Lengthening Reconstruction Surgery for Congenital Femoral Deficiency

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Contents

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13.1 Introduction

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

13.1.1 Classification

Existing classifications of congenital short femur and proximal femoral focal deficiency are descriptive but are not helpful in prescribing treatment. A longitudinal follow-up of different classification systems (1) showed that they were inaccurate in predicting the final femoral morphology based on the initial radiograph. 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. The author's (DP) classification system (Fig. 13.1) is based on the factors that influence lengthening reconstruction of the congenital short femur (2).

13.2 Evaluating the Child with Unilateral CFD

13.2.1 History

Most children born with unilateral CFD have no family history of this or other congenital anomalies. Nevertheless, inquiry should be made into family history, exposure to drugs, medications, radiation, or infectious diseases during the first trimester. Many cases are now identified with prenatal ultrasound early in the pregnancy by measuring the lengths of the two femurs.

13.2.2 Physical Exam

There is an obvious leg length discrepancy. Associated fibular hemimelia and ray deficiency may be present. The hip and knee should be examined for flexion contracture greater than the other side. Neonates and young infants normally have such contractures for the first 3–6 months. The range of motion of the hip, knee, and ankle should be recorded.

Characteristic physical examination findings include the following:

Hip: external rotation (ER) deformity or increased ER vs. internal rotation (IR), fixed flexion deformity (FFD) of hip, and limitation of abduction (when coxa vara is present)

Knee: FFD of knee, no limitation of knee flexion, hypoplastic patella, lateral tracking or subluxed or dislocated patella, anteroposterior instability of knee, rotary instability of knee, anterior dislocation of tibia on femur with knee extension followed by reduction of knee with attempted flexion, hypermobile meniscal clunks, and temporary locking of the knee during flexion

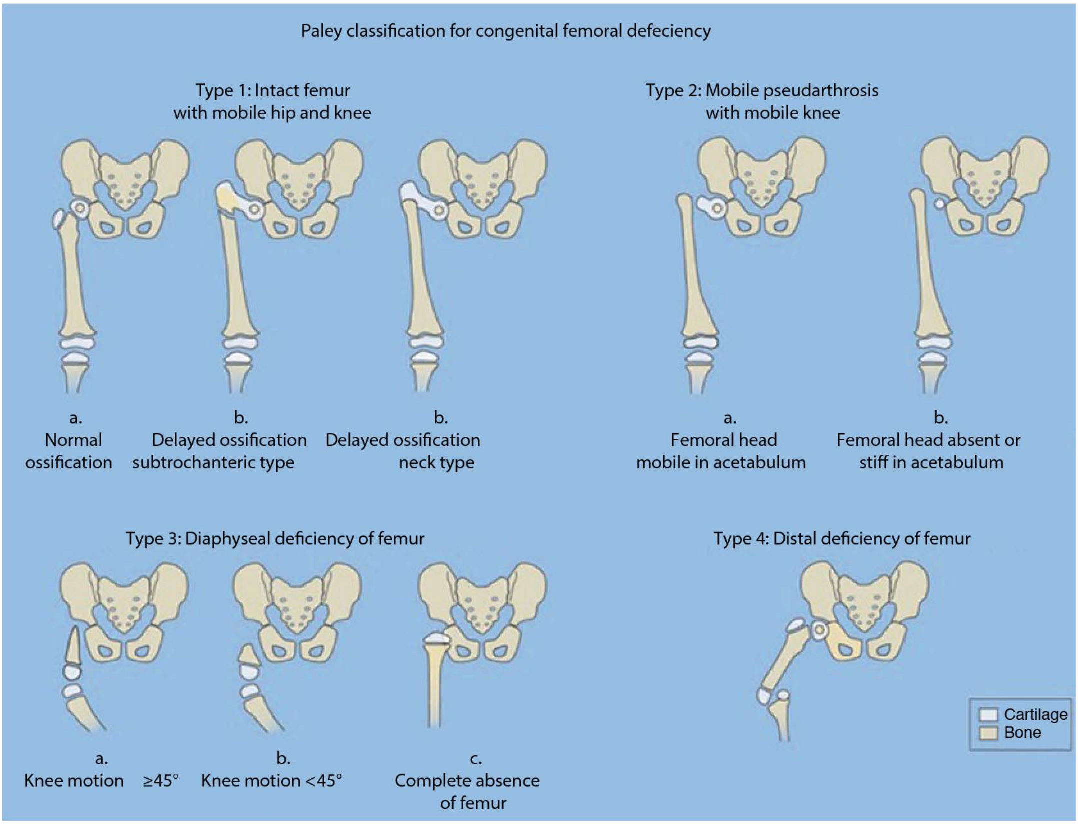

Fig. 13.1 Congenital femur deficiency classification (Conceived by Paley 1998)

Ankle: limitations of ankle dorsiflexion, obligatory eversion with dorsiflexion, hypermobility of ankle to eversion, and lateral malleolus high compared to medial

13.2.3 Radiographic Examination

13.2.3.1 Before Standing Age

Long anteroposterior (AP) pull-down X-ray; this is an AP radiograph of both femurs and tibias with the legs pulled straight and the patellas forward. It allows measurement of length of both femurs and tibia. It does not include foot height.
[AU3] Long lateral radiograph of both lower limbs pull-down with X-ray tube distance the same for both sides; this includes the femur and tibia of each lower limb in maximum extension on the same film. When there is a knee flexion deformity, it also allows accurate measurement of length of femur and tibia on both sides. AP pelvis supine; this allows more accurate measurement of the center-edge (CE) angle of both hips to assess for hip dysplasia. It is also a better-quality X-ray to assess for the ossification of the femoral neck. It is important that the pelvis be level for more accurate measurement.

13.2.3.2 Magnetic Resonance Imaging (MRI)

MRI is useful for assessment of integrity of the proximal femur. It can help determine whether the femoral head is joined to the shaft of the femur via a cartilaginous femoral neck. It can also help determine whether the cartilage of the femoral head is fused to the cartilage of the acetabulum in cases of femoral neck pseudarthrosis. Finally, it helps outline the deformity of the proximal femur. For optimal imaging, the cuts of the proximal femur should be reformatted in an oblique plane to see the entire proximal femur on one cut. MRI can also help outline the intra-articular pathology of the knee identifying deficiency of the cruciate ligament(s) as well outlining the shape of the joint surfaces in frontal and sagittal planes.

13.2.3.3 Computerized Tomography (CT)

CT is only useful at an older age when the acetabulum and proximal femur are nearly fully ossified. Three-dimensional CT reconstruction is useful to compare the normal acetabulum with the dysplastic side. In older children 3D CT can show the pathologic anatomy.

13.3 Surgical Reconstructive Strategy

13.3.1 Step 1: Preparatory Surgery of the Hip and Knee

Prior to lengthening one must determine whether the hip or knee are stable and/or deformed and whether surgical procedures for these joints are [AU4] required before initiating lengthening. At the hip, if the acetabulum has an acetabular index that has a comparable slope to the opposite normal side, a CE angle is ≥20°, and the neck shaft angle (NSA) is ≥110°, no separate hip surgery is required before the first lengthening. If the acetabulum shows signs of dysplasia, then a pelvic osteotomy should be performed prior to lengthening. Objective evidence of dysplasia is defined by a CE angle of less than 20° or a CE angle ≥20° (Suzuki et al. 1994). Increased slope of the "sourcil" (acetabular roof) compared to the other side is often a subtle but very sensitive sign. Similarly increased acetabular index in young children is equivalent to [AU5] increased sourcil slope in older children or adults. Coxa vara should be corrected prior to lengthening if the NSA is less than 120°. Similarly external rotation deformity of the hip is a factor to consider for correction at the same time as the acetabular dysplasia. If a Dega type of osteotomy is chosen, then there is usually a gain of about 1 cm in leg length. Associated hip deformities of retroversion, hip flexion contracture, and hip abduction contracture should be simultaneously addressed. The flexion contracture of the hip is treated by recession of the psoas tendon and release of the rectus femoris tendon. Flexion deformity may also be bony in which case it is treated by extension osteotomy. The abduction contracture is treated by lengthening or resection of the fascia lata and if necessary an abductor muscle slide. When all of these deformities are present together and especially with higher degrees of angulation, the reconstructive procedure is called the "superhip" procedure. Many of these more severe cases also have delayed ossification of the femoral neck or subtrochanteric region. Before lengthening the proximal femur should be as ossified as normal for that age. It is not enough to restore the biomechanics to normal by correcting the femoral and acetabular deformities. If there is a delayed ossification of the femoral neck, BMP should be added to the femoral neck to get it to ossify. It is beneficial to femur lengthening to remove the fascia lata. The fascia lata is a thin but very tough limiting membrane which resists lengthening and applies pressure across the knee joint and the distal femoral growth plate. For this reason, it should always be removed or at least cut before lengthening. Rather than throw away the fascia lata, I prefer to use it to reconstruct the absent cruciate ligaments. Although this is not an essential preparatory procedure, I prefer to reconstruct the knee ligaments at the same time as the hip surgery when indicated. Together with the knee ligament surgery a hemi-physiodesis can be added using a plate. The ideal age for the preparatory procedure is between age 2 and 3 years.

13.3.2 Step 2: Serial Lengthenings of the Femur

To determine the number of lengthening surgeries required, a prediction of leg length discrepancy at maturity is carried out. This can be done using the Paley multiplier method (Paley et al. 2000). The first lengthening of the femur can proceed 12 months after the preparatory surgery assuming the femoral neck has ossified. If the preparatory surgery is performed between 24 and 36 months (age 2–3 years), the first lengthening can follow between ages 3 and 4 years, respectively. The exception to this is if the femur is excessively short for an external fixator and it would be beneficial to wait a year or two to allow it to grow or if the femoral neck fails to ossify in the Paley type 1b case. The lengthening goal depends on the total discrepancy at maturity. Since the total discrepancy is large in most cases, we try and achieve as much length as possible safely. The safe range is 5–8 cm if a good physical therapy program is available. In most cases, we achieve 8 cm as long as the patient is able to maintain adequate knee [AU6] range of motion. As a rule of thumb to make it easy for the parents to remember the age for lengthening, we follow the rule of 4, one lengthening every 4 years (e.g., ages 4, 8, 12). The age for the second lengthening is around 8 years and the final lengthening around 12 years. If each time we can get up to 8 cm, after three lengthenings, the gain is up to 24 cm, and together with the one cm gain from the hip surgery, the total is 25 cm. If more equalization is needed, a physiodesis of the long leg distal femoral growth plate is carried out for an additional 5 cm. In this manner, we can equalize 30 cm of discrepancy with three lengthening surgeries and one physiodesis. If more is required one more lengthening of up to 10 cm can be done, increasing the total to 40 cm. Following the rule of 4, this would be done at age 16 years or older. In some cases, the tibia is also short and contributes to the LLD. During one or more lengthenings of the femur, the tibia would also be lengthened. This will be discussed in more detail later. When the LLD is from both the femur and the tibia, lengthening both at the same time can reduce the total time of external fixation while achieving even more length than is possible with lengthening of only the femur.

13.3.3 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. The acetabulum should be assessed at the age of two by means of a supine AP pelvis radiograph. A CE angle <20° is an indication for pelvic osteotomy. A sourcil angle that is not horizontal ±5° or which is asymmetric from the opposite side is also an indication for pelvic osteotomy even in cases where the CE angle is at 20°. The acetabular dysplasia associated with CFD is not like that associated with developmental dysplasia of the hip. The deficiency is more of a hypoplasia of the entire acetabulum. This is most manifest superolateral and posterior with a hypoplastic posterior lip of the acetabulum. In young children (2–5), the Dega osteotomy is my preferred method to improve coverage (Grudziak and Ward 2001). Although the Dega gives excellent superior and lateral coverage, it is misleading to think that it improves the posterior coverage. Since the posterior lip is an ischial structure located distal to the triradiate, the Dega cannot increase its coverage but rather does not reduce its coverage like a Salter osteotomy does. In older children (6–12 years old) with an open triradiate cartilage, I prefer a periacetabular triple osteotomy, and in adolescents or adults (>13 years old with closed or closing triradiate cartilage), I prefer the Ganz periacetabular osteotomy. In both of these, there is the ability to improve the posterior coverage by internally rotating the periacetabular fragment prior to abducting this fragment for increased lateral coverage. In my early experience I used the Salter or the Millis-Hall modification of the Salter (combining innominate bone lengthening with the Salter) (Salter 1978; Millis and Hall 1979). The problems with this are twofold: (1) the hip becomes more uncovered posteriorly and (2) femoroacetabular impingement tends to develop as they mature since the femoral neck, which is often short and has a poor anterior recess, impinges with the more prominent anterior lip of the acetabulum (which is now retroverted). The Salter osteotomy and its modifications are to be avoided. Similarly, when performing the Dega osteotomy, it is also important to make sure the cut extends posteriorly past the apex of the sciatic notch to end as a T junction with the triradiate as it separates the ischium from the ilium laterally. This will ensure that the Dega osteotomy hinges on the triradiate or medial bone of the ilium rather than rotate the quarter pelvis the way a Salter osteotomy does.

13.3.4 Proximal Femoral Deformities (Fig. 13.2a, b)

There is a wide spectrum of deformity of the proximal femur seen in CFD. This is nicely illustrated in the Pappas classification (Pappas 1983) which is

Fig. 13.2 3D CT reconstruction AP (**a**) and lateral views (**b**) showing typical type 1b deformity. Corresponding radiographs AP (**c**) and lateral (**d**)

very descriptive but not very useful for directing reconstruction. The mildest cases may have coxa valga (Pappas type 9). The majority of cases however have varying degrees of coxa vara. This coxa vara ranges from mild uniplanar deformity to severe triplanar deformity. Until recently, the pathoanatomy of this deformity was not clear. We now understand that it is a complex combination of bony deformities in the frontal, sagittal, and axial planes, combined with soft tissue contractures affecting all three planes. The severity of these deformities is often but not always milder in type 1a cases and more severe in type 1b cases. Since the natural history of delayed ossification of the proximal femur is to ossify, very severe proximal femoral deformities may be seen in type 1a cases which at one point would have been classified as type 1b (Sanpera and Sparks 1994). Therefore, the difference between type 1b and type 1a may be a matter of the timing of the classification. Had the same 1a case been seen young enough, it might have shown a delay in ossification and been called a 1b and vice versa. In other words, the natural history of all 1b cases is to ossify and become 1a.

The proximal femoral deformity of CFD can occur between the level of the femoral neck to and including the subtrochanteric region of the femur. It is a result of a combination of extra-articular hip joint contractures combined with a bony deformity between the proximal diaphyseal part of the femur and the intertrochanteric or subtrochanteric region of the upper femur segment. Due to combination of frontal plane, sagittal plate, and axial plane angular deformities, the deformity appears differently when observed from a proximal vs. a distal reference perspective. This phenomenon is called "parallactic homologues" (Paley 2005a).

To avoid confusion in the description of the deformity, we need to establish a point of anatomic reference. Since the only deformity of the pelvis is hip dysplasia, I will describe the deformity of each of the two segments of the femur relative to the pelvis with the pelvis in an anatomic position. I will also use the convention of describing the distal relative to the proximal segment with the proximal segment being the anatomic pelvis laying flat on a table.

Proximal femoral deformity relative to the pelvis: The proximal femur consists of the femoral head, neck, and greater and lesser trochanter, including a smaller or larger segment of the femoral diaphysis (neck type vs. subtrochanteric type, respectively). The proximal femur relative to the pelvis is in flexion and internal rotation associated with extra-articular contractures of the hip abductors, hip flexors, and piriformis tendons. In the most severe cases, it is common for the hip flexion deformity to be as much as 90° and the internal rotation deformity to be 45°. Purely flexing the femur 90° places the neck of the femur horizontal to the pelvis in the frontal projection but at a 45° angle to the sagittal projection of the neck of a femur with a 135° neck shaft angle. The neck would be oriented anteriorly from the head to the trochanter. The flexion therefore makes the neck appear to be retroverted 45° (apparent retroversion deformity) when viewed from distal to proximal. Internally rotating the femur around its mechanical axis, while the hip is in a 90° flexed position, moves the neck from being horizontal to the pelvis (parallel to the line connecting the two ischial tuberosities), to appear 45° abducted to the horizontal of the pelvis (apparent abduction deformity). The neck moves proximally closer to the iliac wing. The greater trochanter faces posterior, medial, and distal towards the sacrum. Since the tip of the greater trochanter is pointing towards the sacrum, the piriformis muscle which originates at the sacrum will be very short and appear contracted. Similarly for the hip abductor muscles, the [AU7] greater trochanter is so medial and proximal that the hip abductor muscles have never been stretched out to length. Finally the psoas and rectus femoris tendons will also be tight for the same reasons.

13.3.5 Distal Femoral Deformity

The distal femur relative to the pelvis is mostly externally rotated. It may also have some flexion and adduction relative to the pelvis depending on the amount of relative deformities of the proximal and distal segment to each other.

Deformity of the proximal to the distal femur: If we connect the distal femur to the proximal femur, we have a CFD hip deformity. The relationship of the two segments relative to each other is the confusing part. As a reminder, the deformities of the proximal femur to the pelvis include actual flexion and internal rotation deformities, combined with the apparent abduction and retroversion. The deformities of the distal femur to the pelvis include actual neutral to adducted position, neutral to flexed position, and external rotation. In relation to each other, the distal relative to the proximal femur deformities appears very different. The distal femur appears adducted to the proximal segment (coxa vara). I call this apparent since there is no actual abduction of the proximal femur. The proximal femur deformity that appears to be abduction is actually internal rotation of the proximal segment of the femur. There will be a net adduction deformity relative to the pelvis since the adduction of the distal femur relative to the proximal femur is greater than the apparent abduction of the proximal femur relative to the pelvis. Relative to the proximal femur, the distal femur appears to be in extension. I call this apparent extension because the distal femur rotation makes a true flexion deformity of the distal femur appear to be extension. This will become more understandable as we proceed to unravel this complex deformity. Since the apparent extension deformity of the distal femur is smaller than the flexion deformity of the proximal femur, the net is a smaller fixed flexion deformity relative to the pelvis. Relative to the proximal femur, the distal femur appears to be externally rotated. The reason I call this apparent external rotation is because the proximal femur is in apparent retroversion due to its flexion deformity. Because the retroversion is apparent, the relationship between the two is apparent. The smaller apparent retroversion of the proximal segment to the pelvis combined with a larger external rotation deformity of the distal femur leaves a net external rotation deformity relative to the pelvis.

If one could remove all of the soft tissue tethers and place the proximal femur in a normal anatomic position relative to the pelvis but keep the distal femur connected to the proximal femur in its deformed position, what position would the distal femur be in? We start by internally rotating the proximal femur. Because the hip is at 90° of flexion, the internal rotation appears to be abduction. Using the distal femur as a handle, we have to adduct the distal femur until the neck is horizontal. Next we need to extend the proximal femur around the horizontal axis of the pelvis. To rotate around the horizontal axis of the pelvis using the distal femur as a handle, one must consider the apparent adduction deformity of the distal femur (which in extreme cases can make the diaphysis appear parallel to the femoral neck). The maneuver for reduction of the proximal femur to an anatomic position requires the distal femur to rotate externally and flex. Once the femoral neck is reduced to a normal position, the final position of the distal femur relative to the proximal femur and to the pelvis is adduction, flexion, and external rotation. The adduction and external rotation were anticipated, but the flexion was not. This is due to the principle of parallactic homologues mentioned earlier. The rotation deformity makes the angular deformities appear different when the deformity is moved around and viewed from a different perspective.

Chicken and egg question: which came first: the contractures of the proximal femur or the deformity of the bone of the distal femur relative to the proximal femur? We will never know the answer to this question. The net effect in the frontal plane is that the greater trochanter with its insertion of the abductor muscles (gluteus medius and minimus) is abnormally close to the pelvis. This leads to several problems including impingement of the trochanter with the iliac bone and contracture of the gluteal muscles since the distance between their origin and insertion is short. The fascia lata with its iliotibial band extension to the tibia combined with part of the gluteus maximus is the most lateral of the soft tissue structures. They therefore contribute the greatest to the abduction contracture of the hip. Since adduction of the hip is preserved due to the varus of the femur and since the hip cannot be abducted much because of iliotrochanteric impingement, the abduction contracture is not obvious. It is rather stealth and hidden in plain view. If the bony coxa vara is corrected by osteotomy, without soft tissue releases, the abduction contracture will be uncovered. The abduction contracture will prevent the hip from coming back to a neutral position relative to the pelvis producing a fixed pelvic tilt. An abduction pelvic tilt on the short leg makes the limb length discrepancy (LLD) appear less than before surgery. In the face of an open growth plate or a nonossified neck or subtrochanteric segment, as in Type 1b cases, the abduction contracture leads to recurrence of the coxa vara after osteotomy. The mechanism for this recurrence may be differential growth of the physis, bending at the nonossified tissues, or slipped capital femoral epiphysis.

13.4 Superhip Procedure

Positioning, prepping, and draping for the superhip procedure: The patient should be on a radiolucent operating table, positioned supine close to the edge of the table, bumped up 45°, to roll the pelvis towards the opposite side. The entire side should be prepped and draped free from the nipple to the toes.

Step 1: *Incision* (Fig. 13.3a). A long midlateral incision is made from the iliac wing in a straight line to the tibial tuberosity in the anterior midline of the upper leg. The incision is carried down to the depth of the underlying fascia lata and iliotibial band.

Step 2: *Elevation of flap* (Fig. 13.3b). The subcutaneous tissues are dissected off the fascia of the thigh and pelvic region. The fat is adherent to the fascia and should be dissected preferably with a cautery. It is important not to incise the fascia with the cautery if the fascia is to be used for knee ligament reconstruction. Elevate a large flap of skin and subcutaneous tissues anteriorly allowing it to fold upon itself. Posteriorly a subcutaneous flap is elevated for only 1 or 2 cm. Extend the flap dissection medial to the Smith-Peterson interval (interval between the tensor fascia lata (TFL) and the sartorius) proximally. Distally reflect the flap to the patella if no ligament reconstruction is to be done and all the way to the medial side if ligament reconstruction is to be done. The fascia lata is now fully exposed from the patella to a couple of centimeters posterior to the intermuscular septum distally and from the medial side of the TFL to the mid-gluteus maximus proximally.

