# CHAPTER 5

# LOWER LIMB

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#### ANATOMY

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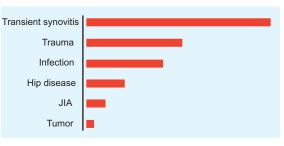
The lower limb spans the head of the femur to the toes. The leg lies between knee joint and ankle joint. Structures of the thigh are referred to as "femoral," from Latin *femur*: "thigh," such as fascia lata femoris: "broad fascia of the thigh," and os femoris: "bone of the thigh," or simply "femur." Because two bones occupy the leg, its structures are referred to as "crural," from Latin *crus*: "shank, leg," such as crural fascia.

Unlike the hip, which commands its own treatise, the knee is difficult to extricate from the lower limb, save by an arthroscope (*cf.* Sports chapter).

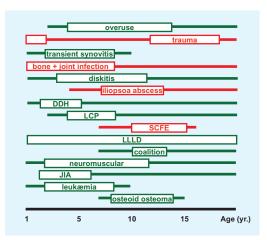
The patella is conferred autonomy due to its size and functional importance. Hence, the term "ligamentum patellæ" when convention dictates "quadriceps tendon," since the patella is lodged in this muscle as a sesamoid. The patella has a proximal base and a distal apex, referred to as superior and inferior poles. The anatomic terms aid understanding why children complain of pain rarely at the broad end (base) but frequently at the pointed end (apex), at which stress is concentrated. Anatomic terminology similarly aids remembering that the site of attachment of the lateral collateral ligament, known as fibular collateral ligament, differs from that of the medial collateral ligament, known as tibial collateral ligament.

Like the femur at the knee, the tibia has two condyles, which by being less rounded are less distinct. The raised space surrendered between the condyles for the cruciate ligaments is the "intercondylar eminence," commonly referred to as "tibial spine" due to its sharp terminus. Like the proximal femur, with its trochanters, the proximal tibia has an apophysis termed "tubercle." Unlike the femur, tibial apophysitis is common during growth.

Knee angle is defined between thigh and leg, or between femur and tibia. Popliteal angle (Latin *poples*: "ham, hollow of the knee") specifies the angle subtended by the leg relative to thigh in the sagittal plane [C], where it is a measure of "hamstring" tension. Full extension is defined as 0 degree, while hyperextension is defined as negative.



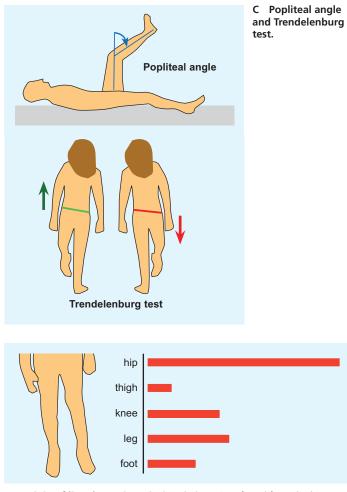
**A** Causes of limp by frequency Infection includes bone and joint, abscess, and diskitis. Hip disease excludes infection.



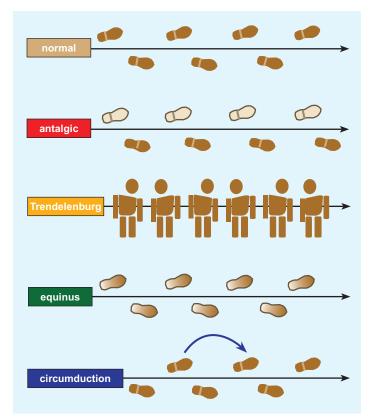
**B** Causes of limp by age Box is peak of presentation. Note that trauma is bimodal, including "toddler" fracture around 2 years. DDH: developmental dysplasia of the hip. (LCP, Legg-Calvé-Perthes; SCFE, slipped capital femoral epiphysis; LLLD, lower limb length discrepancy; JIA, juvenile idiopathic arthritis.) *Red* indicates urgent or emergent condition.

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# 92 Lower Limb / Limp



D Origin of limp by region Thigh includes pain referred from the hip.



E Types of gait.

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# LIMP

A limp signifies abnormal gait, which may be due to pain (conscious or subconscious), deformity, or neuromuscular imbalance. The limping child need not be a diagnostic "black box." It requires a thorough history and physical examination, which may be supplemented by laboratory and imaging studies. Transient synovitis is the most common cause [A]. The condition may be divided according to age and urgency. Different causes occur with different frequencies in different age groups [B]. Some are characteristic of certain ages (e.g., slipped capital femoral epiphysis circa puberty), while others are broadly distributed (e.g., lower limb length discrepancy). The varying causes may be divided into those that require urgent treatment (e.g., hip pyarthritis) and those that may be managed electively (e.g., developmental dysplasia of the hip). The hip dominates anatomically [D].

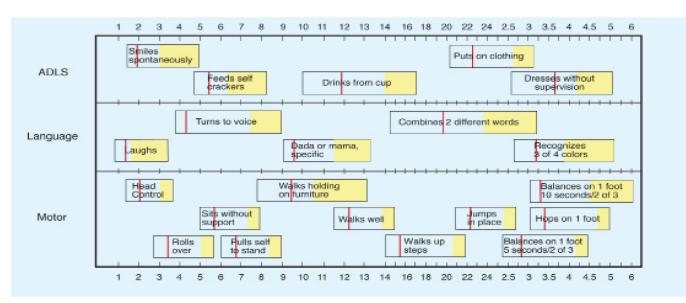
*Gait* Antalgic gait is defined by a shortened stance phase, of which the extreme is a refusal to walk [E]. A common theme in hip deformities producing limp is a Trendelenburg gait [C]. This results from a shift of the body over the affected hip in order to reduce the moment arm exerted on weak abductor muscles, which aids them in maintaining a horizontal pelvis during stance phase. In milder deformities, the gait may be apparent only after several cycles and may give way to pain as the hip abductors increasingly fatigue. This may be distinguished by its more lateral location from painful hip disorders, in which the pain is anterior in the region of the groin. Neuromuscular patients may exhibit an equinus gait, such as a hemiplegic who walks toe–toe. Equinus may drive the knee into recurvatum to plant the foot flat. Circumduction gait is characterized by incomplete knee flexion, actively (e.g., due to pain) or passively (e.g., due to contracture), which functionally lengthens the limb requiring swinging around the direction of walking to clear the ground.

*Evaluation* Age is the single most important discriminator of disease. Obtain a complete history. Is this acute, intermittent, or chronic? Are there associated symptoms (e.g., bruisability on leukæmia)? Is the limp local or a manifestation of a generalized condition (e.g., preceding illness)? Ask about milestones: delay may be an early tip-off of subtle neuromuscular disease [F]. Does the child complain of the limp? Long-standing, intermittent, milder pain may be suppressed in a child's consciousness producing the so-called "painless" limp (e.g., established Legg-Calvé-Perthes disease).

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Look at the whole child. Does she or he seem in distress (which may differentiate synovitis from infection), suggesting an urgent presentation? Are the upper limbs (e.g., hemiplegia) or back (e.g., diskitis) affected? Before examining where it hurts, go to the other side to gain the child's confidence. The physical examination should be performed in both supine and prone positions. The supine position may show hip obligate lateral rotation with flexion as seen in slipped capital femoral epiphysis. The prone position has the distinct advantage of allowing uncoupling of the knee from the hip, which may masquerade one for the other. The knee may be ranged from extension to flexion without moving the hip. In the supine position, moving the knee requires flexion of the hip, making it difficult at times to tell which joint is the offender. In addition, as in examination for torsion, the prone position allows simultaneous comparison of hip rotation (especially medial), which is the most sensitive to disease. Finally, the prone position will reveal a hip flexion contracture that may be concealed by lumbar hyperlordosis without the physician manipulating the child. No room is big enough to evaluate gait in detail. In addition, running amplifies gait disturbances and impedes compensatory mechanisms.

*Management* This is in accordance with condition, of which the different types are discussed independently elsewhere. Röntgenogrammes are readily available, low morbidity, inexpensive, and useful for general screening. Ultrasonography is noninvasive. By contrast, needle aspiration is invasive and stressful to child and parent; however, it is of high diagnostic value. Do not let timidity or logistics delay or dissuade—the emergency setting offers the best opportunity. Specialized imaging focuses and elucidates, such as scintigraphy in radio negative case where occult fracture is



F Denver developmental screening test.

suspected. Obtain laboratory tests judiciously. C-reactive protein is more specific and sensitive for infection than is leukocyte count. Observation is an acceptable and prudent form of management. Before requesting a complex test, which may require anæsthesia or may be expensive, evaluate the child on another day if the presentation is not urgent. A final diagnosis may elude the most thorough investigator in up to 25% of cases.

# LEG ACHES

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Leg aches are "growing pains." This may be explained as overuse in a child who lacks judgment to avoid repetitive injury and lacks consequence when injured (due to rapid recovery). Other mechanisms implicated include rapid, episodic skeletal growth stretching surrounding soft tissues, increasing body weight, participation in sports at higher levels with escalating expectations, and developing body mechanics. One-third of children experience leg aches, in addition to headaches and stomach aches. All of these may be stressful to the family but are physiologically benign, resolving without sequelæ. Extensive evaluation, including MRI of the brain, gastrointestinal contrast study, and multiple röntgenogrammes of the lower limbs, is negative. Recognize and reassure.

*Evaluation* Leg aches usually affect both legs, though one side may predominate [A]. If they produce limp, it is intermittent. They are worst at the end of a busy day or at night, interfering with falling asleep but not awakening from sleep. Long duration presents a paradox: while trying for a family, this lowers the physician's concern (bad things do not linger without causing further trouble). They are not associated with other symptoms. They are poorly localized and may migrate from part to part within a limb or from side to side. This is consistent with benignity, but also calls for a broad physical examination. There are no "hard" objective signs, such as deformity, stiffness, swelling, discoloration.

*Management* Establish this diagnosis of exclusion, in order not to request unnecessary tests or tests that yield equivocal results, which may alarm a family and trigger further testing.

# **TORSION**

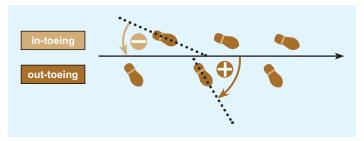
This presents the greatest disparity between familial concern and disease.

