50 Years Ago in CORR

Legg-Calvé-Perthes syndrome (LCPS): An up-to-date critical review

Charles W. Goff, MD CORR 1962;22:93–107

Three independent seminal papers published in 1911 by Arthur Legg [9, 10], Jacques Calvé [3, 4], and Georg Perthes [12, 13] described what we now know as “Legg-Calvé-Perthes disease” (LCPD) and distinguished the entity from other forms of a deforming arthritis that affected children. At that time, arthritis deformans (as it was frequently known) was sometimes not recognized until adolescence and young adulthood. Others [1, 2, 11, 14] had earlier described the entity but had not so clearly distinguished it from other disorders,

particularly tuberculosis of the hip. (The earliest apparent descriptions by Baker [2] and Wright [14] appear as brief notes for the “Section of Diseases of Children” in the British Medical Journal.) By the mid 20th century, the entity was well known.

Charles Weir Goff, in 1962, published in CORR a comprehensive review of the literature with 97 references, dating back to the earliest reference [6]. Goff summarized the then-known population genetics of LCPD. Today, researchers have found a variety of collagen-related genes that

appear to play a role in the development of LCPD [5, 7, 8]. Initial recumbency, plus traction or splinting (by cast or brace) were the preferred methods of treatment at the time. Some surgeons recommended drilling or osteotomy to hasten healing, but Goff noted, “no controlled series (had) been reported...” and if he seemed skeptical he said, “The trend is toward early surgical simulation or revascularization of the upper femur.” The value of Goff’s review was its comprehensiveness and the tables (eg, Tables 1 and 4) that summarized relevant papers on

Table 1

Investigators	Date of Publication	Primary Causes and Pathologic State
Piatt	1922	Mild infectious arthritis
Legg	1927	Trauma
Sundt	1927	Mild, innocuous infectious arthritis
Phemister	1930	Bland emboli, plus an injury cutting off circulation to the epiphysial bone, plus a constitutional factor
Masselli	1934	Trauma, infection, toxemia, local vascular disturbance, endocrine dysfunction. LCPS and congenital coxa vara were varieties of the same basic condition.
Camitz	1934	Satellite of congenital dislocated hip (CDH).
Kisler	1934	Experimentally injected charcoal dust into nutrient arteries of rabbits produced avascular necrosis similar to LCPS.
Gall & Bennett	1942	Generalized disorder not localized to upper femur—renal rickets of subclinical variety
Howorth	1948	Infectious origin with a primary synovitis followed by an avascular necrosis
Tucker	1949	Absence of ligamentum teres blood supply until 6 years of age or older demonstrated definitely
Erlacher	1950	Primary dysplasia of the hip-joint satellite of CDH
Broder	1953	Trauma, 20%, plus other causes

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Table 1 continued

Investigators	Date of Publication	Primary Causes and Pathologic State
Helbo	1953	Trauma, 20%, plus genetic labile joint
Trueta & Harrison	1953	Proposed absence of blood supply through ligamentum teres vessels explained the age incidence of LCPS.
Ferguson, Jr	1954	Soft-tissue origin, beginning with an acute synovitis of the hip
Paltrinieri	1954	Fatigue fracture of epiphysal bone
Ponseti	1954	Experimental feeding of aminonitriles to rats produced disruption of epiphysal cartilage with LCPS-like changes.
Emerick, Corrigan, Joistad & Holly	1954	Hyperfunctional thyroid adenomas produced a hypothyroidal state primary to LCPS.
Goff	1954	Complex, multiple causes, acting on a genetically conditioned individual. Multiple provocative agents, hence affix “syndrome.” Deceleration of growth prior to onset, trauma, general infectious diseases, endocrine imbalance, faulty metabolism or faulty nutrition and hereditary dysplastic derangements (upper femur & hip joint). A juvenile osteochondrosis or enchondral dystosis, sex influenced, racially conditioned, time of growth oriented; a MULTI-SYSTEM DISEASE affecting the hip joint, the epiphysis and the metaphysis less often as the ligamentum teres vessels appear
Golding	1956	Sickle-cell anemia in Negroes
Fox & Griffin	1956	Two of 23 transient synovitis hips developed LCPS.
Slavik	1956	Multiple causes acting on a satellite of a subclinical congenital dysplasia of the hip joint (originally proposed by Maydl, 1897, and advanced by Murk Jansen, 1913)
Ponseti	1956	Two biopsy studies of LCPS showing disruption of the plate and the articular cartilage, a loss of cohesion of the cartilage matrix, ground substance, a chemical alteration similar to that produced in rat studies
Ryder et al.	1957	Trauma, 25%
Papastrovos	1957	Caused frequently by sickle-cell thalassemia
Lemoine	1957	Cut arterial vessels in experimental rabbits to upper femur, producing epiphysal change similar to LCPS. Developed a time-table of reossification and by implication only, established rate in children with LCPS
Canestri & Monzali	1957	Do dysplastic disorders exist from birth and later give rise to LCPS—a satellite of subclinical CDH?
Salvo Legarre	1957	Early synovitis in series of 400 LCPS

