Acta Orthop. Belg., 2009, **75**, 754-760

ORIGINAL STUDY

Clinical presentation of young adults after Legg-Calvé-Perthes disease

Christoph ZILKENS, Bernd BITTERSOHL, Marcus JÄGER, Tanja HAAMBERG, Bettina WESTHOFF, Rüdiger KRAUSPE

From Heinrich-Heine University Medical School, Duesseldorf, Germany

The purpose of the present study was to review the clinical and radiological outcome of young adult individuals who had Legg-Calvé-Perthes disease (LCPD) during child age. After a mean follow-up of 18.7 ± 6.2 years, 25 young adults with various morphological deformity grades were assessed clinically and radiologically. Deterioration of hip joint function and onset of pain were found to manifest early in the intermediate follow-up of LCPD and to correlate with the residual hip deformity.

Keywords : Legg-Calvé-Perthes disease ; osteoarthritis ; Tegner-Lysholm score ; outcome.

INTRODUCTION

Osteoarthritis (OA) of the hip joint is a common sequel of Legg-Calvé-Perthes disease (LCPD). Less joint deformity at skeletal maturity and young age at the onset of the disease are well established predictors for a favorable outcome (*1,3,4,8,9,12,13,15, 16,20,22*). However, several authors have stated that patients do well in early adult age even with a high grade of hip joint deformity (*4,10,12*). These reports are mainly based on clinical and functional results following various methods of treatment up to the age of 40 years. In contrast to these favourable clinical results, radiographic signs of OA are often seen, even in early adulthood. Clarke and Harrison (*2*) noted an incidence of 30% of severe OA at an average age of 27 years. Saito *et al* (*15*) found that 23% of 51 patients had hip OA after a follow-up of 18 years.

The purpose of this study was to review the clinical and radiographic outcome of young LCPD patients and to perform comparative analyses between clinical outcomes and standard radiographic findings in both LCPD hips and the unaffected hips of the same individuals, with no signs of former LCPD or other deformity. We further analyzed the medical history of asymptomatic young adult volunteers in order to outline significant differences between pain, hip joint function, and activity.

PATIENTS AND METHODS

Institutional review board approval and informed consent of all participating individuals were obtained prior to the start of this retrospective study.

■ Christoph Zilkens, MD, Resident.
■ Bernd Bittersohl, MD, Resident.
■ Marcus Jäger, MD/PhD, Attending Orthopaedic Surgeon.
■ Tanja Haamberg, MD, Resident.
■ Bettina Westhoff, MD, Attending Orthopaedic Surgeon.
■ Rüdiger Krauspe, MD, Professor and Chief of the Department.
Department of Orthopaedics, Heinrich-Heine University Medical School, Duesseldorf, Germany.
Correspondence : Christoph Zilkens, MD, Department of Orthopaedics, Heinrich-Heine University Medical School, Moorenstrasse 5, 40225 Duesseldorf, Germany.
E-mail : Christoph.Zilkens@med.uni-duesseldorf.de

This study was supported in part by a grant of the "German Osteoarthritis Aid" ("Deutsche Arthrosehilfe e.V.")

Study population

The study cohort consisted of 25 individuals (16 males, 9 females) who were treated for LCPD in childhood. Individuals were identified through the database of the national LCPD support group. Currently, the LCPD support group includes 805 members, with 95 of them older than 18 years. Twenty-five individuals, who met the inclusion criteria (age 18-35 years, complete medical history, no other hip diseases), agreed to participate in the study. LCPD diagnosis was based on clinical evaluation and standard radiographic assessment. Mean age at diagnosis was 6.3 ± 1.6 years (range : 3.7-9.3). Mean follow-up after diagnosis was 18.7 ± 6.2 years (range : 10.0-28.7). Mean age at the follow-up visit was 25.0 ± 5.3 years (range : 18.0-35.0). Individuals younger than 18 years and older than 35 years were excluded from the study. Further exclusion criteria were any other hip disorders. Both hips were affected in eight individuals. Thus, a total of 50 hips (33 LCPD hips, 17 unaffected contra-lateral hips) were analyzed. As control group for subjective hip joint status and individual activity level, ten healthy and asymptomatic individuals volunteered in the medical history observation.

Clinical assessment

Hip function and clinical symptoms were evaluated with the Harris Hip Score (HHS) (*5*). On physical examination, the range of motion (ROM) of the hip joint in extension, flexion, abduction, adduction, internal and external rotation in 90° of flexion was assessed. In addition, the activity level of each individual was graded according to the Tegner-Lysholm classification which is provided in detail in table I (*18*).

