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LEGG-CALVÉ-PERTHES DISEASE

A Study of Lower Extremity Length Discrepancies and Skeletal Maturation

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Lower extremity length discrepancies and skeletal maturation have been studied in 147 patients with unilateral Legg-Calvé-Perthes disease followed by orthoroentgenograms for 5 or more years. At the time of initial assessment there was a marked delay in skeletal age as related to chronologic age in 83 percent of the patients. Standing height in the majority of patients both at disease presentation and at skeletal maturation was less than the mean. The average maximum total femoral and tibial discrepancy during the course of the disease was 2.14 cm. The maximum femoral discrepancy averaged 1.38 cm and the maximum tibial discrepancy averaged 0.93 cm. (The time of maximum discrepancy differed in the two major lower extremity bones). The extent of tibial discrepancy correlated well with the time of immobilization in the unilateral abduction ischial weight-bearing brace. The discrepancies did not invariably increase with time and many corrected with the repair process. Four developmental patterns of the discrepancy were detected and classified. Epiphyseal arrest was resorted to in 21 percent of the patients.

Key words: Legg-Calvé-Perthes disease; lower extremity length discrepancy; skeletal maturation

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Shortness of the involved extremity in the active stages of Legg-Calvé-Perthes disease is an almost inevitable occurrence, but there is virtually no documentation of the extent and eventual outcome of such discrepancies. An additional growth related matter is the suspected presence of systemic factors in the condition which results in delayed skeletal maturation and shortened stature. This paper reports orthoroentgenographic and anthropometric data from long-term studies of patients with Legg-Calvé-Perthes disease with emphasis on the extent of the lower extremity length discrepancies and on generalized growth parameters involving the rate of skeletal maturation and standing height at disease presentation and maturity.

PATIENTS AND METHODS

All patients referred to the Growth Study Unit of the Children's Hospital Medical Center, Boston, with the diagnosis of Legg-Calvé-Perthes disease from 1940–1972 were reviewed. During this time period virtually all patients with Legg-Calvé-Perthes disease were referred to the Growth Study Unit early in the course of the condition rather than later after an established discrepancy had been detected. Those included in this study had unilateral disease and had been assessed at annual intervals, or more frequently, for a minimum of 5 years prior to skeletal maturity. The treatment regimen used for the patients assessed was a unilateral abduction ischial weight-bearing caliper with a patten bottom such that the foot was suspended 3 inches from the ground (Ferguson 1963).

Unless indicated, the data presented are from 147 patients. The following parameters were assessed: 1. Sex distribution; 2. Side involved; 3. Age at disease presentation; 4. Skeletal maturation (skeletal age), as

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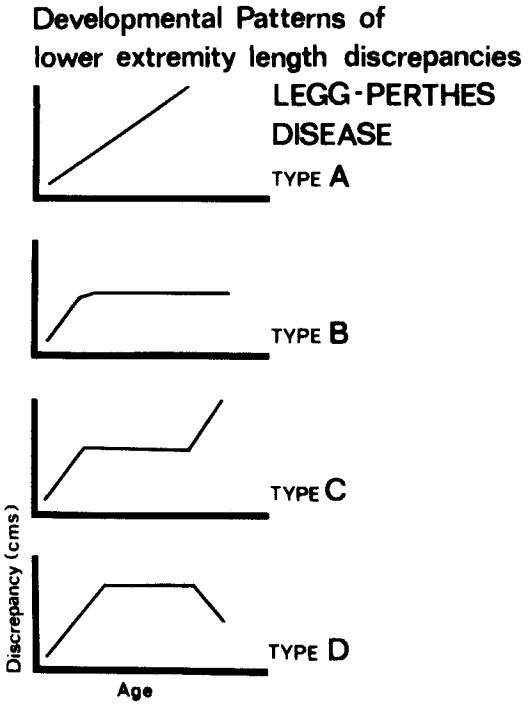


Figure 1. Four characteristic developmental patterns of lower extremity length discrepancies were seen in those with Legg-Calvé-Perthes disease. The patterns described were outlined by the discrepancy in centimeters as related to the chronologic age of the child as determined at each assessment. In the type A group the discrepancies continually increase with time. In type B the discrepancy increase falls off with time often reaching a plateau with no subsequent change for several years regardless of skeletal age maturation. In the type C pattern the discrepancy occurs, reaches a plateau for several years but, owing to premature closure of the capital femoral epiphyseal growth plate, shows an increase again just before skeletal maturity. In the type D pattern the discrepancy occurs and reaches a plateau but then diminishes on its own correcting either partially or fully.