Step 3: *Fascia lata release* (Fig. 13.3c). The fascia is incised at the TFL-sartorius interval making sure to stay on the TFL side in order to avoid injury to the lateral femoral cutaneous nerve. The fascial incision is extended distally to the lateral border of the patella ending at the tibia. The posterior incision of the fascia lata starts distally and posterior at the intermuscular septum and extends proximally to overlie the gluteus maximus in line with the incision. The gluteus maximus (GMax) should be separated from the overlying fascia anterior to the posterior fascial incision. The fascia should be retracted anteriorly and away from the underlying muscle, while the GMax should be dissected off of the fascia and the intermuscular septum that separates it from the TFL. The GMax should not be split in line with the fascial incision to avoid denervating the muscle anterior to the split. It can now be reflected posteriorly to allow exposure of the greater trochanter, piriformis muscle, and sciatic nerve. When ligamentous reconstruction using the fascia lata is planned, the fascia lata is cut proximally at the muscle tendon junction anteriorly. The fascial cut should be sloped posteriorly and proximally to include a longer fascia segment posteriorly from the fascia that was dissected off of the GMax. The fascia lata is reflected distally to Gerdy tubercle. The TFL can be left in place without further dissection. It does not have to be separated from the underlying gluteus medius (GMed). The two muscles are often adherent to each other. In fact, it may be confusing at times which fibers are TFL and which are GMed. The distinguishing feature is that the GMed fibers insert on the greater trochanter while the TFL does not. The distal fascia lata also called the iliotibial band (ITB) blends with the underlying lateral knee capsule. It is common

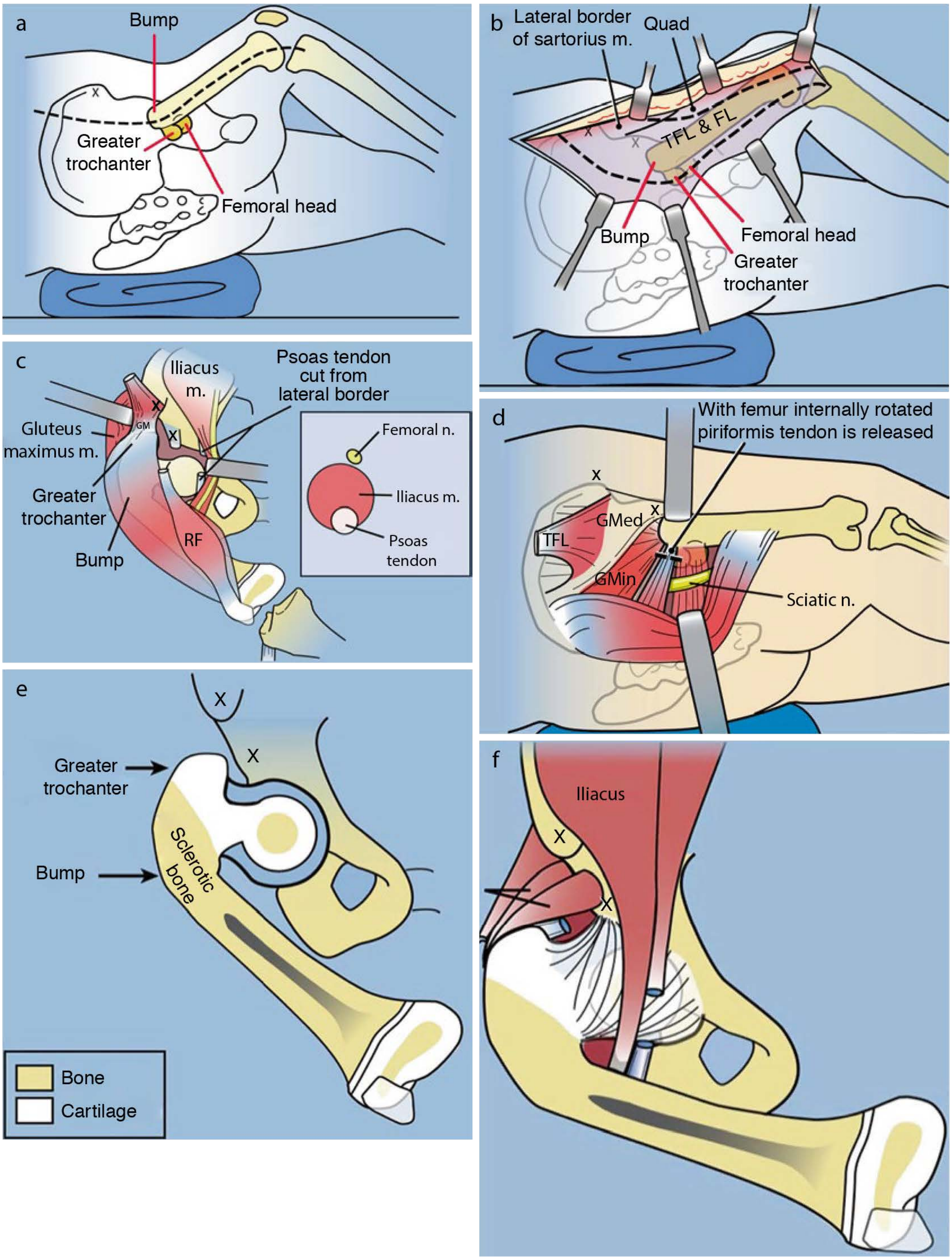

Fig. 13.3 Superhip surgical technique illustrations

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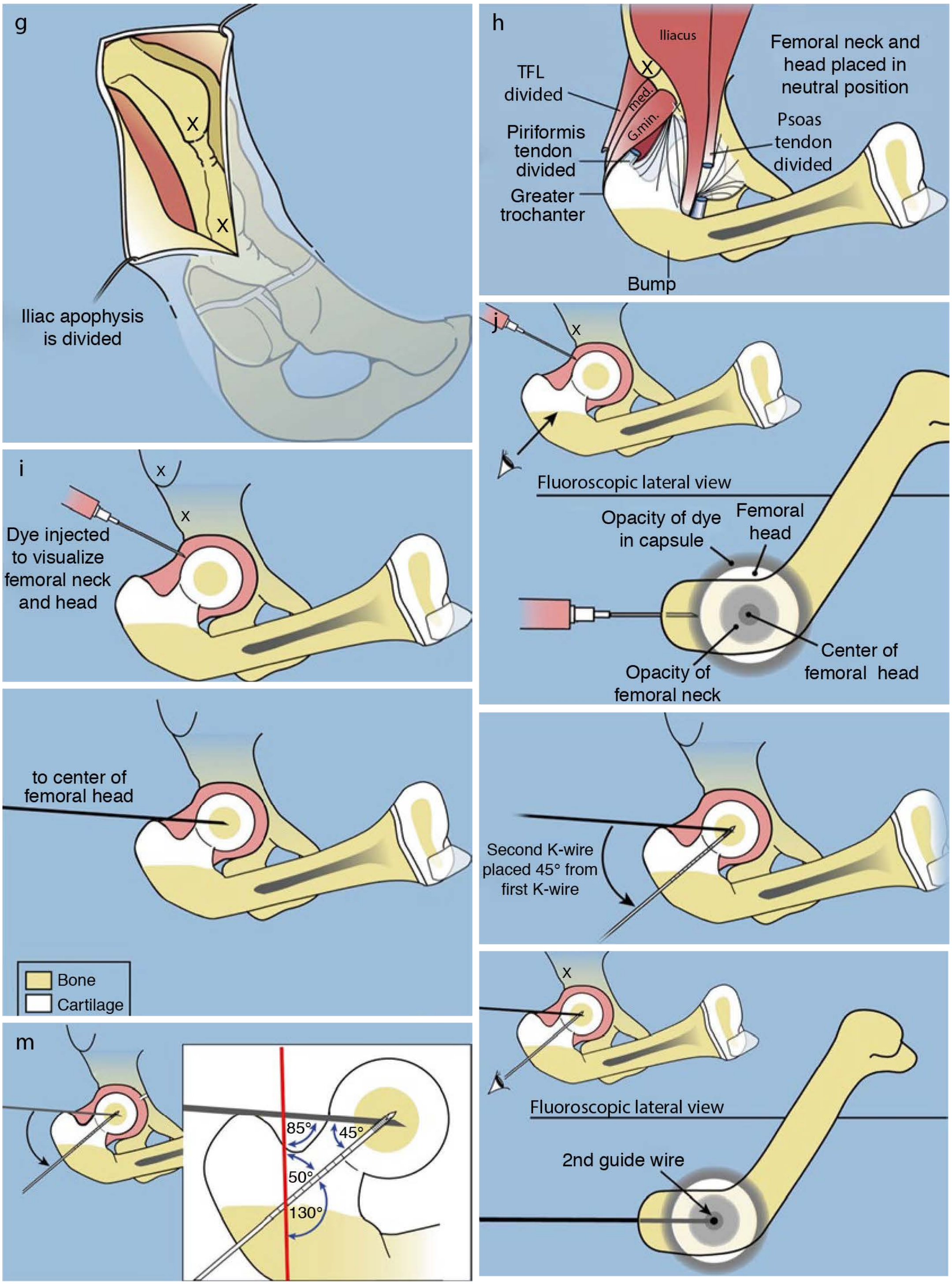

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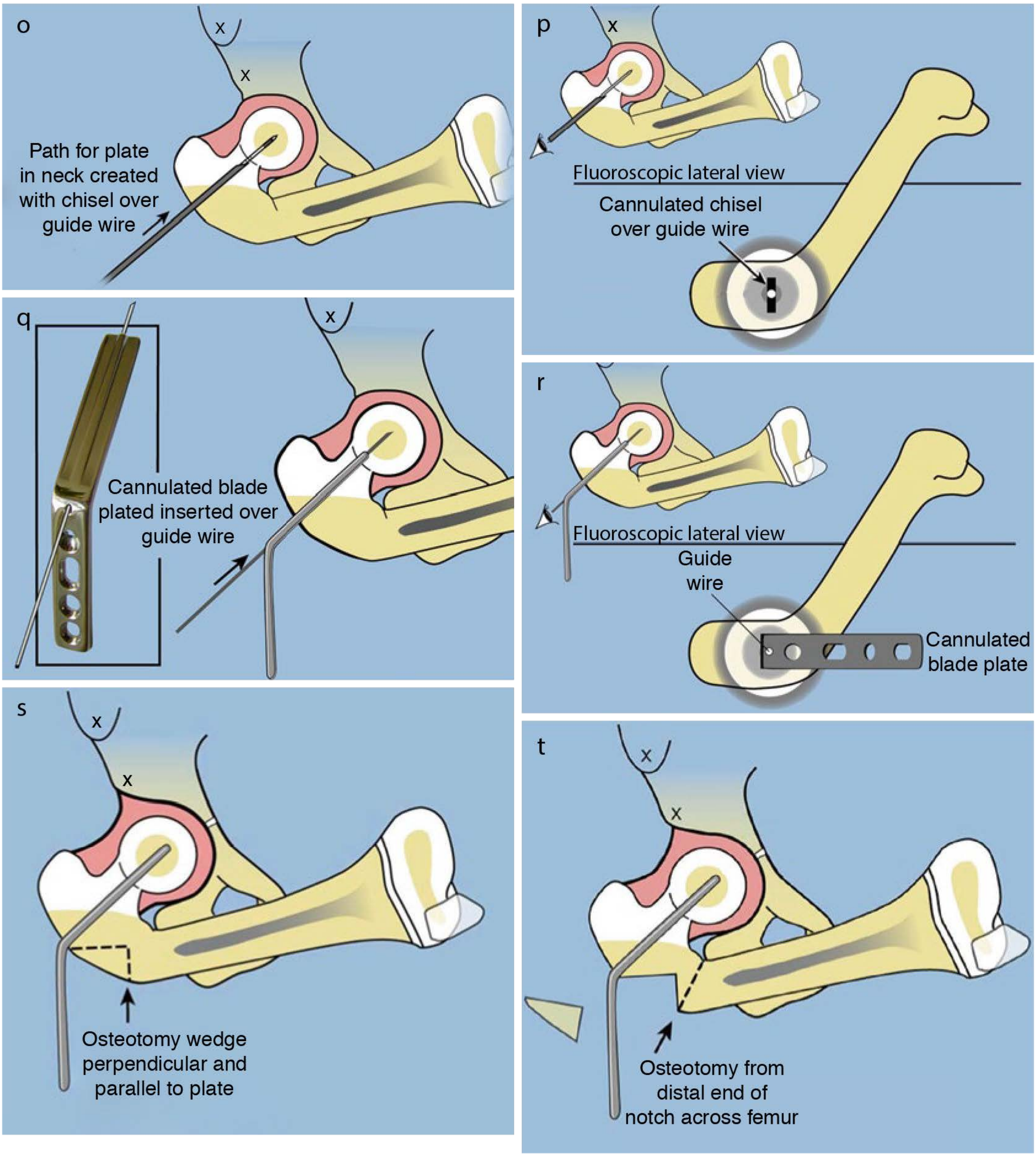

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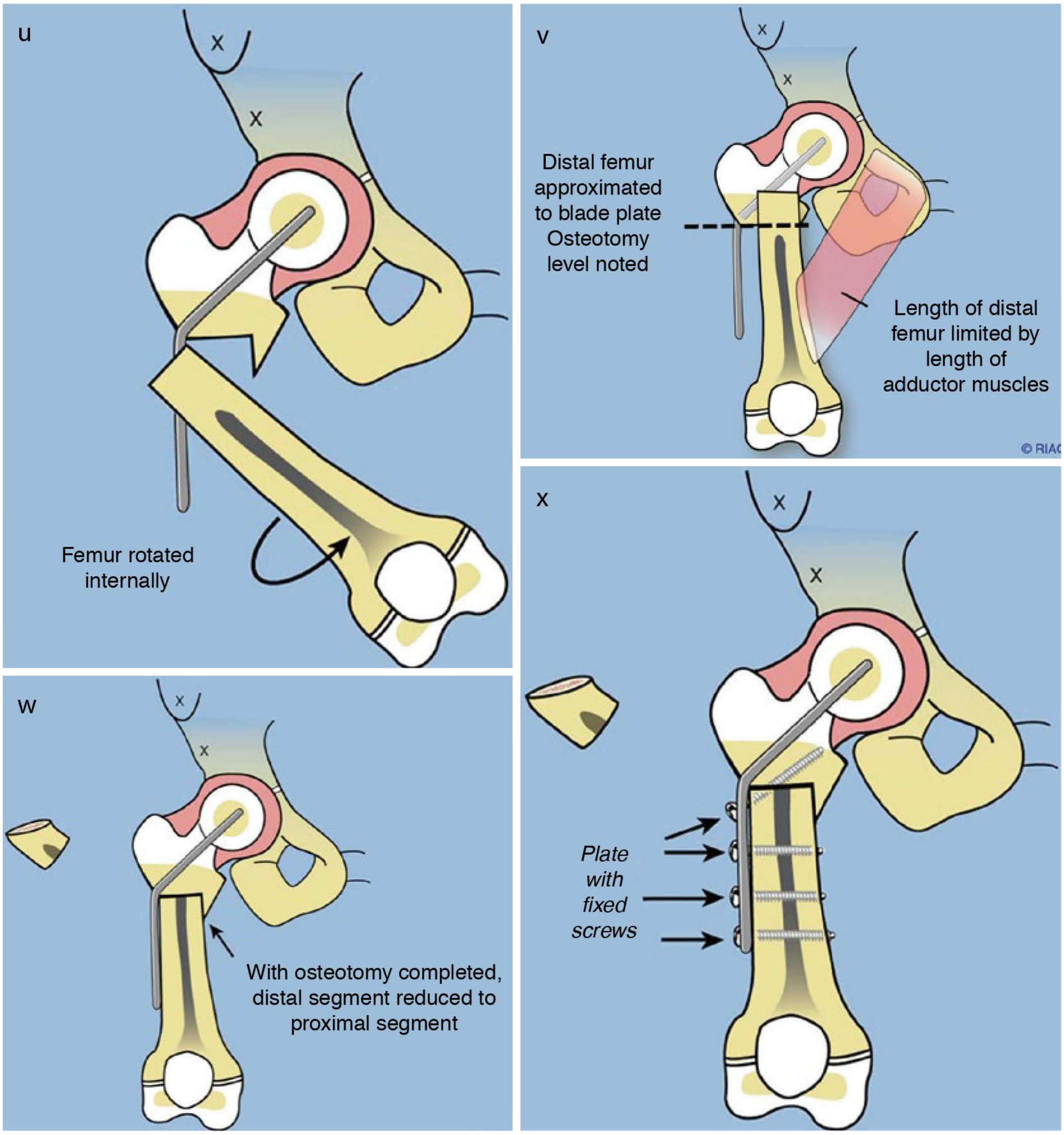

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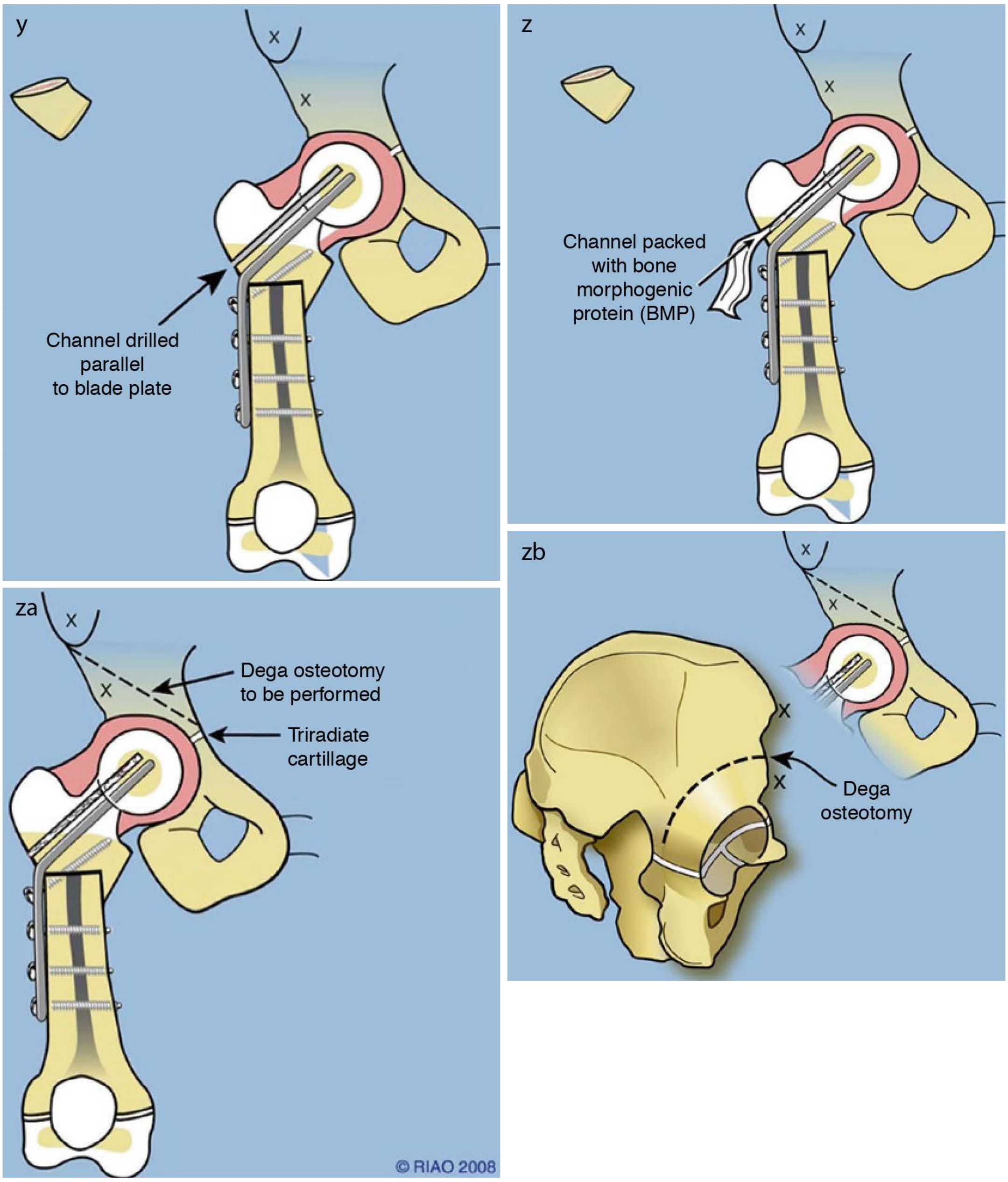

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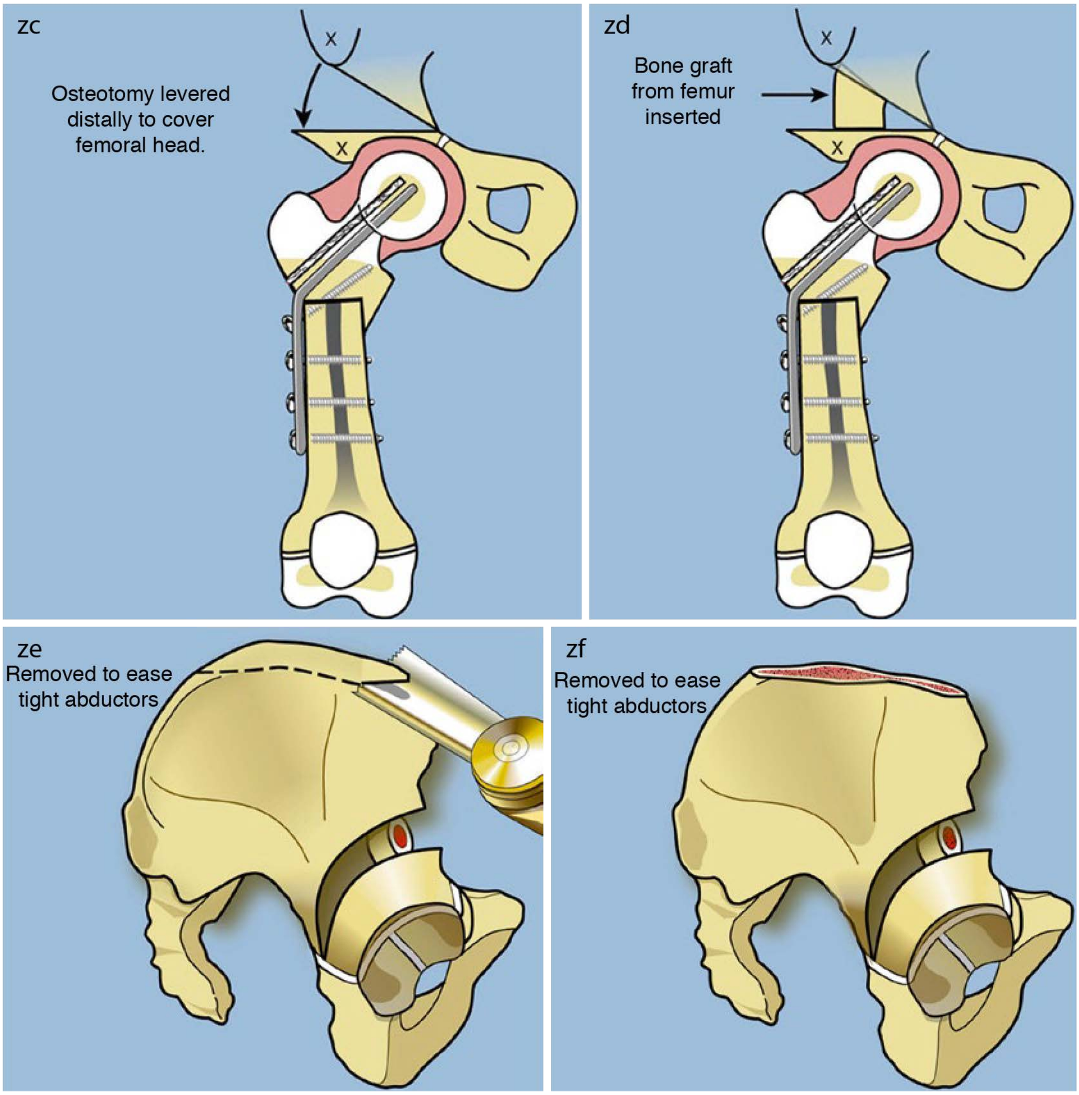

