Radiographic Characteristics of Lower-Extremity Bowing in Children

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Lower-extremity bowing is common in infants and children and can result from a variety of conditions. At radiography, developmental bowing shows varus angulation centered at the knee, “metaphyseal beaking,” thickening of the medial tibial cortices, and tilted ankle joints. Tibia vara (Blount disease) demonstrates genu varum and depression of the proximal tibia medially. Congenital bowing manifests as posteromedial bowing with cortical thickening along the concavity of the curvature and, in some cases, diaphyseal broadening. In rickets, radiographic changes occur primarily at sites of rapid growth and are predominantly metaphyseal, with widening of the zone of provisional calcification. Achondroplasia is characterized by shortening and thickening of the long bones with metaphyseal flaring and cupping. In neurofibromatosis, there may be anterolateral bowing of the tibia, and there is often focal narrowing and intramedullary sclerosis or cystic change at the apex of the angulation. The tibia is typically involved at the junction of the middle and distal thirds. Osteogenesis imperfecta demonstrates bowing from softening due to osteoporosis and multiple fractures and typically involves the entire skeleton. In camptomelic dysplasia, lower-extremity bowing is associated with a short trunk, short limbs, and deficiencies in pelvic bone development. Recognition of these pathologic conditions is important for differentiating those that will resolve spontaneously from those that require surgery or other treatment.

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Introduction

Bowing of the lower extremities is common and is a frequent cause of orthopedic referral (1). The parents of children with bowlegs often want to know if the legs are abnormal. The role of the physician is to determine if the bowing is physiologic or pathologic. In this article, we discuss and illustrate the clinical and radiographic characteristics of various common and uncommon causes of lower-extremity bowing in children, including developmental bowing, congenital bowing, tibia vara (Blount disease) (Figs 1, 2), neurofibromatosis, osteogenesis imperfecta, rickets, camptomelic dysplasia, and achondroplasia.

Causes of Lower-Extremity Bowing

Developmental (“Physiologic”) Bowing

Developmental (physiologic) bowing is a common condition that causes exaggeration of normal age-related angulation changes at the knee joint. Neonates and infants normally have varus angulation of the lower extremities that is believed to be secondary to in utero molding. Gradual correction of this angulation begins as a child starts walking. The bowing is corrected within 6 months of walking or by 18–24 months of age. After this age, there is normally a change to valgus angulation during the 2nd and 3rd years of life that reverts to the adult pattern by age 6 or 7 years (Fig 3). Thus, any varus angulation at the knee joint after the age of 2 years is abnormal.

Exaggerated varus angulation during the 2nd year of life is deemed to be developmental (physiologic) bowing. This condition is typically seen in children who begin walking at an early age and is more common in heavier children as well as in African-American children (1,3). Radiography of developmental bowing shows varus angulation centered at the knee, which is measured by drawing lines parallel to the midshafts of the femur and tibia on a standing anteroposterior radiograph and calculating the angle of intersection. There is mild enlargement and depression of the proximal tibial metaphyses posteromedially without fragmentation (“metaphyseal beaking”). The medial tibial cortices are thickened secondary to buttressing, and the ankle joints are tilted with the medial side higher (Fig 4). Similar but less striking changes may be seen in the femurs (3). Developmental bowing does not require treatment, but follow-up is needed to ensure that the bowing resolves and that tibia vara does not develop.
Figure 3. Graph illustrates how normal varus angulation before 2 years of age changes to valgus angulation after 2 years of age (arrow). (Reprinted, with permission, from reference 2.)

Figure 4. Physiologic bowing in a 16-month-old boy. (a) Initial radiograph demonstrates physiologic bowing. (b) Follow-up radiograph obtained 7 months later shows mild metaphyseal beaking with improvement over time.
Congenital Bowing

Congenital bowing of the tibia is an unusual condition that is believed to result from an abnormal intrauterine position, although localized skeletal dysplasia or fetal vascular insufficiency may also play a role in some cases. This type of tibial bowing is usually convex posteriorly and medially (Fig 5); less commonly, it is convex laterally. The fibula is also bowed. The foot shows marked dorsiflexion at birth. At radiography, there is cortical thickening along the concavity of the curvature, and there may be diaphyseal broadening. There is a good prognosis for remodeling during growth, but protective bracing may be necessary in some cases. Leg-length discrepancy secondary and directly proportional to bowing of the tibial-fibular segment is usually seen (4,5) and may require osteotomies and leg-lengthening procedures. Epiphysiodesis of the contralateral side may also be performed.

Tibia Vara (Blount Disease)

Tibia vara, or Blount disease, is a common condition that is believed to result from abnormal stress on the posteromedial proximal tibial physis, causing growth suppression. The growth at the epiphysis becomes asymmetric, leading to the typical varus angulation. Predisposing factors for this disease are early walking, obesity, and African-American descent. Obesity and early walking exaggerate the physiologic varus angulation. African-American children are believed to have excessive ligamentous laxity and begin to walk earlier than their Caucasian counterparts. Early diagnosis and treatment of this disease is vital to avoid progressive worsening.

