lateral (postaxial) rays missing, a shortened leg and (often) thigh, a valgus knee, and variable anterior bowing of the tibia with a dimple over the apex. Further examination will demonstrate anteroposterior (AP) instability of the knee (but no instability to varus/valgus stress testing) along with a small patella. In more subtle cases, the only clinical feature may be a limb that is mildly shorter than the contralateral extremity. Although equinovalgus foot deformity is usual, occasionally equinovarus can be seen, especially in these cases (78, 79).

Radiographic Features. Radiographically, the fibula will usually be seen to be shortened in relation to the tibia. This may occur either proximally or distally or both. Often a portion of the fibula is absent in part (Fig. 30-7) or in its entirety (Fig. 30-8). In those cases in which the fibula is of normal or near-normal length, the diagnosis can be difficult during the first year of life. Radiographically, the condylar notch of the femur is shallow and the tibial spines are small.

The femoral shortening may be slight to severe, varying from a few centimeters of shortening to a true PFFD. Amstutz (33) reported femoral deficiency in 15% of those with fibular deficiency, whereas Bohne and Root (80) reported femoral deficiencies in almost two-thirds of their patients. Kalamchi noted that 70% of type I and 50% of type II deficiencies were associated with shortening or deformity of the femur (81).

Radiographs of the knee demonstrate a small hypoplastic lateral femoral condyle (82). The ankle may appear normal in some patients with mild deficiencies, but by skeletal maturity there is usually a valgus deformity. The classic appearance of the ball-and-socket ankle joint in these deficiencies is seen in Figure 30-6C,D. There is disagreement about the origin of this abnormality. Some authors feel that it is congenital (83), whereas others feel that it develops secondary to the tarsal coalitions (84). If it is caused by the tarsal coalition, it is difficult to explain its absence in tarsal coalitions without fibular deficiency. In slightly more severe cases, if the fibula is present, it is short and may not reach the level of the ankle joint. The distal tibial epiphysis shows a triangular appearance.

Although not seen on radiographs at birth, tarsal coalitions are present in most of the feet associated with fibular deficiency. Grogan et al. (85) noted such coalitions in 54% of anatomic specimens, although the abnormality could be seen radiographically in only 15% of the specimens because of the cartilaginous nature of the coalition. Finally, radiographs demonstrate missing lateral rays, which are easily seen clinically without radiographs.

Pathoanatomy. The femoral intercondylar notch narrowing and the tibial spine hypoplasia are indicative of dysplasia of one or both cruciate ligaments. Recently, Manner et al. have correlated the degree of plain radiographic abnormality to the absence of one or both cruciate ligaments as seen by magnetic resonance imaging (MRI) (86). In some cases of fibular deficiency, there is a residual fibrocartilaginous anlage present. Whether or not this anlage causes progressive tibial deformity with growth is unresolved in the literature. Some authors suggest it acts as a growth tether and recommend it be surgically removed primarily (87). Other studies showed no evidence that resection of the anlage made a clinical difference (88).

Natural History. In general, the shortening seen in these patients is progressive according to the rule of proportionality as previously mentioned. With regard to the valgus knee, the deformity usually worsens with growth (36, 80, 89, 90). In some cases, the degree of valgus is more severe than can be explained by the smaller lateral femoral condyle alone, and its recurrence after correction speaks of a more dynamic cause. There is inconclusive evidence to suggest that the anteromedial tibial bow, which is occasionally seen, changes over time. However, clinical experience with untreated older children with fibular deficiency often reveals that the valgus deformity of the ankle progresses over time and can become painful. This is likely a consequence of both the lack of lateral ankle supporting structures and a growth asymmetry at the distal tibial physis. It can also be secondary to a progressive valgus deformity in the mid-diaphysis of the tibia in rare cases where the fibular anlage acts as a lateral growth tether.

Treatment Recommendations. The main problems in the treatment of fibular deficiency are the limb-length discrepancy and the deformity and instability of the foot and ankle. It is very important to realize that the discrepancy will become worse with growth, and it is the ultimate discrepancy at maturity that is important.

Nonsurgical Treatment. Although the majority of patient with fibular deficiency are treated surgically, there are rare cases that are not. If the child has a functional foot without significant valgus deformity, and the degree of shortening would result in an ultimate discrepancy of <2 cm, then no surgery is indicated. Rather, a contralateral shoe lift and serial monitoring during growth for progressive knee or ankle deformity is the preferred treatment.

Surgical Treatment. For the vast majority of patients with fibular deficiency, surgical management is indicated. The great difficulty in treating these children is not deciding which patients would benefit from surgery, but rather which surgical treatment pathway is best for any one patient.

Amputation versus Lengthening. Until the 1960s, amputation for fibular deficiency was recommended only as a last resort (91). In a reaction to the results of these early attempts to save the limbs, several reports emphasized the advantages of amputation for severe cases (36, 92–94). The indications are based primarily on the difference in length and the functionality of the foot. Wood et al. recommended amputation for: a discrepancy of 3 in. or more at the time of decision, or if predicted at maturity; for a nonfunctional foot; for a limb that would have severe cosmetic or functional problems; or for children who cannot endure the psychological trauma of repeated hospitalizations and surgery (94). These recommendations were reaffirmed in a