Congenital Femoral Deficiency Reconstruction and Lengthening Surgery

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Introduction

Demographics

The often-cited incidence of congenital femoral deficiency (CFD) is 1 in 52,029 based on a review of the Edinburgh Register of the Newborn by Rogala et al. published in 1974 [1]. In retrospect, this may not be accurate, as the incidence is based on the only case identified during the 4.5-year collection period from 1964 to 1968, notably, after thalidomide use was discontinued. This survey is also representative of a relatively homogenous population, of which no information is given regarding race, age, environmental exposure, medication usage, or socioeconomic status of the population.

Embryology

Fetal growth and development of the lower extremities are controlled by a complex cascade of a multitude of growth factors that are expressed in a particular sequence and at various concentrations during development. In the process of limb patterning, mesenchymal cells in the limb bud integrate positional information from the three axes [2, 3].

Embryogenesis of the extremities occurs between 4 and 8 weeks after fertilization, and the majority of congenital anomalies occur during this period of time. Limb bud development begins with the lateral migration of two layers of mesoderm and outgrowth into the overlying ectoderm. Cells from the underlying somitic mesoderm ultimately form the muscle tissue of the limb, while cells from the lateral plate mesoderm form cartilage and bones. Distinct but coordinated molecular pathways primarily control each axis. In the process of limb patterning, mesenchymal cells in the limb bud integrate positional information from the three axes, indicating a complex interplay of the responsible factors [4, 5].

Development of the proximal-distal axis is at least partially controlled by fibroblast growth factors (FGF) secreted by the apical ectodermal ridge (AER) [4]. The AER is a thickened layer of the ectoderm that forms over the distal edge of the limb bud. FGF secreted by the AER stimulates proximal-distal growth of the limb via differentiation of the underlying mesoderm. As part of a complex positive feedback loop, the signal to produce FGF is supplied by the underlying mesoderm. The AER signaling center is responsible for the differentiation of the underlying mesoderm and the development of the limb in the proximal-to-distal direction. Removal of the AER results in arrest of limb outgrowth. Furthermore, ectopic implantation of the AER results in formation of an extra limb. The AER also contributes to interdigital necrosis, allowing separation of the initially webbed hand [6]. Defects in the AER lead to anomalies such as limb truncation, transverse deficiencies, and syndactyly [7]. The genetics of the apical ectodermal ridge have been further elucidated as a complex interplay of related genes. Five paralogous gene families, named Hox 9 through Hox 13, from Hox A and Hox D gene groups, come together co-linearly (HoxA-9 to HoxA-13 and HoxD-9 to HoxD-13) and play important roles in the AER.

Development in the anteroposterior axis is patterned by secretion of sonic hedgehog (SHH) from the zone of polarizing activity (ZPA), a collection of cells along the posterior

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aspect of the limb bud. Transplantation of the ZPA from the posterior aspect to the anterior aspect of the limb bud causes creation of a mirrored duplication of the ulnar aspect of the hand. In addition to its primary role in development of the anterior-posterior axis, the ZPA also contributes to maintenance of proximal-to-distal limb development and participates in the feedback loop of the AER [4]. Mutations in Indian hedgehog, SHH, and PITX1 in humans have been implicated in lower extremity polydactyly, while mice studies have shown an interaction between the expression of each factor [2, 8].

Development in the dorsal-ventral axis is regulated by the Wingless-type (Wnt) signaling pathway within the dorsal ectoderm [3]. The Wnt pathway induces the underlying mesoderm to develop dorsal characteristics and is blocked in the ventral ectoderm, allowing the development of ventral characteristics. In mice, inactivation of the Wnt signaling pathway results in the development of biventral limbs. The Wnt pathway also contributes to regulation of SHH, reflecting the complex interaction and coordination among the 3-dimensional pathways responsible for limb development [4, 6].

It is unlikely that CFD is caused solely by a defect in the overall function of the AER, as the distal portion of an extremity affected by CFD is often normal. It is more likely that the defect is in the underlying mesodermal layer, within the complex interplay of developmental proteins at a point more downstream in the differentiation of the limb.

Pathophysiology and Genetics

A single underlying genetic cause for CFD has not been elucidated. In the senior author's personal experience (over 1,000 patients with unilateral CFD) there was only one unilateral case that had a parent with unilateral CFD. In contrast, in a smaller group of multi-limb congenital deficiencies including CFD on at least one limb, it was not uncommon to find a history of a first- or second-degree relative with a congenital limb anomaly. Several authors have postulated a genetic cause given that the presence of the condition at birth, its association with other abnormalities that have known genetic conditions, occasional bilateral involvement, and reports of familial cases are supporting evidence for an underlying inheritable genetic defect with incomplete penetration or an autosomal recessive pattern of inheritance [9–11]. It should be noted that phenotype has been used to diagnose and classify patients with CFD, despite quite varied clinical and radiographic presentations. As genetic and molecular understanding of the disease progresses, it is likely that grouping all patients into a single diagnosis of CFD will not be sufficient. Some of the reported abnormalities associated with CFD are fibular hemimelia, femoral-fibular-ulnar complex, patella aplasia/hypoplasia, absence of the fourth or fifth ray, unusual facies, Pierre-Robin sequence, and syndactyly of the toes. Considering the other congenital conditions that present in select individuals with CFD, the underlying pathophysiology and, therefore, genetic causes are likely to be distinct in this group of patients. In other words, one must be careful to assume that all patients *diagnosed* by today's criteria with "CFD" will have the same underlying causality.

An inheritable genetic defect or susceptibility to other risk factors or exposures at a particular stage in lower extremity development of the fetus would commonly present with bilateral involvement; however, bilateral involvement is rare, and associated with other known genetic defects, such as Pierre-Robin sequence. The Pierre-Robin sequence is primarily an anatomical defect of mandibular outgrowth that impacts on oropharyngeal volume and patency of the palate. No single genetic abnormality has been identified in Pierre-Robin, though recent evidence shows that Sox9 may be a crucial step in the pathway [12]. Similarly, it is likely that a variety of inheritable genetic defects are responsible for a common resultant phenotype with similar but varied congenital abnormalities. Conversely, a recent report of a child with Pierre-Robin and bilateral CFD found that array comparative genomic hybridization analysis showed no abnormalities at 1,543 loci while whole-exon analysis identified no mutations suspected to be causative for the patient's condition [13].

A subset of bone morphogenetic proteins (BMPs), known as growth/differentiation factors (GDFs), affect several skeletal processes, including endochondral ossification, synovial joint formation, and tendon and ligament repair. Studies in mice have suggested that GDF-5 deficiency affects the composition and material properties of cortical bone tissue in the femur, but the detailed mechanisms by which this occurs remain to be determined [4]. Paley et al. reported radiographic mineralization of the pseudoarthrotic region of the femur in patients with CFD with the application of exogenous BMP-2 directly to the area [14].

Other investigators have supported a teratogenic cause for CFD as the phenotype is similar to the effects of thalidomide exposure in utero [15]. These effects imply that at a particular stage in fetal growth an exposure in utero occurs causing a downstream effect on the remaining growth of the affected lower extremity. The exact stage in development at which the abnormality occurs could determine the severity of involvement. While a teratogenic exposure is possible as the underlying cause, given the embryology of the limb, the explanation would have to involve a teratogen with transient effect only at the time of exposure. Thus, fetuses exposed to the teratogenic agent at one time point might have only mild CFD, exposed only at the time of limb bud formation of the proximal femur, while those fetuses with more severe involvement have persistent exposure to the teratogenic agent.

Endogenous retinoids, including vitamin A and retinoid acid (the active form of vitamin A), have long been implicated in limb development and abnormalities [5–7]. Embryological studies of quail, rats, and mice demonstrate a role for retinoic acid in the organization of the dorsoventral axis [7, 16]. These results suggest that a combination of maternal vitamin A supplementation and excess vitamin A in the neonatal period could potentially be unfavorable for cartilage development. It can be speculated that neonatal vitamin A supplementation may be beneficial for bone only if the mother's vitamin A status is low.

In summary, the embryologic and genetic data point towards a somatic mutation disorder of cells within the developing limb bud as opposed to a germ cell mutation (hereditary, familial) disorder for unilateral CFD. In contrast, multi-limb cases may be related to a germ cell mutation.

Deformity and Pathoanatomy

Osseous Deformities

CFD was initially grouped with cases of coxa vara, as this is the most obvious deformity in the majority of cases [17]. Cases were often separated into congenital short femur and proximal femoral focal deficiency (PFFD), for mild versus severe cases, respectively. Recognition that these were a spectrum of pathology of the same congenital deficiency leads to a consensus name change to CFD [18]. Although the femoral deformity and deficiency are the most obvious, non-femoral structures are also involved. These include the acetabulum, musculature, vessels, ligaments of the knee, tibia, fibula, and foot. In utero ultrasounds have shown both a shortened femur and a shallow acetabulum [19]. As with other pediatric proximal femur pathologies, dysplasia of the acetabulum whether initially present or not is progressive [20].

The deformities of the femur vary from coxa valga in very mild cases to severe coxa vara in more severe cases (Fig. 22.1). There is a flexion and retroversion deformity of the upper femur. The distal femur demonstrates a valgus deformity often referred to as a hypoplastic lateral femoral condyle since the distance from the physis to the knee joint line is smaller laterally than medially. In other words, there is a greater valgus convergence between the joint line and the distal femoral physis. The deformity of the proximal femur will be discussed in greater detail in a later section.

The ratio of growth in length of the short limb compared to the long limb remains relatively unchanged throughout growth [21–23]. This enables the final discrepancy in leg length to be predicted from the initial radiographs [24]. On this basis the Paley multiplier method is able to accurately calculate the predicted limb length discrepancy (LLD) at skeletal maturity [25, 26].

Ligamentous Structures

Anterior-posterior instability of the knee is common in CFD, but its severity is variable. Multiple modern era studies utilizing MRI have evaluated the ligamentous structures at the knee in patients with various degrees of CFD. The results support the conclusion that like the hip, the distal femur and proximal tibia require interaction of bony and soft tissues to develop normally.

Manner et al. [27] performed a radiographic study of 34 knees associated with CFD with radiographs and MRI. The anterior cruciate ligament (ACL) was affected in all knees studied, with 15 % hypoplastic and 85 % absent. The PCL was hypoplastic in 21 % and absent in 24 %. The most common type (14 knees, 41 %) was aplasia of the ACL and a normal PCL. The cruciate ligament dysplasias were differentiated into three groups (Fig. 22.2). Type I had a hypoplastic or absent ACL and a normal PCL. The intercondylar notch width and height were decreased compared to the normal side, and the lateral tibial spine was hypoplastic. Type II knees had aplasia of the ACL and hypoplasia of the PCL. They had even narrower and shorter intercondylar notches, and both tibial spines were hypoplastic. Two cases had lateral femoral osteochondral lesions. Type III knees had aplasia of both cruciate ligaments. The intercondylar notch was essentially absent and covered with hyaline cartilage, and both tibial spines were aplastic. The distal femoral joint surface is concave, matching the convex tibial plateau like a ball and socket. Three of these cases had discoid meniscus.

Overall, the authors found that on the tunnel-view radiographs, narrowing or absence of the femoral notch and flattening of tibial eminences corresponded to a hypoplasia or deficiency of the cruciate ligaments. Thus, the shape of the distal femur and proximal tibia can be used to predict, with relative accuracy, the presence or absence of the ACL and PCL ligaments.

These radiographic studies have been validated with the use of arthroscopy to directly confirm the presence or absence of the ACL and PCL. Johansson and Aparisi [28] published a case series of six patients with cruciate ligament dysplasia. Three patients had both an anterior and posterior drawer sign, and arthroscopy confirmed aplasia of both the ACL and PCL. Three patients had an isolated anterior drawer, and had ACL aplasia on arthroscopic examination. All knees were found to have hypoplastic tibial spines on radiographs.

Chugiak [29] looked at a larger series of 21 patients with clinical and arthroscopic examinations. All patients were found to have an anterior drawer sign, and nine patients (43 %) had a posterior drawer sign. Four patients (19 %) had medial instability, including one patient who also had lateral instability. The instrumented and clinic drawer tests were not found to be reliable enough, leading the authors to recommend imaging of the cruciate ligaments prior to lengthening

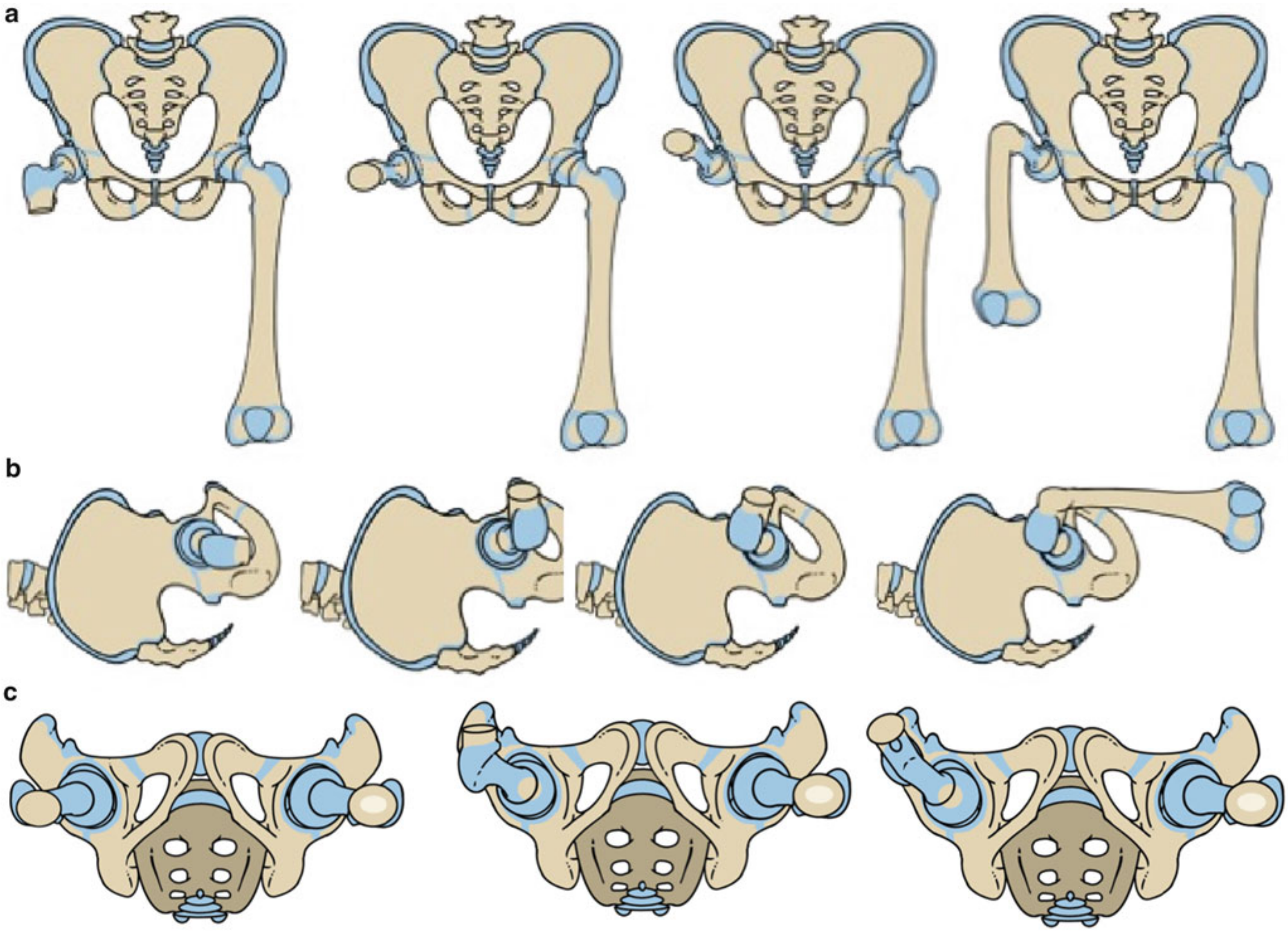

Fig. 22.1 Anteroposterior (**a**), lateral (**b**), and inferior-superior (**c**) views of pelvis and hip joints. These illustrations serve to simulate and recreate the proximal femoral deformity seen with severe CFD on the right hip. The model is based on a normal proximal femur with a 130° neck-shaft angle. First, flex the proximal femur 90° relative to the pelvis. Next, abduct the flexed proximal femur 45° relative to the pelvis. Now reconnect the distal femur to the proximal femur. The distal femur should be placed in 45° of external rotation relative to the pelvis. The resulting deformity is the CFD femur deformity typically seen in most type 1b and some type 1a cases. Note that the femoral neck appears to be retroverted due to the 90° hip flexion of a 130° neck-shaft angle femur. Just the flexion makes it appear to have 50° of retroversion. Since the distal femur is fixed to the proximal femur in external rotation this retroversion is increased even more. Also note that the varus deformity is caused by the abduction of the proximal femur with the hip flexed. The hip flexion places the greater trochanter facing posteriorly. The proximity of the greater trochanter to the ilium and to the sacrum in this position explains why the gluteus medius and minimus, and piriformis muscles, are short

to avoid knee dislocation. The majority of patients (38 %) were found to be completely deficient in both the ACL and PCL. The ACL was completely deficient in 16 (71 %) and hypoplastic in 3 (14 %) patients. The PCL was completely deficient in 10 (48 %) and hypoplastic in 3 (14 %) patients. Only one patient had an intact ACL and PCL.

In the majority of patients, both menisci were intact, with only three (14 %) hypoplastic and unrelated to the cruciate ligaments. Changes in the intercondylar notch and tibial eminences were noted in some patients but it was not specifically studied, as the authors felt that they were not as appreciable in patients under 6 years of age. They did note that the femoral intercondylar notch developed in some patients that had aplasia of both cruciate ligaments.

Muscle Pathoanatomy

Despite the wide spectrum of CFD, the underlying muscular anatomical differences appear relatively consistent. Pirani et al. showed that in patients with Aitken types A through D, the muscles were all present but altered in their size, structure, and location [30]. The study used MRI to qualitatively assess the musculature. The majority of muscles were found to be smaller: gluteus maximus, gluteus medius, gluteus minimus, quadriceps, adductor magnus, adductor longus, adductor brevis, pectineus, semimembranosus, semitendinosus, and biceps femoris. The exception is the sartorius muscle that was found to be hypertrophied, to which Pirani et al. and Panting et al. attributed as the underlying cause of the deformities of the proximal femur, given the sartorius'

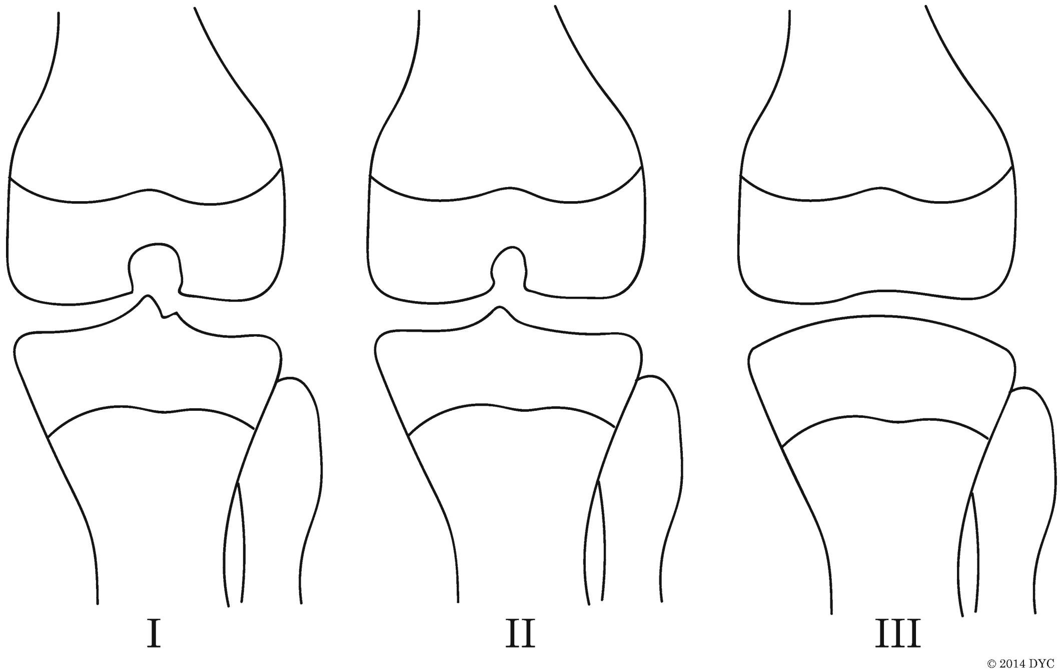

Fig. 22.2 Manner et al. [27] classification of intercondylar notch using the tunnel view correlates to deficiency of the cruciate ligaments. Type I: The intercondylar notch width and height are decreased compared to the normal side. The lateral tibial spine is hypoplastic. This corresponds to a hypoplastic or absent ACL and a normal PCL. Type II: Narrower and shorter intercondylar notch with hypoplasia of both tibial spines. This corresponds to aplasia of the ACL and hypoplasia of the PCL. Type III: The intercondylar notch is absent and covered with hyaline cartilage, and both tibial spines are aplastic. The distal femoral joint surface is concave, matching the convex tibial plateau. This corresponds to aplasia of both cruciate ligaments

orientation [31]. The obturator externus was found to be elongated and the muscle belly extended almost entirely to its insertion. The short external rotators had a larger cross-sectional diameter and were found to insert on the postero-medial greater trochanter. Overall, the course of the muscles was more perpendicular and proximal than normal, inserting onto the proximally migrated femur.

We have also found this to be true in our surgical experience: the musculature is present, the muscle bellies extend nearly to their insertion, and the course of the muscles is proximal. Panting and William had previously noted similar findings, including the hypertrophied sartorius muscle, the relatively normal gluteal muscles, and a hypoplastic quadriceps muscle [31]. Biko et al. also used MRI in seven patients to primarily evaluate the osseous structures, and reported a qualitative decrease in size of the musculature in general. At the level of the triradiate cartilage, the cross-sectional size of the gluteus muscles was significantly smaller in comparison to the uninvolved contralateral limb [32].

Pirani et al. postulated that the muscles became the primary stabilizers of the deformed hip joint, since the osseous structures did not impart inherent stability. However, in our experience the majority of untreated patients with CFD do not report ipsilateral hip pain, even after knee fusion and with prosthetic use and weight bearing.

Vascular Pathoanatomy

The normal embryological development of the arteries of the lower extremity parallels the formation of limb buds, occurring between the 4th and 8th weeks of gestation. The limb begins with a single axial dorsal artery that continues as the ischiadic artery and on to the popliteal artery. The external iliac artery arises at 5 weeks of gestation and bifurcates into the inferior epigastric artery and the femoral artery, which then bifurcates at 6 weeks gestation into the lateral and medial branches. The medial branch becomes the deep femoral artery and gives rise to the medial and lateral circumflex arteries and connects to the ischiadic artery, while the lateral branch develops into the femoral artery. By 7.5 weeks the femoral artery caliber is larger than the ischiadic artery and becomes the major blood supply to the popliteal artery and leg. After full development, the dorsal axial artery remains as the inferior gluteal artery while the ischiadic artery remains as the arteria comitans nervi ischiadici, running as the vasa vasorum of the sciatic nerve [33].

The vascular anatomy is also usually abnormal in an extremity affected by CFD. Chomiak et al. reported the results of 21 patients with various degrees of CFD studied with computed tomography (CT) angiograms to identify vascular abnormalities [34]. The authors found that more severe cases of CFD had a smaller diameter and a shorter length of the femoral artery; however, the severity of osseous abnormalities did not directly correlate with the topographical vascular anatomy abnormalities. All patients, at the least, had differences compared to the contralateral unaffected extremity: mainly with smaller vessel caliber, decreased number of vessels to the thigh, and more proximal bifurcation of the external iliac into femoral artery and deep femoral artery. Despite this, 19 of the 21 patients had the blood supply to the femur and pseudoarthrosis from branches of the deep femoral artery, which originated from the external iliac artery.

Notably, Chomiak found 2 of the 21 patients with a persistent ischiadic artery as the dominant vascular supply to the leg and a diminutive femoral artery that supplied solely the medial thigh [34]. This is an extremely unusual finding in otherwise normal humans, but is seen in lower mammals [35].

It is critical to identify these vascular anomalies preoperatively when reconstruction using rotationplasty is considered. The senior author has identified several cases with a dominant ischiadic artery and absent superficial femoral artery while performing rotationplasty. In our opinion, an MR angiogram (MRA) is indicated in all such cases prior to a rotationplasty.

Box 22.1. CFD Pathology

- The incidence of CFD is approximately 1 in 50,000.
- Limb bud formation is affected by a complex interplay of signaling, including AER, ZPA, and Wnt.
- CFD is likely caused by a somatic mutation during the development of the limb bud, but some cases may be caused by an inherited germ cell mutation.
- CFD usually presents with proximal coxa vara and distal femoral valgus.
- Cruciate ligament deficiency is common and can often be identified on radiographs.
- Generally, most muscles around the pelvis are present and hypoplastic, though the sartorius may be hypertrophied.

  An ischiadic artery may be present and more dominant than the femoral artery in some patients with CFD.

Evaluating the Child with Unilateral CFD

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 of CFD are now identified with prenatal ultrasound early in the pregnancy by measuring the lengths of the two femurs. In such cases the predicted leg length discrepancy at birth and maturity can be calculated using the multiplier method [36].

Physical Exam

There is an obvious leg length discrepancy. Associated fibular hemimelia (FH) and ray deficiency may be present. The hips, knees, and ankles should be examined for range of motion and flexion contractures. Neonates and young infants normally have such contractures for the first 3–6 months. At the hip, patients may have external rotation and fixed flexion deformity of the hip, as well as limited abduction when coxa vara is present. At the knee, patients may have flexion contractures, hypoplastic or subluxation-dislocation of the patella, AP, or rotary instability of the knee.

Finally, muscle length tests are recorded to identify preexisting limits to muscle lengthening. These tests include the Ely test for rectus femoris tightness, popliteal angle measurement for hamstring tightness, and Ober sign for fascia lata or iliotibial band tightness (Fig. 22.3).

Box 22.2. Physical Examination Findings Often Seen in Patients with CFD

- Hip: External rotation (ER) deformity or increased ER versus internal rotation (IR); fixed flexion deformity (FFD) of hip; limitation of abduction (when coxa vara is present).
- Knee: Fixed flexion deformity of knee; no limitation of knee flexion; hypoplastic patella; lateral tracking or subluxated 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; temporary locking of the knee during flexion.
- Ankle: Limitations of ankle dorsiflexion; obligatory eversion with dorsiflexion; hypermobility of ankle with increased eversion; lateral malleolus more proximal than medial malleolus.

Imaging

Radiographic Examination

Radiographs should include a full-length anteroposterior (AP) standing legs with the patellas pointing forward. In children who are unable to stand, a pull-down X-ray may be

Fig. 22.3 Muscle lengthening tests: (**a**) The Ely test checks for rectus femoris tightness, demonstrating pelvic flexion with prone knee bend; (**b**) the popliteal angle is the angle between the vertical and the line of the tibia with the hip at 90° flexion and the knee maximally extended; (**c**) the Ober test checks for fascia lata tightness; a positive sign is the thigh going into an abduction position when the hip is hyperextended and the knee flexed to 90°

performed. It allows measurement of the length of the femurs and tibia, though it does not include foot height. Long lateral leg radiographs in maximum extension allow for evaluation of knee flexion contractures and more accurate length measurement. A supine AP pelvis allows more accurate measurement of the center-edge angle (CEA) of both hips to assess for hip dysplasia. It is also a better quality X-ray to assess for ossification of the femoral neck. It is important that the pelvis be level for more accurate measurement.

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) is useful to assess the 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. In case of a femoral neck pseudoarthrosis, the MRI can help determine if the cartilage of the femoral head is fused to the acetabular cartilage. For optimal imaging, the cuts of the proximal femur should be reformatted in an oblique plane to see the entire proximal femur as a single image. MRI can also help outline the intra-articular pathology of the knee, identifying deficiency of the cruciate ligaments and outlining the shape of the joint surfaces in the frontal and sagittal plane.

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.

Box 22.3. Evaluating the Child with CFD

- CFD can be bilateral; check the contralateral side as well.
- The hip, knee, and ankle should all undergo thorough physical examinations.
- Fibular hemimelia is commonly associated with CFD.
- Radiographic imaging should include a full-length AP legs (standing or pull-down), long leg laterals in maximum extension, and a supine AP pelvis.
- MRI is useful to evaluate soft tissue, ligaments, and a cartilaginous femoral neck.

Classification Systems

Multiple classification systems have been described over the years in attempts to categorize the pathology and direct surgical treatment (Figs. 22.4, 22.5, and 22.6). Most older classification schemes were based on plain radiographs. Newer classification systems incorporate modern imaging modalities, specifically MRI, which allows detailed evaluation of osseous, cartilaginous, and soft-tissue structures, including the non-ossified femoral head, non-ossified acetabulum, labrum, pseudoarthrosis, and musculature [32]. The ability to perform MRI under anesthesia for infants allows for interpretation of these images with better reliability. As a result, MRI dramatically improves the ability to correctly categorize patients over conventional radiographs alone [37]. Table 22.1 shows a comparison of several of the classification schemes further elucidated below.

Aitken, 1959

In 1959 Aitken proposed a four-class schema for PFFD (see Fig. 22.4) [17]. This became the most commonly utilized classification system for PFFD. Aitken focused the classification system on the development of the femoral head, neck, and acetabulum, with progressive dysplasia from one class to the next. The Aitken classification does not include the congenital short femur group discussed earlier.

Aitken initially proposed that Class A patients were not technically PFFD, as all the structures were present, albeit deformed. These patients had a femoral head, a cartilaginous femoral neck, an adequate acetabulum, and a very short femoral segment. Severe subtrochanteric femoral varus and femoral shortening were appreciated. A pseudoarthrosis of the proximal femur was appreciated between the femoral shaft and head, either between shaft and trochanteric component (subtrochanteric pseudoarthrosis) or between trochanteric component and head (femoral neck pseudoarthrosis). Aitken noted that the pseudoarthrosis did not spontaneously resolve in all patients by skeletal maturity, but did eventually heal in some patients.

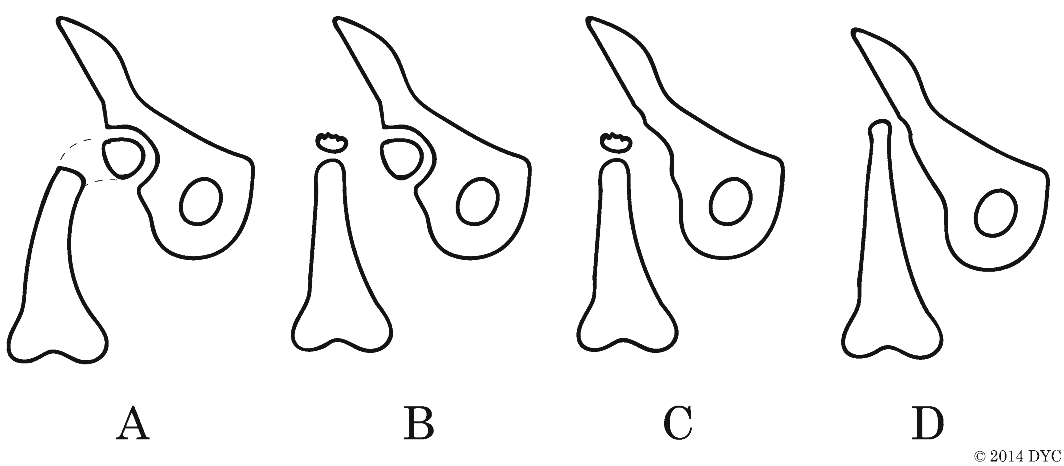

Fig. 22.4 Aitken classification of congenital femoral deficiency. The Aitken classification is based on the relationship of the acetabulum and the femoral head: (**a**) Adequate acetabulum, femoral head is present and attached to shaft; (**b**) adequate acetabulum, femoral head not connected to shaft, which instead has a proximal ossified tuft; (**c**) dysplastic acetabulum, minimal or absent femoral head, disconnected femoral shaft with tuft; and (**d**) no acetabulum, no femoral head, shortened proximal femoral shaft with no tuft

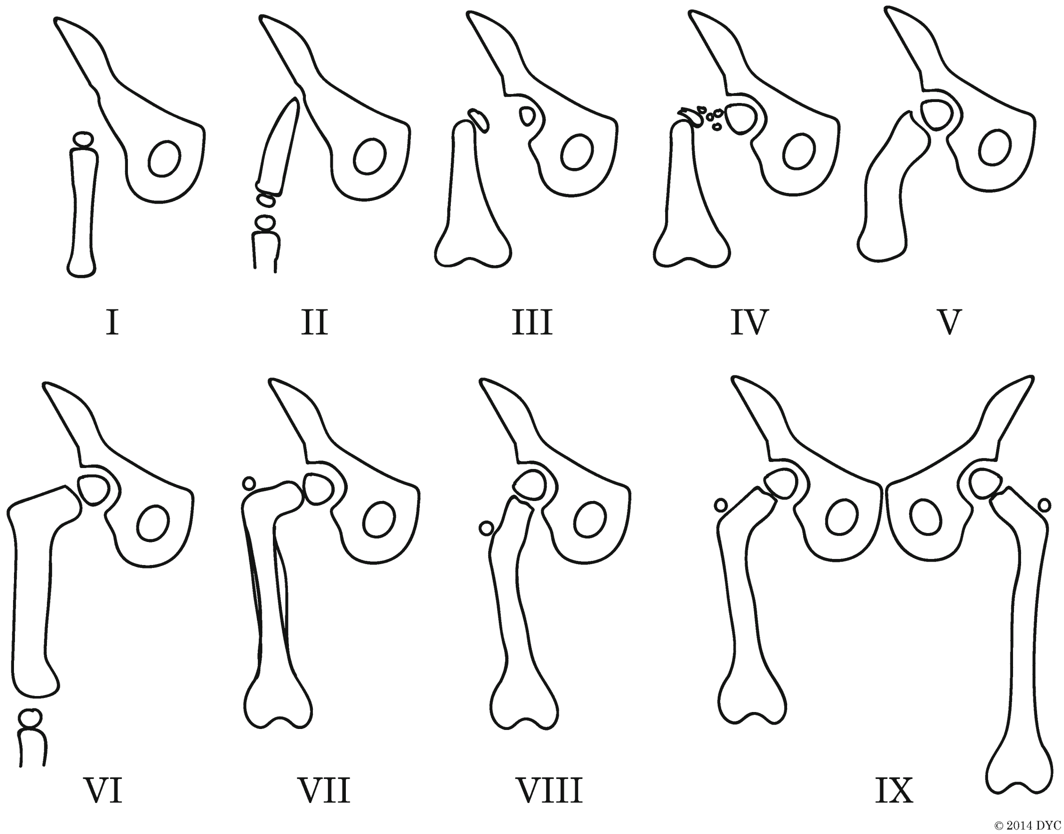

Fig. 22.5 Pappas classification of congenital femoral deficiency. The Pappas classification is divided into nine classes: (I) congenital absence, (II) proximal femoral and pelvic deficiency, (III) proximal femoral deficiency with no osseous connection between femoral shaft and head, (IV) proximal femoral deficiency with disorganized fibrosseous disconnection between femoral shaft and head, (V) midfemoral deficiency with hypoplastic proximal and distal development, (VI) distal femoral deficiency, (VII) hypoplastic femur with coxa vara and sclerosed diaphysis, (VIII) hypoplastic femur with coxa valga, and (IX) hypoplastic femur with normal proportions

The hallmark of the Aitken Class B PFFD was a lack of connection, either bony or cartilaginous, between the femoral shaft and head that persists into skeletal maturity. These patients have a femoral head within an adequate acetabulum and a shortened, deformed femoral shaft. The proximal portion of the shaft often has an ossified tuft. The femoral head and shaft do not move as a unit.

