The following essay represents some of the recent (2001) philosophy of the ICLL for the treatment of congenital short femur. It is intended for physicians and is therefore written in highly technical medical language. We nonetheless advise parents of children with congenital femoral deficiency to read this and encourage them to ask questions. —Dror Paley, MD, FRCSCI

Lengthening Reconstruction Surgery for Congenital Femoral Deficiency
Dror Paley, MD, FRCSC

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 bone malorientation, bone malrotation, and soft tissue contractures of the hip and knee. Both deficiencies and deformities are present at birth, are nonprogressive, and are of variable degrees.

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 systems (1) showed that they were inaccurate in predicting the final femoral morphology based on initial radiographs. Furthermore, previous classification systems were designed with prosthetic reconstruction surgery (PRS) (e.g., Syme's amputation or rotationplasty plus prosthetic fitting) rather than lengthening reconstruction surgery (LRS) (equalization of limb length with realignment of the lower limb and preservation of the joints) in mind. My classification system is based on the factors that influence lengthening reconstruction of the congenital short femur.

Paley's CFD Classification (Fig. 1a)

Type 1: **intact femur** with mobile hip and knee
- normal ossification proximal femur
- delayed ossification proximal femur

Type 2: **mobile pseudarthrosis** with mobile knee
- femoral head mobile in acetabulum
- femoral head absent or stiff in acetabulum

Type 3: **diaphyseal deficiency** of femur
- knee motion > 45 degrees
- knee motion < 45 degrees

Knee joint mobility/deficiency rather than hip joint mobility/deficiency is the most important determining factor for functional outcome and re-constructibility of congenital short femora. Previous classifications (such as the Aitken classification) emphasize the extent of hip deficiency, which is used as a guideline to indications for amputation and prosthetic fitting (PRS) despite that the hip does not change with PRS. Types 1 and 2 are the most re-constructible. 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 subclassification according to factors that

require correction before lengthening can be performed. 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 operations that are required before starting lengthening and therefore may affect the decision of LRS versus PRS.

Type 1 CFD: Intact Femur

Hip and knee considerations: This group is the most re-constructible. Before lengthening, significant bone deformities and soft tissue contractures of the hip and knee should be reconstructed. At the hip, if the acetabulum has a center edge angle greater than 20 degrees, the neck shaft angle is greater than 110 degrees, the greater trochanter is not significantly overgrown such that the medial proximal femoral angle is not less than 70 degrees, then no hip surgery is required before the first lengthening. At the knee, if the fixed flexion deformity is less than 10 degrees and the patella does not track and/or subluxate or dislocate laterally and if there is no evidence of significant rotary subluxation or dislocation of the tibia on the femur with flexion and extension, then the knee does not require surgical reconstruction before lengthening. If, however, any of these criteria are not met, the hip and/or knee should be reconstructed before the first lengthening is performed.

Acetabular dysplasia: It is very common for even mild cases of CFD to have acetabular dysplasia, which predisposes the femoral head to subluxation during lengthening. A center edge angle of less than 20 degrees before femoral lengthening is an indication for pelvic osteotomy. The acetabular dysplasia of CFD is not like that of developmental dysplasia of the hip. The deficiency is not predominantly anterolateral. The deficiency is more superolateral, often with a hypoplastic posterior lip of the acetabulum. Therefore, the Dega osteotomy is our preferred method to improve coverage rather than the Salter or Millis-Hall modification of the Salter (combining innominate bone lengthening with the Salter). This is best done when the patient is 2 years of age but can be done even in adolescents if the triradiate cartilage remains patent.

Proximal femoral deformities: The proximal femoral deformity of CFD is not a simple coxa vara in most cases. Until recently, the pathoanatomy of this deformity was not clear. We now understand that it is a complex combination of bone deformities in the frontal, sagittal, and axial planes combined with soft tissue contractures affecting all three planes. The severity of these deformities is often mild in type 1a cases but is usually severe in type 1b cases.
The obvious coxa vara is associated with an abduction contracture of the hip. If the coxa vara is corrected on its own, the abduction contracture will be uncovered. This contracture will prevent full valgus correction and/or prevent the hip from coming back to a neutral position relative to the pelvis. The abduction contracture causes a fixed pelvic tilt, which makes the limb length discrepancy (LLD) appear less than before surgery. In the face of an open growth plate or a non-ossified neck or subtrochanteric segment, as in type 1b cases, the abduction contracture leads to recurrence of the coxa vara.

Fixed flexion deformity of the hip is also often present in these cases. The magnitude of the fixed flexion deformity is often masked by an extension deformity in the bone of the proximal femur.

External rotation deformity of the distal relative to the proximal femur (retroversion) is always present. This is because of a combination of bone torsion and contracture of the piriformis muscle.

The correction of these deformities is performed with a new surgical procedure developed by Dr. Paley. We call this the super hip procedure because of its complexity.

Super Hip Procedure

Step 1, incision and reflecting anterior flap: A long lateral incision is made over the posterolateral border of the femur, curving gently anteriorly at its proximal end to end at the iliac wing 3 or 4 cm posterior to the anterior superior iliac spine. The distal end of the incision is at the knee joint line. The fatty tissues are dissected off the fascia lata, reflecting the flap of skin and subcutaneous tissues anteriorly. The limit of the dissection is the interval between the tensor fascia lata and the sartorius.

Step 2, reflection fascia lata and tensor fascia lata muscle: The fascia is cut at this interval with care to avoid injury to the lateral femoral cutaneous nerve. The fascial incision is extended to the lateral border of the patella ending at the tibia. The posterior fatty flap is elevated until the intermuscular septum is in view. Because the incision is biased posteriorly, this will usually be in line with the lateral incision or just slightly posterior to it. The fascia lata is cut longitudinally, in line with the intermuscular septum. If knee ligamentous reconstruction is not required, the fascia lata is cut distally at the tibia and is reflected proximally. At the proximal end, the tensor fascia lata muscle is dissected free of the sartorius and is reflected proximally and posteriorly on its posterior pedicle. The anterior vascular pedicle can be cauterized and cut. Care should be taken to separate the tensor muscle from the underlying gluteus medius. The two muscles may be adherent to each other. The interval between the muscles can be discerned because the gluteus inserts on the greater trochanter but the tensor passes over of the trochanter.

Step 3, flexion contracture release: The dissection is carried beneath the sartorius to find the anterior inferior iliac spine. The rectus femoris tendon insertion is identified at the inferior spine. The direct and reflected head rectus femoris tendons are transected. Just medial to the rectus is the psoas muscle and tendon. The tendonous part is cut, leaving the muscle to bridge the gap. The remaining flexion contracture is from the abductor muscles and is addressed in the next step of the procedure.

