1958

A review of Legg-CalvÃ©-Perthes disease with an introduction to an improved method of treatment

Robert Lemoyne Yekel
University of Nebraska Medical Center

A Review Of Legg-Calvé-Perthes Disease With An Introduction To An Improved Method Of Treatment.

Robert Lemoyne Yekel

Submitted in Partial Fulfillment for the Degree of Doctor of Medicine

College of Medicine, University of Nebraska

April 1, 1958

Omaha, Nebraska

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I. Historical Introduction

Knowledge concerning the existence of Legg-Calvé-Perthes disease was made possible only after the discovery of the roentgen ray in 1895. This disease was discovered independently by four men in nearly the same year while they were studying tuberculous coxitis.

Arthur T. Legg(1), a Boston orthopedist, has been accredited with the first publication which appeared in Fubruary 1910, after he reported his findings to the American Orthopedic Association in 1909. While working as Junior Assistant Surgeon at Children's Hospital in Boston, Legg observed a hip disorder which did not follow the pattern expected of tuberculous hip disease. These children previously had been considered as having "atypical tuberculous coxitis", or "the quiet hip disease."

The case which attracted Legg's special attention was that of an eight year old boy who fractured his humerus. Two months later he complained of pain in his right leg and limped. Early observation was possible, and the sequence of changes was followed by x-ray. The opposite hip developed a similar process shortly afterwards with no signs of inflammation. This bilateral involvement stimulated Legg to re-examine many of his former cases. In his original paper Legg

described five cases including four boys and a girl who were between the ages of five and eight.

Jacques Calvé(2), a French orthopedist, prepared his material during the years 1908 and 1909, and he published his paper also in 1910. Using his newly acquired x-ray machine, Calvé had analyzed five hundred cases of children with hip disease. He believed that a number of these children clinically showed differences from the usual tuberculous infections. He selected ten of the five hundred cases which he considered to be of non-tuberculous origin. He originally thought that these children were manifesting signs of recovery from what he called "old rickets" which was associated with "coxa vara." He (3) later withdrew both ideas of the existence of rickets and of coxa vara in the disorder.

Perthes(4), a German surgeon, collected his cases during the years 1903 to 1909, and published his observations also late in 1910. Perthes thought that these cases represented an early form of "juvenile arthritis" in 1910. In 1913 he published his awareness of the disorder as a "self limiting, non-inflammatory condition, affecting the capital femoral epiphysis with stages of degeneration and regeneration leading to a restoration of the bone nucleus, although it

was flattened and yet not destroyed as in tuberculosis." In this publication he described the gross and microscopic pathology for the first time. He studied autopsy material from a child who was killed accidentally while in the early destructive and regenerative stages of the disorder.

A Swedish orthopedic surgeon, Henning Waldenstrom (5), discussed his observation in a paper published in 1909. He described the disease as a benign form of tuberculosis. Since he did not recognize that the disorder was non-tuberculous, he has not been given credit by most writers as contributing to the discovery of this disease.

Many names have been given to this disorder by its many investigators. Legg preferred to call the disease "osteochondral trophopathy", Perthes and Calvé preferred "pseudocoxalgia." The disease was first called Legg-Calvé-Perthes disease by Halfdan Sundt, (4) and this is the term used by most writers today.

II. Theories of Etiology

A. Trauma

Legg (1) in his first publication considered trauma with inadequate blood supply to be the most probable causative factor.

Axhausen(4) was the first to suggest a probable

relationship between an injury and mycotic emboli of blood vessels within the epiphysis. Gill(6) thought that a primary avascular necrosis of the medial portion of the metaphyseal side of the epiphyseal zone occurred which spread by slow progression beneath the growth zone. This indirectly was thought to interfere with the vascular supply to the ossification center of the femoral head.

Jansen(7) attributed the changes within the hip to an inherent increasing declivity of the acetabular roof. He said that this produces a flattening of the head of the femur and is followed by resulting traumatic joint changes. He postulated that during the embryological development, a small amniotic sac bent the fetus into increased flexion, or it would draw tightly over the fetal buttocks with a resulting ischium vacum deformity.

Zemansky(8) also believed that trauma was the causative factor in Legg-Calvé-Perthes disease. He thought that during childhood the blood supply of the epiphysis of the head of the femur by way of the periosteum and ligamentum teres is just adequate to maintain its nutrition. Following trauma one or all of these blood vessels may be sufficiently damaged to interfere with nutrition of the epiphysis, and necrosis

of the subchondral bone takes place. Repair is slow by means of fibrous tissue replacing bone fragments.

Brandes(4) proposed a developmental cause in which a hypolastic or dysytophic condition exists about the cartilaginous head and neck of the femur. This leads to an inadequate ossification, and then slight traumata may compress the cancellous bone and lead to avascular necrosis.

Spisic(4) proposed that an anlage of disproportion between the femoral head and the acetabulum is the primary cause, and that the disorder results from trauma to the capital epiphysis after walking has been learned.

Calot(4) proposed as a primary cause "minute subluxations" after which the trauma of weight bearing might produce avascular necrosis. He based this idea on the fact that trauma resulting in avascular necrosis of the capital femoral epiphysis is a frequent occurrence in a subluxated or dislocated hip which has been reduced either by an open or by a closed method.

Ponseti(9,10,11) in experiments with rats, attemped to prove his theory that diet as well as trauma is instrumental in causing Legg-Calvé-Perthes disease. He related that there was slipping of the epiphyseal plates in young rats which were fed diets containing

aminonitriles, and even where there was no gross evidence of epiphyseal slipping, the femoral head later became necrotic simulating Legg-Calvé-Perthes disease. Microscopic studies of the femoral heads of these rats were similar to studies of biopsy material which was obtained by means of a Phemister punch from a boy and a girl with Legg-Calvé-Perthes disease. Ponseti concluded that faulty epiphyseal plate cartilage was the primary lesion. He believed that necrosis of the epiphysis ensues when its blood supply becomes interrupted. This may occur when the retinacular vessels are occluded or impaired by trauma from jarring where the vessels cross the thick layer of cartilege at the edge of the weakened and badly disrupted epiphyseal plate.

