

Amputation Versus Staged Reconstruction for Severe Fibular Hemimelia

Assessment of Psychosocial and Quality-of-Life Status and Physical Functioning in Childhood

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Background: Fibular hemimelia, a congenital disorder characterized by the partial or complete absence of the fibula, tibial growth inhibition, and foot and ankle deformity and deficiency, is the most common deficiency of long bones. The purpose of the present study of children with congenital fibular hemimelia was to examine the functional and psychosocial outcomes at a minimum of 2 years after treatment either with amputation and a prosthesis or with reconstruction and lengthening.

Methods: Twenty children who were managed with primary amputation were compared with 22 children who were managed with staged limb reconstruction. The average age of the patients at the time of evaluation was 9 years (range, 5 to 15 years). Patients and parents completed psychosocial, quality-of-life, and satisfaction surveys. Patients underwent instrumented gait analysis and a timed 25 or 50-yard dash. The number and nature of surgical procedures were recorded from a retrospective chart review.

Results: Families of children managed with amputation had lower economic and educational levels and were more ethnically diverse compared with the families of children managed with limb reconstruction. Scores on psychosocial and quality-of-life surveys were comparable with those from healthy patient populations. Parents of males treated with amputation perceived a lower school-related quality of life for their child; socioeconomic and ethnic differences between groups might account for this finding. Statistically but not clinically significant differences were measured during instrumented gait analysis at a self-selected walking speed and during a timed 25 or 50-yard dash. The majority of patients and parents reported satisfaction with the treatment method selected and would select the same treatment method again.

Conclusions: At this interim stage of growth, there were no significant functional or psychological differences between groups. Both groups were satisfied with the outcome in mid-childhood, irrespective of the selection of amputation or limb reconstruction.

Level of Evidence: Therapeutic Level III. See Instructions for Authors for a complete description of levels of evidence.

Fibular hemimelia is characterized by partial or complete absence of the fibula, tibial growth inhibition, and foot and ankle deformity and deficiency. The primary problems associated with fibular hemimelia include leg-length discrepancy and equinovagis deformity of the foot. The goal of treatment is restoration of the limb through (1) the production

of a plantigrade, painless foot and equalization of limb length or (2) foot ablation and prosthetic fitting.

Studies have documented the success of amputation as treatment for severe deformity¹⁻⁸. Advances in limb-lengthening surgery, including more versatile external fixators⁹⁻¹² and sophisticated foot and ankle reconstructions¹³⁻¹⁵, have enabled staged

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limb reconstruction to become an alternative to amputation¹⁶. Although such lengthening reconstructive surgery is becoming more common, research regarding psychological outcomes and patient satisfaction is limited. Some studies have documented acceptable patient satisfaction and psychosocial adjustment^{17,18}, whereas others have demonstrated complications and adjustment difficulties¹⁹. A recent evaluation comparing outcomes in adults managed with amputation or staged reconstruction failed to demonstrate clinically meaningful differences on questionnaires measuring function and psychosocial status, although the patients were not physically examined at the time of follow-up²⁰. Other studies have demonstrated a perceived advantage in association with amputation but have focused primarily on functional outcomes and patient satisfaction and have not addressed psychosocial outcomes or prosthetic costs²¹⁻²³. Comparisons between those reports is difficult because of the wide range of ages at the time of the procedures and differences in the duration of follow-up and outcome variables.

Residual or recurrent foot deformities are a common cause of failure following lengthening reconstructive surgery^{22,23}. Patel et al. noted that advances in reconstructive surgery have led to reduced complications and that the costs of prosthetic management may exceed the costs associated with limb-lengthening²⁴.

The purpose of the present study was to examine the functional and psychosocial outcomes at mid-childhood in children who had undergone either amputation or staged reconstruction and lengthening beginning in infancy for the treatment of severe fibular hemimelia.