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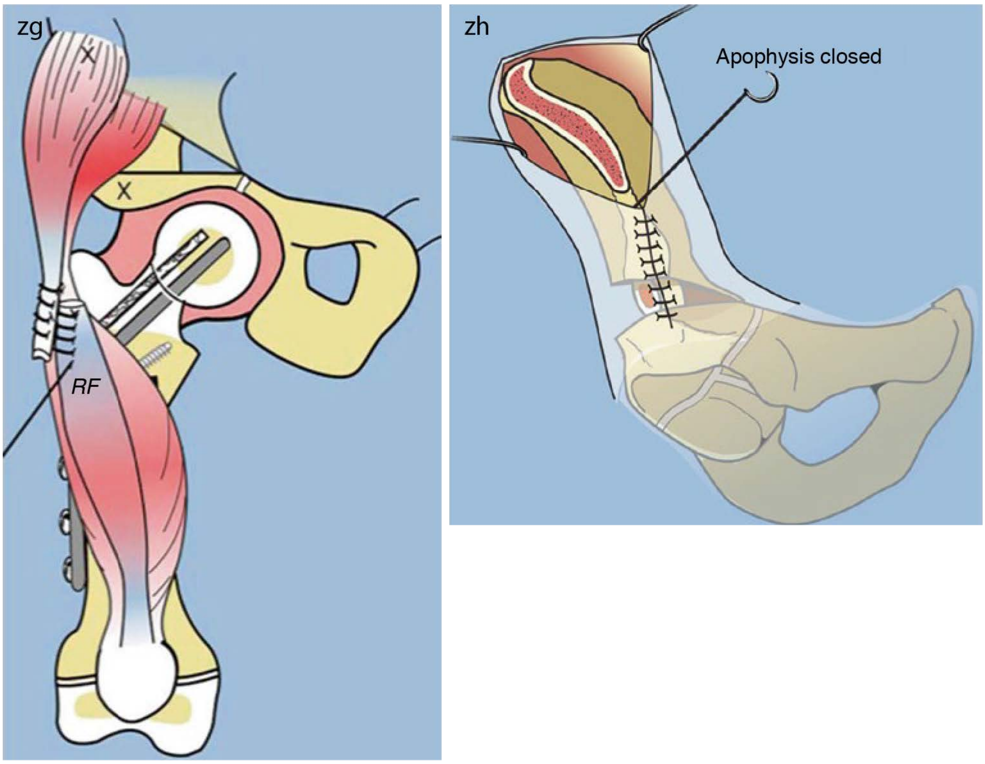


Fig. 13.3 (condinued)

to reflect some capsule with the ITB. The fascia should be mobilized all the way until Gerdy tubercle. The fascia can then be divided into two halves using a straight pair of scissors. It should be kept moist while the rest of the surgery proceeds. The two limbs of the fascia are ready for later use in the superknee procedure.

Step 4: *Hip flexion contracture releases*. The dissection is carried beneath the sartorius to find the rectus femoris tendon. The rectus femoris tendon insertion is identified at the anterior inferior iliac spine. The constant ascending branch of the lateral femoral circumflex artery and vein is cauterized prior to cutting the tendon. The conjoint rectus femoris tendon (distal to the split into reflected and direct heads) is cut and allowed to reflect distally. Care should be taken not to go too distal on the rectus femoris to avoid injury to its innervating branch of the femoral nerve. Just medial to the rectus is the iliopsoas muscle. The iliocapsularis muscle (capsular origin head of iliopsoas muscles) can also be seen here. The femoral nerve lies on the anteromedial surface of the iliopsoas muscle. Before looking for the psoas tendon, the femoral nerve should be identified and decompressed below the inguinal ligament. The posterior aspect of the iliopsoas muscle belly is now elevated from lateral to medial. The psoas tendon is located on the posteromedial surface in the substance of the muscle. The tendon is exposed and cut. Any remaining flexion contracture of the hip is due to the sartorius, gluteus medius and minimus (the part of these muscles originating anterior to the center of rotation of the femoral head in the sagittal plane), and the anterior fascia of thigh. If the anterior thigh fascia is tight, it can be released, taking care not to injure the neurovascular structures. Before releasing the anterior fascia, the lateral femoral cutaneous nerve should be identified and decompressed. It runs inside the fascia covering the sartorius muscle just medial to the anterior superior iliac spine. The next flexors to release are the gluteus medius and minimus muscles. This is accomplished by the abductor muscle slide technique (see step 6).

Step 5: *External rotation contracture release* (Fig. 13.3d). The piriformis tendon is contracted and prevents internal rotation of the hip. It should be released off of the greater trochanter. The greater trochanter should be identified by palpation. The gluteus medius muscle posterior border is very distinct and proceeds down to the greater trochanter where it inserts. Deep to the medius is the gluteus minimus and just distal to the minimus is the piriformis muscle. Its tendon can be palpated through its muscle. It may be difficult to identify the piriformis from the minimus. Care should be taken to avoid dissection at the distal border of the piriformis tendon. This is where the medial femoral circumflex branch anastomoses with the inferior gluteal artery branch. The entire piriformis is transected about one cm from its insertion onto the trochanter. The sciatic nerve can be identified and if necessary decompressed. It is more posterior to the trochanter and runs deep to the piriformis.

Step 6: *Abductor muscle slide*. The abductors may not appear to be tight on first inspection because of the coxa vara. Adduction of the hip into a true AP of the hip with the neck oriented normally in the acetabulum is restricted by the gluteus medius and minimus since the fascia lata has already been cut. Furthermore, the Dega osteotomy which lengthens the height of the ilium makes the abductors even tighter. There are two options to lengthen the abductor mechanism: lengthen it at the tendon end or slide its origin distally. At the tendon end, the conjoint tendon of the glutei and quadriceps can be released and then later reattached to the greater trochanter. I did this for 10 years and found that it lead to permanent weakness due to the change in muscle tendon length ratio. The second option is to detach the abductors from their origin and let them slide distally. This avoids changing the muscle tendon length ratio and avoids weakening the hip abductors. Since switching to the abductor slide, it has eliminated the problem of weakness, lurch, or Trendelenburg gait. To do this in the growing child, we split the iliac apophysis from the anterior inferior iliac spine to the anterior superior iliac spine and then posteriorly along the rest of the iliac crest. It is important to expose the apophysis along the entire length of the desired split. There is a tendency not to elevate the apophysis posteriorly enough. This starts with the reflection of the subcutaneous tissues in step 2 when the anterior and posterior flaps are elevated. The proximal and posterior extent of the dissection should be just beyond the highest point of the apophysis laterally. The anterior extent is just distal to the anterior inferior iliac spine which is exposed for the release of the rectus femoris tendon. The abdominal external oblique muscle is peeled off of the apophysis to expose it along its entire length. The external oblique muscle insertion overlaps the apophysis more laterally and posteriorly than anteriorly. Once the apophysis is bare, it can be split from anterior to posterior. This should be done with a number 15 blade. To know where to split, pinch the apophysis between thumb and index finger of the hand not holding the knife. Then push down on the knife blade until you feel bone. It is important to try and stay in the middle of the apophysis along its entire length. It is also important to push down hard with the knife blade until one feels bone. Using a periosteal elevator, "pop off" the apophysis from the ilium. This should be done at multiple sites to get the entire apophysis to peel back as a unit from the ilium. The apophysis and lateral periosteum are reflected distally, thus relaxing the abductor muscles. Since part of the abductors acts as flexors of the hip, the abductor slide helps eliminate any remaining flexion deformity of the hip. The medial half of the apophysis is reflected medially with the iliacus muscle. This effectively produces a flexor slide effect for additional treatment of flexion contracture of the hip.

Step 7: *Elevation of quadriceps*. The quadriceps are now elevated off of the femur in a subperiosteal fashion. Since the femur is so short, the exposure may extend as far as the distal femoral physis. Proximally, the vastus lateralis should be elevated off of part of the cartilage of the greater trochanteric apophysis by sharp dissection.

Step 8: *Arthrogram*. A hip arthrogram is now performed using a 20 gauge spinal needle. This will outline the femoral head, acetabulum, and femoral neck.

Step 9: *Guidewire insertion*. Since the abduction, flexion, and rotation contractures have all been released, the femoral head and neck can now be placed in a neutral orientation to the pelvis by extending and maximally adducting the lower limb over top the other side. A guidewire should now be drilled up the center of the femoral neck to guide the insertion of a fixed angle fixation device. Since the femoral neck is unossified and short, it is very difficult to drill a guidewire at the correct angle up the femoral neck. The goal is to create a 130° neck shaft angle and a medial proximal femoral angle (MPFA) of 85°. In the normal femur, the angle between the neck shaft line and the tip of the greater trochanter to center of femoral head line is 45°. The first guidewire is inserted from the tip of the greater trochanter to the center of femoral head. Since the tip of the trochanter cannot be seen radiographically in young children because it is cartilaginous, the tip of the trochanter is located by palpation using the wire tip. From this point the wire is then drilled towards the center of the femoral head as shown in the arthrogram. The image intensifier is placed into the lateral view and the leg rotated until a "bull's eye" is seen. This "bull's eye" is formed by the overlapping shadows of three circles. The outermost circle is the dye surrounding the femoral head. The middle circle is the dye surrounding the femoral neck. The innermost circle is the ossific nucleus of the femoral head. All three circles should be seen concentrically. A second wire should be drilled into the center of this "bull's eye" at a 45° angle to the first wire. Using another wire of the same length as the second wire, measure the amount of wire inside the femoral neck by placing it alongside the second wire and measuring the difference in length between the two wires. This will be the length of the blade of the blade plate to be used.

Step 10: *Insert cannulated chisel*. The cannulated chisel for the blade plate should now be hammered up the femoral neck guided by the second guidewire. The chisel should be rotated until it is perpendicular to the back of the edge of the posterior aspect of the greater trochanter. This will guide it to the correct angle in the sagittal plane.

Step 11: *Plate insertion*. Bang the chisel out of the femur and reinsert the guidewire. Insert the appropriate length 130° blade plate along this wire to the depth of the bend of the plate. Make sure on the image intensifier that the tip of the blade is not too deep into the femoral head. Check its position on AP and lateral as well as using the approach withdrawal technique with live fluoroscopy. If the plate is suspected of being too long, then replace it with one with a shorter blade. If the cannulation of the plate is off center, there is greater risk of protrusion into the joint.

Step 12: *First osteotomy*. The femur should be osteotomized with a saw perpendicular to the plate starting at the bend in the plate. The depth of this cut is incomplete and when the deformity angle is very large may be parallel to the lateral cortex. To guide this cut drill a wire perpendicular to the plate. Keep the plane of the saw blade perpendicular to the plate. The width of the perpendicular cut surface is as wide as the width of the femur diaphysis.

Step 13: *Second osteotomy*. A subtrochanteric osteotomy should be made oriented less than 90° to the first osteotomy to minimize the bone protruding medially.

Step 14: *Peel the femur off of the periosteum medially and cut the periosteum*. After the second osteotomy the distal femur can be peeled off of the surrounding periosteum. The periosteum medially is very thick and restricts correction of the varus and rotation deformity. Cut the periosteum by carefully separating it from the surrounding muscle. The profunda femoris and its perforators pass immediately under this periosteum and care should be taken to avoid injury to these vessels. Cutting the periosteum allows the thigh to stretch longitudinally reducing the amount of shortening required of the femur.

Step 15: *Shortening of the femur*. The distal femur is now mobile and can be valgusized and rotated internally. The distal femur is too long to fit end to end with the proximal femoral cut. The two ends should be overlapped. A mark should be made at the point of overlap. The distal femur should be osteotomized at this level. A wire is drilled perpendicular to the femur at the level of the osteotomy. A saw is used to cut the femur at this site. The segment of bone that is removed is stored on the back table in saline. It will be used as a bone graft for the Dega osteotomy.

Step 16: *Fixation of the distal femur*. The femur is now brought to the plate. The bone ends should oppose without tension. The femur is rotated internally to correct the external torsion deformity. To adjust the femur to the correct anteversion, the guidewire should be reinserted into the cannulation of the plate. This wire shows the orientation of the femoral neck. The knee should be flexed to 90° and the angle between the wire and the frontal plane of the femur as judged by the perpendicular plane to knee flexion is observed. This wire should appear at least 10° anteverted relative to the knee. The most distal hole in the plate can now be drilled with the femur held in this rotation. The drill hole should be made at the distal edge of the hole to compress the osteotomy. A depth gauge is used to measure the hole and a screw is inserted. Two more screw holes are drilled and screws inserted into the plate. The most proximal hole in the plate is designed to drill parallel to the blade of the plate. The wire in the plate cannulation is used to guide the drill bit. This screw helps secure the plate to the proximal femur. The other three screws secure the plate to the distal femur. In type 1b cases the blade of the plate goes across the proximal physis into the femoral head as does this oblique screw. In type 1a cases with a horizontally oriented growth plate, neither the blade nor the screw should cross the growth plate of the upper femur. In type 1a cases with a vertically oriented growth plate, the blade but not the screw should cross the physis.

Step 17: *Insertion of bone morphogenetic protein* (*BMP*). In type 1b neck cases, BMP should be inserted into the upper femur to stimulate ossification of the cartilaginous neck of the femur. A wire is drilled proximal and parallel to the guidewire in the cannulation of the plate. A 3.8 mm hole is then drilled overtop this guidewire. The drill hole should extend all the way into the ossific nucleus. BMP-2 (Infuse-Wright Medical) is then inserted in this hole. The BMP-2 is on collagen sponges, which can be pushed into the hole using the tip of a 3.2 mm drill bit in one hand and a forceps in the other hand.

Step 18: *Pelvic osteotomy*. The type of pelvic osteotomy depends on the age of the patient and the degree of dysplasia. In the majority of patients under age 6, I prefer to use the Dega osteotomy to treat the dysplastic acetabulum. In older patients and especially if there is a high grade of dysplasia, I use a periacetabular triple osteotomy (PATO) in children and the Ganz periacetabular osteotomy (PAO) when the triradiate cartilage is closed or nearing closure.

Step 19: *Iliac wing osteotomy and repair of the iliac apophysis*. After the pelvic osteotomy, the apophysis can be sutured back together. Due to the abductor muscle slide, the lateral apophysis cannot reach the top of the iliac crest. Part of the crest has to be resected to allow repair of the apophysis. The bone removed can be inserted into the Dega osteotomy or used to bone graft a PATO or PAO as well as the subtrochanteric femoral osteotomy.

Step 20: *Muscle repairs and transfers*. The TFL muscle should be sutured down to the greater trochanter to act as a hip abductor. The rectus femoris tendon should be sutured to the side of the TFL. The quadriceps should be sutured to the region of the linea aspera. The gluteus maximus should be advanced back to the posterior border of the TFL.

Step 21: *Closure*. If no knee releases or reconstruction are required, the wound can now be closed. The interval between the TFL and the sartorius should be sutured closed with care not to suture the lateral femoral cutaneous nerve. Since there is no fascia lata, the deepest layer is the fat layer. This layer is called the underlayer. It should be sutured with a number one Vicryl. A Hemovac drain should be inserted before closing this layer. If a superknee procedure is performed, a second more medial drain is also used. I prefer to bring the drains out proximally and anteriorly. The drains are usually secured with a clear adhesive sterile dressing (e.g., Tegaderm, 3M, Minnesota). It is important to close the wound in a fashion that the opposite layers get sutured at the same level. The next layer is Scarpa's fascia. It is closed with a 2-0 Vicryl running stitch. The deep dermal layer (subcutaneous layer) is closed with a 3-0 Vicryl and the skin is closed using a subcuticular stitch with 4-0 Monocryl. Sterile dressings are now applied.

Step 22: *Final radiographs*. After the drapes are removed, an AP pelvis to include the femur is obtained. A lateral of the femur relative to the knee joint is also obtained. These X-rays are reviewed before proceeding to the spica cast.

Step 23: *Spica cast*. All infants are placed in a spica cast. The position of the limbs in the cast is important. The operated upon limb should be placed in full hip and knee extension. The opposite limb can be in a flexed, abducted, and externally rotated position. The cast should include the entire affected side but with the foot left free. The opposite side should stop short of the knee joint. The cast should be bivalved before leaving the operating room. In most cases the cast can be converted to a removable spica cast after 5 days.

13.5 Knee Considerations

The knee in CFD may range from a normal stable undeformed knee to an unstable, contracted, deformed joint. The most common deformity of the knee is valgus. The valgus deformity of the knee is usually nonprogressive. The distal femoral physis is usually closer to the knee joint on the lateral side. This is often attributed to hypoplasia of the lateral femoral condyle. CFD cases often have a variable degree of anteroposterior and rotatory instability of the knee related to absent or hypoplastic cruciate ligaments. In some cases, the tibia dislocates anterior or posterior on the femur during extension or flexion, respectively. Furthermore, there may be rotatory instability present. One study has related the radiographic appearance of the tibial spines to the degree of hypoplasia of the anterior cruciate ligament (Manner et al. 2006). The patella is usually hypoplastic and may be maltracking laterally. In some cases it dislocates with flexion. Finally, many cases of CFD have a fixed flexion deformity of the knee.

13.5.1 Indications for Preparatory Surgery of the Knee Prior to Lengthening

Isolated anteroposterior instability is not necessarily an indication for surgery. Grade 3 instability (no endpoint on anterior and posterior drawer tests) will usually become symptomatic as the child gets older. If the child is going to undergo a superhip procedure or Dega osteotomy prior to lengthening, and since the fascia lata is going to be excised, it makes sense to rebuild the knee ligaments and not "waste" the fascia lata. In some children there is a "catch" or "locking" sensation in the knee when going from extension to flexion. This is due to contracture of the iliotibial band. This catching feeling may even be painful and may require a trick motion to release it. In more severe cases, the tibia actually subluxes or dislocates anteriorly on the femur and reduces at about 30° of flexion. Once again the culprit is the iliotibial band combined with an aplasia of the anterior cruciate ligament (ACL). In older patients, the posterior aspect of the tibia may be rounded contributing to anterior dislocation of the tibia on the femur. Whether this is a secondary change due to chronic dislocation or a primary deformity is not clear since the tibia is not ossified posteriorly in infancy.

Patellar hypoplasia and instability is very common. The patella frequently maltracks laterally with flexion. In some cases it even dislocates with flexion. This is due to a combination of factors: valgus distal femur, hypoplastic or absent patellar groove, contracture of the lateral retinaculum with the tight iliotibial band, and external rotatory instability of the tibia on the femur due to cruciate deficiency which lateralizes the patellar tendon insertion. Patellar maltracking or subluxation should be corrected prior to lengthening.

Flexion contracture of the knee is another congenital deformity that may be present and which should be corrected before proceeding with lengthening. When the femur is very short, the acute angle created by the posterior thigh muscles gives the appearance of a flexion contracture. The definition of a flexion contracture however is a flexed angle between the anterior cortical line of the femur and tibia in maximum extension. When the contracture is more than 15°, it should be corrected surgically. Knee flexion contracture can be due to bony or soft tissue causes. In CFD the most common is intra-articular capsular contracture. There may be some extra-articular contribution due to contracture of the hamstring muscles and gastrocnemius muscles. Release of these muscles alone rarely corrects the contracture, while capsular release without complete hamstring release corrects the contracture. In some cases, there is a true bony flexion of the distal femur that may need to be corrected by osteotomy.

The knee reconstruction I developed in 1994 (Paley 1998) is called the superknee procedure. The superknee is a conglomerate procedure combining two or more of the following five procedures, three of which were previously described by other authors and two of which was developed by me: (1) Langenskiöld procedure (Langenskiöld and Ritsila 1992) for congenital dislocation of the patella, (2) MacIntosh procedure (Amirault et al. 1988) for ACL deficiency including extra- and intra-articular anterior cruciate ligament reconstruction using the fascia lata, (3) Grammont procedure (Grammont et al. 1985) for recurrent dislocation of the patella, (4) Paley procedure also referred to as the reverse MacIntosh (Amirault et al. 1988) to prevent external rotatory instability and to act as an extra-articular posterior cruciate ligament, and (5) Paley anterior approach to posterior capsulotomy of the knee. The combinations of these five procedures may be performed at the same time as a pelvic osteotomy or superhip procedure.

The superknee procedure is a combination of some or all of these components including extra- and intra-articular knee ligament reconstruction, patellar realignment, posterior capsulotomy, and knee flexor tendon releases. Typically, the superknee consists of the MacIntosh extra- and intra-articular ACL reconstruction, the reverse MacIntosh (Paley) PCL extra-articular reconstruction, the Grammont patellar tendon realignment, lateral release of the patella, and in some cases the modified Langenskiöld (Paley) procedure for patellar reduction. If performed with a superhip procedure, the incision is a distal extension of the superhip incision. If performed as an isolated procedure, a small second incision can be made to cut the fascia lata at its origin, thus reducing the length of the proximal extent of the midlateral thigh incision. If the superknee is performed without the superhip procedure, the entire surgery can be performed under tourniquet control. The release of the posterior capsule is performed only when there is a significant knee flexion contracture (≥15°).

