Simultaneous Occurrence of Bilateral Legg-Calvé-Perthes Disease in Identical Twins

Sung Man Rowe, M.D., Taek Rim Yoon, M.D., Sung-Taek Jung, M.D., Jong Keun Seon, M.D., and Chang Ich Hur, M.D.
Department of Orthopaedic Surgery, Chonnam National University Hospital, Gwangju, Korea

This report describes simultaneous bilateral Legg-Calvé-Perthes disease (LCPD) development in four-year-old identical male twins. Moreover, the twins showed head involvement patterns with mirror symmetry. We included a review of the literature on this rare condition and discussed the genetic risks associated with LCPD.

Key Words: Legg-Calvé-Perthes disease, Identical twins

Familial and associated genetic interplay in Legg-Calvé-Perthes disease (LCPD) remains a major subject of discussion. Moreover, identical twin studies are of great value in terms of determining the genetic implications of LCPD. However, few reports are available on LCPD in identical twins. Since Giannestras3) first reported of LCPD in identical twins in 1954, only 7 reports have been issued1,2,4-6,8,9). Moreover, hardly any papers have reported the simultaneous occurrence of bilateral LCPD in identical twins. In our review of the literature, only one similar case of a bilateral occurrence was found9).

The present cases are of interest because demonstrated the simultaneous occurrence of bilateral LCPD in identical twins, and furthermore, they showed a mirror patterns of head involvement.

CASE REPORT

A 4-year-old male (H.N.), one of the twins, was initially seen because of pain on the right hip with a mild limp of 10 days duration. He and his brother had been well without any significant illness or trauma. They were delivered by Cesarean section and shared amnion, chorion, and placenta, and were therefore, defined as identical twins. They were of similar height, hair pattern, and facial appearance. A physical examination of one twin (H.N.) showed a slight limp and abduction and internal rotation restriction of the right hip. Initial radiographs showed that the right hip was LCPD of Catterall group III with a fragmentation of almost the entire epiphysis and reactive cystic changes in the metaphysis, and that the left hip was Catterall group II with condensation and a reduced ossific nucleus size (Fig. 2A, B). Bone scan, radiological bone survey and thyroid function testing were performed to differentiate Meyer’s disease, dysplasia epiphysealis multiplex congenita, hypothyroidism and so on. Technetium-99m-methylene diphosphonate (Tc-99m-MDP) pinhole scans of both hip joints demonstrated reduced uptake in the entire femoral heads of both hips, with an increased vascularity in the metaphysis of the femoral necks (Fig. 2C). The bone scan findings were typical of LCPD and differed from those of Meyer’s disease. Thyroid function and the other epiphysis were found normal. He was diagnosed with bilateral LCPD. Considering his age and mild symptoms, we decided to observe him at regular intervals and provided no definite treatment for the LCPD. The course of the illness and final outcome were uneventful. Radiographs of both hip joints during disease healing showed spherical and congruent femoral heads (Fig. 2D, E).

A physical and radiological examination of the other twin (W.N.) was done to evaluate the possible presence of a familial occurrence. A physical examination showed that he had no limp, and had a full range of painless motion of both hips. However, radiographs of both hips disclosed radiological...
findings typical of LCPD, in that the left hip had a LCPD of Catterall group III showing fragmentation of almost the entire epiphysis with a metaphyseal cyst, while the right hip had LCPD of Catterall group II with a lesser degree of involvement (Fig. 3A, B).

The radiological findings in these twins were high symmetrical in a mirror fashion, in terms of the extent of head involvement. The co-twin (W.N.) was also only observed at regular intervals without being given any definite treatment. Follow-up radiographs of both hips made at age 9 years were graded as ‘good’ by both the Mose and Stulberg Classifications, as were those of his twin (Fig. 3C, D).

**DISCUSSION**

The etiology of LCPD is complex and many contributory factors have been suggested. Moreover, its etiology remains obscure despite an increasing volume of literature. Genetic risks in LCPD have also been discussed in the literature. However, some controversy exists regarding the familial nature of the disease.