Many authors have cited that trauma is also a likely cause because in almost all large series of cases, ninety per cent of patients with Legg-Calvé-Perthes disease are boys. They are assuming that boys are much more active than girls during the latent period which is difficult to prove however. Most authors of today believe that trauma does play an important role in the cause of this disorder.

B. Infection

A few men have postulated that a blood stream

infection preceded the joint manifestations in some instances of Legg-Calvé-Perthes disease. Those who followed this infectious theory, especially Phemister (12), often cultured material obtained at operation, and they have obtained positive cultures, usually of Staphalococcus aureus. These cultures were also obtained by Legg(11), but even in his day he considered them to be results of contamination as have most present day investigators, including Goff(4). Syphilis has not been present with Legg-Calvé-Perthes disease except incidentally.

There has been no evidence of seasonal or epidemic relationships to Legg-Calvé-Perthes disease(4).

C. Endocrine Factors

There have been sharp differences of opinion as to whether hypothyroidism is a contributing factor in Legg-Calvé-Perthes disease. Deficient thyroid secretion leads to retarded skeletal maturation. Cretins and children with juvenile myxedema show a femoral capital disorder on x-ray which simulates somewhat the avascular necrosis in Legg-Calvé-Perthes disease.

Cavanaugh, Sheldon, and Sutherland(13) reported cases of Legg-Calvé-Perthes disease which showed delay in bone maturation which was reversed by thyroid therapy. Emerick, Corrigan, Joistad, and

Holly(14) found that all of their last thirty-five patients with Legg-Calvé-Perthes disease had either hypothyroidism or toxic adenomata when radioactive iodine uptake studies, including mapping of the thyroid gland, were done. When these cases were treated with thyroid, or the adenomata were removed along with conventional orthopedic treatment, reversal of the early changes occurred, or the more progressive cases healed more rapidly. In two instances exogenous thyroid therapy was discontinued after a demonstrable early response was obtained but before significant healing had taken place. A "disasterous relapse followed immediately." Others have doubted the accuracy of the diagnosis of Legg-Calvé-Perthes disease in these cases.

Katz(15), using serum protein bound iodine determinations and serum cholesterol levels, could find no evidence of abnormal thyroid function in thirty-two patients with Legg-Calvé-Perthes disease by means of clinical examination, serum protein bound iodine determinations, twenty-four hour thyroid uptake studies, and even salivary radioactive iodine concentration on seven of the ten patients. He also failed to find any evidence of hypothyroidism.

Many authors have showed evidence that the growth rate has been depressed or diminished just prior to

the onset of Legg-Calvé-Perthes disease. It has been suggested that perhaps this might also be due to a less active pituitary growth harmone(4).

D. Nutritional factors

Calvé and Sundt believed that Legg-Calvé-Perthes disease was just a manifestation of old rickets, but this has been refuted by later investigations. Others have suggested that there might be early faulty metabolism in Legg-Calvé-Perthes disease, but they have not been able to prove this idea. As previously mentioned, Ponseti(9) has shown that young rats which were fed aminonitriles, develop aseptic necrosis of the femoral head which was similar pathologically to biopsies taken from the femoral heads of children with Legg-Calvé-Perthes disease. This suggests that perhaps some factor in the diet along with faulty metabolism contributes to causing this disease. There is no evident alteration of calcium or phosphorous metabolism in studies made by most investigators(4).

E. Congenital Malformations theory

Jansen(7) postulates that Legg-Calvé-Perthes disease might be caused from tightness of the amniotic sac over the fetus. He suggested that this disease was a "satellite to a congenital dislocation of the hip.". Adams(4) suggested that it may depend upon a congenitally

abnormal epiphyseal system. Hart(4) also suggested the addition of an "abnormal ossification or congenital dysplasia" to the probable causes.

F. Constitutional Anomaly

Nagasaka(4) found "excitability of muscles, eosinophilia, diminution of inorganic serum phosphorus, delayed endochondral ossification, transient hypertension, normal temperature, and a disturbance of the sympathetic nervous system" to be operating as a complex clinical syndrome in eight cases of Legg-Calvé-Perthes disease. These findings as a group have not been substantiated by other investigators of this disease however.

III. Heredity

Many have tried to link heredity with this disease. Perhaps the most striking study is that presented by Stephens and Kerby(17) when they found the occurrence of Legg-Calvé-Perthes disease in twenty-eight members of a family throughout five generations. Several investigators found cases of Legg-Calvé-Perthes disease in identical twins and in fraternal twins(18,19). Hamsa and Campbell(20) found Legg-Calvé-Perthes disease in three brothers, none of whom were twins. They suggested that this might represent a familial predisposition to an inherent hip weakness or a familial endocrine change.

IV. Nature of the Disease

A. Clinical Features

The age incidence for Legg-Calvé-Perthes disease is usually between two and twelve years of age. The majority of the cases occur in children between the ages of four and nine. Ninety percent of the cases are boys, and ninety per cent of the cases are unilateral; only ten per cent occur bilaterally.

This disease has been found most often in patients who have been otherwise completely well. The first complaints are of indefinite discomfort in and about the affected extremity. The pain is not severe and is often referred to the knee. The patient does not complain of severe pain when pressure is applied to the affected hip, but he only notices a slight discomfort. Limping is common as the child favors the area of discomfort. In some cases the pain and limp will disappear in a few days after its first appearance, only to recur again after overexertion.

There is thought to be a difference in these children for a variable time before the onset of pain. Goff(4) says that the parents will describe a child who has not been as robust as other members of the family, and that frequently they will say that the child has not grown at the same rate as other children. There

is a lack of appetite and a tendency to tire more quickly. These children appear to experience frequent respiratory diseases, and they seem to have a slow recovery rate from their illness. However some investigators say that they fail to find any of these differences in children with Legg-Calvé-Perthes disease and other normal children.

