

## CURRENT CONCEPTS REVIEW

# Blount Disease

By Sanjeev Sabharwal, MD

- Two clinically distinct forms of Blount disease (early-onset and late-onset), based on whether the lower-limb deformity develops before or after the age of four years, have been described.
- Although the etiology of Blount disease may be multifactorial, the strong association with childhood obesity suggests a mechanical basis.
- A comprehensive analysis of multiplanar deformities in the lower extremity reveals tibial varus, procurvatum, and internal torsion along with limb shortening. Additionally, distal femoral varus is commonly noted in the late-onset form.
- When a patient has early-onset disease, a realignment tibial osteotomy before the age of four years decreases the risk of recurrent deformity.
- Gradual correction with distraction osteogenesis is an effective means of achieving an accurate multiplanar correction, especially in patients with late-onset disease.

Blount disease is a developmental condition characterized by disordered endochondral ossification of the medial part of the proximal tibial physis resulting in multiplanar deformities of the lower limb. The first detailed description was provided by Blount in 1937<sup>1</sup>, and this was followed by another extensive study by Langenskiöld in 1952<sup>2</sup>. Although Blount coined the term *tibia vara*, implying a solely frontal plane deformity, subsequent authors noted that multiplanar deformities are commonly seen with this condition<sup>3-5</sup>. Secondary to the asymmetrical growth with relative inhibition of the posteromedial portion of the proximal tibial growth plate, a three-dimensional deformity of the tibia with varus, procurvatum (apex anterior), and internal rotation develops, along with possible limb shortening in unilateral cases. This entity can lead to a progressive deformity with gait deviations, limb-length discrepancy, and premature arthritis of the knee<sup>6-8</sup>.

### Classification

Two clinically distinct forms of Blount disease—early-onset (Figs. 1-A through 1-F), or infantile, and late-onset (Figs. 2-A through 2-G)—have been described, with the classifications

based on whether the limb deformity develops before or after the age of four years<sup>1,2,9</sup>. Thompson and Carter<sup>9</sup> further classified late-onset Blount disease as a juvenile type (onset at the age of four to ten years) and an adolescent type (onset after the age of ten years). Bilateral involvement is common, particularly with an early-onset presentation<sup>4</sup>. Although there are key clinical and radiographic differences between early and late-onset Blount disease, there are several similarities, including a predisposition for obese black children and children of Scandinavian descent. Also, there are comparable histologic findings at the proximal tibial growth plate<sup>9-11</sup>. Langenskiöld<sup>2</sup> described six radiographic stages of progressive changes at the proximal tibial epiphysis and metaphysis in children with early-onset Blount disease (Fig. 3). With advancing age and higher Langenskiöld stages (V and VI), irreversible physeal changes with permanent inhibition of the medial portion of the tibial growth plate can occur. Although the Langenskiöld classification is useful, there is substantial interobserver variability, especially with regard to the intermediate stages<sup>12</sup>. Loder and Johnston<sup>13</sup> studied the applicability of the Langenskiöld classification to a predominantly nonwhite population (one in which 73% of the patients

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Fig. 1-A



Fig. 1-B

Clinical photograph (Fig. 1-A) and standing anteroposterior radiograph (Fig. 1-B) of a five-year-old girl with bilateral early-onset Blount disease.

were black) with early-onset Blount disease. These authors noted that the age at presentation of each of the Langenskiöld stages was much younger than that originally described in Langenskiöld's report from Scandinavia<sup>2</sup>, and the overall outcome was worse despite treatment.

### Etiology and Pathogenesis

Several authors have suggested a mechanical basis for Blount disease given the observation of a predisposition for the disease in children who start walking at an early age and those who are overweight<sup>14-18</sup>. The pathogenesis of the proximal tibial deformity is likely related to excessive compressive forces causing growth inhibition, as suggested by the Heuter-Volkman principle<sup>18,19</sup>. Excessive pressure at the medial portion of the proximal tibial cartilaginous epiphysis causes altered structure and function of the chondrocytes along with delayed ossification of the epiphysis<sup>20</sup>. Obesity can substantially increase the compressive forces generated on the medial compartment of the knee joint in a child with genu varum<sup>14,16,17</sup>. Using finite element analysis, Cook et al.<sup>14</sup> calculated the stresses at the proximal tibial growth plate during simulated single-limb stance and noted that, in a five-year-old obese child, the compressive

forces generated with 10° of varus angulation exceeded those necessary to retard physeal growth. Dietz et al.<sup>16</sup> examined the relationship between body weight and angular deformities in fifteen young children with Blount disease. They observed a significant correlation between body weight and the tibio-femoral shaft angle ( $r = 0.75$ ) and noted an even stronger relationship between body weight and varus deformity when nine obese children were considered independently ( $r = 0.92$ ). Using gait analysis, Gushue et al.<sup>17</sup> studied the effect of childhood obesity on three-dimensional knee joint biomechanics. Compared with children of normal weight, overweight children showed a substantially higher peak internal knee abduction moment during early stance, with increased loading of the medial compartment of the knee joint. My colleagues and I<sup>18</sup> recently reported a linear relationship between the magnitude of obesity and biplanar radiographic deformities in children with the early-onset form of Blount disease and in patients with a body-mass index of  $>40 \text{ kg/m}^2$  irrespective of the age at the onset of the Blount disease. Despite having a lower body-mass index, children with early-onset Blount disease had more severe varus and procurvatum deformities of the proximal part of the tibia than did adolescents with

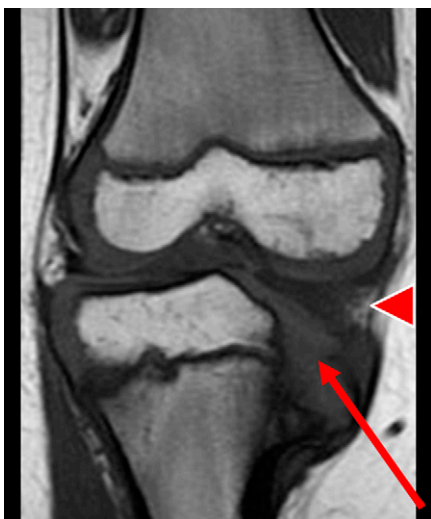


Fig. 1-C

A T1-weighted magnetic resonance imaging scan of the right knee demonstrates the unossified medial aspect of the tibial epiphysis (arrow), which has a lower signal intensity than the adjacent lateral aspect. Also note the hypertrophic medial meniscus (arrowhead), which is often seen in this condition.

Blount disease<sup>18</sup>. Wenger et al.<sup>11</sup> suggested that the proximal tibial growth plate responds differently at various stages of skeletal maturity, with the increased pliability of the unossified epiphyses of younger patients causing more growth inhibition than occurs in adolescents.

Daivids et al.<sup>15</sup> examined gait deviations related to increased thigh girth associated with adolescent obesity. An obese child with large thighs has difficulty adducting the hips adequately and this may result in “fat-thigh gait” by producing a varus moment on the knees, thus increasing pressure at the medial part of the proximal tibial physis. This concept supports the observation that preexisting varus alignment of the knee is not necessary to initiate the pathologic changes seen in some patients with late-onset Blount disease<sup>21</sup>. Recent studies suggest that childhood obesity reduces bone mineral content to levels below what would be predicted on the basis of body weight<sup>22</sup>. Such factors may further predispose obese children with Blount disease to the development of progressive deformities with increasing weight.