Wynne-Davies and Gormley reviewed 310 patients with LCPD and found no evidence of any genetic factors. Their results showed an extremely low frequency of LCPD patients among relatives, with no obvious pattern of inheritance. In contrast, numerous reports have shown a higher incidence of LCPD in families than in the general population. Stephens and Kerby reported a pedigree containing 28
individuals with LCPD among 86 family members in 5 generations. They determined that the pattern of heredity in the family was one of a simple Mendelian dominance with complete penetrance but a variable expression pattern. Wansbrough et al.9) reported a series of 129 cases of LCPD. Of these 12 cases showed a familial incidence, which was considered to be highly significant as compared to the incidence of 1:20,000 in the general population. Harper et al.4) reported two unrelated parents, a male with bilateral disease and a female with unilateral disease, who produced twins, both with LCPD. One was affected unilaterally and the other bilaterally. According to Harper et al.4), the mating of two

<table>
<thead>
<tr>
<th>Authors</th>
<th>Published year</th>
<th>Age (yrs+mos)</th>
<th>Sex</th>
<th>Case number</th>
<th>Simultaneous occurrence</th>
<th>Side of LCPD involvement in two co-twins</th>
</tr>
</thead>
<tbody>
<tr>
<td>Giannestras</td>
<td>1954</td>
<td>9+0</td>
<td>M</td>
<td>1</td>
<td>Yes</td>
<td>Unilateral and unilateral</td>
</tr>
<tr>
<td>Soderberg</td>
<td>1957</td>
<td>5+0</td>
<td>M</td>
<td>1</td>
<td>Yes</td>
<td>Unilateral and unilateral</td>
</tr>
<tr>
<td>Dunn</td>
<td>1960</td>
<td>5+6</td>
<td>M</td>
<td>1</td>
<td>Yes</td>
<td>Unilateral and unilateral</td>
</tr>
<tr>
<td>Inglis</td>
<td>1960</td>
<td>4+6</td>
<td>M</td>
<td>1</td>
<td>No</td>
<td>Unilateral and unilateral</td>
</tr>
<tr>
<td>Tracy</td>
<td>1963</td>
<td>5+0</td>
<td>F</td>
<td>1</td>
<td>Yes</td>
<td>Unilateral and unilateral</td>
</tr>
<tr>
<td>Bernbeck</td>
<td>1967</td>
<td>4+4</td>
<td>F</td>
<td>1</td>
<td>No</td>
<td>Combined⁹</td>
</tr>
<tr>
<td>Harper et al.</td>
<td>1976</td>
<td>-*</td>
<td>-*</td>
<td>1</td>
<td>-*</td>
<td>Combined⁹</td>
</tr>
<tr>
<td>Wansbrough et al.</td>
<td>1959</td>
<td>-*</td>
<td>M</td>
<td>1</td>
<td>-*</td>
<td>Bilateral and bilateral</td>
</tr>
<tr>
<td>Authors</td>
<td>2003</td>
<td>3+7</td>
<td>M</td>
<td>1</td>
<td>Yes</td>
<td>Bilateral and bilateral</td>
</tr>
</tbody>
</table>

Table 1. Cases of Legg-Calvé-Perthes disease in identical twins reported in the literature

Fig. 3. Radiographs of the other co-twin (W.N.). (A, B) Initial radiographs at four years of age showed that the left hip had LCPD of Catterall group III, and that it showed fragmentation of almost the entire epiphysis with a metaphyseal cyst, whereas the right hip had a LCPD of Catterall group II and a lesser degree of involvement. (C, D) Follow-up radiographs at 9 years of age showed a congruent and spherical femoral head in both hip joints.
affected individuals can give rise to affected monozygotic twins, suggestive of an additive genetic effect.

However, despite the fact that many pedigrees have been studied through several generations, few have investigated the occurrence of LCPD in identical twins, particularly bilateral occurrence. After an extensive review of the literature, only 8 reports of LCPD in identical twins were found (Table 1)\textsuperscript{1-6,8,9). Among these, 5 were unilateral, two were combined unilateral/bilateral, and only one was bilateral. Wansbrough et al.\textsuperscript{9) reported bilateral LCPD in monozygotic male twins and suggested that LCPD is a disease of a constitutional origin. After reviewing their 129 cases over a 30-year period, they found a familial incidence of approximately one in 35, while the incidence in the general population is approximately one in 20,000.

This is only the second report on the simultaneous occurrence of bilateral LCPD in identical twins and the ninth on LCPD in identical twins including unilateral involvement.

One interesting finding in the present case was that the pattern of femoral head involvement in these showed a mirror symmetry manifested as a Catterall group III in the right hip and Catterall group II in the left in one twin, and Catterall group III in the left hip and Catterall group II in right hip in the other.

In view of previous reports on the concordance LCPD in identical twins, and the present report of the simultaneous occurrence of bilateral LCPD in a radiologic mirror fashion in identical twins it becomes apparent, that genetic factors are likely to play a role in the development of LCPD.

REFERENCES