

Chapter 11

Lengthening Reconstruction Surgery for Congenital Femoral Deficiency

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Introduction

Congenital femoral deficiency (CFD) is a spectrum of severity of femoral deficiency and deformity. Deficiency implies a lack of integrity, stability, and mobility of the hip and knee joints. Deformity refers to malorientation, malrotation, and soft-tissue contractures of the hip and knee. Both deficiencies and deformities are present at birth, nonprogressive, and of variable degree.

Classification

Existing classifications of congenital short femur and proximal femoral focal deficiency are descriptive but are not helpful in determining treatment. A recent longitudinal follow-up of different classification systems¹ showed that they were inaccurate in predicting the final femoral morphology based on the initial radiograph. Furthermore, previous classification systems were designed with prosthetic replacement surgery (eg, Syme's amputation or rotationplasty) in mind rather than lengthening reconstruction surgery (equalization of limb length with realignment of lower limb and preservation of joints). My classification system (Fig. 1) is based on factors that influence lengthening reconstruction of the congenital short femur.

In type 1, there is an intact femur with mobile hip and knee; ossification of the proximal femur is normal in 1a and delayed in 1b. In type 2, there is a mobile pseudarthrosis with a mobile knee; the femoral head is mobile in the acetabulum in 2a and absent or stiff in the acetabulum in 2b. Type 3 is a diaphyseal deficiency of the femur with knee motion $> 45^\circ$ in 3a and $< 45^\circ$ in 3b.

Knee joint mobility/deficiency, rather than hip joint mobility/deficiency, is the most important factor determining functional outcome and reconstructability of CFD. Previous classifications (such as the Aitken classification) emphasize the extent of hip deficiency. The extent of hip deficiency is used as a guideline to indications for amputation and prosthetic fitting despite the fact that amputation does not improve hip function. Types 1 and 2 are the most reconstructable. A wide spectrum of hip and knee dysplasia and deformity exists in type 1 cases. Because this is the type most amenable to lengthening, it merits a subclassification according to factors that require

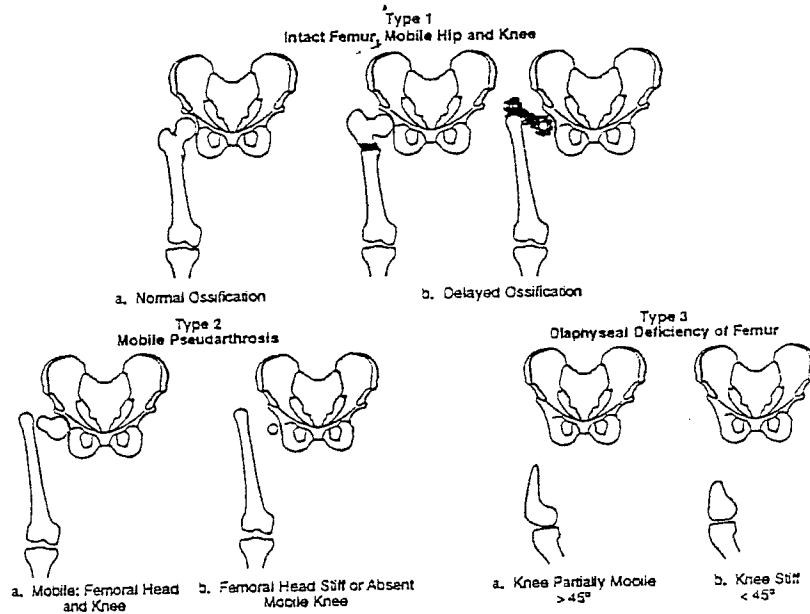


Fig. 1 Paley classification of congenital short femur syndrome, types 1-3.

correction before lengthening can be carried out. These factors affect the age at which the lengthening process can begin because multiple corrections will delay the first lengthening. They also affect the number of surgeries that are required prior to starting lengthening and, therefore, may affect the decision of reconstruction versus amputation or rotationplasty.

Type 1 is subclassified as follows: 0, ready for surgery, no factors to correct before lengthening; 1, one factor to correct before lengthening; 2, two factors to correct before lengthening; 3, three factors to correct before lengthening; and so on. (Factors requiring correction prior to lengthening include neck shaft angle $< 90^\circ$ with or without delayed ossification of the proximal femur, center edge angle $< 20^\circ$, subluxating patella, and/or dislocating knee.)

Neck shaft angle $< 90^\circ$ is corrected by proximal femoral osteotomy; center edge angle $< 20^\circ$ is corrected by pelvic osteotomy. Subluxating patella and/or dislocating knee are corrected by soft-tissue reconstruction.

The strategy of management of all type 1 cases is to convert type 1b into type 1a, and types 1a-1 or worse into 1a-0. Type 1a-0 cases can be treated by one or more lengthenings and/or epiphysiodesis or femoral shortening procedures.

The strategy of management for type 2 cases is to first determine the presence of a mobile pseudarthrosis or the absence of a femoral head. It is important to differentiate between type 1b cases and type 2 cases. In type

2a cases where there is mobility between the femoral head and the acetabulum and between the femoral shaft and the femoral head, union of the pseudarthrosis is an initial goal of treatment, converting type 2a to type 1a. Type 2b cases should not be converted to type 1b because the femoral head has no mobility in the acetabulum. To prevent proximal migration of the femur, the external fixator pins are extended onto the pelvis during lengthening. To stabilize the hip in type 2b cases, a valgus extension proximal femoral osteotomy (pelvic support osteotomy) is performed in conjunction with the final lengthening.

The strategy of management for type 3 cases is determined by preoperative knee and ankle joint motion. If the knee has $< 45^\circ$ of motion (type 3b) and the ankle has a good range of motion, a rotationplasty with prosthetic fitting should be considered. If the ankle has a poor range of motion, a Syme's amputation with prosthetic fitting is preferred. Equalization of limb length by multiple lengthenings is possible but yields the functional outcome of an arthrodesed knee. If there is $> 45^\circ$ of knee motion (type 3a), lengthening can be considered with all efforts made to preserve knee joint function. The goal of surgery in these cases is to convert a type 3a femur to a type 2b femur. The rest of the reconstruction is as for type 2b. This type of reconstruction, while feasible, is very complicated. Prosthetic options may be preferable to minimize the complexity of treatment.