determined from posteroanterior roentgenograms of the left hand and wrist and the Greulich and Pyle radiographic atlas of skeletal development (Greulich & Pyle 1959) (125 patients); 5. Standing height at disease presentation. The height was plotted on the Children's Hospital Medical Center Growth Study charts and the value expressed as standard deviations above or below the mean. The chronologic age was used for this assessment; 6. Standing height at skeletal maturation, expressed as the standard deviation position in relation to the mean; 7. Lower extremity length discrepancies. All measurements were made from orthoroentgenograms (Green et al. 1946). The most proximal bone point of the secondary ossification center of the femoral head was the reference point for both the diseased and normal proximal femur. The discrepancy assessments included (a) The maximum total femoral and tibial discrepancy during the course of the condition; (b) The final total femoral and tibial discrepancy; (c) The maximum and final femoral discrepancies; and (d) The maximum and final tibial discrepancies; 8. The maximum tibial discrepancy related to the length of time of brace immobilization (129 patients); 9. The developmental patterns of the discrepancies and their relationship to the age of onset (140 patients). A classification of the developing pattern types was formulated (Figure 1). The developmental patterns were established by charting the extent of the discrepancy with time; 10. The results of epiphyseal arrests (31 patients).

RESULTS

1. *Sex distribution* – The male/female distribution was 124 to 23 (5.4/1).
2. *Side of involvement* – In the entire series the right side was involved in 90 patients and the left in 57 patients (1.6/1) (Table 1).
3. *Age at disease presentation* – Table 2.
4. *Skeletal maturation* – Only those assessed initially within 1 year of disease presentation (125

Table 1. Sex distribution and side involvement with Legg-Calvé-Perthes disease

	Male 124		Female 23		Total 147 (5.4/1)	
Right	78	1.7/1	12	1.1/1	90	1.6/1
Left	46		11		57	

Table 2. Chronologic age at disease presentation

Age	1^0-1^{11}	2^0-2^{11}	3^0-3^{11}	4^0-4^{11}	5^0-5^{11}	6^0-6^{11}	7^0-7^{11}	8^0-8^{11}	9^0-9^{11}	10^0-10^{11}	11^0-11^{11}	12^0-12^{11}
Number	1	6	18	17	36	25	16	17	7	2	1	1

*Table 3. Skeletal maturation level at or within 1 year of disease presentation**

	Number	Percent
A. Skeletal age less than chronologic age (by 3+ months)	104	83
Skeletal age and chronologic age within 3 months of each other	14	11
Skeletal age greater than chronologic age (by 3+ months)	7	6
	125	

* 22 of the 147 patients assessed were seen 1 year or longer after initial disease presentation and are not considered in this subsection.

B. Extent of skeletal age retardation

0^3–0^{5+}	10 patients	2^0–2^5	19 patients
0^6–0^{11}	23 "	2^6–2^{11}	5 "
1^0–1^5	24 "	3^0–3^5	6 "
1^6–1^{11}	15 "	3^6–	2
			104 "

\+ 0^3 = 0 years 3 months of age
1^6 = 1 year 6 months of age

patients) have been included in this subsection. At the time of initial assessment shortly after disease presentation there was a marked delay in skeletal age as related to chronologic age (Table 3) in the large majority of patients with 83 per cent demonstrating a skeletal age less than the chronologic age by 3 months or more; 11 per cent demonstrating chronologic and skeletal ages within 3 months of each other and only 6 per cent demonstrating a skeletal age greater than the chronologic age. Sixty-eight percent of the patients with skeletal age retardation at the time of presentation or shortly after were from 1 to 3 or more years delayed.

5. *Standing height at disease presentation* – Table 4 illustrates the height distribution of the patients at the time of presentation. The heights represent the chronologic age/height determination. Fifty-three percent of the patients were less than the mean but virtually all remained within normal limits.