The mechanical etiology does not fully explain the unilateral or asymmetrical involvement of the limbs that is often seen in these children or the observation that some children with classic radiographic findings of Blount disease, especially the early-onset type, are not overweight<sup>18,23</sup>. One report showed no difference in terms of obesity or early walking age between black children with early-onset Blount disease and their peers in a local population<sup>23</sup>. However, no details about the children’s weight, body-mass index, or radiographic findings were provided. Currently, the etiology of Blount disease remains unknown and is probably multifactorial. It is likely that various genetic, humoral, biomechanical, and environmental factors control physal growth and influence the development of nor-

mal lower-limb alignment. The clinical manifestations of both forms of Blount disease may represent an alteration in the normal growth and development of the lower limb in genetically predisposed children through distinct but related pathways.

## Imaging Studies

### Plain Radiographs

The classic changes in the proximal part of the tibia in an established case of early-onset Blount disease include a sharp varus angulation of the metaphysis, widening and irregularity of the medial aspect of the growth plate, medial sloping and irregular ossification of the epiphysis, and beaking of the medial part of the epiphysis<sup>19</sup> (Figs. 1-B and 3). In addition to the Langenskiöld classification<sup>2</sup>, there are other radiographic parameters such as the metaphyseal-diaphyseal angle<sup>24-27</sup>, epiphyseal-metaphyseal angle<sup>28</sup>, and relative contributions to the varus deformity by the femur and tibia<sup>29,30</sup> that can help to differentiate physiologic bowing from early-onset Blount disease in children who are less than two years old (Fig. 4). However, none of these radiographic markers can be considered in isolation, and repeated clinical and radiographic examination is essential to establish the diagnosis<sup>31</sup>.

A standing full-length anteroposterior radiograph (teleroentgenogram) of the entire length of both lower extremities with the patellae forward is crucial for a detailed analysis of frontal plane alignment<sup>4,32</sup> (Figs. 1-B and 2-B). It can be challenging to make this radiograph for an obese patient



Fig. 1-D

An intraoperative arthrogram of the right knee was made at the time of a proximal tibial osteotomy to visualize the articular surface of the medial tibial plateau (arrow). Also note the hypertrophic medial meniscus (arrowhead).





Fig. 1-E



Fig. 1-F

Despite satisfactory clinical (Fig. 1-E) and radiographic (Fig. 1-F) outcomes at one year following bilateral tibial realignment with gradual correction with external fixation, the patient is at a high risk for recurrent deformity and will need close follow-up.

because of poor visualization of the osseous details and the tendency for the radiology technician to position the lower limb with the foot forward rather than the patella forward. When the child is young and has not yet had sufficient ossification of the patella, it is useful to place a metal marker over the center of the patella to confirm the knee-forward position. Moreover, no more than 60% of the proximal part of the fibula should be seen to overlap the adjacent tibia on a true anteroposterior radiograph centered at the knee, irrespective of the patient's age<sup>33</sup>.

Despite being referred to as *tibia vara*, Blount disease may involve other sources of medial axis deviation arising from the distal part of the femur<sup>4,34-36</sup> and an intra-articular deformity creating dynamic varus malalignment<sup>4</sup>. A full-length radiograph allows detailed assessment of the mechanical axis

deviation and joint orientation angles, which are crucial for determining the site(s) of deformity correction<sup>32</sup>. Unlike the case in children with early-onset Blount disease, approximately one-third of the varus deformity in adolescents with late-onset disease may be attributable to the distal part of the femur<sup>4,34</sup>. A procurvatum deformity of the proximal part of the tibia is typically noted on the full-length lateral radiograph of the tibia (Fig. 2-F), while the sagittal profiles of the distal parts of the femur and tibia are typically within normal limits<sup>4</sup>. Thus, in order to avoid creating iatrogenic deformities and having incomplete correction, a comprehensive assessment of multiplanar deformities is necessary prior to embarking on operative treatment<sup>4,37</sup>. A scanogram and an assessment of bone age are useful for a complete assessment of current and future limb-length discrepancy.



Fig. 2-A

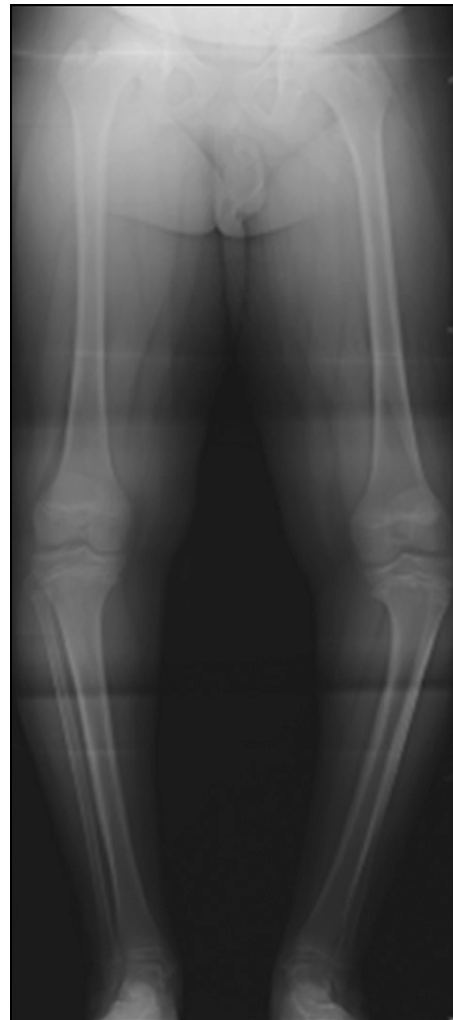


Fig. 2-B

Clinical photograph (Fig. 2-A) and standing anteroposterior radiograph (Fig. 2-B) of a twelve-year-old obese boy with bilateral late-onset Blount disease. The patient underwent realignment of both tibiae a few months apart. (Figs. 2-A through 2-G reprinted, with permission, from: Sabharwal S. Blount's disease. In: Rozbruch SR, Ilizarov S, editors. Limb lengthening and reconstruction surgery. New York: Taylor and Francis; 2006. p 511-20.)

### Advanced Imaging

Plain radiographs may lead to an overestimation of the so-called depression of the medial plateau in advanced stages of early-onset Blount disease<sup>38,39</sup>. An intraoperative arthrogram is helpful for delineating the articular surface<sup>39-41</sup> (Fig. 1-D) and for evaluating dynamic instability of the knee. The knee arthrography is usually done during the same anesthesia session as used for the definitive operative procedure, such as a proximal tibial osteotomy. A magnetic resonance imaging scan can also define intra-articular changes such as posteromedial depression of the tibial plateau and hypertrophy of the medial meniscus in children with early-onset disease<sup>38,42,43</sup> (Fig. 1-C). Fat-suppressed and proton-weighted magnetic resonance images are particularly helpful in detecting growth plate irregularities and early physal bar formation<sup>44</sup>. Although advanced imaging is not routinely indicated for patients with late-onset Blount disease, changes in the adjacent distal femoral epiphysis

and physis have been observed on magnetic resonance imaging scans of adolescents<sup>45</sup>. Computed tomography scanning, particularly with three-dimensional reconstructions, can also be useful for preoperative planning in children with early-onset Blount disease who present with recurrent deformities<sup>46</sup> (Figs. 5-A through 5-E). While advanced imaging techniques provide more detailed information than do plain radiographs, the additional cost, radiation exposure, and potential need for sedation and general anesthesia associated with some of these modalities should be considered.