Materials and Methods

After institutional review board approval, participants who had been managed with amputation were recruited from the Texas Scottish Rite Hospital for Children and participants who had been managed with lengthening reconstructive surgery were recruited from the Rubin Institute of Advanced Orthopedics. Patient databases were surveyed at each institution to identify patients who had been managed between 1996 and 2004. Seventy-five patients who had been managed with amputation and 64 who had been managed with staged reconstruction between 1996 and 2004 were identified. The inclusion criteria for the present study were a diagnosis of severe fibular hemimelia (Achterman-Kalamchi Type II²; Paley Type III^{14,15}), an age of ≥ 5 years at the time of study, and reconstructive surgical treatment performed ≥ 2 years prior to the date of enrollment. Potential subjects were required to be at least 5 years of age and at least 2 years after the most recent reconstructive procedure so that they could fully participate in the functional assessments. Exclusion criteria included bilateral congenital fibular hemimelia, severe femoral abnormality, or any orthopaedic procedure within 12 months prior to enrollment. Of the 24 subjects who met these criteria in the amputation group, 3 declined to participate and 1 was ineligible because of recent revision of the residual limb. Of the 24 subjects in the lengthening reconstructive surgery group, 2 declined to participate. Thus, the present study included 20 patients who had been managed with

amputation and 22 patients who had been managed with lengthening reconstructive surgery. The medical records of the participants were retrospectively reviewed for the nature and number of surgical procedures, the ages at which the procedures were performed, any documented surgical complications, and any other pertinent medical history. The average age of the patients at the time of evaluation was 9 years (range, 5 to 15 years). In the amputation group, the number of prostheses delivered and prosthetic repairs made between the amputation and the date of functional assessment were also recorded.

Surgical Methods

Patients in the amputation group were managed with Syme or Boyd amputation^{25,26}. All patients in the lengthening reconstructive surgery group underwent reconstructive ankle surgery (SUPERankle procedure) as described by Paley and Robbins^{14,15}. The SUPERankle procedure involves soft-tissue releases of the ankle, supramalleolar opening-wedge and/or subtalar osteotomy, a second osteotomy to lengthen the tibia at the level of the diaphyseal angular deformity, and application of a circular external fixator. Second and third lengthenings were planned and/or performed in the lengthening reconstructive surgery group. More detailed descriptions of the reconstructive and lengthening procedures have been previously described by Paley¹⁵.

Functional Evaluation

Participants had physical and prosthetic assessments, completed psychosocial questionnaires, underwent instrumented gait analysis, and completed timed dashes. Physical assessment involved evaluations of lower-extremity alignment, joint range of motion and stability, physical activity, and functional impairment.

Psychosocial Questionnaires

Patients and parents completed standardized psychosocial measures on health-related quality of life, including the Pediatric Quality of Life Inventory (PedsQL) Family Impact Module²⁷, the PedsQL Version 4.0 Generic Core Scale²⁸, and the PedsQL Pediatric Pain Questionnaire-Visual Analog Scale (PPQ-VAS)²⁹. Child self-concept was examined with use of the Piers-Harris Children's Self-Concept Scale, Second Edition (Piers-Harris 2)³⁰. The Behavior Assessment System for Children, Second Edition (BASC-2)³¹ was used to screen for behavioral and emotional disorders via both parent and patient reports.

Parents of patients in the amputation group completed the Childhood Amputee Prosthetics Project-Prosthetics Satisfaction Inventory (CAPP-PSI)³². Supplemental items were added to allow parents to rate their child's as well as their own satisfaction with the amputation and to comment about their choice of amputation over limb salvage. A child-report version of the CAPP-PSI was adapted from the parent version by the authors of the present study. This version included 8 items and utilized 10-cm visual analog scales as the response format (maximum total score, 80).

Parents of patients in the lengthening reconstructive surgery group completed the Limb-Lengthening Satisfaction

Questionnaire (LLSQ) adapted from the CAPP-PSI with permission. Parents rated their child's as well as their own satisfaction with the leg's appearance, function, and amount of lengthening achieved as well as with the child's ability to perform daily activities. Parents also rated their own satisfaction with treatment, including their awareness of treatment options, satisfaction with their treatment decision, and willingness to undertake additional lengthenings in the future. A child version of the LLSQ was created; this version included 5 items and utilized 10-cm visual analog scales (maximum total score, 50).

All parents completed a School and Activities Data Sheet (SADS). On scales ranging from 0 ("not at all") to 4 ("very much"), parents rated the extent to which orthopaedic treatment impacted the child's school performance and activity with peers in the past month, in the past year, and overall. Items were summed to create an "impact" total score, ranging from 0 to 32, with higher scores indicating higher positive impact.

