. He disputes this because dislocation in the fetus is well documented and reports that the dissected specimen showed displacement of both femoral heads within a few hours after birth. He found only one condition in the specimen which was unquestionably abnormal and that was the excessive laxity of the capsule. He concludes that the sequence
of events producing dislocation begins with a primary laxity in utero which permits dislocation of the flexed hip under the influence of fetal posture. Once the head of the femur dislocates the development of the acetabulum is interfered with and some degree of dysplasia will be present at birth. Capsular laxity diminishes after birth, producing a tendency for the hip to become stable. Other factors may oppose the stabilizing tendency, e.g., a short iliopsoas muscle causing a levering action on the femoral head during extension, acetabular or femoral anteversion, and any dysplasia which may have developed. In discussing optimum management of dislocation McKibbin notes that fixation in a cast is usually in one of two positions: full abduction and flexion, or abduction extension and medial rotation. Both have been successful, he believes, because either position eliminates the laxity of the capsule and offsets any defect in orientation of the femoral neck and acetabulum.

Several reports have been published which discuss the significance for CDH of familial generalized joint laxity as distinguished from either temporary hormonal joint laxity or the type of laxity found in certain syndromes. Such heritable disorders of connective tissue as Ehlers-Danlos syndrome, osteogenesis imperfecta, and Marfan's syndrome include a tendency to hip dislocation (Carter and Wilkinson, 1964a). Generalized joint laxity occurs more commonly uncomplicated by other abnormalities and usually with a familial incidence indicating a dominant inheritance of variable expression. In mild form it is present in about 7% of normal school children of either sex. In extreme form it can lead to incapacitating CDH or recurrent dislocation in several
joints. In their study of the prevalence of persistent joint laxity, Carter and Wilkinson (1964b) listed five tests of abnormality:

1) Passive apposition of the thumb to the flexor aspect of the forearm;

2) Passive hyperextension of the fingers to a position parallel with the extensor aspect of the forearm;

3) Ability to hyperextend the elbow more than 10 degrees;

4) Ability to hyperextend the knee more than 10 degrees;

5) An excess range of passive dorsiflexion of the ankle and eversion of the foot.

If more than three tests were positive the diagnosis of persistent generalized joint laxity was made (p.42). A comparison of incidence was made between a random series of 62 CDH patients aged 5 to 14 years and a control group of 285 normal children aged 6 to 11 years. Of the CDH patients with no first degree relative affected nearly one-third of the girls and over three-fourths of the boys showed joint laxity. Of the CDH patients with a first degree relative affected four of seven girls and five of seven boys showed joint laxity. Parents of patients with both CDH and joint laxity were also examined. In five of nine male cases one parent had obvious laxity; in all nine female cases one or both parents had laxity. Carter and Wilkinson conclude that persistent generalized joint laxity is an important predisposing factor for CDH in boys. It is less important for girls except perhaps in familial cases, because there is an alternate cause in the temporary hormonal laxity discussed above.

Kirk et al (1967) reported the results of a study of 24
patients with a "hypermobility syndrome" which they defined as various musculo-skeletal complaints which appeared to be related to generalized joint laxity. They conclude that this condition represents one extreme of a wide normal variation in joint mobility. Another view they suggest is that the isolated laxity is a mild disorder of mesenchymal development which lies at one end of a spectrum of heritable connective tissue disease with Marfan's syndrome and the Ehlers-Danlos syndrome at the other. Although the hypermobility syndrome is fairly common, associated disability is rare except for an apparent predisposition to premature degenerative joint disease (P. 423-5).

Beighton and Horan (1970) discuss the family histories of two patients with loose joints, one with no apparent disability and the other with bilateral hip dislocation and recurrent dislocation in several other joints. In both kindreds other family members were affected in the same way, without disability in the first case, and with many orthopedic problems in the second case. The researchers postulate two disorders representing two distinct and separate genetic entities because of the marked contrast in disability incidence in the two families. Both traits are apparently transmitted as autosomal dominants.