6. *Standing height at skeletal maturation* – Table 5 illustrates the distribution of standing height at maturation. *The same relative distribution as in the initial assessment persisted.* Fifty-nine percent of the patients had a final standing height below the mean although the vast majority still remained within the normal range. Although skeletal maturation occurred late, a compensatory "catch-up" phenomenon as regards total height was not seen.

7. *Lower extremity length discrepancies* – The side with the Legg-Calvé-Perthes disease was always shorter at some time during the disease process.

(a) Maximum total femoral and tibial discrepancy. The maximum average discrepancy during the course of assessment in the 147 patients was 1.5 cm or more in 113 patients (77 percent) and 1.0 cm or more in 139 patients (95 percent). The final average discrepancy in the entire series with or without epiphyseal arrest was 1.5 cm or greater in 50 patients (34 percent). During the course of the condition, the average maximum total femoral and tibial discrepancy in all patients was 2.14 cm; in those requiring epiphyseal arrest (31 patients) it was 2.99 cm; and in those not requir-

Table 4. Standing height at disease presentation

Height in relation to mean	>+2.0	+2.0	+1.5	+1.0	+0.5	Mean	–0.5	–1.0	–1.5	–2.0	<–2.0
Number of patients	0	3	7	14	23	22	32	29	9	5	3
Percent	0	2.0	4.8	9.5	15.6	15.0	21.8	19.7	6.1	3.4	2.0
			32%			15%			53%		

Table 5. Standing height at skeletal maturation

Height in relation to mean	>+2.0	+2.0	+1.5	+1.0	+0.5	Mean	−0.5	−1.0	−1.5	−2.0	<−2.0
Number of patients	0	1	5	13	15	26	36	31	5	10	5
Percent	0	0.7	3.4	8.8	10.2	17.7	24.5	21.1	3.4	6.8	3.4
			23.1%			17.7%			59.2%		

ing epiphyseal arrest (116 patients) it was 1.91 cm.

(b) The average final total femoral and tibial discrepancy in the entire group was 1.21 cm. In the group that had epiphyseal arrest the final average discrepancy was 1.27 cm and in the group that did not it was 1.21 cm.

(c) Maximum femoral discrepancy in all cases averaged 1.38 cm. In those not having arrest the average maximum shortness was 1.18 cm and in those having arrest it was 2.09 cm. The final femoral difference in those not having arrest, which is indicative of the extent of spontaneous correction, averaged 0.92 cm. As the maximum femoral difference was 1.18 cm and the final difference 0.92 cm, the average spontaneous correction was 0.26.

(d) Maximum tibial discrepancy. The average maximum tibial difference in all cases was 0.93 cm. In those not having arrest the average was 0.84 cm; in those having arrest it was 1.28 cm. The final tibial difference in the non-arrest cases

Table 6. Lower extremity length discrepancies in Legg-Calvé-Perthes patients

	cm	Range
a) Maximum total femoral and tibial discrepancy		
All patients (147) (average)	2.14	(0.7–4.9)
Patients without epiphyseal arrest (116)	1.91	(0.7–3.6)
Patients having epiphyseal arrest (31)	2.99	(2.1–4.9)
b) Final total femoral and tibial discrepancy		
All patients (147) (average)	1.21	(0 –2.9)
Patients without epiphyseal arrest	1.21	(0 –2.8)
Patients having epiphyseal arrest	1.27	(0 –2.9)
c) Maximum femoral discrepancy		
All patients (average)	1.38	(0.3–4.0)
Patients without epiphyseal arrest*	1.18	(0.3–2.9)
Patients having epiphyseal arrest	2.09	(1.0–4.0)
d) Maximum tibial discrepancy		
All patients (average)	0.93	(0.1–2.4)
Patients without epiphyseal arrest**	0.84	(0.1–2.1)
Patients having epiphyseal arrest	1.28	(0.2–2.4)

* The final femoral discrepancy in those not having epiphyseal arrest averaged 0.92 cm (range 0–2.9).

** The final tibial discrepancy in those not having epiphyseal arrest averaged 0.30 cm (range 0–1.2).
The final femoral and tibial discrepancies in those having arrest have not been assessed separately as the bone, or bones, chosen for arrest rarely corresponded to the exact discrepancy. For example, a 2.4 cm discrepancy composed of 1.6 cm in femur and 0.8 cm in the tibia was usually corrected with a distal femoral EA (epiphyseal arrest).

