Pathoanatomical Factors Responsible for Femoral Shortening in Legg-Calve-Perthes Disease

Sung-Man Rowe, M.D., Eun-Sun Moon, M.D., Myung-Sun Kim, M.D., Jun-Yub Lee, M.D., Chang-Ich Hur, M.D., and Tae-Yoon Ha, M.D.

Department of Orthopaedic Surgery, Chonnam National University Hospital, Gwangju, Korea

Purpose: The purpose of this study was to determine the relative contributions made by pathoanatomical factors responsible for femoral shortening in Legg-Calve-Perthes disease (LCPD), and to devise a method of reducing the amount of residual shortening based on a better understanding of its pathoanatomy and developmental pattern in LCPD.

Materials and Methods: We measured shortening of three anatomical components, namely, the femoral epiphysis, neck, and diaphysis on the teleoroentgenograms of 106 LCPD patients, comprised of 35 children with active disease, 24 in the healing stage, and 47 at skeletal maturity.

Results: The proportional contributions made by these 3 anatomical components to residual shortening at skeletal maturity were; 20% by the epiphysis (epiphyseal flattening), 53% by the neck (physeal growth retardation), and 27% by the diaphysis (underuse atrophy). These contributions differed according to disease stage and shortening severity. Mean diaphyseal shortening was 3.9 mm at skeletal maturity, but this increased to 5.8 mm when only patients with severe shortening (20 mm or more) were included.

Conclusion: Our findings suggest that diaphyseal shortening is likely to be minimized by the implementation of limb exercise programs.

Key Words: Legg-Calve-Perthes disease, Residual shortening, Pathoanatomical factors

Residual shortening is a common complication in Legg-Calve-Perthes disease (LCPD). The amount of femoral shortening is the sum of reductions due to epiphyseal height and femoral neck shortening, and underuse atrophy of bone.

However, no information is available in the literature on the relative contributions to residual shortening made by these three anatomical components, or on differences in their relative contributions during active disease, disease healing, and skeletal maturity.

To clarify the contributions made by these three anatomical components during each of these three disease periods, we measured femoral length discrepancies in LCPD patients.

In this study, we attempted to identify a means of reducing the amount of residual shortening by examining LCPD pathoanatomy and developmental patterns with respect to these three anatomical components.

MATERIAL AND METHODS

A total of 989 LCPD children were treated at our hospital between 1973 and 2004. We selected 106 LCPD children who had definite shortening of 2 mm or more, of the affected femur on teleoroentgenograms. All of cases were treated conservatively by skin traction, cast, or bracing. They were 35 children with active disease, 24 in the healing stage, and 47 at skeletal maturity. These disease stages were defined as ‘active’ when radiographs showed avascularity, fragmentation, or early healing, as ‘healing’ when radiographs showed reconstitution of the femoral head by new bone, and as ‘skeletal maturity’ in patients of 19 years or over. Mean patient age was 6.8 years at the active disease stage, 9.2 years at disease healing, and 20.2 years at skeletal maturity.

Teleoroentgenograms were taken as a single exposure of both legs on a long (35 × 90 cm) film at a distance of 2 m with a radiopaque ruler placed on the film cassette. Femoral length was measured from the top of the femoral head to...
the subchondral bone of the medial femoral condyle\(^6,7\). Two parallel lines, vertical to the longitudinal axis of the femur, were drawn to divide the femur into 3 components; the epiphysis, neck, and diaphysis with distal femur. One parallel line was drawn to pass through the center of the lesser trochanter and another to pass the center of the physeal line. The length of the epiphysical component was defined as the vertical distance from the top of the femoral head to a line passing the center of the physeal line. The length of the neck component was defined as the vertical distance between the two parallel lines. And, the length of the diaphysial component was defined as the vertical distance from the parallel line passing through the center of the lesser trochanter to the lowest point of the medial condyle (Fig. 1).