Perthes(4) described swelling of the inguinal lymph nodes in the early phase of this disorder, but others have not supported this finding. Detalia(21) describes a muscular spasm about the affected hip. Hip motions show that internal and external rotatory motions exhibit the greatest limitations. There is also a lesser degree of limitation of abduction. Adductor muscle spasm with resistance to abduction is usually present as an early clinical sign. Fawcitt (22) described wasting of the affected hip, a prominent trochanter on the affected side, and limitation of flexion as occurring in some cases also. Gill(23) believed that he found thickening of the involved hip, but this has not been the experience of other investigators.

B. Early Signs for Diagnosis

Early diagnosis of Legg-Calvé-Perthes disease is most desireable if good results from treatment are to

be attained. Since roentgenographic changes in the bone take place slowly, there is usually little change in the bones at the onset of symptoms. Therefore if error is made in not realizing the earliest changes on x-ray, and a child is allowed to walk for two or three months until more definite changes have occurred, irreparable damage is done to the child.

Kite and French(24) have made an extensive investigation for evaluation of the most reliable early signs in Legg-Calvé-Perthes disease. They have come to the following conclusions as a result of their study.

The earliest roentgenographicsign is a slight lateral separation of the head of the femur from the medial wall of the acetabulum. This is present in ninety-nine per cent of cases.

The obturator sign is next most helpful. A smaller obturator foramen is found on the involved side when measurements of the obturator foramina are done on the x-rays. Blanchard (25) thought this was caused by atrophy of the ischium and the pubic bones. Kite however disagrees with this explanation.. He feels that the obturator sign is due to a slight rotation of the pelvis as it is present in other afflictions of the hip. He made numerous x-ray studies of a skeleton

in different positions to prove this. The patient with Legg-Calvé-Perthes disease is not able to balance himself flat on his back, but finds it easier to rotate slightly to the side which is not affected. Kite found the obturator sign to be present in ninety-seven per cent of his patients who had Legg-Calvé-Perthes disease.

Another early sign used by Kite is that the head of the femur is sometimes shaped like the "roof of a house" on x-ray examination. Normally he found that there may be a slight flattening of the medial convexity of the head as seen in the anterior-posterior x-rays, but there is no flattening of the lateral half of the head. In more than a third of the early cases Kite found that the lateral half of the femoral head loses its convexity and is flattened. He calls this the "roof sign".. His reasoning for the development of this sign was that the softening of the head with destruction probably first began in the lateral and anterior surfaces. This results in flattening of the lateral convexity.

The fourth early sign which has been helpful to Kite has been a measurement of bone atrophy on the involved side with a photoelectric light meter. In order to detect the bone atrophy the photoelectric light meter is held closely to the film and the two

hips are compared. The reading may show a difference of only one point in an early case, but may show as much as six points difference in the more advanced stages.

C. Roentgen Features as the Disease Progresses

Careful evaluation of both clinical and roentgenographic findings in Legg-Calvé-Perthes disease have shown that x-rays have been a more reliable criteria for following and treating the patient. Therefore most clinicians have followed the course of the disease with x-rays which are taken every two or three months during the active stages. It has been recommended that anterior-posterior, lateral, and two oblique views be obtained, especially in the early stage, to facilitate accurate diagnosis. Later anterior-posterior and lateral views may suffice.

1. Incipient Stage of Development

The earliest changes are manifested by a synovitis on the x-ray. The capsule is distended. There is prominence of the soft tissues over the capsule of the hip joint laterally with or without increased density of the capsular area. The intermuscular fat line lateral to the hip normally is close to the joint margin and is straight or only slightly curved. Thickening of the capsular tissues and effusion in the joint

cause lateral displacement of this line and shortening of the radius of curvature. Increased density of the involved tissue is usual. It is at this stage where the child warrants a six month follow-up to make certain whether this is a simple synovitis which will resolve spontaneously, or whether it is the preliminary stage of Legg-Calvé-Perthes disease.

2. Early Active Stage

The active stage has been found to last three to eighteen months in most cases. Its onset can be established reliably when widening of the cartilaginous joint space occurs which is often most noticeable inferiorly. It is the most charecteristic feature of Legg-Calvé-Perthes disease. Other early evidence of the active stage is an increased density of the capital femoral epiphysis, osteoporosis of the metaphysis with or without irregularity of the neck, slight flattening of the crest of the head, widening of the epiphyseal line, and beginning shallowness and broadening of the head which is thought to be due to a delayed conversion of cartilage to bone rather than to compression of bone.

3. Later Active Stage

During this stage the greatest damage occurs. One or more areas of increased density appear within

the femoral head. These are believed to result from solution of the calcium in a necrotic area with precipitation of it diffusely in and about that same area. Irregularity of the outline of the head and neck develops which resemble scalloping. There is a greater widening of the joint space, shallowness of the head, and broadening of the head and neck. The neck of the femur may develop translucent, sponge-like areas. There is further compression and thinning of the fragmentary epiphysis with fragmentation of the head and further expansion of the metaphysis. At times the head appears to be almost entirely destroyed. The area of the head which is supplied by the vascular channels of the ligamentum teres may sometimes be spared the aseptic necrosis.

The appearance of the acetabulum indicates a change in contour which suggests a widening associated with unossified cartilage. There are also changes of condensation and rarefaction along the acetabular margin. Ischium varum is also common, but is usually a transient feature, and is not present until late in the active plase. For this reason it has been dismissed as a possible cause for Legg-Calvé-Perthes disease. Sometimes there is protrusion of the head laterally beyond the ilium due to the broadening of the head and the widening of the cartilaginous space inferiorly.

In the transition from the active to the reparative stage there is a fading or beginning absorption in the previously dense areas with appearance of irregular ossification. New areas of density may appear while the first areas are becoming irregularly ossified. The portions of the head and neck that showed the earliest evidence of necrosis are the first to show signs of recovery. There is occasional irregular ossification in the cartilaginous areas at the margins of the epiphyseal line. Obliquity of the roof with lengthening of the acetabulum develops.

