Limb Reconstruction Surgery for Fibular Hemimelia

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Fibular Hemimelia is the most common lower extremity congenital longitudinal deficiency (frequency 1/40,000 live births). . It is associated with a constellation of deformities including foot ray deficiencies, subtalar coalition, ball and socket ankle joint, ankle joint malorientation, diaphyseal angular deformity apex anteromedial, fibular deficiency. .
Congenital shortening of the tibia associated with congenital fibular deficiency is referred to as fibular hemimelia.

The best known classifications of fibular hemimelia focus on the degree of fibular deficiency (1,2,4,10) . Most of these classifications were designed at the time where amputation and prosthetic limb equalization were the only reliable treatment (15). They are descriptive of the fibular pathologic abnormality, which does not require reconstruction, and they lack description of the tibial pathologic abnormality, which is the focus of reconstruction. Furthermore, they do not help in planning reconstruction of the foot. Severe rigid and intractable equino-valgus deformity of the foot in combination with a limb length discrepancy has been the limiting factor for a successful outcome and the primary indication for ablative surgery. There have been concerns about inhibition of the tibia growth after lengthening (ref Sharma).

Paley classification of fibular hemimelia

Type 1: Stable normal ankle

Type 2: Dynamic valgus ankle

Type 3: Fixed equino-valgus ankle

Type 4: Fixed equino-varus ankle (clubfoot type)

The Type 3-fixed equino-valgus ankles are subdivided into 4 types according to ankle-subtalar pathoanatomy.

Type 3a-ankle type: the ankle joint is maloriented into procurvatum and valgus.

Type 3b-subtalar type: the subtalar joint is has a coalition which is malunited in equino-valgus with lateral translation

Type 3 c-combined ankle and subtalar: combination of the ankle and subtalar deformities above.

Type 3d-talar type: malorientation of the subtalar joint.

Surgical technique for type 3 ankles:

THE SUPERANKLE PROCEDURE (developed by Dr. Paley in 1996)

Under tourniquet control, a longitudinal lateral incision is made parallel to the posterior cortex of the tibia and is extended distally to the calcaneus. At the distal end care should be taken not to cut the sural nerve. The distal sensory branch of the superficial peroneal nerve is identified and dissected free. Proximal dissection should not be done before finding this nerve. It runs obliquely across the operative field as it exits the deep fascia. The peroneal tendon(s) are identified, and if both are present, they are sutured together proximally and distally and and the longus cut proximally and the brevis distally. If only one peroneal tendon is present a z-lengthening of this tendon is performed. The fibrous peroneal tendon sheath is excised. The distal fibular cartilaginous anlage, if present, is dissected free from all except a distal flap of soft tissue. It is then reflected distally on this calcaneo-fibular ligamentous pedicle to expose the ankle joint capsule. The fibrous fibular anlage is in continuity with the interosseous membrane. Both are dissected to the level of the apex of the diaphyseal procurvatum. .The interosseous membrane is resected up to this level but the fibrous fibular anlage should be removed along its entire length. The proximal part of the fibrous anlage is exposed using a second incision at the proximal tibia. The second incision is made parallel to the oblique course of the peroneal nerve. The peroneal nerve is identified, decompressed and dissected free of the anlage. The fibular anlage is then dissected from proximal to distal to communicate with the distal dissection. The anlage is dissected free from the overlying muscles and the interosseous membrane by tunneling between the two incisions. It is then detached proximally and removed as a single unit distally. . The anlage is preserved in saline solution for later use. The interosseous membrane distally is also resected The Achilles tendon is then exposed. Before lengthening this tendon the posterior tibial nerve should be identified. It is located immediately anteromedial to the Achilles. The posterior tibial neurovascular structures should be dissected free from the Achilles and then a Z-lengthening of the Achilles tendon is performed, leaving the distal attachment medial if possible. If the tendon inserts laterally and the muscle medially, Z lengthen the tendon and leave it attached laterally distally. Lengthen the Achilles tendon approximately 4 cm in an infant. Identify and resect all fascial bands inserting onto the dorsum of the body of the calcaneus. The fascia surrounding the neurovascular bundle should be freed from its posterior attachment to the calcaneus. This decompresses the tarsal tunnel.