Step 4, abduction and external rotation contracture releases: The abductor tendons (gluteus medius and minimus) insert into the greater trochanter and extend distally to become confluent with the quadriceps origin on the greater trochanter. The tendonous portions of both muscle groups can be sharply dissected and reflected together, maintaining the longitudinal continuity of the musculotendinous units. Neither muscle group can retract or shorten in this manner. In this step, the posterior aspect of the glutei is identified and followed to the posterior border of the greater trochanter. The posterior border of the vastus lateralis at the intermuscular septum is similarly identified and dissected free of the femur extraperiosteally. This line is continued proximally along the posterior aspect of the greater trochanter. Because the tendonous covering of the trochanter is thin, it is important to take a thin layer of cartilage with the flap. The flap of conjoint gluteus-quadriceps tendon is sharply dissected and reflected from posterior to anterior

off the trochanter until the intertrochanteric line. During this release, the piriformis tendon may be identified and should be released from its trochanteric insertion. Release of the piriformis tendon permits the femur to internally rotate. The last step in the abduction contracture release is to release the capsule of the femur from the tip of the trochanter. If one stays on the cartilage, this capsular release will be extra-synovial. Because of the flexion contracture, femoral retroversion, and especially the extension deformity of the femur, the greater trochanter is very posterior. This is the reason the incision is posterior. This also makes the reflection of the conjoint tendon initially difficult.

Step 5, proximal femoral osteotomy and fixation: With the abduction, flexion, and rotation contractures all released, the severity of the coxa vara can finally be fully appreciated by placing the femoral head and neck in neutral orientation to the pelvis. To create a normal neck shaft angle, a 2-mm Steinmann pin is drilled from the lateral cortex of the femur into the piriformis fossa at the desired inclination to the femoral neck (125 degrees). The other reference angle is the line from the tip of the greater trochanter to the center of the femoral head. The angle between the pin and this imaginary line is the medial proximal femoral angle, which should normally measure 84 degrees. To facilitate orienting this pin, an arthrogram should be obtained at this stage. Depending on the diameter of the Rush rod to be used for fixation. an appropriate diameter of cannulated drill is chosen. For a 1/8-in (3 mm) Rush rod, a 3.2-mm cannulated drill is used. For a 3/16-in (4.5 mm) Rush rod, a 4.8-mm cannulated drill is chosen. Subperiosteal dissection around the femur is then performed in the subtrochanteric region. A Gigli saw is passed at this level, and the femur is cut perpendicular to the distal diaphysis. Because the proximal drill hole passes through the lateral femoral cortex, it is decorticated perpendicular to the drill hole. If there is enough bone, this cut can be made, stopping short of the medial cortex and creating a notch with a medial wall. The distal piece is slotted into this notch. The medullary canal of the distal femur is drilled with a solid drill bit of the same diameter as was used for the proximal end. Because correction of the severe coxa vara will significantly lengthen the femur, a 1- to 2-cm segment of femur should be excised from the distal femur to allow reduction of the two segments. The Rush rod is inserted into the proximal segment. The distal segment is reduced to the proximal and the Rush rod advanced across the osteotomy. Although this fixation is probably sufficient, for additional rotatory control, a tension band wire is added. A 1.8-mm drill hole is created in the lateral cortex of the distal femur from anterior to posterior using a K-wire. An 18-gauge wire is then threaded through this hole and twisted in a figure-eight fashion around the Rush rod so that it is medial to the rod. Each limb of the wire is tensioned to compress the osteotomy. If there is delayed ossification, as in the type 1b cases, a cannulated screw is placed up the neck of the femur from the lateral side. This is left in until the ossification of this region is completed. To avoid impeding growth, a partially threaded screw, which is left long at the lateral cortex can be used.

Step 6, pelvic osteotomy: Because of the extensive exposure, it is not necessary to split the iliac apophysis to perform the Dega osteotomy. The ilium is exposed by retracting the hip abductor muscles from anterior without releasing the muscle off the crest. The dissection is carried back to the ischial trunk of the ilium until the triradiate cartilage. The osteotomy is a circular line on the lateral cortex from the anterior inferior iliac spine anteriorly to 2 cm proximal to the joint midlaterally to the triradiate cartilage posteriorly. The osteotomy is inclined toward the triradiate cartilage medially. In the anterior third, the osteotomy is bicortical, whereas in the remaining

part, it is unicortical. The osteotomy is levered distally to cover the femoral head. It is sometimes necessary to re-repair the lateral capsular attachment to prevent lateral subluxation.

Step 7, tendon repair and closure: The conjoint tendon is sutured directly into the cartilaginous greater trochanter with absorbable suture. This is done with the femur in neutral abduction. The tensor fascia lata is also sutured to the greater trochanter to augment the abduction strength of the hip. The incision is closed in layers over a Hemovac drain, including Scarpa's fascia, deep dermis, and subcuticular layers. The patient is placed in a spica cast in full hip extension and neutral abduction and rotation.

Instability of Patella or Tibia and Knee Flexion Contracture

Subluxation or dislocation of the patella or tibia with flexion or extension, respectively, necessitate a stabilizing procedure before lengthening. Isolated anteroposterior instability of the tibiofemoral joint without knee joint dislocation or rotatory subluxation does not need to be addressed before performing lengthening. Isolated subluxation or dislocation of the patella should be treated before lengthening begins. The method of knee reconstruction developed by Dr. Paley is based on a combination of elements from the Langenskiöld procedure (4), designed for congenital dislocation of the patella, the MacIntosh procedure (5), designed for extra-articular reconstruction for anterior cruciate deficiency, and Grammont procedure (6), designed for recurrent dislocation of the patella. This knee stabilization procedure may be performed at the same sitting as the pelvic osteotomy or super hip procedure because both need to be in a long leg cast postoperatively. When performed together with a super hip procedure, the fascia lata is reflected from proximal to distal instead of distal to proximal.

Knee reconstruction surgical techniques: For all these procedures, 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 longitudinally. The fascia lata is transected as proximally as possible and reflected distally until its insertion onto the tibia. The biceps tendon is z lengthened.

Posterolateral tibiofemoral instability pattern: To prevent the tibia from externally rotating on the femur (associated with subluxation/dislocation of the patella), the fascia lata is routed over the patellar tendon and knee joint capsule to insert into the medial femoral metaphysis (we refer to this as a reverse MacIntosh procedure). An 8-mm drill hole is made across the femoral metaphysis from the medial side. The fascia lata is pulled through from the medial side and secured with a ligament screw. This new ligament prevents external rotation of the tibia on the femur and tightens with knee flexion. It does not block knee flexion. Extra-articular ligament reconstructions have the reputation of stretching out with time. Because the fascia lata is anchored proximal to the distal femoral physis, it is continually retensioned by growth.

Anterolateral tibiofemoral instability pattern: If the luxation pattern is anterolateral, the fascia lata is looped over itself after passing under the lateral collateral ligament or the lateral capsule (children with this condition often do not have a well-defined lateral collateral ligament) and through a subperiosteal tunnel just proximal to the insertion of the intermuscular septum and proximal to the growth plate. After passing through this tunnel, it is looped over itself to reattach

to the tibia and form a lateral collateral-like ligament (similar to the MacIntosh procedure [5]) (Fig. 6). This tightens the tibia into external rotation.

