Femoral head necrosis in juvenile chronic arthritis

Masahiro Kobayakawa[1*], Urban Rydholm[1], Hans Wingstrand [1], Holger Pettersson[2] and Lars Lidgren[1]

Radiographic signs of ischemic necrosis of the femoral head in terms of abnormal epiphysis and caput indices were found in 30/72 hips in 36 children with hip pain or limitation of motion among 206 children, consecutively admitted during 15 months because of juvenile chronic arthritis. Nine out of 10 hips with obvious signs of femoral head necrosis showed a sclerotic rim at the base of the femoral neck, confirming an earlier episode of ischemic damage to the epiphysis and growth plate.

Femoral head necrosis in children with juvenile chronic arthritis seems to be more common than previously reported, and may be caused by circulatory disturbance secondary to increased intraarticular pressure due to synovitis and/or effusion, or to treatment with passive extension of the hip. This should be considered in the treatment and the follow-up of these children.

Hip joint involvement is a major cause of disability in children with juvenile chronic arthritis (JCA), and is relatively common in those with seronegative, unremitting polyarthritis and an early onset of disease (Ansell and Unlu 1970, Isdale 1970). Common clinical findings are flexion and adduction contracture, often combined with internal rotation of the hip.

Early radiographic findings are osteopenia and growth disturbance of the proximal femur and the acetabulum, which in young children may result in coxa magna with a short, wide femoral neck. In older children, however, premature closure of the femoral and acetabular growth plates may result in retarded growth with a hypoplastic femoral neck and a dysplastic acetabulum. Growth disturbance may also give rise to increased valgus and anteversion of the femoral neck. Later on, narrowing and irregularities of the joint space – as well as bony erosions, subchondral cysts, and acetabular protrusion – become evident (Gallino et al. 1984; Patriquin et al. 1984, Resnick and Niwayama 1981). Lateral subluxation of the femoral head, probably caused by intense synovitis, soft-tissue contractures, or thickening of the cartilage by hypermetabolism (Gershuni and Kuei 1984), is not unusual and may sometimes result in lateral flattening of the femoral head (Patriquin et al. 1984).

Necrosis of the femoral head has been reported to appear in patients with JCA treated with corticosteroids (Cole and Neal 1976, Gallino et al. 1984, Resnick and Niwayama 1981). In transient synovitis and hemophilia, increased intracapsular pressure may cause tamponade of the nutritional vessels to the femoral head (Patriquin et al. 1984, Wingstrand et al. 1985). Elevated intracapsular pressures, possibly to harmful levels, have recently been demonstrated in children with chronic coxitis (Rydholm et al. 1986).

The purpose of this study was to estimate the frequency of radiographic signs of femoral head necrosis in a consecutive series of children with JCA and clinical signs of hip involvement.

Patients

During 15 months in 1982 and 1983, 206 children were treated as inpatients at the JCA Center of the University Hospital in Lund. Sixteen boys and 20 girls had a radiographic examination of the hips performed because of pain or restricted motion. The radiographs included

Departments of Orthopedics[1] and Diagnostic Radiology[2], University Hospital in Lund, Sweden

*Present address: Department of Orthopedics, Fukoroi shimin Hospital, Kuno 2515-1, Fukoroi city, Shizuoka ken, Japan

Correspondence: Dr. Urban Rydholm, Department of Orthopedics, University Hospital, S-221 85 Lund, Sweden

in every case an anteroposterior and a frog-leg view. Twenty-six children had polyarticular disease and 10 had oligoarticular disease (including 1 child with psoriatic arthritis and 1 with juvenile ankylosing spondylitis). The mean age at onset of the disease was 5 (1–12) years, and at onset of coxitis 7 (1–16) years.

Twenty-four children had been or were on oral corticosteroids at the time of radiographic examination. All of these children had bilateral coxitis, often with one hip symptomatic at the time.

Methods

The radiographs were examined and classified into three groups depending upon the appearance of the femoral head (Figures 1–3).

Group 1: *Obvious* radiographic signs of necrosis. Hips with an appearance similar to Perthes' disease, i.e., condensation, fragmentation, and/or resorption in the epiphysis.

Group 2: Suspect late radiographic signs of necrosis. Hips with flattening and widening of the femoral head similar to the reparative end stage of Perthes' disease.

