Blount's Disease

Abstract

Blount's disease is a pathology of the proximal tibial epiphysis causing outward tibial bowing in children. Physical examinations as well as several imaging modalities are used to diagnose Blount's disease and differentiate from conditions with similar appearances. The Langenskiöld 6-stage classification system, based on the progressive degeneration of the medial joint compartment, is used to assess the severity of the disease. Treatment options include non-surgical and surgical techniques, with specific care hinging on patient age and stage in the Langenskiöld system. Permanent joint damage and deformity can be sustained if left untreated. In order to perform accurate and timely diagnoses, knowledge and understanding of the subtle indications of Blount's disease is indispensable.

Introduction

The growth plates in developing infants and children are crucial sites that, when affected by disease or negative environmental factors, greatly impact the growth and structure of maturing skeletons. Blount's disease (infantile tibia vara), is one such pathology causing outward bowing of the leg in infants and adolescents through damage or displacement of the proximal tibial epiphysis and metaphysis.[1] There are two types of Blount's disease, the primary difference being the age of occurrence: infantile Blount's disease occurring before the age of 4 years and adolescent occurring after the age of 4 years.[2] Multiple factors such as ethnicity, genetics, and mechanical stress are thought to be contributing elements to this disease. Diagnostic radiography is the method of choice in diagnosing Blount's disease in children, and common radiographic manifestations include a rounded, dome-like tibial metaphysis with fracturing or changes in size of the epiphysis.[1] During the earliest stages of infantile Blount's disease, bracing has been utilized in an attempt to correct the varus angle of the proximal tibia. However, a majority of patients require surgical intervention.

Although the clinical and radiographic signs of Blount's disease are commonly known, a previously unreported variant of the disease has been discovered: infero-medial slippage of the tibial epiphysis without change in the size or shape of the growth plate. The multi-faceted nature of Blount's disease warrants the need for continual education and the use of numerous modalities to recognize and treat this disease.

Anatomy of the Adolescent Knee

The adolescent knee differs from a fully-matured joint in that it contains unossified growth plates. One of the first to appear, the proximal tibial epiphysis may be present in some full-term newborns.[3] This secondary site of ossification allows longitudinal growth of the tibia to continue until the epiphyseal plate fuses at about 25 years of age.[3] The metaphysis is the wide, flared portion of the tibia adjacent to the epiphyseal plate where growth in bone length actually occurs. When an unequal distribution of force is continuously applied to this sensitive region of growth, compression or fragmentation of the tibial epiphysis and metaphysis can result.

Throughout childhood, the knee joint compensates for the rapid growth and varying stresses placed on it. At birth, a tibio-femoral angle of 15° varus is common.[4] This angle is formed by the intersection of the long axis of the femur and the tibia (see **Figure 1**). As a child grows, the varus angle decreases, reaching a neutral point at 14-22 months of age. From there, the knee becomes increasingly valgus, peaking at an angle of 10° at the age of 3-4 years before it stabilizes to an adult level of approximately 6° valgus at the age of 6 or 7 years.[4,5] Since young children normally have some degree of physiological varus bowing, it can be difficult to differentiate normal tibia vara from early Blount's disease.

Signs and Symptoms

Children affected by Blount's disease rarely show any signs or symptoms of the disorder before they begin walking. However, after ambulation, most affected children are examined because of a bowleg deformity that worsens as they grow and bear more weight. The varus angulation is more focused in the proximal tibia rather than in the knee itself. Bilateral presentation is most common, especially in infants, yet it is not unusual to have unilaterally-affected limbs.[5]

Risk Factors for Blount's Disease

The exact etiology of Blount's disease remains unclear. Multiple factors are thought to be contributing elements to this disease:

- Ethnicity or race; rates are higher for children of African origin.
- Gender; more common in females.
- Genetics.
- Early walkers (less than 1 year of age).
- Mechanical stress.
- Obesity or increased body mass index (BMI). [1,2,4-6]