### Management Options

Treatment is customized for each patient on the basis of a variety of factors, including the child's age, the magnitude of the deformity, the limb-length discrepancy, psychosocial factors, and the surgeon's training and experience<sup>37</sup>. On the basis of the results of the clinical examination and imaging studies, a



Fig. 2-C  
During the consolidation phase of realignment of the left-sided deformity, the posterior opening of the proximal fixator ring allows knee flexion.

list of current and anticipated deformities is created. Management options include observation with repeat clinical and radiographic examinations; use of long leg orthoses; and various surgical options including realignment osteotomy, lateral hemiepiphysodesis, and guided growth around the knee as well as gradual asymmetrical proximal tibial physal distraction, resection of a physal bar, and elevation of the medial tibial plateau.

### Orthoses

Several authors<sup>47-49</sup> have reported encouraging results with the use of knee-ankle-foot orthoses with a medial upright and drop-lock hinges to unload the medial compartment of the knee in children younger than thirty-six months of age with early-stage (Langenskiöld stage-I or II) Blount disease. The reported risk factors for failure of brace treatment include obesity with a weight greater than the 90th percentile, varus thrust, an age older than three years at the initiation of the brace treatment<sup>47</sup>, bilateral involvement<sup>48</sup>, and Langenskiöld stage-III or higher disease<sup>49</sup>. However, the retrospective case series in which orthoses were used for patients with Blount disease<sup>47-49</sup> have included multiple variables, not included a control group, involved use of various designs of orthoses and various regimens, and provided limited details regarding the actual time that the brace was worn.

Moreover, given the challenge of differentiating between cases of physiologic genu varum and mild Blount disease and issues of compliance with brace wear, the practice of prescribing long leg orthoses for obese children with possible early-onset Blount disease, while based on sound physiologic reasoning, has not been validated with enough scientific scrutiny to recommend its routine use (Table I). In a study in Japan, Shinohara et al.<sup>50</sup> followed twenty-nine patients (forty-six involved limbs) with early-onset Blount disease. The disease resolved without treatment in all twenty-two limbs that had demonstrated Langenskiöld stage-I changes and in eighteen of twenty-four limbs that had showed stage-II or III changes. Because of this high rate of spontaneous correction, these authors questioned the reported efficacy of brace treatment in young patients, including those with moderately advanced radiographic findings.

### Operative Management

To achieve a successful outcome, treatment must be individualized on the basis of a comprehensive analysis of the limb deformities, the amount of growth remaining, the psychosocial status of the patient, and the ability of the surgeon to execute a well-outlined treatment plan with precision and safety.

### Prognostic Factors

Several authors have reported recurrence rates of >50% following valgus osteotomies in children with early-onset Blount disease, with better outcomes occurring when operative realignment had been done prior to the age of four years<sup>13,51-54</sup> (Table I). Loder and Johnston<sup>13</sup> noted that the prevalence of poor results and recurrent deformities following tibial osteotomy for the treatment of early-onset Blount disease increased with increases in the Langenskiöld stage, an older age at the time of the osteotomy, and a lack of postoperative valgus overcorrection. At an average of six years and seven months postoperatively, eighteen (55%) of thirty-three tibiae had recurrence, with an overall satisfactory outcome in six of eight tibiae that had been operated on before the patient was four years old and in fourteen of twenty-five tibiae that had been operated on in older patients. On the basis of their findings on supine intraoperative radiographs, the authors recommended 5° to 10° of valgus overcorrection. Doyle et al.<sup>53</sup> found that, in a series of twenty-six tibiae that had undergone a valgus osteotomy, there was a recurrence in four of eleven children who had had operative realignment before the age of four years compared with nine of fifteen children who had had operative realignment when they were older. While there was no difference in recurrence rates based on the magnitude of the preoperative varus deformity, knees in which the changes were less than Langenskiöld stage III at the time of the osteotomy had a better outcome.

Schoenecker et al.<sup>54</sup> followed twenty-seven patients (forty-four tibiae) with early-onset Blount disease treated with a valgus tibial osteotomy and noted a satisfactory outcome in nineteen (83%) of twenty-three tibiae in which the osteotomy had been performed before the age of five years compared with eight (38%) of twenty-one tibiae treated in





Fig. 2-D



Fig. 2-E

Final clinical (Fig. 2-D) and radiographic (Fig. 2-E) appearance following bilateral staged tibial osteotomy with gradual correction. The mechanical axis has been restored to normal.

older children. Chotigavanichaya et al.<sup>52</sup> reviewed the results of closing wedge valgus tibial osteotomy in seventy-one patients (seventy-four tibiae) with Blount disease. Despite similarities with regard to the Langenskiöld stage, magnitude of preoperative varus deformity, and operative correction, the recurrence rate in children who had had the osteotomy before the age of four years (twelve of twenty-six; 46%) was lower than that in children who had had the procedure when they were older (forty-two of forty-eight; 88%). In a retrospective review of the results of acute correction in twenty-five patients with early-onset Blount disease, Ferriter and Shapiro<sup>51</sup> noted a high rate of recurrent deformity, requiring one to four additional osteotomies, in twenty-one (57%) of the thirty-seven involved tibiae. These authors identified massive obesity (a weight higher than the 97th percentile), a Langenskiöld stage of  $\geq$ III, and an age of more than 4.5 years at the time of the surgery as potential risk factors for recurrence following valgus osteotomy in these children. Hofmann et al.<sup>6</sup> followed twelve patients (nineteen limbs) who had been treated with a proximal tibial osteotomy for early-onset Blount disease. At an average age of 22.4 years, and following a mean of 2.5 operations per limb and four operations per patient, twelve knees were symptomatic and eight knees demonstrated degenera-

tive arthritis. The presence of symptoms in early adulthood and recurrent deformity strongly correlated with an advanced Langenskiöld stage of Blount disease ( $\geq$ IV) at the time of treatment.

#### *Perioperative Considerations*

A high prevalence of sleep apnea in morbidly obese patients with late-onset Blount disease has been noted recently. Gordon et al.<sup>55</sup> reported that eleven of eighteen patients with Blount disease who were older than nine years of age had sleep apnea, diagnosed with polysomnographic assessment, and required preoperative noninvasive positive-pressure ventilation. A high index of suspicion for sleep apnea is necessary, especially for adolescents with a history of snoring, so that appropriate measures are taken preoperatively.

The use of chemical prophylaxis for prevention of deep venous thrombosis and pulmonary embolism, especially in obese adolescents, should also be considered. In a recent presentation of the experience at a large children's hospital over a two-year period, it was noted that seven cases of deep venous thrombosis, including five cases of pulmonary embolism (one of which was fatal), had occurred following a variety of orthopaedic procedures performed for multiple diagnoses<sup>56</sup>.



Fig. 2-F



Fig. 2-G

Preoperative (Fig. 2-F) and postoperative (Fig. 2-G) lateral radiographs of the tibia show adequate correction of the proximal tibial procurvatum deformity.