4. Reparative or Healing Stage

This stage has usually lasted from fourteen to forty-eight months with past methods of treatment. Healing occurs first in the metaphysis and later in the head. The metaphysis is restored to normal long before the regeneration of the head is completed. The dense areas are almost completely replaced by the irregular ossification which is becoming more regular. There is beginning regeneration of the epiphyseal zone. Reossification by a "creeping substitution" takes place with appearance of coarse trabeculations.

The femoral head becomes less flat as the rounded contour is derived from the highest remaining point of the head. The acetabulum tends to conform to the contour

Fig. 1. Incipient Stage

of the head. Distension of the capsule diminishes if it has not already done so, and the joint space becomes less wide.

5. Definitive or Residual Stage

The epiphysis is completely reossified, and normal bone trabeculations are present. The margin of the head where it has grown down over the neck develops a dense line in crossing the neck which is often quite near the trochanters. When this line is definite, the healing process may be regarded as completed although occasionally

Fig. 2. Early Active Stage

there is a little furthur repair to be accomplished. Rarely and only in mild affections is there any length-ening of the neck.

These x-ray descriptions are a compilation of the studies by Eyre-Brook(26), Ferguson(27), Bettman (28), Billing(29), Gill(23), Goff(4), Lewis(30), Levy (31), and Pike(32).

D. Pathological Features

The gross features of Legg-Calvé-Perthes disease have been described from appearance at the time of

Fig. 3. Late Active Stage

operation when the hip joint has been exposed(23,33). In the active stage the synovial membrame appeared thick-ened, soft, fragile, very vascular, and often irregular with villus formation. In hips exposed during the reparative stage, the synovial membrane was smooth, inelastic, thin, and avascular. The periosteum and capsule were scarred
and inelastic also.

Pathological changes in the bone of the femoral head were not synchronous with the changes in the soft tissues. The earliest cases showed no definite gross

Fig. 5. Definitive Stage

within the head and neck of the fenur were constant within fairly normal limits. There were aseptic necrosis or necrobiosis, evidence suggestive of crushing, concurrent degeneration and repair, partial ossification of displaced cartilaginous tissue, loss of polarity of chondrocytes, and areas of cystic formation with giant cells in the walls. There were few lymphocytes, monocytes, or segmented neutrophils. Absence of infection has been supported by negative cultures.

There were no vessels in the degenerated areas.

No consistent vascular changes or thrombi were seen. In some cases large tongues of fibrillated cartilage containing clustered cells, debris, and extravasated blood appeared to penetrate deeply into the femoral neck. The intermingling of the living and the dead tissue would make infarction as a cause of necrosis appear to be unlikely.

E. Laboratory Findings

There have been no consistent reports which relate significiant changes in numerous series of varied laboratory tests(4). Except as incident to other con-comitant disease processes, the serum calcium and serum phosphorous levels have remained unchanged, mantoux tests have been normal, and the white blood cell counts have been normal or very slightly elevated to about ten thousand. Synovial fluid which has been obtained from the affected joint has repeatedly been normal on examination.

V. Differential Diagnoses

Legg-Calvé-Perthes disease was first differentiated from tuberculosis ith which it had been included until the discovery of x-ray. It usually can be differentiated clinically from tuberculosis since the involved hip joint in tuberculosis is markedly thickened and softened.

With an adequate history, clinical and appropriate

laboratory data, and also x-rays, such diseases as acute synovitis, acute transient epiphysitis, acute rheumatic arthritis slipping of the femoral capital epiphysis, congenital coxa vara, and congenital dislocation of the hip can be ruled out without much difficulty.

In Legg-Calvé Perthes disease there is no bony impairment other than in the hip joint which is seen in the skeletal survey. In hypothyroidism there is a generalized delay and irregularity in the calcification of the epiphyses, and there is delayed linear growth in the long bones as well as in the dentition. Dysplasia epiphysialis multiplex is characterized on roentgeno-grams by the mottled irregular density of several epiphyseal centers. Metaphyseal involvement in Legg-Calvé-Perthes diseese causes broadening of the femoral neck, but not the degeneration of the femoral neck which

is found in coxa varum. Chondrodystrophy may cause broadening of the femoral head and neck also with contour defects of the head and widening of the cartilaginous joint spaces. There is however no flattening of the femoral head or increased density within the femoral head. Diagnosis of Legg-Calvé-Perthes disease is dependent upon knowledge of the symptoms and x-ray changes along with a high index of suspicion.

VI. Treatment Methods

Just as we have not advanced far beyond Legg's theory of the etiology of Legg-Calvé-Perthes disease, neither have we advanced far beyond his ideas for treatment(34). In his opinion very little treatment is necessary, and no operation is needed. He felt that just any facility which diminishes strain upon the hip" would be adequate. Danforth(35) found that he obtained completely normal functioning hips with non-weight bearing in patients who were thought to have tuberculosis of the hip in 1909 to 1913, but restudy of these cases showed that they had Legg-Calvé-Perthes disease. The treatment which he used at this very early date was traction in bed, traction in hip splints, or plaster of Paris spicas.

Many methods of facilitating non-weight bearing have been devised with intention to shorten the course of healing and to a low the child to be ambulatory as soon as possible. The object is to minimize both the development of psychological upset from being an invalid and time lost from the child's educational development. The clinical aims of treatment are two-fold, first to maintain full range of movement with no predisposition to osteoarthritis from a misfit in the acetabulum, and second to produce a round head on the

roentgenogram which is adapted to the acetabulum, approximating the normal hip joint as closely as pos-sible(26).

Jansen(7) attempted to explain why non-weight bearing should effect a normal result and also hasten the healing phase. He thought that bone which had become plastic through undue weight bearing will deposit new lime salts and again become firm if relieved of weight bearing for a number of months. He believed that a flattened head would result from a flattened socket if weight bearing were allowed. He thought that the result is nature's way of reestablishing congruency between the head and the acetabulum.