The ankle level is identified but the ankle capsule does not need to intentionally be opened. A small opening in the capsule may occur when the cartilaginous anlage is reflected. An ankle arthrogram can be performed to visualize the orientation of the distal tibial articular surface. If the ankle joint surface is laterally and posteriorly oriented. the equino-valgus should be corrected by supramalleolar osteotomy (Type 3a). To prevent recurrent deformity it is important to pin the ankle and subtalar joints in the position of equino-valgus. Two 1.5mmK-wires are drilled into the calcaneus and across the talus and ankle joint stopping at the distal tibial growth plate. Using an osteotome or a saw a supramalleolar osteotomy is performed from posterolateral to anteromedial. It should converge on the anteromedial distal tibial physis. An acute opening wedge is performed in this oblique plane hinging on the anteromedial physis. A wedge shaped bone graft (autogenous iliac crest or allograft; our preference is adult fibular allograft) is inserted into the opening wedge defect. The K-wires are advanced across the osteotomy and graft into the diaphysis of the tibia. The foot should only be corrected to make the plantar aspect perpendicular to the distal tibial diaphysis.

If the ankle joint orientation is near normal but a subtalar coalition with lateral translation and valgus is present (type 3b), the osteotomy is made through the coalition of the posterior facet of the calcaneus. This osteotomy is made obliquely from superolateral to inferomedial. To displace this osteotomy the calcaneus should be levered distally to correct the equinus and foot height. Medial translation usually occurs automatically. . Although it is preferable to make this cut through bone, it may be necessary to do this through cartilage if there is no ossification of the calcaneus adjacent to the coalition. . The subtalar coalition osteotomy should not be into the neck of the talus. It ends in the sinus tarsi. This osteotomy is fixed with two K-wires from the calcaneus into the distal tibia. In some cases there is both a mal-orientation of the ankle joint and a malunion of the subtalar coalition (type

3c). In such case both the supramalleolar and the subtalar osteotomies should be performed.

In some cases instead of a coalition there is a hypermobile unstable subtalar joint (type 3d). This is due to increased inclination of the subtalar posterior facet. An arthrogram of the subtalar joint can be performed to confirm this deformity To correct this type of valgus deformity, the talus is osteotomized proximal and parallel to the posterior facet. This osteotomy does not extend into the neck of the talus. Reorienting the subtalar joint maintains subtalar joint mobility. The opening wedge is fixed with two K-wires. A bone graft is used to fill the opening wedge site.

After the osteotomy realignment is completed the cartilaginous fibular anlage if present is sutured with absorbable suture to the distal tibial epiphysis such that it abuts against the talus. The Achilles tendon and peroneal tendon(s) are repaired in a Z fashion. In most cases the Achilles tendon repair is end to end with no overlap due to the amount of equines that was corrected. The fibrous fibular anlage that was previously excised is sutured across this repair to help prevent disruption during lengthening. If the Achilles tendon is only laterally attached to the calcaneus, the anlage can be extended and sutured to the medial calcaneus. The incisions are then closed. During the procedure the tourniquet remains inflated only for the first hour and a half. The rest of the procedure is performed with the tourniquet deflated.

The only deformity remaining to be corrected is the diaphyseal one. This can be corrected acutely by removal of a wedge or trapezoidal segment, or gradually with a percutaneous osteotomy and application of an external fixator.

The Ilizarov fixator is applied, and the lengthening osteotomy is performed. If there is a diaphyseal procurvatum deformity, the lengthening osteotomy should be at its'apex. If there is no diaphyseal angulation, the lengthening osteotomy should be more proximal. Lengthening begins 4 days after surgery at a rate of 0.8 mm per day with the pediatric Ilizarov device in children under age six and 1 mm per day with the standard Ilizarov rings for older children and adults. If there is a diaphyseal deformity, it is corrected gradually, simultaneous with the lengthening. This usually requires 0.5 mm per day distraction at the two hinges and 2 mm per day at the posterior distraction rod. In toddlers, no more than 5 cm of lengthening is performed to avoid growth inhibition. If there is an internal rotation deformity, the frame is modified and the leg gradually rotated externally. In children younger than 8 years, hinges are applied to varusize the tibia to compensate for the expected valgus drift. Varusization and complete fibrous fibular excision are two recent modifications that were added to the treatment protocol after the results from this study were analyzed. More recently we prefer to use the Taylor Spatial Frame (Smith and Nephew Orthopedics, Memphis, Tenn.) because of its ability to perform angulation, length, rotation and translation without laborious time consuming modifications of the external fixator.