Aitken Class C is defined by a severely dysplastic acetabulum. The femoral head does not ossify and shaft of the femur is short with an ossified tuft at the proximal end of the shaft. There is no cartilaginous or osseous femoral neck or trochanteric component.

Aitken Class D is the most dysplastic form of PFFD. This class is characterized by the absence of the acetabulum and femoral head and a deformed, shortened femoral shaft without tufting on the proximal shaft of the femur.

Pappas, 1983

The Pappas classification (see Fig. 22.5), published in 1983 [38], was based on the anatomy of 139 cases evaluated in Massachusetts. Pappas describes nine classes based on the femoral deficiency and associated deformity, with diminishing severity as the class number increased. This was the first classification scheme to separate middle and distal femoral deficiencies into their own class. Recommendations were given for treatment for each class; however, treatment was depen-

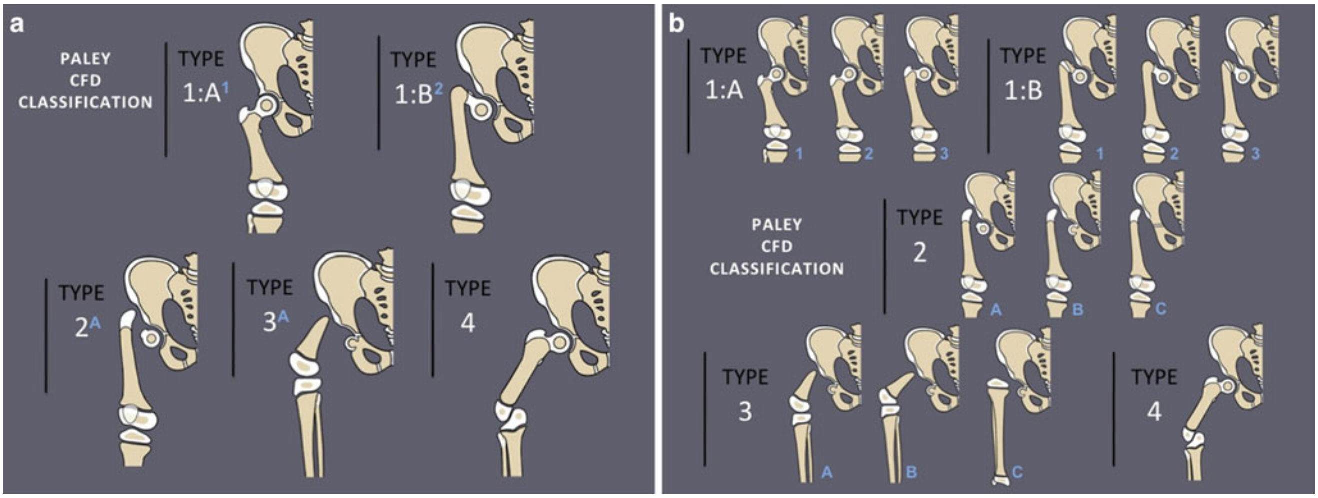

Fig. 22.6 (**a**) Paley classification of congenital femoral deficiency. There are four types of congenital femoral deficiency: (1) intact femur normal ossification (**a**) or delayed ossification (**b**); (2) mobile pseudoarthrosis; (3) diaphyseal deficiency; (4) distal deficiency. (**b**) Each Paley type has several subtypes: $1a_1$: Normal-shaped short femur with mild genu valgum; $1a_2$: normal-shaped short femur with mild genu valgum and retroversion; $1a_3$: coxa vara, retroversion, and mild genu valgum; $1b_1$: subtrochanteric type; $1b_2$: neck type; $1b_3$: combined neck and subtrochanteric type; 2a: mobile femoral head; 2b: partially fused femoral head; 2c: completely fused or absent femoral head; 3a: mobile knee joint with greater than 45° of motion; 3b: stiff knee with less than 45° of motion; 3c: absent femur or fused knee joint

Table 22.1 Comparison of different classification schemes

	Paley [42]	Pappas [38]	Gillespie and Torode [39]	Hamanishi [15]	Aitken [17]
Normal anatomy/minimal deformity/ hypoplasia	1A	7,8,9	Group 1	Type 1, type 2	Class A
Subtrochanteric delayed ossification	1B subtrochanteric	4, 5	Group 1	Type 3a	Class A
Femoral neck delayed ossification	1B femoral neck	3	Group 1	Type 3b	Class A
Combined delayed ossification	1B combined	3		Type 4a	
True pseudoarthrosis with no cartilaginous or osseous femoral neck	2a, 2b	3	Group 2	Type 4b	Class B
Dysplastic acetabulum, no femoral head	2b, 3	2	Group 2	Type 4c	Class C
No proximal femur, short distal femur only	3a,b	2	Group 2	Type 4c	Class D
Femoral absence	3c	1		Type 5	
Distal femoral deficiency only	4	6			

dent on the patient's joint stability and function, which are not part of the classification system. Therefore treatment could not be based directly on the classification schema of Pappas.

Gillespie and Torode, 1983

In 1983, Gillespie and Torode proposed a straightforward division of CFD into two categories: congenital short femur and true PFFD [39, 40]. Each of the two groups had both radiographic and clinical criteria, but were ultimately defined by the function of the hip and knee joints, rather than the type of surgery each would necessitate.

Group 1 was defined as those patients who could have good hip and knee function with or without surgical correction. Radiographically, group 1 patients had coxa vara, lateral femoral bowing, and hypoplasia of the knee with valgus but the predicted leg length discrepancy was no greater than 30 % of the normal femur and femoral deficiency was 40–60 % of the contralateral femur. Clinically this group had laxity in the sagittal plane and mobile joints without flexion deformities at hip or knee.

Group 2 patients had hip and knee joints that were not adequate to support the patient without pain. The appropriate treatment recommendation for this group of patients was amputation and/or rotationplasty of the lower limb. These patients had markedly short femurs. Radiographically, these patients had less than 40 % of the femoral length of the normal side and deficiency or absence of the femoral head and/or neck. The authors defined deficiency of the femoral head as a tenuous cartilaginous connection between femoral shaft and head. Clinically, these patients had fixed flexion deformity at hip and/or knee.

The overall goal of the Gillespie and Torode classification was to simplify the decision making. The authors rationalized for families and surgeons a "life plan" regarding treatment of the affected limb based on each of the two groups. This classification aims to aid families and physicians communicate more effectively and correlate early clinical signs with long-term treatment plans. The value of this classification schema is its simplicity and its guidance in surgical treatment. The drawbacks to this classification scheme are its inability to adapt to improved surgical techniques based on a better understanding of pathoanatomy, as well as advances in biology and technology.

Hamanishi, 1980

Hamanishi [15] published a five-tiered classification system for congenitally short femur. This schema involved a scoring system that included the entire lower limb and upper limb involvement, as opposed to other classification systems, which focus only on the femur. However, the schema does not include specifically the acetabulum, as the author believed that this component is directly related to the femoral head. It was intended to differentiate those patients with the teratologic sequelae of thalidomide exposure from those patients with idiopathic CFD. The diagnosis was based on a series of 60 total patients, 14 of which were known to have had in utero thalidomide exposure. It is an observational classification system and does not aim to guide treatment or prognosis. The study was quite comprehensive as the authors included socioeconomic class, an extensive family history, parental age, birth history, and in utero history in their review. The age of the study subjects at the time of evaluation varied from 6 months to 50 years.

Hamanishi identified several differences between the two groups: overall, the thalidomide exposure group had femora that were more severely affected than the idiopathic group; the thalidomide exposure group more often had concomitant abnormalities of the tibia and radius as opposed to the fibula and ulna in the idiopathic CFD group; the thalidomide group had lower extremity preaxial (tibial) polydactyly, whereas the idiopathic group had lower extremity postaxial (fibular) deficiency. The author defined five groups with subgroups in some utilizing radiographic measurements of the affected femur to quantify the degree of deformity.

Group 1 was defined by hypoplasia of the femur, either without deformity type 1a or with minimal angulation and cortical thickening, type 1b. The type 1a was associated with ipsilateral below the knee deformities, while the type 1b was associated with severe, symmetric bilateral below the knee deformities. The second group was defined by hypoplasia and femoral varus angulation. Type 2a had a normal neck-shaft angle but type 2b had a decreased neck-shaft angle. A transverse subtrochanteric ossification defect was noted in this type as well, but all ossified later in life.

The type 3 group had marked coxa vara from delayed and inconsistent ossification of the femoral neck and subtrochanteric region. This group also has marked retroversion of the femoral neck and distal femur external rotation. Type 3a with a straight femoral shaft was considered "stable" as the proximal femoral varus would not progress, while the "unstable" type 3b with an increased epiphyseal head angle >60° was reported to progress with time.

The type 4 femurs had absent or stunted and tapered proximal femoral shafts with the femoral head attached directly to the shaft, when present. The femoral head was connected to the shaft by a pseudoarthrosis and the entire shaft was proximally migrated. The type 4a had a pseudoarthrosis of the femoral neck and subtrochanteric region. The type 4b had an absent proximal femur with the femoral head connected directly to a tapered proximal shaft. The type 4c had a complete absence of the proximal femur.

Type 5 femurs indicated a rudimentary femur, which ossified later, or complete absence.

One major flaw in this classification system is that it divides the groups based on measurements with arbitrarily chosen cutoffs, such as the neck-shaft angle and femoral varus which change with time. Thus, young patients could move from one group to another without intervention and the treatment plan would also therefore be variable. Additionally, this classification scheme does not include the stability of the hip or knee, and the presence of knee ligaments, and does not guide surgical or nonsurgical treatment.

Some important remarks from this study are that the leg length discrepancy was consistent, when stated as a percentage of the unaffected normal side, which was consistent with older retrospective reports [21, 22] and newer mathematical methods to predict limb leg discrepancy at maturity [26].

Kalamachi, 1985

The Kalamachi classification (1985) [41] attempted to relate the femoral deficiency directly to the limb function. Type I patients had congenital shortening of the femur with a normal hip and knee morphology and function. Type II patients had a congenitally short femur with coxa vara, with some irregularity of the growth plate, a located hip, and a well-developed acetabulum.

Type III patients had a developed acetabulum but a deficient proximal femur. With time, they were separated into two groups. In the IIIA group, the radiographically apparent proximal femoral defect ossifies into a bony bridge, often in varus. In the IIIB group, the defect does not ossify, and develops into a pseudoarthrosis. Type IV patients had no acetabulum and no connection between the pelvis and the distal femoral fragment, which often was a spike proximally. Type V patients had no hip joint and no femur. For all five classification types, the most common treatment was a Boyd or Syme amputation. Knee fusions were used to correct deformity as well as provide a large lever arm for a severely shortened femoral segment. Femoral osteotomies were used mostly in type II and IIIA patients to correct coxa vara. Limb equalization procedures were done in nine patients (four epiphysiodesis, five lengthening) of type I and II. The rest were treated non-operatively or with orthotic and prosthetic fitting.

Paley, 1998

Paley proposed a classification system for CFD (first presented at a symposium in Dallas in 1996 documented in a book published from that symposium [42]) based on the degree of development and integrity of the femur (see Fig. 22.6). Unlike the previous classifications, Paley wanted to create a classification where the type would not change with the degree of ossification of the femur (age dependent). A longitudinal follow-up of different classification systems by Sanpera and Sparks [43] showed that the existing classification systems were inaccurate in predicting the final femoral morphology based on the initial radiographs. Paley also wanted a classification that was oriented towards reconstruction and not amputation. As such, the classification types and subtypes each have a separate surgical reconstructive prescription.

Box 22.4. Femur Deficiency Classification (Paley, 1998) [42]

Type 1: *Intact femur* with mobile hip and knee:
(a) Normal ossification proximal femur
(b) Delayed ossification proximal femur (neck, subtrochanteric, or combined neck subtrochanteric types)

Type 2: *Mobile pseudoarthrosis* (greater trochanteric apophysis present), knee usually mobile:
(a) Femoral head mobile in acetabulum
(b) Femoral head partially fused to acetabulum
(c) Femoral head and acetabulum completely fused or absent

Type 3: *Diaphyseal deficiency* of femur (greater trochanteric apophysis absent):
(a) Distal physis present; knee motion ≥45°
(b) Distal physis present; knee motion <45°
(c) Complete deficiency of distal femur or fusion of distal femoral remnant to tibia (distal physis absent)

Type 4: *Distal deficiency of femur* (proximal end normal)

Treatment Options for Congenital Femoral Deficiency

Rotationplasty

Rotationplasty has been a commonly accepted treatment for patients with CFD with significant proximal deficiency, which is deemed “unreconstructable” [44–48]. The most commonly employed surgical technique is the Van Nes rotationplasty and variations of this technique. In the Van Nes rotationplasty, the lower limb is rotated 180° to use the ankle and foot as a functional knee joint with a prosthesis. The rotationplasty can be combined with a knee fusion when the knee is unstable, or as in the Brown modification the knee is maintained after 180° rotation and the distal femur is fused to the pelvis to substitute for a deficient hip allowing flexion and extension [44]. This will be discussed in more detail in a later section.

Gillespie and Torode reported their surgical recommendations along with their classification schema in 1983 [39, 40]. Prior to the advent of the Wagner device in 1971, patients classified as Group 1 with congenital short femur underwent rotationplasty. After the Wagner technique demonstrated the feasibility of lengthening up to 20 cm, femoral lengthening became the standard recommendation for Group 1, but to a maximum of 20 % of the femur length because the risk of hip and knee subluxation or dislocation was considered too high. Of the five Group 1 patients that underwent rotationplasty, three had good function but two required a Syme amputation and fitting with an above-the-knee prosthesis. Patients classified by Gillespie and Torode as Group 2 had true PFFD and received rotationplasty, Syme amputation for prosthesis fitting, and/or knee fusion. Of 43 Group 2 patients with instability of the hip and knee, 21 underwent rotationplasty, and of those, 8 required de-rotational surgery and 2 required a Syme amputation. They strongly advocated knee fusion for Group 2 patients to correct the valgus and flexion deformities at the knee and tibial rotationplasty to restore “knee flexion” with the ankle dorsiflexion.

Brown reported on three patients with severe CFD who underwent a modification of the Van Nes rotationplasty with mean of 6-year follow-up who had satisfactory results and range of motion of both the hip and knee to 90° of flexion [44]. The Brown modification includes resecting the deficient proximal femur, externally rotating the limb 180°, fusing the femoral remnant to the pelvis, and leaving the muscles distal to the knee undisturbed. Using this technique there was no derotation as previously recurred with the Van Nes rotationplasty which incorporated a midtibial osteotomy for rotation.

Ackman et al. reviewed 12 patients who underwent Van Nes rotationplasty for CFD to determine the long-term outcome [48]. At a mean of 21.5 years (11–45) after their rotationplasty, a total of 12 prosthetic patients were compared with 12 normal age- and gender-matched controls. The authors found no differences between the groups in overall health and well-being on the SF-36, but significant differences were seen in gait parameters in the CFD group. Patients who had undergone Van Nes rotationplasty had a high level of function and quality of life at long-term follow-up, but presented with significant differences in gait and posture compared with the control group.

Syme Amputation

Alman et al. reviewed the results of treatment of 16 patients who had had an isolated unilateral PFFD; 9 were managed with a rotationplasty and 7 with a Syme amputation combined with an arthrodesis of the knee [49]. The perceived physical appearance, gross motor function, and metabolic energy expended in walking were assessed. The mean duration of follow-up was 9.9 years (range, 4–14 years). The mean age of the patients at the time of the study was 13.9 years (range, 8–18.4 years) in the rotationplasty group and 14.8 years (range, 9.5–19.9 years) in the Syme-amputation group. There were three female patients in each group. Roentgenograms showed that the femoral head was in the acetabulum (Aitken class A or B) in four of the seven patients in the Syme-amputation group and in five of the nine patients in the rotationplasty group; the remaining patients did not have this finding (Aitken class C or D). There was no difference in gross motor function or perceived physical appearance between the groups. Rotationplasty was associated with a more energy-efficient gait than was Syme amputation.

Fowler et al. measured lower limb kinematics and kinetics during preferred and fast speeds of walking in persons with PFFD to compare outcomes after Syme amputation (nine subjects) with those after Van Nes rotationplasty (ten subjects) [50]. Subjects with a Van Nes rotationplasty and full tibial rotation (seven subjects) demonstrated prosthetic knee function during stance as they were able to support a flexed-knee posture at both speeds and produced greater knee-extensor moments at preferred speeds as compared with the Syme group. Non-prosthetic limb compensatory mechanics were significantly exacerbated in subjects with a Syme amputation compared with the Van Nes group: (1) stance-phase vaulting, resulting in greater inappropriate ankle-power generation at both walking speeds; (2) excessive hip-extensor moments at fast speeds; (3) excessive hip-power absorption and generation at both speeds; and (4) excessive knee-joint power generation at both speeds. The improved gait after Van Nes rotational osteotomy is one factor that should be considered when making clinical decisions for children with PFFD.

Other studies have found some potential issues with Syme amputation in the long term: heel pad migration, skin sloughs, and problems with prosthetic fitting. Anderson et al. reviewed 69 Syme amputations performed in 62 children with the major indication of leg length discrepancy, due to either paraxial fibula hemimelia (33 cases) or proximal focal femoral deficiency (19 cases) [51]. The average age at amputation was 5.6 years, with an average follow-up of 10.5 years (range 1–25 years). Although the results were assessed by a combination of chart review, patient recall examinations, and questionnaires, satisfaction in adulthood was found to be high. Early complications included three skin sloughs and one infection. Late complications included 2 retained os calcis apophyses, 1 exostosis, and 16 cases of heel pad migration. Only one of the heel pad groups required revision; prosthetic adjustment resolved symptoms in the remaining patients. However, prosthetic knees were often too low because of failure to limit the length of the stump appropriately, though this finding was not as problematic in the CFD group as the residual limb was significantly shorter.

Limb Lengthening

Aston et al. published a series of 27 patients with Paley type 1 CFD (Pappas grades VII, VIII, and IX) who underwent a total of 30 lengthening procedures, with 3 patients undergoing a second femoral lengthening [52]. All patients underwent femoral lengthening with a multiplanar Ilizarov-type external fixator. The mean increase in length was 5.8 cm (3.3–10.4 cm) and 18.65 % of the total length of the femur (9.7–48.8 %). The mean time in the frame was 223 days (75–363) with a mean distraction index of 1.28 months per cm. The authors initially performed the osteotomy distal, but after 17 distal osteotomies changed to a proximal osteotomy for lengthening, noticing a significant increase in mean range of knee motion from 98.1° to 124.2° ($p=0.041$) and a trend towards a reduced requirement for quadricepsplasty, although this was not statistically significant ($p = 0.07$). The overall incidence of regenerate deformation or fracture requiring open reduction and internal fixation was similar in the distal and proximal osteotomy groups (56.7 % and 53.8 %, respectively). However, in the proximal osteotomy group, preplacement of a Rush nail reduced this rate from 100 % without a nail to 0 % with a nail ($p < 0.001$). When comparing a distal osteotomy with a proximal one over a Rush nail for lengthening, there was a significant decrease in fracture rate from 58.8 % to 0 % ($p = 0.043$). This supports the premise of Paley and Herzenberg that CFD patients should be lengthened over a Rush rod or have a Rush rod inserted after frame removal to reduce the risk of regenerate bending or fracture [53].

Aston et al. concluded that lengthening of the femur with an Ilizarov construct should be carried out through a proximal osteotomy over a Rush nail and lengthening to a maximum of 6 cm during one treatment, or 20 % of the original length of the femur, to reduce the risk of complications [52]. Paley and Standard have published on a distal osteotomy for CFD to avoid lengthening through the abnormal, sclerotic bone often seen in the proximal femoral shaft as much as possible with good success [54, 55].

Saghieh and Paley studied a group of 95 CFD patients who had undergone lengthening between the years 1988 and 2000 (Table 22.2). These patients did not undergo the Superhip or Superknee procedures that are described later in this chapter. All femoral lengthenings were with the Ilizarov device with extension of the fixator across the knee to the tibia with hinges. The postoperative result based on knee and hip range of motion score, gait score, lengthening goal score, and alignment score, pain score, and activity level score was excellent and good in over 93 % in all groups, with a mean overall lengthening of 6.0 cm (range, 2.2–12.5). Complications included femur fracture, pin site problems requiring surgery, premature consolidation, nerve irritation/palsy, delayed union, knee subluxation, and hip subluxation. The fracture rate in the older age group was significantly lower than the other groups, but it included several patients that were undergoing lengthening over nails. There was no significant difference in unplanned surgery rate between groups 1 and 2 (see Table 22.2). The difference with group 3 was related to refractures. Prophylactic rodding was not performed at the time of fixator removal in this series of patients, but is now part of the standard treatment algorithm, given the high rate of fractures [56].

The authors have previously presented their results on femoral lengthening after a hip stabilization procedure [57]. A retrospective review was performed of 35 patients with CFD, Paley types 1a and 1b, who underwent femoral lengthening after a pelvic osteotomy, proximal femoral osteotomy, or a combination. Patients underwent a hip stabilization procedure at a mean age of 2.4 years (2–5.5). The mean age at first femoral lengthening was 3.7 years (3–10.7). The mean overall limb length difference prior to femoral lengthening was 64.8 mm (47–100), with a mean postoperative difference of 8.1 mm (−14 to 32). The average amount of time in the external fixator was 186.1 days (107–311), or 1.1 months/cm. Preoperatively, average knee ROM was 2°–131° and hip ROM was −2° extension to 115° flexion. Follow-up at a mean of 14.4 months (5.6–45.2) showed return to baseline ROM in knee of 2°–119° and hip from −1° extension to 91° flexion. The overall rate of obstacles and complications was 37.1 % and 32.4 %, respectively [58].

Table 22.2 Limb lengthening results and complications in 95 patients

		Group 1 (<6 years)	Group 2 (6–13 years)	Group 3 (>13 years)
Number of patients (*n*)		30	40	25
Amount of length achieved				
	Length (cm)	5.4 (2.8–8.5)	6.2 (2.5–11)	6.3 (2.2–12.5)
	Relative (%)	39 (12–71)	24 (7–54)	20 (5–58)
	Lengthening index (month/cm)	1.0 (0.5–1.8)	1.1 (0.5–2.2)	1.1 (0.5–2.2)
Good/excellent outcome score		28/30 (93 %)	37/40 (93 %)	24/25 (96 %)
Problems, obstacles, and complications				
	Femur fracture	13	14	4
	Pin-site problems requiring surgery	4	3	1
	Nerve irritation/palsy	3	3	1
	Premature consolidation	0	3	3
	Delayed union	2	4	3
	Knee subluxation	5	5	3
	Hip subluxation	3	3	1
	Total unplanned surgery	21 (70 %)	22 (55 %)	9 (36 %)

Treatment of CFD has always been divided between amputation and lengthening. In the past, the indications for amputation were much more liberal. Currently, the dividing line can be based more on pathoanatomy. Treatment varies, depending on the severity of the underlying deficiency. The less involved patients, such as Paley type 1a, usually only require leg length equalization surgery, consisting of limb lengthening and/or contralateral epiphysiodesis at the appropriate time. The current treatment of more severely affected patients, such as Paley type 1b and type 2, consists of complex surgical interventions consisting of reconstruction of the hip and knee joints, correction of femoral deformity and length, ossification of the pseudoarthrosis, and reconstruction of lax or absent ligaments. This is discussed in the subsequent sections. The most severely affected patients, Paley type 3, usually undergo surgery to improve prosthetic use, as regaining satisfactory function with limb lengthening and reconstruction is often not feasible.

Box 22.5. Classification and Treatment

- Multiple classification schemes exist, mostly based on their anatomic deformity, though sometimes they are age dependent or have arbitrary cutoffs.
- The Paley classification separates into types and subtypes based on their surgical treatment for reconstruction.
- Van Nes rotationplasty has been used historically on severely shortened femurs with good results.
- Patients undergoing Syme amputation with knee arthrodesis often have more altered gait mechanics with greater energy expenditure compared to those undergoing a rotationplasty.
- In the properly selected patient, limb lengthening often has good outcomes, but can have significant complications. However, these untoward events can be lessened with prophylactic joint stabilization and intramedullary rodding techniques.

Recommended Surgical Reconstructive Strategy for Paley Type 1 CFD

Outlining a "Life Plan" for the Family

At the initial consultation, a surgical reconstructive strategy or "life plan" projected to skeletal maturity should be outlined for the child and the family. It is helpful to write this out on the back of a printed radiograph for the family to put in the child's scrapbook and to refer to in the ensuing years. The strategy is based on the type of CFD, the projected LLD at maturity, and the reconstructive potential of the hip and knee. In cases with combined CFD and FH, the strategy for FH must be combined with that of the CFD [18].

Step 1: Preparatory Surgery for the Hip and Knee

Prior to lengthening one must determine whether the hip and knee joints are stable and/or deformed and whether surgical procedures for these joints are required before initiating limb lengthening. At the hip, if the acetabulum has an acetabular index with comparable slope to the normal contralateral side, a center edge angle (CEA) ≥ 20°, and a neck-shaft angle (NSA) ≥ 110°, no separate hip surgery is required before the first lengthening [59]. If the acetabulum shows signs of dysplasia then a pelvic osteotomy should be performed prior to lengthening. Increased slope of the sourcil (acetabular roof) or acetabular index compared to the other side is a subtle but sensitive sign of acetabular dysplasia. Coxa vara should be corrected prior to lengthening if the NSA is less than 110°. 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 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. A bony flexion deformity may be treated by a proximal femoral extension osteotomy. The abduction contracture is treated by lengthening or resection of the fascia lata and, if necessary, an abductor muscle slide at the iliac crest. When all of these deformities are present together and especially with higher degrees of angulation, the reconstructive procedure is called the Superhip procedure [60].

Other factors should be examined prior to lengthening. The proximal femur should be normally ossified for the patient's age. Often, severe cases of CFD have delayed ossification of the femoral neck or subtrochanteric region, and bone morphogenic protein (BMP) may be added to the femoral neck to promote ossification [14]. The fascia lata is a thin but very tough limiting membrane that resists lengthening and applies pressure across the knee joint and the distal femoral growth plate. Thus, it should always be removed or released before lengthening. It can also be used to reconstruct absent knee cruciate ligaments if procedures are done concurrently. A hemiephysiodesis may also be performed to correct frontal plane deformities. The ideal age for the preparatory procedure is between ages 2 and 3 years.

Step 2: Serial Lengthenings of the Femur and/or Tibia

A prediction of total leg length discrepancy at skeletal maturity helps determine the approximate number of lengthening surgeries required. This can also be done using the Paley multiplier method [26]. The majority of CFD cases will require at least two lengthenings. The goal of each lengthening depends on the total discrepancy at maturity. The safe range of lengthening is from 5 to 8 cm, as long as a good physical therapy program is available. This amount seems to be independent of the initial length of the femur and age of the child, and can be performed safely even in a toddler. In most cases, the maximum of 8 cm is attempted because of a large total discrepancy, as long as the patient maintains adequate knee range of motion.

The first lengthening of the femur can be performed 12 months after the preparatory surgeries, assuming that the femoral neck has ossified. If the preparatory surgery is performed between the ages of 2 and 3 years, then the first lengthening can follow between the ages of 3 and 4 years. However, if the femur is excessively short (total femoral length <75 mm) for an external fixator or if the femoral neck fails to ossify, then it would be beneficial to wait for a year or two for more growth. By beginning lengthening at a young age, the level of prosthetic or orthotic need is reduced earlier in a child's life. In the senior author's experience, the complication rate for limb lengthening is no different for the younger age group [61]. Lengthening of the femur in children younger than 6 years may be associated with sustained growth stimulation [62].

Between 4.5 and 7 years, children may have more difficulty psychologically dealing with limb lengthening. This is related to the normal cognitive stages of children at this age. Children at this age group seem to understand too little and too much at the same time. They are beginning to be more independent and may appear to be mature enough to handle the process. They understand that they have one short leg. They do not connect the recognition of a short leg with the solution of limb lengthening. Their cognitive level is insufficient to understand why their parents allowed someone to do this to them. The younger children do much better because their cognitive level accepts everything their parents decide without questioning. Beyond the age of 6 or 7 years, the child enters the age of reason and begins to understand that there is a problem with a solution. Their cooperation is voluntary rather than coerced. One way to explain this to children is to focus on the size of their shoe lift. It is easier for them to understand that they can wear a normal shoe after the operation instead of understanding that their leg lengths will be equal.

The frequency of lengthening should be spread out to no less than every 3 years and preferably 4 years or greater. The rule of 4 is a good guiding strategy: one lengthening every 4 years starting by 4 years. 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 a 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 fourth lengthening is required it can be done around age 16 years. Patients may also have lengthenings performed later in life as an adult. One severe case treated by the author equalized a 25-cm discrepancy with two lengthenings over a nail. If the tibia is also short, a combined femoral and tibial lengthening can allow for greater total lengthening amount in a shorter time frame. However, a combined lengthening may not be able to achieve as much total length as two separate lengthenings, and growth inhibition in the tibia has been reported [62]. In addition, with the advent of smaller and more reliable implantable nails, in the author's experience, internal lengthening can be started as early as 7 years of age.

Step 3: Hemiepiphysiodesis and Epiphysiodesis

Contralateral epiphysiodesis around the knee is used as an adjuvant method to equalize LLD. It should be calculated into the total strategy of equalization surgeries, and can be used for up to 5 cm of LLD equalization. Judicious use in some cases may avoid the need for one lengthening. For example, a predicted discrepancy of 12 cm may instead be treated with only one 7-cm lengthening and a 5-cm epiphysiodesis around puberty. Calculation of the timing of epiphysiodesis can be achieved quickly and accurately using the multiplier method [26].

Ipsilateral hemiepiphysiodesis is very useful to correct the valgus deformity of the knee from distal femoral or proximal tibial origins. The 8-plate, developed by Peter Stevens, or a similar device, is a simple way to temporarily arrest the one side of physeal growth [63, 64]. Correction of the valgus deformity of the femur permits future implantable lengthening of the femur since there is no angular deformity.

Box 22.6. Planning Surgery for the CFD Patient

- Determine the classification and total discrepancy of the deficiency to plan out a timeline of surgeries.
- Have a low threshold to perform preparatory surgeries of the hip and knee to prevent subluxation or dislocation, which can be disastrous.
- Do not lengthen until the femoral neck has ossified. BMP can be used to promote ossification.
- The fascia lata must be released or resected before lengthening.
- The safe range of each lengthening is 5–8 cm.
- The Rule of 4: First lengthening at 4 years of age and then every 4 years thereafter as needed.