Evaluation of the Infant With CFD

The Hip

Based on the clinical examination and the initial radiograph, the surgeon should try to determine the presence of a femoral head or a pseudarthrosis. If the femoral head is present without a mobile pseudarthrosis, the femur is considered intact. In the intact femur, the neck shaft angle should be evaluated for varus and the acetabulum for dysplasia. A pseudarthrosis, if present, should be examined under anesthesia with an arthrogram and fluoroscopy to determine if it is mobile or stiff. With a stiff pseudarthrosis, the femoral head moves with the rest of the femur; it does not with a mobile pseudarthrosis. If the pseudarthrosis is mobile, then it is important to establish if the femoral head is mobile within the acetabulum. Abduction-adduction, flexion-extension, and internal-external rotation movements under image intensification with the dye in the joint will demonstrate whether the femoral head moves in the acetabulum. The relative movement of the femoral head versus the femoral shaft to the pelvis can be compared on the abduction and adduction radiographs (Fig. 2). Push-pull stress movements with the dye in the joint also help differentiate whether a mobile pseudarthrosis is present. An unossified femoral neck appears like a pseudarthrosis but demonstrates synchronous motion of the femoral head, neck, and shaft. The presence of a well-developed acetabulum is the best clue that the femoral head is present and probably mobile. Magnetic resonance imaging (MRI) and ultrasound may

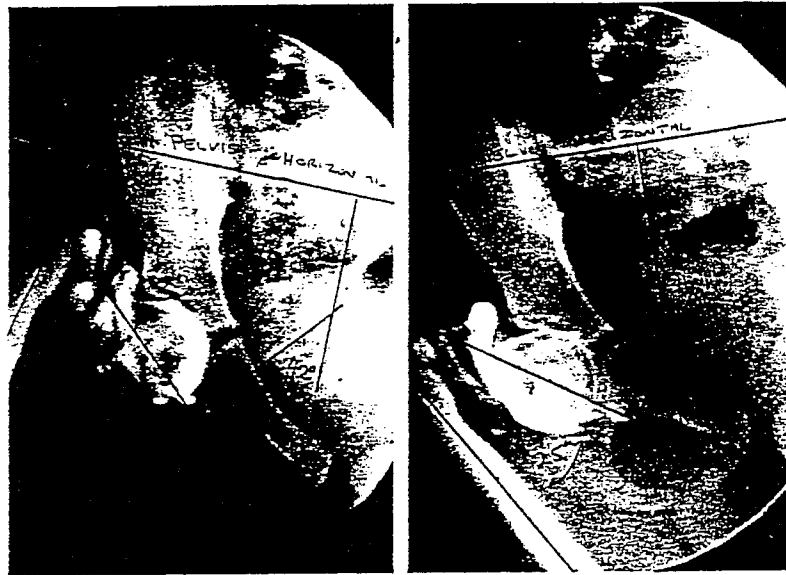


Fig. 2 Five-year-old girl with type 2a congenital short femur. Arthrogram with abduction (left) and adduction (right) stress anteroposterior views of the right hip. The femoral head and the shaft are both seen to move relative to the acetabulum. There is more movement between the femoral head and the femoral shaft than between the femoral head and the acetabulum. This indicates that there is mobile pseudarthrosis with mobility of the femoral head in the acetabulum.

be helpful but are often difficult to interpret in infants with such abnormal anatomy. The arthrogram performed by the orthopaedic surgeon remains my preferred testing modality.

It is important to examine the range of motion of the hip and identify the presence of fixed adduction, flexion, and external rotation of the hip. Fixed flexion deformity of the hip (Thomas test) should be compared to the other side in infants before walking age. There is normally some fixed flexion deformity for many months after birth. In the absence of fixed flexion deformity of the hip, there is usually an absence of hyperextension of the hip on the short side (measured prone). The Ely test is often positive, indicating a tight rectus femoris tendon. Lack of abduction is a sign of coxa vara rather than contracture or dislocation. External rotation of the limb (hip retroversion) is also typical of all grades of CFD. External rotation of the hip is best measured prone with the pelvis held flat.

The Knee

The initial evaluation of the knee is also performed clinically and radiographically. The range of motion of the knee is a critical feature. Fixed flexion deformity on the affected side greater than on a normal contralateral

limb is significant. A stiff knee is rarely found except in patients with severe proximal femoral deficiency. In these patients it may be difficult to assess the range of motion of the knee because of the short, chubby thigh. Examination under anesthesia may be required together with an arthrogram. The arthrogram in patients with a stiff knee may show flattening of the femoral condyles, an absent patellofemoral pouch, and an absent patella.

Mobile knees should also be evaluated for stability of the tibiofemoral joint and tracking of the patella. The tibia may dislocate as it goes into full extension as a result of tight lateral structures (fascia lata and hamstrings), incompetent capsuloligamentous structures, and anterior deficiency of the femoral condyles. There are two patterns of subluxation or dislocation of the tibia on the femur: anterolateral and posterolateral. In the anterolateral pattern, the tibia subluxates or dislocates anteriorly and the foot rotates internally as the knee extends. In the posterolateral pattern the tibia subluxates or dislocates posteriorly and the foot rotates externally as the knee extends. Lateral patellar subluxation or dislocation is associated with both types. Lengthening with the patella laterally subluxated can lead to dislocation or further subluxation of the patella and limitation of knee motion. To identify patellar subluxation, place a thumb over the center of the intercondylar notch of the 90° flexed knee. In a normal knee the thumb will lie over the patella. With patellar subluxation, the thumb will lie medial to the patella and the intercondylar groove can be palpated.

Treatment

Type 1a: Ossified Proximal Femur, Mobile Hip and Knee

This group is the most reconstructable. Lengthening treatment in these patients should not begin until the neck shaft angle is greater than 90°, the center edge angle is greater than 20°, and there is a nonsubluxating patella and nondislocating knee. Each of these prerequisites will be examined separately.

Coxa Vara

If the neck is ossified but the neck shaft angle is less than 90°, a proximal femoral valgus, extension, internal rotation subtrochanteric osteotomy should be performed prior to lengthening. My preference is to perform this osteotomy with external fixation. The hip osteotomy should correct the varus, flexion, and external rotation deformity. If the hip osteotomy is performed with internal fixation, the hardware should be removed prior to lengthening. I prefer to use the Ilizarov device to perform the hip osteotomy to avoid a long scar and hardware removal and to increase the accuracy and magnitude of correction of a complex valgus, lateral translation, extension, and external rotation osteotomy. Following the osteotomy the patient frequently will have an abduction contracture that will stretch out with time.

