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Legg-Calvé-Perthes Disease: A Review of Cases with Onset Before Six Years of Age

By Scott B. Rosenfeld, MD, John A. Herring, MD, and John C. Chao, BA

*Investigation performed at the Texas Scottish Rite Hospital for Children, Dallas, Texas*

**Background:** The prognosis for children in whom Legg-Calvé-Perthes disease develops before the age of six years was initially reported to be good, but later studies demonstrated a less favorable prognosis. To assess the natural history of this condition in this age group, we reviewed a large cohort of children who had received minimal treatment for the disease.

**Methods:** The medical records and radiographs of all children with Legg-Calvé-Perthes disease seen at our institution between 1944 and 2000 were reviewed. The cases of 172 patients with a total of 188 affected hips were studied. The course of the disease and the final clinical and radiographic outcomes were evaluated in all patients.

**Results:** Typical Legg-Calvé-Perthes disease was noted in 164 hips of 160 patients. According to the lateral patellar classification, seven of these hips were in group A; 101, in group B; twenty-seven, in the B/C border group; and twenty-nine, in group C. According to the Stulberg classification, there were 131 Class-I or II (good) results, fourteen Class-III (fair) results, and nineteen Class-IV (poor) results. The lateral pillar classification was highly correlated with the outcome, whereas treatment did not correlate with the outcome. Only lateral pillar group B/C border and C hips with an onset of the disease between the ages of four years and five years and eleven months had a poor prognosis. We unexpectedly encountered a group of twelve patients with bilateral, simultaneous development of apparent Legg-Calvé-Perthes disease. In that group, there were twenty-one Stulberg Class-I or II (good), three Stulberg Class-III (fair), and no Stulberg Class-IV (poor) results, although fourteen of the twenty-four hips were in lateral pillar group C. The outcome in the hips in lateral pillar group C was significantly better when the patient had bilateral simultaneous involvement than when the patient had typical Legg-Calvé-Perthes disease.

**Conclusions:** The prognosis for patients with the onset of Legg-Calvé-Perthes disease before the age of six years is favorable, with 80% having a good result. Only children between the ages of four years and five years and eleven months with a B/C or C lateral pillar classification of involvement have a less favorable prognosis. Treatment was not found to influence outcome. Simultaneous bilateral disease, which had a more favorable prognosis, may represent a unique disorder.

**Level of Evidence:** Therapeutic Level III. See Instructions to Authors for a complete description of levels of evidence.

In the past, Legg-Calvé-Perthes disease with an onset before the age of six years was often thought to be a benign condition requiring little treatment. More recent studies have shown that poor results can occur in this age group, but the prevalence of these more severe cases has not been determined. The prognosis and indications for treatment of patients with Legg-Calvé-Perthes disease have recently been more clearly delineated with use of patient age and the severity of femoral head involvement as guidelines. The lateral pillar classification, used to estimate femoral head involvement, has been shown to correlate strongly with prognosis in children with the onset of Legg-Calvé-Perthes disease after the age of six years, and the system has good interrater reliability. A few investigators have attempted to correlate the extent of femoral head involvement with the outcome in younger patients. In a study of thirty-six hips in patients in whom the

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disease had developed before the age of five years, Fabry et al. reported an association between the lateral pillar classification and outcome in that 78% of group-C hips had a poor outcome\textsuperscript{18}. Their meta-analysis of the literature showed that results are quite mixed with regard to any correlation between age and outcome but that the extent of head involvement was of prognostic importance.

Many of the studies of younger children with Legg-Calvé-Perthes disease have included a small number of patients, and the reported results have been variable. Since 1944,
more than 300 patients with the onset of Legg-Calvé-Perthes disease prior to the age of six years have been treated at the Texas Scottish Rite Hospital. This study was performed to determine the outcome in an untreated or minimally treated group of patients in whom Legg-Calvé-Perthes disease developed before the age of six years and to determine if the lateral pillar classification is predictive of outcome in this younger patient population.

**Materials and Methods**

A search of the Texas Scottish Rite Hospital database identified 373 patients who had been diagnosed, between...
1944 and 2000, with Legg-Calvé-Perthes disease prior to the age of six years, and the medical charts and radiographs of those patients were reviewed.

Criteria for inclusion were a diagnosis of Legg-Calvé-Perthes disease prior to the age of six years, adequate radiographs including initial radiographs made when the disease was in the fragmentation stage or earlier, and later radiographs showing complete healing of the femoral head. When available, radiographs of other joints were reviewed to exclude children with evidence of multiple epiphyseal dysplasia. Patients with bilateral Legg-Calvé-Perthes disease who were included in this study had the onset of the disease in both hips prior to the age of six years. If the onset in the contralateral hip occurred after the age of six, only the first hip was included. Each hip was evaluated separately.