  Contralateral epiphysiodesis can be used to avoid an extra lengthening.

The Superhip Procedure

Is the proximal femur bony deformity (see Fig. 22.1) caused by the hip joint contractures, or vice versa? The net effect in the frontal plane is that the insertion of the abductor muscles (gluteus medius and minimus) on the greater trochanter is abnormally close to the pelvis. This leads to several problems, including impingement of the trochanter on the iliac bone and contracture of the gluteal muscles since the distance between their origin and insertion is short. The gluteus maximus and fascia lata, with its iliotibial band extension to the tibia, are the most lateral of the soft-tissue structures and therefore contribute the greatest to the abduction contracture of the hip. They also contribute significantly to flexion contracture of the hip. The abduction contracture is not obvious, since hip adduction is preserved due to the femoral bony varus deformity, and hip abduction is limited by iliotrochanteric impingement. If the bony coxa vara is corrected by osteotomy, without soft tissue releases, the abduction contracture will be uncovered and 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 LLD appear less than before surgery. In the face of an open growth plate or a non-ossified neck or subtrochanteric segment, as in type 1b cases, the abduction contracture leads to recurrence of the coxa vara after osteotomy. The mechanism for this recurrence may be differential growth of the physis, bending at the non-ossified tissues, or slipped capital femoral epiphysis.

To correct these deformities, a three-dimensional osteotomy combined with a series of selective soft tissue releases was developed by Paley in 1997. It is now known as the *SUPER hip procedure* because of its complexity.

Box 22.7. The Evolution of the Superhip Procedure
Dror Paley, MD

The Superhip name originally arose as a billing code to avoid writing down the multiple CPT codes that comprised this multistep conglomerate procedure. While there was no intention to call this operation the Superhip procedure, the name stuck. To avoid the name being misperceived the SUPER prefix was then made into an acronym: Systematic Utilitarian Procedure for Extremity Reconstruction. Other SUPER joint reconstructive procedures for congenital knee and ankle deformities developed by Paley were subsequently renamed the SUPER knee and SUPER ankle procedures. John Birch of Dallas has expressed an aversion to the SUPER prefix and refers to these

(continued)

Box 22.7. (continued)

procedures as Paley hip, knee, and ankle procedures. As I learned from my mentor Dr. Robert Salter from his “no name no fame” comment regarding the innominate osteotomy now called the Salter osteotomy, it is not for a surgeon to name this procedure after themselves. I have given this procedure a generic name. If others choose to name it after me, then so be it.

My understanding of CFD began with my experience at the Hospital for Sick Children, Toronto, in 1984. I worked with Drs. Robert Gillespie and Ivan Krajbich, who treated the severe CFD cases by rotationplasty [49]. In 1986 and 1987, I witnessed reconstructive options for these deformities in Kurgan, USSR. Drs. Popkov and Maltzev showed me how to do pelvic support osteotomy to bypass the tethered and deformed hip. The message of these experts was that the CFD deformed hip did not lend itself to anatomic reconstruction.

After beginning my practice in 1987 at the University of Maryland, I tried to find alternative reconstructive solutions to the CFD hip. I performed many valgus, extension, and internal rotation osteotomies on young children with this deformity. Initially, they had excellent bony correction with reduction of the leg length difference due to a fixed pelvic obliquity that developed after surgery. Gradually, I watched the deformity recur, presumably through their proximal femoral physis or unossified femoral neck. I realized that the reason for this recurrence was the hip flexion and abduction contractures that existed prior to the surgery. In 1996, I recognized that in order to succeed, I needed to untether the proximal femur from the hip abductors and flexors and then realign the proximal femur to a neutral position at the hip joint. This was a radical concept, and I was left with the challenge of how to safely release the hip abductors. Borrowing from the concept of the Hardinge approach to the hip, which took down the anterior third of the hip abductors as a sleeve with the quadriceps muscle, I decided to take down the entire hip abductors with the vastus lateralis as one sleeve. In 1997, I performed the first such procedure.

While the psoas, rectus, and TFL were all lengthened as they are today, treating the hip abductors by releasing the gluteus and vastus tendons off of the greater trochanter together turned out to be a bad idea. While this freed up the proximal femur to rotate into neutral by extension, adduction, and internal rotation of the hip joint, it also changed the muscle-tendon length ratio, thus permanently weakening the hip abductors.

(continued)

Box 22.7. (continued)

This is discussed in more detail below. I initially fixed the femur using a Rush rod and a tension band wire. There was no fixation up the femoral neck. When this procedure was performed for a subtrochanteric type Paley 1b or for a Paley 1a, no recurrence of the deformity occurred since the delayed ossification part was resected. When it was performed for a neck type Paley 1b, the varus usually recurred and the neck did not ossify.

In 2001, I switched to using a fixed-angle plate. I used the 130° sliding hip screw (Smith and Nephew, Memphis, TN). Some of the cases ossified, but most did not. Some of the plates broke and the deformity recurred, while in others the plate began to cut through the head as the varus recurred. I decided that a blade plate would be the best implant to avoid cutout as well as to control flexion and extension forces. With the engineers at Smith and Nephew, I designed an infant and pediatric 130°-angle cannulated blade plate, to correspond to the normal neck-shaft angle. Since 2004, I have been using this new plate. This minimized recurrent deformities, but incomplete ossification of the femoral neck was the usual result.

It was apparent that while we could correct this complex deformity, we could not get the delayed ossification of the cartilage to ossify. To get some of these failed cases to ossify, I resorted to insertion of BMP up the femoral neck into the non-ossified cartilage. The result was dramatic: the recalcitrant-delayed ossification cases ossified. Since BMP-2 (Infuse, Medtronic, Memphis, TN) was not FDA approved to use in children, we were initially hesitant and reserved in its application. We only used it to salvage previously failed Superhips. With the unexpected success that we saw from such application, I decided to apply it to new cases in 2006. The results were equally remarkable. All of the necks ossified and there were almost no recurrent deformities. Clearly, we needed to combine a mechanical with a biologic solution to solve the CFD deformity puzzle.

The soft tissue part of the operation also went through an evolution to its present state. In the first 5 years, we not only employed release of the conjoint tendon off of the greater trochanter to treat hip abduction contracture, but we also released the hip capsule off of the greater trochanter. This was done in an extra-articular fashion so that the joint was never exposed. No initial consequence of this was observed. However, when we started to lengthen femurs that had previously had a Superhip procedure, we encountered two new complications: hip dislocation and slipped capital

(continued)

Box 22.7. (continued)

femoral epiphysis (SCFE). In retrospect, we realize that both were related to the release of the superior capsular ligament from the pelvis to the greater trochanter. Our selective release of the superior capsular ligament demonstrated the importance of this band. This superior capsular pelvic-trochanteric band is essential to prevent the femoral head from moving laterally relative to the acetabulum. After recognizing this, we stopped releasing this band and we no longer experienced dislocations or slips following a Superhip.

After 10 years of performing the Superhip, we had solved the recurrent varus deformity problem, the delayed ossification problem, and the dislocation/SCFE problem. We noticed, however, that the children walked with a marked lurch or Trendelenburg gait. We attributed this to the abductor tendon release. By detaching the hip abductors from the greater trochanter, we had changed the muscle-tendon length ratio of the gluteus medius and minimus muscles, which was difficult to recover. Essentially, we had added tendon length to the glutei due to the quadriceps tendon moving proximally. To solve this problem in 2008, I began to perform the abductor muscle slide off of the ilium instead of the distal tendon lengthening. This preserved the muscle-to-tendon length ratio, keeping the muscle tendon length and tension constant after surgery. The slide is achieved by shortening the height of the iliac crest. As an added benefit, the glutei are a peculiar group of muscles since they have a growth plate connected to them that grows in a direction that lengthens the muscles. After the repair of the abductor slide, the iliac apophysis can increase the tension and re-elevate the origin of this muscle. Since adding the abductor slide, patients do not have the previously noted lurch or Trendelenburg gait.

Superhip Surgical Technique (Figs. 22.7, 22.8, 22.9, 22.10, and 22.11)

1. *Positioning, prepping, and draping*. An epidural is placed by the anesthesia service with a catheter running up the back on the non-operative side. A Foley catheter is placed and also routed to the non-operative side. The patient should be moved to the edge and foot of the radiolucent table in a supine position. The ipsilateral arm should be appropriately padded and placed across the patient's chest. A radiolucent bump (usually a folded towel or sheet) is placed beneath the ipsilateral ischium to roll the pelvis 45° towards the opposite side.

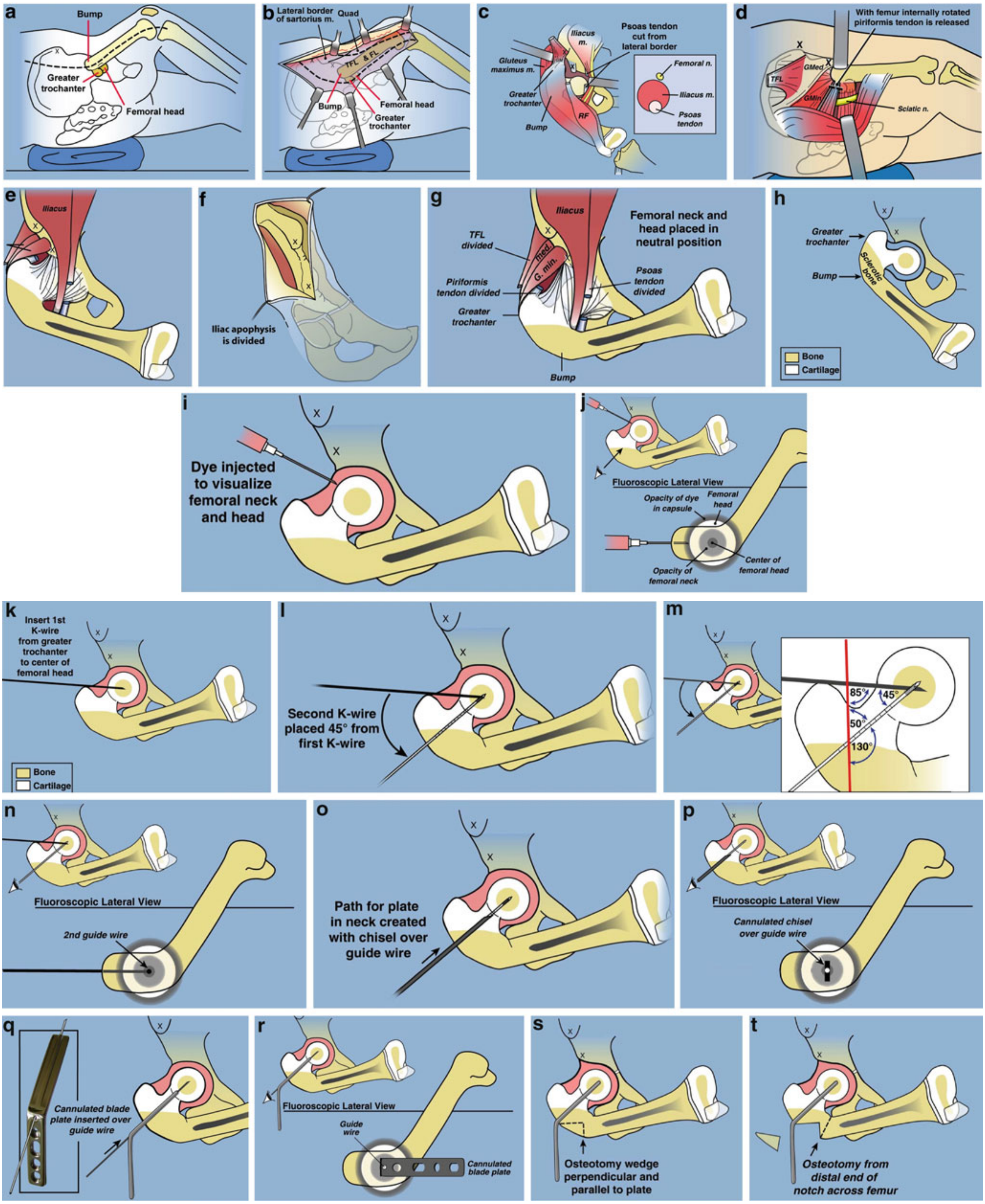

Fig. 22.7 Superhip surgical technique illustrations. (**a**) Straight midlateral incision from top of iliac crest to below tuberosity of knee. (**b**) Reflect fascia lata-iliotibial band distally and leave attached at Gerdy's tubercle. (**c**) Release rectus femoris tendon, decompress femoral nerve, and recess iliopsoas tendon. The TFL muscle remains attached to the ASIS. (**d**) Decompress sciatic nerve and release piriformis tendon at greater trochanter. (**e**) Adduction is limited by hip abductor muscles. (**f**) Split apophysis and do abductor muscle slide. (**g**) All extra-articular tethers released. Hip can be placed in neutral position. (**h**) The quadriceps muscle is elevated to expose the lateral and anterior femur. (**i**) Hip arthrogram to visualize femoral head. (**j**) Lateral view shows concentric rings of ossific nucleus, femoral neck, and femoral head, which should be positioned in a bull's-eye pattern. (**k**) Insert first guide wire from tip of greater trochanter to center of femoral head. (**l**) Insert second guide wire at 45° to first. (**m**) Geometric rationale behind 45° angle. Neck-shaft angle is 130° and medial proximal femoral angle is 85°. Therefore the angle between these must be 45°. (**n**) The second guide wire is centered in the femoral head and neck on both views. (**o**) Insert a cannulated chisel up the femoral neck. (**p**) The chisel should be perpendicular to the posterior aspect of the greater trochanter.

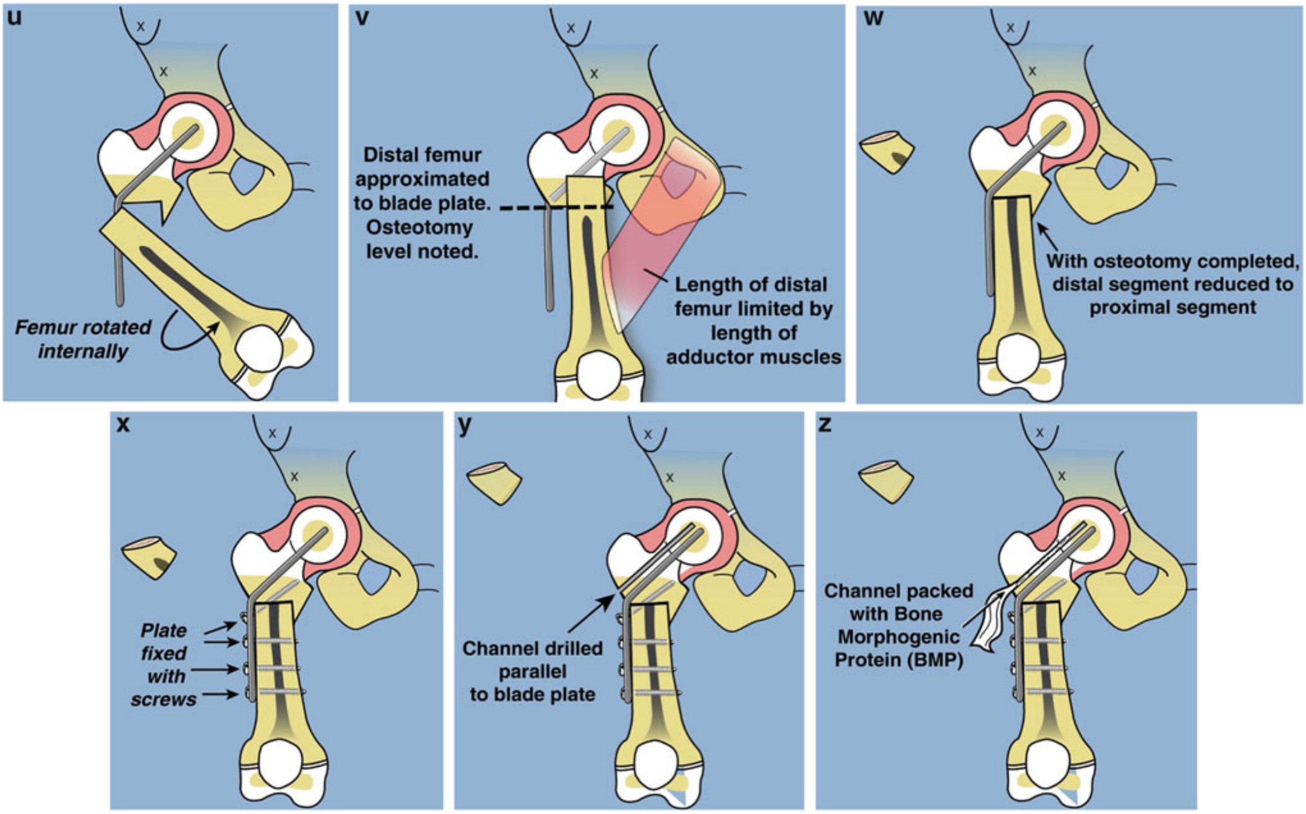

Fig. 22.7 (continued) (**q**) Replace the chisel with a 130° blade plate. (**r**) The blade plate is in place. Note the flexion deformity of the femur shaft relative to the plate. (**s**) Cut perpendicular and parallel to the plate. (**t**) Perform a second osteotomy oblique to the femur. More recently this osteotomy starts inside the notch that was removed eliminating the medial lip. (**u**) Rotate the distal femur internally and abduct it to realign it with the proximal femur. (**v**) Overlap the bone ends and mark the level of overlap. The femur cannot be reduced due to the tethering medial and posterior soft tissues. (**w**) Perform a shortening osteotomy of the femur and reduce the femur to the plate. (**x**) Fix it with three diaphyseal screws and one interfragmentary neck screw. (**y**) Drill a channel parallel to the blade. (**z**) Insert BMP-2 collagen sponges up the femoral neck

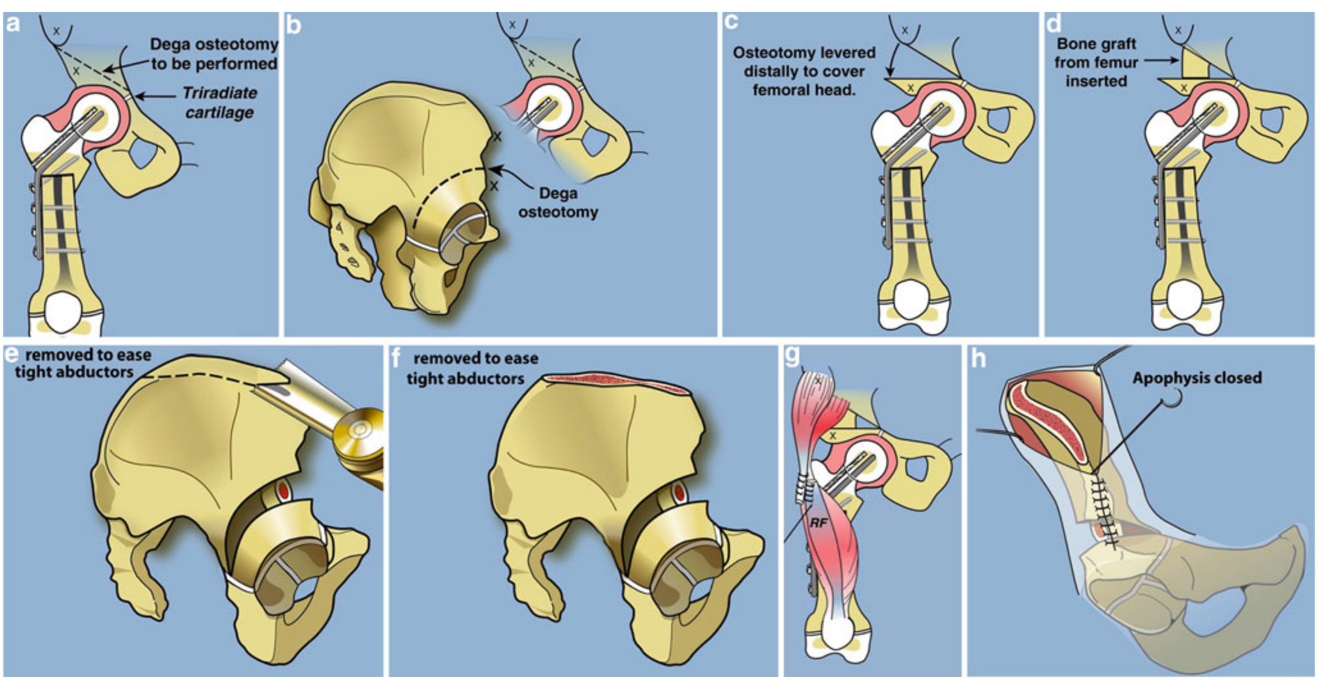

Fig. 22.8 Paley modification of Dega osteotomy. (**a**) The cut should start 2–3 cm proximal to the lateral acetabulum converging on the triradiate cartilage medially. (**b**) The cut should be parallel to the acetabulum circumferentially stopping at the triradiate between the ilium and the ischium. (**c**) The roof of the acetabulum is levered down to cover the femoral head such that the sourcil is horizontal. (**d**) Fashion the bone resected from the femur to fit into the opening wedge space. (**e**) Resect the top of the iliac wing to reduce the tension on the hip abductors so that the apophysis can be sutured closed. This is part of the abductor slide. (**f**) After the resection. (**g**) Transfer the rectus femoris to the TFL. (**h**) Close the apophysis

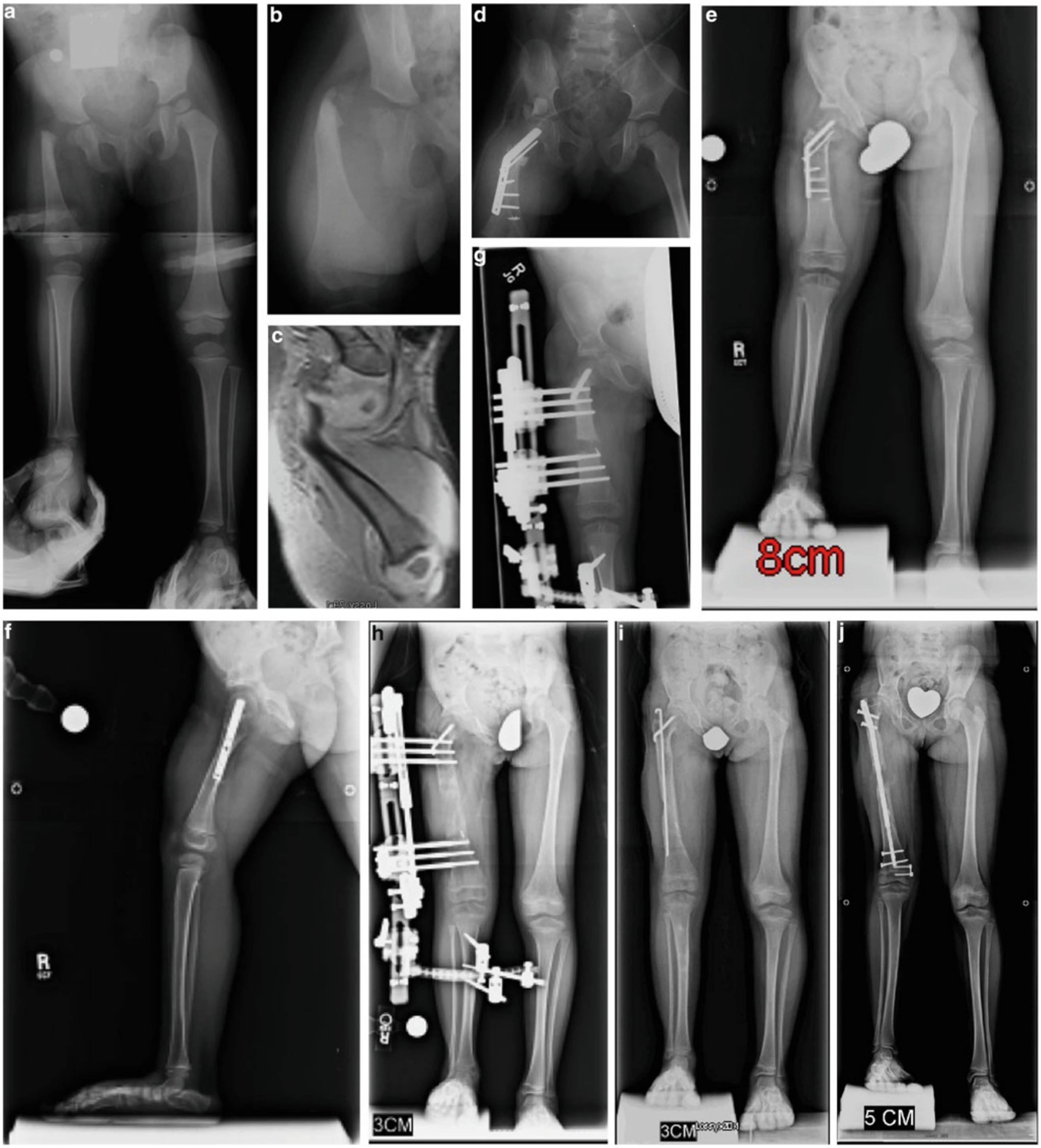

Fig. 22.9 CFD Paley type 1b with delayed ossification of femoral neck (**a**, **b**). MRI showing the cartilaginous femoral neck that remains unossified at 2 years of age (**c**). Superhip with Superknee procedure at 2 years of age, including insertion of BMP in femoral neck (**d**). The neck is fully ossified by 3 years of age. Note the significant growth that has occurred since the year before (**e**, **f**). First lengthening performed at age 4 years with Smith & Nephew Modular Rail System (Memphis, TN) external fixator with articulation across the knee joint (**g**). Eight centimeters of lengthening achieved (**h**). Removal of external fixator with Rush rodding of bone to prevent fracture (**i**). At the age of 8 years she underwent a second lengthening, this time using the PRECICE (Ellipse Technology, Irvine CA) 8.5-mm-diameter lengthening nail (**j**)

Fig. 22.10 Two-year-old girl with CFD Paley type 1b with delayed ossification and severe angulation of the subtrochanteric level of the left femur (**a**). MRI showing the delayed ossification of subtrochanteric region. The neck is normally ossified (**b**). The deformity is fully corrected and the femur is healed after the Superhip surgery. The knee has full extension following posterior capsulotomy and Superknee stabilization. This X-ray is 1 year after the Superhip and Superknee showing significant growth and remodeling which is why the screws are protruding medially (**c**). Lengthening of the femur was performed at age 4 years (**d**). X-rays after lengthening of the femur 6 cm and insertion of Rush rod at the time of removal surgery (**e**)

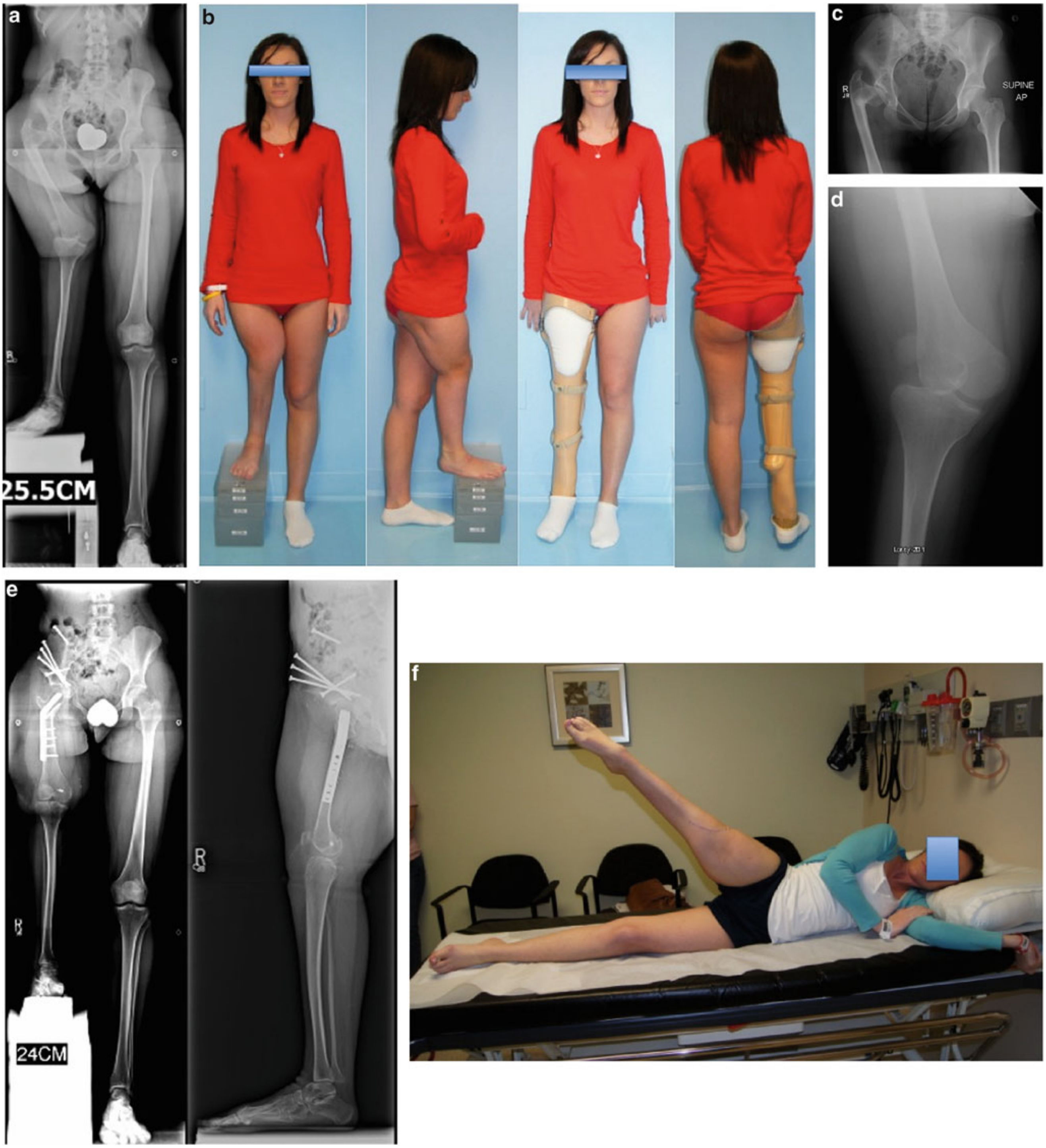

Fig. 22.11 (**a**) Erect leg radiographs of a 22-year-old woman with previously untreated CFD and 25-cm discrepancy. She has marked coxa vara and acetabular dysplasia as well as rotatory subluxation of the knee joint and dislocation of the patella. (**b**) Clinical picture before any surgery with and without her prosthesis. (**c**) AP pelvis X-ray showing the hip deformities. (**d**) AP knee X-ray showing the rotatory subluxation of the knee. (**e**) AP and lateral radiographs after Superhip procedure with Ganz periacetabular osteotomy (PAO) and Superknee procedure for reduction and stabilization of the knee and patella. (**f**) Active hip abduction after recovery. (**g**) Active hip flexion and knee extension after recovery. (**h**) Ganz PAO modified for Superhip procedure. Start by osteotomizing the iliac crest and peeling off the abductor muscles to do an abductor muscle slide. (**i**) The five bone cuts of the Ganz seen from the lateral side. (**j**) The acetabulum is internally rotated to gain posterior coverage. (**k**) The acetabulum is then abducted to gain lateral coverage. (**l**) The iliac wing is shortened, and then the crest is reattached using the same screws that are used to fix the PAO, 3 antegrade and one retrograde. (**m**) Radiograph after first lengthening of femur and tibia (6-cm femur and 5-cm tibia). The femur and tibia were both rodded using Rush rods at the time of fixator removal to prevent fracture

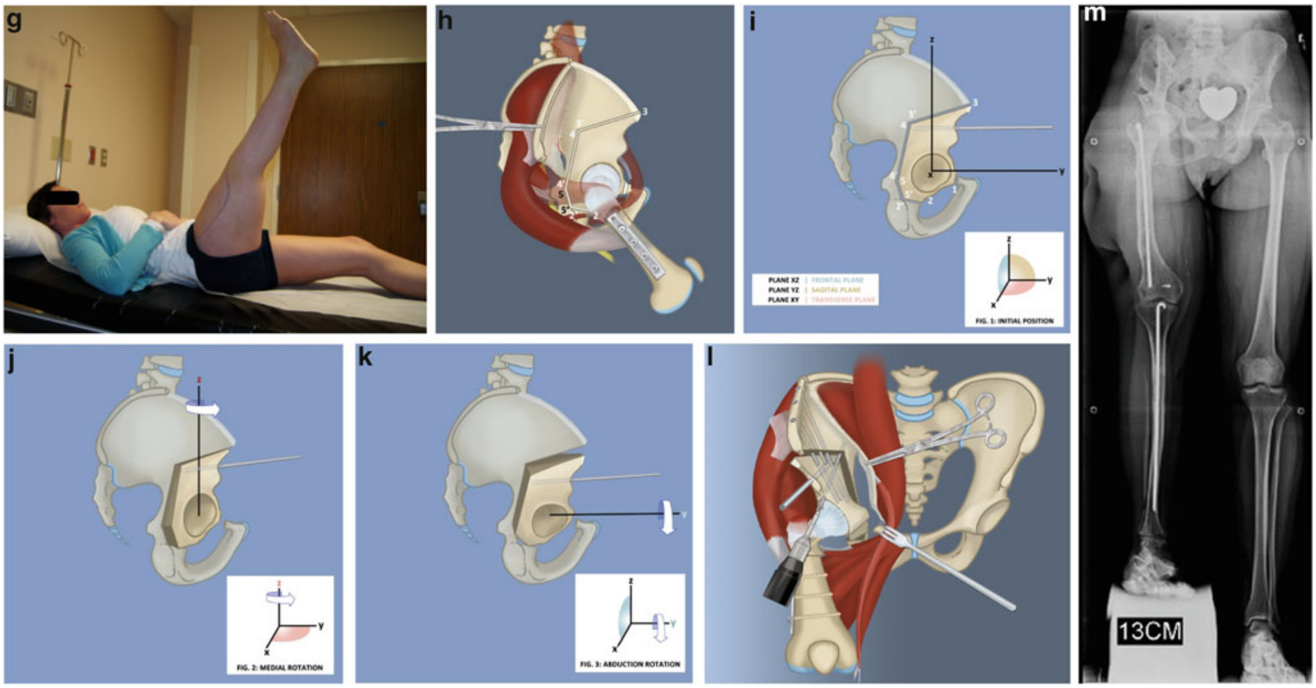

Fig. 22.11 (continued)

The bump should not be beneath the iliac crest or lower back. The entire side should be prepped and draped free from the nipple to the toes. The drapes should extend from the crack of the buttocks to the fold between the scrotum or labia and the thigh. The lower limb should be completely free of the drapes. This is called a forequarter prep.