13.5.2 Superknee Procedure (Ligamentous Reconstruction Only) (Fig. 13.4)

Step 1: *Fascia lata harvest*. The knee is exposed through a long S-shaped incision ending just distal to the tibial tuberosity distally and midlateral proximally. The anterior margin of the fascia lata (iliotibial band) and the posterior margin where it blends with the intermuscular septum are incised longitudinally. The fascia lata is transected at its musculotendinous junction with the tensor fascia lata and reflected distally until its insertion onto the tibia (Gerdy tubercle).

Step 2: *Ligamentization of fascia lata*. The fascia lata should be split into two longitudinal strips to make two ligaments. The posterior half is tubularized using nonabsorbable suture and

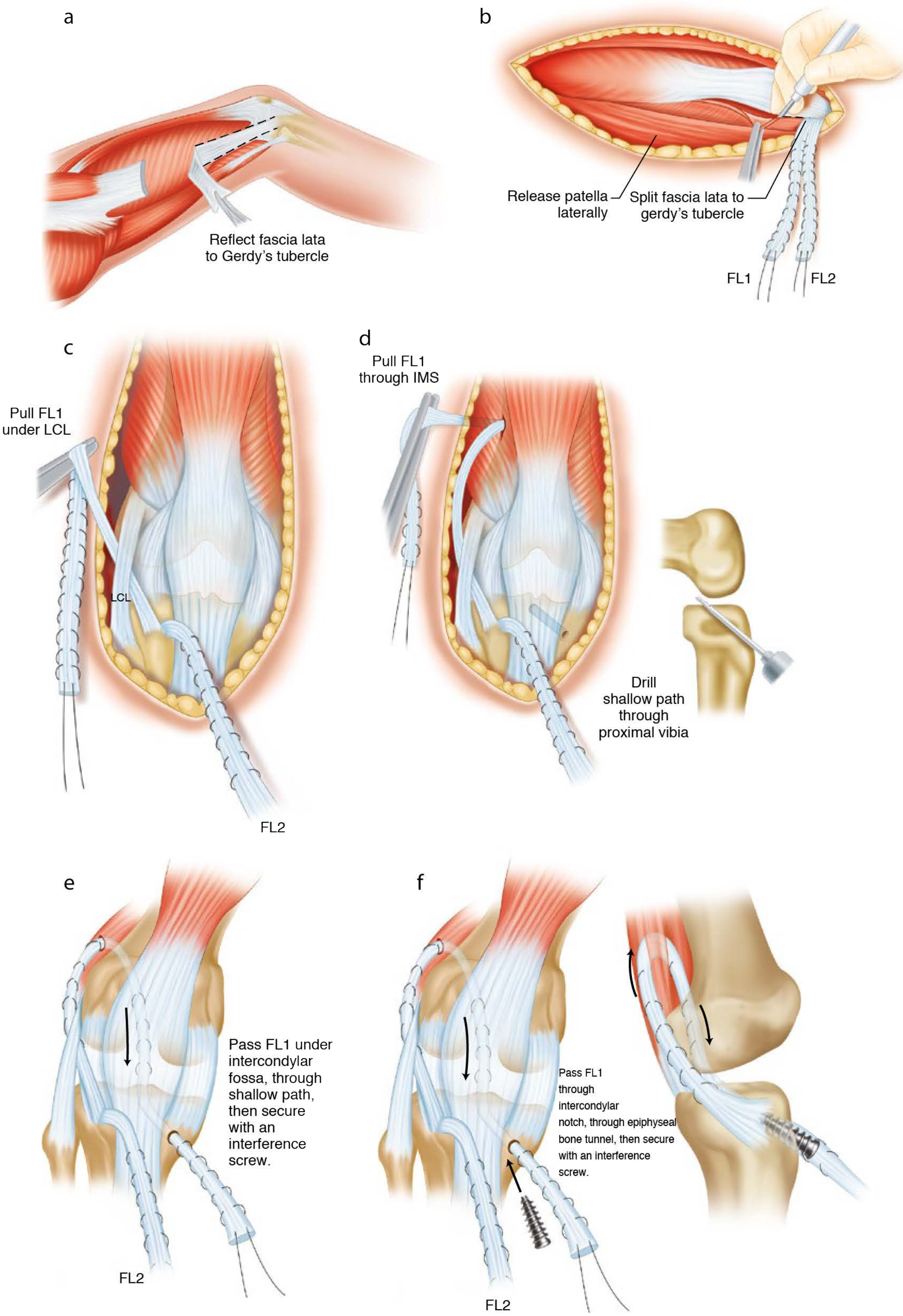


Fig. 13.4 Superknee surgical technique including MacIntosh and Paley reverse MacIntosh illustrations [AU9]

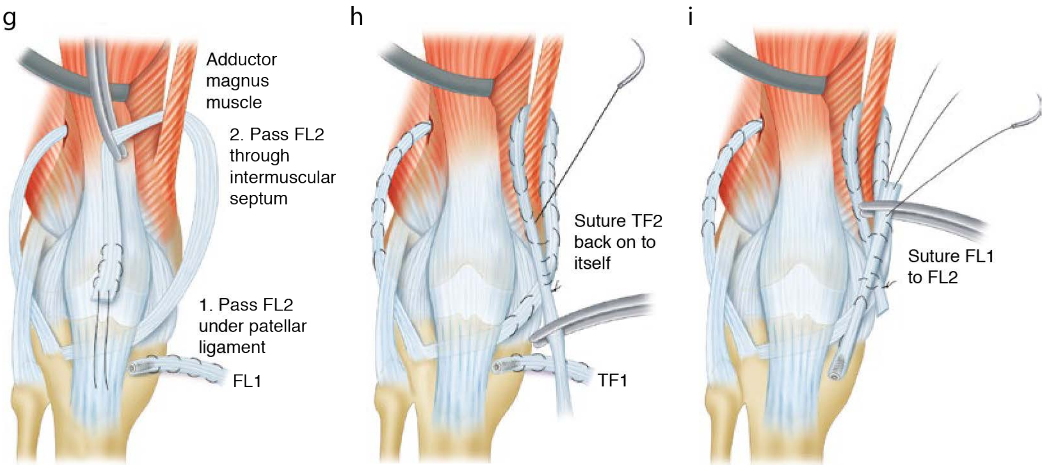


Fig. 13.4 (continued)

the Krackow whipstitch (Krackow et al. 1988) from distal to proximal. The medial half is left flat.

Step 3: *Lateral release and Grammont patellar tendon realignment*: If the patella tracks laterally but is not dislocated or dislocating, a lateral release and patellar tendon transfer should be performed to move the tendon medially. The lateral capsule and edge of the vastus lateralis should be cut to, but not through the synovium. The vastus lateralis is elevated from the lateral intermuscular septum and distal femur. If it is still acting as a major lateral tether of the patella, its tendon should be detached from the patella and then transferred medially at minimal tension. The lateral release is extended distally to the lateral aspect of the patellar tendon. This longitudinal deep incision should be extended past the tibial tuberosity along the crest of the tibia incising the proximal tibial periosteum. A parallel periosteal, para tendonous deep incision is made [AU10] medial to the patellar tendon. Using sharp dissection with a knife blade, the patellar tendon is elevated off the tuberosity trying not to remove any cartilage if possible. Once the tendon is detached, the periosteal extension of the tendon is elevated with the tendon so that the detached tendon remains tethered distally. This procedure (described by Grammont in German) is the pediatric equivalent of the Elmslie-Trillat (Grammont et al. 1985) procedure in adults. The patellar tendon can now be displaced medially and sutured medial to the tuberosity with an absorbable suture.

Step 4: *Extra-articular PCL reconstruction* (*reverse MacIntosh*). To prevent the tibia from rotating externally, which leads to subluxation of the patella and of the tibia on the femur posteriorly, a medial extra-articular ligament can be created using the medial half of the fascia lata. This creates a strap going around the medial tibia tethering it to the medial distal femur. This is the opposite direction of the extra-articular ligament created with the posterior half of the fascia lata (described in the next step). The lateral extra-articular ligament was described by David MacIntosh (one of my former professors from Toronto) for ACL reconstruction. In his honor and memory and in recognition of his idea of a lateral extra-articular ligament, I refer to the medial extra-articular ligament procedure that I innovated as the reverse MacInstosh or extra-articular PCL procedure. To anchor this ligament to the medial femur, it is necessary to elevate the skin flap in a medial direction. The anterior skin flap is kept as thick as possible and is reflected medially until the posterior border of the vastus medialis can be visualized. The medial intermusclar septum (very rudimentary) and the adductor magnus tendon are located posterior to the vastus medialis muscle. Care should be taken dissecting in this area to isolate and not damage the saphenous nerve as it exits from the quadriceps as the terminal branch of the femoral nerve. A subperiosteal tunnel is created around the adductor magnus tendon. The anterior limb of the fascia lata is now passed under the patella tendon, through a tunnel of superficial medial retinaculum and looped around the adductor magnus insertion (from posterior to anterior) and then sutured to itself with nonabsorbable suture. To [AU11] ensure that this new ligament is isometric, a suture anchor can be inserted into the distal tibial epiphysis, just distal to the physis at the posterior junction of the physis with the posterior cortex (this point corresponds to the center of rotation of the knee. I like to use an absorbable anchor for this. The suture from this anchor is then tied around the fascia lata in 90° of knee flexion while the ligament is under tension. After that the part that is looped around the adductor magnus can be sutured to itself using this suture again. This extra-articular ligament should always be tensioned with the knee in 90° of flexion. If it is tensioned in extension, it may restrict flexion. The excess ligament is saved and sutured to the remaining end of the new ACL as it exits the tibia in the next step.

Step 5: *MacIntosh extra-+intra-articular ACL reconstruction*. David MacIntosh described first an extra-articular ligament reconstruction for the ACL-deficient knee and later a combined extra- and intra-articular reconstruction with the "over-the-top" technique. This method although no longer used in sports medicine is a very useful technique for congenitally deficient knees. In CFD the instability pattern is different than in an isolated tear of the ACL. There is more rotary instability in CFD. Therefore, a purely intra-articular ligament reconstruction is insufficient. In my early experience, we made this mistake and had a lot of recurrent instability. The combination of extra- and intra-articular ACL ligament reconstruction is ideal for CFD. Having studied under MacInstosh and having learned this procedure directly from him, it was natural for me to think about its application in the CFD cruciate-deficient knee. I have modified this procedure slightly to adapt it to the skeletally immature knee. It can now be safely done as young as 2 years of age.

The lateral collateral ligament (LCL) is identified using Grant's test. The leg is put in a figure four position, which allows the tensioned LCL to be easily palpated. I dissect the LCL while it is under tension and identify its anterior and posterior borders. An extra-articular tunnel is created under the LCL. The posterior limb of the fascia lata is passed through the LCL tunnel from anterior to posterior. A subperiosteal dissection is done in the over-the-top region preserving the intermuscular septum attachment. A curved tonsil clamp introduced through the "over-the-top" tunnel is used to perforate the posterior knee capsule centrally. Next, a drill hole needs to be made in the anterior tibial epiphysis to anchor the ligament to bone. Drill a guidewire through the anterior tibial epiphysis using image intensifier guidance. The wire should start proximal to the proximal tibial physeal line, lateral to the patellar tendon. The wire should be directed proximal and posterior to exit into the center of the knee about half way back on the tibia. As these patients don't have a true notch and a notchplasty is not an option in children, it is better that the ligament insert more posterior on the tibia than in the normal knee. This avoids damage to the ligament with knee extension. Once the wire is confirmed to be in the correct position in both the AP and lateral views, a hole is drilled with a cannulated ACL reamer. The diameter of the reamer chosen is matched to the diameter of the tubularized fascia lata ligament being passed (use the ligament sizers to determine this). A suture passer is then passed through the tibial epiphyseal tunnel and out the capsular perforation through the "over-the-top" tunnel laterally. The suture connected to the fascia lata is looped through the suture passer and pulled through the knee to exit anteriorly through the epiphyseal tunnel. The tubularized fascia lata is now pulled through the knee capsule and out the epiphyseal tunnel. As was done on the medial side, a suture anchor is placed into the supracondylar region to help anchor the extra-articular ligament and to support the over-the-top point. The extra-articular ligament should be tensioned in full knee extension. Once it is secured, the rest of the graft is anchored to the bony tibial epiphyseal tunnel using an absorbable headless interference screw. It too should be tensioned in full knee extension to prevent a flexion contracture of the knee. As the patient's epiphysis grows, the ligament becomes more taut. Therefore, one does not need to worry that it is not tight enough. I like to secure the ligament by suturing the posterior fascia lata graft to the anterior graft.

13.5.3 Superknee Procedure with Patellar Realignment for Dislocated/Dislocating Patella (Fig. 13.5)

13.5.3.1 Langenskiöld Patellar Realignment

If the patella is dislocated or dislocating, a modified version of the Langenskiöld (Langenskiöld and Ritsila 1992) procedure is performed before the ligament reconstruction. First the capsule is incised and separated from the patella and synovium medially and laterally. On the medial side the two layers are separated all the way to the medial gutter. The medial capsule is cut transversely at its distal end. The patellar tendon and the quadriceps are also separated from the synovium distally and proximally, respectively. The synovium is now incised circumferentially around the patella, separating the patella from the synovium completely. The quadriceps tendon is left attached to the patella proximally, and the patellar tendon remains attached to the patella distally. The synovium is separated from these structures. The synovium now has a patella-sized hole in it. The synovium is sutured closed in a longitudinal direction. This leaves the patella temporarily as an extra-articular bone. The patellar tendon is elevated from the apophysis by sharp dissection after circumscribing a medial and lateral incision extending distally into the periosteum (Grammont procedure). After the tendon is elevated, it is shifted medially at least a centimeter (in the original Langenskiöld, it is detached from the tuberosity). A longitudinal incision is made in the [AU13] synovium over the center of the patella is inserted into this new hole in the synovium, and the synovium is sutured to the patella circumferentially. The medial capsule with the vastus [AU14] medialis is now advanced over the top of the patella and stitched to its lateral border. The lateral capsule is left open. If the Paley reverse MacIntosh procedure is used, the fascia lata should not be fixed in place until after the Langenskiöld repair is completed.

13.5.4 Superknee Procedure with Knee Flexion Deformity (Fig. 13.6)

Knee flexion contracture release: If there is a knee flexion deformity >15°, it can be treated by posterior capsular release. This is often done in combination with a superhip procedure or one of the knee reconstruction procedures described above. 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 common peroneal nerve can be palpated in most people over the neck of the fibula. A fascial incision is made through the superficial fascia that covers the nerve. Define the proximal and distal borders of the nerve. It is not necessary to remove the fat over the nerve. Follow the common peroneal nerve until it disappears into the lateral compartment. Make a transverse incision over the lateral compartment fascia and expose the muscle of the lateral compartment. Retract the muscle of the lateral compartment medially to expose the underlying arcade of fascia covering the common peroneal nerve. This is the decompression of the first tunnel. The common peroneal nerve can be seen dividing into deep and superficial branches, the superficial running down in the lateral compartment, and the deep branch going in the direction of the anterior compartment of the leg.

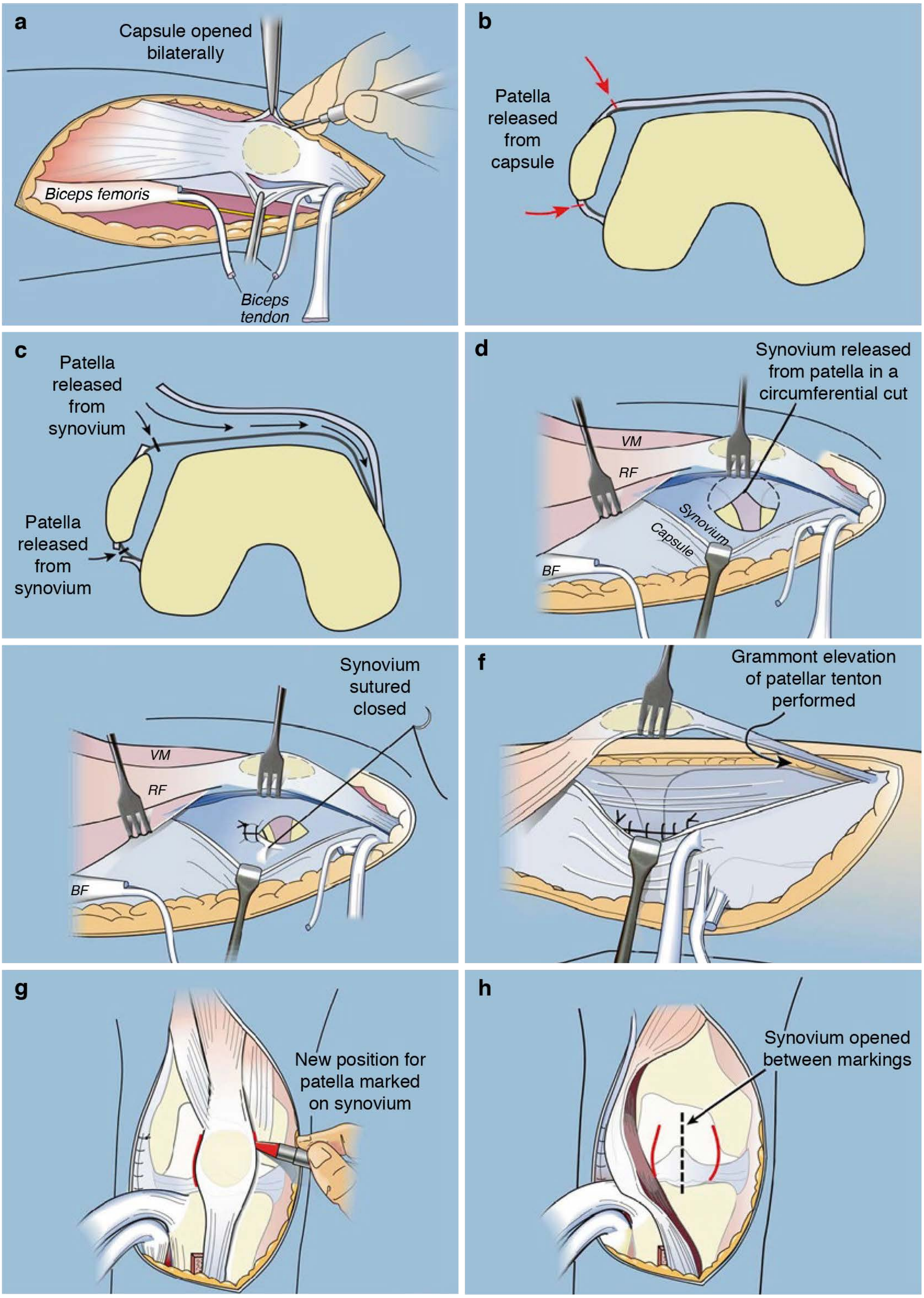


Fig. 13.5 Superknee surgical technique including Grammont, modified Langenskiöld, and Paley reverse MacIntosh procedure illustrations [AU12]

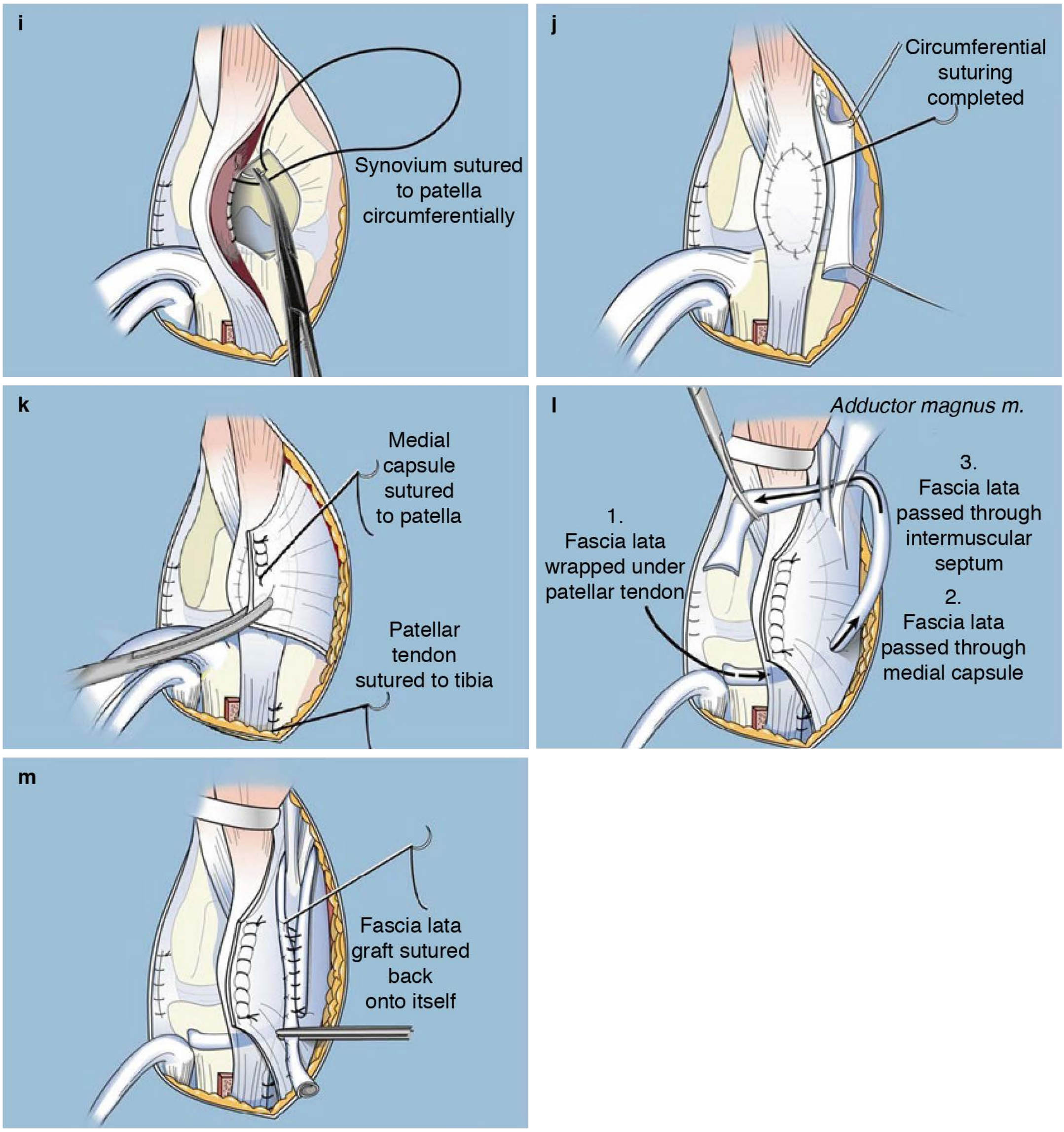


Fig. 13.5 (continued)