Many methods of non-weight bearing have been tried including: (1) complete bed rest; (2) a Wu-splint because of lack of cooperation on bed rest; (3) the Thomas ring caliper brace; (4) an ischial non-weight bearing brace with elevation of the opposite shoe; (5) a Synder sling; (6) crutches; (7) a spica cast for four months and then an ischial non-weight bearing brace with elevation of the opposite shoe; (8) a cast for four months and then a Thomas ring caliper brace; (9) elevation of the opposite shoe and crutches; and (10) continuous traction with Buck's extension(35, 36, 37, 38, 39, 40, 41).

Most of these methods have been carefully studied

by Levy and Girard(31). They concluded that treatment should begin ideally when changes are evidenced as only an increased density in the femoral head. Then the patient should be placed in bed with or without tract-ion until healing is complete. Treatment by rest in bed with Buck's extension traction has lead to such complete restoration that in the final result it was difficult to detect any deformity in the head, neck, or acetabulum(23). This treatment however leads to the problems of cooperation and prolonged recumbency.

The practical treatment of choice which is used by most clinicians appears to be crutches with an ischial non-weight bearing brace on the involved leg, and with a three-inch cork elevation on the shoe of the well leg. This is inexpensive, readily available, and has good results. Full cooperation of the patient is neces-sary or he will need complete bed rest. Where bilateral involvement is found, however, complete bed rest is mandatory.

Many methods have been attempted to hasten the course of healing. Exogenous thyroid(13) and aureo-mycin(42) as growth accelerator factors have been ad-ministered. Drilling (43) two or three channels into the femoral head through a Smith-Peterson incision to increase circulation has been done. Operation(44) to

remove the debris of the necrotic tissue with replacement of healthy bone grafts has been performed. The men who have used these methods to hasten healing have reported successful results, but this has not been corroborated by others. More evaluation and research is necessary before these methods can be accepted.

A return to weight bearing during the course of treatment is justified when no new dense areas have appeared in the femoral head for two months, and when there are no dense areas in the line of stress trabeculae of the head. The patient must have full motion of the hip joint. There must also be evidence that reconstruction is taking place in the head. It is not necessary to wait for a completely remineralized head, but sufficient bone structure should be present to assure confidence in the preservation of that structure(33).

A well balanced diet giving special attention to vitamins is desirable during treatment also to promote healing.

VII. End Results

The final results after being afflicted with Legg-Calvé-Perthes disease appears to depend upon how early the disease is detected and how well treatment prevents weight bearing and muscular spasm. If caught in the incipient or early stage, a very good functional hip

may be expected when properly treated with only coxa magna resulting. Widening of the neck takes place during the stage of necrosis, and then later the regenerating head matches the increased width of the neck. The head which is reformed without deformity other than increased size is consistent with good motion of the hip.

The deformities that arise in the head and neck are due to mechanical crushing of the necrotic tissue which are caused by weight bearing and muscle pull. This results in irregularity of the head with further shortening, flattening, and broadening of the he d and neck. Deformity of the acetabulum is dependent upon the altered shape and position of the head. With advancing age, as puberty is approached during the active stages of the disease, the end results are worse as more weight is carried and there is more mechanical strain.

A disabling feature which occasionally appears in the regenerative plase of this condition is a prominence of the lateral portion of the femoral head projecting beyond the superior acetabular rim. This is due also to weight bearing on the softened head. It results in a mechanical block to abduction, and frequently results in distortion of the acetabulum itself. A reconstruction operation may occasionally correct this lateral

prominence when done after complete regeneration has occurred.

Traum tic arthritis may appear at any time in later years following regeneration and is accompanied by increasing pain and or joint instability. It has also been suggested that osteochondritis dissecans may develop later but his has not been completely substan-tiated.

In ten percent of the cases which have bilateral involvement, usually the second hip becomes diseased within a year after the first has been affected. Results in the second hip have been worse in most instances. Worse results also occur obviously in the non-cooperative child who resists non-weight bearing(33, 45, 46, 47, 48).

VIII. An Improvement in Treatment of Legg-Calvé-Perthes Disease with a Traction Ischial Non-weight Bearing Brace.

Between 1945 and 1955 there h ve been thirty-one cases of Legg-Calv -Perthes disease which h.ve been treated through the definitive stage, and which have been followed carefully by roentgen studies at the Nebraska Orthopedic Hospital at Lincoln, Nebraska. These cases were initially seen in children between the ages of 2.5 to 12 years with an average age of 7.5 years. Twenty-two cases were unilateral, fourteen involving the left hip and eight involving the right hip, while nine cases

contracted bilateral involvement with the disease. When first seen nir ety per cent of the patients com- plained of limping and seventy-four percent complained of having pain, either in the affected hip, or in the knee on the affected side. Nearly all of these cases initially exhibited limitation of abduction and limitation of internal as well as of external rotation. The Wasserman test was negative in all of these cases.

In 1948, the bracemaker at the hospital, Mr. Walter Jackel, conceived the idea of using a traction bar on the ischial non-we ght bearing braces. This bar was attached to the shoe on the foot of the affected side and is tightened by means of a lateral ratchet. Similar traction bars have been used in fracture cases to prevent musclar spasm.

Ferguson(33), Pike(32), and Howorth(40) have suggested that muscle spasm about the hip decreases healing in Legg-Calvé-Perthes disease, and Pike states that this muscle spasm probably decreases circulation. If the muscle spasm were reduced, then circulation should be enhanced and healing should be promoted. The traction ischial non-weight bearing brace is devised to reduce muscle spasm.

The unilateral cases at the Nebraska Orthopedic Hospital were all treated by immobilization in a cast

for one to seven months, and then the patients were fitted with an ischial non-weight bearing brace, crutches, and with elevation of the shoe on the foot of the non diseased side. Since 1948 the traction brace has been used.