2. *Incision.* With the leg fully extended, a long mid-lateral incision is made from the top of the iliac crest to the tibial tuberosity. The incision is kept as straight as possible, passing over the proximal femoral "bump" and continuing longitudinally towards Gerdy's tubercle before curving slightly anteriorly towards the tibial tubercle. The incision is carried down to the depth of the underlying fascia lata and iliotibial band.
3. *Flap elevation.* The subcutaneous tissues and skin are elevated as one large flap anteriorly and posteriorly off the fascia of the thigh and pelvic region. The fat is adherent to the fascia and should be dissected preferably with an electrocautery. The electrocautery should be held flat, parallel to the plane of dissection, and can be quite technically difficult. It is important not to incise or damage the fascia if it is being used for knee ligament reconstruction. Dissection may also be carried out with scissors. Anteriorly, the flap is extended medial to the Smith-Peterson interval (tensor fascia lata (TFL) and sartorius) proximally. Posteriorly, the subcutaneous flap is elevated to just posterior to the intermuscular septum. 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 anterior edge of the TFL to the mid-gluteus maximus proximally.
4. *Fascia lata release.* 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.

If knee ligamentous reconstruction is planned, the fascia lata is cut proximally and anteriorly at the musculotendinous junction. The fascial cut should be a step cut or 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's 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, and it may be difficult to differentiate the muscle fibers. The distinguishing feature is that the GMed fibers insert on the greater trochanter while the TFL does not. The distal fascia lata becomes the iliotibial band and blends with the underlying lateral knee capsule, which may be partially reflected with the iliotibial band. The fascia should be mobilized all the way until Gerdy's 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.

5. *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 retract 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 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. The release of the gluteus medius and minimus muscles is accomplished by the abductor muscle slide technique (see step 7). If the anterior thigh or sartorius fascia is still tight, it can also be released, taking care not to injure the lateral femoral cutaneous nerve, which should be identified and decompressed. It runs inside the fascia covering the sartorius muscle just medial to the anterior superior iliac spine.
6. *External rotation contracture release*. 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 1 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.
7. *Abductor muscle slide.* The abductors may not appear to be tight on first inspection because of the coxa vara. Adduction into a true AP of the hip, with the neck oriented normally in the acetabulum, is now 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. As noted previously, the abductors should be detached at their origin and not their insertion. This avoids changing the muscle-tendon length ratio and avoids weakening the hip abductors, avoiding a lurch or Trendelenburg gait.

The subcutaneous tissue flaps should be elevated to provide adequate exposure to the iliac crest apophysis. There is a tendency to have inadequate posterior exposure, and the flaps should be elevated just beyond the highest lateral point of the apophysis. The anterior extent is the anterior superior and inferior iliac spines, which has been exposed for the release of the rectus femoris tendon. The abdominal external oblique muscles are partially released off of the entire length of the apophysis. Once the apophysis is bare, it can be split from the ASIS to the AIIS, and then split from anterior to posterior along the iliac crest. This should be done with a #15 blade. To know where to split, pinch the apophysis between thumb and index finger of the hand not holding the knife. Stay in the middle of the apophysis along its entire length, pushing 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. The medial half of the apophysis is reflected medially with the iliacus muscle. Since some of the abductors act as flexors of the hip, the abductor slide helps eliminate any remaining flexion deformity of the hip. Furthermore, the iliacus muscle slide also relaxes any residual tension in this muscle.

8. *Elevation of quadriceps.* The quadriceps is 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. The perforator vessels need to be cauterized as the quadriceps is detached from the linea aspera. Proximally, the vastus lateralis should be elevated off of part of the cartilage of the greater trochanteric apophysis by sharp dissection.
9. *Arthrogram.* A hip arthrogram is now performed using a 20-gauge spinal needle. With the trocar inside, the needle is placed into the hip joint from the anterolateral side. Traction of the femur may facilitate placement. Once the needle appears to be in the joint on the image intensifier, the trocar is removed and normal saline is injected. If the needle is in the joint the saline should go in with little pressure, and when the syringe is removed from the needle, the saline should drip back out. These signs confirm that the needle is in the joint space. The arthrographic dye can now be injected into the hip joint to outline the femoral head, acetabulum, and femoral neck.
10. *Guide wire 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 across the opposite leg. A guide wire 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 guide wire 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 guide wire is inserted from the tip of the greater trochanter to the center of femoral head. Since the tip of the trochanter is cartilaginous in young children, it cannot be seen radiographically and 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 of three concentric circles is seen. This is formed by the overlapping dye shadows: the outermost circle is the femoral head, the middle circle is the femoral neck, and the inner circle is the ossific nucleus of the femoral head. A second wire should be drilled into the center of the bull's-eye at a 45° angle to the first wire. Using a depth gauge or 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. The position of the neck wire can also be confirmed by flexing the hip to 90° and looking at a frog leg lateral of the neck.
11. *Blade plate insertion.* The cannulated chisel for the blade plate should now be hammered up the femoral neck guided by the second guide wire. The chisel should be rotated until it is perpendicular to the posterior aspect of the greater trochanter. This will guide it to the correct angle in the sagittal plane. Tap the chisel out of the femur and reinsert the guide wire in its previous position. Insert the appropriate length 130° blade plate over 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 planes, as well as using the approach-withdrawal technique with live fluoroscopy to ensure that the blade is not advanced too close to the articular surface of the femoral head. If the blade is suspected of being too long, then replace it with one with a shorter length. Furthermore, if plate placement is off center, there is greater risk of protrusion into the joint.
12. *Subtrochanteric osteotomy.* The femur should be cut perpendicular to the shaft of the plate, starting just below the bend in the plate. This cut should be below the level of the greater trochanter cartilage. To guide this cut, drill a wire perpendicular to the plate. Keep the plane of the saw blade perpendicular to the plate in all planes. The width of the perpendicular cut surface should be as wide as the femoral diaphysis. If the deformity angle is very large, it may be parallel to the lateral cortex. A second subtrochanteric osteotomy should be made oriented less than 90° to the first osteotomy towards the base of the femoral neck to remove the bone protruding medially. The femoral head and neck can be manually tested for any residual impingement, removing any blocks to achieving 90° of hip flexion. The anteromedial corner often needs to be excised to prevent impingement.
13. *Periosteum release.* After the second osteotomy, the distal femur is easily exposed and the surrounding periosteum is peeled off circumferentially. Medially, it is very thick and restricts correction of the varus and rotation deformity. Cut the periosteum transversely around the femur, carefully separating it from the surrounding muscle. Be careful to avoid injury to the profunda femoris and its perforators, which pass immediately under the periosteum. Cutting the periosteum allows the thigh to stretch longitudinally, reducing the amount of shortening required of the femur.
14. *Femoral shortening.* The distal femur is now mobile and can be corrected into valgus 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,

and a mark should be made at the point of overlap. The distal femur should be shortened at this level. A wire is drilled perpendicular to the femur at the level of the osteotomy and a saw is used to cut, using frequent irrigation to prevent heat necrosis. The segment of bone is kept moist on the back table for use as bone graft for the Dega osteotomy. This is usually a trapezoid-shaped piece about 1–1.5 cm long.

15. *Distal femur fixation.* The femur is now brought to the plate. The bone ends should oppose without tension. The femur is internally rotated to correct the external torsion deformity. To adjust the femur to the correct anteversion, the guide wire should be reinserted into the cannulation of the plate to show 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 with screw insertion. Two more screw holes are drilled and screws are inserted. The most proximal hole in the plate is designed to drill parallel to the blade of the plate and secures the plate to the proximal femur. The wire in the cannulated hole can be used to guide the drill bit. In type 1b cases, the blade of the plate and the oblique screw goes across the proximal physis and into the femoral head. 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.
16. *BMP insertion.* 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 guide wire in the cannulation of the plate. A 3.8-mm hole is then drilled over this guide wire. The drill hole should extend all the way into the ossific nucleus. BMP-2 (Infuse, Wright Medical) is then prepared on collagen sponges that are manually pushed into the hole with a 3.2-mm drill and forceps. A more recent method of insertion of the BMP is to use a cannula, such as the Craig needle biopsy tool, which is 4 mm in its outer diameter. The BMP sponge is labeled by applying radiographic contrast dye onto its surface. The sponge is then inserted into the end of the cannula, and the cannula is inserted into a 4-mm predrilled hole in the femoral neck. Once the cannula is located at the desired depth, a blunt trocar is used to push the sponges out of the cannula into the femoral neck. Since the sponges are labeled by contrast dye, they can be seen radiographically after the cannula is removed. It should be emphasized that such use of BMP (Infuse implant) is an off-label use of this product.
17. *Pelvic osteotomy*. Surgical technique is covered later in this chapter. The type of pelvic osteotomy depends on the age of the patient and the degree of dysplasia. In most patients under age 6 years, the Dega osteotomy is preferred. In hips with severe dysplasia, a periacetabular triple osteotomy or a Ganz periacetabular osteotomy is used depending on the closure of the triradiate cartilage. With both periacetabular osteotomies, the iliac wing shortening osteotomy should be performed prior to fixation of the acetabular fragment. Consider doing the pelvic osteotomy prior to shortening the distal femur segment, as the amount of shortening needed may change after the pelvic osteotomy. Furthermore, the pelvic osteotomy changes the rotation of the hip, since it rotates the capsule. Thus, the amount of rotation changes if the femur fixation is done after the pelvic osteotomy.
18. *Iliac wing osteotomy.* 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. Putting traction on the lateral apophysis, one can see and mark the level to which it reaches. A saw is used to cut and remove the proximal part of the iliac wing, effectively a shortening osteotomy. The removed bone is used as graft for the acetabular osteotomy or the subtrochanteric femoral osteotomy. The apophysis is then repaired with a running #1 Vicryl suture. The medial and lateral halves of the apophysis should be well opposed to avoid a bifid iliac wing. The external abdominal muscles should be advanced and repaired over the apophysis after the closure.
19. *Muscle repairs and transfers.* The TFL muscle is repaired to the rectus femoris tendon. This essentially elongates the rectus femoris while allowing it to maintain its origin on the pelvis. The interval between the TFL and the sartorius is closed, carefully avoiding the lateral femoral cutaneous nerve. The quadriceps is sutured to the region of the linea aspera. Finally, the gluteus maximus is advanced back to the posterior border of the TFL.
20. *Closure.* If no knee releases or reconstruction is required, the fascia lata should be resected from Gerdy's tubercle. The wound can now be closed. Since there is no fascia lata, the deepest layer is the subcutaneous fat layer, called the "underlayer." One or two medium-sized drains are placed, exiting anterosuperiorly. The second drain is laid medially if a Superknee procedure is performed. The drains are usually secured with a clear adhesive sterile dressing (e.g., Tegaderm, 3 M, Minnesota).

It is important to close the wound in a fashion that the opposite layers get sutured at the same level. The deep edges of the subcutaneous underlayer are brought together with a running #1 braided absorbable suture. The Scarpa's fascia is closed with a running 2-0 braided absorbable suture. The deep dermal layer is closed with a running 3-0 braided absorbable suture, and the skin is closed using a subcuticular 4-0 monofilament suture. Dermabond™ may be used, and sterile dressings are applied.

Final radiographs are taken: an AP pelvis and lateral knee, both including the femur. The patient is then placed into spica cast. The operative limb should be placed in full hip and knee extension with the foot and ankle left free. The opposite limb can be in a flexed, abducted, and externally rotated position, with the cast stopping short of the knee. Alternatively, a one-leg spica with a molded pelvic band can be used. The cast should be bivalved before leaving the operating room.

Box 22.8. Superhip Tips and Tricks

- Pay careful attention to soft tissue handling, making sure that the subcutaneous flaps are cleanly dissected to preserve their integrity and prevent fat necrosis.
- The lateral femoral cutaneous nerve is usually found just under the sartorius fascia and originates from deep to the inguinal ligament.
- The femoral nerve lies on the anteromedial side of iliopsoas muscle. The tendon lies posteromedial.
- Do not dissect distal or deep to the piriformis tendon to avoid interrupting the inferior gluteal anastomosis with the medial femoral circumflex artery.
- The apophysis should be split with a single continuous cut, pushing hard down to bone. Multiple passes will piecemeal the apophyseal cartilage.
- Palpate the back of the greater trochanter while placing the chisel and blade plate to make sure that they stay perpendicular to its posterior border in the sagittal plane.
- The extent of the flexion and adduction deformity should be fairly evident by the orientation of the blade plate to the uncut femur. Greater than 45° of flexion deformity and up to 90° of varus deformity are not uncommon.
- Place the guide wire in the cannulated plate to help guide the rotational correction and BMP hole placement, and determining femoral anteversion.

Postoperative Course

Parents are initially educated on transfers, cast care, and hygiene. The bivalved spica cast can be converted into a removable cast in about 1 week. The patient can then start gentle passive flexion and extension of the hip from 0 to 90°, as well as passive abduction. The spica cast is discontinued after 6 weeks, and if radiographs show adequate bony healing, then the patient is progressed to full weight bearing, gait training, strengthening, and active range of motion. The end goal is to restore the child to normal function before they proceed with limb lengthening.

Pelvic Osteotomies

Paley Modification of Dega Osteotomy (Paley-Dega Osteotomy) (See Fig. 22.8)

In the Superhip procedure, the iliac apophysis has already been split and the periosteum from the lateral and medial aspects of the ilium has already been reflected back with its overlying muscles. The lateral periosteum is reflected back to the edge of the acetabulum and to the sciatic notch. The periosteum should be dissected off the anterior wall of the sciatic notch, feeling for the soft cartilage of the triradiate cartilage where it separates the ilium from the ischium.

Using the image intensifier, a guide wire is drilled approximately 2 cm proximal to the lateral edge of the acetabulum towards the triradiate cartilage medially. On the lateral wall of the ilium, the osteotomy is a curved line running perpendicular to the triradiate cartilage, separating the ilium from the ischium. It runs parallel to the anterior wall of the sciatic notch and extends proximally to the level of the guide wire. The cut curves to make a 90-degree bend heading towards the superior border of the anterior inferior iliac spine. In the frontal plane, the osteotomy is inclined from proximal to distal, parallel to the guide wire. The osteotomy ends at the triradiate cartilage medially. It is easier to start distally and work proximally up to the guide wire. At the most posterior distal part of the osteotomy, it is important to stay very near the sciatic notch to avoid entering the hip joint inadvertently.

The osteotomy is levered distally to bring the roof of the acetabulum down. A laminar spreader is used to distract the osteotomy. The vertical limb of the osteotomy extending down to the triradiate cartilage at the ischium allows for greater bending and greater lateral coverage. It is a myth that the Dega osteotomy gives posterior coverage. It cannot do so because it cannot extend distal or posterior than the posterior limb of the triradiate. The original Dega, as well as subsequent publications of this osteotomy, goes through the inner

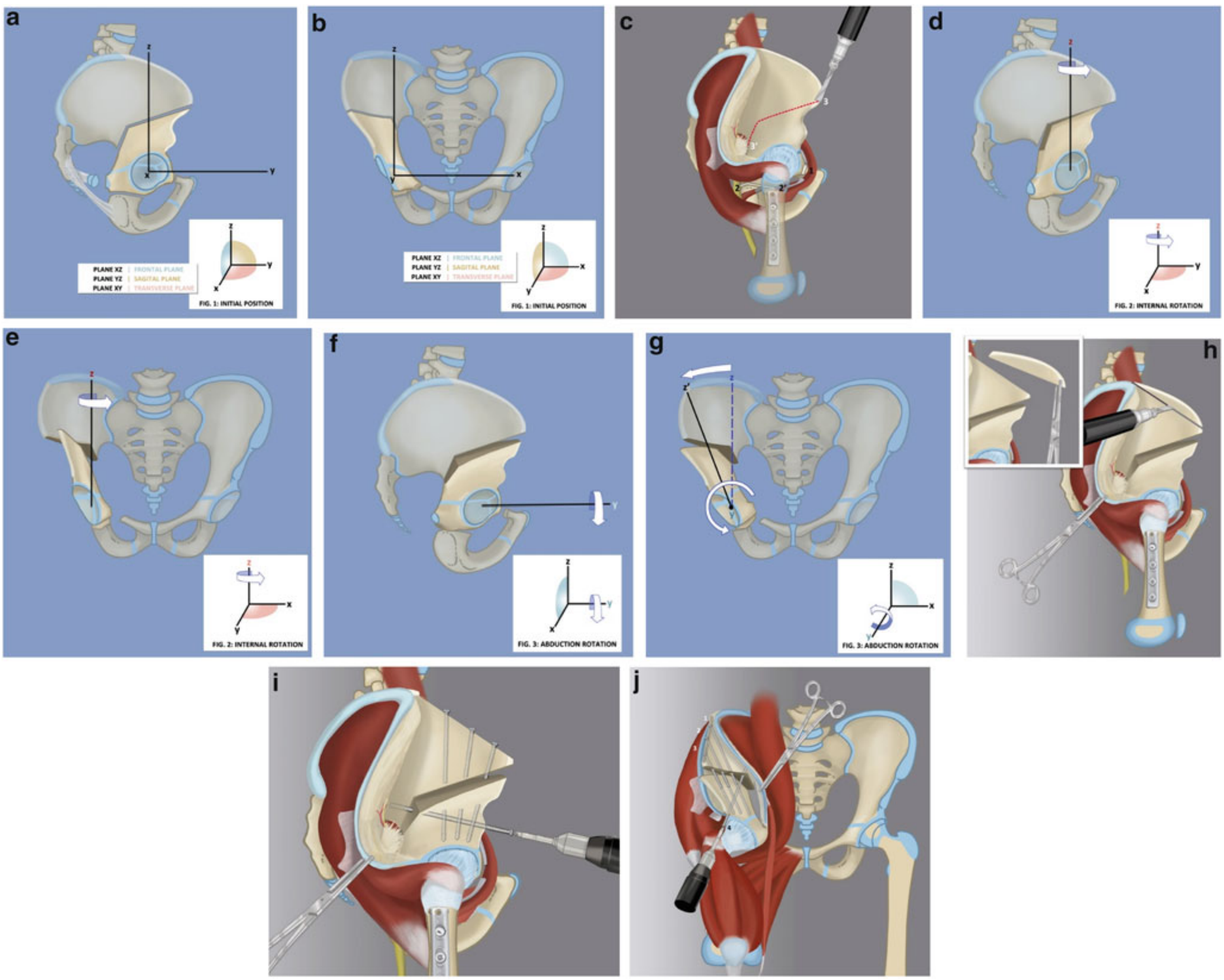

Fig. 22.12 (**a**) Periacetabular triple osteotomy lateral view (Paley). The bone cuts are outlined. The sacrospinous ligament is also released to untether the acetabulum. (**b**) Periacetabular triple osteotomy, AP view (Paley). (**c**) The ischium is cut under direct vision after identifying and decompressing the sciatic nerve. The nerve is not disturbed since the bone is deeper. (**d**) Internally rotate osteotomy to gain better posterior coverage and treat the retroversion of the acetabulum. Lateral view. (**e**) AP view of internal rotation of osteotomy. (**f**) Abduct the osteotomy to gain lateral coverage. (**g**) Abduct the osteotomy to gain lateral coverage. (**h**) Resect the upper end of the iliac wing to allow closure of the hip abductors. (**i**) Fixation is with several antegrade and one retrograde screw. (**j**) Front view of coverage and fixation

table of the pelvis, especially anteriorly. The Paley-Dega modification avoids doing so and also extends farther posterior and distal than other publications of the Dega. In this manner, there is true hinging in a three-dimensional way on the triradiate. Even the anterior part of the cut does not exit the cortex and extends to the triradiate junction between the ilium and pubis.

The bump under the buttocks should be removed at this stage. This allows one to assess the coverage of the femoral head in the true frontal plane. The laminar spreader can be distracted as needed to gain additional coverage and reorientation of the acetabular sourcil. The laminar spreader should be kept posterior to avoid increasing the anterior coverage. In this manner, predominantly lateral coverage is augmented.

At this point, the femoral shortening should be carried out and the excised bone segment cut to fit the opening wedge gap of the pelvic osteotomy. Additional bone from the iliac crest, which is resected during the abductor slide, may be inserted to fill the remaining space.

Paley Modification of Periacetabular Triple Osteotomy (Paley PATO) (Fig. 22.12)

1. Pubis: The pubis is exposed subperiosteally by reflecting the medial iliac apophysis and periosteum more medially. The triradiate cartilage arm at the pubis is exposed and the dissection carried out just distal to it. The superior pubic

ramus can be cut under direct vision under the protection of two Hohmann retractors with an osteotome or a saw.

2. Ischium: The ischium is exposed by returning to the back of the femur and finding the sciatic nerve again. The nerve is followed to the ischium. To avoid stretching the nerve, do not place a retractor between the nerve and the ischium. Subperiosteal elevation of the ischial periosteum is carried out. The ischium should be cut under direct vision near the junction where it forms the inferior wall of the acetabulum, distal to the triradiate cartilage.
3. Sacrospinous ligament release: For large corrections, it is important to release the tether of the sacrospinous ligament. This ligament can be palpated after dissecting the medial iliac periosteum off of the quadrilateral plate inside the true pelvis. A finger can be placed on the tip of the ischial spine, and the ligament is then felt. Using proprioception, a finger from the other hand can be used to palpate the finger that is on the ligament. Make sure that the sciatic nerve, which can be visualized laterally, is not in the way. With the ligament isolated in this manner, it can be cut with scissors from the lateral side.
4. Ilium: The ilium is cut with a saw from the anterior superior iliac spine towards the junction of the true and false pelvis. On the lateral side of the ilium, it should be osteotomized towards and into the sciatic notch. On the medial side it should be cut with an osteotome into the sciatic notch. The osteotomy on the medial side has an angle of about 120°. This bend in the cut rounds off the corner of the distal segment and creates a spike of bone on the proximal segment. This increases the surface area and creates a posterior buttress while making the distal segment rotate more easily with less distraction.

To gain posterior and lateral coverage, the acetabular segment should first be rotated internally along the long axis of the body. This gives posterior coverage. It should then be rotated laterally. This gives lateral coverage. To rotate internally, a lion-jaw bone clamp is placed anteroposteriorly, superior to the acetabulum and the bone fragment rotated internally. It can also be placed around the pubis to lever the acetabulum laterally. If the acaetabular fragment does not move sufficiently, then each osteotomy site should be checked for completeness. The ischial osteotomy is often the culprit, and the periosteum around the ischium may need to be divided. Fixation of the triple osteotomy is achieved using a long 3.5-mm or 4.5-mm screw from the ilium to the acetabular fragment. However, because of the abductor slide, the proximal part of the iliac wing must be resected prior to screw fixation. The description of how to do this is described in a later step. Once complete, multiple drill bits can be placed from the remaining iliac wing towards the acetabular fragment, from proximal lateral to distal medial. One drill bit is placed from the anterior inferior spine, retrograde towards the base of the iliac wing from anterior to posterior. A same sized drill bit is used to compare lengths with the drill bits inside the bone to determine the screw lengths. One by one, the drill bits are removed, and replaced with solid, non-cannulated screws, making sure that they do not enter the triradiate cartilage or the hip joint.

Ganz Periacetabular Osteotomy (PAO) (See Fig. 22.11)

The technique for this has been previously published, though there are some variations when performed with a Superhip procedure. This osteotomy is indicated for patients who are skeletally or near-skeletally mature, usually with quite severe acetabular dysplasia.

To do the abductor slide, an osteotomy of the iliac crest is done from lateral to medial. There is no iliac apophysis to be split, so the proximal 1 cm of crest is cut with a saw after first elevating the medial periosteum with the iliacus muscle. The periosteum of the medial side of the ilium is present at every age and dissects off the bone easily, even in older patients. The periosteum on the lateral side of the ilium will not strip off easily after childhood. The glutei fix to bone with Sharpey's fibers, making subperiosteal dissection impossible in adulthood. Therefore, the glutei are not detached from the top of the crest and are stripped laterally after an osteotomy of the crest of the ilium. This includes the TFL. The sartorius and the iliacus muscles are stripped medially. A small cube of bone is cut with a saw and reflected with the sartorius.

The Ganz PAO consists of five osteotomies of the ilium, ischium, and pubis:

1. The first osteotomy is of the pubis. The pubis is exposed subperiosteally and cut with a saw and an osteotome. A sharp Hohmann retractor can be driven into the distal pubis and retracted to provide better exposure.
2. The second osteotomy involves part of the ischium. The ischium is cut from anterior to posterior through half of its thickness. A special angled osteotome is used for this. A space is created just distal to the pubis between the femur and the ischium. The hip is kept partially flexed at all times to help protect the femoral nerve. A blunt instrument can be inserted to feel for the ischium, which is quite deep. The curved osteotome is inserted down to the ischium. It is then cut part way through the bone.
3. The third osteotomy is partway through the ilium. The periosteum should be dissected off of the quadrilateral plate inside the true pelvis. With a finger, the surgeon can feel and find the ischial spine and feel the sacrospinous ligament. An iliac osteotomy is made from just distal to the anterior superior iliac spine to the brim of the true

pelvis. A mark is placed at the junction of the true pelvis with the false pelvis in line with the sciatic notch. An imaginary line can be drawn from this point parallel and just anterior to the anterior wall of the notch, ending at the level of the ischial spine. This osteotomy can be made with a saw.

4. The fourth bone cut is made with an osteotome from the pelvic brim into the true pelvis to end anterior to the ischial spine. To avoid stretch to the femoral vessels the hip should be flexed at all times during this osteotomy. An osteotome is inserted with its lateral part posterior and its medial part anterior. It should be inclined with its handle towards the belly. Either by feel or using the image intensifier, we need to orient this parallel to the anterior wall of the sciatic notch and stop opposite the ischial spine.
5. The fifth and final osteotomy is the completion of the ischial osteotomy. The same curved osteotome used for the anterior ischium is used here to complete the ischial cut. The osteotome is oriented towards the quadrilateral plate. It should be inserted to start at the end of the fourth cut. Once the ischial cut is complete the acetabular segment is mobilized.

A lion-jaw bone clamp is placed on the pubis and the acetabulum is rotated laterally outwards. The segment may need to be internally rotated due to retroversion. If it will not move sufficiently, there is often a tether at the pubis due to the attached muscles. Releasing these frees the acetabular segment to move. Once the segment is placed in the desired position, screw fixation is used to secure it. Again, the wing of the ilium should be shortened first to allow the abductors to slide distally. Three antegrade screws and one retrograde screw (as in the above periacetabular triple osteotomy) are inserted to secure the fragment. The abductors are secured back with screws through the crest segment into the iliac wing.

Knee Considerations in CFD

The knee in CFD may range from a normal, stable, undeformed knee to a contracted, unstable, deformed joint. The most common deformity of the knee is valgus, which is usually nonprogressive. The distal femoral physis is usually closer to the knee joint on the lateral side, often due 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 ACL [27]. The patella is usually hypoplastic and may maltrack laterally. In some cases, the patella dislocates with flexion. Finally, many cases of CFD have a fixed flexion deformity of the knee.

Indications for Preparatory Surgery of the Knee Prior to Limb Lengthening

Isolated anteroposterior instability is not necessarily an indication for reconstructive knee ligament reconstructive surgery in children with CFD. However, grade 3 ligamentous instability with 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, instead of excising the fascia lata, it can be used to reconstruct the knee ligaments. In some children, there is a catching or locking sensation in the knee when going from extension to flexion. It may be painful and require a trick motion to release it. This is due to contracture of the iliotibial band, combined with aplasia of the ACL. In more severe cases, the tibia actually subluxates or dislocates anteriorly on the femur and reduces at about 30° of flexion. In older patients, the posterior aspect of the tibia may be rounded, contributing to anterior dislocation of the tibia on the femur. It is not clear whether this is a secondary change due to chronic dislocation or a primary deformity, since the tibia is not ossified posteriorly in infancy.

Patellar hypoplasia and instability are also very common. The patella frequently maltracks laterally with progressive knee flexion. In some cases, the patella may dislocate with knee 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 ligament deficiency, which lateralizes the patellar tendon insertion. Patellar maltracking or subluxation/dislocation should be corrected prior to limb lengthening.

Flexion contracture of the knee is another congenital deformity that may be present and should be corrected before proceeding with limb 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 is a flexed angle between the anterior cortical line of the femur and tibia in maximum extension. When the contracture is greater than 15°, it should be corrected surgically. Knee flexion deformity can be due to bony or soft tissue causes. In CFD, it is commonly an 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.

Box 22.9. History of Superknee Procedure

Dror Paley, MD

The knee reconstruction that I developed in 1994 [42] 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 were developed by me: (1) the Langenskiöld procedure [65] for congenital dislocation of the patella; (2) the MacIntosh procedure [66] for ACL deficiency including extra and intra-articular ACL reconstruction using the fascia lata; (3) the Grammont procedure [67] for recurrent dislocation of the patella; (4) the Paley procedure also referred to as the reverse MacIntosh [66] to prevent external rotatory instability and to act as an extra-articular posterior cruciate ligament; and (5) the Paley anterior approach to posterior capsulotomy of knee. Combinations of two or more of these five procedures are referred to as the Superknee procedure and can be performed at the same time as a pelvic osteotomy and/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, it can be done through one midline anterior incision or one medial and one lateral incision. If the Superknee is performed without the Superhip procedure, the entire surgery can be performed under tourniquet control. My personal preference is to use the Hemaclear tourniquet (HEMACLEAR, New Jersey) because of its narrow width. There is typically not enough room in a young patient with a congenital short femur for a pneumatic tourniquet to be placed proximal to the surgical incision. The release of the posterior capsule is performed only when there is a significant knee flexion contracture of greater than 15°.

David MacIntosh first described 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

(continued)

Box 22.9. (continued)

useful technique for congenitally deficient knees. In CFD, the instability pattern is different than in an isolated tear of the ACL. There is more of a rotary instability in CFD. Therefore, a purely intra-articular ligament reconstruction is insufficient and can lead to recurrent instability. Thus, the combination of extra- and intra-articular ACL ligament reconstruction is ideal. Having studied under MacIntosh 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. In my experience, it can now be safely done as young as 2 years of age.

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, which is described in the next step. This lateral extra-articular ligament was described by Dr. David MacIntosh for ACL reconstruction. In his honor and memory (he was one of my professors and mentors in Toronto in 1985) and in recognition of his idea of a lateral extra-articular ligament, I have referred to the medial extra-articular ligament procedure as the reverse MacIntosh, or extra-articular PCL procedure.

Superknee Surgical Technique (Ligamentous Reconstruction Only) (Figs. 22.9 and 22.13)

1. *Fascia lata harvest.* The knee is exposed through a long lateral 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 as at its musculotendinous junction with the TFL and reflected distally until its insertion onto the tibia (Gerdy's tubercle).
2. *Fascia lata preparation.* The fascia lata should be split into two longitudinal strips to make two ligaments. If the fascia is not very wide, bias the posterior limb to be larger. The posterior half is tubularized using nonabsorbable suture (#2 Tiger loop™) and a Krackow whipstitch from distal to proximal. The medial half is left flat.