Each patient’s radiographs were reviewed throughout the course of treatment. Data recorded included the side that was affected, the lateral pillar classification, the Stulberg classification at the time of healing, and whether the triradiate cartilage was closed on the final radiograph. All radiographs were reviewed by the two senior authors (S.B.R. and J.A.H.), and the lateral pillar classification was determined from radiographs made when the disease was in the fragmentation stage with use of the criteria described by Herring et al.19. The Stulberg classification was determined from the radiographs at the time of final follow-up, after the femoral head had completely healed, also with use of the criteria of Herring et al.19. Stulberg Class-I and II results were considered good; Stulberg Class-III, fair; and Stulberg Class-IV, poor. Because the practice at our institution in the early years of this study was to discharge an asymptomatic patient with a healed, round femoral head, many of the patients had not reached skeletal maturity at the time of final follow-up. Previous studies have shown that an immature, round femoral head will remain round and that flat and ovoid femoral heads often remodel to become rounder as the patient approaches skeletal maturity6. Therefore, patients with an open triradiate cartilage and a completely healed femoral head seen on the final radiograph were included in this study (Figs. 1-A through 1-E), although we realize that this may skew our results toward being worse than those that would be seen in a study of skeletally mature hips.

When a patient had bilateral Legg-Calvé-Perthes disease, we compared the two hips to determine if the onset of the disease had been concurrent or sequential (Figs. 2-A through 2-E). If the onset had been concurrent, we compared the disease progression of the two hips. Radiographs of other joints, including the spine, knee, ankle, wrist, and shoulder, were made for the patients with bilateral involvement to exclude a diagnosis of epiphyseal dysplasia.

Medical records of each patient were reviewed to obtain clinical data, including gender, age at diagnosis, treatment, symptoms at the time of final follow-up, and age at the time of final follow-up.
The final study group included 172 patients (152 boys and twenty girls) with 188 affected hips. Two hundred and one patients were excluded from the study. Ninety-seven were excluded because medical records could not be located; sixty-three, because of inadequate radiographs; thirty-one, because the femoral head was not seen to be fully reossified on the final radiograph; six, because the earliest radiographs had been made when the disease was past the fragmentation stage; three, because the femoral head involvement was too slight for it to be classified as Legg-Calvé-Perthes disease; and one, because radiographs of other joints revealed multiple joint abnormalities.

Fifty-four patients (31%) were treated with a brace or cast. An Atlanta brace was used for twenty-six patients; a Craig splint, for ten; an A-frame brace, for four; an unspecified abduction brace, for six; and a Petrie cast, for eight. Seven hips treated with a Petrie cast also had an adductor tenotomy, and three of them also had an iliopsoas tenotomy at the time of cast application. One hundred and fourteen patients (66%) were observed or had treatment of symptoms only. Charts could not be located for four patients.

The Fisher exact test was used to compare rates, and multiple logistic regression methods were used to predict the probability of a particular outcome. Significance was defined as a p value of <0.05.

Results

The entire group included 188 hips diagnosed in 172 patients before they were six years old. The average age at the onset of the Legg-Calvé-Perthes disease was 4.6 years (range, 2.0 to 5.9 years). On the final radiograph, eighty-six

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**TABLE I Outcome Related to Lateral Pillar Group for All Hips***

<table>
<thead>
<tr>
<th>Pillar Group</th>
<th>Good Outcome (Stulberg Class I or II)</th>
<th>Fair Outcome (Stulberg Class III)</th>
<th>Poor Outcome (Stulberg Class IV)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>A or B</td>
<td>109</td>
<td>5</td>
<td>1</td>
<td>115</td>
</tr>
<tr>
<td>B/C border</td>
<td>18</td>
<td>4</td>
<td>8</td>
<td>30</td>
</tr>
<tr>
<td>C</td>
<td>25</td>
<td>8</td>
<td>10</td>
<td>43</td>
</tr>
<tr>
<td>Total</td>
<td>152</td>
<td>17</td>
<td>19</td>
<td>188</td>
</tr>
</tbody>
</table>

*The values are given as the number of hips.