Because the instability pattern is often mixed or unclear and because there is sufficient fascia lata for two extra-articular ligament reconstructions, my preference is to split the fascia lata and take one limb of it anteromedial and the other posterolateral.

If the patella is subluxed but not dislocating, 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 can be displaced medially by using a modification of the Grammont technique. The patellar tendon is elevated off the cartilaginous apophysis but left attached distally to a flap of periosteum. It is sutured 1 cm medial to its insertion with absorbable suture. If the patella tracks well in this position, no further patellar stabilization is required. This assumes that the tibia does not have posterolateral instability. Posterolateral rotatory instability displaces the patellar tendon laterally each time the tibia rotates laterally. This promotes lateral subluxation. This would necessitate the reverse MacIntosh procedure described above. If the patella still tracks laterally, the modified Langenskiöld procedure should be performed.

Modified Langenskiöld procedure (4): 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. The synovium is sutured closed in a longitudinal direction. 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 sutured to the patella circumferentially. The capsule is stitched over the patella on the medial side and left open laterally. If the reverse MacIntosh procedure is used, the fascia lata should not be fixed in place until after the Langenskiöld repair is completed. After multiple layer wound closure, the knee is put in a cylinder cast for 6 weeks and then active and passive motion exercises are performed.

Knee flexion contracture release: If there is a knee flexion deformity greater than 10 degrees, it can be treated by posterior capsular release. This is often performed in combination with a super hip procedure or one of the above knee reconstructions. To avoid direct surgical and indirect stretch injury to the peroneal nerve, this nerve should be identified and decompressed at the neck of the fibula. Next, the biceps tendon should be z lengthened. The lateral head of the gastrocnemius should be released from the femur. The lateral capsule is identified and incised, and the posterior capsule is dissected free of the posterior popliteal fossa contents. This dissection is performed from lateral to medial. An instrument is inserted through this space to tent the skin on the medial side. The medial skin is incised and the medial gastrocnemius head identified and released off the femur. The capsule is dissected free of the medial popliteal fossa. Great care must be taken to avoid injury to the popliteal vessels, which lie adjacent to the medial half of the posterior capsule. The capsule can then be cut under direct vision from both sides. The knee fixed flexion deformity can then be corrected by extending the knee. The collateral capsule and ligaments are left intact. If in addition to this, there is a muscular contracture of the medial hamstrings. A small posteromedial incision can be made to recess the semi-tendinous and semi-membranous tendons. The gracilis can be recessed through the medial incision.

The above hip and knee problems must all be addressed before beginning the femoral lengthening procedures.

Femoral Lengthening of Type 1 CFD
Choice of osteotomy level for lengthening of the congenital short femur: In most cases, we prefer to lengthen the femur using a distal osteotomy. Distal osteotomies have the advantage of a broader cross-sectional diameter for better bone formation and less deforming forces from the adductors and hamstrings. Distal osteotomy lengthening is closer to the knee joint and therefore has greater effect on knee range of motion and on knee subluxation. Proximal osteotomies have less effect on knee range of motion but are more prone to poor bone consolidation, especially a narrow or partially deficient regenerate bone formation. With the external fixator only method of bone lengthening, there is a higher rate of fracture after removal of fixation in the proximal than in the distal lengthening groups. Proximal osteotomies should be reserved for the technique of lengthening over nails because the nail prevents deformation of the proximal femur both during and after removal of fixation and almost eliminate the risk of fracture.

The other considerations for level of osteotomy are the associated deformities of the hip and knee. The external rotation deformity of the femur with CFD should be corrected only by using a proximal osteotomy. Because the quadriceps muscle is in a normal relationship to the knee joint and because most of the quadriceps muscle originates distal to the level of a proximal femoral osteotomy, a proximal femoral derotation osteotomy reorients the quadriceps relative to the knee joint. A distal osteotomy leaves the bulk of the quadriceps muscle attached proximally in a lateral position and rotates the knee medially, thus increasing the effective Q angle and increasing the tendency to lateral subluxation/dislocation of the patella. Varus deformity of the hip or proximal femoral diaphysis is corrected using a proximal osteotomy, whereas valgus deformity of the knee is corrected using a distal osteotomy.

If the femur has undergone previous super hip reconstruction, the proximal femoral deformities should already be corrected and no proximal osteotomy is required at the time of lengthening. If there is previously untreated or residual/recurrent varus, flexion, and/or external rotation deformity despite previous osteotomy, these deformities can be addressed at the time of lengthening by acute correction with a proximal femoral osteotomy. This proximal osteotomy should not be used for lengthening because of the bone healing considerations noted above.
In the distal femur, an osteotomy is made to gradually correct the distal femoral deformities of valgus and flexion. As noted above, this region of the femur has a wider cross-sectional area than the proximal femur and is not in the zone of sclerotic poorly healing bone (Fig. 8). Therefore, the regenerate bone from the distal femur is wider and stronger and subjected to less bending forces than in the proximal femur.

In older children with wider medullary canals (>7 mm), lengthening over nails can be performed (Fig. 8). 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 apophysis (1) and avascular necrosis of the femoral head (1). To avoid the latter, we use a greater trochanteric starting point and a nail with a proximal bend (e.g., humeral or tibial). To avoid a coxa valga deformity, we prefer to use this technique in patients with some coxa vara. The apophysiodesis created by the nail can lead to gradual

correction of the coxa vara. Fixator only lengthening is the method we usually use for the first lengthening. Lengthening over nails is usually the method we chose for the second lengthening, if the anatomic dimensions and deformities mentioned above permit.

Soft tissue releases for lengthening in CFD: Soft tissue releases are essential in conjunction with lengthening to prevent subluxation and stiffness of the knee and hip. If a super hip or knee reconstruction has already been performed, there is clearly no need to release the fascia lata, which was excised. Our approach to muscle-tendon lengthening-releases is to perform the releases at the time of the index procedure if there is evidence of a contracture or muscle tightness at the time of the lengthening surgery (index procedure). If there is no contracture or tightness at the time of the index procedure, the soft tissue releases should be delayed until these soft tissues become contracted.

Before surgery, the range of motion of the hip and knee should be evaluated and the presence of contractures or limitation in muscle lengthening tests should be identified. The muscle lengthening tests are the straight leg raising test (popliteal angle) for the hamstrings and the prone knee flexion test (Ely test) for the rectus femoris. If a patient is able to straight leg raise so that the hip is at 90 degrees of flexion and the knee is in full (popliteal angle = 0 degrees) extension, the hamstring muscles are not tight and require no treatment before lengthening. If the patient is able to fully flex the knee while prone without the pelvis flexing at the hip, the rectus femoris is not tight and requires no treatment at the index lengthening procedure. If there is a popliteal angle greater than 0 degrees or a prone knee bend less than the supine knee bend (or pelvic flexion with prone knee flexion [positive result of Ely test]), the hamstrings and rectus femoris, respectively, are tight and will lead to contractures during lengthening. In such cases, the medial and lateral hamstrings should be recessed through a single midline incision proximal to the knee and to reduce the popliteal angle to 0 degrees and the rectus femoris proximal tendon should be released off the anterior inferior iliac spine through a small groin incision to increase the prone knee bend to the range of the supine knee flexion. If there is no muscle length tightness or contracture initially, the releases should be performed approximately 6 weeks later when these same muscles become contracted. This is the rationale of acute versus delayed releases.