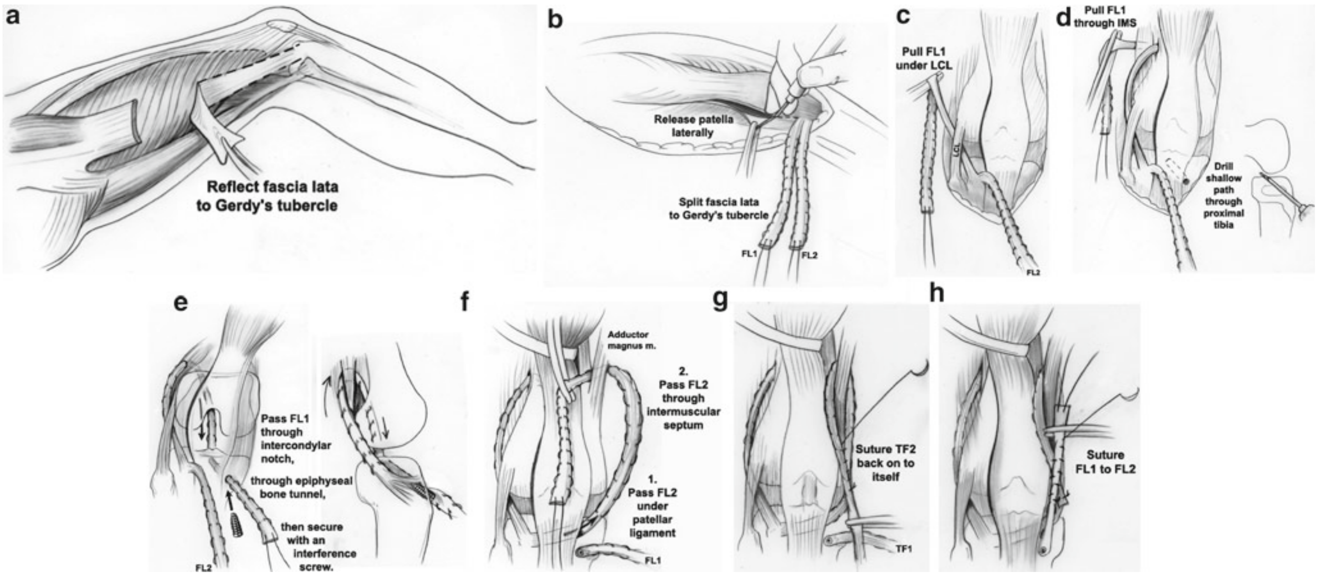

Fig. 22.13 Superknee surgical technique including MacIntosh and Paley reverse MacIntosh illustrations. (**a**) Harvest iliotibial band. (**b**) Divide the ITB into two halves. Do lateral release if maltracking patella. (**c**) Pass the posterior half under the LCL. (**d**) Pass it over the top of the intermuscular septum. (**e**) Drill a hole in the epiphysis of the proximal tibia and pass the ITB through this hole. Secure with interference screw with the knee in extension (MacIntosh). (**f**) Pass the anterior half of the ITB under the patellar tendon and around the adductor magnus and (**g**) suture it back to itself with the knee in 90° flexion (Paley reverse MacIntosh). (**h**) Suture the two new ligaments together

3. *Lateral release and Grammont patellar tendon realignment*: If the patella maltracks laterally, but it 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, 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-tendinous deep incision is made 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 [67]) is the pediatric equivalent of the Elmslie-Trillat [67] procedure in adults. The patellar tendon can now be displaced medially and sutured medial to the tuberosity with an absorbable suture.
4. *Extra-articular PCL reconstruction (reverse MacIntosh).* To anchor the 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 intermuscular 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, looped around the adductor magnus insertion from posterior to anterior, and then sutured to itself with nonabsorbable suture. 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 reconstructed ACL as it exits the tibia in the next step.
5. *MacIntosh extra + intra-articular ACL reconstruction*. The lateral collateral ligament (LCL) is identified using Grant’s test. This is performed by putting the leg in a figure-four position, which allows the tensioned LCL to be easily palpated. Dissect the LCL under tension, identifying 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 reconstructed ligament to bone. Drill a guide wire 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 proximally and posteriorly, exiting into the center of the knee about halfway back on the tibia. As CFD patients lack a true notch and a notch-plasty is not an option, it is better to keep the ligament more posterior on the tibia than 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, which can be measured with a sizer. 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.

A transosseous suture 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 to the anchor, the rest of the graft is anchored to the bony tibial epiphyseal tunnel using an absorbable headless interference screw. Again, tension the ligament in full knee extension to prevent a flexion contracture of the knee. As the patient's epiphysis grows, the ligament will become more taut. Therefore, mild laxity in the ligament is acceptable. The ligament can now be sutured to the anterior limb graft for additional fixation.

Superknee Procedure with Patellar Realignment for Dislocated/Dislocating Patella (Figs. 22.11 and 22.14)

Langenskiöld Patellar Realignment

If the patella is dislocated or dislocating, a modified version of the Langenskiöld [65] procedure is performed before the ligament reconstruction. First, the capsule is incised and separated from the patella and synovium both 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, which 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 periosteum (Grammont procedure).

After the tendon is elevated, it is shifted medially at least a centimeter. In the original Langenskiöld, the patellar tendon is detached from the tuberosity. A longitudinal incision is made in the synovium, the center of the patella is inserted into this new hole, and the synovium is sutured to the patella circumferentially. The medial capsule with the vastus 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.

Superknee Procedure with Knee Flexion Deformity (Figs. 22.10 and 22.15)

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 (see surgical technique below, peroneal nerve decompression).

Next, the biceps tendon should be Z-lengthened. 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 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 and reflect this part proximally. Isolate as much of the tendon as possible with 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

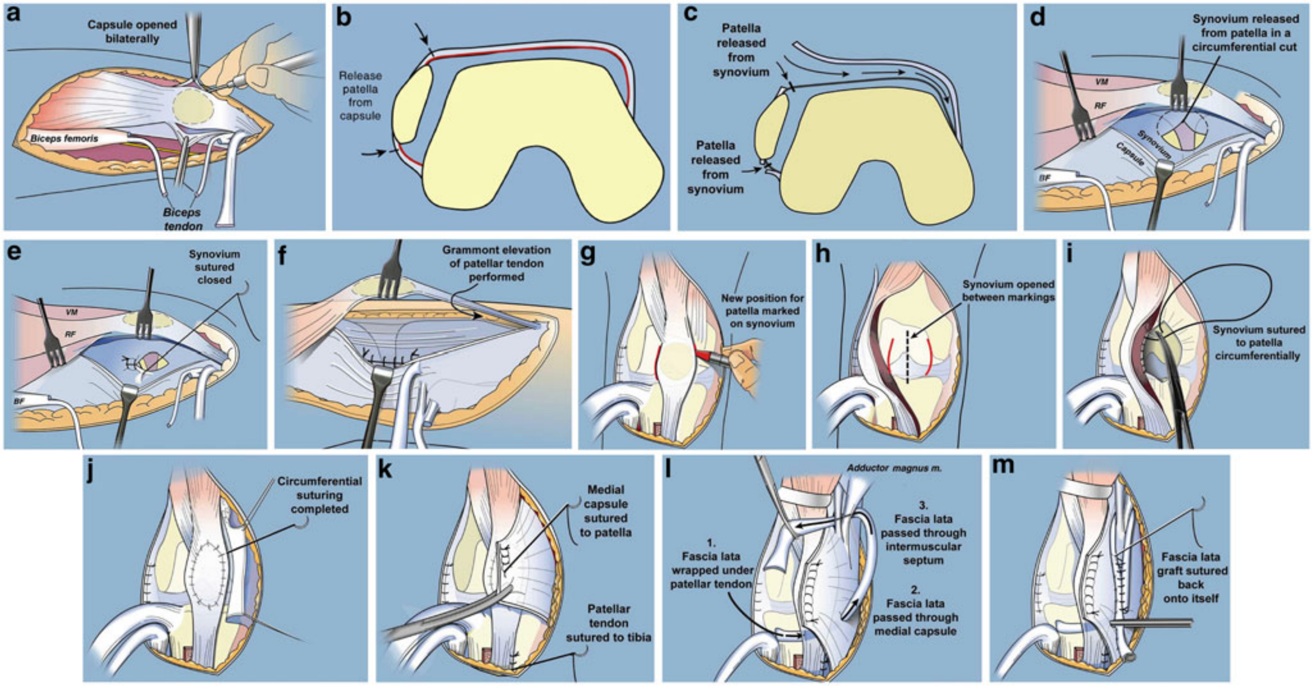

Fig. 22.14 Superknee surgical technique including Grammont, modified Langenskiöld, and Paley reverse MacIntosh procedure illustrations. (**a**) Release capsule on the lateral and medial sides of the patella. Medially cut through the vastus medialis and separate it from the rest of the quadriceps. (**b**) Do not cut through the synovium. (**c**) First separate the capsule from the synovium extensively and circumferentially around the patella and then cut the synovium to release the patella. (**d**) The synovium is also released from the underlying muscle and tendon. This leaves a patella-sized hole in the synovium. (**e**) Suture the hole in the synovium longitudinally. (**f**) Sharply dissect the patellar tendon off of the cartilaginous apophysis extending the dissection distally with a strip of periosteum (Grammont procedure). (**g**) Centralize the patella and mark this new position on the synovium with the knee in extension. (**h**) Incise the synovium between the marks. (**i**) Suture the lateral border of the synovium to the patella and continue circumferentially (Langenskiöld). (**j**) Once the entire synovium is sutured to the edge of the patella the patella will track correctly. The medial capsule with the vastus medialis can now be advanced. (**k**) The medial capsule is advanced and sutured to the lateral side of the patella. The medial patellar tendon is also sutured to secure the medial advancement of the patellar tendon (Grammont). (**l**) Pass the anterior half of the ITB under the patellar tendon and under the medial retinaculum and loop it around the adductor magnus tendon (Paley reverse MacIntosh). (**m**) Secure it back to itself with the knee in flexion. If ACL reconstruction is needed the MacIntosh procedure can be done using the posterior half of the ITB and secured in extension as in Fig. 22.13e

posterolaterally and enter the knee joint. With the knee flexed, the only vascular structures that one should see at the level of the knee joint are the central geniculate artery and vein. These 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 of the procedure. Once the posterior capsule is open, try and extend the knee joint. If there is still too much resistance, then the medial side will need to be exposed.

If performing a Superknee with ligament repair, a medial skin flap will need to be elevated anyway. 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 released from the femur, being careful not to injure the femoral vessels that lie immediately lateral to the muscle. The posteromedial capsule is dissected free from the popliteal fossa to communicate with the lateral dissection. The posterior capsule can then be cut under direct vision from both sides. The knee should now be able to fully extend.

The collateral ligaments are left intact. If the medial hamstrings are felt to be tight after the capsulotomy, 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 doing 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 described above. Occasionally, there is posterior instability that requires an intra-articular PCL reconstruction. This can be easily done using the iliotibial band by passing one limb

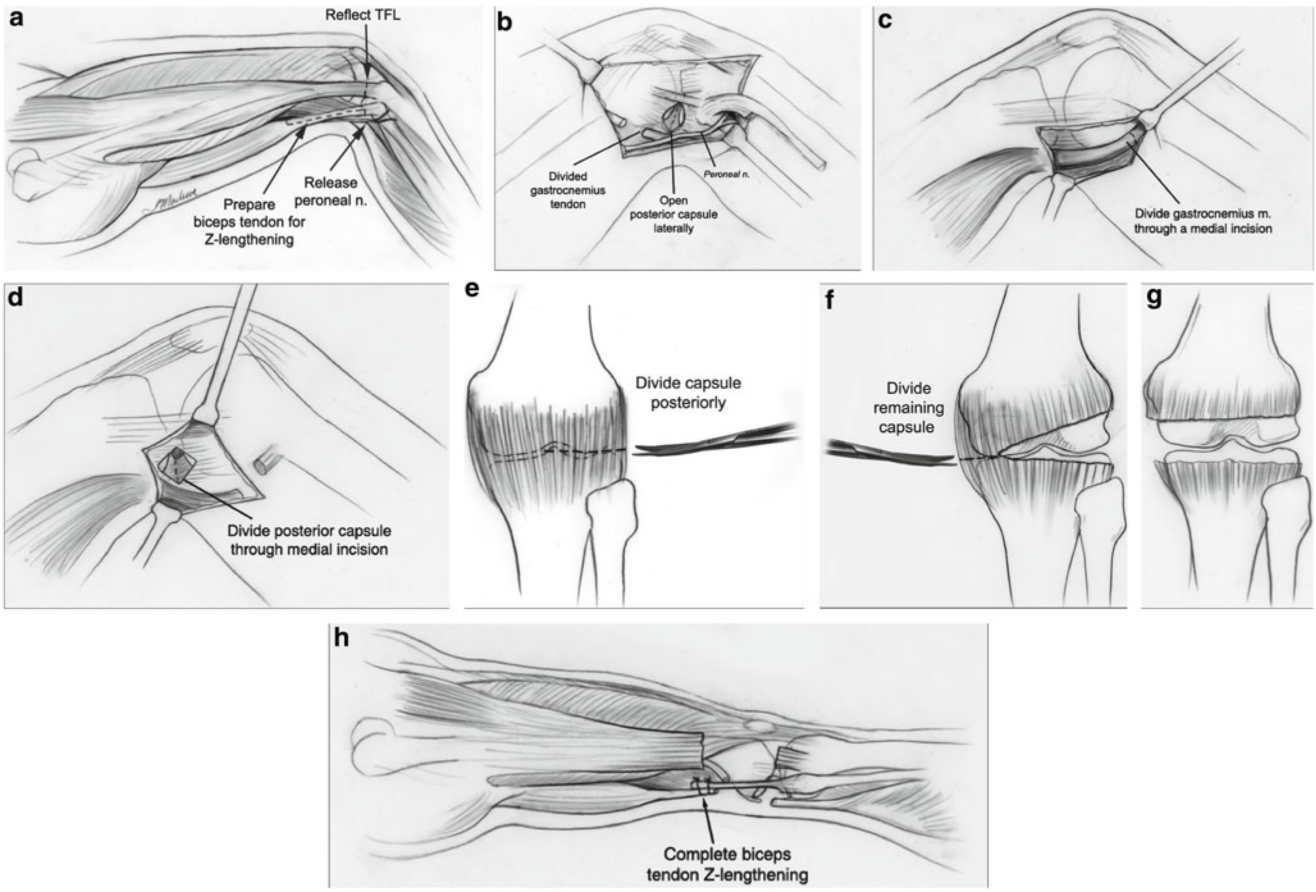

Fig. 22.15 Superknee surgical procedure including posterior capsulotomy, fascia lata, biceps and gastrocnemius tendon lengthening, and peroneal nerve decompression illustrations. (**a**) After lengthening the fascia lata (TFL) the biceps tendon is lengthened in a Z fashion. The peroneal nerve is decompressed as detailed in Fig. 22.16. (**b**) The lateral head of gastrocnemius is released off of the lateral femur. The lateral capsule posterior to the LCL is incised. (**c**) Through a medial incision the medial head of the gastrocnemius is released off of the femur. (**d**) The medial capsule is incised posterior to the medial collateral ligament. (**e**) The popliteal contents are dissected free of the posterior capsule from lateral and medial sides to connect together with the knee in flexion. The central geniculate vessels are cauterized. (**f**) The capsule can now be safely cut from the lateral side. (**g**) The capsular incision is completed from the medial side. (**h**) The biceps tendon is repaired in a lengthened fashion. (**i**) The fascia lata can be used for ligamentous reconstruction as previously described if needed

of it through an epiphyseal drill hole to the back of the knee. The band is then passed from posterior to anterior through a drill hole in the medial femoral epiphysis. It only needs to be secured at the femoral end.

Peroneal Nerve Decompression (Fig. 22.16)

Paley identified two sites of entrapment and described a surgical technique to decompress both tunnels [68]. In a cadaver study, Nogueira demonstrated that nerve tension increases after varus osteotomy and decreases to normal levels after peroneal nerve decompression [76].

The first peroneal nerve tunnel is located at the neck of the fibula. The common peroneal nerve leaves the undersurface of the biceps muscle to enter the lateral compartment of the leg. In order to enter the lateral compartment, the common peroneal nerve must perforate the otherwise intact fascial coverings of this compartment. The lateral compartment fascia folds over itself to form an arcade over the common peroneal nerve. The common peroneal nerve then splits into superficial and deep branches. The superficial peroneal nerve continues unimpeded through the lateral compartment, innervating the peroneal muscles.

The second tunnel is the intermuscular septum. The deep branch passes under the intermuscular septum to enter the anterior compartment of the leg, proximal to the intraosseous membrane. Anteriorly, the septum is confluent with the lateral and anterior compartment fascia. Posteriorly, it abuts but does not become part of the intraosseous membrane.

Surgery can be performed with or without a tourniquet, but without long-acting neuromuscular blockade. The patient is positioned supine, with a bump under the ipsilateral buttock.

The common peroneal nerve can be palpated in most people over the neck of the fibula. To palpate the nerve, roll it with the thumb or finger over the neck of the fibula. Make a 3–5 cm

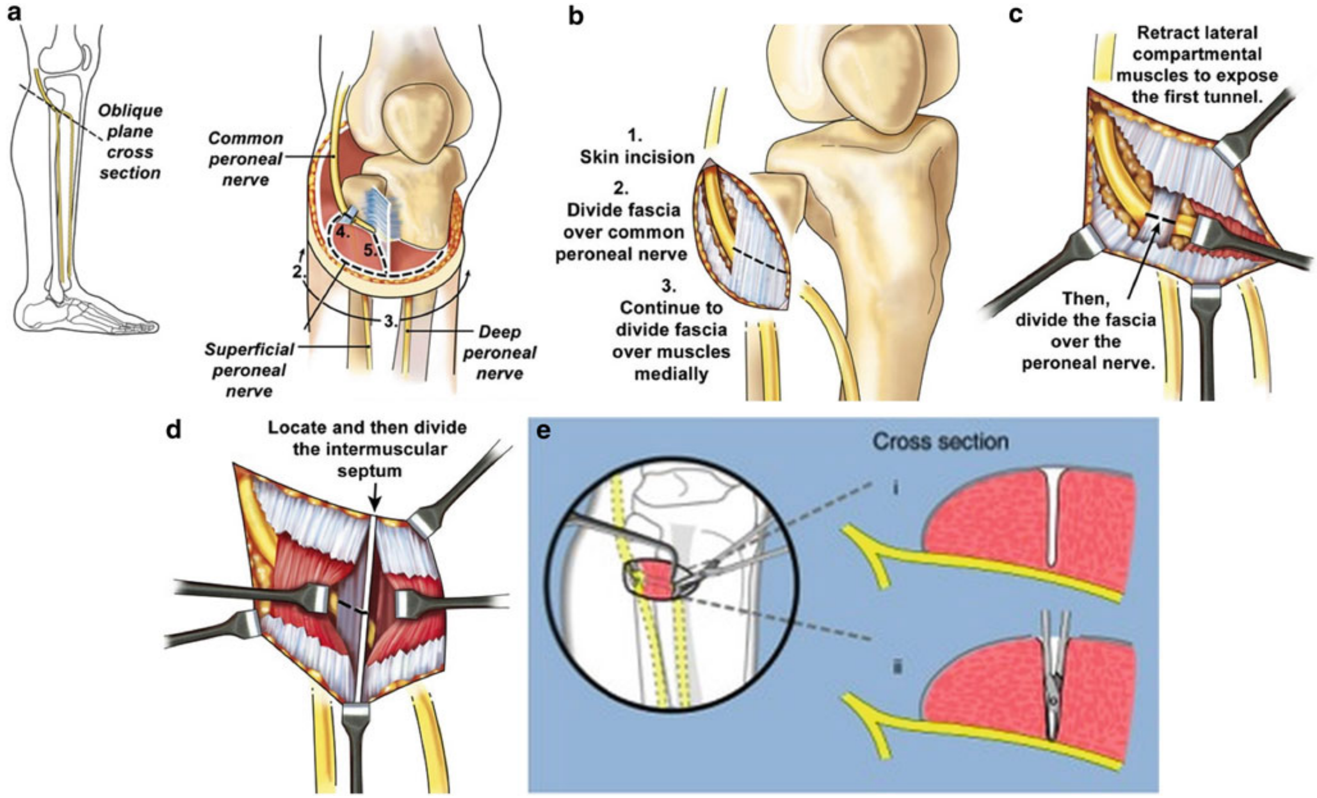

Fig. 22.16 Peroneal nerve decompression. (**a**) The peroneal nerve goes through two very tight fascial tunnels. The first is at the entrance to the lateral compartment and involves the entire nerve. The second is between the anterior and lateral compartments and involves the deep nerve only. (**b**) Start by incising the superficial fascia and identifying the borders of the common peroneal nerve proximal to the first tunnel. Incise the lateral compartment fascia in line with the nerve. (**c**) After incising the fascia retract the peroneal muscles to expose the first tunnel fascia covering the nerve. This fascia can now be cut under direct vision in a retrograde fashion. (**d**) Extend the transverse fasciotomy to the anterior compartment and expose the intermuscular septum between the anterior and lateral compartments. Cut this fascia to complete the decompression of the nerve. (**e**) The intermuscular septum comes down over the deep peroneal nerve like a guillotine. This narrow space under this septum is the second tunnel

oblique incision parallel to the course of the nerve over the neck of the fibula. Cut through skin and subcutaneous tissues down to the fascia, posterior to the lateral compartment. Feel for and identify the nerve again. Dissect and expose the nerve proximally and distally. It is not necessary to remove the fat over the nerve. Follow the common peroneal nerve distally 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 first tunnel, which should be released. The nerve can then be seen dividing into two branches, the superficial running down in the lateral compartment, and the deep branch going in the direction of the anterior compartment of the leg.

Extend the transverse compartment fasciotomy across to the anterior compartment. Notice the intermuscular septum separating the two compartments, confluent with the fascia overlying both compartments. Dissect the muscle off 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 both medial and lateral aspects, cut it from anteromedial to posterolateral. Be careful to stop as soon as the septum ends. Immediately below the septum, a band of fat can often be seen, which contains the deep peroneal nerve. This completes the decompression of the second tunnel of the deep peroneal nerve.

Let down the tourniquet at this point. The nerve can be stimulated with a disposable nerve stimulator. If the tourniquet time exceeds 20 min, the nerve may not initially respond to electrical stimulation due to the tourniquet ischemia. If the nerve was paralyzed before decompression, it can be stimulated before the decompression at the common peroneal nerve and then again after the decompression to evaluate if there is any immediate change.

Box 22.10. Superknee Tips and Tricks

- The stability of the knee cannot be fully assessed until the fascia lata has been released.
- Take a small portion of biceps fascia and intermuscular septum for a wider graft. If the graft is still narrow, bias the more important posterior limb of the graft to be larger.
- Send the ACL reamer all the way into the knee to make sure that the tunnel is adequately drilled for passing the ACL graft.
- Tighten the reverse MacIntosh with the knee in flexion, and the ACL graft with the knee in extension.
- Suture the ends of both limbs of the graft together at the end for additional fixation.

  When cutting the posterior capsule, do not stray distal to the knee joint.

Femoral Lengthening for Paley Type I CFD

Choice of Osteotomy Level

Distal femoral 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. However, the distal osteotomy applies greater forces to the knee joint and increases the risk of knee stiffness and subluxation. These risks can be reduced by articulating across the knee joint and extending fixation to the tibia. Proximal femoral osteotomies have less effect on the knee, but have a greater effect on the hip joint. They 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. Proximal lengthening osteotomies are also more prone to poor bone formation and prolonged consolidation phase in patients with CFD. There is also a higher rate of fracture after removal of external fixation in CFD. For these reasons the author prefers to lengthen at the distal femur in patients with CFD.

However, 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 further increasing the tendency to lateral subluxation/dislocation of the patella in these patients. 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 patient has previously undergone preparatory hip surgery, such as a Superhip reconstruction or proximal femoral osteotomy and Dega pelvic osteotomy, 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 proximal varus, then a proximal internal rotation and valgus osteotomy of the femur are carried out together with a distal lengthening osteotomy.

A distal femoral lengthening osteotomy can be used to gradually (ring fixator) or acutely (monolateral fixator) correct the valgus and any mild flexion deformity of the knee (see Figs. 22.10 and 22.11). 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 poor healing sclerotic bone. Therefore, the regenerate bone in the distal femur is wider, stronger, and subject to more axial deviation muscular forces than in the proximal femur.

In older children with a wider medullary canal, implantable limb lengthening (Figs. 22.9 and 22.17) or lengthening over nail (LON) can be performed (see Fig. 22.17) [69]. A proximal osteotomy can be used for lengthening over nails because there is little risk of refracture. The internal rod supports the bone until consolidation is complete. Intramedullary nailing in children risks disturbing the growth of the trochanteric 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, humeral, or tibial nail). To avoid a coxa valga deformity, we prefer to use this technique in patients with coxa vara. A theoretical epiphysiodesis, created by the nail, may lead to gradual correction of residual coxa vara, although this has not been observed. Fixator-only lengthening is typically used for the first lengthening. LON or implantable lengthening is used for subsequent lengthenings if the anatomic dimensions of the femur permit. In the author's experience, there is a higher risk of deep infection in patients who have had previous external fixators. It is unclear if this is related to the number or degree of previous pin infections. It is suspected that latent bacterial forms remain in the bone in a dormant state and are activated at the time of reaming and nail insertion. During external fixator removal, we recommend aggressive curettage of all granulation tissue in the soft tissues and bone of the pin tract in an effort to reduce such latent infection.

Soft Tissue Releases for Limb Lengthening

In patients with CFD, soft tissue releases in conjunction with femoral lengthening are essential to prevent subluxation and stiffness of the knee and hip. 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, approximately 6 weeks later. Usually, these are done at the index procedure to avoid an additional anesthetic. If a preparatory hip or knee surgery has already been performed, then the fascia lata, rectus femoris, and biceps femoris may have already been lengthened, and there is no need to repeat them. However, if soft tissue releases have not been performed, then they should be carried out at the time of the lengthening surgery.

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 prone knee flexion test (Ely test) for the rectus femoris (see Fig. 22.3a), the straight leg raising test (popliteal angle) for the hamstrings (see Fig. 22.3b), hip abduction range of motion for adductor contractures, and hip adduction range of motion with the knee and hip in extension for the fascia lata. The Ober test is helpful for identifying fascia lata tightness (see Fig. 22.3c).

The rectus femoris is evaluated with the Ely test (see Fig. 22.3a). A positive Ely test demonstrates pelvic flexion at the hip during prone knee bend. The rectus femoris should be released through a small anterior inguinal incision. Even with a negative Ely test, the rectus may be released from anterior inferior spine, as it still tends to tighten with lengthening.

Hamstrings are evaluated by the straight leg raise, measuring the popliteal angle (see Fig. 22.3b). If the knee can fully extend with the hip flexed to 90°, then the popliteal angle is 0° and the hamstrings are not tight and thus such a patient would require no treatment before femoral lengthening. If there is a popliteal angle >0°, the hamstrings are already tight and will lead to contractures during lengthening. The author prefers lengthening the biceps femoris with the iliotibial band through a lateral approach. If needed, the medial hamstrings can be lengthened from the medial side.

If hip abduction is limited, percutaneous adductor tenotomies of the adductor longus and gracilis tendons should be performed. If hip subluxation or more severe adduction contractures develop during lengthening, then an open, more extensive adductor release, including the adductor brevis, is indicated. The adductor release is very important in proximal lengthening, but for distal lengthening, it is usually not necessary. Distal adductor magnus release has been described

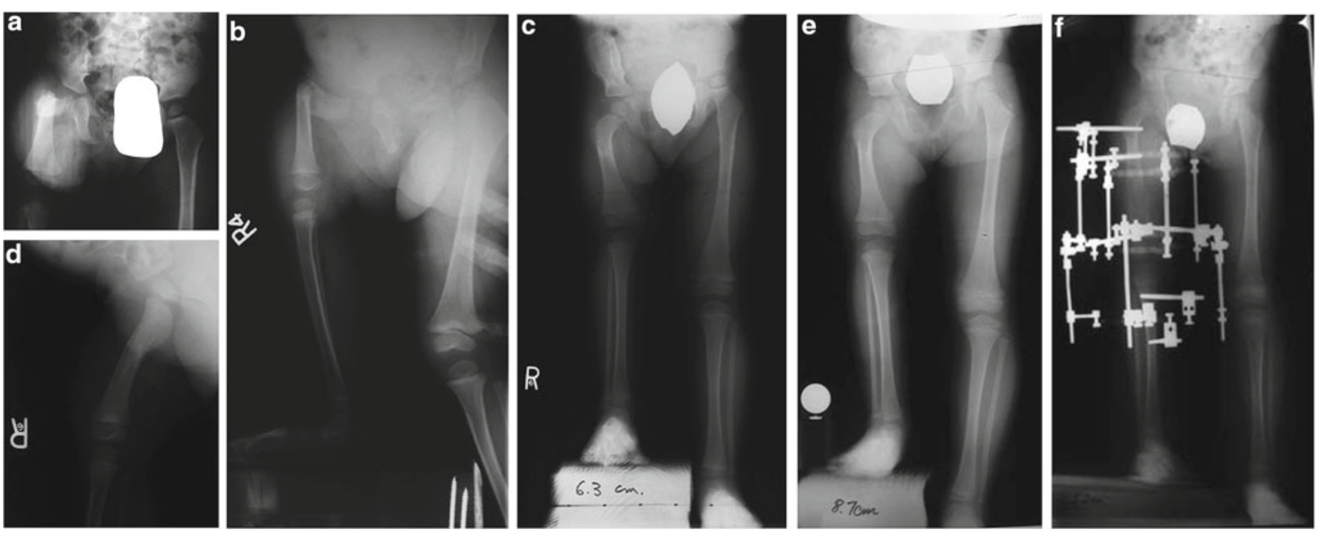

Fig. 22.17 Two-year-old girl born with Paley type 1b subtrochanteric type. AP and lateral radiographs (**a**, **b**) showing characteristic deformity as previous illustrated in Figs. 22.1 and 22.7. After the Superhip procedure performed at age 2 years, the femur appears to have a normal anatomy as shown on AP and lateral views (**c**, **d**). Standing X-ray prior to first lengthening. A 1-cm gain was achieved due to the Dega osteotomy (**e**). First lengthening performed at age 4 years, using Ilizarov apparatus (Smith & Nephew, Memphis, TN) with knee hinges and fixation to the tibia (**f**). After 8-cm lengthening (**g**). Full recovery of hip and knee range of motion and strength following first lengthening (**h**, **i**). Second lengthening performed at age 8 years, using a multiplanar monolateral external fixator (Orthofix LRS and Sheffield components, McKinney, TX). The knee is able to flex and extend with a lateral knee hinge connected to an arch fixed to the tibia (**j**). The femur was lengthened 7 cm (**k**) over a Rush rod similar to Bost's lengthening over nail method [69]. Third and final lengthening of 5 cm at age 15 years (**l**) was performed using an implantable lengthening nail (PRECICE, Ellipse Technologies, Irvine, CA). Note that prior to this a contralateral distal femur epiphysiodesis (5-cm equalization) was performed at age 10 and pelvic osteotomy and distal femoral varus osteotomy were performed at age 14. Total limb length equalization was 26 cm. The hardware was removed 1 year after the lengthening (**m**)

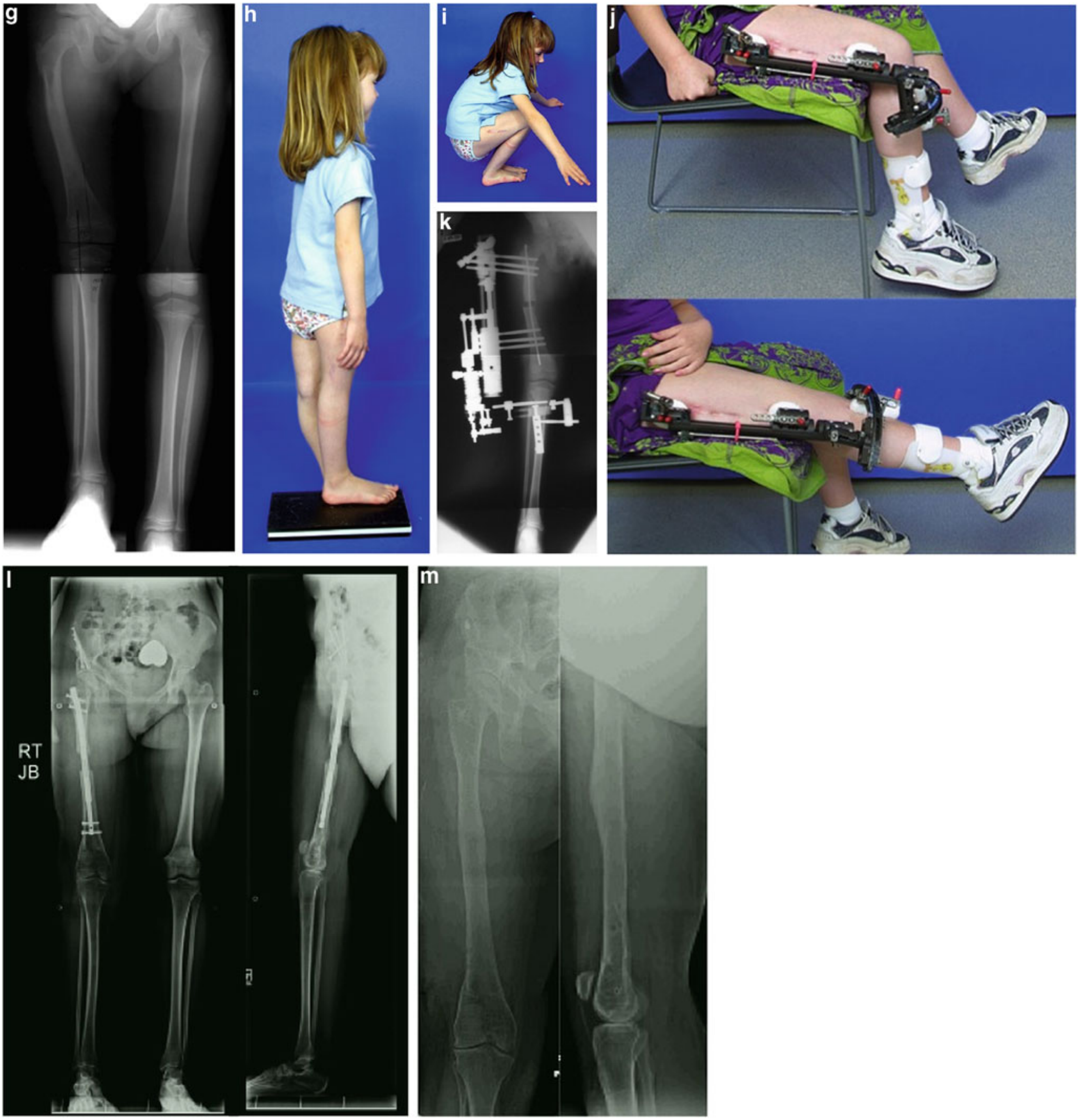

Fig. 22.17 (continued)

for congenital femoral lengthening [70], but this has not been found to be helpful in the senior author's opinion.