Statistical analysis

Statistical analysis was carried out using the Student’s \(t\) test for variables with a normal distribution and using the Mann-Whitney U test for those with non-normally distribution for two-group comparisons. Whereas, one-way ANOVA (analysis of variance) was used to compare differences between three or more groups with respect to variables with a normal distribution and the Kruskal-Wallis test was used for non-normally distributed variables.

Correlations between variables were examined using Spearman’s rank correlation test, and statistical analysis was performed using the SPSS software package (SPSS for Windows Release 10.0, Chicago, Illinois). \(p\)-values of \(<0.05\) were considered statistically significant.

RESULTS

Mean total femoral shortenings in children with active disease, during healing, and at skeletal maturity were 5.8 mm (range, 3.0 to 9.0 mm), 7.9 mm (range, 3.0 to 18.0 mm), and 14.8 mm (range, 3.0 to 28.0 mm), respectively. Anatomic component shortenings at these disease stages were 2.6 mm (44%), 2.9 mm (37%), and 3.0 mm (20%) in the epiphysis, 1.3 mm (22%), 3.0 mm (38%), and 7.9 mm (55%) in the neck, and 1.9 mm (34%), 1.9 mm (25%), and 3.9 mm (27%) in the diaphysis, respectively (Table 1) (Fig. 2).

The slight increase in epiphyseal length shortening was observed on moving from active disease to skeletal maturity, but this was not significant statistically. However, it showed a decreasing contribution to shortening moving from active disease to skeletal maturity, i.e., 44% during active disease, 37% at healing, and 20% at skeletal maturity (\(r=0.493, \(p\)-value 0.036).

<table>
<thead>
<tr>
<th>Period</th>
<th>Case No.</th>
<th>Femoral shortening (mm)</th>
<th>Epiphysis</th>
<th>Neck</th>
<th>Diaphysis</th>
<th>(p)-value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Mean (range)</td>
<td>mm</td>
<td>%</td>
<td>mm</td>
<td>%</td>
</tr>
<tr>
<td>Active disease</td>
<td>35</td>
<td>5.8 (3.0-9.0)</td>
<td>2.6</td>
<td>44</td>
<td>1.3</td>
<td>22</td>
</tr>
<tr>
<td>Disease healing</td>
<td>24</td>
<td>7.9 (3.0-18.0)</td>
<td>2.9</td>
<td>37</td>
<td>3.0</td>
<td>38</td>
</tr>
<tr>
<td>Skeletal maturity</td>
<td>47</td>
<td>14.8 (3.0-28.0)</td>
<td>3.0</td>
<td>20</td>
<td>7.9</td>
<td>53</td>
</tr>
<tr>
<td>(r^*)</td>
<td></td>
<td></td>
<td>0.605</td>
<td></td>
<td>0.102</td>
<td>-0.493</td>
</tr>
<tr>
<td>(p)-value</td>
<td></td>
<td></td>
<td>0.000</td>
<td></td>
<td>0.494</td>
<td>0.000</td>
</tr>
</tbody>
</table>

*Correlation coefficient.
Neck length showed the most remarkable increase in shortening with time ($r_t=0.595$, $p<0.001$).

According to our results, the final proportional contributions made by the three anatomical components at skeletal maturity was 20% for the epiphysis (epiphyseal flattening), 53% for the neck (physeal growth retardation), and 27% for the diaphysis (underuse atrophy). However, this distribution at skeletal maturity also varied according to the severity of shortening, as shown in Table 2.

For all 3 severity groups, the largest contribution to shortening was observed in the neck. Furthermore, both shortening in the neck ($r_t=0.788$, $p<0.001$) and diaphyseal shortening ($r_t=0.621$, $p<0.001$) were significantly increased as the severity of shortening increased. However, no significant change with respect to shortening severity was observed in the epiphysis. Mean diaphyseal shortening of the 47 LCPD patients measured at skeletal maturity was 3.9 mm, but it was 5.8 mm when only 15 cases with severe shortening (20 mm or more) were included.

