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Comprehensive Treatment of Late-Onset Tibia vara

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Background: Late-onset tibia vara (Blount disease) can be difficult to treat because of frequent morbid obesity and associated deformities, including distal femoral varus, proximal tibial procurvatum, and distal tibial valgus, that contribute to lower extremity malalignment. We present a comprehensive approach that addresses all components of the deformity and allows restoration of the anatomic and mechanical axes.

Methods: Fifteen consecutive patients (nineteen lower extremities) with late-onset tibia vara were managed with this comprehensive approach. The mean age of the patients at the time of surgery was 14.9 years, and the mean weight was 113 kg. Standing anteroposterior and lateral radiographs were made preoperatively and at the time of the final follow-up. Preoperatively, the mean mechanical axis deviation was 108 mm, the mean lateral distal femoral angle was 95°, and the mean mechanical medial proximal tibial angle was 71°. In all nineteen extremities, the proximal tibial varus deformity was corrected by means of a valgus osteotomy and application of an Ilizarov ring external fixator. Distal femoral varus was corrected by means of either hemiepiphyseal stapling or valgus osteotomy with blade-plate fixation in thirteen of the nineteen extremities. Distal tibial valgus was treated either with hemiepiphyseal stapling or with varus osteotomy and gradual correction with use of the Ilizarov external fixator in eleven of the nineteen extremities.

Results: After a mean duration of follow-up of 5.0 years, the mean mechanical axis deviation had improved to 1 mm (range, 20 to –30 mm), the lateral distal femoral angle had improved to 87° (range, 83° to 98°), and the mechanical medial proximal tibial angle had improved to 88° (range, 83° to 98°). The mean time required for correction of the proximal tibial varus deformity was thirty-one days, and the external fixator was removed at a mean of 4.5 months postoperatively. All patients had development of one or more superficial pin-track infections (mean, 1.9 pin-site infections per patient). No wound infections, nonunions, or neurovascular complications occurred. Eighteen of the nineteen extremities were pain-free at the time of the final follow-up.

Conclusions: This comprehensive approach allowed restoration of the mechanical and anatomic axes of the lower extremity in patients with late-onset tibia vara, resulting in a resolution of symptoms as a result of normalization of the weight-bearing forces across the knee and ankle. We believe that this approach will decrease the risk of early degenerative arthritis of the knee.

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

Blount described a group of children who had an onset of varus deformity of the proximal part of the tibia in later childhood or adolescence, a condition that he described as adolescent tibia vara. Thompson et al. clarified this classification by categorizing patients who had had a juvenile onset of varus deformity (at the age of four to ten years) along with those who had had a true adolescent onset (at the age of eleven years or more) as having late-onset tibia vara. Early-onset or infantile tibia vara is characterized by a more severe proximal tibial deformity and substantial depression of the medial aspect of the proximal tibial epiphysis. Children with late-onset tibia vara present a challenging combination of marked genu varum deformity, usually accompanied by obesity. The deformity usually develops in middle childhood or early adolescence and is differentiated from infantile tibia vara on the basis of the less severe epiphyseal deformity and the history of normal lower extremity alignment prior to the development of a genu varum deformity.

Although the name of the disorder suggests that varus of the proximal part of the tibia is the only deformity present, this is not the case. The proximal tibial varus deformity is produced by a posteroomedial growth suppression, which initially produces varus and then progressive procurvatum of the proximal part of the tibia. Associated toeing-in gait also worsens as the deformity progresses, initially as a functional maneuver to allow the foot to be placed as close to the line of progression as possible and subsequently as a result of progressive internal tibial torsion. The combination of proximal...
tibial varus and procurvatum as well as internal tibial torsion results in a complex three-dimensional deformity. The progressive proximal tibial varus deformity produces increased forces on the medial aspect of the distal femoral physis, leading to growth suppression and a distal femoral varus deformity as well. In severe cases, the varus deformity in the distal part of the femur can be nearly as severe as that in the proximal part of the tibia. As the proximal tibial and distal femoral varus deformities increase, substantial strain is placed on the lateral collateral ligament of the knee, which can lead to laxity and instability of the knee joint. Finally, in severe cases, compensatory distal tibial valgus may develop, allowing the patient to align the ankle joint parallel to the floor.