Gait Analysis

Fifteen patients in the amputation group and all 22 patients in the limb-lengthening group completed instrumented gait analysis. Five patients in the amputation group did not complete gait testing: 4 declined to participate, and 1 could not complete the testing because of excessive weight, preventing reliable anatomical surface marker placement. Patients in the amputation group used their prostheses and regular shoes. Patients in the lengthening reconstructive surgery group walked barefoot or wore their normal shoes; 5 had shoe lifts ranging from 3.5 to 10 cm and were tested with the lift. Subjects

underwent computerized motion analysis, including anthropometric measurements, passive range of motion, and kinematic and kinetic analysis. In the amputation group, motion analysis was conducted with use of a Motion Capture System (VICON) operating at 120 Hz. Motion capture for the lengthening reconstructive surgery group was conducted using a Motion Analysis system (Motion Analysis). During kinematic data collection, kinetic data were collected simultaneously with use of multiple embedded force plates. Kinematic modeling was done at each institution according to its standard processing protocols; a 15-marker lower-extremity modified Helen Hayes marker set was used for kinematic testing. The data for the amputation group were processed with use of Clinical Manager software (VICON), whereas the data for the lengthening reconstructive surgery group were processed within the Cortex software (Motion Analysis). Subjects completed a timed 25-yard (23 m) lengthening reconstructive surgery group) or 50-yard (46 m) (amputation group) dash. Times for the 25-yard dash were doubled to allow for comparison between groups.

Statistical Methods

Chi-square tests were utilized to examine differences between the groups in terms of parent education, income, ethnicity, and sex. T tests were used to examine differences in age. Multivariate analysis of variance (MANOVA) was used to evaluate differences between the groups in terms of outcomes variables. Demographic variables related to group membership were included within MANOVA tests when group effects were detected.

TABLE I Treatment Group Characteristics

	Amputation Group (N = 20)	Limb Reconstruction Group (N = 22)	P Value	Power Value
Age at time of review* (yr)	10.0 ± 3.0 (5-14)	8.7 ± 2.6 (5-15)	0.057	0.482
Male:female ratio (no. of patients)	14:6	9:13	0.059	0.555
Ethnicity (Caucasian:non-Caucasian) (no. of patients)	9:11	16:6	0.07	0.494
Highest parent education (no. of patients)			0.003	
Less than college	7	0		
Some college or college	12	15		
Graduate school	1	7		
Family income (no. of patients)			0.002	
<\$50,000	10	1		
\$50,000-\$100,000	6	8		
>\$100,000	4	13		
Time since amputation or first limb-lengthening* (yr)	8.3 ± 3.5	6.3 ± 3.0	0.065	0.458
No. of procedures per patient*†	1.2 ± 0.4	3.5 ± 1.9	<0.001	
Surgical complications‡	0	9 (41%)‡	—	—
Limb-length discrepancy at time of review* (cm)	NA	6.1 (0.6-15.0)	—	—

*The values are given as the mean, with or without the standard deviation, with the range in parentheses. NA = not applicable. †Obtained from retrospective medical record review. ‡Joint contracture (5), nerve injury (1), premature consolidation (2), pin-site infection (1).

TABLE II Comparable Items from the Amputee and Limb-Lengthening Satisfaction Scales*

	CAPP-PSI† (Amputation Group)	LLSQ† (Limb-Lengthening Group)	MANOVA P Value	Power Value
Parent-rated child satisfaction			0.151	0.491
Appearance	2.6 ± 1.0	2.3 ± 1.1		
Limb function	3.0 ± 1.1	3.3 ± 0.9		
Limb's ability to aid daily activities	3.3 ± 1.0	3.4 ± 0.7		
Overall result of treatment	2.9 ± 1.2	3.4 ± 0.8		
Parent satisfaction			0.638	0.188
Appearance	3.2 ± 1.0	2.8 ± 1.0		
Limb function	3.2 ± 1.1	3.4 ± 1.0		
Limb's ability to aid daily activities	3.7 ± 0.8	3.6 ± 0.7		
Overall result of treatment	3.4 ± 1.1	3.5 ± 1.1		
Child-reported satisfaction			0.130	0.521
Appearance	8.9 ± 1.3	7.2 ± 2.7		
Limb function	9.5 ± 0.7	8.4 ± 2.7		
Limb's ability to aid daily activities	9.3 ± 1.8	8.8 ± 2.3		
Overall result of treatment	8.9 ± 2.3	8.8 ± 1.3		

*Copies of the LLSQ and child form of the CAPP-PSI can be obtained from the authors of the present study. †The values are given as the mean and the standard deviation.