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Table 1 continued

Investigators	Date of Publication	Primary Causes and Pathologic State
Hulth	1958	Any extracoxal disturbance of arterial blood supply and venous blood flow. Experimental rabbits fed papain showed disruption of cartilage, ground substance, LCPS like.
Portych	1958	Interference in upper femoral circulation
Mau	1958	Complex causal factors, an enchondral dystosis, effected by mechanical endogenous and exogenous factors, with dwarfism or deceleration of growth generally present. Metaphysial zone frequently affected, together with epiphysis
Howorth	1959	Not caused by infection, cultures sterile
Trueta, Pinto de Lima	1959	<ol style="list-style-type: none"> 1. Early—to about 4 years of age no ligamentum teres vessels develop, but lateral epiphysial and cartilage penetrating vessels from metaphysial branches are present. 2. Middle—4 to 8 years of age, when lateral epiphysial vessels alone nourish epiphysis 3. Ligamentum teres vessels penetrate to epiphysis after 8 years of age. Peak age incidence from 4 to 8 years of age
Laurent	1959	General deceleration of growth with shortening of neck in severe cases. Believes accompanying overgrowth of trochanter produces a coxa vara.
Spock	1959	Acute transient synovitis a precursor in 6% of cases; others abort. Obesity a precipitating cause in 20%, rarely in Negroes. Trauma and allergy causal in 30%, yet he does not consider them important. Respiratory infections in 70% preceded disturbance.
Yamoguchi	1959	Disruption of growth of epiphysial and articular cartilage plus metaphysial zone
Durbin	1959	May follow undisplaced cervical fracture of femur
Schuster	1959	May follow lateral fractures of hip, 1% of 800 children. Degree of circulatory development prime causal factor. Older children nearly always develop a coxa plana with accompanying acetabular changes, plus an early osteoarthritis of hip joint.
Katz	1959	Trauma, 25%
Sherman	1959	Sickle-cell anemia produced disintegration of hip joint in Negroes.
Pylkkänen	1960	No common cause or pathology between LCPS and coxa vara biopsy studies
Lima, Esteve & Trueta	1960	In a CDH study found same 3 stages of avascular necrosis as in LCPS: 65% slight, 25% greater and 8% extensive, as in LCPS

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Table 1 continued

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Salter & Field	1960	Continuous compression of living cartilage under forced positioning produced a range of necrosis from slight to great, depending on duration. Even growth may be arrested.
Bozdech & Bozdechova	1960	Sickle-cell anemia may produce bony infarcts.
Jacobs	1960	Three of 25 cases of acute synovitis of the hip developed LCPS.
Greenlaw, Kohn & Petrie	1960	A loop tied round the femoral neck of experimental rabbits produced an avascular necrosis, as in LCPS.
Nogai	1960	No increase in the incidence of LCPS in children exposed to the effects of the atomic bomb in Nagasaki or in children born to mothers who had suffered such exposure
Bobechko & Harris	1960	Experimentally produced avascular necrosis of femoral heads of rabbits showed increased x-ray density, while both the dead and the living (reossifying) bone were present. If no reossification was taking place, there was no change in x-ray pattern or density. They do not believe in “creeping substitution.”