(Figs. 2-A through 2-E) A child with the bilateral onset of Legg-Calvé-Perthes disease at the age of five years and four months. Fig. 2-A: Initial radiograph showing that both femoral heads are in the early stage of Legg-Calvé-Perthes disease. Both hips show a metaphyseal reaction. The right hip has increased density in the portion of the femoral head next to the physis.)
patients were seen to have an open triradiate cartilage and eighty-six had a closed triradiate cartilage. According to the lateral pillar system, seven hips (4%) were in group A, 108 (57%) were in group B, thirty (16%) were in the B/C border group, and forty-three (23%) were in group C. Since there were so few group-A hips and all had a good outcome, they were included with the group-B hips (for a total of 115 group-A or B hips [61%]) for statistical analysis (Table I). There were 156 patients with unilateral disease and sixteen with bilateral disease. One hundred and fifty-two results (81%) were good (Stulberg Class I or II), seventeen (9%) were fair (Stulberg Class III), and nineteen (10%) were poor (Stulberg Class IV). There was a significant correlation between the lateral pillar classification and the outcome (p < 0.001).

The 172 patients were divided into two groups on the basis of the radiographic appearance and progression of the disease. The first group consisted of patients with unilateral or bilateral Legg-Calvé-Perthes disease with typical characteristics. The second group comprised those with concurrent onset and progression of bilateral Legg-Calvé-Perthes disease.

**Typical Legg-Calvé-Perthes Disease (Group I)**

This group consisted of 160 patients with 164 affected hips. According to the lateral pillar classification, there were seven group-A hips (4%), 101 group-B hips (62%), twenty-seven group-B/C-border hips (16%), and twenty-nine group-C hips (18%) (Table II). One hundred and thirty-one results (80%) were good (Stulberg Class I or II), fourteen (9%) were fair (Stulberg Class III), and nineteen (12%) were poor (Stulberg Class IV). The Fisher exact test showed a significant correlation between the lateral pillar classification and the final outcome (p < 0.001).

When the patients were divided into two groups according to their age at onset (newborn to three years and eleven months or four years to five years and eleven months), the

![Fig. 2-B](https://i.imgur.com/2.png)

Initial frog-leg lateral radiograph showing the femoral head to have a crescent sign of a subchondral fracture. Both hips show a metaphyseal reaction.

<table>
<thead>
<tr>
<th>Pillar Group</th>
<th>Good Outcome (Stulberg Class I or II)</th>
<th>Fair Outcome (Stulberg Class III)</th>
<th>Poor Outcome (Stulberg Class IV)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>A or B</td>
<td>102</td>
<td>5</td>
<td>1</td>
<td>108</td>
</tr>
<tr>
<td>B/C border</td>
<td>15</td>
<td>4</td>
<td>8</td>
<td>27</td>
</tr>
<tr>
<td>C</td>
<td>14</td>
<td>5</td>
<td>10</td>
<td>29</td>
</tr>
<tr>
<td>Total</td>
<td>131</td>
<td>14</td>
<td>19</td>
<td>164</td>
</tr>
</tbody>
</table>

*The values are given as the number of hips.*
combination of the younger age group and a lateral pillar group-A or B classification was found to be significantly correlated with a better outcome ($p = 0.002$) with use of logistic regression methods. The resulting logistic regression model correctly classified the hips in terms of outcome 83.7% of the time and also estimated the probability of a good outcome as shown in Table III.

A logistic regression model was used to determine whether treatment (bracing with or without adductor tenotomy) affected the Stulberg outcome (good, fair, or poor) after adjustment for the lateral pillar classification (A or B, B/C, or C). The lateral pillar classification significantly affected outcome ($p < 0.0001$), but brace treatment did not regardless of the lateral pillar classification ($p = 0.75$) (Table IV). We also could not identify a correlation between gender and outcome.

**Bilateral Legg-Calvé-Perthes Disease with Concurrent Onset and Progression (Group II)**

Twelve patients were diagnosed with bilateral Legg-Calvé-Perthes disease with a concurrent onset prior to the age of six years. Each patient had also had concurrent bilateral progression through the stages of fragmentation and reossification. Ten of these twelve patients had radiographs of the spine and other joints, which did not reveal any abnormalities. Of the twenty-four hips, none were in group A, according to the lateral pillar classification, seven were in group B, three were in the B/C border group, and fourteen were in group C. Final radiographs revealed twenty-one good results (Stulberg class I or II), three fair results (Stulberg Class III), and no poor results (Stulberg Class IV). All three fair results occurred in group-C hips. With the numbers studied, we could not identify a correlation between the lateral pillar classification and outcome.