**DISCUSSION**

Limpness caused by residual shortening of the affected limb is the most common and important finding in patients with LCPD. In the early stage of the disease, limpness is a combination of an antalgic and a Trendelenburg gait. In contrast, during the late stage of the disease, it is generally due to a combination of residual shortening and a Trendelenburg gait$^{10,13}$.

It is generally accepted that the amount of the leg shortening in LCPD depends almost exclusively upon the severity of endochondral ossification impairment in the proximal femoral growth plate$^{1,2,5,11}$. In addition, loss of epiphyseal height, underuse atrophy of the diseased limb$^{5,11,12}$, and femoral varus osteotomy (FVO) in older children are considered to contribute to shortening of the affected leg$^{3,5,9}$.

Diminution in the height of the epiphysis is caused by multiple mechanisms, i.e., growth retardation in cartilage cell duplication of the capital femoral epiphysis, retardation in ossification of the proximal capital epiphysis, and bone resorption after restoration of vascularity$^8$.

Shortening of the femoral neck is a result of blood supply deprivation to the proximal longitudinal growth plate$^9$.

Grzegorzewski et al.$^5$ studied 261 LCPD patients with unilateral involvement who had reached skeletal maturity at final follow-up. Leg length discrepancy (LLD) was found in 33 patients and ranged from 1 to 5.2 cm (average 2.51 cm). They found that the extent of disease involvement in the femoral head was associated with LLD, and that LLD was greater and more frequent in patients in Herring group C. They believed that necrosis involving the epiphysis and the metaphysis, may inhibit the growth of the proximal

### Table 2. Shortening distributions at skeletal maturity according to shortening severities

<table>
<thead>
<tr>
<th>Grade</th>
<th>Case No.</th>
<th>Femoral shortening (mm)</th>
<th>Epiphysis</th>
<th>Neck</th>
<th>Diaphysis</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Mean (range)</td>
<td>mm</td>
<td>%</td>
<td>mm</td>
<td>%</td>
</tr>
<tr>
<td>Mild (2-9 mm)</td>
<td>13</td>
<td>6.8 (3.0-9.0)</td>
<td>2.1</td>
<td>32</td>
<td>3.1</td>
<td>46</td>
</tr>
<tr>
<td>Moderate (10-19 mm)</td>
<td>19</td>
<td>13.4 (10.0-19.0)</td>
<td>3.4</td>
<td>26</td>
<td>5.9</td>
<td>44</td>
</tr>
<tr>
<td>Severe (≥20 mm)</td>
<td>15</td>
<td>24.0 (20.0-28.0)</td>
<td>3.1</td>
<td>13</td>
<td>15.2</td>
<td>63</td>
</tr>
</tbody>
</table>

$r_t$ Correlation coefficient.
femur. They commented along the following lines- "The amount of residual shortening of the affected limb in LCPD at the end of skeletal growth seems to depend on the extent of necrosis of the femoral head and on the inhibition of enchondral ossification in the proximal femur growth plate. Also, bone bridge formation between the epiphysis and the metaphysis increases leg shortening, and varus osteotomy produces more significant shortening than other methods of treatment. Neither age at symptom onset nor sex influences the amount of LLD; however, an early onset can result in more severe limb shortening.

Underuse atrophy of the bone is a result of reduced weight bearing, caused by an attempt to protect the affected limb or to relieve pain (antalgic gait), and by treatment applied by cast or orthosis. In conservatively treated children prolonged splinting of the affected limb contributes to delayed growth. Sahpiro documented that the treatment method chosen in Perthes can have an additional effect on length discrepancy at skeletal maturity. During the era in which unilateral abduction non-weight bearing bracing was prominent, reduced function of the affected limb increased discrepancy because normal growth stimuli to the distal femur and the tibia were absent.

Therefore, the sites responsible for residual shortening in the femur can be divided into three anatomical components; the epiphysis, neck, and diaphysis including the distal femur.

Despite copious number of reports regarding residual shortening in LCPD, reported results are inconclusive and conflicting in terms of the factors responsible, the role of causative factors during disease periods, and the amount of the shortening caused by causative factors.