For neck shaft angles < 90°, especially in infants and young children, I prefer to perform the hip osteotomy separate from the lengthening procedure (Fig. 3) because both acute valgus hip corrections and lengthening

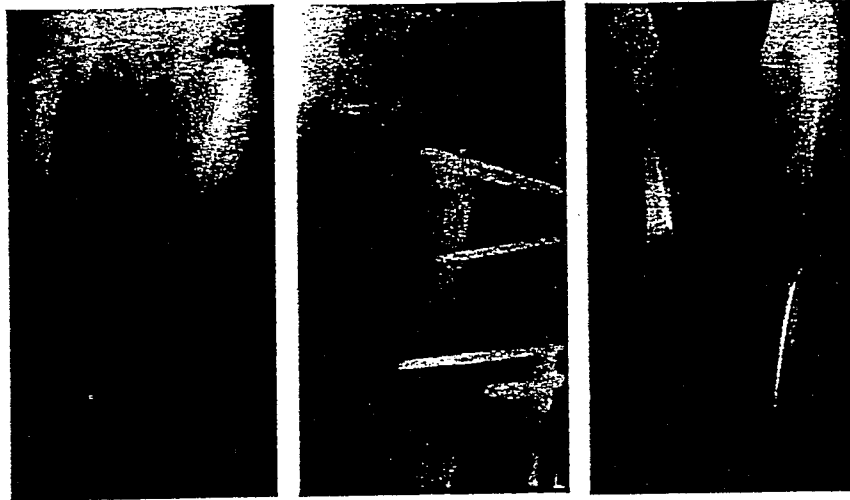


Fig. 3 Left, Anteroposterior (AP) radiograph of a 2-year-old girl with congenital short femur and coxa vara; neck shaft angle = 80°. The proximal femoral physis is vertically inclined. Center, Acute correction osteotomy stabilized with four half pins and the Ilizarov apparatus. The correction includes internal rotation, valgus with lateral translation, and extension with posterior translation. The neck shaft angle is restored to normal on the AP view and the femoral head is now anteverted on the lateral view. Right, AP radiograph 1 year after correction. The neck shaft angle is 124° and the proximal femoral physis is more horizontally inclined.

of the femur apply significant pressure to the hip joint. When the neck shaft angle is $> 90^\circ$ and/or if diaphyseal varus deformity is present, these can be corrected acutely together with lengthening of the femur (Fig. 4). Small degrees of coxa vara (neck shaft angle 110° to 120°) stabilize the hip in the face of a dysplastic acetabulum and should not be corrected prior to lengthening.

Acetabular Dysplasia

If the center edge angle is $< 20^\circ$, a pelvic osteotomy should be performed to stabilize the hip before a lengthening procedure.² If a proximal femoral osteotomy is necessary to correct a severe hip varus, it should be performed separately prior to the pelvic osteotomy. A three-dimensional (3-D) reconstruction computed tomography (CT) scan of both hips is useful in deciding which pelvic osteotomy to perform. The inferior and posterior 3-D views are the best to assess posterior coverage. If the 3-D scan shows good coverage of the affected hip posteriorly and deficiency anterolaterally, the Millis-Hall modification of the Salter osteotomy for coverage and pelvic lengthening is performed.³ If the 3-D scan shows decreased coverage posteriorly as a result

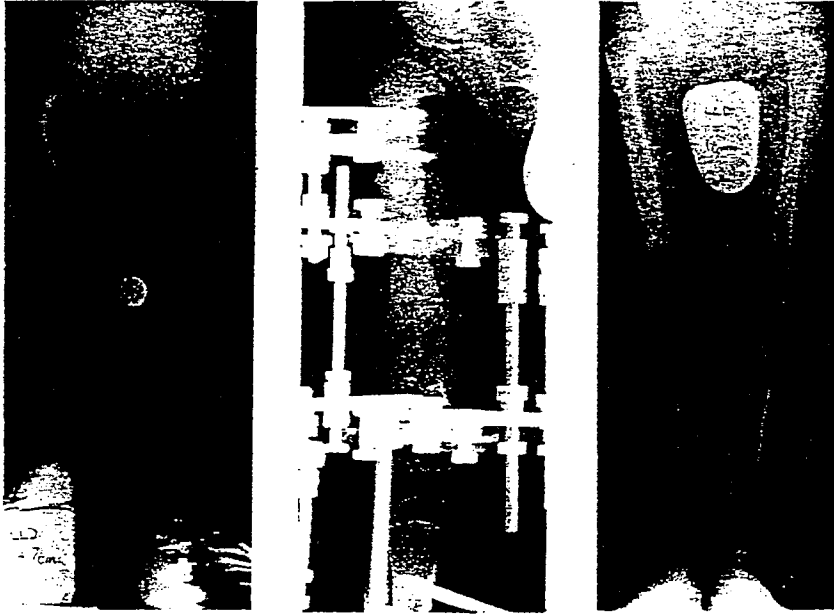


Fig. 4 Left, Two-year-old boy with congenital short femur (leg-length discrepancy = 7 cm) and coxa vara. The neck shaft angle = 95° relative to the proximal shaft of the femur. The proximal femoral physis is relatively horizontally inclined. There is a diaphyseal varus of 20° . Center, Treatment by proximal femoral valgus external rotation osteotomy. The proximal coxa vara was not corrected but the diaphyseal varus was corrected. The lengthening is performed through a distal femoral osteotomy. Right, Final anteroposterior radiograph after (6 cm) limb lengthening. The residual leg-length discrepancy is 1 cm.

of a hypoplastic posterior lip of the acetabulum, my preference is a Dega osteotomy with anterior shelf augmentation if necessary.

Dislocation of Patella or Tibia

Dislocation of the patella or tibia with flexion or extension, respectively, necessitates a stabilizing procedure prior to lengthening. Isolated anteroposterior (AP) instability of the tibiofemoral joint without dislocation does not need to be addressed before lengthening. Isolated subluxation or dislocation of the patella should be treated before lengthening. The knee reconstruction that I developed is based on a combination of elements from the Langenskiöld,⁴ MacIntosh,⁵ and Grammont⁶ procedures. This procedure may be performed at the same sitting as a pelvic osteotomy because both need to be in a long leg cast postoperatively.