If the fascia lata has not been excised, it should be released, usually on a delayed basis. The entire fascia lata is transected at the level of the proximal pole of the patella. The lateral biceps can be safely recessed through the same incision. The fascia of the tensor fascia lata muscle may lead to hyperlordosis and hip flexion contracture. This can be released through the same small incision used to release the rectus femoris tendon. For distal femoral lengthenings, it is not necessary to release the hip adductors. For proximal lengthenings, adductor release is very important. Most of these tissues are released on a delayed basis unless a contracture is present. Botulinum toxin injected at the time of surgery is useful to temporarily weaken or paralyze the quadriceps, hamstrings, and adductors. It seems to reduce muscle spasm and pain and thereby improve the ease of knee flexion range of motion at physical therapy.

Knee instability consideration: Almost all cases of CFD can be assumed to have hypoplastic or absent cruciate ligaments with mild to moderate anteroposterior instability. Some also have mediolateral and torsional instability. Despite this, the knee tracks normally preoperatively and there is no indication to perform ligamentous reconstruction in most cases. The significance of the knee instability to lengthening is the tendency toward subluxation of the knee with

lengthening (Fig. 8). Knee subluxation with lengthening is usually posterior or posterior plus external rotation but can also be anterior. Posterior subluxation can occur only with knee flexion. Therefore, to prevent posterior subluxation, some people recommend splinting the knee in extension throughout the distraction phase (1). This promotes knees stiffness while protecting the knee from subluxation. Dr. Paley prefers to protect the knee by extending the fixation to the tibia with hinges. The hinges permit knee motion while preventing posterior and anterior subluxation. This is easily performed 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. 6). 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 partly caused by an anterior deficiency of the distal femur (the lateral view radiograph of the knee shows a lack of the anterior protuberance 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 flexion required to relocate the knee; Dr. Paley, however, prefers the soft tissue reconstruction procedures described above better than extension osteotomy in these instances because knee extension osteotomy leads to loss of knee flexion.

Distal femoral lengthening technique: All the acute soft tissue releases are performed first. If soft tissue releases are to be performed on a delayed basis, proceed directly with the frame application. If a proximal femoral derotation, valgus, and/or extension osteotomy is needed, the proximal pin is inserted into the proximal femur with the hip in the position in which it will lie after the correction. For example, for an internal rotation osteotomy, the proximal pin should be inserted with the knee in external rotation. For a valgus osteotomy, the proximal pin should be inserted with the hip adducted. For an extension osteotomy, the proximal pins should be inserted with the hip flexed. For correction of varus, flexion, and external rotation, the femur should be externally rotated and crossed over the other thigh to adduct and flex the hip. This places the hip in the true neutral position. The first half-pin is placed from lateral to medial in the frontal plane, parallel to the line from the tip of the greater trochanter to the center of the femoral head. The plan is to attach the proximal arch parallel to the line from the tip of the trochanter to the center of the femoral head , the middle ring perpendicular to the mechanical axis of the shaft of the femur (7 degrees to the shaft), and the distal ring parallel to the knee joint line. After the osteotomies are performed, when all the rings and the arch are parallel, the mechanical axis of each segment will be aligned and the joint orientation of the hip and knee will be parallel. A second proximal half-pin is inserted on the proximal arch from 30 degrees anterolateral to the first pin. The proximal arch is perpendicular to the floor with the leg is crossed over and rotated as described above for correction of deformity. Two Ilizarov rings properly sized for the distal femur are applied to a distal femoral reference wire, which is parallel to the knee joint. For young children, we obtain arthrograms to better outline the cartilaginous femoral condylar line. The arthrograms are also useful to visualize the posterior femoral condyles for hinge placement. Conical washers or hinges are used between the two distal rings because of the valgus of the distal femur. The rings are at the valgus deformity angle to each other. A lateral half-pin is inserted into the mid-segment of the femur. This pin is at 7 degrees to the shaft of the bone. At that point, the proximal subtrochanteric osteotomy can be performed. This is performed percutaneously by making multiple drill holes and then using an osteotome. At the osteotomy

site, the bone is internally rotated, laterally translated, and then angulated into valgus and extension to correct all components of the deformity. The order of correction is important to achieve the necessary displacement without loss of bone-to-bone contact and stability. Two additional half-pins are inserted and fixed onto the distal ring, one from posteromedial and one from posterolateral between the quadriceps and the hamstring muscles. One more middle pin is inserted. In small children, all half-pins are inserted by using the cannulated drill technique. This involves insertion of a wire first, then a cannulated drill, and then a half-pin. This technique permits very accurate placement of large diameter pins in narrow bones to avoid eccentric placement. Eccentric placement of drill holes and half-pins in the femoral diaphysis can lead to fracture. The distal femoral osteotomy is performed percutaneously, using multiple drill holes and an osteotome. The only wire used is removed to avoid tethering of the quadriceps and fascia lata. The last step is to extend the fixation to the tibia using hinges. The center of rotation of the knee is located at the virtual intersection of the posterior femoral cortical line and the distal femoral physeal line (1). When the two posterior femoral condyles are seen to overlap on the lateral view, it is important that the distal femoral ring, which is parallel to the knee, appear to be perpendicular to the x-ray beam. The medial and lateral skin is marked at the location of the planned hinge placement. A single half-ring is attached to two threaded rods from the hinges. This half-ring is oriented perpendicular to the tibia with the knee in full extension. The first half-pin is inserted from anterior to posterior into the tibia. After fixing this pin to the proximal tibial half-ring, the knee is flexed and extended through a range of motion. If this range feels frictionless, a second tibial half-pin is added. Finally, a removable knee extension bar is inserted between the distal femoral and the tibial half-ring.

Rehabilitation and follow-up during lengthening: Femoral lengthening requires close follow-up and intensive rehabilitation 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. Radiographically, the distraction gap length, regenerate bone quality, limb alignment, and joint location are assessed.

Physical therapy [link to physical therapy section] 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, physical therapy is resumed. During the distraction phase, one to two formal sessions each day (45 to 60 minutes each session) with a physical therapist are required. In addition, at least two home sessions per day (30 minutes each session) are recommended. The more therapy the patient receives, the better is the potential functional result and the faster is the rehabilitation after fixator removal. Inpatient rehabilitation is often the only practical method of achieving this quantity of therapy. The philosophy of therapy for lengthening is very different from that for other orthopaedic surgical procedures. After most orthopaedic procedures, the patient is at his or her worst after surgery and gradually recovers. One week after surgery, patients undergoing lengthening 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. One can think of the lengthening surgery 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, we 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. Weight bearing is encouraged and allowed as tolerated.

