Adaptive Changes in the Ipsilateral Femur of Patients with Unilateral Infantile and Adolescent Blount Disease

Abstract

Introduction: Pediatric patients with Blount disease frequently demonstrate secondary adaptive deformities in the adjacent distal femur. This study evaluates adaptation of longitudinal and angular proportions of the ipsilateral healthy femur to progressive leg length discrepancy in unilateral cases.

Methods: The study included 55 children with unilateral Blount disease. Preoperative radiographs were analyzed to characterize the condition as infantile or adolescent and measure femoral/tibial lengths and mechanical lateral distal femoral angles (mLDFA).

Results: There were 26 patients with infantile and 29 with adolescent Blount disease. Adolescent patients were significantly older (14.4 ± 2.0 vs. 9.2 ±2.4; p<0.01). Black race was prevalent in both groups (69-79%). The adolescent group was predominantly male (25/29; 86%), while the infantile group was predominantly female (15/26; 58%, p<0.01). Leg length inequality in adolescent patients was significantly greater than in the infantile group (2.8 ± 2.0 vs.1.5 ± 1.1cm; p<0.01) with ipsilateral femoral shortening (1.8 ± 1.8 cm) accentuating tibial shortening (1.0 ± 1.1cm). Patients with infantile Blount disease had more pronounced tibial discrepancy (2.0 ± 1.1 cm; p<0.01) but modest overgrowth of the ipsilateral femur (0.5 ± 0.7; p<0.01) partially compensating ipsilateral tibial shortening. There was a significant difference in tibial:femoral ratios between the groups (p<0.01). The infantile group had on average normal mLDFA (88°), most adolescent patients had accentuating distal femoral varus deformity (96° ± 5°; p<0.01).

Conclusions: Patients with unilateral infantile and adolescent Blount disease demonstrated distinctly different adaptation of the ipsilateral femur. Concomitant ipsilateral femoral changes aggravate angular deformity and leg length discrepancy in adolescent Blount disease.

Keywords: Genu Varum, infants, Adolescents, Developmental Bone Disease, Femur

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Introduction

Blount disease is a developmental disturbance of normal growth of the posteromedial aspect of the proximal tibial physis of unknown etiology [1-3]. At least two clinically and morphologically distinct forms ("infantile" and "adolescent") are recognized [1-4]. Although some cases of early-stage infantile Blount disease may resolve spontaneously or with brace treatment [1-4], progressive angular deformity due to medial proximal tibial physeal growth deceleration is typical. In patients with unilateral involvement, deceleration of growth of the affected tibia often results in progressive moderate leg length discrepancy in addition to angular deformity. Secondary adaptive deformity in the adjacent distal femoral epiphysis may also occur [1-5].

How the ipsilateral femur responds to progressive tibial growth deceleration and developing leg length discrepancy in unilateral cases of Blount disease has not been studied previously to our knowledge, and is the main purpose of this investigation. We hypothesized that the ipsilateral femur in patients with unilateral Blount disease would compensate for tibial growth deceleration by femoral growth acceleration to ameliorate the overall leg length discrepancy, identified as increased length compared to the contralateral femur of the unaffected limb. We also sought to determine if the femoral response to leg length discrepancy was different between the infantile and adolescent forms of Blount disease.

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Methods

With IRB approval, we reviewed the clinical and radiographic records of patients treated surgically at our hospital for Blount disease between 2000 and 2014. Inclusion criteria for this study was defined as no prior lower extremity surgery; unilateral involvement; and adequate preoperative medical records and radiographs to confirm a diagnosis of Blount disease and calculate angular and segmental length relationships. Preoperative standing radiographs of the lower extremities were analyzed to categorize the deformity as infantile or adolescent Blount disease, measure angular deformity of the distal femur, measure the length of the femoral and tibial segments of both the affected and contralateral extremities, and calculate the tibial:femoral ratio for the affected and contralateral legs. A femoral length difference between the affected and unaffected limbs ≥ 0.5 cm was chosen as evidence of acceleration or deceleration phenomena. Published normal values of mechanical lateral distal femoral angles (mLDFA) for skeletally mature patients range from 87 to 89° 1,2. We selected a difference in mLDFA between the affected and unaffected sides of ≥5° as beyond measurement error and clinically significant.

A chi-square test was used to compare categorical variables between infantile and adolescent groups of patients and for the small sample size a Fisher's exact text was utilized. Continuous variables were first examined for normality of data distribution and then a nonparametric test such as Mann-Whitney was executed for comparison between these two groups of patients. Statistical significance was set at $p < 0.05$.

Results

Of 232 patients undergoing surgery for Blount disease during the time interval studied, 55 patients met study criteria: 29 with adolescent and 26 with infantile Blount disease. Black ethnicity was predominant in both patient groups: 23 of 29 adolescent Blount patients (79%) and 18 of 26 infantile Blount patients (69%) were Black. The right and left legs were equally affected in the adolescent group (15R:14L), while left-leg involvement was more common in the infantile group (4R:22L). The adolescent group was predominantly male (25M:4F, 86% male), whereas infantile Blount disease patients were predominantly female (11M:15F, 58% female). Comparative analysis of these and other categorical variables is shown in Table 1.