Table 7. Tibial discrepancy related to time of brace immobilization

	Less than 2 years	2^0–2^{11} years	3^0-3^{11} years	4 or more years
# Patients (129)*	28	63	26	12
Average tibial shortening (cm)	0.72 (±.07 SEM)	0.94 (±.05 SEM)	1.01 (±.08 SEM)	1.35 (±.14 SEM)

* 129 patients with well documented adherence to the brace-treatment regimen were used for this subsection. In those where brace use was sporadic, the tibial growth inhibition phenomenon was not assessed.

averaged 0.30 cm, indicating average spontaneous correction of 0.54 cm. The time of maximum femoral discrepancy rarely coincided with the time of maximum tibial discrepancy. The results and ranges of discrepancies are tabulated in Table 6.

8. *The maximum tibial discrepancy related to time of immobilization in the patten bottom splint* – There were 129 patients with well documented adherence to the brace regimen who were included in the assessment for this subsection. There was good correlation between the extent of tibial discrepancy and the length of immobilization; the longer the immobilization, the greater the tibial discrepancy (Table 7). The average maximum tibial shortening in 28 patients immobilized for less than 2 years was 0.72 cm (SEM ± 0.07), while the 12 patients immobilized for 4 years or more had an average shortening of 1.35 cm (SEM ± 0.14).

9. *Developmental patterns of discrepancies* – (Figure 1). When the extent of the discrepancy in centimeters was charted in relation to age it became evident that not all discrepancies had increased continually with time. A series of patterns of the developing discrepancies was identified and a classification made (Figure 1). Case studies illustrating each type are demonstrated in Figure 2a–d. The type A developmental pattern (discrepancy increasing continually with time) occurred in 21; 60 showed a type B pattern; 10 a type C pattern; and 49 a type D pattern, where the discrepancy occurred, reached a plateau and then partially or completely corrected without surgery.

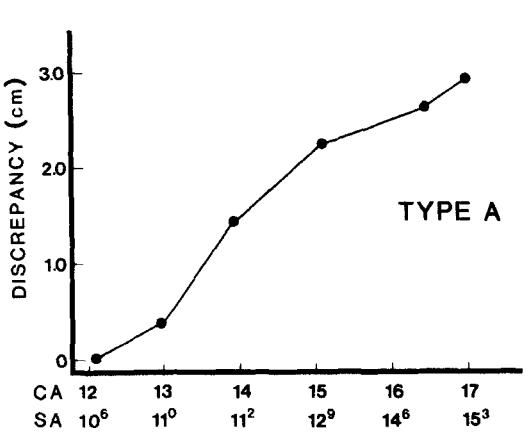
10. *Epiphyseal arrest* – Thirty-one patients had epiphyseal arrest. The average preoperative discrepancy was 2.99 cm and the average discrepancy at maturation was 1.27 cm.

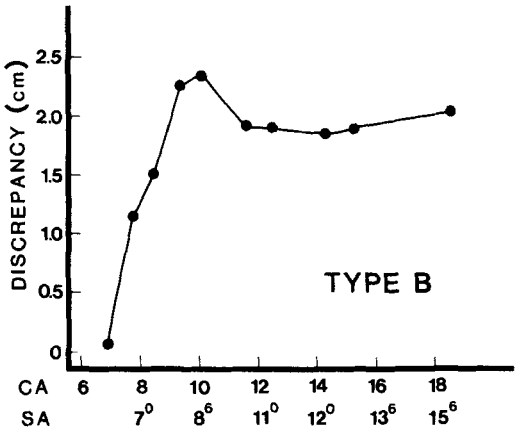
Table 8. Chronologic age of presentation in relation to discrepancy developmental pattern

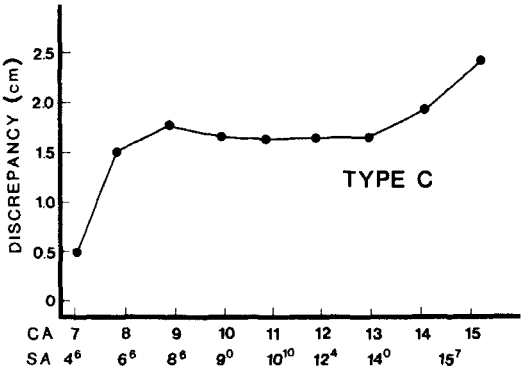
Developmental pattern	A	B	C	D
Average age at presentation (years)	8.7	6.5	5.6	5.3
No. of patients (140)	21	60	10	49