More diagnoses of Blount's disease are made in children who begin walking early and in those who have increased BMI. According to Ferguson et al[4], "In early walkers the physiological varus has not yet resolved by the time they start to weight-bear, so the mechanical axis will pass medial to the knee. This may result in excess loading of the medial tibial physis." (p.31) An undue pressure on the medial portion of the knee leads to disruption of ossification and chondrocyte function which can instigate damage. Early walking can cause undue stress on the tibial physis, but obesity can also be the source of excessive loading. For susceptible individuals, obesity greatly increases medial compartment pressure and contributes to the development of Blount's disease.[1,2] As the prevalence of obesity has risen, so too has the incidence of Blount's disease.[2]

Typical Presentation and Detection

Blount's disease can be difficult to diagnose due to the similarities it shares with other pathologies causing outward bowing of the lower limb. Fibular hemimelia, rickets, various skeletal dysplasias, and normal physiological bowing are several disorders commonly mistaken for Blount's disease.[4] Presently, physical examinations along with diagnostic x-ray and magnetic resonance imaging are used synergistically to detect changes in the proximal tibia typical of Blount's disease.

Physical

Several factors are physically evident in patients with Blount's disease. These include a visible varus angle and lateral concave bending of the knee and tibia. In clinical assessment, proper positioning of the patient is essential in diagnosing Blount's disease. Toddlers tend to stand with externally rotated legs and slightly flexed hips and knees.[4] This posture gives the impression of outward tibial bowing and therefore may result in a false reading of the varus angulation. Achieving a true impression of the measurement requires rotating the hips until the patella is facing forward and the knee is fully extended. Serial clinical photography may be used in determining and documenting the severity of tibial bowing.

Radiography

Diagnostic radiography is the main tool for accurately diagnosing Blount's disease. Many changes in anatomy can be visualized on an anterior projection of the lower limb. Langenskiöld classified these changes into 6 different stages or types of Blount's disease that progress over time (see **Figure 2**).[4,5,7] Each stage has distinct radiographic characteristics:

- Type I: Medial metaphyseal beaking.
- Type II: Saucer shaped defect of medial metaphysis.
- Type III: Saucer deepens into a step.
- Type IV: Epiphysis bent down over medial beak.
- Type V: Double epiphysis.
- Type VI: Development of medial physeal bony bar.[4]

These six stages have again been divided into low-grade Blount's disease, type I-IV, and high-grade Blount's disease, type V and VI. Furthermore, in a study of 20 children with high-grade tibia vara, Khanfour[5] found that the Langenskiöld system is reliable and reproducible in determining a prognosis for late onset Blount's disease.

Radiography is not only used to visualize epiphyseal changes, but also to determine precise degrees of angulation in the lower limb. These measurements can then shape treatment plans and establish baseline comparisons for future reference. One method of determining the presence and severity of tibia vara is to measure the angle formed between the long axis of the femur and tibia, the tibio-femoral angle, on a radiograph. Another angle shown to help distinguish between normal tibial bowing and Blount's disease is the tibial metaphyseal-diaphyseal angle, TMDA (see **Figure 3**). "This is calculated by the angle subtended by a line perpendicular to the long axis of the tibia and a line connecting the most prominent beaking on the medial and lateral sides of the proximal metaphysis."[4(p.32)] This measurement has been found to play an important role in diagnosing Blount's disease: if the TMDA is less than 11°, there is a 95% chance that the tibial bowing is physiological, but if the angle is greater than 11°, Blount's disease is more likely.[4]

MRI

"The knee is the most often evaluated portion of the lower limb, and MRI is invaluable in detecting ligament damage or meniscal tears of the joint capsule."[3(p.222)] In a retrospective analysis by Sabharwal et al[6], the intra-articular variations of the menisci, articular surface, and proximal tibial epiphysis in children with Blount's disease were studied to quantify any abnormal changes when compared with a control. The MRI scans of 26 children with Blount's disease were analyzed along with those in a control group of 20 children. The height and width of the menisci, the thickness of the unossified tibial epiphysis, and the presence of tears in the menisci were among the measurements recorded for both groups. The height and width of the medial menisci were found to be significantly greater in affected limbs and strongly correlated to the BMI of the patient.[6] The unossified proximal tibial cartilage was thicker and more medial meniscal tears were discovered in children with Blount's disease compared with those in the control. Neither group presented with abnormalities in the lateral compartment of the knee joint.