X-rays were taken monthly until evidence of healing was seen. Then the cases were followed on roentgenograms which were taken every three to six months until the definitive stage had been reached. I have been able to review eighteen of these twenty-two unilateral cases. The definitive x-rays on four of the cases were taken elsewhere and were not available. Since the patients were admitted to the hospital at many different phases of the active stage, I have used the duration of time between when evidence of healing was first noted on the x-rays and when the definitive stage was described to compare cases where the traction brace was used with those where the usual ischial non-weight bearing brace had been used.

The average time for healing of eight cases with the usual ischial non-weight bearing brace was 37;5 months, while the average time in using the traction brace was 22.5 months. This also compares favorably with the series of cases which were treated by Levy and Girard(31) with the usual ischial non-weight bearing brace where

thirty months was the average time when healing became definitive.

I have also carefully measured the hips on the defi .itive x-ray ccording to the excellent method proposed by Heyman and Herndon(49). The average of the epiphseal quotient, the head-neck quotient, the acetabular quotient, and the acetabular-head quotient establishes the overall comprehensive quotient which is an excellent index for evaluating the end results of the treatment. The immediate results by clinical evaluation may appear to be excellent, but on x-ray the damage which has occurred will be revealed, and the end results in later years may be foreseen. A comprehensive quotient above 90 indicates an excellent to a normal result· between 80-90, good; between 70-80, fair; bet veen 60-70, poor; and below 60, very poor to a bad result.

Most of the comprehensive quotients in this series of cases revealed a fair to a good result. This is readily explainable as so many of the cases were seen so late in the active stages that irreparable damage has already occurred. More excellent results may be obtained if the patients can be seen in the incipient or very early stage of the disease.

TABLE COMPARING TREATMENT METHODS

Usual Case Non-Weight Bearing Brace		Age at Onset	Healing Time	Comprehensive Quotient	Immediate Clinical Result*
	1	6	49 mos.	80	---
	2	7	35 "	81	--
	3	7	38 "	91	--
		7	62 "	91	--
	5	9	30 "	68	---
	6	7	18 "	70	---
		8	32	85	--
		7	36	79	-
Tract-	9	6	26 mos.	74	--
ion	10	9	30 "	86	---
Ischial	11	9	28 "	64	---
Non-	12	6.5	16	83	--
Weight	13	8	29 "	69	--
Bearing	14	8	17 "	6	--
Br ce	15	7	35 "	71	---
	16	6.5	16 "	75	---
	17	8	25 "	77	---
	18	8	23 "	73	---

*

--- Full Motion

-- Sight Loss of Motion

- Moderate Loss of Motion

This table represents eighteen unilateral cases which have been followed to the definitive stage between 1945 to 1955 at the Nebraska Ortho-pedic Hospital at Linco n, Nebraska.

IX. Summary

Legg-Calvé-Perthes disease was differentiated from tuberculous coxitis in 1910 by three men independently including an Ame.ican, a Frenchman, and a German whose names were iven in that order for the disease by Halfdan Sundt. The discovery of the roentgen ray in 1895 made possible the acknowledgment of Legg-Calvé-Perthes disease.

Legg's theory for the etiology of the disease is still accepted by most of the writers today. Legg thought that trauma associated with an inadequate blood supply to the hip joint was the causative factor. Many ideas including mycotic emboli of blood vessels within the epiphysis, trauma during embryological development, minute subluxations, caused by dietary factors, infection, hypothyroidism, or a constitutional anomaly have been proposed as etiological factors. None of these have been proved.

The disease has been found to be familial in some instances. It has been suggested that Legg-Calvé-Perthes disease might represent a familial predisposition to an inherent hip weakness or a familial endocrine change. Ninety percent of the cases are found in boys, and ninety percent are unilateral. The patient complains

of mild pain in the affected hip or in the knee on the affected side, or he demonstrates a limp on the affected side, sometimes without pain. Hip motions often show a limitation of internal rotation, external rotation, and abduction. There is a definite history of trauma in many of the cases.

Early aids for dia nosis of this disease are (1) increase in the separation of the head of the femur from the medial wall of the acetabulum; (2) the obturator sign; (3) the "roof sign"; and (4) the measurement of bone atrophy on the involved side with a photoelectric light meter. These aids have been proposed by Kite and French(24) in order to aid the clinician in making an early correct diagnosis.

The progression of the disease is shown on roentgenograms. This is described in the following stages.

1. Incipient stage of development

This stage is mani ested by a synovitis on the x-ray.

2. Early Active Stage

During this stage there is an increase in the density of the capital emoral epiphysis, an apparent increase in the jo nt space, and an osteoporosis of the metaphysis. It lasts two to three months.

3. Late Active Stage

During this stage there is compression of the epiphysis, beginning of fragmentation, furthur com-pression and thinning of the fragmentary epiphysis, fading or beginning absorption of the dense portions, and expansion of the metaphysis with widening of the femoral neck. This stare lasts usually from three to eighteen months.

4. The Healing Stage

At this time there is a beginning regeneration of the epiphyseal zone, absorption has been completed, areas of rarefied reoss fied bone are appearing, and there is reossification by creeping substitution". This stage usually lasts from fourteen to forty-eight months.

5. The Definitive St ge

At this sta e the epiphysis has reossified and normal bone trabeculations are present. It usually occurs during or after t e fourth year following the onset of the disease.

Gross and microscopic studies of material obtained at autopsy from operation, or by biopsy of the femoral head reveal aseptic necrosis. Cultures of staphalo-coccus aureus have been obtained in some cases, but most investigators believe that it was present due to con-tamination. There have been no consistent significant changes in laboratory tests.

Many methods of treatment have been used including complete bed rest, Wu-splints, the Thomas ring caliper brace, an ischial non-weight bearing brace with elevation of the opposite shoe, a Synder sling, crutches, casting, and traction with Buck's extension. The practical treatment by choice by most clinicians seems to be the ischial non-weight bearing brace with elevation of the opposite shoe.

The end results of Legg-Calvé-Perthes disease without early and adequate treatment may be appalling. Traumatic arthritis may occur at any age. Muscular contractures and severe limitation of motion of the involved joint may occur also. However with an early diag-nosis and proper treatment, a completely normal hip joint may be the expected result.