Results

Participants underwent amputation or first lengthening at an average age of 1.7 and 1.9 years, respectively; in most cases, such treatment was performed at least 7 years prior to the age at which the functional, movement science, and psychosocial investigations were performed. There were significant differences between the groups in terms of parent education ($p = 0.003$) and income ($p = 0.002$), with a near-significant difference in terms of ethnicity ($p = 0.07$) (Table I). The amputation group tended to have lower income and education levels relative to the lengthening reconstructive surgery group. Differences between the groups approached significance in terms of sex ($p = 0.059$) and age ($p = 0.057$). To account for observed differences in outcomes, we included education, ethnicity, and income variables as fixed factors in analyses of variance.

Medical and Clinical Comparisons

In the amputation group, 19 patients had a Syme amputation and 1 had a Boyd amputation. On physician physical examination at the time of review, 5 patients were noted to have valgus knee deformity. All prostheses included a dynamic-response foot. In the lengthening reconstructive surgery group, 16 patients (73%) had completed 1 lengthening at the time of the study (mean, 1.4 lengthenings; range, 1 to 3 lengthenings) and all 22 patients were expected to require at least 1 further lengthening procedure.

Treatment Satisfaction, School, Activities

For the amputation group, the mean total score (and standard deviation) on the parent form of the CAPP-PSI was 44.2 ± 9.6

(range, 26 to 56) and the mean score on the child form was 68.2 ± 6.7 (range, 49 to 80). For the lengthening reconstructive surgery group, the mean parent score on the LLSQ was 25.1 ± 5.9 (range, 13 to 32) and the mean child score was 41.7 ± 7.2 (range, 19 to 48). For parents, the total score averaged 79% of the maximum score of 56 for the amputation group and 63% of the maximum score of 40 for the lengthening reconstructive surgery group. For children, the total score averaged 85% of the maximum score of 80 for the amputation group and 84% of the maximum score of 50 for the lengthening reconstructive surgery group. The total scores on these measures were not compared directly across treatments because many items on each measure were treatment-specific.

Table II presents mean values (and standard deviations) for the comparable items between the CAPP-PSI, LLSQ, and related child adaptations. There were no significant differences between treatment groups for the parent-proxy child ratings ($p = 0.151$), parent self-reported items ($p = 0.638$), and child self-reported items ($p = 0.130$). Ethnicity, parent education, and income did not have significant effects on these measures. Parents and children in both groups tended to report satisfaction with the treatment method selected. On the SADS, no difference in the average total score was found between the amputation group (18.1 ± 7.8) and the lengthening reconstructive surgery group (19.7 ± 5.6) ($p = 0.48$).

Health-Related Quality of Life, Pain, Self-Concept, Behavior

Health-related quality-of-life scores are shown in Table III. For the parent-proxy ratings of child health-related quality of life, the males in the amputation group tended to have lower scores