Group 3: No radiographic signs of necrosis. Hips with the femoral head spherical in both projections, including hips with radiographic findings typical of JCA – i.e., narrowing and irregularities of the joint space, erosions, subchondral cysts, and acetabular protrusion–were classified as normal in this context.

The shape of the femoral head was defined by measurements with a digitizer connected to a microcomputer. The *epiphysis index* and the *caput index* were calculated according to Heyman and Herndon (1950) and Jonsäter (1953). The epiphysis index is the relation between the height of the epiphysis and the width of the growth plate (Figure 4), and indicates any abnormal proportion of the epiphysis. The caput index is the relation between the height of the femoral head and the radius of its greatest width (Figure 4), and thus expresses the sphericity of the femoral head. Finally, the presence of a sclerotic rim at the base of the femoral neck ("sagging-rope"; Apley and Wientroub 1981), most likely caused by previous ischemic damage to the growth plate and epiphysis, was recorded.

The extension deficit as given in the medical records at the time of the radiographic examination was recorded.

Results

Obvious radiographic signs of necrosis of the femoral head were found in 10 hips in 6 children with a mean age of 3 (1–6) years at the onset of the disease. Suspect

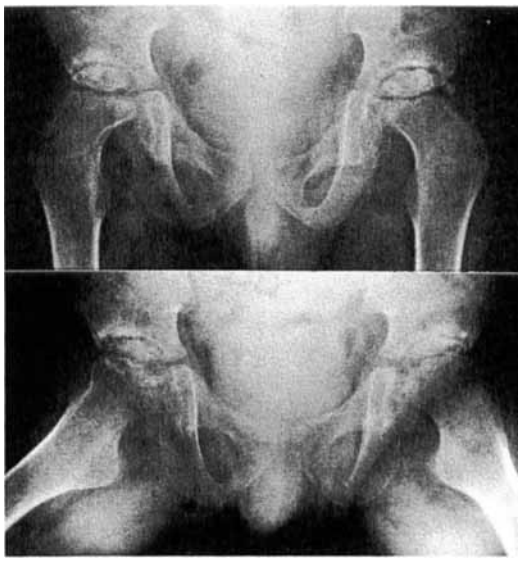

Figure 1. Group 1. Case 5. A 7-year-old boy with obvious early bilateral signs of ischemic necrosis of the proximal femoral epiphysis resembling Legg-Calvé-Perthes disease, i.e., sclerosis, flattening, and subchondral fractures (AP and frog-leg views).

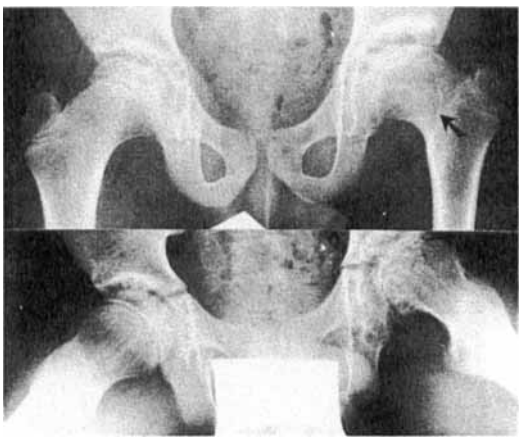

Figure 2. Group 2. Case 7. A 12-year-old girl with suspect, late changes of ischemic necrosis of the left hip, i.e., flattening of the epiphysis, widening of the metaphysis, and a sclerotic rim ("sagging rope") at the base of the femoral neck (arrow).

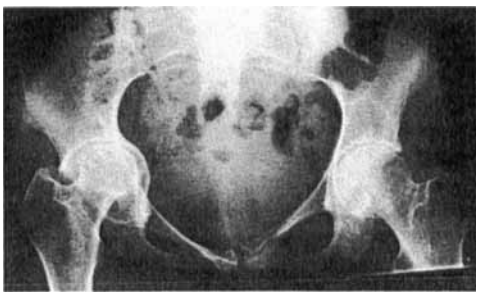

Figure 3. Group 3. Case 35. A 20-year-old girl without signs of previous ischemic necrosis of the epiphysis, although with other radiographic changes typical of the JCA hip.

radiographic signs were found in 20 hips in 13 children aged 4 (2–11) years, and no radiographic signs of necrosis in 42 hips in 23 children aged 7 (2–13) years at the onset of the disease. The mean age at the onset of hip pain or reduced mobility was 3, 6, and 9 years, respectively. The mean duration of coxitis at the time of the first accessible radiographic examination was 8 (4–12) years in Group 1, 8 (2–12) years in Group 2, and 6 (0–16) years in Group 3.