Although the formation of an osseous bar across the medial aspect of the proximal tibial physis along with medial epiphyseal depression is not uncommon in patients with severe infantile tibia vara, similar bar formation in patients with late-onset tibia vara has not been reported, to our knowledge. Depression of the medial aspect of the proximal tibial epiphysis requiring medial plateau elevation has not been reported in patients with late-onset tibia vara.

Although late-onset tibia vara occasionally may occur in thin or normal-sized patients, the majority of patients are obese. These patients typically are in the ninetieth percentile of weight for both age and height. During normal walking, nonobese individuals place the foot close to the midline of foot progression, thereby minimizing weight transfer and energy expenditure. Obese individuals with large thighs have a very difficult time adding the hip adequately to place the foot for midline progression. Davids et al. speculated that this “fat thigh syndrome” produces a varus moment on the knee that leads to increased pressure on the medial aspect of the proximal tibial physis and inhibits growth in accordance with the Hueter-Volkmann law.

Since 1994, we have addressed all of the deformities associated with late-onset tibia vara with a comprehensive approach. Proximal tibial varus is treated with a valgus osteotomy followed by gradual correction with use of an Ilizarov external fixator. If the patient has open physeal and adequate growth remains for correction of the deformities of the distal part of the femur and distal part of the tibia, hemiepiphyseal stapling is performed. When the physeal are closing, these deformities are corrected with osteotomy. Distal femoral varus is treated with a valgus osteotomy and stabilization with a blade plate. Distal tibial valgus is treated with a varus osteotomy followed by gradual correction with use of an Ilizarov external fixator. All procedures are performed simultaneously. The purpose of the present study was to evaluate the results of this comprehensive approach to the deformities in patients with late-onset tibia vara.

**Materials and Methods**

Fifteen consecutive patients (nineteen lower extremities) with late-onset tibia vara were managed with our comprehensive approach between January 1, 1994, and December 31, 1998. Medical records, hospital charts, and radiographs were retrospectively reviewed with regard to demographic characteristics, operative procedures, complications, and functional and radiographic data. All nineteen extremities underwent a proximal tibial valgus osteotomy followed by gradual correction of the proximal tibial varus deformity with use of an Ilizarov circular external fixator (Smith-Nephew-Richards, Memphis, Tennessee). This device was also used to correct associated proximal tibial procurvatum and internal tibial torsion. The distal femoral varus deformity was corrected with use of a valgus osteotomy and stabilization with a 95° adult condylar blade plate (Synthes, Paoli, Pennsylvania) if insufficient growth remained to allow lateral hemiepiphyseal stapling. Distal tibial valgus deformity was treated either with medial hemiepiphyseal stapling or with varus osteotomy and gradual correction with use of an extension of the Ilizarov frame. All patients were followed for a minimum of two years postoperatively.

The lower extremities were evaluated preoperatively and at the time of the final follow-up with use of standard radiographs. These included a standing anteroposterior radiograph of both lower extremities; a standing anteroposterior radiograph of both ankles, showing the distal half of the tibia; and a lateral radiograph of the knee, showing the proximal half of the tibia. Line measurements were performed according to the system of Paley et al. (Figs. 1-A and 1-B). These measurements included the mechanical axis deviation, the mechanical lateral distal femoral angle (normal, 88°; range, 86° to 89°), the mechanical medial proximal tibial angle (normal, 87°; range, 85° to 89°), the mechanical lateral distal tibial angle (normal, 91°; range, 88° to 95°), and the joint-line congruency angle (normal, 2°; range, 1° to 3°). Lateral radiographs were evaluated to determine the posterior proximal tibial angle (normal, 80°; range, 77° to 84°).

All patients complained of pain in the involved knee or knees preoperatively and were observed to have a lateral knee thrust when walking.

The mean age at the time of surgery was 14.9 years (range, 10.6 to 18.1 years). The etiology was juvenile onset tibia vara for three patients and adolescent tibia vara for the remaining twelve patients. The mean weight was 113 kg (range, 43 to 178 kg). Thirteen patients (87%) were above the ninety-fifth percentile for weight according to their age, and fourteen patients (93%) were above the ninety-fifth percentile for weight according to their height. Four patients were so obese that they had sleep apnea that necessitated nighttime use of continuous positive airway pressure.