To confirm that the fascia lata is tight, perform an Ober test (see Fig. 22.3c). The patient lies on his or her side, and the knee is flexed and hip extended by pulling back on the foot and pushing forward on the buttock. The knee will stay up in the air and not fall down due to contracture of the fascia lata. This is called a positive Ober sign and indicates that the fascia should be excised or cut. In every case of CFD lengthening, if it has not already been excised, the fascia lata should be lengthened. A 3-cm longitudinal incision is made over the posterior edge of the fascia lata, where it connects with the intermuscular septum. The entire fascia lata is transected at the level of the proximal pole of the patella by dissecting anterior to the incision. If the incision is made more posteriorly, the biceps femoris can be easily exposed on the posterior side and the lateral biceps fascia can be safely recessed.

Fig. 22.18 Intra-articular osteotomy. (**a**) CFD Paley type 2 with rotatory subluxation of the knee joint. The posterolateral tibial plateau is round (*arrow*) and contributes to the subluxation. (**b**) The Superknee procedure was performed with decompression of the peroneal nerve and taking down the proximal tibiofibular joint to expose the lateral plateau. A hemi-plateau osteotomy of the posterior plateau was performed, with allograft and internal fixation. (**c**) After complete bone healing the lateral plateau is now flat and not round. The subluxation was eliminated by the Superknee procedure

Patients may develop hyperlordosis or a hip flexion or abduction contracture secondary to the femoral lengthening. If so, the proximal fascia lata is cut transversely through a small lateral incision just distal to the greater trochanter.

Botulinum Toxin Injection

The effect of botulinum toxin has been shown to be helpful in prevention of muscle contractures [71, 72]. In the author's experience, it decreases muscle stretch pain due to muscle spasm, which is primarily in the quadriceps muscle during physical therapy. Botulinum toxin can be injected into the quadriceps up to a limit of 10 U/kg body weight. The added volume of the saline is limited to avoid systemic toxicity, so injection is usually done with a total of 3–4 mL.

Knee Instability

Almost all cases of CFD can be assumed to have hypoplastic or absent cruciate ligaments, with mild to moderate anteroposterior instability. Some patients 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 knee instability is the tendency of the knee to subluxate with lengthening. The subluxation is usually posterior, or posterolateral (posterior plus external rotation of the tibia on the femur), but it 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 [42]. This promotes knee stiffness while protecting the knee from subluxation. The author prefers to protect the knee by extending the external fixation to the tibia with a hinge. The hinge permits knee motion while preventing posterior as well as anterior subluxation. It also prevents pressure being transmitted to the knee joint cartilage. Hinges are an integral part of circular external fixators such as the Ilizarov device, as well as some monolateral external fixators such as the Orthofix Limb Reconstruction System (LRS), Smith and Nephew Modular Rail System.

Less common is anterior subluxation or dislocation of the tibia on the femur. This occurs as the knee goes into extension. It is important to document the angle of flexion the knee relocates or, conversely, at which angle short of full extension the knee dislocates. These dislocations are more commonly due to a posterior deficiency with rounding of the tibial plateau, but they can also be due to an anterior deficiency of the distal femur, in which the lateral radiograph of the knee shows a lack of the anterior protuberance of the femoral condyles. If the posterior condyle is rounded but the anterior part of the knee is straight, an intra-articular osteotomy is required to elevate the posterior condyle: intra-epiphyseal in children and metaphyseal in adults (Fig. 22.18). This can also be combined with a PCL reconstruction as noted above.

Surgical Technique

There are currently a multitude of different devices used for external fixation and distraction osteogenesis. The original Ilizarov fixator technique requires the use of rings or arches. It allows multiplanar fixation in small segments of bone and can incorporate hinges to correct angular deformity gradually or acutely. It allows for hinged articulation across the hip and knee joints, thus protecting these joints from subluxation during distraction without restricting joint motion. However, it is bulkier than a monolateral external fixator and forces the legs to be abducted due to the ring protruding medially.

Monolateral external fixators traditionally lacked the ability to span across joints. They have evolved to permit articulated fixation to the tibia and pelvis. They also allow locking of the hinge to prevent contracture. Deformities can also be corrected acutely. Monolateral fixators are less bulky and easier to wear for the patients. Nevertheless, they are more constrained in their pin location options and are therefore less space efficient than the circular fixators. The Orthofix LRS (Orthofix, Lewisville, TX) and the Smith & Nephew Modular Rail System (MRS, Smith & Nephew, Memphis, TN) are two examples of currently available monolateral external fixators that allow articulated spanning of joints. Their application for CFD lengthening together with the application of the Ilizarov device will be described.

In the femur, half-pin fixation is the rule, even with the circular fixator. Half-pin size is determined by the diameter of the bone, with 6.0-mm pins used for most older children and 4.5-mm pins for younger children with smaller bones. For very young children, small tapered 3.5- to 4.5-mm pins can be used. The author prefers to use hydroxyapatite-coated pins to decrease the chance of loosening and to lower the risk of pin-site infection [73].

Distal Femoral Lengthening: Ilizarov™ Fixator Technique (See Fig. 22.17)

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 second-level, 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 proximal arch is attached parallel to a line from the tip of the greater trochanter to the center of the femoral head, the middle ring is perpendicular to the mechanical axis of the shaft of the femur (7° to the shaft), and the distal ring is parallel to the knee joint line. After the osteotomies, when 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. The first half-pin is from lateral to medial in the frontal plane, parallel to a line from the tip of the greater trochanter to the center of the femoral head. 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 an arthrogram to better outline the cartilaginous femoral condylar line and 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 a valgus deformity angle to each other. A lateral half-pin is inserted into the mid-segment of the femur. This pin is at 7° to the shaft of the bone.

At this 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 extended to correct all components of the proximal femoral 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 the other 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, especially anteriorly, 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.

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 in the plane where the two posterior femoral condyles are seen to overlap on the lateral view (Fig. 22.19) [42]. 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 distal femoral condyles, 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. A drop test is performed to see if the tibia flexes through the hinge without catching. If this range feels

Fig. 22.19 Lateral fluoroscopic view of knee arthrogram. The center of rotation knee axis pin has been inserted at the level of distal physis and the intersection with the posterior cortex. The medial and lateral femoral posterior femoral condyle outlines can be seen because of the radiographic dye. They both overlap each other

frictionless, a second and third tibial half-pin are added. Finally, a removable knee extension bar is inserted anteriorly between the distal femoral and the tibial half-ring.

Box 22.11. The Drop Test

- A half-pin is placed in the proximal tibia from anterior to posterior.
- The half-pin is attached to the proximal tibia ring and secured.
- The tibia is raised up in full extension with the hip flexed.
- While supporting the femur, the tibia is released to flex with gravity.
- A positive drop test and a well-placed knee hinge allow the tibia to flex to 90° without catching.
- If there is significant friction, make adjustments to the half-pin.
- Securing the half-pin with the knee in full flexion or full extension or 45° may improve the drop test.
- Do not add any more pins until the drop test demonstrates no friction.
- The success of the drop test depends on the initial accurate placement of the hinge.

  Perform the drop test after each pin is added. Use a total of three pins in the tibia.

Distal Femoral Lengthening: Orthofix™ Fixator Technique (See Fig. 22.17)

1. 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 comes in pediatric and adult sizes. The hinge axis is lined up through its most distal hole.
2. A commercially available "sandwich" clamp is used, or 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 the most proximal half-pin inserted.
3. The most distal 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 the distal femoral valgus is to be corrected acutely, a 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.
4. 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 that protrudes laterally. A Sheffield clamp from Orthofix™ is applied to this pin to act as a surrogate knee hinge. It is locked in place by putting a Rancho™ cube laterally with a set screw to prevent it from moving outward. Conical washers are used between the Sheffield clamp and the LRS to prevent friction. The Sheffield clamp is left partially loose to permit motion.
5. 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 for friction, two more oblique pins are inserted into the tibia and connected to the Sheffield arch using cubes.
6. A removable knee extension bar is fashioned from Ilizarov parts to be used especially at nighttime. 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.

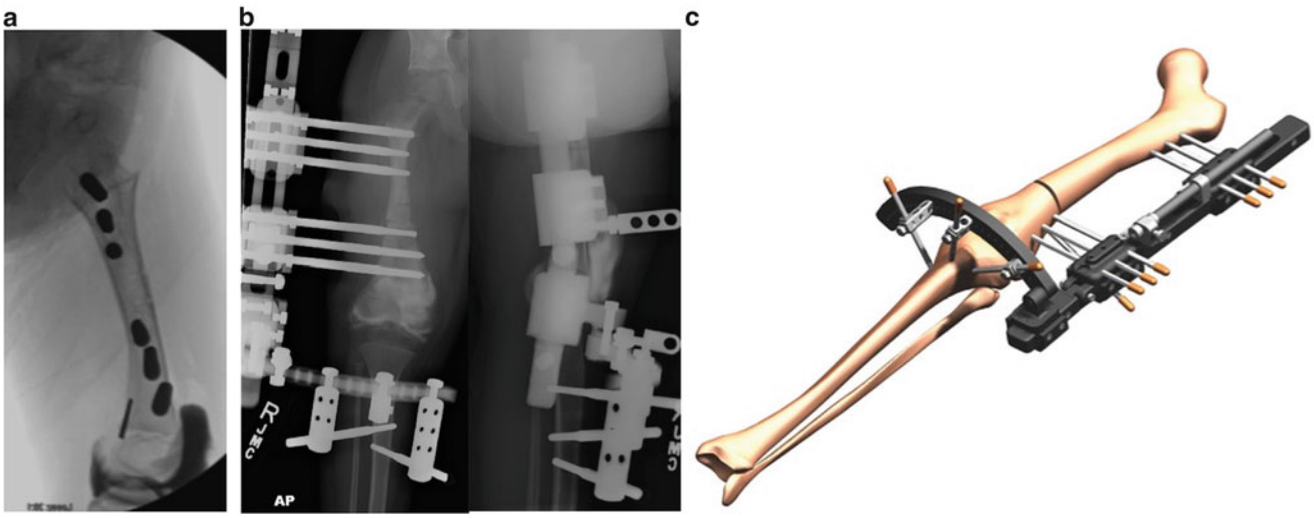

Fig. 22.20 Modular Rail System (MRS, Smith & Nephew, Memphis, TN) technique. (**a**) The external fixation pin is inserted parallel to the knee axis pin. The external fixator is used as a template. (**b**) AP and lateral radiographs showing the external fixator in place and the articulated spanning across to the tibia. The arthrographic dye is in place. The fixator hinge line corresponds to the knee hinge center of rotation axis. (**c**) Illustration of the Modular Rail System external fixator articulated across the knee joint. Note location of knee hinge

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. Thus, 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.

Distal Femoral Lengthening: Modular Rail System (Smith & Nephew, Memphis) Technique (Figs. 22.9, 22.10, and 22.20)

This external fixator was designed 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.

1. Pre-construct the external fixator using pediatric or adult rail segments with two pin clamps, knee hinge, and tibial rail segment.
2. Inject radio-contrast solution into the knee joint and obtain a true lateral projection, rotating the femur until the posterior aspect of the medial and lateral femoral condyles overlap.
3. Insert a 2-mm Steinmann pin into the center of rotation of the knee joint. This is defined as the point of intersection of the posterior femoral cortex with the distal femoral physis. In adults, it is the intersection of the posterior cortex of the femur with Blumensaat's line. Confirm on the AP view that the pin is parallel to the joint line.
4. If there is no valgus or flexion deformity, proceed to drilling a 1.8-mm wire into the proximal 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, and 3.2 mm for a 3.5- to 4.5-mm pin).
5. Insert a pin of the appropriate thread length corresponding to the diameter of the bone at that level. The bending strength of the pin is related to the smallest diameter protruding from the near cortex. It is better not to leave threads outside of the near cortex to obtain the maximum bending strength out of each pin.
6. Mount the pre-constructed MRS onto the Steinmann pin and the proximal half-pin. The Steinmann pin goes through the cannulated hinge of the fixator.
7. Drill a 1.8-mm wire through the distal most pin hole of the clamp. An overhang clamp can be used 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.
8. Insert the other two pins into the clamp, making sure that the holes line up with the bone. All the pins should be parallel to each other. Make sure that the distal pin clamp is locked down to the fixator with 7-mm red bolts. Slide off the fixator from the pins.
9. 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.
10. Reapply the fixator to the pins. Make sure that the osteotomy does not displace or translate, making

adjustments as needed. If AP clamps are being used, drill at least one hole for an oblique posterolateral proximally and distally. There should be three or four total number of pins per segment, depending on the size of the femur and the thigh being lengthened.

11. Drill and place an anterior to posterior pin in the proximal tibia, making sure that it is distal to the tip of the proximal tibial apophysis. 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 can be done if there is subluxation. Apply a set screw to the pin once the knee motion is free. Confirm with a drop test: if the leg drops with no friction from 0° to 90°, then the hinge is perfect. Apply two more pins with a different orientation in the proximal tibia.
12. Apply a distractor bar between the two clamps, making sure that it turns freely and that the osteotomy is distracting appropriately. The bar distracts 1 mm for every full turn of 360°.
13. 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 fix 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 full time at night and half time during the day. It is easily removable for exercise.
14. Incisions are closed and sterile dressings applied around the pins. Consider Botox to the quadriceps muscles to diminish muscle spasm. Final radiographs are taken prior to leaving the operating room.

Box 22.12. Lengthening Tips and Tricks

- A knee arthrogram is helpful in young children to visualize the posterior condyles or center of rotation.
- The first half-pin or the hinge pin sets up the rest of the pins. Spend extra time getting the first one perfectly right, and the rest will follow.
- For smaller bones, use the cannulated wire technique to ensure proper placement of pins. Eccentric cortical drilling can lead to fractures or pin pullout.
- Use off-axis pins for stronger fixation if possible.

  Make sure that the osteotomy is completed and distracts appropriately without too much resistance before leaving the operating room.

Modification for Valgus ± Flexion Deformity of the Distal Femur

If there is a valgus or flexion deformity of the distal femur, changing the pin placement will provide correction. The first wire is placed into the distal femur, using the MRS as a guide to keep the wire parallel to the knee hinge pin. A cannulated drill is then used and a half-pin is inserted. The most proximal pin is then placed into the proximal femur at about 7° of valgus in relation to the shaft. In reference to the distal pin, it should be in valgus angulation and in the same rotational orientation as the distal pin. A distal femoral osteotomy is performed at the distal femur at the planned level, leaving enough room to add two more pins distal to the osteotomy. Once the pins are connected to the external frame and parallel to each other, the valgus deformity is corrected. If there is also a flexion deformity of the knee, extend the knee with the tibia to level the distal segment into extension before adding the remaining frontal plane pins.

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. If the lengthening rate is 1 mm per day, then follow-up is usually every 2 weeks. If there is delayed healing of the regenerate bone, or if knee motion is decreasing, then the rate is often decreased. If the lengthening rate is decreased to 0.75 mm or 0.5 mm per day then the follow-up can be increased to 3 or 4 weeks, respectively. Rarely, if there is concern for premature consolidation and the lengthening rate is increased, then follow-up is more frequent.

Patients are allowed to weight-bear as tolerated with their external fixator. Physical therapy is begun within 1 or 2 days after surgery and continues daily throughout the distraction and consolidation phase. In the author’s opinion, the amount of therapy is directly related to a better functional result and faster rehabilitation after removal. During distraction, patients have one to two formal physical therapy sessions daily (45–60 min each). In addition, at least two home sessions (30 min each) are recommended to the parent. Patients undergoing lengthening are at their best immediately after surgery and have difficulty with muscle tightness and range of motion only after lengthening has started. It is not until the consolidation phase that the usual orthopedic pattern of rehabilitation and recovery occurs. Thus, lengthening should be thought of as a prolonged procedure that lasts until the end of the distraction period, and not even be considered in the absence of therapy.

The majority of therapy time is spent obtaining knee flexion and maintaining knee extension. Knee flexion should be maintained as close to 90° as possible. It is important to start pushing the extremes of passive knee flexion (especially prone) during the 1-week latency period after surgery. Once distraction begins, there is more resistance from muscle spasm. If knee flexion begins to diminish (<75°), then lengthening should be stopped or slowed to rehabilitate the knee better, sometimes waiting for improvement before resuming lengthening. Function should never be sacrificed for length, and preserving the knee joint and motion is paramount. A knee extension bar is used at night and part-time during the day to prevent a knee flexion contracture [42]. Posterior subluxation can occur with a fixed flexion deformity and is suspected clinically based on a change in the shape of the front of the knee. The patella becomes very prominent and the tibia appears depressed (ski hill sign).

Weight bearing is encouraged throughout the lengthening as long as the patient has normal proprioception. In younger children, walking without support can be achieved immediately because the fixator is very stiff relative to the patient's weight. When the child is older and heavier, crutches or a walker is preferred until there is sufficient consolidation.

During the distraction phase, therapy is focused on passive range of motion exercises to maintain hip and knee range of motion. During the consolidation phase, the priority shifts to active range of motion and strengthening to regain any motion lost during distraction. Hip abduction and extension exercises help prevent adduction and flexion contractures that can lead to hip subluxation. Release of the adductors, rectus femoris, and TFL during lengthening may need to be considered to allow further lengthening and to prevent hip subluxation.

After the lengthening regenerate is healed and the external fixator removed, recovery is limited by two factors: joint range of motion and muscle weakness. In most cases, joint motion is regained faster than muscle strength. The three antigravity muscles are the limiting factor to full recovery: the hip abductors, quadriceps, and gastro-soleus muscles. To minimize disability, these muscles should have resistance training throughout the lengthening program to prevent atrophy from prolonged external fixation and lengthening. Other modalities such as electrical stimulation or aquatic therapy may be helpful. Please also refer to Chap. 12 for more details regarding physical therapy in such patients.

Fixator Removal and Rodding of Femur (See Figs. 22.9 and 22.10)

The external fixator can be removed once the regenerate bone is fully healed radiographically. The biggest risk after removal is fracture of the femur, which can manifest as painless bending of the femur or as a completely displaced fracture. The use of a spica cast does not reduce the fracture rate. A study, performed by the author in 1999, showed a fracture rate of 34 % after CFD lengthening, compared to 9 % for other etiologies [84]. The regenerate bone should show no gaps, the interzone should be closed, and based on AP and lateral views there should be evidence of corticalization on at least three sides of the lengthened regenerate. Since this refracture rate was so high and unacceptable, Paley started prophylactic rodding of the femur with a Rush rod in 2000. The fracture rate dropped to almost zero in the senior author's personal experience and somewhat higher in a multi-surgeon series [53]. In the senior author's personal series, the rate of infection after such prophylactic rodding was 4 %. These were easy to eliminate by removal of the rod and insertion of an antibiotic cement rod [74]. This was considered a reasonable trade-off: trading 34 % fracture rate for 4 % infection rate.

Surgical Technique for Prophylactic Rodding of Femur at Time of External Fixator Removal

1. The external fixator is removed under general anesthesia, ensuring that the patient is deeply asleep (to avoid involuntary muscle spasms which could lead to fracture of the regenerate at the time of removal). Slightly loosen all of the pins while they are still supported by the frame, which is especially important in hydroxyapatite-coated pins. Remove the middle pins while the fixator is still on. The most distal and proximal pins are removed while an assistant provides bimanual support of the thigh above and below the lengthening zone.
2. After prepping the thigh, curettage any loose granulation tissue from the pin holes. The pin holes in the bone are also curetted in the event of osteolysis or infection. Cover and seal the pin holes with an adherent plastic dressing (e.g., Tegaderm, 3 M, Minneapolis, MN). In the event of a severe infection, the surgeon must weigh risks of a fracture and loss of length versus hardware infection and secondary surgeries.
3. Prep and drape the lower limb free, being careful not to manipulate the leg. A bump may be placed under the ischium to raise the femur for lateral viewing on image-intensifier radiography.
4. Insert a 1.8-mm bayonet-tipped Ilizarov wire at the tip of the greater trochanter. Advance the wire by tapping, rotating the curved tip of the wire to bounce off the inner cortex. Use the drill only if the wire will not advance. A slight bend in the wire may be necessary to direct the wire. The wire is passed through the center of the intramedullary canal and regenerate bone, all the way to the distal femoral physis if possible. A second wire of

equal length can help measure the length by inserting it to the greater trochanter and measuring the difference in length of the two wires.

5. Drill over the wire with a cannulated drill bit. The 3.2-mm, 4.8-mm, and 6.4-mm drill bits correspond to the 1/8″, 3/16″, and 1/4″ Rush rods, respectively. Long cannulated drills are available from Pega Medical (Montreal, Canada) and are preferred since they slightly overdrill above the designated size making rod insertion easier. They are also longer and can reach the full length of almost any femur.
6. Insert an appropriate length Rush rod. As an alternative, a stainless steel flexible nail can be used instead, but they are more difficult to insert and direct. The end of the Rush rod is better designed for directing the implant tip. Recently, a lockable rod (the SLIM, Pega Medical, Montreal, Canada) has been developed for this purpose with a tip similar to the Rush rod but with a threaded proximal part similar to the Fassier-Duval nail (Pega Medical, Montreal, Canada).
7. After rodding, the patient goes home with sterile, dry dressings covering the pin sites and a 10-day course of oral antibiotics, usually a first-generation cephalosporin. Weight bearing and physical therapy are restricted for 1 month. Gentle range of motion of the knee is allowed at home.

Box 22.13. Fixator Removal Tips and Tricks

- Do not remove fixator until three sides have corticalized.
- Use a rush rod or intramedullary device to prevent fracture through the regenerate bone.
- Slightly turn or "crack" all the half-pins while in the fixator prior to removing the frame. The hydroxyapatite bond can be quite strong.
- Curettage out any hypertrophic granulation tissue. Loose pins warrant more thorough curettage of the bone.
- It may not be possible to direct a thin Ilizarov wire all the way down the femur. Drill just the proximal portion and then proceed to the Rush rod.
- If needed, put a small bend in the Rush rod in line with the bevel to augment guided insertion.

  Hold off on physical therapy for a month after fixator removal.

Specific Complications and Their Treatment for Congenital Femoral Deficiency Lengthening

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 dysesthesia in the distribution of the peroneal nerve or extensor hallucis longus weakness. A nerve conduction study may show evidence of nerve injury, but most cases will be negative since too many fibers are conducting normally. Quantitative sensory testing using the pressure-sensitive sensory device (PSSD), if available, is the most sensitive test to assess for nerve involvement [75]. Near-nerve conduction using very fine electrodes is also very accurate.

If a nerve problem is identified early, it can be treated by slowing the rate of distraction. However, if symptoms persist or motor signs develop, then peroneal nerve decompression should be performed [76, 77]. The timing of nerve decompression affects the rate of recovery of neuropraxia, and an early decompression leads to faster recovery. The peroneal nerve should be decompressed at the neck of the fibula, including transverse fasciotomy of the anterior and lateral compartment and release of the intermuscular septum between these compartments [68] (see Fig. 22.16). When peroneal nerve decompression was performed on patients undergoing limb lengthening, intraoperative findings included hemorrhage, nerve flattening, narrowing of the nerve at the entrance of the fascial tunnel, and reduction of the perineural vascularization at the site of compression. These findings are typical of nerve entrapment and not of stretch injury. In addition, no relationship was found between nerve injury and the amount or percent of lengthening.

Poor or Failed 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 to continue lengthening. If the defect in the distraction gap does not fill, it will need bone grafting. Bisphosphonate infusion (e.g., zoledronic acid) can be used to prevent bone resorption while permitting bone formation [78, 79].

Complete failure of bone formation is very unusual. Partial defects, especially laterally, are common. Dynamization of the fixator and bone growth stimulators (e.g., 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.

There are two methods of bone grafting the regenerate:

1. *Autogenous cancellous bone grafting*. The distraction gap should be opened and the fibrous tissue excised, leaving behind any regenerate bone. Once all of the fibrous tissue has been removed, cancellous autogenous bone graft can be harvested and inserted. In most children, this is obtained from the iliac crest. In young children there may not be enough bone from the standard anterior iliac crest approach. More bone graft can be obtained by splitting the tables of the iliac crest and taking graft from above the acetabulum. This allows harvesting of a large cache of bone that is normally not available for grafting, providing enough bone to fill the distraction gap.
2. *Cortical allograft*. This technique is used when the distraction gap is very long and exceeds the available autogenous cancellous bone graft. The technique is credited to Wasserstein [80], originally from Latvia and later from Germany. He described inserting a slotted allograft into a distraction gap to create early structure and bridging of the gap. A non-slotted allograft can be used instead, stabilized with an intramedullary nail. BMP-2 can be added for both cancellous and cortical grafts.

Incomplete Osteotomy and Premature Consolidation

If there is a lack of separation of the osteotomy site after a week of distraction, it may be due to an incomplete osteotomy or a periosteal hinge that will not separate. Continued distraction can lead to an acute separation of the bone ends, which is usually very painful and may have an audible pop. The pain will continue unabated until the bone is acutely shortened by a few millimeters. It is important to advise the patient of this possibility. If the bone does not separate, or if the patient or parents wish to avoid a painful separation, a repeat osteotomy at the same site should be performed.

Overabundant bone formation may lead to premature consolidation. Watch for a mismatch between the distraction performed and the radiographic distraction. Increasing the distraction rate for a few days may prevent a premature consolidation in such cases. Patients should be checked weekly during this time. If premature consolidation occurs, a repeat osteotomy should be performed at a new location, as a repeat osteotomy through the original site is more likely to lead to failure of bone formation. Leave the fixator in tension, as the osteotomy will separate easier when cut under tension. After the osteotomy, the tension in the frame should be reduced to normal.

Hip Subluxation/Dislocation

There is often mild or moderate acetabular dysplasia in patients with CFD. The pattern is different than 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 [59]. The orientation of the sourcil should also be horizontal. If it is inclined superiorly, then the hip joint is potentially unstable even with a normal CE angle. It is always safer to err on the side of performing a pelvic osteotomy prior to femoral lengthening than to end up with a hip subluxation.

Hip joint subluxation or dislocation during lengthening is a dreaded complication. It is diagnosed radiographically. The earliest sign is a break in Shenton's line or increased medial head-teardrop distance indicates subluxation of the hip. The hip usually has an adduction and flexion contracture and may also exhibit stiffness with flexion and extension. Hip subluxation does not usually occur if there is adequate coverage of the femoral head, but they can occur if there are adduction and flexion contractures.

If hip subluxation occurs, distraction must stop. The patient should be taken to the operating room to reduce the hip. Release the adductor longus and gracilis, and the TFL and rectus femoris if needed. Abduction should reduce the hip subluxation. If not, 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. Pelvic fixation consists of at least two anterolateral pins between the two tables above the anterior inferior iliac spine followed by two off-axis lateral pins. The femur should be in 15–20° of abduction to the pelvis to maintain the reduction. A hip extension bar should be added to prevent flexion contracture with a hinge. Lengthening may resume with caution (Figs. 22.21, 22.22, and 22.23).

If a pelvic osteotomy is performed after lengthening, care must be taken not to force the hip into the acetabulum. Since the femoral head is osteoporotic, it can be easily crushed with attempts at reduction. If an acute reduction is quite difficult, then it may be safer to do a gradual reduction with the external fixation if the external fixator is still in place. A hip dislocation that occurs after frame removal should be treated with an open reduction, capsulorrhaphy, and femoral shortening and pelvic osteotomy. The femur should be osteotomized after the capsulotomy and the femoral head reduced into joint. The bone ends of the femur are overlapped to determine the amount of shortening required. It is essential to shorten. There must be no tension on the femoral head if it is to remain in the acetabulum. It is important to preserve the superior capsule; if released, it will lead to recurrent dislocation. If the hip is difficult to stabilize despite all of these measures, it can be tethered in place using a suture anchor in the cotyloid notch, with the suture passing through the fovea and out the lateral neck. This creates a mobile tether that prevents dislocation while limiting motion. This technique was developed by the senior author and has been used since 2002 (see Fig. 22.23).

Knee Subluxation/Dislocation

Most cases of CFD usually have hypoplastic or absent cruciate ligaments in the knee. The tendency towards flexion contracture during lengthening predisposes the tibia to posterior subluxation, which is the most common. Posterolateral or external rotatory subluxation may occur with a tight fascia lata and biceps. Anterior or anteromedial subluxation can result from extension contractures and patella alta.

A more proximal femoral lengthening is less likely to cause knee subluxation, but it often has narrower and less well-formed regenerate bone, leading to higher fracture risk.

To prevent knee subluxation, the distal fascia lata should be released and the external fixator should span the knee and include the tibia. If the fascia is not released, then the knee can begin to subluxate after only 2–3 cm of lengthening. Without tibia fixation, the knee will often start to subluxate after 4 cm of lengthening. The spanning fixator also protects the knee cartilage and physis from increased forces. An articulated hinge can be used to allow the patient to maintain knee range of motion. Subluxation may be prevented by keeping the knee in full extension throughout lengthening, but it will result in a stiff knee. In the knee, posterior or anterior subluxation can be monitored on a lateral knee radiograph in full extension [81]. If the knee begins to subluxate, the rate of lengthening may need to be decreased as well as more intensive physical therapy for stretching.

If the knee subluxates or dislocates after removal of the external fixator, it should initially be treated by aggressive physical therapy, traction, and splinting. Physical therapy measures include pulling the knee forward and rotating it inwards during knee extension and flexion range of motion. If the knee remains subluxated despite an adequate trial of several months of therapy, it requires a version of the Superknee procedure called a "rescue knee." This involves lengthening of the biceps femoris and the iliotibial band if it is intact. In some cases, the Langenskiöld is done to reduce the patella into joint. If there is a fixed flexion deformity, the femur must be shortened proximally to avoid creating a quadriceps lag. In the most severe cases, a posterior capsulotomy is required. If the knee also has an extension contracture, this can be combined with a Judet quadricepsplasty (Fig. 22.24).