All the hips in Group 1 were painful and had an extension deficit; at the time of radiographic examination, it was 31° (20–40°). Eighteen out of 20 hips in children with suspected necrosis were painful, and the extension deficit in this group was 17° (0–35°). Twenty-four out of 42 hips with normal radiographs had pain, and the extension deficit in this group was 3° (0–5°).

All the children with obvious unilateral or bilateral signs of necrosis were or had been on long-term (> 3 months) treatment with corticosteroids, whereas this was the case in 8 out of 11 children with suspected signs of necrosis in one or both hips, and in 10 out of 18 children without signs of necrosis in either hip.

The results of the radiographic measurements expressed as epiphyseal and caput indices and the relationship between these indices and the radiographic classification are shown in Figure 4. The epiphyseal indices were close to 0.5 in Group 3, but lower in Groups 1 and 2. The caput indices were close to 1.0 in most hips in Group 3, but lower in most hips in Groups 1 and 2. A sclerotic rim in the femoral neck was evident in 9 out of 10 hips in Group 1, 2 out of 20 hips in Group 2, and 1 out of 42 hips in Group 3.

Discussion

The frequency of hip involvement in JCA is at least 20 percent (Martel et al. 1962, Mogensen 1982). The frequency of hip symptoms, obvious enough to lead to radiographic examination was 17 percent in our series. It is probable that radiographic changes may exist in children without clinical signs or symptoms and that our figures should thus be considered as minimum values.

Many children with polyarthritis develop progressive, destructive changes with subsequent need for total hip replacement at an early age (Ruddlesdin et al. 1986). Such destructive changes as growth disturbance of the proximal femur and acetabulum, lateral subluxation of the femoral head, erosions, joint space narrowing, and acetabular protrusion are all well known and seem to be correlated with the age at onset and type of disease, and have a worse prognosis in young children with polyarticular disease that has

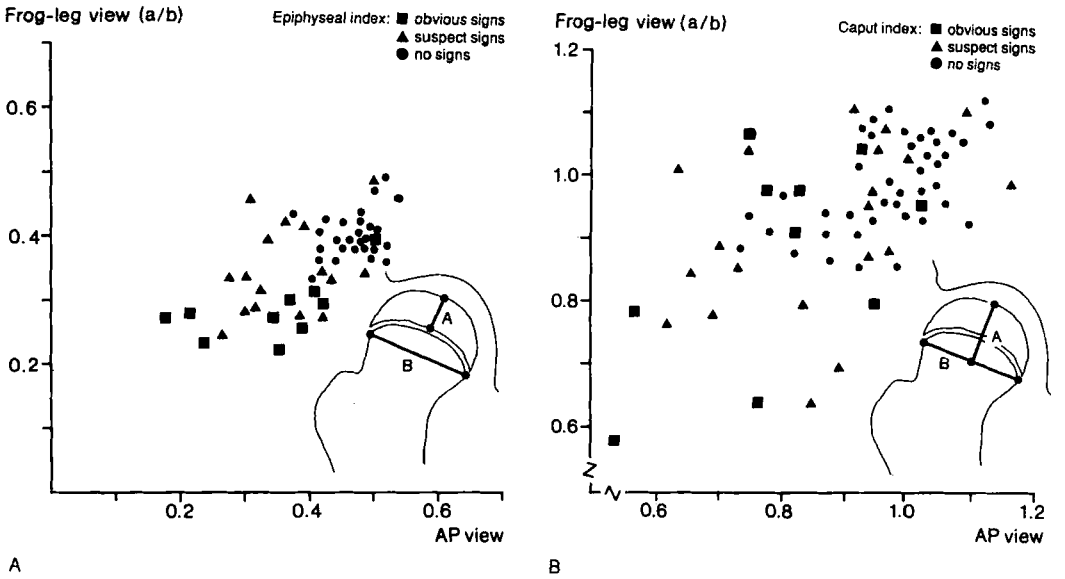

Figure 4. Radiographic indices of femoral head deformation in patients with radiographic signs of ischemic necrosis of the proximal femoral epiphysis in juvenile chronic arthritis.