Table 1. Ethnicity, Gender, and Incidence of Ipsilateral Femoral Overgrowth in Infantile and Adolescent Blount Patients

Categorical variables	Characteristic	Infantile Blount disease (n=23)	Adolescent Blount disease (n=24)	P-value
Ethnicity	Black Other	15 (65%) 8 (35%)	18 (75%) 6 (25%)	0.53
Affected side	Right Left	4 (17%) 19 (83%)	12 (50%) 12 (50%)	0.03
Gender	Female Male	13 (56.5%) 10 (43.5%)	3 (12.5%) 21 (87.5%)	<0.01
Femoral Overgrowth*	Yes No	16 (70%) 7 (30%)	5 (21%) 19 (79%)	<0.01
Femoral Overgrowth (≥ 0.5 cm)	Yes No	7 (30%) 16 (70%)	2 (8 %) 22 (92%)	0.07

*Affected side femoral overgrowth of any amount
p≤0.05 is statistically significant

As expected, patients with adolescent Blount disease were significantly older (14.4 ± 2 years; range, 10.8-17.3) than patients with infantile Blount disease (9.2 ±2.4 years; range, 5.3- 15.5; p<0.01) at the age of preoperative radiographic analysis. Patients with unilateral adolescent Blount disease had significantly greater leg length discrepancy than patients with infantile Blount disease (2.8 ± 2.0 cm vs.1.5 ± 1.1 cm; p<0.01). The discrepancy in adolescent Blount disease consisted of a combined tibial shortening (1.0 ± 1.1 cm) and concomitant ipsilateral femoral shortening (1.8 ± 1.8 cm). In contrast, patients with unilateral infantile Blount disease, while having more pronounced absolute tibial shortening (2.0 ± 1.0 cm, p<0.01), had on average modest overgrowth of the ipsilateral femur compared to the contralateral side (0.5 ± 0.7 cm) resulting in partial compensation of the tibia-induced leg length discrepancy. The difference in femoral longitudinal growth behavior (modestly accelerated or normal in the infantile group, but decelerated in the adolescent group) resulted in a significant tibial:femoral segment ratio difference between the groups. Patients with infantile Blount disease presented with significantly smaller tibial:femoral ratio (0.75± 0.04) than patients with adolescent Blount disease 0.81± 0.04, p<0.01). These continuous variables are summarized in Table 2.

Table 2. Comparison of tibial, femoral, and total LLD discrepancy; and mLFDA* between patients with infantile and adolescent Blount disease

Continuous variables	Infantile Blount disease (n=23) Mean±SD	Adolescent Blount disease (n=24) Mean±SD	P-value
Chronological Age (years)	7.0 ± 2.2	14.0 ± 2.0	<0.01
LLD (cm)	0.6 ± 0.8	1.8 ± 1.9	<0.01
Tibial discrepancy (cm)	0.8 ± 0.7	0.7 ± 1.3	0.5
Femoral discrepancy (cm)	-0.2 ± 0.7**	1.2 ±1.2	<0.01
Tibial:Femoral ratio	0.78 ± 0.04	0.81 ± 0.04	<0.01
mLFDA affected side (°) mLFDA contralateral side (°) Δ mLFDA** (°)	87 ± 2.4 88 ±2.8 -1.1	95 ± 4.1 88 ±2.3 6.5**	<0.01 0.11 <0.01

p≤0.05 is statistically significant
*mLFDA: mechanical lateral femoral distal angle
**Calculations of limb length discrepancy made by subtracting affected leg segment lengths from those of normal (contralateral) leg. Negative values indicate affected leg segment overgrowth.
***Δ mLFDA: Difference in mLDFA calculated by subtracting mLDFA of contralateral (normal) femur from that of the affected femur. Negative value indicates relative valgus deformity, and positive value increased varus deformity of the affected femur.

The majority of adolescent Blount patients had an accentuating distal femoral varus deformity, while none demonstrated an ipsilateral compensatory distal femoral valgus deformity. The average mLDFA of unaffected side was normal (88°; range, 85-96°) but the affected extremity mLDFA averaged 96° (range, 84-108°; p<0.01). This included 26 patients with varus deformity (mLDFA increased 5° or more), three with mLDFA within 5° of the contralateral, and none with ipsilateral valgus deformity (mLDFA decreased 5° or more). In the infantile group, the average mLDFA for both extremities were similar, normal, and measured 88° (range, 83-94°) and 89° (range, 85-94°) for affected and contralateral side, respectively. This included five patients with affected side valgus deformity (mLDFA decreased 5° or more), two with varus deformity (mLDFA increased 5° or more), and 19 with mLDFA within 5° of the contralateral. These findings are also summarized in Table 2.

Discussion

Since the original descriptions of Blount[3] and Langenskiöld[2], so-called "Blount disease" or "tibia vara (deformans)" has been generally accepted to present as one of two types in children without other obvious systemic illness or skeletal dysplasia: infantile and adolescent. This study was carried out in a group of patients prior to their first surgical intervention to determine the femoral response to tibial shortening typically seen in patients of either type with unilateral leg involvement.