Preoperative anteroposterior radiographic measurements of the lower extremities demonstrated a mechanical axis deviation of 108 mm (range, 41 to 208 mm), a mean mechanical lateral distal femoral angle of 95° (range, 82° to 102°), and a mean mechanical medial proximal tibial angle of 71° (range, 61° to 77°). These measurements all indicated substantial lower extremity malalignment. The mean joint-line congruency angle was 3° (range, −2° to 12°). Radiographs of the ankles revealed a mean mechanical lateral distal tibial angle of 85° (range, 79° to 93°). Lateral radiographs revealed a mean posterior proximal tibial angle of 71° (range, 58° to 88°). The
mean preoperative lower limb-length discrepancy in the eleven patients with unilateral disease was 14 mm (range, 0 to 30 mm) as assessed either with scanography or on the standing anteroposterior radiograph of both lower extremities. None of the four patients with bilateral disease had a substantial limb-length discrepancy.

All nineteen extremities had a proximal tibial osteotomy with placement of an Ilizarov circular external fixator. Correction of the distal femoral varus was indicated if the lateral distal femoral angle was >95° (representing >5° of mechanical varus). According to this criterion, thirteen of the nineteen femora had a distal varus deformity that required correction. Eleven of these thirteen extremities had a distal femoral varus osteotomy with blade-plate fixation, whereas the other two had enough remaining growth to undergo lateral distal femoral hemiepiphyseal stapling.

Correction of the distal tibial valgus deformity was performed if the mechanical lateral distal tibial angle was ≤86° (representing ≥5° of mechanical valgus). According to this criterion, eleven of the nineteen tibiae had a distal valgus deformity that required correction. Eight of these extremities had a distal tibial varus osteotomy with gradual correction, and the other three were corrected by means of medial hemiepiphyseal stapling. Two extremities had lateral instability of the knee secondary to lateral collateral ligament laxity and were treated with concomitant distal transport of the proximal part of the femur.
fibula to tighten the ligament. Four extremities underwent only a proximal tibial osteotomy, six extremities underwent a proximal tibial osteotomy with correction of either the distal part of the femur or the distal part of the tibia, and seven extremities underwent deformity correction at the proximal part of the tibia, the distal part of the femur, and the distal part of the tibia. Two extremities required deformity correction at the proximal part of the tibia, the distal part of the femur, and the distal part of the tibia as well as fibular transport to tighten the lateral collateral ligament.

Surgical Technique
Preoperatively, radiographs were analyzed and planning was performed according to the techniques described by Paley and Tetsworth. An Ilizarov circular external fixator was preconstructed to lengthen the tibia as needed and to simultaneously correct varus and procurvatum through an oblique-plane placement of the hinges. After correction of the angulatory deformity and restoration of length, the fixator was modified in situ to correct rotational malalignment. In two patients (three tibiae) with greater rotational deformity, an inclined oblique-plane hinge initially was constructed to provide simultaneous correction of alignment in the frontal, coronal, and rotational planes, with restoration of length. If the patient was skeletally mature, a proximal two-ring block, with the rings located approximately 2 cm apart, was utilized to stabilize the proximal part of the tibia. If the patient was skeletally immature, a single proximal ring was used to avoid performing the osteotomy.
distally in the diaphysis of the tibia. This proximal ring block was connected to the distal ring segments with appropriately placed hinges or struts. Two distal rings were used; the rings were locked with rods if a distal tibial osteotomy was not required, and they were connected with appropriately placed hinges or struts if a distal osteotomy was to be performed. If clinically important lateral collateral ligament laxity was present, a separate block of pins was used to secure the distal end of the proximal fibular fragment and to transport it distally, thereby tightening the lateral collateral ligament.