Fig. 22.21 The MRS articulated across both the hip and the knee with fixation to the pelvis and tibia, respectively. Both the knee and hip external fixation hinge axes correspond to the knee and hip center of rotation axes in the frontal plane

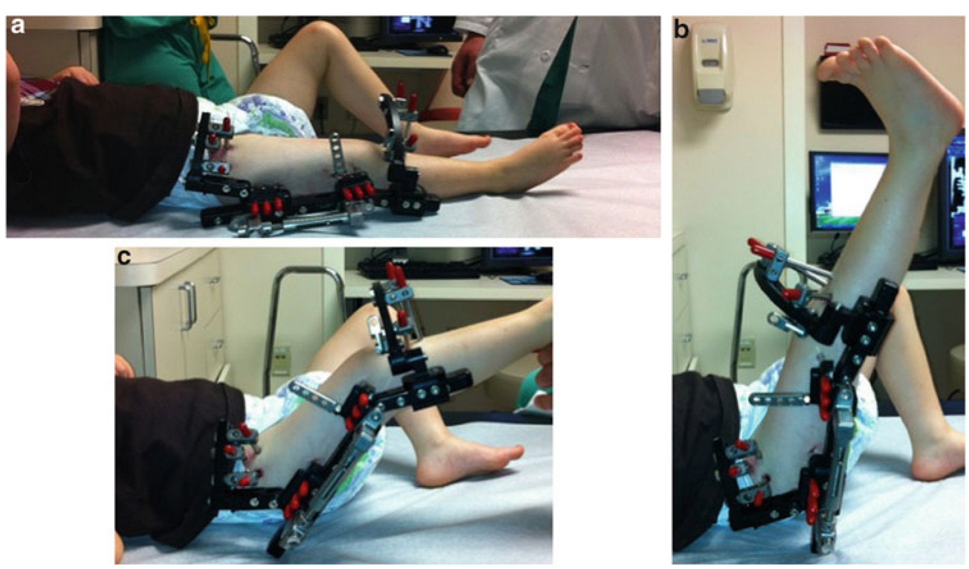

Fig. 22.22 MRS device in place on a 2-year-old boy being lengthened for CFD. The external fixator spans the hip and knee with a hinge connection to the femoral rail. This construct is shown with the hip and knee in extension (**a**), hip flexion (**b**), and hip and knee flexion (**c**)

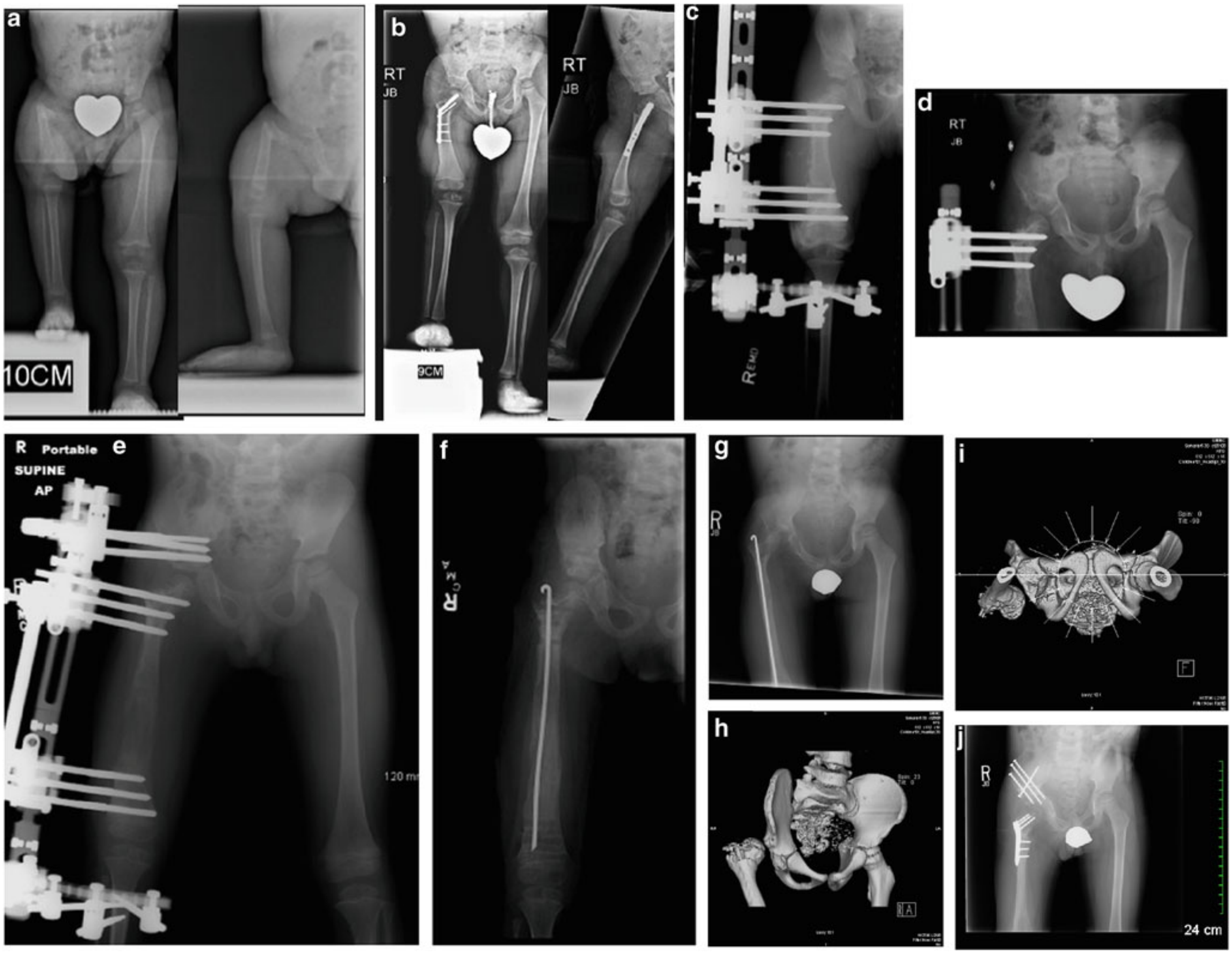

Fig. 22.23 Radiographs of a 2-year-old boy with CFD Paley type 1b subtrochanteric type deformity (**a**). Following Superhip and Superknee reconstruction including Dega pelvic osteotomy (**b**) at age 2. Lengthening of the femur at age 3 years, using the MRS device articulated across to the tibia (**c**). Radiograph after 4 cm of lengthening demonstrating early lateral subluxation of the hip. Note the increased femoral head-tear drop space (**d**). Extension of the external fixator to the pelvis with articulated spanning fixation (**e**). The femoral head is well reduced and stabilized permitting continued lengthening up to 8 cm. Radiograph at the time of removal of the external fixator with Rush rodding to prevent fracture. Note that the hip appears well reduced and stable (**f**). Radiograph 3 months later showing dislocation of the hip joint (**g**). Three-dimensional CT scan showing that the femoral head is dislocated posteriorly (**h**, **i**). Note on the axial view (**i**) that the posterior wall of the acetabulum is hypoplastic. Thus, the acetabulum is retroverted despite excellent superior and lateral coverage from the previous Dega. This confirms the statement that the Dega osteotomy cannot improve the posterior coverage of the hip. A "rescue hip" procedure was performed. The femur was shortened 2 cm and fixed with a plate. An open reduction of the hip was performed. To tether the femoral head into the acetabulum a suture anchor was used from the cotyloid notch through the femoral head and secured over a washer laterally. This washer can just be seen protruding at the upper end of the plate. A periacetabular triple osteotomy (according to Paley's modification, Fig. 22.12) was performed and secured with screws. The hip remains stable and mobile. Illustration of the open reduction, shortening, and tethering technique (**k**–**t**). After the pelvic osteotomy, capsulotomy, and subtrochanteric osteotomy the femoral head can be reduced into joint. The quadrilateral plate is drilled (**k**). The perforation seen from the inside of the pelvis (**l**). The femoral head is drilled through the fovea (**m**). After drilling both the femur and the acetabulum (**n**). The suture anchor is introduced into the medial wall of the acetabulum (**o**). The author's current preference is to use the Juggerknot (Biomet, Warsaw, IN). This has a soft anchor made of nonresorbable suture material that bunches up when pulled back through the hole (**p**) locking on the inside of the quadrilateral plate of the pelvis. A Keith needle is used to pull the suture through the femoral head and neck and greater trochanter (**q**, **r**). The suture is tied over a washer to secure the reduction of the femoral head. The hip remains mobile but not dislocatable. The distal femur can be overlapped on the proximal femur to determine how much shortening is required (**s**). The distal femur is shortened and fixed to the plate

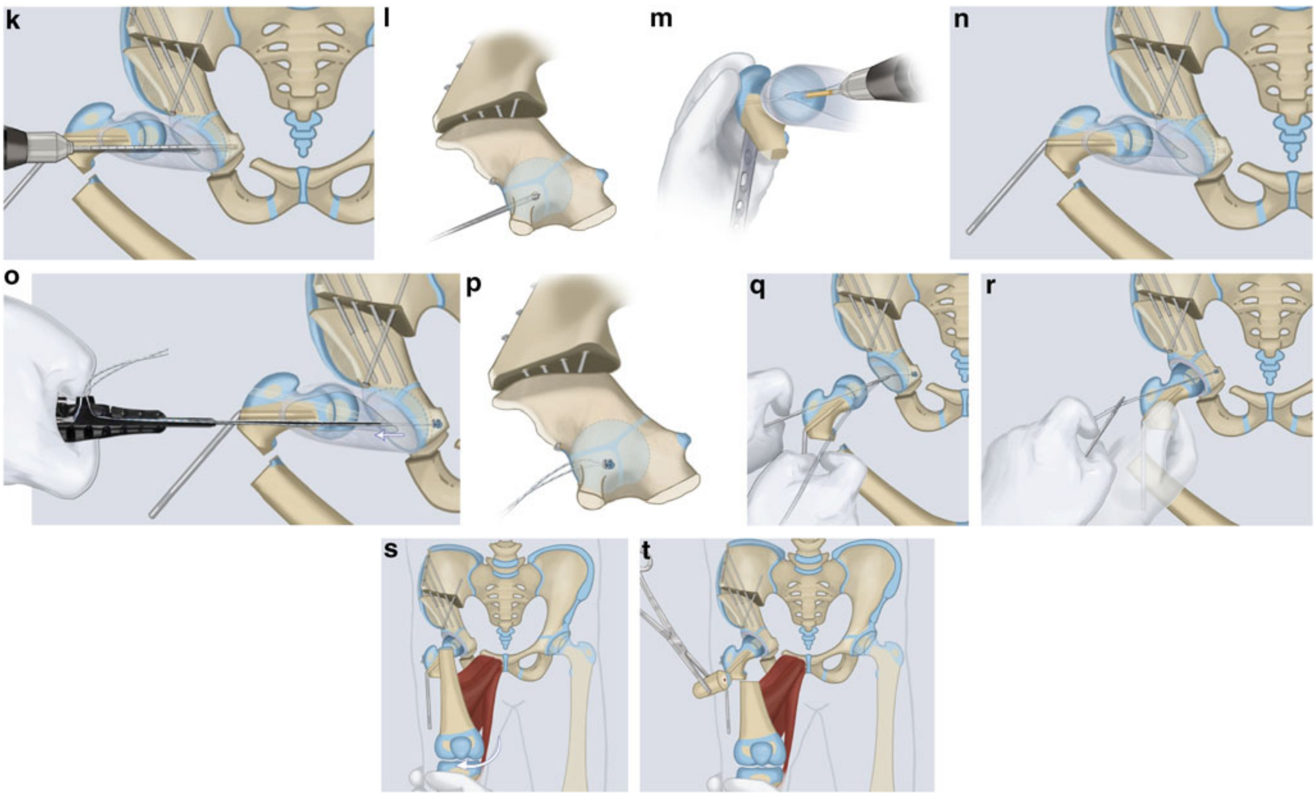

Fig. 22.23 (continued)

Limb Malalignment

Limb length equalization should be based on full-length standing radiographs. Limb alignment is assessed for the femur and tibia both separately and in combination. The joint orientation of the knee should be measured using the malalignment test [82].

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. Residual malalignment after the external fixator is removed and can usually be addressed with hemiepiphysiodesis or repeat osteotomy.

Fractures

Fractures associated with limb lengthening can occur throughout and after the lengthening process. Fractures can occur through pin holes or be pathologic secondary to disuse osteoporosis. A fall on a frame can cause a femoral neck fracture or a pin hole fracture, usually the second most proximal pin. Physical therapy manipulation can cause flexion buckle fractures of the proximal tibia or distal femur. An unexpected muscle spasm during frame removal can cause flexion failure of the femur through a pin hole or the regenerate. After frame removal, fractures can occur through pin holes or the regenerate bone, usually at the mid-regenerate area or host bone junction. The incidence of all these type of fractures associated with CFD lengthening was 34 % compared to 9 % for non-congenital femoral lengthening [83–85]. In many cases, this was despite the use of a spica cast after removal.

By prophylactically rodding the femur at the time of removal, the risk of refracture is much lower [53]. There is still a concern for intramedullary infection, so proper irrigation, debridement, and curettage before nail insertion are warranted. Patients are given intraoperative antibiotics and a 10-day course of oral antibiotics.

Prophylactic rodding permits continuation of knee mobility after removal. To protect from osteoporotic stress fractures through the tibial pin holes we hold formal physical therapy for a month but permit the patient gentle range of knee motion. Rodding also permits weight bearing with a removable spica cast right after frame removal. Fractures may still occur after rodding if the rod is too small or too short. The appropriate diameter rod should be used in each case. In most small children the 1/8″ (3.1 mm) Rush rod is used. In larger children the 3/16″ (4.65 mm) rod is preferable and in older children the ¼″ (6.2 mm) Rush rod is chosen. In general, the Rush rod is passed as distal as possible to the edge of the distal femoral physis.

Alternatively, a prophylactic submuscular plate can be used when the pin sites are too infected to allow safe rodding (Fig. 22.25).

Fig. 22.24 Thirteen-year-old girl with CFD undergoing lengthening over nail using Orthofix LRS device articulated across to the tibia to prevent knee subluxation (**a**). After the nail was locked and the fixator removed, the knee dislocated by an external rotatory subluxation including dislocation of the patella. Note that this radiograph is a true AP of the femur but an oblique of the externally rotated tibia (**b**). This radiograph is a true AP of the tibia but an oblique of the femur, which is relatively internally rotated to the tibia (**c**). AP and lateral radiographs of the knee after a Superknee procedure was performed to "rescue" the knee. AP and lateral radiographs of the knee (**d**) after a Superknee procedure was performed including the Paley reverse MacIntosh procedure, which in this case was secured with two medial suture anchors in the medial femoral condyle. Because she was skeletally mature an Elmslie-Trillat transfer of the tuberosity was performed instead of a Grammont. Note that the tibia is congruently reduced to the femur. The patella is back in place using the Langenskiöld procedure. This surgery was performed by the author in 1994. The patient maintains excellent stable and painless range of motion to the present date (**e**, **f**)

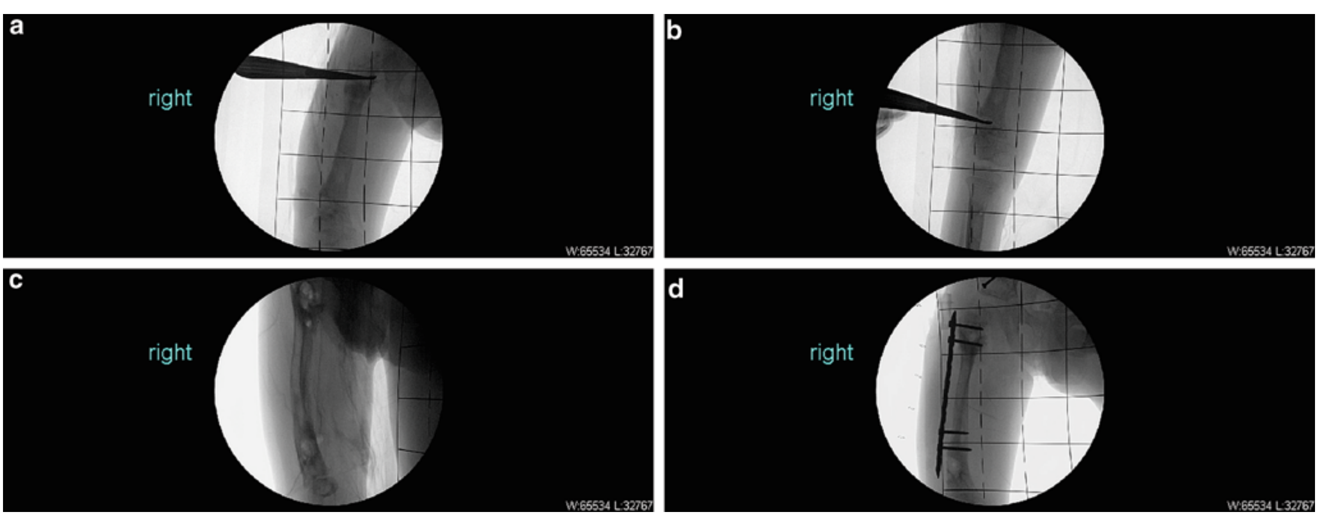

Fig. 22.25 Significant pin-site osteolysis after 8-cm femoral lengthening in a 3-year-old boy after previous Superhip and Superknee procedures. The pin sites are debrided including curettage of the bone, at the time of removal of the external fixator (**a**, **b**). Lateral radiograph showing the holes in the bone that make it at high risk of fracture. Due to the contamination of the bone by the pins the risk of intramedullary infection with Rush rodding was considered to be too high in this case. To prevent fracture a submuscular locking plate was used instead. This avoids contact with the pin sites

Joint Stiffness and Contracture

The knee can develop both flexion and extension contractures during lengthening. Although transfixing pins or wires may contribute to difficulty in flexion, even lengthening with internal distractors may still lead to loss of knee flexion with increased length. However, stiffness of the knee is preventable. Surgical release and lengthening of specific soft tissues (fascia lata, rectus femoris) reduce the joint reactive forces on the knee due to lengthening. They can be done acutely with the index procedure, but the soft tissues may heal together prior to the end of distraction. A delayed release at about 6 weeks allows more soft tissue distraction since the release occurs when the soft tissues are taut from lengthening.

Dynamic splinting can be done by using commercially available products or office-customized splints such as developed by Bhave [86]. The custom knee device consists of two fiberglass casting material cuffs for the thigh and lower leg connected by cast brace hinges. The elastic forces are applied using elastic bandages anteriorly off of various towers suspended proximal and distal to the knee. This is a very efficient and inexpensive type of dynamic splinting.

Physical therapy is essential to successful CFD lengthening. This is especially true for the knee, and one should not consider lengthening a CFD case without outpatient PT. If the knee gets stiff in flexion despite adequate rehabilitation, then a quadricepsplasty should be performed. Rozbruch recommends doing this at or shortly after frame removal [87]. He cuts the central tendon of the quadriceps while leaving the medial and lateral musculature intact. He claims that this does not lead to a quadriceps lag. This author prefers to do a full Judet quadricepsplasty [88] instead. With a concomitant flexion contracture, either an external fixator with gradual distraction or an open posterior capsule release can be performed.

Difference in Treatment of Types 1a and 1b

In general, Paley CFD type 1a (normal ossification) has less deformity, deficiency, and discrepancy in the hip and knee, in comparison to type 1b (delayed ossification). The distinction between types 1a and 1b should be made while the patient is in infancy, because the natural history of type 1b is to ossify. Therefore, adult type 1b cases may appear to be severe type 1a cases. Most type 1a cases do not require the complex Superhip reconstruction. In the author's experience, approximately half do require pelvic osteotomy before lengthening, and all cases should have extension of the fixator across the knee to protect the knee joint.

The strategy of treatment for type 1b is to correct all the associated deformities, which will allow the proximal femur and hip to be more normally oriented and accept more axial loading. The response to the anatomic change is ossification of the proximal femur, converting the CFD from type 1b to type 1a. This conversion usually occurs within 2 years of the Superhip procedure, and no lengthening is attempted until conversion. The first lengthening can be performed between the ages of 2 and 4, once the femur is of adequate size. If initially seen in infancy, patients with type 1a CFD typically undergo their first lengthening at age 2 years, whereas patients with type 1b CFD typically undergo lengthening closer to age 4 years.

Treatment of Paley CFD Type 2 (Fig. 22.26)

In comparison to the Paley type 1b neck type, which is sometimes referred to as a stiff pseudoarthrosis of the femoral neck, the Paley type 2 CFD is a mobile pseudoarthrosis of the femoral neck. In type 2 there is a fibrous anlage of the femoral neck present extending from the base of the greater trochanter to the femoral head. There may be a well-formed synovial joint present with a mobile femoral head (type 2a), partially formed synovial joint with a partial fusion of the femoral head to the acetabulum at one spot (type 2b), or an absent or circumferentially fused femoral head (type 2c). The defining anatomic feature that characterizes all three of these is the presence of a greater trochanteric apophysis, which is missing in Paley type 3. Radiographically, the apophysis can be recognized when the proximal end of the femur appears rounded, while in most Paley type 3 cases, the upper end of the femur appears sharp and pointed. An MRI makes this distinction much more definitive.

There are three approaches for treatment for Paley type 2: (1) rotationplasty versus Syme amputation and prosthetic fitting, (2) serial lengthenings with pelvic support osteotomy at skeletal maturity, or (3) Superhip 1.5 or Superhip 2 procedures for selective cases of types 2a and some type 2b.

Superhip 1.5 Procedure for Treatment of a Mobile Proximal Femoral Pseudoarthrosis with a Mobile Femoral Head with Cartilaginous Remnants of the Femoral Neck Present (Fig. 22.27)

This procedure was developed in 2010. It is used when there is a type 2b with part of the femoral neck present on the femur and the femoral head. It is carried out similar to the Superhip 1, but with exposure of the anterior hip capsule. An arthrotomy is made, and the ends of the femoral neck are identified. A blade plate is used as described previously,

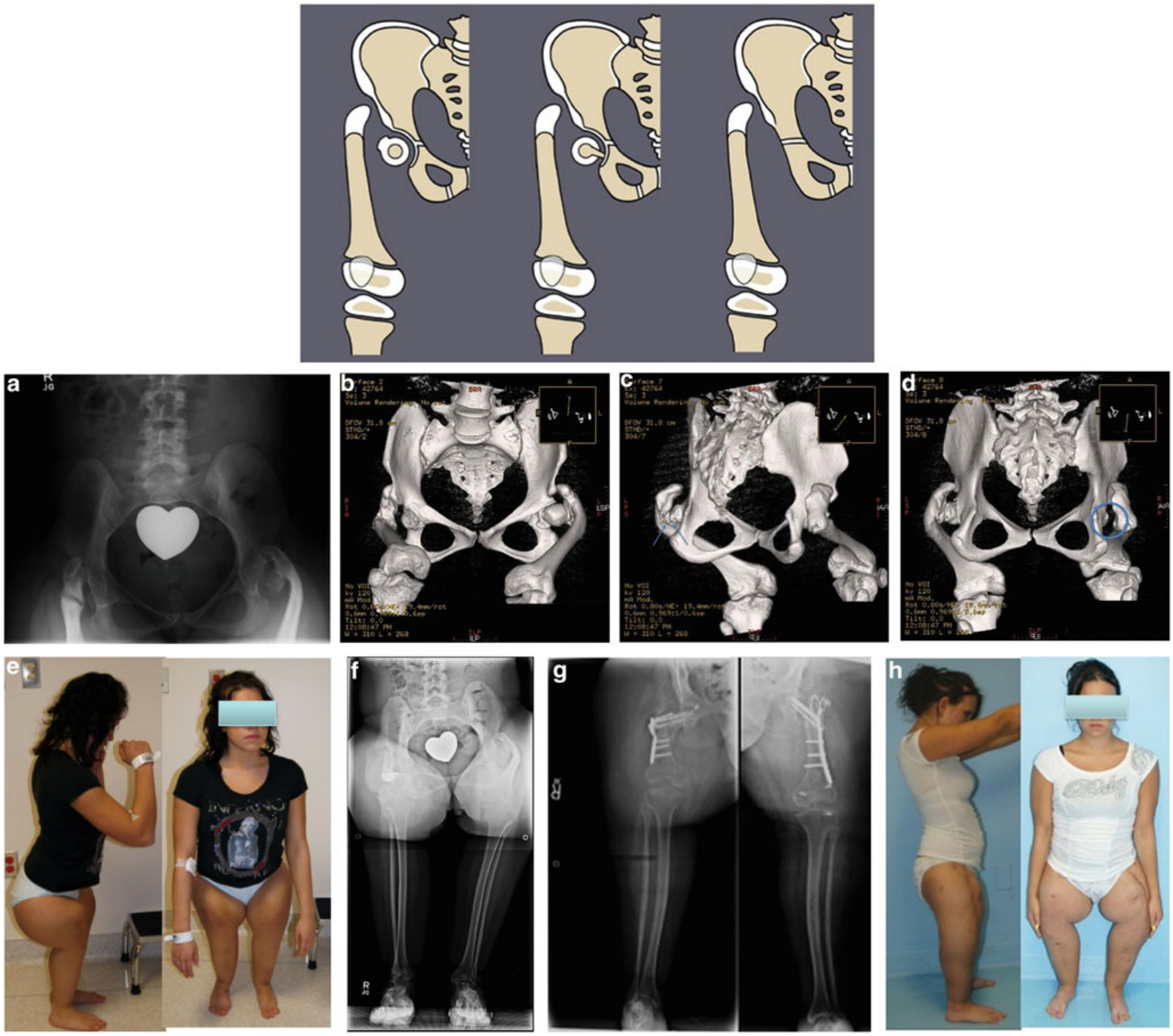

Fig. 22.26 An 18-year-old girl with bilateral Paley type 2a CFD. AP radiograph of pelvis shows femoral neck remnants on either side of the pseudoarthrosis while on the left side there is no femoral neck (**a**). Three-dimensional CT scan cuts show the femoral neck remnants from the front (**b**), oblique (**c**, *arrows*), and back (**d**) views of the right hip compared to a pseudoarthrosis without any neck remnant on the left (**d**, *circle*). Her standing posture is shown clinically (**e**) and radiographically (**f**). She has fixed hip and knee flexion contractures and very disproportionately short thighs. Note that her hands reach down below her knees. Due to the fixed flexion deformities, she fatigues when walking in a crouch gait. The goal of treatment was to make her stand and walk erect by eliminating the flexion deformities of both hips and knees. This was achieved by the Superhip 1.5 on the left and, the Superhip 2 on the right, and the Superknee with capsulotomy bilaterally. The posttreatment radiographs showing the new erect posture and the healed femoral neck pseudoarthrosis are also seen (**g**). Her clinical photograph after recovering from bilateral Superhip procedures. She now walks longer distances without fatigue (**h**). Since she stands and walks without a crouch gait her upper extremities hang to a more normal level than before

except that the cannulated chisel is introduced to each segment independently. The blade plate is then inserted across to stabilize the two parts. BMP-2 is used to induce the two parts of the cartilaginous femoral neck to ossify and unite. This is referred to as "cartilage welding." This is a unique feature of BMP-2 that was first identified by the senior author when treating the unossified femoral neck of type 1b [14] (see Fig. 22.24) [14].

Superhip 2 Procedure for Treatment of a Mobile Proximal Femoral Pseudoarthrosis with or Without Partial Fusion of the Femoral Head (Figs. 22.28 and 22.29)

The same approach as the Superhip is used. The operation remains the same until the subtrochanteric osteotomy. Before performing the osteotomy, the region of the femoral neck is

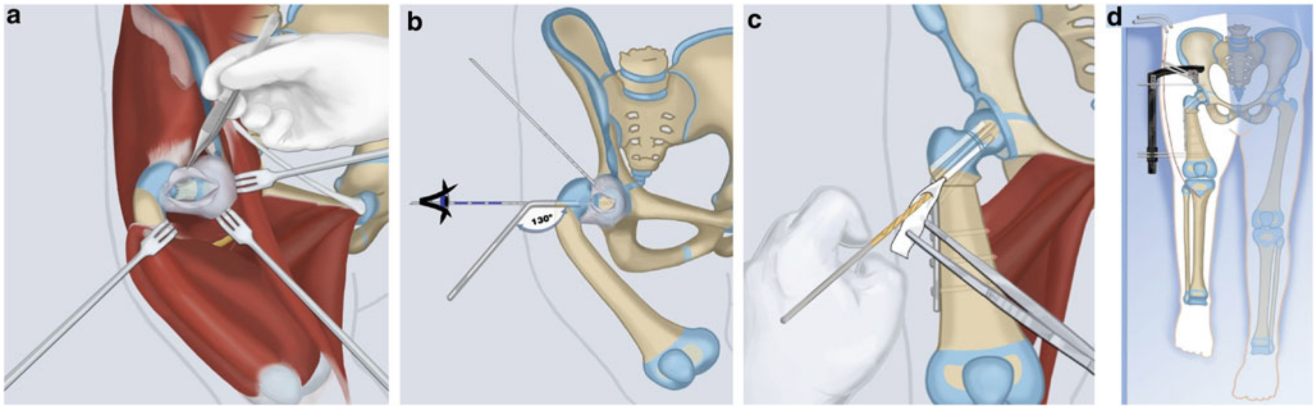

Fig. 22.27 Superhip 1.5 technique. (**a**) This is indicated for Paley type 2a when there is a remnant of cartilaginous femoral neck remaining on the proximal femur and femoral head. Through the same surgical exposure of the Superhip procedure (see Fig. 22.7) the anterior capsule of the hip is exposed by first separating the quadriceps from the gluteus medius. The capsule is then opened in line with the femoral neck. (**b**) The fibrous anlage is resected and the cartilaginous parts of the femoral neck are approximated. (**c**) The femur is osteotomized, shortened, and corrected as for the Superhip procedure and fixed with a blade plate. BMP-2 is inserted into the superior and inferior neck across the cartilaginous pseudoarthrosis. It is used to "weld" the cartilage together by inducing it to form bone. This is an off-label use of BMP-2. (**d**) The forces on the femur are neutralized by a spanning external fixator from the pelvis to the femur

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 inferiorly. The femoral head is moved in the acetabulum with forceps. If it is type 2a, it will be mobile. If it is type 2b, it will not move. If there is a cartilaginous or bony bridge, it is always located between the femoral head and the ischium. The cartilage of the head is cut back to the ossific nucleus to expose bone. Small 1-mm slices are cut back until the ossific nucleus is widened to about 2/3 of the femoral head. Dental wire may be passed through the nucleus for additional fixation.

The femoral neck is made from the proximal femur and greater trochanter. Posteriorly, the interval between the vastus lateralis and the abductors is found, and all of the abductors are released off of the proximal femur. The vastus lateralis and medial femoral circumflex artery serve as the pedicle for this bony segment. The external rotators are also removed. The proximal lateral cortex is partially decorticated, as it will eventually be oriented inferiorly and attached to the femoral shaft. The first cut is made about 4–5 cm long, perpendicular to the femoral shaft, creating the new femoral neck. This segment is rotated 135° on its soft tissue pedicle; the greater trochanter moves distal and lateral, and the distal cut end of the subtrochanteric osteotomy is rotated superiorly to fix to the ossific nucleus of the femoral head.

The trochanteric segment is predrilled with a 1.5-mm drill in four places around its cortical periphery, and 5/64″ threaded K-wires are run retrograde through the femoral neck and set to the same length. This bone is then drilled at 45° to its long axis for insertion of a Rush rod. This is drilled in a way to miss the path of the four wire holes and into the area previously decorticated. The distal femur intramedullary diaphysis is prepared with a 3.2-mm drill bit. Dental wire is also placed around this segment underneath the soft-tissue pedicles, in preparation for fixation.

The new femoral neck is then attached to the head. The four threaded K-wires are advanced carefully, holding the head with the dental wire. Verify that the wires are not intra-articular with fluoroscopy and a Freer elevator. After the threaded K-wire fixation, carefully tighten down the dental wire, compressing the femoral neck to head junction.

The second subtrochanteric osteotomy is then made at 45° to the shaft of the femur, removing a trapezoidal piece to provide adequate shortening of the femur. The cut should be made with the leg internally rotated to create femoral neck anteversion. The Rush rod is then placed through the trochanteric piece into the femoral shaft, and the dental wire tightened down. A cannulated 4.0 screw may be placed perpendicular to the shaft cut at 45° to augment fixation if there is adequate room. The abductor slide is performed as needed, and the abductors are repaired to the greater trochanter. To neutralize the forces on the femoral head and neck, an external fixator is applied from the pelvis to the tibia for 3 months.

Pelvic Support Osteotomy

A pelvic support osteotomy is used to treat most type 2c and some type 2b cases, combined with a distal femoral lengthening and realignment osteotomy (Fig. 22.30). This combination is called the *Ilizarov hip reconstruction* [89]. Pelvic support osteotomy is usually enough to prevent proximal migration of the femur during lengthening for non-congenital pathologies.