Knee flexion should be maintained at greater than 45 degrees. If knee flexion is 40 degrees or less, the lengthening should be stopped and the knee rehabilitated more. If after a few days the knee flexion improves, lengthening may resume. Our motto is to never sacrifice function for length. More length can be obtained with an additional lengthening, but we 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 part time 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 knee cap and a depression of the tibia relative to the kneecap (ski hill sign). Extension of the external fixation across the knee with hinges prevents posterior subluxation (1).

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

Nerve injury from surgery or distraction is unusual with femoral lengthening. To avoid peroneal nerve injury from the pins, the posterolateral pin should not enter posterior to the biceps tendon. During distraction, if the patient complains of pain in the dorsum of the foot or asks for frequent massage of the foot, this is most likely referred pain from stretch entrapment of the peroneal nerve. More advanced symptoms include hyper- or hypoesthesia in the distribution of the peroneal nerve or weakness of the extensor hallucis longus muscle. A nerve conduction study (our 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. If the nerve problem is identified early, it can be treated by slowing the rate of distraction. If, despite that, symptoms continue or motor signs develop, the peroneal nerve should be decompressed at the neck of the fibula, including transverse fasciotomy of the lateral and anterior compartment and release of the intermuscular septum between these compartments.

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 achieved during 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 (11). Limb length equalization should be based on full length standing radiographs. Limb alignment is assessed for the femur and tibia both separately and in combination. Separately, the joint orientation of the knee should be measured using the malalignment test (1). Axial deviation from lengthening (procurvatum and valgus for distal femoral lengthening and procurvatum 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 under-corrected to compensate for the tibial deformity. The tibia should be corrected separately, either during the same treatment or at a later treatment.

Complete failure of bone formation is very unusual. Partial defects are not uncommon. Resection of the fibrous tissue in these defects and cancellous bone grafting may become necessary to reduce the external fixation time and prevent fracture after frame removal.

After removal of the frame, a one-legged spica cast is used. Depending on the degree of healing no, partial, or complete weight bearing are permitted with the cast on. The cast is usually removed 1 month later, and physical therapy to regain knee motion is begun. The knee should be in full extension in the cast to prevent contracture and subluxation.

Difference in treatment of types 1a and 1b: In general, CFD type 1a (normal ossification) has less proximal femoral, hip, and knee deformity, deficiency, and discrepancy than does CFD type 1b (delayed ossification). This is not always the case. Most type 1a cases do not require the complex super hip reconstruction. Approximately half of the type 1a cases do require pelvic osteotomy before lengthening. All type 1a cases require extension of the fixator across the knee to protect the knee joint. The distinction between types 1a and 1b should be made in infancy because the natural history of type 1b is to ossify. Therefore, adult type 1b cases generally appear to be severe type 1a cases. The strategy of treatment for type 1b is to correct all the associated deformities, which will allow the proximal femur and hip to be more normally oriented and accept more axial loading. The response to the anatomic change is ossification of the proximal femur and conversion from type 1b to type 1a. We do not lengthen type 1b cases until they convert to type 1a. This conversion usually occurs within 2 years of the super hip procedure. Our preference for the first lengthening is between the ages of 2 and 4 years. Patients with type 1a CFD typically undergo their first lengthening at age 2 years, whereas patients with type 1b CFD undergo their lengthening closer to age 4 years.

Treatment CFD Type 2

The goal of treatment in this group is to reconstruct the hip and then perform lengthening. Although type 2a differs from type 2b by the presence of a mobile femoral head, attempts at connecting these together is often met with failure or stiffness of the hip. Therefore, the preferred option is to reconstruct the hip without directly joining the proximal femur to the femoral head and neck. This is accomplished by combining a pelvic support osteotomy for hip reconstruction

with a distal femoral lengthening and realignment osteotomy. This combination is called the *Ilizarov pelvic support osteotomy* to distinguish it from other pelvic support osteotomies that do not include a second osteotomy for distal femoral lengthening and realignment.

In very young children with very short femora, the femur may be too small to perform both the pelvic support and the distal lengthening osteotomies. In such cases, pins are extended to the pelvis to prevent proximal migration during lengthening. The lengthening is then performed through a distal femoral osteotomy, much in the way described previously. In older children, the pelvic support osteotomy is performed at the level at which the proximal femur crosses the ischial tuberosity in the maximum cross-legged radiograph. The amount of valgus is equal to the total amount of adduction of the hip plus 15 degrees of overcorrection. The proximal osteotomy should also be internally rotated and extended. The amount of rotation is judged by the position of the knee relative to the hip in maximum adduction. The amount of extension depends on the amount of hip fixed flexion deformity. The level of the distal osteotomy is planned by extending the line of the tibia proximally and seeing where it intersects a line perpendicular to the pelvis passing through the midproximal segment of femur. In general, a distal osteotomy is preferred because of bone healing considerations, even if the hinges have to be placed more proximal. The external fixator must be extended to the tibia with hinges, as previously discussed.

Treatment of Type 3a: Diaphyseal Deficiency, Knee Range of Motion > 45 Degrees

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 greater than 45 degrees of motion (type 3a), usually with a 45-degree knee flexion deformity, whereas others have a stiff knee with a less than 45-degree range of motion (type 3b). The most predictable and reliable treatment option in these cases remains PRS (rotationplasty or Syme's amputation). LRS has a role in these cases and can equalize LLD. Because the numbers of these patients treated by LRS is small, the ultimate functional result for these cases is still not predictable or reliable. There are several new and promising reconstructive methods that we are pursuing at the **ICLL**. There is also a new and improved method of rotationplasty that we are using, which even improves the results of PRS. These are described below.

LRS Approach for Diaphyseal Deficiency Cases: Type 3

LRS is most applicable to type 3a cases in which there is a functional range of knee motion present. In type 3 cases, there is usually a knee flexion deformity present. In addition, there is a hip flexion contracture present. Both of these are treated by soft tissue release using the same approach as that described above for type 1 cases. The fascia lata is reflected proximally, and the quadriceps and abductors are elevated off the proximal femur. The psoas tendon is absent, but the rectus and sartorius are present. Any fibrous femoral anlage is resected, and frequently, some of the cartilaginous femoral anlage may need to be trimmed. The proximal femur is freed from these attachments, including the hip capsular remnants, permitting it to move proximally without a soft tissue tether. This is important, especially for the acute correction of the knee contracture. The long lateral incision is extended distally, and the peroneal nerve decompressed and protected. Because the femur is so short, it is advisable to explore the peroneal nerve before the proximal release and then follow it proximally to the hip joint region. This will prevent injury to

the sciatic nerve as it passes very near the dissection around the hip capsular remnant. The medial and lateral approach knee flexion contracture release of the posterior capsule is performed, and the knee joint is fully extended. A 2-mm Steinmann pin is drilled across the knee joint from the femur into the tibia to maintain full knee extension. During the same operation, a monolateral fixator is placed from the pelvis to the femur and tibia. The most important and strongest pelvic pin is one that is placed from the anterior inferior iliac spine and oriented toward the greater sciatic notch (using the cannulated drill technique). Another pin from the lateral side in the supra-acetabular region is attached to the fixator. One pin is used in the distal femur and two in the tibia.