While the total number of patients who had undergone orthopaedic surgery during the study period was not mentioned in the available abstract, all seven patients with deep venous thrombosis had a body-mass index  $\geq 25$  kg/m<sup>2</sup> and six of seven had a body-mass index  $>30$  kg/m<sup>2</sup>. The authors recommended that, in addition to the routine use of intermittent pneumatic compression devices for prophylaxis against deep venous thrombosis in patients over fourteen years of age who have immobilization of the limb after the operation, low-molecular-weight heparin should be given to adolescents with certain risk factors, including obesity, estrogen contraception, and a family history of thrombosis. Additional studies are needed to better define the role of chemical prophylaxis against deep venous thrombosis in adolescents undergoing lower-extremity surgery for the treatment of Blount disease.

### Hemiepiphyseodesis

Several authors of retrospective case series<sup>57-62</sup> have reported on hemiepiphyseodesis of the lateral aspect of the proximal part of the tibia and/or distal part of the femur, with mixed results. Since a hemiepiphyseodesis relies on the growth of the remaining healthy part of the physis for angular correction, given

the unhealthy medial aspect of the growth plate, achievement of limb realignment is not as predictable in children with Blount disease as it is with children with other disorders<sup>45,58,61</sup>. In a retrospective case series of hemiepiphyseodeses performed for correction of angular deformities about the knee related to a variety of causes, Castañeda et al.<sup>58</sup> reported an improvement of only 3° in patients with Blount disease compared with 19° in those with genu varum related to other causes. In a retrospective study of twenty-six children (thirty-three extremities) with late-onset tibia vara, Park et al.<sup>60</sup> reported the findings on full-length standing radiographs following nineteen proximal tibial and fourteen combined proximal tibial and distal femoral hemiepiphyseal stapling procedures performed when the patients were an average of 11.8 years of age. At a mean of 3.8 years postoperatively, radiographs demonstrated restoration of the mechanical axis to within the central half of the knee in twenty (61%) of the thirty-three extremities. Untoward events included residual limb-length discrepancy of  $>3$  cm (four patients), backing out of staples (five extremities), valgus overcorrection (two extremities), and transient peroneal neurapraxia (one extremity). The authors recommended hemiepiphyseal stapling in children younger than ten years old in whom the preoperative mechanical axis of the lower extremity is within the medial half of the medial compartment (mild varus) or medial to the knee joint but by a width no larger than the width of the medial compartment (moderate varus). Westberry et al.<sup>62</sup> performed lateral hemiepiphyseodesis in twenty-three patients (seven with early-onset and sixteen with late-onset Blount disease) and found more predictable correction in extremities with less preoperative varus angulation (median, 15°). Recently, McIntosh et al.<sup>63</sup> presented their experience with hemiepiphyseodeses in forty-nine patients (sixty-four extremities) with late-onset Blount disease. At an average of 3.3 years postoperatively, 66% of the patients had substantial residual medial mechanical axis deviation ( $>40$  mm). Factors associated with failure to obtain satisfactory alignment following a lateral hemiepiphyseodesis included a body-mass index  $>40$  kg/m<sup>2</sup>, a weight of  $>100$  kg, a preoperative mechanical axis deviation of  $>60$  mm, and a medial proximal tibial angle<sup>32</sup> of  $<76^\circ$ .

### Guided Growth

Stevens<sup>61</sup> popularized the concept of guided growth with use of a nonlocking titanium plate with screws placed extraperiosteally across the convex growth plate. A nonrigid extraperiosteal plate with two screws, one in the metaphysis and the other in the epiphysis, serves as a focal hinge at the perimeter of the physis (Figs. 6-A through 6-D). Once the mechanical axis of the limb has been restored or slightly overcorrected, the implant can be removed, with the anticipation of resumption of the previous growth rate at the involved physis. In the implant designer's original series of thirty-four consecutive patients who had undergone insertion of the tension band plate to correct a total of sixty-five deformities related to a variety of pathologic conditions (with five patients having Blount disease), the only two patients who were reported to have insufficient correction had



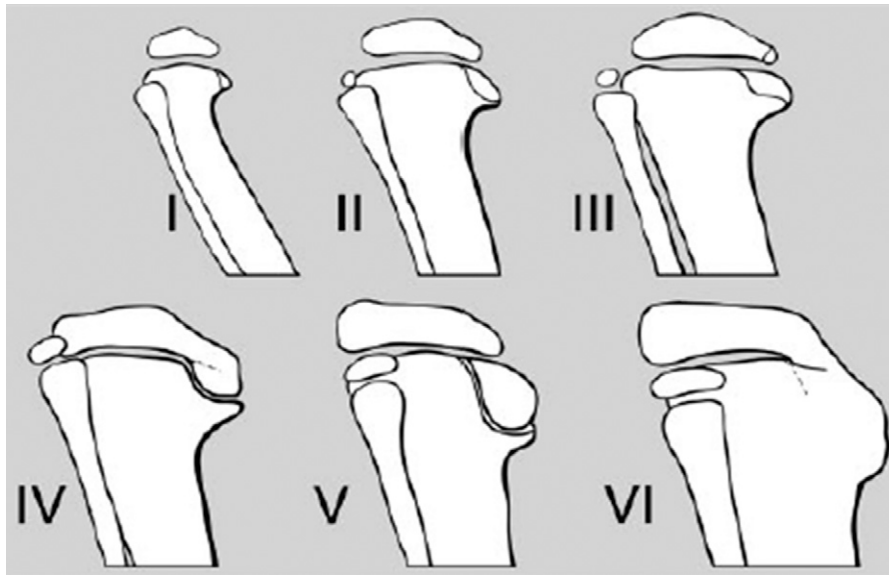


Fig. 3

Diagram of the six stages of progressive radiographic changes seen in early-onset Blount disease, as described by Langenskiöld<sup>2</sup>. (Reprinted, with permission, from: Schoenecker P, Luhmann S. Knee and leg: pediatric aspects. In: Koval KJ, editor. Orthopaedic Knowledge Update 7. Rosemont, Illinois: American Academy of Orthopaedic Surgeons; 2002. p 465-78.)

a diagnosis of late-onset Blount disease<sup>61</sup>. Moreover, the one reported hardware failure (screw loosening) was noted in a patient with Blount disease. In a recent presentation, Schroerlucke et al.<sup>64</sup> reported that five of their ten patients with late-onset Blount disease required revision of the tension band plate because of hardware failure. The average body-mass index of the patients with hardware failure was 37 kg/m<sup>2</sup> compared with 26 kg/m<sup>2</sup> in those with intact hardware. Schroerlucke et al. recommended use of a stronger implant in patients with a body-mass index of >31 kg/m<sup>2</sup>.

Although hemiepiphysodesis and guided-growth systems are relatively safe options with low overall morbidity rates and they allow rapid postoperative mobilization, these interventions do not address limb shortening and they require careful patient selection and vigilant follow-up. While it may be possible to correct multiplanar deformities with strategic placement of a staple or plate across the growth plate, to my knowledge no one has reported sagittal and axial deformity parameters following the use of hemiepiphysodesis or guided-growth implants for the treatment of Blount disease. Currently, hemiepiphysodeses and guided-growth systems may be considered for adolescents with late-onset Blount disease who have <15° of varus deformity, have at least two years of skeletal growth remaining, and have no more than 1 cm of limb shortening (Table II). While recently popularized nonlocking-plate designs may represent a technical improvement over staples, perhaps use of larger-diameter screws, stronger implant materials, noncannulated screws, and two adjacent plates in heavy adolescents may further diminish the prevalence of implant failure in this population. Whether the principles of guided growth can be applied safely and effectively in much younger children with early-onset Blount disease needs further investigation (Table I).