DISCUSSION

Lengthening reconstruction surgery for fibular hemimelia can be divided into two steps: foot deformity correction and limb length equalization. In the absence of foot deformity, limb lengthening can be performed in one or more stages depending on the amount of lengthening required. Most cases are treatable with one, two, or three lengthenings (7). If the foot deformity of fibular hemimelia is present without limb length discrepancy, the treatment consists of soft tissue and bone procedures to correct the deformity. Because the deformity is in a growing child, some recurrence rate is expected, as with clubfoot. When foot deformity is present in association with limb length discrepancy, the techniques used to correct foot deformity and the methods used for limb length equalization are combined (21).

Surgical releases for clubfoot correction have evolved from the Turco posteromedial release to the circumferential Cincinnati approach. The correction of equinovalgus for fibular hemimelia has also undergone evolution. Gradual distraction of the foot deformity, in our experience, led to a high rate of recurrence and degenerative changes. Extra-articular soft tissue releases (includingAchilles tendon, peroneal tendons, fibrous anlage, interosseous membrane remnant, and fascial bands) led to some improvement in foot position but was frequently associated with residual dynamic valgus deformity. Extra-articular soft tissue release combined with intra-articular release (posterolateral or circumferential ankle arthrotomy) provided a better correction but still led to recurrence and/or stiffness. Recurrence is not surprising, considering that soft tissue release does not address malorientation of the tibial ankle joint surface. The tibial plafond is oriented laterally and posteriorly, producing valgus and procurvatum deformity. This can be seen preoperatively using magnetic resonance imaging or arthrography. In older children in whom ossification of the distal tibial epiphysis has already occurred, direct measurement using the malorientation test for the ankle (measuring the anterior distal tibial angle on the lateral radiograph [normal range = 80–82] and the lateral distal tibial angle on the anteroposterior radiograph [normal range = 87–92]) objectively shows how much of the equinus and valgus is due to ankle joint malorientation (28). In our study, this was confirmed either by arthrography or by direct observation at the time of arthrotomy. More recently, we have begun using magnetic resonance imaging to visualize the cartilage orientation of the distal tibial articulation and the relationship between the talus and calcaneus .

Realizing that the ankle joint was maloriented led to increasing use of supramalleolar osteotomy in the latter patients of this study. One shortcoming of supramalleolar osteotomy is that after the ankle joint is reoriented, the distal tibial growth plate may be maloriented. Theoretically, this could lead to gradual recurrence. In practice mild recurrence of the valgus is occasionally seen and can be addressed at the time of the next staged lengthening. One way to avoid this theoretical problem is to perform an acute intraepiphyseal opening wedge osteotomy. Since the realignment is distal to the growth plate it does not malorient the physis. The limitation of the latter technique is the size of epiphysis. In most cases, it is too small at two years of age to perform and intraepiphyseal osteotomy.

The best treatment of foot deformity is a combination of soft tissue lengthening of the Achilles tendon and peroneal muscles and resection of the interosseous membrane, aberrant fascial bands, and fibrous fibular anlage combined with supramalleolar, subtalar coalition, and/or talus osteotomy to correct valgus and procurvatum. There is no need to resect any cartilaginous anlage. Instead, it can be moved distally and sutured to the distal tibial epiphysis to add to ankle stability or else discarded. Equinovalgus foot correction can be performed separately or in combination with lengthening. We prefer to combine it with lengthening because the soft tissues are loosest after release and, therefore, the lengthening is easier and less painful because muscle tension is reduced. An argument can be made that preservation of ankle motion might be better if the two procedures are staged. Furthermore this would permit foot deformity correction at an earlier age, followed by staged lengthening. Certainly this is the approach we take with the superhip procedure.