A. Epiphysis index (A/B).

B. Caput index (A/B).

Table 1. Clinical data for 36 children with juvenile arthritis

A	B	C	D	E	F	G	H	I	J
1	M	6	8	12	2	1	O	+	20
2	F	1	2	8	1	2	P	+	40
3	M	3	3	12	1	2	P	+	40
4	M	4	4	16	1	1	P	+	40
5	M	1	1	7	1	1	P	+	20
6	F	1	1	13	1	1	P	+	25
7	F	4	4	12	3	3	P	+	25
8	M	1	1	13	2	2	P	+	35
9	F	5	5	14	2	2	P	+	20
10	F	3	3	18	2	2	P	+	15
11	F	3	13	15	2	2	P	+	10
12	F	2	2	12	2	2	P	+	25
13	M	11	11	15	2	2	O	+	20
14	F	2	6	13	2	2	P	+	10
15	F	3	3	13	2	3	P	+	30
16	M	2	5	8	3	2	O	–	0
17	M	2	8	14	3	2	P	–	0
18	M	2	3	18	3	3	P	–	15
19	F	6	6	22	3	3	P	–	10
20	F	9	10	16	3	3	J	+	
21	F	9	10	18	3	3	P	+	15
22	F	8	10	13	3	3	P	–	0
23	F	8	16	18	3	3	P	–	0
24	F	8	8	14	3	3	O	+	15
25	F	2	2	17	3	3	P	+	0
26	M	3	13	15	3	3	P	+	0
27	M	7	8	14	3	3	P	+	0
28	M	5	5	5	3	3	O	+	0
29	M	6	6	22	3	3	O	+	0
30	M	10	14	16	3	3	P	–	0
31	M	5	8	11	3	3	P	+	0
32	F	7	7	19	3	3	P	+	0
33	F	3	7	19	3	3	O	+	0
34	M	8	9	9	3	3	P	–	0
35	F	12	14	20	3	3	Ps	–	0
36	M	10	13	14	3	3	O	–	0

A Case.
B Sex.
C Age (yr) at the time of onset of the disease.
D Age (yr) at the time of onset of hip symptoms.
E Age (yr) at the time of first hip radiograph.
F Signs of epiphyseal necrosis right hip; 1 Obvious necrosis, 2 Suspect necrosis, 3 No signs of necrosis.
G Left hip as in F.
H Type of disease: P: Polyarticular; O: Oligoarticular; Ps: Psoriasis; J: Juvenile ankylosing spondylitis.
I Hip pain at the time of examination.
J Hip extension deficit (°).

been persistently active over several year (Ansell and Unlu 1970, Gallino et al. 1984, Isdale 1970, Patriquin et al. 1984). Ischemic necrosis of the femoral head is much less frequently reported. Because we found obvious or suspect radiographic signs of this complication in 30 out of 72 hips in children with JCA, and who had been radiographically examined owing to hip pain, we believe it is important to focus interest on this phenomenon and to discuss its possible etiology.

The radiographic appearance of ischemic necrosis in Group 1 is very similar to that observed in idiopathic Legg-Calvé-Perthes disease in its early stages, with signs of condensation, fragmentation, resorption, and flattening of the epiphysis, as we found in the youngest children in this study. Group 2 changes resembling Legg-Calvé-Perthes disease in later stages, with a disproportion of the epiphysis and widening of the metaphysis, were found in older children. A sclerotic rim at the base of the femoral neck indicating a period of ischemia with subsequent injury to the growth plate, the superior layers of which are also supplied via branches from intraarticular vessels (Trueta and Amato 1960), was also more common in the older children. This sclerotic rim, initially described in Legg-Calvé-Perthes disease (Apley and Wientroub 1981), has recently also been reported in 9 patients with JCA (Patriquin et al. 1984), and it is strong circumstantial evidence of an earlier ischemic damage of the growth plate and epiphysis.