In the Paley knee construction technique, the knee is exposed through a long S-shaped incision. The anterior margin of the fascia lata and the posterior margin where it blends with the intermuscular septum are incised lon-

gitudinally. The fascia lata is transected as proximally as possible and reflected distally until its insertion onto the tibia. The biceps tendon is lengthened if needed.

In the posterolateral tibiofemoral instability pattern, if the patella is subluxated but not dislocating then a lateral release is performed by cutting the capsule laterally without cutting the synovium. The vastus lateralis tendon is released off the patella and attached to the rest of the quadriceps muscle. The patellar tendon is displaced medially by internally rotating the tibia on the femur. If the patella tracks well in this position following the releases noted above, no further patellar stabilization is required. To prevent the tibia from externally rotating on the femur (leading to subluxation of the patella), the fascia lata is routed overtop the patellar tendon and knee joint capsule to insert into the medial femoral condyle. A drill hole is placed in the femoral epiphysis from a medial stab incision. A 6-mm hole is drilled into the condyle through to the lateral side. The fascia lata is anchored on the lateral side after passing through the bony tunnel. The knee is mobilized after 6 weeks in an extension cast.

In the anterolateral tibiofemoral instability pattern, if the subluxation pattern is anterolateral, the fascia lata is looped over itself after passing under the lateral collateral ligament or the lateral capsule (these children often do not have a well defined lateral collateral ligament) and through a subperiosteal tunnel proximal to the growth plate to reattach to Gerdy's tubercle (MacIntosh⁵) (Fig. 5). This procedure tightens the tibia into external rotation, and it can make the patellar subluxation worse by lateralizing the patellar tendon insertion site. The patellar tendon insertion site can be moved medially. The patellar tendon is sharply elevated off the apophyseal cartilage and is left connected to periosteum distally. The patellar tendon can be moved medially as much as needed pivoting on the distal periosteum. The medial side of the tendon is stitched medially (Grammont and associates⁶).

If the patella tracks in a laterally subluxated position even after the lateral releases and medialization of its tendon or if the patella is dislocated to begin with, then a Langenskiöld⁴-type reconstruction is performed. The capsule is separated from the patella and synovium medially and laterally. The synovium is cut from the patella circumferentially. The quadriceps tendon is left attached to the patella proximally and the patellar tendon remains attached to the patella distally. The synovium now has a patella-sized hole in it. A longitudinal incision is made in the synovium more medially. The patella is inserted into this new hole in the synovium and the synovium is sewn to the patella circumferentially. The original synovial hole is closed laterally where the patella was removed. The capsule is stitched overtop the patella on the medial side and left open laterally. After multiple-layer wound closure the knee is put in a cylinder cast for 6 weeks followed by active and passive motion exercises.

The above hip and knee problems must all be addressed before beginning the femoral lengthening. Once they are corrected, the femur is considered a type 1a-0, which is ready for lengthening.



Fig. 5 Left, Anterolateral dislocation of the knee. Left center, The tibia is internally rotated on the femur. Right center, After reconstruction; the knee is reduced and stable. Right, The fascia lata was looped around the lateral collateral ligament to externally rotate the tibia.

Type 1b Femora: Delayed Ossification Proximal Femur, Mobile Hip and Knee

The natural history of the intact, unossified femoral neck is usually to eventually ossify. Radiographically, the lack of ossification is often interpreted as a pseudarthrosis. Arthrographic examination reveals that the neck and shaft move as one. The coxa vara associated with these unossified femoral necks probably contributes to the shear forces on the neck that delay its ossification. Therefore, the treatment of the delayed ossification of the femoral neck is a valgus proximal femoral osteotomy, which is performed in the manner described above for the ossified proximal femur.

One particular type of delayed ossification is a stiff nonunion line in the inter- or subtrochanteric region. There is no movement seen at this nonunion line under fluoroscopy or stress radiographs. This type of nonunion always is associated with coxa vara. It can be ignored and the rest of the treatment carried out as for the delayed ossification cases with coxa vara. One alternative treatment I have used in these cases is distraction of the pseudarthrosis to correct deformity and lengthen. Although this method works, the regenerate produced is narrow. An acute correction by osteotomy as previously described is preferable and less complication-ridden.

Lengthening of Type 1 Congenital Femoral Deficiency

Choice of Osteotomy Level for Lengthening of the Congenital Short Femur Distal osteotomies have the advantages of a broader cross-sectional diameter for better bone formation and lesser deforming forces from the hamstrings and adductors. Proximal osteotomies have less effect on knee range of motion, but are more prone to poor bone consolidation, and they should be reserved for the technique of lengthening over nails (Fig. 6).

Deformations that require simultaneous correction must also be considered. Derotation and coxa vara correction are performed proximally. Valgus deformity of the knee is corrected distally. If both are needed, a proximal osteotomy is performed for derotation and varus correction, and a distal osteotomy for valgus correction and lengthening (Fig. 4). If the femur is completely straight with only some rotational deformity, the osteotomy can be made in the mid-diaphysis, which has a wider cross-sectional area than the proximal femur and is not in the zone of sclerotic, poorly-healing bone.

In older children with a wider medullary canal (> 7 mm), lengthening over a nail can be performed⁷ (Fig. 6). A proximal osteotomy can be used for lengthening with this technique because there is little risk of refracture with a rod in the medullary canal. Intramedullary nailing in children adds the risk of disturbance of growth of the apophysis³ and osteonecrosis of the femoral head.⁹ To avoid the latter, I use a greater trochanteric starting point and a nail with a proximal bend (eg, humeral or tibial). To avoid a coxa valga deformity, I prefer to use this technique in patients with some coxa vara. The apophyseodesis created by the nail can lead to gradual correction of the coxa vara. Fixator-only lengthening is usually used for the first lengthening. Lengthening over a nail is usually the method chosen for the second lengthening if the anatomic dimensions and deformities mentioned above permit.