Two early reports of hypermobile joints were those of Whitney (1932) and Sturkie (1941). Whitney studied the incidence of double-jointed thumbs, apparently through a classroom survey. He reported a prevalence of 5 - 10% for the ability to bend the thumb at the second (basal) joint. He concluded that it behaves as a typical Mendelian recessive trait, but his research was fairly superficial. Sturkie examined the pedigrees for two cases of joint hypermobility and concluded that transmission of the trait is irregular. All
descendants of a single female in one pedigree exhibited the trait to varying degrees in different joints, although the propositus was affected only in fingers and thumbs. Her first generation descendants all showed the trait in joints of the fingers, thumbs, knees, and elbows, and 4 of 5 showed marked flexibility in hip and toe joints. Sturkie concluded that there is a great deal of variation of expression of the trait.

McKusick (1972) describes a simple test of joint mobility in which the 5th finger is bent backward and the angle with the proximal phalanx is measured. He reports a survey of 500 subjects which showed that most children have a high degree of joint mobility and that females have a higher degree of mobility between the ages of 15 and 50 years than do males (P. 352).

In a report on recurrent dislocations of the patella and shoulder, Carter and Sweetnam (1960) review the literature on familial joint laxity. Isolated familial joint laxity was first reported in the medical literature (in English) by Finkelstein in 1916, and since that time there have been numerous reports of joint laxity associated with various dislocations. In each report the pattern of inheritance was that of a dominant gene. Carter and Sweetnam surveyed 111 patients with recurrent dislocations to determine the relationship to joint laxity and its occurrence in affected relatives. Twelve of their patients had near relatives with similar dislocations. Of the ten having recurrent patellar dislocations in more than one family member two had familial joint laxity, and of the two with recurrent shoulder dislocations in more than one family member, both had familial joint laxity. In one family a mother and daughter both had bilateral CDH.
Carter and Sweetnam conclude that familial joint laxity is a sufficient cause for recurrent dislocation of the shoulder in more than one member of a family, but that other factors contribute to familial recurrent patellar dislocation.

According to James (1972) there are still a great many cases of CDH in which diagnosis is missed at birth. Several clinical studies have emphasized the high rate of success with conservative treatment of CDH identified in the neonate. In these cases, treatment has consisted of maintaining the hips in flexed abduction with a type of pillow splint. James raises the question as to whether the cases being splinted are not those which would have corrected themselves. The incidence of missed diagnosis of dislocation has approximated the previous expected incidence in several surveys. Late-diagnosis cases still require more aggressive treatment such as fixation of the hip in abduction through various cast techniques and/or osteotomy to reconstruct the acetabular roof.

Two recent papers summarize the changes secondary to CDH and the late results of inadequate treatment (Specht, 1974, and Primer on the Rheumatic Diseases, 1973). The principal secondary changes are a relative shortening of the adductor muscles which limits passive abduction, an increased slope to the acetabular roof, stretching of the supporting capsule and ligaments, anteverision of the proximal end of the femur, false socket development, and premature degenerative joint disease. In addition, late results of inadequate treatment include abnormal function of the gluteus medius causing a lurch to the lesion side (Trendelenburg gait), a shortened extremity, instability, and limited motion.
In a study of 124 hips requiring surgery for osteoarthritis 24 (20.9\%) were considered secondary to congenital dysplasia (Lloyd-Roberts, 1955). In this study the average age at onset of symptoms was 46.6 years. Others have reported from 31 - 40\% of osteoarthritis of the hip apparently related to CDH.

Woolf (1971) summarized the implications for genetic counseling in the genetic components of CDH. Existing data support the possibility of both polygenic and dominant types. Exogenous factors are apparently more important in females than in males in determining whether CDH will occur, and genetic factors are more significant in males. The risk to the sibs of a propositus is influenced by the sex of the propositus, whether the propositus was in breech malposition, the family history of the disorder, and the season of birth of the propositus. Nongenetic factors must be considered in counseling.

Improvement in the quality of screening for CDH is urged by Moore (1974) who states that too many cases are still being missed because of the lack of well-qualified examiners. He estimates the cost to the State (Ireland) to be in excess of 2,000 pounds for the treatment of one established CDH case, to say nothing of the cost to the patient of a life-time disability.

I have summarized the findings of several papers on the incidence and etiology of CDH. In spite of the success in recent years in identifying CDH in newborns and returning most of them to normal, hip disabilities are still being identified in about the same proportion as before in the populations surveyed.
Genetic counseling which takes account of both the polygenic and dominant factors discussed may reduce the incidence somewhat, but the relatively high frequency of these factors makes it unlikely that CDA will disappear, at least in the populations studied.
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