In a survey of eighteen unilateral completely healed cases between 1945 and 1955 at the Nebraska Orthopedic Hospital at Lincoln, Nebraska, I compared the length of time taken for the healing stage in eight cases where the usual ischial non-weight bearing braces were used with the length of time taken for healing in ten cases

where traction ischial non-weight bearing braces were used. The traction bar is applied to the shoe on the affected side. This is to decrease the musclar spasm about the affected hip joint which should enhance the

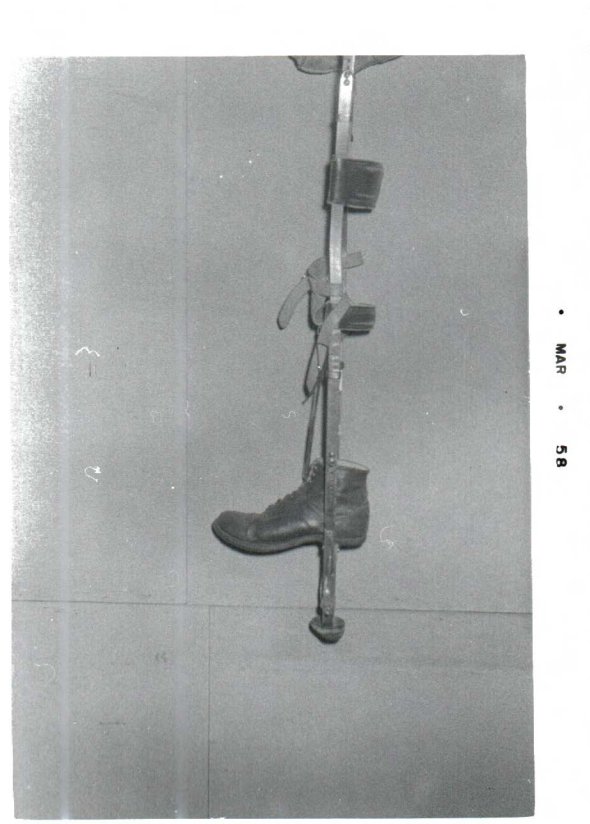
blood supply to the area, and time for healin should be decseased. Int ese ten ceses the average heeling time was 22. 5 months while in the eisht ceses where the usual ischial non-weight bearin$_u$ brace were used, the aver a e healing tirr e we.s 37. 5 months. This also compares favorable to a large series which were treated b-Levy and G1rard(31) wbere the aver ge healing time was reported as being 30 months.

X.Conclusion

Since 1910 wren Legg-Calve-Perthes disease was first described we have rot improved much on Legg's theory of etiology and idea of treatment. Various methods have now been de ised uhich enable the affected child to be ambulatory. This permits him to continue his education, develop more normally socially, and often to remain in his home en ironment. This decreases his becoming an invalil while simultaneously he may heal physically with hope for an excellent result.

The traction ischial non-weight bearing brace which has been described has decreased the heeling time by six to twelve months in ten unilateral cases as compared to eight unilateral cases where the usual ischial non-weight bearing brace was used. From this result in this small series I think that the traction ischial non-weight bearing brace warrants further use to establish

whether or not it will significantly shorten healing time in a large number of cases.



The Traction Ischial Non-Weight Bearing Brace

Bibliography

1. Legg, A. T.: An Obscure Affection of the Hip Joint. Boston Med. and Surg. J., 162:202-204, 1910.

2. Calvé, Jacques: Osteochondritis of the Upper Extremity of the Femur. J. Orthop. Surg., 3:489-493, 1921.

3. Calvé, Jacques: Pathogenesis of the Limp due to Coxalgia.. Bone and Joint Surg., 21:12-25, 1939.

4. Goff, C. W.: Legg-Calvé-Perthes Syndrome and Related Osteochondroses of Youth. Charles C. Thomas. Springfiel , Illinois, 1954.

5. Waldenstrom, Henning: The First Stage of Coxa Plana. J. Bone and Joint Surg., 20:559-566, (July) 1938.

6. Gill, A. B.: Relationship of Legg-Perthes Disease to the Function of the Thyroid Gland. J. Bone and Joint Surg., 25:892-901, (Oct) 1943.

7. Jansen, Murk: On Coxa Plana and its Causation. J. Bone and Joint Surg., 5:265-277, 1923.

8. Zemansky, A. P., Jr.; and Lippmann, R. K.: The Importance of the Vessels in the Round Ligment to the Head of the Femur During the Period of Growth, and Their Possible Relationahip to Perthes Disease. Surg., Gyne. and Obst., 48:461-469, 1929.

9. Ponseti, I. V.; and Shepard, R. S.: Lesions of the Skeleton and of Other Mesodermal Tissues in Rats Fed Sweet-Pea (Lathyrus odaratus) J. Bone and Joint Surg., 36:A:1031-1058, (Oct) 1954.

10. Ponseti, I. V.: Legg-Perthes Disease, Observation on Pathological Changes in Two Cases. J. Bone and Joint Surg., 38-A-2:739-50, (July) 1956.

11. Ponseti, I. V.; and McClintock, Robert: The Pathology of Slipping of the Upper Femoral Epiphysis. J. Bone and Joint Surg., 38-A:71-83, (Jan) 1956.

12. Phemister, D. B.: Perthes Disease. Surg., Gyne, and Obst., 33:87, 1921.

13. Cavanaugh, L. A.; Shelton, E. K.; and Sutherland, Ross:

13. (cont) Metabolic Studies in Osteochondritis of the Capital Femoral Epiphysis. J. Bone and Joint Surg., 18:957-968, (Oct) 1936.

14. Emerick, R. W.; Corrigan, K. E.; Joistad, A. H. Jr.; and Holly, L. E.: Thyroid Function in Legg-Calve-Perthes Disease. Clinical Orthopedics, No. 4:160-5, 1954.