Extend the transverse compartment fasciotomy across to the anterior compartment. Notice the intermuscular septum separating the two compartments and confluent with the fascia overlying both compartments. Dissect the muscle of both sides of the septum. Be careful not to cut any superficial sensory branches of the nerve that may ascend the septum and innervate the skin overlying the compartments. With the septum exposed on its medial and lateral aspects, cut it from anterior to posterior. Be careful to stop as soon as the septum ends. Immediately below the septum a band of fat can often be seen. This fat band contains the deep peroneal nerve. This completes the decompression of the deep peroneal nerve (second tunnel). Extend the decompression of the common peroneal nerve under the biceps muscle. With the nerve protected and visualized, Z-lengthen the biceps muscle and tendon. In congenital cases, there is little tendon and mostly

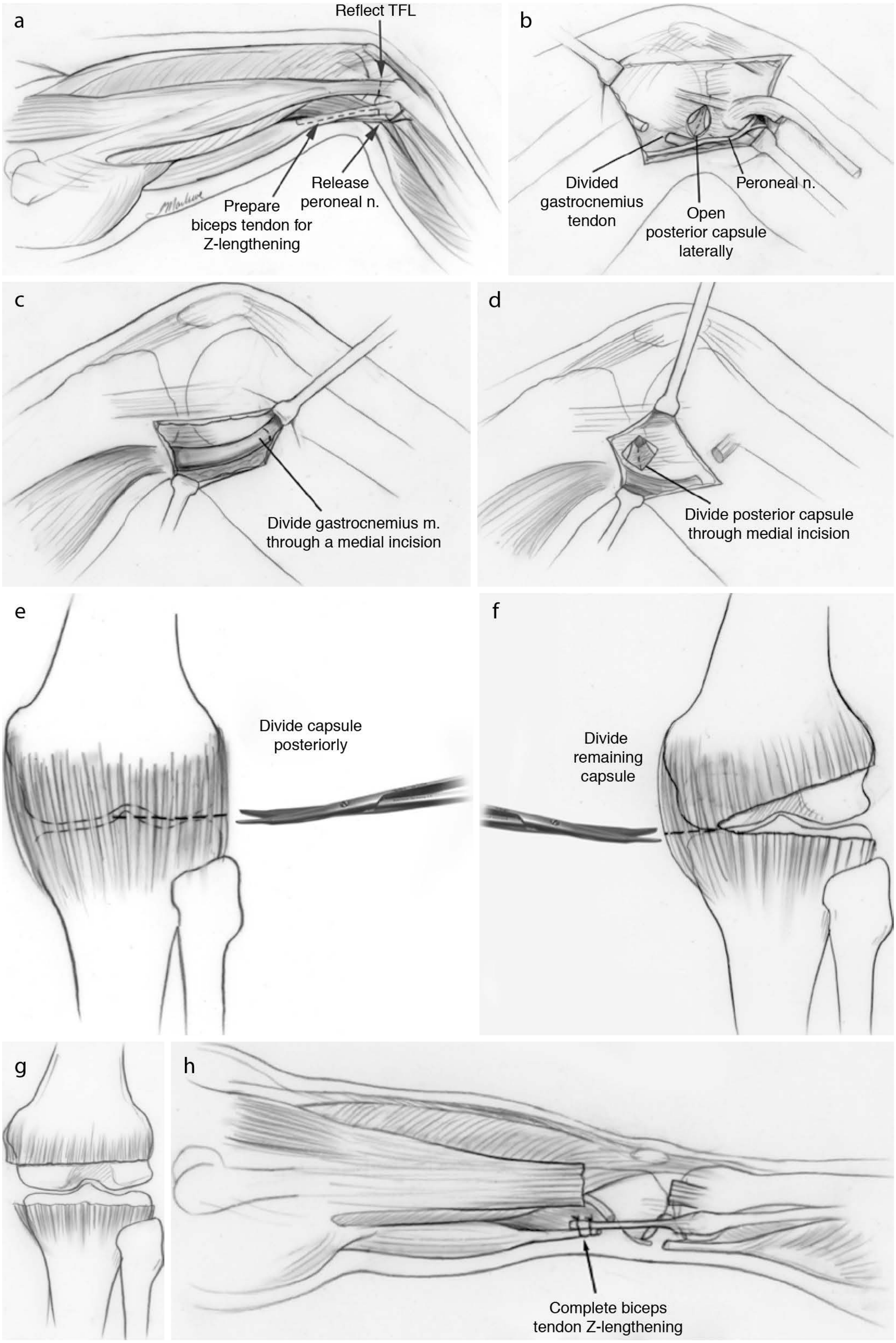


Fig. 13.6 Superknee surgical procedure including posterior capsulotomy, fascia lata, biceps and gastrocnemius tendon lengthening, and peroneal nerve decompression illustrations [AU15]

muscle. The biceps muscle consists of two parts: the short and long heads. To avoid damage to this muscle, the short head should be dissected off of the periosteum of the femur. Try and keep as much of the muscle together for the Z-lengthening and reflect this part proximally and isolate as much of the tendon by sharp dissection off of the muscle and reflect this part distally.

The lateral head of the gastrocnemius should be released from the femur. It has a very broad insertion of muscle and tendon. The lateral capsule can now be identified. Dissect the contents of the popliteal fossa away from the posterior capsule. To confirm that it is the posterior capsule, incise it posterolaterally and enter the knee joint. With the knee flexed the only vascular structure that one should see at the level of the knee joint is the central geniculate artery. This can be dissected free and cauterized. Care should be taken to make sure that the dissection does not inadvertently go distal to the level of the knee joint. If the dissection is behind the tibia instead of the knee joint line, the anterior tibial vessels may be encountered or injured. The rest of the popliteal soft tissues can be carefully dissected off of the capsule all the way to the medial side. The capsule can be cut under direct vision. A head lamp may be useful for this part. Once the capsule is open, try and extend the knee joint. If there is still too much resistance, then the medial side should be exposed. When performing a superknee with ligament repair, a medial skin flap needs to be elevated. If the posterior capsulotomy is being performed without the rest of the superknee, then the dissecting scissors can be inserted from lateral to medial until they can be seen under the medial skin. A separate small skin incision can be made medially to visualize the medial capsule. The medial head of the gastrocnemius is identified as the only structure with a muscle inserting proximally. The medial head should be cut from the femur, being careful not to injure the femoral vessels that lie just lateral to the medial head of gastrocnemius. The capsule is dissected free of the medial popliteal fossa to communicate with the lateral dissection. The capsule can then be cut under direct vision from both sides. The knee FFD can be corrected by extending the knee. The collateral ligaments are left intact. If after the capsulotomy the medial hamstrings are felt to be tight, they can be lengthened through the medial part of the dissection. I prefer to recess the aponeurosis of the semitendinosus and semimembranosus tendons rather than do a Z-lengthening.

After doing the knee capsular release, the knee is examined for instability. If it is unstable, then the ligamentous reconstruction of the superknee procedure is carried out as in Fig. 13.4.

The above hip and knee problems must all be addressed before beginning femoral lengthening procedures. These are called preparatory surgery for lengthening. By performing preparatory surgery, we prevent complications that occur during lengthening: Pelvic osteotomy prevents hip subluxation/dislocation; proximal femur reconstruction (superhip) prevents worsening of coxa vara, proximal migration of femur, and dislocation of hip; patellar realignment prevents patellar dislocation and extension contracture of knee; ACL-PCL reconstruction prevents knee subluxation/dislocation and late problems of knee instability in adolescence; and fascia lata excision prevents knee stiffness and contracture, pressure on joint and physis, valgus deformity, and patellar subluxation.

13.6 Rehabilitation After Superhip and Superknee Surgery

13.6.1 Preoperative

An evaluation can be performed preoperatively when warranted to establish a relationship and determine any special concerns that the patient may have while in the hospital. Evaluation should include past medical and surgical history, pain level, preoperative range of motion (ROM), strength, sensation, limb length discrepancy and/or deformity, posture, coordination, and mobility including transitions and gait. Initiation of an exercise program may be needed prior to surgery to address deficits identified. The preoperative visit is also useful to educate the family and patient on what to expect after surgery and establish realistic postoperative goals.

13.6.2 Acute

The goal of inpatient PT after a superhip/knee surgery is to teach the parents how to manage with a child in a spica cast. Necessary skills include bed mobility, carrying and holding their child, transfers to wheel chair and car seat, equipment procurement (wheel chair, safety belt for car seat, sliding board if needed), and peroneal hygiene (diaper vs. toilet) including how to keep the spica cast clean.

13.6.3 First 6 Weeks After Surgery

After the patient is discharged, they return to the hospital to have the spica cast made into a removable cast. The spica can be removed for gentle passive range of motion (PROM) of flexion and extension and abduction of the involved hip and flexion and extension of the knee not to exceed 0–90°. Therapist should be very gentle and not push the extremes of motion. It is important to support the limb both proximally and distally at all times when performing PROM. Parents will also be instructed in PROM techniques with handover hand training and video recordings for reinforcement of proper technique at home.

13.6.4 After 6 Weeks

The spica cast is discontinued after 6 weeks. If the 6-week radiographs show adequate bony healing, then the patient can progress to weight bearing and active range of motion as well as gait training. A PT evaluation is performed after the SPICA cast is removed and includes assessment of strength, range of motion (ROM), proprioception, motor planning, mobility, transitions, gait, and developmental milestones. Treatment will aim to gently restore passive and active (P/A) ROM in order to regain independent mobility including rolling, transitions, standing, and walking with assistive device as needed. A gradual return to independent walking with a shoe lift will occur.

The goal at this stage is to regain mobility, strength, and gait training and restore range of motion. The end goal is to restore the child to normal function before they proceed with limb lengthening.

13.7 Femoral Lengthening of Type 1 CFD

13.7.1 Choice of Osteotomy Level for Lengthening of the Congenital Short Femur

Distal osteotomy for lengthening has the advantage of a broader cross-sectional diameter for better bone formation and less deforming forces from the adductors and hamstrings. If distal femoral valgus is present, it can be corrected using a distal lengthening osteotomy. Distal osteotomy lengthening is closer to the knee joint and therefore applies greater forces to the knee joint. This increases the risk of knee stiffness and subluxation. These risks can be reduced by articulating across the knee joint with extension of the fixator to the tibia. Proximal osteotomies have less effect on the knee, but are more prone to poor bone formation and consolidation in patients with CFD. There is also a higher rate of fracture after removal of external fixation in the proximal than in the distal lengthening groups. Proximal osteotomies have a greater effect on the hip joint and produce a much higher risk of adduction contracture, hip subluxation, and dislocation. Extension of the external fixator across the hip is more complex and less desirable than articulated distraction of the knee. For all of these reasons, I prefer to lengthen at the distal femur in patients with CFD.

The external rotation deformity of the femur with CFD should be corrected only by using a proximal osteotomy. 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 internal rotation osteotomy does not change the orientation of 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 valgus osteotomy, whereas valgus deformity of the knee is corrected using a distal varus osteotomy.

If the femur has undergone previous hip preparatory surgery such as superhip reconstruction or combination of Dega and proximal femoral osteotomy for milder cases, the proximal femoral deformities will already have been corrected and no proximal osteotomy is required at the time of lengthening. If hip preparatory surgery was not required, but there is external femoral torsion and perhaps mild proximal varus, then a proximal internal rotation-valgus osteotomy of the femur is carried out together with a distal lengthening osteotomy.

The distal femoral lengthening osteotomy should also acutely correct the valgus and any mild flexion deformity of the knee. For valgus correction alone no peroneal nerve decompression is required. However, if a flexion deformity is to be corrected acutely, the peroneal nerve should be decompressed just prior to the correction at the same surgery. As noted above, this distal 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. Therefore, the regenerate bone in the distal femur is wider and stronger and subjected to axial deviation muscular forces than the proximal femur.

In older children with a wider medullary canal, implantable limb lengthening or lengthening over nail can be performed. A proximal osteotomy can be used for lengthening with nails because there is little risk of refracture and bone does not extend external fixation treatment time since the internal rod supports the bone until bone consolidation is complete. Intramedullary nailing in children adds the risk of disturbance of growth of the apophysis and avascular necrosis of the femoral head. To avoid the latter, we use a greater trochanteric starting point and a nail with a proximal bend (e.g., trochanteric entry femoral, or humeral or tibial). To avoid a coxa valga deformity, we prefer to use this technique in patients with some coxa vara. A theoretical apophysiodesis created by the nail can lead to gradual correction of residual coxa vara. I state theoretical because I have not observed this complication yet. Fixator only lengthening is the method we usually use for the first lengthening. LON or implantable lengthening is used for the second or third lengthening if the anatomic dimensions of the femur permit.

13.7.2 Soft Tissue Releases for Lengthening in Cases of CFD

Soft tissue releases are essential in conjunction with lengthening to prevent subluxation and stiffness of knee and hip. If a preparatory hip-knee surgery has already been performed, then the fascia lata and rectus femoris and in some cases biceps have already been lengthened. There is no need to repeat soft tissue releases that were already performed.

If soft tissue releases have not been performed, they should be carried out at the time of the lengthening surgery. If there is no contracture or tightness at the time of the index procedure, the soft tissue releases can either be performed at the index procedure or delayed until these soft tissues become contracted (6 weeks later). I usually prefer to do the soft tissue releases at the time of the index surgery to avoid an additional anesthetic.

Before surgery, the range of motion of the hip and knee should be evaluated using muscle lengthening tests and the presence of contractures or limitation identified. The muscle lengthening tests are the straight leg raising test (popliteal angle) for the hamstrings, the prone knee flexion test (Ely test) for the rectus femoris, hip abduction range for the adductors, and hip adduction range with knee and hip in extension for the fascia lata. One can also do an Ober test for the fascia lata. If a patient is able to straight leg raise so that the hip is at 90° of flexion and the knee is in full extension (popliteal angle = 0°), the hamstring muscles are not tight and require no treatment before lengthening. If there is a popliteal angle >0°, the hamstrings are already tight and will lead to contractures during lengthening. The medial and lateral hamstrings should be fractionally lengthened through a single midline posterior incision proximal to the knee, to reduce the popliteal angle to 0°.

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 (negative Ely test). I still prefer to release the rectus femoris (RF) from the anterior inferior spine since it will still become tight during lengthening. Obviously, if the Ely test is positive before surgery (pelvic flexion with prone knee bend), the RF should be released through a small anterior inguinal incision.

If hip abduction is limited, especially with proximal lengthening, percutaneous adductor tenotomies should be performed of the adductor [AU16] longus and gracilis tendons. During lengthening, if more severed adduction contracture or hip subluxation develops, an open more extensive adductor release (including adductor brevis). Distal adductor magnus release has been described for congenital femoral lengthening (ref Richard Gross). I have not found this to be useful or necessary. For distal femoral lengthening, it is not necessary to release the hip adductors in most cases. For proximal lengthening, adductor release is very important.

In every case of CFD lengthening, the FL should be lengthened if it has not already been excised previously. The entire fascia lata is transected at the level of the proximal pole of the patella. I prefer to make a 3 cm longitudinal incision at the posterior edge of the FL where it connects with the intermuscular septum. The FL is transected by dissecting anterior to the incision. By making the incision more posterior, the biceps muscle can be easily exposed by retracting the incision posteriorly. The lateral biceps can be safely recessed.

Proximal release of the FL is almost never done at the index procedure. If however hyperlordosis develops, the fascia lata is released as it passes over the greater trochanter through a small lateral incision. This is almost never done at the index procedure but is performed during the lengthening or in conjunction with frame removal, to treat the hyperlordosis, abduction contracture, and hip flexion contracture secondary to the lengthening. We have noticed that patients who have undergone the superhip never develop this complication, while those who have just had distal incision of the fascia lata may develop this complication. They also have less difficulty maintaining knee range of motion during lengthening. We assume this is due to the fascia lata resection. We are therefore opting for limited incision complete FL resection in non-superhip patients. This can be achieved through two or three small incisions with tunneling between them.

13.7.2.1 Botulinum Toxin Injection

Botulinum toxin should be injected into the quadriceps to a limit of ten units of Botox per kg body weight. I prefer to limit the volume of the saline into which we mix the Botox to avoid systemic toxicity. I therefore usually inject with a total of 3–5 cc. The effect of Botox has been shown to be helpful in prevention of muscle contracture in an animal model of lengthening (Sam Rosenfeld). In humans I have observed that the role of Botox is to decrease muscle stretch pain especially due to muscle spasm and during physical therapy. The Botox should be injected only to the quadriceps muscles both because the amount allowable during one injection is only sufficient for the quadriceps and also because the quads are the primary muscle to have spasm during therapy.

13.7.3 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 medial-lateral 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 of the knee to subluxate with lengthening. Knee subluxation with lengthening is usually posterior or posterolateral (posterior plus external rotation of the tibia on the femur), but can also be anterior. Knee extension usually reduces posterior subluxation before lengthening. Therefore, to prevent posterior subluxation, some surgeons recommend splinting the knee in extension throughout the distraction phase (1). This promotes knees stiffness while protecting the knee from subluxation. I prefer to protect the knee by extending the external fixation to the tibia with hinges. The hinges permit knee motion while preventing posterior as well as anterior subluxation. They also prevent pressure being transmitted to the knee joint cartilage. Hinges are an integral part of circular external fixators such as the Ilizarov device. They are now also integral parts of some monolateral external fixators (e.g., Orthofix LRS, Smith and Nephew Modular Rail System).

A less common knee instability is anterior subluxation/dislocation of the tibia on the femur. 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). These dislocations can be due to anterior deficiency of the distal femur (the lateral radiograph of the knee shows a lack of the anterior protuberance of the femoral condyles) or posterior deficiency or rounding of the posterior tibial plateaus. The posterior rounding of the tibial plateau is the more common bony deficiency. The treatment of such instability is by the superknee procedure combined with a posterior elevation osteotomy of the tibial plateau. In growing children, the osteotomy is epiphyseal, while in skeletally mature patients, it starts in the metaphysis.

13.7.4 Distal Femoral Lengthening-Ilizarov Fixator 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 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° to the shaft), and the distal ring parallel to the knee joint line. After the osteotomies, when all the rings and 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° anterolateral to the first pin. The proximal arch is perpendicular to the floor with the leg 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 arthrogram is 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 midsegment of the femur. This pin is at 7° to the shaft of the bone. At that point, the proximal subtrochanteric osteotomy can be performed. This is done percutaneously by making multiple drill holes and then using an osteotome. The osteotomy 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, with multiple drill holes and an osteotome. The only wire used is removed to avoid tethering of the quadriceps and fascia lata.

13.7.4.1 Knee Hinges

The last step is to extend the fixation to the tibia using hinges. The center of rotation of the knee is located at the intersection of the posterior femoral cortical line and the distal femoral physeal line (1) in the plane where the two posterior femoral condyles are seen to overlap on the lateral view. For younger children, it is helpful to inject arthrographic dye into the knee to visualize the posterior femoral condyles. It is important that the distal femoral ring, which is parallel to the knee, appears 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 (perform a drop test: drop the tibia and see if it flexes without any catch), a second and third tibial half-pin are added. Finally, a removable knee extension bar is inserted between the distal femoral and the tibial half-ring.

13.7.5 Distal Femoral Lengthening-Orthofix Fixator Technique

One needs to start by identifying the center of rotation axis of the knee joint (see description of this above). A 1.8 mm wire is drilled into the lateral edge of the physis at the intersection of the posterior cortex of the femur with the physis in line with the plane of overlap of the posterior femoral condyles. The Orthofix LRS (pediatric or adult depending on the size of the child) is lined up with the hinge axis through its most distal hole. A commercially available sandwich clamp is used or if one is not available an extra lid is used in the pin clamp to create a second layer of pin holes more anteriorly. The fixator bar is lined up with the shaft of the femur and [AU17] the most proximal-most half-pin inserted. The distal-most pin is then drilled one hole proximal to the center of rotation pin. The LRS without sandwich clamps is then used to place the rest of the pins (three proximal and three distal). If there is a distal femoral valgus that is to be corrected, an acutely swivel clamp is used for distal pin placement. When using a pediatric LRS, the three-hole pin clamp contains two half-pins only, since one hole is used up for the knee center of rotation. A third pin is added using Ilizarov cubes connected via these two pins. Before reapplying the sandwich clamps, the osteotomy is performed and the distal valgus corrected acutely. After the correction the fixator can be exchanged for one with straight clamps and with the sandwich attachments. All the pins should be in the upper deck of the double-decker sandwich clamps. The only pin in the lower deck is the knee axis pin. This pin does not enter the patient's leg. It is a 6 mm segment of pin which protrudes laterally. A Sheffield clamp from Orthofix is applied to this pin to act as a hinge. It is locked in place by putting a cube lateral to it with a set screw to prevent it moving outward.

Conical washers are used between the Sheffield clamp and the LRS to prevent stiction friction. The Sheffield clamp is left partially loose to permit motion. A 1/3 Sheffield arch is attached to the clamp arching towards the tibia. An anteroposterior pin is inserted and the drop test (see above) is performed. If there is friction, the Sheffield clamp should be loosened. If friction persists adjust the connection of the pin to the Sheffield arch. If friction persists the axis pin may need to be bent slightly to alter the axis of rotation. Once the drop test is negative, two more oblique pins are inserted into the tibia and connected to the Sheffield arch using cubes. A removable knee extension bar is fashioned from Ilizarov parts to be used especially at night time. If there is an unstable hip, an axis pin for the hip can also be fashioned and attached from the proximal clamp. The same Sheffield clamp and arch arrangement are used. Two pins are placed in the pelvis from the anterior inferior and superior spines extending posteriorly. These are fixed to the Sheffield clamp to prevent proximal subluxation of the hip during lengthening. As one can see, the same principles are applied when using monolateral as with circular fixation, i.e., hinge fixation across joints when there is a joint at risk.

13.7.6 Distal Femoral Lengthening-Modular Rail System (Smith and Nephew, Memphis) (Figs. 13.7 and 13.8) Technique

This external fixator was designed by me specifically for the CFD patient. It is able to articulate and span across the hip and knee joint either separately or at the same time. It also has clamps that allow fixation in the oblique plane in addition to the frontal plane producing delta fixation.

Step 1: Preconstruct the external fixator using pediatric or adult rail segments, two pin clamps, knee hinge, and tibial rail segment.

Step 2: Inject radiocontrast solution into the knee joint.