External Rotation Deformity Most congenital short femora have an external rotation deformity (Figs. 3 and 4). This deformity should not be corrected through a distal femoral osteotomy for fear of subluxation of the patella. If the derotation osteotomy is performed proximally (subtrochanteric), the entire quadriceps mass attached to the shaft of the femur rotates medially, decreasing the Q angle.

Distal Femoral Valgus Deformity The distal femur usually has a nonprogressive valgus deformity resulting from hypoplasia of the lateral femoral condyle. The center of rotation of angulation of this valgus deformity is at the level of the knee joint line. Therefore, any osteotomy to correct the valgus in the supracondylar region needs to angulate into varus and translate laterally to avoid creating a secondary translational deformity. This deformity does not need to be corrected before lengthening but should be corrected at the time of the lengthening. The fascia lata should be transected or lengthened at the time of correction to help prevent recurrence, increased pressure on the lateral compartment of the knee, knee subluxation during lengthening, and loss of knee motion.

Soft-Tissue Releases for Lengthening of the Congenitally Short Femur Patients with congenitally short femur may have a fixed flexion deformity of the hip or knee, have an increased popliteal angle without fixed deformity, a



Fig. 6 A, Preoperative radiograph of 8-year-old girl with congenital short femur with a leg-length discrepancy of 14.5 cm. Predicted leg-length discrepancy at skeletal maturity is 20.8 cm. B, Due to the dysplastic acetabulum a Millis-Hall modification of the Salter osteotomy was performed prior to lengthening. This procedure gained 2.5 cm of length. At age 9 she underwent her first lengthening and acute derotation through a mid-diaphyseal osteotomy using the Orthofix™ apparatus. The apparatus was not extended across the knee because her knee joint was noted to be stable preoperatively. Note the well-developed tibial spines that imply the presence of cruciate ligaments. C, Final radiograph following the first lengthening. Note the overgrown greater trochanter and the coxa vara. D, At age 12 she underwent simultaneous lengthening of her femur and tibia. The femur was lengthened over a nail, and the femur and tibia frame were linked with hinges. E, Her final result following two lengthenings demonstrates near equalization of limb length (limb-length discrepancy = 1.9 cm) at bone and chronologic age 14. Total length gain = 2.5 cm (pelvis) + 4 cm (femur) + 8 cm (femur) + 5 cm (tibia) = 19.5 cm. The remaining leg-length difference will be made up by valgus osteotomy of the coxa vara which will acutely lengthen the leg by about 2 cm. F and G. Knee extension and flexion range of motion 3 months after removal of apparatus.

positive Ely test, and a thick, tight fascia lata. Soft-tissue releases are essential in conjunction with lengthening to prevent subluxation and stiffness of the knee and hip. I always release the rectus femoris tendon at its origin and the fascia lata distally. Through the same distal incision, the anterior fascia of the thigh and the lateral hamstring tendinous portion are cut, leaving the underlying muscle in continuity. If there is a popliteal angle $> 10^\circ$, a separate incision is made to fractionally lengthen the semimembranosus and gracilis tendons. The semitendinosus tendon is transected. The adductor muscles are also released. For distal femoral lengthenings, a percutaneous release is sufficient. For proximal femoral lengthenings, I prefer an open more extensive adductor release.

Botulinum toxin injected at the time of surgery is useful to temporarily weaken or paralyze some of the hamstrings and adductors and the rectus femoris. It seems to reduce muscle spasm and pain and may increase knee range of motion.

The timing of soft-tissue release may be important. Although I usually perform the release at the time of application of fixator and lengthening osteotomy, I have recently observed that in a few cases where the soft-tissue release was performed as a planned second-stage procedure (4 to 6 weeks after the lengthening began), I was able to achieve greater lengthening because the knee motion was better maintained for a longer period of time. Delayed soft-tissue release cuts the soft tissues when they are under tension and prevents them from healing before the distraction is over.

Knee Instability Consideration Almost all congenitally short femurs can be assumed to have hypoplastic or absent cruciate ligaments with mild to moderate anteroposterior (AP) instability. Some also have mediolateral (ML) and torsional instability. Nevertheless, the knee tracks normally preoperatively and there is no indication to do a ligamentous reconstruction in most cases. The significance of the knee instability to lengthening is the tendency to subluxation of the knee with lengthening (Fig. 6). Knee subluxation with lengthening is usually posterior or posterior plus external rotation, but can also be anterior. Posterior subluxation can only occur with knee flexion. Therefore, to prevent posterior subluxation some people recommend splinting the knee in extension throughout the distraction phase.¹⁰ Splinting promotes knee stiffness while protecting the knee from subluxation. I prefer to protect the knee by extending the fixation to the tibia with hinges. The hinges permit knee motion while preventing posterior as well as anterior subluxation. Fixation can be extended easily with the Ilizarov circular fixator but not as readily with the monolateral fixators.

A less common knee instability is anterior dislocation of the tibia on the femur (Fig. 5). This type of dislocation occurs as the knee goes into extension. It is important to document at which angle of flexion the knee relocates (conversely at which angle short of full extension the knee dislocates). The dislocation is caused by an anterior deficiency of the distal femur (the lateral radiograph of the knee shows a lack of the anterior protruberance of the femoral condyles). One treatment of this instability is extension osteotomy of the knee. The distal femur is extended by the number of degrees of flex-

ion required to relocate the knee, but I prefer the soft-tissue reconstruction procedures described, because knee extension osteotomy leads to loss of knee flexion.

Rehabilitation and Follow-up During Lengthening

Femoral lengthening requires close follow-up and intensive rehabilitation in order to identify problems and maintain a functional extremity. Follow-up is usually every 2 weeks for radiographic and clinical assessment. Clinically the patient is assessed for hip and knee range of motion, knee subluxation, nerve function, and pin-site problems. The distraction gap length, regenerate bone quality, limb alignment, and joint location are assessed radiographically.