### Proximal Tibial Metaphyseal Osteotomy

#### Acute Correction with Conventional Techniques

Acute correction of angular and rotational deformity in Blount disease can be accomplished with a proximal tibial metaphyseal osteotomy. A variety of techniques have been advocated, including closing wedge<sup>13,65</sup>, opening wedge<sup>32,66</sup>, dome<sup>67,68</sup>, serrated<sup>69</sup>, and inclined<sup>70</sup> osteotomies. Furthermore, different fixation methods have been reported, including cast immobilization<sup>51,71</sup>, smooth pins and wires<sup>51</sup>, interfragmentary screws<sup>70</sup>, plates and screws<sup>65,66,72</sup>, and external fixators<sup>52,73-75</sup>. Several authors have reported on the use of monolateral<sup>73-76</sup> and circular<sup>67</sup> external fixators to maintain acute deformity correction with the potential for gradual lengthening at the osteotomy site in patients with >1.5 cm of limb-length discrepancy<sup>75</sup>. Overall, the choice of the osteotomy technique and fixation method should be based on several factors, including the patient's age and body habitus, the magnitude of the deformity, the presence of associated sagittal and axial plane deformities, and the surgeon's training and experience in safely executing acute realignment of the lower extremity.

Irrespective of the type of osteotomy and fixation device, there is a potential for neurologic injury and compartment syndrome with acute correction<sup>13,51,73,74,76-79</sup>. A prophylactic anterior compartment fasciotomy and insertion of a drain should be strongly considered for patients with Blount disease who are undergoing acute deformity correction. Also, vigilant postoperative management with frequent clinical examinations is warranted. However, despite the use of such measures, up to one-third of patients can have transient or permanent neurologic injury, which typically presents as weakness of the extensor hallucis longus<sup>13,51,70,73,74,76-79</sup>. Although there are several reports of encouraging short-term results after acute cor-

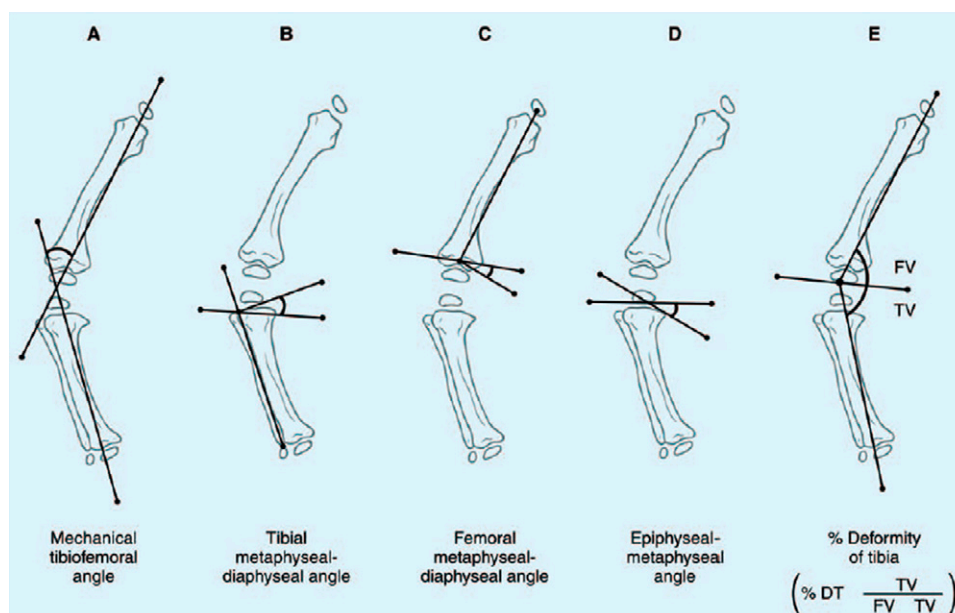


Fig. 4

Radiographic indices used in the evaluation of lower-extremity bowing in infants and young children. The mechanical tibiofemoral angle (A) is the angle between a line drawn from the center of the hip to the center of the knee and a line drawn from the center of the knee to the center of the ankle. The tibial metaphyseal-diaphyseal angle (B) is the angle between a line drawn through the most distal aspects of the medial and lateral beaks of the proximal tibial metaphysis and a line perpendicular to a line drawn along the lateral aspect of the tibial diaphysis. The femoral metaphyseal-diaphyseal angle (C) is created by a line drawn perpendicular to the anatomic axis of the femur and a line drawn parallel to the distal femoral physis. The epiphyseal-metaphyseal angle (D) is created by a line drawn through the proximal tibial physis, parallel to the base of the epiphyseal ossification center, and a line connecting the midpoint of the base of the epiphyseal ossification center with the most distal point on the medial beak of the proximal tibial metaphysis. The percentage deformity of the tibia, % DT (E), is calculated as the degree of tibial varus (the medial angle between the mechanical axis of the tibia and a line drawn parallel to the distal femoral condyles) divided by the total amount of limb varus (femoral varus [FV] + tibial varus [TV]). Femoral varus is represented by the medial angle between the mechanical axis of the femur and a line parallel to the distal femoral condyles. (Reprinted, with permission, from: Kayes KJ, Spiegel DA. Knee and leg: pediatrics. In: Vaccaro AR, editor. Orthopaedic knowledge update 8. Rosemont, Illinois: American Academy of Orthopaedic Surgeons; 2005. p 745-55.)

rection of tibial deformities in patients with Blount disease<sup>13,32,51,52,65,67-75</sup>, most of these studies had several limitations, such as inclusion of patients with early and late-onset Blount disease together with those who had other etiologies, a lack of consistent and reliable intraoperative and postoperative measurement of limb alignment, creation of iatrogenic translational deformities (Figs. 7-A through 7-D), the lack of set criteria for defining recurrence of the deformity, and short follow-up.

#### External Fixation with Gradual Correction

Gradual correction with distraction osteogenesis appears to be a safe and reliable means of treating multiplanar deformities, including limb-length discrepancy, even in obese patients<sup>80-84</sup>. The reported prevalences of neurovascular injury, compartment syndrome, and loss of correction following gradual correction of these deformities in patients with Blount disease have been substantially lower than the prevalences following acute deformity correction in Blount disease<sup>80-85</sup> (Table II). The average time with the external fixator in place has ranged from twelve to eighteen weeks<sup>80-85</sup>. The lack of acute neurovascular injuries in the reports on distraction osteogenesis for patients

with Blount disease is likely related to the avoidance of acute intraoperative traction on the neurovascular structures.