TABLE III Health-Related Quality of Life Scores

	Amputation Group*	Limb Reconstruction Group*	MANOVA P Value	Univariate P Value†	Power Value
Parent-proxy health-related quality of life	—	—	0.006 (treatment main effect)	—	—
Total	73.9 ± 18.2	86.3 ± 13.4	—	NS	
Physical	76.4 ± 23.6	82.9 ± 19.0	—	NS	
Emotional	76.3 ± 24.1	86.7 ± 14.7	—	NS	
Social	73.8 ± 22.2	87.6 ± 15.8	—	NS	
School	66.2 ± 23.0	90.0 ± 14.6	—	<0.001	
Child-form health-related quality of life	—	—	0.014 (treatment-by-income interaction)	—	—
Total	81.6 ± 10.9	83.0 ± 12.5	—	NS	
Physical	81.7 ± 18.9	83.4 ± 16.7	—	NS	
Emotional	83.2 ± 16.5	85.5 ± 14.0	—	NS	
Social	87.8 ± 14.8	83.6 ± 17.6	—	NS	
School	74.5 ± 15.8	78.9 ± 19.8	—	NS	
PedsQL Family Impact Module	—	—	0.311	—	0.491
Total	78.6 ± 24.8	85.4 ± 13.4	—	NS	
Physical	84.1 ± 25.9	83.3 ± 15.3	—	NS	
Emotional	73.9 ± 26.0	83.6 ± 19.5	—	NS	
Social	82.6 ± 28.9	91.7 ± 20.0	—	NS	
Cognitive	79.3 ± 27.6	92.1 ± 14.6	—	NS	
Communication	77.5 ± 28.0	87.3 ± 16.6	—	NS	
Worry	69.3 ± 25.5	74.5 ± 22.1	—	NS	
Daily activities	81.8 ± 27.7	89.7 ± 20.7	—	NS	
Family relations	81.0 ± 26.2	85.2 ± 24.0	—	NS	
PPQ-VAS	—	—	0.359	—	0.316
Child—current pain	1.0 ± 1.7	0.9 ± 1.7	—	NS	
Child—worst in past week	2.0 ± 2.9	2.1 ± 3.1	—	NS	
Parent-proxy—current pain	0.4 ± 0.7	0.3 ± 0.5	—	NS	
Parent-proxy—worst in past week	1.4 ± 2.4	0.4 ± 0.8	—	NS	
Piers-Harris self-concept scales	—	—	0.909	—	0.134
Total	52.8 ± 9.9	55.9 ± 11.9	—	NS	
Physical	49.8 ± 9.6	51.4 ± 9.3	—	NS	
Behavioral	49.4 ± 9.7	52.7 ± 9.3	—	NS	
Intellectual	52.1 ± 8.6	55.8 ± 8.7	—	NS	
Freedom from anxiety	53.5 ± 7.8	55.1 ± 8.6	—	NS	
Popularity	51.3 ± 9.3	53.5 ± 11.4	—	NS	
Happiness	50.9 ± 7.9	53.3 ± 9.2	—	NS	
BASC-2: parent report	—	—	0.104	—	0.560
Internalizing	52.3 ± 10.7	45.9 ± 10.1	—	NS	
Externalizing	52.7 ± 11.9	46.6 ± 8.5	—	NS	
Attention problems	53.6 ± 11.5	46.1 ± 9.4	—	NS	
Behavioral symptoms index	53.0 ± 12.8	44.4 ± 8.3	—	NS	
BASC-2: self-report	—	—	0.498	—	0.233
Internalizing	45.5 ± 5.8	44.9 ± 11.9	—	NS	
Inattention-hyperactivity	50.6 ± 12.2	46.9 ± 12.8	—	NS	
Attention problems	51.4 ± 10.7	46.4 ± 13.5	—	NS	
Emotional symptoms index	45.5 ± 6.4	44.1 ± 13.6	—	NS	

*The values are given as the mean and the standard deviation. †NS = not significant.

TABLE IV Gait Functioning

	Amputation Group*	Limb Reconstruction Group*	Controls*
Knee extension during mid-stance (°)	9.0 ± 9.9†	13.5 ± 10.1†	0.3 ± 5.2
Ankle range of motion (°)	13.7 ± 6.5†	21.2 ± 6.7††	32.0 ± 7.8
Mean ankle angle over gait cycle (°)	12.0 ± 3.6†	8.7 ± 4.1††	0.1 ± 3.5
Peak ankle moment (Nm/kg)	0.9 ± 0.3†	0.6 ± 0.2††	1.2 ± 0.2
Ankle moment impulse (Nm/kg·s)	1.7 ± 0.8†	1.4 ± 0.6†	2.5 ± 0.7
Peak ankle power (W/kg)	0.8 ± 0.4†	0.4 ± 0.3†	3.1 ± 1.0
Cadence (steps/min)	123 ± 9§	129 ± 18	134 ± 12
Walking speed (m/s)	1.20 ± 0.12	1.13 ± 0.16#	1.25 ± 0.16