Biomechanics of Fibular Hemimelia foot Deformity Correction

In the normal ankle joint, the ground reaction force vector passes antero-medial to the ankle at the time of peak loading in single leg stance. The lateral path of this force vector is a product of the lateral offset of the calcaneus to the talus and ankle joint. This produces a lateral moment arm that is countered by the posterior tibial tendon. It also produces a lateral displacement force on the talus which is countered by the lateral malleolus. It is well recognized that even small amounts of shortening or lateral translation of the lateral malleolus lead to a lateral shift of the talus in the mortis (Yablon). Valgus tilt of the ankle plafond increases the valgus moment arm and force of lateral translation. With a combination of valgus of the plafond, lateral translation of the calcaneus and an absent fibula the ankle joint is potentially unstable and will eventually sublux or dislocate laterally especially if the extra-articular muscular forces increase due to lengthening.

Rationale of Osteotomy Realignment for Fibular Hemimelia

In fibular hemimelia the ankle joint is usually ball and socket. If one considers the tibial plafond like an acetabulum, the talus is only covered medially in such ankles. Varus osteotomy covers the talus. More importantly, supramalleolar varus osteotomy, medially displaces the entire foot including the calacaneus. The ground reaction vector is moved medially. The valgus moment arm on the ankle is reduced or eliminated. Despite the absence of the lateral malleolus there is no tendancy for the talus to shift laterally. Similarly, when there is a lateral translation malunion of a subtalar coalition the valgus moment arm is increased. Without a lateral malleolus the foot wants to rest in valgus and the talus wants to shift laterally. Medial translation of the calcaneus reduces the moment arm. In more severe cases a combination of medial calcaneal shift with supramalleolar osteotomy is required to fully reduce the valgus moment arm. Weber described a lateral malleolarplasty to treat fibular hemimelia. This does not reduce the valgus moment on the ankle. As discussed above, the lateral malleolus is not essential to the function of the ankle if the valgus moment is reduced. We see no indication for lateral malleolarplasty since it does not address the rest of the pathoanatomy of the equino-valgus deformity.

Finally, genu valgum also contributes to the valgus moment arm on the ankle. Therefore despite complete correction of the foot deformity, if the knee valgus is not corrected, the foot deformity may recur. It is therefore important to determine if the valgus knee is due to deformity of the distal femur, proximal tibia or both. Correction of these can be carried out using osteotomy or more simply by means of temporary hemi-epiphysiodesis. The latter is accomplished either by means of Blount’s staples or Peter Steven’s Eight Plate (Orthofix, Bussolongo, Italy).

Lengthening Reconstruction Surgery vs Amputation

Most authors agree that lengthening is the preferred treatment for patients with mild to moderate leg length discrepancy with mild foot deformities (Paley types 1 and 2). The controversial cases are those that include more severe foot deformities (Paley type 3, a–c) and greater leg length discrepancies due to more severe tibial inhibition or combined femoral and tibial discrepancy. Syme’s or Boyd amputation has been the conventional recommendation for these more severe cases.

The justification for amputation for the more severe cases has been the failure of most authors to obtain satisfactory results after limb lengthening. No one would dispute that amputation with prosthetic fitting requires fewer surgical interventions, requires fewer days of hospitalization, and is associated with a lower complication rate. Furthermore, no one would dispute that with availability of modern prosthetics, limb length equalization with excellent function can be achieved reliably in patients who have undergone Syme's or Boyd amputations (3,13,15). This does not prove, however, that the best treatment for severe cases of fibular hemimelia is amputation with prosthetic fitting. Excellent function could also be obtained if amputation and prosthetic fitting were used as the treatment of clubfoot, ankle arthritis, or other disabling foot conditions. This is a testimony to the excellence of modern prosthetics and nothing more.

The challenge, therefore, is not to compare the function achieved in cases of Syme's or Boyd amputation with that achieved in cases of lengthening but rather to improve the results of lengthening. Why are the results that are reported by many authors so poor? Is it because these cases are unreconstructable or is it because of fundamental errors in the treatment strategy? We think the latter is the case. An analysis of the unsatisfactory results reported in different series in the literature clarified that the overriding factor associated with poor results is recurrent or residual foot and tibial deformities. It is not the inability to obtain equalization of limb length. The few series in which good results were obtained, even in severe cases of fibular hemimelia, reported that the final result was a stable plantigrade foot (7,16,22). The total amount of discrepancy can always be equalized by serial moderate-sized lengthenings rather than by one very large lengthening. The foot deformity can be treated by various methods, including soft tissue and bone procedures. If these fail, ankle arthrodesis is a very successful way of permanently stabilizing the foot (7). It is clear that ankle arthrodesis should not be the indication for amputation. Therefore, because the worst-case analysis in stabilizing and correcting the foot deformity is ankle arthrodesis , there is no reason that the foot cannot be made stable in a plantigrade position.