De Pablos et al.<sup>82</sup> performed a percutaneous osteotomy with progressive opening wedge correction using a modified Wagner monolateral fixator in ten patients (twenty tibiae) with late-onset Blount disease. Except for one case of under-correction, no major complications were noted. Coogan et al.<sup>81</sup> reviewed their experience with gradual correction with use of a circular external fixator in eight obese adolescents (twelve tibiae) with late-onset disease. One case of premature consolidation required a repeat osteotomy. Stanitski et al.<sup>84</sup> reported on the use of the Ilizarov circular external fixator with gradual correction in seventeen obese adolescents (twenty-five tibiae) with late-onset Blount disease. Despite one case each of delayed union and premature consolidation, alignment within 5° of normal was achieved in all patients. Alekberov et al.<sup>80</sup> reported on the use of the Ilizarov fixator in forty-five patients (sixty-nine tibiae) with early or late-onset Blount disease. Frontal and rotational deformity parameters were corrected in the majority of patients, with six tibiae requiring repeat osteotomies for treatment of residual deformity. Gordon et al.<sup>83</sup> advocated comprehensive treatment for late-onset Blount disease, in-

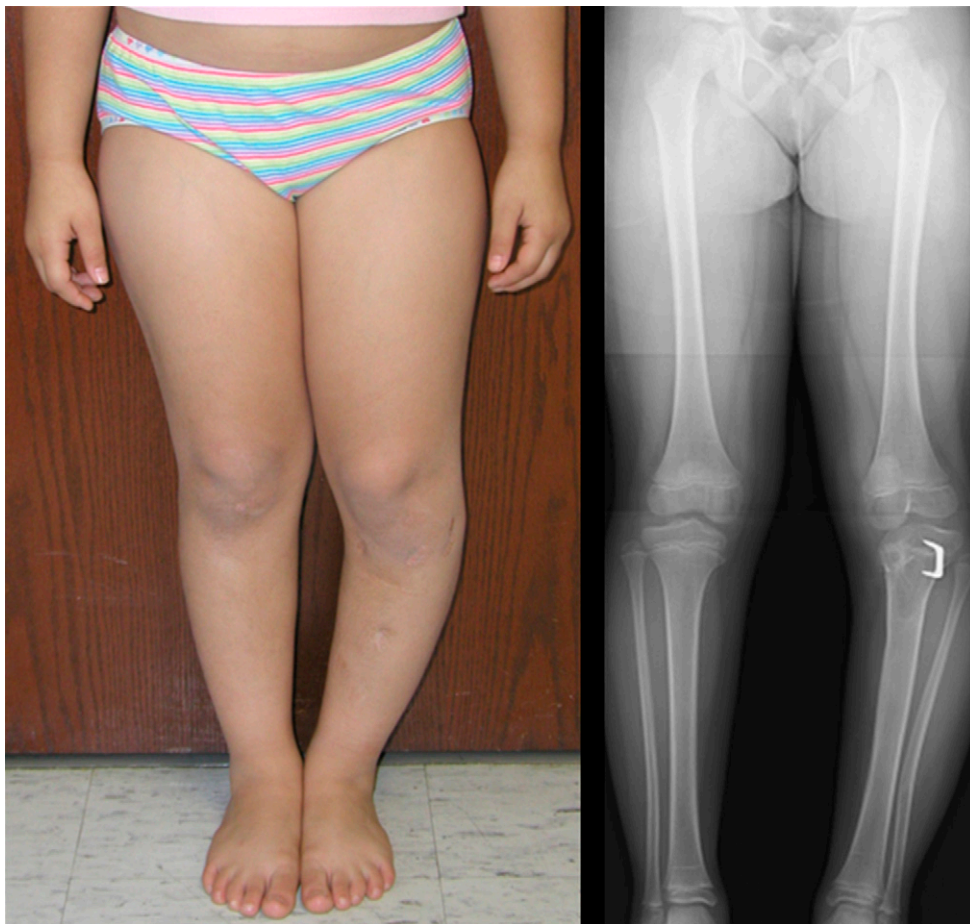


Fig. 5-A



Fig. 5-B

**Fig. 5-A** An eight-year-old girl with a recurrent deformity following a proximal tibial osteotomy done three years previously to treat left-sided early-onset Blount disease. (Figs. 5-A through 5-E reprinted, with permission, from: Sabharwal S. Blount's disease. In: Rozbruch SR, Ilizarov S, editors. Limb lengthening and reconstruction surgery. New York: Taylor and Francis; 2006. p 511-20.) **Fig. 5-B** A single extraperiosteal staple placed across the anterolateral portion of the proximal tibial growth plate failed to correct the deformity because of an osseous physeal bar, as seen on the computed tomography scan.



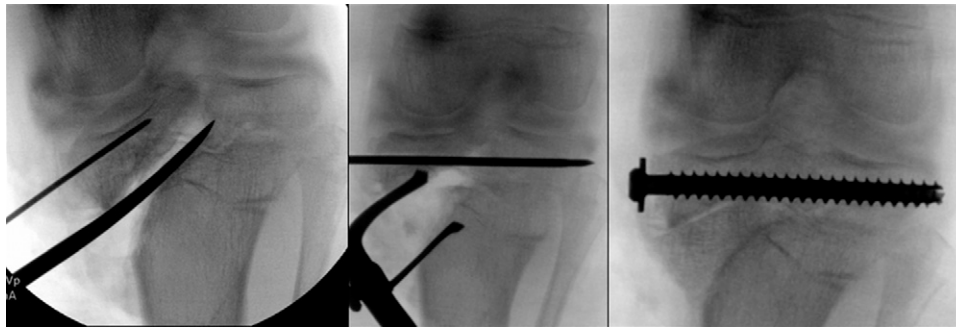


Fig. 5-C

Intraoperative fluoroscopic images demonstrate the technique of medial tibial plateau elevation with internal fixation and use of a structural allograft. Following exposure of the posteromedial portion of the proximal part of the tibia and placement of multiple anterior-to-posterior drill holes outlining the curved osteotomy site, a guidewire is inserted into the medial part of the tibial epiphysis. A curved osteotome is then utilized to complete the multiple-drill-hole osteotomy, hinging at the articular surface between the tibial spines. A lamina spreader is inserted posteromedially. Following complete elevation of the medial tibial plateau, the guidewire is advanced into the lateral part of the tibial epiphysis. A fully threaded cannulated screw with a washer is placed over the guidewire to stabilize the tibial plateau elevation. The posteromedial gap beneath the plateau is supported by a structural allograft that is stabilized by a second lag screw or a buttress plate (not shown in this image). A concomitant drill epiphyseodesis of the lateral aspects of the proximal tibial and fibular physes was performed.

cluding anatomic correction of distal femoral and proximal and distal tibial deformities. They reviewed the outcomes in a consecutive series of fifteen obese adolescents (nineteen limbs) who had undergone an osteotomy followed by gradual correction of the proximal tibial deformity with use of a circular external fixator. Thirteen of the extremities had associated distal femoral varus and also underwent either hemiepiphyseal stapling or acute correction with plate fixation, and eleven extremities underwent simultaneous correction of a distal tibial valgus deformity. At a mean of five years after treatment, satisfactory deformity correction and frontal plane alignment were maintained in all patients.

With the introduction of the Taylor Spatial Frame (Smith and Nephew, Memphis, Tennessee) and the ability to perform six-axis deformity correction on the basis of a computer-generated schedule, the well-trained clinician has the opportunity to correct multiplanar deformities with greater accuracy<sup>37,85-87</sup> (Figs. 2-A through 2-G). Feldman et al.<sup>85</sup> reported on nineteen obese patients (twenty-two tibiae), including six children (eight tibiae) with early-onset Blount disease and thirteen adolescents (fourteen tibiae) with late-onset Blount disease. On the basis of radiographic measurement of the mechanical axis, twenty-one (95%) of the twenty-two tibiae were considered to be corrected to within 3° of normal alignment with <0.5 cm of limb-length discrepancy.