#### Nomenclature

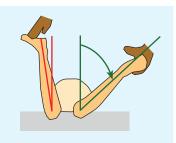
Version refers to normal axial rotation of a long bone. In the femur, this is the angle subtended between the neck and condyles, normally anteriorward 30 degrees at birth declining to 12 degrees at maturity. In the tibia, it is measured as the transmalleolar axis, with the knee as a neutral

History				
Geography	Non focal			
Limp Occasional, intermittent				
Timing	End of day			
	Interferes with falling asleep			
	Long duration			
Associated symptoms	None			
Physical examination				
Tenderness	No			
Deformity	No			
Stiffness	No			
Other soft tissue change	No			
Tests				
Imaging	Normal			
Laboratory	Normal			

A Leg aches Distinguishing characteristics.



A Foot progression angle.

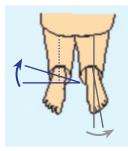


**B Hip rotation test** Prone position allows simultaneous evaluation to expose asymmetry from side to side and of all sites (hip, transmalleolar axis, and foot) of rotation. Asymmetric loss of medial rotation of the hip (*red*) is a concerning sign. In the patient pictured below, the feet easily rest on the table due to severe antetorsion (90 degrees+) ۲

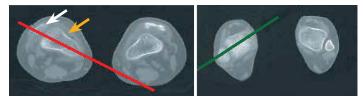


Site	Range	of Rotation	Age
Нір	Medial:	30–70 degrees	V
	Lateral:	30-70 degrees	
Transmalleolar axis	0–30 deg	grees	

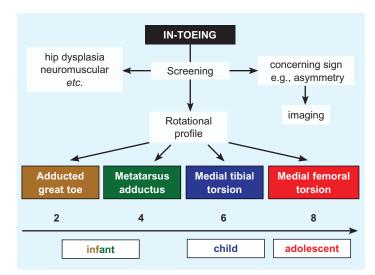
**C** Range of rotation for hip and leg Ranges by the end of the first decade. Total arc at the hip is 100 degrees. Medial rotation of the hip declines while lateral rotation increases with age, as degenerative disease contracts the capsular fibers.



**D** Measurement of tibial rotation Thighfoot angle is subtended between the long axis of the thigh and of the foot (*gray*). Transmalleolar axis is the angle subtended by a line drawn through the malleoli and a line drawn perpendicular to the axis of the thigh (*blue*).



**E CT** in preparation for operative correction of torsion **Rotational** malalignment results in lateral patellar dislocations. The femora are rotated medialward (*red*), while the tibiae rotate lateralward (*green*). The opposite rotation wrings the lower limb to drive the patella (*white*) out of the trochlea (*orange*).



**F** Algorithm for in-toeing Numbers indicate age in years by which component of in-toeing resolves spontaneously in most cases. Boxes below abscissa indicated period of treatment if indicated.

reference: normal is 5 degrees at birth increasing to 15 degrees lateralward at maturity.

Torsion refers to abnormal version, > or < 2 standard deviations from the mean. For example, in the proximal femur, "antetorsion" is preferable to "excessive anteversion." "Retrotorsion" signifies neck inclination 15 degrees to 0 degree. Retroversion is abnormal *per se*.

Rotation may be described as medial and lateral, or internal and external. The latter distinguishes rotation as a movement.

# Development

In the fetus, the lower limb rotates medialward to bring the apex of the knee anterior and the hallux medial. With growth, the lower limb unwinds lateralward, with declining femoral anteversion and increasing transmalleolar axis. Thus, in-toeing tends to resolve with growth, whereas out-toeing may worsen.

#### Evaluation

History This is essential to acknowledge the concerns of the family.

*Physical examination* Determine the rotational profile, which has the following components.

FOOT PROGRESSION ANGLE This is the angular difference between the axis of the foot and line of progression walking [A].

FEMORAL VERSION Measure hip rotation prone [B]. Significant asymmetry may be a sign of focal disease (e.g., slipped capital femoral epiphysis). Identify the midpoint between medial and lateral rotation, a measure of resting rotation. Normal medial rotation is <70 degrees; >90 degrees, which requires moving the limb off the side of the table, is considered severe [C]. A child with femoral antetorsion sits in a W position. The patellæ "squint" or "kiss" in the standing position. Running is characterized by an "eggbeater" pattern, as the legs flip out during swing phase.

ANGLE OF TIBIAL ROTATION This may be determined by thigh-foot angle or transmalleolar axis [D]. Thigh-foot angle is a measure of both leg (ankle) and foot (subtalar) rotation, whereas transmalleolar axis isolates the leg. In an infant, thigh-foot angle has wide variation due to ligamentous laxity: minimize this by guiding the foot to its neutral position rather than manipulating it into position. In an older child, it may be compared with transmalleolar axis to estimate contribution of hind foot rotation.

Foor Examine the lateral border of the foot. This may be convex in metatarsus adductus, thereby producing in-toeing. It may be concave in forefoot abductus, as in flatfoot or overcorrected clubfoot. An adducted hallux, dynamic or static, may give the appearance of in-toeing.

*Imaging* Consider imaging for concerning sign, such as asymmetry of hip rotation, or as part of operative planning. For the former, start with screening röntgenogrammes. For the latter, CT measures rotation [E].

*Management* The natural history of torsion is unaffected by manipulative therapy or bracing. Sitting in the W position is OK. Twister cables are not OK. Each component of torsion resolves spontaneously in the majority of patients with growth over the first decade [F]. In the young child, it is difficult to determine the functional impact of torsion, for example, frequent falling is more likely due to judgment and development of gait mechanics than to torsion, unless severe. In fact, medial torsion may be advantageous in rectifying the course of tibialis posterior and poising the subtalar joint to lock, thereby expediting push-off. Conversely, increasing lateral rotation reduces lever arm of the foot, thereby weakening push-off. Appearance is determined by the family and often is a significant cause for consultation.

There is no evidence that persistent femoral antetorsion accounts for long-term morbidity such as osteoarthritis. Femoral retroversion is abnormal and is associated with slipped capital femoral epiphysis and femoroacetabular impingement.

Only operation can change long bone torsion. This is indicated in < 1% of patients, and after 8 years of age. Torsion must be severe, clearly a cause of dysfunction, and natural history must be allowed to complete

its course. This is most likely in opposing or "miserable" malalignment [E], which is deleterious to the patellofemoral articulation, accounting for pain and patellar instability.

FEMORAL OSTEOTOMY This may be performed proximal or distal. Proximal incision may be covered more readily by clothing. Intertrochanteric osteotomy allows level proximal to trochanter minor, which heals readily and takes advantage of iliopsoas to add compression. Operation in the prone position, while requiring familiarity ("upside down"), allows comparison of both lower limbs for symmetry. Use a high angle plate (120 to 130 degrees) with longest blade into the head of the femur. This will provide an internal strut in the neck as protection against future osteoporosis.

TIBIAL OSTEOTOMY This is performed proximal to the tibial tubercle to correct patellofemoral malalignment. Distal metaphysial osteotomy is easier [G]. Cut the fibula for severe deformity. Add fasciotomy of the anterior crural muscle compartment to reduce risk of compartment syndrome. Plate fixation forgoes cast. Wire fixation allows more distal osteotomy for healing, and implant removal in clinic.

# GENU VALGUM AND GENU VARUM

Anatomic description of angulation at a joint or fracture addresses the distal skeletal element. Genu valgum refers to pointing of the leg away from the midline, or apex medial angulation of the knee, in the coronal plane. Genu varum is the opposite. Genu valgum is known as "knock-knee," due to overlap of the joints during walking. Genu varum is known as "bowlegs," after the arc formed by the lower limb.

#### Evaluation

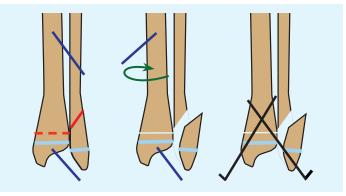
There is a normal evolution with age, and a normal range, of knee shape [A]. Knee shape may be measured as an angle or indirectly by distance between the ankles (genu valgum) or knees (genu varum).

*History* Is this acute (e.g., after fracture) or chronic (e.g., in rickets)? Does it disturb the child, such as pain or limp, or is it primarily a parental concern?

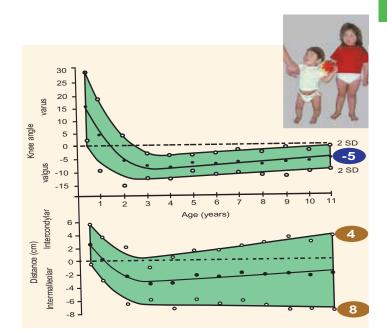
Physical examination Look at the entire child: is this focal, regional, or part of a generalized condition [B]? Short stature is common in rickets and various syndromes, which also may be characterized by other deformities, cutaneous stigmata, or dysmorphic features. Is it symmetric? Asymmetric deformity is likely morbid and not a physiologic variant. Determine the rotational profile, and neutralize this to isolate the coronal plane. Measure knee angulation with the patellæ pointing straight forward, by turning the hips as necessary. Most infants referred for genu varum have medial tibial torsion instead. In order to compensate for in-toeing, such children rotate the hips lateralward, presenting an oblique view of the knee. This will display a normal bend as the knee flexes during walking, which is interpreted as bowing by an observer who assumes an anteroposterior view of the knee. A goniometer aids angular measurement. Intermalleolar and intercondylar distance may be an easier measurement. Obesity influences measurements. Examine the child supine (static), as well as standing and walking (dynamic), which may expose soft tissue laxity.

Laboratory analysis This is indicated for a metabolic disorder, such as rickets.

*Imaging* Röntgenogrammes are indicated if disease is suspected. Weight-bearing views simulate the functional position. Educate the technician to obtain a relaxed view with the feet or knees together. Do not distort the image by forcing the limbs together. Include hips to ankles, in order to view mechanical as well as anatomic axes [C]. Mechanical axis identifies deformity: in genu valgum, the axis passes lateral to the knee or medial to the ankle, and genu varum is characterized by the reverse. Anatomic axis identifies site of deformity, for



**G** Osteotomy of the tibia and fibula Wires are placed (*blue*) remote from osteotomy of the tibia (*red*) to facilitate measurement of derotation, in this case medialward (*green*) for lateral torsion. For large derotation, cut fibula and consider fixation if fragments are widely displaced.