Johnson and Haideri (20), using gait analysis, showed that patients in whom lengthening has resulted in plantigrade feet and well-aligned tibiae have better ankle push-off strength and better knee flexion strength than do patients who have undergone Syme’s amputation. They noted that the lengthened limb, even if it was stiff and weak, was less different from its opposite normal limb than was the prosthetic side in cases of Syme’s amputation, as compared with its opposite normal limb. They reported that the lengthened limb with a plantigrade foot was “clearly more functional than a prosthetic ankle.”

Naudie et al. (25) achieved satisfactory results in only four of 10 cases after lengthening. They compared their group with an amputation group and concluded that amputation was preferable to lengthening. The reason for the unsatisfactory outcomes was residual or recurrent foot and tibial deformities. Cheng et al. (8), in a small prospective group of four lengthenings, had the same experience with unsatisfactory results secondary to recurrent tibial and foot deformities. Both groups succeeded in achieving the limb lengthening amounts desired using the Ilizarov apparatus. These results using the Ilizarov apparatus are not much different from those reported by Choi et al. (9), who used the older Wagner method. All of the cases of higher grades of fibular hemimelia had unsatisfactory results in the study presented by Choi et al. These unsatisfactory results were attributed to rigid uncorrected equinovalgus deformity of the foot. They achieved satisfactory results in all except one of the patients with mild fibular hemimelia, a patient who had a rigid equinovalgus foot. They, too, concluded that the more severe grades of fibular hemimelia are not candidates for lengthening surgery and would be best served with amputation and prosthetic fitting.

After reviewing these reported results, we think that although the limb length can be successfully corrected in most patients, if the foot deformity is left uncorrected initially or after recurrence, the final functional outcome will be unsatisfactory (6,14). This conclusion is also valid for the treatment of clubfoot and vertical talus deformities.

If one examines the few series in the literature that report good functional results after limb lengthening, the predominant difference is that in the final result, not only was the leg length discrepancy addressed successfully but the foot deformities were also addressed successfully. Miller and Bell (22) reported the outcomes of 12 lengthenings in cases of fibular hemimelia. At the time of final follow-up, all limbs had regained full knee motion and all feet were plantigrade. All except three limbs had regained their preoperative range of ankle motion. None of the ankles had residual instability. Despite these excellent final results, 25 complications occurred in 12 lengthenings. These patients required eight secondary procedures to treat and correct complications.

Gibbons and Bradish (16) lengthened 10 tibiae in cases of fibular hemimelia. In all cases, the desired lengthening was achieved and all patients were able to wear normal shoes without orthoses. A plantigrade position was achieved in all feet without persistent ankle instability. Complications occurred in nine of the 10 cases. These were all resolved either surgically or nonsurgically. Several required foot deformity correction with soft tissue or bone procedures.

Perhaps the largest series in the recent literature with the longest follow-up duration is that presented by Catagni and Guerreschi (7). Using the modified Dal Monte classification (12), they reported 32 grade 1, 37 grade 2, and 20 grade 3 cases of fibular hemimelia that were treated with lengthenings, all of which led to completed reconstruction. Of the 32 grade 1 cases, 31 required only one lengthening each and one required a second lengthening. Equal leg lengths with a plantigrade foot were achieved in each of these patients. In the grade 2 group, five patients required three lengthenings each, nine required two lengthenings each, and 23 required one lengthening each. Thirty-five of the patients each ended the reconstruction with a plantigrade, functional foot. Two have residual valgus deformity and require shoes with orthoses. None have undergone ankle arthrodesis. Thirty-two of the 37 participate in recreational sports, and five limit their activities as a result of knee stiffness or instability. In the grade 3 group, two required six stages of reconstruction (a stage referred to as a lengthening or a deformity correction), four required five stages, six required four stages, three required three stages, four required two stages, and one required one stage. Eight of these 20 underwent foot deformity correction as a separate procedure before the age of 3 years. Sixteen of the feet are plantigrade, stable, and asymptomatic. Five have residual valgus with stiffness and require an orthosis to alleviate the symptoms. Although most of the patients could bike or swim, athletic pursuits were more limited than in the grade 1 and 2 patients. There were no permanent sequelae of knee subluxation, hip subluxation, nerve injury, nonunion, or osteomyelitis in any patient. All of the patients were satisfied with the functional results of their reconstruction.