*The values are given as the mean and the standard deviation. †Significantly different from the control group ($p < 0.001$). ††Significantly different from the amputation group ($p < 0.001$). §Significantly different from the control group ($p = 0.033$). #Significantly different from the control group ($p = 0.022$).

relative to both standardized norms and the scores for the lengthening reconstructive surgery group on the school domain. For child-reported health-related quality of life, post-hoc analysis showed no significant differences between treatment groups. For example, the tendency for patients in higher-income families to have higher scores was not significant ($p = 0.087$). On the PedsQL Family Impact Module, while many parents expressed some worry about their child's condition, there were no significant differences when comparing treatments ($p = 0.311$).

Data from the PPQ-VAS revealed that most patients perceived pain as a non-factor in their functioning at the time of the study (Table III). As with the PPQ-VAS, scores from the BASC-2 and Piers-Harris instruments were within normal limits, with no clinically meaningful differences between groups.

Gait Analysis

Several significant differences were noted between the amputation and lengthening reconstructive surgery groups, and both groups were significantly different from normal age-matched controls (Table IV). While there were slight differences in cadence (amputation group, $p = 0.033$) and walking speed (lengthening reconstructive surgery group, $p = 0.022$) compared with the controls, both groups were within 90% of their age-matched healthy peers.

Kinematic and kinetic analysis indicated that both groups had slight crouch at the knee during midstance. The patients in the lengthening reconstructive surgery group had greater ankle range of motion ($p < 0.001$) and less calcaneus ($p < 0.001$) compared those in the amputation group; however, both groups demonstrated significantly decreased ankle range of motion compared with controls. Despite the decreased ankle range of motion seen in the amputation group compared with the lengthening reconstructive surgery group, there was no difference between the groups in terms of peak ankle push-off power at toe-off ($p = 0.279$). Both groups showed significantly decreased peak ankle moment ($p < 0.001$) and peak ankle power ($p < 0.001$) compared with the control group.

Discussion

The data in the present study are preliminary as the information presented here does not represent outcomes at skeletal maturity, and we believe that it will be important and informative to repeat these assessments in this cohort after the subjects achieve skeletal maturity. Our findings mirror those of other studies that have suggested that psychosocial adjustment to either amputation or limb-lengthening treatment is possible and probable²⁰. Participants from both groups reported satisfaction with treatment and functioning, and health-related quality-of-life scores were generally consistent with those of other individuals who reported no serious health conditions. Self-concept scores were within normal limits and behavior problems were largely denied.

Analyses failed to find significant differences between the groups in terms of contrasting psychosocial adjustment or physical functioning. While parent-proxy health-related quality-of-life scores were higher in certain instances, it is important to note the considerable difference in demographic characteristics of the 2 treatment groups. The lengthening reconstructive surgery group was appreciably more affluent and educated and included more Caucasians, while the amputation group contained more ethnic minorities and families with lower income and education levels. It is possible that the sample of patients in the lengthening reconstructive surgery group was a product of those families' efforts to obtain additional opinions for treatment after having been presented with recommendations for amputation. The means to acquire and implement additional treatment options may be reflective of the higher socioeconomic status among those families. We also observed a higher number of females within the lengthening reconstructive surgery group. This finding may have been due to an increased willingness of parents of females to lengthen a limb because of the negative perception associated with wearing a prosthesis. At any rate, neither treatment group appeared to be at a significantly greater risk for psychosocial problems relative to the other at this stage of maturation.

As might be expected, both treatment groups demonstrated significant functional differences relative to normal controls on gait analysis. Despite these kinematic differences, cadence parameters showed that both groups were generally able to keep up with their peers. The majority of participants reported no pain, and virtually every family in the study reported to investigators that they would choose the same mode of treatment again. This finding is consistent with those of Ramaker et al.¹⁸, who reported an 88% rate of patient satisfaction with the results of Ilizarov treatment. Both groups will require further surgical procedures during the remainder of skeletal maturation, with all of the participants in the lengthening reconstructive group expected to have at least 1, and in many cases 2, future lengthening procedures. As such, it is conceivable that such intervention could contribute to more impaired performance within the group in the future.