15. Katz, J. F.: Protein Bound Iodine in Legg-Calvé-Perthes Disease. J. Bone and Joint Surg., Am. Vol. 37-A:842-6, (July) 1955.

16. Beiler, D. D.; and Love, W. H.: Thyroid Function in Legg-Perthes Disease. Journal of Bone and Joint Surg., Am. Vol, 1320-2, (Dec) 1956.

17. Grannestras, Nicholas: Legg-Perthes Disease. J. Bone and Joint Surg., 36-A-1:149-52, (Jan) 1954.

18. Burrows, H. J.: Coxa Plana With Special Reference to its Pathology and Kinship. Brit. J. of Surg., 29:23-46, 1941.

19. Derian, P. S.· Coxa Plana in Dizygotic Male Twins. J. Bone and Joint Surg., 38-A-2:901-7, (July) 1956.

20. Hamsa, W. R.; and Campbell, L. S.: Osteochrondritis Deformans Coxa Juvenilis. Am. J. Dis. Child, 86: 54-59,1953.

21. Detalia, Francesco: A Typical Disease of the Femur. Am. J. Orth. Surg., 12:555-588, (April) 1915.

22. Fawcitt, John: The Radiological Pattern of Pseudo-coxalgia. Brit. J. Radiology, 504-510, (Sept) 1954.

23. Gill, A. B.: Legg-Perthes Disease of the Hip, Its Early Roentgenographic Manifestations and Its Clinical Course. J. Bone and Joint Surg., 22: 1013-1047, (Oct) 1940.

24. Kite, J. H.; and French, G. O.: The Early Diagnosis of Flat Headed Femur. Southern Medical Journal, 45:581-5, (July) 1952.

25. Blanchard, Wallace: Treated and Untreated Osteochondritis Juvenilis of the Hip. J.A.M.A., 69:1060, 1917.

26. Eyre-Brook, A. L.: Osteochondritis Deformans Coxae Juvenilis or Perthes Disease. The Results of Treatment by Traction in Recumbency. Brit. J. Surg., 24·166-182, (July) 1936.

27. Ferguson, A. B., Sr.: Early Roentgenographic Changes in Perthes Disease. Clinical Orthopedics, No. 1:33-42, 1953.

28. Bettman, E. H.; and Siffert, R. S.: Roentgen Examination of Hip in Legg-Perthes Disease. Radiology, 53:548-551, (Oct) 1949.

29. Billing, Lars: Viewpoints on Roentgen Examination of Coxa Plana. Acta Radiological Supplementum, No. 110:66-71, 1954.

30. Lewis, R. W.: Correlation of Roentgen and Pathologic Findings in Perthes Disease. Radiology, 22:188, 1934.

31. Levy, L. J.; and Girard, P. M.: Legg-Perthes Disease, A Comparative Study of Various Methods of Treatment. J. Bone and Joint Surg., 24:663-671, (July) 1942.

32. Pike, M. M.: Legg-Perthes Disease. Connecticut M. J., 4:5-11, 1940.

33. Ferguson, A. ., Jr.: Synovitis of the Hip and Legg-Perthes Disease. Clinical Orthopedics, No. 4:180-8, 1954.

34. Legg, A. T.: Osteochondral Trophopathy of the Hip Joint. Surg. Gyne. and Obst., 22:307-323, 1916.

35. Danforth, M. S.: The Treatment of Legg-Calvé-Perthes Disease without Weight Bearing. J. Bone and Joint Surg., 16:516-534, (July) 1934.

36. Bozsan, E. J.: A New Treatment of Intracapsular Fractures o the Neck of the femur and Legg-Calvé-Perthes Disease. J. Bone and Joint Surg., 14:884-887, (Oct) 1932.

37. Bozsan, E. J.: A New Treatment of Intracapsular Fractures of the Neck of the Femur and Legg-Calvé-Perthes Disease, Technique. J. Bone and Joint Surg., 16:75, (Jan) 1934.

38. Herndon, C. H ; and Heyman, C. H.: Legg-Perthes Disease, an Evaluation of Treatment by Traction and Ischial Weight-Bearing Brace. J. Bone and Joint Surg., 24-A:25, 1952.

39. Pedersen, H. E.; and Mc Carrol, H. R.: Treatment in Legg-Perthes Disease. J. Bone and Joint Surg., 33-A, 3:591-600, 1951.

40. Pike, M. M.: Legg-Perthes Disease. A Method of Conservative Treatment. J. Bone and Joint Surg., 32-A:663-370, 1950.

41. Pike, M. M.: Legg-Perthes Disease with Particular Regard to Treatment. Conn. State Med. J., 4:5-11, 1940.

42. Goff, C. W.: Growth Acceleration in Legg-Calvé-Perthes Disease Syndrome by Complementary Feedings of Aureomycin. Clinical Orthopedics, Philadelphia.

No. 6:95-109, 1955.

43. Siewers, C. F.: The Diagnosis of Legg-Perthes Disease. North Carolina Med. J., 14:11, 560-4,(Nov) 1953.

44. Steele, P. B.: Further Report on Operative Treatment of Perthes Disease. Instr. Course Lecture, Am. Academy of Orthopedic Surg., 136-143, 1943.

45. Legg, A. R.: End Results of Coxa Plana. J. Bone and Joint Surg., 9:26-33, 1927.

46. Mindell, E. R.; and Sherman, M. S.: Late Results in Legg-Perthes Disease, J. Bone and Joint Surg., 33-A:1-23, (Jan) 1951.

47. Caldwell, G. A.: End Results of Coxa Plana as Related to Treatment. South M. J., 27:402-207, 1943.

48. Ratliff, A. H.: Pseudocoxalgia; A Study of Late Results in the Adult. J. Bone and Joint Surg., 38-B: 498-512, (May) 1956.

49. Heyman, C. H.; and Herndon, C. H.: Legg-Perthes Disease, A Method for the Measurement of the Roentgenographic Result. J. Bone and Joint Surg., 32-A:767-778, (Oct) 1950.