Plain radiographic evaluation

A standard anteroposterior pelvic radiograph and a lateral Lauenstein radiograph were obtained for each LCPD case. Radiographic analysis included the Stulberg classification (*16*) and the osteoarthritis (OA) grading according to the Tönnis OA evaluation system (*19*). The study cohort was further sub-classified based on the Stulberg classification. In order to maximize the power of statistical analysis, we combined Stulberg classes I + II (group 1) and Stulberg classes III + IV (group 2) in order to increase the sample sizes for analysis. Group 3 consisted of Stulberg V cases while group 4 included the non-affected, morphologically normal appearing contralateral hips. Radiographic analyses and subsequent classification were performed by two senior and experienced orthopaedic surgeons (MJ and RK) in consensus, who were blinded to medical history and clinical findings.

Statistical analysis

SPSS® software (Version 16.0, SPSS Inc., Chicago, IL, USA) was used for statistical analysis. Continuous data were expressed as mean and standard deviations (SD). The Spearman's rho test was used to determine if correlation between Stulberg and Tönnis classes and between Tönnis and age was present. In order to reveal statistically significant differences between the various groups of LCPD hips (group 1-3) and the control group (group 4), Student's t-test (parametric variables, independent samples) or Wilcoxon's rank sum test (non-parametric variables, exact significance, two-sided) was applied. P-values below 0.05 were considered to be statistically significant.

RESULTS

Overall 25 LCPD patients with 33 affected hips and 17 morphologically normal appearing unaffected contra-lateral hips were studied. A total of 10 hip joints were classified as group 1 (Stulberg I or II), 16 as group 2 (Stulberg III or IV) and 7 as group 3 (Stulberg V). Group 4 consisted of the 17 normal hips. Ten hips revealed no radiographic signs of OA (Tönnis grade 0), while Tönnis grade 1 changes were noted in six hips, Tönnis grade 2 changes in 14 hips and Tönnis grade 3 in 3 hips. There was a high correlation ($r = 0.934$) between the modified Stulberg classification system and the Tönnis grading, which was statistically significant ($p < 0.001$). No correlation was found between Tönnis grade and age ($r = -0.146$, p-value = 0.374).

Except for hip joint extension we noted a trend of decreasing ROM for the various groups, which was consistent with the morphological grade of joint deformity (table I). In group 1 a significant difference was noted between the affected hips and the control group only in terms of a decreased external rotation ($p = 0.002$) while in group 2 there was a significant decrease in hip flexion ($p = 0.008$), abduction ($p = 0.001$), and external rotation ($p < 0.001$). In group 3, all values for ROM except for extension ($p = 0.266$) and adduction ($p = 0.078$)

Table I. — Tegner-Lysholm activity level score

Level	Description
0	Sick leave or disability pension
1	Work : sedentary Walking on even ground possible
2	Work : Light labour Walking on uneven ground possible but impossible to walk in forest
3	Work : light labour (e.g. nursing) Competitive and recreational sports : swimming Walking in forest possible
4	Work : moderately heavy labour (e.g. truck driving, heavy domestic work) Recreational sports : cycling, cross-country skiing Recreational sports : jogging on uneven ground at least twice weekly
5	Work : heavy labour (e.g. building, forestry) Competitive sports : cycling, cross-country skiing Recreational sports : jogging on uneven ground at least twice weekly
6	Recreational sports : tennis and badminton, handball, basketball, downhill skiing, jogging (at least five times per week)
7	Competitive Sports : tennis, athletics (running), motocross/speedway, handball, basketball Recreational sports : soccer, bandy and ice hockey, squash, athletics (jumping) Cross-country track findings both recreational and competitive
8	Competitive sports : bandy, squash or badminton, athletics (jumping etc), downhill skiing
9	Competitive sports : soccer (lower divisions), ice hockey, wrestling, gymnastics
10	Competitive sports : soccer (national and international elite)

were significantly decreased with p-values ranging from $p < 0.001$ to $p = 0.008$.

In terms of HHS analysis we used the classification for pain and function only, because ROM was assessed in detail and the HHS for deformity was not relevant in this study cohort. When both sides were affected ($n = 8$) we used the side with more deformity for comparison. Thus, seven hips in group 1, 11 hips in group 2, seven hips in group 3 and ten hips in the control group were compared for HHSs and Tegner-Lysholm grades.