The present study does not fully support the findings reported by McCarthy et al., who concluded that children with fibular hemimelia who were managed with amputation were more satisfied than those who were managed with lengthening²¹. McCarthy et al. compared groups at different ages, whereas we evaluated patients of approximately the same age at the time of follow-up. In a comparison of amputation versus reconstruction, Naudie et al. concluded that, because of recurrent residual foot deformity, amputation was the preferred option²³. It is likely that the ankle reconstructive procedure described by Paley and Robbins^{14,15} may avoid the recurrent residual foot deformities that have resulted in reports of poor outcomes after lengthening reconstructive surgery^{22,23}. Patel et al. also noted that the costs associated with prosthetic management may substantially exceed those associated with limb-lengthening over a patient's lifetime²⁴. The outcomes of limb reconstruction seen in the study by Patel et al. have been corroborated in more recent studies involving the use of the SUPERankle procedure for staged reconstruction in patients with fibular hemimelia³³.

In conclusion, we have demonstrated that psychosocial adjustment and health-related quality of life after primary amputation or limb salvage and reconstruction in patients with

severe fibular hemimelia are comparable and frequently within normal limits for a healthy population at this stage of development and treatment. The quality of performance on gait tests was similar between the groups, although in most cases it was significantly different from that for age-matched controls. Parents and surgeons must weigh life-long prosthetic requirements against significantly greater number of surgical interventions for limb salvage and reconstruction when selecting a treatment strategy for severe fibular hemimelia. ■

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References

- Westin GW, Sakai DN, Wood WL. Congenital longitudinal deficiency of the fibula: follow-up of treatment by Syme amputation. *J Bone Joint Surg Am.* 1976 Jun;58(4):492-6.
- Achterman C, Kalamchi A. Congenital deficiency of the fibula. *J Bone Joint Surg Br.* 1979 May;61-B(2):133-7.
- Epps CH Jr, Schneider PL. Treatment of hemimelias of the lower extremity. Long-term results. *J Bone Joint Surg Am.* 1989 Feb;71(2):273-7.
- Kruger LM, Talbott RD. Amputation and prosthesis as definitive treatment in congenital absence of the fibula. *J Bone Joint Surg Am.* 1961 Jul;43-A:625-42.
- Oberc A, Sulko J. Fibular hemimelia - diagnostic management, principles, and results of treatment. *J Pediatr Orthop B.* 2013 Sep;22(5):450-6.
- 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 Nov;81(11):1511-8.
- Herring JA, Barnhill B, Gaffney C. Syme amputation. An evaluation of the physical and psychological function in young patients. *J Bone Joint Surg Am.* 1986 Apr;68(4):573-8.
- Horgan O, MacLachlan M. Psychosocial adjustment to lower-limb amputation: a review. *Disabil Rehabil.* 2004 Jul 22;26(14-15):837-50.
- Bishay SNG. Simultaneous femoral and tibial lengthening in combined congenital complete fibular hemimelia and congenital short femur using Ilizarov ring external fixator. *J Orthop.* 2014 Jul 18;11(4):183-7.
- Catagni MA, Radwan M, Lovisetti L, Guerreschi F, Elmoghazy NA. Limb lengthening and deformity correction by the Ilizarov technique in type III fibular hemimelia: an alternative to amputation. *Clin Orthop Relat Res.* 2011 Apr;469(4):1175-80. Epub 2010 Oct 21.
- Changulani M, Ali F, Mulgrew E, Day JB, Zenios M. Outcome of limb lengthening in fibular hemimelia and a functional foot. *J Child Orthop.* 2010 Dec;4(6):519-24. Epub 2010 Sep 19.
- Cheng JCY, Cheung KW, Ng BKW. Severe progressive deformities after limb lengthening in type-II fibular hemimelia. *J Bone Joint Surg Br.* 1998 Sep;80(5):772-6.
- Shabtai L, Specht SC, Standard SC, Herzenberg JE. Internal lengthening device for congenital femoral deficiency and fibular hemimelia. *Clin Orthop Relat Res.* 2014 Dec;472(12):3860-8.
- Paley D, Robbins CA. Fibular hemimelia: Paley type 3. In Rozbruch SR, Hamdy RC, editors. *Limb lengthening and reconstruction surgery case atlas.* Cham: Springer International Publishing; 2015. p 1-18.
- Paley D. Surgical reconstruction for fibular hemimelia. *J Child Orthop.* 2016 Dec;10(6):557-83. Epub 2016 Dec 1.