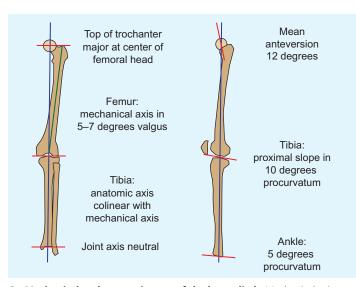
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A Normative data for knee angle and intercondylar/malleolar distance Children are born bowlegged, becoming maximally knock-kneed at 3 years before ending at mild knock-knee by the end of the first decade. The siblings pictured show genua vara (12 months) evolving to genua valga (3 years).

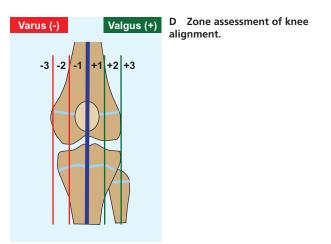
	Туре	Sub-type			
	Physiologic	Normal developmental variation			
Р	Focal growth disturbance	Blount			
a t	(over-/undergrowth)	Trauma			
h		Infection			
0	D I	Inflammatory arthritis			
0	Regional growth disturbance	Limb deficiency			
g	Primary bone disease	For example, osteogenesis imperfecta			
c	Other syndrome	For example, achondroplasia			

B Classification of knee angle Be systematic in approach.

# **96** Lower Limb / Genu Valgum and Genu Varum

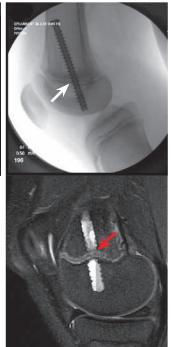


**C** Mechanical and anatomic axes of the lower limb Mechanical axis passes through joint centers, a measure of bones in combination. Anatomic axis is bone centered.





**E** Screw physiodesis Cannulated (for percutaneous use) full thread (to ease extraction) transphysial screw is placed in the medial physis (*orange*) of the distal femur for genu valgum and centrally in the lateral plane to avoid pro- or recurvatum deformation MRI after implant removal shows intact physis with no osseous bridge.



example, hypoplasia of lateral condyle of the femur producing genu valgum, or growth disturbance of medial condyle of the tibia producing genu varum. Coronal deformity also may be assessed by noting through which zone of the knee the mechanical axis travels [D]. Eliminate anisomelia, so that the child does not bend the knee, by a block under the foot of the short limb. What is the quality of bone? Is deformity diffuse or focal? Are the physis normal? Is there evidence of previous injury or disease? Focus the assessment with CT or MRI as indicated, to provide greater detail and for operative planning. Consider obtaining serial photographs.

#### Management

Treat the primary disorder first in generalized conditions, for example, vitamin D for rickets (*cf.* Syndromes chapter). Treat the primary musculoskeletal cause of knee deformity as indicated, for example, Blount disease (q.v.). For physiologic knee (mal)alignment, consider natural history during growth. Because children are maximally knock-kneed at 3 years and stabilize to mild knock-knees toward the end of the first decade, that is the time to intervene. Management earlier consists of education and reassurance. Because children become maximally knock-kneed at 3 years never to return to bowlegs, bowlegs at this age are abnormal and should be treated.

Like torsion, nothing influences natural history of genu valgum or genu varum except surgery. Determine the site of deformity by complete imaging. Harness the physis to guide growth: tether the medial physis in genu valgum and the lateral in genu varum, until correction, at which point remove the tether. Temporary hemiphysiodesis may be achieved by staples, plate, or transphysial screw [E]. Staples are the original. Early bending failure under force of growth has been addressed by corner reinforcement. They may dislodge due to smoothness of the tines. Plate and screws are more stable than staples, and span the physis. Screws may be placed percutaneously, are most stable, but are transphysial; however, removal after correction has not been associated with permanent growth disturbance. This approach is low morbidity, ambulatory, and predictable. For angular correction, remove implants within 2 years, before a physis gives up growth completely.

Deformity in the mature child, in whom growth modulation is not possible, or deformity that is not focal or uniplanar, occurring in the setting of broader complex deformity such as limb length discrepancy, requires more powerful reconstruction. Plan an osteotomy, including making cutouts or with graphics that allow operative simulation. The closer the osteotomy to the site of deformity, the less the requirement for translation to compensate for distance away from the center of rotation. Be prepared to perform an osteotomy at more than one level. Primary bone disease can deform the entire structure. Plan the fixation, internal or external, and location. External fixators are more potent in allowing three-dimensional correction. They enable gradual correction, which reduces the need for reconstruction of, and injury to, surrounding soft tissues. They are more forgiving because they may be manipulated after operation.

# Lower Limb / Blount Disease 97

# **BLOUNT DISEASE**

Blount called this "tibia vara." It represents a growth disturbance of the posteromedial proximal physis of the tibia. The growing tibia spirals into varus (medial tether) as well as medial rotation and procurvatum (posterior tether), thereby forming a complex multiplanar deformity. While the condition originates in the tibia, where signs are most striking, it also may involve the distal femur.

#### Cause

Familial clustering suggests a genetic predisposition. Race is confusing: it has been reported most in Black Americans [A] and Scandinavians, with a worse prognosis in the former. A mechanical cause is suggested by association with early walking and obesity: a heavy child whose physiologic genu varum is greater the earlier is walking and who does not fully extend the knees during gait concentrates load at the posteromedial physis of the proximal tibia. Compression retards or arrests growth according to the Hueter-Volkmann principle. Wide gait due to obesity increases the varus moment at the knee, squeezing the medial physis. Asymmetric or unilateral disease suggests that other, local factors may play a rôle.

#### Evaluation

*Classification* Like idiopathic scoliosis, Blount disease may be divided by age into infantile (<4 years), juvenile (4 to 10 years), and adolescent (>10 years). Severity is inversely proportional to age of onset.

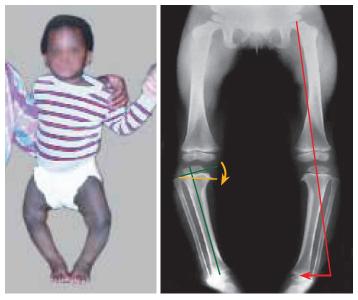
The condition also has been classified according to progressive physial irregularity [B]. However, this system has high interobserver variability, in part due to variable ossification over a wide age range and to segmentation of a continuous process. As a result, it may be simplified into I-III and IV-VI, based upon likelihood of spontaneous recovery and bridge formation, which impacts operation and outcomes.

*Natural bistory* The most important predictor of severity and poor outcomes is age of onset: early in growth, the more cartilaginous proximal tibia is more fragile. Knee degeneration is proportional to severity of deformity. Body weight is an independent predictor of outcome, with body mass index >40 kg/m<sup>2</sup> being a critical value for severity.

*History* Ask about walking age. Also enquire about nutrition and exercise, as well as efforts at weight reduction.

*Physical examination* It often is difficult to distinguish physiologic varus from Blount disease before 2 years of age: bring such patients back at 3 years of age. Calculate body mass index. Measure the intercondylar distance standing. Examine the knee dynamically: is there lateral thrust at the knee during walking? The lateral soft tissue become so attenuated over time that they are unable to support fully the knee during stance phase, allowing the tibia to be displaced lateralward. The deformity may be exaggerated, associated with a clunk, by varus stress of the attenuated lateral soft tissues. Determine the rotational profile of the tibia, which produces in-toeing and must be addressed by osteotomy.

*Imaging* Obtain röntgenogrammes standing with the ankles together and the patellæ forward to obtain orthogonal views of the knees. Alert the technician to allow the feet to turn inward as much as necessary. Projection is critical to isolating the coronal plane. Röntgenogrammes may be challenging in the adolescent due to obesity and size, including getting

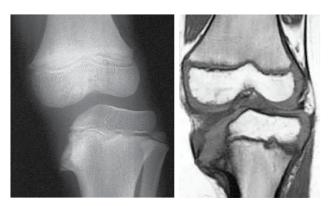


**A** Infantile tibia vara Mechanical axis misses the ankle laterally (*red*). Beaking of medial metaphysis indicates growth disturbance. Its effect is measured as the metaphysial–diaphysial angle.

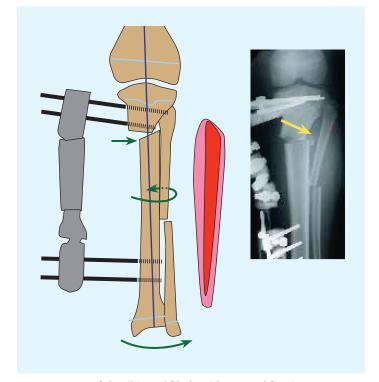




# **98** Lower Limb / Blount Disease



**C MRI in Blount disease** The medial condyle, absent on röntgenogramme, is visible on MRI, where depression explains medial subluxation of the femur.



**D** Osteotomy of the tibia and fibula with external fixation Transverse osteotomy distal to tibial tubercle and more distal to fibula. Correction includes valgus, lateral rotation, and lateral displacement to bring ankle into line with mechanical axis (*blue*). Deformity reduced and osteotomy fixed with external fixator (*gray*). Anterior crural compartment fasciotomy (*red*) to reduce the risk of compartment syndrome.

the width of the knees on a single view. The earliest sign is varus with metaphysial beaking [A]. Growth disturbance of the proximal physis of the tibia may be characterized according to the Langenskiöld classification. In long-standing disease, a significant (i.e., worthy of inclusion as part of surgical correction) proportion of the coronal deformity in adolescent presentation arises from the distal femur.

METAPHYSIAL–DIAPHYSIAL ANGLE This is the most established and useful measurement, formed by a line drawn through the metaphysis and one drawn orthogonal to the anatomic axis of the tibia in the coronal plane [A]. Substitute the axis of the fibula if the tibia is too deformed. In the incompletely ossified skeleton, a critical value is >15 degrees.

Arthrography outlines articular surfaces, often showing development of the medial condyle of the tibia invisible to röntgenogrammes because of a delay in ossification. MRI gives details of delayed ossification and deformation; articular changes, such as medial meniscal hypertrophy; and physial changes, such as bridge formation [C].