Mean values and range for HHS and Tegner-Lysholm grades are given in tables II and III. Significant differences were noted (p-values ranging from < 0.001 to 0.013) for the HHS for pain and function in all grades of deformity (group 1-3) and for the Tegner-Lysholm grades in group 3 ($p = 0.015$).

DISCUSSION

Residual hip joint deformity after LCPD may cause joint degeneration and premature OA. Many efforts were made to define clinical and radiographic characteristics, prognostic factors, treatment and follow-up outcome *(1,6-8,12,13,15,16,21)*. However, detailed reports on the clinical outcome and activity level at mid-term follow-up of LCPD are rare. Since LCPD patients likely present with a limited ROM and treatment outcome is also measured upon hip joint function, this knowledge may be of clinical interest. Therefore, the aim of the present study was to assess the clinical outcome at mid-term follow-up (18.7 ± 6.2 years) in young adult individuals with different morphological deformity grades after LCPD in childhood. ROM, HHS for pain and function and activity level according to the Tegner

Table II. — Comparison of range of motion (ROM) between LCPD patients with different grades of hip joint deformity (group 1 = Stulberg class 1 + 2 ; group 2 = Stulberg class 3 + 4 ; group 3 = Stulberg class 5) and asymptomatic morphologically normal appearing contralateral hip joints (group 4). Note : EXT = extension, FLEX = flexion, ABD = abduction, ADD = adduction, IRO = internal rotation in 90° of hip flexion, ERO = external rotation in 90° of hip flexion, SD = standard deviation

		Group 1 (10 hips)	Group 4 (17 hips)	p-value
EXT	(°) (mean ± SD)	12.0 ± 4.2	14.7 ± 4.2	0.116
FLEX	(°) (mean ± SD)	109.5 ± 7.6	111.8 ± 5.8	0.393
ABD	(°) (mean ± SD)	31.5 ± 6.7	33.8 ± 6.0	0.360
ADD	(°) (mean ± SD)	18.2 ± 4.5	16.2 ± 5.2	0.313
IRO	(°) (mean ± SD)	37.5 ± 8.9	37.9 ± 10.9	0.915
ERO	(°) (mean ± SD)	31.0 ± 8.4	41.8 ± 7.1	0.002**
		Group 2 (16 hips)	Group 4 (17 hips)	p-value
EXT	(°) (mean ± SD)	14.1 ± 3.3	14.7 ± 4.2	0.625
FLEX	(°) (mean ± SD)	104.0 ± 9.5	111.8 ± 5.8	0.008**
ABD	(°) (mean ± SD)	26.9 ± 4.8	33.8 ± 6.0	0.001**
ADD	(°) (mean ± SD)	14.1 ± 4.2	16.2 ± 5.2	0.207
IRO	(°) (mean ± SD)	33.4 ± 14.5	37.9 ± 10.9	0.318
ERO	(°) (mean ± SD)	27.8 ± 6.3	41.8 ± 7.1	< 0.001**
		Group 3 (7 hips)	Group 4 (17 hips)	p-value
EXT	(°) (mean ± SD)	13.7 ± 3.9	14.7 ± 4.2	0.266
FLEX	(°) (mean ± SD)	107.2 ± 9.6	111.8 ± 5.8	0.007**
ABD	(°) (mean ± SD)	29.5 ± 7.1	33.8 ± 6.0	< 0.001**
ADD	(°) (mean ± SD)	15.3 ± 4.9	16.2 ± 5.2	0.078
IRO	(°) (mean ± SD)	22.1 ± 14.4	37.9 ± 10.9	0.008**
ERO	(°) (mean ± SD)	29.3 ± 9.6	41.8 ± 7.1	0.002**

Lysholm classification were compared for different grades of joint deformity. Regardless of clinical or radiographic findings, young adults were included who were not drawn from the clinic but were drafted through the database of the national LCPD support group. Therefore, a high range of clinical and radiographic findings could be analyzed.

Based on our findings, we conclude that the clinical outcome does depend on the degree of morphological deformity, which is in line with previously reported studies (*3,8,9,12,15,16,20,22*) and contrary to others (*4,10-12*). In our study cohort of young adults the ROM decreased with higher grades of deformity, starting with a decrease in external rotation followed by a decrease in flexion and abduction in group 2 while all ROM values were significantly decreased in the group with severe deformity (group 3) except for extension and adduction.