- 16.** El-Sayed MM, Correll J, Pohlig K. Limb sparing reconstructive surgery and Ilizarov lengthening in fibular hemimelia of Achterman-Kalamchi type II patients. *J Pediatr Orthop B*. 2010 Jan;19(1):55-60.
- 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 Oct;78(10):1480-5.
- 18.** Ramaker RR, Lagro SW, van Roermund PM, Sinnema G. The psychological and social functioning of 14 children and 12 adolescents after Ilizarov leg lengthening. *Acta Orthop Scand*. 2000 Feb;71(1):55-9.
- 19.** Hrutkay JM, Eilert RE. Operative lengthening of the lower extremity and associated psychological aspects: the Children's Hospital experience. *J Pediatr Orthop*. 1990 May-Jun;10(3):373-7.
- 20.** Walker JL, Knapp D, Minter C, Boakes JL, Salazar JC, Sanders JO, Lubicky JP, Drvaric DM, Davids JR. Adult outcomes following amputation or lengthening for fibular deficiency. *J Bone Joint Surg Am*. 2009 Apr;91(4):797-804.
- 21.** McCarthy JJ, Glancy GL, Chnag FM, Eilert RE. Fibular hemimelia: comparison of outcome measurements after amputation and lengthening. *J Bone Joint Surg Am*. 2000 Dec;82-A(12):1732-5.
- 22.** Choi IH, Kumar SJ, Bowen JR. Amputation or limb-lengthening for partial or total absence of the fibula. *J Bone Joint Surg Am*. 1990 Oct;72(9):1391-9.
- 23.** Naudie D, Hamdy RC, Fassier F, Morin B, Duhaime M. Management of fibular hemimelia: amputation or limb lengthening. *J Bone Joint Surg Br*. 1997 Jan;79(1):58-65.
- 24.** Patel M, Paley D, Herzenberg JE. Limb-lengthening versus amputation for fibular hemimelia. *J Bone Joint Surg Am*. 2002 Feb;84(2):317-9.
- 25.** Davidson WH, Bohne WHO. The Syme amputation in children. *J Bone Joint Surg Am*. 1975 Oct;57(7):905-9.
- 26.** Eilert RE, Jayakumar SS. Boyd and Syme ankle amputations in children. *J Bone Joint Surg Am*. 1976 Dec;58(8):1138-41.
- 27.** Varni JW, Sherman SA, Burwinkle TM, Dickinson PE, Dixon P. The PedsQL Family Impact Module: preliminary reliability and validity. *Health Qual Life Outcomes*. 2004 Sep 27;2(55):55.
- 28.** Varni JW, Seid M, Kurtin PS. PedsQL 4.0: reliability and validity of the Pediatric Quality of Life Inventory version 4.0 generic core scales in healthy and patient populations. *Med Care*. 2001 Aug;39(8):800-12.
- 29.** Varni JW, Thompson KL, Hanson V. The Varni/Thompson Pediatric Pain Questionnaire: I. Chronic musculoskeletal pain in juvenile rheumatoid arthritis. *Pain*. 1987 Jan;28(1):27-38.
- 30.** Piers EV, Harris DB, Herzberg DS. Piers-Harris 2: Piers-Harris Children's Self-Concept Scale. 2nd ed. Los Angeles: Western Psychological Services; 2002.
- 31.** Reynolds CR, Kamphaus RW. BASC-2: Behavior Assessment System for Children. 2nd ed. Circle Pines: American Guidance Service; 2004.
- 32.** Pruitt SD, Varni JW, Seid M, Setoguchi Y. Prosthesis satisfaction outcome measurement in pediatric limb deficiency. *Arch Phys Med Rehabil*. 1997 Jul;78(7):750-4.
- 33.** Kulkarni RM, Arora N, Saxena S, Kulkarni SM, Saini Y, Negandhi R. Use of Paley classification and SUPERankle procedure in the management of fibular hemimelia. *J Pediatr Orthop*. 2017 May 26. Epub ahead of print.