# Management

Do not forget the child: make sure a specialist is in charge of weight control. Orthopædic management is observation or surgical. Initially observe Langenskiöld I-III, as many improve spontaneously.

Osteotomy (infantile) Consider realignment osteotomy of the proximal tibia at 3 years of age [D]. Include osteotomy of the fibula distal to its proximal third to reduce risk of common fibular nerve injury. Factors associated with recurrence of deformity after operation include the following:

- Age >4 years
- Langenskiöld > III
- Body mass index >40 kg/m<sup>2</sup>

Osteotomy realigns the tibia, unloading and protecting the knee. It decompresses the proximal physis but does not change the fundamental growth disturbance directly; as a result, the tibia may grow back into deformity. Inform the patient and family that index operation may be the first stage in treatment: potential for recurrence is part of preoperative expectations. Prolong interval to recurrence by measured overcorrection to 10-degree valgus. Fixation is by wires supplemented with cast, or a blade plate with offset applied medially to displace the distal fragment lateralward, which has the advantage of no external immobilization.

Growth modulation (juvenile) Lateral physial tethering is indicated for:

- Juvenile onset, in which there is significant growth remaining.
- Mild deformity, which does not expect big correction. This is defined as Langenskiöld I-III, where the medial physis has reasonable growth potential, and a mechanical axis that traverses the medial condyle of the femur (does not miss medially).
- Body mass index <40 kg/m<sup>2</sup>.

Confirm that there is no physial bridge by MRI: if there is, lateral tether will arrest growth, achieving an even shorter limb.

Osteotomy (adolescent) If MRI shows a discreet physial bridge in the adolescent, this may be resected with placement of a fat graft before performing an osteotomy. External fixation has advantages.

• Because of long-standing growth disturbance, and because of patient size, it allows stable correction of coronal (varus), transverse (medial torsion), as well as sagittal (procurvatum) deformities.

# Lower Limb / Anterior Knee Pain 99

- Gradual rather than acute correction is possible, which may reduce neurovascular risk. Postoperative adjustment of correction compensates for any limitations of intraoperative imaging (e.g., not weight bearing, not full length).
- A limb shortened by severe disease may be lengthened in addition to corrected by distraction osteogenesis.

Level of osteotomy is distal to tibial tubercle: because this is remote from the site of deformity, the distal fragment must be translated lateralward as well as into valgus to compensate. Other osteotomy techniques include opening *versus* closing wedge and transverse *versus* oblique *versus* dome cuts. Opening lengthens the tibia, which partially addresses limb length discrepancy. Closing wedge may be safer by relaxing the common fibular nerve. Oblique and dome cuts increase surface area for union. Other fixation techniques include blade plate. The irregular shape of the tibia makes fitting this implant difficult if derotation and extension are included in correction. The subcutaneous medial face of the tibia makes implants prominent in this location. Add fasciotomy of the anterior crural muscle compartment by long scissors inserted through a small proximal incision and passed along the subcutaneous crest of the tibia to the extensor retinaculum.

*Articular reconstruction* For severe dysplasia of the medial condyle of the tibia in the adolescent, in whom remodeling potential is limited, an osteotomy may be performed to elevate the medial articular surface of the tibia to restore a horizontal joint line. Supplement the open procedure with arthroscopy to evaluate articular surface and guard against disruption or step-off [E].

# ANTERIOR KNEE PAIN

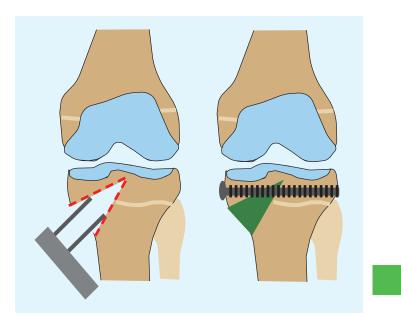
This is one of the common "growing pains." Some call it the "headache of the knee." Do not attach the word "syndrome." There are similarities with the Minnesota Multiphasic Personality Inventory for back pain.

#### Evaluation

This is a diagnosis of exclusion. Pain may be projected to the anteromedial knee from a condition of the hip, after the law of Hilton (John Hilton, surgeon at Guy's Hospital London, 1860), which states that "a nerve which innervates a muscle that acts on a joint will innervate that joint and the skin overlying the muscle's insertion." Examine the hips, in particular rotation. Knee examination does not suggest focal disease [A]. The pain is associated with a period of rapid growth and certain activities such as walking on an incline, sitting, and squatting. It is poorly localized: the patient cups the knee cap in the hand or circles the front of the knee when asked to point to the locus of pain. There are no "hard" objective signs such as swelling or persistent gait disturbance and no specific antecedent event such as trauma.

#### Management

Educate the patient and family that this is a normal albeit frustrating occurrence in childhood, with a natural history of spontaneous resolution without sequelæ over months or even years. The patellofemoral articulation and core strength are the focus, without or with the aid of a physiotherapist, in order to address subclinical instability of weakness. Include a coach or athletic trainer for proper sports mechanics. Do not neglect general health measures such as stretching, activity modification, and weight reduction if indicated.



#### E Articular reconstruction of medial condyle of the tibia

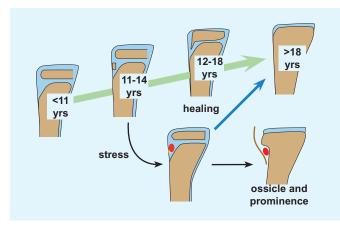
Osteotomy is opened (*red*) to elevate the medial plateau. The gap is filled with a structural tricortical osseous graft (*green*) and fixed with cannulated screw(s).

Examination	Finding
Gait	Normal, including no limp
Deformity	Normal alignment
Inflammatory signs	None, including no swelling, warmth
Pain	Diffuse; no focal tenderness
Knee motion	Full, supple
Patella motion	Normal stable tracking
Crepitus	Patellofemoral in some
Stability	Normal
Rotational profile	Normal
Muscle tightness	Hamstring, quadriceps
Single limb deep bend	Valgus may expose core weakness
Нір	Full supple motion, no pain

A Checklist for evaluation of anterior knee pain There are no focal signs of disease.



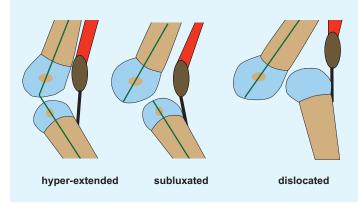
A Osgood-Schlatter condition The presentation is classic.



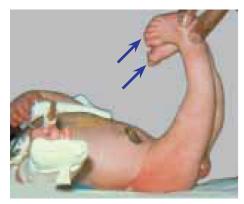
**B** Natural history of Osgood-Schlatter condition. Normal development of the apophysis (*green*) may be interrupted by excessive traction (*red*). Most heal and normalize (*blue*), but some remain as a separate ossicle or a prominence, which may be symptomatic.



**C** Sinding-Larsen-Johansson condition Separate ossicle at the apex of patella (*red*).



A Congenital dislocation of the knee.



# B Congenital knee dislocation

Hyperextension of the knees was accompanied by postural clubfoot due to packaging pressure.

# **OSTEOCHONDROSIS OF THE KNEE**

#### Osgood-Schlatter Condition

This represents a traction apophysitis of the tibial tubercle due to repetitive tensile microtrauma. It presents early in the second decade, during the period of prepubertal growth acceleration. The natural history is resolution with growth deceleration or cessation. In 10% of patients, a residual prominence or ununited ossicle may remain after maturity. The condition may result in a site of weakness that predisposes to fracture of the tibial tubercle.

*Evaluation* There is swelling and focal tenderness over the tibial tubercle [A]. The symptoms and signs are so classic that no imaging is necessary. Röntgenogrammes may not show the apophysis before ossification in a young child, or they may show normal fragmentation that becomes a cause of concern for the family. Röntgenogrammes also may show elongation of the patella, consistent with traction on the anterior knee during rapid growth.

*Management* Take the time to reassure the patient, as pain may be disabling. Modify activities. The patient may take anti-inflammatory agents *pro re nata*. Stretching of quadriceps femoris will reduce traction on tibial apophysis.

In the event of a healed though prominent tibial tubercle, which may be traumatized or hurt when pressed such as in kneeling, or an ununited ossicle in the ligamentum patellæ [B], operative treatment is an option. This consists of simple excision of ossicle(s) and shaving of the tibial tubercle.

#### Sinding-Larsen-Johansson Condition

This is a traction apophysitis at the other end of ligamentum patellæ, at the apex of the patella [C]. There is activity-related pain that correlates with tenderness at the apex of the patella. It is the ossified end of a spectrum that includes quadriceps tendinitis or "jumper's knee."

Evaluation and management resemble Osgood-Schlatter condition, with the exception that persistence beyond maturity does not occur.

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# DISLOCATION OF THE KNEE

#### Congenital Dislocation of the Knee

This may occur in isolation, or it may be associated with other condition, such as Larsen syndrome. It is divided into three types [A]:

- Hyperextension, which is characterized by recurvatum with normal contact of distal femoral and proximal tibial epiphysis
- Subluxation, where articular surfaces are partially in contact
- · Dislocation, in which the articular surfaces are not in contact

The condition is caused by extrinsic *in utero* pressure, which also increases the risk of hip dysplasia.

*Evaluation* The physical appearance is classic [B]. Look for signs of a generalized disorder or for other packaging signs. Because the proximal epiphysis of the tibia is unossified at birth, röntgenogrammes may be difficult to interpret. Ultrasonography will better reveal the knee epiphysis and is useful to screen for hip dysplasia. Determine passive flexion of the knees: fixed subluxation or dislocation is a sign of quadriceps fibrosis, which has a poor prognosis for resolution with closed methods, and is more likely to be associated dislocation of the hips.

*Management* For supple hyperextension, defined as the ability to flex the knee, allow passive stretching by parents for the first 2 weeks because often this suffices. Serial casting accelerates resolution. For a stiff knee that cannot be flexed, surgical lengthening of the extensor mechanism may be necessary. Include lysis of adhesions to achieve unencumbered glide of the muscle. Aim for flexion to 45 degrees followed by postoperative manipulation to recover more motion, rather than overlengthening, in order to avoid weakness of the quadriceps femoris. Operation at 3 months is within the window for closed management of hip dysplasia, and postoperative spica cast cares for both problems.