The issue of whether to use acute or gradual correction in patients with angular deformities and limb-length discrepancy has been examined recently. Matsubara et al.<sup>88</sup> reviewed the cases of twenty-eight patients (thirty-four limbs) with lower-limb deformities and limb-length discrepancy related to multiple etiologies. The distraction index (number of days of use of the external fixator per centimeter gained) and the total duration of external fixation were substantially less for limbs treated with gradual correction of the deformity and limb-length discrepancy than they were for limbs in which the de-

formity had been treated with acute correction followed by gradual lengthening despite similar magnitudes of pretreatment deformity and shortening in the two groups. Feldman et al.<sup>89</sup> compared the accuracy of acute correction with that of gradual correction in thirty-two patients (thirty-two tibiae) with Blount disease. Despite similar age ranges and magnitudes of preoperative tibial deformities in the two groups, the fourteen patients who had undergone acute deformity correction with application of a monolateral fixator had greater postoperative residual mechanical axis deviation, sagittal plane angulation, translational deformity, and limb-length discrepancy than did the eighteen patients who had undergone gradual correction with a circular fixator. No neurovascular injury or compartment syndrome was noted in either group, although one patient with acute correction had a delayed union. The authors concluded that gradual correction was a more accurate method of correcting multiplanar deformities in patients with Blount disease. Nevertheless, whether correction of associated sagittal and rotational deformities in addition to the varus malalignment will lead to better long-term outcomes remains unknown.

With proper insertion techniques, the improved design of half-pins, and the use of hydroxyapatite coating, the prevalence of pin-site infections can likely be lowered, although not completely avoided. Other issues such as the psychosocial impact of using external fixators and concerns about the cosmetic appearance of pin sites should be discussed with the family preoperatively. However, a detailed discussion of this subject is beyond the scope of the current review<sup>37</sup>.

#### Postcorrection Assessment of Limb Alignment

There is no consensus in the literature regarding the ideal alignment of the lower extremity following operative reconstruction in a patient with Blount disease. Some have advocated normalization of the mechanical axis<sup>2</sup>, whereas others have thought that some degree of overcorrection should be



Fig. 5-D



Fig. 5-E

Clinical appearance (Fig. 5-D) and standing radiograph (Fig. 5-E) made six months postoperatively showed improved limb alignment and healing of the tibial plateau osteotomy site. The patient required a contralateral epiphyseodesis closer to skeletal maturity to equalize the limb lengths.

attempted<sup>52,71,90</sup>. A major limitation of the studies by these authors is that the operative correction was assessed on non-weight-bearing radiographs of the knee, often with an overlying cast, without the visualization of the entire femur and tibia that is required for performing a comprehensive analysis of multiplanar deformities in Blount disease<sup>4</sup>.

Accurate assessment of limb alignment following acute or gradual correction can be challenging. Several authors have used intraoperative fluoroscopy<sup>73-75,91</sup> with the electrocautery cord stretched across the skin overlying the center of the hip and ankle and visualization of the relationship of the cord to an anteroposterior view of the knee. Zhao and I<sup>91</sup> compared this supine fluoroscopic view with a standing full-length radiograph for the measurement of lower-limb alignment and found a linear relationship ( $r = 0.88$ ) between the two methods. However, the fluoroscopic method was less reliable for obese patients, especially those with residual ligamentous laxity and mechanical axis deviation.

Although full-length standing radiographs of the entire lower extremity can be utilized for patients undergoing gradual correction, a recent study showed that the assessment of lower-limb alignment in patients with an overlying circular external

fixator was not very reliable<sup>92</sup>. This inaccuracy is probably related to the patient's tendency to flex the knee and externally rotate the lower limb and have difficulty in maintaining a patella-forward position, especially when the patient is obese and has a bulky external fixator in place. The accuracy of assessment of lower-limb alignment when there is an overlying external fixator can be enhanced by paying attention to patient positioning for the standing radiograph, imaging one limb at a time, and supplementing the standing radiograph with information obtained from the clinical examination and full-length biplanar radiographs of the tibia<sup>92</sup>.

#### *Asymmetrical Physeal Distraction*

De Pablos and Franzreb<sup>93</sup> utilized asymmetrical physeal distraction with a modified Wagner monolateral fixator in twelve adolescents who had bilateral late-onset Blount disease. Two 6-mm half-pins were placed into the proximal tibial epiphysis and two pins were placed into the diaphysis; this was followed by gradual distraction without fibular osteotomy at a rate of 1.5 mm/day in two installments. A medial opening wedge correction at the site of the deformity achieved realignment of the tibia in all cases, with an average of 13° of angular cor-

**TABLE I Grades of Recommendation for Treatment of Early-Onset Blount Disease**

| Type of Treatment  | Grade of Recommendation* |
|--|--------------------------|
| Knee-ankle-foot orthosis   | I                        |
| Lateral proximal tibial hemiepiphysodesis/guided growth  | I                        |
| Valgus proximal tibial osteotomy prior to age of 4 yr  | B                        |
| Resection of physeal bar   | C                        |
| Elevation of medial plateau  | C                        |
| *A = good evidence (Level-I studies with consistent findings) for or against recommending intervention, B = fair evidence (Level-II or III studies with consistent findings) for or against recommending intervention, C = poor-quality evidence (Level-IV or V studies with consistent findings) for or against recommending intervention, and I = there is insufficient or conflicting evidence not allowing a recommendation for or against intervention. |                          |

rection of the varus deformity. The proximal tibial growth plate closed following distraction in all of these adolescent patients. There were no reported cases of septic arthritis or neurovascular injury, and the treatment was reportedly well tolerated. However, this technique has not gained popularity, possibly because of concerns about septic arthritis, pain during distraction, and premature closure of the proximal tibial growth plate (Table II).

#### Physeal Bar Resection

Medial proximal tibial epiphysolysis in children with advanced stages of early-onset Blount disease has been reported by a few authors<sup>94-96</sup> (Table I). Beck et al.<sup>96</sup> performed resection of a physeal bar at the medial aspect of the proximal part of the tibia with interposition of fat or silicone and a simultaneous valgus osteotomy in three young children with recurrent deformity and advanced (Langenskiöld stage-VI) early-onset Blount disease. None of these children were followed to skeletal maturity, but short-term follow-up at an average of twenty-two months demonstrated continued growth of the tibial physis and maintenance of alignment. Andrade and Johnston<sup>95</sup> recently reported on twenty-four patients (twenty-seven tibiae), five to ten years of age, who had undergone medial proximal tibial epiphysolyses with a valgus osteotomy for treatment of moderate-to-advanced early-onset Blount disease (a Langenskiöld stage of  $\geq$ III). The children who underwent the epiphysolysis before the age of seven years and had correction of varus malalignment with a concomitant osteotomy had a more favorable outcome, especially when they had not had prior surgical procedures. Given the availability of alternatives for older children, the authors did not recommend this procedure in children older than seven years of age<sup>95</sup>.

Unlike patients who have a posttraumatic bar in the periphery of the growth plate, children with Blount disease

do not usually have a discrete area of osseous tether that lends itself to operative excision. Moreover, given that children with Blount disease commonly have coexisting varus deformity and limb shortening that may require an osteotomy, an epiphysolysis as a stand-alone procedure has limited application in these patients.