Knee flexion should be maintained at $> 45^\circ$. If knee flexion is 40° or less, the lengthening should be stopped and the knee rehabilitated more. If after a few days knee flexion $> 45^\circ$ is regained, lengthening may resume. Never sacrifice function for length; an additional lengthening can provide more length but surgeons cannot recreate a knee joint. A flexion contracture may develop during lengthening. To prevent this, a knee-extension bar may be used at night and for 1 or 2 hours during the day. A fixed flexion deformity of the knee places it at risk of posterior subluxation. Subluxation of the knee can be suspected clinically based on a change in shape of the front of the tibia relative to the kneecap. Posterior subluxation of the tibia presents with a very prominent kneecap and a depression of the tibia relative to the kneecap (skihill sign). Extension of the external fixation across the knee with hinges prevents posterior subluxation.^{11,12}

Hip motion may become more limited with lengthening. Adduction and flexion contractures are the most significant because they lead to hip subluxation and dislocation. Rerelease of the adductors and the rectus, sartorius, and the tensor fascia lata during lengthening may need to be considered to allow further lengthening.

The deep peroneal nerve is the nerve at greatest risk with femoral lengthening. Pain referred to the anterior distal leg or dorsum of the foot should be considered peroneal nerve related until proven otherwise. Hyper- or hypoesthesia in the distribution of the peroneal nerve or weakness of the extensor hallucis longus muscle are corroborative evidence of nerve entrapment. A nerve conduction study (my preference is near nerve conduction using very fine needle technique at the level of the fibular neck) may show evidence of nerve injury. Quantitative sensory testing, if available, is the most sensitive test to assess for nerve involvement. With quantitative sensory evidence and sensory signs only, the distraction is slowed to see if the referred pain goes away. If the referred pain does not dissipate or if motor signs or positive nerve conduction evidence of nerve injury are present, nerve decompression at the neck of the fibula is carried out. Lengthening may continue after the decompression at 0.75 or 0.5 mm/day.

Hypotrophic regenerate formation requires slowing of the distraction rate. Overabundant bone formation may lead to premature consolidation and requires increasing the distraction rate for a few days. A mismatch between

the increase in the distraction gap from one visit to the next and the number of millimeters of distraction carried out in the same time period is a sign of an impending premature consolidation. Radiographs are also used to assess joint location. A break in Shenton's line or increased medial-lateral head-teardrop distance indicates subluxation of the hip. In the knee, posterior or anterior subluxation can be monitored on the lateral full knee extension radiograph.¹² Limb-length equalization should be based on full-length standing radiographs. If there is a knee flexion deformity, a scanogram with the knees equally flexed and the hip, knee, and ankle equally positioned to the radiographic plate is used instead. Limb alignment is assessed for femur and tibia separately and in combination. The joint orientation of the knee should be measured separately using the malalignment test.¹³ Axial deviation from lengthening (procurvarum and valgus for distal femoral lengthening and procurvarum and varus for proximal lengthening) is identified and corrected at the end of the distraction phase when the regenerate bone is still malleable. When there is malalignment of the femur and tibia, the femoral malalignment is corrected to a normal distal femoral joint orientation. The femur is not over- or undercorrected to compensate for the tibial deformity. The tibia should be corrected separately either during the same or at a later treatment.

Physical therapy starts within 1 or 2 days from surgery and should continue daily throughout the distraction and consolidation phase. It stops briefly after removal of the external fixator to avoid a fracture through the regenerate bone or a pin hole. Once the bone is strong enough, it continues. During the distraction phase, one to two formal sessions each day (45 to 60 minutes each) with a therapist are required. In addition, at least two home sessions per day (30 minutes each) are recommended. The more therapy the better the potential functional result and the faster the rehabilitation following removal. Inpatient rehabilitation is often the only practical method of achieving this quantity of therapy. The philosophy of therapy for lengthening is very different than for other orthopaedic surgical procedures. Following most orthopaedic procedures the patient is at his or her worst after surgery and gradually recovers. One week following surgery, lengthening patients are at their best. Thereafter, because of the distraction, the muscles become tighter and range of motion of joints more limited. It is not until the consolidation phase that the usual pattern of rehabilitation and recovery occurs. The lengthening surgery can be thought of as ending at the end of the distraction phase: a surgical procedure that can be measured in months rather than hours. In the absence of a therapy program, I will not even consider femoral lengthening.

The majority of the therapy time should be spent obtaining knee flexion and maintaining knee extension. Passive exercises are the most important during the distraction phase and passive plus active exercises during the consolidation phase. Hip abduction and extension are the two important hip exercises. Strengthening exercises should be focused on the hip abductors and the quadriceps. Electric muscle stimulation is used on the quadriceps. Upper extremity strengthening is helpful for use of walking aids and transfers. Weightbearing is encouraged and allowed as tolerated.

Type 2a Femora: Femoral Head Mobile in Acetabulum, Mobile Pseudarthrosis and Knee

The goal of treatment in this group is to convert the femur into a type 1. This requires obtaining union of the pseudarthrosis. To classify this group into type 2a it is necessary to do an arthrogram and fluoroscopic examination to demonstrate differential motion of the mobile nonunion and mobile femoral head.

In order to obtain union of the head to the femur it is necessary to open the pseudarthrosis, bone graft it, and reorient it. A proximal femur valgus osteotomy is performed to reorient the nonunion and the coxa vara. If the femur is proximally migrated, it may be necessary to pull down the femur relative to the pelvis as a first stage. If this is not possible acutely, it should be done gradually. It is important to extend the fixation to the pelvis in order to neutralize the forces at the pseudarthrosis site. Once the pseudarthrosis is united the rest of the treatment is as per type 1.

Type 2b Femora: Femoral Head Stiff or Absent in Acetabulum, Mobile Pseudarthrosis and Knee

If the femoral head is stiff or absent but the knee is mobile, reconstruction can still be performed combined with lengthening. Motion of the hip in these patients comes from the mobile pseudarthrosis. The proximal femoral shaft should not be fixed to the femoral head because this will result in a stiff hip. To preserve hip motion but create stability for lengthening and gait, the proximal femur should be osteotomized into valgus under the femoral head and acetabulum (pelvic support osteotomy) between 10 and 16 years of age (Fig. 7).

Prior to this age, the femur can be lengthened once or twice. Frequently the femur is so short that it cannot be lengthened together with hinging of the knee. In these cases, the knee can be locked in extension for the first lengthening. Either femoral lengthening alone or simultaneous femur and tibial lengthening may be performed. Fixation must extend to the pelvis to prevent proximal migration of the femur.