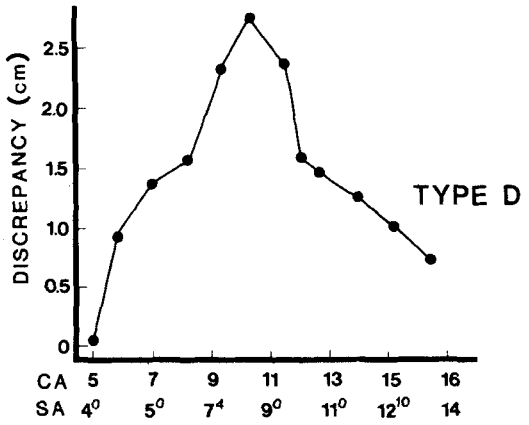
DISCUSSION

At the time of presentation of Legg-Calvé-Perthes disease delayed skeletal maturation was both frequent and considerable. The skeletal age was less than the chronologic age by 3 months or more in 83 percent and in 68 percent of those the retardation was 1 year or more. The patients were relatively short in stature with 53 percent being less than the mean, 15 percent at the mean and 32 percent above the mean. This finding indicates that overall growth retardation had occurred in advance of the clinical and radiologic changes which allowed the diagnosis to be made. Assessment of height at skeletal maturation showed persistence of the relative shortness indicating that the slight but definite growth retardation was not a transient phenomenon. Similar conclusions about a generalized growth slowdown have been reached by others (Goff 1954, Ralston 1955, Weiner & O'Dell 1970, Fisher 1972, Harrison et al. 1976, Burwell et al. 1978)









Figures 2a–d. CA-Chronologic age; SA-Skeletal age. The orthoroentgenograms were taken at yearly intervals based on the patients' chronologic age. The corresponding skeletal age for each time period is shown immediately beneath the chronologic age.

a. A representative case study from a patient with the type A pattern of discrepancy development is shown. The patient was male and developed Legg-Perthes disease at 12 years of age. His skeletal age was retarded by 18 months at the time of initial assessment. The discrepancy continued to increase with time and was 3.0 cm at maturation. Owing to the late onset of disease, sufficient time for repair and reversal of the discrepancy was not available.

b. A representative case study from a patient with the type B developmental pattern of discrepancy is shown. The male patient developed Legg-Perthes disease at chronologic age 6 years 10 months. Comparison of chronologic and skeletal ages indicates persisting retardation of the skeletal age. The maximum discrepancy reached was 2.3 cm at 10 years of age. The discrepancy remained virtually unchanged over the next 8 years and was 2.0 cm at skeletal maturity.

c. A representative case study of a patient with a type C developing discrepancy is shown. The boy developed Legg-Perthes disease at 6 years 4 months of age. The initial assessment at 7 years of age indicated 0.5 cm of shortening on the involved side. This increased rapidly during the first year, reaching 1.5 cm at 8 years of age. Between the chronologic ages of 9 and 13 the discrepancy was unchanged even though the growth was considerable as the skeletal age maturation increased from 8^6 to 14^0 during this 4 year period. Premature closure of the proximal femoral capital epiphysis then occurred and the discrepancy increased to 2.4 cm at maturity.

d. The type D developmental pattern is demonstrated. A 5-year-old boy developed Legg-Perthes disease which resulted in a maximum discrepancy of 2.6 cm attained shortly before 11 years of age. Over the subsequent 5 years the discrepancy diminished such that it was only 0.8 cm at skeletal maturity.

and detailed studies have indicated that genetic factors and familial short stature are not involved (Wynne-Davies & Gormley 1978). We must continue to postulate the presence of a systemic disorder of undefined nature in Legg-Calvé-Perthes disease making the already vulnerable proximal capital femoral epiphysis more susceptible to insults which render it necrotic.