Our results are similar to those of these last three studies. We were able to achieve good or excellent functional results, including the desired goal of lengthening, in 36 of the 38 legs lengthened. The one patient who was rated as having achieved only a fair results was lost to followup and had a residual equinus deformity with a painful arthritic ankle. This patient had an ankle fusion at another center but we were not able to evaluate the final result. Many of our patients are involved in recreational and/or competitive athletics. All of the adults in our series are gainfully employed, including the one with a fair result due to ankle arthritis. We, too, experienced a high complication rate, and our study (as did the last three studies mentioned above) showed that the final result is not related to the complication rate.

Few of the complications lead to major sequelae; those that do can usually be resolved surgically (26).

One of the other criticisms of lengthening is the psychologic impact on the child. Although lengthening is undisputedly stressful for the child and the family, two recent studies have shown that the majority of the problems were transitory and remitted with appropriate treatment (17,23). The lengthening treatment did not cause long-term psychologic maladjustment (17). Although most patients in our experience tolerated the lengthening process well, some patients did develop loss of appetite, weight loss, and difficulty sleeping. We have found that a single small dose of amitriptyline before bed time is useful in helping these patients. Contrary to popular belief, lengthening should not be an excruciatingly painful experience. If a patient is complaining of a lot of pain, especially during the day while at rest, the cause of the pain should be sought. Pain may be related to pin infection, pin loosening or cutting out, frame instability, nerve stretch, reflex sympathetic dystrophy, rupture of the regenerating bone after premature consolidation, etc. Appropriate treatment, such as antibiotics, pin removal, wire retensioning, slowing distraction, replacing pins, and backing up the distraction, should be administered as soon as the problem is recognized.

Peroneal nerve release should be considered if evidence of peroneal nerve stretch does not respond to slowing distraction.

To minimize the psychologic impact of lengthening, serial lengthenings and surgical reconstructions should be spaced apart according to the patient's age to give the child as much time as possible without surgery between sessions. We prefer to lengthen when the patient is between the ages of 1.5 and 4 years for the first lengthening, 6 and 10 years for the second lengthening, and 12 and 14 years for the final lengthening. We have found that children between the ages of 4.5 and 6 years have the most psychologic difficulty with lengthening, whereas children 4 years and younger have the easiest time with lengthening. We prefer to complete the last lengthening before the patient is in high school, for social reasons, if possible.

Cost is another argument for reconstruction rather than amputation. The cost of amputation and prosthetic fitting from age 1 to 18 years is $81,000 per patient (20). Projected lifetime total costs are $373,051 per amputee (34). In comparison, the cost of surgical reconstruction is $40,000 to 50,000 for a single surgical lengthening reconstruction (20). Even three such reconstructions cost less than the lifetime cost per amputee. Therefore, limb salvage is more cost-effective than amputation.

In our opinion, there are few contraindications to lengthening. We think that all patients should be given the option of lengthening versus amputation. If the lengthening option is not available at the treating center, patients should be offered a second opinion at a referral center that has expertise in lengthening reconstruction surgery. Socioeconomic factors may limit such second opinion options. Nevertheless, this should be the patient’s decision and not the doctor’s. Currently, one of the biggest limitations is the availability of pediatric orthopaedic surgeons who are trained in advanced lengthening reconstruction techniques. Finally, in many developing nations, amputation may be culturally unacceptable and good prosthetics unobtainable. In such situations, amputation is contraindicated.