After appropriate general endotracheal anesthesia had been induced and the patient had been carefully positioned on a radiolucent surgical table (Orthopedic Systems, Hayward, California), the entire lower extremity was prepared and draped and a sterile tourniquet was placed about the proximal part of the thigh. The distal femoral varus deformity was corrected first, when necessary. A supracondylar distal femoral opening-wedge valgus osteotomy was performed through a lateral approach. A large Kirschner wire was first placed perpendicular to the lateral cortex of the femur to serve as a reference wire. A second Kirschner wire was then placed into the distal metaphysis at or just proximal to the physis. It was oriented such that the subtended angle between it and the initial Kirschner wire was 5° less than the preoperatively measured deformity (to allow for the placement of the 95° condylar blade-plate). The initial reference Kirschner wire was then removed. The chisel was then placed into the distal femoral metaphysis, just proximal and parallel to the second Kirschner wire and the blade length. An osteotomy was then created 2 cm proximal to the chisel with use of an oscillating saw, and the chisel was withdrawn. The five-hole 95° blade-
plate was then inserted into the slot created by the chisel and was impacted into place. The second Kirschner wire was then removed. The distal part of the femur and the attached blade-plate were then reduced to the femoral shaft, creating a stable opening-wedge osteotomy.

After the femoral osteotomy but prior to closure of the femoral wound, while the tourniquet was still inflated, a posterolateral incision was made over the fibula at the junction of its middle and distal thirds. Subperiosteal exposure of the fibula was performed to avoid injury of the branches of the deep peroneal nerve to the extensor hallucis longus or the peroneal artery and vein. A 1-cm section of the fibula was removed with use of an oscillating saw and, if necessary, was cut into pieces and placed into the femoral osteotomy site as bone graft. Both femoral and fibular osteotomy wounds were then thoroughly irrigated and closed. The femoral wound was closed over a drain.

If hemiepiphyseal stapling of the distal part of the femur or the distal part of the tibia was planned, the staples were placed extraperiosteally under fluoroscopic guidance as described by Zuege et al. prior to application of the external fixator.

The tourniquet was deflated, the preconstructed circular fixator was placed onto the lower extremity, and suction tubing was used to suspend the leg within the external fixator. This tubing was placed under the leg and over the external fixator and was clamped into place with use of hemostats to secure the leg appropriately within the fixator. Transverse and transfibular wires initially were placed both proximally and distally along the osteotomy site, and the construct was complete.

**Fig. 2-F** Lateral radiograph of the knee, showing partial correction of the proximal tibial procurvatum with a healed osteotomy site. The posterior proximal tibial angle is 76°.

**Fig. 2-G** Standing anteroposterior radiograph of the right ankle, made seven months postoperatively, showing partial correction of the distal tibial valgus with a lateral distal tibial angle of 88°.

**Fig. 2-H** Standing anteroposterior long-cassette radiograph, made four years postoperatively, showing maintenance of the correction and a neutral mechanical axis. The mechanical axis deviation is 7 mm, the lateral distal femoral angle is 90°, the medial proximal tibial angle is 88°, and the joint-line congruency angle is 2°. At the time of the most recent follow-up, the patient was asymptomatic.
Results

The fifteen patients were followed postoperatively for a mean of 5.0 years (range, 2.3 to 9.8 years). All patients were skeletally mature at the time of the last follow-up. The mean time required for correction of the proximal tibial varus deformity was thirty-one days (range, eleven to sixty-eight days). The external fixator was removed after a mean of 4.5 months (range, 2.9 to 6.4 months).

All patients had clinical improvement. At the time of the last follow-up, fourteen patients (eighteen extremities) were asymptomatic and one patient (one extremity) continued to complain of knee pain, the etiology of which could not be determined. The latter patient had a final mechanical axis deviation of 7 mm lateral to the joint center, with good alignment of the distal part of the femur and the proximal part of the tibia. Magnetic resonance imaging demonstrated no evidence of intra-articular abnormality. The pain did not limit the patient’s activities, and the patient declined additional intervention.

The lateral knee thrust was eliminated in all patients. All patients had regained full ranges of knee and ankle motion at the time of the last follow-up. No patient had clinically important knee laxity to varus or valgus stress at the time of the last follow-up.

