

publication from the same institution, following up many of the same patients (36).

More recent recommendations begin to stretch the extent to which length can be restored, reflecting improvements in limb lengthening. Westin et al. (36) suggested amputation for any discrepancy that would be >7.5 cm at maturity (36). For Letts and Vincent (21), the number was >10 cm, and for Hotnick et al. (95) it was between 8.7 and 15 cm.

Although modern prosthetics have made amputation a somewhat more acceptable alternative, the improved ability to lengthen limbs has also made limb salvage a more feasible option. The recommendations of Birch et al. are an effort to account for these changes (76). They would recommend amputation for those with a nonfunctional foot, regardless of leg length, unless the upper extremities were nonfunctional. For those with a functional foot, but a leg-length discrepancy of 30% or more, amputation would be recommended. For those with a functional foot and a discrepancy of $<10\%$, epiphysiodesis or lengthening is reasonable. There is little disagreement about these indications today.

It is between those two groups that the controversy regarding treatment lies, and the greater the discrepancy in length, the greater the controversy. According to Birch et al., those patients with a functional foot and a discrepancy between 10% and 30% are candidates for either amputation or lengthening (76). The parents, who are the decision makers, are weighing the hope for their child to retain the limb against what that will entail. Without knowing what a child with an amputation and prosthesis versus a lengthened limb is like, their first response is almost always to lengthen the limb. They most likely have never seen a child or adult with an amputation; they visualize something horrible. At the same time, they cannot really know what a lengthened limb will be like at the end of treatment; they imagine the limb will be normal. Although they may understand that they will need two or three lengthening procedures, they cannot know what the impact will be on their child or their family, what complications they will encounter on the way, or how their child will look or function at the end of the treatment.

As yet, there are but a few preliminary reports of lengthening in fibular deficiencies with predicted discrepancies >10 cm. These preliminary reports, using the Ilizarov methods, deal mainly with the extent of length achieved, often before maturity, but with little information on cosmetic and functional result (96–100).

One way to begin to assess the problem is to look at what amount of length is required. The combined femoral and tibial length for a girl of average height at maturity will be approximately 80 cm (37) (Table 30.2). A 10% discrepancy would be approximately 8 cm, a 20% discrepancy would be 16 cm, and a 30% discrepancy would be 24 cm. To achieve >10 cm of length in a congenital limb deficiency with AP knee instability, ankle instability, foot deformity, and congenitally short soft tissues are a significant undertaking (100–102).

Reports comparing Syme amputation with lengthening are few and incomplete, but begin to give an appreciation

of the problems associated with lengthening severe deficiencies (71, 103–105). These reports conclude that lengthening should be reserved for those with more normal feet and less discrepancy in length, although early Syme amputation is the best treatment for the more severe problems. Herring gives a philosophical perspective on the dilemma (106). Birch et al. (107) reviewed a series of adults who were treated with Syme amputation in childhood. These authors conducted physical examination, prosthetic assessment, psychological testing, and physical performance testing and commented that the results of multistaged lengthenings for this condition would have to match these results to be justified. They currently offer lengthening to patients whose limb-length discrepancy is 20% or less.

Bilateral. In patients with bilateral fibular deficiency, the three problems are the foot deformity, the discrepancy in length between the two limbs, and the overall shortening in height because of two short limbs. Without extenuating circumstances (e.g., nonfunctional upper extremities), disarticulation of the foot and prosthetic fitting is the best solution. For those children with nonfunctional upper extremities who will use their feet for many of the activities of daily living (ADL), amputation of the feet is not an option.

In children with bilateral fibular deficiency, there is usually little discrepancy between the two limbs, but rather a discrepancy between their height and what their normal height should be. As they enter into their peer group, this becomes an increasing problem. This problem is most easily solved by the prosthetist. If there is a significant difference between the length of the two limbs that cannot be solved by prosthetic adjustment, lengthening of the short limb becomes an attractive option.

Syme and Boyd Amputation. The amputation described by Syme (108) seems to have been accepted for adults before it was accepted for children, and its use in boys was advocated before its use in girls because it was said that the Syme amputation produced an unsightly bulkiness around the ankle. This resulted in many children receiving a transtibial amputation rather than a Syme amputation. It was subsequently learned, however, that the ankle does not enlarge following amputation in a young child, and the cosmetic appearance is excellent as the child grows.

Thompson et al. were the first to recommend the Syme amputation, rather than transtibial amputation, although only as a last resort (91). Subsequent reports by Kruger and Talbott (93) and Westin et al. (36) not only confirmed the advantages of the Syme amputation in both boys and girls but also advocated its early use for severe deficiencies. Several studies now confirm the value of Syme amputation (90, 93, 106, 109–113).

One of the major advantages of the Syme amputation is the ability to bear weight on the end of the residual limb. This is important both for prosthetic use and for instances in the home when the child will walk short distances without the prosthesis (for instance, going to the bathroom in the middle of the night). It is also relatively technically easy to perform.