The various clinical and radiographic signs of Blount's disease are relatively well-known. However, some characteristics of this disorder are not visualized well by radiographs or physical examination alone. The MRI analysis by Sabharwal et al[6] confirmed the existence of some controversial signs of Blount's disease: enlarged height and width of the medial menisci, greater thickness of unossified proximal tibial cartilage, and increased prevalence of medial meniscal tears. A positive correlation, not previously evaluated, between the intra-articular changes and the BMI of the patient has also been established.[6]

Disc Slippage Case Reports

Three cases of tibia vara were studied by Sanghrajka et al[1] in which each child presented with bowing of the leg due to infero-medial slippage of the proximal tibial epiphysis. Although different ages, each patient was in the 97th percentile for their weight. Radiographs confirmed the medial displacement of the epiphyses and the resulting gap on the lateral edge. Corrective surgical procedures using external fixation hardware were performed to straighten the leg and prevent further slippage of the epiphyses. Follow-up radiographs show the effectiveness of treatment as the varus angle was corrected, the slippage reversed, and the space between the epiphysis and metaphysis opened. These three cases are unique in that a slippage of the growth plate without fragmenting or compression has not yet been associated with Blount's disease.[1]

This previously unreported variation of the disease, infero-medial slippage of the tibial epiphysis with no change in the size or shape of the growth plate, warrants treatment quite different from characteristic Blount's disease. A link has also been hypothesized between this slippage and excessive stresses on the growth plate due to obesity. These findings are significant since early detection of epiphyseal slippage can potentially be treated with corrective tensile forces.[1] As the rate of childhood obesity increases, it is likely that the incidence of this variation will also increase. Therefore, early recognition is essential for the appropriate medical care and corrective action to occur.

Treatment

The purpose and goal in treating Blount's disease is the unloading of the medial tibial growth plate to prevent multi-planar deformity and degenerative arthritis later on.[8] There exist several different treatment options divided into two main categories: non-surgical and surgical. The type of treatment and its probability of success can be predicted by the age of the patient and what stage the affected joint falls under in the Langenskiöld system. Non-operative techniques are often used for treating stages I and II in children less than 3 years of age, while stages III-VI in children older than 3 years (and those who failed non-surgical treatment), commonly require intraoperative procedures.[5] According to Ferguson et al[4], in Langenskiöld's study following treatment, "Restoration to normal was common in stages I and II and possible in stages III and IV. However, stages V and VI were associated with recurrent deformity and often permanent sequelae."[(p.33)]

Non-Surgical

During the earliest stages of infantile Blount's disease, bracing has been utilized to attempt to correct the varus angle of the proximal tibia. Children under the age of 3 are prime candidates for bracing since physiological bowing is difficult to distinguish from early Blount's disease and unnecessary operations should be avoided. A knee ankle foot orthosis (KAFO) is a common method of bracing. This type of support fixes the knee in extension and contains a medial bar that applies valgus pressure to the joint.[4,9] The medial joint line is then relieved of excess pressure and normal growth along the epiphysis will hopefully resume. For best results, it is recommended that the brace be worn 23 hours a day for approximately 2 years depending on the degree of varus angulation.[4,5]

The actual effectiveness of this method is controversial however. Bracing is not successful for every patient. Severe obesity, bilateral deformity, and age greater than 3 years are factors found to increase the rate of brace failure.[5,9] Additionally, a lack of scientifically-founded results supporting the efficacy of bracing has caused many physicians to disregard this method. A retrospective observation by Shinohara et al[10] discovered that in a study of 46 limbs left untreated for Blount's disease stages I-III, 40 limbs resolved spontaneously. Recovery time was also found to be no sooner for patients using a brace than for those without.[10] Critics of bracing believe that patients who responded positively to non-surgical methods would have likely improved without it.