If there is a major deficiency of the proximal femoral diaphysis, the first lengthening is performed to grow the femoral shaft. Obviously the more deficient the proximal femur the more complicated the lengthening program becomes. The risk of losing knee motion in this group is very high. Knee flexion deformity is very common in this group. An arthrogram should be done to determine if the femoral condyles are round. If the femoral condyles are round, the knee can be extended by soft-tissue releases and distraction. If the femoral condyles are flat, the knee should be extended by osteotomy. As the degree of deficiency increases, the appeal of a prosthetic option of treatment increases. This issue will be addressed later.

Type 3a: Diaphyseal Deficiency of Femur, Knee Motion $> 45^\circ$

Deficiency of the proximal femur with absent femoral head, greater trochanter, and proximal femoral metaphysis results in a mobile pseudarthrosis and a very short femoral remnant. Some cases have a mobile knee with $> 45^\circ$

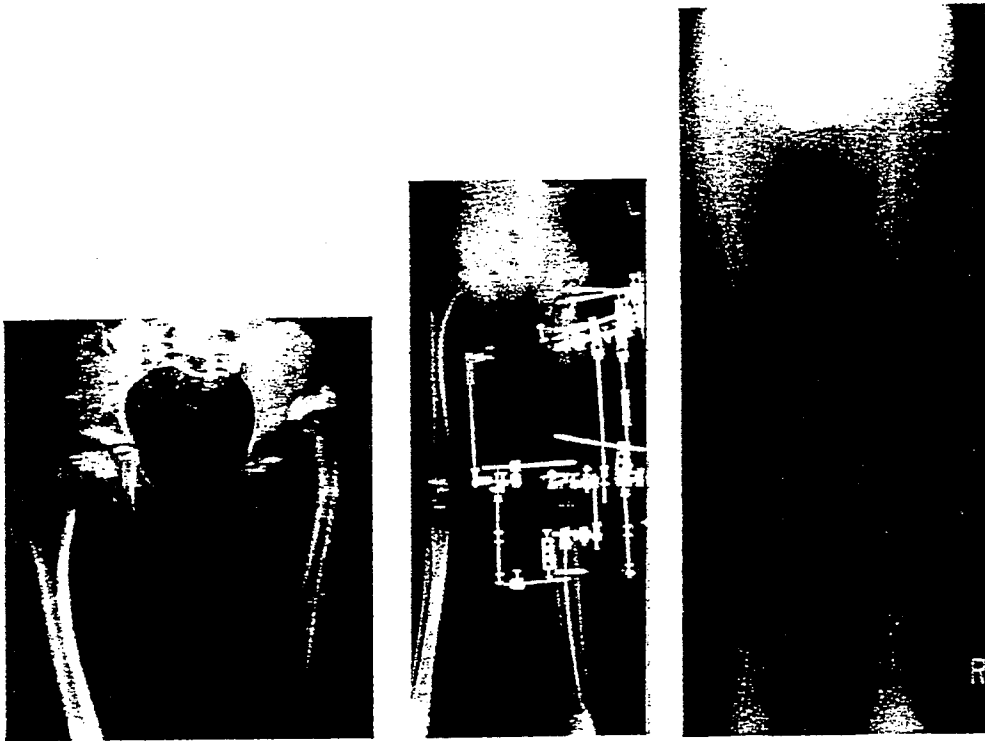


Fig. 7 Left, Radiograph of a 12-year-old girl with type 2b congenital short femur. There is no acetabulum or femoral head and the proximal femur is proximally dislocated. Center, A valgus-extension proximal femoral pelvic support osteotomy was performed with distal femoral varus realignment. No fixation to the pelvis was used during the 13-cm femoral lengthening. Note that fixation is extended with hinges across the knee to the tibia. Right, Final standing anteroposterior radiograph shortly after removal of the fixator.

of motion, usually with a 45° knee flexion deformity. The treatment option in these cases includes lengthening or prosthetic reconstruction (van Nes or Syme's). To lengthen a type 3a femur deficiency, the femur should be converted to a type 2b. After that, treatment is as described for type 2b. The knee flexion deformity in these cases can be addressed by extension osteotomy of the distal femur. Because of the severe discrepancy from this type of deficiency, a combined femur and tibia lengthening can be carried out. The frame is extended across the knee joint without hinges. The fixator is also extended across the hip to the pelvis to prevent proximal migration of the femur.

Type 3b Femora: Diaphyseal Deficiency, Knee Motion $< 45^\circ$

Reduction of knee motion to $< 45^\circ$ is due to malformation of the lower end of the femur and abnormalities and deficiencies of some of the soft tissues. The femoral condyles are flat and the tibia articulates in flexion with the squared-off end of the femur. There is an absence of the suprapatellar pouch. The quadriceps muscle and tendon are usually present although the patella is usually absent. Complete rigidity of the knee is frequently related to an abnormal ligament running from the insertion site of the anterior cruciate to the anterior surface of the distal femoral condyles. Resection of this ligament frequently results in 45° or more of knee flexion. Because the femur is so short, it is very difficult to exercise the knee; there is no proximal lever to stabilize. The best result that can be hoped for in these limbs with a lengthening program is equalization of limb length, a mobile hip, a mobile ankle, and a stiff knee with at the most 45° of motion. Functionally this is probably not as good as a van Nes rotationplasty or Syme's amputation and prosthetic fitting. With either of these procedures the hip is still deficient and unstable. Ilizarov hip reconstruction to minimize limp and give better pelvic support can be considered. A lengthening program requires multiple procedures to achieve limb length equality, and should only be considered in patients who absolutely refuse a prosthetic option, have bilateral disease, or have phocomelia affecting the upper extremities.

Age Strategies

The majority of type 1 congenital short femurs require at least two lengthenings. As the expected discrepancy at skeletal maturity increases, the number of lengthenings required to equalize limb length discrepancy increases (Table 1). The amount of lengthening that can be performed in the femur at any one stage depends on the initial length of the femur. Generally 4 to 6 cm can be performed safely in toddler (age 2 to 4 years) femurs. In children older than age 6 years, at least 6 to 8 cm is usually possible. In adolescents and young adults, 8 to 12 cm may be possible. Combined femur and tibia lengthenings allow greater lengthening amounts. Tibial lengthening of up to 5 cm can be combined with the above femoral lengthening amounts. Toddler lengthening should be considered only in children with a well developed hip joint including an ossified femoral neck. Toddler lengthening is usually limited to 4 to 6 cm although I have safely performed up to 8 cm in the older toddlers if knee motion is well maintained. The two main advantages of toddler lengthening are growth stimulation and reduction of prosthetic needs. Growth stimulation was seen consistently in my toddler femoral lengthenings. It is a progressive stimulation in some cases, whereas in others it returns to the previous growth rate. With a reduction of the leg length difference, toddlers are able to reduce one level of prosthetic/orthotic need. This means going from a long leg prosthesis to an ankle-foot orthosis (AFO) and shoe lift, from an AFO and shoe lift to only a shoe lift, or from a shoe lift to no lift. The complication rate in this group is no higher than in older children in my experience.