As demonstrated by the present study, the total amount of residual shortening is the sum of diminution in the epiphyseal height, shortening of the femoral neck, and underuse atrophy of bone.

However, no information is available in the literature on the relative contributions to residual shortening made by these three anatomical components, and on different contribution made during different disease periods, i.e., active disease, disease healing, and skeletal maturity.

In this study, we determined the contributions of three anatomical components to residual shortening at skeletal maturity, and also compared these measurements with those at the periods of active disease and disease healing. Mean residual shortening at skeletal maturity indicated that 20% of the residual shortening of the femur developed as a result of flattening of the capital femoral epiphysis, 53% from growth retardation of the proximal femoral physis, and 27% from underuse atrophy. In addition, the relative contributions made by these three anatomical components differed in each stage of the disease.

The percentage contribution of epiphysal shortening was greatest during the stage of active disease and it gradually reduced until skeletal maturity. However, there was no significant change in the epiphysal shortening amount between the three disease stages.

The shortening of the neck component, in contrast, showed a continued increase with time in terms of its absolute and relative contribution, and indicating that the largest contribution to residual shortening was physeal growth retardation.

Underuse atrophy in the femoral diaphysis showed significant change, from only 1.9 mm in both the active and healing periods to 3.9 mm (27%) at skeletal maturity.

Furthermore, mean diaphysal shortening increased to 5.8 mm in cases with severe shortening of the femur (20 mm or more) at skeletal maturity (Table 2).

Knowledge of the degree to which shortening can be prevented by the efforts of doctors or patients is of substantial value. Both epiphyseal and neck shortening are caused by the disease process itself. Therefore, the prevention of the shortening of these two components is not possible. However, as underuse atrophy is partly caused by reduced use of the diseased limb, it can be minimized or prevented by efforts, and thus an appropriate exercise program for the affected limb is a prerequisite for minimizing residual shortening.

The limitation of the present study is the lack of investigation about relations between the femoral head involvement of the disease and LLD.

CONCLUSION

We determined the proportional contributions to residual shortening made by the three anatomical components responsible in LCPD by different mechanisms. Total residual shortening of the femur in LCPD is the sum of diminution of epiphyseal height (20%), physeal growth retardation (53%), and atrophy of the femur (27%).
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목적: Legg-Calve-Perthes 병(LCPD) 환자에서 대퇴골의 단축에 관여하는 병리해부학적 원인들의 상대적인 기여도를 결정하여 그 병리해부와 단축진행과정을 이해하고, 이를 통해 지단축을 감소시킬 수 있는 방법을 찾아보고자 하였다.

대상 및 방법: 106명의 LCPD 환자를 대상으로 원격 X-선 촬영법을 통해 대퇴골의 골두, 경부 및 간부의 단축정도를 측정하였으며, 질병 시기별 분포는 활동기가 35명, 치유기가 24명 그리고 골성숙기가 47명이었다.

결과: LCPD 환아에 있어서 골성장 완료 후 대퇴골 단축은 골두 단축에서 20% (골두 편평화), 경부에서 53% (성장판의 성장 지연), 그리고 간부에서 27% (저사용에 의한 골위축)로 기여하였으며, 이러한 부위별 기여도는 질병의 시기 및 단축의 정도에 따라 차이가 있음을 확인할 수 있었다. 또한, 대퇴골 간부의 단축은 골성숙기 총 47명의 환아에 있어서는 평균 3.9 mm였으나, 47명 중 20 mm 이상의 고도의 단축을 보인 15명의 환아에 있어서는 평균 5.8 mm의 단축을 보였다.

결론: 단축된 하지의 저사용 위축에 의한 대퇴골 간부의 단축은 환자의 운동부족이 원인의 하나로 생각되므로 적절한 운동처방으로 이를 예방할 수 있을 것으로 생각된다.

색인 단어: Legg-Calve-Perthes 병, 지단축증, 병리해부학적 원인

REFERENCES