Surgical

There are some cases in which spontaneous resolving of the deformity is very improbable and surgical measures are necessary. Children older than 3 years with advanced-stage Blount's disease are typical recipients of operative procedures. A somewhat uncommon practice, lateral hemiepiphysiodesis is a surgical technique used to direct physeal growth through manipulation of the growth plate.[9] With a drill or curette, the surgeon permanently ablates the lateral portion of the epiphyseal plate, thereby halting growth of the bone in that compartment. This technique relies on the potential for further medial growth to correct the varus angulation. However, because growth is inhibited, hemiepiphysiodesis is only recommended for children close to skeletal maturity. A variation of this method for younger children involves placing staples, screws, or non-locking plates across the lateral physis with the potential for growth resumption once removed.[9]

The preferred surgical procedure to correct tibia vara caused by Blount's disease is a valgus proximal tibial osteotomy. A wedge of bone is excised from the tibia allowing the limb to return to physiological alignment. Several different osteotomy techniques have been used with success: opening wedge, closing wedge, dome, inclined, and serrated osteotomies.[9] The primary difference between these methods is the shape and location of the bone excision. In addition, fixation by cast, pins and wires, plates and screws, external fixators, or any combination of these can be utilized depending on the patient's unique presentation. According to Ferguson et al[4], "the method employed seems less important than attaining the goal of adequate correction of the mechanical axis."[(p.33)]

Recurrence

An osteotomy with an excessive valgus angulation of 15° is favored by many surgeons as it is thought to prevent recurrence, minimize the compression across the physis, and allow time for normal growth to start again.[11] However, recurrence of Blount's disease, even with excessive valgus angulation, still occurs and in some instances causes deformity in patients.[11] A group of 38 patients with second stage Blount's disease who had failed from brace treatment were studied by Eamsobhana et al[11] to assess the correlation between overcorrection and recurrence. Radiographs of the affected limbs confirmed the varus angulations, and each patient underwent osteotomy of the proximal tibia and fibula. To study the effects, patients were divided into two groups based on their post-op femorotibial angles. The differing ages and BMI measurements of the patients in each group were not statistically significant. Group 1 had post-op valgus angles of 7-13º, and group 2 had post-op angles of more than 13º. Follow-up measurements were recorded periodically and after four years, 28.6% had recurrence in group 1 and 12.5 % had recurrence in group 2.[11] Recurrence is diagnosed when the femorotibial angle measures 10º varus at follow up exams. Although there were less cases of recurrence in group 2, the statistical difference was non-significant.

After careful analysis of the data, overcorrection of more than 15° valgus was found to show no benefit in preventing recurrent Blount's disease.[11] Although many surgeons recommend valgus overcorrection in order to relieve stress on the medial aspect of the tibia, no correlation between the degree of overcorrection and recurrence was found. These results suggest that the current practice of over-angulating, which can cause deformity, is not necessary. An osteotomy of 7-13° would be just as effective in preventing recurrence as using an angle greater than 15° would be, except with less chance of crippling deformity.

Conclusion

Although the clinical presentations of Blount's disease are commonly-known, this pathology remains a concern for healthcare professionals as it affects a vital portion of a child's joint: the epiphyseal growth plate. Through displacement or damage of the proximal tibial epiphysis, a progressive outward bowing of the lower limb occurs. Furthermore, the formerly unreported symptom of Blount's disease, slippage of the tibial epiphysis without a change in size or shape of the growth plate, reaffirms the importance of understanding the signs and indications of this disease process. With careful consideration for each patient's unique circumstance, treatment through corrective bracing, growth plate manipulation, or tibial osteotomy can effectively repair the deformity. Left untreated, abnormal tibia vara can eventually lead to multi-planar deformity and degenerative arthritis later in life, so it is essential that all modalities are employed and all efforts are made to recognize and educate about this disease.