The marked slowdown of growth in the femoral head in Legg-Calvé-Perthes disease associated with necrosis, subchondral fracture and collapse leads to shortening, the occurrence of which has been recognized for decades (Legg 1927). It is documented here that the average maximum femoral and tibial shortening in all patients was 2.14 cm. In the 21 percent of patients who eventually had epiphyseal arrest to correct discrepancies the average maximum femoral and tibial discrepancy was 2.99 cm. A significant contribution to the lower extremity shortening was from the ipsilateral tibia with the average maximum tibial discrepancy being 0.93 cm. This appeared due to disuse related to unilateral immobilization during treatment. The large majority of patients in this series were treated with the abduction patten bottom splint for 1½ to 4 years with good correlation noted between the tibial discrepancy and the time of immobilization.

This study has demonstrated that the repair process can lead to meaningful correction of discrepancies. Table 6 documents the extent of maximum and final femoral and tibial discrepancies. When the discrepancies were plotted against age, differing developmental patterns were outlined. These have been referred to as types A, B, C, and D (Figure 1) and illustrated by four case studies (Figure 2a–d). It is also shown that the pattern types relate directly to the disease and repair phenomenon rather than to skeletal age. This classification, in slightly expanded form, has been applied to discrepancy development in many childhood musculoskeletal conditions (Shapiro 1981). The four patterns seen indicate that careful continuing assessment of lower extremity length discrepancies should be done in an effort to improve the long-term results in such patients. A good correlation was found between the average age at presentation and the final developmental pattern of the discrepancy which developed. In the type A pattern the discrepancy continues to increase with time. The average age at presentation in this type was the oldest of the four groups, 8.7 years. The older patients have a relatively larger amount of necrosis and more importantly less time for repair. The type D pattern where the discrepancy was partially or entirely corrected by the repair process was associated with the youngest average age at presentation, 5.3 years. In these patients there was adequate time to allow for generally excellent repair to occur. In type B, where the discrepancy reached a plateau, the average patient age at presentation was intermediate to types A and D, being 6.5 years. These numbers do not represent absolute correlations; they do, however, provide an indication of how awareness of the developmental pattern classification can help in determining the outcome of discrepancies with time. The type C pattern if not appreciated can worsen a discrepancy just before skeletal maturation due to premature closure of the proximal femoral capital epiphysis. There were 10 instances of this phenomenon documented when both femoral and tibial measurements were plotted. The femoral discrepancy alone showed this conformation in 14 (10 percent).

Those patients having distal femoral epiphyseal arrest (31) had an average maximum total femoral and tibial discrepancy of 2.99 cm with a final total discrepancy averaging 1.27 cm. The existence of a large head with lateral extrusion or subluxation was such that the physician planning arrest frequently wished the involved extremity to remain slightly shorter such that, in gait, head coverage would be greater. The sluggish skeletal age maturation documented above appeared to have made timing prediction more difficult than in other diseases and failure to appreciate the differing developmental patterns outlined here led on occasion to inaccuracies in timing. This was especially true with group C patients where arrest was done occasionally late or not at all with the end-stage increase in discrepancy missed, owing to failure to appreciate the early closure of the proximal femoral plate. Change in the femoral head – greater trochanter relationship is an early radiologic indicator of this occurrence.

This review of the extent and developmental

patterns of lower extremely length discrepancies has proven useful in monitoring patients. Changing modes of therapy will alter the factors which contribute to discrepancy in Legg-Calvé-Perthes disease serving either to increase or decrease their extent. Tibial shortening is a factor in a unilateral abduction brace regimen but should be less where bilateral abduction plasters (Petrie & Bitenc 1971) or bilateral abduction splints (Lovell 1978) are used. The increasing use of operation to obtain containment (Axer et al. 1973, Salter 1973, Lloyd-Roberts et al. 1976) also affects femoral and lower extremity length and accurate assessment of discrepancy prior to and after operation is important. Innominate osteotomy will lengthen the lower extremity whereas proximal femoral varus osteotomy produces shortening. If varus osteotomy is done in an older patient, (9 years of age or greater), considerable shortening may occur as a result of the shortening already present, the small amount of growing time remaining for correction (type A developmental pattern) and the shortening produced by the osteotomy. Tibial discrepancy, however, should not be a factor in those having early surgical intervention since prolonged unilateral immobilization is not required.

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