# Lower Limb / Other Conditions of the Knee **101**

# Dislocation of the Patella

There are two types [A].

**Congenital**. While named for the patella, the bone is a passenger in congenital dislocation of the quadriceps femoris muscle.

**Habitual**. The fundamental abnormality is osseous deformity, including dysplasia of the femoral trochlea and genu valgum.

*Evaluation* Congenital may be isolated or associated with other condition, such as Rubinstein-Taybi syndrome or nail–patella syndrome. It presents after walking due to gait disturbance. The quadriceps is short and dislocated lateralward posterior to the center of rotation of the knee, where it acts as a flexor and pulls the tibia into valgus and lateral rotation. The patella cannot be reduced. Röntgenogrammes are helpful after ossification of the patella (4 to 6 years of age). Ultrasonography reveals a cartilaginous dislocated patella.

Habitual presents later and throughout childhood. There is tenderness around the patella with apprehension to lateral force. In knee extension, the patella may be laterally subluxated such that, when reduced abruptly with knee flexion the patella traces a J track into and along the femoral trochlea. The lateral margin of patella may resist elevation due to retinacular contracture. The medial soft tissues may be attenuated. The knee may be in excessive valgus. There may be flattening or even a medial scoop of the medial soft tissues at the patella, suggesting hypotrophy of vastus medialis. Röntgenogrammes may show a shallow or flat femoral trochlea, lateral displacement of patella in trochlea, lateral patellar tilt, and a patella that is proximally displaced (where it does not engage the trochlea). Obtain full length lower limb standing röntgenogrammes if there is significant valgus, to evaluate the mechanical axis.

*Management* Congenital dislocation of the patella is a surgical problem: the entire quadriceps femoris must be reduced (Stanisavljevic) [B].

- The incision extends from trochanter major to apophysis of the tibia.
- Mobilize the quadriceps from lateral intermuscular septum and subjacent femur.
- Release the lateral retinaculum.
- Advance vastus medialis to the lateral margin of patella.
- Imbricate the medial soft tissues.
- Medialize the lateral half of ligamentum patellæ to periosteum (Roux-Goldthwaite). Transfer of tibial tubercle violates the physis, risking recurvatum deformity.
- Support the reduction by semitendinosus tenodesis through an osseous tunnel in patella drilled form distal medial to proximal lateral (Galeazzi). An alternative medial support for reduction is the adductor magnus terminal tendon, which is harvested at the musculotendinous junction and sewn to the patellar retinaculum adjacent the medial patellofemoral ligament.

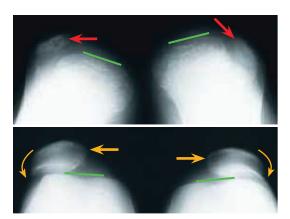
Management of habitual dislocation of the patella is discussed in Sports chapter.

# OTHER CONDITIONS OF THE KNEE

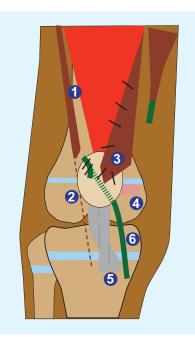
#### **Popliteal Cyst**

The term refers to a "fluid-filled mass" (Greek  $\kappa \upsilon \sigma \tau \iota \varsigma$ : "bladder, fluid-filled sack") that appears in the posterior knee (Latin *poples*). This more properly should be referred to as a popliteal bursa (Greek  $\beta \upsilon \rho \sigma \alpha$ : "sack, *purse*"): it occurs subjacent to semimembranosus or medial head of gas-trocnemius (or both) and fills with fluid in response to overuse. As a bursa, it is related to the joint by geography but not function. Language is important to distinguish this from other juxta-articular cysts (Baker), which communicate with the joint from which they originate, and represent a sign of internal derangement.

*Evaluation* Presentation is classic: location in medial popliteal fossa, fusiform shape, firm but not hard, nonreactive surrounding tissues, non-tender [A]. Transillumination confirms that the mass is cystic and not



**A Dislocation of patella** Merchant views. In congenital dislocation (*red*), patella rests against the lateral surface of the lateral femoral condyle and does not engage the trochlea (*green*), due to a dislocation of the entire quadriceps mechanism. In habitual dislocation (*orange*), the patella rests laterally displaced, due to hypoplasia of the trochlea, without or with (*orange*) lateral tilt, due to contracture of the lateral retinaculum. In both, the trochlea is hypoplastic.

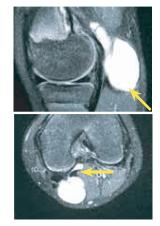


# B Reconstruction for dislocation of patella.

- 1. Release and mobilization of guadriceps
- 2. Release of lateral
- retinaculum 3. Advancement of vastus
- medialis 4. Imbrication of medial soft
- tissues
- 5. Medialization of lateral half of ligamentum patellæ
- 6. Semitendinosus tenodesis

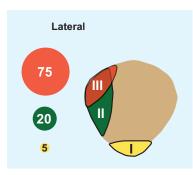
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A Popliteal cyst This is located in the medial popliteal fossa (*red*). It is filled with clear gelatinous fluid (**inset**). MRI shows no communication with the joint (*yellow*).

# **102** Lower Limb / Other Conditions of the Knee



**B** Classification of bipartite patella Secondary ossification of the patella takes place from superolateral to inferior. Numbers represent % of total.



**C Bipartite patella** Superolateral (*white*) is the most common site.

solid. The mass disappears with knee flexion, as it has more room to flatten. Gait is normal. There is no deformity, crepitus, or other knee abnormality. Although unnecessary for diagnosis or treatment, aspiration yields clear gelatinous material. While ultrasonography and MRI will show the mass, they add little and may tempt intervention.

*Management* The child has no symptoms and normal function. Natural history is spontaneous resolution over months with no sequelæ. It may recur but remains benign. Because of such benignity, no active treatment is indicated. Focus on education of parents, who may worry about a pernicious process.

#### **Bipartite Patella**

The primary ossification center of patella appears at 4 to 6 years of age. Failure of coalescence of a secondary ossification center, which appears at the turn of the decades, results in a bipartite patella. Incidence is 2%, and bilaterality is 50%, with no gender predilection. Three types have been distinguished [B]. Significant trauma, or repetitive microtrauma, may disturb a quiescent synchondrosis to elicit pain, akin to a traction osteochondrosis. Watershed blood supply to the (supero)lateral patella impedes healing once injured.

*Evaluation* Most are asymptomatic, found incidentally on röntgenogramme [C]. Smooth borders and typical location distinguish this from fracture. In symptomatic bipartite patella, there is focal tenderness and enlargement of the patella. A cleft may be palpable due to the subcutaneous nature of the bone. A stress manœuvre in the squatting position may show separation compared with anteroposterior röntgenogramme. Scintigraphy, by showing increased uptake, and MRI, by showing œdema, may be useful to confirm the diagnosis.

*Management* Most respond to conservative measures, including activity modification, quadriceps stretching, anti-inflammatory agents, and if necessary a short course of bracing (30-degree knee flexion). This approach may be supplemented with steroid injection.

( )

For intractable pain >6 months, consider operation.

- Fragment excision. This may be open or arthroscopic. While definitive, there is no consensus on fragment size limit.
- Vastus lateralis release, open or arthroscopic. This relieves pain by removing traction forces on the part. This is simplest, but quadriceps weakness is a concern.
- Débridement of the synchondrosis, followed by open reduction and internal fixation, is indicated for large fragments or when it is difficult to distinguish an acute fracture.

# Lower Limb / Lower Limb Length Discrepancy 103

# LOWER LIMB LENGTH DISCREPANCY

Lower limb length discrepancy (LLLD), or *anisomelia*, may be structural [A] or apparent. Contracture, dynamic asymmetry in function or remote deformity may produce an apparent discrepancy in limb length. Structural discrepancy may occur at the pelvis, femur, tibia, and foot, all of which should be included for complete assessment.

# Cause

The fundamental basis for hypertrophy is hyperperfusion [B]. The causes of limb shortening are diverse. They may be primary osseous, or secondary, such as limb shortening in hemiplegic cerebral palsy.

## Natural History

LLLD may be constant, such as shortening after malunion of femur fracture, or variable. Variable LLLD may be temporary, such as while a disease is uncontrolled (e.g., hyperæmia of inflammatory arthritis) or constant, as in limb deficiency. Constant LLLD is measured in millimeters or inches. Constantly variable LLLD is measured in % of limb length.

Up to 1/2" LLLD is normal in the general population. Between 1/2" and 1" is a gray zone. Work of gait (joules) increases significantly for LLLD >5%, while oxygen consumption (ml/kg/min) has been shown to increase significantly beyond 1" LLLD. Both result in visible alteration of gait. Mechanisms to compensate for LLLD include circumduction of the long limb, vaulting over the long limb, and equinus of the short limb. Children adapt to LLLD when adults are unable; hence, a 10-mm difference after arthroplasty may be a source of consternation.

It is impossible to arrive at consensus on relationship of LLLD to back pain. Similarly, reduction of center–edge angle secondary to titling of the pelvis has not been linked to hip degeneration. Pelvic obliquity may produce scoliosis, which is compensatory and remains supple and stable because time spent *per diem* standing erect with hips and knees extended represents such a small proportion of the day.

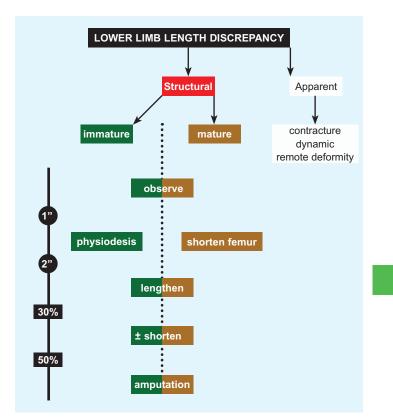
#### Evaluation

*History* Does the patient complain of pain, of inability to play or participate in sports, of appearance? Most patients will elucidate most primary disease, such as prior infection. How mature is the child? The primary physician can provide Tanner stage. Ask about familial stature. Discuss minimal acceptable height, starting at range for the population: women 4'11" to 5'9" (150 to 175 cm) and men 5'5" to 6'4" (165 to 193 cm). This may be influenced by multiple factors, including culture and surgical tolerance.