Progressive genu valgum after lengthening in patients younger than 12 years is a previously under-recognized problem that can occur as a result of limb lengthening. Seventy-five per cent of our patients younger than 12 years and essentially all of our patients younger than 4 years developed this problem. The deformity recurs through the proximal tibia. The origin is unclear but follows a pattern similar to that seen with the Cozen phenomenon (11) after proximal tibial metaphyseal fractures. In fibular hemimelia, the progressive tibial valgus may be related to the lack of lateral growth by the fibula or may be due to soft tissue tethers on the lateral side. Intentionally deforming the tibia into 10° to 15° of varus at the end of the lengthening balances out with the expected rebound leaving the leg well aligned within the first year. A similar valgus tendency is observed with progressive valgus deformity in children with fibular hemimelia after amputation (13). Femoral valgus is also associated with fibular hemimelia but is nonprogressive (5). Femoral valgus may contribute to valgus overload, which may be a factor for valgus rebound in the tibia. For this reason we currently prefer to perform distal femoral hemiepiphsiodesis or varus osteotomy at the time of the index procedure. Complete fibrous anlage resection compared to partial anlage resection has reduced the frequency and degree of rebound but has not eliminated the problem. We prefer to perform complete resection instead of partial resection.

Some authors have shown growth inhibition after tibial lengthening for fibular hemimelia (30,31). Sharma et al. (31) concluded that this is related to complete (type 2) fibular hemimelia. Most of the cases presented by Sharma et al. (31) were treated with double-level or combined femur and tibial lengthening without soft tissue release. Hope et al. (19), who used only single-level lengthening, could not demonstrate any growth inhibition. From our center Sabharwal et al. (29) showed that growth inhibition occurred only if there had been a second tibial lengthening performed within a year of the first. In a larger followup study, Sagieh et al, found evidence of mild stimulation and mild inhibition in one-third of the cases.

Our classification according to foot pathology is very surgically oriented. It does not classify the amount of leg length discrepancy. Several authors include the leg length discrepancy as part of their classification (7,12,21). We prefer to classify the case according to the hindfoot deformity and to classify the lengthening according to how many lengthenings will be required to complete limb length equalization. By combining the foot classification with this lengthening classification, we have a realistic picture of how many operations to need to be planned and what type of foot reconstruction is required.

CONCLUSION

In conclusion, the final result of lengthening for fibular hemimelia is dependent on the final foot position after reconstruction. It is essential to obtain a plantigrade stable foot to ensure a satisfactory result. Extra-articular soft tissue release combined with osteotomy is the best method to obtain a plantigrade stable foot. Some cases will require ankle fusion to maintain this result.

REFERENCES

1. Achterman C, Kalamchi A. Congenital deficiency of the fibula. *J Bone Joint Surg Br* 1979;61:133–7.
2. Amstutz HC. Natural history and treatment of congenital absence of the fibula: Proceedings of the American Academy of Orthopaedic Surgeons. *J Bone Joint Surg Am* 1972;54:1349
3. Birch JG, Walsh SJ, Small JM, Morton A, Koch KD, Smith C, Cummings D, Buchanan R. Syme amputation for the treatment of fibular deficiency: an evaluation of long-term physical and psychological functional status. *J Bone Joint Surg Am* 1999;81:1511–8.
4. Birch J, Lincoln T, Mack P. Functional classification of fibular hemimelia. In *Seventh Annual Scientific Meeting, ASAMI North America, The Limb Lengthening and Reconstruction Society*. 1997:18.
5. Boakes JL, Stevens PM, Moseley RF. Treatment of genu valgus deformity in congenital absence of the fibula. *J Pediatr Orthop* 1991;11:721–4.
6. Bohne WH, Root L. Hypoplasia of the fibula. *Clin Orthop* 1977;125:107–12.

7. Catagni MA, Guerreschi F. Management of fibular hemimelia using the Ilizarov method. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:179–93.
8. Cheng JC, Chueng KW, Ng BK. Severe progressive deformities after limb lengthening in type-II fibular hemimelia. *J Bone Joint Surg Br* 1998;80:772–6.
9. Choi IH, Kumar SJ, Bowen JR. Amputation or limb-lengthening for partial or total absence of the fibula. *J Bone Joint Surg Am* 1990;72:1391–9.
10. Coventry MB, Johnson EW. Congenital absence of the fibula. *J Bone Joint Surg Am* 1952;34:941–55.
11. Cozen L. Fracture of the proximal portion of the tibia in children followed by valgus deformity. *Surg Gynecol Obstet* 1953;97:183–8.
12. Dal Monte A, Donzelli O. Tibial lengthening according to Ilizarov in congenital hypoplasia of the leg. *J Pediatr Orthop* 1987;7:135–8.
13. Davidson WH, Bohne WH. The Syme amputation in children. *J Bone Joint Surg Am* 1975;57:905–9.