References

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Figures and Captions

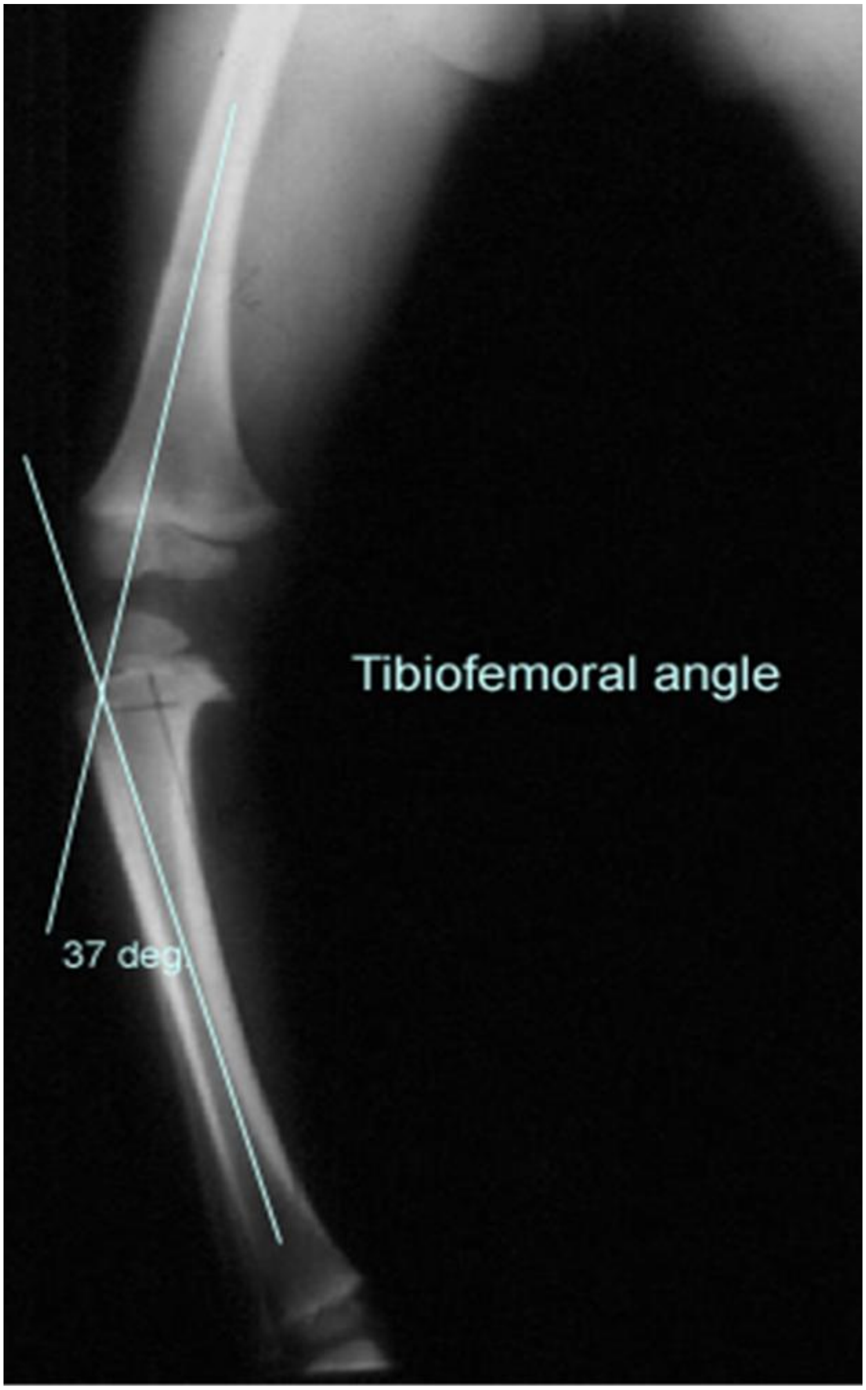

Figure 1. *Radiograph demonstrating the method of measuring the tibio-femoral angle used in diagnosing Blount's disease. Image courtesy of:* Orthobullets. Infantile blount's disease (tibia vara). http://www.orthobullets.com/pediatrics/4050/infantile-blounts-disease-tibia-vara. Updated August 30, 2014. Accessed October 23, 2014.