Three major types of tibia vara have been recognized, depending on age at presentation: infantile, juvenile, and adolescent. Infantile type tibia vara is the most common. The later-onset types may represent an unrecognized or untreated variant of the infantile form (1).

Tibia vara is best diagnosed with standing anteroposterior radiography of both legs. The radiographs will demonstrate genu varum, measured as described earlier, and abnormality of the proximal tibia consisting of depression and irregularity or fragmentation of the metaphysis posteromedially and deficiency of the epiphysis medially. The physis may be widened medially due to arrested growth or laterally due to traction injury. More advanced cases will show lateral subluxation of the tibia, and genu recurvatum may also be seen.

### Table: Metaphyseal-Diaphyseal Angle Measurements for Physiologic Bowing and Tibia Vara

<table>
<thead>
<tr>
<th>Patient Age (mo)</th>
<th>Metaphyseal-Diaphyseal Angle (degrees)</th>
<th>Average</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physiologic bowing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11–20</td>
<td>5.1 ± 2.8</td>
<td>0–11</td>
<td></td>
</tr>
<tr>
<td>21–30</td>
<td>3.7 ± 3.1</td>
<td>0–10</td>
<td></td>
</tr>
<tr>
<td>Tibia vara</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11–20</td>
<td>18.4 ± 4.3</td>
<td>8–22</td>
<td></td>
</tr>
<tr>
<td>21–30</td>
<td>13.7 ± 4.3</td>
<td>7–22</td>
<td></td>
</tr>
</tbody>
</table>

Source.—Reference 6.
Note.—Angulation greater than 11° indicates tibia vara.
Whereas developmental bowing is typically symmetric, Blount disease is usually unilateral or asymmetric. Measurement of the metaphyseal-diaphyseal angle has been used to more accurately diagnose this condition and to differentiate it from developmental bowing (Fig 6). The metaphyseal-diaphyseal angle is the angle between a line drawn parallel to the top of the proximal tibial metaphysis and another line drawn perpendicular to the long axis of the tibial shaft. In physiologic bowing, this angle is 5° ± 2.8 (range, 0°–11°). In Blount disease, the angle is 16° ± 4.3 (range, 8°–22°). It has been suggested that children with angles greater than 11° have Blount disease (Table). Children with indeterminate angles (8°–11°) may require follow-up radiography to clarify the diagnosis (6).

A six-stage classification system for Blount disease, first proposed by Langenskiold in 1952, is still the system most commonly used by orthopedic surgeons (Fig 7). Langenskiold himself cautioned that this classification system was not intended to help determine prognosis or treatment. However, surgery consisting of osteotomies of the tibia and fibula is the usual treatment of choice in stage 3 disease or higher (7). Bowing can recur

Figure 6. Infantile Blount disease in a 23-month-old boy. (a) Radiograph shows asymmetric depression of the proximal tibial metaphyses, particularly on the affected left side. (b) Radiograph obtained 7 months later demonstrates increased metaphyseal depression bilaterally and fragmentation of the medial tibial metaphysis on the left side. Black lines illustrate abnormally increased metaphyseal-diaphyseal angles measuring 13° and 26° on the right and left sides, respectively.

Figure 7. Drawings illustrate the six stages of the Langenskiold classification system for Blount disease. Note the progressive depression, beaking, and fragmentation of the medial tibial metaphysis, also involving the epiphysis in the higher stages. (Reprinted, with permission, from reference 7.)
postoperatively, requiring repeat surgery. Magnetic resonance (MR) imaging (Fig 8) has been performed to evaluate the growth plate in Blount disease and may be useful in surgical planning (8,9). It may also be used to predict development of Blount disease in patients with severe physiologic bowing (10).

**Neurofibromatosis**

Neurofibromatosis is a common genetic disorder. In addition to cutaneous, nervous system, and ocular abnormalities, osseous lesions are seen in neurofibromatosis type 1. These osseous lesions affect the skull, spine, ribs, pelvis, and long bones. There may be anterolateral bowing of the tibia with or without a hypoplastic fibula, and there is often focal narrowing and intramedullary sclerosis or cystic change at the apex of the angulation, a finding that is consistent with hamartomatous fibrous tissue. The tibia is typically involved at the junction of the middle and distal thirds (Fig 9). The underlying mechanism is mesodermal dysplasia. Pathologic fracture with nonunion may result in pseudarthrosis of the tibia and sometimes of the fibula, with pencil pointing of the bone fragments. When dysplasia without fracture is diagnosed, prophylactic bracing may be used to prevent development of pseudarthrosis. Once fracture has occurred, treatment consists of osteotomy with grafting with the goal of achieving union (11). The precise mechanism for defective healing and pseudarthrosis is not known. Distal to the pseudarthrosis, disuse osteoporosis and secondary deformities of the talus and calcaneus are seen. Aberrations in limb growth can also be seen in neurofibromatosis (12,13).
Osteogenesis Imperfecta

Osteogenesis imperfecta is one of the more common heritable disorders of the connective tissue. Traditionally, four major types of osteogenesis imperfecta have been described, each with a different mode of inheritance depending on the basis of the specific collagen defect. Type 1 is characterized by fractures of varying severity, blue sclerae, abnormal tooth development, and wormian bones in the skull. Type 1 osteogenesis imperfecta comprises most of the cases previously classified as osteogenesis imperfecta tarda. Types 2 and 3 are characterized by severe skeletal involvement and poor postnatal survival rates. These cases may correspond to the cases previously classified as osteogenesis congenita. Patients with type 2 or 3 have blue sclerae and fractures at birth or even in utero. Type 4 exhibits normal sclerae and variable skeletal findings.