The demographic and radiographic findings in our patient population confirmed previously known distinctions between infantile and adolescent Blount patients: infantile patients were younger, more frequently female, and had considerable medial physeal changes of the affected tibia, whereas adolescent patients were older, mostly male, and with less severe changes of proximal tibia. Although infantile Blount disease typically affects both lower extremities, our study indicates more frequent involvement of the left side in unilateral cases [(22 of 26)] Black ethnicity predominance in both groups correlated with other observations [1,2,6,7,8.] In both groups, patients with unilateral Blount disease had an associated leg length discrepancy. In patients with adolescent Blount disease, the total leg length discrepancy was, on average, greater than in patients with infantile Blount disease due to an accentuation of the discrepancy by concomitant ipsilateral femoral shortening. In our patient population tibial shortening of infantile Blount patients was partially ameliorated by modest "overgrowth" of the ipsilateral femur. This overgrowth, however, was not sufficient to offset the overall shortening of the affected leg compared to the contralateral side but significantly reduced the tibial: femoral segment ratio to 0.75± 0.04. Skeletal surveys have identified a normal ratio between 0.78 and 0.85[1,5,9]. The functional consequences of tibial: femoral segment disproportion has not been thoroughly investigated, although Liu et al identified an association between an increased tibial: femoral segment ratio and an increased incidence of hip and knee arthritis in a cadaver study[9.]

We found that the majority of patients with infantile Blount disease had a normal mLDFA, while several had either partially compensating valgus deformity (5/26) or accentuating varus deformity (2/26). In contradistinction, almost all patients with adolescent Blount disease in our study [(26 of 29)] had an associated varus deformity of the distal femur averaging 7° to aggravate the proximal tibial varus deformity. None of these 29 patients demonstrated compensatory distal femoral valgus, while three had a mLDFA comparable to the unaffected extremity.

We believe that the findings in our study support several assumptions. First, the more severe tibial shortening in infantile Blount disease, and the modest compensatory changes in the femur suggest a growth-decelerating injury localized to the posteromedial tibia with corresponding partial compensatory overgrowth in an otherwise healthy femoral segment. In contrast, the accentuating varus deformity and concomitant shortening of the ipsilateral femur in adolescent Blount disease suggests a more systemic etiology. Lastly, the modest and incomplete compensation of tibial shortening by femoral overgrowth in infantile group of patients and concomitant shortening of the femur along with tibial length deficit in adolescent group may suggest that maintenance of proper tibial: femoral segmental proportion may be more important to lower limb function than amelioration of leg length discrepancy by ipsilateral segment overgrowth.

This study has several shortcomings. Differences in femoral and tibial lengths that were documented in our patient population cannot be considered as representative of a natural history or longitudinal study of patients until skeletal maturity, since essentially none of our patients were skeletally mature at the time of our analysis. It is possible then that during the remainder of skeletal growth without surgical intervention differences in the length of the femur could change. On the other hand, to our knowledge, there are no natural history studies of Blount disease of either type, untreated to skeletal maturity, since effectively all affected patients undergo surgery of some nature prior to maturity, and are rarely observed prospectively to maturity before undergoing corrective surgery [10-14].

In conclusion, this study identified that patients with unilateral infantile and adolescent Blount disease have a completely different adaptation of the ipsilateral femur to the associated tibial deformities of shortening and varus. These findings remind the treating surgeon that careful assessment of the angular relationship and length of the ipsilateral femur is important in patients with Blount disease, particularly adolescent Blount disease, where accentuating shortening and varus deformity are common features.

Figure 1A Standing anteroposterior radiograph of a 8-year-old boy with left-sided infantile Blount disease. The right and left tibial lengths were 32.6 and 31.1 cm., whereas the right and left femoral lengths were 39.9 and 40.2 am, and the total LLD was 1.2 cm, left side shorter than right. The right and left mechanical distal femoral angles (mLDFA) were 91° and 88°, respectively.

Figure 1B. Standing anteroposterior radiograph of a 9-year-old girl with left-sided infantile Blount disease. The right and left tibial lengths were 30.8 and 26.5 cm., whereas the right and left femoral lengths were 37.5 and 38.7 cm., respectively. Despite 1.2 cm femur overgrowth on the affected side, the total leg length was 3.1 cm shorter than that of contralateral side. The right and left mechanical distal femoral angles (mLDFA) were 91° and 89°, respectively.

Figure 2. Standing anteroposterior radiograph of an 11+6-year-old boy with left-sided adolescent Blount disease. The right and left tibial lengths are 34.0 and 33.3 cm. and the right and left femoral lengths are 41.3 and 37.5 cm., respectively, resulting in a total leg length discrepancy of 4.5 cm., left side shorter than the right. The right and left mechanical distal femoral angles (mLDFA) are 87° and 96°, respectively.

Iranian Journal of Orthopaedic Surgery
Vol 19, No 1 (Serial No 72), winter 2021, p 1-6

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