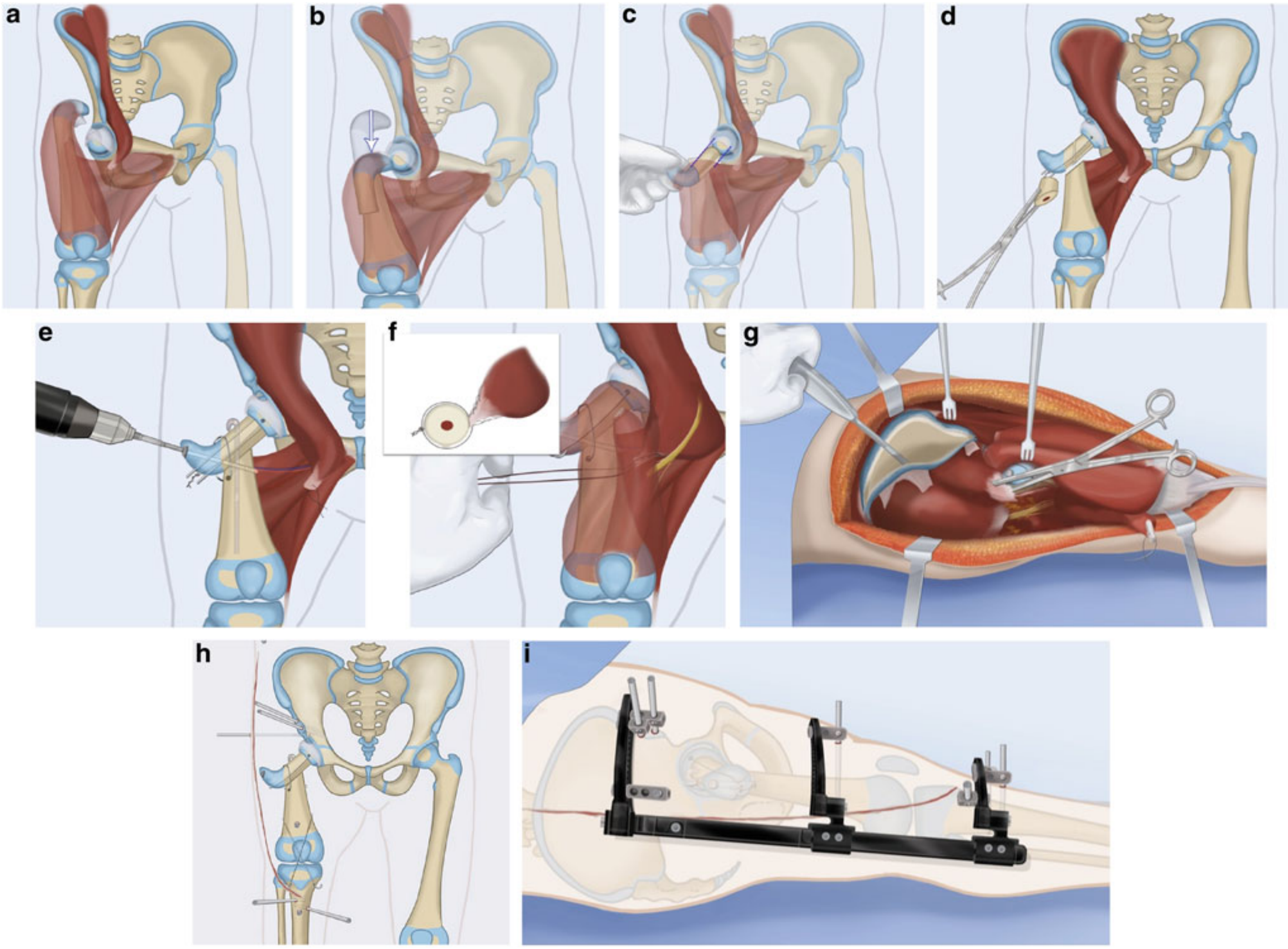

Fig. 22.28 Superhip 2 (SH2) technique. (**a**) After the same exposure as in the Superhip (see Fig. 22.7). The capsule of the hip is exposed without taking down the quadriceps muscle. The psoas tendon is released from the lesser trochanter. The femoral neck anlage is resected. The inferior capsule is opened. (**b**) All of the muscles inserting into the proximal femur are detached including all of the glutei, external rotators, and psoas. The muscles that originate on the proximal femur such as the quadriceps are not disturbed. The quadriceps becomes the vascular pedicle for the proximal femur and greater trochanter. The femur is osteotomized in the subtrochanteric region. It is then mobilized and moved distally. (**c**) The proximal femur is rotated 135° to form a femoral neck. (**d**) The femoral neck is fixed to the head using threaded K-wires and a cerclage wire. The second osteotomy of the femur is at 45° with the distal femur rotated 15° internally for anteversion. The femur is shortened according to the soft-tissue tethers. (**e**) The femur is fixed using a Rush rod, tension band wire, and compression screw technique. (**f**) The psoas tendon is reattached. (**g**) The hip abductors and rotators are reattached. If the abductors are too short, then the apophysis is split and an abductor slide is carried out. (**h**) A spanning external fixator from the pelvis to the femur to the tibia is used. In most cases there is a fixed flexion deformity of the knee. A posterior capsulotomy Superknee procedure is performed and pinned with cross K-wires to hold the extension correction. The capsulotomy and knee extension should be performed after the first osteotomy but prior to the shortening osteotomy of the femur. (**i**) MRS device used to span the hip and knee joints and neutralize the forces on the femoral neck for 4 months

Fig. 22.29 (continued) contraindication for the Superhip 2 procedure. Modification of the technique has made it a viable option in the author's current clinical practice. The critical consideration is rotating the fused area of the femoral head to face and join to the "new" femoral neck. Intraoperative fluoroscopic image (**g**); during this procedure, after fixation with threaded K-wires, cerclage wire and Rush rod are in place. After application of the spanning external fixator (**h**, **i**). Knee capsulotomy and acute extension of the knee flexion contracture were carried out and temporarily fixed with cross K-wires (**h**, **i**). Final radiograph after removing the external fixator (**j**). Note that the Rush rod reaches to the distal femoral physis. Radiograph 2 years later showing that there has been a lot of growth of the distal femur as seen relative to the end of the Rush rod (**k**). Hip and knee range of motion after recovery from Superhip 2 procedure (**l**, **m**). First lengthening of the femur with MRS and articulated fixation spanning the knee joint. The hip joint was left free (**n**). The femur was successfully lengthened 8 cm with no loss of hip or knee motion

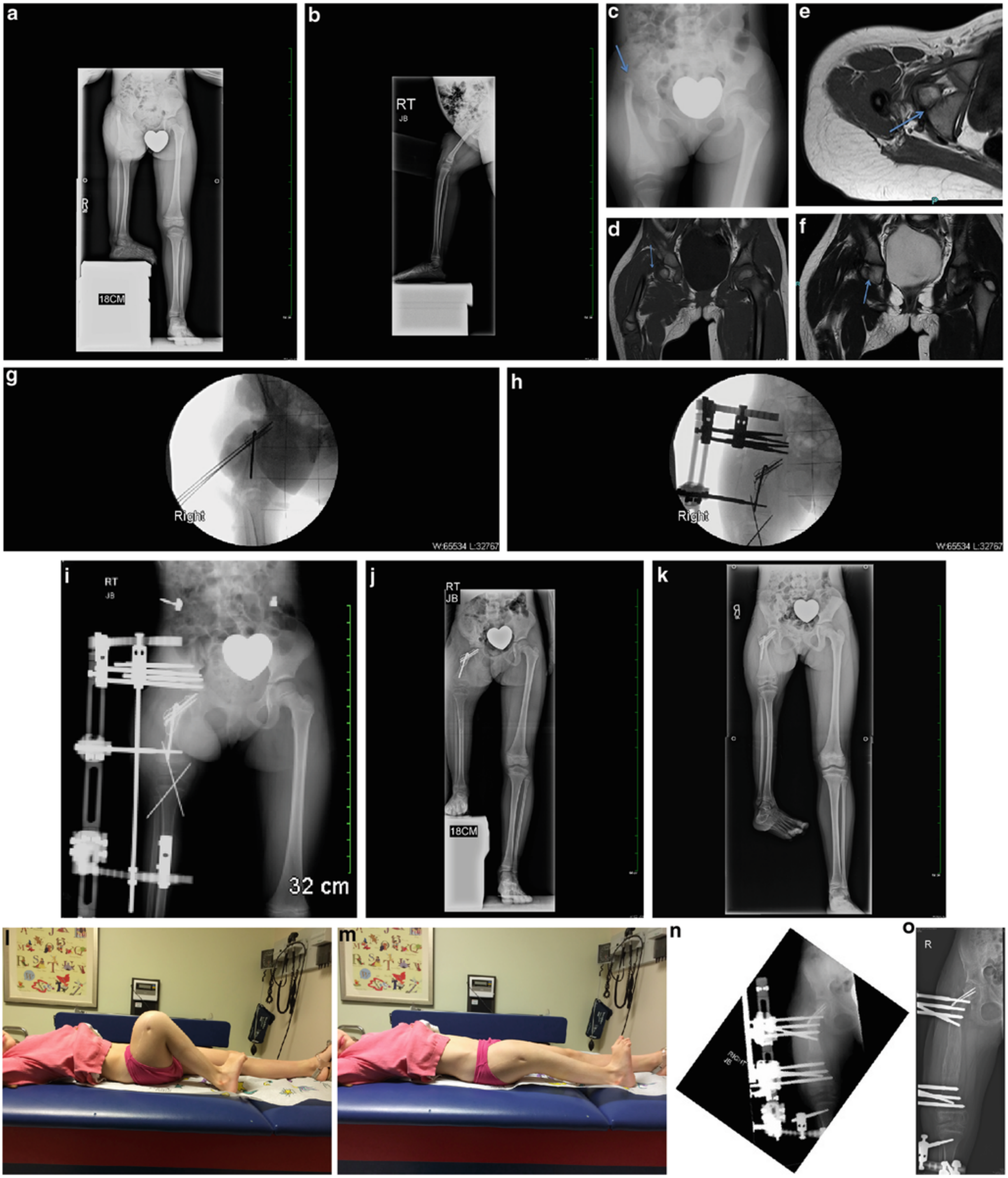

Fig. 22.29 Standing AP and lateral radiographs of patient with Paley type 2b CFD (**a**, **b**). AP pelvis radiograph showing the rounded proximal end of the femur indicating that it has an apophysis. It is proximally migrated. There is an obvious femoral head ossific nucleus with a well-formed acetabulum present. The *arrow* is pointing to the greater trochanteric apophysis (**c**). MRI coronal cut, showing the ossific nucleus with no bony or cartilaginous connection to the proximal femur (*arrow*). The greater trochanter is out of plane for this cut (**d**). MRI transverse cut showing the femoral head in the acetabulum (**e**). Note the bridge of bone posteriorly between the femoral head and the ischium (*arrow*). The rest of the femoral head is covered with cartilage and remains unfused. The same can be seen on the coronal cut of the posterior femoral head (*arrow*) (**f**). This is therefore classified as a partial fusion of the femoral head (Paley type 2b). Type 2b was originally thought to be a

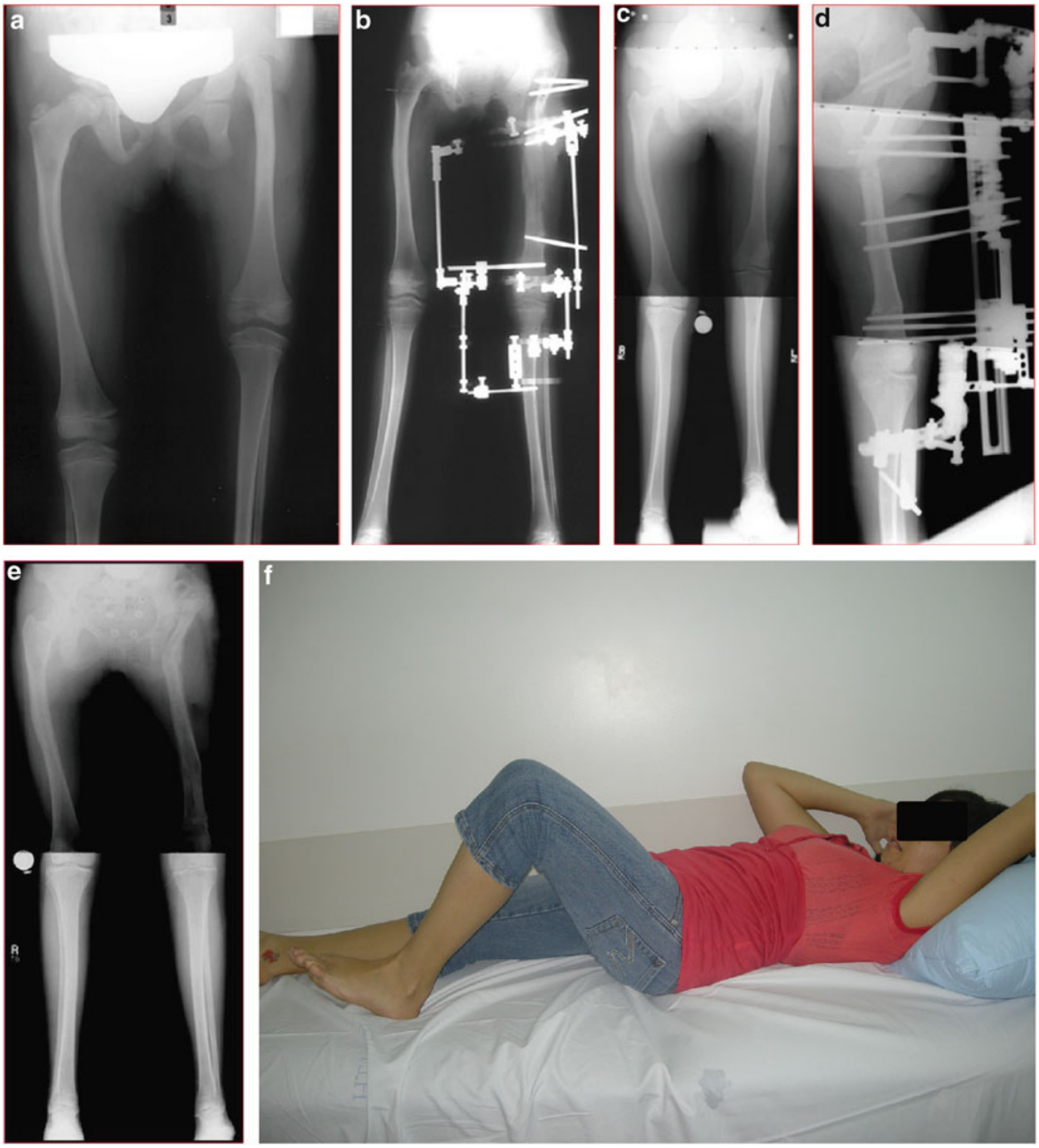

Fig. 22.30 AP radiograph (**a**) of 10-year-old girl with Paley type 2c CFD left side (absent femoral head). She was treated by pelvic support osteotomy (**b**) and distal femoral lengthening (13 cm). This is an unusual and extreme amount for one lengthening. The only reason we exceeded the 8-cm limit is that she was able to maintain knee range of motion to 90° and for financial reasons was not sure if she would ever be able to afford travelling for another lengthening. We therefore continued lengthening as long as she was able to maintain knee motion. 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 (**e**). She has excellent function (**f**) and walks with no appreciable limp. She is now 20 years from her original surgery

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. In addition, the valgus component of the pelvic support osteotomy may remodel straight.

The proximal osteotomy is performed at the level at which the proximal femur crosses the ischial tuberosity in maximum cross-legged adduction. The amount of valgus is equal to the total amount of adduction of the hip plus 15° of overcorrection. The proximal osteotomy should also be internally rotated and extended. The amount of rotation is judged by the position of the knee relative to the hip in maximum adduction. The amount of extension depends on the amount of hip fixed flexion deformity.

The level of the distal osteotomy is determined by the intersection of the proximal and distal mechanical axes. The proximal mechanical axis passes through the proximal

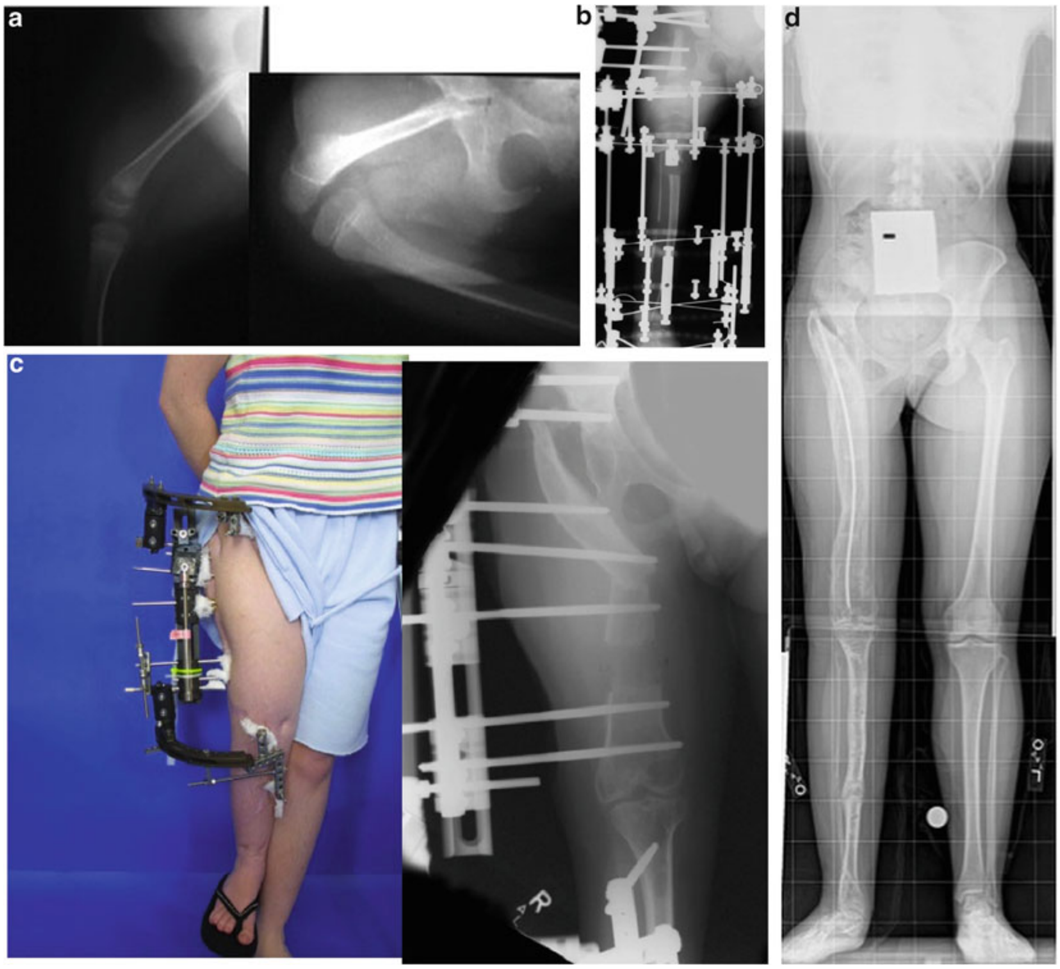

Fig. 22.31 (**a**) CFD Paley type 3A. Note the initial knee flexion and extension for an arc of motion of 70°. Parents refused to have a Syme's or a rotationplasty. She was treated at age 7 with a lengthening of the femur and tibia for a total of 12.5 cm (**b**). At age 10 she underwent the same treatment 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 (**c**). At age 18 she underwent her fourth femoral plus tibial lengthening of 12.5 cm. She had a final tibial lengthening of 2.5 cm to correct deformity and equalize her leg lengths (**d**). Her total lengthening was 50 cm. She maintains 70° of knee motion and good ankle and hip motion. She has excellent clinical function, has no pain, and walks with minimal limp. She is now 18 years following her first procedure

formal osteotomy site and is perpendicular to the horizontal line of the pelvis. The distal axis is from the center of the ankle to the center of the knee. Varus correction can be made through the distal osteotomy. The external fixator must still be extended to the tibia with hinges, as previously discussed.

Treatment of CFD Type 3

Deficiency of the proximal femur with absent femoral head, greater trochanter, and proximal femoral metaphysis results in a mobile pseudoarthrosis and a very short femoral remnant. The Paley type 3a CFD has a mobile knee with at least 45° of motion, usually with a 45° knee flexion deformity. The type 3b has a stiff knee with less than 45° of motion and usually has a greater flexion deformity. The most predictable and reliable treatment option in these cases remains prosthetic reconstruction surgery with either rotationplasty or a Syme amputation. Limb reconstruction surgery has a role in these cases and can equalize an LLD. Because the number of these patients treated this way is small, the ultimate functional result for these cases is still not predictable.

Limb Reconstructive Surgery for Type 3

Limb reconstructive surgery is most applicable to type 3a cases that have functional knee range of motion (Fig. 22.31). Many of these patients have knee and hip flexion contractures. These are treated by soft tissue releases, using the same approach as described above for type 1 CFD. 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.

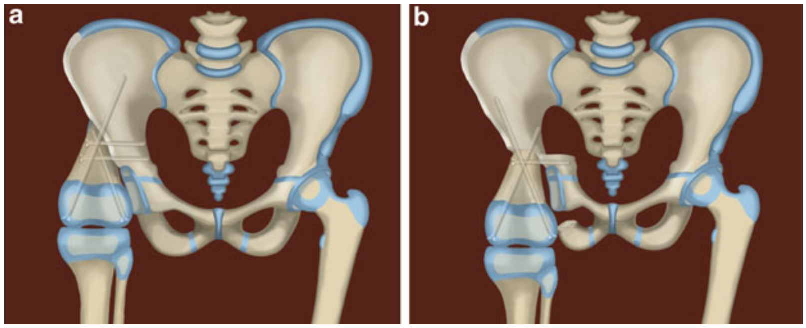

Fig. 22.32 Rotationplasty for CFD type 3. (**a**) Brown rotationplasty. The femur is fused to the side of the pelvis. It is difficult to align the femur perpendicular to the pelvis. The hip protrudes laterally due to its lateral fusion position. (**b**) Paley-Brown rotationplasty. A Chiari osteotomy is used to fuse to the femur. The ischium is notched to make room for the sciatic nerve

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 for the acute correction of the knee contracture. The long lateral incision is extended distally and the peroneal nerve is decompressed and protected. Because the femur is so short, it is advisable to explore the peroneal nerve and follow it proximally to the hip joint region prior to significant release. This will prevent injury to the sciatic nerve, as it passes very near the dissection around the hip capsular remnant.

The lateral approach to knee flexion contracture release of the posterior capsule is performed, and the knee joint is fully extended. A 2-mm Steinmann pin may be 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. Once fixation is removed, range of motion exercises are started to regain knee motion.

After the above procedures, the femur can be 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 too 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 the same as a type 2b: serial lengthenings and eventually a pelvic support osteotomy (Fig. 22.31) combined with a 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.

Prosthetic Reconstructive Surgery for Type 3

Prosthetic fitting is possible without any surgery. However, this can be difficult with a hip and knee flexion contractures, as well as a foot not in equinus. Surgical releases as described above may help with prosthetic fitting. A Syme or Boyd amputation may also help create a residual limb that is fitted easily. In some cases, even a pelvic support osteotomy may be considered to decrease limp and stabilize the hip joint.

The other approach to prosthetic reconstruction is with rotationplasty [49, 90]. Torode 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°, such that the foot points backward and the ankle can 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 [44] modified the rotationplasty approach, using a racquet-like incision, performing the rotation between the femoral remnant and the pelvis (Fig. 22.32a). 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. However, one problem with the Brown method is excessive shortening of the hip muscles and lateralization of the hip and lower limb. It is also challenging to adjust the alignment correctly.

Paley modified the Brown technique in 1997 by adding a Chiari osteotomy of the pelvis and fusing the femoral remnant to the cancellous roof of ilium (Figs. 22.32b and 22.33). The detached muscles are not removed and are instead transferred distally as much as possible. Care is taken to appropriately shorten the knee muscles so that they can function adequately as hip flexors and extensors. The fascia lata connected to the gluteus maximus is reattached to the tibia to serve as a hip abductor. Posterior capsule release of the knee is performed as the knee is often contracted as much as 90°. The peroneal nerve is decompressed to prevent injury and

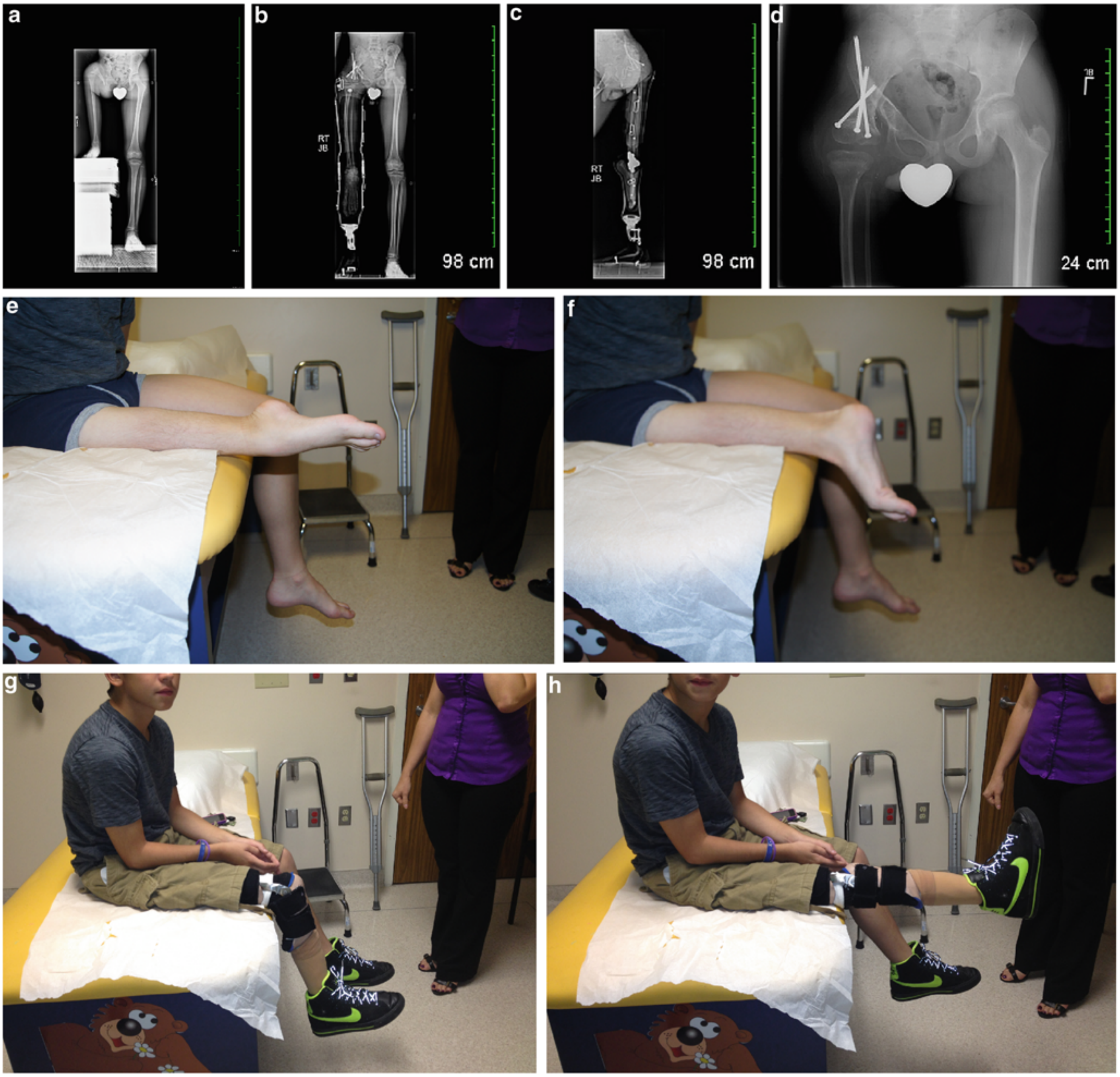

Fig. 22.33 (**a**) Preoperative radiograph of a 10-year-old boy with CFD Paley type 3A. Note how the knee is fully flexed and the hip is rotated into a frog leg position. Standing AP (**b**) and lateral (**c**) radiographs after Paley-Brown rotationplasty using the prosthesis. The ankle is at the level of the opposite knee. Closeup radiograph (**d**) showing the fusion to the pelvis and the closure of the distal femoral physis. Sitting posture with excellent hip flexion and active "knee extension" (via ankle plantar flexion) (**e**), and "knee flexion" (via ankle dorsiflexion) (**f**). Active extension (**g**) and flexion (**h**) of the knee wearing the prosthesis

allow greater rotation. A supramalleolar osteotomy may be performed to correct the internal rotation deformity of the tibia. Epiphysiodesis is done with the same screws used for fixation of the femur to the pelvis. This modified Paley-Brown rotationplasty provides good functional results with better hip function and stability than the Van Nes and the Brown rotationplasties.

The senior author has recently developed a new rotationplasty variant, which for labeling purposes will be called the Paley rotationplasty (Fig. 22.34). The steps are the same as in the Brown, but instead of fusing the femur to the pelvis, the femoral segment is fixed to a mobile femoral head. This gives better hip reconstruction with active abduction motion present. Even if the femoral head is fused to the acetabulum, as in

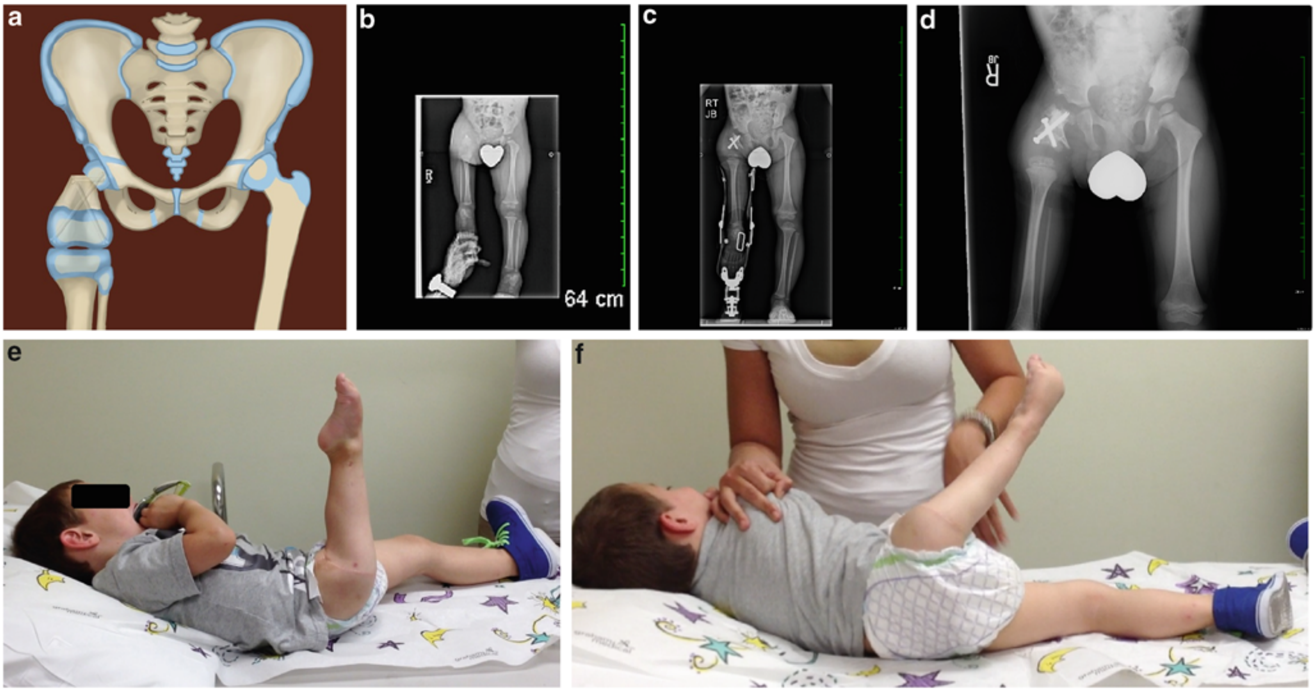

Fig. 22.34 (**a**) Paley rotationplasty. The femoral remnant is fused to the femoral head, which is made mobile in the acetabulum. (**b**) 2-year-old boy with type 3b. (**c**) Paley rotationplasty was performed and the femoral head was fused to the femoral head (**d**). He has excellent active hip flexion (**e**) and abduction (**f**) of the hip joint. Hip flexion is usual but hip abduction is not usually possible after a Brown rotationplasty

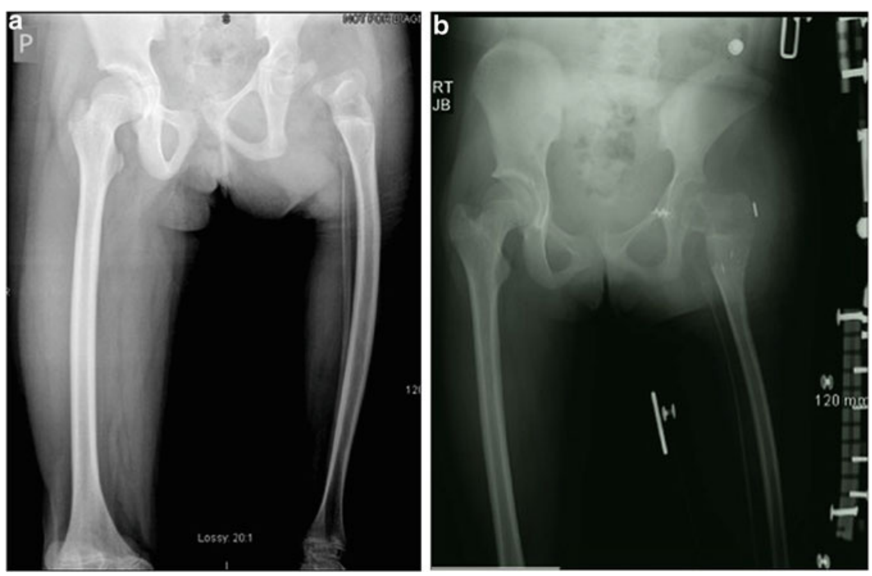

Fig. 22.35 Twelve-year-old girl with CFD Paley type 3c (**a**). She had a rotationplasty performed with insertion of the proximal medial tibia into the acetabulum (**b**). This gave her a stable hip with excellent function of the hip and knee joints

type 2b, it can be freed up similar to the Superhip 2 and then fused to the femoral remnant (see Fig. 22.34). Finally, in cases where there is no knee joint present (type 3c), the proximal tibia or fused distal femoral remnant can be fused to the residual femoral head. If there is no femoral head, then a roof can be created by a Chiari osteotomy or by creating a depression into the pelvis (Fig. 22.35). This is stabilized by a suture ligament tether technique as previously shown (see Fig. 22.23).

Summary

CFD is a spectrum of congenital deficiency, deformity, and discrepancy of the femur that involves not only the osseous part of the femur, but also the surrounding musculature, ligaments, and joints. All of these components must be addressed if one is to be successful in the treatment of CFD. Though the definitive origin of CFD is still not known, it is a problem that can affect the function of a patient for his or her entire life. Depending on the severity, there are several treatment strategies, reconstructive surgeries, and prosthetic options, all of which can improve a patient's gait, function, and quality of life. As each deformity is different, so also is each patient and family, and therefore treatment should be tailored to their individual needs and cultural expectations. Advances in surgical techniques, new biologic agents, and new technologies have expanded the reconstructive indications and options in lieu of amputation. Though more research of newer treatments and their long-term outcomes are needed, there are now more options and hope for a brighter future for a child with CFD.

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