Table 1 Treatment strategies and timing of reconstructive stages in management of congenital short femur

Discrepancy at Maturity	Treatment Strategy Options	Timing
≤ 6 cm	1 lengthenings	over age 6
7–12 cm	2 lengthenings	toddler (< 5 cm) + age 8–10 (< 8 cm)
	1 lengthening	toddler (< 5 cm) or age 6 (< 8 cm) + epiphyseodesis (< 5 cm)
12–16 cm	2 lengthenings	toddler (< 5 cm) or age 6–8 (< 8 cm) + age 10–12 (8–10 cm)
16–20 cm	2 lengthenings	toddler (< 5 cm) or age 6–8 (< 8 cm) + age 10–12 (8–10 cm) + tibial (< 5 cm) during one of the femoral lengthenings
	3 lengthenings	toddler (< 5 cm) + age 8–10 (6–8 cm) + age 10–14 (8–10 cm)
	2 lengthenings	toddler (< 5 cm) or age 6–8 (< 8 cm) + age 10–12 (8–10 cm) + epiphyseodesis (< 5 cm)
21–25 cm	3 lengthenings	toddler (< 5 cm) + age 8–10 (6–8 cm) + age 12–16 (10–12 cm)
	3 lengthenings	age 6–8 (< 8 cm) + age 10–12 (8–10 cm) + age 12–16 (8–12 cm) + tibia (< 5 cm) during one of the femoral lengthenings
	2 lengthenings + epiphyseodesis	age 6–8 (< 8 cm) + age 10–12 (8–10 cm) + epiphyseodesis (< 5 cm), tibial lengthening (< 5 cm) with one of the femoral lengthenings > 25 cm
	3 lengthenings + epiphyseodesis	
	4 lengthenings	

Lengthening Reconstruction Surgery Versus Prosthetic Replacement Surgery

My results in 54 patients to date with congenital short femur syndrome are given in Table 2. Many of these patients have completed only one lengthening, whereas others have completed as many as three lengthenings. In a separate study of 70 Ilizarov femoral lengthenings, clinical and radiographic results were compared between congenital, posttraumatic, and developmental cases undergoing lengthening (unpublished data, 1996). There was no significant difference in results based on etiology.

Because of the improvements in results of lengthening with the introduction of Ilizarov's techniques, more authors are recommending limb reconstructive surgery. However, currently the presence of a pseudarthrosis and the status of the hip are used as the primary deciding factors for limb reconstructive surgery versus amputation and prosthetic fitting. It should be emphasized that the hip status does not change after amputation and prosthetic fitting. I argue, therefore, that the status of the hip should not be a

Table 2 Results of lengthening

Type	No.	Results*			
		Excellent	Good	Fair	Poor
1a	45	32	10	3	0
1b	2	0	1	1	0
2a	1	0	1	0	0
2b	3	1	1	1	0
3a	1	0	1	0	0
3b	2	0	2	0	0

* Result score is based on clinical subjective, clinical objective, and radiographic criteria

major deciding factor for amputation and prosthetic fitting. In fact, hip procedures used for limb reconstructive surgery are useful to stabilize the hip and improve gait even after amputation and prosthetic fitting. For me, the status of the knee is the deciding factor to recommend limb reconstructive surgery versus amputation and prosthetic fitting. Therefore, my absolute indications for amputation and prosthetic fitting are primarily in type 3 cases. In type 2 cases it should also be considered depending on how functional the knee is and on the magnitude of the predicted discrepancy. Type 1a and b should rarely be considered for amputation and prosthetic fitting, unless there is an associated stiff knee. Finally, in type 1 congenital femoral deficiency, limb reconstructive surgery is so reliable in my hands that amputation and prosthetic fitting should be considered only when psychologic or socioeconomic reasons prevail.

One of the arguments for amputation and prosthetic fitting is the contention that limb reconstructive surgery leads to psychologic scarring and loss of childhood. In my experience, limb reconstructive surgery if properly conducted with an appropriate rehabilitation program and surgeries strategically spaced apart does not lead to obvious psychological scarring to the child. It can truly be a 'growing experience.' Limb reconstructive surgery is an investment. The child invests part of his or her childhood in order to live the majority of life as an adult with as near normal an extremity as possible. I try to complete the limb reconstructive surgery before the child enters high school whenever possible so that the formative years of body image at the time when the children are most self-conscious occur with both limbs of equal length and near normal function. In this manner, most go through a normal adolescence. The psychologic stress of wearing a prosthesis during adolescence is not well quantified by psychologic profiles performed on these individuals as adults. Therefore, it is difficult to compare limb reconstructive surgery versus amputation and prosthetic fitting.

Psychosocioeconomic stresses can play a major role in deciding between limb reconstructive surgery and amputation and prosthetic fitting. Single parents, marital difficulties, financial difficulties, drug problems, behavioral problems, learning disabilities, mental capacity, and other problems may interfere with compliance, maturity, and home stability required to undergo

limb reconstructive surgery. Amputation and prosthetic fitting is easier, more painless, and requires far less treatment assistance by the family. In situations where the family would find it difficult to comply or too stressful for the other family members, amputation and prosthetic fitting is the preferable option. Finally, successful limb reconstructive surgery requires a team dedicated to this type of treatment. It is not a procedure that should be performed casually or by surgeons inexperienced in the treatment of these patients. Experience in limb lengthening for other conditions is not sufficient to know how to successfully perform lengthening in children with congenital femoral deficiency. It requires a long commitment of time on the surgeon's part and on the part of the surgeon's team. It requires appropriate rehabilitation services. If all of these facilities are not available, limb reconstructive surgery should not be considered at that venue. The latter is perhaps the main limiting factor today in the availability of limb reconstructive surgery.

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