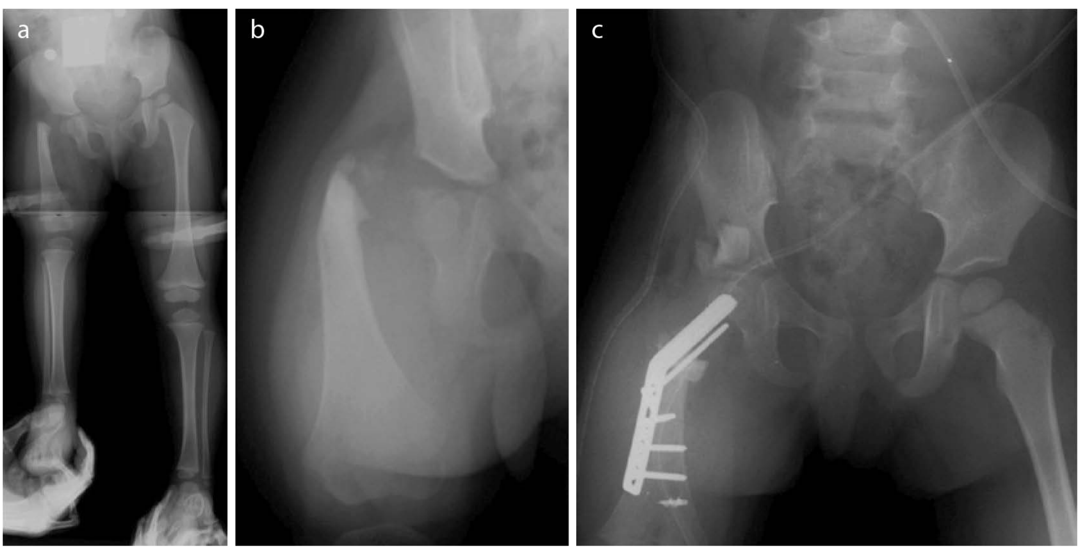


Fig. 13.7 CFD Paley type 1b with delayed ossification of femoral neck (**a**, **b**). Superhip procedure at age 2 including insertion of BMP in femoral neck (**c**). The neck is fully ossified by age 3 (**d**). First lengthening performed at age 4 with Smith and Nephew Modular Rail System external fixator with articulation across the knee joint (**e**, **f**). Eight centimeters of lengthening achieved (**g**). Removal of external fixator with Rush rodding of bone to prevent fracture (**h**)

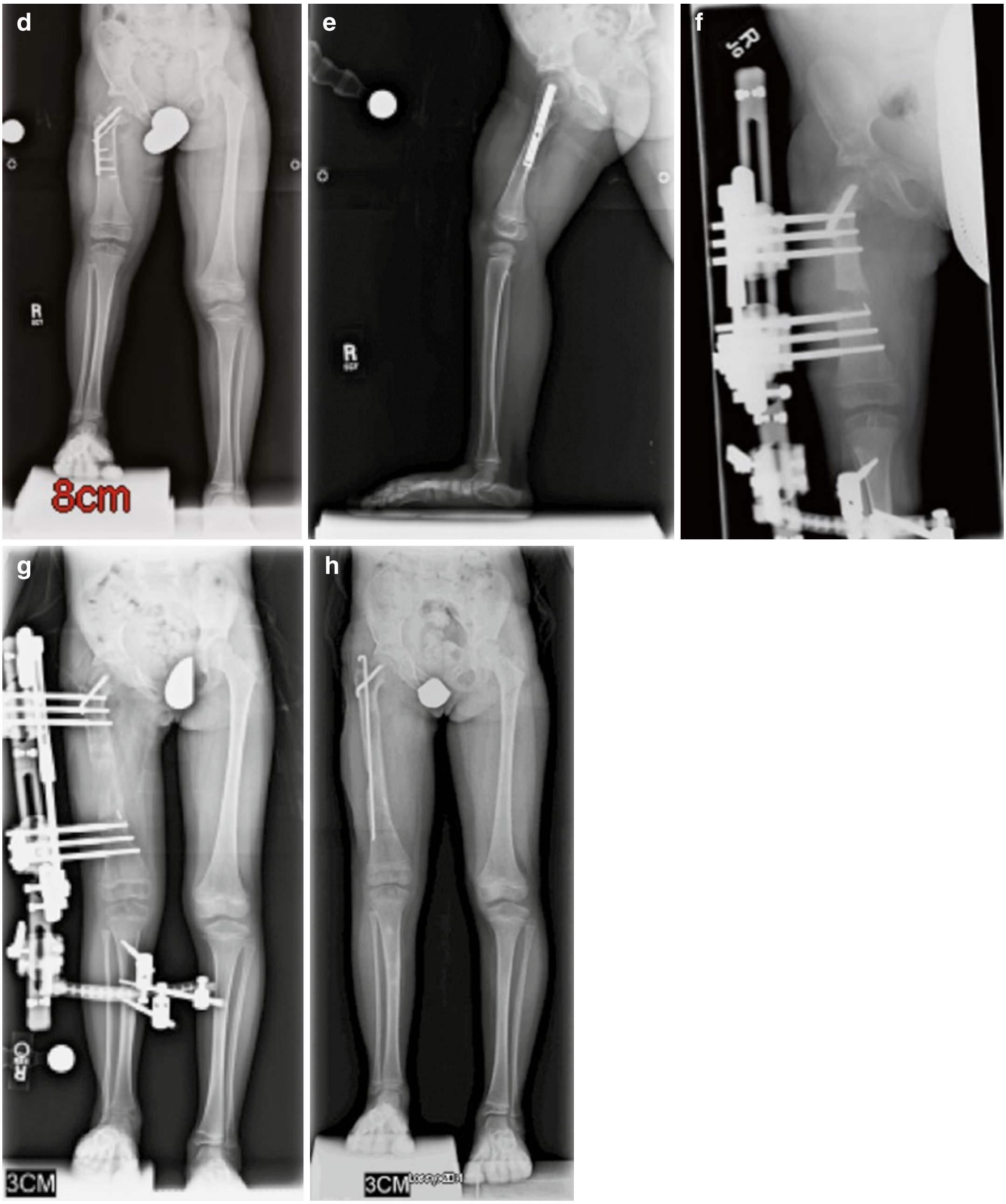


Fig. 13.7 (continued)

Step 3: Bring the image intensifier to the lateral projection and rotate the femur until the posterior aspect of the medial and lateral femoral condyles overlap.

Step 4: Insert a 2 mm Steinmann pin into the center of rotation (COR) of the knee joint. The COR is defined as the point of intersection of the posterior femoral cortex with the distal femoral physis. In adults it is the intersection of Blumensaat's line with the posterior cortex of the femur. Confirm on the AP view that the pin is parallel to the joint line.

Step 5: If there is no valgus or flexion deformity, proceed to drilling a 1.8 mm wire into the prox-

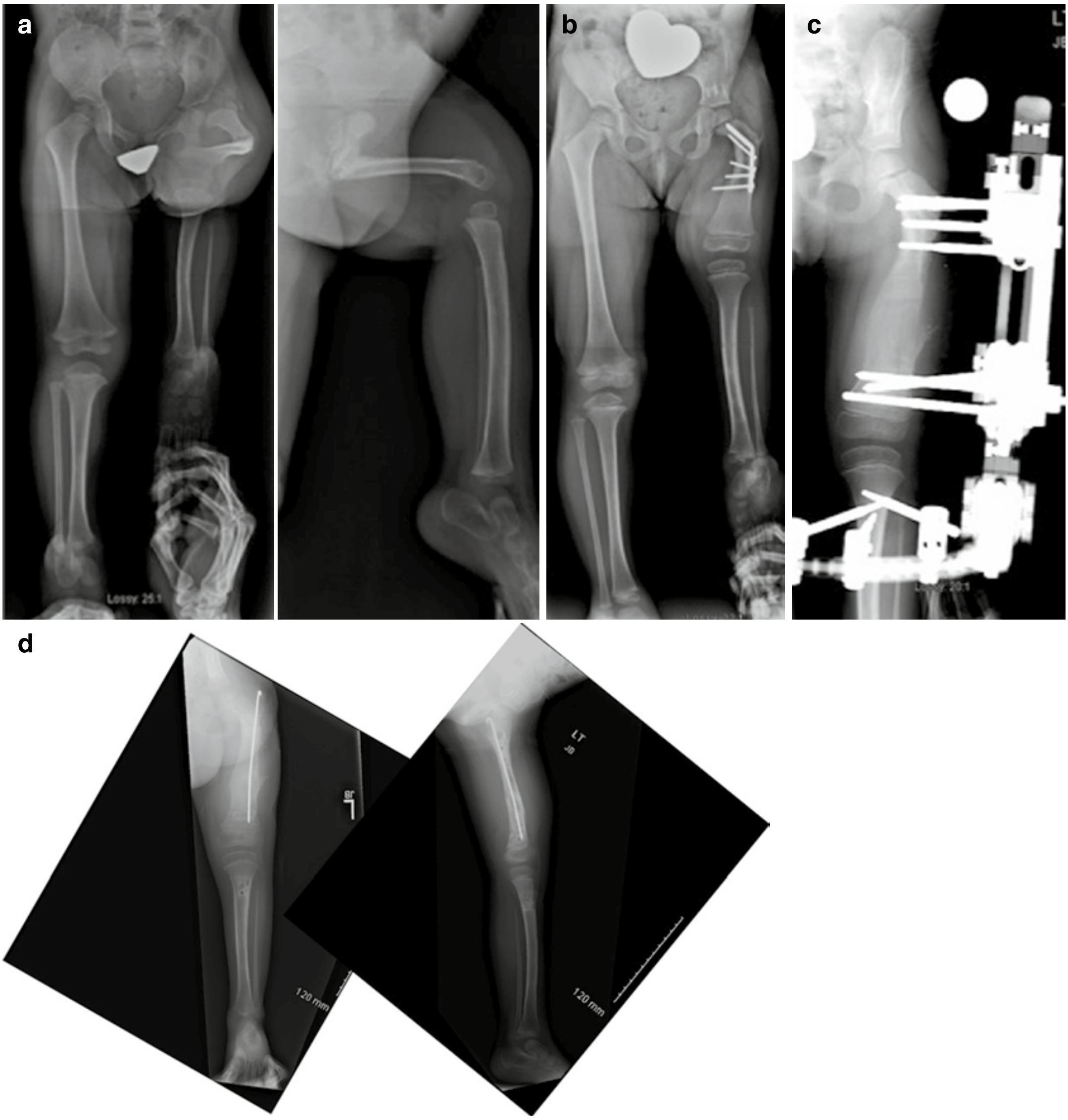


Fig. 13.8 A two-year-old girl with CFD Paley type 1b with delayed ossification and severe angulation of the subtrochanteric level of the femur (**a**). The deformity is fully corrected and the femur is healed after the superhip surgery (**b**). Lengthening of the femur was performed at age 4 years (**c**). X-rays after lengthening of the femur 7 cm and insertion of Rush rod (**d**)

imal femur parallel to the Steinmann pin. This wire should be as proximal as possible in the femur but distal to the trochanteric apophysis. Confirm the location of this wire making sure that it is located in the mid-diaphysis or slightly posterior to the midline. Drill a hole using a cannulated drill appropriate to the pin size preferred (e.g., 4.8 mm for a 6 mm pin, 3.8 mm for a 4.5 mm pin, 3.2 mm for a 3.5–4.5 mm pin).

Step 6: Insert a pin of the appropriate thread length corresponding to the diameter of the bone at that level. It is better not to leave threads outside of the near cortex to obtain the maximum bending strength out of each pin. The bending strength of the pin is related to the smallest diameter protruding from the near cortex.

Step 7: Mount the preconstructed MRS onto the Steinmann pin and the proximal half-pin. The

Steinmann pin goes through the cannulation in the hinge of the fixator.

Step 8: Drill a 1.8 wire through the distal-most pin hole of the clamp. I prefer to use an overhang clamp to minimize the distance from the most distal pin to the hinge. This pin should be just proximal to the physis. Overdrill the wire with a cannulated drill and insert the pin.

Step 8: As long as the other pin clamp holes line up with the bone, use all three to fix the bone. All the pins should be parallel to each other.

Step 9: Make sure that the distal pin clamp is locked down to the fixator with red bolts (7 mm). Slide off the fixator from the pins.

Step 10: Make a 7 mm incision just proximal to the most proximal-distal pin. Drill the femur at this level with multiple drill holes. Complete the osteotomy using a sharp osteotome.

Step 11: Reapply the fixator to the pins. Make sure the osteotomy did not displace. If it did, adjust it in the frontal plane. If AP clamps are being used, drill at least one hole for posterolateral oblique pin proximally and distally. The total number of pins per segment should be 3 or 4 depending on the size of the femur and the thigh being lengthened.

Step 12: Drill a hole in the upper tibia from anterior to posterior using a 3.2 mm drill bit. Insert a pin into this hold. Make sure that the drill hole is distal to the tip of the proximal tibial apophysis.

Step 13: Connect the tibial pin to an arch mounted off of the tibial rail extension using a cube. Test the motion of the knee to confirm that the knee moves freely. By placing the first tibial pin in the sagittal plane, anteroposterior adjustment of the tibia in case of subluxation can be done. Apply a set screw to the pin once the knee motion is free. To confirm it is free, do the “drop test.” If the leg drops with no friction from 0 to 90°, then the hinge is perfect.

Step 14: Apply two more pins in different orientation in the proximal tibia.

Step 15: Inject Botox to the quadriceps muscles.

Step 16: Apply a knee extension bar constructed from Ilizarov parts. This includes a post proximally and a twisted hinge distally. Off of each of these, insert a post and fixate it with a 20 or 30 mm socket. Connect a threaded rod between the two posts. The knee extension bar should be placed in maximum extension of the knee and locked down so that the rod length cannot be changed. The patient uses the bar all night long and half the time during the day. It is easily removable for exercise.

Step 17: Apply a distractor.

Step 18: Apply sterile dressings.

Step 19: Obtain final radiographs before leaving the operating room.

Modification for valgus ± flexion deformity of the distal femur

Step 5a: Insert a wire into the distal femur using the MRS as a guide. This wire should be parallel to the Steinmann pin. Use a cannulated drill and then insert a half-pin.

Step 6a: Drill a wire into the proximal femur at about 7° to the shaft. This proximal wire should subtend a valgus angulation with the distal pin. It should be in the same rotational orientation as the distal pin.

Step 7a: Perform an osteotomy of the distal femur at the planned level leaving enough room to add two more pins distal to the osteotomy.

Step 8a: Correct the valgus deformity by making the pins parallel to each other in the frontal plane. Reapply the external fixator to the pins.

Step 9a: If there is also a flexion deformity of the knee, extend the knee using the tibia to lever the osteotomy into extension.

Step 10a: Add the remaining frontal plane pins.

Step 11a: Add the remaining AP clamp pins.

Step 12–19: same as before.

13.7.7 Rehabilitation and Follow-Up During Lengthening

Femoral lengthening requires close follow-up and intensive rehabilitation to identify problems and maintain a functional extremity. Clinically, the patient is assessed for hip and knee range of motion, nerve function, and pin site problems. Radiographically, the distraction gap length, regenerate bone quality, limb alignment, and joint location are assessed.

Knee flexion should be maintained as close to 90° as possible. The minimum goal is >45°. If knee flexion is ≤45°, the lengthening should be stopped or at least slowed and the knee rehabilitated more. If after a few days the knee flexion improves, lengthening may resume. It is critical to never sacrifice function for length. More length can be obtained with subsequent lengthenings, but we cannot recreate a new knee joint. Therefore, everything should be done to preserve the knee joint and its motion. 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 (FFD) 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 (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 can lead to hip subluxation and dislocation. Release of the adductors and the rectus and the tensor fascia lata during lengthening may need to be considered to allow further lengthening and to prevent hip subluxation.

Weight bearing (WB) is permitted all throughout the lengthening. In younger lighter children walking without support can be achieved immediately because the fixator is so stiff relative to the patient's weight. When the child is older and heavier, they prefer to walk with crutches or a walker until there is sufficient consolidation. We therefore permit WB as tolerated (WBAT) as long as they have normal proprioception.

13.7.8 Fixator Removal and Rodding of Femur (Figs. 13.7 and 13.8)

The fixator can be removed once the regenerate bone is fully healed radiographically. The regenerate bone should show no gaps (interzone closed) and evidence of corticalization on three or four cortices of the AP and lateral views.

The external fixator should be removed under anesthesia as an outpatient procedure. Remove all the middle pins and leave only the most distal and proximal pin in place. Using image intensifier radiography, first drill or ream a hole for a Rush rod and then insert a Rush rod into the femur from the greater trochanter to the distal femoral physis. The hooked tip of the rod should embed into the tip of the greater trochanter for ease of later removal.

13.8 Specific Complications and Their Treatment for Congenital Femoral Deficiency Lengthening

13.8.1 Nerve Injury

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 may show evidence of nerve injury but, in most cases since too many fibers are conducting normally, will likely be negative. Quantitative sensory testing using the Pressure Sensitive Sensory Device (PSSD), if available, is the most sensitive test to assess for nerve involvement (Nogueira et al. 2003). Near nerve conduction using very fine electrodes is also very accurate. If the nerve problem is identified early, it can be treated by slowing the rate of distraction. If despite slowing the distraction, 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 (Paley [AU18] 2005b; Nogueira and Paley 2011) found that peroneal nerve decompression is efficacious for the treatment of peroneal nerve injury secondary to lengthening.

13.8.2 Poor or Failure of Bone Formation

Hypotrophic regenerate formation requires slowing of the distraction rate. The rate can be slowed to ¾, ½, or ¼ mm per day. If the regenerate bone does not improve, a decision needs to be made as to whether lengthening may be continued knowing that the bone defect being created may need to be bone grafted. Biphosphonate infusion (e.g., zoledronic acid) can be used to prevent bone resorption while permitting bone formation. At the end of lengthening if the distraction gap does not fill, it can be bone grafted.

13.8.3 Incomplete Osteotomy and Premature Consolidation

Lack of separation of the osteotomy site after a week of distraction may be due to an incomplete osteotomy or at least a periosteal hinge that will not separate. Continued distraction can lead to an acute separation of the bone ends. There may be an audible pop associated with this. Such an acute separation is usually very painful. This pain continues unabated until the bone is acutely shortened by a few millimeters. It is important to advise the patient of this. If the bone does not separate or if the patient or parents wish to avoid a painful separation, a reosteotomy should be performed.

13.8.4 Hip Subluxation/Dislocation

Hip instability is judged radiographically. A break in Shenton's line or increased medial-lateral, head-teardrop distance indicates subluxation of the hip. Although the diagnosis is radiographic, the hip usually has an adduction and flexion contracture and may also exhibit stiffness to flexion-extension. Hip subluxation does not usually occur if there is adequate coverage of the femoral head. However, adduction and flexion contracture predisposes the hip to subluxation even in the presence of adequate coverage.

If hip subluxation occurs during lengthening or consolidation phases, the patient should urgently be taken to the operating room for soft tissue releases of the adductor longus and gracilis and in some cases the tensor fascia lata (assuming the rectus femoris has already been lengthened). A closed reduction of the hip should be performed by abducting the hip. If the hip will not reduce, the distraction gap should be shortened to loosen the hip joint. The external fixator should be extended to the pelvis with or without a flexion-extension hinge. With a hinge a hip extension bar should be added to prevent flexion contracture. The pelvic fixation consists of at least two anterolateral pins between the two tables and between the anterior inferior and superior spines and two pins more lateral. The femur should be in 15–20° of abduction to the pelvis to maintain the reduction.

13.8.5 Knee Subluxation/Dislocation

CFD knees usually have hypoplastic or absent cruciate ligaments. In the knee, posterior or anterior subluxation can be monitored on the lateral view full knee extension radiograph (Paley 2005c). Posterior subluxation is the most common, although in most cases, this is combined with an external rotation subluxation (posterolateral rotatory subluxation). If the fascia lata and biceps are intact, tension in these structures is the culprit.

Limb length equalization should be based on full length standing radiographs. Limb alignment is assessed for the femur and tibia separately and in combination. Separately, the joint orientation of the knee should be measured using the malalignment test (Paley 2005c, d). 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. The most common location is lateral. Dynamization of the fixator should be carried out and bone growth stimulators (our preference is the Exogen) can be used. 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.

13.8.6 Fracture

Fractures associated with limb lengthening can be divided into those that occur while the external fixator is still on, those that occur at the time of removal, and those that occur after removal. Fractures can also be classified as to their location: regenerate or remote. The incidence of all these types of fractures associated with CFD lengthening was 34 % compared to 9 % for noncongenital femoral lengthening. In many cases this was despite the use of a spica cast after removal.

Since this rate was unacceptably high, we started prophylactically Rush rodding the femur at the time of removal. This new protocol virtually eliminated the complication of refracture after lengthening.

13.8.7 Hip and Knee Joint Luxation

Hip joint subluxation or dislocation is a dreaded complication associated specifically with congenital femoral lengthenings. The hip joint is often mildly or moderately dysplastic in patients with CFD. The acetabular dysplasia has a different pattern than that of DDH. The femoral head is usually uncovered laterally rather than anterolaterally. If the CE angle is less than 20°, the hip joint is considered at risk for dislocation (Suzuki et al. 1994). The other parameter to look at is the orientation of the sourcil (dome). The sourcil should be horizontal. If it is inclined laterally, then the hip joint is potentially unstable even if the CE angle is greater than 20. It is always safer to err on the side of performing a pelvic osteotomy than to end up with a hip luxation.

During lengthening the hip abduction and flexion range of motion should be checked. Hip luxation always occurs together with adduction or flexion contractures of the hip. Hip luxation is diagnosed radiographically. The earliest signs are widening of the medial head teardrop distance and a break in Shenton's line.

Once luxation is diagnosed, distraction must stop. If it is early in the lengthening process, the patient can be taken to the OR, the hip reduced acutely, and the external fixator extended to the pelvis to stabilize the hip. Lengthening can proceed under such protection. If this occurs after at least 5 cm of lengthening, it is wiser to stop the lengthening completely and reduce the luxation by means of an abduction cast or fixator. Adductor, flexor release may be required including the proximal tensor fascia lata.