Radiographs that were made at the time of the last follow-up revealed improvement in lower extremity alignment. The mechanical axis deviation improved to a mean of 1 mm (range, 20 to –30 mm) (Table I). The lower extremities were essentially equal in length, with a mean discrepancy of 1 mm (range, –22 mm to 6 mm) at the time of the final follow-up (Figs. 2-A through 2-H).

The mean mechanical lateral distal femoral angle improved to 87° (range, 83° to 98°). In the two patients who underwent hemiepiphyseal stapling, the mechanical lateral distal femoral angle improved from 92° to 87° in one patient and from 96° to 87° in the other. The mechanical lateral distal femoral angle in the eleven patients who underwent distal femoral osteotomy improved from a mean of 97° (range, 94° to 102°) to a mean of 88° (range, 84° to 96°).

The mean mechanical medial proximal tibial angle improved to 88° (range, 83° to 98°). The mean joint-line congruency angle improved to 2° (range, 0° to 5°). The mean posterior proximal tibial angle improved mildly from 71° (range, 58° to 88°) to 77° (range, 57° to 89°). The mean mechanical lateral distal tibial angle improved slightly from 85° (range, 79° to 93°) to 89° (range, 82° to 93°). The mean mechanical lateral distal tibial

<table>
<thead>
<tr>
<th>TABLE I Radiographic Measurements</th>
<th>Preoperative*</th>
<th>Postoperative*</th>
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<tbody>
<tr>
<td>Mechanical axis deviation (mm)</td>
<td>108 (41 to 208)</td>
<td>1 (20 to –30)</td>
</tr>
<tr>
<td>Mechanical lateral distal femoral angle (deg)</td>
<td>95 (82 to 102)</td>
<td>87 (83 to 98)</td>
</tr>
<tr>
<td>Joint-line congruency angle (deg)</td>
<td>3 (–2 to 12)</td>
<td>2 (0 to 5)</td>
</tr>
<tr>
<td>Mechanical medial proximal tibial angle (deg)</td>
<td>71 (61 to 77)</td>
<td>88 (83 to 98)</td>
</tr>
<tr>
<td>Mechanical lateral distal tibial angle (deg)</td>
<td>85 (79 to 93)</td>
<td>89 (82 to 93)</td>
</tr>
<tr>
<td>Posterior proximal tibial angle (deg)</td>
<td>71 (58 to 88)</td>
<td>77 (57 to 89)</td>
</tr>
</tbody>
</table>

*The values are given as the mean, with the range in parentheses.
angle in the three patients who underwent hemiepiphyseal stapling of the distal part of the tibia improved from 84° (range, 83°, 86°, and 83°) to 89° (86°, 88°, and 92°). The mean mechanical lateral distal tibial angle in the eight patients who underwent osteotomy of the distal part of the tibia improved from 83° (range, 79° to 86°) to 90° (range, 86° to 92°).

All patients had development of one or more superficial pin-site infections (mean, 1.9 infections per patient), which resolved with oral antibiotics. No patient required intravenous antibiotic therapy. No neurovascular complications were encountered. No patient had development of a deep infection, osteomyelitis, or nonunion.

Postoperatively, the need for additional procedures required fourteen of the fifteen patients to return to the operating room between one and three times per involved extremity. Removal of the external fixator under a general anesthetic was requested by the patient for eighteen of the nineteen extremities. Other indications for additional procedures included wire removal (thirteen patients), fixator modification (two patients), and staple removal (two patients). In all, only one extremity required no additional procedures, six required one additional procedure, nine required two additional procedures, and three required three additional procedures.

**Discussion**

Patients with late-onset tibia vara typically are obese and can present with complaints of genu varum, knee pain, toeing-in, or knee instability. The natural history of this condition is unclear, but residual infantile tibia vara leads to chronic knee pain and degenerative arthritis. Varying amounts of respiratory distress may be present, depending on the patient’s obesity and activity level. Occasionally, even the walk from the waiting room to the examination area can be exhausting for these patients. Patients with knee pain often describe medial pain and symptoms of increased pressure in the medial compartment of the knee as well as symptoms of anterior knee pain, probably secondary to maintenance of the knee in a flexed position during gait.