**Comparison of Groups I and II**

Ten of the twenty-nine lateral pillar group-C hips in the group with typical Legg-Calvé-Perthes disease (Group I) and none of the fourteen group-C hips in the group with concurrent bilateral disease (Group II) had a poor outcome. This difference was significant ($p = 0.038$).

**Typical Features of Group II**

There were radiographic changes that could be interpreted as indicating Meyer dysplasia in two patients (four hips) in Group

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**Table III Probability of a Good Outcome (Stulberg Class I or II) Relative to Lateral Pillar Group and Age Group**

<table>
<thead>
<tr>
<th>Age (yr + mo)</th>
<th>Lateral Pillar Group</th>
<th>Probability</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 to 3 + 11</td>
<td>A or B</td>
<td>0.99</td>
</tr>
<tr>
<td>4 to 5 + 11</td>
<td>A or B</td>
<td>0.93</td>
</tr>
<tr>
<td>0 to 3 + 11</td>
<td>B/C border or C</td>
<td>0.84</td>
</tr>
<tr>
<td>4 to 5 + 11</td>
<td>B/C border or C</td>
<td>0.43</td>
</tr>
</tbody>
</table>

---

Fig. 2-C

Initial abduction, internal rotation radiograph showing a distinct metaphyseal cyst on the right.
II. In six patients (twelve hips), there was a distinct increased density phase not seen in Meyer dysplasia (Figs. 2-A through 2-E). Two patients (four hips) had metaphyseal changes, and another two had radiographic changes in the femoral head that did not resemble those of Meyer dysplasia.

| TABLE IV Outcome According to Lateral Pillar Group and Whether Bracing Was Used* |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
|                                 | Stulberg Class I | Stulberg Class II | Stulberg Class III | Stulberg Class IV |
| Bracing                        |                 |                 |                  |                  |
| Lateral pillar A               | 0               | 0               | 0                | 0               |
| Lateral pillar B               | 2 (7%)          | 25 (89%)        | 1 (4%)           | 0               |
| Lateral pillar B/C border      | 0               | 8 (57%)         | 2 (14%)          | 4 (29%)         |
| Lateral pillar C               | 0               | 8 (57%)         | 2 (14%)          | 4 (29%)         |
| No bracing                     |                 |                 |                  |                  |
| Lateral pillar A               | 3 (43%)         | 4 (57%)         | 0                | 0               |
| Lateral pillar B               | 3 (4%)          | 65 (89%)        | 4 (5%)           | 1 (1%)          |
| Lateral pillar B/C border      | 0               | 7 (54%)         | 2 (15%)          | 4 (31%)         |
| Lateral pillar C               | 0               | 8 (53%)         | 1 (7%)           | 6 (40%)         |

*The values are given as the number of hips with the percentage in parentheses. There was no significant difference in outcomes between the group treated with and the group treated without bracing.

Discussion

No strong conclusions can be drawn from the studies published to date regarding the treatment and outcomes of Legg-Calvé-Perthes disease in patients with an onset prior to the age of six years\(^{6,10,18}\). Small sample sizes and variable results...
have made interpretation of these data difficult, and to our knowledge there has been no large multicenter prospective study of this age group. With use of the large database of young patients with Legg-Calvé-Perthes disease at our institution, we attempted to answer three questions. First, do patients who have the onset of Legg-Calvé-Perthes disease prior to the age of six years have good outcomes? Second, is the lateral pillar classification predictive of outcome in this younger population? Third, does treatment affect the outcome in this group of patients?

Fabry et al. performed a meta-analysis of the literature in an attempt to support or refute the belief that a young age is predictive of a good outcome. They concluded that the reported results were too variable to support any correlation between a young age and a good outcome. Furthermore, their own study of thirty-six patients who had had the onset of Legg-Calvé-Perthes disease prior to the age of five years suggested that the extent of femoral head involvement was of greater prognostic importance than age. One-third of the patients in their series had a poor result. Of nineteen hips that were classified as being in lateral pillar group C, only four had a good result and nine had a poor result. Fabry et al. concluded that, as evidenced by the high percentage of hips in lateral pillar group C, patients diagnosed prior to the age of five years were not protected from severe femoral head involvement and that greater involvement was predictive of a poor outcome.

Schoenecker et al. reported similar results in their review of the cases of 109 young patients; they concluded that the degree of femoral head involvement was the single most important variable predicting outcome.