Comparative analysis of HHSs for pain and function for various deformity grades revealed a statistically significant difference in all LCPD groups. This knowledge points towards the conclusion that individuals even in their early ages (range : 18 years to 35 years) and even hips with less deformity are

Table III. — Comparison of HHS for pain and function and Tegner-Lysholm activity levels between the different LCPD groups and asymptomatic young-adult volunteers (pts = points)

		Group 1 (n = 7)	Volunteers (n = 10)	p-value
HHS Pain	(pts) (mean ± SD)	40.0 ± 8.9	44.0 ± 0.0	0.013*
HHS Function	(pts) (mean ± SD)	44.3 ± 4.9	47.0 ± 0.0	< 0.001**
		Group 2 (n = 11)	Volunteers (n = 10)	p-value
HHS Pain	(pts) (mean ± SD)	32.6 ± 8.5	44.0 ± 0.0	< 0.001**
HHS Function	(pts) (mean ± SD)	43.8 ± 7.6	47.0 ± 0.0	< 0.001**
		Group 3 (n = 7)	Volunteers (n = 10)	p-value
HHS Pain	(pts) (mean ± SD)	28.6 ± 12.2	44.0 ± 0.0	< 0.001**
HHS Function	(pts) (mean ± SD)	37.9 ± 8.9	47.0 ± 0.0	< 0.001**
		Group 1 (7 hips)	Volunteers (n = 10)	p-value
Tegner-Lysholm	(pts) (mean)	4.38	4.60	0.410
		Group 2 (11 hips)	Volunteers (n = 10)	p-value
Tegner-Lysholm	(pts) (mean)	4.06	4.60	0.165
		Group 3 (7 hips)	Volunteers (n = 10)	p-value
Tegner-Lysholm	(pts) (mean)	3.50	4.60	0.015*

significantly impaired in the intermediate follow-up. This is to some extent consistent with the study of Pécasse *et al* who performed 15 intertrochanteric osteotomies in 14 symptomatic LCPD patients and analyzed the clinical outcome at an average follow-up of 11.3 years (*14*). A mean value of the total HHS of 78.5 was noted and the ROM in these patients was still limited with a mean hip flexion of 95°, extension of -1°, abduction of 15° and adduction of 14°. However, this study consisted only of a small study cohort and focused on the follow-up of one therapeutic procedure.

Using the Tegner-Lysholm classification system, there was a statistically significant difference only for the group with severe morphological deformity (group 3). One explanation for this may be the relatively high weighing of regularly performed competitive sports in this system, which we believe does not truly reflect the activity level distribution of the normal active young adults. On the other hand, slight but remarkable changes might have been underestimated. Thus, similar activity grades within the LCPD groups (group 1-3) and within normal asymptomatic volunteers (group 4) were noted.

As a function of time and deformity, LCPD can cause premature OA if appropriate intervention is not performed (*2,7,15-17*). Aids to diagnosis of OA include a detailed medical history, physical exami-

nation, plain radiographs and magnetic resonance imaging (MRI).

Hip joint preserving procedures such as pelvic osteotomies or arthroscopic or open osteochondroplasties are more likely to be effective in the absence of severe joint damage.

The search for both clinical and radiological diagnostic markers that would identify early joint degeneration as a sequel of LCPD is warranted. We explored in this present study the clinical outcome and subjective problem staging in different grades of hip deformity, which may be of clinical interest.

This study has limitations. Our study population was relatively small. Second, the included individuals were collected from the LCPD-support group regardless of their current symptoms and LCPD treatment in childhood. Thus, we analyzed an inhomogeneous group of LCPD patients. However, the clinical findings of our study reflect those of previously reported studies. In addition, the amount of radiographic OA correlated well with the degree of deformity while there was no correlation noted between OA and age, which is a well known finding with respect to sequels of LCPD. Therefore, we believe that our data reflect those of a larger LCPD cohort. Furthermore, by including individuals irrespective of their symptoms, we were able to correlate a large variety of clinical outcomes with various grades of hip joint deformity. One major limitation was the absence of a diagnostic gold standard such as intra-operative correlation. Thus, hip joint damage that could not be recognized with standard radiography may have biased the comparison analysis.

In conclusion, this study showed that deterioration of hip joint function and onset of pain manifest early in the intermediate follow-up of LCPD.

Acknowledgements

We thank Mr. Wolfgang Strömich from the German LCPD support group for the good cooperation ("Deutsche Morbus Perthes Initiative DMPI").