13.8.7.1 Knee Luxation

As previously noted, most cases of CFD have hypoplastic or absent cruciate ligaments. Most patients have a more dominant instability pattern (e.g., posterior vs. anterior vs. posterolateral). The tendency towards flexion contracture during lengthening predisposes the tibia to posterior subluxation. If the fascia lata remains intact the knee will sublux posterolaterally. Extension contracture leads to patella alta and anterior or anteromedial subluxation. Knee subluxation depends on the level of the lengthening. Proximal femoral lengthening is less likely to sublux the knee while distal femoral lengthening is more likely to sublux the knee. Proximal femoral lengthening has a narrower and often less well-formed regenerate bone with a much higher risk of fracture than distal femoral lengthening. Prevention of knee subluxation involves a combination of soft tissue release of the distal fascia lata together with extension of the external fixator to the tibia. The latter is best done with [AU19] hinges to permit and maintain knee motion. One of the problems is that if the tibia is left unfixed with distal femoral lengthening without release of the fascia lata, the knee will begin to sublux after only 2–3 cm. If fascia lata release is performed without fixation to the tibia, the knee will start to sublux after about 4 cm. To permit greater lengthening and to protect the knee from pressure and subluxation, it is best to articulate across the knee. This protects the cartilage of the knee and physis from undue forces. In the past extending the frame across the knee with hinges was considered something that could only be done with circular external fixation. With the modularity of monolateral external fixators, the same can be achieved with monolateral external fixators. It is important to preserve the principal of articulated fixation to the tibia with all lengthenings of congenitally short femurs of 5 cm or more whether one prefers a monolateral or circular fixator. The biggest mistake is to become a slave to the fixator rather than the fixator becoming a slave to the surgeon. I have seen so many knee subluxations and dislocations secondary to lengthening without soft tissue releases and without protection of the knee by means of articulated distraction. One approach that has been used to protect the knee is to keep the knee in full extension without bending throughout the lengthening. While this may avoid subluxation in most cases, it may also result in a stiff knee.

13.8.8 Joint Stiffness and Contracture

The tendency is for the knee to lose flexion during lengthening. This is combined with the tendency for a flexion contracture. These problems can occur together or separately. Although the transfixing pins or wires may contribute to difficulty in flexion, lengthening with internal distractors (e.g., ISKD) still leads to loss of knee flexion with increased length. Stiffness of the knee is preventable. Surgical release/lengthening of specific soft tissues (fascia lata, rectus femoris) reduces the joint reactive forces on the knee due to lengthening. Physical therapy is essential to successful CFD lengthening. This is especially true for the knee. I am not prepared to lengthen most CFD cases if they cannot organize outpatient PT.

If the knee gets stiff to flexion despite adequate rehab, then a quadricepsplasty should be performed. When there is a concomitant flexion contracture, I will choose either open or closed treatment: Closed requires an external fixator and gradual distraction. Open means posterior capsule release. Physical therapy including dynamic splinting can be used first to obtain extension of the knee before considering any type of quadricepsplasty.

Joint stiffness due to damage to the articular surface occurs due to unprotected compressive forces across the knee joint combined with immobility and subluxation. This irreversible complication can be prevented by means of distraction across the knee joint combined with protection against subluxation and maintenance of knee joint range of motion.

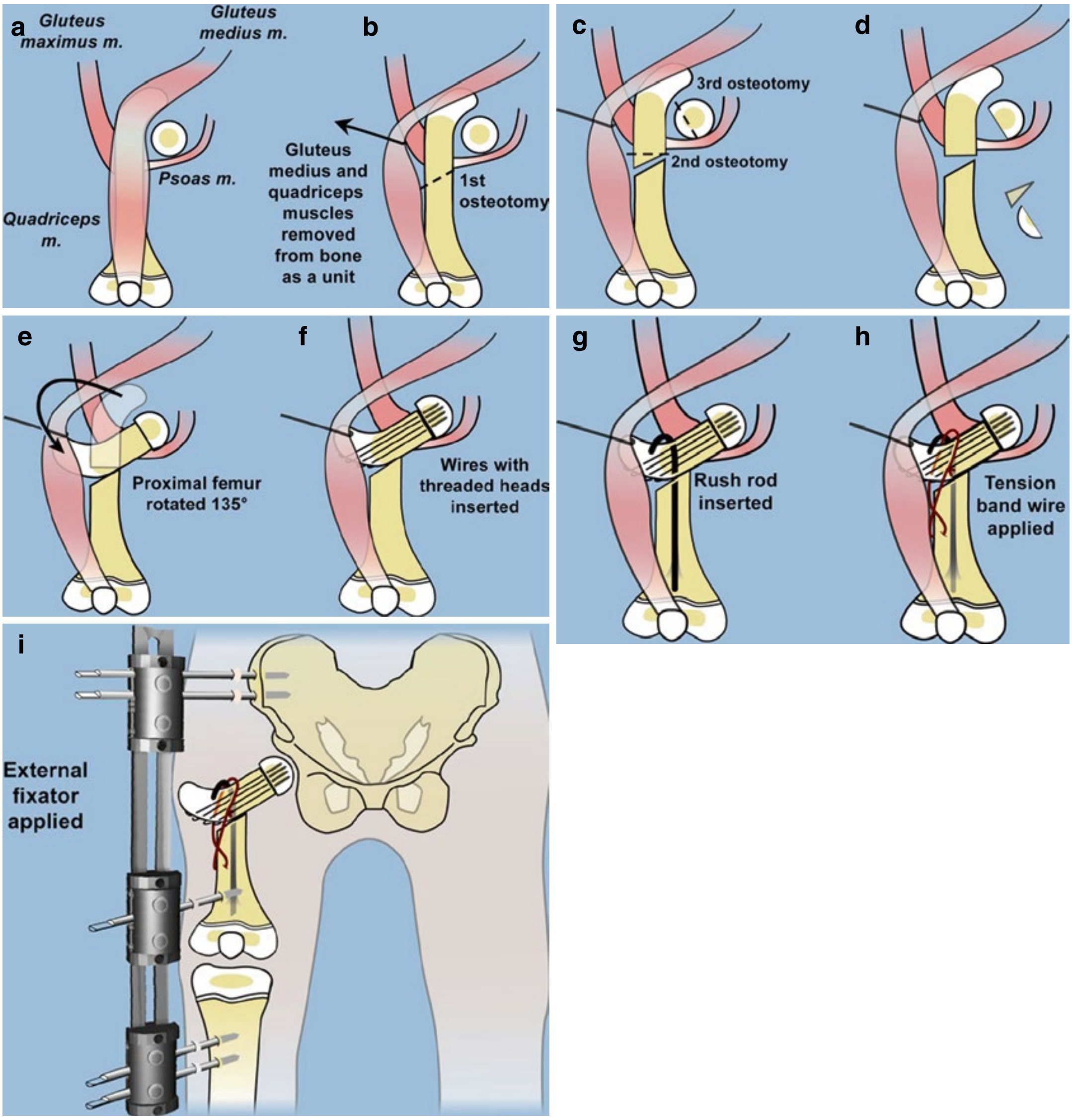
13.9 Treatment CFD Types 2 and 3

13.9.1 Treatment CFD Type 2a

Type 2a differs from type 2b by the presence of a mobile femoral head. If there is a mobile femoral head, an attempt should be made to obtain union between the femoral head and upper femur. Attempts at connecting these together are often met with failure or stiffness of the hip. I have developed a new way of reliably achieving union between the upper femur and the femoral head with the creation of a new femoral neck.

13.9.1.1 Superhip 2 Procedure for Treatment of a Mobile Proximal Femoral Pseudarthrosis (Figs. 13.9 and 13.10)

The same approach as the superhip is used. The operation remains the same until the subtrochanteric osteotomy. Before performing



[AU20] **Fig. 13.9** Superhip 2 surgical technique illustrations

the osteotomy, the region of the femoral neck is dissected. The level of the acetabulum is identified with the image intensifier and the femoral head and acetabulum are identified. The capsule of the femoral head is opened. The femoral head is moved in the acetabulum using a needle as a joystick through its cartilage. If there is a cartilaginous femoral neck, it is excised down to the ossific nucleus of the femoral head. If there is no neck, the cartilage of the head is cut back to the ossific nucleus to expose the bone. The femoral neck is made from the proximal femur and greater trochanter segment. This segment is rotated 135° on a soft tissue pedicle so that the greater trochanter goes distal and lateral and the distal cut end of the subtrochanteric osteotomy is fixed to the ossific nucleus of the femoral head. The subtrochanteric osteotomy is made by taking a trapezoidal segment of femur out. The distal cut is at 45° to the shaft of the femur, while the proximal cut is perpendicular. All of the soft tissue is dissected off the trochanteric segment

Fig. 13.10 X-ray (**a**) and MRI (**b**) of a girl with CFD Paley type 2a with a true pseudarthrosis of the femoral neck. X-ray after superhip 2 procedure with external fixator in place (**c**). The femoral neck is reconstructed and intact after removal of the external fixator (**d**). X-ray (**e**) after first lengthening surgery (6 cm). Second lengthening surgery (**f**). After (**g**) second lengthening surgery (8 cm). Two more lengthening surgeries are needed

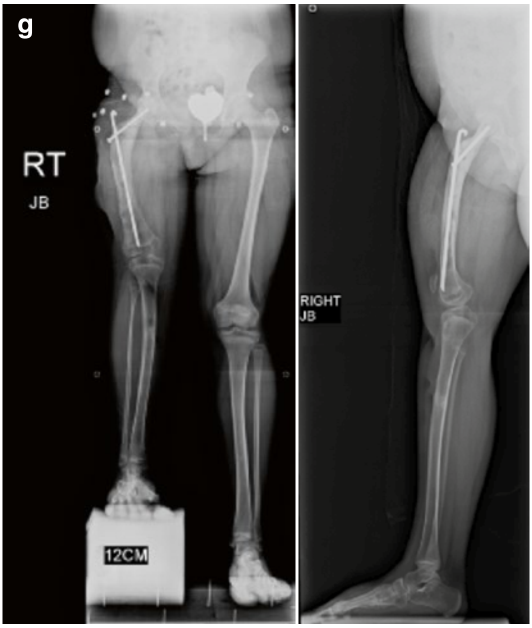
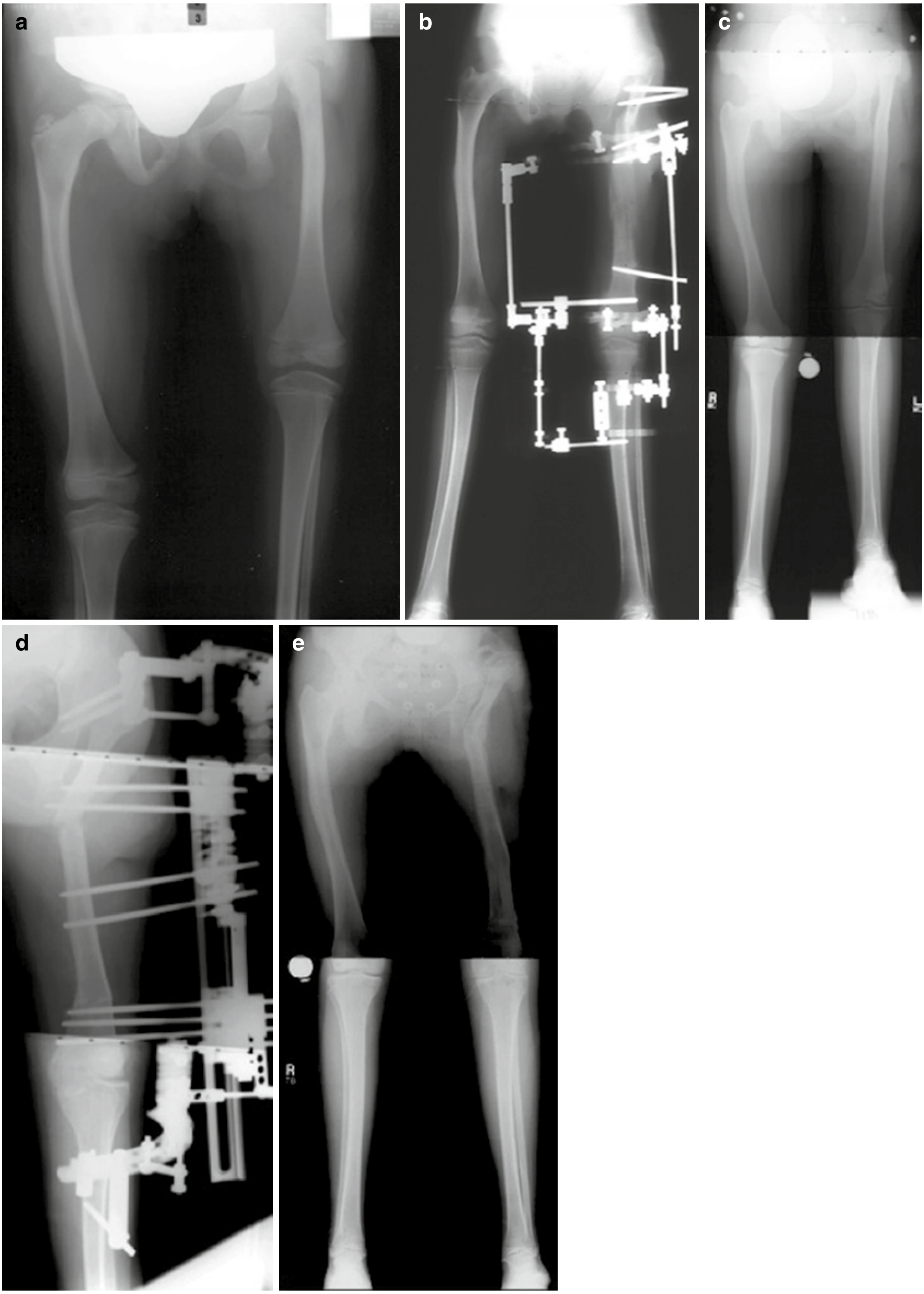


Fig. 13.10 (continued)

except the gluteus maximus and the psoas tendon. Care should be taken next to the psoas tendon not to injure the profunda artery and other perforators at its inferior edge. The trochanteric segment is predrilled with a 1.8 mm wire in four places around its periphery. This bone is also drilled at 45° to its long axis for insertion of a Rush rod. This is drilled in a way to miss the four wire holes path. The distal femur diaphysis is also drilled with a 3.2 mm drill bit. The lateral cortical surface of the trochanteric segment which will be oriented inferiorly is slightly decorticated. The Rush rod is now fixed in place with the distal femur articulating against the decorticated surface. A tension band wire is applied similar to the one described for the superhip. This new femoral neck which now subtends an angle of 135° to the femoral shaft is brought up in line with the femoral head. The cannulated wire is inserted into the femoral head and drilled with the 3.2 cannulated drill bit. An appropriately sized partially threaded 4.5 or 5.5 mm screw with washer is inserted to compress the femoral neck-head site. This osteosynthesis is stable for gentle abduction and adduction but cannot control flexion and extension since the single screw acts as a hinge for flexion and extension. An additional threaded k wire is inserted parallel to the screw into the femoral head. To neutralize the forces on the femoral neck-head junction, an external fixator is applied from the pelvis to the tibia. The external fixator is applied before closure to ensure that there is no loss of fixation. Two anterior external fixation pins are put into the pelvis between the anterior inferior and superior spines in an oblique anteroposterior direction following the orientation of the ilium. The tibial pins are also anteroposterior. A pediatric Orthofix LRS with cubes as needed to get fixation of the pelvic pins is applied for neutralization. Prior to closure any residual bone graft can be morselized around the osteotomy site. The abductor-quadriceps muscle tendon unit followed by the tensor fascia lata is sutured to the greater trochanter. The rest of the closure is per routine. It is important to use suction drains to prevent a hematoma under the anterior flap. The fixator is removed after 3 months. Physical therapy for the hip joint can begin at that time.

13.9.2 Treatment CFD Type 2b

In this pathology, there is either a hypoplastic femoral head which is fused to the acetabulum (Fig. 13.11). If the fusion region is small (usually posteroinferior), it can be broken and converted and a superhip 2 performed. If the fusion area is large, the superhip 2 is not an option. What distinguishes this from the diaphyseal deficiency of type 3 is the cartilaginous cap of the greater trochanter which is present in type 2b. To reconstruct the hip without directly joining the proximal femur to the femoral head requires a pelvic support osteotomy. The treatment of type 2b cases is accomplished by combining a pelvic support osteotomy proximally with a distal femoral lengthening and realignment osteotomy (Fig. 13.8). This combination is called the *Ilizarov hip reconstruction*. Pelvic support osteotomy is usually enough to prevent proximal migration of the femur during lengthening for noncongenital pathologies (Rozbruch et al.



[AU21] **Fig. 13.11** X-ray (**a**) of 10-year-old girl with Paley type 2c CFD (absent femoral head). She was treated by pelvic support osteotomy (**b**) and distal femoral lengthening (13 cm). The pelvic support angulation remodeled and the leg length difference was 10 cm at skeletal maturity (**c**). At age 16 she underwent a second pelvic support osteotomy with 10 cm lengthening (**d**). Final radiographic result showing limb length equalization. She has excellent function

2005). For congenital pathologies, the fixation should be extended to the pelvis to prevent proximal migration of the femur.

In young children with very short femora, the femur may be too small to perform both the pelvic support and the distal lengthening osteotomies. Furthermore, the valgus component of the pelvic support osteotomy will remodel straight within a year in most cases. In such cases, pins are extended to the pelvis to prevent proximal migration during lengthening. 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 X-ray. The amount of valgus is equal to the total amount of adduction of the hip plus 15° 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 FFD. 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 mid-proximal segment of femur. The distal osteotomy is usually mid-diaphyseal. The external fixator must still be extended to the tibia with hinges, as previously discussed.

13.9.3 Treatment Type 3a: Diaphyseal Deficiency, Knee ROM >45°

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° of motion (type 3a), usually with a 45° knee flexion deformity, whereas others have a stiff knee with <45° range of motion (type 3b) and usually a greater flexion deformity. The most predictable and reliable treatment option in these cases remains prosthetic reconstruction surgery (PRS) (e.g., 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.

13.9.3.1 LRS Approach for Diaphyseal Deficiency Cases: Type 3a

LRS is most applicable to type 3a cases in which there is a functional range of knee motion present (Fig. 13.12). In type 3 cases, a knee flexion deformity is usually present. In addition, a hip flexion contracture is 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 from the anterior inferior iliac spine oriented towards 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 fixation is kept in place for 6 weeks to maintain knee and hip extension. Range of motion exercises are then [AU22]

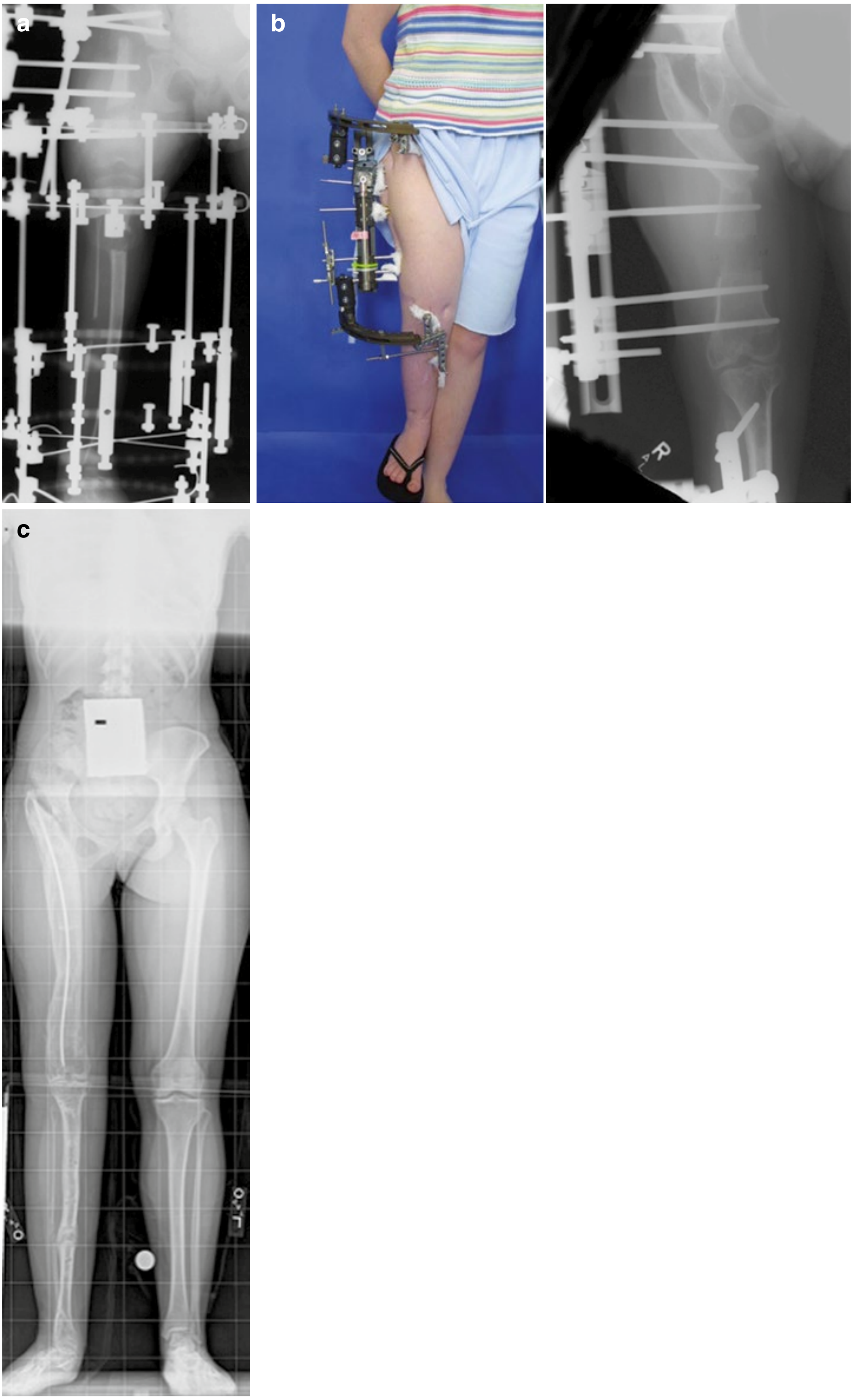


Fig. 13.12 CFD Paley type 3A in a patient whose parents refused a rotationplasty. She was treated at age 7 with a lengthening of the femur and tibia for a total of 12.5 cm (**a**). At age 10 she underwent the same treatment again and achieved another 12.5 cm of lengthening. At age 14 when she was skeletally mature, she underwent a pelvic support osteotomy together with a 10 cm femur lengthening (**b**). At age 18 she underwent her fourth femoral plus lengthening of 12.5 cm. She had a final tibial lengthening of 2.5 cm to correct the deformity and equalize her lengths (**c**). She has excellent clinical function

begun to regain the knee motion in its new more functional arc (arc extending to full knee extension).