*Pbysical examination* Examine the whole child, in order to rule out primary disease, including musculoskeletal, neural, vascular, cutaneous, and abdominal examinations, for example, vascular malformation may manifest at the skin by discoloration or warmth. Take the height and plot it serially to calculate growth velocity. Upper and lower segment lengths aid in distinguishing hyper- from hypotrophy. Analyze gait, including walking and running.

The clinical discrepancy is the height of a block under the foot that levels the iliac crests. Pay attention to the height of tibial malleolus off the floor, which is a reflection of foot height. The supine position is necessary before the age of standing and allows differentiation of the femur and tibia [C]. Iliac spine to tibial malleolus, while technical in depending on a tape measure, is prone to inaccuracy due to difficulty in normalizing the pelvis and obscurity of its landmarks.

**C Clinical assessment of LLLD** Comparison of upper:lower segment lengths reveals right lower limb hypertrophy. The pelvis is leveled by standing on a lift (*white*), calculating total discrepancy. Femoral component of the discrepancy (*red*) is determined by comparing knee heights with hips flexed to 90 degrees and feet off table. Tibial component (*yellow*) is determined by comparing knee heights with hips flexed to 45 degrees and knees flexed to 90 degrees.

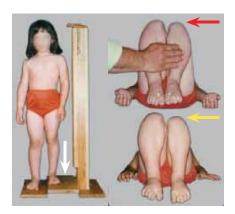


**A** Algorithm for lower limb length discrepancy (LLLD) Ordinate indicates the calculated amount of LLLD at maturity.

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Туре	Short	Long
Congenital	<i>Mild</i> : Hip dysplasia Clubfoot	Vascular anomaly
	<i>Severe</i> : limb deficiency skeletal dysplasia	
Other musculoskeletal	LCP SCFE	
Neural	Imbalance Paralysis	
Tumor	Physial injury	Physial hyperæmia
Infection	Physial injury	Physial hyperæmia
Trauma	Physial injury Malunion	Physial hyperæmia
Arthritis	Physial maturation	Physial hyperæmia

B Causes of lower limb length discrepancy Causes are varied.



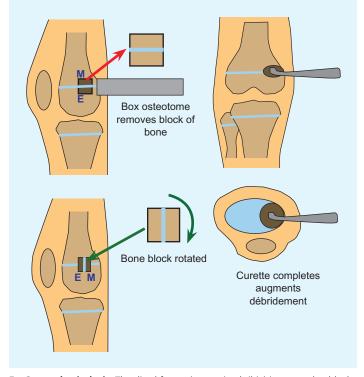
# **104** Lower Limb / Lower Limb Length Discrepancy



D Arithmetic method Distal femoral physis contributes 10 mm/year to growth, while the proximal tibia contributes 6 mm/ year. Girls stop growing at 14 years, while boys stop at 16 years.

LOWER LIMB Multiplier for BOYS				LOWER LIMB Multiplier for GIRLS				
Age (yr + m	o) M	Age (yr + m	o) M		Age (yr + mo)	M	Age (yr + mo	) M
Birth	5.080	7+6	1.520	]	Birth	4.630	6+0	1.510
0+3	4.550	8 + 0	1.470	]	0+3	4.155	6+6	1.460
0+6	4.050	8+6	1.420		0+6	3.725	7 + 0	1.430
0 + 9	3.600	9 + 0	1.380		0+9	3.300	7+6	1.370
1+0	3.240	9+6	1.340		1+0	2.970	8+0	1.330
1+3	2.975	10 + 0	1.310		1+3	2.750	8+6	1.290
1+6	2.825	10 + 6	1.280		1+5 1+6	2.600	9+0	1.260
1+9	2.700	11 + 0	1.240		1+0	2.490	9+6	1.200
2+0	2.590	11 + 6	1.220					
2+3	2.480	12 + 0	1.180		2+0	2.390	10 + 0	1.190
2+6 2+9	2.385	12 + 6	1.160		2+3	2.295	10+6	1.160
$\frac{2+9}{3+0}$	2.300	13 + 0	1.130		2+6	2.200	11 + 0	1.130
3+0 3+6	2.230	13+6 14+0	1.100		2+9	2.125	11+6	1.100
$\frac{3+6}{4+0}$	2.000	14 + 0 14 + 6	1.080		3 + 0	2.050	12 + 0	1.070
4+0 4+6	1.890	14 + 6 15 + 0	1.060		3 + 6	1.925	12+6	1.050
5+0	1.890	13 + 0 15 + 6	1.040		4 + 0	1.830	13 + 0	1.030
5+6	1.740	16+0	1.020		4+6	1.740	13+6	1.010
$\frac{5+0}{6+0}$	1.670	16 + 6	1.010		5 + 0	1.660	14 + 0	1.000
6+6	1.620	10 + 0 17 + 0	1.000	1	5+6	1.580		
7 + 0	1.570	Mature Leng		1	Modified from Dror Paley et al., JBJS Am 2000			

E Multiplier method The multiplier varies by age and by gender.



**F Open physiodesis** The distal femur (or proximal tibia) is exposed, a block of bone and physis is excised (*red*), rotated 90 degrees (green), and replaced, thereby forming a bridge of metaphysial (M) and a bridge of epiphysial (E) bone across the physis.

Imaging

ULTRASONOGRAPHY Start with ultrasonographic screening of the abdomen to rule out Wilms tumor in the setting of hemihypertrophy.

RÖNTGENOGRAPHY Röntgenogrammes may be generalized or focused. Generalized röntgenogrammes are used to calculate discrepancy. Teloröntgenogrammes (from Greek  $\tau\eta\lambda\epsilon$ : "at a distance, far off"), which represents a single exposure on a long film, risk inaccuracy due to parallax. Orthoteloröntgenogrammes (green) are a synthesis of separate orthogonal exposures of the hip, knee, and ankle (emitter moves) on a single film, giving a full view of the lower limbs to permit complete assessment of coronal alignment without terminal distortion. Scanogrammes are focused views of the hip, knee, and ankle (small cassette moves) with a ruler for reference but with intervening segments excluded; as a result, they are useful for length but not deformity. With computed röntgenography, teloröntgenogrammes have become standard. Focused röntgenogrammes are requested to view site of deformity and for operative planning.

Röntgenogrammes aid maturity assessment, including left hand-wrist films (Greulich and Pyle atlas), as well as physial appearance. The latter has a practical influence on operative options.

OTHER MODALITIES CT (scout view to limit radiation exposure) and MRI may be used in cases where computed röntgenography is difficult or confusing.

#### Management

Balance the broad target of  $<1^{"}$  and the lack of clarity regarding long-term outcomes against treatments of questionable benefit and significant risk.

*Orthotics* Do not underestimate the costs of orthotics. The obvious cost is pecuniary, from "custom" fabrication, from the need for multiple orthotics for different shoes and occasions, as well as when an orthotic is lost. A more insidious cost is the definition of a problem for a child who may have no complaint or disability. Furthermore, what is the end point if the discrepancy will not resolve? A lifelong intervention is a formidable prospect.

INTERNAL LIFTS These may be placed in the shoe, where they are discreet. However, not more than 10 mm can be placed before the heel will slip out of the cup, and this is within the normal range for the population.

EXTERNAL LIFTS These may compensate for a large discrepancy. They are benign in avoiding surgery. They are not benign in their conspiculty and become less functional with increasing height. They may be combined with internal lifts to mitigate these negatives.

*Physiodesis* Obliteration of the growth plate is indicated in the immature child for LLLD 1" to 2". It is performed at the distal femur and proximal tibia, where physis are most rapidly growing and accessible. Timing is essential to ensure a discrepancy at maturity <1". Longitudinal data reduce error. Physiodesis of the proximal tibia should be accompanied by physiodesis of the proximal fibula before age 10 years, to avoid relative overgrowth of the fibula.

SIMPLE METHOD This is applied to congenital discrepancy in the perinatal period or in infancy. Begin a conversation about amputation for a limb that is <50% the length of the contralateral side at birth. Final discrepancy may be estimated as double the discrepancy in the 3rd year of life, when half of adult height has been reached.

ARITHMETIC METHOD This makes the assumptions that growth is linear and that girls and boys cease growth consistently [D]. It has the advantages of practicality with the recognition that the goal is a broad target.

MULTIPLIER METHOD This is complex for the promise of greater accuracy [E]. It is based upon normative data for femoral and tibial lengths during growth as well as for growth remaining (Anderson).

• LLLD:

Dm [limb length discrepancy at skeletal maturity] = D [current limb length discrepancy] × M [multiplier].

Length at Skeletal Maturity (Lm):

Lm = L [current length of long limb]  $\times M$ .

# Lower Limb / Lower Limb Length Discrepancy **105**

- Timing of Epiphysiodesis:
  - Le [desired length of bone to undergo epiphysiodesis at time of epiphysiodesis] = Lm – Ge [amount of femoral or tibial growth remaining at age of epiphysiodesis]. Ge = e/0.71 for femur and e/0.57for tibia.

There are three principal techniques of epiphysiodesis. A cube of bone centered on the physis may be excised, rotated 90 degrees, and reinserted, disrupting the physis and creating two bridges of bone between metaphysis and epiphysis (Phemister) [F]. The physis is identified by direct visualization and needle palpation.

The physis may be drilled and débrided percutaneously with the aid of image intensification (Canale) [G]. Use of cannulation allows accurate placement of wires: these are overdrilled, and intervening physis may be excised with curette and rongeur. This is effective and requires less exposure.

Because of the inherent inaccuracies of predicting growth and discrepancy, the physis may be fixed to arrest growth. If limb equalization occurs before maturity, the implants may be removed to prevent overcorrection and shortening of the longer limb. Implants are the same as for angular correction, including staple, plate, and cannulated screw.

*Shortening* In the mature patient with LLLD 1" to 2", an equivalent segment of the longer femur may be excised open or closed. Blade plate fixation allows the most proximal shortening in the subtrochanteric region, which has less impact on quadriceps femoris and hamstring strength than diaphysial level. In the closed method [H], two cuts are made in the diaphysis separated by a distance equivalent to the desired discrepancy by a circular saw blade that moves eccentrically as the axis is turned. A fragmenter splits the ring of bone and a hook pushes the fragments out, after which the femur is shortened and nailed. This technique has not received wide adoption.