REFERENCES

1. **Catterall A.** Legg-Calve-Perthes disease. *Instr Course Lect* 1989 ; 38 : 297-303.
2. **Clarke NM, Harrison MH.** Painful sequelae of coxa plana. *J Bone Joint Surg* 1983 ; 65-A : 13-18.
3. **Engelhardt P.** [Late prognosis of Perthes' disease : which factors determine arthritis risk ?] (in German). *Z Orthop Ihre Grenzgeb* 1985 ; 123 : 168-181.
4. **Gower WE, Johnston RC.** Legg-Perthes disease. Long-term follow-up of thirty-six patients. *J Bone Joint Surg* 1971 ; 53-A : 759-768.
5. **Harris WH.** Traumatic arthritis of the hip after dislocation and acetabular fractures : treatment by mold arthroplasty. An end-result study using a new method of result evaluation. *J Bone Joint Surg* 1969 ; 51-A : 737-755.
6. **Herring JA, Kim HT, Browne R.** Legg-Calve-Perthes disease. Part I : Classification of radiographs with use of the modified lateral pillar and Stulberg classifications. *J Bone Joint Surg* 2004 ; 86-A : 2103-2120.
7. **Herring JA, Kim HT, Browne R.** Legg-Calve-Perthes disease. Part II : Prospective multicenter study of the effect of treatment on outcome. *J Bone Joint Surg* 2004 ; 86-A : 2121-2134.
8. **Ippolito E, Tudisco C, Farsetti P.** The long-term prognosis of unilateral Perthes' disease. *J Bone Joint Surg* 1987 ; 69-B : 243-250.
9. **Lecuire F.** The long-term outcome of primary osteochondritis of the hip (Legg-Calve-Perthes' disease). *J Bone Joint Surg* 2002 ; 84-B : 636-640.
10. **Lloyd-Roberts GC.** The management of Perthes' disease. *J Bone Joint Surg* 1982 ; 64-B : 1-2.
11. **Lloyd-Roberts GC, Catterall A, Salamon PB.** A controlled study of the indications for and the results of femoral osteotomy in Perthes' disease. *J Bone Joint Surg* 1976 ; 58-B : 31-36.
12. **McAndrew MP, Weinstein SL.** A long-term follow-up of Legg-Calve-Perthes disease. *J Bone Joint Surg Am* 1984 ; 66 : 860-869.
13. **Mose K, Hjorth L, Ulfeldt M *et al.*** Legg Calve Perthes disease. The late occurence of coxarthrosis. *Acta Orthop Scand Suppl* 1977 ; 169 : 1-39.
14. **Pecasse GA, Eijer H, Haverkamp D, Marti RK.** Intertrochanteric osteotomy in young adults for sequelae of Legg-Calve-Perthes' disease – a long term follow-up. *Int Orthop* 2004 ; 28 : 44-47.
15. **Saito S, Takaoka K, Ono K *et al.*** Residual deformities related to arthrotic change after Perthes' disease. A long-term follow-up of fifty-one cases. *Arch Orthop Trauma Surg* 1985 ; 104 : 7-14.
16. **Stulberg SD, Cooperman DR, Wallensten R.** The natural history of Legg-Calve-Perthes disease. *J Bone Joint Surg* 1981 ; 63-A : 1095-1108.
17. **Stulberg SD, Cordell LD, Harris WH *et al.*** Unrecognized childhood hip disease : a major cause of idiopathic osteoarthritis of the hip. *Proceedings of the Third Open Scientific Meeting of the Hip Society.* CV Mosby, Saint-Louis, 1975 : 212-228.

18. Tegner Y, Lysholm J. Rating systems in the evaluation of knee ligament injuries. *Clin Orthop Relat Res* 1985 ; 198 : 43-49.

19. Tonnis D. Normal values of the hip joint for the evaluation of X-rays in children and adults. *Clin Orthop Relat Res* 1976 ; 119 : 39-47.

20. Weinstein SL. Natural history and treatment outcomes of childhood hip disorders. *Clin Orthop Relat Res* 1997 ; 344 : 227-242.

21. Yrjonen T. Long-term prognosis of Legg-Calve-Perthes disease : a meta-analysis. *J Pediatr Orthop* 1999 ; 8-B : 169-172.

22. Yrjonen T. Prognosis in Perthes' disease after noncontainment treatment. 106 hips followed for 28-47 years. *Acta Orthop Scand* 1992 ; 63 : 523-526.