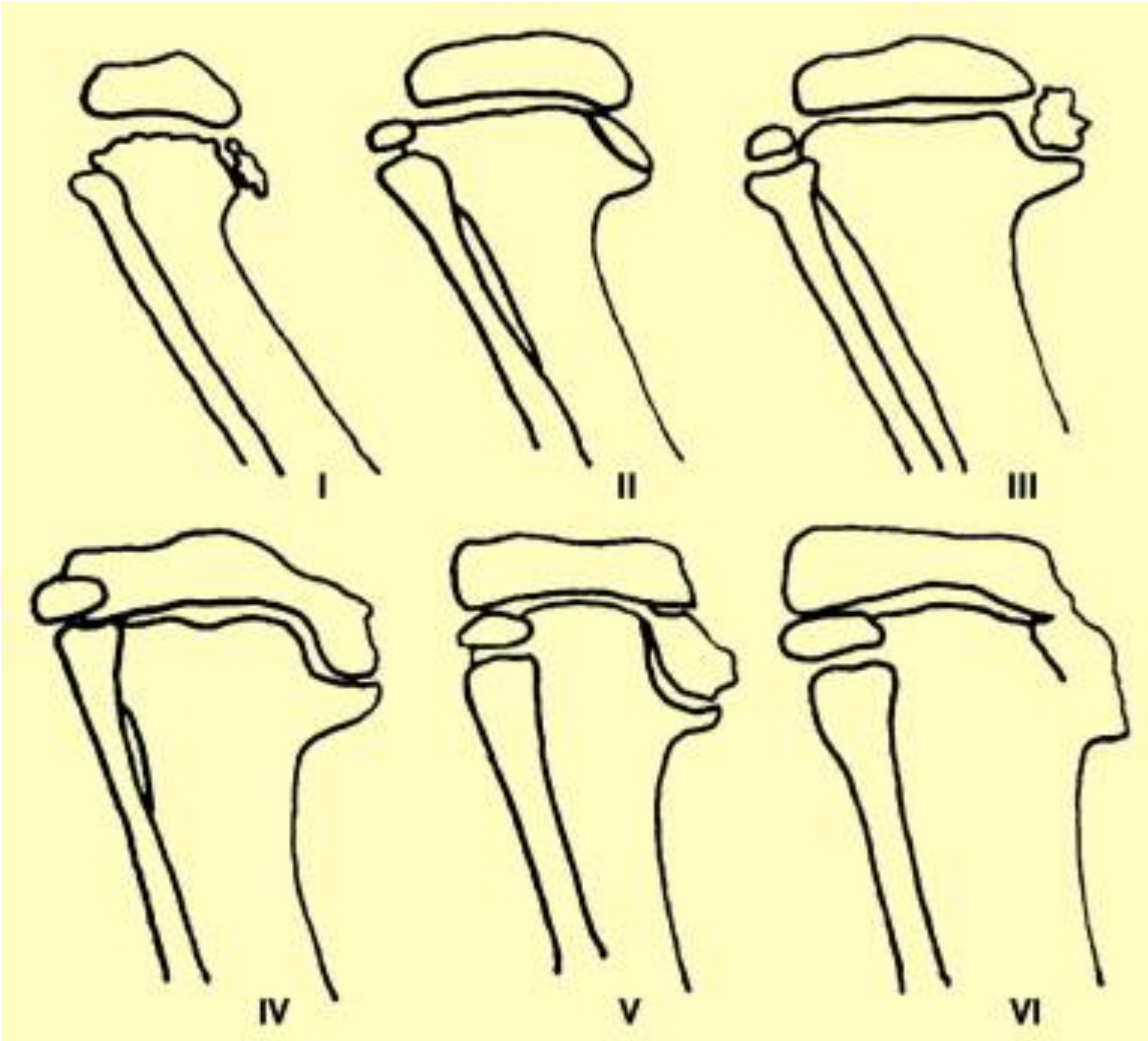

Figure 2. *The six stages of Blount's disease as classified by* Langenskiöld*. Stage I: Medio-distal beaking of the upper proximal tibial metaphysis. Stage II: Saucer-shaped defect of the metaphyseal beak. Stage III: Saucer defect deepens into step. Stage IV: Epiphysis bends over and fills medial beak. Stage V: Development of double epiphysis. Stage VI: Development of medial physeal bony bar. Image courtesy of:* Ferguson J, Wainwright A. Tibial bowing in children. *Orthopaedics & Trauma*. 2013;27(1):30-41.

Figure 3. *Various angles used in diagnosing Blount's disease. A. An AP radiograph demonstrating tibial bowing deformity. B. The tibio-femoral angle measured between the intersection of the tibial and femoral axis. C. The tibial metaphyseal-diaphyseal angle, TDMA, subtended by a line perpendicular to the axis of the tibia and a line connecting the most prominent beaking on the medial side of the proximal metaphysis. D. The epiphyseal-metaphyseal angle (EMA) measured between a parallel line along the tibial physis and another line from its mid-point to the most distal medial beaking. Image courtesy of:* Ferguson J, Wainwright A. Tibial bowing in children. *Orthopaedics & Trauma*. 2013;27(1):30-41.