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Table 4

Investigators	Date of Publication	Form of Treatment
Danforth	1934	Recumbency, 12 or more months with traction in a Thomas splint or spica
Waldenström	1938	Recumbency, a few months
Gill	1940	Recumbency, 12 or more months
Howorth	1948	Drilling the epiphysis shortens recovery.
Mendell & Sherman	1951	Bed rest or ambulation with sling. Saw no difference in end-result study.
Jonsäter	1953	Recumbency, but results not the best. Recommends drilling across epiphysal plate into head to shorten recovery time
Broder	1953	Bed rest, 24 months or less, plus crutch-walking. Subtrochanteric osteotomy in 10 cases plus ischial weight-bearing brace. Tried several varieties of management

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Table 4 continued

Investigators	Date of Publication	Form of Treatment
Helbo	1953	Bed rest or cast, 12 to 24 months, 80% good results. Prefers bed rest for 15 to 18 months, plus crutches for 3 to 6 months. No surgery was tried.
Larcomb, Perry & Peterman	1954	B ₁₂ additive to stimulate statural growth
Emerick, Corrigan Joistad & Holley	1954	Surgical removal of thyroidal adenomas, plus recumbency and oral thyroid substance
Ferguson, Jr	1954	Traction with bed rest
Paltrinieri	1954	Abduction casts or braces; never surgery
Giannestras	1954	Hip plaster spica for long recumbency
Novo Monteiro	1954	Bed rest plus plaster spica; physical exercises when cast is removed
Goff	1954	Iron-frame recumbency, average 27 months, plus crutch-walking, NO SLING plus aureomycin, 50 mg., daily. No deleterious effects. Stimulated growth and hastened reossification
Peter	1955	Early cases: nail driven from subtrochanteric area through neck and plate and into head of femur. Method also used by Pitzen, Hauberg and others. Immobilize in brace and walking splint. Grafting and drilling on occasion. Abduction spica preferred. Time for recovery, 9.7 months average. Ischial weight-bearing brace tried
Goff	1955	Abduction frame average recumbency reduced to 19 months plus daily 50 mg. of aureomycin, plus high caloric diet. Crutch-walking 6 months. Accelerated recovery by 8 months
Slavik	1956	Derotation femoral osteotomy shortens recovery by 1 year. Crutch-walking 3 to 6 months
Fevre & LeGrange	1956	Recumbency until reossification observed on roentgen plate. Long periods, time not stated
Ratleff	1956	Recumbent frame, average 13 months, plus crutches and sling to follow
Katz	1957	Bed rest with bilateral traction, 18.3 months average, plus crutches and sling to follow

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Table 4 continued

Investigators	Date of Publication	Form of Treatment
Prochazka	1957	Bone grafts of cancellous bone placed in epiphysial bony nucleus, plus spica. Hastens reossification by many months, time not given
Scapaticci & Caccarini	1957	Administer tetracycline, 100 mg. daily, for 30 days for weight and general health gain
Ryder et al.	1957	Bed rest "few" months to 2 years, plus crutches. Drilling (Howorth manner) but "benefits not proven." Crutches plus sling in some cases. Prefer bed rest for best results
Masse	1957	Recumbency in plaster spica 18 to 36 months
Camargo	1957	Large plug cut from neck and reversed, then driven up to and sometimes across epiphysial plate into head. Performed in early stage of LCPS, followed by bed rest for 10 months. Crutches
Petrocini & Bullio	1958	Stimulated growth in stature and weight by administering 4 chlorotestosterone acetate; no side effects
Evans	1958	Bed rest plus simple leg traction or plaster spica, or bed rest alone, or drilling and grafting into neck and head. Recumbent 2 to 24 months. Today prefers only sling and crutches on outpatient basis
Mau	1958	Plaster abduction spica for many months in some cases. Average not stated
Glimcher	1958	Patent bottom or hip traction, ischial weight-bearing brace reduces compressive forces on femoral head by 10% compared with full weight-bearing.
Bette	1959	1. Spica immobilization, internal rotation for many months, plus ischial weight-bearing splint and crutches. 2. Drilling or grafting or nailing across plate into head, plus cast for 2 to 3 months and crutch-walking. Prefers the latter method
Goff	1959	Recumbency in abduction frame, average reduced to 9 months, plus crutch-walking 6 months, plus additive of 50 mg. achromycin daily for 18 months
Yamaguchi	1959	Removed plug of bone and reversed it as a bone graft across plate into head in early stages
Wagner	1959	Same method
Gueritee, Ducroquet & Ducroquet	1959	In severe cases use a daily dose of lyophilized thyroid extract, 0.5 to 0.2 Gm., plus testosterone base and estradiol, plus recumbency