#### Elevation of the Medial Plateau

In the advanced stages of early-onset Blount disease, the tibia can translate laterally with the medial femoral condyle falling into the posteromedial depression, causing a varus thrust in the stance phase of gait<sup>5</sup>. Physical examination typically demonstrates increased instability to varus stress with the knee flexed 20°, as compared with the instability at full extension, and this is related to secondary laxity of the medial collateral ligament. Elevation of the medial tibial plateau is recommended for the few children older than six years of age who have severe early-onset Blount disease (Langenskiöld stage V or VI) and substantial posterior depression of the medial tibial plateau (Figs. 5-A through 5-E) (Table I). Advanced imaging studies such as arthrography<sup>39</sup>, magnetic resonance imaging<sup>39</sup>, and three-dimensional computed tomography<sup>97</sup> can be very helpful for preoperative planning. Authors have described either an intraepiphyseal<sup>5,8,98</sup> or a transepiphyseal osteotomy<sup>65,90,97,99-101</sup> hinging at the articular cartilage of the intercondylar notch with insertion of structural bone graft to support the elevated medial tibial plateau. Care should be taken to simultaneously correct the posterior depression of the medial plateau by inserting a larger portion of the graft posteriorly. It is imperative to perform a lateral proximal tibial and fibular epiphysodesis at the same time to prevent recurrent deformity<sup>101</sup>. However, the epiphysodesis may lead to a change in limb length in a young child. The shortening may be addressed by an appropriately timed contralateral epiphysodesis<sup>101</sup>.

**TABLE II Grades of Recommendation for Treatment of Late-Onset Blount Disease**

| Type of Treatment  | Grade of Recommendation* |
|--|--------------------------|
| Lateral proximal tibial hemiepiphysodesis/guided growth  | C                        |
| Proximal tibial osteotomy with acute correction  | C                        |
| Proximal tibial osteotomy with gradual correction  | B                        |
| Asymmetrical physeal distraction   | I                        |
| *A = good evidence (Level-I studies with consistent findings) for or against recommending intervention, B = fair evidence (Level-II or III studies with consistent findings) for or against recommending intervention, C = poor-quality evidence (Level-IV or V studies with consistent findings) for or against recommending intervention, and I = there is insufficient or conflicting evidence not allowing a recommendation for or against intervention. |                          |





Fig. 6-A



Fig. 6-B

Preoperative photograph (Fig. 6-A) and radiograph (Fig. 6-B) of a thirteen-year-old girl who had late-onset Blount disease with distal femoral varus and proximal tibial deformity as well as 2 cm of limb shortening.



Fig. 6-C



Fig. 6-D

The patient underwent gradual correction with distraction osteogenesis at the proximal part of the tibia and application of an extra-periosteal plate across the lateral aspect of the distal femoral physis for guided growth. There was satisfactory restoration of limb length (Fig. 6-C) and alignment (Fig. 6-D) at skeletal maturity.



Fig. 7-A



Fig. 7-B



Fig. 7-C



Fig. 7-D

**Fig. 7-A** A forty-five-year-old woman who had undergone six prior surgical procedures for treatment of early-onset Blount disease presented with activity-related medial knee pain and 2 cm of limb shortening.

**Fig. 7-B** An anteroposterior radiograph demonstrates degenerative arthritis, primarily involving the medial compartment of the knee; residual varus malalignment; and iatrogenic translational deformity of the proximal part of the tibia.

**Fig. 7-C** The patient was deemed a poor candidate for knee arthroplasty and underwent gradual realignment with use of an external fixator. **Fig. 7-D** An early postoperative radiograph demonstrates appropriate lateral translation and realignment of the distal fragment. The knee symptoms had decreased substantially at the time of short-term follow-up, six months following removal of the fixator.



or a metaphyseal tibial lengthening, especially if there is a secondary metaphyseal tibial deformity<sup>65,97,99</sup>. The metaphyseal osteotomy, with or without lengthening, can be performed at the time of the plateau elevation<sup>65,90,99,101,102</sup> or in a staged fashion<sup>65,97</sup>.

### Long-Term Follow-up Studies

There are few long-term follow-up studies of patients with Blount disease<sup>6-8,103</sup>. Most of the long-term data on the natural history of Blount disease are derived from a Swedish national registry of a largely white nonobese population. Some authors from the United States<sup>13,19,51,104</sup> and Jamaica<sup>3</sup> have speculated that the clinical form of the disease seen in black children has a worse prognosis than that reported from Scandinavia<sup>8,94,103</sup>. Ingvarsson et al.<sup>103</sup> followed forty-nine patients (eighty-six affected knees) with early-onset Blount disease; thirty-eight knees had had no prior surgery. At an average age of thirty-eight years, eleven (13%) of the knees had arthritis, which was mild in nine of them. Of the eleven knees with arthritis, two had been treated nonoperatively and the remaining nine had undergone a variety of surgical interventions. In another study, Zayer<sup>8</sup> reported on eighty-six patients (133 affected knees) with early or juvenile-onset Blount disease. None of the patients who were younger than thirty years old at the time of follow-up had radiographic evidence of arthritis, whereas arthritis was noted in eleven (41%) of twenty-seven knees in the older patients. Although increasing varus deformity was associated with arthritic changes in the older patients, a constant relationship between varus malalignment and the severity of the osteoarthritis could not be established. In another long-term study, Ingvarsson et al.<sup>7</sup> reported on twenty-three patients (twenty-seven affected knees) with late-onset Blount disease; nine knees were treated nonoperatively. At the time of follow-up, when the average age was forty-seven years, fifteen (65%) of the patients reported no knee symptoms. Standing full-length radiographs were made for eighteen patients, and nine knees showed mild-to-moderate arthritis. Although arthritis was more common in the nonoperatively treated patients (seen in four of these eight patients) than it was in those treated with surgery (seen in five of fourteen), given the retrospective study

design and the small number of patients in both groups, firm recommendations could not be made.

### Overview

In summary, the goal of treatment of Blount disease is to attain a normally aligned lower extremity with normal joint orientation<sup>32,92</sup> and equal limb lengths at skeletal maturity. Observation with repeat clinical and radiographic examination is recommended for children under the age of two years, especially if the diagnosis is uncertain and the varus deformity is mild. Although the clinical effectiveness and feasibility of bracing are debatable, some advocate the use of a knee-ankle-foot orthosis for children younger than three years of age<sup>13,19,47</sup>. In addition to a proximal tibial metaphyseal osteotomy, with either acute or gradual correction, several realignment strategies are available for very specific indications. These procedures include a lateral hemiepiphysodesis and guided growth around the knee, a distal femoral osteotomy, elevation of the medial tibial plateau, resection of a physeal osseous bar, and gradual asymmetrical proximal tibial physeal distraction. Occasionally, two or more of these operative modalities are applied simultaneously or in a staged manner. Given the possibility of recurrent deformity, progressive limb-length discrepancy, and overcorrection, it is imperative to follow patients with Blount disease at least until skeletal maturity, irrespective of the age at onset and the treatment strategy. Further research is needed to delineate the etiology of Blount disease so that appropriate preventive measures as well as more predictable and less invasive means of management can be utilized in the future. ■

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