14. Fulp T, Davids JR, Meyer LC, Blackhurst DW. Longitudinal deficiency of the fibula: operative treatment. *J Bone Joint Surg Am* 1996;78:674–82.
15. Gaine WJ, McCreath SW. Syme's amputation revisited: a review of 46 cases. *J Bone Joint Surg Br* 1996;78:461–7.
16. Gibbons PJ, Bradish CF. Fibular hemimelia: a preliminary report on management of the severe abnormality. *J Pediatr Orthop B* 1996;5:20–6.
17. Ghoneem HF, Wright JG, Cole WG, Rang M. The Ilizarov method for correction of complex deformities: psychological and functional outcomes. *J Bone Joint Surg Am* 1996;78:1480–5.
18. Hootnick D, Boyd NA, Fixsen JA, Lloyd-Roberts GC. The natural history and management of congenital short tibia with dysplasia or absence of the fibula. *J Bone Joint Surg Br* 1977;59:267–71.
19. Hope PG, Crawfurd EJ, Catterall A. Bone growth following lengthening for congenital shortening of the lower limb. *J Pediatr Orthop* 1994;14:339–42.
20. Johnson CE, Haideri NF. Comparison of functional outcome in fibular deficiency treated by limb salvage versus Syme's amputation. In: Herring JA, Birch JG, ed. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998: 173–7.

21. Letts M, Vincent N. Congenital longitudinal deficiency of the fibula (fibular hemimelia): parental refusal of amputation. *Clin Orthop* 1993;287:160–6, 1993.

22. Miller LS, Bell DF. Management of congenital fibular deficiency by Ilizarov technique. *J Pediatr Orthop* 1992;12:651–7.

23. Morton A. Psychologic considerations in the planning of staged reconstruction in limb deficiencies. In: Herring JA, Birch JG, ed. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:195–204.

24. Nakamura E, Mizuta H, Takagi K. Knee cartilage injury after tibial lengthening: radiographic and histological studies in rabbits after 3–6 months. *Acta Orthop Scand* 1995;66:313–6.

25. Naudie D, Hamdy RC, Fassier F, Morin B, Duhaime M. Management of fibular hemimelia: amputation or limb lengthening. *J Bone Joint Surg Br* 1997;79:58–65.

26. Paley D. Problems, obstacles and complication of limb lengthening by the Ilizarov technique. *Clin Orthop* 1990;250:81–104.

27. Paley D, Herzenberg JE, Paremain G, Bhave A. Femoral lengthening over an intramedullary nail: a matched-case comparison with Ilizarov femoral lengthening. *J Bone Joint Surg Am* 1997;79:1464–80.

28. Paley D, Herzenberg JE, Tetsworth K, McKie J, Bhave A. Deformity planning for frontal and sagittal plane corrective osteotomies. *Orthop Clin North Am* 1994;25:425–65.

29. Sabharwal S, Paley D, Bhave A, Herzenberg JE. Growth patterns after lengthening of congenitally short lower limbs in young children. *J Pediatr Orthop* 2000;20:137–45.

30. Shapiro F: Longitudinal growth of the femur and tibia after diaphyseal lengthening. *J Bone Joint Surg Am* 1987;69:684–90.

31. Sharma M, MacKenzie WG, Bowen JR. Severe tibial growth retardation in total fibular hemimelia after limb lengthening. *J Pediatr Orthop* 1996;16:438–44.

32. Stanitski DF. The effect of limb lengthening on articular cartilage: an experimental study. *Clin Orthop* 1994;301:68–72.

33. Stanitski DF, Rossman K, Torosian M. The effect of femoral lengthening on knee articular cartilage: the role of apparatus extension across the joint. *J Pediatr Orthop* 1996;16:151–4.

34. Williams MO. Long-term cost comparison of major limb salvage using the Ilizarov method versus amputation. *Clin Orthop* 1994;301:156–8.