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Table 4 continued

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Koskinen	1959	Evidence from experimental animals suggests use of human growth hormone and thyrotropin daily to accelerate reossification. He believes that an endocrine imbalance exists.
Stamp, Canales & Odell	1959	Originally used bed rest plus traction, then crutch-walking on elevated shoe. Now use bed rest plus traction, average 17.7 months, plus limited weight-bearing with sling, average 11.2 months. Bilateral cases, bed rest of 56 months. Early weight-bearing produces flattening?
Torto	1959	Crutch-walking with special traction brace designed by Marino-Zucco
Fields	1959	Daily doses of tri-iodo-thyronine produced earlier physical maturation and an earlier recovery.
Braadt	1960	Recumbency, hip spica with knee bent to exert traction in unilateral cases and quickly permitting weight-bearing without crutches for 10 months average. Results not reported for comparative analysis
Carpenter & Powell	1960	Recumbency plus crutches with or without sling. Good results, but slow to obtain
Williamson	1960	Suggests daily doses of gonadal steroids for more rapid reossification
Koenig & Heller	1960	In experimental animals administration of a sterile bone extract accelerated reossification in fractures with necrosis (interesting correlation).
Lima, Esteve & Trueta	1960	No weight-bearing allowed “during the evolution of the osteochondritis”??
Jacobs	1960	Early, prolonged recumbency
Inglis	1960	Recumbency in spica, average 19 months, plus crutch-walking
Craig, Kramer Luque & Watanabe	1961	Osteotomy, abduction type or a shelf overhead to prolong acetabulum and buttress femur, or selected drilling and abduction splints
Ponseti & Cotton	1961	Tri-iodothyronine administrations had no beneficial effects. Plaster abduction cast for recumbency, plus crutches with or without sling for as long as necessary? Time average not reported

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each issue; the reader readily captures the array of treatments that were advocated at that time.

Since Goff's time, many researchers have identified risk factors (age at onset, "at risk" radiographic signs) that influence repair, subsequent growth, and development of the hip. Because the condition is self-limited (in the sense that the necrotic regions of the bone repair albeit sometimes with deformity), and most patients with the milder forms of the disease do well without surgery (at least for many decades), earlier nonoperative treatments, such as prolonged recumbency and bracing, have largely been abandoned. Nonetheless, treatment remains controversial. Most agree that children with symptoms (pain and limited motion) should be treated acutely by short-term bedrest and traction until the symptoms are relieved. Today, surgery during childhood is typically reserved for older children (over age 8) with more severe forms of the disease and hips at risk for deformities and subsequent osteoarthritis. Since if untreated these children not infrequently require reconstructive surgery as young adults owing to early onset osteoarthritis, the goal of surgery is generally to prevent or minimize the ultimate deformity. As in Goff's day, we still lack controlled trials of the

surgical options, and it remains unclear whether or to what degree these procedures delay the development of subsequent osteoarthritis and the need for reconstruction.

Richard A. Brand MD
Editor-in-Chief
Clinical Orthopaedics
and Related Research

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