It has generally been accepted that the cause of Legg-Calvé-Perthes disease is an interruption of the blood supply to the proximal femoral epiphysis. The vascular supply of the epiphysis varies with age, as clarified by Trueta (1957) and Ogden (1974). Before closure of the growth plate, the epiphysis is supplied mainly by lateral intracapsular branches from the medial circumflex artery with no contribution across the growth plate and a very small contribution via vessels in the ligamentum teres. The vessels, being located intracapsularly, should theoretically be vulnerable to an increase in the intracapsular pressure, as has been shown in puppies (Lucht et al. 1983, Woodhouse 1964) and clinically in septic arthritis and intracapsular fractures (Minikel et al. 1983, Wingstrand et al. 1986), as well as in transient synovitis of the hip (Kloiber et al. 1983, Wingstrand et al. 1985). In patients symptomatic before closure of the growth plate, an intracapsular hematoma or effusion with subsequent increase in the intracapsular pressure may thus compromise the blood flow through the proximal femoral epiphysis, as has been demonstrated with ^{99}Tc-MDP-scintigraphy (Wingstrand et al. 1985). In children with JCA, synovitis of the hip may result in an increase in the intracapsular pressure, as reported recently by Rydholm et al. (1986). The magnitude of the pressures recorded (mean 11 kPa in extension and inward rotation) in these children was in many cases well above the pressure in the venous drainage vessels from the epiphysis and would thus decrease the pressure gradient across the capillary bed, possibly to a level where the metabolism of the epiphyseal cells is impaired. Synovitis, with subsequent increase in the intracapsular pressure tamponading the blood supply of the proximal femoral epiphysis, may be a causative mechanism. This is supported by experimental and clinical findings of femoral head ischemia in other conditions with an in-

creased intracapsular pressure, such as transient synovitis of the hip, septic arthritis, posttraumatic hematoma, or hemophilic bleeding (Ahlberg 1965, Minikel et al. 1983, Wingstrand et al. 1985, Wingstrand et al. 1986).

Symptoms of hip synovitis in JCA are often intermittent. The children have often been treated with recommendations of rest in the prone position aiming at passive extension of the hip. The intraarticular pressure of the hip is highly dependent on the position of the hip, with a sharp increase in extension and inward rotation, whereas flexion causes a pronounced decrease of the pressure and reduced pain (Wingstrand et al. 1985). It is likely that this mechanism is, at least partly, responsible for the the common flexion contracture of the hip. These children would probably benefit from decompression of the hip, i.e., tenotomy of the iliopsoas and adductor tendons (Swann and Ansell 1986) and anterior capsulotomy, especially prior to extension treatment. Traction applied for flexion contracture may cause ischemia of the femoral head if the intraarticular pressure is not released.

Sonography gives information on the presence of joint effusion or synovitis (Rydholm et al. 1986), and should, if possible, be performed prior to traction. Apart from providing information on the stage of synovitis and cartilage destruction (Holgersson et al. 1981), arthroscopy, with joint lavage and possibly administration of intraarticular corticosteroids, may be used to decompress the joint before beginning physical therapy, including traction and extension. Severe hypertrophic synovial inflammation found at arthroscopy may also be an indication for synovectomy (Mogensen et al. 1982) to reduce the intracapsular pressure.

Necrosis of the femoral head in JCA was described by Cole and Neal (1976) who attributed it to treatment with systemic corticosteroids. We found long-term treatment with systemic corticosteroids to be more common in the children with radiographic signs of necrosis than in those without such signs. The mean age at onset of disease was also considerably lower for both children with obvious and with suspect signs of necrosis than for the children without such signs. Obviously, in younger children the disease has a more destructive course, and these children are thus more likely to receive treatment with corticosteroids earlier in life and for a longer period. Necrosis of the femoral head following corticosteroid treatment is a well-known phenomenon in adults (Fischer and Bickel 1971, Pietrograndi and Mastromarine 1957), and a similar complication in children cannot be ruled out.

References

Ansell B M, Unlu M. Hip involvement in juvenile chronic polyarthritis. Ann Rheum Dis 1970;29(6):687–8.

Ahlberg Å. Haemophilia in Sweden. VII. Incidence, treatment and prophylaxis of arthropathy and other musculoskeletal manifestations of haemophilia A and B. Acta Orthop Scand 1965:(Suppl 77):3–132.

Apley A G, Wientroub S. The sagging rope sign in Perthes' disease and allied disorders. J Bone Joint Surg (Br) 1981;63(1):43–7.

Cole W G, Neal B W. Corticosteroids and avascular necrosis of the femoral head in childhood. Aust Paediatr J 1976; 12(1):37–42.

Fisher D E, Bickel W H. Corticosteroid induced avascular necrosis. A clinical study of seventy-seven patients. J Bone Joint Surg (Am) 1971;53(5):859–73.

Gallino L, Pountain G, Mitchell N, Ansell B M. Developmental aspects of the hip in juvenile chronic arthritis. A radiological assessment. Scand J Rheumatol 1984;13(4):310–8.