The femur and tibia are distracted away from the pelvis. Although distraction may begin at 1 mm per day, to achieve the greatest lengthening possible requires decreasing this rate progressively to ¾ mm per day and finally to ½ mm per day. There is no risk of premature consolidation because there is no osteotomy. It is usually possible to transport the femur distally 10 cm below the triradiate cartilage.

At that point, a second stage procedure is performed to fill the gap. The femoral diaphysis from the other femur is resected subperiosteally and transplanted to the gap between the pelvis and femur. If a femoral head remnant is present, the graft is inserted between the femoral head and the proximal femur with an attempt to fuse the two. If not, then a fusion is attempted between the femur and the pelvis. The fixator is left in place to neutralize the forces on this fusion. The normal femur is stabilized with a monolateral external fixator. This fixator is applied before the resection and the resection incision performed between the inner two pins. The long femur is then shortened by 5 cm, and the periosteum is sutured closed over a bone graft substitute material, which is injected into the periosteal sleeve. The shortened long femur heals very quickly. Both fixators are removed at the same time. The LLD is corrected by a total of 15 cm. Most of these children, have a 30- to 35-cm predicted discrepancy. Because half of the LLD is present by age 3 years in girls and age 4 years in boys, this method of lengthening usually equalizes the limbs of children younger than 4 years. With rehabilitation, knee motion is regained. Further lengthening of the femur can be performed either by standard lengthening methods, accepting the hip fusion at least until skeletal maturity, or by means of the new Phenix self-lengthening hip prosthesis. The Phenix prosthesis was designed by Arnaud Sourieran from France as a custom lengthening prosthesis for patients with bone tumors. With this prosthesis, either the bone or the prosthesis can be lengthened to equalize the limb length. The application of this prosthesis for type 3 CFD is new, and the long-term results remain to be seen. The advantage of this approach is clearly the implantable nature of the lengthening device and restoration of mobility of the hip joint. The prosthesis will need to be changed as the child grows in size and as the discrepancy changes. At skeletal maturity, the Phenix can be replaced with a standard hip replacement prosthesis. For a patient with a stiff knee (type 3b) in whom soft tissue release is unable to reestablish knee motion, it may be possible to replace the femur with a Phenix self-lengthening total femur replacement, as is done for patients with sarcoma.

PRS Approach to CFD with Diaphyseal Deficiency: Type 3

Prosthetic fitting is possible without any surgery. The difficulties with this approach are the fixed flexion deformity of the hip and knee and the need to put the foot in equinus. For better

prosthetic fitting, a Syme's or Boyd amputation of the foot can be performed to create a better fitting stump. Furthermore, surgical correction of the hip and knee flexion contractures, as described above, can also help with prosthetic fitting. In some cases, even a pelvic support osteotomy may be considered to decrease the limb and stabilize the hip joint.

The other approach to PRS is with rotationplasty. Kraibach and Gillespie popularized the Van Ness rotationplasty for CFD patients. They used a long oblique incision. The goal was to fuse the residual knee and rotate the limb 180 degrees so that the foot pointed backward and the ankle could function as a knee joint. This was most applicable to cases in which the ankle was already at the level of the opposite knee joint. Brown modified the rotationplasty approach using a racquet-like incision and performing the rotation between the femoral remnant and the pelvis. He then fused the femur to the pelvis, thus converting the knee into a hip joint and the ankle into a knee joint. This provided improved hip stability over the Van Ness rotationplasty. One problem with the Brown method is excessive shortening of the hip muscles and lateralization of hip and lower limb. It is also more difficult to fuse the side of the femur to the side of the pelvis. Dr. Paley modified this technique by combining it with a Chiari osteotomy of the pelvis and fusing the femoral remnant to the cancellous roof of ilium. Furthermore, great care is taken to appropriately shorten the knee muscles so that they can function adequately as hip flexors and extensors. Finally, reattach the tensor fascia lata to the tibia to serve as a hip abductor. Because the knee in these patients is often contracted, release the posterior capsule and patch the previously posterior aspect of the knee (now facing anteriorly) with the excised fascia lata. Also decompress the peroneal nerve to prevent injury and allow greater rotation. Because the tibia in these patients typically has an internal rotation deformity, perform a supramalleolar osteotomy to derotate the tibia and fibula. This can be done during a second operation. A second operation is always needed to perform epiphysiodesis the distal femoral physis so as to prevent distal migration of the new hip joint away from the pelvis. The epiphysiodesis is usually performed a year after the rotationplasty. The modified Brown rotationplasty provides very functional results with better hip function and stability than does the Van Ness rotationplasty.

Age Strategies

The majority of cases of type 1 CFD require at least two lengthenings. As the expected discrepancy at skeletal maturity increases, the number of lengthenings required to equalize LLD increases. Generally, we prefer to perform the first lengthening when the patient is between the ages of 2 and 4 years. We have found that children between the ages of 4.5 and 6 or 7 years often are not prepared psychologically to deal with limb lengthening. Their cognitive level is insufficient to understand why their parents allowed someone to do this to them despite that they are beginning to be more independent and may appear to be mature enough to handle the process. The younger children do much better because their cognitive level accepts everything their parents decide without questioning it. Dr. Paley terms this understanding too little and too much at the same time. Older than 6 or 7 years, children enter the age of reason and begin to understand that they are different from other children and that they have a problem and that there is a solution. They learn to accept the solution by reason rather than by faith. The amount of lengthening that can be performed in the femur at any one stage is dependent on the initial length of the femur. Generally, 5 to 7 cm can be achieved safely in the femora of toddlers (age, 2 to 4 years). In children older than 6 years, at least 6 to 8 cm is usually possible. In adolescents and young adults, 8 to 12 cm may be possible in the femur. Combined femoral and tibial

lengthenings allow greater lengthening amounts. Tibial lengthening of up to 5 cm can be combined with the above femoral lengthening amounts. Lengthening of the femur in children younger than 6 years may be associated with sustained growth stimulation. By beginning lengthening at a young age, we are able to reduce one or more levels of prosthetic/orthotic needs. This means going from an HKAFO prosthesis to an ankle-foot orthosis and shoe lift or from an ankle-foot orthosis and shoe lift to a shoe lift only or from a shoe lift to no lift. The complication rate in this young age group is no higher than in older children, in our experience.

Dr. Paley has had the opportunity lengthen the limbs of several older patients (age, 15 to 60 years) with CFD whose parents refused PRS when they were children. He was able to successfully equalize their leg lengths in one or two lengthenings, depending on the discrepancy (the most severe case had 25 cm of equalization in two lengthening over nail treatments). Therefore, adult CFD residua are not contraindications to treatment.