After the preparatory work described above, the femur is lengthened. The external fixator is placed from the pelvis to the femur to the tibia. The femur is lengthened up to 8 cm. The knee joint is distracted, but since no knee or hip hinges are used when the femur is so short, there is little need for physical therapy during this first lengthening. The goal of this first lengthening is to convert a type 3a femur to a type 2b femur. The rest of the treatment is for type 2b, with serial lengthenings and finally a pelvic support osteotomy combined with the final lengthening. Since the predicted discrepancy ranges between 30 and 40 cm, at least four lengthenings and an epiphysiodesis are required to equalize limb lengths.

13.9.3.2 Prosthetic Reconstructive Surgery Approach to CFD with Diaphyseal Deficiency: Type 3

Prosthetic fitting is possible without any surgery. The difficulties with this approach are the FFD 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 residual limb that can more easily be fitted. 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 limp and stabilize the hip joint.

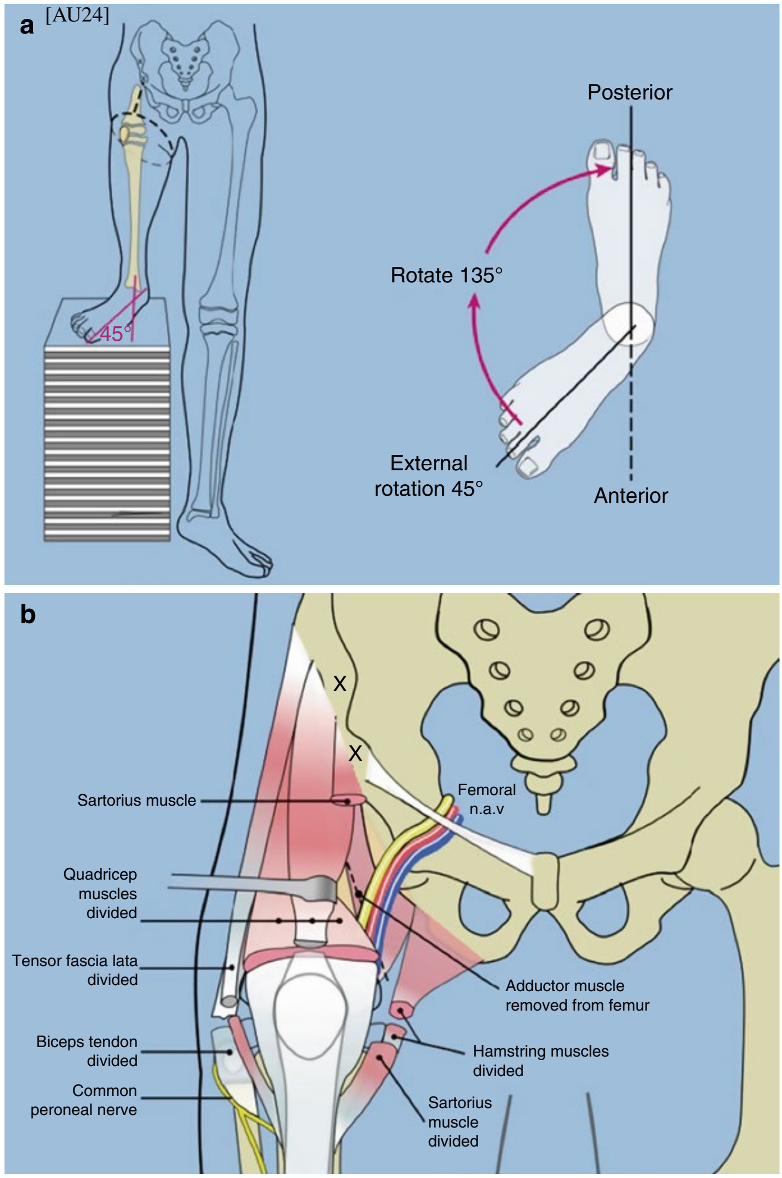
[AU23] The other approach to PRS is with rotationplasty (Alman et al. 1995; Torode and Gillespie 1983) popularized the Van Nes rotationplasty for patients with CFD. They used a long oblique incision. The goal was to fuse the residual knee and rotate the limb 180° so that the foot is pointed backward 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 (2001) modified the rotationplasty approach using a racquet-like incision, performing the rotation between the femoral remnant and the pelvis. He then fused the femur to the lateral aspect of the ilium, thus converting the knee into a hip joint and the ankle into a knee joint. This provided improved hip stability over that provided by the Van Nes rotationplasty. One problem with the Brown method is excessive shortening of the hip muscles and lateralization of the hip and lower limb.

I modified the Brown technique by combining it with a Chiari osteotomy of the pelvis and fusing the femoral remnant to the cancellous roof of ilium (Figs. 13.13 and 13.14). I also don't remove any of the detached muscles and I take care to transfer all of them as distal as possible. I refer to this procedure as the Paley-Brown rotationplasty to distinguish it from the original published version. 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 (as much as 90°), release the posterior. Decompress the peroneal nerve to prevent injury and allow greater rotation. Because the tibia in these patients typically has an internal rotation deformity, a supramalleolar osteotomy should also be performed to derotate the tibia and fibula. Although epiphysiodesis can be done at a second surgery, I prefer to achieve this with the same screws used for fixation of the femur to the pelvis (fig). The modified Brown rotationplasty provides very functional results with better hip function and stability than does the Van Nes rotationplasty.

13.10 Age Strategies

The majority of type 1 CFD cases 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 at the optimal age 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

[AU24] **Fig. 13.13** Paley modified Brown rotationplasty surgical technique



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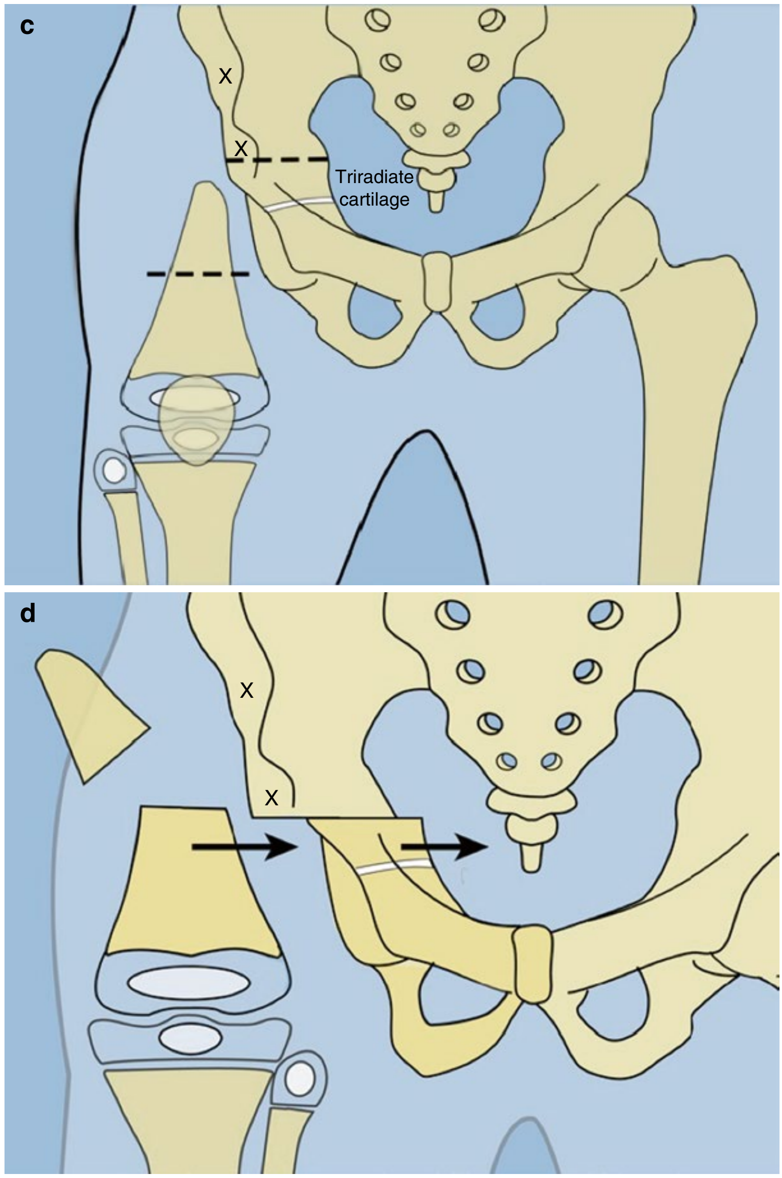

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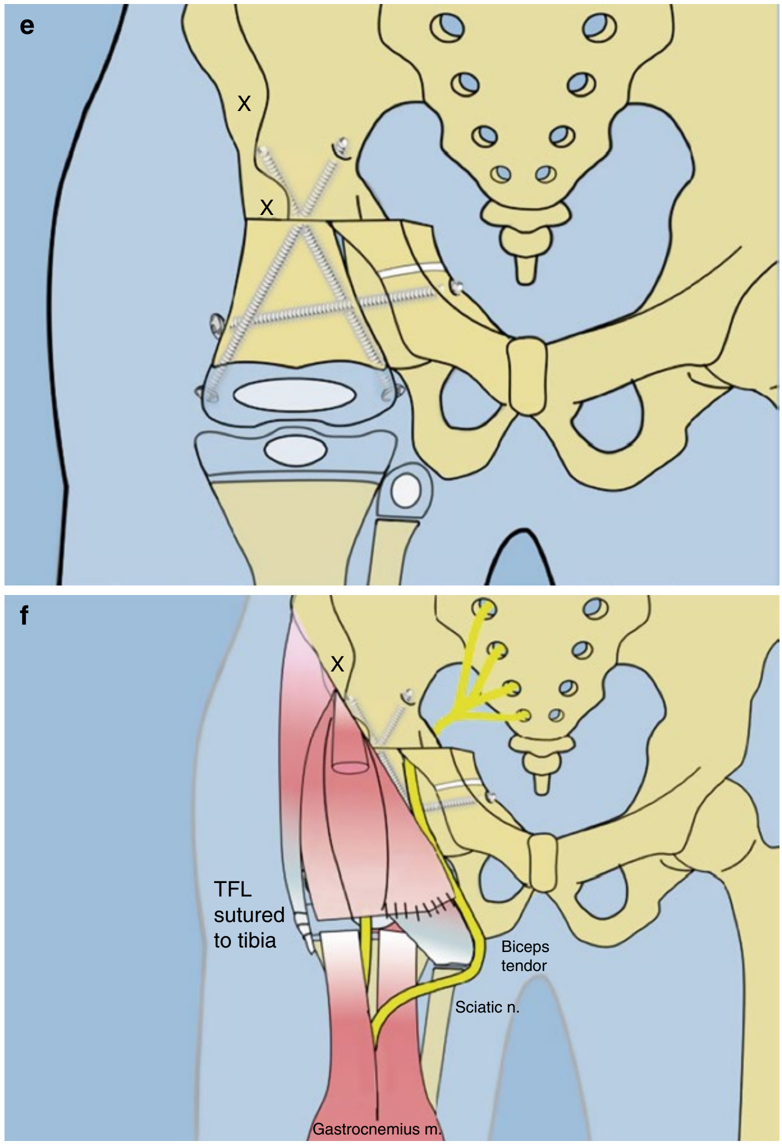

cognitive level accepts everything their parents decide without questioning it. Children at this age group seem to understand too little and too much at the same time. They don't connect their recognition that they have a short leg with the solution of limb lengthening. Beyond the age of 6 or 7 years, the child enters the age of reason and begins to understand that he or she is different from other children and that he or she has a problem for which there is a solution. They learn to accept the solution by reason rather than by faith. Their cooperation is voluntary rather than coerced. The amount of lengthening that can be performed in the femur at any one stage is usually between 5 and 8 cm. This lengthening amount seems to be independent on the initial length of the femur and age of the child. Generally, 5–8 cm can be performed safely in toddler (age 2–4 years), as well as in older children and adults. Combined femoral and tibial lengthenings allow greater total lengthening amounts. Tibial lengthening ≤5 cm can be combined with a 5 cm

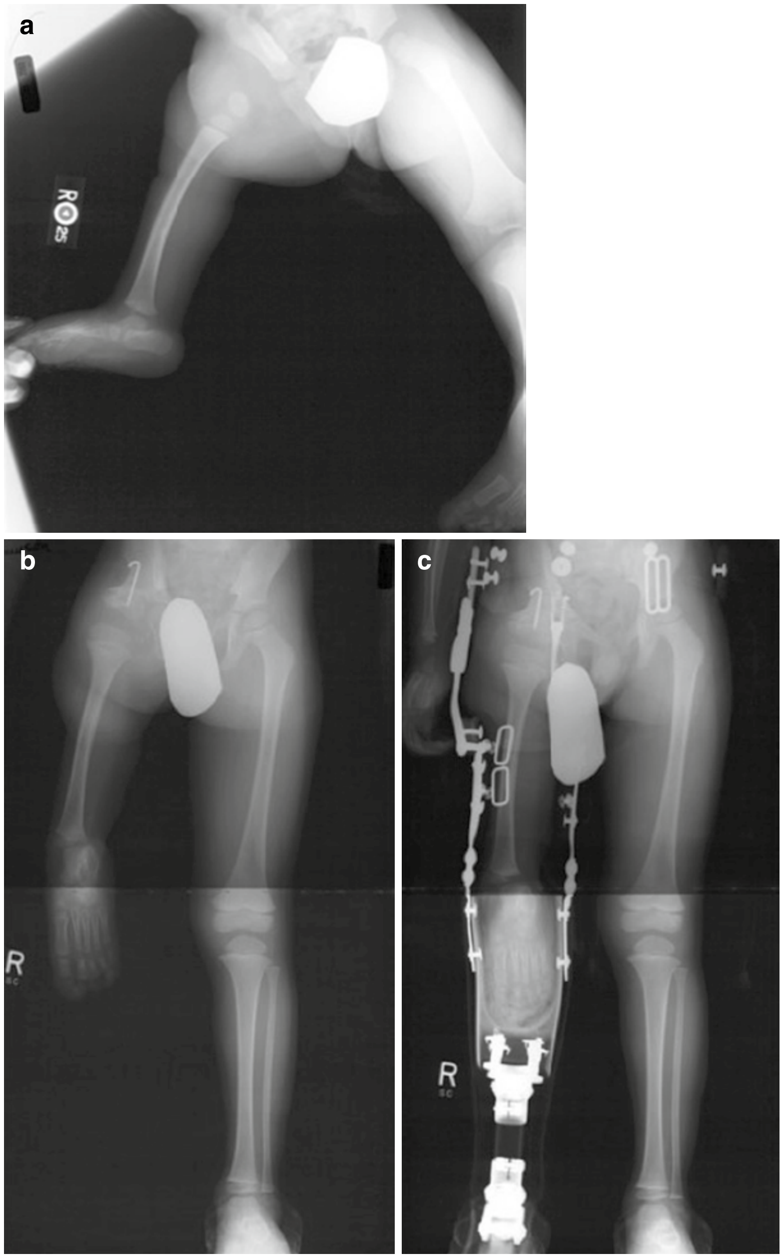

Fig. 13.14 X-ray of Paley type 3b CFD with fibular hemimelia (**a**). X-ray after rotationplasty age 4 (**b**) and age 10 (**c**)

femoral lengthening. 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 need. This means going from a hip-knee-ankle-foot orthosis 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.

We have also lengthened adults (age 15–60 years) with CFD whose parents refused PRS for them when they were children. We were able to successfully equalize their leg lengths with one or two lengthenings, depending on the discrepancy (the most severe case underwent 25 cm of equalization with two LON treatments) (Fig. 13.11). Therefore, adult CFD residua are not contraindications to treatment.

The frequency of lengthening should be spread out to no less than every 3 years and preferably every 4 years. The strategy of rule of 4 is a good guiding strategy. Assuming that a preparatory surgery is done between ages 2 and 3 years, the first lengthening can be done between ages 3 and 4 years. The second lengthening would occur around age 8 years and the final third lengthening around age 12 years. For psychosocial reasons it is preferable to complete all the lengthenings by age 14 when the child starts high school education. If a forth lengthening is required, it is done around age 16 years.

13.10.1 Role of Epiphysiodesis and Hemiepiphysiodesis

Epiphysiodesis is used as an adjuvant method to equalize limb length discrepancy. It should be calculated into the total strategy of equalization surgeries. Epiphysiodesis should be used for up to 5 cm of LLD equalization. Judicious use in some cases may avoid the need for one lengthening (e.g., predicted LLD = 12 cm; plan 7 cm lengthening before age 4 and 5 cm epiphysiodesis around puberty). Calculation of the timing of epiphysiodesis can be achieved quickly and accurately using the multiplier method (Paley et al. 2000).

Hemiepiphysiodesis is very useful to correct the valgus deformity of the knee from distal femoral or proximal tibial origins. I prefer to use the epiphysiodesis plate devices the concept of which was developed by Peter Stevens (Burghardt et al. 2008). Correction of the valgus deformity of the femur permits implantable lengthening of the femur since there is no angular deformity.

[AU25] References

Alman BA, Krajbich JI, Hubbard S (1995) Proximal femoral focal deficiency: results of rotationplasty and Syme amputation. J Bone Joint Surg Am 77: 1876–1882

Amirault JD, Cameron JC, MacIntosh DL, Marks P (1988) Chronic anterior cruciate ligament deficiency: long-term results of MacIntosh's lateral substitution reconstruction. J Bone Joint Surg Br 70:622–624

Brown KL (2001) Resection, rotationplasty, and femoropelvic arthrodesis in severe congenital femoral deficiency: a report of the surgical technique and three cases. J Bone Joint Surg Am 83:78–85

Burghardt RD, Herzenberg JE, Standard SC, Paley D (2008) Temporary hemiepiphyseal arrest using a screw and plate device to treat knee and ankle deformities in children: a preliminary report. J Child Orthop 2(3):187–197

Grammont PM, Latune D, Lammaire IP (1985) 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

Grudziak JS, Ward WT (2001) Dega osteotomy for the treatment of congenital dysplasia of the hip. J Bone Joint Surg Am 83-A(6):845–854

Krackow KA, Thomas SC, Jones LC (1988) Ligament-tendon fixation: analysis of a new stitch and comparison with standard techniques. Orthopedics 11(6): 909–917

Langenskiöld A, Ritsila V (1992) Congenital dislocation of the patella and its operative treatment. J Pediatr Orthop 12:315–323

Manner HM, Radler C, Ganger R, Grill F (2006) Dysplasia of the cruciate ligaments: radiographic assessment and classification. J Bone Joint Surg Am 88(1):130–137

Millis MB, Hall JE (1979) Transiliac lengthening of the lower extremity. A modified innominate osteotomy for the treatment of postural imbalance. J Bone Joint Surg 61A:1182

Nogueira MP, Paley D (2011) Prophylactic and therapeutic peroneal nerve decompression for deformity correction and lengthening. Oper Tech Orthop 21:180–183

Nogueira MP, Paley D, Bhave A, Herbert A, Nocente C, Herzenberg JE (2003) Nerve lesions associated with limb-lengthening. J Bone Joint Surg Am 85-A(8):1502–1510

Paley D (1990) Problems, obstacles, and complications of limb lengthening by the Ilizarov technique. Clin Orthop Relat Res (250):81–104. Review

Paley D (1998) Lengthening reconstruction surgery for congenital femoral deficiency. In: Herring JA, Birch JG (eds) The child with a limb deficiency. AAOS, Rosemont, pp 113–132

Paley D (2005a) Chapter 12. Six -axis deformity analysis and correction. In: Principles of deformity correction, 1st edn. 2002. Corr. 3rd printing 2005. Springer, pp 411–436 [AU26]

Paley D (2005b) Chapter 10. Lengthen consideration: gradual versus acute correction of deformities. In: Principles of deformity correction, 1st ed. 2002. Corr. 3rd printing 2005. Springer, pp 269–289

Paley D (2005c) Chapter 3. Radiographic assessment of lower limb deformities. In: Principles of deformity correction, 1st edn. 2002. Corr. 3rd printing 2005. Springer, pp 31–60

Paley D (2005d) Chapter 2. Malalignment and malorientation in the frontal plane. In: Principles of deformity correction, 1st edn. 2002. Corr. 3rd printing 2005. Springer, pp 19–30

Paley D, Bhave A, Herzenberg JE, Bowen JR (2000) Multiplier method for predicting limb-length discrepancy. J Bone Joint Surg Am 82-A(10):1432–1446

Pappas AM (1983) Congenital abnormalities of the femur and related lower extremity malformations: classification and treatment. J Pediatr Orthop 3(1):45–60

Rozbruch SR, Paley D, Bhave A, Herzenberg JE (2005) Ilizarov hip reconstruction for the late sequelae of infantile hip infection. J Bone Joint Surg Am 87(5):1007–1018

Salter RB (1978) The classic. Innominate osteotomy in the treatment of congenital dislocation and subluxation of the hip by Robert B. Salter, J. Bone Joint Surg. (Brit) 43B:3:518, 1961. Clin Orthop Relat Res (137):2–14

Sanpera I Jr, Sparks LT (1994) Proximal femoral focal deficiency: does a radiologic classification exist? J Pediatr Orthop 14:34–38

Suzuki S, Kasahara Y, Seto Y, Futami T, Furukawa K, Nishino Y (1994) Dislocation and subluxation during femoral lengthening. J Pediatr Orthop 14:343–346

Torode IP, Gillespie R (1983) Rotationplasty of the lower limb for congenital defects of the femur. J Bone Joint Surg Br 65:569–573

Author Queries

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Queries	Details Required	Author's Response
AU1	Please provide department name for Dror Paley. Also Provide complete affiliation details for Fran Guardo.	
AU2	Please confirm if the identified head levels are correct.	
AU3	Please check sentence starting "Long lateral radiograph…" for clarity.	
AU4	Please check if edit to sentence starting "At the hip…" is okay.	
AU5	Closing parenthesis has been deleted. Please check if okay.	
AU6	Please check if edit to sentence starting "As a rule…" is okay.	
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AU8	Please provide caption for part figures "a–z, za–zh" in Fig. 13.3.	
AU9	Please provide caption for part figures "a–i" in Fig. 13.4.	
AU10	Please check the term "para tendonous" for correctness.	
AU11	Please provide closing parenthesis for sentence starting "To ensure that…"	
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AU16	Please check sentence starting "During lengthening, if…" for completeness.	
AU17	Please check sentence starting "The fixator bar…" for clarity.	
AU18	Please check the statement "… found that peroneal…" for completeness.	
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