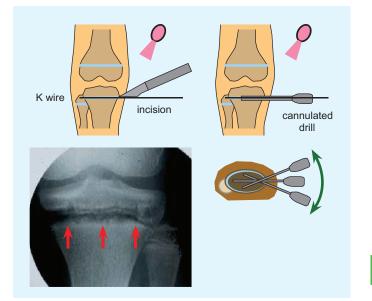
Shortening will relax the muscle envelope of the thigh, which cannot adapt to reduction beyond 10% of the total length of the femur, resulting in weakness.

*Lengthening* This is indicated for LLLD >2'', which is too much to lose functionally due to distortion of the surrounding soft tissue, and socially, including body proportions.

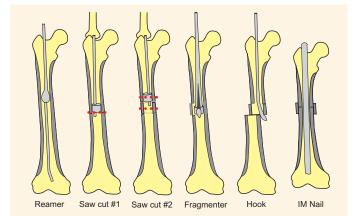
Lengthening device may be external or internal. External fixator may be unilateral [I] or circular (e.g., Ilizarov). It may be supplemented with a medullary nail or a plate, which reduce time of external fixator, obviate the need for casting after external fixator removal, and support the lengthening. The most recent internal device is a medullary nail controlled with an external magnet. This eliminates pin site care and complications and may be more comfortable because it does not tether the surrounding skin and muscle. However, in the femur such lengthening occurs along the mechanical axis, driving the knee medial, increasing lateral compartment stress. Previous devices, such as the ratchet nail, have been problematic, including binding limiting lengthening. "Run away" or over lengthening also is a potential complication of a remotely controlled device.

Operative principles to enhance bone regeneration include the following:

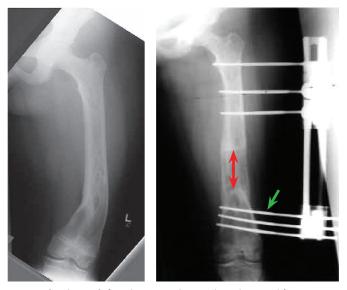
- Metaphysial osteotomy
- Protection of the soft tissue envelope, including minimizing incision, dissection, periosteal stripping
- · Corticotomy to leave the medulla in continuity
- Osteotomy with a mechanical device such as Gigli saw rather than a motorized device, which may burn the bone



**G** Percutaneous physiodesis Under image intensification (*pink*), cannulated drill is fanned (*green*), leaving a shadow across the proximal physis of the tibia (*red*).



**H** Closed femoral shortening A segment of medulla is cut (*red*) by an eccentric saw blade. Once the ring of bone is driven out, the femur is shortened and nailed.



I Lengthening + deformity correction Unilateral external fixator corrected deformity immediately at index operation for a 15-year-old girl with Ollier disease. Forces generated by lengthening bend distal pins (*green*).

TERMINAL Transverse Paraxial Complete Amelia Ulnar Radial Incomplete hemimelia fibula tibial INTERCALARY Incomplete phocomelia Ulnar Radial Complete phocomelia fibular tibia

 $( \blacklozenge )$ 

A Classification of congenital limb deficiencies.

After operation, a 1-week latency period is observed to allow the bone to mount a healing response. Distraction begins at a rhythm of 1/4 mm four times *per diem* for a rate of 1 mm/day. This is followed by serial röntgenogrammes: the rate may be adjusted based upon quality of regenerate, for example, slowed or even reversed temporarily in the event of discontinuity. After the desired lengthening or deformity correction, distraction stops and consolidation begins. Consolidation is twice the length of distraction; hence, the estimate that total time of lengthening is 1 cm/ month (10-mm distraction for 10 days, 20 days for consolidation). If external fixation is used alone, the barrel is removed at the end of consolidation, leaving the pins in place, to allow a period of dynamization (2 to 4 weeks): problems can be rectified by reapplying the barrel. Finally, pins are removed, in office if the patient can tolerate this and if cone shaped, which become loose after the first revolution.

Lengthening is complicated.

- Pin site infection. Educate patients and parents about pin care, including mobilization and prevention of sealing of skin around pins. Superficial infections may be treated by ambulatory oral antibiotics. Less often, incision and drainage are necessary.
- Delayed union may require arrest or compression of lengthening device, or lengthening of the consolidation phase. Nonunion may require grafting.
- Premature consolidation is treated by accelerating the rate of lengthening; rarely, osteotomy is required.
- Deformity of the callus. This is a reflection of soft tissue forces. It may be corrected by adjusting the frame during or at the end of distraction.
- Muscle contracture. These include triceps suræ and toe flexors in tibial lengthening, and rectus femoris and hamstrings for femoral lengthening. Instruct patients about daily stretching exercises. Other modalities include static or dynamic splints.
- Nerve injury. This is more frequent in tibial than femoral lengthening. Flex the knee for peroneal nerve symptoms. Reduce the rate of distraction. Occasionally, nerve decompression may be necessary.

 $( \bullet )$ 

 Arthritis. Lengthening increases joint forces. Joint stability, bony and soft tissue, is a prerequisite to successful lengthening. The effects of prolonged joint compression remain unclear.

# LOWER LIMB DEFICIENCIES

Congenital limb deficiencies occur in about 1 in 10,000 children, or about one-tenth the frequency of clubfoot or hip dysplasia. Boys outnumber girls (3:2), the lower limb is twice as affected as the upper limb, and >80% of cases are single limb.

Limb deficiencies may be congenital or acquired. Most congenital deficiencies are sporadic, occurring in children who are otherwise normal, and they have no genetic basis. Other causes include syndromic, for example, thrombocytopenia-absent radius (TAR) syndrome, and environmental, such as thalidomide phocomelia. Acquired limb deficiency may result from injury, as in trauma, or disease treatment, such as tumors.

Congenital limb deficiencies generally may be divided into terminal, in which all distal elements are absent, or intercalary, in which a segment between others is absent [A]. Deficiencies may be transverse, crossing the entire limb, or paraxial, affecting one side of the longitudinal axis of a limb.

Involve others early, including geneticist, orthotist–prosthetist, occupational and physical therapists, social worker, psychologist, patient support groups. Recognize the rôle of culture, in particular with regard to amputation. Remember that children can adapt. Save the knee whenever possible. Transarticular amputations are preferable, in order to avoid osseous overgrowth and subsequent stump problems. Prepare the patient and family for staged procedures. Align expectations and incentives so that the correct balance struck between risks of multiple and complex procedure and rewards of function and appearance.

#### Femoral Deficiency

"Proximal femoral focal deficiency" (PFFD) focuses on deformity of the hip. "Congenital short femur" represents a milder form on a continuum of femoral deficiency (Hamanishi). The spectrum of disease may be divided into:

- Presence or absence of a hip joint (Aitken) [B]. This influences salvage. Hip deformity correlates with femoral shortening.
- A femur that is too short or long enough for salvage (Gillespie).
- Associated fibular deficiency (q.v.).

*Evaluation* A limb >50% length of contralateral side may be salvageable. On röntgenogrammes, there appears to be discontinuity between proximal epiphysis and rest of femur. A femur <50% length of contralateral side, with a tapering proximal end and no epiphysis or acetabulum, is too short and lacks a hip joint to salvage. Arthrography may be necessary to determine hip integrity [C]. How unstable is the knee? Cruciate absence requires spanning of the knee during lengthening of the femur.

*Management* Have a clear treatment plan by the end of the first year, to allow prosthetic fitting by walking age if indicated.

ABSENT HIP, FEMUR < 50% Convert the limb to above-knee amputation. Syme amputation to improve prosthetic fitting. Fuse the knee, without or with excision of the distal epiphysis. The former addresses the fact that with growth the residual stump will become too long for an above-knee prosthetic. The latter addresses that issue by a delayed timed amputation. Leave the proximal femur free against ilium, where it will provide three-dimensional motion.

An alternative approach for very short femora involves fusing the hip and leaving the knee free to function as a hinge hip joint.

An alternative approach is to convert to a below-knee amputation. This requires a good ankle. Instead of Syme amputation, perform a Van Nes rotationplasty [D]. This is complex to perform and complicated for the patient. It substitutes the hinged ankle for the hinged knee, which is significantly more functional than an above-knee amputation. It requires fortitude to become accustomed to the appearance of a foot pointing backward.

PRESENT THOUGH DEFORMED HIP, FEMUR > 50% Reconstruct the hip, including valgus osteotomy for coxa vara. This will stabilize the hip and add length in anticipation of lengthening. It also may accelerate ossification, thereby limiting deformity.

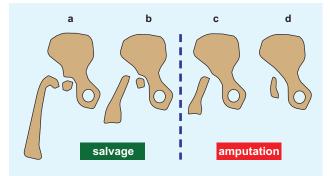
GOOD HIP, FEMUR > 50% Lengthen and correct deformity as necessary.

#### Fibular Hemimelia

This postaxial longitudinal deficiency is the most common lower limb deficiency. There is shortening or partial or complete absence of the fibula [E]. A fibrous analogue may replace the osseous fibula. The tibia, tethered by the short fibula, is short and bowed anteromedially. Ankle deformities include ball and socket in milder cases, and increasing valgus with instability and equinus as the fibula shortens taking with it the lateral malleolus. The foot shows loss of postaxial rays and talocalcaneal coalition. Hypoplasia of the femur and lateral condyle together with absent anterior cruciate ligament complete the spectrum. Deformity is proportional to amount of shortening.

Like tibial hemimelia (q.v.), most cases are sporadic. It has been reported in association with varus femur, hip dislocation, tarsal coalition, and digital anomalies as Fuhrmann syndrome, and linked to a mutation in WNT7A gene on 3p25.1.

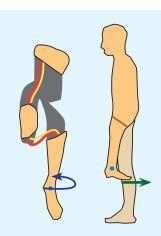
*Evaluation* How many rays are present? How deformed is the ankle? Is the foot displaced proximalward such that it rests against the distal lateral leg? These will determine whether the foot is salvageable. Is the



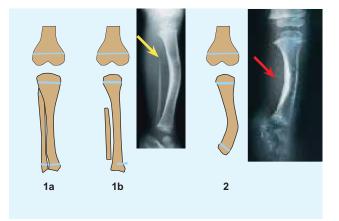
**B** Classification of femoral deficiency Like other classifications, this may be grouped into two (a + b; c + d), based upon outcomes.