Table 1: Treatment Strategies and Timing of Reconstructive Stages in Management of CFD

Predicted LLD at Maturity (cm)	Number of Lengthenings	Timing and Amount
<= 6	one	age > 6 years
7-12	two	age 2-4 yr (< 7 cm) + age 8-14 yr (< 8 cm)
	one	age 2-4 yr (< 7 cm) or age 6-10 yr (< 8 cm) + epiphysiodesis (< 5 cm)
12-16	two	[age 2-4 yr (< 7 cm) or age 6-8 yr (6-8 cm)] + age 10-14 yr (8-10 cm)
16-20	two	age 2-4 yr (< 5 cm) or age 6-8 yr (< 8 cm) + age 10-14 yr (8-10 cm) + tibia (< 5 cm) during one femoral lengthening
	three	age 2-4 yr (< 7 cm) + age 8-10 yr (6-8 cm) + age 10-14 yr (8-10cm)
	two	age 2-4 yr (< 7 cm) or + epiphysiodesis age 6-8 yr (< 8 cm) + age 10-14 yr (8 cm) + epiphysiodesis (< 5 cm)
21-25	three	age 2-4 yr (< 5 cm) + age 8-10 yr (6-8 cm) + age 12-14 yr (10-12 cm)
	three	age 6-8 yr (< 8 cm) + age 10-12 yr (8-10 cm) + age 12-16 yr (8-12 cm) + tibia (< 5 cm) during one femoral lengthening
	two	age 6-8 yr (< 8 cm) + age 10-12 yr (8-10 cm) + epiphysiodesis + tibia (< 5 cm) during one femoral lengthening + epiphysiodesis (< 5 cm)
> 25	three + epiphyseodesis	
	four	

LRS versus PRS

Our results in 54 patients with congenital short femur syndrome are as follows (result score is based on clinical subjective, clinical objective and radiographic criteria):

Type 1a (45 patients):   excellent, 32; good, 10; fair, 3; poor, 0

Type 1b (2 patients):   good, 1; fair, 1
Type 2a (1 patient):   good, 1
Type 2b (3 patients):   excellent, 1; good, 1; fair, 1
Type 3a (1 patient):   good, 1
Type 3b (2 patients):   good, 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 among congenital, posttraumatic, and developmental cases undergoing lengthening. There was no significant difference in results based on cause.

Because of the improvement in results of lengthening with the introduction of Ilizarov's techniques, more authors are recommending LRS. However, currently, the presence of pseudarthrosis and the status of the hip is used as a primary deciding factor for LRS versus PRS. It should be emphasized that the hip status does not change after PRS. We argue, therefore, that the status of the hip should not be a major deciding factor for PRS. Hip procedures used for LRS are useful to stabilize the hip and improve gait, even if PRS is chosen. The status of the knee for us is the deciding factor to recommend LRS versus PRS. Therefore, our absolute indications for PRS are primarily in type 3 cases. In type 2 cases, it should also be considered, depending on how functional the knee is and depending on the magnitude of the predicted discrepancy. Types 1a and 1b should rarely be considered for PRS, unless there is an associated stiff knee. Finally, in cases of type 1 CFD, LRS is so reliable in our hands that PRS should be considered only when psychological or socioeconomic reasons prevail.

One of the arguments for PRS is the contention that LRS leads to psychological scarring and loss of childhood. In our experience, LRS, if properly conducted with an appropriate rehabilitation program and operations strategically spaced apart, does not lead to obvious psychological scarring to the child. It can truly be a "growing experience." LRS is an investment. The child invests part of his or her childhood to live the majority of adult life with as nearly normal an extremity as possible. When multiple lengthenings are required, we space out the lengthenings so that the child does not feel as though he or she is undergoing one procedure after another. In this way, we minimize the interruption of activities. We prefer to complete the hip and knee reconstruction and one lengthening before the patient is 4 years old. In this manner, most children will have no conscious memory of these procedures. Whenever possible, we try to complete the LRS before the child enters high school so that the formative years of body image at the time when the children are most self-conscious are with both limbs of equal length and nearly normal function. In this manner, most go through normal adolescence. The psychological stress of wearing a prosthesis during adolescence is not well quantified by psychological profiles obtained from these persons as adults. Therefore, it is difficult to compare LRS versus PRS. Psycho-socio-economic stresses can play a major role in deciding between LRS and PRS. Single parents, marital difficulties, financial difficulties, drug problems, behavioral problems, learning disabilities, mental capacity, and other factors may interfere with the compliance, maturity, and home stability required for optimal success and support during LRS. PRS is easier and less painful and requires far less treatment assistance by the family. In situations in which the family would find it difficult to comply or too stressful for other family members, PRS may be the preferable option. Successful LRS 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

such patients. Experience in limb lengthening for other conditions is not sufficient to know how to successfully lengthen children with CFD. 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 these facilities are not available, LRS should not be considered at that venue. The latter is perhaps the main limiting factor today in the availability of LRS.

Figure 1. *a*, Paley classification of congenital short femur syndrome, types 1 through 3. *b*, Subclassification according to factors that need to be corrected before lengthening in cases of type 1 CFD.

Figure 2. 5-year-old girl with type 2a congenital short femur. Arthrogram with abduction (*a*) and adduction (*b*) 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.

Figure 3. *a*, Anteroposterior view radiograph of a 2-year-old girl with CFD and coxa vara. Neck shaft angle = 80 degrees. The proximal femoral physis is vertically inclined. *b*, Lateral view radiograph shows the marked degree of retroversion of the proximal femur. *c* and *d*, 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 translation is intentionally built into the correction to maintain the alignment of the head with the shaft of the femur. The neck shaft angle is restored to normal on the anteroposterior view, and the femoral head is anteverted on the lateral view. e, Anteroposterior view radiograph obtained 1 year after correction. Neck shaft angle = 124 degrees. Proximal femoral physis is more horizontally inclined.

Figure 4. *a*, 2-year-old boy with congenital short femur (LLD = 7 cm) and coxa vara. Neck shaft angle = 95 degrees relative to the proximal shaft of the femur. The proximal femoral physis is relatively horizontally inclined. There is a diaphyseal varus of 20 degrees. *b*, Treatment was by proximal femoral valgus external rotation osteotomy. The proximal coxa vara was not corrected, but the diaphyseal varus was corrected. The lengthening was performed through a distal femoral osteotomy. Final anteroposterior (*c*) and lateral (*d*) view radiographs obtained after 6 cm of limb lengthening. Residual LLD = 1 cm.

Figure 5. *a* and *b*, Posterolateral dislocation of the knee with patellar dislocation. The tibia is externally rotated on the femur. *c* and *d*, Radiographs obtained after knee reconstruction procedure. The knee is well reduced and stable. Because this patient is skeletally mature, the tuberosity was transferred instead of detaching and moving the patellar tendon. The fascia lata was fixed to the medial femoral condyle by means of suture anchors. Active flexion (*e*) and extension (*f*) of the knee joint are shown. g, Lateral view of the knee in 45 degrees of flexion. Note how well the tibia and patella track.

Figure 6. *a* and *b*, Anterolateral dislocation of the knee. The tibia is internally rotated on the femur. *c* and *d*, After the knee reconstruction procedure, the knee is reduced and stable. The fascia lata was looped around the lateral collateral ligament to externally rotate the tibia.