With advances in genetics, many specialists now believe that this classification system is too narrow and misleading and that osteogenesis imperfecta is actually a more complex abnormality that is more accurately characterized in terms of clinical findings. In all four types of osteogenesis imperfecta, bowing of the long bones results from softening caused by osteoporosis and multiple fractures. Bowing typically involves all the long bones (Fig 10) (14), and osteotomies and pinning are commonly performed for bowing. Recently, some success has been achieved with medical treatment with bisphosphonates (15). However, although the density of the bones increases and clinical improvement is seen, fractures continue to occur (16).

Rickets

Rickets results from deficient mineralization of normal osteoid and interruption of the normal orderly development and mineralization of growth plates. Vitamin D–resistant (hypophosphatemic) and nutritional rickets are the most common types. The bones are soft, with consequent bowing of long bones on weight bearing. However, the predominant changes are metaphyseal, with widening of the zone of provisional calcification due to the presence of unmineralized osteoid. Cupping, fraying, and splaying of metaphyses occurs with growth and continued weight bearing.

Radiologic changes in rickets occur predominantly at sites of rapid growth, including the proximal humerus, distal radius, and distal femur and both ends of the tibia. Thus, a skeletal survey for rickets can be accomplished with anteroposterior radiography of the knees, wrists, and ankles. The radiologic appearance of rickets varies somewhat depending on the cause of the disease. For example, in renal rickets, the metaphyseal changes are usually less severe, varying with the
growth rate of the child (Fig 11). Renal rickets is usually seen in older children, whereas nutritional rickets is more common in infants (17–19). Treatment is dietary and medical unless the bowing is severe, in which case valgus osteotomies may be performed.

**Camptomelic Dysplasia**

Camptomelic dysplasia is a rare inherited disorder that is often fatal in infancy. In newborns, it leads to a large head, a short trunk, and short, bowed limbs. There is angular anterolateral bowing of the limbs that is most marked in the legs, as well as involvement of both femurs and tibias (Fig 12). The hips are frequently dislocated. Clubbed feet and joint contractures may be seen. There is bowing of the forearm bones with small hands. The pelvis will show absence of sacral alae, narrow iliac bones, acetabular hypoplasia, and poor pubic ossification as well as widely separated, short ischial bones. The chest is bell shaped with thin, wavy ribs and slender clavicles. The tracheal caliber is narrow owing to defective tracheobronchial cartilage, and characteristic vertebral anomalies are present. Camptomelic dysplasia is classified as a bent-bone dysplasia (20). Treatment is usually supportive.

**Achondroplasia**

Achondroplasia is the most common form of short-limbed dwarfism. It is most often autosomal dominant in transmission but with a high rate of spontaneous mutation. Achondroplasia is characterized by shortening and thickening of the long bones with metaphyseal flaring and cupping. The lower extremities are bowed, and treatment of severe bowing requires valgus osteotomies. The phalanges are short, broad, and cupped. The iliac bones are short and rectangular with narrow sacrosciatic notches and short, wide pubic and
ischial bones (Fig 13). There are bullet-shaped vertebral bodies with posterior scalloping and narrowing of lumbar interpedicular distances. The intervertebral disks are widened, resulting in normal trunk length, and the thorax is slender due to the short ribs with flared anterior ends. The head is large, with frontal bossing and a depressed nasal bridge (1).

Surgical Correction of Bowing

Surgical treatment of bowing depends on the age of the patient and the cause and stage of the condition (21). The goal of treatment is restoration of satisfactory mechanical alignment. Osteotomies are often performed for Blount disease (Figs 14, 15), achondroplasia, vitamin D–resistant rickets, and osteogenesis imperfecta. When reviewing postoperative studies, the radiologist should look for recurrence of bowing, which can be varus or valgus in direction. Subsequent asymmetric growth plate closure can also affect surgical results.

Figure 13. Achondroplasia in a 6-year-old boy. Radiograph shows short bones, metaphyseal flaring, and genu varum. The acetabula are squared, and the sacrosciatic notches are narrowed.

Figure 14. Blount disease in a 3-year-old girl who was treated with proximal tibial osteotomies. (a) Preoperative radiograph demonstrates bilateral metaphyseal deformity and marked left-sided metaphyseal depression and varus angulation. (b) Radiograph obtained 5 months after surgery shows improved angulation.
Conclusions

Bowing of the lower extremities is present in a wide variety of pathologic conditions. Recognition of these conditions is important for differentiating those that will resolve spontaneously from those that require surgery or other treatment.

References