A standing long-cassette anteroposterior radiograph of both lower extremities is essential for the evaluation of these patients. If the patient appears to have unilateral bowing, careful attention should be paid to the contralateral limb because the patient’s obesity frequently can mask mild contralateral genu varum. Because of the obesity of these patients, we have found it necessary to make preoperative preparations for special equipment, including a large tourniquet and a radiolucent table that can support the patient’s weight.

Although some authors have recommended physeal distraction as a means of achieving correction, this is not possible in skeletally mature adolescents. Effectively utilized an oblique, laterally based closing-wedge osteotomy that hinges at the intact cortex just distal to the proximal tibial physis for acute correction of mild deformities. Most patients with late-onset tibia vara require at least proximal tibial osteotomy to correct the deformity. Osteotomy with limited internal fixation, the mainstay of treatment of infantile tibia vara has little application in the treatment of late-onset tibia vara.

The obesity of the patient makes the application of a well-fitting above-the-knee cast difficult, if not impossible, and prevents the adequate control of the position of the osteotomy site. In addition, the older age and size of the patient make patient mobility highly desirable. The problems associated with non-weight-bearing on the affected extremity make walking extremely difficult for these patients. Furthermore, the difficulties associated with assessing limb alignment either intraoperatively or postoperatively make residual proximal tibial varus or undercorrection a possibility. External fixation of the tibia is indicated for the correction of more severe deformities. This allows either acute or gradual correction with a monolateral or circular fixator. Price et al. demonstrated that monolateral fixation with either acute or gradual correction can effectively correct the tibial varus deformity in the frontal plane. Alignment in the sagittal plane and rotational correction are more difficult to obtain with use of the monolateral fixator.

A detailed preoperative plan is essential for any surgical procedure. The magnitude and location of the various osseous deformities, laxity of the lateral collateral ligament, the presence of joint contractures, and lower limb-length discrepancy should all be assessed and incorporated into the preoperative plan. The overall plan should correct each of the constituent deformities, normalizing the mechanical and anatomic axes in order to avoid increased stresses resulting from chronic malalignment as well as increased shear forces on the knee or ankle resulting from joint-line obliquity. Although the long-term results of our comprehensive technique are not known, this approach has provided pain relief, a stable correction, and restoration of normal lower extremity alignment in this difficult patient population.

During the planning and execution of the varus correction, particular attention should be paid to normalizing distal femoral and proximal tibial alignment. Because of the obesity of these patients, joint-contact forces are excessive even when normal mechanical alignment is present. Although the mechanical alignment in these patients could be corrected through a single tibial osteotomy that compensates for existing femoral varus, this would leave the knee joint oblique to the mechanical axis and would introduce shear forces during weight-bearing. Because of these concerns, we believe that it is essential to correct all of the deformities separately in order to provide optimal mechanical alignment and to delay the onset of degenerative arthritis.

Circular external fixation with gradual correction of the proximal part of the tibia allows for maximal adjustability of the alignment in all planes and is indicated for more severe deformities and the most obese patients. Other advantages include stable fixation with improved patient mobility; the ability to evaluate alignment in a functional, standing position; and the ability to accurately correct all deformities of the tibia, including proximal tibial varus and procurvatum, internal tibial torsion, and distal tibial valgus.

After correction of the genu varum deformity, the distal
tibial valgus, if uncorrected, can produce a pes planus deformity with hindfoot valgus. This persistent distal tibial valgus leaves the ankle in a position in which it is oblique to forces involved in walking, leading to sheer forces at the joint surface. This can become painful, particularly in obese patients with late-onset tibia vara. Wagner et al. found decreased tibiotalar contact areas at the ankle in patients with distal tibial deformities of ≥10°. Ting et al. found that this effect was exacerbated in patients with decreased subtalor motion who were unable to compensate for the valgus. This has led us to correct deformities of >5° at the distal part of the tibia.

In conclusion, this comprehensive approach to late-onset tibia vara is effective, but careful preoperative planning and an analysis of all of the deformities are essential. In particular, correction of each of the deformities in the lower extremity allows the mechanical alignment of the lower extremity to be optimized with the restoration of anatomic and mechanical axes. This approach has led to relief of pain through adolescence, and we are hopeful that it will eliminate the onset of degenerative arthritis in the knee and ankle, although the long-term results remain to be seen.

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