Figure 7. *a*, Preoperative anteroposterior view radiograph of 8-year-old girl with congenital short femur with an LLD of 15.5 cm. Predicted LLD at skeletal maturity is 20.8 cm. *b*, Because of the

dysplastic acetabulum, a Millis-Hall modification of the Salter osteotomy was performed before lengthening. The procedure gained 2.5 cm of length for the patient. At age 9 years, 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 the knee joint was noted to be stable preoperatively. Note the well-developed tibial spines, which imply the presence of cruciate ligaments. *c*, Despite the apparently stable knee, operative release of the fascia lata and hamstrings, daily physical therapy, and dynamic knee extension splinting, the patient's knee joint began to posterolaterally subluxate after 4 cm of lengthening. The lengthening was stopped and the knee reduced [Dror: Is it the knee that reduced?] with intensive physical therapy. This case illustrates the need to always cross the knee with the external fixation, even when it seems clinically stable. *d*, Final radiograph obtained after the first lengthening. Note the overgrown greater trochanter and the coxa vara. *e*, At age 11, the patient had a nail inserted with simultaneous transfer of the greater trochanter. The trochanter was fixed with the proximal locking screw of the nail. This nail was inserted in preparation for the subsequent lengthening over nail procedure that was performed 6 months later. If the trochanter did not have to be transferred, the nail would have been inserted at the time of the lengthening surgery. *f*, Preoperative radiograph obtained after healing of the transferred trochanter. *g*, At age 12 years, the patient underwent a simultaneous lengthening of her femur and tibia. The femur was lengthened over the nail while the tibia was lengthened with the Ilizarov apparatus only. *h*, The femur was lengthened 8 cm and the tibia 5 cm. Radiograph of the femur at the end of the lengthening over the nail. The regenerate new bone is healing well. *i*, The femoral and tibial frames were linked together by a monolateral hinge. A 45-degree flexion contracture developed while a flexion range to 95 degrees was maintained. The flexion contracture was treated by distraction of the femoral and tibial frames from each other. Note the distraction gap in the tibia and the extension of fixation to include the foot for prevention of equinus contracture. *j*, Final result after two lengthenings shows near equalization of limb length (LLD = 1.9 cm) at bone and chronological age 14 years. Total length gain = 2.5 cm (pelvis) + 4 cm (femur) + 8 cm (femur) + 5 cm (tibia) = 19.5 cm. The remaining LLD will be made up by valgus osteotomy of the coxa vara, which will acutely lengthen the leg by approximately 2 cm. *k* and *l*, Photograph of knee extension and flexion range of motion seen on the front and side views 3 months after removal of the apparatus.

Figure 8. *a*, 2-year-old girl with type 1b congenital short femur with delayed ossification of stiff proximal pseudarthrosis and of femoral neck, coxa vara, and fixed flexion deformity of the hip. Her predicted LLD at skeletal maturity is 30 cm. b, Radiographs obtained when the patient was 6 years old, after proximal femoral deformity correction and 5 cm of lengthening through the pseudarthrosis and pelvic osteotomy, which were performed before age 4 years. The pseudarthrosis is well healed, and the femur is now a type 1a. The patient will require at least two more femoral lengthenings.

Figure 9. *a*, 14-year-old skeletally mature girl with type 2b congenital short femur and a 19-cm LLD. She was previously treated by contralateral epiphysiodesis of the distal femur. *b*, Anteroposterior view radiograph of the left hip shows the pseudarthrosis and the overgrown greater trochanter. *c*, Lengthening of the femur and tibia with fixation to the pelvis. The femoral neck was opened and bone grafted and fixed internally with a screw. The fixation to the pelvis neutralizes the forces on the nonunion. *d*, Radiograph obtained after lengthening of the femur (8

cm) and tibia (6 cm). The remaining LLD is 5 cm and can be equalized either by a contralateral closed femoral shortening or by a second lengthening. The patient had 90 degrees of knee flexion before and after the lengthening. *e*, Final radiograph of the hip shows union of the femoral neck nonunion.

Figure 10. *a*, 12-year-old girl with type 2b CFD. There is no acetabulum or femoral head and the proximal femur is proximally dislocated. *b*, There is a 16-cm LLD. *c*, 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 of femoral lengthening. Note that the fixation is extended with hinges across the knee to the tibia. *d*, Final standing anteroposterior view radiograph obtained shortly after removal of the fixator. *e*, Lateral view radiograph obtained after removal of the fixator shows the extension component of the osteotomy.

References

1. Sanpera I Jr, Sparks LT: Proximal femoral focal deficiency: Does a radiologic classification exist? *J Pediatr Orthop* 14:34-38, 1994.
2. Suzuki S, Kasahara Y, Seto Y, Futami T, Furukawa K, Nishino Y: Dislocation and subluxation during femoral lengthening. *J Pediatr Orthop* 14:343-346, 1994.
3. Millis MB, Hall JE: Transiliac lengthening of the lower extremity: A modified innominate osteotomy for the treatment of postural imbalance. *J Bone Joint Surg Am* 61:1182-1194, 1979.
4. Langenskiöld A, Ritsila V: Congenital dislocation of the patella and its operative treatment. *J Pediatr Orthop* 12:315-323, 1992.
5. Grammont PM, Latune D, Lammaire IP: Treatment of subluxation and dislocation of the patella in the child. Elmslie technic with movable soft tissue pedicle (8 year review) [in German]. *Orthopade* 14:229-238, 1985.
6. Sisk DT: Knee Injuries. In, Crenshaw AH (ed): *Campbell's Operative Orthopedics*. St. Louis, CV Mosby, 1987, pp 2399-2400.
7. Gonzalez-Herranz P, Burgos-Flores J, Rapariz JM, Lopez-Mondejar JA, Ocete JG, Amaya S: Intramedullary nailing of the femur in children: Effects on its proximal end. *J Bone Joint Surg Br* 77:262-266, 1995.
8. Astion DJ, Wilber JH, Scoles PV: Avascular necrosis of the capital femoral epiphysis after intramedullary nailing for a fracture of the femoral shaft: A case report. *J Bone Joint Surg Am* 77:1092-1094,1995.
9. Pouliquen J, Gorodischer S, Verrevet, Richard L: Femur lengthening in children and adolescents: Comparative study of a series of 82 cases [in French]. *Rev Chir Orthop Reparatrice Appar Mot* 75:239-251, 1989.
10. Hollister AM, Jatana S, Singh AK, Sullivan WW, Lupichuk AG: The axes of rotation of the knee. *Clin Orthop* 290:259-268, 1993.
11. Paley D: Problems, obstacles, and complications of limb lengthening by the Ilizarov technique. *Clin Orthop* 250:81-104, 1990.
12. Paley D, Tetsworth K: Mechanical axis deviation of the lower limbs: Preoperative planning of uniapical angular deformities of the tibia or femur. *Clin Orthop* 280:48-64,1992.