**C Imaging** Arthrography clarifies what röntgenogrammes suggested. There is severe coxa vara, still unossified.

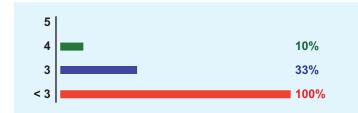


**D** van Nes rotationplasty The leg is rotated 180 degrees (*blue*), converting the ankle into an active knee joint. A below-knee prosthetic is applied to the foot, which is turned backward.

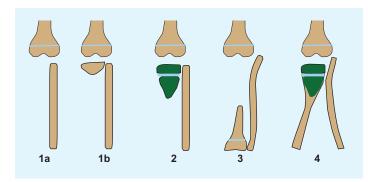


**E** Fibular deficiency classification The fibula may be short, very short (*yellow*), or absent (*red*). A system based upon fibula has limitations in not taking into account the ankle and foot or the femur.

# **108** Lower Limb / Tibial Bowing

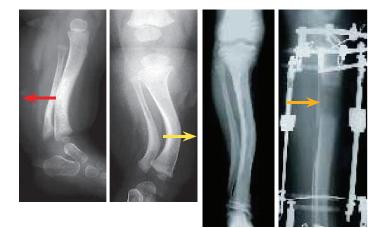


**F** Amputation for fibular hemimelia Amputation is correlates with number (ordinate) of rays of the foot.



**G** Classification of tibial hemimelia The presence of proximal tibia (*green*) correlates with quadriceps function, which improves outcomes of reconstruction.

 $(\mathbf{\Phi})$ 



**A Posteromedial bowing** This is defined by the direction of the apex posterior (*red*) and medial (*yellow*). In the second decade, persistent bowing and shortening (4.5 cm) was treated by correction and lengthening with circular external fixator (*orange*).



**B** Anterolateral bowing This may be associated with neurofibromatosis type I.  $\Psi$ : pseudarthrosis.

fibula palpable, including malleolus? Is there a cutaneous dimple overlying anteromedial bowing of the tibia? Examine the entire limb to determine the width of the spectrum. Is the knee stable to cruciate ligament testing? This impacts lengthening.

Röntgenogrammes elucidate shortening of the fibula, bowing of the tibia, architecture of the ankle, condition of subtalar joint, and number and anomalies of rays in the foot.

*Management* Amount of shortening of the fibula does not correlate with foot salvage. The two principal questions are as follows:

- Can the foot be saved? A foot with less than three rays will be too thin to support weight effectively [F]. Severe valgus and equinus deformity of the ankle will be too unstable and may become stiff and painful even if reconstructable. In such cases, Syme amputation is indicated.
- How is knee stability? For salvageable limbs, deformity correction of the tibia followed by lengthening requires external fixator spanning of the unstable knee to the femur.
- A stable ankle in the setting of mild shortening may require no treatment (≤5% shortening of lower limb) or lower limb length equalization by timed contralateral physiodesis (≤10% shortening of lower limb).

#### **Tibial Hemimelia**

Tibial hemimelia is a preaxial longitudinal deficiency classified based upon the extent of loss, without or with fibular deformity [G]. In type 1, the tibia is absent or demonstrates a nonfunctional remnant. I type 2, the proximal epiphysis and tibial tubercle are present, predictive of growth and preservation of knee function. Type 3 is rare and lacks a functional quadriceps mechanism. While type 4 has a complete tibia, distal divergence from the fibula results in proximal migration of the talus, which impacts ankle function.

Most cases are sporadic. Autosomal dominant and recessive transmissions have been described. Association with Langer-Giedion syndrome and evidence from a murine model suggest that tibial hemimelia may be caused by deletion of a gene involved in limb development contiguous with the gene for Langer-Giedion syndrome at 8q24.1. ( )

*Evaluation* History will reveal other syndrome. Determine how much tibia is present. Ultrasonography may be helpful at birth when röntgenogrammes are negative. Is quadriceps femoris function present? This is essential to preservation of the knee. Is the ankle salvageable in a type 4?

*Management* Surgical decision is based principally around quadriceps function and secondarily upon ankle stability.

ABSENT TIBIA, ABSENT QUADRICEPS Transgenual amputation with shortening of the femur by timed physiodesis to prepare for an above knee prosthetic.

ABSENT TIBIA, PRESENT QUADRICEPS Centralization of the fibula (Brown procedure) with Syme amputation. The fibula may be stable enough, and the quadriceps may be functional enough, that the patient may function like a below-knee amputee.

PROXIMAL TIBIA, ACTIVE QUADRICEPS Synostosis of the tibia and fibula with Syme amputation is highly functional and durable.

TIBIOFIBULAR DIASTASIS, PRESENT QUADRICEPS Consider reconstruction of the ankle to stabilize talus and preserve foot. The rarity of type 4 precludes consensus.

# **TIBIAL BOWING**

There are three types, named according to the apex of the tibia:

- Anteromedial. This results from fibular hemimelia (*q.v.*), in which the short fibula tethers the tibia like a string on a bow.
- Posteromedial.
- Anterolateral.

There is no bowing posterolaterally, where the fibula lies to obstruct this direction.

## Lower Limb / Tibial Bowing **109**

#### **Posteromedial Bowing**

Because the deformity improves with time, because intrauterine position has been implicated as a cause rather than intrinsic disease, and because outcomes are favorable, it may be referred to as "physiologic." However, it is not entirely benign, potentially ending with shortening (2 to 7 cm) requiring limb equalization [A], mild bowing, and calcaneovalgus foot deformity that may require reconstruction.

*Evaluation* Recognize it so that you may recommend to the family observation for spontaneous improvement. Degree of bowing correlates with amount of shortening. Röntgenogrammes quantify bowing (up to 70 degrees) and percentage of shortening, which remains constant throughout growth and allows prognostication.

*Management* Observe for improvement. Stretching of the foot by parents is benign. Treat lower limb length discrepancy according to general principles, including shoe lift if mild, contralateral physiodesis if moderate, and lengthening—with deformity correction if there is residual posteromedial bowing—for a discrepancy >2".

#### Anterolateral Bowing

This may progress to a fracture at the apex of the bow that tends not to heal, hence the term "pseudarthrosis of the tibia." In 60% of cases, this is a sign of neurofibromatosis [B]. Of all patients with neurofibromatosis, 10% will develop pseudarthrosis of the tibia.

Pseudarthrosis of the tibia has been classified [C], based upon radiographic changes at the apex, presence or absence of a pseudarthrosis of the fibula, and associated deformity of the foot.

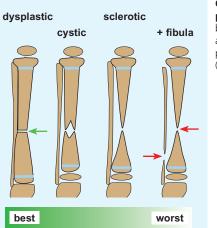
*Evaluation* Enquire about a family history of neurofibromatosis. Examine the whole child, including the skin for café au lait markings. Most pseudarthroses develop by 3 years of age; later onset is less severe and associated with more favorable prognosis. Röntgenogrammes show primary osseous changes that distinguish an acute traumatic fracture. Distinguish focal fibrocartilaginous dysplasia by its milder deformity, more benign radiographic appearance, lack of fibular involvement, and spontaneous improvement.

#### Management

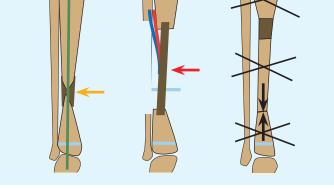
No FRACTURE The first goal is to prevent fracture. Once anterolateral bowing has been identified, apply a brace, which is worn for all weight-bearing activity throughout childhood.

FRACTURE There is no consensus. The pseudarthrosis does not want to heal, so simple fixation without or with bone grafting is insufficient. The pseudarthrosis is surrounded by a cuff of hamartomatous tissue, including fibroblasts and osteoclasts but lacking neural elements. This soft tissue must be excised in addition to all abnormal bone, in order to maximize the environment for healing. The residual defect may be filled with autogenous bone graft, a free vascularized fibula, or by transporting bone from the proximal healthy tibia [D]. Fixation may be internal, such as a medullary nail passed from proximal epiphysis to calcaneus for stability [E], or external, which though difficult in the younger child has the added benefit of allowing compression at the pseudarthrosis docking site. Radical resection with internal fixation and interposition of a cement spacer for to induce a foreign body reactive membrane (Masquelet), followed by removal of the cement and autogenous osseous grafting of the cavity, has shown early success in the same manner as with large osseous defects from trauma or débridement of infected nonunion. Supplementation with recombinant human bone morphogenetic protein-2 (rhBMP-2) is clinically controversial. While this may not increase the rate of union (quantitative), it may reduce the rate of refracture, suggesting a better qualitative response to treatment. Combining rhBMP-2 with bisphosphonate increases union rate while reducing callus fibrosis in a murine model of neurofibromatosis with pseudarthrosis.

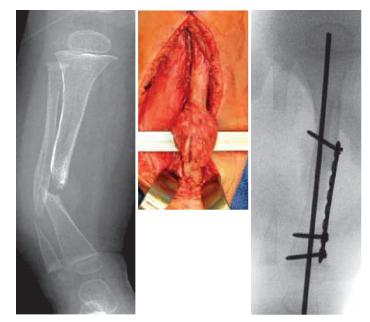
Despite union (mean time 15 months), some refracture, such that only half of cases heal long term. Union that survives into the second decade is durable. For nonunion, perform a Syme amputation and fit the patient with a below-knee prosthetic. It is a painful irony that some nonunions unite once amputation is performed.



C Classification of tibial pseudarthrosis Prognosis is best for simple type (*orange*) and worst when there is a pseudarthrosis of the fibula (*red*).



**D** Surgical options for pseudarthrosis of the tibia These include medullary nail and autogenous bone graft (*green*), free vascularized fibula (*red*), or bone transport by external fixator (*black*).



**E** Pseudarthrosis tibia treatment methods Pseudarthrosis excised *in toto*, including surrounding soft tissue mass leaving muscle and wide segment of bone leaving a gap. A vascularized fibula was transferred into gap and secured by medullary interference fit in proximal and distal tibia fragments. Construct was fixed with medullary nail from proximal epiphysis to calcaneus, as well as plate for rotational control.

# **110** Lower Limb / Tibial Bowing

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