Gershuni D H, Kuei S C. Articular cartilage deformation following experimental synovitis in the rabbit hip. J Orthop Res 1984;1(3):313–8.

Heyman C H, Herndon C H. Legg-Perthes' disease. A method for the measurement of the roentgenographic result. J Bone Joint Surg (Am) 1950;32:767–78.

Holgersson S, Brattström H, Mogensen B, Lidgren L. Arthroscopy of the hip in juvenile chronic arthritis. J Pediatr Orthop 1981;1(3):273–8.

Isdale I C. Hip disease in juvenile rheumatoid arthritis. Ann Rheum Dis 1970;29(6):603–8.

Jonsäter S. Coxa plana. A histo-pathologic and arthrographic study. Acta Orthop Scand 1953;24(Suppl 12).

Kloiber R, Pavlosky W, Portner O, Gartke K. Bone scintigraphy of hip joint effusions in children. AJR 1983;140(5): 995–9.

Lucht U, Bünger C, Krebs B, Hjermind J, Bülow J. Blood flow in the juvenile hip in relation to changes of the intraarticular pressure. An experimental investigation in dogs. Acta Orthop Scand 1983;54(2):182–7.

Martel W, Holt J F, Cassidy J T. Roentgenologic manifestations of juvenile rheumatoid arthritis. Am J Roentgenol 1962;88:400–23.

Minikel J, Sty J, Simons G. Sequential radionuclide bone im-

aging in avascular pediatric hip conditions. Clin Orthop 1983;(175):202–8.

Mogensen B. Hip surgery in juvenile chronic arthritis. Thesis, University of Lund, Lund, Sweden 1982.

Mogensen B, Brattström H, Ekelund L, Svantesson H, Lidgren L. Synovectomy of the hip in juvenile chronic arthritis. J Bone Joint Surg (Br) 1982;64(3):295–9.

Ogden J A. Changing patterns of proximal femoral vascularity. J Bone Joint Surg (Am) 1974;56(5):941–50.

Patriquin H B, Camerlain M, Trias A. Late sequelae of juvenile rheumatoid arthritis of the hip: a follow-up study into adulthood. Pediatr Radiol 1984;14(3):151–7.

Pietrogrande V, Mastromarino R C. Osteopatia da prolongato trattamento cortisonico. Ortop Traum Appar Mot 1957;25:791–810.

Resnick D, Niwayama G. Diagnosis of bone and joint disorders. W.B. Saunders Co, Philadelphia 1981.

Ruddlesdin C, Ansell B M, Arden G P, Swann M. Total hip replacement in children with juvenile chronic arthritis. J Bone Joint Surg (Br) 1986;68(2):218–22.

Rydholm U, Wingstrand H, Egund N, Elborg R, Forsberg L, Lidgren L. Sonography, arthroscopy, and intracapsular pressure in juvenile chronic arthritis of the hip. Acta Orthop Scand 1986;57(4):295–8.

Swann M, Ansell B M. Soft tissue release of the hips in children with juvenile chronic arthritis. J Bone Joint Surg (Br) 1986;68(3):404–8.

Trueta J. The normal vascular anatomy of the human femoral head during growth. J Bone Joint Surg (Br) 1957;39: 358–94.

Trueta J, Amato V P. The vascular contribution to osteogenesis. III. Changes in the growth cartilage caused by experimentally induced ischemia. J Bone Joint Surg (Br) 1960;42:571–87.

Wingstrand H, Bauer G C, Brismar J, Carlin N O, Pettersson H, Sundén G. Transient ischaemia of the proximal femoral epiphysis in the child. Interpretation of bone scintimetry for diagnosis in hip pain. Acta Orthop Scand 1985;56(3):197–203.

Wingstrand H, Egund N, Carlin N O, Forsberg L, Gustafson T, Sundén G. Intracapsular pressure in transient synovitis of the hip. Acta Orthop Scand 1985;56(3):204–10.

Wingstrand H, Strömqvist B, Egund N, Gustafson T, Nilsson L T, Thorngren K G. Hemarthrosis in undisplaced cervical fractures. Tamponade may cause reversible femoral head ischemia. Acta Orthop Scand 1986;57(4):305–8.

Woodhouse C F. Dynamic influences of vascular occlusion affecting the development of avascular necrosis of the femoral head. Clin Orthop 1964;(32):119–29.

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