Similar conclusions can be drawn from the results of the present study. The results for the entire group of patients were very good, with 81% having a good outcome and only 10% having a poor outcome. Further analysis showed that, among these younger children, age was not a significant independent variable. We found that the severity of femoral head involvement, as assessed with the lateral pillar classification, had a stronger independent correlation with outcome than did the age at the time of the disease onset. While 95% of the lateral pillar group-A and B hips had a good outcome, only 58% of the lateral pillar group-C hips had a good outcome in this early-onset population (p < 0.001). Excluding the bilateral cases with simultaneous onset, 48% of the group-C hips had a good result. Furthermore, we found that the combination of age and the lateral pillar classification was an even stronger predictor of outcome. Patients diagnosed with a lateral pillar group-A or B hip when they were between the ages of zero and three years and eleven months had a 99% probability of having a good outcome, whereas those diagnosed with a lateral pillar group-B/C or C hip when they were between the ages of four
years and six years and eleven months had only a 43% probability of having a good outcome.

While analyzing the radiographs, we noted that there was a distinct group of patients whose disease pattern and progression did not fit that of “typical” Legg-Calvé-Perthes disease. Initially, we identified thirteen patients who had bilateral Legg-Calvé-Perthes disease with concurrent onset and simultaneous progression through the radiographic stages. Typically, in patients with bilateral Legg-Calvé-Perthes disease, the onset of disease in the two hips occurs at different ages and there is asymmetric progression. Analysis of radiographs of the spine and other joints revealed only one patient who had features consistent with multiple epiphyseal dysplasia, and this patient was then excluded from the study. The remaining twelve patients had normal findings on radiographs of the spine and the other extremities and had no other disorders. Fourteen of these twenty-four hips were classified as being in lateral pillar group C, and many would have been expected to have a poor radiographic outcome. However, none had a poor result, and twenty-one (88%) of the twenty-four hips had a good result. Since these results were much better than expected, one may question the diagnosis of Legg-Calvé-Perthes disease in those patients. All patients who had radiographs of the spine or other joints showed none of the features of an epiphyseal dysplasia. As described by Meyer in 1964, dysplasia epiphysealis capitis femoris is characterized as a delayed appearance of bilateral capital femoral epiphysial ossification. Once the nucleus does appear, the epiphysis is fragmented but then it demonstrates constant improvement to the end point of a normal, round femoral head, and increased density is never seen on radiographs. This disorder is also characterized by a normal femoral neck without metaphyseal reaction or widening, and there is never flattening of the femoral head. As described by Rowe et al., the average age of onset of Meyer dysplasia (2.5 years) is earlier than that of Legg-Calvé-Perthes disease. Taking these factors into consideration, we do not believe that these twelve patients should be classified as having Meyer dysplasia. Our theory is that this group of atypical Legg-Calvé-Perthes disease may have had an as yet undescribed disorder of the femoral head, which will require further study.

The third question that we hoped to answer with this study was whether treatment affected the outcome in this young population. In a multicenter prospective study, Herring et al. demonstrated that patients who are over eight years of age at the time of the onset of Legg-Calvé-Perthes disease and have a lateral pillar classification of B or B/C have a better outcome with surgical treatment than they do with nonoperative treatment. These authors noted that nonoperative treatments such as bracing or casts had no influence on outcomes. Since the predominant approach to treatment for the patients in our study was observation and treatment of symptoms, all were treated essentially nonoperatively. Those that did receive some form of treatment underwent one of several forms of abduction bracing. The Craig brace, used in the early years, is an abduction bar attached to the shoes with which the patient can flex the knees and adduct the hip and hyperabduct the unaffected hip. Of the 172 patients studied, fifty-four were treated with bracing or splinting. Of these, seven had an adductor and/or iliopsoas tenotomy prior to bracing. One hundred and fourteen patients had only observation or treatment of symptoms. We could not detect a significant effect of bracing on outcome, regardless of the lateral pillar classification, compared with the outcomes in the untreated hips. We found that hips in lateral pillar groups B/C and C had a higher risk of a poor outcome. We cannot comment on the use of surgical containment of the femoral head for these lateral pillar groups since none of the hips in our series were treated surgically. On the basis of the very good results in this age group with lateral pillar group-A or B hips, it seems that more aggressive treatment is not indicated for these hips.

In conclusion, most patients with the onset of Legg-Calvé-Perthes disease before the age of six years have a good result. The lateral pillar classification is a strong predictor of outcome in this group of patients, and only those with a lateral pillar group-B/C border or C hip are at risk for a poor outcome. The age at onset is not an independent predictor of outcome in this group of younger patients. However, the combination of the age of onset and the lateral pillar classification is strongly correlated with outcome.

References


